Introduction

CLEFT LIP AND CLEFT PALATE FACTS

• Cleft lip and palate are developmental defects of the upper lip and roof of the mouth that are present at birth (congenital malformations).
• Cleft lip and palate may occur separately and in combination.
• Both malformations occur as a consequence of incomplete fusion of the developing lip (normally occurs by 35 days of gestation) or of incomplete fusion of the hard or soft palate (normally occurs by the eighth to ninth week of gestation).

What is a cleft palate?

A cleft lip is an opening extending through the upper lip. It may be in the midline (center) or left and/or right side of the lip. A cleft palate is an opening of the hard palate (the bony front portion of the roof of the mouth) or the soft palate (the muscular non-bony region in the rear of the roof of the mouth. Similar to a cleft lip, a cleft palate may be midline and/or to either right of left side of the palate. A cleft palate may extend from the upper jawbone to the rear of the throat. Since development of the lip and palate occur at different times during gestation, an infant may have either a cleft lip or cleft palate or clefting of both regions.
How often do cleft lip and cleft palate occur?

Cleft lip, either as a sole malformation or coupled with cleft palate occur in approximately one in 700 live births. Boys are twice as likely to have a cleft lip with or without an associated cleft palate. On the other hand, girls are more likely to have a cleft palate alone (those not associated with a cleft lip malformation). Ethnic background has been shown to affect the frequency of clefts.

SYMPTOMS OF CLEFT PALATE AND LIP

In most cases, the split in the lip that is characteristic of a cleft is the most noticeable sign of the defect. Milk may come out of your baby’s nose while feeding because the barrier between the mouth and nose is abnormal. It’s also possible for children with a cleft to have dental problems, such as missing teeth or having extra teeth. A cleft palate can also trigger frequent middle ear infections and problems with your child’s Eustachian tubes. These tubes help to drain fluid out of the ears and are responsible for making sure that the amount of pressure is equal on both sides of your eardrum. If your child has problems with ear infections and their Eustachian tubes don’t drain properly, hearing loss can result.

Your child may also have problems with speech. This is more common in cases of cleft palate than in cases of cleft lip. Speech problems caused by a cleft are usually characterized by a nasal quality in the voice.

Causes of Cleft Palate and Lip

The cause of cleft palate and lip isn’t known, but doctors believe that the defects occur because of both genetic and environmental factors. Genetics can play a role in the development of clefts if one or both parents pass down a gene that makes a cleft palate or lip more likely. What you do during your pregnancy can also increase the likelihood that your child will have a cleft palate
or lip. Factors that scientists believe may cause a cleft to develop include:

- cigarette smoking
- drinking alcohol
- taking illegal drugs
- being diabetic
- not getting enough prenatal vitamins, like folic acid.

A cleft can occur as an isolated birth defect or as part of a larger genetic syndrome, such as van der Woude syndrome or velocardiofacial syndrome, which are both genetic malformation disorders.

**Diagnosis of Cleft Palate and Lip**

It’s possible for cleft palate and lip to be diagnosed while your baby is still in the womb through the use of an ultrasound. An ultrasound uses high frequency sound waves to create an image of your baby inside your abdomen. If your doctor discovers that your baby has a cleft palate or lip while in the womb, they may want to remove some of the amniotic fluid that surrounds your baby to have it tested for other genetic abnormalities, such as van der Woude syndrome.

**TREATMENT OF CLEFT PALATE AND LIP**

The treatment for your child’s cleft palate or lip will depend on the severity of the condition. Treatment often involves several surgeries to close the opening and reconstruct the face. A team of specialists may work with you and your child. For example, if your child has problems with speech because of their cleft, they might work with a speech pathologist. Your child’s team may also include a plastic surgeon, an oral surgeon, and/or an orthodontist.

**Surgery**

Surgery to repair a cleft palate or lip is ideally performed in your child’s first year. However, additional reconstructive surgeries may be required well into your child’s teenage years.
Cleft Lip

It’s recommended that you get your baby’s cleft lip repaired when they’re between 10 weeks and 1 year old. If your child has a particularly wide cleft, your child’s doctor may want to do a lip adhesion that involves sewing the cleft together temporarily until a surgeon performs the repair. During surgery, your child will be put under anesthesia and a surgeon will use tissue and skin from both sides of the cleft lip to make the lip wider and close the gap.

Cleft Palate

Doctors usually perform surgery to repair a cleft palate when a child is between 6 and 18 months old. During surgery, the muscles and tissue on the two sides of the palate are connected together to close the cleft.

COPING WITH YOUR CHILD’S BIRTH DEFECT

Ask your child’s treatment team about support groups in your area for families of children with birth defects. Communicate openly with your child and help to build their self-esteem by letting them know that they are not defined by their defect.

PROBLEMS RELATED TO CLEFT LIP AND PALATE

Cleft lips and cleft palates can sometimes cause a number of issues, particularly in the first few months, before surgery is carried out.

Problems that can occur include:

- difficulty feeding – babies with a cleft palate may be unable to breastfeed or feed from a normal bottle because they can’t form a good seal with their mouth
- hearing problems – some babies with a cleft lip are more vulnerable to ear infections and a build-up of fluid in their ears (glue ear), which may affect their hearing
- dental problems – a cleft lip and palate can mean a child’s
teeth don’t develop correctly and they may be at a higher risk of tooth decay

- speech problems – if a cleft palate isn’t repaired, it can lead to speech problems such as unclear or nasal-sounding speech when a child is older

Most of these problems will improve after surgery and with treatments such as speech and language therapy.

Causes of cleft lip and palate

A cleft lip or palate happens when the structures that form the upper lip or palate fail to join together when a baby is developing in the womb. The exact reason why this doesn’t happen in some babies is often unclear. It’s very unlikely to have been the result of anything you did or didn’t do during pregnancy.

In a few cases, cleft lip and palate is associated with:

- the genes a child inherits from their parents (although most cases are a one-off)
- smoking during pregnancy or drinking alcohol during pregnancy
- obesity during pregnancy
- a lack of folic acid during pregnancy
- taking certain medicines in early pregnancy, such as some anti-seizure medications and steroid tablets

In some cases, a cleft lip or palate can occur as part of a condition that causes a wider range of birth defects, such as 22q11 deletion syndrome (sometimes known as DiGeorge or velocardiofacial syndrome) and Pierre Robin sequence.

Diagnosing cleft lip and palate

Cleft lips are usually picked up during the mid-pregnancy anomaly scan carried out when you’re between 18 and 21 weeks pregnant. Not all cleft lips will be obvious on this scan and it’s very difficult to detect a cleft palate on a routine ultrasound scan. If a cleft lip or palate doesn’t show up on the scan, it’s normally
picked up immediately after birth or during the newborn physical examination done within 72 hours of giving birth.

When a cleft lip or palate is diagnosed, you’ll be referred to a specialist NHS cleft team who will explain your child’s condition, discuss the treatments they may need and answer any questions you have. You may also find it useful to contact a support group, such as the Cleft Lip and Palate Association, who can offer advice and put you in touch with parents in a similar situation.

**Treatments for cleft lip and palate**

Cleft lips and cleft palates are treated at specialist NHS cleft centres. Your child will usually have a long-term care plan that outlines the treatments and assessments they’re likely to need as they grow up.

The main treatments are:

- surgery – surgery to correct a cleft lip is usually carried out at 3-6 months and an operation to repair a cleft palate is usually performed at 6-12 months
- feeding support – you may need advice about positioning your baby on your breast to help them feed, or you might need to feed them using a specially-designed bottle
- monitoring hearing – babies born with cleft palates have a higher chance of glue ear, which may affect hearing; close monitoring of their hearing is important and if glue ear affects their hearing significantly, a hearing aid may be fitted or small tubes called grommets may be placed in their ears to drain the fluid
- speech and language therapy – if your baby is born with a cleft affecting their palate (cleft palate or cleft lip and palate) a speech and language therapist will monitor your child’s speech and language development throughout their childhood; they will help with any speech and language problems as necessary
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- good dental hygiene and orthodontic treatment – you’ll be given advice about looking after your child’s teeth, and braces may be needed if their adult teeth don’t come through properly

Outlook for cleft lip and palate

The majority of children treated for cleft lip or palate grow up to have completely normal lives.

Most affected children won’t have any other serious medical problems and treatment can normally improve the appearance of the face and resolve issues such as feeding and speech problems.

Surgery to repair the cleft may leave a small pink scar above the lips. This will fade over time and become less noticeable as your child gets older. Some adults who’ve had a cleft lip or palate repair may be self-conscious or unhappy about their appearance. Your GP may refer you back to an NHS cleft centre for further treatment and support if there are any ongoing issues.

Will a cleft lip and palate happen again?

Most cleft lips and palates are a one-off and it’s unlikely you’ll have another child with the condition. The risk of having a child with a cleft lip or palate is slightly increased if you’ve had a child with the condition before, but the chances of this happening are thought to be around 2-8%.
If either you or your partner were born with a cleft, your chance of having a baby with a cleft is also around 2-8%. Most children of parents who had a cleft will not be born with a cleft. The chances of another child being born with a cleft or of a parent passing the condition to their child can be higher in cases related to genetic conditions. For example, a parent with 22q11 deletion syndrome (DiGeorge syndrome) has a 50% chance of passing the condition to their child.

**Information about your child**

If your child has a cleft lip or palate, your clinical team will pass information about him or her on to the National Congenital Anomaly and Rare Diseases Registration Service (NCARDRS). This helps scientists look for better ways to prevent and treat this condition. You can opt out of the register at any time.

**CLEFT LIP AND CLEFT PALATE**

Cleft lip and cleft palate are birth defects that occur when a baby’s lip or mouth do not form properly during pregnancy. Together, these birth defects commonly are called “orofacial clefts”.

**What is Cleft Lip?**

The lip forms between the fourth and seventh weeks of pregnancy. As a baby develops during pregnancy, body tissue and special cells from each side of the head grow toward the
center of the face and join together to make the face. This joining of tissue forms the facial features, like the lips and mouth. A cleft lip happens if the tissue that makes up the lip does not join completely before birth. This results in an opening in the upper lip. The opening in the lip can be a small slit or it can be a large opening that goes through the lip into the nose. A cleft lip can be on one or both sides of the lip or in the middle of the lip, which occurs very rarely. Children with a cleft lip also can have a cleft palate.

What is Cleft Palate?

The roof of the mouth (palate) is formed between the sixth and ninth weeks of pregnancy. A cleft palate happens if the tissue that makes up the roof of the mouth does not join together completely during pregnancy. For some babies, both the front and back parts of the palate are open. For other babies, only part of the palate is open.

Other Problems

Children with a cleft lip with or without a cleft palate or a cleft palate alone often have problems with feeding and speaking clearly and can have ear infections. They also might have hearing problems and problems with their teeth.

Occurrence

CDC recently estimated that, each year in the United States, about 2,650 babies are born with a cleft palate and 4,440 babies are born with a cleft lip with or without a cleft palate. Isolated orofacial clefts, or clefts that occur with no other major birth defects, are one of the most common types of birth defects in the United States. Depending on the cleft type, the rate of isolated orofacial clefts can vary from 50% to 80%.

Causes and Risk Factors

The causes of orofacial clefts among most infants are unknown. Some children have a cleft lip or cleft palate because of changes
in their genes. Cleft lip and cleft palate are thought to be caused by a combination of genes and other factors, such as things the mother comes in contact with in her environment, or what the mother eats or drinks, or certain medications she uses during pregnancy.

Like the many families of children with birth defects, CDC wants to find out what causes them. Understanding the factors that are more common among babies with a birth defect will help us learn more about the causes. CDC funds the Centers for Birth Defects Research and Prevention, which collaborate on large studies such as the National Birth Defects Prevention Study (NBDFS; births 1997-2011) and the Birth Defects Study To Evaluate Pregnancy exposureS (BD-STEPS; began with births in 2014), to understand the causes of and risks for birth defects, including orofacial clefts.

Recently, CDC reported on important findings from research studies about some factors that increase the chance of having a baby with an orofacial cleft:

• Smoking ~ Women who smoke during pregnancy are more likely to have a baby with an orofacial cleft than women who do not smoke.
• Diabetes ~ Women with diabetes diagnosed before pregnancy have an increased risk of having a child with a cleft lip with or without cleft palate, compared to women who did not have diabetes.
• Use of certain medicines ~ Women who used certain medicines to treat epilepsy, such as topiramate or valproic acid, during the first trimester (the first 3 months) of pregnancy have an increased risk of having a baby with cleft lip with or without cleft palate, compared to women who didn’t take these medicines.

CDC continues to study birth defects, such as cleft lip and cleft palate, and how to prevent them. If you are pregnant or thinking about becoming pregnant, talk with your doctor about ways to increase your chances of having a healthy baby.
Diagnosis

Orofacial clefts, especially cleft lip with or without cleft palate, can be diagnosed during pregnancy by a routine ultrasound. They can also be diagnosed after the baby is born, especially cleft palate. However, sometimes certain types of cleft palate (for example, submucous cleft palate and bifid uvula) might not be diagnosed until later in life.

Management and Treatment

Services and treatment for children with orofacial clefts can vary depending on the severity of the cleft; the child’s age and needs; and the presence of associated syndromes or other birth defects, or both.

Surgery to repair a cleft lip usually occurs in the first few months of life and is recommended within the first 12 months of life. Surgery to repair a cleft palate is recommended within the first 18 months of life or earlier if possible. Many children will need additional surgical procedures as they get older. Surgical repair can improve the look and appearance of a child’s face and might also improve breathing, hearing, and speech and language development. Children born with orofacial clefts might need other types of treatments and services, such as special dental or orthodontic care or speech therapy.

CAUSES

The cause of this failure of fusion is not known in most cases, and a cleft lip and/or palate usually occurs as a “one off” within a family. It may happen as a result of a number of genetic and environmental factors which occur together in a way that could not have been predicted or prevented in advance. Smoking in the first weeks of pregnancy, however, is linked with a slightly higher risk of having a child with a cleft. There are clear links between high levels of drinking alcohol during pregnancy and having a child with a cleft. Some types of medicines taken in pregnancy may also increase the risk of having a baby with a cleft.
These include anticonvulsants, medicines for insomnia, medicines for anxiety (such as diazepam) and corticosteroids. In a minority of families there may be a genetic cause for clefting, which may result in a higher chance of cleft lip and/or palate happening again within the family. Where there is no known cause of the cleft lip and/or palate and no other member of the family is affected, the risk of another baby being born to that family with a cleft will be small (less than 5%), but a little higher than for families where no child has ever been born with a cleft.

**Types of clefts**

There are two major types of clefts:

- cleft lip with or without a cleft palate
- isolated cleft palate.

Cleft lip with or without cleft palate occurs more frequently in boys than girls whereas isolated cleft palate occurs more evenly amongst girls and boys, with some studies showing more girls than boys affected. Cleft lip with or without cleft palate also occurs more frequently in some races than others. Asian populations have a higher incidence than Caucasian populations, who in turn are more frequently affected than Afro-Caribbean populations. Isolated cleft palate occurs evenly in all races.

Up to 50% of the babies referred to a Cleft Service will have an isolated cleft palate. An isolated cleft of the palate involves some or all of the soft palate and may also go into the hard palate. A cleft lip is a gap in the lip extending into the nostril on one side. The babies referred with a cleft lip with or without cleft palate may only have a cleft of the lip or they may have a cleft affecting the lip, the upper gum (alveolus) and the palate. The cleft of the lip and gum may be complete (leaving no part of the lip or gum across the gap) or incomplete (leaving some tissues intact) and may affect only one side (unilateral) or both sides (bilateral) of the nose. The proportions of children affected by each type of cleft will vary from year to year to some extent.
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**Submucous cleft palate**

Some children may have a condition where the palate appears to be intact, but there are underlying muscle and bone defects. This is known as a submucous cleft palate. It is often not diagnosed until a child begins to speak, but there may be a history of early feeding difficulties. A submucous cleft palate may happen in association with a cleft lip, but most happen with no involvement of the lip.

** Syndromes **

Cleft lip and/or palate may occur on its own with no other problems, or as part of a syndrome. There are a very large number of syndromes where a cleft lip and/or palate may be a feature. These are most frequently associated with isolated cleft palate and up to 50% of babies with an isolated cleft palate may have another congenital anomaly or a named syndrome. It is not possible to discuss all the syndromes associated with cleft lip and palate in this chapter, but it is important to look for other anomalies in a baby with a cleft lip and/or palate. Syndromes that may involve a cleft of the lip and/or palate and are most frequently encountered by a cleft team include 22q11 deletion, Van der Woude syndrome and Stickler syndrome.

**22q11 Deletion**

This syndrome is the result of a deletion on the long arm of chromosome 22. It is also known as DiGeorge syndrome, Shprintzen syndrome and Velocardiofacial syndrome. It can cause a very large number of anomalies including characteristic facial features, cardiac anomalies, palate problems, low blood calcium levels and low immunity. People with this syndrome may have many or only some of the possible problems and each of these to a varying degree. So, for example, the palatal problems may be an isolated cleft palate; a submucous cleft palate; a soft palate that looks normal but does not function normally, causing problems with speech and sometimes feeding; or no palate problems at all.
Van der Woude syndrome

Van der Woude syndrome may cause a cleft lip and/or palate or an isolated cleft palate. It can also cause pits in the lower lip and this is usually the clue to the person having the syndrome. This syndrome is also variable in how it affects people who may only have pits in the lip that they are not really aware of, or they may have lip pits and a very severe cleft lip and/or palate. The syndrome is inherited in an autosomal dominant way, meaning that if a person has Van der Woude syndrome they will have a 50% chance of passing the condition on to their children.

Stickler syndrome

Stickler syndrome also has an autosomal dominant pattern of inheritance and is made up of isolated cleft palate, high myopia with a risk of retinal detachment, hearing loss and arthropathy. It is also variably expressed. It is thought that up to 30% of infants with Pierre Robin Sequence may have Stickler syndrome.

Pierre Robin sequence

Some babies born with an isolated cleft palate also have a small lower jaw. In Pierre Robin sequence it is thought that a very small lower jaw results in the formation of the cleft palate during embryological development.

The diagnosis of Pierre Robin sequence is made when an isolated cleft palate occurs in association with a small lower jaw and a tongue that tends to fall back in the mouth, the position of the tongue resulting in feeding and breathing problems of varying severity. Such a newborn baby may need to be monitored in hospital and may need intervention to assist feeding and maintain the airway. Intervention may include feeding the baby via a nasogastric feeding tube and the use of a nasopharyngeal airway to relieve upper airway obstruction. Most babies with these problems will grow out of them by the age of six months, with lower jaw “catch up” in growth in the first two years of life.
Organisation of cleft care in the UK

Babies born with a cleft of the lip and/or palate and their families will have a number of problems to overcome. The degree and nature of the difficulties experienced will vary from child to child but may include problems with feeding, appearance, hearing, speech, dental development and social/psychological issues.

These problems are often complex and interlinked. Appropriate care requires a large multidisciplinary team to work closely together in the best interests of the patient and their family throughout the growth and development of the child to adulthood. The treatment of cleft lip and palate will be discussed in more detail in the following articles. In 1998 the Department of Health commissioned a national review of cleft services in England and Wales by the Clinical Standards Advisory Group (CSAG). This led to the reorganisation of commissioning and the development of specialised Regional Cleft Centres.

All such centres provide a highly multidisciplinary approach to treatment and care with a team of specialists. Teams are led by a clinical director and a cleft services manager. Information on the location of cleft services in Scotland and Northern Ireland, as well as in England and Wales, is available from the voluntary organisation CLAPA. All Cleft Centres in the UK deliver care based on a hub-and-spoke principle. The details of the way in which care is delivered differs throughout England and Wales, with some centres providing more outreach services and others using a more centralised approach. The way in which each service is organised is largely governed by regional geographical considerations. For all centres, however, planning and monitoring of care and cleft surgery is provided in the specialist centre.

The Cleft Care Pathway

In the Cleft Centres patients and their families are seen according to nationally agreed standards and care pathways. Patients and their families are seen by the Cleft Team from the
time of diagnosis and are followed up until adulthood. During this time treatment will include primary surgical repair of the lip and/or palate, usually in the first year of life. Over time the children may also undergo a number of further interventions (including surgery) to improve, for example, speech, hearing, dentition and appearance. As much treatment as possible (for example, speech and language therapy) will be provided locally, and throughout treatment the Cleft Team maintain close links with health professionals in local hospitals and community services. Where children have complex and multiple health needs in addition to their cleft, close collaboration with other specialties both in the community and during hospital admissions is required.

Cleft Teams also provide care for adults who develop problems related to the fact they were born with a cleft lip and/or palate, or to the treatment they had when younger. Adults may be motivated to seek help at any stage in life. This may occur as a result of changes in social circumstances, as a result of continued development (for example loss of teeth and problems with dental rehabilitation), or as a result of increased awareness of changing treatments. Cleft teams regularly collect information on the outcomes of cleft care according to the requirements of the CSAG report. Such audit data includes information about care at the time of diagnosis and birth, speech, facial growth, appearance of lip and nose, hearing and psychological well-being. This data is currently audited at a local, multicentre and national level.

**TREATMENTS FOR CLEFT LIP AND PALATE**

The cleft is usually treated with surgery. Other treatments, such as speech therapy or dental care, may be needed for associated symptoms.

**Your child’s care plan**

Children with clefts will have a care plan tailored to meet their individual needs. A typical care plan timetable for cleft lip and palate is described below:
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- birth to six weeks – feeding assistance, support for parents, hearing tests and paediatric assessment
- 3-6 months – surgery to repair a cleft lip
- 6-12 months – surgery to repair a cleft palate
- 18 months – speech assessment
- three years – speech assessment
- five years – speech assessment
- 8-12 years – bone graft to a cleft in the gum area
- 12-15 years – orthodontic treatment and monitoring jaw growth

Your child will also need to attend regular outpatient appointments at the cleft clinic so their condition can be monitored closely and any problems can be dealt with. These will usually be recommended until they’re around 21 years of age, when they’re likely to have stopped growing.

Surgery

Lip repair surgery

Lip repair surgery is usually carried out when your child is around three months old. Your child will be given a general anaesthetic (where they’re asleep) and the cleft lip carefully repaired and closed with stitches. The operation usually takes one to two hours.

Most children are in hospital for a day or two. Arrangements may be made for you to stay with them during this time. The stitches are removed after a few days or may dissolve on their own depending on the type of stitches used. Your child will have a slight scar, but the surgeon will attempt to line up the scar with the natural lines of the lip to make it less noticeable. It should fade and become less obvious over time.

Palate repair surgery

Palate repair surgery is usually carried out when your child is 6-12 months old. The gap in the roof of the mouth is closed and
the muscles and the lining of the palate are rearranged. The wound is closed with dissolvable stitches.

The operation usually takes about two hours and is carried out under general anaesthetic. Most children are in hospital for one to three days, and again arrangements may be made for you to stay with them. The scar from palate repair will be inside the mouth.

Additional surgery

In some cases, additional surgery may be carried out at a later stage to:

- repair a cleft in the gum using a piece of bone (bone graft) – usually done at around 8-12 years of age
- improve the appearance and function of the lips and palate – this may be necessary if the original surgery doesn’t heal well or there are any ongoing speech problems
- improve the shape of the nose (rhinoplasty)
- improve the appearance of the jaw – some children born with a cleft lip or palate may have a small or “set-back” lower jaw

Feeding help and advice

Many babies with a cleft palate have problems breastfeeding because of the gap in the roof of their mouth. They may struggle to form a seal with their mouth – so they may take in a lot of air and milk may come out of their nose. They may also struggle to put on weight during their first few months.

A specialist cleft nurse can advise on positioning, alternative feeding methods and weaning if necessary. If breastfeeding isn’t possible, they may suggest expressing your breast milk into a flexible bottle that is specially designed for babies with a cleft palate. Very occasionally, it may be necessary for your baby to be fed through a tube placed into their nose until surgery is carried out.
Treating hearing problems

Children with a cleft palate are more likely to develop a condition called glue ear, where fluid builds up in the ear. This is because the muscles in the palate are connected to the middle ear. If the muscles aren’t working properly because of the cleft, sticky secretions may build up within the middle ear and may reduce hearing.

Your child will have regular hearing tests to check for any issues. Hearing problems may improve after cleft palate repair and, if necessary, can be treated by inserting tiny plastic tubes called grommets into the eardrums. These allow the fluid to drain from the ear. Sometimes, hearing aids may be recommended.

Dental care

If a cleft involves the gum area, it’s common for teeth on either side of the cleft to be tilted or out of position. Often a tooth may be missing, or there may be an extra tooth. A paediatric dentist will monitor the health of your child’s teeth and recommend treatment when necessary. It’s also important that you register your child with a family dentist.

Orthodontic treatment, which helps improve the alignment and appearance of teeth, may also be required. This can include using braces or other dental appliances to help straighten the teeth. Brace treatment usually starts after all the baby teeth are lost, but may be necessary before the bone graft, to repair the cleft of the gum.

Children with a cleft are more vulnerable to tooth decay, so it’s important to encourage them to practise good oral hygiene and to visit their dentist regularly.

Speech and language therapy

Repairing a cleft palate will significantly reduce the chance of future speech problems, but in some cases, children with a repaired cleft palate still need some form of speech therapy. A speech and
language therapist (SLT) will carry out several assessments of your child’s speech as they get older.

If there are any problems, they may recommend further assessment of how the palate is working and/or work with you to help your child develop clear speech. They may refer you to community SLT services nearer to your home. The SLT will continue to monitor your child’s speech until they are fully grown and they will work with your child for as long as they need assistance.

Further corrective surgery may sometimes be required for a small number of children who have increased airflow through their nose when they’re speaking, resulting in nasal-sounding speech.
Types of Cleft Palate

There are several main types of cleft palate, and these can affect the type of surgery required. When a cleft palate is associated with a cleft lip, there is most often a complete gap from the front to the back, joining with the gap in the gum and lip.

A cleft palate that occurs without a cleft of the lip almost always affects the back of the palate, but the amount of the rest of the palate that is affected is very variable. It can affect only part of the soft palate, the whole soft palate or the whole soft palate and part of the hard palate. There is a type of cleft that can be difficult to see.

A submucous cleft palate is when the muscles of the soft palate have a gap, but the lining of the palate doesn’t have a gap, so it looks similar to a normal palate. There are often a few signs that something is wrong:

• The uvula, or dangling bit at the very back of the palate, is often in two pieces rather than one.
• There can be a bluish color to the soft palate.
• You can feel a notch in the bone at the very back of the hard palate.

Most children with a submucous cleft palate have no problems from it and need no treatment, but occasionally, there can be speech problems that surgical repair can help.
Examples of these are illustrated here:
1) Submucous cleft palate
2) Cleft of the soft palate only
3) Cleft of the hard and soft palate
4) Cleft of the hard and soft palate associated with a cleft lip on one side – this is the most common type
5) Cleft of the hard and soft palate associated with a bilateral (both sides) cleft lip.

CLEFT LIP AND CLEFT PALATE

Cleft lip and cleft palate are birth abnormalities of the mouth and lip. These abnormalities affect about one in every 700 births and are more common among Asians and certain groups of American Indians than among Caucasians.

They occur less frequently among African-Americans. Cleft lip and cleft palate occur early in pregnancy when the sides of the lip and the roof of the mouth do not fuse together as they should.

A child can have cleft lip, cleft palate, or both. Cleft lip and cleft palate together are more common in boys. It is also important to know that most babies born with a cleft are otherwise healthy with no other birth abnormalities.

Cleft palate

Cleft palate occurs when the roof of the mouth does not completely close, leaving an opening that can extend into the nasal cavity.

The cleft may involve either side of the palate. It can extend from the front of the mouth (hard palate) to the throat (soft palate). Often the cleft will also include the lip. Cleft palate is not as noticeable as cleft lip because it is inside the mouth. It may be the only abnormality in the child, or it may be associated with cleft lip or other syndromes. In many cases, other family members have also had a cleft palate at birth.
Cleft lip

Cleft lip is an abnormality in which the lip does not completely form during fetal development. The degree of the cleft lip can vary greatly, from mild (notching of the lip) to severe (large opening from the lip up through the nose). As a parent, it may be stressful to adjust to the obvious abnormality of the face, as it can be very noticeable.

There are different names given to the cleft lip according to its location and how much of the lip is involved. A cleft on one side of the lip that does not extend into the nose is called unilateral incomplete. A cleft on one side of the lip that extends into the nose is called unilateral complete. A cleft that involves both sides of the lip and extends into and involves the nose is called bilateral complete.
Cleft lip and cleft palate may occur together in an infant, or separately. The degree of the abnormality of both cleft lip and cleft palate can vary greatly. The most common early problem associated with these abnormalities is feeding your baby.

**What causes cleft lip and cleft palate?**

The exact cause of cleft lip and cleft palate is not completely understood. Cleft lip and/or cleft palate are caused by multiple genes inherited from both parents, as well as environmental factors that scientists do not yet fully understand. When a combination of genes and environmental factors cause a condition, the inheritance is called “multifactorial” (many factors contribute to the cause). Because genes are involved, the chance for a cleft lip and/or cleft palate to happen again in a family is increased, depending on how many people in the family have a cleft lip and/or cleft palate.

If parents without clefts have a baby with a cleft, the chance for them to have another baby with a cleft ranges from 2 to 8 percent. If a parent has a cleft, but no children have a cleft, the chance to have a baby with a cleft is 4 to 6 percent. If a parent and a child have a cleft, the chance is even greater for a future child to be born with a cleft. Genetic consultation is suggested.

**What are the symptoms of cleft lip and cleft palate?**

The symptoms of these abnormalities are visible during the first examination by your infant’s physician. Although the degree
of the abnormality can vary, upon inspection of the mouth and lips, the abnormality can be noted, as there is an incomplete closure of either the lip, roof of the mouth, or both.

**Possible complications associated with cleft lip and cleft palate:**

Beyond the cosmetic abnormality, possible complications that may be associated with cleft lip and cleft palate include, but are not limited to, the following:

- **Feeding difficulties:** Feeding difficulties occur more with cleft palate abnormalities. The infant may be unable to suck properly because the roof of the mouth is not formed completely.

- **Ear infections and hearing loss:** Ear infections are often due to a dysfunction of the tube that connects the middle ear and the throat. Recurrent infections can then lead to hearing loss.

- **Speech and language delay:** Due to the opening of the roof of the mouth and the lip, muscle function may be decreased, which can lead to a delay in speech or abnormal speech. Referral to a speech therapist should be discussed with your child’s physician.

- **Dental problems:** As a result of the abnormalities, teeth may not erupt normally and orthodontic treatment is usually required.

- **Feeding my baby with cleft palate:** The most immediate concern for a baby with cleft palate is good nutrition. Sucking for children with a cleft palate is difficult because of the poorly formed roof of the mouth. Children with just a cleft lip (without a cleft palate) do not routinely have feeding difficulties. The following are suggestions to help aid in the feeding of your infant:

- **Breastfeeding is allowed.** It will take extra time and patience. Be open for alternatives if this is not providing adequate nutrition for your infant. You may still pump
your breast and feed your infant breast milk through other techniques.

- Hold your infant in an upright position to help keep the food from coming out of the nose.

- Other feeding devices may be utilized.

- Small, frequent feedings are recommended. This can be a frustrating and slow process, however, your infant will receive more calories, and therefore, gain weight.

- There are many types of bottles and nipples on the market that can assist with feeding an infant with cleft palate. Consult with your child’s physician regarding which type is most appropriate for your child. The following are a few examples:
  
  o NUK nipple
  
  o This nipple can be placed on regular bottles or on bottles with disposable bags. The hole can be made larger by making a criss-cross cut in the middle.

  o Mead Johnson Nurser
  
  o This is a soft, plastic bottle that is easy to squeeze and has a large crosscut nipple. You may use any nipple that the infant prefers with this system.

  o Haberman Feeder
  
  o This is a specially designed bottle system with a valve to help control the air the baby drinks and to prevent milk from going back into the bottle.

  o Syringes
  
  o These may be used in hospitals following cleft surgery and may also be used at home. Typically, a soft, rubber tube is attached on the end of the syringe, which is then placed in the infant’s mouth.

In some cases, supplements may be added to breast milk or formula to help your infant meet his/her calorie needs. Consult your child’s physician regarding other home devices (i.e., small paper cups) for feeding your child with a cleft palate.
The team approach for managing and correcting cleft abnormalities: There may be many people involved in management of a cleft abnormality for your child, because the skills of many different areas are needed to help with the problems that can occur with cleft abnormalities. The following are some of the members of the team:

- Plastic/craniofacial surgeon - a surgeon with specialized training in the diagnosis and treatment of skeletal abnormalities of the skull, facial bones, and soft tissue; will work closely with the orthodontists and other specialists to coordinate a surgical plan.
- Pediatrician - a physician who will follow the child as he/she grows and help coordinate the multiple specialists involved.
- Orthodontist - a dentist who evaluates the position and alignment of your child’s teeth and coordinates a treatment plan with the surgeon and other specialists.
- Pediatric dentist - a dentist who evaluates and cares for your child’s teeth.
- Speech and language specialist - a professional who will perform a comprehensive speech evaluation to assess communicative abilities and who will closely monitor your child throughout all developmental stages.
- Otolaryngologist (ear-nose-throat specialist) - a physician who will assist in the evaluation and management of ear infections and hearing loss that may be side effects of your child’s cleft abnormality.
- Audiologist (hearing specialist) - a professional who will assist in the evaluation and management of hearing difficulties your child may have.
- Genetic counselor - a professional who reviews the medical and family history, as well as examines your child to help in diagnosis. A genetic counselor also counsels your family regarding risk for recurrence in future pregnancies.
• Nurse team coordinator - a registered nurse who combines experience in pediatric nursing with specialization in the care of your child and acts as liaison between your family and the cleft team.

• Social worker - a professional who provides guidance and counseling for your child and your family in dealing with the social and emotional aspects of a cleft abnormality and assists your family with community resources and referrals (i.e., support groups).

Treatment for cleft lip and cleft palate: Treatment for these abnormalities includes surgery and a complete team approach to help with the multiple complications that can occur. Specific treatment will be determined by your child’s physician based on:

• Your child’s age, overall health, and medical history
• Specific qualities of your child’s abnormality
• Your child’s tolerance for specific medications, procedures, or therapies
• Involvement of other body parts or systems
• Your opinion or preference

For most infants with cleft lip alone, the abnormality can be repaired within the first several months of life. This will be decided by your child’s surgeon. The goal of this surgery is to fix the separation of the lip. Sometimes, a second operation is needed. Cleft palate repairs are usually done between the ages of 9 to 12 months. This is a more complicated surgery and is done when the baby is bigger and better able to tolerate the surgery. The exact timing of the surgery will be decided by your child’s physician. The goal of this surgery is to fix the roof of the mouth so that your child can eat and learn to talk normally. Sometimes, a second operation is needed.

Surgery:

At your first visit with the plastic surgeon, he/she will discuss with you the details of the surgery, risks, complications, costs,
recovery time, and outcome. At this time, your child’s surgeon will answer any questions you may have.

**After the surgery for cleft lip:**

Your child may be irritable following surgery. Your child’s physician may prescribe medications to help with this. Your child may also have to wear padded restraints on his/her elbows to prevent him/her from rubbing at the stitches and surgery site. Stitches will either dissolve on their own or will be removed in approximately five to seven days. Specific instructions will be given to you regarding how to feed your child after the surgery. The scar will gradually fade, but it will never completely disappear.

During the surgery, and for a short time after surgery, your child will have an intravenous catheter (IV) to provide fluids until he/she is able to drink by mouth. For a day or two, your child will feel mild pain, which can be relieved with a non-aspirin pain medication. A prescription medication may also be given for use at home. Your child’s upper lip and nose will have stitches where the cleft lip was repaired. It is normal to have swelling, bruising, and blood around these stitches.

**After the surgery for cleft palate**

This surgery is usually more involved and can cause more discomfort and pain for the child than cleft lip surgery. Your child’s physician may order pain medicine to help with this. As a result of the pain and the location of the surgery, your child may not eat and drink as usual. An intravenous (IV) catheter will be used to help give your child fluids until he/she can drink adequately.

- Your child will have stitches on the palate where the cleft was repaired. The stitches will dissolve after several days and they do not have to be taken out by the physician. In some cases, packing will be placed on the palate. Do not take the packing out unless you are told to do so by your child’s physician.
• There may be some bloody drainage coming from the nose and mouth that will lessen over the first day.
• There will be some swelling at the surgery site, which will diminish substantially in a week.
• For two to three days, your child will feel mild pain that can be relieved by a non-aspirin pain medication. A prescription medication may also be given for use at home.
• Many infants show signs of nasal congestion after surgery. These signs may include nasal snorting, mouth breathing, and decreased appetite. Your child’s physician may prescribe medication to relieve the nasal congestion.
• Your child will be on antibiotics to prevent infection while in the hospital. Your child’s physician may want you to continue this at home.
• Your child may be in the hospital for one to three days, depending on your child’s physician’s recommendation.
• A small amount of water should be offered after every bottle or meal to cleanse the incision. You can continue to rinse this area gently with water several times a day, if necessary.

**Diet after surgery**

Your child’s physician may allow breastfeeding, bottle-feedings, or cup-feedings after surgery. Your child should be placed on a soft diet for seven to 10 days after surgery.

For older infants and children, age-appropriate soft foods may include strained baby foods, popsicles, yogurt, mashed potatoes, and gelatin. Note: your child should not use a straw or pacifier, as both could damage the surgical repair.

**Activity after surgery**

Your child can walk or play calmly after surgery. He/she should not run or engage in rough play (i.e., wrestling, climbing) or play with “mouth toys” for one to two weeks after surgery.
Your child’s physician will advise you when your child can safely return to regular play. Follow-up with your child’s surgeon and the cleft team is very important. This will be discussed with you. Your child’s physician will also be an important part of the child’s overall health management after the surgery.

**WHAT HAPPENS TO A KID WITH CLEFT LIP OR CLEFT PALATE?**

A baby with cleft lip and cleft palate may have other difficulties that include feeding problems, middle ear fluid and hearing loss, dental problems, and speech problems.

**Feeding problems**

Have you ever laughed so hard while drinking milk that it came out of your nose? Because there is a hole between the nose and mouth in babies with a cleft palate, they have the same problem, but it can happen every time they drink and not just when they laugh. Fortunately, there are feeding specialists and special baby bottles that can help.

**Ear and hearing problems**

Kids with cleft palate can have hearing loss. This may be caused by fluid building up inside the ear.

They usually need very small special tubes placed in their eardrums to help them hear better. Therefore, children with cleft palate should have their ears and hearing checked about once or twice a year.

**Dental problems**

Children with cleft lip or palate often have dental problems. These problems can include small teeth, missing teeth, extra teeth, or crooked teeth. All kids need to see the dentist regularly and keep their teeth clean. A kid with a cleft might need to go to the orthodontist for braces, too, to make sure his or her teeth grow in straight.
Speech problems

Kids with cleft lip or cleft palate may also have problems with speech. When you talk, the muscles of your soft palate help to keep air from blowing out of your nose instead of your mouth. Kids with cleft palate find their soft palate sometimes does not move well and lets too much air leak out the nose while speaking. That gives them problems with certain sounds and it makes them sound like they are speaking partly through their nose.

What Do Doctors Do?

Treating cleft lip or cleft palate takes a team of different types of specialists (in this case, people who are experts in helping kids with clefts). This team includes a plastic surgeon, a speech-language pathologist, an orthodontist, an otolaryngologist (a doctor trained in ear, nose, and throat problems — say oh-toe-lar-un-GOL-uh-jist), an oral surgeon, a dentist, a geneticist, a social worker, a psychologist, an audiologist, and other health care specialists. Because there are so many different people for a kid to see, the team has a coordinator who works with the parents to help organize everything.

Kids with cleft lip and palate will need several surgeries to fix the cleft. The cleft lip is usually repaired by the time the baby is between 3 to 6 months old. During surgery, the doctor closes the gap in the lip. A person who has cleft lip repaired as an infant will have a scar on the lip under the nose.

The cleft palate is usually repaired at age 9 to 12 months. During surgery on cleft palate, doctors close the hole between the roof of the mouth and the nose and reconnect the muscles in the soft palate.

After surgery, kids will get regular hearing tests to check for hearing problems caused by fluid building up in the ears. If they have special tubes placed in their eardrums, the otolaryngologist will check to see if they are working properly in keeping the fluid from building up.
Types of Cleft Palate

Lots of kids, including those with cleft lip or palate, need orthodontics or braces after their permanent teeth grow in. Braces can straighten crooked teeth. Kids with cleft lip and cleft palate may also need a bone graft when they’re about 8 years old. In a bone graft, a surgeon takes some bone, usually from the kid’s hip, and uses it to fill in the gap in the upper gum area. This allows the upper gum area to hold the permanent teeth better and keep the upper jaw steady.

As kids with cleft lip or cleft palate grow older and become teenagers, some may want to have their scars made less noticeable, their jaws aligned, or their noses straightened. Operations to do these can improve a person’s bite, speech and breathing, and appearance.

Living With Cleft Lip or Cleft Palate

Some kids with severe cleft lip or cleft palate have a flattened nose or small jaw. Other kids with just cleft palate may look just like everyone else. Either way, kids with cleft lip or cleft palate want to be treated like everyone else. A person might have cleft lip, but also have beautiful eyes, a great sense of humor, or a terrific slam-dunk!

If you have cleft lip or cleft palate, there may be some things about your face you can’t change and some that you can. Doctors can do amazing things to make you feel good about the way you look on the outside, and you can do things to make yourself feel good about the way you are on the inside. Even with the many successful operations and treatment for cleft lip and palate, some kids have a hard time growing up with this condition. They may have classmates who tease or bully them or who are just curious and want to know more about it.

Number of Surgeries Needed to Correct Different Types of Cleft Lip

The number and type of surgery used to correct a cleft lip depends on how much of the lip is involved, whether it occurs...
with cleft palate, and whether the nose is affected. Surgery for a child with an isolated cleft lip (not occurring with cleft palate) may require:

- A single surgery if the cleft is small and affects only one area of the lip (unilateral).
- Two surgeries, if it is a wide unilateral cleft. The first surgery on the lip usually is done as soon as possible (generally when the baby is between a few days to 6 weeks old). The second surgery on the lip usually is done when the baby is about 2 to 3 months old.
- Two surgeries for a cleft affecting two areas of the lip (bilateral). Each area is usually surgically repaired in separate procedures.

The type of surgery needed for a child with a cleft lip and/or other related deformities depends on the specific combination of problems.

- A child with cleft lip and a defect of the nose usually needs two surgeries. However, some doctors prefer to correct the nose defect during the same surgery to correct the cleft lip; they believe that this will cause fewer problems with speech later in life. Other doctors prefer to wait until the child is 5 or 6 years old, believing that waiting avoids problems with uneven facial growth because the nose grows more slowly than the rest of the face.
- A child with a cleft lip and a cleft palate may need two or more surgeries.
- Additional surgeries may be needed to correct any unevenness of the lip line or scars that formed on the lips from previous surgery. These surgeries may be done as late as the teen years.

**CLEFT LIP AND/OR PALATE**

Cleft lip and cleft palate are birth defects that occur when the lip or mouth do not form properly during pregnancy. This type
of birth defect also is called a facial anomaly. A child can have only a cleft lip, only a cleft palate, or both together. A cleft lip involves an opening from the upper lip to one or both nostrils. With a cleft in the palate, the opening in the roof of the mouth connects the oral and nasal cavities. Children with cleft lip or cleft palate often have problems eating and talking.

**How common is clefting?**

Clefting of the lip is a relatively common facial anomaly. It occurs in approximately 1 in every 700 live births. Most cases (80 percent) occur in males. There is a wide variation in occurrence in different racial and ethnic groups. The African-American population has a lower incidence (1 per 2,300), and the Japanese and Native Americans have an increased incidence (1 per 580 and 1 per 280 respectively).

In 80 percent of cases, only one side of the face is affected. Twice as many of one-sided clefts occur on the left side than on the right side. A cleft lip can occur alone or together with a cleft palate. The palate is the roof of the mouth. A cleft palate with or without a cleft lip occurs in approximately 1 per 2,500 births. An isolated cleft palate (meaning the cleft palate occurs without a cleft lip) is more common in females than males. It is also more frequently associated with other anomalies.

**What causes cleft lip and palate?**

These birth defects occur very early in the pregnancy, and we do not know exactly what causes them. However, we know that cleft lip and palate is not caused by anything the mother did or didn’t do during pregnancy. If your child is born with this birth defect, it is not your fault.

**Prenatal diagnosis of cleft lip and/or palate**

Even with modern ultrasound technology, we are not able to detect every cleft lip and/or palate before babies are born. If your doctor discovers that your baby has a cleft lip or palate, he or she
may refer you to a maternal-fetal medicine specialist (a doctor who handles high-risk pregnancies). Your maternal-fetal medicine specialists will perform additional ultrasounds to confirm the diagnosis and to evaluate for any other associated anomalies.

**How does cleft lip and/or palate affect my baby?**

The major effect on newborns is feeding issues. Some infants have only mild trouble, and others have more significant problems. Special bottles and careful positioning of your baby is usually helpful until the lip and/or palate is repaired. Your child’s pediatrician and the craniofacial disorder program at Children’s may be helpful if you have any feeding issues. If you had planned to breastfeed, Children’s has lactation consultants who are experienced dealing with babies with special needs.

**How does the diagnosis of cleft lip and/or palate affect the pregnancy?**

In most cases, the only way a clefting diagnosis affects pregnancies is that mothers will have to undergo some additional tests.

- **Ultrasound:** Approximately 25 percent of infants with clefts have an associated condition. Therefore, we always recommend a thorough ultrasound exam when we suspect a cleft lip or palate.
- **Amniocentesis:** Associated conditions can include chromosomal disorders. Your maternal-fetal medicine specialist may recommend an amniocentesis to evaluate your baby’s chromosomes. We perform this test at 16 to 18 weeks gestation. During an amniocentesis, a physician will insert a needle through the abdominal wall into the uterus to remove a small amount of amniotic fluid. We then send the fluid to the lab for testing. Final test results are usually available in 10 to 14 days.
- **FISH test:** A quick response test, called fluorescence in situ hybridization or FISH, will give preliminary results in 24
to 48 hours. It is quite accurate for the diagnosis of the three most common chromosomal disorders: trisomy 13, trisomy 18 and trisomy 21.

Otherwise, management of your pregnancy will be routine. There are no common issues or complications for mothers when their babies have clefting. Unless your baby has additional issues that will require immediate care after birth, you should be able to deliver your child at your community hospital. We do recommend that you bring your child to see a pediatric craniofacial surgeon after he or she is born so we can talk about treatment options.

**How are cleft lip and/or palate treated?**

Your baby will need surgery to correct the cleft lip/palate. The exact timing will depend upon the type of cleft.

- We can repair a one-sided lip within the first month, with follow-up surgery at about 6 months of age.
- We normally repair a one- or two-sided cleft lip with partial palate involvement before 6 months of age.
- If your baby has two-sided clefting of the lip and palate, he or she may need some type of orthodontic device to help him or her eat. You will likely need to schedule that surgery within the first 6 months.
- Depending on whether there is involvement of the gums, your baby may eventually need oral surgery to help with dental issues. If the defect is confined to the palate only, this surgery may not be done until 1 year of age.

The Craniofacial Disorder Program at Children’s Hospital of Wisconsin includes physicians from multiple specialties who can help with all of these treatment options.

**What can I expect after surgery?**

After surgery, your baby will have an IV to provide fluids, medication and nourishment until he or she is able to eat. Some of your baby’s medications may include antibiotics to prevent
infection and pain medication. We will immobilize your baby’s arms using sleeves that keep the elbows from bending. You can remove these sleeves to bathe and/or exercise the arms, but you should leave them on at all other times. The sleeves keep your child from touching his or her face, which helps protect the repair and encourages proper healing. With a cleft lip repair, you will see a line of stitches on the upper lip. The lip will appear swollen for several days. There may be some oozing of blood from the incision line. With cleft palates, all stitches are inside the mouth.

**Will I be able to help care for my baby after birth?**

Yes. We will treat your baby in the newborn nursery if cleft lip/palate is his or her only problem, and you will be able to visit the nursery and have the baby come to your room. If you had planned to breastfeed your baby, a lactation consultant can answer any questions you may have. She can assist with determining if your baby will be able to take directly from your breast or if he or she would do better with a bottle with a special nipple. She can help you to pump your breasts while you are still in the hospital. Your milk can be frozen and stored until your baby is ready for it. Breast pumps are available for use while you are in the hospital.

**When can my baby go home?**

Some babies with clefting have minimal problems and may go home when their mothers are discharged. Others, however, have more difficulties eating, and they may require appliances to assist them with sucking and swallowing. This is usually dependent on the amount of involvement with the palate. Once you’ve left the hospital, your baby may require more frequent follow-up with a pediatrician to ensure he or she is eating enough and gaining enough weight.

If you deliver at Froedtert & The Medical College of Wisconsin Froedtert Hospital Campus, the plastic surgeon may see your baby in the hospital. If you do not deliver at Froedtert, or the plastic surgeon does not see your baby before he or she goes home,
please call to set up an appointment as soon as possible. After surgery, your baby will be able to go home when he or she is able to take in enough food to maintain weight and grow.

What is my baby's long-term prognosis?

Long-term prognosis for isolated cleft lip/palate is good. However, your baby may have several issues that require follow-up.

• Dental concerns: One issue could be dental problems, such as missing, extra or malpositioned teeth. Almost all children with a cleft palate will require braces on their permanent teeth. Also, eruption of the permanent teeth is often delayed.

• Speech problems: As many as 25 to 35 percent of children with cleft lip and palate have speech problems that necessitate a secondary palate surgery and speech therapy. Any baby with a cleft palate should be evaluated by a speech therapist.

• Nasal and septum deformities: There may be some deformities of the nose and septum (cartilage in the nose that divides it into two sides).

• Hearing problems: Many infants with cleft lip and palate also will have problems with hearing. Most will also need to have tubes placed in their ears to help with chronic ear infections and drainage of fluid. This also helps with their hearing and, ultimately, their pronunciation of words.

GENES THAT PLAY A ROLE IN PALATE DEVELOPMENT

The secondary palate develops as an outgrowth of the maxillary prominences at about embryonic day (E) 11.5 in the mouse. The palate shelves initially grow vertically down the side of the tongue (E12.5) and then elevate above the tongue as it drops in the oral cavity (E13.5). With continued growth, the shelves appose in the midline (E14.5) and fuse (E15.5). Growth of the palate shelves
depends on the survival and continued proliferation of mesenchymal cells that originate from neural crest and mesodermal cells of the first pharyngeal arch. In this issue of the JCI, using transgenic animal models Rice and coworkers provide details of the interactions between the epithelium and mesenchyme that lead to palate growth and development (2). They demonstrate a signaling process in which Fgf10 is expressed in the mesenchyme, then activates its receptor, FGF receptor 2b (Fgfr2b), which is located in the epithelium. Finally, Fgfr2b mediates expression of sonic hedgehog (Shh) in the epithelium.

Additional genetic factors involved in palate development have been described using mouse transgenic models; in particular, mice lacking the muscle segment-specific homeobox Msx1 or the signaling molecule Tgfb3 exhibit cleft palate. While many other gene knockouts also result in palate or other craniofacial defects, in most cases the gene deletions and/or insertions cause multiple structural or functional defects. Consequently, evaluation of the role of a particular gene in palate formation has not been possible. The function of Msx1 and Tgfb3 in palate development was extended to isolated forms of clefting in humans. Point mutations and/or statistical analyses have indicated a role for these factors in cases of cleft lip and/or palate in which the only other feature was dental abnormalities (1, 5). In parallel with the advances made from the study of animal models, complementary progress has been made to identify additional genes that play a direct role in human palate development.

Two recent gene discovery reports are particularly relevant to human palate development. In the first, mutations were identified in the gene that encodes the transcription factor interferon regulatory factor 6 (IRF6), resulting in the autosomal dominant disorder Van der Woude syndrome (VWS). VWS is an especially important model for isolated cleft lip and palate. In the clinic, the only difference between individuals with VWS and those with isolated cleft lip and palate is the presence of pits in the lower lip of most VWS cases. In addition, VWS is caused by mutations in
Types of Cleft Palate

a single gene, whereas the more common isolated cleft is a complex trait caused by multiple gene mutations and/or environmental insults. Very recently it was demonstrated that a common haplotype associated with IRF6 contains a mutation that provides an attributable risk of approximately 12% to all common forms of cleft lip and palate. In a second report, nonsense mutations and deletions in the FGFR1 gene were identified in cases of Kallmann syndrome, an autosomal dominant disorder typically characterized by infertility and anosmia. However, approximately 5% of Kallmann syndrome cases have clefts of the lip and/or palate and, as with VWS, some individuals may present with clefts as the only component of the phenotype. Other genes that play a role in human palate development were reviewed recently (P63, PVRL1, TGFA, and TBX22; ref 1) or were reported (SATB2).

One remarkable feature of the genes IRF6, MSX1, and FGFR1 is that mutations in any of the three are associated with dental anomalies and “mixed clefting.” Mixed clefting refers to disorders in which cases of isolated cleft palate and cleft lip (with or without cleft palate) occur in the same pedigree. Clefts of the lip or clefts of the palate arise in the primary palate, whereas clefts of the palate alone occur in the secondary palate. Mixed clefting disorders suggest that identical mechanisms cause these two forms, which previously had been separated based on embryologic and genetic evidence. The presence of dental anomalies in some individuals who have mutations in each of these three genes suggests that these same pathways are common to tooth development.

Pathways in palate development

In addition to demonstrating the essential role of the Fgf10/Fgfr2/Shh signaling pathway in palate development, Rice and coworkers integrate this model into the Msx1 pathway. Zhang et al. previously demonstrated that Msx1, bone morphogenetic protein 4 (Bmp4), Shh, and Bmp2 constitute a pathway that is essential for palate development in mice. An expansion of the models
presented in those papers and incorporates known and speculative interactions between these and other signaling pathways in lip and palate development. First, we show other proteins in pathways, including Shh. These pathways drive the epithelium and mesenchyme interactions that support cell proliferation and palate growth. In addition, we posit that FGFR1, SATB2, and TBX22 are also involved in palate growth in humans and/or mice, although their exact placement in a known pathway remains to be determined. Second, we show a more speculative pathway that attempts to connect molecules that are involved in palate fusion. Solid evidence supports a role in palate fusion in mice for Ahr, Tgfb3, Alk5, Smad2, Gabrb3, and in humans for IRF6. The involvement of these genes and their hypothesized interactions suggest that a broader view of the major players in palate development is coming into focus and represents additional candidate genes that can be investigated by DNA resequencing and/or statistical analyses.

**Pursuit of gene-environment interactions**

Although genes play a substantial role in facial embryogenesis, the role that the environment plays in modulating genetic effects is equally critical. At least three major classes of environmental triggers have been studied. One of these is teratogens. Maternal smoking, for example, has been recognized as an important covariate in clefting. Other teratogens that increase the risk of cleft lip and palate through maternal ingestion include pharmaceuticals, such as the anticonvulsant phenytoin and benzodiazepines, or pesticides, such as dioxin. The effect of a second class of environmental trigger, infection, is less clear. However, we wish to point out that two genes that are essential for palate development, IRF6 and PVRL1, are members of gene families that modulate the immune response to infection. These findings suggest that we need a more critical examination of whether infectious agents increase the risk of clefting after exposure during the first trimester. Finally, both nutrients (e.g., vitamins or trace elements)
and cholesterol metabolism also are increasingly seen as being important in influencing embryonic development. Folate in particular is recognized as playing an important role in neural tube formation. The recognition that folic acid supplementation can decrease the risk of neural tube defects represents, along with the treatment of Rh disease and phenylketonuria, one of the great genetic public health successes of the twentieth century. Cholesterol is an essential component of Shh signaling.

The central role for Shh presented by Rice and colleagues in this issue of the JCI provides further support for the idea that normal variations in cholesterol metabolism and/or disruptions in cholesterol levels through pharmacological intervention might also be risk factors for facial birth defects. Recently, Edison and Muenke provided preliminary data suggesting that early embryonic exposure to the cholesterol-lowering statin drugs may confer a risk for a wide range of birth defects of the midline, including clefts of the lip and palate. The integration of Shh into signaling pathways that include Egf, Fgf, Tgfb or Wnt molecules provides strong justification for critical investigation of the role of cholesterol metabolism in human facial embryogenesis.
Cleft Lip and Palate Repair

Cleft lip is a separation in one or both sides of the lip that is present at birth. Early in the development of the baby inside the mother, the left and right sides of the face and the roof of the mouth join together or “fuse.” If the two sides do not come together correctly, an opening in the lip may occur. This opening can be on just one side of the face, called a unilateral (yoon-ill-lat-er-ool) cleft, or on both sides of the face, called a bilateral (by-lat-er-ool) cleft. A cleft lip that goes up to the nose is called a complete cleft lip; otherwise the cleft is called an incomplete cleft lip.

Left untreated, a child born with a cleft lip may face problems with feeding, growth, development, ear infections, hearing, speech and facial appearance. The cleft lip usually is corrected early in a child’s life, between 3 to 6 months of age, but sometimes later.

Cleft lip surgery will correct the cleft and usually will leave minimal scarring. Virtually every child born with a cleft lip is able to lead a healthy, happy life once the cleft has been repaired.

Home Preparation

When general anesthesia is needed, there are important rules for eating and drinking that must be followed in the hours before the surgery. One business day before your child’s surgery, you will receive a phone call from a nurse between the hours of 1 and 9 p.m. (Nurses do not make these calls on weekends or holidays.)
Please have paper and a pen ready to write down these important instructions. If these instructions are not followed exactly, it is likely your child’s surgery will be cancelled.

- The nurse will give you specific eating and drinking instructions for your child based on your child’s age. Following are the usual instructions given for eating and drinking. No matter what age your child is, you should follow the specific instructions given to you on the phone by the nurse.

For children older than 12 months:
- After midnight the night before the surgery, do not give any solid food or non-clear liquids. That includes milk, formula, juices with pulp, and chewing gum or candy.

For infants under 12 months:
- Up to 6 hours before the scheduled arrival time, formula-fed babies may be given formula.
- Up to 4 hours before the scheduled arrival time, breastfed babies may nurse.

For all children:
- Up to 2 hours before the scheduled arrival time, give only clear liquids. Clear liquids include water, Pedialyte®, Kool-Aid® and juices you can see through, such as apple or white grape juice.
- In the 2 hours before the scheduled arrival time, give nothing to eat or drink.
- You may bring along a “comfort” item — such as a favorite stuffed animal or “blankie” — for your child to hold during the surgery.
- You should bring a long-sleeve T-shirt, slightly larger than your child’s usual size, to the hospital on the day of surgery. It will help make your child more comfortable on the day you take your child home from the hospital.
- You may want to purchase a bottle of hydrogen peroxide, a tube of antibiotic ointment, such as bacitracin, and a box
of Q-Tips to have on hand so you can take care of your child’s lip and nose when you get home from the hospital.

The Surgery

Your child’s cleft lip repair will be done at the Same Day Surgery Center at Children’s Hospital in Lawrenceville. When you have checked in at the Same Day Surgery Center, you and your child will be called to an examination room where your child’s health history will be taken and vital signs will be checked.

You will meet with one of the doctors on your child’s surgical team to go over the surgery. He or she will answer any last-minute questions you might have at this time. A member of the anesthesia staff also will meet with you and your child to review his or her medical information and decide which kind of sleep medication he or she should get. As the parent or legal guardian, you will be asked to sign a consent form before the anesthesia is given.

When it is time for your child to go the operating room, you will be asked to wait in the surgical family waiting area.

• If your child is very scared or upset, the doctor may give a special medication to help him or her relax. This medication is flavored and takes effect in 10 to 15 minutes.

• If relaxation medicine is needed, you may stay with your child as he or she becomes drowsy; you will be asked to wait in the surgical waiting area when your child is ready to move to the operating room.

• Young children get their sleep medication through a “space mask” that will carry air mixed with medication. Your child may choose a favorite scent to flavor the air flowing through the mask. There are no shots or needles used while your child is still awake.

• Once your child is asleep, an intravenous (in-tra-VEE-nuss) or IV line will be inserted into a vein in your child’s arm or leg so that medication can be given to keep him or her sleeping throughout the surgery. Your child will
have no pain during the surgery and no memory of it afterward.

A Parent's/Guardian's Role

The most important role of a parent or guardian is to help your child stay calm and relaxed before the surgery. The best way to help your child stay calm is for you to stay calm. During the surgery, at least one parent or guardian should remain in the surgical family waiting area at all times, in case the family needs to be reached.

While Asleep

While your child is asleep, his or her heart rate, blood pressure, temperature and blood oxygen level will be checked continuously. To keep your child asleep during the surgery, he or she may be given anesthetic medication by mask, through the IV or both. When the surgery is over, the medications will be stopped and your child will begin to wake up.

Waking Up

When your child is moved to the recovery room, you will be called so that you can be there as he or she wakes up.

- Your child will need to stay in the recovery room to be watched until he or she is alert and vital signs are stable. The length of time your child will spend in the recovery room will vary because some children take longer than others to wake up after anesthesia.
- Your child will still have the IV in. A nurse will remove it before your child leaves the hospital, when he or she drinking well.
- Your child will have set of padded arm restraints called “no-no’s” placed on his or her arms to prevent them from bending at the elbow. These no-no’s will need to stay in place for about 2 weeks as the surgical scar heals.
- Children coming out of anesthesia may react in different ways. Your child may cry, be fussy or confused, feel sick
Cleft and Cleft Palate: Causes and Treatments

to his or her stomach, or vomit. These reactions are normal and will go away as the anesthesia wears off.

- You may notice some swelling around your child’s mouth, lips and eyes, as well as some dried blood or oozing where the cleft lip was repaired. The swelling may look worse on the day after the surgery, but it will go down over the next weeks.

- You may see sutures (SOO-chers) or stitches on the outside of the skin. Sometimes, only “dissolvable” sutures will be used, and these sutures do not need to be removed. As the skin heals, the parts of the sutures on the inside of the lip and mouth will dissolve on their own, and the parts you can see on the outside of the skin will dry up and fall off. If non-dissolvable sutures were used, they will be removed by the doctor at your child’s first follow-up visit.

- Your child may have a nasal retainer in place to help reshape the nose during healing. The retainer, which acts as a splint inside the nose, may stay in place for up to 3 months.

- Your child can be given pain medication every 4 to 6 hours, as needed, when he or she wakes up.

- When your child is alert, he or she will be moved to a hospital room so the nursing staff can continue his or her care. If you need help, the nurse will show you how to feed your child and clean his or her scar so that you will become comfortable caring for your child at home.

Going Home

After the surgery, and for the weeks afterward at home, your child will only be allowed to drink liquids or semi-liquids from a bottle or cup. No utensils or straws should be used until your child’s surgeon says it is OK.

- Within the first 24 hours after the surgery, while your child is still in the hospital, he or she will be allowed to drink clear liquids from a bottle or cup.
Your child will stay in the hospital until he or she is drinking well and urinating normally.
When your child is drinking well, the IV will be removed by a nurse before you leave the hospital.
Some children will need to stay for more than 1 day if they are not drinking and urinating normally, or if their parents need extra time to learn how to care for them.
Your child may be given Tylenol with codeine prescription medication for pain relief when he or she leaves the hospital. Over-the-counter Motrin can be combined with the Tylenol with codeine if your child is still having pain, but most children only need over-the-counter Tylenol once they get home. DO NOT give your child any over-the-counter Tylenol while he or she is still on Tylenol with codeine.
The long-sleeve T-shirt you brought from home will be used on the day your child goes home. Place the T-shirt on your child, then put the no-no’s on. Roll the cuff of the T-shirt over the edge of the no-no’s and pin them to the T-shirt with a safety pin. Additional safety pins can be used to pin the no-no’s to the T-shirt at the shoulders as well. A nurse will show you what to do if you have any questions.
As soon as you get home, you should call to make an appointment for your child to be checked 1 week after surgery, and to have any sutures removed, if needed.

At-Home Care

A complete list of instructions for taking care of your child at home will be given to you before you leave the hospital. The main things to remember are:
• If you notice any of the following changes in your child, call the surgeon right away:
  o Fever higher than 101.4°F
  o Trouble breathing or skin color changes (pale, blue or gray)
Bleeding or foul-smelling drainage from the scar or nose

Signs of dehydration, including lack of energy, sunken eyes, dry mouth or not urinating enough/fewer wet diapers

Any redness, swelling, or any “in-and-out movement” of the nasal retainer

- Your child can drink any kind of liquid once he or she gets home. Your child may also eat any kind of food that can be watered down and poured from a cup or bottle, including yogurt (such as Go-GURT®), pudding, milkshakes, or anything that you can grind in a blender to be as smooth as baby food. Remember, though—no utensils or straws!

- The no-no’s must stay on your child’s arms for at least 2 weeks. You should check them every 2 to 4 hours to make sure they are not too tight, and take them off briefly several times a day to allow your child to bend and move his or her arms. Change the long-sleeve T-shirt after you bathe your child.

- Your child may be given an antibiotic for the first couple of weeks after surgery.

- Using a clean jar with a lid, mix up a solution of 50 percent hydrogen peroxide and 50 percent water. Using a clean Q-Tip, gently clean the lip area as often as needed, at least 2 or 3 times each day.

- If your child had a nasal retainer placed during surgery, you will need to clean it the same way you clean the lip area.

- If your child has any non-dissolvable sutures, they will be taken out at the first follow-up visit, approximately 4 to 5 days after surgery. You should make sure that your child has had nothing to eat and is very hungry at the time of the appointment. Bring a bottle or cup of liquid with you to the appointment. The sutures will be taken out while
you feed your child so that he or she be distracted and will not feel the sutures being removed.

• After the sutures are taken out, continue to wash the lip and nose with the hydrogen peroxide/water mix and apply a light layer of antibiotic ointment for the next 2 days. You will be told when to begin to use a moisturizing lotion or scar cream to moisturize and massage the scar.

UNILATERAL CLEFT LIP REPAIR

The presence of unilateral cleft lip is one of the most common congenital deformities. A broad spectrum of variations in clinical presentation exists.

Unilateral cleft lip involves deformity of the lip in addition to the alveolus and nose. Patients with this deformity require short-term care and long-term care and follow-up from practitioners in multiple specialties. Patients may need multiple surgical interventions, from infancy to adulthood, in order to achieve necessary function and aesthetic quality.

No universal agreement has been reached as to the timing and technique of repair. Several methods are used with comparable long-term results, which serves as an indication that more than one treatment option exists for definitive repair. Treatment goals include the restoration of facial appearance and oral function, improvement of dental skeletal and occlusal relationships, improvement of speech, and the psychosocial state.

History of the Procedure

In 1843, closure of the unilateral cleft lip with local flaps was described by Malgaigne. The following year, Mirault modified Malgaigne’s technique by using the lateral lip flap to fill the medial defect. All future methods of cleft lip closure are based on Mirault’s technique. LeMesurier and Tennyson modified this technique with a quadrilateral and triangular flap, respectively. In 1976, Millard published his definitive repair in which the lateral flap
advancement into the upper portion of lip was combined with downward rotation of medial lip. Other modifications have been published by Noordhoff, Mohler, and Onizuka. Fisher has described an anatomical subunit approximation for definitive cleft lip repair.

Millard’s methods, including variations, remain among the most popular method for unilateral cleft lip closure.

Cleft lip surgery has evolved from a simple adhesion of paired margins of the cleft to an understanding of the various malpositioned elements of the lip to a more complicated geometric reconstruction using transposition, rotation, and advancement flaps.

**Problem**

The cleft affects the facial form as an anatomic deformity and has functional consequences.

These include the child’s ability to eat, speak, hear, and breathe. Consequently, rehabilitation of a child born with a facial cleft must involve a multidisciplinary approach and staged appropriately with the child’s development, balancing the timing of intervention against its effect on subsequent normal growth.

**Epidemiology**

**Frequency**

The overall occurrence of cleft lip with or without cleft palate is approximately 1 in 750-1000 live births. Racial differences exist, with the incidence in Asians (1:500) greater than in Caucasians (1:750) greater than in African Americans (1:2000). The incidence of cleft lip/palate is more common in males.

The most common presentation is cleft lip and palate (approximately 45%), followed by cleft palate alone (35%) and cleft lip alone (approximately 20%). Unilateral cleft lips are more common than bilateral cleft lips and occur more commonly on the left side (left cleft lip:right cleft lip:bilateral cleft lip = 6:3:1).
The risk of a newborn having a cleft lip increases if a first-degree relative also has a cleft. If one child already has a cleft lip, the chance of a second child being born with the deformity is 4%. If a parent has a cleft lip, the chance of a newborn having a cleft is 7%. If both a parent and a sibling have a cleft lip, the newborn’s risk rises to 15%.

**Etiology**

Clefting has a multifactorial basis, with both genetic and environmental causes cited.

The observation of clustered cases of facial clefts in a particular family indicates a genetic basis. Clefting of the lip and/or palate is associated with more than 150 syndromes. The overall incidence of associated anomalies (eg, cardiac) is approximately 30% (more common with isolated cleft palate).

Environmental causes, such as viral infection (eg, rubella) and teratogens (eg, steroids, anticonvulsants), during the first trimester have been linked to facial clefts.

The risk also increases with parental age, especially when older than 30 years, with the father’s age appearing to be a more significant factor than the mother’s age. Nevertheless, most presentations are of isolated patients within the family without an obvious etiology.

Midfacial development involves several sets of genes, including those involved in cell patterning, proliferation, and signaling.

Mutations in any of these genes can change the developmental process and contribute to cleft development. Some of these genes include the DIX gene, sonic hedgehog (SHH) gene, transforming growth factor (TGF) alpha/beta, and interferon regulatory factor (IRF6).

**Classification**

Kernahan developed a classification scheme in which the defect can be classified onto a Y-shaped symbol. In this diagram, the incisive foramen is represented as the focal point. This system has been applied to both cleft lip and palate.
Millard modification of Kernahan striped-Y classification for cleft lip and palate. The small circle indicates the incisive foramen; the triangles indicate the nasal tip and nasal floor.

**Pathophysiology**

While the normal embryologic development of the face is detailed in Head and Neck Embryology, a brief outline relevant to the formation of facial clefts follows. In short, the branchial arches are responsible for the formation of several areas, including the mouth and lip. Mesenchymal migration and fusion occurs during weeks 4-7 of gestation. The first branchial arch is responsible for the formation of the maxillary and mandibular processes. The maxillary and mandibular prominences form the lateral borders of the primitive mouth or stomodeum.

Mesenchymal migration and fusion of the primitive somite-derived facial elements (central frontonasal, 2 lateral maxillary, mandibular processes), at 4-7 weeks gestation, is necessary for the normal development of embryonic facial structures. When
migration and fusion are interrupted for any reason, a facial cleft develops along embryonic fusion lines. The embryonic development of the primary palate (lip and palate anterior to the incisive foramen) differs from the secondary palate (palate posterior to the incisive foramen).

The developing processes of the medial nasal prominence, lateral nasal prominence, and maxillary prominences form the primary palate. Fusion occurs, followed by “streaming” of mesodermal elements derived from the neural crest. In contrast, the secondary palate is formed by the fusion of palatal processes of the maxillary prominence alone. The difference in embryonic development suggests the possibility of differing degrees of susceptibility to genetic and environmental influences and accounts for the observed variation in incidences.

In summary, unilateral cleft lip results from failure of fusion of the medial nasal prominence with the maxillary prominence.

**Presentation**

For treatment purposes, unilateral cleft lip can be placed into one of three categories: microform/forme fruste, incomplete, or complete cleft lip.

- **Microform cleft (forme fruste):** This defect is characterized by a “light” furrow along the vertical length of the lip with a small vermilion notch and minor imperfections in the white roll. A small component of vertical lip length deficiency and associated nasal deformity may be present.

- **Incomplete cleft lip:** This defect is characterized by the varying degree of vertical lip separation. By definition, it has an intact nasal sill, commonly termed the Simonart band.

- **Complete cleft lip:** This involves the full-thickness defect of the lip and alveolus (primary palate), extends into the base of the nose (no Simonart band exists), and is often accompanied by a palatal cleft (secondary palate). The premaxilla is typically rotated outward and projects
anteriorly in relation to a relatively retropositioned lateral maxillary alveolar element.

As a consequence of the clefting of the lip, an associated nasal deformity occurs. The structures of the ala base, nasal sill, vomer, and septum are distorted significantly. The lower lateral cartilage on the cleft side is positioned inferiorly, with an obtuse angle as it flattens across the cleft. The alar base is rotated outward. The developing nasal septum pulls the premaxilla away from the cleft, and the septum and the nasal spine are deflected toward the noncleft side. The cleft may continue through the maxillary alveolus and palatal shelf, extending to the palatal bone and soft palate.

A study by Buyuk et al found that patients with unilateral cleft lip and palate had a higher rate of dehiscence around the anterior maxillary teeth on the cleft and noncleft sides of the mouth than did normal controls. The study also found that the rate of fenestrations around these teeth was higher on the cleft side of patients than in controls.

**Further treatment planning**

Orthodontic treatment can be initiated a few weeks following birth, prior to surgical intervention. Other adjunct procedures include lip adhesion, presurgical orthopedics, primary nasal correction, and nasoalveolar molding. These procedures attempt to reduce the deformity. Nasoalveolar molding is the active molding and repositioning of the nasal cartilage and alveolar processes with an appliance. This orthodontic intervention takes advantage of the plasticity of the cartilage. Presurgical nasal alveolar allows repositioning of the maxillary alveolus and surrounding soft tissues in hopes of reducing wound tension and improving results.

Definitive repair is delayed until approximately 3 months of age; this varies, depending on physician comfort. A multidisciplinary approach should be carried out over several years for patients with unilateral cleft lip. This team should include practitioners from audiology, otolaryngology, and speech therapy, among other specialties.
CLEFT LIP AND PALATE REPAIR

Cleft lip and cleft palate repair is surgery to fix birth defects of the upper lip and palate (roof of the mouth).

Description

A cleft lip is a birth defect:

• A cleft lip may be just a small notch in the lip. It may also be a complete split in the lip that goes all the way to the base of the nose.
• A cleft palate can be on one or both sides of the roof of the mouth. It may go the full length of the palate.
• Your child may have one or both of these conditions at birth.

Most times, cleft lip repair is done when the child is 6 to 12 weeks old. For cleft lip surgery, your child will have general anesthesia (asleep and not feeling pain). The surgeon will trim the tissues and sew the lip together. The stitches will be very small so that the scar is as small as possible. Most of the stitches will absorb into the tissue as the scar heals, so they will not have to be removed later.

Most times, cleft palate repair is done when the child is older, between 9 months and 1 year old. This allows the palate to change as the baby grows. Doing the repair when the child is this age will help prevent further speech problems as the child develops. In cleft palate repair, your child will have general anesthesia (asleep and not feeling pain). Tissue from the roof of the mouth may be moved over to cover the soft palate. Sometimes a child will need more than one surgery to close the palate. During these procedures, the surgeon may also need to repair the tip of your child’s nose. This surgery is called rhinoplasty.

Why the Procedure is Performed

This type of surgery is done to correct a physical defect caused by a cleft lip or cleft palate. It is important to correct these conditions as they can cause problems with nursing, feeding, or speech.
Risks

Risks from any anesthesia and any surgery include:
• Breathing problems
• Reactions to the medicines
• Bleeding
• Infection
• Need for further surgery

Problems these surgeries may cause are:
• The bones in the middle of the face may not grow correctly.
• The connection between the mouth and nose may not be normal.

Before the Procedure

You will meet with a speech therapist or feeding therapist soon after your child is born. The therapist will help you find the best way to feed your child before the surgery. Your child must gain weight and be healthy before surgery.

Your child’s health care provider may:
• Test your child’s blood (do a complete blood count and “type and cross” to check your child’s blood type)
• Take a complete medical history of your child
• Do a complete physical exam of your child

Always tell your child’s provider:
• What medicines you are giving your child. Include drugs, herbs, and vitamins you bought without a prescription

During the days before the surgery:
• About 10 days before the surgery, you will be asked to stop giving your child aspirin, ibuprofen (Advil, Motrin), warfarin (Coumadin), and any other drugs that make it hard for your child’s blood to clot.
• Ask which drugs the child should still take on the day of the surgery.
On the day of the surgery: Most times, your child will not be able to drink or eat anything for several hours before the surgery.

- Give your child a small sip of water with any drugs your doctor told you to give your child.
- You will be told when to arrive for the surgery.
- The provider will make sure your child is healthy before the surgery. If your child is ill, surgery may be delayed.

After the Procedure

Your child will probably be in the hospital for 5 to 7 days right after surgery.

Complete recovery can take up to 4 weeks. The surgery wound must be kept very clean as it heals. It must not be stretched or have any pressure put on it for 3 to 4 weeks. Your child’s nurse should show you how to take care of the wound. You will need to clean it with soap and water or a special cleaning liquid, and keep it moist with ointment.

Until the wound heals, your child will be on a liquid diet. Your child will probably have to wear arm cuffs or splints to prevent picking at the wound. It is important for your child not to put hands or toys in the mouth.

Outlook (Prognosis)

Most babies heal without problems. How your child will look after healing often depends on how serious the defect was. Your child might need another surgery to fix the scar from the surgery wound. A child who had a cleft palate repair may need to see a dentist or orthodontist. The teeth may need correcting as they come in.

Hearing problems are common in children with cleft lip or cleft palate. Your child should have a hearing test early on, and it should be repeated over time. Your child may still have problems with speech after the surgery. This is caused by muscle problems in the palate. Speech therapy will help your child.
Cleft and Cleft Palate: Causes and Treatments

RECOVERY

Parents generally request the specific "care path" for their child so that expectations for cleft lip and cleft palate repair plastic surgery can be defined. These treatment protocols vary between centers and among surgeons; there is no absolute right or wrong. Nevertheless, we believe that any protocol should deliver a clear vision of care from infancy to adulthood.

Our usual sequence of care is as follows:

<table>
<thead>
<tr>
<th>Age</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth to 3 mos</td>
<td>presurgical orthodontics</td>
</tr>
<tr>
<td>3 mos</td>
<td>cleft lip and soft palate repair</td>
</tr>
<tr>
<td>18 mos</td>
<td>hard palate repair (columella lengthening)</td>
</tr>
<tr>
<td>4 yrs</td>
<td>pharyngeal flap (if needed)</td>
</tr>
<tr>
<td>7-8 yrs</td>
<td>alveolar bone graft/orthodontics</td>
</tr>
<tr>
<td>14-16 yrs</td>
<td>orthodontics/orthognathics/rhinoplasty</td>
</tr>
</tbody>
</table>

Many parents are very concerned about the presence of scars. Unfortunately, all cleft lip repairs leave visible scars. Every effort is made to keep scars to a minimum and to place scars so that they are easily concealed. Secondary (or "redo") surgeries at any stage may be necessary. Our occurrence of secondary surgery is less than 5%.

After Surgery Care

The goal after surgery is to protect the new repair and stitches. For this reason there will be some changes in the child’s feeding, positioning, and activity for a short time. Remember, these are only temporary!

Infants will not be able to suck on a nipple/bottle or pacifier for 10 days after surgery. A syringe with a short piece of soft rubber tubing will be used for feeding. Older children may drink from a cup. It is helpful if the child has practiced drinking from the syringe before surgery. As soon as the infant awakes from anesthesia and acts hungry they may be offered a feeding of clear
liquid (Pedialyte, sugar water, apple juice). When this is tolerated, they may resume their regular formula. Infants who have already begun cereal or baby foods may be offered diluted feedings with the syringe. Older children will be on a blenderized diet that pours easily from a cup.

There may be some discomfort as the child swallows so they may not drink much the first evening. This is why IV fluids are continued until their drinking improves. Pain medicine will also be given to relieve distress.

A child who has had a cleft lip repair should be positioned on their side or back to keep them from rubbing their face in the bed. A child with only a cleft palate repair may sleep on their stomach. It is important to keep the stitches clean and without crusting. Parents are shown how to clean the suture line and apply ointment while in the hospital. This will continue until the stitches are removed about a week later.

It is important to keep the child from hurting the incision or putting hands or toys in their mouth. For this reason they will wear arm restraints (NoNo’s) which keep them from bending their elbows. These are also used for 10 days after surgery.

Children usually spend one night in the hospital and are discharged when they begin to drink an adequate amount of fluids. Parents are encouraged to stay with their child and participate in their care. Chair beds are available in the rooms for overnight sleeping.

**SURGICAL REPAIR OF CLEFT LIP AND PALATE**

**Treatment overview**

The surgical procedures required for each patient with cleft lip and palate will vary depending upon the type and severity of the deformity. Timing and treatment will be adjusted based on each patient’s overall medical needs, but treatment typically includes a combination of the procedures explained here, performed within general time frames based on development.
The Division of Plastic and Reconstructive Surgery at The Children’s Hospital of Philadelphia is one of the nation’s leading centers for treating cleft lip and palate. Our team performs more than 1,000 surgical repairs each year.

Read on below for an introduction to the various procedures that may be included in the surgical repair of cleft lip and cleft palate at Children’s Hospital. Surgery for both cleft lip and cleft palate require general anesthesia.

**Nasoalveolar molding**

1 week to 3 months of age (if needed)

Babies born with unilateral cleft lip or unilateral cleft lip and palate have the option of nasoalveolar molding (NAM), a procedure performed by an orthodontist who specializes in treating craniofacial deformities. Beginning in the first few weeks after birth and continuing until the patient is ready for cleft lip repair, NAM gradually brings the palate and lip together and provides symmetry of the nose, preparing the patient for optimal surgical outcomes.

The process uses an appliance consisting of a palatal plate and nasal stent which is made based on an impression of the patient’s mouth. Frequent adjustments gradually tighten the device to slowly mold the palate. Nasoalveolar molding is performed by our specialized orthodontists.

**Cleft lip repair**

3 to 6 months

The goal of cleft lip surgery is to repair the separation of the lip. Cleft lip is typically repaired between 3 and 6 months of age. During those first few months, your child is monitored closely for adequate weight gain and nutrition, and to make sure that there are no issues relative to breathing while eating.

There are a variety of techniques that may be used to repair a cleft lip. The most common type of cleft lip repair is a rotation
advancement repair. The plastic surgeon will make an incision on each side of the cleft from the lip to the nostril. The two sides of the lip are then sutured together, using tissue from the area to rearrange and close the lip as needed. In addition to closing the lip, cleft lip repair realigns the muscle of the upper lip to provide normal lip function and facilitate suckling. In some cases, a second operation is needed. For example, patients with a bilateral cleft lip may require a short hospital stay in order to complete two surgeries, about a month apart.

A primary nasal repair is often performed at the time of lip repair. Although the type of repair differs from surgeon to surgeon, this procedure involves liberating some of the nasal elements and realigning them to a more normal configuration with the use of stents or sutures. Nasoalveolar molding is often used after surgery to maintain this correction.

After surgery for cleft lip:
• Your child may be irritable and feel mild pain.
• Your child may have to wear padded restraints on his elbows to prevent rubbing at the surgery site.
• Swelling, bruising and blood around stitch sites are normal. Stitches dissolve or will be removed in five to seven days.
• Scars will gradually fade but will not completely disappear.
• An intravenous (IV) catheter will be used to give your child fluids until he can drink adequately.

Cleft palate repair

9 to 18 months

The goal of cleft palate surgery is to fix the roof of the mouth so that your child can eat and talk normally. Cleft palate repair is a more complicated surgery and has the best outcome when the child is slightly older and better able to tolerate the surgery, but before significant speech development occurs. Surgical repair of the palate generally occurs around 1 year of age, following the successful repair of cleft lip if present. In some cases, a second
operation is needed. There are a variety of different techniques that may be used to repair the cleft palate, such as a Z-plasty or a V-Y-pushback. These procedures close the palate in three layers: the inner layers that form the nasal lining; the middle layers, consisting of the muscles at the back of the palate; and the final layer, which includes the oral mucosa.

Palate repair closes these layers while also realigning the palatal muscles, a procedure called an intravelar veloplasty. This puts the muscles in a normal position that allows for the best function of the palate during speech, eating and swallowing.

Surgical repair of cleft palate separates the oral and nasal cavities. This separation involves the formation of a watertight and airtight valve that is necessary for normal speech. The repair also helps with preserving facial growth and proper dental development.

Once the lip and palate are repaired, typically no further surgery is performed for several years. A portion of the palate is usually left open to allow room for the mouth, palate and jaw to grow.

After surgery for cleft palate:

• Your child may experience more discomfort and pain with cleft palate repair than cleft lip repair.
• Your child may have nasal congestion. This can be relieved with medication.
• Your child may stay in the hospital for one to three days and will be given antibiotics to prevent infection.
• Your child will have stitches on his palate. Stitches will dissolve after several days. If packing is placed on the palate, do not remove the packing until instructed.
• There may be bloody drainage from the nose and mouth. It is also normal to have temporary swelling, bruising and blood at the surgery site.
• An intravenous (IV) catheter will be used to help give your child fluids until he can drink adequately.
Palatal expansion

5 to 7 years (if needed)

Due to the clefting of the alveolus, or gum, and the cleft itself, approximately 25 percent of patients with cleft lip and palate will require palatal expansion as a pre-surgical procedure prior to bone grafting.

The palates in patients with clefts tend to be narrow and collapsed. Palatal expansion prepares your child for subsequent bone grafting by pushing out and aligning the alveolar segments, creating space for permanent teeth. A device is fixed to either the bone or teeth, and with a jack screw, the palate is transversely widened to a normal state. This is done during the mixed dentition phase, when your child’s permanent teeth are beginning to develop. The procedure is done by your orthodontist, working closely with your plastic surgeon. Once palatal expansion is complete, alveolar fistulas that were left open at the time of initial lip and palate repair are generally addressed.

Alveolar bone graft and fistula repair

6 to 9 years

Alveolar bone grafting creates a more complete dental arch, and space for permanent teeth to erupt, by placing bone along the alveolus where it is deficient. Soft bone, generally taken from the hip, is packed in to any remaining opening of the palate. The bone graft is secured with a surgical splint as it heals and solidifies.

At this time, procedures are also performed to close any fistulas (openings) between the gum and nose. Closing the fistulas with local tissue prevents the escape of fluid into the nose which leads to nasal regurgitation and leakage of fluids during eating.

After the bone graft is placed, permanent teeth may erupt in abnormal positions. Once the bone graft is placed and any fistulas are closed, orthodontic treatment can move teeth into the space created.
Tip Rhinoplasty

6 to 9 years

If the patient has a significant nasal deformity, an intermediate rhinoplasty may be performed. This is a procedure in which the nose is opened and the cartilage is rearranged to improve the nasal shape and airway.

In cases where a less significant nasal deformity is present, your plastic surgeon may perform a tip rhinoplasty. This procedure addresses just the tip of the nose, providing greater symmetry and improving the nasal airway.

Phase I orthodontics

6 to 9 years

Orthodontic treatment may consist of several phases of treatment, lasting several years each. Phase I orthodontics typically occurs during the mixed dentition phase, when patients begin to lose their baby teeth. In patients with cleft lip and palate, permanent teeth commonly erupt in abnormal positions, so minor orthodontic movement may be required to align teeth.

Planning for orthodontic treatment is generally assessed 6 months after the bone graft is done, and treatment is determined based on each patient’s healing and growth.

Phase II orthodontics

14 to 18 years

Patients generally begin Phase II of their orthodontic treatment during early adolescence or adolescent years. During Phase II orthodontics, teeth are leveled and aligned, missing teeth may be replaced, and teeth that are out of position or fail to erupt may be brought down into the dental arch or removed. This phase of orthodontia includes treatment for atopic eruption of teeth and other potential complications that emerge as a result of bone grafting.
In general, patients with cleft lip and palate may have missing or displaced teeth that have to be removed. Many of these patients, even after final orthodontic treatment and restorations, are missing teeth and may need prolonged orthodontia to move and shape teeth into a more appropriate position.

Patients may also need a bridge constructed or dental implants placed. The need for long-term orthodontic treatment varies by the patient.

**Orthognathic surgery (jaw surgery)**

**14 to 18 years**

Cleft palate patients commonly have underdevelopment of the maxilla (upper jaw), resulting in maxillary retrusion. In these cases, the upper jaw is situated behind the lower jaw, which is the reverse of the normal jaw placement in which the upper jaw and teeth project further than the lower jaw.

In severe cases of maxillary retrusion, the upper jaw may need to be cut and brought forward in a procedure called a LeFort I osteotomy and advancement. This surgery is generally done at skeletal maturity, when the patient is between 14 and 18 years old. Follow-up care will include any necessary orthognathic surgical orthodontics related to jaw discrepancies.

Children seen at a younger age with severe maxillary retrusion may undergo an intermediate phase called a distraction osteogenesis.

This procedure is reserved for younger patients or adolescents with a severe maxillary retrusion that prevents the jaw from being moved forward in a single stage.

Distraction for severe deformities involves cutting the jaw, applying a halo device called a distractor, and then gradually pulling the jaw forward over a several week period. The jaw is then held in this position for six to eight weeks while the new bone that has been created solidifies.
Final touchup surgery

Adolescence or adulthood

As the patient grows, secondary speech procedures and secondary palatal or lip procedures may be done based on the function, appearance and scarring.

Some patients will undergo a final cleft rhinoplasty once they have reached skeletal maturity. This procedure may need to be done in stages. Secondary lip revision to improve the scars and correct irregularities may also be performed at this time.

The goal of treatment is to complete all procedures by the time a patient reaches skeletal maturity (usually around age 18). This process sometimes extends well into the late teens or early 20’s due to the complexity of the cases, but the goal is to have the patient finished with their cleft care at this point in time.

CLEFT LIP REPAIR

Cleft lip repair (cheiloplasty) is surgical procedure to correct a groove-like defect in the lip.

Purpose

A cleft lip does not join together (fuse) properly during embryonic development. Surgical repair corrects the defect, preventing future problems with breathing, speaking, and eating, and improving the person’s physical appearance.

Demographics

Cleft lip is the second most common embryonic (congenital) deformity. (Club foot is the most common congenital deformity.) Cleft lip occurs in approximately one in 750–1,000 live births. The highest incidence exists in North American Indians and Japanese (approximately one in 350 births). African Americans and Africans represent the lowest incidence of cleft lip deformity (approximately one in 1,500 births). There is a higher frequency of clefting in certain populations of Scandinavia and Middle European countries.
Cleft lip occurs more commonly in males, while cleft palate is more likely to occur in females. Cleft lip alone (without cleft palate) occurs in approximately 20% of cases across both genders. The majority of cases—80%—have both cleft lip and cleft palate. A unilateral cleft lip, commonly occurring on the left side, is more common than a bilateral cleft lip.

**Potential causes**

Most cases of cleft lip have no known cause. However, there is a strong genetic correlation. Other single gene defects that are associated with cleft lip include: Van der Woude syndrome, Opitz Syndrome, Aarskog syndrome, Fryns syndrome, Waardenburg syndrome, and Coffin-Siris syndrome. Approximately 5% of cleft conditions are associated with a genetic syndrome. Most of these syndromes do not include mental retardation.

Facial cleft has been implicated with maternal exposure to environmental causes, such as rubella or medications that can harm the developing embryo. These medications include steroids, antiseizure drugs, vitamin A, and oral anti-acne medications (such as Acutane) taken during the first three months of pregnancy. Cleft lip is also associated with fetal alcohol syndrome and maternal diabetes.
Risk of cleft lip increases with paternal age, especially over 30 years at the time of conception. Generally, the risk is higher when both parents are over 30 years of age. However, most cases seem to be isolated within the family with no obvious causation.

When the affected child has unilateral cleft lip and palate, the risk for subsequent children increases to 4.2%. Advances in high resolution ultrasonography (prenatal ultrasound exam) have made it possible to detect facial abnormalities in the developing embryo (in utero).

Description

Developmental anatomy

Important structures of the embryo’s mouth form at four to seven weeks of gestation. Development during this period entails migration and fusion of mesenchymal cells with facial structures. If this migration and fusion is interrupted (usually by a combination of genetic and environmental factors), a cleft can develop along the lip. The type of clefting varies with the embryonic stage when its development occurred.

There are several types of cleft lip, ranging from a small groove on the border of the upper lip to a larger deformity that extends into the floor of the nostril and part of the maxilla (upper jawbone).

Unilateral cleft lip results from failure of the maxillary prominence on the affected side to fuse with medial nasal prominences. The result is called a persistant labial groove. The cells of the lip become stretched and the tissues in the persistent groove break down, resulting in a lip that is divided into medial (middle) and lateral (side) portions. In some cases, a bridge of tissue (simart band) joins together the two incomplete lip portions.

Bilateral cleft lip occurs in a fashion similar to the unilateral cleft. Patients with bilateral cleft lip may have varying degrees of deformity on each side of the defect. An anatomical structure (intermaxillary segment) projects to the front and hangs unattached.
The edges of the cleft between the lip and nose are cut (A and B). The bottom of the nostril is formed with suture (C). The upper part of the lip tissue is closed (D), and the stitches are extended down to close the opening entirely (E).

Defects associated with bilateral cleft lip are particularly problematic due to discontinuity of the muscle fibers of the orbicularis oris (primary muscle of the lip.) This deformity can result in closure of the mouth and pursing of the lip.

Classification

In addition to classification as unilateral or bilateral, cleft lips are further classified as complete or incomplete. A complete cleft involves the entire lip, and typically the alveolar arch. An incomplete cleft involves only part of the lip. The Iowa system (which also classifies cleft palate) classifies cleft lip in five groups:

- Group I—clefts of the lip only
• Group II—clefts of palate only
• Group III—clefts of lip, alveolus, and palate
• Group IV—clefts of lip and alveolus
• Group V—miscellaneous

Another widely accepted cleft lip classification is based on recommendations of the American Cleft Palate Association. This classification divides cleft lip into unilateral or bilateral (right, left or extent) in thirds—(i.e., one-third, two-thirds, three-thirds), or median cleft lip, the extent of which is also measured in thirds.

**Surgical procedure**

Cleft lip repair can be initiated at any age, but optimal results occur when the first operation is performed between two and six months of age. Surgery is usually scheduled during the third month of life.

While the patient is under general anesthesia, the anatomical landmarks and incisions are carefully demarcated with methylene blue ink. An endotracheal tube prevents aspiration of blood. The surgical field is injected with a local anesthetic to provide further numbing and blood vessel constriction (to limit bleeding). Myringotomy (incisions in one or both eardrums) is performed, and myringotomy tubes are inserted to permit fluid drainage.

There are several operative techniques for cleft lip reconstruction. The Millard rotation advancement (R-A) technique is the most widely accepted form of repair. This method involves rotation of the entire philtral dimple (groove in the upper lip) and Cupid’s bow (double curve of the upper lip). The scar falls along the new philtral column, and is adjusted as required since the procedure allows for flexibility.

The Millard procedure begins with an incision on the edge of the cleft side of the philtrum, and the cutting continues upward, medially, and to the side. A second incision extends to the buccal sulcus (top part of the upper jaw). The length of this incision depends on the size of the gap to be closed. In this second incision,
the surgeon frees soft tissue, which allows him or her to completely lift the lip from the underlying bone. This dissection should be tested to ensure free advancement toward the middle (inadequate dissection is the root cause of poor results). Nasal deformity can be dealt with by a procedure known as the McComb nasal tip plasty, which elevates the depressed nasal dome and rim. Cartilage from the cleft side is freed from the opposite side, and is positioned and reshaped using nylon sutures.

Advantages of the Millard rotation advancement technique (include:

• It is the most common procedure (i.e., surgeons more familiar with it).
• The technique is adaptable and flexible.
• It permits construction of a normal-looking Cupid’s bow.
• A minimal amount of tissue is discarded.
• The suture line is camouflaged.

The disadvantage of the Millard rotation advancement technique is the possible development of a vermilion notch (shortening of the entire lip in the vertical direction), resulting from contracture of the vertical scar.

Cupid’s bow is a critical part of the repair, making it very important to accurately determine the high point of Cupid’s bow on the lateral lip.

**Diagnosis/Preparation**

Facial clefting has a wide range of clinical presentations, ranging from a simple microform cleft to the complete bilateral cleft involving the lip, palate, and nose. A comprehensive physical examination is performed immediately after birth, and the defect is usually evident by visual inspection and examination of the facial structures. Care must be taken to diagnose other physical problems associated with a genetic syndrome. Weight, nutrition, growth, and development should be assessed and closely monitored. Presurgical tests include a variety of procedures, such
as hemoglobin studies. It is important for the patient’s parents and physician to discuss the operation prior to surgery.

**Aftercare**

The postoperative focus is on ensuring proper nutrition, as well as lip care and monitoring the activity level. Breast milk or full-strength formula is encouraged immediately after surgery or shortly thereafter. Lip care for patients with sutures should include gentle cleansing of suture lines with cotton swabs and diluted hydrogen peroxide.

Liberal application of topical antibiotic ointment several times a day for 10 days is recommended. There will be some scar contracture, redness, and firmness of the area for four to six weeks after surgery. Parents should gently massage the area, and avoid sunlight until the scar heals.

The patient’s activities may be limited. Some surgeons use elbow immobilizers to minimize the risk of accidental injury to the lip. Immobilizers should be removed several times a day in a supervised setting, allowing the child to move the restricted limb(s). Interaction between the orthodontist and surgeon as part of the treatment team begins in the neonatal period, and continues through the phases of mixed dentition.

**Risks**

There may be excessive scarring and contraction of the lips. Two types of scars, hypertrophic or keloid, may develop. Hypertrophic scars appear as raised and red areas that usually flatten, fade in color, and soften within a few months. Keloids form as a result of the accelerated growth of tissue in response to the surgery or trauma to the area.

The keloid can cause itching and a burning sensation. Scratching must be avoided because it can lead to healing problems. Some patients require minimal revision surgery, but in most cases, the initial redness and contracture is part of the normal healing process.
Normal results

Ideal surgical results for cleft lip include symmetrically shaped nostrils, and lips that appear as natural as possible and have a functional muscle. Many characteristics of the natural lip can be achieved; however, the outcome ultimately depends on a number of factors, including the skill of the surgeon, accurate presurgery markings, alignment of bones within the affected area, uncomplicated healing of the initial repair, and the effect of normal growth on the repaired lip. Additional surgical correction to reconstruct nasal symmetry is sometimes necessary.

Morbidity and mortality rates

Generally, cleft lip repair is well-tolerated in healthy infants. There are no major health problems associated with this reconstructive surgery. Depending on the results, it may be necessary to perform additional operations to achieve desired functional and cosmetic outcomes.

Alternatives

There are no alternatives for this surgery. Obvious deformity and impairments of speech, hearing, eating, and breathing occur as a direct result of the malformation. These issues can not be corrected without surgery.

CLEFT LIP REPAIR DISCHARGE INSTRUCTIONS

Following the instructions below will prevent injury to your child’s newly repaired lip and help in the healing process.

Cleansing the Lip

Your child’s lip has been repaired with Dermabond. You may begin cleaning the lip on the third day after surgery. The lip may be cleansed as needed with a solution of \( \frac{1}{2} \) strength hydrogen peroxide. This is made by mixing equal amounts of hydrogen peroxide with water. Do not use any Neosporin, Vaseline or ointments to the surgical site as these will cause the Dermabond
to melt. Following surgery, the lower teeth may be brushed, and you may begin to brush the upper teeth 2 weeks after surgery. Make sure to use only a foam or soft bristle toothbrush for at least the first 2 weeks of brushing.

**Tube Care**

Your child will also have tubes in his or her nose. These tubes are called nasal stents. It is important to keep these tubes clean and prevent them from becoming clogged. You may clean the tubes by using a Q-tip that has been moistened with water.

**Restraining and Positioning**

Your child should wear arm restraints (no-no’s) at all times to keep him or her from touching or putting anything in their mouth. The arm restraints should be removed one at a time twice daily for 15-30 minutes to allow for elbow exercising. This should be done only under careful observation. These restraints are not uncomfortable and are for your child’s safety. They will be necessary for 2-3 weeks after surgery.

It is also important to keep your child from rolling over onto his or her side or stomach, as pressure against the surgical area could cause injury. Positioning your child on his or her back in an infant seat or a seat supported on both sides will prevent rolling over.

**Feeding**

To help healing and decrease tension to the suture line, your child will need to be fed using only the tip of the nipple in his or her mouth. NO pacifiers should be used during this time. We suggest you place the nipple on the unaffected side. If your child has a bilateral cleft repair, the nipple should be placed in the middle of the mouth. Your child should be fed in a sitting position, fed slowly and carefully, allowing time for burping. Feeding this way is necessary for 2-3 weeks. If you are breastfeeding, you may continue to do so at this time.
Cleft Lip and Palate: Comprehensive Treatment and Technique

CLEFT LIP / CLEFT PALATE

At Plastic and Craniofacial Surgery for Infants and Children, we provide compassionate care for children with congenital disorders and birth defects. Cleft lip and cleft palate deformities can be successfully corrected through advanced surgical techniques.

Types of Cleft Lip

A cleft lip can range in severity from a slight notch in the red part of the upper lip to a complete separation of the lip extending into the nose. Clefts can occur on one or both sides of the upper lip. The term “incomplete” refers to deformity that does not go through the floor of the nose and the incisive foramen, while the term “complete” includes the floor of the nose and incisive foramen and the entire primary and secondary palates. The laterality of the anomaly is described as right, left, or bilateral. Cleft lip and/or palate may present in any combination. The least affected lip is known as a “forme fruste” or microform cleft lip and may be exhibited as a depression in the lip and slight alteration of the nostril floor or alar shape. This variant may still require definitive
Cleft and Cleft Palate: Causes and Treatments

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lip and nasal repair. Surgery is generally done when the child is 10-12 weeks old.

Types of Cleft Palate

A cleft palate may occur as part of the cleft lip deformity (cleft lip/cleft palate) or as an isolated midline entity involving the secondary palate (isolated cleft palate or ICP). These two occurrences of cleft palate are distinctly different embryologic events and should be viewed as different congenital anomalies even though the surgical correction and goals of treatment are very similar.

It is important to recognize that the risk of occurrence for ICP and CL/CP is very different with CL/CP being far more common. Furthermore, multiple malformation syndromes are far more likely in ICP (42%) as compared to CL/CP where multiple malformation syndromes occur in only 14%. By far the most common associated disorder with ICP is the Pierre Robin variant while the most frequently occurring syndrome is Stickler Syndrome (17.5% of syndromic clefts, autosomal dominant, severe myopia, retinal detachment, and glaucoma).

Embryology

Normal facial development is completed in the first three months of life. The events that occur in-utero and result in clefting are not clear. The cleft anterior to the incisive foramen (Primary) and that posterior to the foramen (Secondary) define the anatomical delineation of cleft palate. The primary palate refers to that area that forms the upper lip, columella, maxillary alveolus, and the hard palate anterior to the incisive foramen. The secondary palate forms the soft and hard palate posterior to the incisive foramen. Between the fourth and eighth week, the primary palate formation is usually complete, while the secondary palate formation is completed between the eighth and twelfth weeks. These distinctly different times for the development of the primary and secondary palate form the basis for considering cleft lip with cleft palate and
isolated cleft palate as different developmental deformities. The significance of this distinction is underscored when one envisions the difference in familial inheritance as well as the difference in risk for inheriting associated disorders with other organ systems.

**Etiology**

The exact cause for cleft lip and cleft palate is not known. It is postulated that environmental, genetic chromosomal factors may be responsible for the occurrence of this anomaly with all its variations. Animal experiments have produced clefting in the offspring by varying diets, drug exposures, radiation and vitamin dosages. A protective influence in the prevention of clefting with folic acid supplementation before and during the first trimester is now being postulated. Families known to be at risk for clefting have been used for these studies. Environmental factors in humans have been limited to rubella, in which offspring may demonstrate clefting.

**HEALTH PROBLEMS RELATED TO CLEFT LIP AND PALATE**

**Feeding**

Feeding difficulties may occur in newborns with cleft lip and/or palate as the normal anatomy of the mouth is disrupted. Each baby is different in their ability to feed. In the presence of a cleft lip, the child may be unable to form a seal around the nipple of a bottle. Children with cleft palates have difficulty in generating suction because of the opening between the mouth and the nose. During your child’s first clinic visit, the baby will have a feeding assessment with the occupational therapist present, who will provide information and demonstrate how to feed the baby.

**Tooth Development**

A cleft lip and/or palate can affect the development of teeth. The type of cleft the child has will have an impact on how the teeth
will be affected. Missing teeth, small teeth, extra teeth and crooked teeth are common. Regular dental care is important for healthy teeth and gums.

Orthodontics

A baby with a cleft lip and palate may have the upper jaw divided into two or three parts separated by wide gaps. Orthodontic treatment for a child with cleft lip and palate could start as early as a few days old, depending on the child’s type of cleft. A custom-fitted orthodontic appliance is used to bring the parts of the upper jaw, lips and nose closer together and in a better position. This is called Nasoalveolar Molding (NAM). Not all infants will need this treatment. The orthodontist at Children’s Hospital Los Angeles will direct fitting of the appliance. Most children with cleft lip and palate will have problems with the alignment of the teeth and jaws. The orthodontist can correct this problem with regular treatments starting at age seven for some children.

Hearing

Proper hearing is important for normal speech development. In children with clefts involving the soft palate, the middle ear may be affected. This occurs when the eustachian tube, which connects the middle ear with the back of the throat, does not function properly. This structure allows fluid to drain from the middle ear into the back of the throat. When its function is compromised, fluid may build up in the middle ear, interfering with hearing. The child may develop a temporary hearing loss. This fluid can also get infected and ear infections may be frequent. For this reason, many children have tubes inserted into the eardrum to drain the fluid. This results in improved hearing and fewer ear infections.

Speech

Children with unrepaired cleft palates will have speech difficulties. Normal speech is the goal of surgery. Many children will require speech therapy after the operation, and some may
need a second procedure if speech difficulties persist. The speech therapist will regularly assess speech development and will arrange for speech therapy in the community if necessary.

**Social and Emotional Impacts**

There can be a considerable social and emotional toll associated with having a child with cleft lip and/or palate. The infant’s appearance, feeding difficulties, frequent trips to the doctor and surgeries can be quite stressful. This may, in turn, have an impact on the adaptation process of both child and parents.

**CLEFT LIP AND PALATE TREATMENT**

Cleft lip, and especially cleft lip and palate, are associated with sucking and swallowing difficulties. Affected infants need to be followed closely to ensure adequate weight gain. Multiple ear infections, often with resulting hearing loss, are not uncommon, and the presence of fever needs to be evaluated for possible inflammation of the middle ear. Difficulty with speech articulation is common and may necessitate speech therapy.

The goal of surgical repair of cleft lips is to restore the normal appearance and muscular anatomy of the upper lip. At our institution, this is done typically between three and six months of age. Cleft palate repair proceeds at approximately 10 to 12 months of age. The goal is to close the gap between the mouth and nose, in addition to restoring the muscular function of the soft palate. The initial cleft lip repair is often combined with placement of ear tubes by the otolaryngologist. The child is monitored closely following repair of the palate to determine whether additional assistance is warranted for the proper development of speech and hearing.

Since each child’s cleft is unique, different surgical techniques may be used to repair the cleft. Also, secondary procedures may be required to correct speech, fill in the bony gap between the teeth, improve the relationship of the upper jaw to the lower jaw or refine the appearance of the nose. Your plastic surgeon will
discuss the surgeries that will be performed on your child at each stage with you.

CLEFT LIP AND PALATE

Cleft lip and palate is the result of abnormal fusion of the facial structures during pregnancy. Patients are diagnosed at birth by the presence of a gap or crack of the lip and/or palate. The range of severity for this defect can be quite wide. Cleft lip and/or palate occurs in approximately 1 in 1000 births. Children can have cleft lip, cleft palate, or both. The most common type is cleft lip and palate (46%), followed by isolated cleft palate (33%) and isolated cleft lip (21%).

In addition to cosmetic problems, these children can have difficulty with feeding and development of speech. Early referral to a craniofacial clinic is needed to ensure the best outcomes for these children.

Causes & Risk Factors

Smoking during pregnancy and fetal exposure to phenytoin (a drug used to treat seizures) can cause cleft lip and palate. Children of Asian descent have a higher incidence of cleft lip and palate. A family history of cleft lip and/or palate is also a risk factor for the condition. If the parent or sibling has a cleft lip and palate, the risk that your next child will have the disorder is 4%. If two siblings have cleft lip and palate, the risk of a subsequent child increases to 9%. If the parent and a child have cleft lip and palate, then the risk of the next child having the disorder goes to 17%. For isolated cleft palate, the risk of subsequent pregnancies with the disorder is 2% with one affected child and 6% if the parent had a cleft palate.

Symptoms & Types

Cleft Lip

The easiest way to classify cleft lip is either as a unilateral or bilateral cleft.
A unilateral cleft lip only involves one part of the lip that extends toward one of the nostrils.
• A bilateral cleft lip involves two parts of the upper lip and extends towards both nostrils.

Clefts can be considered complete or incomplete.
• Complete cleft lips are clefts that extend up and into the nostril and will involve the tooth bearing structures in the mouth (known as the alveolus).
• An incomplete cleft lip extends toward the nostril, but the skin of the upper lip is not fully separated.

Complete cleft lips usually have a more severe deformity than incomplete clefts. Complete and incomplete clefts can be seen in both unilateral and bilateral cleft lips. A microform cleft lip is the least severe form of cleft lip and is usually only noticeable as lip notching.

Cleft Palate

Cleft palates occur with cleft lips or by themselves. A cleft palate can involve the uvula (the small tissue that hangs down in the back of the mouth), or a bifid uvula (one that looks like it has split in 2). It can involve only the soft palate, or extend into the soft and hard palate. If a cleft palate involves both the soft and hard palate and extends into the tooth baring portion of the mouth (alveolus) it is considered a complete cleft palate. Just like in a cleft lip, there can be unilateral and bilateral cleft palates. Unilateral cleft palates involve only one side of the hard palate and alveolus, while bilateral cleft palates involve both sides. A submucous cleft palate is a unique type. Although the roof of the mouth appears structurally intact, the muscles of the submucous cleft palate are not in the correct position or orientation. This may contribute to speech problems as a child grows.

Diagnosis & Tests

The diagnosis of cleft lip and/or palate is a clinical one, made by physical examination. Genetic testing is frequently done to
screen for syndromes that can be seen associated with cleft lip and palate.

**Treatment & Care**

Care of patients born with a cleft lip or cleft palate begins at birth and continues until they are fully grown. These patients are best treated in by multidisciplinary team. Treatment can begin as early as 1 week of age for severe cleft lip patients. A technique known as Nasoalveolar Molding (NAM) is used for very severe clefts to decrease the severity of the deformity, provide for an easier repair and increase chances for good outcomes. The craniofacial orthodontist on the cleft team will perform this operation.

Surgical repair of the cleft lip is usually performed in the first 3-6 months of life. The precise timing depends on whether the child needs NAM treatment or not and whether she has any other medical issues that may prevent her from being operated on at such a young age. The cleft lip surgery usually is a day surgery or an overnight admission. Children can resume feeding with a bottle after the procedure.

Doctors usually recommend cleft palate repair when the child is 9 -12 months old. Because this surgery is performed on the child’s airway, proper precautions are taken. These precautions include overnight hospital admission and continuous monitoring of patient oxygenation. The cleft palate surgery is also delayed until this age because the child must be old enough to handle the surgery safely, but not too old where they have begun to produce many sounds.

The cleft palate operation is a functional: its goal is to repair the hole in the palate as well as the muscles in the palate so that appropriate speech production can be achieved. After this repair, constant speech therapy is needed so that any speech problems can be identified early and addressed. Secondary speech surgery is sometimes necessary if the cleft palate is not functioning so that all appropriate sounds can be produced.
After the cleft lip and palate have been repaired, each patient is followed by the Cleft Lip and Palate Clinic on to make sure all issues associated with the cleft have been addressed. These issues can include speech production, feeding issues and sleeping problems. Further surgical intervention is not necessary until the permanent teeth begin to replace the baby teeth. This usually occurs around 7-9 years of age. The craniofacial orthodontist will work closely with the cleft surgeon to determine the appropriate age for alveolar bone grafting. The cleft that is in the tooth baring segment of the mouth needs to be filled with bone so that when the adult teeth emerge, they have bone to hold them in place. If this is not done, the teeth will simply become loose and fall out. Proper timing of this intervention is critical to preserving as many permanent teeth as possible.

After alveolar bone grafting is done, the craniofacial orthodontist will work to straighten all of the teeth. This is done with braces, just as with children who do not have a cleft. Future surgical intervention is delayed until adulthood is reached, usually around 16-18 years old. Further surgery to straighten the nose and open the airway should be considered. Some cleft patients need to have their jaw bones adjusted so that their teeth will come out straight. This type of surgery is called oral surgery or orthognathic surgery. Not every cleft patient needs this surgery, so it is important to be evaluated by the clinic team members so that all information can be gathered.

Patients who are in the Cleft Lip and Palate Clinic are usually discharged from care after the age of 18 or when they go to college. All of their care should be concluded by this time. If, however, there are things that they are unhappy with, such as the appearance of their lip or nose, surgical revisions can always be considered.

Living & Managing

Living with cleft lip and/or palate can be challenging for children. The stigma of an abnormal appearing lip or speech production can be difficult to cope with. With attentive team care
and intervention (usually surgical), these children do very well and develop normally. It is a long road of treatment, and it is important for both the patient and family to understand that they are not alone.

The Cleft Lip and Palate Clinic team has the resources and time required to help them through the process. We offer a support group for parents of children with cleft lip and palate and a camp for children with cleft lip and palate.

COMPLETE CLEFT LIP AND PALATE

- From birth to 3 months of age, affected infants receive support from the feeding nurse specialist, and airway management if required.
- In wide (>1 cm) clefts, pre-surgical lip taping, oral appliance insertion, or pre-surgical nasal alveolar moulding (PNAM) is also done between birth and 3 months of age prior to definitive cleft lip repair, which is performed as the second stage of cleft lip repair at 6 to 7 months of age. Infants with narrow (<1 cm) clefts receive definitive cleft lip repair without any prior pre-surgical procedure at around 3 months of age.
- In the presence of Eustachian tube dysfunction, bilateral myringotomy and tympanostomy tube (T-tube) (Shepard type) placement is performed concurrently with definitive cleft lip repair.
- Between 10 and 14 months of age, affected infants undergo palatoplasty and insertion of a long-lasting T-tube, with additional V-Y columellar advancement (lengthening of short columellar skin, performed by advancing skin from the central lip on to the columella with a V-shaped end and closing the lip, resulting in a Y configuration) in those with bilateral complete cleft lip and palate who have not received or responded to primary techniques (e.g., PNAM).
- Between the ages of 2 and 5 years, speech and language therapy is initiated, with additional surgery aimed at
improvement of speech for those who find initial therapy insufficient in the management of hyper-nasal speech.

• If there is gross aesthetic distortion or nostril stenosis with complete nasal obstruction, intermediate rhinoplasty is performed at any time after definitive cleft lip repair and prior to definitive septorhinoplasty.

• Between the ages of 8 and 11 years, alveolar cleft bone grafting with preparatory orthodontics is performed.

• Those with dentofacial malocclusion go on to receive definitive septorhinoplasty with additional orthodontics and orthognathic surgery at skeletal maturity.

**Isolated cleft palate**

• From birth to 3 months of age, affected infants receive support from the feeding nurse specialist, and airway management if required.

• Between 3 and 6 months of age, bilateral myringotomy and tympanostomy tube (T-tube) (Shepard type) placement is performed if Eustachian tube dysfunction is present.

• Between 10 and 14 months of age, affected infants undergo palatoplasty and insertion of a long-lasting T-tube.

• Between the ages of 2 and 5 years, speech and language therapy is initiated, with additional surgery aimed at improvement of speech for those who find initial therapy insufficient in the management of hyper-nasal speech.

• Those with dentofacial malocclusion go on to receive preparatory orthodontics with or without orthognathic surgery at skeletal maturity.

**Isolated cleft lip**

• From birth to 3 months of age, affected infants receive support from the feeding nurse specialist.

• In wide (>1 cm) clefts, pre-surgical lip taping, oral appliance insertion, or pre-surgical nasal alveolar moulding (PNAM) is also done between birth and 3 months of age prior to
definitive cleft lip repair, which is performed as the second stage of cleft lip repair at 6 to 7 months of age. Infants with narrow (<1 cm) clefts receive definitive cleft lip repair without any prior pre-surgical procedure at around 3 months of age.

- Between 10 and 14 months of age, those with isolated bilateral cleft lip who have not received or responded to primary techniques (e.g., PNAM) undergo V-Y columellar advancement (lengthening of short columellar skin, performed by advancing skin from the central lip on to the columella with a V-shaped end and closing the lip, resulting in a Y configuration).
- If there is gross aesthetic distortion or nostril stenosis with complete nasal obstruction, intermediate rhinoplasty is performed at any time after definitive cleft lip repair and prior to definitive septorhinoplasty in select cases of lip clefting.
- Those with isolated unilateral cleft lip go on to receive definitive septorhinoplasty at skeletal maturity.
- In isolated bilateral cleft lip, between the ages of 8 and 11 years, alveolar cleft bone grafting with preparatory orthodontics is performed, and at skeletal maturity, definitive septorhinoplasty is performed with additional orthodontics and orthognathic surgery in the presence of dentofacial malocclusion.

**SPECIALISED FEEDING**

Neonates with any form of oro-facial clefting may require extended postnatal hospitalisation due to feeding difficulties, although NG tube feeding is seldom required. Breastfeeding is possible with an isolated cleft lip. However, neonates with a cleft palate often cannot produce the negative pressures necessary for suction. Evidence-based guidelines exist for the use of breastfeeding in CLP patients, both pre-operatively and after cleft repair. These advocate both individualised support for nursing mothers and
monitoring newborn weight gain and hydration status. Nasal regurgitation is addressed with upright positioning of the infant during breast and bottle feeding. Feeding nurse specialists teach parents how to optimise bottle feeding using specialised fissured nipples and bottles controlling the flow rate of pumped breast milk or formula. Soft-bodied bottles are squeezed in synchronisation with infant sucking to reduce the effort of feeding and maximise the amount of feed entering the mouth. The infant should be burped during pauses, and feeding should not be continued for more than 30 minutes to avoid fatigue.

A Cochrane review of 5 randomised controlled trials examined various feeding interventions and their effects on weight of children at 6 weeks after surgery for repair of cleft lip and palate. While squeezable bottles appeared easier to use than rigid bottles, no differences in growth outcomes were noted. Maxillary plates also showed no evidence for improved growth at 6 weeks after surgery. Weak evidence exists to show that breastfeeding had a positive effect on weight gain after surgery when compared to spoon feeding.

The neonate will lose weight after birth (up to 10% of birth weight), but the birth weight is expected to be re-established within the first 2 postnatal weeks and the infant should go on to gain at least 25 g (1 ounce) per day thereafter.

**Airway management**

A small proportion of infants with complete cleft lip and palate and isolated cleft palate present with symptoms of severe airway obstruction requiring immediate airway management. Neonates with Pierre Robin sequence (triad of cleft palate, microgenia, and glossoptosis) may have upper airway obstruction. This is initially treated non-invasively with prone positioning, nasopharyngeal trumpet, and/or nasal CPAP. However, up to 23% of infants with micrognathia have a tongue-related obstruction requiring invasive intervention with endotracheal intubation if prone positioning, nasopharyngeal trumpet, and nasal CPAP are
ineffective. Surgical management of airway obstruction in micrognathia includes tongue lip adhesion, mandibular distraction osteogenesis (to move the base of tongue forwards and open the airway), and tracheostomy, which should only be undertaken once all other options have been exhausted.

A paediatric otolaryngologist can perform an airway evaluation with a dynamic flexible laryngoscopy to rule out other airway abnormalities and manage associated laryngopharyngeal reflux.

**Pre-surgical lip taping, oral appliances and pre-surgical nasal alveolar moulding (PNAM)**

Wide (>1 cm) clefts in complete cleft lip and palate and isolated cleft lip require a 2-stage cleft lip repair. This involves pre-surgical lip taping, oral appliance insertion, or PNAM in order to narrow the cleft prior to definitive cleft lip repair.

Daily lip taping with steristrips and benzoin to enhance adherence (as feeding causes wetting of the tape) is undertaken by the parents following instruction, and is used to protect the cheek skin in both bilateral and unilateral cleft lip deformities.

Although a variety of oral devices has been described, the Latham device is used to actively re-position the lateral alveolar cleft segments, while de-projecting the protruded pre-maxilla. The merits of pre-surgical pre-maxillary positioning using an intra-oral device are still debated. Associated maxillary growth inhibition is often cited, although there are insufficient long-term data to support either side.

PNAM is performed by the orthodontist and cleft surgeon.

- An appliance is placed within the cleft and adjusted weekly to approximate the alveolar segments, thus reducing the width of the cleft alveolus and corresponding soft tissues of the cleft lip. This is done prior to surgical reconstruction with definitive cleft lip repair, which is delayed during PNAM.
- The objectives of the PNAM technique are to elongate the columella, expand the cleft nasal mucosa, and improve
nasal tip symmetry. It takes advantage of the plasticity of the elastic cartilage of the nose, surrounding soft tissues (skin, muscle, mucosa), and bony maxilla in the early neonatal period. Serum maternal oestrogens in the infant are thought to induce a transient window of nasal cartilage pliability similar to that of the pubic symphysis in preparation for childbirth. Thus, PNAM should ideally be initiated within the first 6 postnatal weeks in order to utilise the early plasticity of the nasal cartilages.

- The PNAM appliance differs from traditional intra-oral alveolar moulding devices through the addition of nasal prongs. An orthodontist adjusts the acrylic intra-oral appliance by differential addition and removal of the leading edge of the maxillary segments at weekly appointments in order to move the alveolar segments together. Nasal stents are added to the intra-oral appliance when the alveolar segments are within 6 mm of one another. These stents are adjusted weekly to create a tissue expander effect on the length of the cleft-side columella and re-position the alar (from the Latin ‘ala’ or wing; the soft tissue and cartilaginous components of the nostril, which includes the lower lateral [alar] cartilages) hooding. This is purported to lead to improved nasal appearance, limited maxillary growth disturbance, and fewer subsequent procedures.

- Parental compliance is the most important factor in the successful completion of the PNAM treatment programme.

DEFINITIVE CLEFT LIP REPAIR

Narrow clefts (<1 cm) receive definitive cleft lip repair without any pre-surgical treatment (pre-surgical lip taping, oral appliance insertion, or PNAM).

Repair of the unilateral or bilateral cleft lip involves approximation of the 2 sides of the cleft lip using precisely designed segments of tissue, creating exact proportions of the underlying
oral mucosa, muscle, and lip contours. One Cochrane review suggests that the evidence supports the intra-operative use of an infra-orbital nerve block during cleft lip repair to assist with postoperative pain management. Additional well-designed studies would be needed to compare pain management options.

Unilateral cleft lip repair
- Surgical approach to the repair of a unilateral cleft lip is similar to that of a bilateral cleft lip except for the inherent asymmetries of a unilateral defect.
- The most important feature of unilateral cleft lip repair is creating symmetrical lip contours by lengthening the medial lip segment using a variety of techniques, including rotation-advancement flaps (Millard), triangular (Tennison/Skoog) designs, geometric designs, or a sub-unit (Fisher) approach.

Bilateral cleft lip repair
- The timing and technique for repair of the bilateral cleft lip are related to the extent of the deformity and surgeon preference. Numerous techniques have been described, including those of Millard, Cutting, and Mulliken.

Intermediate rhinoplasty

Although infrequently performed, alterations to the cleft lip nasal deformity can be made with intermediate rhinoplasty at any time after definitive cleft lip repair and prior to definitive septrhinoplasty in select cases of lip clefting: for example, where there is gross aesthetic distortion or nostril stenosis with complete nasal obstruction.

Intermediate rhinoplasty can be performed using an external or endonasal surgical approach. The nasal cartilages and soft tissue of the nostril can be treated with suturing and repositioning. Most surgeons delay septal surgery until skeletal maturity is reached, to minimise growth impairment of the nose and maxilla.
Bilateral myringotomy and tympanostomy tube (T-tube) placement

Infants with a cleft palate often have Eustachian tube dysfunction. Thus, all neonates with oro-facial clefts receive hearing screening. Bilateral myringotomy and T-tube (Shepard type) placement is performed following tympanograms and head and neck surgery assessment if there is evidence of Eustachian tube dysfunction. Audiometry is undertaken before and after T-tube placement.

Palatoplasty, long-lasting T-tube placement, and V-Y columellar advancement

The placement of longer-lasting T-tubes following bilateral myringotomy is performed concurrently with palatoplasty (cleft palate repair). Audiometry is undertaken before and after T-tube placement. Chronic otorrhoea is managed with antibiotic drops or tube change if bio-film accumulation is suspected. Any hearing loss not improved with T-tube placement is treated with hearing amplification following neuro-otological consultation to consider the various available options (e.g., hearing aid, FM system, soft-band bone-anchored hearing aid, cochlear implant).

Palatoplasty may be undertaken as a 1- or (rarely) 2-stage repair, and is performed through creation of oral tissue flaps from the palate and rotation of these palatal flaps to allow 3-layer closure (oral mucosa, soft palate muscles, and nasal layer).

- The 2-flap technique is the mainstay of unilateral palatoplasty, with the addition of a vomer (bony component of the nasal septum inferior to the perpendicular plate of the ethmoid bone and posterior to the quadrangular cartilage of the anterior nasal septum) dissection in bilateral palatoplasty.
- The 2-flap technique is undertaken as follows: after the palate is marked with the chosen design, the oral mucosal flap is incised down to the palatal bones and elevated in a sub-periosteal plane. The greater palatine vessels are
preserved and the flaps are mobilised to allow the oral layers to contact one another medially. The muscles of the soft palate (tensor veli palatini and levator veli palatini) are dissected to allow tension-free closure after the nasal layers are incised and closed. Absorbable sutures are used in closure, with attention given to minimising tension on the suture lines by appropriate flap mobilisation.

- Historically, a 2-stage (Schweckendiek) repair was undertaken. The first stage involved repair of the soft palate with placement of a prosthetic obturator for several years before completing the hard palate repair. The philosophy behind this technique was that maxillary growth would be improved without the development of early scarring around the palatal bony shelves.

- The type of palatoplasty (2 flap vs Furlow palatoplasty) has no effect on the incidence of otitis media or post-operative audiogram results. Furlow palatoplasty has been shown to exhibit less velopharyngeal insufficiency (VPI) but increased incidence of fistula.

V-Y columellar advancement (lengthening of short columellar skin, performed by advancing skin from the central lip onto the columella with a V-shaped end and closing the lip, resulting in a Y configuration) is undertaken concurrently in complete bilateral cleft lip and palate and isolated bilateral cleft lip where primary techniques (e.g., PNAM) have not been done or have been unsuccessful.

Speech and language therapy

Speech dysfunction related to oro-facial clefting is complex and should be analysed with the input of a speech and language pathologist.

Between the ages of 2 and 5 years, children with complete cleft lip and palate and isolated cleft lip therefore receive speech and language assessment with fluoroscopic speech examination and
nasopharyngoscopy in order to observe for velopharyngeal insufficiency (VPI) or dysfunction (VPD). Speech and language therapy is instituted if VPD is present.

Secondary speech surgery

If hyper-nasal speech is not responsive to speech and language therapy, secondary speech surgery is warranted between the ages of 2 and 5 years, following a VPD evaluation by the surgeon and speech pathologist.

Surgical options for the treatment of VPD include a superiorly based pharyngeal flap or dynamic pharyngoplasty. One meta-analysis, performed in 2012, examined the incidence of persistent VPI associated with either pharyngeal flap surgery or sphincteroplasty. Results showed slightly less VPI with pharyngeal flap surgery. Occasionally a palate-lengthening procedure (Furlow double-opposing Z-plasty) is performed.

A patient with a sub-mucous cleft palate may develop velopharyngeal dysfunction requiring surgical intervention. This may include a lengthening palatoplasty (Furlow double-opposing Z-plasty) or pharyngeal surgery (sphincter pharyngoplasty or pharyngeal flap procedure). Both are considered equally effective, although comparative studies have not yet been well-designed or powered enough to differentiate between the effectiveness.

Secondary speech surgery, which may be complicated by obstructive sleep apnoea, is performed in 10% to 30% of cases of complete cleft lip and palate.

Alveolar cleft bone grafting with preparatory orthodontics

Preparatory orthodontic maxillary expansion is performed prior to alveolar cleft bone grafting with iliac crest bone on eruption of the key permanent dentition. If maxillary segments and dentition on either side of the alveolar clefts are aligned, orthodontics can be postponed until bone grafting has been done and most of the permanent dental eruption is complete.
Orthodontics, orthognathic surgery, and definitive septorhinoplasty

Orthodontics and orthognathic surgery for dentofacial malocclusion, which may be necessary in order to obtain Angle Class I molar tooth relationships, are performed prior to definitive septorhinoplasty when the child has reached skeletal maturity.

In Angle Class I occlusion (named after the orthodontist Edward Angle), there is a normal relationship between the maxillary and mandibular first molars with the mesiobuccal cusp of the maxillary first molar resting in the mesiobuccal groove of the mandibular first molar.

LeFort advancements are sometimes performed to adjust the dentofacial relationship, particularly in the case of mid-face hypoplasia. Although providing increased mid-face protrusion, it is important to note that a moderate amount of relapse in the horizontal and vertical plane does occur.

Definitive septorhinoplasty is performed without orthodontics and orthognathic surgery in isolated unilateral cleft lip when the child has reached skeletal maturity.

Definitive septorhinoplasty is completed using an open approach in order to correct the asymmetrical upper and lower lateral cartilages and re-align the caudal septum that is deviated to the non-cleft side.

Symmetry of the alar base (from the Latin ‘ala’ or wing; the soft tissue and cartilaginous components of the nostril, which includes the lower lateral [alar] cartilages) is corrected with alar base excisions and, on the cleft side, is augmented with cartilage, bone, or allograft. Lateral crural strut grafting and nasal tip refining techniques (interdomal sutures and tip shield grafts) improve symmetry. Alar rim grafts and excision of nostril hooding are also effective in select cases. Osteotomies and dorsal refinement (excision or augmentation) are combined with spreader grafting between the upper lateral cartilages for enhanced support.
MAXILLARY GROWTH IN UN-OPERATED CLEFT PATIENTS

Patients with un-operated cleft palate demonstrate reduced maxillary length and retruded premaxillary position relative to the cranial base, which become increasingly obvious as the patients grow. In a study of 39 un-operated cleft palate cases from West China Stomatology Hospital, we observed reduced sagittal length and retruded position of the maxilla at the end of the growth period. Most un-operated cleft lip patients with or without alveolar involvement demonstrate normal facial projection, exhibiting only dental arch malalignment in the cleft region. In the case of un-operated unilateral cleft lip and palate, the maxilla demonstrates normal growth potential in the sagittal dimension, although the dental arch is typically straighter. By studying 24 un-operated unilateral complete cleft lip and palate patients, Capelozza Júnior et al. suggested that the position and growth amount of these cleft maxillae were similar to normal controls and that the dental arch was normal on the non-cleft side but collapsed medially on the cleft side. These authors further compared the maxillofacial growth of all types of un-operated clefs and found that un-operated cleft lip and alveolus patients demonstrated greater premaxillary projection, maxillary length (Ans-Ptm), labial tipping of the anterior teeth, ANB angle, and maxillary projection (NA-PA) with normal mandibular position and dimensions. In another study, un-operated unilateral cleft lip and palate patients demonstrated similar or even more protruded maxillary growth when compared to normal controls. These finding suggest that un-operated cleft patients possess the normal potential and mechanism for growth.

The impact of pre-surgical intervention on maxillary growth

Currently, pre-surgical nasal alveolar moulding (PNAM) is the most widely used orthopaedic technique for cleft correction. The alar cartilage is more pliable to the orthopaedic manoeuvre soon after birth, whereas by the age of 3 months, the cartilage
becomes rigid with reduced plasticity. PNAM can significantly improve the nasal symmetry, elongate the columella, bolster the alae, narrow the cleft and restore the arch form, demonstrating favourable immediate- and long-term outcomes.

In a follow-up study over 6–9 years, Bennum et al. found that patients who were treated with PNAM within 15 days after birth all maintained satisfactory nasal symmetry, requiring no further revision. Ezzat et al. compared measurements from 12 unilateral cleft lip and palate patients before and immediately after PNAM treatment and found that PNAM narrowed the alveolar cleft, increased the posterior width of the dental arch, uprighted the columella and improved the nasal symmetry. In particular, the increase in the height of the cleft side nostril was closely related to the PNAM treatment. Yang et al. reported similar results from 45 unilateral complete cleft lip and palate patients. Moreover, it was reported that 60% of cleft alveolus patients treated with PNAM did not require secondary bone grafting, and early restoration of the dental arch facilitated normal facial growth.

PNAM devices developed for bilateral clefts can hold back the protruded premaxillae, reduce the alveolar gap and non-surgically elongate the columella. In a large multicentre study sample, Ross concluded that orthopaedic correction of the premaxillae failed to stimulate maxillary growth, and thus was not necessary, and that the reduction of the cleft was due to the transverse growth of the maxillae. In contrast, Ras et al. considered distraction forces that may disturb the growth centre in the premaxillae and interfere with midfacial growth.

**Surgical timing and maxillofacial growth**

**The timing of cleft palate repair**

In a large sample study including over 2,000 cases, Koberg and Koblin reported similar postoperative maxillary growth rates among patients who took either early (<1 year old) or delayed palatoplasty, and most of the observed midfacial retrusion occurred
between 8 and 15 years of age. However, this surgery should generally be postponed until 15 years of age in order to completely avoid growth interference, including impacts on speech development and sociopsychological health. Early palatoplasty produces maximal growth inhibition in all dimensions, and the surgical region has been shown to grow more slowly than the surrounding tissue. In particular, the severity of growth inhibition is positively related to the timing of surgery and the extent of scar contracture.

**The timing of cleft alveolar repair**

Initially, Ross considered the alveolar bone graft procedure to be harmless to maxillary growth because the grafting area was not a growth site. However, in his follow-up large-sample, multicentre study, he found that cleft alveolar repair resulted in reduced maxillary height. Thus, Ross proposed postponing cleft alveolar repair until after 9 years of age.

By the 1970s, secondary bone grafting had been accepted by most surgeons for correction of the alveolar cleft. The best timing for this procedure is approximately 9–11 years of age, at which time the root of the permanent canine has formed 1/3 to 2/3 and the crown is still partially covered by bone. In a cephalometric study, Gesch et al. suggested that secondary bone grafting has no influence on sagittal growth of the maxillae. Levitt et al. reported that maxillae tended to retrace after alveolar bone grafting, although such trends existed prior to the bone graft and did not change significantly after the secondary bone graft.

**Surgical design and maxillofacial growth**

**Cleft lip repair**

In a comparison of 84 cleft lip patients (with or without cleft palate) and normal controls, we found that the extent of growth inhibition after primary lip repair was related to the severity of the original deformities. Among patients with cleft lip and alveolus, the influence of primary cheiloplasty was mainly restricted to the
incisors and alveolus in the cleft site, and the shape and position of the maxillae were similar to those of controls. However, in the case of cleft lip and palate, maxillary retrusion and reduced maxillary length were observed after primary lip repair, whether the cleft palate was repaired or not. Thus, we inferred that the severity of the original defect and displacement of the cleft maxillae was associated with more significant growth inhibition after primary cheiloplasty.

In animal models, we found that both Millard and Tennison lip repairs produced shorter, wider, and posteriorly displaced maxillae, and Tennison’s technique tended to cause more problems to the anterior tooth and alveolus.

Shortly after bilateral cleft lip repair, the protruded premaxillae move backwards very rapidly and reach a normal position by adulthood. The posterior part of the maxillae is somewhat retruded but shows normal dimensions. Specifically, this moulding effect is a result of the lip pressure from suturing bilateral lateral labial components to the middle. Secondary alveolar deformities due to inappropriate lip pressure may be extremely difficult to correct.

**Cleft palate repair**

Koberg and Koblin closely examined the maxillofacial growth of 1,033 cleft palate patients and found that Veau’s pushback technique and Langenbeck’s technique with relaxing incisions were most detrimental to growth. Pichler introduced the vomer flap into cleft palate repair in 1926 (ref. 45) but reported a high incidence of premaxillary retrusion, which was avoided when the flap elevation area was restricted away from the vomeropremaxillary suture.

In a large-sample, multicentre study in 1987, Ross found that repairing the soft palate only resulted in decreased posterior maxillary height but normal sagittal length and position of the maxillae. In addition, he suggested that the technique used for soft palate repair was unrelated to maxillary growth. In 2013, Jackson et al. examined 1,500 patients treated with Furlow palatoplasty and
reported no significant midfacial retrusion or crossbite; only 14% patients in this study required LeFort I advancement. In another consecutive series of 33 double-Z palatoplasty-treated patients from Florida, only 1 bilateral case required maxillary advancement.

Chate et al. reported that patients treated with his intravelar palatoplasty without lateral relaxing incisions demonstrated more favourable maxillary growth when compared to European data.

**Pharyngoplasty**

Currently, data discussing the relationship between pharyngoplasty and growth remain limited. Long et al. found that patients who underwent pharyngoplasty between the ages of 5–7 years demonstrated increased lower facial height, posteriorly inferiorly rotated mandibles and lingually tilted incisors. Voshol et al. studied 580 fully developed cleft patients and found that 19% of those who underwent pharyngoplasty required LeFort I surgery, while this percentage among those who did not undergo pharyngoplasty was only 8%. In contrast, in a series of 48 cleft palate only patients, Heliövaara et al. found no significant difference in maxillofacial growth between patients who received pharyngoplasty and those who did not.

**Treatment protocol and maxillofacial growth**

Schweckendiek first proposed repairing the soft palate first and delaying the hard palate closure. After modifications over half a century, this protocol has achieved satisfactory results in preventing growth inhibition. In Schweckendiek’s 25-year follow-up study, over 60% of his patients demonstrated normal maxillary growth, and Olin reproduced Schweckendiek’s success. The Zürich centre (hard palate closure at 7 years) and the Göteborg centre (hard palate closure at 9 years) both achieved satisfactory facial growth using their modified two-stage palate repair protocols.

In 1991, Semb evaluated another two-stage protocol, the Oslo protocol, in which hard palate closure was performed at the same
time as lip repair at 3 months, and then the soft palate was repaired at 18 months. He found that patients treated this way tended to have retruded maxillae and mandibles and reduced posterior facial height. In contrast, Mølsted et al. found that the Oslo protocol produced the most favourable maxillofacial contour in comparison to other Eurocleft centres. del Guercio et al. compared patients from Oslo and Milan (where lip repair and soft palate closure were performed at 4–6 months, and hard palate repair and gingivoperiostoplasty were performed together at 18–36 months) and found no difference in maxillofacial growth at 5 years of age.

In a 5-year study, we found that early soft palate closure significantly reduced the width of the hard palate cleft, but did not reduce the final growth inhibition. In addition, sagittal and vertical growth inhibition was similar between one-stage and two-stage treated patients.

This result suggested that it was the timing of hard palate closure, instead of the sequence of hard or soft palate repair, that determined the postoperative growth. Data from both Mommaerts et al. and Richard et al. further support this statement, as these authors found that one-stage and two-stage protocols showed no difference in postoperative maxillary growth because the hard palate was repaired at the same time in both protocols.

**CLEFT LIP AND PALATE CLINIC TAKES A WHOLE-CHILD TREATMENT APPROACH**

Cleveland Clinic Children’s provides comprehensive, sophisticated care for youngsters with all types of cleft lip and palate abnormalities. Its multidisciplinary Cleft Lip and Palate Clinic brings together a team of experts to address diverse needs associated with these deformities while minimizing stress on patients and their families.

“Children with cleft lip and palate may require multiple operations by their teenage years,” says pediatric otolaryngologist Brandon Hopkins, MD, one of several at Cleveland
Clinic Children’s with fellowship training in cleft and craniofacial care. “We keep the number of operations as low as possible by coordinating tests and procedures.

“Our overall goal is to deliver complete care from birth, enabling the child to graduate into life without a noticeable cleft or nasal deformity.”

Enduring care from a wide-ranging team

A simple cleft lip may be repaired in a single operation performed by a cleft surgeon. More complex deformities, often including a cleft palate, require care from a team of specialists throughout childhood. Cleveland Clinic Children’s is structured to provide all services required by patients with any level of deformity. The Cleft Lip and Palate Clinic team includes:

• Cleft surgeons (plastic surgeons and pediatric otolaryngologists) to ensure the child’s cosmetic appearance and functionality by repairing the cleft lip, performing palate repair, reconstructing ear deformities, correcting velopharyngeal insufficiency, addressing nasal deformities and performing orthognathic surgery. The pediatric otolaryngologist also may insert and replace ear tubes to minimize hearing loss, address nasal breathing issues and sleep apnea, and manage voice and swallowing concerns.
• Orthodontists to evaluate for braces, timing of alveolar bone grafting and orthognathic surgery
• A general pediatrician, who helps oversee the patient’s overall health during cleft therapy
• A speech pathologist to assist with feeding concerns, promote normal speech and help determine the procedures necessary to produce a normal voice
• An audiologist to conduct hearing tests and optimize hearing with hearing aids
• A geneticist to check for potential underlying causes of the cleft and provide comfort and counseling to the family
• Psychologists and social workers to address the often complex social situations and psychological stresses families face

Families meet with all specialists and receive a recommended treatment plan in a single morning.

**Sophisticated repair techniques**

When performing cleft lip repair, Dr. Hopkins re-establishes continuity between the mucosa, muscle and skin of the lip and then recreates the floor of the nose. A primary tip rhinoplasty may be done at the same time as cleft lip repair to improve cosmetic appearance and function of the deformed nasal cartilages. The primary goal of cleft palate repair is to reconstruct an intact palate to allow for normal speech and swallowing development while ensuring harmonious facial growth and minimizing the incidence of oronasal fistulae. This is done by closing the cleft palate with oral and nasal mucosal flaps while reorienting the muscles of the soft palate.

In patients with cleft palate, hearing loss is often addressed with ear tubes. Multiple tubes may be needed over the first several years of life. Dentition is managed as the patient grows. Interventions may include palatal expanders, braces and alveolar bone grafting using bone harvested from the hip to close the defect in the upper dental arch. Patients may also need orthognathic surgery to correct malocclusion inherent in their deformity or related to a previous surgery. The jaw may be lengthened to provide more room for the tongue. All surgeries and procedures, including hearing testing, are carefully coordinated and performed under one administration of anesthesia whenever possible.
Preoperative and postoperative photos of two infants with cleft lip and palate who underwent repair surgeries at Cleveland Clinic Children’s. The top patient (left complete cleft) was photographed at 2 weeks of age and then at eight months after surgery. The bottom patient (left incomplete cleft) was photographed at 3 weeks of age and then at four months after repair.

Outcomes to stand by

Although consensus is lacking on which palate repair technique yields optimal speech results with the lowest likelihood of oronasal fistulae, a review of all 64 patients who underwent cleft palate repair by the Cleft Lip and Palate Clinic team between September 2010 and December 2013 revealed no fistulae.

APPLYING THE PRINCIPLES OF ANATOMIC SUBUNIT REPAIR FOR VERY SHORT LIPS

The lateral lip element is commonly short in vertical height, especially in adults with untreated clefts. It has been pointed out that in order to gain the necessary height, it may be necessary to move the position of Noordhoff’s point more laterally, and in doing so compromise lateral lip transverse length to achieve vertical height. Pool stated that vertical height of the lateral lip is difficult to obtain with the rotation-advancement repair only when the lateral lip is short in both its horizontal dimension and its vertical height. It was concluded, that 63% patients had combined height and transverse length deficiencies of the lateral lip. For these patients with considerable deficiencies, an inferior triangle, needs to be incorporated for successful management of the lateral lip. As described by Noordhoff, when the discrepancy of the lip height is large (>3 mm in our experience), a triangle needs to be incorporated above the white roll as it is not possible to achieve the necessary vertical height by only rotation flap. This lower triangle also decreases the tendency for peaking of the lateral bow. Fisher has previously highlighted the key concept that design of
the triangle should vary according to the length of the lateral lip element. Three situations can exist: Lateral lip is normal, long or short. shows the design and placement of the triangle in different situations. If the lateral lip is normal, which means that adequate tissue is present in the lateral lip element, and the necessary lip height can be achieved without excessive lateral shift of the Noordhoff’s point, then the triangle is incorporated in line with the future philtral column. This will avoid the use of superior tissue instead of lateral tissue, which is adequate in this situation. If the lateral lip is very long, meaning that excessive tissue is present on the lateral lip element, then the triangle is sloped down, so that it will also marginally negate the amount of gain from the Rose Thompson effect. Furthermore, a wedge of tissue may be excised at the superior margin as needed to match the necessary height. The most challenging, and most common, situation is when the lateral lip is very short. In this situation, the lateral lip is deficient and hypoplastic. In this case if the Noordhoff’s point is moved too laterally to gain the necessary height, it will further compromise the already deficient lateral lip. Hence, in this situation, the design of the triangle is done to utilize the superior tissues rather than the lateral tissues to avoid hypoplastic lip. This helps to maintain Noordhoff’s point medially, saving the precious lateral lip vermilion. Again, it has to be noted that the above mentioned three situations are only pertaining to the lateral lip and the discrepancy in lip height on the medial element in all the three situations is more than 3 mm. shows the application of a lower triangle to the advancement flap. In this patient, the greater lip height is 10 mm and lesser lip height is 6 mm. The discrepancy is 4 mm, and the lateral lip element is also very short. With the rotation of the medial flap, 1.5 mm will be gained from the columellar extension and backcut, and 1 mm will be gained from the Rose Thompson effect. This necessitates incorporation of a triangle of 1.5 mm to gain the necessary length. In a very short lip such as this, the design and placement of a triangle above the white roll are the key to manage the lateral lip without sacrificing
excess tissue. The triangle is placed as shown in , which utilizes the tissue superior to the white roll instead of the lateral lip element. This technique of placement of the triangle has resulted in saving 1.5 mm of lateral lip element (distance X in the , which will be equal to the base of the triangle), which otherwise would have been sacrificed to gain the necessary length shows the application of this principle and the final result with 1 year follow-up. Note the balance of the lip is maintained even in very short lateral lip element. In this case, the surgeon tried to gain the necessary length by compromising the lateral lip.

**Avoiding whistle deformity**

Vermillion notching is most often caused by inadequate excision of the cleft tissue and persistent tissue deficiency at the vermillion border. The best way of preventing this deformity is to mark Noordhoff’s point to have adequate vermilion at the line of repair. Alignment and repair of the pars marginalis portion of the orbicularis muscle at the lip margin is essential in gaining adequate volume. Furthermore important are precise closure of the vermillion, while incorporating a lateral “v” flap, and accurate re-approximation of mucosa.

**Managing excess vermilion**

The problem of excess vermilion on the lateral lip element arises most often in incomplete clefts. This often results in a bulky lateral lip. In most instances, this problem is noted during the final stages of the repair, and there is evident mismatching of the vermilion. This problem can be solved by resecting a wedge of mucosa in the sulcus on the advancement flap and not near the junction of wet and dry vermilion, thus removing the excess. Care has to be taken that the mucosa and muscle have been adequately dissected. Furthermore, it is prudent not to inject excess local anesthetic solution into the lip tissues prior to surgery, causing increased tissue swelling and difficulty in assessing amount to be resected.
Results

The unilateral cleft lip repair and technical refinements have been incorporated in more than 500 patients by the primary author. The specific technique for saving tissue and volume of the lateral lip element in wide clefts has been practiced by the primary surgeon in 100 patients with discrepancy between the greater and the lesser lip of more than 3 mm. Although, measurements were not analyzed to note whether the difference after using this modification was statistically significant, the results with this technique were visually more pleasing. A more balanced lip is achieved, as proper design of the triangle saves precious tissue on the lateral lip element.

Discussion

A new era began in the art of cleft surgery when Millard introduced his “rotation - advancement” technique at the First International Congress of Plastic Surgery in Stockholm in the year 1955. This technique has been modified countless times, but the principles of rotation and advancement have persevered as surgeons have sought to refine various elements in order to achieve an optimal repair.

Historically most of the modifications have been focused on gaining the necessary length on the medial side. There is far less literature on the management of the lateral lip element, especially in situations where the discrepancy between lip height of the cleft side and noncleft side is high. It has been observed at our center, where we do not practice presurgical orthopedics, that rotation of the medial segment is often not enough to gain the necessary length.

When a large discrepancy exists between the greater lip height and lesser lip height, a lateral triangle is inserted above the white roll to gain necessary length on the medial segment and prevent a short repair. This also produces a small amount of tension on the lip and accentuates the pout. Fisher has previously described various methods of incorporating this triangle in varying lengths
of the lateral lip as part of his anatomic subunit repair. This principle can be utilized in rotation-advancement technique as well when the discrepancy between the greater and lesser lip length is more than 3 mm.

If the lateral lip is normal in length, then the triangle is incorporated along the future philtral column. If the lateral lip is very long, then the triangle is sloped downwards to negate the additional gain by Rose Thompson effect. In very short lips, the triangle is placed such that it utilizes the superior tissue rather than lateral tissue and prevents excessive resection of the valuable lateral lip element.

The incisions can also be sloped more, almost 90° as they cross the white roll, which provides with an additional 1 mm length of the lateral lip element due to Rose Thompson effect. Now the question is, why not follow anatomic subunit principle for all lips where the vertical and transverse discrepancy is more? In most cleft centers, especially in developing world, presurgical orthopedics is not practiced. Hence, very high discrepancy is observed between the greater lip length and the lesser lip length, sometimes more than 3 mm.

In such situations, it may not be possible to gain the necessary length by using the principles of anatomic subunit closure alone. Combination of rotation-advancement and anatomic subunit principles can offer a better result than just one technique; this includes the preservation of the lateral lip. Alternatively, two triangular flaps can be inserted, one above the cutaneous roll, and the other below columella as described by Skoog.

This technique gives a straight line closure and can avoid the drawbacks of rotation-advancement technique. Cutting and Dayan reported that although the lateral lip element is observed to be significantly short immediate postoperatively, this deficiency significantly improves in long-term follow-up. The results were reported using black and white photographs. The use of photographs for reporting results has long been debated for
accuracy as it is very challenging to get exact photographs, especially in children. We also believe that this improvement will be marginal and all efforts have to be made to conserve the lateral lip element. This fact was also pointed by Farkas et al., who concluded in their anthropometric studies that the total growth remaining in the upper lip height as well as vermillion was very small after 1 year of age which explain that a cleft lip repair in early life may retain its quality fairly well throughout life. But, the problem of adult untreated cleft lip repair is highly prevalent in the developing world. In such situations, all efforts need to be made to save all the tissue and simultaneously achieve a pleasing result.

TREATMENT OPTIONS

The birth of a child is a joyous event full of new challenges and difficulties for the parents. For the parents of a child born with a facial deformity such as cleft lip and/or palate, the challenges may seem overwhelming. To provide parents with hope and expert medical care, Plastic and Craniofacial Surgery for Infants and Children specializes in the treatment of children born with cleft lip, cleft palate and other facial abnormalities.

The Importance of Early Treatment

A child born with a cleft lip and/or palate can begin early treatment to enable the restoration of as normal an appearance as possible before the child begins peer interaction. The early treatment minimizes the social discomfort a child born with a facial deformity may feel as the child grows. In addition to correction of lip, palate, nose and facial structures, the team will work to prevent hearing and speech difficulties that may accompany such anomalies. The specialized approach to correcting deformities of the lip and palate is unique in the care and attention given to enhancing a child’s quality of life as soon as possible.

While it is known how cleft lip and palate deformities occur before birth, it is not known why they occur. Yet, one in 500 births
results in a deformity of the lip and/or palate. As the cause and possible preventative treatment for this facial deformity is sought, the team at Medical City Children’s Hospital of Dallas is ready to provide the best possible treatment for your child and to make possible the smile he or she was meant to have.

Children with a cleft palate are particularly prone to ear infections because the cleft can interfere with the function of the middle ear. To permit proper drainage and air circulation, the ear-nose-and-throat surgeon on the Cleft Palate Team may recommend that a small plastic ventilation tube be inserted in the eardrum. This relatively minor operation may be done at the same time of the cleft repair.

Cleft Lip Repair

To repair a cleft lip, the surgeon will make an incision on either side of the cleft from the mouth into the nostril. The goal in lip repair is to create a structure of normal appearance and function. This is accomplished by reconstructing the normal anatomical landmarks such as the philtral ridge, vermilion cutaneous border, nostril floor, and orbicularis muscle for lip function. The inclusion of muscle in the prolabium of the bilateral cleft lip is important to bring motion to a structure which otherwise would remain virtually without animation. The surgeon will then turn the dark pink outer portion of the cleft down and pull the muscle and the skin of the lip together to close the separation. Muscle function and the normal “cupid’s bow” shape of the mouth are restored. The nostril deformity often associated with cleft lip will also be improved at the time of lip repair.

Cleft Palate Repair

Repair of the palate is directed at producing normal speech, restoring Eustachian tube function, attaining closure of oronasal fistulas, and minimizing alterations in maxillary growth. In some children, a cleft palate may involve only a tiny portion at the back of the roof of the mouth; for others, it can mean a complete
separation that extends from front to back. Just as in cleft lip, cleft palate may appear on one or both sides of the upper mouth. The soft palate may be repaired at the time of lip repair if it is involved, also. This is usually done at 3 months. The hard palate is done when the baby is older and the teeth have erupted (avoiding growth disturbance to the teeth and maxilla), usually at the age of 18 months.

To repair a cleft palate, the surgeon will make an incision on both sides of the separation, moving tissue from each side of the cleft to the center or midline of the roof of the mouth. This rebuilds the palate, joining muscle together and providing enough length in the palate so the child can eat and learn to speak properly.

**Alveolar Bone Grafts**

Children with clefts including the alveolar dental arch will require bone grafting to maintain the dental arch and allow the ingrowth of teeth immediately adjacent to or within the cleft. The timing of this procedure varies but is at approximately age six to eight and is determined by dental x-rays which show the development of the permanent teeth. Cancellous bone from the iliac crest will be inserted into the alveolus once the dental team has aligned the arch or growth and lip closure have brought the alveolar ends into approximation. At this time, any residual oronasal fistulas can be closed as well.

Children generally spend one night in the hospital in order to insure they are taking fluids and ambulating with help. Complaints of hip discomfort and reluctance to walk are common. A soft, blenderized diet and restriction from strenuous activities is recommended for 10 days.

**Velopharyngeal Insufficiency**

In a small percentage of cases, some children, in spite of cleft palate repair, will continue to exhibit hypernasal speech. This defect can be demonstrated by good physical examination, speech pathologist evaluation, cine-fluoroscopy and nasal endoscopy. The
fogged mirror test, conducted with the patient’s nose alternately open and occluded, is one of the simplest methods used to document nasal air escape, which in turn demonstrates soft palate dysfunction. The condition known variously as velopharyngeal insufficiency (VPI) or velopharyngeal dysfunction (VPD) may be seen after cleft palate surgery or noted after adenoidal tissue undergoes involution as the child grows. In some patients who have undergone palatal repair, short sentences may sound relatively normal. Long, sustained speech, however, may deteriorate, resulting in increasing hypernasality as the palate tires. In those cases, consultation with the “Cleft Palate Team” will most likely result in a recommendation for some type of surgical intervention to help with the soft palate closure.

Surgical procedures for correction of velopharyngeal insufficiency include pharyngeal flaps, in which the posterior pharyngeal wall is elevated and sutured to the soft palate, thereby reducing the gap present from the short or poorly functioning soft palate. Other surgical approaches may be used which involve some alteration of the anatomy surrounding the soft palate and posterior pharyngeal wall (described as a “pharyngoplasty”). A variety of approaches have been described and the choice rests with the surgeon in consultation with the speech pathologist. In the past, injectable Teflon into the posterior pharyngeal wall has been described, but is not currently of significant use.

The age and timing of this surgery varies and has been reported in very young children before school age and late in teen years. Early diagnosis with good speech therapy to produce maximal function of the soft palate, however, is important as a prelude to surgical intervention. Once the speech pathologist can no longer produce correction in speech patterns and sounds, consideration for surgical repair should be made. This again is the advantage of a coordinated approach to children with clefting deformities by the coordinated and active “Cleft Palate Team.”
INTRODUCTION

Cleft lip and palate is a major congenital structural anomaly that is notable for significant lifelong morbidity and complex etiology. The prevalence of orofacial clefts varies from 1/500 to 1/2500 births depending on geographic origin, racial and ethnic backgrounds and socioeconomic status. In many parts of the world orofacial clefts go unrepaired. Individuals who do have their clefts repaired undergo surgical, speech, dental and psychological therapies. These outcomes, along with the relatively high prevalence of orofacial clefts, emphasize the importance of understanding the underlying causes of orofacial clefts.

The causes of orofacial clefts are complex, involving both genetic and environmental factors. That genes play an almost deterministic role in the development of normal craniofacial structures is evident from observations of monozygotic twins, where the majority are phenotypically indistinguishable. At the same time, this genetic program is exquisitely sensitive to postconception disturbances in genes or the environment, as evidenced by the identification of numerous teratogens that lead
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to orofacial clefts. Thus, the complex etiology of clefts affords ample opportunities to identify genes, explore gene-environment interactions and learn more about human embryology and its disturbances.

Facial Development

Normal facial development begins with migrating neural crest cells that combine with mesodermal cells to establish the facial primordia. The growth of facial primordia from undifferentiated mesenchymal cells into the finely detailed structures of the mature head and face is largely determined genetically. These processes are known to be dependent on a spectrum of signaling molecules, transcription factors and growth factors. A subset of genes already shown to play an important role in the development of the head and with particular relevance to development of the lips and palate.

Additional growth and signaling factors that play a role in facial development include JAGGED1, sonic hedgehog, patched, CREB-binding protein, GLI3, FGFR1, CASK, treacle and FGFR2. Other transcription factors involved include DLX5/6 and PAX3. Extracellular matrix proteins such as COL2A1, COL1A2, COL11A2, PIGA, ±V integrin, glypican 3, fibrillin and aggrecan are essential as well. This ever-expanding catalog of molecules highlights the complex genetics of facial clefts.

When the structure or expression of these genes is altered, a cleft of some type may occur. Orofacial clefts can be divided into four groups: non-syndromic cleft lip with or without cleft palate, non-syndromic cleft palate only, syndromic cleft lip with or without cleft palate and syndromic cleft palate only. The term ‘non-syndromic’ is restricted to cleft cases where the affected individuals have no other physical or developmental anomalies and no recognized maternal environmental exposures. Cleft cases are further divided by which palate is affected. Genetics and embryology suggest that clefts of the primary (hard) palate that involve the lip and/or palate are different in mechanism from
clefts that affect the secondary (soft) palate. At the present time, most studies suggest that <"70% of cleft lip with or without cleft palate (CL/P) cases are non-syndromic . The remaining 30% of syndromic cases can be subdivided into chromosomal anomalies, >300 different recognizable Mendelian syndromes, teratogens and uncategorized syndromes. Recent reviews of birth defects secondary to chromosomal anomalies identified five regions in which there was a significantly higher frequency of clefting associated with either specific deletions (4p16–14, 4q31–35 or 1q25) or duplications (3p26–21 or 10p15–11) than would be expected from the background frequency. Nonetheless, it is also apparent that deletions or duplications of portions of every chromosomal arm, including the X chromosome, have been associated with clefts, suggesting that many genes are involved in facial development and that cleft lip and palate can represent a common end-point for disruption of facial processes. It is likely that in the future CL/P will be subdivided further based on underlying genetic etiologies or better phenotypic descriptors. Some attempts at clinical distinction already include associations with hypoplasia of the orbicularis oris muscle or handedness .

Genetics of Orofacial Clefts

The study of orofacial clefts has a rich history in human genetics and provides a model for complex disease study in general. An inherited component for clefts was first widely recognized through the work of Fogh-Andersen in his thesis of 1942 . Genetic factors in clefting are now well established through segregation analysis as well as through twin studies . Additional genetic linkage and association studies are now being used to identify these genetic factors.

Although genetic linkage studies of CL/P have been limited by insufficient numbers of families and genotyping resources, a few efforts using candidate genes or loci have been reported. Studies using from 1 to 40 families have suggested loci for clefts on chromosomes 2, 4, 6, 17 and 19. Linkage has been excluded at
these same loci in other data sets. These inconsistent linkage results reflect the small number of families studied and probable locus heterogeneity. Thus, while the studies are useful for preliminary data, they need to be replicated on far larger sample sets. Only loci on 6q have consistently shown linkage to CL/P, beginning with studies from Denmark and subsequently in Italy.

CLEFT PALATE

Cleft palate is a relatively common congenital malformation with an incidence of between one and two in 1000 live births. The severity is variable and ranges from occult to overt; the causes are numerous and range from syndromic/genetic to teratogenic/environmental to isolated/idiopathic; the management is often complex and ranges from straightforward surgical closure to multidisciplinary surgical and cleft team care. Furthermore, the socioeconomic burden posed by orofacial clefting is significant; according to the Healthcare Cost and Utilization Project, these costs often exceed $100 million annually.

Surgical repair of cleft palate is currently the clinical standard of care. In these cases, affected children often require multiple physiologically challenging operations to address not only palate closure, but also associated problems with speech, dental occlusion, fluid buildup within the ears, and maxillary growth deficiency. As such, recent research has made strides at elucidating both the biology underlying normal palate development and the pathogenesis of cleft palate in attempts to improve the way cleft palate is managed in the future.

The process of palatogenesis depends on highly coordinated, anatomically specific and precisely timed molecular signals for normal development. Among them, cell migration, proliferation, fusion, apoptotic, and differentiation events contribute to the complexity of craniofacial organization. In addition, multiple signaling pathways including sonic hedgehog, FGF, and transforming growth factor-α (TGF-α) signaling complement each
other. Aberration from any of this programming is likely to lead to pathogenesis of the palate, namely cleft palate. Much of our knowledge of craniofacial clefting arises from patient studies and selected animal models. Study of these models has revealed a well orchestrated sequence of events that has now been well documented.

Throughout the process of palatogenesis, molecular signaling between the mesenchymal and epithelial layers guides appropriate development. This paradigm of development is not specific to the palate, but the pathways underlying these interactions are temporally and spatially distinct and have not been clearly elucidated in the past. Learning what molecules are involved during the well-described classical stages of palate development allows for identification of missteps that may arise.

In an effort to further understand one aspect of the complex biology underlying normal palatogenesis, we recently investigated the role of GSK-3β in the process. In this system, we showed that cleft palate resulting from loss of GSK-3β could be rescued by protein stabilization during a short, specific window in embryogenesis in the mouse model. In this series of experiments, a mutant mouse carrying alleles for GSK-3β<sup>FRB*</sup> was injected with rapamycin to inducibly stabilize GSK-3β during various 2-d windows in embryogenesis within the timeline of palate development. This transgenic mouse was engineered to carry alleles for GSK-3β such that without drug addition the unstable FRB* tag would necessarily cause protein degradation, and the mouse would exhibit a null mutation phenocopy (i.e., cleft palate). Subsequent histologic analysis revealed that without rapamycin, no GSK-3β<sup>F+/F−</sup> embryos were able to rescue the cleft palate; however, with rapamycin injection of the pregnant dam between E13.5 and E15.0, the majority of conditional GSK-3β<sup>F+/F−</sup> mutant animals were able to be partially or completely rescued from their cleft palate in utero. Rescue was not seen in other injection windows during palatogenesis, suggesting a critical role for GSK-3β function in normal palatogenesis between E13.5 and E15.0 in the mouse model.
GSK-3β has been implicated as a key regulator of a wide variety of developmentally important molecular pathways including Wnt, nuclear factor of activated T-cells (NFAT), Hedgehog, and insulin signaling. These signaling pathways are essential components of many biologic responses and associated diseases, including embryonic development and cell fate determination, diabetes, neurodevelopment and neurodegeneration, psychiatric disorders, cell cycle regulation and cancer, hematopoiesis, and immunity. GSK-3β has not previously been implicated in the development of the mammalian palate. However, because it is positioned at the “node” of so many significant developmental pathways, analysis of GSK-3β function during palatogenesis will likely provide important insight into this common birth defect. In addition, because of the “promiscuous” nature of GSK-3β, it has become a potentially important therapeutic target. Thus, many potent and selective inhibitors of GSK-3β function are being developed by the pharmaceutical industry.

Although GSK-3β mutations have not been documented to be a cause of human orofacial clefting, our recent findings suggest it is clinically relevant because of the potential to devise methods for improved treatments, including in utero rescue, for human orofacial clefting. Ongoing investigations of GSK-3β’s role in palatogenesis promise future clinical applicability, because it has the potential to reveal signaling pathways underlying cleft formation and lay the groundwork for potentially improved treatments using small molecules.

Within the last several years, there have also been numerous reports of TGF-α3’s role in palatogenesis. In 2001, Koo et al. first reported a novel mutant mouse that could potentially be used as an animal model for the study of cleft palate. In this report, they described a mutant mouse that was homozygous null for TGF-α3 on both alleles. They reported 100% clefting of the palate in the homozygous TGF-α3 knockout pups. Changes in TGF-α3 have also been associated with orofacial clefting in humans, further bolstering the significance of Koo et al.’s report. Subsequent to the
development of the animal model for TGF-\(\alpha\)-mediated clefting, several reports have demonstrated that restoration of TGF-\(\alpha\) signaling was sufficient to rescue at least part of the cleft phenotype. In particular, Cui et al. demonstrated that downstream TGF-\(\alpha\) signaling element Smad2 expression in the palatal shelf medial edge epithelia rescued much of the cleft secondary palate in TGF-\(\alpha\)3 null mice.

Yang and Kaartinen have also recently reported rescue by TGF-\(\alpha\) signaling. In this report, TGF-\(\alpha\)1 was knocked into the TGF-\(\alpha\)3 locus in TGF-\(\alpha\)3 null mice; the result was similar to the aforementioned report by Cui et al. in that a significant portion of the secondary palate was rescued.

Finally, Spivak et al. have reported rescue of the cleft palate phenotype in TGF-\(\alpha\)3 null pups in utero by viral-mediated delivery of TGF-\(\alpha\)3 during palatogenesis with substantial success, perhaps setting the stage for future in utero gene therapy for craniofacial disease processes. In summary, TGF-\(\alpha\) signaling has long been recognized as a critical mediator of successful palatogenesis, and it will be interesting to follow further research in this field toward clinical translation into alternative strategies for the management of cleft palate.

Finally, Wnt signaling has recently received considerable attention for its role in craniofacial morphogenesis, including orofacial clefting. Several reports discuss changes in Wnt family member gene expression in association with cleft palate, but only recently was loss of Wnt9b purported to be causal in the etiology of cleft palate in a mouse model. In this report, the authors confirmed that the previously described mutation clf1 in A/WySn mice was a mutation of the Wnt9b gene by a standard genetic test of allelism. The authors conclude by suggesting future examination of Wnt9b loci in humans with nonsyndromic cleft lip with or without cleft palate. Indeed, it appears that modulation of Wnt signaling holds promise for more effective management strategies in cases of orofacial clefting in the future, and it will be exciting to follow this line of research as it matures.
CRANIOSYNOSTOSIS

Advances in genetics and the advent of transgenic mice have contributed greatly to the fund of knowledge regarding specific pathways that control both normal and abnormal cranial suture fusion. Cranial sutures, which form as the bones of the skull vault approximate one another during development, serve as sites of bone growth. The patency of these bony joints during development allows the cranial vault to expand and accommodate the growing brain. Under physiologic conditions, bone deposition at the cranial sutures is regulated by molecular boundaries. Under pathologic conditions, these boundaries become obscured and premature fusion of one or more cranial sutures, or craniosynostosis, can occur. An understanding of the molecular mechanisms dictating these events carries important implications for the development of novel therapies for craniosynostosis. However, given that premature suture fusion can be thought of in its most basic terms as abnormal bone formation, the molecular lessons gleaned from an understanding of cranial suture biology are also broadly applicable toward skeletal tissue engineering applications.

Craniosynostosis has a reported incidence of approximately one in 2000 to 2500 live births world-wide. Premature fusion of cranial sutures leads to a restriction of brain growth and can result in a dysmorphic cranial vault, as well as a multitude of serious functional and morphologic consequences. Current surgical approaches to this disorder consist primarily of performing linear craniotomies to excise the synostosed suture or sutures and cranial vault remodeling procedures, such as fronto-orbital advancement, early in life. However, these procedures are physiologically challenging for young children and are often associated with refusion of the suture after surgical correction. In addition, these highly invasive operations are associated with a number of complications such as infection, bleeding, and the need for frequent blood transfusions. Thus, there is a great demand for improved strategies for treating craniosynostosis. Ultimately, the goal of tissue engineering in this context is to convert our understanding
of the molecular mechanisms controlling suture biology into minimally invasive, molecular-based therapies to correct and prevent premature cranial suture fusion.

Our laboratory has been particularly interested in noggin, a secreted antagonist of bone morphogenetic proteins (BMP). Upon evaluating levels of BMP in fusing and patent sutures of mice, Warren et al. found no difference. However, a screen of BMP antagonists revealed noggin to be expressed in patent, but not fusing, cranial sutures. This differential expression suggested that the fate of a cranial suture, that is fusion or maintenance of patency, might be controlled by the relative amounts of antagonist to agonist, rather than by the absolute amount of agonist. To test this hypothesis, Warren et al. injected a noggin-expressing adenovirus into normally fusing posterior frontal sutures in both an in vitro calvaria culture system and an in vivo mouse model. In both models, the injected sutures were found to be abnormally patent. Thus, the mis-expression of noggin had profound consequences on cranial suture fate, raising the possibility that noggin could be exploited for therapeutic purposes. Recently, Cooper et al. demonstrated the potential clinical application for noggin in their rabbit model of familial nonsyndromic craniosynostosis. After performing suturectomy on the fused sutures, the authors implanted noggin-loaded gels into the suturectomy sites. They subsequently found significantly decreased rates of suture refusion in rabbits treated with noggin, compared with sham-treated rabbits.

Interestingly, Warren et al. also found that noggin expression is suppressed by FGF-2. Based on this finding, Warren et al. proposed that syndromic, FGF receptor (FGFR)-mediated craniosynostosis might be the result of inappropriate down regulation of noggin expression. When examining the known mutations of craniosynostosis, gain-of-function mutations of the FGFR family have been shown to be the cause of approximately 20% of all known craniosynostosis disorders, including Crouzon, Apert, Pfeiffer, and Jackson-Weiss. Mutations of the FGFRs in these syndromes have been described to cause aberrant
signaling via three main mechanisms: receptor dimerization resulting in constitutive activation, increased ligand affinity, and removal of inhibition. As an example, Crouzon syndrome can be the result of constitutive activation of FGFR-2 through receptor dimerization of its free cysteines.

Given the prevalence of FGFR mutations, several investigators have attempted to manipulate this receptor in animal models to nonsurgically mitigate the premature fusion of sutures associated with FGFR mutations. In an organ culture model, Eswarakumar et al. described the application of PLX052, a small-molecule inhibitor of FGFR, to calvaria harvested from embryonic day 18.5 Crouzon-like mutant mice and wild-type mice. PLX052 is a novel inhibitor created by chlorination and a chemical substitution. With the addition of this drug, the autophosphorylation of Fgfr2c is inhibited, which in turn blocks the phosphorylation of Frs2á, ultimately preventing dimerization of the mutant receptor.

After 2 wk of exposure to PLX052 in organ culture, the authors found the premature fusion of sutures in calvaria of Crouzon-like mice to be prevented, whereas the growth of the wild-type calvaria was unaffected. Although performed in an organ culture system, these results highlight specific, small-molecule inhibitors that can be used to pinpoint signaling elements involved in the pathologic state. Such information will be critical for future development of targeted molecular therapies of craniosynostosis.

This point was further demonstrated by Perlyn et al. who used a similar pharmacological strategy, using PD173074, a selective FGFR tyrosine kinase inhibitor, as a treatment for craniosynostosis syndromes caused by constitutive FGFR activation. The authors also used a mouse whole calvaria culture system to compare skulls of mutant mice with a Crouzon-like phenotype versus skulls of wild-type mice in the presence of the inhibitor for 2 wk. Upon histologic analysis, they found that mutant calvaria exposed to PD173074 demonstrated patency of the coronal sutures with its characteristic overlapping pattern.
Parallel to efforts aimed at disrupting aberrant FGF signaling at the receptor level, efforts have also been directed at investigating strategies that block abnormal signaling at the gene transcript level. With the recent explosion of RNA interference technology, this tool has now been applied to craniosynostosis. Shukla et al. generated a small hairpin RNA targeted at the mutant form of Fgfr2 (Fgfr2<sup>S252W</sup>), which is responsible for the Apert-like craniosynostosis syndrome in mice. When transgenic mice expressing this small hairpin RNA were crossed with mutant mice carrying the constitutively activated form of FGFR2, the Apert-like phenotype in the progeny was prevented. The authors also demonstrated that in calvarial cultures from the mutant Fgfr2<sup>S252W</sup> mice, greater amounts of phosphorylated extracellular signal-related kinase (ERK)1/2, a downstream mediator of FGF signaling, were present in comparison to calvarial cultures from wild-type mice. As further confirmation that the aberrant FGF signaling in these Fgfr2<sup>S252W</sup> mice was due to ERK1/2 signaling, the authors administered U0126, a pharmacological inhibitor of ERK phosphorylation. They found that administration of the drug to the mice during embryonic and early postnatal stages significantly repressed the craniosynostosis phenotype in the FGFR2 mutant mouse model. This study elegantly demonstrates both the utility of RNA interference for probing specific signaling pathways and the potential for its eventual application in the clinical realm.

Taken together, these experiments indicate the importance of continued investigation of the complex pathways and relationships among the genes and mitogens responsible for craniosynostosis. These studies may have potential for exploitation in tissue engineering strategies to treat diseases caused by specific mutations. For instance, noggin, or specific FGFR inhibitors could be locally delivered to prevent premature suture fusion and also refusion, commonly associated after correction of synostosed sutures. Furthermore, an understanding of the molecular pathways guiding cranial suture biology can also provide insight into the mechanism that regulates bone formation.
Distraction Osteogenesis

DO is now accepted as the standard for correction of a wide range of craniofacial skeletal hypoplasias. DO can be viewed as a form of endogenous tissue engineering, whereby the discrete application of force vectors results in robust bone formation. First described by Alessandro Codivilla in 1905 and later popularized by Gavril Ilizarov, DO was initially a modality applied for the treatment of long bone deficits. During the 1970s, studies were initiated to investigate the application of DO to the craniofacial skeleton in animal models. Translation of this work to the clinical arena was finally realized in 1992 when McCarthy et al. reported the application of DO to treat a hypoplastic mandible. DO, in its most basic form, involves an osteotomy and application of distraction hardware to the bone of interest. After the osteotomy, the two bone edges are left unperturbed during the latency period, allowing an initial fracture callus to form and the regional accumulation of cytokines and growth factors to recruit and organize osteoblast and osteoclast activity. This is followed by the activation phase, when the bony segments are moved apart from one another, which usually progresses at a rate of 0.5 to 2 mm per day, until the desired degree of distraction is obtained. Complications arising from an improper rate of distraction include fibrous union and premature consolidation, depending on whether the process was too expeditious or delayed, respectively. Activation is finally followed by a period of consolidation, allowing for maturation of the skeletal regenerate.

Today, DO has revolutionized the treatment of both syndromic and nonsyndromic congenital craniofacial malformations. The technique has been successfully applied to the treatment of skeletal hypoplasias involving the mandible (hemifacial microsomia, Pierre Robbins Sequence, Treacher Collins Syndrome, Stickler Syndrome, Nager Syndrome), midface (cleft lip and palate, Crouzon Syndrome, Apert Syndrome, Pfeiffer Syndrome), upper face, orbits, and cranial vault (craniosynostosis). The severity of anatomical derangement and resultant functional sequelae of patients eligible for craniofacial
DO is highly variable. In cases of mandibular hypoplasia, or situations in which the mandible is posteriorly displaced from proper anatomic position, an increasing portion of the tongue becomes resident in the oropharynx and hypopharynx. This retroversion of oral soft tissue can result in significant airway obstruction, often mandating tracheostomy or endotracheal intubation. Additionally, these anatomical irregularities are the source of considerable feeding difficulties, leading to failure to thrive. Newborns afflicted with craniosynostosis can experience multiple physiologic and developmental derangements because of their anatomical anomalies. Elevated intracranial pressures, visual disturbances, as well as impaired cognitive maturation can result, and the aim of surgical intervention is to alleviate these symptoms, while restoring normal intracranial volume and skeletal structure. Proponents of craniofacial DO argue that it is both less invasive than traditional reconstructive procedures and has the added advantage of gradually extending the accompanying soft tissue envelope along with the underlying bone.

Research in the field of DO has most importantly served to highlight the influence of mechanical forces on osteogenesis. The observation made by Carter et al. that tensile forces drive osteogenesis, whereas compressive forces promote chondrogenesis, has provided a useful lens with which to view DO. Expounding on this concept, Loboa et al. reported that tensile strain ranging from 10 to 12.5% during distraction yielded an environment advantageous for de novo bone regeneration. Subsequently, by implementing three-dimension finite element analyses and making comparisons to histologic patterning in bony regenerates, regional tissue responses to tensile and hydrostatic forces across the regenerate were defined. Loboa et al. found that regions exposed to tensile strains of 13% or less corresponded to bone regeneration, whereas low periosteal hydrostatic pressures were associated with cartilaginous differentiation. In efforts to further dissect the influence of mechanical forces on osteogenesis at a cellular level in vitro, Gabbay et al. used a microdistraction device capable of
applying linear forces in a three-dimensional environment. MC3T3 preosteoblasts were suspended in three-dimensional collagen gels and exposed to either continuous distraction, or cyclical distraction and compression. It was observed that continuous distraction drove cellular proliferation, whereas cyclical distraction and compression encouraged the progression to a differentiated phenotype.

Necessary to successful endogenous bone tissue engineering in the setting of DO is the transduction of biomechanical forces into molecular signals that orchestrate bone regeneration. Extending our knowledge of this intricate process, Rhee et al. have identified signaling pathways that appear to function specifically in the process of translating mechanical strain into guided osteogenesis. They observed that expression of both c-Src, a kinase in the integrin mediated signaling pathway, as well as ERK1/2, a key modulator of mesenchymal stem cell differentiation, were up-regulated during distraction. Concurrently, expression of these factors was not significantly elevated in the healing of critical and noncritical sized osteotomies. Additionally, elevated levels of both of these proteins coincided with elevated levels of BMP 2/4, suggesting that signaling pathways responsible for the mechanotransduction of external forces resulting from DO may play a role in the resultant process of organized skeletal regeneration.

The importance of angiogenesis, and the forces driving circulating progenitor cells to localize to wounds and promote neovascularization, has garnered considerable interest from researchers. Sojo et al. were able to demonstrate that neovascularization precedes osteogenesis. After femoral distraction in rats, vascular endothelial growth factor (VEGF) and BMP immunohistochemical staining was performed, revealing that induction of angiogenesis occurred before bone regeneration. Fang et al. further established the importance of angiogenesis to endogenous bone regeneration by demonstrating that decreased angiogenesis led to impaired healing. Subsequent to the
administration of the angiogenic inhibitor TNP-470, fibrous nonunion in rat mandibles undergoing distraction was observed. In light of such findings, the potential to derive enhanced osteogenesis by means of augmenting angiogenesis will serve as a target for future studies.

Of note, recent work by Ceradini et al. has furthered our understanding of the role of ischemic signals in eliciting circulating progenitor cells in the setting of DO. In their studies, they outlined the process by which progenitor cells are recruited to areas of tissue ischemia through elevated expression of hypoxia-inducible factor-1 in endothelial cells. Increased hypoxia-inducible factor-1 induces expression of stromal cell-derived factor-1, potentiating the conscription of progenitor cells to hypoxic tissue. In an effort to elucidate microenvironmental cues driving neovascularization during DO, Cetrulo et al. examined whether endothelial progenitor cells responded to local ischemia produced by distraction. The authors found that by injecting fluorescently labeled endothelial progenitor cells at the start of activation, it was revealed that they are sequestered to the relatively ischemic environment of the regenerating tissue. This finding reinforces the microniche created by a region of injury and identifies specific factors that are required for successful bone formation.

A Cell-based Approach

Our discussion has heretofore focused on the molecular mechanisms involved with the pathologic processes of cleft palate and craniosynostosis or environmental factors dictating successful bone formation. Arguably, however, application of these factors toward a cell-based approach may offer the best solution. As pediatric craniofacial surgeons treating cleft palates, craniosynostosis, and craniofacial skeletal hypoplasias are commonly confronted with the challenge of replacing or reconstructing tissue deficits, cell-based therapies offer a paradigm shift as to how these tissue deficits should be addressed. Strategies that simply seek to repair or reconstruct missing tissue are no
longer adequate. Instead, the overarching goal is to regenerate the missing tissue in a patient specific manner.

Advances in our understanding of multipotent cells over the last half century have fueled a cell-based approach. At the core of such a strategy is the ability to harvest a sufficient quantity of progenitor cells, which when given the proper environmental cues are capable of regenerating the missing tissue such that it functionally and structurally mimics endogenous tissue. Because lineage-committed cells are often limited in quantity and in their potential for expansion, attention has turned to stem cells to fill this void. When considering stem cells, a pyramid of cellular pluripotency exists, with embryonic stem cells undoubtedly occupying the apex. Derived from the inner cell mass of the blastocyst, embryonic stem cells possess the ability to differentiate along endodermal, ectodermal, and mesodermal lineages. However, because of the ethical and political debate currently surrounding the use of embryonic stem cells, substantial efforts have been directed at characterizing alternative sources of stem cells, which maybe more limited in their multilineage potential. These include amniotic fluid, umbilical cord blood, bone marrow, subcutaneous (s.c.) fat, and dental pulp, among others. The recent description by two independent groups, of induced pluripotency in adult, somatic cells by turning on select genes, has added yet another exciting twist to the burgeoning stem cell field.

Missing bone is a common challenge that spans all three of the previously discussed congenital disorders, and our laboratory has taken particular interest in a cell-based approach to this problem. Although other tissue deficits, involving mucosa, cartilage, and muscle exist, the broad lessons gleaned from cell-based, skeletal tissue engineering can also be applied to regeneration of these other tissues. In cleft palate patients, the surgeon is often confronted with bony defects of the alveolus. Similarly, in syndromic forms of craniosynostosis where complex remodeling procedures are performed, sizeable calvarial defects can often result. The gold standard material for reconstructing
these defects remains autogenous bone grafts. Autogenous bone grafts, however, are accompanied by concerns for donor site morbidity and limited quantities. A host of allogeneic and synthetic materials are available as well, but they also have their inherent disadvantages, including risk of infection, immunologic issues, structural integrity, and contouring abnormalities. Finally, although DO has tremendously improved outcomes in the treatment of hypoplasias of the craniofacial skeleton, it is also not without its own morbidities including soft tissue infection, osteomyelitis, pin-tract loosening or infection, hardware failure, and patient discomfort.

In terms of skeletal tissue engineering in the craniofacial region, substantial research has been directed at investigating two cell sources, bone marrow and adipose tissue. Since Pittenger et al.’s description of the multipotent nature of bone marrow-derived mesenchymal cells, bone marrow has served as the traditional source of skeletal progenitor cells. When seeded on a variety of scaffolds in both endochondral and membranous bone defects, bone marrow mesenchymal cells have demonstrated the potential for regenerating skeletal tissue.

Since Zuk et al.’s first description of multipotent cells within s.c. adipose tissue, our laboratory and others have taken interest in the potential of adipose-derived stromal cells (ASC) for skeletal tissue engineering applications. Because of the relative accessibility, safety of harvest, and abundance of s.c. fat, ASC are an attractive candidate for cell-based therapies. We have demonstrated as proof of principal the ability of these cells to regenerate bone in critical-sized calvarial defects. Mouse-derived ASC were seeded on apatite-coated, poly(lactic-co-glycolic acid) (PLGA) scaffolds and implanted into 4 mm, parietal bone defects. By 12 wk, substantial bone formation was observed in calvarial defects treated with ASC, comparable to the amount of bone formation observed in groups treated with bone marrow stromal cells and osteoblasts. This finding has recently been generalized to human-derived ASC in a nude mouse model. Yoon et al. reported on greater calvarial
healing in nude mice where defects were implanted with human ASC-seeded PLGA scaffolds in comparison to mice treated with scaffolds alone.

The identification of postnatal sources of multipotent cells has been instrumental in the development of cell-based approaches for tissue engineering. As evidenced by the recent descriptions of induced pluripotency, this field continues to evolve. Ultimately, efforts are geared toward understanding what cues guide these multipotent cells along a given lineage. Molecular lessons gleaned from studies in cleft palate, cranial suture biology, and DO can be similarly applied to these multipotent cells.

DEVELOPMENTAL FIELD REASSIGNMENT IN UNILATERAL CLEFT LIP: RECONSTRUCTION OF THE PREMAXILLA

In the 21st century the greatest stimulus for progress in cleft surgery will come from more a more accurate model of facial development and how clefts originate. Victor Veau accurately predicted this: “All cleft surgery is merely applied embryology.” The revolution in developmental biology has not yet been incorporated into surgical practice. The drawings and terminology used in textbooks today are based on the work of Wilhelm His in the 1870’s. Cleft repairs are therefore designed based on the anatomy as it appears in the newborn. Measurements are taken and the tissues are geometrically manipulated. But the anatomy of a cleft as seen at the time of birth is far different from its original configuration in the embryonic face. From its onset at gastrulation, the clefting event unleashes pathologic processes that predictably alter the original embryonic anatomy over time to produce what we recognize at term as a cleft lip. Left uncorrected, these processes will remain operative throughout facial growth. This explains why geometric cleft repairs relapse over time, requiring revision.

Developmental field repair (DFR) is based upon the neuromeric model of craniofacial embryology. The goals of DFR surgery are: (1) resolution of all pathologic processes of clefting (deficiency,
displacement, division and distortion); (2) dissection along embryonic separation planes (subperiosteal release); (3) preservation of blood supply to the alveolar mucoperiosteum; (4) primary unification of the dental arch; and (5) reassignment of all developmental fields to their correct relationships. Before describing the surgical technique, certain basic concepts of field theory should be understood.

**Rubenstein**

Applications of neuromeric anatomy provide a potential embryonic “map” of all craniofacial structures with important implications for diagnosis and surgery. Exclusive of the cranial base (basisphenoid and posterior) and parietal bone, the craniofacial skeleton is made from exclusively from neural crest. Thus the cell populations producing the ethmoid, presphenoid, premaxilla and vomer all originate in antero-posterior order from the neural folds in genetic register with the 1st rhombomere (abbreviated r1). The inferior turbinate, palatine bone, maxilla, alisphenoid (greater wing) and zygoma arise from the neural crest of the 2nd rhombomere. The squamous temporal, mandible, malleus and incus are r3 neural crest bones.

Non-neural crest craniofacial bones come from paraxial mesoderm (PAM) lying immediately adjacent to the neural tube. PAM is divided into individual units shaped like popcorn balls called somitomeres, each one in register with its corresponding neuromere. The first seven somitomeres contain the myoblasts for all muscles of the orbit and the first three pharyngeal arches. For example, the mandible comes from r3 neural crest and all muscles originating from it arise from Sm3. Beginning with Sm8 all somitomeres undergo a further transformation into somites, each having a dermatome, myotome and sclerotome. The first four somites (derived from Sm8-Sm11) produce the cranial base posterior to the sphenoid, the muscles of the tongue and part of the sternocleidomastoid and trapezius. These are called occipital somites.
Disturbances at a particular neuromeric level can affect individual or multiple fields to be deficient or absent. Thus, *isolated cleft palate* (unassociated with cleft lip) represents a deficiency state of the vomer.

This occurs as a spectrum. As the vomer is progressively smaller it lifts away from the plane of the palatal shelves and the cleft extends forward toward the incisive foramen. In mild cases of cleft palate associated with *Pierre Robin sequence*, a reduction in the horizontal plate of the r2 palatine bone is seen. Soft palate muscles are consequently normal but divided. As the pathology worsens, reduction in the horizontal plate of the r2 maxilla creates the well-known “horse-shoe” palate cleft. *Submucous cleft palate*, on the other hand, involves pathology in the 3rd pharyngeal arch. Somitomeres 6 and 7 contain the myoblasts of levator, uvulus, palatopharyngeus and superior constrictor. These can be globally affected. Frequently, persistent VPI follows a seemingly simple palatoplasty, requiring further surgery. *Failure to stratify cleft palate by embryologic mechanism explains much of the confusion currently extent in the speech and surgical literature*.

Finally, *Treacher-Collins syndrome* provides an example in which all r2 developmental fields of the midface are affected: the maxillary, palatine and zygomatic bones are all small. The septum, vomer and premaxilla (being r1 structures) are unaffected. For this reason, the central midface projects normally while the dimensions of the palate, maxilla and zygoma are constricted.

Developmental fields form in a specific spatio-temporal sequence. Each one builds upon its predecessors. Making a face is much akin to assembling a house with magical pieces of Lego®, each one of which will grow over time. Imagine a Lego house made from 20 pieces (4 on the floor and 5 stories high). All pieces are growing independently. If a cornerstone piece is removed, the 19 remaining pieces undergo a deformation and the house tilts into the deficiency site. *The missing Lego piece in cleft lip is the premaxilla*. The physiologic basis of DFR is the reconstruction of the premaxillary field. This chapter presents the reconstructive
application of these principles to the surgery of labiomaxillary clefts.

The pathologic anatomy of cleft formation

The pathologic anatomy of unilateral and bilateral labiomaxillary clefts stems from a tissue deficiency state localized to the lower lateral piriform fossa. The tissue at fault is the mesenchyme of the ipsilateral premaxilla. Neural crest stem cells responsible for synthesis of the presphenoid and ethmoid arise from the mesencephalic neural folds and are genetically identified with the 1st rhombomere (r1). Note that the basisphenoid is not neural crest in origin; it comes from paraxial mesoderm from somitomere 1 (Sm1). Sm1 lies just outside the neural tube at level r1 and is in register with it. Immediately caudal to this population are neural crest cells immediately above the rostral rhomboencephalon with the 2nd rhombomere (r2). The most rostral zone of r2 (herein referred to as r2') is the likely source material for the vomer and PMx. The more caudal zone of r2 produces inferior turbinate (IT), palatine bone (P), maxilla (Mx), alisphenoid (AS) and zygoma (Z).

These cell populations migrate forward into the developing face in a strict temporo-spatial order. The sphenoid is laid down first, followed by the ethmoid. In like manner, formation of PMx is the prerequisite for the appearance of V. Formation of the PMx and V requires the pre-existence of the perpendicular plate of the ethmoid (PPE). The function of the PPE is to provide a cellular scaffold by which r2' neural crest cells can reach the midline. In holoprosencephaly (HPE) the PPE can be absent. PMx and V cannot develop correctly; a wide bilateral cleft results. The piriform fossa of humans and some high primates is assembled as the fusion of the frontal process of the premaxilla (PMxF) and the frontal process of the maxilla (MxF). In all other vertebrate skulls PMxF and MxF are readily visible as two distinct entities. Evolution foreshortened the human snout. The two fields became superimposed, PMxF becoming telescoped internal to MxF. This
lamination is responsible for the strength of the piriform rim. Plating of the piriform “buttress” in fracture repairs takes advantage of this bicortical anatomy.

**Piriform**

The developmental field in which the PMx resides consists of an epithelium and a mesenchyme. Formation of the PMx results from interactions between these tissues. The premaxilla has several anatomic subcomponents, these are assembled in a strict sequence. In dental terminology the central incisor is called “A” and the lateral incisor is called “B.” A erupts before B. Therefore the central incisor field (PMxA) is biologically “older” than the lateral incisor field (PMxB). Neural crest mesenchyme flows forward along the previously-established PPE. It first encounters the epithelium corresponding to PmA and then “spills over” into zone PMxB. The time sequence of dental eruption (central incisor A > lateral incisor B) is a manifestation of the relative biologic “maturity” of the mesenchymal field within which each tooth develops. The frontal process field (PMxF) is a vertical offshoot of PMxB; this subfield is the biologically “newest” tissue.

Pathology affecting the PMx occurs as a spectrum based on this original developmental pattern. A deficiency state of the PMx will first occur in the most distal aspect of the frontal process (ie. at its most cranial extent). As the mesenchymal deficit worsens, frontal process will be reduced in a cranial-caudal gradient. “Scooping out” of the piriform rim results; the nasal lining is pulled down as well. This causes depression of the alar base and a downward-lateral displacement of the lateral crus. Biologic signals from PMxF regulate epithelial stability and therefore affect lip formation (vide infra). When the signal strength is minimally disturbed the lip is normal despite the piriform distortion. Therefore the *forme fruste* manifestation of premaxillary deficiency is a cleft lip nose with a perfectly normal lip.

Once the frontal process is eliminated, the deficiency state shows up in the lateral incisor field. Progressive degrees of PMx
deficiency in the lateral incisor field cause incremental loss of alveolar bone. Normal alveolar development follows a gradient. It begins at the incisive foramen and progresses forward. Mild deficiency causes notching on the labial surface. As the deficiency worsens the notch deepens backward toward the incisive foramen. A critical lack of alveolar bone mass results in outright failure of lateral incisor development.

BMP-4 signals from this field are directly implicated in the mechanism of fusion between the lateral lip element and the prolabium. BMP-4 emanating from PmxL forms a cranio-caudal chemical gradient. The strength of this gradient depends upon the total amount of available BMP-4; this in turn is proportional to the overall mesenchymal mass of PMxB. Reduction in mass of the lateral incisor field results in a diminution of the total BMP-4 signal. Lip fusion follows this same gradient. Mild weakness of the BMP-4 gradient will result in notching of the vermilion. As the situation worsens the extent of the lip cleft worsens in a cranial direction. The clinical spectrum of the so-called minimal cleft lip deformity faithfully reproduces this biologic sequence.

In summation, variations in clefts involving the primary palate and the lip can be understood as interactions between deep plane fields of the premaxilla, the maxilla and superficial plane field of the lateral lip element. The mesenchyme of the lip has a different embryologic origin. It is genetically identified with the 2nd rhombomere. Neural crest cells from r2 provide the dermis and the subcutaneous tissue of the alar base, while paraxial mesoderm from the 2nd somitomere forms the anterior half of the squamous temporal bone and the cranial half of the parotid gland. All derivatives from level r2 can be mapped out within the sensory distribution of V2, the nucleus of which resides within r2.

The developmental anatomy of the nose, prolabium and premaxilla has been previously described in terms of neuromeric theory by this author. In contrast to the rest of the body, all dermis and submucosa of the head originates from neural crest cells, not from a dermatome associated with a somite. Pre-dermal neural crest
arises from three distinct zones of the embryo. The dermis of the forehead, nose and vestibular lining come from the caudal prosencephalon (above prosomeres p4-p1). This prosencephalic neural crest (PNC) migrates forward like a gigantic glacier to occupy the neural folds above prosomeres p6 and p5. The alar half of p6 and p5 creates the telencephalon (cerebrum). The basal halves of p6 and p5 plus all remaining prosomeres synthesize the diencephalon (epithalamus, thalamus and hypothalamus). The neural folds above p6 and p5 are “sterile.” They contain the pituitary, olfactory and optic placodes, but no neural crest. When PNC flows forward into this zone the placodes are activated and dermis is formed. Nasal vestibular lining from the cribriform plate forward to the internal nasal valve comes from p6 PNC. All remaining frontonasal dermis from the internal nasal valve forward to the hairline comes from p5 PNC.

PNC skin shares sensory innervation with the dura of the underlying frontal lobe. V1, the sensory nerve for this common zone has its nucleus with the 1st rhombomere (r1). The neuroanatomy is analogous for the rest of the face. Rhombomeres r2 and r3 contain neurons supplying all skin and dura innervated by V2 and V3. This is the embryologic basis for the treatment of migraine headaches with Botox® injected into peripheral trigger points.

Design of surgical incisions for cleft repair follows this embryology. The boundary between these two fields is sharply demarcated within the naris. The skin of the anterior columella and philtrum is thus a p5 derivative. This skin is supplied by terminal branches of the internal carotid artery, the anterior ethmoid arteries and innervated by V1. The skin making up the floor of the nose has a different origin. It extends from the base of the columella laterally and makes contact with the alar base. The medial (terminal) branch of the sphenopalatine artery innervates this skin. The innervation is from V2 and is shared with the ipsilateral incisors. Continuity between the p5 skin of the lateral columella and the r2' skin of the nasal floor makes possible the elevation of a skin-
cartilage flap containing the medial crura with long skin flaps. Many years ago Vissarianov described this flap as a means of secondary cleft reconstruction.

Based on signals from the underlying premaxilla, the r2 alar base produces a tongue of tissue that makes contact with the r1 lateral prolabium. This skin bridge sets up the floor of the nose. It also provides a mechanical platform by which mesenchyme from the lateral lip element can make contact with the p5 mesenchyme of the prolabium. Lip closure thus occurs. This process involves mesenchymal “flow” from the lateral lip element toward the prolabium. For this to occur, the epithelium covering the lateral lip element, the r2-r1 skin bridge and prolabium must undergo a genetically-induced breakdown. BMP-4 produced by the premaxillary field causes de-repression of Sonic hedgehog (SHH), a gene localized to these overlying epithelia. The protein product of SHH causes epithelial breakdown. Thus, absence or deficiency of an appropriate BMP-4 signal will lead to restricted expression of SHH, abnormal persistence of epithelium and failure of mesenchymal fusion.

The volume of the PMx determines whether or not lip closure can occur. First, a small premaxillary field manufactures small amounts of BMP-4. The amount of BMP-4 produced is critical to produce the epithelial breakdown necessary to permit mesenchymal merger. Second, when the premaxilla is too small, the physical distance between it and maxilla will exceed a critical dimension. Epithelial bridge formation between the alar base and the prolabium cannot occur. If this critical distance exists at the level of the incisive foramen, a cleft of the secondary palate will form. This is because the horizontal repositioning of the palatal shelf from the maxilla must make contact with the vomer just posterior to the incisive foramen. The process is just like a zipper. If initial contact is not made, fusion of the palatal shelf to the vomer cannot take place. Even if initial contact is made, a secondary palatal cleft can still result due to displacement of the vomer away from the midline. The vomer can become warped by the inequality
of growth forces on either side of the cleft. Thus, the zipper may get started anteriorly but, as the process proceeds posteriorly, when it encounters the deviated vomer, a palatal cleft will open up.

Reconstruction of the premaxilla

Cleft surgery that does not reconstruct the missing PMx does not solve the biologic problem. The pediatric face is a set of Lego® pieces, all growing over time. If one piece of the set is missing, with subsequent growth the remaining pieces will collapse into the deficiency site. Only when all the Lego pieces are in place can harmonious facial development occur.

The premaxillary developmental field consists of lining (present in its totality) and mesenchyme (missing). Lining can be recreated by: (1) subperiosteal dissection of the primary palate (r2’ premaxilla and r2 maxilla); (2) subperiochondrial dissection of the p6 septum (sufficient to reduce the septum into the midline; (3) subperiosteal elevation of r2’ vomer to close the nasal floor (sufficient to reduce the septum into the midline); and (4) subperiosteal rescue of the r2’ nasal floor skin from the lateral prolabium. When these flaps are elevated and sutured, the primary palate becomes a box lined with neural crest stem cells, all of which carry membrane-bound BMP receptors.

Mesenchymal replacement can be undertaken using two basic mechanisms. Autogenous bone graft from rib or hip provides mesenchymal cells originating from paraxial mesoderm. Incorporation of the graft into the primary palate occurs by osteoconduction. Implantation of recombinant human bone morphogenetic protein-2 (rhBMP-2) in combination with an activated collagen sponge (ACS) results in morphogen-based recruitment of stem cells from the environment into the sponge. Stem cell concentration and differentiation into osteoblasts results in the formation of bone native to the site. This process is known as osteoinduction. Extensive pre-clinical work by Boyne demonstrated the ability of rhBMP-2 to form membranous bone,
including reconstitution of surgically-created cleft palate defects in primates. Studies demonstrating efficacy in maxillary lift combined with absence of donor site morbidity resulted in FDA approval for this indication.

The technique of facial bone reconstruction using rhBMP-2/ACS implantation is known as in situ osteogenesis (ISO) and has been previously reported by this author and co-workers. Resynthesis of a 12 cm mandibular defect in a 9 year old demonstrated the ability of ISO function effectively outside the range of blood supply associated with a critical-size defect. BMP-2 mediated osteoinduction is accompanied by extensive recruitment of blood vessels from local environment. For this reason selected bone defects can be resynthesized using ISO alone, without recourse to microsurgical tissue transplantation.

Distraction techniques have been successfully applied to ISO-regenerated bone. In a number #7 lateral cleft with a foreshortened mandibular body and absent ramus, distraction of the recipient periosteal chamber in a posterior and superior direction permitted synthesis of 3.5 cm of mandibular ramus with eventual articulation with the skull base via a pseudoarthrosis. Distraction-assisted in situ osteogenesis (DISO) will be an alternative treatment to rib grafts in the reconstruction of the Pruzansky III mandibular defects in craniofacial microsomia. The bone produced will be membranous. The dissection is less problematic. There is avoidance of unpredictable growth of the rib graft outside the natural periosteal environment. Finally, the chest wall donor site is obviated. Histology of ISO-produced membranous bone is indistinguishable from that of the recipient site in both mandible and maxilla.

Alveolar clefts are lined with mucoperiosteum containing neural crest stem cells. Blood supply is excellent and the dimensions modest. For this reason, this author and co-workers reported 50 alveolar clefts successfully treated with rhBMP-2. Precise surgical technique of implant placement and of soft tissue closure was emphasized in the current series of 200 cleft sites. Long-term outcome of 43 cleft sites was assessed at one year post grafting.
Mechanisms of Cleft Palate: Developmental Field Analysis

with 3-D CT. The series was comprised of 23 unilateral clefts (6 primary and 17 secondary) and 12 bilateral clefts (3 primary and 9 secondary). Complete transverse fill (unification of the dental arch) was achieved in all cases. Vertical fill was improved to 75-100% when an inert bulking agent (tricalcium phosphate) was included. For these reasons, primary cleft repair using developmental field reassignment technique can ideally be combined with ISO to achieve primary unification of the dental arch without donor site morbidity.

Philosophy of Developmental Field Reassignment (DFR): the 4 D’s

Faulty embryogenesis of the premaxillary field affects the position and shape of surrounding fields in four distinct ways: Division of force vectors causes unequal growth, Deficiency-related collapse of partner fields into the void, Displacement of partner fields away from the midline. Distortion over time of structures such as the septum. DFR repair addresses these “4 D’s” in reverse order. All distortions should be corrected. The displaced soft tissues if the face should be centralized into the midline. The deficiency site should be restored, using neighboring soft tissues and bone graft or ISO; and (4) division should be closed via unification of soft tissues. The execution of DFR surgery is based upon the concepts of Sotereanos. Its guiding principles are five. (1) Correct all pathologic states in the first operation (as above). (2) Respect embryonic developmental planes during dissection, ie. subperiosteal release of the soft tissue envelope for a tension-free closure. (3) Conserve blood supply to the periosteum, safeguarding it for future membranous bone synthesis. Fourth, align and unite the dental arch into a normal relationship. Fifth, reassign all developmental fields into correct relationships with each other. When properly executed, these principles result in the restoration of the functional matrix. When all the bone-forming soft tissue fields are spatially correct, all force vectors exerted upon bone will be correctly aligned. Subsequent growth of the face is directed back toward the normal.
A new algorithm: Is lateral nasal wall deficiency relative or absolute?

In keeping with Victor Veau, cleft repair is a constant exploration of nature’s experiments. Developmental field reassignment surgery is deliberately designed to address a tissue deficit of the lateral nasal wall and alveolus, the product of a congenitally small premaxillary field. That the lateral crus be entrapped cannot be in doubt. Its release into a normal position occasions a triangular tissue gap that must be filled. Proper airway reconstruction is the name of the game. At the same time, the alveolar cleft (a six-sided box) must be reconstructed. This requires flap coverage for its nasal surface (the “top” of the box). Can these two goals be accomplished with the same tissue?

A skin graft alone (particularly anterior auricular skin and cartilage) will effectively support lateral alar crus advancement. It cannot provide vascular coverage for the alveolar cleft “roof.” At first blush, the LLC-NPP flap would seem big enough to accomplish both goals. After 7 years of work with this technique this author has concluded that paring from the prolabium is not sufficient. Premaxillary field soft tissues are not only relatively deficient from the lateral nasal wall: an absolute tissue deficit exists. Use of the premaxillary tissue mismatched to the prolabium is not always sufficient to solve the problem. The new algorithm of DFR surgery now calls for optional composite grafting of the lateral wall, followed by elevation and transposition of the LCC-NPP flap across the nasal surface of the alveolar cleft. The decision to graft is based upon the relative size of the defect versus that of the flap.

Surgical Technique of DFR

This operation consists of markings, a five-step dissection sequence and a five-step closure.

Preparation and marking

The DFR operation is comprehensive, providing simultaneous
correction of the lip, nose and primary palate. A more extensive dissection is required. The operation takes longer to perform (about 3-4 hours for a unilateral cleft). For this reason, patients come to surgery at 4-6 months of age. Prior to operation the dental arch is prepared with splinting (a form of infant presurgical orthopedics); this is begun as early as two weeks after birth. The emphasis of presurgical splinting is: (1) promoting anterior growth of the retro-displaced cleft maxilla; and (2) maintenance of the space in the alveolar cleft. If satisfactory maxillary shelf repositioning does not result by age 3 months, a lip adhesion procedure is carried out. When satisfactory dental arch correction is achieved, DFR repair is performed. This usually occurs 3-4 months after the lip adhesion.

The patient receives antibiotics and corticosteroids for swelling. A central V2 block is performed at the pterygopalatine fossa using 0.25% Marcaine® (bupivicaine hydrochloride). Approximately 3-5 cc per side is sufficient (maximum dose for 0.25% bupivicaine being 1 cc/kg). At the end of the case, the central block is reinforced by blocking the infraorbital nerves with bupivicaine, 1-2 cc per side. The child returns to recovery pain-free. The initial block ensures that substance P (a critical mediator in the pain cascade released in response to surgical trauma) will not be produced. In the absence of substance P the entire postop pain response is altered.

The surgical fields of the unilateral cleft are defined as follows. (In neuromeric terms: A = p6 (red) + p5 (turquoise), B = r2' (pale gold), C is r2 (yellow) and D is r2 + r4 (yellow + orange). The prolabium is divided into two zones. Zone A is the true philtrum; it measures the width of the columella. It contains paired anterior ethmoid arteries (the terminal branches of the internal carotid system) and paired terminal branches of V1. Thus zone A contains two neurovascular developmental fields. Zone B of the prolabium is all tissue lateral to the philtrum: the non-philtral probium (NPP). The nasopalatine (medial sphenopalatine) artery, the terminal branch from the internal maxillary system, supplies the NPP. Bilateral clefts have two NPP zones. The cleft-side alar base is zone
C. The lateral lip element below the alar groove is zone D. It contains a sphincter layer, the deep orbicularis oris (DOO) and the dilator layer, the superficial orbicularis oris (SOO) muscles. DOO develops in conjunction with the oral mucosa while SOO develops with the skin. A layer of fat conveniently separates these two layers.

The true destination of NPP soft tissue is in the nasal floor and the lateral nasal wall. NPP represents the soft tissue envelope corresponding to the lateral zone of the premaxilla, PMxB + PMxF. Nasal floor soft tissue corresponds to the lateral incisor field, PMxB. Soft tissue of the lateral nasal wall between the inferior turbinate and the lateral crus corresponds to the frontal process field, PMxF. Lateral wall soft tissue corresponds to the frontal process field, PMxF. Nasal floor soft tissue corresponds to the lateral incisor field, PMxB. The rationale of DFR surgery is to reassign the misplaced fields of the premaxilla into correct position. When the NPP flap is combined with the reflected mucoperiosteum from the margins of the bony cleft a surgical “pocket” is created. This site is subsequently filled with bone graft or a bone-producing cytokine (rhBMP-2) such that missing premaxillary fields are re-synthesized.

The NPP flap functions just like a “boxtop” to cover the alveolar cleft. It is almost always adequate. I am convinced that the “secret identity” of NPP is the original PMxB. Because of the cleft this tissue is “shipwrecked” on the premaxilla. Unfortunately, NPP is frequently insufficient to replace PMxF. Every time the lateral crus is released and advanced a soft tissue defect appears. The defect represents missing PMxF. In my clinical experience this is almost always the size of the auricular cymba. A composite graft of anterior auricular skin and cartilage from the cymba is the most reliable replacement for this tissue. The cymba graft, in combination with the NPP flap, provides more than enough tissue to reconstruct the missing premaxilla.

Markings are carried out using a modification of the Millard system. (Paired anterior ethmoid neurovascular bundles define
the true philtrum; this equals the width of the columella.). First to be marked is point 2, the junction of the normal philtral column with the white roll. The width of the columella at its base is then measured. This distance (usually 6-10 mm) is subsequently marked along the white roll medial to the philtral column as point 3. Distance 2-3 is the width of the philtrum; this contains the two anterior ethmoid arteries (approximately 4-6 mm apart). The midpoint of the philtrum (point 1 in the Millard system or nadir of cpid’s bow) is irrelevant. Point 13, defined by the bulge of the footplate of the medial crus on the non-cleft side, marks the “shoulder” of the columella. The corresponding point 12 on the cleft side will be found displaced caudally and internally with respect to point 13. Total philtral height 2-13 should equal 3-12. Points 4 and 10 mark the midpoint of the alar bases on non-cleft and cleft sides, respectively.

On the lateral side, points 9 and 11 mark the terminus of skin within the nasal cavity. This is located just anterior to the inferior turbinate. Point 9 is the tip of flap D while point 11 is the tip of the future nostril sill C’, so-called because it is in continuity with the alar base C. The dimensions of C’ can be roughly mapped out by measuring the nostril sill on the non-cleft side. This is the distance from the midpoint of the non-cleft alar base (point 4) to the ipsilateral footplate point 13. Point 8 marks the natural transition of the white roll. Distance 8-9 should match 3-12.

**Dissection sequence**

**Step 1. Lateral dissection: rescuing the nostril sill**

The lateral lip is tensed with a single hook and the skin-mucosa margin is incised proceeding upward from 8 to 9, located just below and anterior to the inferior turbinate. From here the incision swings around laterally to 10, the midpoint of the alar base, (but not beyond it at this point). In this manner, the lateral lip flap D is separated from the alar base C. This step separates orbicularis from the nasalis. From point 9, a second, more internal, incision defines the triangular flap C’. The base width of C’ can
be deduced by measuring the dimensions on the non-cleft side. From the lip a lateral vermilion flap (L) is pared off and dissected down to the alveolar cleft margin. Proper paring of L includes a small strip of lip skin 2-3 mm wide because it is “rolled-in,” with an abnormal relationship to the underlying muscle. If the surgeon does not do this, the skin will pucker inward at the final closure. The L flap is the most optional tissue of the entire DFR operation. It is best brought medially and interposed between the nasal vestibular lining and the B flap. It can also be used as a free graft to the lateral nasal wall. No tissue is sacrificed in DFR.

With its vermilion stripped off, the lateral lip margin is now entered and the orbicularis is split into its deep (constrictor) and superficial (dilator) components. The deep orbicularis oris (DOO) is shaped like the letter J and separated from the superficial orbicularis oris (SOO) by a layer of fat. The caudal margin of the two muscles defines the white roll and contains the labial artery.

**Step 2. Medial dissection: the NPP-lateral columellar advancement flap**

The medial margin of the cleft (zone B in our diagram) consists of prolabial tissue lateral to the true philtrum. This is in continuity with lateral columellar skin and the medial crus of the alar cartilage. Conveniently, the blood supply of these two units is a watershed permitting dissection of a very long flap. The B flap has two parts: (1) a skin/mucosa flap of the non-philtral philtrum (NPP) and (2) a chondrocutaneous flap containing the lateral columellar skin and medial crus (LCC). \[ B = NPP + LCC \]

B has a rich blood supply. The NPP component is irrigated by the nasopalatine artery. The LCC component gets supply from 2-3 lateral branches of the ipsilateral anterior ethmoid. In addition, vessels in continuity with the lateral nasal system from the facial artery run along the surface of the alar cartilage itself. Once elevated, B is surprisingly long, reaching all the way across the cleft to the lateral nasal rim. is inset into the gap created by advancement of the p5 lateral vestibular lining (and the lateral crus). This achieves
two important surgical goals: (1) cephalic advancement of the medial crus with increased tip projection; and (2) soft tissue augmentation of the lateral nasal wall. Inset of B permits release of the tethered lateral vestibular lining. Repositioning the lateral crus now occurs in what would otherwise be a Y-V pattern without the Y-V closure. Natural alar cartilage anatomy is accomplished without pinching together an already-deficient lining.

The future cleft-sided philtral column is determined by understanding the embryology of the prolabium. The true philtrum lies between points 2 and 3 and consists of two fields (each supplied by a separate branch of the anterior ethmoid artery). The philtral dermis comes from p5 prosencephalic neural crest and is innervated by V1. The width of the philtrum is roughly equal to that of the columella, generally measuring 6-10 mm. Because the philtrum develops on top of the r2' premaxilla it receives additional blood supply from the terminal branches of the sphenopalatine artery (SPA) emerging at the level of the septopremaxillary junction. Thus, the philtrum has a dual blood supply. All remaining prolabial skin and mucosa (the NPP) is an r2' derivative, supplied by the nasopalatine artery and innervated by V2. The prolabium of a bilateral cleft thus consists of four developmental units based on embryology, blood supply and innervation: two central philtral A fields and two lateral B fields.

The embryology of the columella is as follows. The anterior centrally-located skin comes from p5 and contains the paired anterior ethmoid arteries. On either side of that central swatch, the columellar skin extends backwards toward the septum. Just beneath the lateral columellar pillars lie the medial crura of the lower lateral cartilages (LLC). Because these pillars serve as the biologic "template" for the cartilage to form, dissection of skin away from the crura is extremely difficult. The LLCs are thus p5 neural crest derivatives. The upper lateral cartilages lie above the p6 vestibular lining and are therefore of p6 neural crest origin.

Correction of the cleft lip nose requires releasing the alar cartilage from two points of entrapment. The deficiency state of
the premaxilla causes a mechanical deformation of perfectly normal p5 skin on both sides of the nasal introitus. In the lateral nasal wall, the r2' skin overlying the premaxillary frontal process (PMxF) is reduced or absent. Consequently, p5 skin containing the lateral crus of the alar cartilage is dragged down into this “sinkhole” located just in front of the inferior turbinate. The lateral crus is therefore flattened. On the medial side, the deficiency of the r2' premaxillary lateral incisor field (PmxB) creates a “sinkhole” into which the p5 lateral columellar skin is displaced. This displaces the medial crus of the alar cartilage downward and inward compared with the non-cleft side.

The LCC-NPP flap is elevated as follows. Under tension, the NPP skin is elevated in continuity with the lateral columellar wall. The lateral (internal) incision is made first, separating the skin from the mucosa all the way to the junction between the premaxilla and vomer. Although this incision will eventually be carried up in front of the septum, it is advisable to stop here. Tension can thus be maintained on the NPP while it is separated from the philtrum proper.

Next, a second incision, parallel to the first, ascends from point 3 to the base of the columella. It then extends into the nose along the lateral sidewall of the columella. Previously I would continue this incision along the side of the columella (just anterior to the edge of the medial crus) and thence bring it directly into the nose as an infracartilagenous incision. This design made me concerned about possible scar contracture. At the suggestion of Dr. John Reinisch, I began to break up the incision by elevating a medially-based rectangular skin flap occupying the caudal half of the columella. Just beneath the Reinisch flap lies the footplate of the lateral nasal cartilage. At its cephalic margin, the incision is resumed along lateral columella. At the level of the intermediate crus the incision transitions beneath the soft triangle into a standard infracartilagenous incision and is then continued all the way to the piriform margin. The anterior incision of the NPP-LCC flap is not sufficient to advance it. A second parallel incision is required.
Mechanisms of Cleft Palate: Developmental Field Analysis

It starts in the membranous septum about halfway up. It is then carried downward to the vomer and then re-directed forward to meet the lateral margin of the B flap. Note that the membranous septal incision provides immediate exposure of the cartilaginous septum.

The NPP-LCC flap now resembles a long boot, shaped like Italy. The “toe” of the boot extends along the prolabial margin. Beneath the “heel” of the boot lies the footplate of the alar cartilage. This landmark corresponds to point 12. Grasping the heel of B provides instant access to the medial aspect of the medial crus. This is a safe plane permitting dissection of the alar cartilage right into the nasal tip, with the following caveat. The B flap gets blood supply from the nasopalatine artery via 2-3 branches emerging at the junction of the premaxilla and vomer. Gentle spreading along the medial border of the cartilage will reveal these branches and preserve them. Additional blood supply descends along the skin. Formerly I would elevate these NPP-LCC flaps completely, never having an issue with ischemia. Now however, I think it prudent to preserve the nasopalatine branches when possible. This type of blunt dissection is more than sufficient to advance the nasal tip.

The septum is now dissected out and freed from the maxillary crest until it sits passively in the midline. As growth proceeds the centralized septum will no longer constitute an abnormal force vector tethering the nasal tip.

Step 3. Nasal dissection: “open-closed” rhinoplasty

At this juncture, a standard infracartilagenous incision is made. This incision is brought all the way to the piriform rim, following the natural fold between the nasal skin and the vestibular skin. The caudal extent of this incision terminates at the internal border of the triangular nostril sill flap B’. The exposure gained via the lateral columella-infracartilaginous incision allows a complete dissection of the dorsal nasal skin envelope as described by McComb. The success of the McComb dissection is really an embryonic field separation. The deep layer comprises the p6
vestibular epithelium and p6 neural crest upper lateral cartilages, the blood supply to which comes from below via the ICA. The superficial layer is p5 nasal skin and the lower lateral nasal cartilages, the blood supply to which is also of ICA derivation. Interposed between these two layers, like a sandwich, is the SMAS layer of facial muscles derived from the 2nd pharyngeal arch (r4-r5). The myoblasts come from somitomere 4. The nasal muscles are compartmentalized by r4 neural crest fascia (the SMAS). The blood supply to this intermediate 2nd arch layer is from the facial artery (ECA). This mesenchyme provides an additional source of blood supply to the alar cartilages. Careful dissection of the lateral columellar walls from the columella discloses vessels running along the medial surface of the medial crura. These anastomose the lateral nasal vessels with the nasopalatine vessels.

Along the lateral nasal rim the dissection is carried right down to the piriform margin. The proper plane for separation is achieved by hugging the surface of the cartilage. The overlying r4 SMAS layer and the p5 skin layer are left in anatomic continuity. Very little bleeding is occasioned by this approach. As one reaches the piriform rim the periosteum is incised and stripped vertically. Subperiosteal elevation of the soft tissues lateral to the piriform rim preserves the facial artery arcade. The nasal skin envelope is then liberated cephalically all the way to the nasal bones.

Despite these maneuvers, the vestibular lining is still tight! The media crus has been completely released and advanced into the nasal tip but the lateral crus being remains splayed out and tethered. Releasing the lateral crus from the vestibular lining has been advocated in the past. This is technically difficult because it violates the embryology (recall that the alar cartilage arises as a neural crest response to a pattern embedded in the vestibular epithilium). The size and shape of the cleft-side alar cartilage has been demonstrated to be normal compared with the non-cleft side. Consequently, the p5 vestibular lining “program” must be normal as well. Studies at UNC demonstrate that the overall surface area of the repaired cleft nostril is reduced by 30%. The
site and dimensions of the deficit correspond to the soft tissues of PMxF.

The soft tissues of the alveolar cleft PMxB can be found on the prolabium. The LCC-NPP flap effectively provides a vascularized “roof” for the bony cleft. Lateral nasal wall release “uncovers” a defect that frequently exceed the reach of LCC-NPP. The release begins at the inferior turbinate and continues along the junction of vestibular and nasal skin. At the apex of the lateral crus it becomes V-shaped. Due to the prior McComb dissection the lateral crus is advanced cephalically into symmetry with the normal side. The internal nasal valves will appear equal. Left behind is a raw spot roughly the size of the ear cymba. A composite graft using anterior skin and cartilage from the cymba provides a sturdy reconstruction for the airway. The LCC-NPP flap, inset without tension across the alveolus, comes to rest at the foot of the cymba graft.

Exit the turbinate flap

As the DFR evolved, I employed various solutions to patch the defect created by the vestibular release. One of these solutions made use of an anteriorly-based inferior turbinate flap as described by Noordhoff. Although this tissue worked adequately, I had several reservations about it: (1) the dissection is subtle and difficult to teach; (2) the tissue type is distinct and not native to the nasal rim; and (3) healing of the donor site can be accompanied by crusting and bleeding; and (4) it did not make embryologic sense. Proper dissection and inset of flap NPP combined with use of a composite for FTSG made the turbinate flap unnecessary. The key to getting the most out of NPP is to include the prolabial vermilion with the skin.

Step 4. Intraoral dissection: sliding sulcus S flap

The rationale and design of the sliding sulcus mucoperiosteal flap stem directly from pioneering work at the University of Pittsburgh by Sotereanos. This technique involves a gingival release
on the cleft side carried out from the alveolar cleft to the buttress. This permits wide subperiosteal dissection over the entire face of the maxilla below the infraorbital foramen as described by Delaire. A 45 degree backcut up the buttress is performed. The attached gingival is released. The flap is covered on its undersurface by a sheet of periosteum, rendering it rather stiff. The Sotereanos maneuver is a mobilization of the S flap using a counter-incision in the periosteal sheet itself parallel to the gum line. The counter-incision, located half-way from the gingival margin to the infraorbital foramen, is made by just scoring through periosteum. It extends from the piriform margin straight lateral to the buttress. At this point it joins up with the previous backcut. These two incisions make a right angle: the periosteal counter-incision is transverse across the maxilla and the buttress incision is vertical. The combination of these two incisions releases the lower mucoperiosteal flap S from the upper mucoperiosteum attached to the orbit.

When S is released, it advances mesially about two tooth units. The margin of S that was freed from the lateral border of the cleft now extends across the alveolar cleft without tension and is sutured to the mucoperiosteum of the premaxilla. In this way, like a sliding door, the S flap seals up the anterior aspect of the alveolar cleft.

On the noncleft side, a similar subperiosteal dissection is done without recourse to a gingival release. A large bipedicle flap is created, permitting centralization of the previously lateralized soft tissue envelope. Note that when cleft lip occurs without an alveolar cleft, bilateral S flaps, elevated without gingival release, permit proper tension-free centralization of midface soft tissues.

The S flap then comes in two varieties, each of which has a specific benefit for the patient. The simple S flap is elevated in the subperiosteal plane across the entire face of the maxilla between two points of vertical release: the alveolar cleft and lateral wall of the buttress. It remains attached to the teeth. A gingival releasing incision is not required. By virtue of its ability to centralize the
soft tissues of the entire midface, the simple S flap fulfills two important functions: (1) the creation of an esthetic soft tissue “drape”; and (2) the biologically active osteogeneic potential of the mucoperiosteal stem cells is transferred forward, thus enabling bone deposition to take place in centrically for better facial skeletal projection. The drawback of the simple S flap is that it cannot produce a mucoperiosteal flap to cover over the anterior aspect of the alveolar cleft. In such cases, a rectangular mucoperiosteal flap can be harvested immediately lateral to the alveolar cleft and brought over for closure. This design works for narrow clefts.

The compound S flap (as described by Sotereanos) implies a full gingival releasing incision all the way from the alveolar cleft back to the tuberosity. This involves additional operating time. Gingival release from an alveolus containing erupted primary or secondary dentition is technically simple. The bone is solid and the surgical plane easy to follow. Infants are a different matter. Prior to the 4th month of life maxillary and alveolar bone is too soft to work with. DFR should be performed between ages 4-6 months. Dissection is facilitated using loupe magnification, a #15C or Beaver blade, an amalgam packer and Molt (#9) or Louisville periosteal elevators. A compound S flap can be mobilized two full tooth widths. The main advantage of the compound S flap is the dramatic increase in mucoperiosteal flap length it provides. This is generally the width of two dental units. The anterior aspect of the alveolar cleft is thus covered with stem cell-containing mucoperiosteum. Disadvantages of S flap relate to the temporary anatomic disruption of pericoronal attachments.

The S flap has distinct indications for use in primary and secondary cleft repair. These are of vital importance, particularly when one must operate under conditions that are less than ideal. In a primary cleft, whenever possible, pre-surgical orthodontic control of the arch should be accomplished. This may also involve a lip adhesion. The entire effort is directed toward normalizing arch dimensions. In these cases, compound S flaps are not required. Closing the anterior wall of the alveolar cleft can be accomplished
with a rectangular superiorly-based mucoperiosteal flap elevated directly off the lateral incisor. The space will fill in nicely. If one desires, the flap donor site can be grafted with Alloderm®.

Limited resources and difficult logistics in economically developing countries force surgeons treating cleft patients to adopt different and creative strategies, particularly when the alveolar cleft is wide. These children often have no access to pre-surgical orthopedics.

They may never receive orthodontics. They do possess a ready source of bone graft from the rib. In such cases, achievement of a consolidated arch and elimination of oronasal fistula are reasonable goals for primary cleft surgery. The two key factors for success in these patients are: (1) ability to make a simple acrylic splint and secure it—2 mm screws work perfectly well; and (2) meticulous dissection of the alveolar cleft with attention to its vascular anatomy.

If a dental splint can be fabricated, lip adhesion can narrow the cleft down to dimensions permitting simultaneous DFR repair and alveolar grafting with a simple S flap. When this is not possible, a compound S flap will close the gap. In some very wide primary unilateral clefts I have used bilateral compound S flaps to successfully close gaps of 14-16 mm. Of course, this maneuver disconnects the frenulum from the midline. Postoperative remodeling of the alveolus reestablishes symmetry. In such cases, arch stability justifies the extensive intraoral dissection.

Secondary cleft reconstruction follows the same rules but with the proviso that gingival release incisions are fast and easy to accomplish. The floor of the alveolar reconstruction has already been provided by a pre-existent palatal repair. When orthodontic capability exists, such clefts should first be expanded to the original width prior to grafting. But when the patient has no access to ortho, S flap alveolar reconstruction can be combined with DFR to produce a permanent, aesthetically pleasing and physiologic result in one stage.
Step 5. Dissection of the primary palate

Beginning with the lateral nasal wall, the mucoperiosteum is elevated off the maxillary alveolar bone. The palatal margin is undermined as well. This results in a large mucoperiosteal envelope. This must be separated into nasal and oral components. This is done at the level of the “shoulder” of the alveolus. The incision can be extended backward a few mm along the edge of the palatal shelf. On the medial side, the septal mucoperichondrium and the vomerine mucoperiosteum are elevated in continuity. The surgeon then proceeds forward to elevate the mucoperiosteum off the premaxilla. Separation of the envelope occurs at the “shoulder” of the premaxilla. A vomer mucoperiosteal flap is elevated and sutured to the nasal mucosa of the cleft maxilla.

A six-sided “box” is created. The medial and lateral walls are raw bone. The floor is oral mucoperiosteum. The anterior wall is the sliding sulcus mucoperiosteum. The roof is the nasal mucoperiosteum + the NPP flap. The posterior wall is the vomer flap. These flaps are loaded with undifferentiated mesenchymal stem cells (MSC). The cambium layer of the periosteum is especially rich in stem cells. All neural crest MSCs contain membrane-bound receptors for BMP. This pocket is now ready to be filled with rib graft (iliac crest graft in older children) or with an rhBMP-2/ACS implant.

Alveolar reconstruction, both primary and secondary, demands precise knowledge regarding the vascular anatomy of mucoperiosteum bordering the alveolar cleft. This is a neglected area of cleft surgery. The blood supply to the premaxilla is the most complex (and potentially treacherous). It is derived from three sources. (1) Premaxilla is an r2’ derivative. Like all mesenchymal structures caudal to r1, premaxilla is supplied from the external carotid. The vascular axis of the vomer and premaxilla is the medial sphenopalatine (nasopalatine) artery. PMx mucoperiosteum supplied by the SPA envelops the bone from above-downward, like a cloak thrown over a chair. (2) From below, PMx mucoperiosteum is continuous with that of the r2 maxillary
palatal shelf (MxP), the arterial axis of which is the external carotid-based greater palatine artery. (3) a second anastomosis exists within the mucoperiosteum covering the anterior superior aspect of the premaxilla. This zone is in contact with the soft tissues of the prolabium, a p5 derivative supplied by the internal carotid-based anterior ethmoid arteries.

Premaxillary survival depends upon maintenance of sufficient contact between the bone and its enveloping mucoperiosteum. Alveolar cleft reconstruction cannot be accomplished without stripping away the vascular coverage of PMx, to a greater or lesser degree. Thus, premaxillary vascular anatomy dictates the direction in which the mucoperiosteum must be elevated. Surgical strategy for this maneuver is a direct consequence of the cleft type.

In unilateral palate clefts, the medial alveolar cleft margin is a continuous zone of external carotid from the SPA to the GPA. This permits downward dissection of the mucoperiosteum from the shoulder of the premaxilla or upward dissection from the oral margin. The premaxilla remains alive, based on its other sources. In bilateral palate clefts, no such palatal anastomosis exists. The mucoperiosteum can only be reflected upward. If this is done on both sides and if the prolabium is simultaneously elevated, premaxillary necrosis can ensue.

This principle is exactly the same as in prolabial death reported by Millard, a combination of bilateral rotation backcuts (destroying the anterior ethmoid supply to the philtrum) and elevation away from the underlying premaxilla. In bilateral cleft lip and palate, the anatomy of both prolabium and premaxilla consists of paired angiosomes. One can disrupt one or the other with impunity, but not both.

Fortunately, the robust blood supply of the maxilla comes to our rescue. Injection studies demonstrate the maxilla to be supplied by the pharyngeal branch of the facial artery and facial branch of the ascending pharyngeal artery. It is independent of its mucoperiosteal cover. Meticulous dissection of the lateral alveolar cleft creates a continuous mucoperiosteal sheet uniting the lateral
nasal wall with that of the hard palate. This contains a mirror-image external carotid anastomosis, this time between the medial sphenopalatine and greater palatine arteries. This can be stripped downward with impunity. It is a long flap, extending from just in front of the inferior turbinate all the way to the hard palate. The lateral alveolar mucoperiosteal flap is of sufficient size to close the entire floor of the alveolar cleft.

What then should be done for children BCL(P)? Must one sacrifice arch reconstruction to facial aesthetics or vice versa? The answer (no!) lies in a staged approach with careful dissection. Primary lip reconstruction must not violate the tissue plane separating the prolabium and mucoperiosteum. Even in a bilateral dissection, 50% of the premaxilla will remain perfused by both SPA and AEA vessels.

Lateral nasal wall reconstruction with the cymba composite graft is appropriate. Liberation of the medial crura must be done without entering the territory of the medial sphenopalatine artery. If this appears difficult, nasal elevation and lip revision may have to be staged later. Alveolar cleft closure should be done by elevating the premaxillary mucoperiosteal flap upward and the maxillary mucoperiosteal flap downward.

Wide bilateral clefts require alternative strategies. Two principles to avoid trouble in these patients are: (1) meticulous primary closure of the nasal floor; and (2) delayed grafting of the alveolar cleft. A wide alveolar “roof” defect requires big nasal floor flaps. These are best harvested at the primary surgery, before other tissues get in the way. The key point here is to not cut the lateral mucoperiosteal flap until the medial flap is harvested from the premaxilla. Once the dimensions of medial flap are known, a superiorly-based lateral flap of sufficient size is elevated to complete the anterior nasal closure. At this point we are still left with a wide alveolar “floor” defect and not enough tissue to close it. What does one do? One waits it out, using a palatal acrylic splint (secured with pins or 2 mm screws) is maintain the transverse arch dimensions and prevent collapse.
6 months later, at palatoplasty, the long mucoperiosteal flaps elevated from the hard palate provided ideal coverage for the alveolar cleft “floor.” Bilateral compound S flaps are raised. These will adequately seal off the anterior face of the alveolar cleft. The graft (using rhBMP-2 or rib) is placed. Once again, care must be taken to prevent transverse collapse until the bone graft has hardened. Intraoral splinting is thus continued for another 2-3 months.

Secondary grafting in bilateral cases follows the same principles. The vascular anatomy may have been altered (primary surgery may have elevated the prolabium away from the premaxilla). In such cases some degree of revascularization to the premaxilla occurs over time. Nonetheless, dissection of the premaxillary mucoperiosteum must be sparing. All that is required is elevation of sufficient tissue to close the soft tissue defect and place the graft. Nasal tip elevation can be safely accomplished using DFR technique and cymba grafts. The key point is to preserve soft tissue continuity between the vomer, the septal mucoperichondrium and the premaxilla: this is the site of entry of blood supply into the premaxilla.

**Closure sequence**

**Step 1. Elevation of the nasal tip**

The lateral crus is stabilized into symmetry with the normal side using a transcutaneous 4-0 chromic mattress stitch. The V-shaped limbs of the releasing incision are closed. The donor site is reconstructed using a full-thickness retroauricular graft or a composite graft of anterior ear skin and cartilage. By trial and error I prefer the cymba as the donor site.

The tip is positioned anatomically using the Cronin nasal retractor (Padgett Instruments). The medial crura are battened together with 5-0 PDS. Suture suspension and modification of the alar cartilage can be readily executed by means of the open-closed approach as per the surgeon’s preference. The author approach
is predicated on the establishment of normal field relationships. Wide dissection of the nasal soft tissues combined with release from their “piriform prison” allows all fields to be passively held in position by a nasal stent inserted at the conclusion of the procedure. The author finds the Koken-type silicon stents (Porex Corp, Newman, Georgia) easy to use. The stent is placed at the end of the surgery. Closure of the nostril incision starts at the intermediate crus working medially down to the Reinisch flap. One then proceeds laterally to the margin of the lateral nasal wall. This involves placing 2-3 sutures over 5 mm.

The remainder of the lateral nasal incision will be filled by inset of the B flap. Reconstruction of the frontal process of the premaxilla adds the missing tissue to the lateral nasal wall. This frequently involves placement of a composite for full thickness graft into the defect. The medial crural complex the then elevated with respect to the septum with 4-0 vicryl. This also closes the membranous septum counter-incision.

Step 2. Soft tissue reconstruction of the premaxilla

The roof of the missing premaxillary field is reconstructed based on meticulous closure of the nasal floor. This is carried out using a mouth gag, starting anteriorly at the incisive foramen. Because the space is tight using 5-0 Vicryl on a small P-2 needle is helpful. The medial vomerine and lateral nasal mucoperiosteal flaps are closed all the way posterior to the end of the vomer flap. This provides correct orientation for inset of the B flap. The posterior margin of B is sutured from medial to lateral along the newly-united nostril floor. The tip of B is eventually inset into the donor site of the lateral crural advancement flap. Next, the alar nasal skin flap C is sutured to the anterior margin of B. The surface area of the lateral nasal wall is now restored.

The floor of the missing premaxillary field is reconstructed when the medial and lateral mucoperiosteal flaps harvested from the walls of the alveolar cleft are turned down into the mouth using 4-0 vicryl. The cleft side sliding sulcus flap S is advanced
and secured using 4-0 Vicryl PS-2 around the dental units. Three or four such sutures will suffice. Optional suspension of the sulcus to maintain height can be done by passing a 3-0 vicryl up to the nasal floor and then back down into the sulcus as a mattress suture.

**Step 3. Osseous reconstruction of the premaxilla: In situ osteogenesis (ISO)**

The primary palate is reconstructed with Infuse bone graft (Memphis, Tennessee Sofamor-Danek). The implant is prepared by soaking a Helistat® activated collagen sponge (ACS) (Plainsfield, N.J. Integra Life Sciences) of preselected size with reconstituted rhBMP-2 applied uniformly over the ACS.

The minimum time required for binding is 15 minutes; however, I usually wait 30 minutes. The Infuse® comes in three kit sizes 4.2 cc, 5.6 cc and 8.4 cc. These are used as follows: primary unilateral (small), primary bilateral (medium), secondary unilateral (medium) and secondary bilateral (large).

The volume of the implant should match that of the defect. Mastergraft® tricalcium phosphate is useful for “bulking up” the implantation site. It acts passively, a space-occupying agent. One could use freeze-dried bone as well (the ideal bulking agent does not yet exist.)

The Mastergraft is wrapped with the ACS collagen sponge like a fajita or sushi roll and placed into the defect. I like to place a vertical component into the alveolar walls and a horizontal component below the nasal floor. The pocket created by DFR dissection can be filled with iliac crest graft as well.

**Step 4. Closure of the lip: Delaire concepts modified**

Many years ago pioneering work by Delaire demonstrated that wide subperiosteal dissection yield significant aesthetic benefits of tissue draping and, at the same time, did not impair maxillary growth. Indeed, when compared with supraperiosteal release, this approach is developmentally correct because it separates the
osteogenic functional matrix (the periosteum and overlying soft tissues) from the product (the premaxilla and maxilla).

The suture sequence is as follows:

_Delaire 1_: Centralization of the bilaterally displaced soft tissues is accomplished with 4-0 nylon sutures placed from below the flap to the paranasalis (levator) insertion into the superficial orbicularis oris (SOO). This insertion does not involve the alar base. It is generally located one fingerbreadth lateral to the alar base. The non-cleft side is sutured first to below the anterior nasal spine. This suture serves as a reference point for a similar suture from the cleft side.

_Suspension of the DOO_: This suture sets the depth of the sulcus on the cleft side. At the most cephalic margin the DOO is suspended from the septum with a 4-0 PDS.

_Delaire 2_: The purpose of this suture is to control the height and curvature of the cleft side nostril. This is accomplished by connection of the nasalis to the anterior nasal spine. However this step is _optional_ at this point. If performed, care should be taken to not tighten this and inadvertently narrow the nostril. When the remaining sutures are placed a decision can be made if a Delaire 2 is warranted. I usually decide upon a Delaire 2 at the end of the case to accentuate curvature.

_Orbicularis closure_: Three or 4 sutures of 4-0 PDS are required for the DOO layer. The SOO layer is closed with 5-0 PDS making sure the loop is at the level of the dermal-epidermal junction.

_Delaire 3_: The oblique head of SOO sets the aesthetic drape of the lip. The 5-0 PDS is obliquely passed upward from the cephalic edge through the base of the columella and then back down to the SOO as a mattress suture.

**Step 5 Final adjustments: finessing the nostril floor**

After closure of the lip, the alar base C is at time compressed medially by the movement of the neighboring D flap (lateral lip element). If so, the alar base must be translocated laterally. This
can be accomplished by excision of a crescent of skin from the lateral lip element. C is then elevated and secured to the lateral via a buried 5-0 PDS suture.

At times it is necessary to elevate the ala completely in order obtain sufficiently lateralization. The tip of the nostril sill flap C’ is now sutured just posterior to the columellar shoulder. Continuity between the columellar shoulder and the nostril sill is now reestablished. Perialar suturing with inverted 5-0 PDS sutures restores the alar crease very nicely.
Unilateral Cleft Lip and Nose Repair

The face is composed of soft tissue over an underlying skeletal framework. This construct allows for complex facial animations and expressions. The nose, positioned at the center of the face, is the most prominent facial feature.

*Typical appearance of cleft nasal deformities.*

The face of a child with cleft lip and nose often draws attention that negatively impacts the child’s psychosocial development. In current practice, plastic surgeons are better able to reconstruct the faces of children with cleft lip and nose to near-normal anatomic
form and physiologic function. With early surgical correction, children born with cleft lip and nose have the chance to develop positive social interactions and better self-esteem early in life, along with good speech and attractive smiles.

While multiple techniques are available for the repair of cleft nasal deformity, the universal surgical principles are to restore the normal nasal anatomy and function affected by clefting and to anticipate the secondary deformities that appear with subsequent growth and development.

The optimal timing for the reconstruction of the cleft lip nasal deformity was once unresolved. Multiple long-term studies published by experienced cleft surgeons now show that early nasal reconstruction has no adverse effects on growth. The current standard of care is to reconstruct the nose at the time of primary cleft lip repair.

Although plastic surgeons continue to improve outcomes in the management of cleft lip and nose defects, secondary corrections and revisions are still common.

The families of children with cleft lip and nose should be counseled that while primary nasal repair sets up a “more normal” 

*Cleft nasal deformity can be seen even in patients with incomplete cleft lip.*
vector for growth, their child may benefit from a touch-up revision before the school years and, ultimately, a formal rhinoplasty at the conclusion of orthodontic/orthognathic care.

The ultimate goals of surgical care for children with cleft lip and nose are an aesthetic facial appearance, a beautiful smile, acceptable speech, and normal psychosocial development.

**SURGICAL THERAPY**

**Surgical approach to the nasal defect**

“A well mended harelip would pass unnoticed at a cocktail party were it not for the nose.” Despite multiple technical procedures described, no one protocol has proven to be completely satisfactory in the repair of all cleft lip nasal deformities. Still, controversy remains as to the optimum corrective approach, the best techniques for exposure and repair, and, most significantly, the timing of the correction.

Some surgeons believe that early nasal surgery (1) interferes with growth, resulting in nasal hypoplasia; (2) introduces scars, making secondary correction difficult; (3) damages infantile cartilage; and (4) makes repair technically harder because of the small size of the nose and immature cartilage.

Reconstructive surgeons historically have been reluctant to perform rhinoplasty on a growing nose; however, the use of prudent operative techniques has created growing acceptance to correcting nasal deformities prior to puberty.

With less traumatic techniques, a loss of integrity of the cartilaginous nasal framework does not usually result in growth inhibition in the region of the mid face when the septum is not subjected to aggressive resection.

While primary repositioning and manipulation of the nasal septum and changing its abnormal position in infancy have a positive effect on nasal development, it may have a negative effect on maxillary growth.
PRIMARY RHINOPLASTY AT THE TIME OF UNILATERAL CLEFT LIP REPAIR

The nose in a unilateral cleft lip child has some tell-tale deformities that have been well documented by Huffman and Lierle. However, for a long time most cleft surgeons were reluctant to attempt a correction of the nasal deformity during primary cleft lip surgery. This was due to a notion that early nasal repair would cause a detrimental effect on the growth of the nose and the maxilla.

Vilray Papin Blair and James Barrett Brown (1930) were among the first to attempt primary nasal correction with unilateral cleft lip repair. They undermined the skin, thus, separating it from the cartilage and used mattress sutures tied over a bolster on the skin.

Gillies and Millard in 1953 also performed alar cartilage dissection and repositioned the septal cartilage after freeing it from its deviated position in the vomerine groove, straightened it, and sutured it to the lip muscle on the cleft side.

Berkeley (1959) followed an aggressive primary correction of the cleft lip nose. He stressed the importance of straightening the septum. He resected the nasal spine and performed a rotation of the nose on the cleft side. He used a mid-line columellar incision for his primary repair.

The Cleft Lip Nasal Deformity

These have been well documented by Huffman and Lierle. We shall list some of the significant components:

- The columella on the cleft side is short.
- The anterior nasal spine is displaced to the noncleft side.
- The anterior part of the nasal septal cartilage is also deviated to the noncleft side.
- The cleft side nostril is wider.
- The cleft side ala is buckled inward.
- The cleft side alar dome is retroplaced.
Unilateral Cleft Lip and Nose Repair

- The angle between the cleft side medial and lateral crura is obtuse.

Figure: Child with the classical unilateral cleft lip nasal deformity

Evolution of the Concept of Repair of the Cleft Lip Nose

Early cleft surgeons faced the dilemma on whether to repair the cleft lip nose primarily. Surgeons who shied away from primary correction feared that they would cause harm to the growth of the nose and the maxilla. There have been some animal studies to substantiate this claim. However, this fear has been repeatedly been countered by numerous studies showing that primary repair causes no such deleterious effects.

As Millard notes, Blair, Brown and McDowell, and their disciples have undermined thousands of alar cartilages without stunting nasal growth, and hence that aspect need not be a deterrent. Many early surgeons used external incisions. The results were generally unsatisfactory. Interest in primary rhinoplasty was rekindled by McComb and Coghlan. When his procedure was first presented on 1975, some surgeons predicted drastic interference with subsequent development of the nose. On review,
there did not appear to be any interference with growth. To a large extent, the children were spared the embarrassment of a deformed nose through their childhood years.

McComb felt that more harm was probably done by failing to correct the nasal deformity at the time of lip repair. The alar cartilage became locked and tethered in its displaced position by scar and the transverse shortage of nostril lining and growth of the nasal tip was altered. Secondary correction of the cleft lip nose was always more difficult to achieve.

This has been supported by others - Byrd et al., Haddock et al., Cussons et al. as well. Cutting believed that primary nasal repair is durable and decreases the extent of secondary surgery in adolescents.

Anthropometric assessment has also shown no interference with nasal growth when the lip and nose were repaired simultaneously. This study also showed better symmetry of the nostril and nasal dome projection and better correction of the alar flaring and overall balanced growth and development of the alar complex with primary nasal repair.

This has been our belief too. The senior author has performed an aggressive correction of the cleft lip nose from the late 1960s on thousands of unilateral cleft lip children, and we have not found any detrimental effect on the growth of the nose. In fact, the overall shape and symmetry of the nose is better and the extent of secondary deformity is much less when compared to the patients who come to our center after having had primary surgery without any nasal intervention at some other centers.

Despite the plethora of evidence in favor of primary nasal correction, there continues to be a minority of cleft surgeons who believe that primary repair does not diminish the need for further operations and also that the magnitude of the secondary deformity is also not decreased.

The advent of the modality of nasoalveolar molding (NAM) has made the cleft lip nasal deformity less severe during primary
repair. However, NAM without any nasal surgical correction does not produce any lasting results.

**Open/Closed/Semi-Open Rhinoplasty**

An open approach has been advocated by some authors. However, many others, including us, use a closed approach. A semi open method is also in vogue. We prefer the closed technique because we believe that we obtain results comparable with the open techniques.

**The Charles Pinto Centre Protocol for Primary Correction of the Cleft Lip Nasal Deformity**

At our center, the senior surgeon has been performing an aggressive closed rhinoplasty with septal repositioning in all complete unilateral cleft lips at the time of the primary repair. All such lips are repaired using the Millard rotation-advancement procedure.

*Figure: Closed alar dissection on the cleft side*

The ala is approached from both the medial and the lateral
aspects using a pair of curved Kilner Scissors. The scissors are introduced medially through the incision at the base of the columella and laterally through the perialar incision. Dissection is carried out in the plane between the dorsal skin and the lower and the upper lateral cartilages on the cleft side, so that these cartilages are completely devoid of any skin attachments from the alar rim up to the nasal bones. A more limited dissection is also carried out on the noncleft side up to the dome. We do not attempt to separate the lower lateral cartilage from the vestibular lining as the cartilage is firmly adherent to the lining and we believe that it is almost impossible to separate the two in a closed dissection. The freed cleft side lower lateral cartilage is hitched to the noncleft side upper lateral cartilage using a bolster suture. An interdomal suture is also introduced through the noncleft side nostril to narrow the tip.

We use the Millard cinch suture to correct the alar flare. This suture (5-0 polypropylene) is taken from the noncleft side nostril through the membranous part of the septum and then through the paranasal muscles at the base of the ala and then again through the membranous septum to the noncleft side and tied there as a mattress suture. The perialar incision helps in identifying the paranasal muscles well, and aids in obtaining a good bite on them in order to correct the alar flare better. We have observed that when this perialar incision is avoided by some surgeons, the postoperative results do show some degree of alar flare in most patients.

Primary alar cartilage dissection has gained wide acceptance now. However, when it comes to the repositioning of the deviated nasal septal cartilage primarily, there is still widespread reluctance on the part of cleft surgeons.

As already mentioned, Gillies and Millard (1953) described the separation of the deflected septum from the vomerine groove, the straightening of the septum, and its suture to the cleft side lip muscles. Berkeley (1959) also stressed the straightening of the septum and resection of the anterior nasal spine.
However, the septal correction has still not gained wide acceptance. The Senior Surgeon at our center and his mentor Prof. Charles Pinto have aggressively addressed the problem of the deviated nasal septum from the late 1960s. This was not accepted by cleft surgeons for a long time.

The nasal septum in unilateral cleft lip patients is always deviated to the noncleft side anteriorly, and the anterior nasal spine is similarly displaced to the noncleft side. We approach the septum by incising the mucoperichondrium on the cleft side over the groove at the base of the septum.

The mucoperichondrium is carefully stripped off the underlying septal cartilage. The septospinal ligament is then identified anteriorly and divided to expose the anterior border of the septal cartilage. The cartilage is separated at its anterior border from the mucoperichondrium on the noncleft side. This is essential to avoid inadvertent shearing of the cartilage during the next step, which is the separation of the mucoperichondrium on the noncleft side from the cartilage.

Separating the septal cartilage from the mucoperichondrium on both sides does no harm. The septal cartilage is freed from the perpendicular plate of the ethmoid and the vomer. The cartilage thus freed of its inferior and posterior attachments, still tends to buckle when repositioned toward the midline. To avoid this, a sliver of cartilage is excised off the inferior border.

Previously, we also used to excise a wedge of cartilage from the anterior border. However, we have discontinued this now as we have realized the importance of the anterior most part of the septum for support of the tip of the nose in these patients. The cartilage still has a bow-stringing effect, and this is overcome by making scoring incisions on the cartilage on the noncleft side to make it flail.

The final step in the cartilage repositioning is the suturing of the cartilage to the newly constructed nasal floor on the cleft side. Ideally, the cartilage should be anchored to bone or periosteum
in the mid line. However, in the cleft child, there is no such structure in the mid line. The anterior nasal spine is itself displaced to the noncleft side, as already mentioned. Hence, we compromise and overcorrect by suturing it to the soft tissue on the cleft side. We believe that there will be a drift the cartilage medially with time. We can acknowledge that in most of our patients we do not need to perform any further correction of the septal position subsequently. We advocate this aggressive septal repositioning as we believe that it has not caused any adverse effects on nasal or maxillary growth. While we have not yet been able objectively to substantiate this belief due to logistical reasons, there are enough studies in literature to confirm our philosophy.

Anderl et al. and his group have published their results after 35 years of septal repositioning and shown that there is no late damage and that secondary correction is necessary only in 20% of their patients. They also stress the functional improvement in breathing in their patients. They separated the septal cartilage from mucoperichondrium on both sides in their patients.

Smahel et al. used X-rays to study the effects of primary septal cartilage separation from the maxilla and the nasal cartilages 15 years after primary correction. They concluded that after primary repositioning of the nasal septum patients had a more favorable nasal prominence and better vertical growth of the face than patients who did not have a primary septal correction. The maxillary retrusion in the two groups was similar.

Mulliken and Martínez-Pérez have added septal work in his primary unilateral cleft lip repairs since 1995. His group has studied these patients using posteroanterior cephalograms retrospectively and found less sepal deviation and smaller contralateral turbinates in these patients. They agree with us that there is no growth disturbance to the nose from primary sepolasty.

Other studies have established using cephalograms that there is a long-term overall beneficial effect of primary septal correction on nasal symmetry and tip projection with no negative growth effects.
Columella

As mentioned earlier, the cleft side hemicolumella is always shorter in unilateral cleft lip patients. As advocated by Millard, we use the C flap to lengthen the hemi columella in all our unilateral cleft lip patients. We have observed that most of our patients have symmetrical columella as a result of this. We use the excised sliver of septal cartilage as a columellar strut graft. When many of these patients undergo rhinoplasty subsequently, we do observe the presence and actual growth of this cartilage graft in many of them.

Other Deformities

There are some other associated deformities observed in the cleft lip noses.

High Riding Nostril

When there is a wide alveolar anteroposterior disparity between the medial and lateral elements, we note that the nostril base comes to lie at a more superior level than its counterpart on the noncleft side. We have used the unequal Z-plasty described by Jackson to prevent this deformity. Ever since we commenced the use of this refinement, the incidence of such high riding nostrils has diminished. We believe that this deformity can be completely eliminated only when the medial and lateral maxillary segments are at the same anteroposterior plane and this may be possible using NAM.

Vestibular Web

This is a nagging problem during cleft lip repair. Patel and Mulliken believes that this has bony, cartilaginous, and soft tissue components. He uses intercartilaginous sutures under vision in his semi open approach and releases the attachments of the lateral crus from its piriform ligamentous attachments. In addition, he uses a lenticular excision of the web as he believes that there is an excess. This is in contrast to the belief of others that there is actually less of vestibular lining. We too believe that the vestibular
lining is to be preserved and are not sure if there is actual excess. Moreover, there is a need for all the available soft tissue lining during future rhinoplasties to avoid alar rim notching. The mentor of the Senior Surgeon Prof. Charles Pinto devised a Z-plasty that helps to address the web. One has to be very meticulous during the dissection of the lining to avoid damage to the underlying cartilage.

However, of late we do not perform this Z-plasty often as most of these children undergo preschool rhinoplasty and the web tissue is then used to build up a notch-free alar rim.

With such an extensive closed primary alar cartilage shift and septal repositioning, we are able to obtain consistent long-term results. The classical cleft lip nose stigmata of the grossly deviated septum, the grooved, slumped ala with a wide flare is almost never seen in our patients. However, most of them almost 80% have a slight droop in the soft triangle. While this is not gross by any standards, it is still noticeable enough to invite ridicule from peers at school. Hence, we perform an open rhinoplasty at 5½-6 years in these children using a sutural technique to provide a symmetrical and stable cartilaginous framework. This has given good long-term results.

We conclude that the components of the unilateral cleft lip nasal deformity have been well documented and that these can and should be addressed during the primary lip repair itself. Such intervention does not cause any growth disturbance in the long-term. There should no longer be a shadow of doubt about this. Similarly, repositioning of the nasal septum should also be taken up more enthusiastically by all cleft surgeons as this too has been shown to only produce better noses and it causes no harm even after 40 years of follow-up. The actual modality of intervention must of course be left to the virtuosity of the individual surgeon. We have described our protocol in detail.

Conflicts of interest

There are no conflicts of interest.
CLEFT LIP RHINOPLASTY

The secondary cleft lip nasal deformity presents an extreme challenge to the facial plastic surgeon.

The deformity is complex and involves all tissue layers, including skeletal platform, inner lining, osseocartilaginous structure, and overlying skin. It is often the characteristic cleft nasal deformity that is noticeable to the observer after a well performed cleft lip repair.

Secondary repair of the cleft lip nasal defect requires an understanding of the pathological nasal anatomy associated with congenital clefting. The basic cleft nasal deformity is characteristic and dependent upon the original extent of clefting of the lip. However, the secondary nasal defect varies greatly and is a result of: 1) the original malformation, 2) any interim surgery performed, and 3) growth of the nose and face.

The cleft surgeon must therefore have a treatment philosophy and technique flexible enough to reconstruct a variable range of associated nasal problems. This chapter describes the pathological anatomy associated with cleft deformities, and describes approaches and techniques designed to improve form and function of the cleft nose.

Anatomy of the Unilateral Cleft Nasal Deformity

The etiology of the primary unilateral cleft nasal deformity is a lack of skeletal support of the cleft alar base. The unilateral cleft malformation also includes a hypoplastic and malaligned orbicularis oris muscle on the involved side.

The combination of the lack of cleft side skeletal support and abnormally oriented muscle results in a characteristic caudal deviation of the nasal septum to the noncleft side.

Lack of muscular continuity often is associated with an abnormal configuration of the cleft nasal sill. The unilateral cleft side nasal base is lateral, posterior, and inferior to its noncleft counterpart alar base.
Inadequate skeletal support to the cleft alar base causes asymmetry and displacement of the cleft lower lateral cartilage (LLC). The medial crus of the cleft LLC is shorter than the noncleft LLC, while the lateral crus of the cleft LLC is longer than its noncleft counterpart. The cleft side dome is wide and the tip is relatively underprojected on the affected side. Although underprojection of the nasal tip accompanies the unilateral cleft lip, the amount of the underprojection is less than it is in bilateral clefts. There is always asymmetry of the nasal tip and the alar base; the extent of the asymmetry is related to the extent of the original deformity.
Anatomy of the Bilateral Cleft Nasal Deformity

The nasal deformity associated with bilateral cleft lips is usually more symmetric than is the unilateral nasal deformity. Nasal asymmetry in bilateral clefts is only present if significant differences exist in the severity of the lip clefting. The bilateral nasal deformity usually includes a deficiency in skin and soft tissue between the vermillion-cutaneous junction of the lip and the nasal tip. This results in a short columella and underprojection of the nasal tip. The extent of the underprojection of the tip is related to the amount of projection of the premaxilla and the underlying skeletal deformity.

The nasal tip in the bilateral deformity is broad and flat. Both medial crura of the LLC are shorter than normal, while the lateral crura in bilateral clefts are longer than normal. The nasal septum in bilateral clefts is usually in the midline, but the septum is often wide and reduplicated. The alar bases are poorly supported due to skeletal deficiency, and this causes lateral and posterior malposition of both the cleft alar bases.

Primary Management of the Cleft Nasal Deformity

Traditional management of the cleft lip deformity often does not focus attention on the nose. However, it is clear that early management of the nasal deformity minimizes nasal asymmetries and allows the nose to grow in a symmetric fashion. Primary treatment of the cleft nasal deformity includes pre-surgical nasoalveolar molding (PNAM) and primary rhinoplasty (at the time of lip repair).

PNAM is a continual low-level pressure on the cleft alveolar segments. This non-surgical technique is initiated before one month of age and exerts a measurable orthopaedic effect. Non-surgical repositioning of the bony cleft segments helps to: 1) narrow the cleft gap, 2) improve alar base symmetry in asymmetric clefts, 3) expand the soft tissue envelope, and 4) elongate the columella.

Although PNAM is not used in all clefts, the technique is very helpful in wide clefts with significant asymmetry. In bilateral
clefts with a “locked-out” premaxilla, PNAM can facilitate retropositioning of the premaxilla, allowing a single stage definitive cleft repair.

Successful use of PNAM requires a team approach with a dedicated orthodontist, and a compliant and understanding family.

Figure: The effects of 6 weeks of PNAM (from left to right).

Primary Cleft Nasal Repair

Various philosophies and techniques of primary cleft rhinoplasty have been described.

Approaches usually include some form of caudal septal repositioning and nasal tip reorientation.

Most cleft surgeons do not advocate removal of cartilage in early childhood, but rather use repositioning of cartilage with suture techniques. These maneuvers can usually be accomplished with the standard incisions used to close the lip cleft.

After the lip incisions are made and the primary lip dissection is completed, the muscle and soft tissues of the alar base are separated from their maxillary attachments.

The malpositioned alar base is freed by creating an internal alotomy at the anterior head of the inferior turbinate.

If adequate soft tissue dissection of the alar base is performed, the cleft alar base can be repositioned (during closure) in the optimal 3-dimensional position.
Alotomy being performed to reposition the alar base.

The cleft side LLC is then dissected from its cutaneous attachments by creating a medial and a lateral tunnel. These subcutaneous tunnels are connected and allow the cleft LLC to be repositioned into a more symmetric fashion. Care is taken to not violate the vestibular skin, avoiding the complication of secondary adhesions and nostril stenosis.

The cleft side medial crura is dissected from its cutaneous attachments.

The cleft side lateral crura is dissected from its cutaneous attachments.
Primary cleft rhinoplasty begins with closure of the nasal floor and sill. This closure is first started with reapproximation of the musculature of the nasal base. The skin of the nasal sill is then closed with attention paid to making the cleft nasal sill look like the noncleft side. Closure of the nasal sill is performed with #5-0 chromic catgut suture. It is important to not narrow the sill too much. A nasal base which is too wide is easy to narrow secondarily, while a too narrow sill is difficult to widen. The other component of primary cleft rhinoplasty is to reposition the cleft nasal tip into a more projected, symmetric position. Many surgeons have been reluctant to perform rhinoplasty at the time of lip repair to avoid theoretic growth inhibition of the nose. However, McComb and Coughlan have observed no nasal or facial growth inhibition in patients followed for 18 years after undergoing primary cleft rhinoplasty. The initial alar base and tip symmetry obtained in their patients was obtained during the entire study period.

After the nasal sill is reestablished and the lip is repaired in a layered fashion, the cleft LLC is repositioned to create a narrowed, projected location. This is achieved with internal mattress, or tie-over external bolsters. A new dome occurs, with a lengthened medial crus and a shortened lateral crus. If nasal bolsters are used, they are removed in 7 to 10 days. The resulting nasal tip is more symmetric, defined, and projected.

Figure: Pre and post primary cleft rhinoplasty.

Secondary Cleft Septorhinoplasty

The timing of definitive septorhinoplasty in cleft patients should take into consideration both psychological aspects and
physical growth maturation of the nose and the overall facial skeleton. In cleft patients with significant malocclusion and dentofacial deformity (usually class III secondary to maxillary hypoplasia), definitive nasal reconstruction is usually delayed until after the orthognathic surgery is completed.

This often postpones the surgery until 17-18 years of age in females and until 19 years in males.

Figure: Patient with unilateral cleft lip and maxillary hypoplasia. LeFort I osteotomy with distraction osteogenesis being performed prior to secondary cleft rhinoplasty.

Figure: Before and after LeFort I osteotomy with distraction osteogenesis and subsequent secondary cleft rhinoplasty with autologous costal cartilage.

The goals of definitive nasal surgery are to maximize nasal function and appearance. This requires total septal reconstruction, and often necessitates significant cartilage grafting for support and camouflage of deformities.
Incisions and Approaches

Both endonasal and external approaches can be used for cleft septorhinoplasty. The open, or external approach is usually preferred as this approach allows superior visualization for diagnosing deformities, and better exposure for suturing structural grafts. In addition, the open approach provides excellent access to the caudal septum, which is usually significantly deviated and poorly supported.

If the skin and columellar soft tissues are adequate, a standard columellar incision (either a stairstep or an inverted notched-V)
is utilized. However, if inadequate skin and soft tissue exists, recruitment of columellar skin is required. In the unilateral cleft deformity, an asymmetric V-to-Y is used. In the bilateral deformity, a midline V-to-Y or forked flaps can create additional columellar soft tissue necessary to increase projection.

**Septal Reconstruction**

The nasal septum associated with cleft deformities is often severely deviated, with the caudal septum deflected to the noncleft side, and the posterior septum bowing into the cleft side. In order to reconstruct the cleft septum, complete mobilization of the caudal septum must be performed. Access graft to its caudal border. This can be performed in an overlapping fashion to the cleft septum is usually obtained through an open approach, by separating the intracrural ligament between the feet of the medial crura of the LLCs. After detaching the caudal septum from its maxillary attachments, the septum is usually strengthened by affixing a cartilage, or the cartilage graft can be sutured end-to-end.

*Figure: Correction of the deviated septum in the unilateral cleft lip patient.*

The posterior septum is then straightened with standard incisional and excisional techniques. Once the septum is
straightened and adequately supported, the anterior portion is sutured to the anterior nasal spine with long-acting resorbable monofilament suture. The well-supported septal construct is then used as a platform upon which to anchor the nasal tip complex.

**Cleft Rhinoplasty**

After the nasal septum is stabilized and repositioned, the outer nasal structure can be reconstructed. This usually requires structural grafting to support the underprojected and inadequately supported cleft side infrastructure.

The cartilage grafts are obtained from the posterior septum, the conchal bowl of the ear, and the rib. In most instances, sufficient graft material can be harvested from the septum. In secondary cases, however, costal cartilage is often required for adequate supporting cartilage.

**Upper Nasal Third**

The upper one-third of the nose is usually not affected by the cleft malformation. Standard nasal osteotomies, lateral +/- medial, are performed to narrow the nose, close an open roof deformity, or to straighten a deviated nasal pyramid. It is important to expect a weak middle third on the cleft side. This will, of course, affect the method and type of osteotomy performed.

**Middle Nasal Third**

The middle nasal third in patients with clefts is usually weak as a result of inadequate skeletal support on the cleft side. Reconstruction of the cleft side middle third can be performed with a spreader graft (placed between the upper lateral cartilage (ULC) and the nasal septum, an onlay cartilage graft, or flaring sutures. In some instances, more than one technique can be used (e.g. flaring sutures and spreader grafts). Spreader grafts are frequently used bilaterally, and can be placed in an asymmetric manner. In bilateral cleft nasal deformities, bilateral spreader grafts are often used.
Figure: Placement of asymmetric spreader grafts.

It is important to anticipate functional consequences from bony or cartilaginous hump reduction in cleft patients. The underlying weakness in skeletal support (secondary to the alveolar bony cleft) of the cleft side alar base results in a deficient platform supporting the lateral nasal soft tissues. This bony weakness, along with the significant underlying septal deformity, predisposes the cleft patient to nasal obstruction. For this reason, any patient with short nasal bones or weak ULCs requires support in the middle nasal third.

Lower Nasal Third

The lower third of the nose always exhibits some abnormality in patients with congenital clefts of the lip. In patients with unilateral clefts, there is always some asymmetry of the alar base and nasal tip. In patients with bilateral deformities, there is lack of normal tip projection and flaring of the lateral crura of the LLCs. These deformities can be partially corrected with primary
rhinoplasty. However, residual secondary deformities of the nose almost always are present. For this reason, all patients and families should expect to have definitive secondary septorhinoplasty.

The caudal nasal septum is stabilized with structural grafting (either extended spreader grafts or a septal extension graft) and repositioned into the midline. The nasal tip can then be positioned into the optimal 3-dimensional position by suspending the tip complex to the caudal septum. The cleft side LLC is advanced anteriorly, creating improved tip projection on the cleft side. This advancement equalizes and defines the nasal tip. After creating improved nasal tip projection and symmetry, a nasal tip graft can be fashioned and sutured-in-place to further increase projection and camouflage any residual asymmetry.

Treatment of the Malpositioned Alar Rim

The cleft LLC is poorly supported and always malpositioned. This malposition results from lack of alar base support. The contour of the lateral crus of the LLC is concave, as opposed to the normal (noncleft) relative convexity. Additionally, the lateral crus is typically inferiorly displaced, or hooded.

Treatment of the hooded lateral crus of the LLC can be performed by suturing the superior, or cephalic edge of the LLC to the periosteum of the ipsilateral nasal bone. Restoring the normal convexity to the lateral crural contour can be achieved with several methods. These include: 1) using the lateral crus as a free graft and flipping the concave cartilage into a convex position, 2) alar strut grafts, 3) alar rim grafts, or 4) onlay camouflage grafts. At the conclusion of the alar rim repositioning, maximal 3-dimensional alar base and tip symmetry should be present.

Summary

The cleft nasal deformity is extremely challenging and presents both functional and aesthetic problems. The tissue deficiencies associated with congenital clefting of the lip and palate involves all tissue layers, from the skeletal framework to the skin. Primary
rhinoplasty can help minimize the secondary deformity and allow more symmetric nasal growth. However, most patients have significant secondary aesthetic and functional nasal problems, requiring complex nasal reconstruction. If the patient has an associated dentofacial deformity requiring orthognathic surgery, this bony surgery should be completed prior to rhinoplasty surgery.

Secondary septorhinoplasty is performed after full nasal growth is completed. Attention should be paid to adequate straightening and support of the nasal septum. Nasal surgery usually requires structural grafting to the middle and lower nasal thirds. Structural grafting will enable the surgeon to maximize nasal function and aesthetics.

**NASAL PLASTY IN PRIMARY UNILATERAL CLEFT LIP REPAIR**

Reconstructive surgery in a third world country is what organized plastic surgery is all about. Humanitarian aid is irreplaceable, and the demand is always there. In many countries there is a limited supply of skilled, trained surgeons to help deformed children. Much of the work involves unreported cleft lip/palate cases and other birth defects. The facilities are meager and anesthesia supplies hard to arrange. The outreach programs that get the children to a local facility are limited, and secondary surgeries are often excluded due to the large amount of work on a trip. Therefore, the most surgery than can be performed on the patients is done in a single setting.

Most of the children on these trips have unilateral lip/palate clefts with significant nasal asymmetry. Much of our emphasis on past trips is to repair the primary clefts and leave the nasal tip asymmetry for another day. The author feels if a tip nasal plasty can be added to the initial unilateral cleft lip repair, the patients would not need additional anesthesia and surgery. A simple and effective technique is presented to accomplish nasal correction and prevent the need for secondary surgery.
Millard has been credited with many innovations in plastic surgery especially the repair of a unilateral cleft lip. He describes the medial rotation of the cleft lip segment with a fill of the tissue gap with a lateral advancement flap. The surgery can be used on most any cleft lip repair and offers a cut as you go technique. The surgeon can adapt these principles to any patient and obtain an excellent result. Refinements in this technique have also been published for complicated cases.

The classical surgery to repair the nasal imbalance was typically delayed until the patient was school aged. Numerous publications on these repairs demonstrate successful results. Salyer frees the alar cartilage and skin over the nasal dome. Additional mobilization is accomplished laterally with release of the soft tissue of the nose and mucosal lining. The alar cartilage is then maintained in the new position with a tie over bolus with a red rubber tube with the cartilage moved in a medial and cephalic direction. Additional suspension sutures can be utilized with his technique, and minimal external scarring results.

Cronin and Denkler presented a series of 53 patients where he used external “V” incision across the columella. The vestibular incisions went along the lower border of the lateral crus connected to an intercartilagenous incision on the cleft side. This allows excellent exposure of the alar domes for suture repair. Cross hatching the cartilage allows for flexion of the malformed cartilage. The nasal defect on the cleft side is closed with V-Y advancement. The basic goal is to secure the cleft side ala in a more medial and cephalic position.

Stal and Hollier described the nasal support structure as a tripod with lateral displacement in the unilateral cleft lip and loss of tip definition. The cleft side tip defining point is inferiorly displaced with a wide intra alar distance. Early repair with an intra dermal suspension suture with limited alar dissection is advised through an infra cartilagenous incision. The repair has the advantage of time and simplicity, and good symmetry is accomplished. External scarring is avoided.
Technique

At the time if the lip repair, an infra cartilaginous incision is made adjacent to the caudal margin of the greater alar cartilage on the cleft side. The incision is extended into the columella. The vestibular skin is left attached to the cartilage, and the dorsal skin on the cleft side is elevated. The dissection is carried across to the junction of the upper lateral cartilage on the non cleft side creating a space over the cleft side alar cartilage. A PDS suture is used and the needle is easily bent. The suture is inserted through the skin and alar cartilage through the vestibule on the cleft side. The needle is then passed through the skin above the junction of the upper and lower lateral cartilages for a maximum correction of the displaced ala. The needle is then rotated through the same skin hole but passed through a different dermal tract. The suture technique is completed when the needle is passed back through the alar cartilage leaving the knot inside the nose. The vector is medial and cephalic. The ala is then rotated to the nostril sill. You will know if you have an excellent correction as the negative space created by the nostril is even to the non cleft side. With time and experience, the added correction takes only a few minutes, and symmetry is easily accomplished. External splints are not used.

Results

The author presents four cases for review, all performed at the Obras Sociales Hermano Pedro Hospital in Antigua, Guatemala.

Discussion

The author presents a short series of successful tip rhinoplasties at the time of a primary cleft lip repair. The procedure is technically simple and gives good results even in complicated cleft lip repairs. The patients in third world countries don’t always have an opportunity for multiple surgeries so it is beneficial to the children to add the nasal tip surgery. There is minimal anesthesia time needed and little difference in recovery for the added benefit. Because these children are lost to follow up, there are no long term
photos available. The immediate post op photos, however, do show a successful improvement in nasal symmetry.

Other authors have delayed the nasal tip plasty in a cleft patient, or relied on external incisions that leave a permanent scar to solve the facial asymmetry. These procedures are more difficult and less successful. The technique presented is uncomplicated and has an immediate result that does not leave the external scars. The techniques presented by Cronin and Denkler and Salyer seem too extensive for a neonatal repair, and the time commitment in a third world country is limited due to the large case load in a one week trip for more extensive secondary surgery.

Kim et al. presented a series of Asian children with tip rhinoplasty at the time of a unilateral cleft lip repair. The average age of the patients was 3 months. Of the 412 cases they performed surgery on, 217 had additional tip rhinoplasty. The author reviewed nasal tip projection, columellar length and nasal width. The results on the rhinoplasty treated patients were compared to the non rhinoplasty patients with a long term study following cleft lip repair with anthropometric evaluation. The results demonstrate that the simultaneous correction of the cleft lip with tip rhinoplasty does not interfere with nasal growth and gives a well balanced nasal appearance.

PRIMARY REPAIR OF THE UNILATERAL CLEFT LIP AND NOSE

Analysis of the specific cleft deformity is important for surgical design. Formal anthropometric measurement is useful to objectively document the deformity and the severity. At minimum, analysis considers the lateral lip height, medial lip height, horizontal lip length, and nostril dimensions.

Planning and Design

“Make a plan and a pattern for this plan” – Sir H. Gillies

An ideal technique should facilitate the creation of a balanced lip, allow for adjustments, and produce a favorable pattern of scar.
Although each method has its own merits, the surgeon should select one that compliments his or her style. In *Cleft Craft*, Millard details much of the history of cleft lip repair. Recognizing the need to lengthen the lip, Rose and Thompson designed concave excisions of the cleft margins that provided length when closing in a straight line. This is now known as the Rose-Thompson effect. LeMesurier lengthened the lip with a Z-plasty, placing the peak of the lateral lip into the center of Cupid’s bow. Although the lip form produced was favorable, the orientation and position of scar was not ideal. Modern techniques of cleft lip repair incorporate some form of Rose-Thompson effect, Z-plasty, or both.

![Fig. : Designs for cleft lip repair and expected lines of closure: (A) LeMesurier; (B) Tennison-Randall; (C) Millard II; (D) Mohler; (E) Fisher.](image)

**The Tennison-Randall Approach**

Tennison was inspired by LeMesurier, but moved the Z-plasty to the cleft side Cupid’s bow peak. Randall built on the design using anatomic landmarks and a geometric pattern. The Tennison-Randall technique involves a back-cut that extends from the cleft Cupid’s bow peak toward the center of the philtrum that is filled by a laterally based triangular flap whose width is the measured deficiency in lip height. Two points of closure along the nostril floor are designed so that when they are brought together the nasal deformity is corrected. From these two points, corresponding lines are dropped to the cleft Cupid’s bow peak medially and to the base of the triangular flap laterally. Calipers can be used to
facilitate the final design by making intersecting arcs swung from the lateral lip (the selected Cupid’s bow peak) and lateral nostril point of closure. Cronin suggests placing the triangular flap 1mm above the vermillion to optimize definition of the repaired white roll. Brauer suggests making the repaired side 1mm shorter than the noncleft side to avoid making the lip too long. In the case of incomplete cleft lips, the lateral lip element may be too long and can be shortened by full-thickness excision below the ala. The Tennison-Randall repair relies upon rigid geometric design rather than surgeon experience and is particularly useful for wide clefts with severe vertical deficiency. However, the technique has been criticized for producing lips that are too long and the closure does not follow borders of anatomic subunits.

The Millard Approach

With the goal of preserving the philtral dimple, Millard described the rotation-advancement repair that emphasized minimal tissue discard, a “cut as you go” approach, and placement of scars that better respect anatomic borders. On the medial side, a curvilinear incision extends upward from Cupid’s bow peak toward the noncleft philtral column. Downward rotation of the philtrum corrects the deformity and leaves a gap. Advancement of the lateral lip fills the defect, corrects the alar flare, and narrows the nostril floor. Finally, a superiorly-based C-flap is elevated and transposed for nasal floor closure. The overall tissue rearrangement is much like a Z-plasty.

Although the Cupid’s bow peak on the medial side of the cleft is fixed, selection of the corresponding point on the lateral lip considers the available lateral lip height. Measurement and transposition of the horizontal lip length from the normal side tends to produce a point that is very medial and incorporates deficient cleft tissues. Noordhoff’s point is further lateral and ensures adequate tissue quality, but not necessarily the required lip height. If further height is required, the upper end of the advancement flap is limited by nasal sill and the design is moved
lateral on the lip until sufficient height to match the medial lip incision is attained. Although sacrifice of horizontal length can give the vermilion a thinned appearance, leaving a deficiency in vertical height is a much more obvious asymmetry.

Numerous modifications of Millard’s original technique have been described. A back-cut at the end of the rotation incision allows greater rotation. Another small back-cut, in or above the white roll, can be filled with a lateral triangular flap to drop the Cupid’s bow further. In the case of a vertically oriented philtrum, the rotation incision can be kept on the cleft side to avoid crossing anatomic borders. Millard described extending the advancement incision around the alar base; however, this should be abandoned as it is unnecessary and produces a conspicuous scar. Millard also described using the C-flap to lengthen the columella, especially if a back-cut is added to the rotation incision. Stal has compiled a comprehensive description of the many subtle variations used by notable surgeons. An important modification is that described by Mohler.

The Mohler Modification

Dissatisfied with a scar that traverses the upper third of the philtrum, Mohler modified Millard’s repair and used the columella to lengthen the lip. The rotation incision is designed to mirror the normal philtral column and extends onto the columella. A back-cut is designed to end at the lip-columellar junction and the C-flap is used to both fill the columellar defect and abut the rotated lip segment. Lip closure follows anatomic subunits and the concept of using the columella to lengthen the lip has gained popularity.

The Fisher Approach

Fisher recently described another approach to cleft lip repair that avoids scars on or under the columella and is not limited by deficiencies of lateral lip height or width. The design is measured and geometric, but uses anatomic landmarks to place closure along borders of anatomic subunits. Lip length is attained by the
Rose-Thompson effect and a small triangle placed within the concavity immediately above the white roll. Compared with other techniques, it is a "measure twice, cut once" style of repair. The design relies upon 25 landmarks and can be time consuming.

The sequence of landmarks begins with central and noncleft side points so that the corresponding cleft side points can be measured and identified. Three points are placed along the crease between the lip and columella: the center and the two peaks of the philtral columns. While manually correcting the nasal deformity, two points are placed at each alar base: the subalare (lowest part of the ala) and the alar insertion point (junction of ala and sill). An arbitrary point is identified within the noncleft nostril that is collinear with the two noncleft alar base and the two noncleft columellar landmarks. The arbitrary point can then be transposed to the cleft side to produce two points along a line of closure. By manually bringing the points of closure together, the nasal deformity should be corrected.

On the medial side of the lip, the center and two peaks of the Cupid’s bow are identified along the vermilion border, above the white roll, and along the red line. The medial incision runs along the base of the medial footplate, down the philtral column, and perpendicular to the white roll and red line. A back-cut is designed above the white roll to augment lip height and along the red line to augment vermilion. On the lateral side, Noordhoff’s point and the corresponding points above the white roll and along the red line are identified. An incision is designed perpendicular to the white roll and down the vermilion to match the medial lip vermilion height. The remaining vermilion is incorporated into a flap for augmentation. The point above the white roll defines one fixed point; the previously identified lateral point of closure within the nostril floor defines the other fixed point. Between these two points, three components need to be designed to fit the medial lip markings: the limb along the medial footplate, the length of the cleft-side philtral column, and a small triangular flap (whose width is defined by the relative deficiency in philtral height minus 1mm
Unilateral Cleft Lip and Nose Repair

because of the Rose-Thompson effect). The angle between each limb can be varied much like the limbs of an articulating ruler so that the components span the two fixed points. Although the planning for a Fisher repair is extensive, there is less reliance on surgeon experience, and the anatomic basis allows it to be reliably applied to a wide spectrum of clefts.

Comparison of Techniques and Changes with Growth

It is difficult to compare different designs of lip repair due to variations in cleft severity and surgeon expertise. Although outcomes of traditional triangular and rotation-advancement repairs have been found to be similar, rotation-advancement tends to produce short lips when used for wide clefts. For this reason Meyer uses a Tennison-Randall repair for wide clefts and a Millard repair for narrow clefts. The suggestion that imbalances occur from differential growth has been challenged by studies that have found relative lip dimensions to be stable with both triangular and rotation-advancement repairs. The immediate result is likely the best predictor of eventual outcome, and the results of surgery rely on more factors than just the surgical markings.

Wide Surgical Release

“Treat the primary defect first” – Sir H. Gillies

Although Gillies’ notion of wide surgical release is based upon traumatic deformities, the principle is well applied to clefts. The lip and nose are tethered to the distorted underlying anatomy; much like a burn contracture, there is a point of maximal tension that can be clearly visualized when traction is applied to the lip and nose. Adequate release allows three-dimensional (3D) correction. Wide mobilization over the maxilla permits medial and superior movement, whereas release along the piriform rim allows anterior movements. Correction of the nasal deformity requires that the alar base, lower lateral cartilage, and accessory cartilages are free from the maxilla. Wide muscle release permits functional OOM reconstruction, but dissection should be
discriminating. Care must be taken to preserve the philtral depression and the J shape of the orbicularis along the lower lip margin.

**Component Reconstruction**

"Losses must be replaced in kind" – Sir H. Gillies

**Nasal Floor**

Repositioning of the alar base is crucial in correcting the nasal deformity. In the case of a bony defect, nasal floor closure provides a stable platform for accurate 3D repositioning and rotation of the ala. Lateral vestibular skin can be apposed to skin along the medial footplate; more posteriorly, lateral vestibular mucosa can be apposed to septal mucosa. Closure even further posterior requires an extended incision along the palatal shelf for elevation of the nasal mucoperiosteum. Single- and double-layer closures of the nasal floor extending into the palate have also been described. An alternate method that preserves the palatal mucoperiosteum uses an anteriorly based turbinate flap transposed 90 degrees. In addition to stabilizing the nose, nasal floor closure facilitates subsequent palatoplasty and alveolar bone grafting by sealing the nasal mucosa along the alveolus when the exposure is wide and easy.

**Nasal Sidewall**

With great anterior movement of the lateral nose, release of the mucoperiosteum leaves a potential space along the piriform rim. This defect can be addressed in several ways depending upon surgeon preference or the clinical scenario. The turbinate flap is anteriorly based and rotates 90 degrees to fill the defect after release of the lateral nose. Harvest requires an open cleft palate for posterior access. It replaces like with like tissue and preserves all of the nasal mucoperiosteum that may be used for palatoplasty. The L-flap is the marginal lateral lip vermilion and mucosa that would otherwise be discarded with cleft lip repair. Blood supply can be robust if it is based upon periosteum of the lateral nasal wall. The flap is transposed into the defect along the nasal vestibule
while more posterior mucoperiosteum is mobilized to close the nasal floor. Although nasal mucosa is replaced by lip vermilion and mucosa, the L-flap is versatile and can be used in any scenario. Lateral nasal wall advancement involves movement of mucoperiosteum in continuity with the rest of the nose as a broad flap. Incision along the palatal shelf allows elevation of mucoperiosteum and a back-cut posterior to the piriform aperture leaves the defect along the bony nasal wall. Although the flap is robust, the release is posterior to the site of greatest tension and a low-lying turbinate can limit the extent of the back-cut.

Following wide release of the lateral nose and component reconstruction, absorbable quilting sutures along the vestibule and alar crease can be used to obliterate the vestibular web, support the lower lateral cartilage, and create better definition for the nose.

**Nasal Septum**

Disruption of the palatal arch results in untethered growth of palatal segments and rotation of the anterior nasal spine away from the cleft. Displacement of the caudal septum has a ripple effect on the rest of the septum and nasal cartilages. Smahel described correcting the position of the caudal septum at the time of cleft lip repair to improve nasal form. No alteration in maxillary growth was reported and other surgeons report similar favorable results. The caudal septum is approached via the medial lip incision and is found behind an often bifid anterior nasal spine. Firm attachments on the noncleft side need to be released to unfurl the cartilage and reposition it to the midline of the face.

**Nasal Tip Cartilages**

The nasal tip cartilages sit on top of a deformed nasal base. Dissection of the nasal tip was once criticized for potential growth disturbance, but short-term anthropometrics and long-term subjective analyses have demonstrated no alteration in growth. McComb describes suspension of the cleft alar dome via long sutures tied over bolsters at the glabella, whereas Tajima describes
suspension to the upper lateral cartilage and the contralateral lower lateral cartilage. Many surgeons have incorporated nasal tip dissection and have used limited vestibular incisions, an extensive intranasal approach, or an open external approach for exposure. Although the greater dissection affords the ability to manipulate and modify anatomy, it also risks iatrogenic insult. Warnings of scarring, vestibular stenosis, micronostril, and other iatrogenic deformities have accompanied reports of favorable outcomes. Proponents of primary nasal tip rhinoplasty admit that nasal correction can be limited and that there is a “perverse tendency for the genu to slump with time.” Objective long-term audit demonstrates deterioration of alar symmetry over time, especially with wide clefts. Nasal revision is performed in 20 to 74% of patients and at some centers most patients go on to definitive septrhinoplasty. As such, the balance of surgical manipulation against surgical insult with nasal tip correction at lip repair must be considered.

Controversies in Correction of the Cleft Lip Nasal Deformity

“Never do today what can honourably be put off till tomorrow” – Sir H. Gillies

The composite tissues and complex shape make the nose a difficult structure to correct. With presurgical molding, various forms of primary rhinoplasty, and variations in postoperative stenting, the relative impact of each intervention on the ultimate result is unclear. For example, NAM has been associated with improved outcomes without any nasal dissection, with primary rhinoplasty, and with varying durations of postoperative nasal stenting. Likewise, septal repositioning has been associated with improved nasal form with and without nasal tip dissection. Analysis needs to consider early results, late results, deterioration over time, and treatment outcome at completion. The lack of any universally accepted objective assessment makes comparison of the various components of treatment difficult. While the relative
merits of molding and various forms of primary rhinoplasty remain unclear, surgeons need to constantly reassess their outcomes as they relate to their treatment protocols.

Alveolus

Gingivoperiosteoplasty (GPP) is a mucoperiosteal flap closure of the cleft alveolus that is typically performed following NAM if the alveolar segments are in close proximity. Adequate bone can form within the constructed cavity in up to 73% of patients. Although GPP is used with good bone production and no apparent alteration in facial growth by some centers, GPP has not gained widespread use due to reported concerns of facial growth disturbance and variable quality of alveolar bone.

Lip Mucosa

Adequate upper buccal sulcus incision and release allows the lateral lip mucosa to advance to meet the medial lip mucosa. If the cleft side buccal sulcus hangs low on the alveolus, the mucosa can be secured to periosteum higher up. Final inset of mucosa requires accurate alignment of the red line.

Lip Musculature

Anatomic studies have emphasized the importance of accurate muscle repair. On the medial side, release of muscle from the columella lengthens the lip and opens a space. On the lateral side, downward rotation of muscle from the alar base creates an “empty triangle.” When the lateral muscle is inserted into the base of the columella, a muscular sling for the nasal sill is created. At the same time, the empty triangle docks against the ala at the nose–cheek junction and the height of the medial lip muscle is augmented. Further muscle repair establishes the oral sphincter, aligns the overlying structures, and reduces tension on skin repair. Particular care should focus on aligning the J shape of the caudal OOM as it contributes to the lip’s natural pout. If a traction stitch is used at the lower end of the muscle, the surgeon must ensure that muscle form is not distorted and the pout is not obliterated.
Cleft and Cleft Palate: Causes and Treatments

Lip Skin and Vermillion

Final adjustments are well worth the investment in time as the form achieved at the completion of the procedure predicts the ultimate outcome. The white roll and vermilion should be perfectly aligned and the lip and nose should have balanced form. Adjustments will vary according to the technique used.

PRIMARY UNILATERAL CLEFT LIP REPAIR

The unilateral cleft lip in its varying manifestations of shape, size and asymmetry is a complex deformity. To obtain consistent results one requires basic training in soft tissue handling, an understanding of the bony foundations of the face, followed by experience and a fair amount of craftsmanship.

In the late 1950s the senior author was introduced by his mentor Charles Pinto to the straight repair of Rose and Thompson as modified by Peet, who called it the “Oxford modification of the straight repair”. In the hands of the artistic Peet it gave good results.

In his search for something better, Charles Pinto brought back from Barrett Brown’s unit at St Louis, Missouri, a form of the triangular flap of Mirault that had been modified by Vilray Papin Blair, Brown and Mc Dowell into a smaller triangular flap. The Blair-Brown-McDowell plan held centre stage for a good 10 years. The stature of these three great men and their artistry was probably one of the reasons why this procedure flourished. In our hands the results were no better than the straight repair. There was not the slightest semblance of a Cupid’s bow in these repairs; instead there was an unnatural central peak and in most cases a tight lip resulted. Secondary corrections of these lips were always difficult.

A major breakthrough in cleft surgery took place when Le Mesurier, an orthopaedic surgeon working at the Hospital for Sick Children at Toronto, used Werner Hagedorn’s quadrilateral flap and for the first time created a Cupid’s bow. No surgeon at the time could ignore the positive advantages of having a nice
Cupid’s bow. As time went on and the long-term results of the Le Mesurier repair were shown at conferences, it became obvious that the lip on the cleft side became long and over-hanging and the scar, like the triangular flap was unnatural and did cut across the normal philtral line.

As the ‘Le Mesurier’ began to fade out, Tennison’s modification with a Z-plasty began to be accepted. Peter Randall did to the Tennison what Blair and Brown had done to the Mirault - he made his triangle smaller and marked his points with greater precision. Sawhney of Chandigarh improved on the Tennison-Randall’s operation, making the cutting of the triangular flap almost geometrical in its precision. With Sawhney’s contribution, the triangular flap became easy to teach and easy to execute and is still quite popular with surgeons in North India. When well executed, the Tennison-Randall-Sawhney procedure gives good results. The scar however is unacceptable and, when not properly executed, secondary repairs are difficult.

Somehow, we at the Charles Pinto Centre, missed out on the Tennison-Randall-Sawhney improvements and went straight on to the rotational advancement technique of Millard. In 1958, on his last visit to India, Sir Harold Gillies demonstrated the rotational advancement technique to a group of Indian surgeons at Pune. He turned around to the fascinated audience and said “Gentlemen, try this one - I think it has merit, but I must warn you that it has not yet been published!” The Millard procedure broke like dawn on the Indian horizon and caught the imagination of surgeons the world over by its clear, logical thought process.

Millard said that:

- All the previous flap procedures based their logic on the false premise that the actual defect in the cleft is in the lower third of the lip, which is not so. Discarding precious tissue in Tennison’s approach when there was already poverty, is against all established plastic surgical principles.
- Three quarters of the Cupid’s bow is present on the non-cleft side, but is riding high. What better way of bringing
it down in a horizontal line with its fellow than by a rotational flap? No rotational flap is complete without a back cut and this not only further helps to drop the obliquely oriented Cupid’s bow, but compensates for the contracture of the straight line of the Millard procedure.

• This main rotational flap is taken from the rich non-cleft side and not from the poverty stricken cleft side as in the triangular and quadrilateral flap procedures. (“It is unwise to borrow from Peter to pay Paul when Peter can ill afford it”).

• The defect thus created is in the upper part of the lip and can be hidden under the overhanging nostril.

• What better way of filling this defect than by advancing a flap from the cleft side.

• The advancement flap gives the additional bonus of correcting the nostril flare.

• The “C” flap helps to lengthen the short columella.

• The scar imitates the philtral line, creates a philtral column, a philtral dimple and a slight pout which adds charm to the finished result. The scars of both the triangular and quadrilateral flaps crisscross Langer’s lines, which again is contrary to basic tenets of plastic surgery.

This to our mind is the eight-fold path to the ‘Cleft Nirvana’ that the reconstructive surgeon wants to achieve. The authors would not like to give an impression that mere reading of these eight points would ensure a good result. The Millard procedure needs to be taught on the table, needs a considerable amount of virtuosity on the part of the surgeon and it needs a fair amount of experience. Unlike the ‘Tennison-Sawhney’ there are very few mathematically precise points to mark and you can “cut as you go” depending upon the needs of the case, keeping your eye on shape and symmetry. As Millard remarked “all art depends on freedom for its vitality for no two lips are identical - they may be similar but never identical”. The straight line part of the Millard incision often contracts and pulls the Cupid’s bow up in the first
Unilateral Cleft Lip and Nose Repair

A few months, but in a year’s time it descends without any further intervention.

Critics of the Millard operation have often said in publications that the rotational advancement procedure is only suitable for partial clefts and not for complete ones. This statement is far from the truth. To the original Millard theorem, in this presentation, we have added our own original method of correcting the nostril deformity and our method of avoiding a notch or whistle deformity on the vermillion.

Protocol

Timing of Surgery

The “rule of tens” has been followed widely in many parts of the world. However, this does not apply to our country. We are physically a smaller people and many of our children are undernourished. We undertake surgery for these children when they are at least 5 Kg in weight. On an average, our children attain this weight by five to six months. Neonatal surgery is not recommended in view of the risks involved and the need for a compromise on the surgical procedure to minimise the time and extent of the surgery. Miniature tissues are difficult to work on and work in the nose is well nigh impossible.

Pre-surgical orthodontics

This is being followed in many centres across the world. We believe that without any pre-surgical intervention, we are able to achieve results at least on par with those from centres using some form of pre-surgical orthodontics in unilateral cleft lips. Expense and patient compliance are also factors to be taken into account. Hence we do not use any orthodontic intervention prior to surgery.

Procedure

We use the standard Millard incisions. The rotation flap at its superior end hugs the base of the columella. We always make an ample back cut, taking care not to encroach onto the philtral
column on the non-cleft side. An adequate rotation incision with a back-cut is required to get the Cupid’s bow points at the same horizontal level. If the back cut were to transgress the philtral column on the non-cleft side this would cause a lengthening of the lip on that side.

We do use the peri-alar component of the Millard incision for the advancement flap. Many contemporary authors have abandoned this, as they are apprehensive about the visibility of the scar. We have, however, used this in more than 7,000 cleft lips and we are entirely satisfied that the scar is not obvious if the incisions are placed precisely at the base of the ala in the alar groove. The advantage of the peri-alar incision is that one can dissect the paranasal muscles under vision and include them in Millard’s Cinch suture. This suture traverses the membraneous septum and takes a bite on the paranasal muscles before going back through the septum. This helps in correcting the alar flare. However, one should be careful when tightening this suture, as one can easily cause extreme narrowing and deformity of the nostril base by excessive tightening. In addition to the Millard Cinch suture, we use an additional Cinch suture with 5.0 prolene at the nasal sill. This goes through the subcutis medially, and laterally through the dermis. When this is tightened, the shape of the nostril improves significantly.

While we basically follow the Millard technique, the senior author has included several technical refinements to the procedure. These have been in relation to the primary correction of the nasal deformity and in producing a notch-free vermillion.

**Notch-free vermillion**

A notch is a common blemish following cleft lip repair. The senior surgeon at our centre analysed the main causes leading to the formation of a notch and addressed them using a protocol which has been adhered to on all unilateral cleft lip patients operated on at our centre. As a result, we have been able to consistently obtain a notch-free vermillion.
Causes of vermillion notching

- Inadequate rotation of the medial element of the lip resulting in a tented-up Cupid’s bow point on the cleft side and a notch on the vermillion
- Turning-in of the sutured edges around the vermillion
- Deficiency of bulk of the orbicularis oris at the vermillion
- Contracture of the straight line scar on the mucosal aspect of the lip

Having noted the above causes, an attempt was made to correct each of them.

- Adequate rotation of the medial element with an ample back-cut in all patients.
- Undermining the skin and mucosal edges to prevent their turning in. This undermining is limited to a few millimetres from the cleft edges.
- While paring the vermillion, an excess of muscle tissue is retained on both the medial and lateral elements. As a result, there is a good bulk of muscle tissue that acts as a filler. At least three 6/0 Nylon (Ethilon) sutures are placed to bring this muscle together, thus creating the appearance of “a roll” or “sausage”.
- To counteract the straight line scar contracture, a Z plasty is mounted on the mucosal aspect of the lip and an attempt is made to align Noordhoff’s red line.

All the above mentioned steps are done in all our unilateral cleft lip patients.

Primary closed rhinoplasty and extensive septal correction

Primary correction of the nasal deformity associated with the unilateral cleft lip has come to be accepted as the norm today. Many authors have recommended primary nasal correction. Some of them have used a closed approach. Others have used an open technique. There is also a group of authors who recommend a semi-open approach. However, there is a consensus that some
form of primary nasal correction must be done. We use a closed rhinoplasty technique.

With the help of Kilner’s scissors, we approach the ala from both the medial and lateral aspects. The medial approach is from the incision at the base of the columella. Laterally, the scissors are introduced at the base of the ala through the peri-alar incision.

The scissors are used to dissect in the plane between the dorsal skin and the alar cartilages - both the lower and upper lateral cartilages are completely separated from the skin. The dissection is carried out till the nostril rim to free all superficial attachments of the alar cartilages.

A more limited dissection is carried out on the non-cleft side up to the dome. The freed lower lateral cartilage is fixed to the upper lateral by means of bolster sutures.

The correction of the associated septal deviation is yet to gain universal acceptance. It is well documented that the septum is deviated towards the non-cleft side anteriorly. The anterior nasal spine is itself similarly displaced to the non-cleft side.

We approach the septum by incising over the mucoperichondrium on the cleft side on the groove at the base of the septum. The mucoperichondrium is carefully stripped off the septal cartilage.

We then proceed to divide the septo-spinal ligament in order to expose the anterior border of the septal cartilage. This is an important step to avoid shearing of the septal cartilage when we proceed to strip it off the mucoperichondrium on the non-cleft side. This is done after incising the junction of the cartilages with the underlying maxillary crest.

The septal cartilage is also freed from the vomer and the perpendicular plate of the ethmoid. The cartilage thus freed will buckle when repositioned in the midline. Hence, a sliver of cartilage is excised inferiorly. We believe in Sir Harold Gillies’ philosophy that all cleft lip noses require some shortening. Hence
Unilateral Cleft Lip and Nose Repair

we excise a thin wedge of septal cartilage anteriorly. This causes an upward recoil of the nasal tip, enhancing its projection. There is usually a residual bow-string effect to the cartilage even after all these manoeuvres. This is nullified by scoring with a knife on the non-cleft concave side of the cartilage until it is flail. Finally, it is hitched to the newly reconstructed nasal floor on the cleft side to overcorrect the deviation, and with time it comes to lie in the midline. A sliver of the excised cartilage is used as a vertical strut graft in the columella.

Many cleft surgeons have shied away from primary septal correction following apprehensions regarding the effect of this on subsequent nasal and maxillary development. However, the senior author has been following this radical septal correction for the past 40 years and we have not found any detrimental effect on any of our patients on long-term follow-up.

In fact, we strongly believe that this has helped the overall functional outcome of the nose in our unilateral cleft lip patients. This has also been the view of other exponents like Samahel et al. who have objectively studied the long-term effect using cephalograms. Other authors like Anderl have confirmed that there is no additional deleterious effect in the long-term to maxillary or nasal growth from septal cartilage repositioning.

Other Refinements

High-riding nostril

Often, in patients who have a wide alveolar disparity between the medial and the lateral elements, we note that the nostril base on the cleft side comes to lie at a more superior level than its counterpart on the non-cleft side. This discrepancy has been corrected using an unequal Z plasty on the nasal floor. Ever since we commenced using this additional procedure, the incidence of such high-riding nostrils has diminished dramatically. In children with severe alveolar disparities, sometimes we have had to perform two such Z plasties.
Another common deformity after a unilateral cleft lip repair is a deficiency in height of the lateral vermillion on the cleft side. This deformity has also been noted with other types of cleft lip repairs including the triangular repair. We believe that it is an inherent component of the unilateral cleft lip deformity and has nothing to do with the method of correction. As far as we know, there is no procedure documented so far to prevent this occurrence. However, it can secondarily be corrected either by a Gillies hemi-Cupid’s bow procedure or by a V-Y mucosal advancement.

Unilateral partial cleft lip

The technique is essentially the same as for the complete variety. There is obviously no need for a nasal floor repair. However, there is some element of nasal deformity in most of these patients. Hence, we perform a closed alar cartilage dissection in all these patients. One needs to be wary while making the rotation and the back-cut as it is possible to lengthen the lip excessively.

Microform cleft lip

These have variously been referred to as “a minimal cleft”, “occult cleft” “forme fruste cleft” and “nature’s union”. The deformity always includes a vermillion notch. There may be, in addition, a white roll mal-alignment, a scar or a furrow on the body of the lip and a flattened alar cartilage with a wide nostril. When the deformity is confined to a notch of the vermillion, a notch correction procedure including muscle build-up and a Z plasty on the mucosa are all that is required. When there is no upward displacement of the Cupid’s bow point, a simple straight repair (Rose-Thompson) would suffice.

However, in the more significant deformities that require downward rotation of the Cupid’s bow point and closed nasal dissection, we follow Millard’s procedure. But in the majority of these patients that have good muscle continuity across the cleft, we restrict the Millard incisions to the skin and subcutis and do
not cut into the muscle. This form of a “cutaneous Millard’s” rotation advancement procedure minimises the trauma inflicted on these patients with trivial deformities and helps in better scarring post-operatively. A muscle build-up is sometimes necessary. This innovation has been used by the senior author for many years now.

**Vestibular web**

This is yet another vexing problem encountered during unilateral cleft lip repair. Some surgeons indulge in excision of the webbed vestibular skin and mucosa in the belief that there is an excess. However, we believe that there is no real excess of vestibular lining. This is also the view held by other exponents. The fold forms at the upper border of the lower lateral cartilage and can only be eliminated if the lower lateral cartilage is hitched to the upper lateral. This may be done blindly in the closed technique or under vision in the open technique of primary nasal correction.

A Z plasty was described by Charles Pinto, mentor to the senior author, but remained unpublished till the present. The vertical limb of the Z is along the web. The two oblique (60°) limbs are then marked with the upper limb on the medial, and the lower limb on the lateral aspect. Care must be taken when elevating the vestibular lining flaps so that the underlying cartilage is not damaged. This procedure also helps in reorienting the axis of the nostril.

**Soft triangle deformity**

With good primary nasal correction we have been able to consistently obtain acceptable results. However, in most of these cases, there remains a residual soft triangle droop. In many this is trivial. In some patients it is significant enough to require correction by a secondary rhinoplasty. With the present improved state of the art of secondary rhinoplasty, a good percentage of our patients are subjected to this procedure in an attempt to achieve well nigh perfection.
No cleft surgeon should forget the pathos of this deformity and the severe psychological trauma that it inflicts on parent and child. A plastic or reconstructive surgeon is really a general surgeon with a hobby and that hobby lies in the aesthetic realm of a refined reverence for tissue and the true appreciation of the dignity and beauty of the normal human form. His art would be quite meaningless if he reconstructed a face but failed to put a smile on it. The true plastic surgeon must always hope that the skill of his surgery will help towards the healing of all the internal scars that external wounds do cause.
Bilateral Cleft Lip and Nose Repair

Although multidisciplinary care for the people affected by orofacial clefting has undergone many advances, surgical correction of the nasal deformities associated with bilateral cleft lip remains a challenge. Various single- and multiple-stage procedures have been used. Deformities may become apparent after further growth and development of the nose, making the bilateral cleft lip nasal deformity a 4-dimensional problem.

Many early surgical approaches for repairing the bilateral cleft lip and its nasal deformity in a single stage resulted in scarring and corrections that did not last. This led to the belief that primary repair may interfere with growth of the nasal cartilages and that the nasal deformity should not be corrected in a secondary staged procedure after nasal growth is complete. Soon, evidence began to refute this belief, and some have claimed that early surgery may assist growth.

Tan et al found in a survey of surgeons listed in the American Cleft Palate/Craniofacial Association that 108 (52%) of 210) performed primary nasal repair, and 102 (48%) of 210) preferred secondary nasal repair.

Many centers recently have presented results of children treated with preoperative orthopedic management followed by
single-stage correction. Tan et al found in their survey that 71% of surgeons would perform some form of preoperative dentofacial orthopedics for a complete bilateral cleft lip; the most common technique was nasoalveolar molding (NAM) at 55% of those administering preoperative treatment.

The history of the surgical treatment of the bilateral cleft lip nose is long and fascinating; for a more complete treatment, the reader is directed to Millard’s *Cleft Craft*. Below are some of the influential surgeons and a brief summary of their contributions.

**Manchester**

Manchester described fairly simple technique for repair of the bilateral cleft lip, in which the nose is minimally repaired, if at all.

**Millard**

Millard maintained that the columella is actually shortened because the dislocated alar cartilages have not stretched the columella properly, and he designed a repair with this in mind. He begins by presurgical active orthopedics with an intraoral fixed device. If insufficient prolabium is available, lip adhesions may be used to enlarge the prolabium. At the time of primary surgery, he is then able to make gingivoperiosteoplasties on the alveolar segments. During the lip repair, he banks prolabial forked flaps in the nasal floor (“whisker” flaps). When the patient is about 4 years old, Millard advances the forked flaps into the columella.

**Broadbent and Woolf**

Broadbent and Woolf were among the first to describe simultaneous primary repair of the lip and nose. They believed that early surgery certainly did not retard growth and might actually assist it. Their experiences taught them that a good primary repair endures; however, any deformities not fully repaired do not improve with time. Their method consists of making an incision in the superior cleft defect extending between the upper lateral cartilage (ULC) and lower lateral cartilage (LLC) to the tip. The
superior border of the LLC is undermined, and the skin of the nasal tip and over the ULCs is freed. They made a midline incision of the nasal tip and sutured together the domes of the LLCs, which lengthened the columella. They also relocated the LLCs by pulling them upward and medial to the ULCs with sutures.

**Cronin**

Cronin advocated using floor of the nose tissue to lengthen the columella as a secondary procedure.

**Mulliken**

Mulliken advocates lengthening the columella and reconstructing the nasal tip by suturing the genu of the LLCs together. He initially used a single central vertical incision over the tip of the nose to expose the genu of the LLC, but later found that incision was unnecessary, as the LLCs could be approached through rim incisions. He now advocates bilateral vestibular rim incisions and alar base incisions. The LLCs are freed on their anterior surfaces through the nasal incisions. He then elevates and sutures the genu of the LLCs together. The lateral portion of each dome is suspended to the ipsilateral ULC near the septum. The freed alar bases are held to the prolabium medially via a mattress suture from alar base to base. Excess tissue in the soft triangle is excised. When necessary, a ridge of vestibular lining is excised.

**Nakajima**

Nakajima reported a series of 169 patients who had primary nasal cartilages repair through rim incisions similar to Mulliken’s approach. Nakajima brought his incision out onto the external skin near the tip. As the LLCs are moved superiorly, this external skin is folded under to form a “soft triangle.” Fifteen-year follow-up showed good results.

**McComb**

In 1986, Harold McComb published a 10-year follow-up study of repairs he had accomplished using a 2-stage procedure. It
involved lengthening the columella with forked flaps taken from the prolabium when the patient was aged 6 weeks, with repair of the lip and nasal deformity performed 6 weeks later. Later, he began to notice complications associated with this type of columellar lengthening and abandoned this technique in favor of another 2-stage procedure. In describing his new procedure, he noted that, embryologically, the prolabium belongs to the lip and therefore should not be used to reconstruct the columella.

In McComb’s current technique, preoperative orthopedic appliances are used. In his first stage, when the patient is aged 6-8 weeks, the nasal floor is repaired and lip adhesions are performed. A V-shaped incision is made above the nostril rim with the tip of the V ending over the dorsal columella. The nasal skin is widely undermined over the LLCs through the incision in the cleft margin. The soft tissue between the domes is removed, and the domes are then sutured together. The nasal flap is closed in a V-Y advancement, lengthening the columella by approximately 5 mm. Mattress sutures over bolsters are placed to eliminate any dead space in the tip. At the second stage, 1 month later, the prolabium is lifted and mucosal-muscular flaps are sutured behind it, completing the lip repair.

**Salyer**

Salyer performs his lip repair procedure, along with a limited nasal repair, when the patient is aged approximately 3 months. Initial surgery entails bilateral superiorly based prolabial flap elevation developed around the flap to be used for philtral reconstruction. These are rotated into the nasal floor after bilateral vertical intranasal alar and alar base incisions allow freeing of the LLCs and rotation of the bases medially. The lip repair is then completed.

Further nasal reconstruction is achieved when the patient is aged 12-15 months. Incisions are made below each alar base running toward the midline. Bilateral horizontal rim incisions meet in the midline and extend up the columella. Through this incision, the
alar cartilages and columellar skin are freed. The alar cartilages are advanced, and the domes are sutured together. This lengthens the columella and redefines the tip through advancement of nasal floor tissue. No permanent sutures are described between ULCs and LLCs. Instead, Dacron bolsters are placed and left for 6-7 days to promote LLC support, nasal tip projection, and lateral crux stabilization, which is thought to eliminate vestibular webbing.

Salyer and Genecov have published 40-year follow-up studies of techniques, with good long-term results.

Noordhoff

Initially an advocate of a 2 stage procedure with forked flaps, Noordhoff now uses a 1-stage lip and primary nasal repair, performed when the patient is aged 3 months. The alar bases and orbicularis are sutured to the nasal spine to attempt to prevent columellar drift. The domes are also sutured together at the tip to improve tip projection.

Cutting and Grayson

In 1993, Cutting and Grayson described a prolabial unwinding flap method as a single-stage reconstruction of the bilateral cleft lip and nasal deformity, relying on presurgical orthopedic appliances and sufficient prolabial size. The repair involves an oblique, asymmetrical incision beginning at one side of the columellar base and continuing inferiorly and medially. This creates a flap that forms the inferior columella and philtrum. The asymmetrical nature of this reconstruction, however, led to a very high incidence of revision surgery, and they ultimately abandoned it.

In 2004, Cutting and Grayson described the use of nasoalveolar molding (NAM), using a very elegant but work-intensive molding device. Their protocol begins with a passive presurgical orthopedic device, fitted to the patient’s alveoli and adjusted weekly. They progress to add wire outriggers that press up on the collapsed nasal vestibule. This is felt to remodel the cartilage and even create
increased tissue for the subsequent less-involved surgical repair of the nose. Cutting addresses the nasal tip by extending the prolabial flap via a transmembranous septal incision. The medial crura of the LLC are then elevated with the columellar flap. This avoids the problem of flap ischemia with the McComb approach, and it allows suturing the genu of the LLC together from a retrograde approach.

**Delaire**

Delaire, on the other hand, offered the opinion that the best orthopedic treatment is an anatomic and functional surgical repair achieved in a single stage at approximately age 6 months. The columella can be lengthened at the time of the lip repair by precise repositioning of the lower lateral cartilages and good control of the healing process. He maintains that there is no skin deficiency of the columella, but he uses a nasal retainer to preserve the patency of the nasal valve.

**REPAIR OF THE PRIMARY DEFORMITY**

The goals of primary bilateral cleft lip nose surgery are:

- Closure of the nasal floor and sill
- Lengthening of the columella
- Repositioning of the alar base
- Achieving nasal tip projection
- Repositioning of the lower lateral cartilages
- Reorienting the nares from horizontal to oblique position.

**Timing for correction**

There are differences of opinion regarding the optimal timing for nasal repair. All surgeons perform limited nasal repair like nasal floor reconstruction and narrowing of alar base at the time of primary lip repair. The timing of final revision of soft tissue nasal deformity differs. Some surgeons take the child up for surgery at one year of age and others at four to seven years of age.
Limited correction of the nasal deformity done at the time of primary lip repair improves results of second stage of primary repair. At our centre, Bardach’s method is commonly used. We perform limited nasal correction, that is, reconstruction of nasal floor and sill at the time of lip repair. We correct the lower lateral cartilages before the child goes to school. Some centres prefer Salyer’s two-stage technique. The first stage is performed at three months of age. It consists of lip repair along with banking of tissue into the nasal floor. Nasal soft tissue correction is done at one year of age. It is considered to be the second stage of primary bilateral cleft lip-nose repair. Early correction of the nasal deformity would assure a more normal relationship of the columella-lip angle, better projection and definition of the tip, and also allow subsequent growth in a more normal anatomic relationship.

Mulliken believes that the columella is not short but lies within the nose. He advocates primary repair of the nasal cartilage along with the lip repair at three to five months of age. We have currently adopted this principle. We perform total primary nasal repair at the time of lip repair. We have also incorporated presurgical nasoalveolar moulding (NAM) in our protocol to aid in single stage repair. This is because the size of the premaxillary segment and the extent of its protrusion vary considerably. In complete clefts, it is often necessary to retroposition the premaxilla before definitive lip repair.

The principle objective of presurgical NAM is to reduce the severity of the initial cleft deformity enabling the surgeon to enjoy the benefits associated with repair of an infant presenting with a minimal cleft deformity. The goals of NAM include lip segments that are almost in contact at rest, symmetrical lower lateral alar cartilages, and adequate nasal mucosal lining, which permits postsurgical retention of the projected nasal tip. Presurgical NAM also includes the nonsurgical elongation of the columella, centering of the premaxilla along the midsagittal plane, and retraction of the premaxilla in a slow and gentle manner to achieve continuity with the posterior alveolar cleft segments. Presurgical nasoalveolar
moulding uses both an intraoral alveolar moulding device and nasal moulding prongs. Successful use of any presurgical orthopaedic devices requires a team approach.

If properly performed, presurgical nasoalveolar moulding can provide soft tissue expansion and mould the nasal architecture, thereby decreasing nasal deformity. Our initial results incorporating NAM with primary lip and nose repair have been encouraging.

PREOPERATIVE DENTOFACIAL ORTHOPAEDICS

Alignment of the maxillary segments sets the stage for synchronous, bilateral, nasolabial repair. Retrusion and centralization of the premaxilla permits design of the philtral flap in proper proportions, facilitates nasal correction, and allows soft tissue closure of the alveolar clefts, which stabilizes the maxillary arch and eliminates oronasal fistulas. Furthermore, premaxillary retropositioning minimizes the nasolabial distortion that occurs during the rapid growth of early childhood.

There are two basic dentofacial orthopedic strategies: passive and active. A passive appliance maintains arch width, but requires some type of external force to retract the premaxilla. Bilateral labial adhesions have been tried for this purpose since the mid-19th century; however, they often dehisce because of tension and absent muscle in the prolabium. Other methods include traction by an elastic band attached to a custom-fitted head cap or application of pressure on the prolabium by cheek-to-cheek tape. Cutting and Grayson have described a more sophisticated variation on a passive plate and taping called “nasoalveolar molding” (NAM). After the alveolar gap is reduced, an acrylic extension is added to the palatal plate that pushes the nostrils upward against a counterforce of soft material across the nasolabial junction, thus stretching the diminutive columella. This apparatus must be re-taped and adjusted frequently, necessitating repeated visits over several months. Another type of passive plate and nasal outrigger, without any need for tape, has been reported by Bennun and Figueroa.
The Latham appliance is the best-known active-type device for premaxillary moulding. It is constructed from a plaster cast of the upper jaw. Most centers have the appliance fabricated in Ontario, Canada, although it can also be done locally. The device is pinned to the maxillary shelves, and the parents turn a ratcheted screw daily to expand the anterior palatal segments. Visits are necessary at one, three, and five weeks to tighten the bilateral elastic chain that retroclines the premaxilla. Usually, the premaxilla is aligned within 6–8 weeks.

The merits of active versus passive dentofacial orthopedic methods continue to be discussed by proponents of each approach. Cleft lip centers differ in their capability to provide this service. Dentofacial orthopedics may not be available in developing countries. Furthermore, infants in these nations often present when they are older than 6–12 months of age; by when, the premaxilla is rigid and manipulation is not possible. Another dilemma, sometimes seen in every land, is the twisted premaxilla that fails to be centralized despite several weeks of dentofacial manipulation. In these situations, the surgeon may resort to staged repair (with/without labial adhesion) or closure of the labial clefts over the procumbent premaxilla. Premaxillary ostectomy/setback and gingivoperiosteoplasties should also be considered in these predicaments.

Premaxillary retropositioning must be done with great care. Premaxillary circulation can be compromised by the mucosal incisions and dissection necessary for resection of the premaxillary neck and inferior septal cartilage, which is required to permit alveolar closure. With careful attention to mucosal blood supply, premaxillary retropositioning can be accomplished in an infant at the same time as nasolabial repair. If the child is near one year or older, then speech becomes the first priority. The strategy for these children is to first close the secondary palate and retroposition the premaxilla (with its labial blood supply), along with soft tissue closure of the alveolar clefts. Nasolabial correction is scheduled later on a solid maxillary foundation. Primary premaxillary
retropositioning might be called heretical because it is likely to accentuate midfacial retrusion, but the child’s nasolabial appearance takes precedence. The majority of children with repaired bilateral complete cleft lip/palate will need maxillary advancement anyway, after completion of skeletal growth.

The Operation

Bilateral cleft lip presents in three major anatomic forms: bilateral symmetrical complete (50%); bilateral incomplete (25%), and asymmetrical bilateral (complete/incomplete) (25%). The technical steps are first described for the most common, the bilateral complete type, after which, modifications are suggested for the major variants.

Note: The day of bilateral cleft lip repair is the most important day in the child’s life. It should be the first case of the morning. The surgeon must work slowly, carefully, and take as much time as necessary to do the very best operation. There should be no distractions such as scheduled meetings, patients waiting in the office, or other obligations for that day.

Bilateral Complete Cleft Lip and Palate

Markings

The anatomic points are designated using standard anthropometric initialisms. The philtral flap is drawn first while the nostrils are held upward with a double-ball retractor. A sharpened tooth-pick is used for drawing; brilliant green dye (tincture) is preferred over methylene blue (aqueous). The dimensions are determined by the age of the child and ethnicity. The average age at primary repair is five months. At this time, the length of the philtral flap is usually the same as the height of the cutaneous prolabium (usually 6–7 mm). If the prolabium is overly long, the philtral flap should be shortened to this length. Philtral flap width is set at 2 mm at the columellar-labial junction (cphs-cphs) and 3.5–4 mm between the proposed Cupid’s bow peaks (cphi-chpi). The sides of the philtral flap should be drawn slightly
concave because the scars tend to bow. The dart-like tip of the philtral flap should not be overemphasized. A thin rectangular flap is drawn on each side of the philtral flap. These side flaps will be de-epithelialized and will come to lie beneath the lateral labial flaps in an effort to simulate the elevation of philtral columns.

The proposed Cupid’s bow peaks are carefully sited on the lateral labial elements and marked just atop the white roll above the vermilion-cutaneous junction. These points are situated so that there is some medial extension of the white roll that will form the handle of the Cupid’s bow and sufficient vermilion height to form the central raphe and median tubercle. Curvilinear incisions are drawn at the juncture of the alar bases and lateral labial elements. Nostril rim incisions are marked and extended along the inside edge of the upper columella. Nasal and labial segments are infiltrated with lidocaine (0.5%) / epinephrine (1:200,000). After waiting for five minutes, the critical points including the lateral vermilion-mucosal junctions, are tattooed with tincture of brilliant green.

Labial Dissection

First, all labial incisions are lightly scored. The flaps flanking the philtral flap are de-epithelialized, the remaining prolabial skin is discarded, and the philtral flap is elevated (including subdermal soft tissue) off the premaxilla up to the anterior nasal spine. The white-roll-vermilion-mucosal flaps are incised (just short of the tattooed lateral Cupid’s bow point) and the lateral labial elements are disjoined from the alar bases. These basilar flaps are freed from the piriform attachments by incision along the inferior cutaneous-mucosal (inter-cartilaginous) junction. The mucosal incisions on the underside of the lateral elements are extended distad, on the anterior side of the gingivolabial sulcus, to the premolar region. With a double-hook on the muscle layer, the lateral labial elements are widely dissected off the maxilla in the supraperiosteal plane. This permits greater advancement of the cheek than possible using subperiosteal dissection. A protective ring finger is held on the
infra-orbital rim as this dissection extends over the malar eminence. Releasing the lip from the maxilla is a critical maneuver; this is needed to minimize tension on the muscular closure and permit tension-free cutaneous closure. The orbicularis oris bundles are dissected in both the subdermal and submucosal planes for 7–10 mm.

**Nasal dissection**

Using a semi-open approach through bilateral rim incisions, the anterior surface of the slumped and splayed lower lateral cartilages is exposed by scissor-dissection, aided by elevation with a cotton-tipped applicator on the mucosal underside. This dissection continues superiorly over the upper lateral cartilages and across the dorsal septal junction. Interdomal fatty tissue is elevated and partially excised. Perichondrium-to-perichondrium heals more securely without intervening soft tissue.

**Alveolar Closure**

Vertical incisions are made on each side of the premaxilla and on the facing gingiva of the lesser segments. Alveolar gingivoperiosteoplasties are completed. The nasal floors are closed using a lateral mucosal flap raised from below the inferior turbinate and a medial flap from the premaxilla. The alar base flaps are advanced and sutured to the edge of the constructed nasal floor. The vermillion component of the premaxillary mucosa is trimmed and the remaining mucosal flange is secured high to the premaxillary periosleum to construct the posterior side of the central gingivolabial sulcus.

**Labial Closure**

Advancement of the lateral labial elements during closure of the sulcus is difficult to illustrate. Nevertheless, this maneuver is critical to muscular closure, tension-free philtral closure, and a protrusive posture of the lip. A back-cut is made at the distal end of the sulcal incision, and the sulcus is closed while the labial flap
Bilateral Cleft Lip and Nose Repair

is pulled mesially with a double hook. The lateral labial mucosal lining forms the anterior wall of the central gingivolabial sulcus.

The orbicular bundles are apposed, end-to-end (inferiorly-to-superiorly), using either a vertical mattress or simple polydioxanone sutures. Polypropylene suture is used to suspend the pars peripheralis and nasalis to the periosteum of the anterior nasal spine.

Construction of the median tubercle begins with the insertion of a fine chromic suture to join the white-roll-vermillion flaps at the midline; this is placed about 3 mm medially from the tattooed lateral Cupid’s bow point. Excess vermillion-mucosa is trimmed from each flap and the junction aligned to form the median raphe. There is a natural inclination to save too much vermillion-mucosa in these flaps resulting in a central furrow.

Attention is turned to correction of the nose before insetting the philtral flap.

Nasal Correction

Three techniques have been described to suspend and secure the displaced lower lateral cartilages: 1) bolster sutures; 2) transfixion sutures, and 3) intercartilaginous sutures. The first two suture methods are done blindly; the third semi-open approach is preferred. Under vision, the genua are apposed with a 5-0 polydioxanone (1/2 circle cutting needle). The lateral crura are elevated and secured to the ipsilateral upper lateral cartilage with a 5-0 polydioxanone mattress suture; a cotton-tipped applicator in the nostril beneath the genu facilitates inserting and tying these sutures. In an infant, usually only one upper-to-lower lateral suture is necessary (or possible), whereas 2–3 such suspension sutures are placed in an older child.

The c-flap on each side of the columellar base is trimmed to 3 mm in length. The alar bases are advanced, rotated endonasally, and sutured side-to-end to the c-flaps. Usually, the tip of the alar base flap is conservatively trimmed as closure of the sill is
completed. A polydioxanone suture is placed through the maxillary periosteum in the region of the depressor alae nasi and left untied. This is best done prior to apposition of the upper orbicularis oris. A “cinch” suture of polypropylene is brought through the dermis of each alar base, passed beneath the philtral flap, and tied to narrow the interalar distance (al-al) to less than 25 mm. The previously placed maxillary periosteal suture is brought above the pars peripheralis and nasalis, then though the alar base, and tied. This suture simulates the depressor alae nasi and: 1) gives a cymal shape to the sill; 2) prevents alar elevation with smiling; and 3) minimizes postoperative nasal widening.

**Final Touches**

In an effort to form the dimple, a suture is brought through the dermis in the lower one-third of the philtral flap and through the underlying orbicular muscular layer. The tip of the philtral flap is inset into the handle of Cupid’s bow. In a complete bilateral cleft lip, it is unnecessary to adjust the leading edge of the lateral labial flaps before apposing them to the philtral flap with interrupted, fine dermal and percutaneous sutures. The cephalic margin of the labial flaps must be trimmed to correspond to the position and cymal configuration of the alar bases. Labial flap-to-sill closure proceeds laterally-to-medially.

After anatomic placement of the lower lateral cartilages, it is obvious that there is redundant domal skin in the soft triangles and upper columella. A crescentic excision of this extra skin is drawn along the leading edge of the rim incisions, extending inferiorly along each side of the columella. This resection narrows the nasal tip, defines and tapers the mid-columella, and elongates the nostrils. Interdomal apposition also accentuates the extra lining in the lateral vestibule; this is corrected by lenticular excision.

An internal resorbable nasal splint is used rather than an external splint. A short, curved polylactic-polyglycolic resorbable plate is inserted into the nasal pocket above the newly positioned lower lateral cartilages. Immediate postoperative nasolabial
anthropometry is documented and placed in the child’s record. After measuring, a strip of 1/4 inch Xeroform® gauze is wrapped around a 19 gauge silicone catheter and a 1 cm segment is inserted into each nostril.

Postoperative Care

A Logan bow is taped to the cheeks to: 1) protect the labial repair and 2) hold an iced-saline sponge for 24 hours postoperatively. The infant is usually discharged from hospital on postoperative day #2. The parents are instructed in suture-line care. The percutaneous sutures are removed and the nostrils are cleaned 5–6 days postoperatively under general anesthesia using mask induction and insufflation. A ½ inch transverse Steri-Strip™ (3M Health Care, St. Paul, Minnesota) is trimmed to fit the sn-ls dimension and placed over the labial scar; this tape is changed as needed for six weeks. Thereafter, parents are instructed on how to perform digital massage and warned about the importance of the application of a sunblock ointment.

Figure: (A) Preoperative image of newborn with bilateral complete cleft lip/palate prior to dentofacial orthopaedic manipulation, (B) Intraoperative markings: note philtral flap designed 2.0 mm at cphs-cphs and 4.5 mm at cphi-cphi with lateral tabs.
BILATERAL CLEFT LIP VARIATIONS

Binderoid bilateral complete cleft lip and palate

The nasal features in this rare variant are: orbital hypotelorism, hypoplastic bony/cartilaginous elements, conical columella, short septum, and absent anterior nasal spine. The labial features are: hypoplastic prolabial-premaxillary segment (containing a single tooth bud) and thin vermilion in the lateral labial elements. Dentofacial orthopedic manipulation is usually not possible because the premaxilla is floppy; furthermore, it is also unnecessary because the premaxilla is not protuberant. Synchronous repair is accomplished as described above. Sometimes, the premaxilla is so small that alveolar gingivoperiosteoplasties cannot be accomplished. The philtral flap need not be made overly small because it will expand very little with growth (because the premaxilla is so tiny). The lower lateral cartilages are very small; however, they can usually be dissected, positioned, apposed, and elevated. Specific secondary procedures in this variant include dermal grafting to augment the median tubercle and to widen the narrow columellar base; possible cartilage grafting of the tip and nasal dorsum; and maxillary advancement along with augmentation of the fossae praenaseale.

Bilateral complete cleft lip and intact secondary palate

Approximately 10% of infants born with bilateral complete cleft lip and cleft alveolus have an intact secondary palate. The premaxilla is procumbent and unyielding to any attempts at dentofacial orthopedic manipulation. This situation is another indication for premaxillary set-back undertaken synchronously with nasolabial repair. There are two alternatives to ensure adequate premaxillary blood supply through the mucosa/periosteum: 1) postpone attachment of the posterior edge of the premaxilla to the anterior edge of the hard palate, or 2) delay alveolar gingivoperiosteoplasties. Midfacial retrusion is very unlikely to evolve in the presence of an intact secondary palate.
Bilateral incomplete cleft lip

One-fourth of bilateral cleft lips are bilaterally incomplete; most are symmetrical. This variant is the most easily corrected of all. The concepts for design and execution are the same as for a complete bilateral cleft lip, including the need for adjustments based on expected nasolabial changes with growth. Nevertheless, there are crucial technical points that must be considered. The first relates to the construction of the median tubercle. Usually, the tubercle should be formed using the lateral white roll-vermilion-mucosal flaps. However, if the clefts are minor (< 50% of the cutaneous lip) and the central white-roll is prominent, the prolabial vermilion-mucosal may be retained as the central segment with flanking scars. The next consideration is the columella: measure its height (sn-c). If the columellar length is normal and the lower lateral cartilages are in reasonable position, then it may be unnecessary to adjust the lower lateral cartilages and sculpt the nasal tip. Interalar narrowing is always needed as this dimension is overly wide preoperatively and increases with growth. If there is any separation of the alar domes, they should be apposed through the semi-open approach. In contrast to the complete deformity, both the leading and superior edges of the lateral labial flaps may have to be trimmed.

Asymmetrical bilateral (complete/incomplete) cleft lip

One-fourth of bilateral cleft lips are asymmetrical: complete on one side and incomplete on the other side. Severity of the cleft on the incomplete side determines the operative strategy. The following classification system of incomplete clefting is useful in categorizing the types of repair for these asymmetrical variants.

“Incomplete” is a general term applied to a cleft lip in which there is cutaneous continuity between the medial (nasomedial process) and lateral (maxillary process) elements. Incomplete cleft lips present in a wide spectrum, beginning at the severe end with those with a thin cutaneous band (that some would argue constitutes a “complete” cleft lip) to lesser-forms that have been
called by various names, e.g., microform, minimal, and occult. Yurzuriha and Mulliken subdivided these lesser-forms into minor-form, microform, and mini-microform as determined by the degree of disruption at the vermillion-cutaneous junction. Minor-form cleft lip extends 3–5 mm above the normal Cupid’s bow peak, i.e., 50% or less of the normal cutaneous labial height (sbal-cphi). Microform cleft lip is characterized by a notched vermillion-cutaneous junction, whereas the Cupid’s bow peak is elevated < 3 mm above normal. The mini-microform cleft lip is distinguished by a discontinuous white-roll without elevation of the Cupid’s bow peak. The severity of the nasal deformity, muscular depression, and mucosal notching correspond in these three categories of lesser-form cleft lip.

Symmetry, the first principle of bilateral labial repair, is the foremost concern in planning correction of these asymmetrical variants. An algorithm for timing and techniques for repair of asymmetrical bilateral cleft lip. If both greater and lesser sides are incomplete clefts or the lesser side is minor-form, then synchronous bilateral repair is indicated. However, if the greater side is incomplete, it alone is first repaired if the contralateral side is a microform or mini-microform. If the greater side is complete, it is initially addressed by unilateral dentofacial orthopedics and followed by nasolabial adhesion and alveolar gingivoperiosteoplasty; this levels the surgical field. If the contralateral (lesser side) cleft lip is a minor-form or a more severe incomplete form, the next procedure is simultaneous bilateral nasolabial repair. During this second stage, technical maneuvers on the complete side are exaggerated because the distortions and tensions are greater than on the incomplete side. If the contralateral cleft is a microform, it is best to close only the complete side, following the rotation-advancement principle, along with primary nasal repair. After the scar has remodeled, the contralateral microform is repaired using a double unilimb z-plasty, which includes muscular reapposition, dermal graft, and nasal correction. This three-stage stratagem is most likely to result in acceptable mirror-image nasolabial symmetry.
If the contralateral cleft lip is a mini-microform, this can be addressed along with repair on the greater side, although nothing is lost by waiting. Often, only minor adjustment of the nasal tip is needed if there is a contralateral mini-microform. Augmentation of the median tubercle is almost always necessary.

**TREATMENT OF COMPLETE BILATERAL CLEFT LIP-NASAL DEFORMITY**

**Recording Of Pathology**

There are many possible variations of the bilateral cleft lip. The morphology can vary from being complete on both sides to asymmetric with a complete cleft on one side and incomplete on the other. Kernahan’s “striped Y” method cannot fully illustrate the range and diversity of the asymmetric cleft. The double-Y numbered classification, reported by Noordhoff in 1990, is a more accurate method for recording as well as a more suitable system for computer database documentation. For patients with a complete cleft of primary and secondary palate on one side and a complete cleft of the secondary palate on the other side, this classification can record the pathology in a more accurate way than the single striped Y classification.

**Evaluation Of Pathology**

There is a wide variation in the quality and amount of tissue in the prolabium, premaxilla, nasal cartilages, vomer, and lateral lip elements. All bilateral clefts have some amount of asymmetry in their horizontal or vertical dimensions. All cleft patients have a certain amount of tissue deficiencies. These deficits are most severe in bilateral medial facial dysplasia patients, who, therefore, have a less than optimal outcome after lip and nose repair. They always have a significant growth disturbance and require orthognathic surgery when they reach skeletal maturity. It is important to document these deformities or any preexisting asymmetry, or both, prior to surgery to assess the postoperative results more accurately.
General Plan Of Management

Presurgical orthopedics, nasoalveolar molding, is started on the first visit. The aim of this molding process is to centralize the premaxilla, narrow the alveolar gaps, match the alar cartilages, and elongate the columella.

This process usually takes 3 to 4 months to achieve an optimal outcome. The initial surgery is usually performed at 3 to 4 months of age, depending on the result of the molding process. The palate is repaired at about 12 months of age together with the insertion of grommet tubes.

Speech assessment is started at 2.5 years. If the patient requires speech therapy, it starts at 3.5 years. Velopharyngeal insufficiency is diagnosed by nasoendoscopy at 4 years old and corrective surgery for velopharyngeal insufficiency is performed as soon as the diagnosis is made. Residual alveolar clefts are closed before the eruption of canine teeth, usually when the child is 9 to 11 years old. If the patient has any psychological problems related to any residual lip or nasal deformity, a revision surgery is usually done before the child enters primary school.

Presurgical Orthopedics/Nasoalveolar Molding

The purpose of presurgical orthopedics or nasoalveolar molding is to restore a more normal nasal shape and a balanced skeletal base. The following techniques have all been used in Chang Gung Craniofacial Center for the past 20 years.

Presurgical Orthopedics

The protruding premaxilla may be gradually pushed back by applying micropore tapes across the lip with or without traction rubber bands. It is suggested that the patient sleep in either the prone or side-lying position to increase pressure on the cheeks. The movement of the alveolar segments is controlled by an acrylic plate. This simple technique is effective in expanding the prolabial tissue and places the premaxilla in a better position.
Nasoalveolar Molding

Silicone Nasal Conformer

A silicone nasal conformer can be used as a tool for presurgical nasal molding when the patient has an incomplete cleft lip. The height of the conformer can be adjusted by gradually adding some soft resin or flat silicone sheets on the domes.

Grayson’s Technique

A passive-type orthopedic appliance is used together with taping of the lip for premaxilla and alveolar molding. The protruding premaxilla is molded first into a proper position. When the alveolar gap is approximated and the arch is aligned, a nasal molding device is added to the orthopedic appliance to increase the columellar length as well as to reshape the alar dome. A nonsurgical lip adhesion is performed by placing tape across the upper lip. The tape aids in the closure of the clefts, decreases the width of the base of the nose, and helps to approximate the lip.

Figueroa’s Technique

Alveolar molding and nasal molding are performed simultaneously using an acrylic plate with rigid acrylic nasal extension. Rubber bands are connected to the acrylic plate for gentle retraction of the premaxilla backward. A soft resin ball attaching to the acrylic plate across the prolabium is sometimes used to maintain the nasolabial angle.

Liou’s Technique

The nasoalveolar molding device is composed of a dental plate, two nasal components for nasal molding, and several micropore tapes for premaxillary retraction. Denture adhesive (Poligrip, Australia) keeps the dental plate on the maxillary lateral segments. The nasal components are made up of 0.028-inch stainless steel wire projecting forward and upward bilaterally from the anterior part of the dental plate. The top portion contains a soft resin molding bulb that fits underneath the nasal cartilages for
nasal molding. Micropore tapes are placed across the cleft lips and prolabium to minimize the alveolar cleft and retract the premaxilla. At the same time, they pull both alar bases medially. Retraction of the premaxilla and lengthening of the columella are performed at the same time. The columella is lengthened and stretched by pulling on the premaxilla backward. The nasal tip is kept at the same height while the premaxilla is pulled back. Rather than pushing forward, the soft resin molding bulbs basically support the nasal cartilages and nasal tip.

The key point of nasal molding in bilateral clefts is to push the alar domes forward in a sagittal direction for columellar lengthening instead of pushing the domes upward in a cephalic direction into a turned-up nasal tip. Nasoalveolar molding techniques require regular patient follow-up with an interval of 1 to 2 weeks. Grayson’s technique approximates the alveolar cleft before the nasal molding. Both Figueroa’s and Liou’s method achieve nasal and alveolar molding at the same time.

**General Surgical Principles**

There are several surgical principles that need to be stressed. They are as follows: (1) preserve the presurgical columellar length; (2) keep the width of the central lip segment narrow without compromising the blood supply; (3) advance the columella prolabium complex superiorly to allow reconstruction of the orbicularis oris muscle behind the prolabium; (4) release the alar cartilage attachment from the pyriform rim and provide additional coverage of this soft tissue deficiency with the use of inferior turbinate flaps; (5) release and reposition the lower lateral cartilage; (6) adequately dissect above the maxillary periosteum; (7) reconstruct the nasal floor by local mucosal flaps; (8) reconstruct the prolabial buccal sulcus with tissue from the prolabium; (9) reconstruct the orbicularis muscle sphincter and attach it to the anterior nasal spine; (10) reconstruct a new Cupid’s bow, central vermillion, and lip tubercle with tissue from lateral lips; balance the height of both lateral lips without an incision around the ala; and maintain the presurgical nasolabial angle.
Surgical Procedure

Markings and Measurements

The landmarks of the lip are marked out on the prolabium and both lateral segments. The various vertical and horizontal measurements are evaluated for any asymmetry. The width between CPHL and CPHR is usually maintained at 5 to 6 mm. The central segment is gradually narrowed toward the columellar base and maintained at 4 mm in width at the level of the columellar base. Traction applied to the alae is usually needed to identify the nasolabial junction. The incision lines are kept straight, not curvilinear. The proposed peak of the Cupid’s bow on the lateral lips (CPHR’ and CPHL’) is marked at the point where the vermilion first becomes widest and usually would be 13 to 15 mm from the commissure or 3 to 4 mm lateral to the converging junction of the red line and white skin roll (WSR).

Central Segment

A double hook is used to retract the columella up, and a small single hook is used to stretch the prolabium. The central segment is developed by laying a number 11 blade on the incision line of the prolabium to give a straight cut. The two forked flaps are developed with lateral incisions on the skin-vermilion junction extending behind the columella up into the membranous septum and continuing up along the skin-mucosa junction to the dome area, then along the lower border of the lower lateral cartilages (LLCs) as a gull wing open rhinoplasty incision or outside the alar rim as a Trott incision. The central segment, the forked flap, and the columella are raised as a unit to expose the cartilaginous framework. The central part of the vermilion and mucosa of the prolabium is used for the lining of the raw surface on the premaxilla. The lateral parts of the prolabial mucosa flaps (PM flaps) are used for nasal floor reconstruction.

Lateral Segments

The incision is made from the proposed peak of Cupid’s bow
along the cleft edge to the edge of the alveolar cleft. The incision is right above the WSR to develop a WSR-vermillion-free border flap. This flap will be used for reconstruction of the central Cupid’s bow. An L-mucosal flap is raised along the cleft edge. The incision is then turned upward along the pyriform rim and then around the inferior turbinate to be incorporated with the inferior turbinate flap. The dissection is carried above the periosteum on the maxilla. The abnormal muscle insertion on the lateral segment is released adequately until the lateral segment can be brought medially to touch the medial segment without tension. The cleft edge is then opened to develop the WSR-vermillion-free border flap. The dissection on the mucosal side is limited to 2 mm, and the dissection on the skin side is quite extensive to separate the abnormal muscle insertion from the skin. The dissection is carried below the alar base to release the abnormal muscle component that inserts to the alar base.

Nasal Floor and Muscle Reconstruction

The inferior turbinate flap is used to fill in the defect on the pyriform area after the LLCs are advanced. The turbinate and L flaps are sutured together, brought across the cleft, and sutured to the septal incision to reconstruct the nasal floor. Special attention must be focused on the width of the nostril. The PM flap is sutured below the L-flap for lining. The orbicularis muscles are approximated with 4-0 polyglactin sutures with the upper edge sutured to the anterior nasal spine.

Nasal Reconstruction and Cupid’s Bow Reconstruction

The separated LLCs are approximated by absorbable sutures, 5-0 polydioxanone, or nonabsorbable sutures, 5-0 polypropylene, depending on the surgeon’s preference. The fibrofatty tissue on the nasal tip is brought to the top of the approximated nasal tip. The skin flap of the central segment is then sutured to the lateral lip. Through-and-through alar transfixion sutures are placed on the alar-facial groove to provide further support to the LLCs. The excessive tissue on the nasal floor is adequately trimmed and the
floor is closed. The full-thickness WSR, vermillion, and free border flap are brought together below the central segment to reconstruct the central lip. Excessive orbicularis marginalis muscle on the tip of the WSR–vermillion–free border flaps is preserved for augmentation of the lip tubercle.

**Postoperative Care**

The wounds on lip and nose are covered by antibiotic ointment without any dressing. The sutures are removed 5 to 7 days after surgery at the outpatient clinic. The lip scar is supported by micropore tapes as well as silicone sheets for 6 months. A silicone nasal splint is needed for 6 to 9 months. Throughout this period, the height of the splint is gradually increased by adding silicone sheets to the domes of the splint. The central prolabial portion of the lip will gradually widen and lengthen by the age of 3 years. The nasal width will also increase, similar to the central prolabial portion of the lip width. The columella length will shorten slightly after the primary lip repair and then remain stable without further growth, while the rest of the nose will grow significantly in both height and width.

**Discussion**

**Comparison of the Molding Techniques**

The different techniques of alveolar or nasoalveolar molding are used in Chang Gung Craniofacial Center. Grayson’s technique, with emphasis on approximating the alveolar clefts before nasal molding, achieves the best preoperative nasal shape symmetry and skeletal base balance. However, it is also the most expensive and time-consuming method. Figueroa’s and Liou’s techniques of performing alveolar and nasal molding at the same time are simpler and less expensive methods. A study comparing the three techniques in unilateral clefts showed that the latter two techniques tend to result in a larger diameter in the cleft side nostril postoperatively.
Gingivoperiosteoplasty

Millard, Mulliken, and Cutting all advocated the importance of primary gingivoperiosteoplasty. The long-term result from Cutting and Grayson’s report showed that 60% of the patients who received primary gingivoperiosteoplasty do not need alveolar bone grafting later on. However, it is very difficult to perform a primary gingivoperiosteoplasty unless the alveolar gap is around 1 to 2 mm. Figueroa’s and Liou’s techniques tends to leave the alveolar gap larger, 3 to 4 mm, which limits the possibility of a primary gingivoperiosteoplasty.

Central Segment Height And Width

There is a significant difference in the outcome of the shape of the central lip in the bilateral cheiloplasty with or without muscle approximation. In the technique without muscle approximation, the central lip tends to become wider and remains short. With muscle approximation, the central lip segment has less widening but more lengthening. Mulliken advocated narrowing the central lip width down to 2 to 3 mm for a better long-term result. Noordhoff, in attempting a primary elongation of the columella by interdigitating the forked flaps into a transverse incision in the columella, found two vessels running from the columella to the prolabium. A central segment that is 2 mm wide at the columellar base may injure the vessels. A 4-mm-wide base of the prolabium includes both columellar vessels, providing a good blood supply to the prolabium. The long-term result shows a tendency of widening as well as lengthening of the central segment. A wide central segment in primary lip repair, although maintaining a good blood supply, may result in an unnaturally wide Cupid’s bow.

Regarding the vertical height of the central lip, Lee advocated that the vertical height of the central lip in a 3-month-old baby should be around 7 mm. However, the height is somewhat predetermined by the size of the prolabium. A prolabium shorter than 7 mm should not be lengthened on the operating table, as
it will always lengthen vertically with muscle approximation and have a satisfactory appearance. A relatively longer prolabium can provide additional tissue for the columella and make it easier to achieve a better nose. The critical problem in determining the vertical height of the lip occurs when there is a marked discrepancy between the vertical height of the central lip and the lateral lips. In this situation, the surgeon should vertically shorten the lateral lip to match the vertical height of the central prolabial portion of the lip. Otherwise, the nasal tip will be pulled downward because of the tension in the central segment. Even a short vertical length of 4 to 5 mm will elongate adequately with muscle repositioning.

**Forked Flap**

Millard suggested preserving the prolabial tissue lateral to the central segment as forked flaps that are banked on the nasal floor. These banked forked flaps are used for columellar lengthening in secondary revisions. The experience in Chang Gung Craniofacial Center does not support his concept in Oriental patients. Pigott studied the ratio between dome component and columellar component in Caucasians of varying ages. The dome columellar ratio is much greater in Orientals than in their Caucasian peers. A nose with a disproportionately long columella often results after a columellar elongation procedure using the banked forked flaps. The banked forked flaps also end up with unsightly scarring on the nasal floor. The authors do not bank these forked flaps. They are trimmed to an adequate size and sutured backward to the septum to improve the nasolabial angle. The report from Nakajima et al suggested a similar approach.

**Septal Incision – In Front Or Behind The LIC’s**

Cutting et al raised the central segment tissue behind the medial crura of LLCs and reported that it has a safer blood supply to the prolabium. Trott and Mohan used a technique of raising the central segment in front of the LLCs. The Chang Gung experience comparing the two techniques shows that there is no difference in terms of blood supply to the central prolabium between these
two techniques. Cutting and Noordhoff believe that the medial crura need to be elevated superiorly on the septal cartilage, and Trott and Mulliken leave the LLCs attached to the septum. In the authors' experience, the two techniques offer a similar early result. Technically, the retrograde dissection and approximation as advocated by Cutting is more difficult compared with the technique of approximation of cartilages under direct vision as advocated by Trott.

**Nasolabial Angle**

In his anatomical dissection around the nasolabial angle, Wu showed that the angle is maintained by a ligament from the subcutaneous tissue to the anterior nasal spine. Whenever the columella-prolabium complex is raised, the nasolabial angle tends to be flattened after operation. However, this procedure is definitely necessary as the separated orbicularis muscles need to be approximated under the prolabium to achieve an anatomical repair. The technique with the placement of the incision behind the medial crura tends to maintain a better nasolabial angle postoperatively than the technique with the incision located in front of the medial crura. Restoration of the ligament by a tuck-down suture from skin to anterior nasal spine may jeopardize the blood supply to the prolabium. Suturing the tips of the forked flaps backward to the septum may be more helpful in maintaining the nasolabial angle.

**Open Versus Closed Rhinoplasty**

The approximation of the LLCs can be achieved through either an open or closed rhinoplasty. The authors' experience shows a similar result with the two techniques. Technically, a closed rhinoplasty with two rim incisions or bilateral Tajimaincisions is simpler than the open rhinoplasty through a gull wing or Trott incision. However, an open gull wing incision, approximating the LLCs through direct vision with nonabsorbable sutures, is the author's preferred method. This allows the surgeon better visualization for accurate approximation of the LLCs. The open
technique also provides a better approach for redraping or redistributing the central segment tissue.

**Postoperative Nasal Shape Maintenance**

Friede et al used a postoperative acrylic molding splint to improve nasal configuration. Other reports used a similar concept for postoperative maintenance. In Chang Gung Craniofacial Center, a silicone conformer is routinely used after surgery and proved its efficacy in maintaining the postoperative nasal shape in unilateral clefts as well as in bilateral clefts. It is necessary to use the splint for at least 6 months postoperatively while waiting for scar maturation.

**Muscle Dissection**

Delaire suggested wide subperiosteal dissection on the maxilla to achieve a functional closure. There is still controversy about whether a subperiosteal or supraperiosteal muscle dissection is better in terms of function or subsequent facial growth. There are no scientific data supporting the concept that a subperiosteal dissection results in less scarring or better facial growth. A muscle dissection above the periosteum seems to offer a better release of the abnormal muscle insertion around the alar base from both the skin side and periosteal side. The technique presented here keeps the extent of muscle dissection as minimal as possible but still adequate for muscle approximation at the center. This should create minimal scarring or muscle tension in front of the maxilla.

**Muscle Reconstruction**

Manchester felt that the orbicularis muscle should not be reconstructed as it would cause too much tension and growth disturbance. Nagase et al showed that there was no significant growth disturbance after muscle reconstruction. There is a definite difference in the appearance of the bilateral lip repair with or without muscle reconstruction. Muscle reconstruction produces a much better result both functionally and aesthetically.
Cupid’s Bow And Lip Tubercle

The techniques leaving the prolabial WSR and vermilion on the prolabium to reconstruct the Cupid’s bow result in a Cupid’s bow with abnormal peaking, indistinct prolabial WSR, and irregular vermilion with a depressed scar at the central lip. The quality of the WSR from the lateral lips is much better compared with the WSR of the prolabium. The reconstruction of the central lip by advancing the WSR–vermilion–orbicularis marginalis flaps from the lateral lip beneath the prolabial segment gives a continuous WSR, underlying vermilion, parallel red, and a full central prolabial tubercle without notching.

HORIZONTAL INCISION ON LATERAL LIPS

From the experience in unilateral cleft lip repair, the horizontal incision below the nasal floor is usually unnecessary. Nevertheless, the alar-facial groove has a better appearance if the skin is kept intact. The surgeon needs only to approximate the orbicularis muscles. However, in the presence of a vertical discrepancy between the central lip and lateral lips, a horizontal incision below the nasal floor may be needed. The lateral incision is used for shortening of the longer lateral lip.
A cleft lip is a form of lip malformation that occurs very early in pregnancy, when the baby’s facial features are just beginning to develop. The tissues that create the top lip do not come together and fuse normally. This results in an abnormal slit, or “cleft”, in the upper lip. The nostrils and tip of the nose are also usually affected. Cleft lip deformities can affect one or both sides of the upper lip. They can also occur in conjunction with a cleft in the roof of the mouth. This is known as a cleft palate. Cleft palates can also occur in isolation.

Surgery can correct a cleft lip, but leaves behind a facial scar on the top lip and around the base of the nose that can be quite visible. Most parents are understandably very eager to fade and minimize their child’s scar after cleft lip surgery. The fresh scar is usually red, which is normal. As any wound begins to heal, the body creates new tiny blood vessels to bring extra blood to the area to help healing. This extra blood flow can make new scars appear quite red. Unfortunately, in some cases, the scar redness continues beyond this initial healing phase. The new scar may also become raised and firm.

It can take several months before the scar begins to soften, flatten, and fade. However, with proper care, the scar can eventually become much lighter, thinner and softer.
When Will the Scar Go Away?

Unfortunately, most surgical scars are permanent so there is no way to get rid of a cleft lip scar completely. It can take a year or longer for the scar to fade, soften and look its best.

Some scars don’t mature well and develop into more prominent scars over time. The other issue with children is that they grow. As they grow, their facial features (and facial scars) grow too. While the amount of scarring in proportion to the facial features stays the same, the absolute size of the scar gets bigger, and potentially more visible, as children grow into adults.

Will Vitamin E Help?

Many parents prefer to use a “natural” scar treatment and choose vitamin E. However, vitamin E has not actually been shown to reliably improve scars, and may even cause skin irritation in as many as 1/3rd of users. Aloe vera is a much safer “natural” choice.

Topical silicone is the gold standard in scar care; a product that combines silicone with natural ingredients like aloe vera is the most logical (and effective) choice for most parents.

The preferred scar therapy can be started as soon as the initial wound has healed, often within just a couple of weeks after surgery, as long as this is cleared by the surgeon ahead of time.

What Can I Do to Maximize the Results?

As you wait for your scar therapy to work, there are several things you can do to ensure the best results. First and foremost, a healthy diet is essential for normal healing, something you’ll already be providing for your little one!

Keep your child’s scar out of the sun as much as possible. Sun exposure increases scar pigmentation and can make scars permanently dark or red. Use a good tear-free sunscreen every time your child goes outside. Just be sure to put the sunscreen on after the scar therapy; the scar therapy should be applied directly
Laser Treatment of Cleft Lip Scars

to the skin. Scar massage can also really help soften firm scars but this can be very tough to do in children. Try to apply pressure to the upper lip in a circular motion for as long as your child tolerates. This helps break up scar tissue and softens firm scars. One technique is to grasp the upper lip between the thumb and index finger and then lightly squeeze the scar in a circular motion.

**SCAR TREATMENT OPTIONS AFTER CLEFT LIP AND PALETTE SURGERY**

One of the more common birth defects in babies is a cleft lip and palette. This is a gap in the natural formation of the upper lip that is treatable with surgery. According to the Centers for Disease Control and Prevention (CDC), an estimated 2,651 babies are born in the US each year with a cleft palate; about 4,437 are born with a cleft lip. This malformation of the lip and/or mouth can cause problems with breathing, nursing, eating, drinking, speech and hearing when it is not corrected. When surgery is an option it is typically done before the child is a few months old, and is recommended before one year for a cleft lip and 18 months for a cleft palette. Some children require repeated surgeries as they grow older. This can affect the lip and mouth. Like all facial surgeries on the skin’s exterior, cleft lip surgery leaves a visible scar. It is typically visible between the upper lip and bottom of the nose.

New scars tend to have a red appearance, since more blood vessels are produced when the body is trying to heal itself. Some scars are more visible than others. You will see the scar fading over time, eventually creating a pale line. Visibility is dependent on the child’s skin type, the cleft size, the body’s ability to heal itself and the placement of the sutures. After the wound has had a chance to heal for at least two to three weeks, a topical agent to treat scars can be applied.

There are a number of treatment options available for cleft lip scars. One option is to perform scar reduction surgery to make the scar thinner and flatter. Although it may become fairly thin, it will
still be visible without further scar treatment. Some people apply Vitamin E for faster healing; others use sunscreen to reduce permanent discoloration from ultraviolet rays.

Using Scarfade is also an option to further reduce cleft palette scars. It is safe to use on children of all ages. Best results occur when Scarfade is applied to scars less than a year old, since children’s scars tend to reach full maturity after 12 to 18 months in duration.

Scarfade C accelerates scar healing and is another option. Sunscreen can be applied over Scarfade for added protection from skin discoloration without lowering its effectiveness.

**SCAR TREATMENT**

When the skin is in the process of recovering from an injury, whether the result of an accident, surgery, a burn, or acne, scarring will occur wherever multiple layers of the skin have been affected. Once a scar forms, it is permanent but may be made less visible or relocated surgically.

Different scars require different treatments. A few common scars include:

- **Burn scars.** Severe burns that destroy large sections of skin cause the skin to heal in a puckered way. As the skin heals, muscles and tendons may be affected in this “contracting” movement.

- **Keloid scars** are a result of the skin’s overproduction of collagen after a wound has healed. These scars generally appear as growths in the scar site.

- **Hypertrophic scars,** unlike keloids, do not grow out of the boundaries of the scar area, but because of their thick, raised texture, can be unsightly and may also restrict the natural movement of muscles and tendons.

- **Facial scars** can be unattractive simply because of where they appear on the face, while others affect facial expressions.
All surgical possibilities will be discussed in the initial consultation along with risks involved for each type of scarring.

**Is Facial Scar Treatment for You?**

The goal of facial scar treatment is to disguise the scar, relocate it, or minimize its prominence.

Important factors to be discussed with your surgeon include:
- Skin type and color
- Ethnic background
- Individual healing rates
- Age.

Different types of scars respond to different plastic surgery techniques. A person considering facial scar revision must understand that there is no way to remove scars completely.

Timing of surgery is another important choice. Some surgeons advise against any scar revision in cases of injury for a period that might extend up to a year after the injury. This interval allows the body enough time to heal fully.

**Making the Decision for Scar Treatment**

What you should expect:
1. Your surgeon will examine the scar in order to decide upon the proper treatment and inform you of outcomes that can be expected from facial scar revision surgery.
2. The agreement between you and your surgeon on how to proceed is a prerequisite for successful surgery. After you both decide to proceed with scar revision, your surgeon will inform you about:
   a. Anesthesia
   b. Surgical facility
   c. Supportive surgery options
   d. Costs

Because scars are highly individualistic and the patient’s attitude toward scars is so personal, maximum improvement in
facial scars may require more than one procedure. More than one technique may be employed.

**Understanding Scar Revision Surgery**

Following is a general description of the surgery. Remember, each patient’s individual needs and features are considered before surgery.

1. When a scar is of the contracture type, surgery generally involves removing the scar tissue entirely.
2. Skin flaps, composed of adjacent healthy, unscarred skin, are then lifted and moved to form a new incision line.
3. Where a flap is not possible, a skin graft may be used. A graft involves taking a section of skin tissue from one area and attaching it to another. Time must be allowed following surgery for new blood vessels and soft tissue to form.
4. Z-plasty is a method to move a scar from one area to another, usually into a natural fold or crease in the skin to minimize its visibility. While Z-plasty does not remove all signs of a scar, it does make it less noticeable.

Dermabrasion and laser resurfacing are methods a surgeon uses to make “rough or elevated” scars less prominent by removing part of the upper layers of skin with an abrading tool or laser light. Clearly, the scar will remain, but it will be smoother and less visible.

Keloid or hypertrophic scars are often treated first with injections of steroids to reduce size. If this is not satisfactory, the scars can be removed surgically, and the incisions closed with fine stitches, often resulting in less prominent scars.

**What to Expect After the Surgery**

Note: These are general guidelines. Please ask your doctor to fully explain what your expectations should be post-surgery.

1. You can expect to feel some discomfort after facial scar revision surgery. Some swelling, bruising and redness are generally unavoidable.
2. It is important for you to follow your surgeon’s after care recommendations to the letter. Though the sutures will be removed within days after the surgery, your skin needs time to heal.

3. Surgeons generally insist on decreased activity after surgery and instruct the patient to keep the head elevated when lying down, to use cold compresses to reduce swelling, and to avoid any activity that places undue stress on the area of the incision.

4. Depending on the surgery performed and the site of the scar, the facial plastic surgeon will explain the types of activities to avoid.

5. No medication should be taken without first consulting the surgeon. It is important to remember that scar tissues require a year or more to fully heal and achieve maximum improved appearance.

**Follow-up Care**

Follow-up care is vital for this procedure to monitor healing. Obviously, anything unusual should be reported to your surgeon immediately. It is essential that you keep your follow-up appointments with your surgeon. Read more about what you should do before and after your scar treatment.

**SCAR REVISION SURGERY**

Scars can be the result of trauma, prior surgery, or even a childhood injury. While no scar can be removed completely, plastic surgery can often improve the appearance of a scar by making it less obvious. How much the appearance of a scar bothers you is a personal matter. Dr. Hughes will examine the scar and discuss at length the treatment choices including the risk and benefit of each option. Whether it’s revision surgery, steroid injections, laser treatments or dermabrasion, many scars can be improved dramatically with minimal downtime. If you are bothered by a scar, your first step should be to consult with a doctor regarding...
treatment options. Scars can be the result from an injury or a previous surgical procedure. Proper facial plastic surgical principles will minimize scarring however healing, genetics, and certain medical conditions can result in less than optimal scars. Certain steps can be taken to improve bad scars and include silicone sheeting therapy, steroid injections, dermabrasion, and laser treatments. However when these techniques are not enough surgical scar revision is required. Dr. Hughes has expert training in scar revision surgery. The latest techniques to improve scars include geometric broken line closures (GBLC), W-plasty, and Z-plasty. If you have a scar you are unhappy with and are considering revision surgery a consultation with Dr. Hughes will outline all of the available options and customize a treatment program that is best suited for you.

When is the best time to undergo scar revision?

Often patients are told to wait at least one year before considering scar revision. In many cases this may be good advice however in other cases this may not. Depending on the type of scar you have, early intervention can in some cases stop poor scar formation before it even occurs. Certain scars may improve dramatically with time however other types of scars may never get better no matter how long one waits. Steps which can be taken while in the ‘waiting period’ include; silicone sheeting therapy, steroid injections, and laser treatments. Depending on the type of scar, it may dictate when intervention is best taken. Many scars that appear unattractive at first may become less noticeable with time. Dr. Hughes will outline all of the available options and customize a treatment program that is best suited for you.

What is a hypertrophic scar?

A hypertrophic scar is often confused with a keloid scar. Both tend to be thick, raised and red. However hypertrophic scars remain within the boundaries of the original scar and typically will improve with time. Treatment options include waiting, steroid injections, silicone sheeting, laser treatments or scar revision
surgery. Depending on the size and location of the scar, surgery may be performed in the office with local anesthesia or in the operating room under sedation anesthesia.

**What is a keloid scar?**

Keloids are thick, hard, raised scars that tend to itch and in some cases hurt. The skin color can be red or darkened causing the appearance of the scar to be worse. Keloids can develop soon after surgery or in many cases months after the initial incident. Keloids can occur anywhere on the body but are more common on earlobes, shoulders and chest. They occur more commonly in darker skinned people, but can occur in any skin type. Keloids will grow outside the boundaries of the original scar, almost like a tumor.

Keloids are often treated with pressure, silicone sheeting and steroid injections. More radical treatments include surgery and in some cases radiation therapy. Keloids are stubborn and can recur, in some cases even larger than before. If you have a keloid and undergo treatment, this may require close observation for long periods of time so that if the keloid does recur it is treated properly early so that scarring is minimal.

**What is a Z-plasty?**

A Z-plasty is a type of scar revision surgery in which the direction of a scar is changed in order to camouflage the scar or diminish a scar contracture. A scar contracture is a scar that is pulled in an unusual way causing an unnatural appearance to the scar. Contractures are often seen with burns or traumatic accidents, but can in some cases be the result of poor wound healing after surgery.

A Z-plasty gives the scar a Z pattern appearance which in many cases can diminish the appearance of a straight line scar. Other types of procedures that are used in scar camouflage surgery include W-plasty, running W-plasty, multiple Z-plasties or Geometric broken line closure (GBLC).
What is a skin flap or graft?

In some cases the entire scar needs to be surgically removed. By removing the scar the surgeon then creates a new scar which need to be covered with normal skin. This can be accomplished by stretching nearby normal skin, a method known as a skin flap. Or the scar can be covered with new skin from another location, much like a patch, which is called a skin graft. Skin grafts are typically taken from more obscure locations so that the donor site scar is minimilized. Areas such as behind the ear or under the arm are often used. Skin graft donor sites are chosen based to best match the skin and are determined based on size, color, and location of the scar.

What is Scar Revision?

Surgical scar revision is an option available to patients faced with scars originating from injury or through surgery. Depending on the severity of the scar, revision of the scarred tissue may aid in the restoration of both form and function.

Who is a Good Candidate for Scar Revision?

First and foremost, you must be in good health, have no active diseases or serious, pre-existing medical conditions, and you must have realistic expectations of the outcome of your surgery. Whether caused by injury, surgery, or burn, scars can be disfiguring. Scars stand out against the rest of the skin because scar tissue is made of collagen cells rather than ordinary skin cells. As a result, scars are usually a different color, and do not have sweat glands or hair follicles. The severity of a scar depends on many factors, including the size and depth of the wound, the blood supply to the area, and the thickness and color of the skin. Some people - especially those with deeper skin tones - have a tendency to produce prominent, raised scars called keloids.

Examples of scars most commonly treated include: hypertrophic scars, keloid scars, wide scars and contracture scarring. Hypertrophic scars occur within the boundaries of the
incision or wound and are generally thick, red, and raised in appearance. Keloid scars are somewhat similar in appearance to hypertrophic scars. Although, keloids generally grow beyond the boundaries of the incision or wound. Keloids are commonly found on the earlobe, shoulder, and over the breastbone and appear most commonly in dark-skinned individuals. Spread scars are widened and the skin is thinned. Contracture scars are the most severe forms of a scar and usually occur as a result of a loss of a large area of skin. Contracture scars are most commonly found in patients that have experienced burn injuries. In this instance, the scars that have formed cause the edges of skin to pull together affecting the adjacent muscles and tendons, which in turn, causes the restriction of normal movement. Most minor scars can be treated by injecting a steroid medication directly into the scarred tissue. This form of treatment generally results in a reduction of redness, and size.

How is the Procedure Performed?

Based on the initial consultation, your surgeon will determine which procedure is best for you. Here are some possible options:

**Steroid Applications and Injections**

Steroids can help flatten and reduce the redness of hypertrophic and keloid scars. Steroids are applied or injected into the scar to break down the skin’s collagen. This is especially effective on hypertrophic and keloid scars, both of which continue to form collagen after the wound has healed. These injections can also help reduce the itching and/or pain associated with these scars.

**Silicone Gel Sheets**

Silicone gel sheets can help flatten hypertrophic and keloid scars. These clear sheets are placed on the scar and worn 24 hours a day.

**Z-Plasty and Related Tissue-Rearrangement Techniques**

Z-Plasty is a technique used to re-orient scars. The scar is oriented by cutting the skin around the scar in small triangular
flaps. These flaps usually follow a Z-shape (hence the name), but the technique your doctor chooses will depend on the shape of the scar.

The flaps are repositioned to follow natural lines and creases of the skin. The new scar is often less noticeable. Z-plasty can also help relieve the pressure of contracture scars.

**Dermabrasion**

Dermabrasion smoothes out surface irregularities such as deep lines or scars by removing the topmost layers of the skin. The afflicted area will be injected with anesthetic and then carefully "sanded" with a rotating wire brush or a diamond wheel until the desired amount of skin is removed.

**Vascular Laser**

A vascular laser works by shrinking the blood vessels that feed the scar, thus improving the coloration of red scars. This treatment is done without anesthetic. During the process, the sensation is much like that of a rubber band snapping on the skin.

**Tissue Expansion**

In this procedure, a “balloon” is inserted under a patch of healthy skin near a scar. The balloon is filled with a saline solution to stretch the skin. When the skin has been adequately stretched, which can take several weeks or months, the balloon is removed. The scar is then surgically removed, and the balloon-stretched skin is pulled over the previously scarred area and carefully closed.

**Skin Grafts**

In this procedure, doctors take skin from a healthy part of the body and transplant it to the injured area. Grafts aren’t always cosmetically pleasing because the grafted skin may not match the surrounding skin’s color or texture. The area where the graft came from will also scar — but skin grafts can greatly restore function to a severely scarred area.
Collagen Injections

Collagen Injections are used to rise, or fill in, sunken scars. Collagen is a natural animal protein. Before using collagen, you should take an allergy test to ensure that you are not allergic to the substance. The results of collagen injections are immediate but not permanent. The scars will eventually have to be re-filled as the body slowly absorbs the collagen.

Laser Skin Resurfacing

There are two types of lasers used for reducing the uneven surface of scars: the CO2 Laser and the Erbium: YAG Laser. The CO2 laser is typically used for deeper scars, while the Erbium is used for superficial scars and deeper skin tones. Both lasers remove the topmost layers of skin, allowing new, smooth skin to form. There are some color lasers that can be effectively treat the abnormal red pigmentation of hypertrophic and keloid scars as well.
Cleft Lip and Cleft Palate

Cleft lip and cleft palate, also known as orofacial cleft, is a group of conditions that includes cleft lip (CL), cleft palate (CP), and both together (CLP). A cleft lip contains an opening in the upper lip that may extend into the nose. The opening may be on one side, both sides, or in the middle. A cleft palate is when the roof of the mouth contains an opening into the nose. These disorders can result in feeding problems, speech problems, hearing problems, and frequent ear infections. Less than half the time the condition is associated with other disorders.

Cleft lip and palate are the result of tissues of the face not joining properly during development. As such, they are a type of birth defect. The cause is unknown in most cases. Risk factors include smoking during pregnancy, diabetes, obesity, an older mother, and certain medications (such as some used to treat seizures). Cleft lip and cleft palate can often be diagnosed during pregnancy with an ultrasound exam.

A cleft lip or palate can be successfully treated with surgery. This is often done in the first few months of life for cleft lip and before eighteen months for cleft palate. Speech therapy and dental care may also be needed. With appropriate treatment outcomes are good.

Cleft lip and palate occurs in about 1 to 2 per 1000 births in the developed world. CL is about twice as common in males as
females, while CP without CL is more common in females. In 2013 it resulted in about 3,300 deaths globally down from 7,600 deaths in 1990. The condition was formerly known as a hare-lip because of its resemblance to a hare or rabbit, but that term is now generally considered to be offensive.

**Signs and symptoms**

If the cleft does not affect the palate structure of the mouth it is referred to as cleft lip. Cleft lip is formed in the top of the lip as either a small gap or an indentation in the lip (partial or incomplete cleft) or it continues into the nose (complete cleft). Lip cleft can occur as a one sided (unilateral) or two sided (bilateral). It is due to the failure of fusion of the maxillary and medial nasal processes (formation of the primary palate).

**Cleft palate**

Cleft palate is a condition in which the two plates of the skull that form the hard palate (roof of the mouth) are not completely joined. The soft palate is in these cases cleft as well. In most cases, cleft lip is also present. Cleft palate occurs in about one in 700 live births worldwide.

Palate cleft can occur as complete (soft and hard palate, possibly including a gap in the jaw) or incomplete (a 'hole' in the roof of the mouth, usually as a cleft soft palate). When cleft palate occurs, the uvula is usually split. It occurs due to the failure of fusion of the lateral palatine processes, the nasal septum, and/or the median palatine processes (formation of the secondary palate).

The hole in the roof of the mouth caused by a cleft connects the mouth directly to the inside of the nose.

A result of an open connection between the mouth and inside the nose is called velopharyngeal inadequacy (VPI). Because of the gap, air leaks into the nasal cavity resulting in a hypernasal voice resonance and nasal emissions while talking. Secondary effects of VPI include speech articulation errors (e.g., distortions, substitutions, and omissions) and compensatory
misarticulations and mispronunciations (e.g., glottal stops and posterior nasal fricatives). Possible treatment options include speech therapy, prosthetics, augmentation of the posterior pharyngeal wall, lengthening of the palate, and surgical procedures.

Submucous cleft palate (SMCP) can also occur, which is a cleft of the soft palate with a classic clinical triad of a bifid, or split, uvula which is found dangling in the back of the throat, a furrow along the midline of the soft palate, and a notch in the back margin of the hard palate.

Psychosocial issues

Most children who have their clefts repaired early enough are able to have a happy youth and social life. Having a cleft palate/lip does not inevitably lead to a psychosocial problem. However, adolescents with cleft palate/lip are at an elevated risk for developing psychosocial problems especially those relating to self-concept, peer relationships and appearance. Adolescents may face psychosocial challenges but can find professional help if problems arise. A cleft palate/lip may impact an individual’s self-esteem, social skills and behavior. There is research dedicated to the psychosocial development of individuals with cleft palate. Self-concept may be adversely affected by the presence of a cleft lip and/or cleft palate, particularly among girls.

Research has shown that during the early preschool years (ages 3–5), children with cleft lip and/or cleft palate tend to have a self-concept that is similar to their peers without a cleft. However, as they grow older and their social interactions increase, children with clefts tend to report more dissatisfaction with peer relationships and higher levels of social anxiety. Experts conclude that this is probably due to the associated stigma of visible deformities and possible speech impediments. Children who are judged as attractive tend to be perceived as more intelligent, exhibit more positive social behaviors, and are treated more positively than children with cleft lip and/or cleft palate. Children with clefts tend to report feelings of anger, sadness, fear, and
alienation from their peers, but these children were similar to their peers in regard to “how well they liked themselves.”

The relationship between parental attitudes and a child’s self-concept is crucial during the preschool years. It has been reported that elevated stress levels in mothers correlated with reduced social skills in their children. Strong parent support networks may help to prevent the development of negative self-concept in children with cleft palate.

In the later preschool and early elementary years, the development of social skills is no longer only impacted by parental attitudes but is beginning to be shaped by their peers. A cleft lip and/or cleft palate may affect the behavior of preschoolers. Experts suggest that parents discuss with their children ways to handle negative social situations related to their cleft lip and/or cleft palate. A child who is entering school should learn the proper (and age-appropriate) terms related to the cleft. The ability to confidently explain the condition to others may limit feelings of awkwardness and embarrassment and reduce negative social experiences.

As children reach adolescence, the period of time between age 13 and 19, the dynamics of the parent-child relationship change as peer groups are now the focus of attention. An adolescent with cleft lip and/or cleft palate will deal with the typical challenges faced by most of their peers including issues related to self-esteem, dating and social acceptance. Adolescents, however, view appearance as the most important characteristic above intelligence and humor. This being the case, adolescents are susceptible to additional problems because they cannot hide their facial differences from their peers. Adolescent boys typically deal with issues relating to withdrawal, attention, thought, and internalizing problems and may possibly develop anxiousness-depression and aggressive behaviors. Adolescent girls are more likely to develop problems relating to self-concept and appearance. Individuals with cleft lip and/or cleft palate often deal with threats to their quality of life for multiple reasons including: unsuccessful
social relationships, deviance in social appearance and multiple surgeries.

Complications

A baby being fed using a customized bottle. The upright sitting position allows gravity to help the baby swallow the milk more easily.

Cleft may cause problems with feeding, ear disease, speech and socialization. Due to lack of suction, an infant with a cleft may have trouble feeding. An infant with a cleft palate will have greater success feeding in a more upright position. Gravity will help prevent milk from coming through the baby’s nose if he/she has cleft palate. Gravity feeding can be accomplished by using specialized equipment, such as the Haberman Feeder, or by using a combination of nipples and bottle inserts like the one shown, is commonly used with other infants. A large hole, crosscut, or slit in the nipple, a protruding nipple and rhythmically squeezing the bottle insert can result in controllable flow to the infant without the stigma caused by specialized equipment.

Individuals with cleft also face many middle ear infections which may eventually lead to hearing loss. The Eustachian tubes and external ear canals may be angled or tortuous, leading to food or other contamination of a part of the body that is normally self-cleaning. Hearing is related to learning to speak. Babies with palatal clefts may have compromised hearing and therefore, if the baby cannot hear, it cannot try to mimic the sounds of speech. Thus, even before expressive language acquisition, the baby with the cleft palate is at risk for receptive language acquisition. Because the lips and palate are both used in pronunciation, individuals with cleft usually need the aid of a speech therapist.

CAUSE

The development of the face is coordinated by complex morphogenetic events and rapid proliferative expansion, and is thus highly susceptible to environmental and genetic factors,
rationalising the high incidence of facial malformations. During the first six to eight weeks of pregnancy, the shape of the embryo’s head is formed. Five primitive tissue lobes grow:

a) one from the top of the head down towards the future upper lip; (Frontonasal Prominence)

b-c) two from the cheeks, which meet the first lobe to form the upper lip; (Maxillar Prominence)
d-e) and just below, two additional lobes grow from each side, which form the chin and lower lip; (Mandibular Prominence)

If these tissues fail to meet, a gap appears where the tissues should have joined (fused). This may happen in any single joining site, or simultaneously in several or all of them. The resulting birth defect reflects the locations and severity of individual fusion failures (e.g., from a small lip or palate fissure up to a completely malformed face).

The upper lip is formed earlier than the palate, from the first three lobes named a to c above. Formation of the palate is the last step in joining the five embryonic facial lobes, and involves the back portions of the lobes b and c. These back portions are called palatal shelves, which grow towards each other until they fuse in the middle. This process is very vulnerable to multiple toxic substances, environmental pollutants, and nutritional imbalance. The biologic mechanisms of mutual recognition of the two cabinets, and the way they are glued together, are quite complex and obscure despite intensive scientific research.

Genetics

Genetic factors contributing to cleft lip and cleft palate formation have been identified for some syndromic cases, but knowledge about genetic factors that contribute to the more common isolated cases of cleft lip/palate is still patchy.

Many clefts run in families, even though in some cases there does not seem to be an identifiable syndrome present, possibly
because of the current incomplete genetic understanding of midfacial development.

A number of genes are involved including cleft lip and palate transmembrane protein 1 and GAD1, one of the glutamate decarboxylases. Many genes are known to play a role in craniofacial development and are being studied through the FaceBase initiative for their part in clefting. These genes are AXIN2, BMP4, FGFR1, FGFR2, FOXE1, IRF6, MAFB (gene), MMP3, MSX1, MSX2 (Msh homeobox 2), MSX3, PAX7, PDGFC, PTCH1, SATB2, SOX9, SUMO1 (Small ubiquitin-related modifier 1), TBX22, TCOF (Treacle protein), TFAP2A, VAX1, TP63, ARHGAP29, NOG, NTN1, WNT genes, and locus 8q24.

Syndromes

• The Van der Woude Syndrome is caused by a specific variation in the gene IRF6 that increases the occurrence of these deformities threefold.

• Another syndrome, Siderius X-linked mental retardation, is caused by mutations in the PHF8 gene (OMIM 300263); in addition to cleft lip and/or palate, symptoms include facial dysmorphism and mild mental retardation.

In some cases, cleft palate is caused by syndromes which also cause other problems.

• Stickler’s Syndrome can cause cleft lip and palate, joint pain, and myopia.
• Loey-Dietz syndrome can cause cleft palate or bifid uvula, hypertelorism, and aortic aneurysm.
• Hardikar syndrome can cause cleft lip and palate, Hydronephrosis, Intestinal obstruction and other symptoms.
• Cleft lip/palate may be present in many different chromosome disorders including Patau Syndrome (trisomy 13).
• Malpuech facial clefting syndrome
Cleft Lip and Cleft Palate

- Hearing loss with craniofacial syndromes
- Popliteal pterygium syndrome
- Treacher Collins Syndrome

Specific genes

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Many genes associated with syndromic cases of cleft lip/palate have been identified to contribute to the incidence of isolated cases of cleft lip/palate. This includes in particular sequence variants in the genes IRF6, PVRL1 and MSX1. The understanding of the genetic complexities involved in the morphogenesis of the midface, including molecular and cellular processes, has been greatly aided by research on animal models, including of the genes BMP4, SHH, SHOX2, FGF10 and MSX1.

Environmental factors

Environmental influences may also cause, or interact with genetics to produce, orofacial clefting. An example of how environmental factors might be linked to genetics comes from research on mutations in the gene PHF8 that cause cleft lip/palate. It was found that PHF8 encodes for a histone lysine demethylase, and is involved in epigenetic regulation. The catalytic
activity of PHF8 depends on molecular oxygen, a fact considered important with respect to reports on increased incidence of cleft lip/palate in mice that have been exposed to hypoxia early during pregnancy. In humans, fetal cleft lip and other congenital abnormalities have also been linked to maternal hypoxia, as caused by e.g. maternal smoking, maternal alcohol abuse or some forms of maternal hypertension treatment. Other environmental factors that have been studied include: seasonal causes (such as pesticide exposure); maternal diet and vitamin intake; retinoids — which are members of the vitamin A family; anticonvulsant drugs; nitrate compounds; organic solvents; parental exposure to lead; alcohol; cigarette use; and a number of other psychoactive drugs (e.g. cocaine, crack cocaine, heroin).

Current research continues to investigate the extent to which folic acid can reduce the incidence of clefting.

DIAGNOSIS

Traditionally, the diagnosis is made at the time of birth by physical examination. Recent advances in prenatal diagnosis have allowed obstetricians to diagnose facial clefts in utero with ultrasonography.

Clefts can also affect other parts of the face, such as the eyes, ears, nose, cheeks, and forehead. In 1976, Paul Tessier described fifteen lines of cleft. Most of these craniofacial clefts are even rarer and are frequently described as Tessier clefts using the numerical locator devised by Tessier.

TREATMENT

Cleft lip and palate is very treatable; however, the kind of treatment depends on the type and severity of the cleft.

Most children with a form of clefting are monitored by a cleft palate team or craniofacial team through young adulthood. Care can be lifelong. Treatment procedures can vary between craniofacial teams. For example, some teams wait on jaw correction until the child is aged 10
to 12 (argument: growth is less influential as deciduous teeth are replaced by permanent teeth, thus saving the child from repeated corrective surgeries), while other teams correct the jaw earlier (argument: less speech therapy is needed than at a later age when speech therapy becomes harder). Within teams, treatment can differ between individual cases depending on the type and severity of the cleft.

Cleft lip

Within the first 2–3 months after birth, surgery is performed to close the cleft lip. While surgery to repair a cleft lip can be performed soon after birth, often the preferred age is at approximately 10 weeks of age, following the “rule of 10s” coined by surgeons Wilhelmmesen and Musgrave in 1969 (the child is at least 10 weeks of age; weighs at least 10 pounds, and has at least 10g hemoglobin). If the cleft is bilateral and extensive, two surgeries may be required to close the cleft, one side first, and the second side a few weeks later. The most common procedure to repair a cleft lip is the Millard procedure pioneered by Ralph Millard. Millard performed the first procedure at a Mobile Army Surgical Hospital (MASH) unit in Korea.

Often an incomplete cleft lip requires the same surgery as complete cleft. This is done for two reasons. Firstly the group of muscles required to purse the lips run through the upper lip. In order to restore the complete group a full incision must be made. Secondly, to create a less obvious scar the surgeon tries to line up the scar with the natural lines in the upper lip (such as the edges of the philtrum) and tuck away stitches as far up the nose as possible. Incomplete cleft gives the surgeon more tissue to work with, creating a more supple and natural-looking upper lip.

Pre-surgical devices

In some cases of a severe bi-lateral complete cleft, the premaxillary segment will be protruded far outside the mouth. Nasoalveolar molding prior to surgery can improve long-term
nasal symmetry among patients with complete unilateral cleft lip-cleft palate patients compared to correction by surgery alone, according to a retrospective cohort study. In this study, significant improvements in nasal symmetry were observed in multiple areas including measurements of the projected length of the nasal ala (lateral surface of the external nose), position of the superoinferior alar groove, position of the mediolateral nasal dome, and nasal bridge deviation. “The nasal ala projection length demonstrated an average ratio of 93.0 percent in the surgery-alone group and 96.5 percent in the nasoalveolar molding group” this study concluded.

**Cleft palate**

A repaired cleft palate on a 64-year-old female.

Often a cleft palate is temporarily covered by a palatal obturator (a prosthetic device made to fit the roof of the mouth covering the gap). Cleft palate can also be corrected by surgery, usually performed between 6 and 12 months. Approximately 20–25% only require one palatal surgery to achieve a competent velopharyngeal valve capable of producing normal, nonhypernasal speech. However, combinations of surgical methods and repeated surgeries are often necessary as the child grows. One of the new innovations of cleft lip and cleft palate repair is the Latham appliance. The Latham is surgically inserted by use of pins during the child’s 4th or 5th month. After it is in place, the doctor, or parents, turn a screw daily to bring the cleft together to assist with future lip and/or palate repair.

If the cleft extends into the maxillary alveolar ridge, the gap is usually corrected by filling the gap with bone tissue. The bone tissue can be acquired from the patients own chin, rib or hip.

**Speech and hearing**

A tympanostomy tube is often inserted into the eardrum to aerate the middle ear. This is often beneficial for the hearing ability of the child.
Children with cleft palate typically have a variety of speech problems. Some speech problems result directly from anatomical differences such as velopharyngeal inadequacy. Velopharyngeal inadequacy refers to the inability of the soft palate to close the opening from the throat to the nasal cavity, which is necessary for many speech sounds, such as /p/, /b/, /t/, /d/, /s/, /z/, etc. This type of errors typically resolve after palate repair.

However, sometimes children with cleft palate also have speech errors which develop as the result of an attempt to compensate for the inability to produce the target phoneme. These are known as compensatory articulations. Compensatory articulations are usually sounds that are non-existent in normal English phonology, often do not resolve automatically after palatal repair, and make a child’s speech even more difficult to understand.

Speech-language pathology can be very beneficial to help resolve speech problems associated with cleft palate. In addition, research has indicated that children who receive early language intervention are less likely to develop compensatory error patterns later.

**Hearing loss**

Hearing impairment is particularly prevalent in children with cleft palate. The tensor muscle fibres that open the eustachian tubes lack an anchor to function effectively. In this situation, when the air in the middle ear is absorbed by the mucous membrane, the negative pressure is not compensated, which results in the secretion of fluid into the middle ear space from the mucous membrane. Children with this problem typically have a conductive hearing loss primarily caused by this middle ear effusion.

**Sample treatment schedule**

Note that each individual patient’s schedule is treated on a case-by-case basis and can vary per hospital. The colored squares indicate the average timeframe in which the indicated procedure occurs. In some cases this is usually one procedure (for example
lup repair) in other cases this is an ongoing therapy (for example speech therapy).

**Craniofacial team**

A craniofacial team is routinely used to treat this condition. The majority of hospitals still use craniofacial teams; yet others are making a shift towards dedicated cleft lip and palate programs. While craniofacial teams are widely knowledgeable about all aspects of craniofacial conditions, dedicated cleft lip and palate teams are able to dedicate many of their efforts to being on the cutting edge of new advances in cleft lip and palate care.

Many of the top pediatric hospitals are developing their own CLP clinics in order to provide patients with comprehensive multidisciplinary care from birth through adolescence. Allowing an entire team to care for a child throughout their cleft lip and palate treatment (which is ongoing) allows for the best outcomes in every aspect of a child’s care. While the individual approach can yield significant results, current trends indicate that team based care leads to better outcomes for CLP patients.

**Epidemiology**

Cleft lip and palate occurs in about 1 to 2 per 1000 births in the developed world. Rates for cleft lip with or without cleft palate and cleft palate alone varies within different ethnic groups.

The highest prevalence rates for (CL ± P) are reported for Native Americans and Asians. Africans have the lowest prevalence rates.

- Native Americans: 3.74/1000
- Japanese: 0.82/1000 to 3.36/1000
- Chinese: 1.45/1000 to 4.04/1000
- Caucasians: 1.43/1000 to 1.86/1000
- Latin Americans: 1.04/1000
- Africans: 0.18/1000 to 1.67/1000
Rate of occurrence of CPO is similar for Caucasians, Africans, North American natives, Japanese and Chinese. The trait is dominant. It caused about 4,000 deaths globally in 2010 down from 8,400 in 1990. Prevalence of “cleft uvula” has varied from .02% to 18.8% with the highest numbers found among Chippewa and Navajo and the lowest generally in Africans.

**SOCIETY AND CULTURE**

**Abortion controversy**

In some countries, cleft lip or palate deformities are considered reasons (either generally tolerated or officially sanctioned) to perform an abortion beyond the legal fetal age limit, even though the fetus is not in jeopardy of life or limb. Some human rights activists contend that this practice of what they refer to as “cosmetic murder” amounts to eugenics.

**Works of fiction**

The eponymous hero of J.M. Coetzee’s 1983 novel *Life & Times of Michael K* has a cleft lip. However, cleft lip is more often portrayed negatively in popular culture. Examples include Oddjob, the secondary villain of the James Bond novel *Goldfinger* by Ian Fleming (the film adaptation does not mention this but leaves it implied); the fanciful portrayal of Roman Emperor Commodus in the 2000 film *Gladiator*; and serial killer Francis Dolarhyde in the film *Red Dragon*.

In the 1920 novel *Growth of the Soil*, by Norwegian writer Knut Hamsun, Inger (wife of the main character) has an uncorrected cleft lip which puts heavy limitations on her life, even causing her to kill her own child, who is also born with a cleft lip. In contrast, the protagonist of the 1924 novel *Precious Bane*, by English writer Mary Webb, is a young woman living in 19th-century rural Shropshire who eventually comes to feel that her deformity is the source of her spiritual strength. The book was later adapted for television by both the BBC and ORTF in France.
Compassion for those with cleft palates has also been used as the theme of young adult novels such as *Words in the Dust* by Trent Reedy and *Whisper* by Christina Struyk-Bonn.

**OTHER ANIMALS**

Cleft lips and palates are occasionally seen in cattle and dogs, and rarely in goats, sheep, cats, horses, pandas and ferrets. Most commonly, the defect involves the lip, rhinarium, and premaxilla. Clefts of the hard and soft palate are sometimes seen with a cleft lip. The cause is usually hereditary. Brachycephalic dogs such as Boxers and Boston Terriers are most commonly affected. An inherited disorder with incomplete penetrance has also been suggested in Shih tzus, Swiss Sheepdogs, Bulldogs, and Pointers. In horses, it is a rare condition usually involving the caudal soft palate. In Charolais cattle, clefts are seen in combination with arthrogryposis, which is inherited as an autosomal recessive trait. It is also inherited as an autosomal recessive trait in Texel sheep. Other contributing factors may include maternal nutritional deficiencies, exposure in utero to viral infections, trauma, drugs, or chemicals, or ingestion of toxins by the mother, such as certain lupines by cattle during the second or third month of gestation. The use of corticosteroids during pregnancy in dogs and the ingestion of *Veratrum californicum* by pregnant sheep have also been associated with cleft formation.

Difficulty with nursing is the most common problem associated with clefts, but aspiration pneumonia, regurgitation, and malnutrition are often seen with cleft palate and is a common cause of death. Providing nutrition through a feeding tube is often necessary, but corrective surgery in dogs can be done by the age of twelve weeks. For cleft palate, there is a high rate of surgical failure resulting in repeated surgeries. Surgical techniques for cleft palate in dogs include prosthesis, mucosal flaps, and microvascular free flaps. Affected animals should not be bred due to the hereditary nature of this condition.
Orofacial clefts—including cleft lip (CL), cleft lip and palate (CLP), and cleft palate (CP) alone, as well as median, lateral (transversal), and oblique facial clefts—are among the most common congenital anomalies. Approximately 1 case of orofacial cleft occurs in every 500-550 births. The prevalence varies by ethnicity, country, and socioeconomic status. Nonsyndromic CLP, which forms the largest subgroup of craniofacial anomalies, occurs in the range of 1.5-2.5 cases per 1000 live births. In the United States, 20 infants are born with an orofacial cleft on an average day, or 7500 every year.

Children who have an orofacial cleft require several surgical procedures and multidisciplinary treatment and care; the conservative estimated lifetime medical cost for each child with an orofacial cleft is $100,000, amounting to $750 million for all children with orofacial cleft born each year in the United States. In addition, these children and their families often experience serious psychological problems.

With rapidly advancing knowledge in medical genetics and with new DNA diagnostic technologies, more cleft lip and palate anomalies are diagnosed prenatally and more orofacial clefts identified as syndromic. Although the basic rate of clefting (1:500
to 1:550) has not changed since Fogh-Andersen performed his pioneering 1942 genetic study distinguishing two basic categories of orofacial clefts—namely, CL with or without CP (CL/P) and CP alone—these clefts can now be more accurately classified.

The correct diagnosis of a cleft anomaly is fundamental for treatment, for further genetic and etiopathologic studies, and for preventive measures correctly targeting the category of preventable orofacial clefts.

**PATHOPHYSIOLOGY**

**Embryology**

In facial morphogenesis, neural crest cells migrate into the facial region, where they form the skeletal and connective tissue and all dental tissues except the enamel. Vascular endothelium and muscle are of mesodermal origin.

The upper lip is derived from medial nasal and maxillary processes. Failure of merging between the medial nasal and maxillary processes at 5 weeks' gestation, on one or both sides, results in cleft lip. CL usually occurs at the junction between the central and lateral parts of the upper lip on either side. The cleft may affect only the upper lip, or it may extend more deeply into the maxilla and the primary palate. (Cleft of the primary palate includes CL and cleft of the alveolus.) If the fusion of palatal shelves is impaired also, the CL is accompanied by CP, forming the CLP abnormality.

CP is a partial or total lack of fusion of palatal shelves. It can occur in numerous ways:

- Defective growth of palatal shelves
- Failure of the shelves to attain a horizontal position
- Lack of contact between shelves
- Rupture after fusion of shelves

The secondary palate develops from the right and left palatal processes. Fusion of palatal shelves begins at 8 weeks' gestation
and continues usually until 12 weeks’ gestation. One hypothesis is that a threshold is noted beyond which delayed movement of palatal shelves does not allow closure to take place, and this results in a CP.

**Classification**

The group of orofacial cleft anomalies is heterogeneous. It comprises typical orofacial clefts (eg, CL, CLP, and CP) and atypical clefts (eg, median, transversal, oblique, and other Tessier types of facial clefts). Typical and atypical clefts can both occur as an isolated anomaly, as part of a sequence of a primary defect, or as a multiple congenital anomaly (MCA). In an MCA, the cleft anomaly could be part of a known monogenic syndrome, part of a chromosomal aberration, part of an association, or part of a complex of MCA of unknown etiology.

**ETIOLOGY**

Most orofacial clefts, like most common congenital anomalies, are caused by the interaction between genetic and environmental factors. In those instances, genetic factors create a susceptibility for clefts. When environmental factors (ie, triggers) interact with a genetically susceptible genotype, a cleft develops during an early stage of development.

The proportion of environmental and genetic factors varies with the sex of the individual affected with cleft. In CL and CP, it also varies with the severity and the unilaterality or bilaterality of the cleft anomaly; the highest proportion of genetic factors are in the subgroup of females with a bilateral cleft, and the smallest proportion is in the subgroup of males with a unilateral cleft. Thus, the classic multifactorial threshold (MFT) model of liability can be applied to CL/P as the multifactorial model of liability with four different thresholds.

Theoretically, the subgroup of clefts closest to the population average should have the highest population prevalence, the lowest value of heritability, and thus the lowest risk of recurrence. This
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was confirmed in a large, population-based study of whites with clefts. The value of heritability expresses a ratio of genetic and nongenetic factors. Heritability is equal to 1 for conditions completely controlled by genetic factors and equal to 0 for conditions completely controlled by environmental factors.

A higher proportion of environmental factors indicates a lower risk of recurrence and also gives a better chance to act in prevention, because the only etiologic factors that can be changed are environmental factors. Thus, the subgroup whose average prevalence is closest to the population average represents males affected with a unilateral CL/P. This subgroup is most common among orofacial clefts; the risk of recurrence for siblings and for offspring of an individual with cleft is the lowest, the value of heritability is the lowest, and efficacy of primary prevention is the highest.

A cleft develops when embryonic parts called processes (which are programmed to grow, move, and join with each other to form an individual part of the embryo) do not reach each other in time and an open space (cleft) between them persists. In the normal situation, the processes grow into an open space by means of cellular migration and multiplication, touch each other, and fuse together. In general, any factor that could prevent the processes from reaching each other—for instance, by slowing down migration or multiplication of neural crest cells, by stopping tissue growth and development for a time, or by killing some cells that are already in that location—would cause a persistence of a cleft. Also, the epithelium that covers the mesenchyme may not undergo programmed cell death, so that fusion of processes cannot take place.

DNA studies

Considerable interest has developed in the identification of genes that contribute to the etiology of orofacial clefting. Advances in modern molecular biology, newer methods of genome manipulation, and availability of complete genome sequences led
to an understanding of the roles of particular genes that are associated with embryonic development of the orofacial complex.

The first candidate gene was transforming growth factor-á (TGFA), which showed an association with nonsyndromic CLP in a white population. Lidral et al investigated five different genes (TGFA, BCL3, DLX2, MSX1, TGFB3) in a largely white population from Iowa. They found a significant linkage disequilibrium between CL/P and both MSX1 and TGFB3 and between CP and MSX1. The TGFB3 gene was identified as a strong candidate for clefting in humans based on both the mouse model and the linkage disequilibrium studies.

Other candidate genes that show an association with nonsyndromic CLP include D4S192, RARA, MTHFR, RFC1, GABRB3, PVRL1, and IRF6.

MSX1 was found to be a strong candidate gene involved in orofacial clefts and dental anomalies. Analysis of the MSX1 sequence in a multiplex Dutch family showed that a nonsense mutation (Ser104stop) in exon 1 segregated with the phenotype of nonsyndromic cleft lip and palate. Some have proposed that cleft palate in MSX1 knock-out mice is due to insufficiency of the palatal mesenchyme.

Zucchero et al reported that variants of IRF6 may be responsible for 12% of nonsyndromic cleft lip and palate, suggesting that this gene would play a substantial role in the causation of orofacial clefts. A meta-analysis of all-genome scans of subjects with nonsyndromic cleft lip and palate, including Filipino, Chinese, Indian, and Colombian families, found a significant evidence of linkage to the region that contains interferon regulatory factor 6 (IRF6).

Also, gene-gene interactions have been examined. A complex interplay of several genes, each making a small contribution to the overall risk, may lead to formation of clefts. Jugessur et al reported a strong effect of the TGFA variant among children homozygous for the MSX1 A4 allele (9 CA repeats).
Evaluation of gene-environment interactions is still in a preliminary stage. Studies of the role of smoking in TGFA and MSX1 as covariates suggested that these loci might be susceptible to detrimental effects of maternal smoking. Folate-metabolizing enzymes such as methylenetetrahydrofolate reductase (MTHFR), which is a key player in etiology of neural tube defects, and RFC1 are considered candidate genes on the basis of data that suggest that folic acid supplementation can reduce incidence of nonsyndromic cleft lip and palate.

More than 30 potential candidate loci and candidate genes throughout the human genome have been identified as strong susceptibility genes for orofacial clefts. The MSX1 (4p16.1), TGFA (2p13), TGFB1 (19q13.1), TGFB2 (1q41), TGFB3 (14q24), RARA (17q12), and MTHFR (1p36.3) genes are among the strongest candidates.

The TGFB3 gene was identified as a strong candidate for clefting in humans based on a mouse model. Generally, palatogenesis in mice parallels that of humans and shows that comparable genes are involved. Kaartinen demonstrated that mice lacking the TGFB3 peptide exhibit cleft palate. In addition, the exogenous TGFB3 peptide can induce palatal fusion in chicken embryos, although the cleft palate is a normal feature in chickens.

In humans, association studies between the TGFB3 gene and nonsyndromic CL/P showed conflicting results. Lidral reported failure to observe an association of a new allelic variant of TGFB3 with nonsyndromic CL/P in a case-control study of the Philippines’ population. Another study by Tanabe analyzed DNA samples from 43 Japanese patients and compared results with those from 73 control subjects with respect to four candidate genes, including TGFB3. No significant differences in variants of TGFB3 between case and control populations were observed.

On the other hand, subsequent case-control association studies, family-based studies, and genome scans supported a role of TGFB3 in cleft development. Beaty examined markers in five candidate genes in 269 case-parent trios ascertained through a
child with nonsyndromic orofacial clefts; 85% of the probands in the study were white. Markers at two of the five candidate genes (\textit{TGFB3} and \textit{MSX1}) showed consistent evidence of linkage and disequilibrium due to linkage.

Similarly, Vieira attempted to detect transmission distortion of \textit{MSX1} and \textit{TGFB3} in 217 South American children from their respective mothers. A joint analysis of \textit{MSX1} and \textit{TGFB3} suggested a possible interaction between these two genes, increasing cleft susceptibility. These results suggest that \textit{MSX1} and \textit{TGFB3} mutations make a contribution to clefts in South American populations.

In a study of the Korean population, Kim reported that the G allele at the SfaN1 polymorphism of \textit{TGFB3} is associated with an increased risk of nonsyndromic CL/P. The population study consisted of 28 patients with nonsyndromic CL with or without CP and 41 healthy controls.

In 2004, Marazita performed a meta-analysis of 13 genome scans of 388 extended multiplex families with nonsyndromic CL/P. The families came from seven diverse populations including 2551 genotyped individuals. The meta-analysis revealed multiple genes in 6 chromosomal regions including the region containing \textit{TGFB3} (14q24).

In the Japanese population, blood samples from 20 families with nonsyndromic CL/P were analyzed by using \textit{TGFB3} CA repeat polymorphic marker. On the basis of the results of the study, the investigators concluded that either the \textit{TGFB3} gene itself or an adjacent DNA sequence may contribute to the development of cleft lip and palate.

A study by Ichikawa et al investigated the relationship between nonsyndromic CL/P and seven candidate genes (\textit{TGFB3}, \textit{DLX3}, \textit{PAX9}, \textit{CLPTM1}, \textit{TBX10}, \textit{PVRL1}, \textit{TBX22}) in a Japanese population. The sample consisted of 112 patients with their parents and 192 controls. Both population based case-control analysis and family based transmission disequilibrium test (TDT) were used.
The results showed significant associations of single nucleotide polymorphisms (SNPs) in TGFB3 and nonsyndromic CL/P, especially IVS+5321(rs2300607). Although IVS-1572 (rs2268625) alone did not show a significant difference between cases and controls, the haplotype “A/A” for rs2300607-rs2268625 showed significant association. The author concluded that the results demonstrated positive association of TGFB3 with nonsyndromic CL/P in Japanese patients.

A study by Bu et al found evidence of an association between nonsyndromic CLP and SNPs in FOXC2 (6p25.3).

Several micromanifestations of orofacial clefts have been studied, and additional candidate genes associated with these minimal, clinically less significant anomalies have been suggested. Associations of specific candidate genes with nonsyndromic CL/P have not been found consistent across different populations. This may suggest that multiplicative effects of several candidate genes or gene-environmental interactions are noted in different populations.

The identification of factors that contribute to the etiology of nonsyndromic CL/P is important for prevention, treatment planning, and education. With an increasing number of couples who seek genetic counseling as a part of their family planning, the knowledge of how specific genes contribute to formation of nonsyndromic CL/P has gained an increased importance.

EPIDEMIOLOGY

Reported data on the frequency of orofacial clefts vary according to the investigator and the country. In general, all typical orofacial cleft types combined occur in white populations with a frequency of 1 per 500-550 live births. Although the total combined frequency of CL, CLP, and CP is often used in statistics, combining the two etiologically different groups (ie, CL/P and CP alone) represents a misclassification bias similar to that of combining clefts with other congenital malformations.
The sex ratio in patients with clefts varies. In whites, cleft lip and cleft lip and palate occur significantly more often in males, and cleft palate occurs significantly more often in females. In CL/P, the sex ratio correlates with the severity and laterality of the cleft. A large study of 8952 orofacial clefts in whites found the male-to-female sex ratio to be 1.5-1.59:1 for CL, 1.98-2.07:1 for CLP, and 0.72-0.74:1 for CP. The prevalence of clefts varies considerably in different racial groups. The lowest rate is for blacks. A high prevalence of CL/P was found for the Japanese population, and the highest prevalence was found for the North American Indian populations. In contrast, no remarkable variation among races was found in isolated CP. In particular, its prevalence did not significantly vary between black and white infants or between infants of Japanese and European origin in Hawaii. Leck considered that such findings may reflect a higher etiologic heterogeneity of CP than of CL/P. Methods of ascertainment and classification criteria undoubtedly influence prevalence figures.

In a large population-based study of 4433 children born with orofacial cleft (ascertained from 2,509,881 California births), the birth prevalence of nonsyndromic CL/P was 0.77 per 1000 births (CL, 0.29/1000; CP, 0.48/1000), and the prevalence of nonsyndromic CP was 0.31 per 1000 births.

In that study, the risk of CL/P was slightly lower among the offspring of non-US-born Chinese women compared to US-born Chinese women and slightly higher among non-US-born Filipinos relative to their US-born counterparts. For CP, lower prevalences were observed among blacks and Hispanics than among whites. The risk of CP was higher among non-US-born Filipinos compared to US-born Filipinos. These prevalence variations may reflect differences in both environmental and genetic factors affecting risk for development of orofacial cleft.

Risk of recurrence

Genetic factors (ie, genes participating in the etiology of nonsyndromic orofacial clefts) are passed to the next generation,
thus creating an increased risk for such anomaly in offspring. The risk of recurrence also differs with respect to proportion of genetic and nongenetic factors. In CL/P, the hypothetical four-threshold model closely corresponds with differences in the risk of recurrence.

From a clinical point of view, the following two factors are most important in evaluating the risk of recurrence for CL/P:

- Sex of the individuals (ie, patient and individual at risk)
- Severity of the effect in the patient (eg, unilateral vs bilateral)

The lowest recurrence risk for CL/P is for the subcategory of male patients with unilateral cleft and, within this category, for sisters of males with a unilateral cleft and for daughters of fathers with a unilateral CL/P. The highest risk of recurrence of CL/P is for the subcategory of female patients affected with a bilateral CL/P.

CAUSES

There are many causes of cleft lip and palate. Problems with genes passed down from 1 or both parents, drugs, viruses, or other toxins can all cause these birth defects. Cleft lip and palate may occur along with other syndromes or birth defects.

A cleft lip and palate can:

- Affect the appearance of the face
- Lead to problems with feeding and speech
- Lead to ear infections

Babies are more likely to be born with a cleft lip and palate if they have a family history of these conditions or other birth defects.

SYMPTOMS

A child may have 1 or more birth defects. A cleft lip may be just a small notch in the lip. It may also be a complete split in the lip that goes all the way to the base of the nose. A cleft palate can
be on 1 or both sides of the roof of the mouth. It may go the full length of the palate.

Other symptoms include:

- Change in nose shape (how much the shape changes varies)
- Poorly aligned teeth

Problems that may be present because of a cleft lip or palate are:

- Failure to gain weight
- Feeding problems
- Flow of milk through nasal passages during feeding
- Poor growth
- Repeated ear infections
- Speech difficulties

Exams and Tests

A physical examination of the mouth, nose, and palate confirms a cleft lip or cleft palate. Medical tests may be done to rule out other possible health conditions.

Treatment

Surgery to close the cleft lip is often done when the child is between 6 weeks and 9 months old. Surgery may be needed later in life if the problem has a major effect on the nose area. A cleft palate is usually closed within the first year of life so that the child’s speech develops normally. Sometimes, a prosthetic device is temporarily used to close the palate so the baby can feed and grow until surgery can be done. Continued follow-up may be needed with speech therapists and orthodontists.

Outlook (Prognosis)

Most babies will heal without problems. How your child will look after healing depends on the severity of their condition. Your child might need another surgery to fix the scar from the surgery wound. Children who had a cleft palate repair may need to see
a dentist or orthodontist. Their teeth may need to be corrected as they come in. Hearing problems are common in children with cleft lip or palate. Your child should have a hearing test at an early age, and it should be repeated over time.

Your child may still have problems with speech after the surgery. This is caused by muscle problems in the palate. Speech therapy will help your child.

**When to Contact a Medical Professional**

Cleft lip and palate is usually diagnosed at birth. Follow your health care provider’s recommendations for follow-up visits. Call your provider if problems develop between visits.
Treatment for a cleft lip/cleft palate usually involves reconstructive surgery and several key support services. Here at Boston Children’s Hospital, our Cleft Lip and Palate Program uses a multidisciplinary team approach—not only to repair the child’s specific defect, but also to address any and all related complications. While your child’s treatment plan will depend on his individual circumstances—as well as your family’s preferences—here are the basics of treating cleft lip/cleft palate:

**SURGICAL REPAIR: CLEFT LIP**

What is a “lip-nasal adhesion” procedure? Does my child need one before his cleft lip repair? For some children with a unilateral cleft lip, the first operation they need is a lip-nasal adhesion. This operation is performed at about 3 months of age, and involves:

- a simple closure of the lip
- the first stage of nasal correction
- when possible, closure of the cleft in the upper gum (this procedure is called gingivoperiosteoplasty)
How and when is an incomplete cleft lip typically repaired?

An incomplete cleft lip, either unilateral or bilateral, is usually repaired with one operation when the child is between 3 and 5 months of age. During this operation, the baby’s nasal asymmetry (unevenness in the shape of the nose) is also corrected.

During the repair procedure, a plastic surgeon uses the existing muscle and tissues of the child’s lip and nose to close the cleft. Repair of a unilateral cleft lip is typically performed in the operating room under general anesthesia. The child will stay in the hospital for one to two nights after the operation.

How and when is a complete cleft lip typically repaired?

A child with a complete cleft lip typically needs two operations. The second operation is a more comprehensive repair of the cleft lip and correction of the nose. This procedure usually takes place at 5 to 6 months of age.

How will my child look after his cleft lip is repaired?

After the operation, your child’s lip, nose and face will be swollen for a few days. His scar may be red for several weeks. It will take 6 to 12 months for the scar to soften and fade.

Although it will never completely disappear, in time, the scar will become difficult to see. Your child’s lip and nose will be nearly normal in appearance after the swelling and scar have subsided.

As my child gets older, will he need another operation on his lip or nose?

Although some children need to have another procedure on their lips and/or nose before they begin school, or as they enter adolescence, other children never need further surgeries.

Children whose cleft lips involve the alveolus, or gum line, typically need another operation to help their permanent teeth come in and to make it easier for orthodontic treatments to improve
their bite and jaw function. This operation is called an alveolar bone graft, and is usually performed when the child is 8 to 10 years old.

**Surgical repair: Cleft palate**

*How is a cleft palate usually repaired?*

A plastic surgeon brings together the separated muscles and tissue from the two halves of the palate to close the opening. This procedure is performed in the operating room under general anesthesia. Your child will be in the hospital anywhere from one to three nights after the operation.

*When will my child’s cleft palate be repaired?*

A cleft palate is typically closed between the ages of 8 and 11 months, before a baby makes his first attempt to speak. Your child’s plastic surgeon will discuss the best repair plan for his needs and circumstances.

*What precautions do I need to take after my child’s cleft palate repair?*

You should give your child soft foods, using the side of a soft-tipped baby spoon. After each feeding, be sure to rinse your child’s mouth with water. Rinsing is very important, especially for the first 10 to 14 days after surgery. Ask your nurse about using a special syringe for rinsing.

*Will my child experience any side effects after the surgical repair?*

Your child may regurgitate some food and liquid through her nose for up to three months after the operation.

This is normal. It takes time for the swelling to diminish and for the muscles in the palate to begin working properly. You’ll be given further post-operative instructions when your child is discharged.
As my child gets older, will he need another operation on his palate?

Approximately 5 to 15 percent of all children who have undergone a cleft palate repair will need a second operation to correct “nasal”-sounding speech. The most common procedure used to fix this problem is called a pharyngeal flap, which is performed when the child is about 5 years old.

Managing feeding issues

How do I feed my newborn with a cleft lip/cleft palate?

Your baby’s ability to feed, whether by breast or bottle, is determined by the extent or severity of her cleft lip/cleft palate. Right after birth, your cleft team nurse will determine the type of feeding method that’s best for you and your baby.

- If your baby has a cleft palate that only involves her soft palate, a nipple shield might be helpful in assisting with breastfeeding.
- If your baby is working too hard to suckle, or if her cleft palate involves the hard palate, she may need a device such as a VentAire® feeder with a small cross cut in the silicone nipple.
- If your baby has both a cleft lip and a cleft palate, she will most likely need a special feeding device. A Haberman® feeder is usually recommended, because it allows milk to be pumped in time with the baby’s suck-swallow sequence. A Ross® nipple might be added to help deliver the flow of milk.

How much milk does my baby need?

Your cleft team nurse will help you determine the total volume of milk your infant needs to consume over a 24-hour period.

- Feedings should last no more than 30 minutes; prolonged feeding can exhaust you and the baby, and infants spend calories very quickly.
However, it’s important to feed your baby every 3 to 4 hours. Never let him go more than four hours without a feeding—unless he is close to meeting his volume quota for the 24-hour period.

Weigh your infant once per week. If he is not gaining more than one ounce per day, you should talk to your pediatrician about increasing the caloric content of the milk.

How do I keep my baby sucking sufficiently?

• Establishing a “rhythm” with your infant is paramount: Watch for your infant’s “hunger cues” and do not interrupt a sucking pattern.
• When the sucking stops, burp your infant while holding him upright and supporting his lower jaw.

Is there a positioning technique for bottle feeding?

Yes: Wrap your infant, enclosing the hands, in a blanket. This is called “swaddling.”

• Sit in a comfortable chair, like a rocking chair or a “glider” chair, with a footstool.
• Hold the baby upright in your arms or hold his head from behind.
• Relax your arms, and place the nipple gently into the baby’s mouth.
• Lay the nipple on top of the baby’s tongue.
• Rotate your arm so that the underside of your hand is holding the bottle. Put your ring finger under the baby’s chin.
• With firm pressure, keep your ring finger in place so as the baby suckles you feel pressure against your finger. Your infant should feel comfortable while suckling (i.e., no straining or squirming to access the nipple or to swallow).
• If your infant has both a cleft lip and a cleft palate, position the nipple so that his upper and lower gums connect with it.
• With gentle pressure under the chin, push up to start your baby’s sucking.
• Maintain this pressure. If, after a minute of sucking, there is little flow of milk, rotate the nipple to a longer line or compress the nipple with gentle pressure in rhythm with your baby’s suck-swallow reflex.
• Watch for cues that your infant is either satiated or needs burping (“bubbling”).

What should my baby and I do right after feeding?

• Keep the baby upright for about 20 minutes, either by holding him or by placing him in a seat.
• If you place your baby in a bed, slightly turn his body to the side with a wedge.
• Elevate the bed by 20 degrees: his chest should be higher than his stomach.
• An infant with a cleft palate may exhibit some esophageal and nasopharyngeal reflux (milk coming out through the nose), or he might regurgitate shortly after feeding has ended. You should always keep a suction bulb handy for these instances.
• Record the time, length and amount of feeding.

How can I adjust the feeding process if my baby is not gaining sufficient weight?

If your baby is gaining less than one ounce per day, calories in the formula or breast milk need to be increased. This can be accomplished by concentrating the formula or adding powdered milk to your breast milk. Your pediatrician and cleft team nurse will help you with these steps.

If a particular feeding device is not working for you or for your infant, your cleft team nurse will give you alternative feeders to try. If your baby’s weight gain is insufficient even after increasing the calories per volume, you may need to consult with a pediatric gastrointestinal/nutrition specialist.
Are there ways to supplement or replace oral feeding?

When oral feedings are not sufficient for your baby, there are other methods that can be used to deliver the needed calories:

- Your child’s treatment team may recommend a naso-gastric tube, which is passed through the baby’s nostril into his stomach to provide a direct source of nutrients. However, this is only a temporary solution until the baby is able to consume all necessary calories by mouth.

- If your baby is having trouble with both eating and breathing, your doctor may recommend a gastrostomy tube, which is placed directly into his stomach from the outside of the belly under general anesthesia. The food source is delivered to the stomach through a syringe or mechanical device. A gastronomy tube is normally used only until the child is old enough to eat by mouth.

How do I feed my child in preparation for her cleft palate repair?

Some—but not all—babies are introduced to cup feeding before surgical repair of their cleft palate. It will take several months for your baby to get used to cup feeding, so it’s good to start early (around 6 months of age).

- Begin by using the cup to replace one feeding a day, and gradually increase the number of cup feedings.

- Over several weeks, you should be able to completely transition to a cup.

- Begin spoon feedings, using a soft-tipped spoon, when your baby is 6 months old. As directed by your pediatrician, you may give her cereal, fruits, vegetables and other foods with the spoon. You may also use the spoon to feed her liquids.

How do I feed my child after her cleft palate repair?

Your cleft team nurse will review feeding instructions at the time of your baby’s cleft palate repair.
• In some instances, your child may return to using a cleft feeder. If the cleft feeder is not appropriate for your child, a “sippy cup” will be introduced prior to the surgery.

• Your child should use a cup that either has a very short spout, or doesn’t have a spout at all. Your nurse will show you the type of cup that’s best for your child.

• Remember that you will need to use a cup for all feedings during the first 10 to 14 days after your child’s cleft palate repair.

• If your child is having difficulty getting enough fluid with the cup, it is permissible to use a silicone nipple with a large cross cut. This can be accomplished by using the Haberman® bottle or a standard soft silicone nipple. It may instead be necessary to feed your child with the Ross® nipple.

Ten days after surgery, you can begin to give your child soft foods with the side of a soft-tipped baby spoon. After each feeding, be sure to rinse your child’s mouth with a small amount of water from a cup or special syringe.

How do I ensure that my baby is nurtured as well as nourished?

It’s important to remember that your infant is a normal baby who just happens to have an anatomic defect that can be surgically corrected. Once you feel comfortable with the feeding method your cleft team nurse teaches you, you will be more at ease with nurturing. You will see your baby thriving, smiling and responding to your touch.

It is very important that at least three people you trust with supporting and assisting you in feeding your baby learn and feel comfortable with the chosen feeding method, too. Your infant can sense when someone is confident with feedings, and will be more relaxed as a result.
MANAGING HEARING DIFFICULTIES

Will my child have difficulty hearing?

Many—but not all—children born with a cleft palate experience temporary hearing loss because of fluid in the middle ear and recurring ear infections. This hearing loss may last for a short time, or it can go on for a number of months.

Since speech and language development are influenced by a child’s ability to hear well, any hearing problems he has can also cause speech difficulties.

How and when will my child’s hearing be tested?

Your child will have his first hearing test early in life, and again prior to surgery to repair his cleft palate. His age and developmental level will determine which hearing test method will be used:

• Very young infants (newborn to 6 months) undergo a special hearing evaluation called an Auditory Brainstem Response Evaluation (ABRE). This test is performed while the child is asleep.
• Older infants (at a developmental level of 6 months or more) can undergo a hearing evaluation with behavioral audiometric test methods. This type of test is conducted while the child is awake and able to participate.
• Children between the ages of 7 months and 2 ½ years typically undergo a Visual Reinforcement Audiometry (VRA) hearing test.
• A child who is between 2 ½ and 5 years old should have a hearing test by conventional audiometric testing methods (also known as the “hand-raise response” test).
• Tympanometry testing is often performed at the time of hearing evaluation. This is a test of middle ear function, and can also be used to check the function of ear tubes and to detect the presence of an eardrum perforation.
Your child’s audiologist (hearing specialist) will assess his hearing every 6 to 12 months, and will work closely with an otorhinolaryngologist (ORL)—an ear/nose/throat specialist. Before his cleft palate repair, your child will be seen by the ORL specialist to discuss his ear function and any special care he may need.

**How is persistent middle ear fluid and associated hearing loss treated?**

Because the tube that connects the middle ear to the throat (called the Eustachian tube) does not drain normally in an infant with a cleft palate, fluid collects in the baby’s middle ear space. This fluid—also known as effusion—is present in virtually every baby with an unrepaired cleft palate who is younger than 1 year of age. The accompanying hearing loss can cause difficulties with speech, language and cognitive development as the child grows.

Persistent fluid in the middle ear is also associated with a recurring infection called otitis media. Infants with a cleft palate, middle ear effusion and hearing loss will require an operation to remove the fluid and to insert a ventilation tube. This operation is performed under general anesthesia, usually at the same time as the cleft palate repair. The ventilation tubes will stay in place for 9 to 12 months, and your child’s ORL specialist will check them every 6 months. The tubes usually fall out on their own, and do not require another surgery for removal.

As many as half of all infants who undergo cleft palate repair will need a repeat insertion of ventilation tubes. Although they unfortunately carry a risk of eardrum scarring and perforation, the tubes are necessary to ensure normal long-term hearing, and are crucial to healthy speech and language development.

**Managing speech/language issues**

**How does the palate affect speech?**

The hard and soft palate separate the mouth from the nose. When we breathe, the air flows in and out of our lungs through
the nose and throat. When we talk, the muscles in the soft palate move the palate to the back of the throat to seal off the nose (this is called the velopharyngeal valve). This sealing allows air to flow through the mouth alone when we speak; there are only three speech sounds in the English language (“m,” “n” and “ng”) that are made through the nose instead.

**Will my child experience difficulty speaking?**

A cleft lip alone does not cause speech problems. A child born with a cleft palate, however, is unable to make normal speech sounds—other than those three nasal sounds rendered through the nose—until the defect is repaired. Some children with a cleft palate experience both speech and language delays. These may be related to the temporary hearing loss associated with the cleft palate and middle ear fluid. Children may also exhibit difficulty with speech if their palate is not effectively closing off the nose from the mouth while they are speaking.

The good news is that most children will acquire speech and language skills at a normal pace after their cleft palates are repaired, and once middle ear tubes are placed for drainage.

**How will my child’s speech change after repair of his cleft palate?**

Right after his cleft palate repair, your child’s sounds may decrease in frequency and variety. It can take up to 6 weeks for him to resume his normal vocalization.

A speech pathologist will work with you to design home activities that can help your child build and maintain normal speech and language capabilities. For example, you should encourage him to make “lip” sounds (“p” and “b”) and “front of the tongue” sounds (“t”, “r” and “d.”) You can do this with playful “lip-popping” games—like mimicking a fish—and tongue-clicking games—like mimicking a horse’s trot. One of our former patients even reports practicing with peanut butter in his mouth! Sounds made in the throat, such as “uh oh,” and animal roaring
sounds should be discouraged, as they can lead to poor speech habits in a child recovering from cleft palate repair.

**Will my child need speech therapy?**

Speech therapy teaches children to make and express sounds in a normal manner. The speech pathologist on your child’s cleft treatment team will determine whether she needs this type of therapy. If so, the pathologist will usually recommend treatment in either an early speech intervention program or a community school program.

**Managing dental and orthodontic issues**

**Will my child’s cleft lip/cleft palate affect his teeth?**

Your child’s first tooth (usually the lower incisor) may appear between 4 and 14 months of age. By age 3, all children usually have their primary (baby) teeth.

A child with cleft lip and/or cleft palate may have poorly formed enamel (outer tooth layer) on some of his teeth, especially those near the cleft. Teeth in this region may also be out of alignment, partially erupted and, therefore, difficult to clean. All of these factors make children with a cleft lip/cleft palate more susceptible to developing cavities.

**How should I take care of my child’s teeth?**

- Brush your child’s teeth at least twice a day to minimize the likelihood of cavities.
- Avoid foods with a lot of sugars and starches. Frequent snacking is especially harmful to the teeth, since the bacteria in dental plaque produce cavity-causing acids each time food enters the mouth.
- Fluoride, whether through the water supply or through prescribed supplements, has been proven to reduce the amount of decay in the baby teeth and permanent teeth. The greatest benefits from fluoride occur between 6 months and 8 years of age. Therefore, a child with a cleft lip/cleft...
palate should be placed on the optimal fluoride dosage early in life.

When should my child see the dentist for the first time?

If your child has a complete cleft lip/cleft palate and has needed a dental appliance, you probably have already met our dental team! Your child should visit a pediatric dentist between the ages of 12 and 18 months, or earlier if you have any questions or concerns.

What dental issues should I expect as my child gets older?

As your child grows and his teeth and bite develop, your pediatric dentist and orthodontist will periodically evaluate the need for treatment.

• Common problems include missing, malformed or extra teeth in the region of the cleft.
• Absent teeth may need to be replaced artificially or by moving teeth into the space with orthodontics.
• A dental implant can be inserted if a tooth is missing. This may be an option for your child once his dental growth is complete.

Will my child need orthodontic therapy?

Since a cleft palate almost always affects a child’s bite, most children with the condition will need at least one phase of orthodontic treatment. The decision to receive orthodontic care should be made by your child’s pediatric dentist, or by an orthodontist who has specialized expertise in treating children with cleft lip/cleft palate.

Phase I orthodontics

Phase I orthodontics are used when the child still has some of his “baby teeth” (typically between the ages of 7 and 10). During Phase I for a child with a cleft lip/cleft palate, an orthodontist uses an appliance to widen the palate in preparation for the alveolar bone graft procedure.


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Preface

A cleft lip is an opening in the lip. A cleft palate is an opening in the roof of the mouth. The palate is made up of two parts—the hard palate and the soft palate. The hard palate is made of bone and is towards the front of your mouth. The soft palate is made up of muscle and tissue and is towards the back of your mouth. Most people have a piece of tissue hanging down from the back of their soft palate that can be seen when you open your mouth. This is called the uvula.

A child can have a cleft lip, cleft palate, or both. Clefts can happen on only one side of the face or on both sides. A cleft can go only part way through the lip or palate or all the way through. Sometimes there is an opening in the bony part of the palate that is covered by a layer of thin tissue. You may not be able to see this opening because it is covered. This is called a submucous cleft palate. A cleft palate leaves an opening between the roof of the child’s mouth and his nose.

The cleft lip nasal deformity has been well described. However, for a long time, cleft surgeons feared that repair of the cleft lip nose at the time of primary repair would cause a growth disturbance especially of the nose. Hence the nasal deformity was not repaired until later. However, from the time of Blair and Barrett Brown, it has been shown that there are no deleterious growth effect from primary nasal interventions. At our centre the senior surgeon has performed primary nasal correction including septal repositioning from the late 1960s. There has been no deleterious growth effect and the overall appearance of the nose has actually improved. This is now well established through many objective studies. Hence
it is now imperative that the deformity of the nose including the septum be addressed at the time of primary unilateral cleft lip repair.

Cleft lip and palate is a common human birth defect, and its causes are being dissected through studies of human populations and through the use of animal models. Mouse models in particular have made a substantial contribution to our understanding of the gene pathways involved in palate development and the nature of signaling molecules that act in a tissue-specific manner at critical stages of embryogenesis. Related work has provided further support for investigating the role of common environmental triggers as causal covariates.

Human birth defects arise from many etiologies, including single-gene disorders, chromosome aberrations, exposure to teratogens, and sporadic conditions of unknown cause. Birth defect syndromes include multiple structural abnormalities and/or cognitive delays. However, most human birth defects affect a single organ system, and those disrupting facial structures are found in approximately 1% (or 1 million) of infants born worldwide each year. The most common of these birth defects is cleft lip and/or palate, a complex trait caused by multiple genetic and environmental factors. All the matter is just compiled and edited in nature. Taken from the various sources which are in public domain.

It is hoped that the book will serve the purpose of students and scholars on the subject and can be useful to them in allied fields.

—Editor
ABOUT THE BOOK

A cleft lip is an opening in the lip. A cleft palate is an opening in the roof of the mouth. The palate is made up of two parts—the hard palate and the soft palate. The hard palate is made of bone and is towards the front of your mouth. The soft palate is made up of muscle and tissue and is towards the back of your mouth. Most people have a piece of tissue hanging down from the back of their soft palate that can be seen when you open your mouth. This is called the uvula. Cleft lip and palate is a common human birth defect, and its causes are being dissected through studies of human populations and through the use of animal models. Mouse models in particular have made a substantial contribution to our understanding of the gene pathways involved in palate development and the nature of signaling molecules that act in a tissue-specific manner at critical stages of embryogenesis. Related work has provided further support for investigating the role of common environmental triggers as causal covariates. It is hoped that the book will serve the purpose of students and scholars on the subject and can be useful to them in allied fields.

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