# Medicine for Managers

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# Cleft Lip and Palate

**A cleft palate is the result of failure of elements of the face to join appropriately during development in the womb leading to a gap between parts of the upper lip or the palate. These developmental anomalies are the most common facial birth defect. The incidence is about one in 700 births.**

Of the babies born with a cleft lip or palate, about a half have a cleft palate only, about a quarter have a cleft lip and the remaining quarter have both a cleft lip and palate.

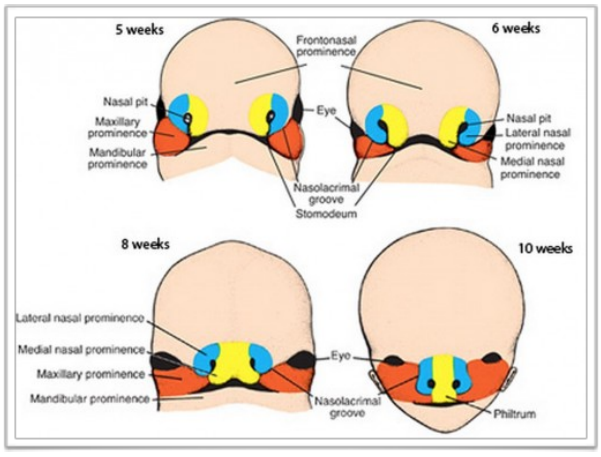
The degree of severity varies from case to case. There may be a small hole in the palate or it may be a split running front to back of the palate, with all variations in between. A cleft lip may affect one or both sides and again may vary from a small notch-like deformity to a split extending from the lip to the nose.

***How does a cleft occur?***
The nose and the mouth of a baby develop between the fifth and the twelfth weeks of pregnancy.

The lips and palate develop from two areas of the developing face

1. An orange-coloured ***maxillary prominence*** (destined to become the cheek bone) on each side of the face.
2. **A pink-coloured *frontonasal prominence*** located at the upper front area of the face.

The maxillary prominences grow to form the lower face, lower lip and jaw, outer part of the upper lip and the hard and soft palate (see the progression of the orange-coloured segments)




The frontonasal prominences grow to form the forehead and, with the ***medial nasal***

***prominences***, the nose and the middle portion of the upper lip.

This process is complete by 12 weeks pregnancy

The various components join together by a process called ***fusion*** so that the face with nose, lips and palate is complete. Sometimes, the process is disturbed and the development is incomplete or the fusion does not occur to some extent resulting in a cleft palate, cleft lip or both.

The aetiology (cause) of a cleft palate is unknown in most cases. In general it is not thought to be anything to do with what was done or not done during the pregnancy. In a few cases, however, there is believed to be an association with smoking, alcohol consumption, obesity, or the use of particular medications.

In a few other cases there may be a genetic component and a cleft may develop in families where a close relative also has a cleft. In a small number of cases, a cleft is one part of a more complex syndrome of several anomalies.

A cleft lip, with  or without a cleft palate, is more common in boys. Cleft palate alone is more common in girls.

***Diagnosis of cleft palate***

It is increasingly possible to diagnose a cleft lip on ultrasound although it may not be apparent on a routine ante-natal scan. In appropriate scans it can be seen after the 13th week of pregnancy.

Cleft palate, especially if occurring as the only abnormality, may be very difficult to see on ultrasound.

If a cleft is identified on ultrasound, it is possible to have an ***amniocentesis*** (taking a sample of the fluid surrounding the baby) to see if the anomaly is part of a wider syndrome, but this is rare.

Of course, cleft lip and palate are rapidly identified after birth during the initial examination of the newborn.

Diagnosis is rapidly followed by a referral to one of the specialist cleft teams in the UK, such as at Great Ormond Street, Bristol Royal Infirmary, Guys, Manchester Children's Hospital, Alder Hey, Nottingham or at Addenbrookes.

The team is multidisciplinary because the problem cannot be cured simply by surgery but will need the intervention of other specialists.

In the early weeks, prior to surgery, there will be difficulty feeding because the baby will be unable to breastfeed or use a bottle because of the inability to form a seal for sucking, resulting in milk and air coming out through the nose. The child is at risk of failing to thrive. Specialist cleft nurses can

advise on feeding, including positioning and alternative methods if necessary.

Children with clefts are more likely to develop hearing problems. The musculature of the palate and ear are linked and disturbances in the anatomy can result in accumulation of thick fluid in the ear reducing hearing. Such children may require grommets.

Dental problems are common resulting in teeth being out of position or sometimes missing altogether. During the childhood years there may be need for treatments including extraction of misplaced teeth, bone grafting and orthodontic treatment to move the teeth into a desirable position.

This is commonly done with fixed orthodontic appliances (braces) and the result of such treatment in the above patient produced an excellent result.

Children may also experience speech problems and, although the problems are reduced by the successful repair of the cleft, nonetheless speech therapy may often be required.

Assessments are carried out by speech and language therapists until they reach adulthood.

The plan for repair is normally to provide surgery for a cleft lip at 3-6 months, repair of a palate at 6-12 months, management of dental, auditory or speech problems from 12 months to adulthood, bone grafting in the mouth if required from 9-14 years and orthodontic treatment if required from 12-18 years.

The final outcome for children with cleft lip and palate is generally excellent. Facial and oral defects are normally considerably improved and most speech or hearing difficulties are resolved. Following surgery any facial scarring fades with time.

***The History of Clefts***

Since ancient times, ignorance of the anatomy and cause of clefts has resulted in behaviour towards those with the defects based on ignorance and superstition.

**Literature suggests that the Greeks ostracised them, whilst the Romans would kill the children who were believed to be harbouring evil spirits.**

Curiously, Hippocrates made no reference to facial clefts in his writings.

Archbishop Magnus of Uppsala in Sweden announced that children with clefts were born to mothers who had leapt over the head of a hare (hare lip). Other equally fanciful theories were published well into the nineteenth century.

Early treatments were largely disfiguring and unsuccessful but in 1460 Pfolsprundt

devised a technique of stitching the layers of the lip achieving a muscle repair.

By 1500, the technique was refined by Brunschwig, a military surgeon from Alsace, using clamps and interrupted waxed sutures.

**Further progress occurred in the 16th Century through the work of probably the greatest of the Renaissance surgeons, Ambroise Paré who undertook detailed studies of the anatomy, improved suturing techniques and first illustrated the procedure in his work *Les Oeuvres*.**

Pierre Franco (1505-78), taught by Paré, further developed the suturing technique to produce unobtrusive scars, so important, he said, for young girls.

Other surgeons developed techniques using flaps to improve the outcome and, in 1848, Hagerdon of Magdeburg introduced a rectangular flap.

He developed a single stage repair for bilateral (both sides of the upper lip) clefts and the technique was still being modified and improved as recently as 1960.

Palatal cleft work is naturally much more recent.

The Finnish surgeon Faltin published details of surgical technique in 1935 but, because he published in Swedish, it took thirty years for the details to be widely known and the Americal surgeon Millard developed a procedure (called the Millard Repair).

He published extensively, including a compendium of surgical techniques in the 1950s and 60s.

**I worked in a maxillo-facial unit in London at the beginning of the 1970s and can say that, in the last fifty years, the results have become considerably better both functionally and cosmetically.**

The expectation now is of an excellent cosmetic and functional result in the vast majority of cases.

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