Plastic and Reconstructive Surgery

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CLEFT LIP AND PALATE

Incidence

Classification	
Cleft lip	
• Incomplete	Unilateral
• Complete	Bilateral
• Plus alveolus (gum)	Median
Cleft palate	
• Soft palate and posterior third of hard palate	Unilateral
• Soft and hard palate	Bilateral
• Submucous cleft	

Cleft lip occurs in one in every 750 live births, affecting males more often than females and the left side more often than the right. Cleft palate affects one in every 2000 live births, and females are affected more often than males. In half of all cases a cleft of the lip and palate occur together. Other cases are roughly divided between cleft lip and cleft palate alone. Overall, the incidence is slowly increasing and it is the most common congenital abnormality of the head and neck. The aetiology of cleft lip with or without cleft palate is multifactorial. It is determined by a genetic predisposition involving several genes plus intrauterine exogenous factors. Cleft palate alone is genetically and embryologically distinct. When parents who are both unaffected have one child with a cleft lip the risk of a subsequent child having a cleft lip or palate is about 5%, rising to 9% if there are two affected siblings. When one parent and one child are affected the risk is about three times greater than in the normal population. Exogenous factors, including the use of thalidomide, phenytoin, or anticancer drugs during pregnancy, or possibly folic acid deficiency, have been incriminated in the formation of facial clefts. In about 3-5% of cases cleft lips and palates are associated with congenital syndromes affecting other parts of the body.

Embryology

The cleft deformity is established in the first six to eight weeks of pregnancy. Two causes have been postulated: (1) a failure of fusion of the maxillary processes with the nasal processes; or (2) incomplete penetration of the mesoderm into the epithelial membranes of the medial and lateral nasal processes and maxillary processes. When the epithelial membranes break down this results in a cleft of the lip or gum, or both.

The palate develops from two shelves from the inner aspect of the maxillary processes, initially separated by the tongue, which with age descends and allows the two shelves to fuse in the midline by the end of the eighth week of pregnancy, joining from front to back. Failure of the fusion of the shelves, or again mesodermal penetration into them, results in formation of a cleft palate.

Management

Patients with cleft lip and palate will require medical supervision from birth through to adult life and the attention of many different specialists including plastic surgeons, orthodontists, oral surgeons, ear, nose, and throat surgeons, paediatricians, speech therapists, and child psychiatrists. Obviously these patients are best looked after under the umbrella of a combined team, where all or most of these specialists can be seen in one visit as necessary, making treatment more convenient for the patient and permitting an interchange of ideas between the members of that team. Such an interchange may lead not only to an improvement in the management of the individual patient but also to improvement in the overall management of other patients with cleft lip and palate.

Few things can be more devastating for a mother than to go through a normal pregnancy only to give birth to a deformed child. A member of the team should always be available to see any such mother as early as possible so that she can have some of her anxieties dealt with and be reassured that help is at hand. Of equal help at this time can be a counsellor of the Cleft Lip and Palate Association (CLAPA). This is a self help organisation run by parents of children with cleft lip and palate, who are able to counsel other parents in this difficult initial period by visiting them in hospital and also by advising them about such problems as feeding. CLAPA has regional groups throughout the country, usually associated with plastic surgery centres. Its headquarters' telephone number is 01-405 9200 x 256.

Repair of cleft lip

Modern techniques of repairing cleft lip appreciate that the deformity affects not only the skin but also the orbicularis muscle, the underlying maxilla, which tends to be hypoplastic, and the nose, in which the septum is deviated and the alar cartilage is hypoplastic on the side of the deformity. The emphasis is to correct as many of these deformities as possible at the initial operation, and as a result each surgeon tends to have his own modifications of one or other of a few standard techniques. My own personal technique at the initial operation consists of:

- (1) Repair of the nasal layer of the anterior alveolar cleft.
- (2) Correction of the nasal deformity by repositioning the alar cartilage.
- (3) Muscle dissection on both sides of the cleft to reconstitute the orbicularis oris muscle.
- (4) Millard rotation advancement to repair the skin.

Most surgeons still depend on the rule of 10 for assessing the time of closure—that is, the child should weigh 10 lb (4535 g) and have a haemoglobin concentration of 10 g/dl; these usually occur together when the child is about 10 weeks old. This delay allows the lip and nose to increase in size for more accurate surgery and the child to withstand safely a longer operation. There are some surgeons who undertake lip repair (usually a simple lip adhesion) within the first 48 hours after birth. This obviously gives great psychological support to the parents but requires a neonatal surgical unit with experienced anaesthetists and does not give the conditions for a more radical closure; either final closure or additional procedures may consequently be required later.

Repair of cleft palate

Modern techniques of palate repair appreciate that the major abnormality, apart from the cleft, is that the velar muscles in the cleft are abnormally inserted into the back of the hard palate. The soft palate being a muscular organ, the velar muscles have to be dissected free and repaired in the midline of the reconstructed soft palate. In addition, radical dissections, particularly the stripping of the oral mucosa and periosteum off the hard palate, may result in scarring and interference with the growth potential of the maxilla, which can later result in collapse of the palatal arch and a flat face. Surgery for the hard palate is therefore becoming less radical, and indeed some surgeons now close only the soft palate at the initial operation, leaving closure of the hard palate to be performed much later.

Cleft palates are now being closed much earlier and should be repaired between the ages of 6 months and 1 year. This gives the child the correct anatomical and physiological apparatus with which correctly to mimic sounds that he hears, which is the start of speech development.

Clefts of the palate may be associated with considerable underdevelopment of the mandible, resulting in the tongue prolapsing into the oropharynx and thus interfering with breathing—the Pierre Robin syndrome. This may delay surgery until the mandible has caught up in its growth. Submucous cleft palate is a condition in which superficially there does not appear to be a cleft of the palate but in which the uvula may appear bifid and a notch on the midline of the back of the hard palate may be palpated. The essential defect is that the muscles, as in cleft of the soft palate, are inserted incorrectly into the back of the hard palate. Patients with this condition usually present with either regurgitation of food or difficulties in speech and require a repair of the muscle sphincter of the soft palate.

Loss of hearing

Children with cleft palates show a high incidence of loss of hearing. Two thirds of patients aged between 5 and 13 are affected. The muscle tensor palati arises from the eustachian tube and inserts abnormally into the back of the hard palate in patients with cleft palates. This may be a reason for drainage problems of the middle ear occurring, leading to an increased incidence of middle ear infection and thus to conductive hearing loss. Ear infections should therefore be treated promptly and the ear drums inspected regularly. Any loss of hearing may have a deleterious effect on the development of speech. Treatment is by decongestants, antibiotics, and myringotomy.

Speech

Roughly three quarters of children with a cleft palate will eventually speak normally. The palate, being a muscular organ, moves upwards and backwards during speech and completes a sphincter with the lateral and posterior walls of the oronasal pharynx, stopping air going up the nose and diverting it out of the mouth. In children with repaired cleft palates the sphincter may not be completely occluded and air will therefore escape up into the nose, producing nasal escape. Children with nasal escape may try to compensate for this by abnormal movements of the tongue to stop loss of air. This and abnormal dentition will produce errors in articulation. Abnormalities causing nasal escape can be corrected by surgery, but articulation can be corrected by the patient alone. The ability to talk normally depends on: the surgical success of closing the soft palate and producing a competent muscular sphincter; the presence of normal hearing; the intelligence and motivation of the patient; and parental encouragement to speak correctly.

Basal view of palate showing incomplete closure of sphincter

Children who do not speak correctly are usually referred to the speech therapist, who makes the initial assessment. The speech therapist can alter articulation patterns and give the patient exercises to improve movement and coordination of the palate using artificial speech aids. Most severe cases of nasal escape may well require surgery, with about 20% of patients with cleft palates undergoing pharyngoplasty.

The assessment of the palate includes an oral examination of the soft palate to see, on movement, whether the levator muscles have been successfully repaired or whether they remain inserted into the back of the hard palate and whether oronasal fistulas are present. X ray examination of the palate may be undertaken but requires cinefluoroscopy and includes lateral and basal views of the sphincter. Lastly, a nasendoscope may be passed down the anaesthetised nose and the palate viewed from above during normal speech. The exact position and size of the gap between the soft palate and the rest of the sphincter may be seen and the appropriate type of pharyngoplasty designed for the patient. There are many different designs of pharyngoplasty to achieve the desired effect of diverting air into the oral cavity. It is important to note that in a patient who has just undergone pharyngoplasty the voice may initially appear to have changed very little; it takes a trained ear to appreciate that the nasal escape has indeed been cured. The patient, if he had abnormal articulation patterns before surgery, will then need to learn correct articulation.

Later procedures

The orthodontist is an important member of the cleft palate team, who, as a specialist dentist, acts firstly as the archivist. Using plaster study models he can record the initial deformity and follow the growth of the palate from birth to maturity, recording the effects of operative procedures. Secondly, by the use of lateral skull radiographs, he can analyse the growth and development of the jaws and maxilla and advise his plastic and oral surgical colleagues on the exact physical abnormality in terms of size, shape, and position for bones of the developing face. Thirdly, he is called on at various times, from infancy onwards, to manipulate the teeth and jaws into better alignment by the use of appliances.

In some patients disturbance of growth may be so great that orthodontic treatment alone will not suffice. Such patients may have considerable hypoplasia of the maxilla, extreme malocclusion of the teeth, and occasionally an overdeveloped mandible. Once the exact abnormality has been diagnosed major osteotomies can be performed by an oral surgeon, usually when growth of either the maxilla or mandible is considered to be complete, to realign the facial bones. These major movements of the facial bones permit correction of the dental occlusion and also improve the harmony of the face.

LATE COSMETIC SURGERY

Small adjustments may be required to the lip scar, including the production of a nice smooth vermilion. Apart from any osteotomies that may be required, the most common cosmetic abnormality in these patients may well be the nose. Many patients who are now reaching adulthood have not had primary correction of the nose, principally because it was thought that any early surgery might interfere with its growth potential. The nose slumps down on the side of the cleft, and the septum is deviated. At about the age of 16 a correction of the tip of the nose is undertaken together with surgery to the septum, which may be combined with formal rhinoplasty.

I thank my colleagues of the combined lip and palate clinic at the West London Plastic Surgery Centre, West Middlesex University Hospital.

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Timing of procedures

0- 3 months	Repair of cleft lip
6- 18 months	Repair of cleft palate
4 years	Preschool "touch up" of lip
6- 9 years	Pharyngoplasty for nasal escape
9- 11 years	Alveolar bone graft Orthodontic procedures on secondary dentition
15- 18 years	Maxillary osteotomies Adjustment of tip of nose and rhinoplasty