Management of cleft lip and palate

Alex Habel, Debbie Sell, Michael Mars

Cleft lip and/or palate (CL/P) malformations are the most common congenital abnormalities in the craniofacial region (about one in 600 live births in the UK) and present a serious problem for health delivery systems throughout the world. The condition requires multiple surgical procedures from birth to maturity and frequent outpatient attendances. Many patients suffer impaired facial growth, dental anomalies, speech disorders, poor hearing, and difficulties in psychological wellbeing and social relationships. A recent study from Denmark showed that subjects with CL/P aged between 20 and 30 years have double the incidence of suicide when compared with normal subjects (A M Herskind et al, paper given at 7th International Congress on Cleft Palate and Related Craniofacial Anomalies, Queensland, Australia, 1993). A significant number of associated syndromes (now over 342) result in problems for cardiac, limb, ophthalmological, and multiple other systems; many have genetic implications. All these features dictate that this condition should be managed by specialist multidisciplinary teams whose members consult with the patients together. Ideally their services should be provided in dedicated specialist centres.

Most of the literature stresses the surgical timing and techniques required for primary lip and palate closure. This has unfortunately led to the view that this is simply a cosmetic problem wherein once surgery in early infancy has been undertaken the problem is resolved. In other words it is inappropriately perceived, not only by some clinicians, but also by parents, as the surgical closure of a wound. This concept has historically resulted in the development of a narrow surgical perspective of the problem, which is still perpetuated in some quarters. This article deliberately avoids discussion of precise surgical protocols but aims to provide an overview with special emphasis on outcome measures from birth to maturity. The paediatrician is ideally placed to offer a global approach for the child with CL/P. His/her involvement should start in the immediate neonatal period through to the completion of physical growth, and during the emotional and intellectual hazards of adolescence.

Team structure

The optimum is for the following clinicians to be available in a multidisciplinary CL/P team: paediatrician, plastic surgeon, oral-maxillofacial surgeon, ear, nose, and throat (ENT) surgeon, audiologist, orthodontist, speech and language therapist, specialist nurse, clinic co-ordinator, psychologist, social worker, and geneticist. Maximum benefit can only be achieved by a referral rate of no fewer than 30 new cases per year which would justify the establishment of such a team; this ensures clinical expertise and experience, as well as the opportunity to provide meaningful audit of outcome (report to the Audit Committee of the Royal College of Surgeons of England, 1995). It is disturbing that the Royal College of Surgeons’ recent investigation of services to this client group in the UK found that far too many surgeons are treating too few patients without proper team support. This may account for the poor results in some British centres when compared with their European counterparts.

Timetable of care

Antenatal care

Embryologically, the primary palate (alveolus and lip) fuses from the incisive foramen anteriorly at about 5–6 weeks of intrauterine life, and the secondary palate from the incisive foramen posteriorly at about 7–8 weeks. These separate processes may fail completely or be arrested at any stage. Clefts occur in a variety of clinical presentations. Diagnosis of cleft lip in utero is now possible by ultrasound scanning from about 17 weeks. Such diagnosis however is not simple, and even in the most experienced centres these defects may be missed (C Welch, personal communication, 1994). False positives have been reported. The diagnosis of cleft palate is even more difficult. When a CL/P is diagnosed antenatal it is generally the paediatrician’s role to identify and confirm the defect and to determine if it is an isolated cleft palate or potentially syndromic. The whole of the palate as far back as the tip of the uvula should be examined with a tongue depressor and a bright light. Palpation with the finger may detect a notch in the posterior border of the hard palate suggestive of submucous cleft palate (fig 1). Other signs of this may be a bifid uvula and/or a translucent central zone. Representing 3% of all clefts, submucous cleft palate is often
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missed or diagnosed late. Nasal regurgitation of fluids may alert the paediatrician.

Cleft size of itself is not necessarily correlated with the extent of dysfunction. Indeed some physically small clefts of the palate and submucous cleft palate may have serious implications for speech function and hearing. Where paediatricians are in doubt early referral to a specialist team is advised.

It is important that the parents are advised promptly and sensitively about the birth of a child with a congenital defect. Ideally a consultant or senior paediatrician should be involved in imparting the news in the first few hours after delivery. Most subsequent criticisms arise where the newborn baby is ‘whisked away’ from the parents and a prolonged delay ensues before the parents are informed. Ideally the parents should be put in touch with a member of the multidisciplinary team within 24 hours of birth (report to the Audit Committee of the Royal College of Surgeons of England, 1995).

The first appointment with the CL/P team provides an opportunity for each member to explain his role and involvement in long term management. Parents should be encouraged to be actively involved with decisions regarding management.

Respiratory difficulties

Children with the Pierre-Robin sequence (small mandible: micrognathia, cleft of the hard and soft palate, small repositioned tongue: glossoptosis) may present with airway problems, which on occasion may be so severe as to render intubation in the newborn extremely difficult. The posteriorly attached tongue may occlude the airway if the infant is laid on the back. The prone position often suffices to overcome this problem. This condition is one of the named exceptions to the recommended supine posture in the ‘back to sleep’ campaign to reduce cot deaths. In more severe cases with persistent soft tissue indrawing, cyanosis or apnoea, a nasopharyngeal airway overcomes the obstruction, improves feeding, relieves associated congestive cardiac failure, and results in improved weight gain. A ‘cut down’ Portex 3·5 mm endotracheal tube is used. Careful positioning of the tube just above the epiglottis is required but once in place it only needs replacement every two weeks until the surrounding structures have grown sufficiently (usually within four to 12 weeks). Such tubes can be satisfactorily managed at home in many cases. Rarely is tracheostomy required.

Feeding difficulties

Feeding difficulties due to CL/P leading to inanition and death of affected infants were recognised almost 400 years ago by Fabricus of Aquapendente. About 25% of CL/P infants have early feeding difficulties with poor weight gain for the first two to three months (145 g/week compared with an average 200 g/week in babies without CL/P). Feeds are also often prolonged, in part due to ulceration of the nasal mucosa. Some infants have increased metabolic needs, for example due to congenital heart disease or airway obstruction.

Successful breast feeding requires negative intraoral pressure which cannot be generated with a cleft palate, but is usually possible with an isolated cleft of the lip where the breast tissue fills the defect. Some mothers are very anxious to breast feed. These mothers should be encouraged to do so, but should be warned that it may be difficult. Otherwise failure may lead to distress and disappointment. This is sometimes resolved to some extent by mothers expressing their milk. A wide range of special bottles and teats are available from CLAPA (Cleft Lip and Palate Association, a nationwide parents’ association) which produces a book Help with Feeding. The continued production of a remarkable variety of teats (some 30 at last count) attest to the persistent and varied difficulties experienced by carers of these babies. Generally it is advised that bottle feeding is kept as simple as possible. Soft easily squeezed bottles, and/or teats with enlarged holes, are usually effective. More complex bottles such as the Haberman feeder may sometimes be helpful. Usually nasogastric feeding is not required and should be avoided if possible. Should this be necessary the involvement of a speech and language therapist is advocated to maintain and develop oral motor skills, and prevent behavioural problems.

Counselling, advice, and support ideally should be available from a specialist nurse (report to the Audit Committee of the Royal College of Surgeons of England, 1995). This helps to promote adequate intake, and reduces parental anxiety about weight gain. Further work is required to put nutritional advice on a truly scientific basis.

Baby plates

A major area of controversy is the fabrication of baby plates (presurgical orthopaedics). This has been common place in many British centres for 50 years. Advocates claim that they help with feeding, facilitate lip and palate repair, and encourage the parent. Further claims are that speech and facial growth are enhanced. To date there is no evidence to support or indeed refute any of these claims. The practice remains empirical.
Lip and palate repair

Protocols for lip and palate repair vary from centre to centre and are empirical. A review of 34 European centres revealed 34 different approaches. The optimum timing and nature of surgery remains elusive. As a general rule, in most British centres lip repair is undertaken at 3 months and palate repair between 6 and 12 months. The recent fashion for neonatal lip repair on the basis that this will improve bonding of the child and mother has not been proved. There is a current fashion for the so-called ‘functional repair’ first described by Delaire in Nantes (see Le Fanu and Markus and Ward11,12). This is advocated by many British oral-maxillofacial surgeons who claim better outcome for midface growth with this technique, compared with more established techniques usually undertaken by plastic surgeons. Cross centre studies between two historical British cohorts and four European cohorts did indeed show that the British centres demonstrated the poorest results.13 No British or indeed European centre (including Delaire’s) where the functional repair is practised, has submitted its results to the same rigorous audit. Further the European ‘good results’ in the cross centre study were in cases where surgery had been performed by plastic surgeons. Claims for superior outcome by British oral-maxillofacial surgeons using the Delaire technique must therefore be considered to be premature. It will also be appreciated that this claim is made for only one of the outcome measures of facial growth – no reference to speech outcome is made, and yet this is recognised as an equally important indicator of the success of surgery.

PRESCHOOL YEARS

The major areas of concern in the preschool years are: speech and language development, ENT monitoring and hearing, somatic growth and development, and general dental welfare.

Speech and language development

Even when the palate is repaired children are still at risk for subsequent speech disorders. The incidence of speech disorders varies considerably. Authors have reported that approximately 25% of children with CL/P develop normal speech after primary surgery while approximately 75% require many interventions throughout childhood and adolescence to achieve acceptable speech production and language competence.14 The causes of speech disorders are often multifactorial and complex. Persistent structural anomalies such as velopharyngeal insufficiency (VPI, see below), dental and occlusal problems, oronasal fistulas, and hearing problems may contribute. Other associated anomalies, such as syndromes or social and psychological factors may be causative factors. In addition, children born with cleft palate are subject to those same factors that influence the development of speech and language in the child without a cleft, such as neurological, cognitive, developmental, environmental, and emotional influences.

Although expressive language development is often slower in these children, the speech problems usually associated with cleft palate include disturbed resonance or tone, abnormal consonant production, excessive nasal air emission/turbulence during consonant production, and disturbed laryngeal voice quality.15 The commonest disorder of tone is that of hypernasality: it occurs when the sound waves produced by the vocal folds enter both the oral and nasal cavities during speech, causing both cavity chambers to vibrate and enhance the sound waves. The overall sound of the patient is ‘nasal’. The main cause of hypernasality is VPI, in which there is inadequate closure of the velopharyngeal valve during speech. Approximately 15–20% of patients who have repair within the first 12–15 months of life have VPI. Sometimes an oronasal fistula may be significant; air enters the nasal cavity through a fistula in the hard or soft palate. When VPI is suspected two simple investigations are undertaken: multiview videofluoroscopy (moving x rays while speaking) and nasopharyngoscopy. The latter involves fibreoptic examination of the structure and function of the soft palate and lateral and posterior pharyngeal walls during speech.16 Treatment is usually surgical, often described as a pharyngoplasty, the exact nature of which is defined by the nature of the defect that is observed through these techniques.17 The treatment options include prosthetic appliances such as a palatal training appliance or speech bulb, or biofeedback speech treatment particularly applicable where an inconsistent pattern of closure is found. An obturator consists of a dental retainer with an ‘extension’ that is designed to occlude the residual velopharyngeal gap that occurs during speech.

A significant problem for patients with cleft palate is difficulty in consonant articulation. Consonant errors related to the cleft palate, that is ‘cleft type errors’, need to be distinguished from those related to other aetiological factors, such as hearing problems, or developmental factors.15 The major input of the speech and language therapist is during the period of normal speech development, in particular the child’s first seven years. The speech and language therapist assesses, provides a differential diagnosis, advises other team members and parents/caregivers, and provides treatment. When regular treatment is required this is usually undertaken at the child’s local health centre, in close collaboration with the team. The monitoring of speech continues into maturity alongside active orthodontic and surgical management. The timing of speech therapy intervention is often determined in collaboration with other members of the team.

Hearing

Patients with cleft palates are at increased risk from middle ear effusions. This is because the
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Attentions of the levator palatini muscle around the entrance to the eustachian tube are abnormal leading to poor aeration and drainage of the middle ear. Repair of the palatal defect is therefore unlikely to result in improved function. Frequently grommet insertion is recommended. Regular review by the ENT surgeon and audiologist is recommended so that poor hearing is not a contributory factor to compromised speech.

Somatic growth and development
The nutritional, early component of the infant-child-puberty model of growth is arguably the most important phase. Fortunately the initial poor weight gain usually resolves after lip closure, and catch up growth occurs by 6 months. Ideally children with cleft palate should have their height measured annually. As most children attend cleft palate clinics where height measurement may not be routinely measured, cases of impaired growth may be missed until late. If a child with cleft palate fails to achieve a normal growth velocity, septo-optic dysplasia and growth hormone deficiency should be suspected. Although it is reportedly 40 times commoner in cleft palate, this is still not frequent enough to account for short stature below the fifth centile being five times commoner in children with isolated cleft palate. The peaks and pulsatile release of growth hormone may be relevant but have not been evaluated in this group if children.

Alerting parents to the potential difficulties in development can bring earlier referral for appropriate action. In the preschool years in the non-syndromic population impaired concentration and delay in achieving milestones other than those of speech and language is reported to be commoner in children with cleft palate. This may be a valid association with clefting, or multifactorial, and include impaired hearing, multiple hospitalisations, recurrent ear infections, and overprotectiveness by the parents limiting social interaction and playgroup attendance.

General dental welfare
Children with CL/P are at risk from disordered dental occlusions. If the cleft involves the dental bearing portion of the palate (the alveolar process) there is the possibility of congenitally absent, malformed, or supernumerary teeth. No active orthodontic intervention is required in the preschool years. Regular attendance at the dentist is recommended and the importance of a balanced diet with a minimum of refined carbohydrates and good oral hygiene is stressed. Where the local water supply is not fluoridated fluoride supplements should be prescribed. The periods of active occlusal manipulation should be deferred until the permanent dentition begins to establish (see below).

The school aged child
The major areas of concern in the school years are: orthodontic management, the surgical procedure of alveolar bone grafting, academic achievement, and psychological issues. When appropriate the team works in close liaison with the community child health services.

Alveolar bone grafting
Autologous bone grafting for patients with CL/P has been practised for many decades. Early attempts to fill in the bone defect used rib struts at the time of lip repair - so called 'primary bone grafting'. The results were generally poor, and this practice is now obsolete.

Secondary bone grafting, or alveolar bone grafting, has been practised in Great Britain since 1982; it was first described in America in 1972. It is one of the major technical breakthroughs in recent years. At ages 9–11 years

Figure 3 (A) Before and (B) after alveolar bone graft occlusion showing cleft defect and its closure; fixed appliance in place.
cancellous bone, usually taken from the iliac crest, is placed in the alveolar process defect. A period of six to nine months of orthodontic alignment and expansion precedes the operation, which is a combined surgical/orthodontic exercise. Patients are admitted for about three days.

After three months the grafted bone is indistinguishable from maxillary bone on radiography and the bony defect is completely filled in (fig 2). Alveolar bone grafting permits the creation of a normal alveolar architecture through which teeth can erupt and subsequently be moved orthodontically (fig 3A). This surgery has other benefits: residual fistulas can be repaired simultaneously and more successfully. Sometimes the appearance of the nose can be improved because of augmented nasal base infrastructure. However the major advantage of alveolar bone grafting is that the need for future replacement of missing teeth is considerably reduced – dentures and bridges are much rarer than 15 years ago and, when required, are much smaller.

Great Ormond Street Hospital for Children introduced alveolar bone grafting to the UK based on work developed in Oslo, Norway by Bergland et al. The success rate at Great Ormond Street NHS Hospital Trust is over 95% in the audit reports of the first 200 patients (M Collins et al, paper given at the Craniofacial Society of Great Britain, Cambridge, 1994 and Y L Jia et al, paper given at the Craniofacial Society of Great Britain, Glasgow, 1995). No significant untoward effects on facial growth have been observed.

After alveolar bone grafting the dentition is allowed to erupt relatively normally until definitive orthodontic treatment is required.

Definitive orthodontic treatment
Three to four years after alveolar bone grafting, at around 12–13 years, definitive orthodontic treatment can proceed. Treatment usually takes two to two and a half years with out-patient appointments monthly. Alignment is nearly always by means of fixed appliances (fig 3B) and arch expansion is required. In the majority of cases good interdental alignment and interdigitation is now possible.

Academic achievement
Reading difficulties are found in a third of primary schoolchildren with cleft palate, caused or compounded by impaired hearing, deviant articulation, and delayed language development.

Psychological issues
Emotional difficulties may emerge from middle childhood; poor self esteem and impaired social relationships leading to depression in adolescence may be improved by midfacial surgery. Peer acceptance and positive parent-child relationships are the best prophylactics, though not always available. School refusal, even if minor, or marked overdependence on adult company, should lead to a review of social function and discussion with child guidance. Workshops for the facially disfigured teenager are offered free of charge by Changing Faces, 27 Cowper Street, London EC2A 4AP.

Child abuse is common in children with disability, and children with CL/P are probably at particular risk before closure of the defects, though this has not been quantified in a non-syndromic cleft population.

ADOLESCENCE
Compromised facial growth and the need for major maxillary advancement
A significant number of patients with CL/P experience distorted midfacial growth. Midfacial retrusion may not become apparent until the pubertal growth spurt, at which time gross facial disharmony may become evident. Studies on adult subjects from the less developed world who have received no surgery whatsoever have shown that palatal surgery in early infancy is responsible for such growth disruption. Unfortunately, palatal repair is essential for normal speech development.

No ideal timing of surgical intervention that minimises facial growth disturbance while permitting normal speech development is presently known. The two are closely primary objectives of cleft lip and palate management. Benefits for one should not be obtained with detriment to the other as may be the case with delayed hard palate closure.

Treatment of midface retrusion
Surgical advancement of the maxilla is the only viable treatment for midface retrusion. This is usually delayed until facial growth has ceased so that the facial position can be achieved without further adverse growth (fig 4). Unfortunately, this requirement means that patients have to go through their vulnerable adolescent years with a worsening facial appearance. Occasionally for this reason
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surgery may be undertaken during the early teenage years if it is considered that delay might cause considerable psychological harm to the patient. Such patients must be warned that a second major repeat operation may be required.

Rhinoplasty
The nose is one of the last components of the face to cease growing. For this reason wherever possible rhinoplasties are deferred until the very late teenage years.

Genetic advice and referral
Three types of genetic risk groups are present. The syndromic group are most easily identified by examination, followed by the familial, by history: the isolated defect is identified by exclusion. Among the 40% of children with an associated major malformation is the syndromic group. This is a large group that may have been relatively ignored in the past, accounting for almost 25% of a large geographically representative Scottish series. More than 340 syndromes are associated with clefting, so this alone is not a particularly good ‘handle’ by itself. When found with other less common conditions, lip pits for example, a diagnosis can be made of Van de Woude’s syndrome, an autosomal dominant condition. Other syndromes may be chromosomal, for example trisomies 13 and 18, the DiGeorge sequence, and velocardiofacial cardiac or Shprintzen’s syndrome. The latter two conditions are now thought to share a gene deletion at the 22q11 region and may also be transmitted from parent to child.

In cases of isolated defects, cleft palate alone is associated with the likelihood of recurrence of 2% whereas CL/P has a 4% risk overall, varying between 2–6% depending on the severity of the defect. In familial cases the risk for subsequent pregnancies is one in 10 with two siblings, or sibling and parent affected. Apparently isolated cleft palate may be the pointer to more serious conditions, for example Stickler’s syndrome, an autosomal dominant condition requiring careful ophthalmological follow up, with characteristic long bone changes on radiography that are not always present in early childhood.

Outcome measures
The principal outcome measures are facial attractiveness, speech, facial growth, and psychosocial wellbeing (figs 5 and 6). Teams of sufficient size with large volumes of patients are in a position to make meaningful audit of their results. For example a retrospective audit of children born at Great Ormond Street Hospital for Children in 1960–70 showed that 50% required major reconstructive surgery in late adolescence. A more recent study of children born in 1976–9 showed that this had reduced to 35% compared with 10–12% in Copenhagen and Oslo. These results have led to changes in surgical technique that should result in better outcome. The results of mid-face osteotomy in 30 patients one year after surgery are as stable as other published series. Studies of speech and velopharyngeal function reveal that this surgery is free of complications with respect to speech (D Sell et al, paper given at the 7th International Congress on Cleft Palate and Related Craniofacial Anomalies, Queensland, Australia, 1993).

Conclusion
The complex nature of treatment for CL/P, a condition that requires a large multidisciplinary team treating patients from birth to maturity, has been outlined. Subjecting centres’ outcomes to audit should precede heeding the current siren calls for paediatricians to refer children exclusively to a particular surgical speciality. A growing body of evidence has shown a close correlation between quality of outcome and the availability of high volume centralised care by dedicated teams, as has been proved and accepted for years in other fields such as surgery in infancy, childhood malignancies, and cystic fibrosis.
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