Cleft lip and palate: current management

Tim Goodacre
Marc C Swan

Abstract
Cleft lip and palate are the most common presenting congenital conditions of the face and cranial bones. This article describes current understanding of the aetiology and presentation of the deformity and management of the child from prenatal diagnosis until maturity. Principle concerns include correction of the physical defect with the best possible functional and cosmetic outcome, optimal speech correction, satisfactory feeding and hearing, and dental and orthodontic health. The value of comprehensive management of all aspects of care within a multidisciplinary team including clinical psychology support for child and family is discussed.

Keywords alveolar bone graft; cleft lip; cleft palate; naso-alveolar moulding; orthognathic; pharyngoplasty; postoperative emergencies; submucous

Definitions
Cleft lip (CL) is defined as a congenital abnormality of the primary palate (i.e. anterior to the incisive foramen). It may be complete, incomplete or microform, unilateral or bilateral, and may involve a palatal cleft (CL±P) (See Figure 1).

A cleft palate (CP) is a congenital abnormality of the secondary palate and may be complete or incomplete, unilateral or bilateral, or submucous.

CL±P is epidemiologically and aetiologically distinct from isolated CP.

Epidemiology
The overall incidence of orofacial clefting is approximately one in 700 live births, amounting to approximately 1000 new cases per annum in the UK. However, the incidence varies with ethnicity, geography and the nature of the cleft itself.

In the context of CL±P, the incidence is approximately 0.3 per 1000 in African American populations, 1.0 per 1000 in Caucasian populations, and 2.1 per 1000 in Japanese populations. The incidence of isolated CP is racially homogeneous at approximately 0.5 per 1000 live births.

Unilateral clefts are nine times as common as bilateral clefts, and occur twice as frequently on the left than the right. The ratio of left:right:bilateral clefts is 6:3:1. Males are predominantly affected by CL±P (M:F 2:1) whereas females are more commonly affected by isolated CP.

Aetiology
Over 300 syndromes are associated with orofacial clefting and most occur as an isolated abnormality – the so-called non-syndromic CL±P. Isolated CP is more likely to be syndromic than CL±P.

The cause of isolated clefting is multifactorial involving a complex influence of environmental and genetic factors. There is a predisposition for familial clustering. In one Danish study the concordance rate for CL±P was 60% in monozygotic twins and 10% in dizygotic twins.

There is a dose–response relationship between maternal periconception smoking and orofacial clefting. Maternal alcohol consumption is also associated with an increased risk of isolated CP. Other maternal risk factors include diabetes, nutritional factors (e.g. vitamin A, folic acid), and anticonvulsant medication.

Genetics
Inheritance may be chromosomal, Mendelian or sporadic (Table 1). With respect to non-syndromic clefts, the risk of unaffected parents with one child with CL±P having a second affected child is 4%, while with two affected children, this risk increases to 9%. If one parent has a CL±P, the risk of having an affected child is 4%, which increases to 17% for a second affected child.

A total of 35% of CL±P patients and 54% of isolated CP patients are associated with another anomaly, although less than 3% of these is due to a single gene disorder.

Numerous ‘candidate’ genes/loci have been proposed on the basis of linkage and/or association studies and include TGF-α TGF-β-3 MSX-1 and IRF-6.
A recent longitudinal population based study from Norway demonstrated that the risk of recurrence of an isolated cleft in first degree relatives does not seem to be related to the anatomical severity of the defect. Furthermore, the relative risk of cleft recurrence in first degree relatives was 32 for any cleft lip and 56 for isolated cleft palate – thus indicating that genetics contribute more to cleft palate alone than to cleft lip. There was a low (three-fold) crossover risk between the incidence of cleft lip and isolated cleft palate in families, which implies that genes such as MSX-1 and IRF-6 may participate in all forms of oral clefting.

Antenatal diagnosis

Since first reported in the prenatal diagnosis of facial clefting, most centres performing 20-week foetal anomaly ultrasound scanning now include observation of the facial elements as a routine. Detection of cleft lip and alveolus (gum) is around 70% cases in the best series, but the sensitivity is generally in the order of 20%, although there is high specificity. Missed cases will inevitably occur due to foetal movement and adverse position during the scan. Isolated CP is particularly difficult to diagnose on account of the acoustic shadow created by the facial bones. Currently up to 25% of cleft lips (with or without CP) are diagnosed antenatally. The addition of three-dimensional (and now four-dimensional) ultrasound methods gives better quality pictures for parents’ benefit, but do not significantly improve the ability to predict foetal palate status. To date, only foetal magnetic resonance imaging offers a realistic means of predicting important additional information about the palate, which has a bearing upon the future child’s feeding, speech, and facial growth capacity (Figure 2).

Prenatal diagnosis has been described as a ‘mixed blessing’. Psychological studies of parents indicate the appreciation of preparatory knowledge, but an increased anxiety level during the remaining pregnancy. Diagnosis, therefore, carries with it a considerable obligation for parental support and counselling – now offered routinely by most of the newly configured cleft teams. Training for ultrasonographers involved in the first moments of detection has been shown to be beneficial. Outcome following prenatal diagnosis of clefting across UK maternity units is unknown, but carefully organised support for parents has avoided the high levels of termination of pregnancy for isolated clefting that have been reported elsewhere.

Cleft types

Most clefts fall within the fusion lines of the fronto-nasal process and lateral maxillary elements and the midline of the palatal shelves in the mouth once posterior to the incisive foramen. CLEFTs in other lines are rare and were classified by Paul Tessier as craniofacial clefts no. 0–14. They are not the subject of this review, and are almost always best managed by referral to a specialist paediatric craniofacial team.

‘Typical’ clefts may involve all or part of one or both lip philtral columns, alveolar (gum) bone, hard palate or soft palate. The cleft can also be complete, incomplete, or a forme fruste involving muscle dehiscence only. In the lip, these latter ‘near misses’

Table 1

<table>
<thead>
<tr>
<th>Chromosomal Trisomy 13 or 21</th>
<th>Cleft lip and palate; CP, cleft palate; AD, autosomal dominant.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Single gene</td>
<td>Treacher Collins Syndrome</td>
</tr>
<tr>
<td>Van der Woude (Chromosome 1, AD)</td>
<td>Stickler Syndrome</td>
</tr>
<tr>
<td>EEC (ectrodactyly, ectodermal hyperplasia) and CL±P Syndrome (Chromosome 3, AD)</td>
<td>Velocardiofacial Syndrome</td>
</tr>
<tr>
<td>EEC (chromosome 12, AD)</td>
<td>Opitz GBBB Syndrome (AD)</td>
</tr>
<tr>
<td>Sporadic</td>
<td>Pierre Robin Sequence</td>
</tr>
</tbody>
</table>

Figure 2

Intra-uterine magnetic resonance imaging scan (sagittal view) demonstrating normal a and cleft b palates. The absent palatal stripe (red arrow) in b is a pathognomic sign of cleft palate.
can produce asymmetry of smile and nasal shape and thus are of cosmetic importance. In the soft palate, a *forme fruste* is of even greater importance, presenting as a submucous cleft palate.

The approximate distribution of the major cleft types is as follows:
- cleft lip and palate 46%
- isolated cleft palate 33%
- isolated cleft lip 22%

A total of 86% of bilateral cleft lips and 68% of unilateral cleft lips are associated with a cleft palate deformity.

**Submucous cleft palate**

Submucous cleft palate (SMCP) is sufficiently important to merit further mention. It may present ‘overtly’ with a classical triad of signs:
- notched hard palate posterior margin (however small)
- bifid uvula
- lucency of midline of palate (the ‘zona pellucida’ – due to muscular diastasis) (Figure 3).

Even the most marginal of hard palate notches is a hard sign of possible SMCP, in contrast to the bifidity of the uvula, which is common in those with no other signs of muscle dehiscence, and may be of no clinical consequence.

‘Occult’ presentation of SMCP presents with the speech and swallowing difficulties of SMCP, but none of the classical triad of signs. It can be confirmed by observing a characteristic ‘grooved’ surface on the dorsum of the soft palate during nasendoscopy.

SMCP presents a management conundrum. Most – detected by neonatal examination or as a consequence of early feeding difficulties – will progress to severe speech dysfunction if left untreated. For these children, the only effective treatment is surgical muscle dehiscence, and may be of no clinical consequence.

Diagnosis of SMCP is often delayed. Awareness of the presenting features should enable detection during all neonatal screening examinations if the mouth is examined correctly. Casual slipping of a finger into the mouth is not adequate. Suspicion should always be raised by neonates who fail to suck with good pressure, and in older toddlers whose speech develops with cleft type characteristics (see speech section).

**Bilateral clefts**

Bilateral cleft lip presents a difficult problem when associated with alveolar +/− more posterior clefting. The bony element (the ‘premaxilla’) may be unrestrained by the normal ring of lip muscle, and protrude in a much distorted and upwardly rotated position (Figure 4a). Presurgical orthopaedics (using a dental plate, and sometimes lip strapping) is often helpful in such cases and may make the primary surgery a much easier (and, therefore, successful) procedure.

**Early care**

**Presurgical orthopaedics and naso-alveolar moulding**

There is a long history of the use of dental devices to assist cleft lip and palate management. Prospective randomised trial data now shows that there is no benefit to infant feeding with the use of such treatment. The term ‘feeding plate’ is, therefore, now defunct.

Moulding of the dental arch form with orthopaedic devices is more controversial, and the subject of a large multicentre trial still accruing data. Contradictory data exist for whether such bony manipulation improves outcome, affects growth, or makes surgery more straightforward. Improved surgical ability is likely to be the most consistent and valuable effect, especially if it produces more satisfactory cosmetic outcomes.

A variety of appliances have been described, which if ‘active’ may contain springs or other mechanisms to gently oppose the gum ridges. No UK unit currently uses the more severe Latham device, which requires pin fixation to the jaw arch line.

Naso-alveolar moulding involves adding an extension to the orthopaedic device, to exert moulding pressure on the distorted nasal margin. The principle is similar to that espoused for ear cartilage moulding in early months (‘ear buddies’). It requires much additional work (and cost) from the orthopaedic team, and adds considerably to the burden of care for the new parents. No UK unit currently offers this service routinely, but the best series in the United States have impressive results, which would be expected to lead to better cosmetic outcomes.

**Postoperative nasal splints**

Along similar lines, post lip repair nostril splinting (using conformers) is thought by many to be beneficial. Use of progressively enlarging conformers to shape the nostril aperture and lift the slumped rim has a good evidence base, but again requires considerable commitment from the parents in conjunction with good nurse specialist support.

**Specialist nursing care**

One of the indisputable advances in UK cleft care over the past 15 years has been the widespread development of highly skilled nurse specialists to offer support to expectant parents, peri-natal care, complex feeding advice, home visits and peri-operative care. The success of such specialists has improved continuity of care from the more centralised teams, and raised standards from previously *ad hoc* support structures. Arguably, this development has had a far greater impact on outcomes than any specific surgical methodological change.
Pierre Robin sequence
The sequence of cleft palate associated with micrognathia, glossoptosis, and respiratory difficulty, was described by Robin in 1923. The incidence is approximately one in 14,000 births.27 Not indicative of a specific syndrome, the spectrum of severity ranges greatly. The most severe forms exhibit wide clefts with gross maxillary shelf and muscle hypoplasia, the tongue prolapsing as a ‘ball valve’ into the posterior nasopharynx, a minute jaw, and major problems with airway maintenance. Management of Pierre Robin sequence is concentrated on establishing a secure airway at all times and satisfactory oral feeding (Figure 5).

Current best practice is invariably to use a nasopharyngeal airway for the mainstay of airway protection. The widespread adoption of this method has obviated the need for more invasive techniques of the past, such as glossopexy (fixation of the tongue tip to the lip/jaw) or the Burston frame (a prone positioning frame to allow forward head projection). Very early surgical distraction osteogenesis of the hypoplastic mandibular arch has been advocated by several current authorities in the United States.28 However, the method has found few devotees elsewhere, where surgical enthusiasm is more reasonably balanced by wise, less invasive, medical support. No case in the past 17 years in Oxford (a major referral centre for such cases) has indicated such surgical treatment.

Feeding support for such children can be problematic. Nasogastric supplementation may be required, but every effort is made to prevent the child becoming overly dependent upon nasogastric feeds in preference to using normal oral sucking.

Postoperative emergencies
Primary surgery
The most commonly encountered problems after primary cleft surgery are airway obstruction and bleeding. Airway obstruction usually follows narrowing of the nostril apertures in lip/anterior palate closure, in the child who is still an obligate
nasal breather. Use of the nostril conforming splint, together with perhaps a nasogastric tube, can further obstruct the nasal airway. Relief of such obstruction is usually easy with gentle exterior suction and use of a nasopharyngeal airway if required.

Airway obstruction following posterior palate repair can be more difficult. If anticipated, a nasopharyngeal airway can be left in situ at the end of the procedure, together with placement of a temporary tongue suture to assist with positioning. If a nasopharyngeal airway requires placement on the ward postoperatively, it should be passed with the utmost care to avoid suture line disruption with inevitable additional bleeding and functional consequences for the repair.

Significant postoperative bleeding is a surgical emergency. Some bleeding in the first 12 hours is expected, although the surgeon should be alerted if it is fresh or brisk. Later bleeding can be reactionary or secondary in nature. Either way, the child will require some sedation (usually morphine 0.1 mg/kg), and a topical adrenaline soaked gauze swab is used to apply pressure onto the roof of the mouth. The most common bleeding site is one of the lateral releasing incisions used to close the cleft palate and such digital pressure can establish control remarkably quickly. Unnecessary staff and/or parents should be removed from the room and very great care given to any suction (avoided if at all possible). The child should be prepared for urgent return to theatre, although often ward control obviates the need for any more active surgical intervention.

Secondary surgery
Secondary procedures include pharyngoplasty, alveolar bone grafting and osteotomy-orthognathic surgery. Bleeding is the most common later emergency. It should be managed with all usual supportive measures (ABC, intravenous fluids and cross matching if appropriate) and early return to theatre. Airway obstruction can frequently follow the less physiological forms of pharyngoplasty.

Current treatment protocols
Timing of cleft repair
The late 20th century UK-based controversy surrounding the value of neonatal cleft lip repair has now almost disappeared; the pur-ported benefit of improved lip scarring from foetal wound healing patterns is disproven, and no benefit to parental bonding from early restoration of ‘normality’ demonstrated. Most UK centres (and similarly almost all world centres) now undertake first surgery once feeding patterns have been established and birth weight regained.

The main difference in timing protocol in major centres is found in the sequence in which the lip and palatal elements are operated upon. The more frequent pattern used is lip and anterior palate as a primary procedure around 2–3 months, with soft palate closure once the airway is more secure – from 4 to 12 months. The opposing view (common in France – sometimes termed the ‘Malek sequence’) aims to avoid any early surgical interference with the hard palate growth centres, and repairs the lip +/− soft palate at around 2–3 months, followed by delayed hard palate closure at times varying from 6 to 60 months. Those centres adopting palate repairs later than 12 months frequently use hard palate cover plates to reduce abnormal airflow and permit better speech development than would occur in the presence of an oronasal fistula.

The dilemma of the mutually opposing benefits of early and later hard palate repair on palatal growth versus speech development remains one of the most controversial and difficult aspects of cleft management. Robust evidence accounting adequately for all variables and cleft types is lacking, and many opinions accept that it is the quality of overall surgical tissue handling rather than defined technique or timing that has most influence on long-term outcome for growth and speech.

Cleft lip repair
Lip repairs adopt a form of lengthening of the greater segment margin, the rotation advancement being the most popular.29
Almost all authorities now include some form of primary nasal tip correction in the primary lip repair, results usually improving upon the status quo if nothing is done.\textsuperscript{30} Radical muscle repositioning is also widely adopted, and many surgeons now use a subperiosteal dissection of the muscle away from the maxilla, in order to minimise deep seated scar tissue and offer the potential to generate more bone from the under surface of the periosteum. Subtle corrections of the lip scar with the interposition of small flaps above the white roll, and within the dry vermilion mucosa as well as the avoidance of cuts around the base of the lateral nostril margin are all advances that improve long-term outcome (Figure 6).

Cleft palate repair

The most significant advance in palate repair over the past 15 years has been the widespread adoption of the radical levator palate muscle repositioning procedure described by Brian Sommerlad.\textsuperscript{31} The outcome of this procedure appears to have no adverse growth effect and produces the very best speech capability with lowest incidence of velopharyngeal incompetence. It also has potential benefits on Eustachian tube function. The technique involves closure of the nasal mucosa, followed by transposition of the medial insertion of the levator muscles by 90 degrees so that the two mobilised ends can be sutured together to form a new, extensible muscle ‘sling’, which is capable of velopharyngeal closure (Figure 7).

The remaining development in palate surgery is the trend towards minimising lateral releasing incisions (as with the Von Langenbeck procedure, now in use for over 100 years) by radical undermining of the palatal flaps. Self-inflating tissue expanders might hold some improvement in this respect in the coming 10 years.\textsuperscript{32}

Foetal surgery

The advent of improved antenatal diagnosis of intrauterine pathology has made foetal surgery a feasible option, however the standard ‘open’ techniques are associated with significant morbidity and mortality.\textsuperscript{33} Thus, the development of less invasive feto-endoscopic techniques appears encouraging, and has been demonstrated to be effective \textit{in vivo} using cleft animal models. The major advantage is the ‘holy grail’ of scarless wound healing, which has been reported at mid-gestation and would have clear functional and aesthetic advantages.\textsuperscript{34} Consensus criteria exist as to which congenital malformations are considered appropriate for intrauterine surgery. At present such surgery is purely experimental with respect to orofacial clefting and there is little prospect of clinical trials commencing in the foreseeable future.\textsuperscript{34}

Speech

Cleft palate (overt or submucous) carries an inevitable potential for speech to develop abnormally due to the position of the soft palate musculature. Of the five known soft palate muscles, the palatoglossus and palatopharyngeus are principally involved in swallowing. The tensor and levator veli palatini muscles are speech motors, acting to extend and lift the soft palate in a ‘knee’ shaped valvular action, which closes the posterior nasopharynx from the oropharynx. This action is essential if air pressure is to be raised in the mouth – a necessary component of normal speech in most languages.

Raised oral pressure is needed particularly for fricative sounds (such as ‘s’, ‘f’, ‘sh’) and plosives (‘p’, ‘m’, ‘b’). Failure of this...
action results in the speech pattern described as velopharyngeal incompetence (VPI) the component parts of which are hypernasality, nasal emission and nasal turbulence or resonance.

When young children have incompetent palatal musculature, some will develop very substantial compensatory misarticulations in order to attempt normal speech. These can lead to severely compromised intelligibility, and in some places have resulted in erroneous association with learning disorders.

Recent evidence supports the adoption of therapeutic intervention at an early stage (the babbling phase) to counter adverse speech development. Once normal speech is developing, it is essential that specialised speech and language therapists are involved to monitor and guide speech development. Significant abnormalities in palatal function can then be identified at an early stage, and investigated to ascertain whether secondary speech surgery will offer any benefit to the child.

**Audiology**

Most children with cleft palate would develop glue ear without intervention. This relates to the abnormal positioning of the stylopharyngeus muscle. Some authorities, therefore, advocate early grommet insertion by way of prophylaxis for this condition, to improve hearing, and to prevent chronic secretory otitis media and worse (e.g. cholesteatoma). However, there is no consensus for early middle ear management for cleft children, other than agreement that careful audiological prolonged assessment is mandatory, with appropriate intervention (either grommet insertion or use of hearing aids) as required. It is possible that the newer, more radical, palatal muscle repositioning techniques will lead to ‘normalisation’ of middle ear Eustachian ventilation and reduce the need for external ventilating grommets. However, no published evidence currently exists to support this view.

**Secondary speech surgery**

Although primary cleft palate repair techniques have improved early outcomes, a substantial number of children will remain with deficient palatal function despite optimal speech therapy. Those demonstrated to have VPI are considered for either palatal muscle re-repair (along the lines described by Sommerlad[31] for primary repair) or pharyngoplasty (an operation performed on the pharyngeal wall in order to improve closure of the velopharyngeal orifice).

**Pharyngeal flap**

The oldest, and to some extent most consistently reliable, means of improving velopharyngeal closure is by elevating a myomucosal flap from the posterior pharyngeal wall and attaching it to the posterior soft palate. The static flap acts as a broad ‘wafer’ of tissue tethering the palate and also obstructing the widest portion of the airway.

The pharyngeal flap – of which there are numerous anatomic variations – is also the most obstructive, and can produce significant sleep apnoea as well as other adverse sequelae, such as severe snoring, mucous obstruction, and long-term airflow obstruction (implicated rarely in right heart failure).
Pharyngoplasty procedures
In an attempt to reduce such symptoms, the use of lateral pharyngeal wall or tonsillar pillar tissue to constrict the nasopharyngeal valve was developed. Lateral pharyngeal wall flaps set high in the adenoidal area act as a form of ‘speed bump’ to bring the posterior pharyngeal wall closer to the soft palate and assist closure of the valve (the ‘Hynes’ pharyngoplasty procedure). Posterior tonsillar pillar flaps, set lower, act as a form of dynamic sphincter, and tend to be somewhat more obstructive to airflow (the ‘Orticocchoea’ pharyngoplasty procedure).

Posterior pharyngeal wall augmentation
In an attempt to avoid all adverse obstructive consequences of pharyngoplasty, some authorities create the ‘speed bump’ effect on the posterior pharyngeal wall entirely by placing a piece of material (cartilage or alloplastic) beneath the pharyngeal mucosa. Such secondary speech procedures are now only performed with full pre- and postoperative support by a specialised speech and language therapy team. Additional, non-surgical, methods for the infrequent severe and resistant cases include the use of prosthetic devices such as ‘speech bulbs’ attached to permanently worn dental plates, and biofeedback therapy techniques.

Alveolar bone grafting
Primary cleft repair does not correct the bony deformity in the gum ridge, although recent work on primary gingivoperiosteoplasty is an attempt to address this. Since the 1970s, it has been understood that the bone defect is best filled with cancellous (marrow) bone graft in the secondary dentition phase, before the eruption of the secondary canine tooth, which usually lies adjacent to the cleft gum (Figure 8). The timing of this procedure is determined by the specialist orthodontist and relates to dental maturity. It is usually between the ages of 7 and 14 years and involves reopening the cleft bony line and packing the mucosally lined cavity with chips of graft harvested from the iliac crest or tibial plateau. This procedure also offers the best opportunity to close any residual oronasal fistula in the anterior palatal region. It is a highly effective operation, and enables the subsequent orthodontic management of what are frequently much distorted teeth into a well-corrected arch form.

The necessity for such bone grafting is well established. Careful paediatric dental health is a mandatory part of early cleft care, and later specialist restorative dental expertise enables all but the most resistant cases to obtain a high level of dental health and appearance.

Orthognathic surgery
Despite the best outcomes available, a certain proportion of children with complete cleft lip and palate will develop secondary maxillary hypoplasia following primary repair. The characteristic ‘dish face’ appearance is often the subject of severe teasing and self-consciousness, and is best addressed by a combination of psychological support with orthognathic (jaw moving) surgical correction in the late teenage years.

Orthognathic surgery involves moving the maxilla forwards by means of controlled bone cuts (‘osteotomies’) in patterns described by René Le Fort in the 19th century. Maxillary advancement may be combined with surgical correction of the mandible by sliding it backwards again using bone cuts. Recent use of bone distraction osteogenesis has improved the long-term outcome of such procedures.

The cause of mid-facial growth failure remains controversial. The adverse effect of primary surgical intervention is indisputable (as shown by careful analysis of adult unrepaired clefts in Sri Lanka over many years) but the relative impact of various forms of primary management are still unclear. It would appear that, despite the best efforts to minimise adverse sequela of surgery, some children are ‘poor growers’ with inherent hypoplasia, and are destined to require significant secondary surgery regardless of their primary treatment protocol.

Clinical psychology
Throughout this description of interventional medical care for cleft lip and palate children, it will be evident that the goal of all care should be a well-rounded and healthy child able to achieve his/her full potential in life. They should be able to enjoy a normal childhood as little disrupted by treatment or adverse consequences of the cleft as possible.

The input from skilled specialist psychologists to cleft management from prenatal diagnosis to adulthood is invaluable, and essential as a guide to achieve holistic care from all team members.
the Clinical Standards Advisory Group (CSAG) report and reorganisation, a full psychology service has become a mandatory part of the team structure. It is probable that the number of unnecessary or ill-advised secondary procedures is reduced by such input, as well as demonstrable improvements in individual and family wellbeing and dynamics. Future developments in the area of psychological input will focus on the potential value of early intervention in families at high risk of adverse psychological consequences when older.

CSAG

Perhaps the single most effective change in cleft care standards in the UK has been brought about by reorganisation and service centralisation stimulated by the 1996 report from the CSAG - now a defunct organisation. Service provision was reduced from 57 centres in the early 1990s to nine centres (some ‘twin site’) in the UK from 2006. This centralisation has been accompanied by more careful financial investment by regional commissioners, and has brought UK clinical outcomes to a level that is arguably among the best in the world.

REFERENCES


Practice points

- Cleft lip and palate are the most commonly encountered anomalies of the craniofacial region
- The genetic basis of non-syndromic clefting is complex and poorly understood. Environmental factors are involved in a proportion of cases
- Antenatal diagnosis of lip clefts can be expected in about two-thirds of cases. Palatal clefts cannot be diagnosed before birth other than with magnetic resonance scanning. Prenatal diagnosis carries with it a responsibility for rapid access to specialist counselling and advice. Outcome following such support is very good
- The spectrum of cleft types is considerable. Incomplete and forme fruste conditions include submucous clefting of the palate, which is frequently missed in postnatal examinations
- The triad of notched posterior hard palate, bifid uvula, and midline zona pellucida are diagnostic of submucous cleft palate, which should be referred for specialist opinion
- The role of pre-surgical orthopaedics to improve surgical repair of wider cleft lips remains controversial. It has no beneficial effect on feeding, but many surgeons value the outcome
- There is little (if any) role for surgical intervention in early severe Pierre Robin sequence. Nasopharyngeal airway management is highly successful in support during the early months
- Cleft care has been centralised into less than 10 units in the UK. This has enabled coordinated nursing, surgical, and other specialist service provision, with a rising of standards since instigated. All newly diagnosed or suspected clefts and related conditions should be referred to these teams as early as possible
- Optimal surgical technique remains elusive. However, radical palatal muscle repositioning has improved speech outcomes considerably. Mid-facial growth disturbance remains the most common long-term adverse outcome of surgical intervention
- Comprehensive and ‘holistic’ cleft care involves mandatory clinical psychology support of children and their families. Unnecessary surgical interventions can be reduced, and long-term global health outcomes are improved