50 Developmental abnormalities of the face, mouth

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Aetiology

Aetiology

Non-syndromic cleft lip and/or palate may present as new diagnosis within a family or with a clear family history . A family history of cleft lip and palate in which a first-degree relative is a ff ected increases the risk of subsequent cleft cases in the family , supporting the theory that there are underlying genetic mutations contributing to the aetiology . Isolated cleft palate is more commonly associated with a syndrome than cleft lip and palate and isolated cleft lip. Over 150 named syndromes are associated with cleft lip and palate, although Stickler (ophthalmic and musculoskeletal abnormalities), DiGeorge (cardiac/thymic anomalies), Down, Apert and Treacher Collins syndromes are most frequently encountered. aetiology of cleft lip and/or palate, including maternal epilepsy (and associated medication) and drugs (e.g. steroids, diazepam, sodium valproate and phenytoin). The role of antenatal folic acid supplements in preventing cleft lip and/or palate remains equivocal. Pierre Robin sequence is a condition worth considering in specific terms. This sequence comprises isolated cleft palate, retrognathia and a posteriorly displaced tongue (glossoptosis), w hich is associated with early airway and feeding di ffi culties. Although airway obstruction does not commonly occur in babies with a non-syndromic cleft lip and/or palate, in babies with an airway obstruction, e.g. Pierre Robin sequence hypoxic episodes during sleep and feeding can be life-threat ening. Intermittent airway obstruction is more frequent and is managed conservatively . In more severe cases the children will often require adjunctive support for their airway compro mise suc h as supplemental oxygen, nasopharyngeal airway and even tracheostomy . More controversially , surgical adhesion of the tongue to the lower lip (labioglossopexy) in the first few da ys after birth is an alternative but less commonly practised method of management. Mandibular distraction surgery has advocates but numerous attempts at developing a consensus view in support of this procedure for airway compromise in cleft have been unsuccessful. Summary box 50.2 Aetiology of cleft lip and palate /uni25CF /uni25CF /uni25CF

The cause of cleft lip and/or palate is multifactorial Most cases occur without a clear family history or known risk factors Clefts can be associated with many craniofacial/medical syndromes

Age 1–7 years early years care follow-up

Age 1–7 years: early years care/ follow-up

Following primary surgery , regular review by an MDT is essential. Many aspects of cleft care require review during the early years of childhood: /uni25CF hearing; /uni25CF speech; /uni25CF dental development; /uni25CF wound healing and aesthetics. Hearing Eustachian tube dysfunction plays a central role in the patho -). genesis of otitis media with e ff usion (OME) in babies and children born with a cleft palate. Children with a cleft lip alone exhibit the same frequency of otitis media as their age-matched counterparts, whereas those c hildren with palatal involvement may have an increased incidence of OME. Regardless of the hearing is within normal limits. All children are screened at birth, but those who have a cleft palate are monitored regularly with audiological screening. Speech In the UK, specialist SLTs involved in cleft care engage at an early stage with families and children. Some teams will run group sessions to encourage speech development. Speech is constantly monitored during development and early interven tion is advisable if speech pathology is suspected or diagnosed. The problems that may present can be considered in two broad groups: 1 Velopharyngeal incompetence (VPI). Where the soft palate fails to achieve adequate velopharyngeal clo sure, which is required for certain sounds in speech, air escape occurs, leading to the resonance issue of hyper nasality . This can lead to unintelligible speech because of either the hypernasality itself or the adaptations made by the child in an attempt to achieve velopharyngeal closure. 2 Articulation errors . These either arise as a compen satory mechanism, as stated above to overcome VPI, or, less commonly , are caused by jaw/dental and occlusal abnormalities. To investigate these problems, the cleft team relies upon the specialist SLT assessment and investigations such as lateral videofluoroscopy and nasendoscopy . These investigations ar used to visualise the palate as it moves in real time during speech. Secondary speech surgery may be o ff ered when there are structural issues to overcome such as VPI. Cleft palate repair is carried out when the palatal function is assessed to be sub optimal but other procedures that alter the dynamics of airflow during speech to reduce nasal escape may also be employed. These interventions are broadly termed pharyngoplasty procedur es. Dental Dental anomalies are common findings in children with cleft lip and/or palate. Various phenomena, including delayed tooth development, delayed eruption of teeth and morpho logical abnormalities, are well documented. The number of teeth may be reduced (hypodontia) or increased (hyperdontia), occurring most commonly in the region of the cleft alveolus and involving the maxillary lateral incisor tooth. These abnor malities can occur in both primary and secondary dentition. All children with cleft lip and palate should undergo regular dental examination. Dental management should also include preventive measures such as dietary advice, fluoride supple ments and fissur e sealants. A well-maintained and disease-free dentition in childhood provides the optimal situation for successful orthodontic treatment. Wound healing/aesthetics Wound infections are rare but if they occur may lead to revi sion surgery . If this is a lip wound infection then revision

can be timed to be pre-school or, if the problem is subtle, the lip can be revised opportunistically at the time of, for example, ABG. ryngoplasty as described earlier in Speech .

Age 7–12 years late childhood care follow-up

Age 7–12 years: late childhood care/ follow-up

Alveolar bone grafting - ABG is a key surgical intervention for patients with alveolar involvement. The procedure can be carried out at the same time as primary cleft lip surgery and is defined as primary bone grafting. More commonly the procedure is a separate surgical intervention later in development. In this case the term secondary bone grafting is used. Secondary bone grafting is - timed in relation to the development of the underlying adult dentition in the region of the cleft. Dental development can - be assessed radiographically and the optimal window for bone grafting is thus easily defined. The lateral incisor tooth is commonly absent or diminutive but, if present and of normal morphology , the bone graft can be timed around the root - development of this tooth (often described as early secondary grafting at age 5–7 years). The canine tooth is most commonly used in assessment and timing. The optimal timing for intervention is at the point when the canine root is one-half to two-thirds formed (often described as late secondary grafting at age 8–11 y ears). As e there is wide variation in the rate of dental development it is better to assess each patient and their dentition on an individ - ual basis and tailor the treatment to this. Patients may undergo a short period of orthodontic treat - ment prior to bone grafting. Less than 50% of patients with - UCLP will require this. When carried out, the aim is to expand the alveolar cleft to improve surgical access. Occasionally the adjacent teeth may be aligned in advance of surgery if they are interfering with access. It is vital in a bilateral cleft to be able to stabilise the mobile anterior (premaxillary) segment to facilitate bone healing. Adjunctive secondary procedures can be carried out simultaneously , e.g. cleft lip revision. The success rate of ABG is high. There are a variety of scoring systems used to measure outcome. Close teamwork - between the cleft surgeon and the orthodontist is vital. In the situation where there is significant hypodontia in the region of the cleft a decision may be taken not to perform ABG. In this case the missing teeth can be replaced with a variety of - restorative options, including a denture, an adhesive bridge or an implant-retained prosthesis. Bone grafting will be required for implant placement but this is better carried out when the patient is skeletally mature. - The primary objectives when performing ABG are to: /uni25CF provide adequate bony support for the adult teeth to enable subsequent orthodontic alignment; /uni25CF enable the eruption of adult teeth into the line of the dental arch; /uni25CF stabilise the premaxilla in bilateral clefts; - /uni25CF definitively close the residual alveolar cleft. The secondary objectives or associated benefits may include aesthetic improvements to the nasolabial region.

Figure 50.9 Nasal asymmetry.

CLASSIFICATION OF CRANIOFACIAL ABNORMALITIES

CLASSIFICATION OF CRANIOFACIAL ABNORMALITIES

- van der Meulen and his colleagues proposed a classification system that has significant utility in helping to understand the variety and complexity of craniofacial malformations. This classification considers the embryological development of the - craniofacial region. First, in terms of the formation and fusion of the processes (branchial arches): the failure of the fusion

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the failure of fusion between the frontonasal process and the maxillary process results in a cleft lip, either unilaterally or bilaterally . Second, in the formation of bone and cartilage; if this is abnormal it is termed dysostosis or dyschondrosis. Third, the formation and growth at the sutures between the various bones of the craniofacial skeleton: premature fusion leads to synostosis. Superimposed on this concept is the consideration of the development of the central nervous system; this leads to a number of types of abnormality , as outlined in Table 50.1 In addition, and in common with all classification systems, there is another large group of conditions that do not sit within the system outlined above and also listed in Table 50.1

CLEFT LIP AND PALATE

CLEFT LIP AND PALATE

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Classification of cleft

Classification of cleft

Cleft lip and/or palate presents in a heterogeneous manner. In simple terms these conditions can be divided into two clinical types (phenotypes): - 1 isolated cleft palate; 2 cleft lip with or without involvement of the alveolus (tooth-bearing portion of the jaw) or palate. Within these broad classifications a variety of combinations of cleft type can exist. These all aim to define the extent and laterality of the cleft (left/right/bilateral) (Figure 50.2). This - information is both diagnostic and, increasingly , of prognostic value. Many have argued for a single classification system to be Victor Veau , 1871–1949, French surgeon and author of several books on cleft lip and cleft palate surgery . Gunnar B Stickler , 1925–2010, born in Germany , Chair of Section of Paediatrics and later Paediatric Cardiology , The Mayo Clinic, Rochester, MN, USA. Angelo Mario DiGeorge , 1921–2009, Italian American physician and pediatric endocrinologist. John Langdon Haydon Down (sometimes given as Haydon-Down), 1828–1896, physician, The London Hospital, London, and Superintendent, Earlswood Asylum for Idiots, Surrey , UK, described this syndrome in 1866. adopted: even when the same system is being used, clinicians may interpret the findings inconsistently . In the UK, national audit data are collected for outcomes in unilateral cleft lip and palate (UCLP), thus allowing intercentre comparison. Cleft lip and/or palate is more common in males, whereas isolated cleft palate is more common in females. In UCLP the condition a ff ects the left side in 60% of cases. Summary box 50.1 Overview of cleft lip and palate /uni25CF /uni25CF /uni25CF /uni25CF

(a) Figure 50.1 (a) Unilateral cleft lip. (b) Bilateral cleft lip. (c) Isolated cleft palate. (a) L L Lip Lip A A Alveolus Alveolus Left Right H Hard palate S Soft palate (b) Class Description I Soft palate only II Hard and soft palate to the incisive foramen III Complete unilateral of soft, hard, lip and alveolar ridge IV Complete bilateral of soft, hard and/or lip and alveolar ridge Figure 50.2 (a) The LAHSHAL code. (b) The Veau classi /f_i cation system. (b) Cleft lip and/or palate has two main phenotypes Cleft palate is more common in females and cleft lip and/or palate is more common in males The incidence/prevalence demonstrates geographical variation There are simple classi /f_i cation systems that describe phenotype

Cleft lip and or palate embryology and pathogenes

Cleft lip and/or palate: embryology and pathogenesis

Embryologically , the lip and palate are derived from facial prominences/processes. 1 The lip/nose complex is derived from a mixture of the median nasal process and the maxillary processes. 2 The primary palate is derived from the median nasal pro cess and consists of all anatomical structures anterior to the incisive foramen, namely the alveolus and philtral por tion of the upper lip. The remainder of the lip is derived from the maxillary processes. 3 The secondary palate is derived from the maxillary pro cesses and is defined as the remainder of the palate behind the incisive foramen, which is divided into the hard palate and, more posteriorly , the soft palate. Cleft palate results in failure of fusion or descent of the two palatal shelves. This failure to descend, fuse or remain fused can result in a cleft a ff ecting any part of the palate. Pierre Robin , 1867–1950, Professor, The French School of Dentistry , Paris, France, described this syndrome in 1929. , - - Clinical anatomy The muscle chains of the face are shown in Figure 50.3 . Their disruption in unilateral cleft lip is shown in Figure 50.4 . Summary box 50.3 Embryology and pathogenesis of cleft lip and/or palate /uni25CF /uni25CF /uni25CF

1 A 2 3 4 B 5 6 7 8 C 9 Figure 50.3 The muscle chains of the face: frontal view. The nasal cartilages are represented in blue. A, nasolabial (muscles 1–3); B, /uni00A0 bila

bial (muscles 4–6); C, labiomental (muscles 7–9); 1, transverse nasalis; 2, levator labii superioris alaeque nasi; 3, levator labii superioris; 4, /uni00A0 orbicularis oris (oblique head) – upper lip; 5, orbicularis oris (hori

zontal head) – upper lip; 6, orbicularis oris – lower lip; 7, depressor anguli oris; 8, depressor labii inferioris; 9, mentalis. Clefts occur at the points of fusion of facial processes Normal anatomical structures are displaced and disrupted Abnormal muscle insertion results in aesthetic and functional sequelae

Cleft lip nose revision

Cleft lip/nose revision

Indications for revisional surgery to a previously repaired cleft lip are dependent on the site and severity of the residual deformity . Relative indications for lip revision include: /uni25CF misaligned vermilion; /uni25CF lip asymmetry . Relative indications for residual nasal deformity include: /uni25CF incorrect alar base position; /uni25CF poor nasal tip projection; /uni25CF deviation of cartilaginous nasal septum into the non-cleft nostril. Residual nasal deformity is an external manifestation of incomplete reconstruction of the nasolabial muscle ring (see Clinical anatomy). It is thought less than ideal to sur gically interfere with the nasal septum in the growing child. Minor adjustments are possible before the age of 14–15 year (Figures 50.9 and 50.10), but more major nasal surgery is usually delayed until after this age. Open septorhinoplasty may be considered for definitive surgical nasal correction. In patients with cleft lip and palate, open surgery is preferred to gain access to the external cartilag inous framework, w hich is frequently a ff ected by the primary issues of muscle attachment related to the cleft. One common feature is collapse of the lower lateral cartilage on the cleft side together with a dislocation of the cartilaginous septum into the non-cleft nostril. The open method ensures adequate access and repositioning of the cartilaginous framework as a tertiary procedure to improve nasal tip projection, correct sep - tal deformity and relocate alar cartilages. Grafting techniques are often employed using harvested septal (nasal) cartilage or conchal (ear) cartilage.

Figure 50.10 Nasal asymmetry addressed by open surgical revision.

Cleft lip

Cleft lip

The abnormalities in cleft lip are the direct consequence of disruption of the muscles of the upper lip and nasolabial region. The muscle continuity is disrupted, leading to the cleft lip and also abnormal insertions of the muscle at the cleft edge. - The e ff ect of this can be seen on the nasal septum and the nose itself. - Unilateral cleft lip In the unilateral cleft lip, the muscle rings are disrupted on - one side, resulting in an asymmetric upper lip and/or nose. This involves the external nasal cartilages, nasal septum and anterior maxilla (premaxilla). This influences the mucocuta - neous tissues, causing a displacement of nasal skin onto the lip and a retraction of labial skin, as well as changes to the vermil - ion and lip mucosa. All these changes need to be considered in planning the surgical repair of the unilateral cleft lip. (a) Bilateral cleft lip In the bilateral cleft lip the disruption is greater but often symmetrical. Muscular continuity is disrupted bilaterally , producing a flaring of the nose (caused by a lack of nasolabial muscle continuity), a protrusive premaxilla and an area of skin in front of the premaxilla devoid of muscle, known as the prolabium. As in the unilateral cleft lip, the muscular, carti laginous and skeletal deformities influence the mucocutaneous tissues, which must be respected in planning the repair of the bilateral cleft lip.

A B C (b) Figure 50.4 (a) Schematic representation of disruption of the nasola bial and bilabial muscle chains in unilateral (right) cleft lip. A, nasola bial; B, bilabial; C, labiomental. (b) Unilateral cleft lip before muscular reconstruction (courtesy of William P Smith).

Cleft palate

Cleft palate

Embryologically , the primary palate consists of all anatomical structures anterior to the incisive foramen, namely the alveolus and upper lip. The secondary palate is defined as the remainder of the palate behind the incisive foramen, divided into the hard palate and, more posteriorly , the soft palate. Cleft palate results in failure of fusion of the two palatine shelves. This failure may be confined to the soft palate alone or involve both hard and soft palate. When the cleft of the hard palate remains attac hed to the nasal septum and vomer, the cleft is termed incomplete. When the nasal septum and vomer are completely separated from the palatine processes, the cleft palate is termed complete. Soft palate In the non-cleft soft palate, closure of the velopharynx, which is essential for normal speech development, is achieved by elevation of the soft palate. Although this is achieved by coor - dinated muscular activity , it is the levator veli palatini that is the key muscle in achieving this. In general, the muscle fibres of the soft palate are orientated transver sely with no significant attachment to the hard palate. In a cleft palate the muscle fibres are orientated in an anteroposterior direction, inserting into the posterior edge of the hard palate. Hard palate The hard palate can be divided into three anatomical and physiological zones (Figure 50.5). The central palatal fibromu - cosa is very thin and lies directly below the floor of nose. The maxillary fibromucosa is thick and contains the greater palatine neurovascular bundle. The gingival fibromucosa lies mor e lateral and adjacent to the teeth. In performing surgical closure of a cleft palate, the changes associated with the cleft must be understood to obtain an anatomical and functional repair. In complete cleft palate the median part of the palatal vault is absent and the pala - tal fibromucosa is reduced in size. T he maxillary and gingival - fibromucosa are not modified in thickness, width or position.

1 2 3 Figure 50.5 The three mucosal zones of the hard palate. 1, palatal /f_i bromucosa; 2, maxillary /f_i bromucosa; 3, gingival /f_i bromucosa.

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DIAGNOSIS

DIAGNOSIS

The diagnosis of the craniofacial anomalies has, in recent years, undergone a massive change on two fronts: first, advances in ultrasonography have increased the rate of prenatal diagnosis and impacted management significantly; second, the rapid expansion in genetic understanding has led to many more mutations being linked to particular phenotypes. Despite these advances the diagnosis of the majority of these conditions remains clinical.

EPIDEMIOLOGY

EPIDEMIOLOGY

The incidence of congenital craniofacial anomalies varies in di ff erent parts of the world and is often not easy to quantify . Table 50.2 outlines the various incidences of the more common craniofacial abnormalities.

TABLE 50.2 Approximate incidence data from multiple sources. Condition Incidence Apert syndrome 1 in 100 /uni00A0 000 Pfeiffer syndrome 1 in 100 /uni00A0 000 Crouzon syndrome 1 in 62 /uni00A0 500 Treacher Collins syndrome 1 in 50 /uni00A0 000 Unicoronal synostosis 1 in 10 /uni00A0 000 Metopic synostosis 1 in 7000 Sagittal synostosis 1 in 5000 Hemifacial microsomia 1 in 3500 Neuro /f_i bromatosis 1 in 2600 Cleft lip and palate 1 in 600

Immediate neonatal care

Immediate/neonatal care

- Feeding Babies born with a cleft involving the palate will feed well and thrive, provided that they receive the appropriate CNS input. The feeding aids for a child with a cleft palate aim to improve the e ffi ciency of delivery of milk, reducing the e ff ort of feeding. Expressed breast milk is best. A range of modified bottles and teats are available. Soft bottles allow the parents to do much of the work of milk delivery for the child by synchronising their ‘squeeze’ to the baby’s ‘suck’. Feeding plates, constructed from a dental impression of the upper jaw , were used in the past in the UK and may still be used in other parts of the world. In some units, babies are provided with an active plate that aims not only to improve feeding but also to reduce the width of the cleft and improve the shape of the nose prior to surgery – nasoalveolar moulding (NAM). The e vidence in the literature of long-term benefit using such a regime is conflicting. - Summary box 50.4 Immediate/neonatal care for a patient with a cleft and/ - or palate /uni25CF /uni25CF /uni25CF - -

Babies born with a cleft may have issues with feeding and airway A team of clinicians is required to meet all the needs of a child with a cleft Most of the care delivered to a child with a cleft lip and/or palate is non-surgical in the initial phase

Incidence

Incidence

The incidence of cleft lip and/or palate is around 1:600 live births. There are geographical and ethnic variations, with a higher incidence among the South East Asian and Native American populations than elsewhere in the world. The accu - racy of these figures may be questionable owing to a variance in reporting and healthcare infrastructure. The typical distribution of cleft types is: /uni25CF cleft lip alone: 15% (Figure 50.1a,b); /uni25CF cleft lip and palate: 45%; /uni25CF isolated cleft palate: 40% (Figure 50.1c).

Introduction

INTRODUCTION

Congenital abnormalities of the head and neck are complex and often confusing. For these reasons it is helpful to have a classification system that helps to understand the variety of conditions. For any classification system to be useful it should ideally help to explain the aetiology and pathogenesis of the abnormality and to determine treatment. For these multi faceted and multifactorial conditions an ideal classification system is not available. Consequently , there are a number of di ff erent systems available: some are purely descriptive (e.g. Tessier' s classification of clefts), while others apply only to single conditions, such as the OMENS (O, orbital abnormal ities; M, mandibular deformity; E, ear deformity; N, nerve /uni25CF /uni25CF /uni25CF Paul Tessier , 1917–2008, French maxillofacial surgeon, considered the 'father of modern craniofacial surgery'. Jacques C H van der Meulen , 1929–2017, Professor in Plastic and Reconstructive Surgery , Erasmus University , Rotterdam, The Netherlands. Kar l Heinz Binder , 1923–2016, German dentist, documented the facial features of three children with the condition that now bears his name. Louis Edouard Octave Crouzon , 1874–1938, neurologist, Paris, France, described this syndrome in 1912. Eugene Apert , 1868–1940, physician, L'Hôpital des Infants Malades, Paris, France, described this syndrome in 1906. Rudolf Arthur Pfei ff er , 1931–2012, geneticist, Münster, Germany , described this syndrome in 1964. involvement; and S, soft-tissue abnormalities) classification of hemifacial (craniofacial) microsomia, which has utility in instituting treatment protocols.

TABLE 50.1 Types of developmental abnormalities of the face, mouth and jaws. Type Examples Cerebrocranial dysplasias Anencephaly, microcephaly Cerebrofacial dysplasias Rhinencephalic and oculo-orbital dysplasias Craniofacial dysplasias with clefting Lateronasomaxillary, medionasomaxillary, intermaxillary, maxillomandibular clefting Craniofacial dysplasias with dysostosis Sphenoidal, sphenoidal frontal, frontal, frontofrontal, frontonasoethmoidal, internasal, nasal, premaxillomaxillary, nasomaxillary, maxillozygomatic, zygomatic, zygoauromandibular, temporoauromandibular, mandibular, intermandibular Craniofacial dysplasias with synostosis Craniosynostosis: lambdoid and sagittal Craniofaciosynostosis: metopic, coronal, bicoronal Faciosynostosis: vomeropremaxillary (Binder syndrome) Craniofacial dysplasias with dysostosis and Crouzon, Apert and Pfeiffer syndromes synostosis Craniofacial dysplasias with dyschondrosis Achondroplasia After van der Meulen JC, Mazzola R, Vermey-Keers C et al. A morphogenetic classi /f_i cation of craniofacial malformations. 1983; 71 (4): 560–72. In more depth the epidemiology, pathogenesis and • management of cleft lip and palate

Introduction

Cleft lip and/or palate is the most common congenital abnormality a ff ecting the orofacial region. These conditions most commonly occur as isolated deformities but can also be associated with other medical conditions, e.g. congenital heart disease. They are also an associated feature in over 300 recognised syndromes. All children born with a cleft are screened for other congenital

abnormalities. Where the cleft is thought to be associated with a syndrome any appropriate further investigations, including genetic counselling, will be organised.

Learning objectives

Learning objectives

To understand: The range and complexity of craniofacial anomalies • The principles driving interventions for the developing • child with a craniofacial anomaly

MANAGEMENT

MANAGEMENT

In considering the management of this vast range of heteroge neous congenital abnormalities it is very di ffi cult to generalise about management protocols. The vast majority of manage ment is delivered by multidisciplinary teams (MDTs) within specialist centres. Edward Treacher Collins , 1862–1932, ophthalmic surgeon, Royal London Ophthalmic Hospital and Charing Cross Hospital, London, UK, described this syndrome in 1900. There have been a few reported cases of prenatal surgery when a diagnosis was made or suspected prenatally . However, these procedures remain at present experimental; in general, the options open are for termination or best supportive care in preparation for the birth. This can often provide the parents with a period of time to adjust to the impending birth of a child with additional demands and needs. The opportunity to meet parents, adults and children who have experienced . the same condition is often very valuable. Termination and its therapeutic uses is obviously a contentious and very personal issue. However, some parents may request this for very treat - . able conditions (e.g. isolated cleft lip); in these circumstances, the local ethics board must be involved and ultimately on occasions the advice of the courts must also be sought.

Management in early childhood (1–12 years)

Management in early childhood (1–12 years)

In early childhood management should be aimed at dealing with functional problems – airway obstruction, speech and feeding issues – but there is an increasing imperative for surgery to address the appearance of the child. There is no doubt that visible di ff erences can a ff ect a child’s development, both socially and emotionally; however, there is a significant role for psychological and emotional support for the whole family and in some cases for the school community to help the child, family and school understand and deal with the additional pressures that visible di ff erence makes. Surgery can make a significant di ff erence for some patients, but for many surgery should be delayed as long as possible for an optimal outcome in the long term. In the older child airway issues can become a problem and their identification is more di ffi cult. The usual presentation is of sleep apnoea, which often has an insidious onset; the his tory should be actively sought as parents may be accustomed to noisy snoring and daytime tiredness in the child and may not consider it abnormal. Initial investigation is with a home overnight oxygen saturation monitor, w hich, if abnormal, should trigger a comprehensive sleep study . The management of obstructive sleep apnoea includes the use of tonsillectomy/ adenoidectomy , midface advancement and mandibular dis traction as well as a variety of ventilator support devices.

Management in infancy (0–12 months)

Management in infancy (0–12 months)

At this age treatment falls into two categories: that directed at - major functional issues as for neonatal care and that directed at skull surgery in cases of craniosynostosis. same; however, there is the possibility of surgery to advance the mandible in the severely retrognathic patient. This can be used to obviate the need for a tracheostomy or to allow for early decannulation. The most e ff ective technique is distrac tion osteogenesis (or distraction histogenesis), which utilises the same basic principles as in limb lengthening. The bone is cut and a device placed across the osteotomy site; after a short latent period the bone ends are gradually separa ted, dis tracting the callus. In the mandible, unlike the long bones, it is not necessary to limit the bone cut to the cortex (corticotomy) and a complete osteotomy is used. The technique allows for a lengthening of approximately 1 /uni00A0 mm/day , after which ther retention period to allow for consolidation of the callus. Craniosynostosis results in premature fusion of one or more of the skull sutures. The conditions may be isolated or part of a syndrome. This can result in abnormalities of both the skull and, particularly in syndromic cases, the facial skel eton. In 10–20% of single-suture cases and a higher propor tion of syndromic multisuture cases the infants develop raised intracranial pressure, which presents as episodes of distress, listlessness and disturbed sleep . This may be associated with papilloedema and, untreated, can lead to visual failure. The diagnosis is confirmed with intracranial pressure monitoring. Some congenital lesions may obstruct the vision of one or both eyes and this type of problem needs to be addressed to minimise the chances of amblyopia developing. An example of this would be the development of a large true haemangioma of the eyelids threatening to obscure the child’s vision out of one eye.

Neonatal management

Neonatal management

In the neonatal period management is aimed at addressing the urgent issues relating to the airway , breathing, eye protection and establishing feeding. In many of the craniofacial conditions the airway can be a ff ected and may be fully or partially obstructed. This may be because of a retropositioned hypoplastic maxilla – the tongue falling back to close o ff the upper airway; this is often com - pounded b y a hypoplastic mandible. The trachea itself may also be abnormal and tracheomalacia can lead to respiratory problems. Neonates are obligate nasal breathers and some forms of nasal obstruction can pr ecipitate airway symptoms. In the most severe cases intubation is not possible as a result of the abnormal anatomy and a tracheostomy may be necessary . In emergency situations it may be helpful to nurse the baby prone, allowing the tongue to fall forwards. In some cases, particularly the syndromic craniosynostoses such as Apert syndrome, Pfei ff er syndrome or Crouzon syndrome, the combination of midface retrusion and brachycephalic forehead shape can lead to severe exorbitism. In the worst cases this can cause ocular dislocation with the eyelids closing behind the globe. In severe exorbitism the eyelids do not close adequately to moisturise and protect the cornea; without intervention this may lead to irreversible corneal damage. In neonates with airway embarrassment, even without ana - tomical abnormalities, the e ff ort of breathing can be exhaust - ing and this can significantly compromise the ability to feed. Structural anomalies can also a ff ect the ability to feed; expert input from a specialist feeding nurse is often helpful. The use of specialised teats may be helpful but in some cases naso- or orogastric feeding may be necessary . -

Orthodontic treatment

Orthodontic treatment

Children with cleft lip and alveolar involvement will often benefit from orthodontic treatment. Orthodontic treatment is commonly carried out in two phases: 1 Mixed dentition (8–10 years): to prepare the alveolar cleft for ABG (see Alveolar bone grafting). 2 Permanent dentition (12–18 years): to definitively align the dental arches, aiming for a normal functioning occlusion. This phase of treatment may be linked to preparation for orthognathic surgery (jaw alignment surgery).

Orthognathic surgery

Orthognathic surgery

Impaired growth of the midface (maxilla) is a consequence of a number of factors, which are poorly understood. Genetic factors as well as local factors following primary surgery may be involved. Elective maxillary advancement or bimaxillary surgery may be indicated to restore aesthetics and dental occlusal harmony . Orthognathic surgery is usually performed when facial growth is complete (16–17 years in female patients, 17–19 years in male patients). The principal dentofacial deformity associated with cleft lip and palate is underdevelopment in both the horizontal and vertical direction of the maxilla. This jaw size discrepancy can be corrected with orthognathic surger y (Figure 50.11).

PRINCIPLES OF CLEFT SURGERY

PRINCIPLES OF CLEFT SURGERY

The ultimate aim in cleft lip and palate management is to facilitate normal development and well-being. In seeking this, surgical repair is aimed at producing normal anatomy in the lip, nose and palate. Essentially , oral and dental health should - also be optimised in the management. Key outcomes measured include speech, facial growth, general well-being and dental health. With the exception of rare conditions such as holopros - - encephaly , there is no true hypoplasia of the tissues involved on either side of the cleft. There is, however, displacement, deformation and underdevelopment of the muscles and facial skeleton. Emphasis is placed on muscular reconstruction of the lip, nose and face as well as muscles of the soft palate. Normal or near-normal anatomy promotes normal func tion, thereby encouraging normal growth and development of lip, nose, palate and facial skeleton. An in-depth understanding of the anatomy of the cleft is invaluable if the surgeon is to achieve normal, or near -normal, anatomical reconstruction.

Figure 50.6 Postoperative unilateral cleft lip repair. Figure 50.7 Postoperative bilateral cleft lip repair.

SECONDARY REVISION SURGERY

SECONDARY/REVISION SURGERY

These procedures are undertaken to improve aesthetics and/ or function. They may be considered as procedures that were unplanned at the time of primary surgery . Specific examples are as follows.

SUMMARY

SUMMARY

Cleft care has been the subject of significant reorganisation in recent years. Coordinated care is provided in most countries by MDTs. Specific training pathways exist in many countries for cleft surgery . Better collection and collation of outcome data will drive evidence-based improvements in care and service development. Summary box 50.6 - Summary of care for patients with cleft lip and/or palate /uni25CF s /uni25CF /uni25CF /uni25CF - /uni25CF

Cleft surgery in infants is time sensitive Aesthetic and functional outcomes are important and are measured Surgery involves restoration of muscle position to as close to normal as possible Planned surgery includes bone grafting in children with alveolar involvement Revision/secondary surgery optimises aesthetic and functional outcomes

(b) Figure 50.11 (a) Pro /f_i le of a class III skeletal relationship and maxillary hypoplasia and mandibular prognathism. (b) Lateral skull radiograph. (c) Pro /f_i le following bimaxillary osteotomy. (d) Postoperative radio graph following bimaxillary osteotomy demonstrating internal /f_i xation.

(e) Schematic representation of bimaxillary osteotomy with maxillary advancement and mandibular retrusion (courtesy of William P Smith). (d) (e)

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Surgical techniques

Surgical techniques

Much debate and variation exist across the world in the timing and techniques employed in cleft repair. All have the common aims stated above. Restoration of form and function can be achieved using many of these protocols, but the following protocol is that which is used in the UK and was popularised in Norway . /uni25CF Cleft lip/nose and anterior palate repair is performed between 3 and 6 months of age (Figures 50.6 and 50.7 /uni25CF The anterior palate closure is achieved by using a single-layer mucosal flap from the vomer. The lip is closed using a variety of described techniques but most surgeons believe that the muscle repair is more important than the skin incision, hence the variation. Bartolomeu Eustachio (Eustachius) , 1513–1574, Professor of Anatomy , appointed physician to the Pope in 1547. /uni25CF Definitive cleft palate repair is carried out between 6 and 12 months. There is conflicting evidence within the pub - lished literature relating to optimal timing of palate repair. The principle applied in the UK is that of closure during the early stages of speech development. /uni25CF The most common surgical approach in cleft palate repair is the intravelar veloplasty (IVVP), in which incisions along the cleft edge provide access to the soft palate muscle. The levator muscles are dissected free (Figure 50.8) and su - tured together in the midline to recreate a muscular sling. Summary box 50.5 Primary surgery for cleft lip and/or palate /uni25CF /uni25CF /uni25CF -

Figure 50.8 Dissection of the levator muscles. Treatment staged from anterior (lip) to posterior (soft palate) in the UK Multiple eponymous skin incisions for lip repair but muscle reconstruction is key Management of the levator sling is key in cleft palate repair

The cleft multidisciplinary team and primary manag

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The cleft team Modern cleft services rely on well-coordinated patient path - ways. The pathways and protocols may vary from country to country but the aims of treatment are consistent. Care is with cleft lip and/or palate has appropriate access to the correct clinician and care at the optimal time. In the UK, most children with a cleft involving the lip are diagnosed antenatally . Scanning protocols now include an ‘anomaly scan’ at around 20 weeks. Isolated cleft palate cannot be diagnosed antenatally using routine scanning techniques. Some researchers have suggested that Doppler studies may help in diagnosing isolated cleft palate. When an antenatal diagnosis is confirmed, referral to a cleft team is appropriate. Clinical nurse specialist involve ment would commence from this point onwards. The cleft MDT therefore has a range of clinical expertise and specialisms within it. These are: /uni25CF Cleft coordinator/administrator . This is vital to ensure that patients and families have clinical episodes organised as per the protocol of the service. Responsive administrative support is vital for patients, families and cli nicians. /uni25CF Clinical nurse specialist (CNS). The role of the CNS is central to the safe and e ff ective delivery of cleft care. These clinicians will, in most cases, be the first clinical contact with the team. The CNS will assess the child and provide initial support to the family . Assessment of feeding, airway and general well-being is carried out. The role of the CNS is vital in ensuring that each child is optimally prepared for surgery . /uni25CF Paediatrician . Most children who have a cleft will be otherwise well. In some cases there may be associated or coexisting medical problems, e.g. cardiac or respiratory . These will require appropriate specialist input and perhaps coordination of care by a paediatrician. /uni25CF Speech and language therapist (SLT). The input of an SLT is vital where palatal involvement exists in the cleft type. Assessment and therapy are provided where re quired. Outcome measurements and diagnosis of palatal dysfunction are key elements of the SLT’s role in cleft care. /uni25CF Ear–nose–throat (ENT)/audiology . Regular hear ing tests and e ff ective intervention for hearing loss are vital in ensuring speech development. This is a key part of early cleft care. /uni25CF Paediatric dentist . Traditionally dental/oral health has been poor for this patient group. A greater emphasis on disease prevention has resulted in much improved den tal outcomes. A key part of early health care would involve a paediatric dentist. /uni25CF Orthodontist . The role of the orthodontist varies in dif ferent services. Some services will have early orthodontic intervention to mould the anterior cleft presurgically . This is not undertaken in many countries, e.g. the UK. The orthodontist, therefore, becomes a key figure at around 7 years of age as the child enters the early ‘mixed denti tion’ phase. Assessment and preparation for alveolar bone grafting (ABG) as well as definitive orthodontic alignment are undertaken where required. The orthodontist is a key member of the team delivering orthognathic (jaw align ment) sur gery at the point of skeletal maturity if required. Christian Johann Doppler , 1803–1853, Professor of Experimental Physics, Vienna, Austria, enunciated the ‘Doppler principle’ in 1842. throughout the clinical

pathway , providing support to pa - tients, families and team members. Key outcomes in rela - tion to quality of life are assessed by these clinicians. /uni25CF Cleft surgeon . The cleft surgeon’s role is to provide as - sessment and intervention to patients. The main aim of cleft surgery is to correct the underlying anatomical abnor - malities that can lead to issues with appearance and func - tion. Optimal clinical outcomes can be achieved for most - patients with limited surgical intervention. One to three opera tive interventions (depending on the type of cleft) in childhood are all that would be planned as part of a cleft pathway/pr otocol. Outcomes of surgery/cleft care are au - dited annually in most countries.

maturity

maturity

Airway and other functional issues are usually stabilised by - this time and interventions are aimed at optimising the overall appearance. The transition from primary school to secondary school is often a period of distress for patients with visible di ff erences and their families. If there are pressing psycholog - - ical reasons corrective surgery can be o ff ered, although this is usually best postponed until growth is complete. In general, a comprehensive integrated corrective plan should be de veloped within the MDT . This would usually address the skeletal and e is a dental abnormalities first and then address the soft tissues. The majority of the major craniofacial abnormalities should be managed by a formal MDT . -