Chapter

23 Anaesthesia for Cleft Lip and Palate Surgery in Children*

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Introduction

Successful anaesthesia for cleft lip and palate surgery requires an understanding of the cleft condition, associated abnormalities and the surgery involved. Close cooperation with the surgeon is vital to manage the child's airway during and after surgery. The aim is to provide balanced anaesthesia and optimum analgesia, so a child starts feeding and is able to return to normal activities as soon as possible after surgery.

Incidence, Type of Cleft and Embryology

Cleft lip and palate is a common congenital abnormality with an incidence of one in 700 live births, with around 1,100 affected children born in the United Kingdom each year. It encompasses two distinct conditions: cleft lip with or without cleft palate making up 55% of cases, and isolated cleft palate making up 45% of cases (Figure 23.1). The embryological abnormality is a failure of fusion at four to eight weeks of fetal life of the five facial prominences that form the facial structures. The medial nasal process and lateral maxillary process fuse to form the upper lip and front of the mouth. The embryonic palatal shelves fuse to form the palate– the primary palate anterior to the incisive foramen and the secondary palate posterior to this.

Cleft Lip with or without Cleft Palate

In cleft lip with or without cleft palate (CL +/–P), the anomaly can be a cleft lip alone (CL), a unilateral cleft lip and palate (UCLP) or bilateral cleft lip and palate (BCLP). The condition varies in severity from a small notch caused by a defect in the muscle of the upper lip, called a microform cleft lip, to a wide bilateral cleft lip and palate. Cleft lip is termed complete if it goes to the nasal sill and incomplete if not. A scheme for surgical classification is shown in Figure 23.2.

Cleft lip is more often left-sided and has a male preponderance. It is more common in Chinese people than Caucasians and less common in the African-Caribbean population. The aetiology is both genetic and environmental. The presence of cleft lip with or without cleft palate in a first-degree relative increases the risk of a cleft in a pregnancy to 3–4%. Environmental factors that have been implicated are maternal diabetes, alcohol, folic acid deficiency and drugs, including phenytoin, corticosteroids and diazepam.

Isolated Cleft Palate

Isolated cleft palate (iCP) is less common, accounts for 45% of clefts and has a female preponderance. The genetic risk of recurrence depends on the risk of recurrence of any associated syndrome.

The cleft may be of the soft palate or extend into the hard palate (Figure 23.2). A submucous cleft palate (SMCP) is a defect in the musculature of the soft palate; the palate may look normal, or there may be a bifid uvula or central lucency of the soft palate. A submucous cleft may present with speech abnormality or nasal regurgitation of food.

Velopharyngeal Incompetence without Cleft Palate

A cleft service may encounter several children who do not have a cleft palate but who are referred with velopharyngeal incompetence (VPI). This condition is due to a relative mismatch between the palate and pharynx whereby speech sounds resonate in the nasal space producing, amongst other

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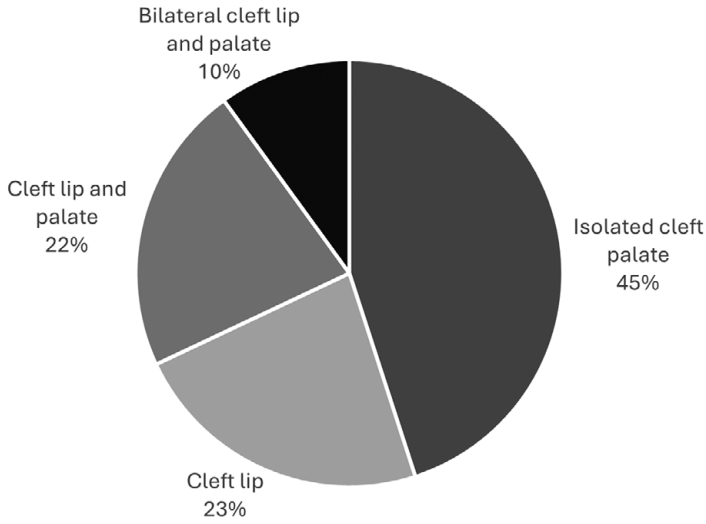


Figure 23.1 The relative incidence of the common cleft types in the United Kingdom.

features, hypernasal speech. These children as a group are more complex. They are more likely than the cleft population to have associated syndromes, such as 22q deletion, other anomalies, feeding issues or learning difficulties. They may have an SMCP or a seemingly normal palate, and the VPI may be due to a large pharynx or neuromuscular weakness or poor coordination within the muscles responsible for speech.

Association between Cleft Conditions and Other Congenital Abnormalities

There is an association between cleft conditions and other congenital abnormalities, especially iCP, which should be borne in mind during anaesthetic assessment. Infants with clefts are more likely to be born preterm with low birth weight. Up to 150 different congenital syndromes have been associated with clefts, and some of the more commonly occurring syndromes are listed in Table 23.1. Syndromes causing mandibular hypoplasia (micrognathia) result in airway problems, and those with associated cardiac defects are of particular relevance to the anaesthetist. Reviews of children with clefts have quoted an incidence of congenital heart disease from 3.7% to 15%, much higher than that in the general population.

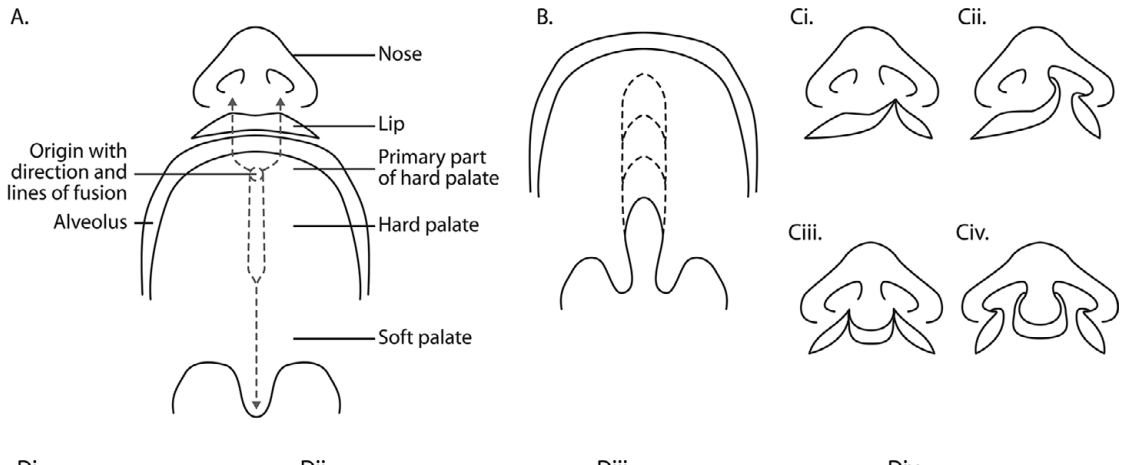
Pierre Robin Sequence

Pierre Robin sequence (PRS) is a combination of cleft palate, micrognathia and glossoptosis (posterior displacement of the tongue) and is present in 20–30% of children with isolated cleft palate. It may be associated with airway obstruction from the neonatal period through to early infancy in severe cases. Disease severity is classified as type 1–3. Babies with type 1 PRS have mild symptoms, and any airway obstruction may be relieved by side-to-side positioning rather than nursing supine. Babies with type 2 PRS have associated feeding difficulties requiring nasogastric feeding. Babies with type 3 PRS have moderate respiratory distress and are managed in the United Kingdom with a nasopharyngeal airway. A minority will require tracheostomy. Airway and feeding problems resolve with age as mandibular growth occurs.

22q11.2 Deletion Syndrome

22q deletion syndrome (previously called velocardiofacial or Di George syndrome) is caused by a microdeletion on chromosome 22. It usually occurs as a new mutation, although in some cases there is autosomal dominant inheritance.

It is the syndrome most commonly associated with clefts, and children may present with isolated cleft palate or submucous cleft palate, causing velopharyngeal incompetence and hypernasal speech. There is a wide spectrum of associated abnormalities, including cardiac defects (interrupted aortic arch, truncus arteriosus, tetralogy of Fallot or pulmonary atresia with VSD), typical facial characteristics of a broad nose and



	Left	Right	Midline
Lip			*
Alveolus			*
Hard palate			
Soft palate	N/A	N/A	

*Very rare

Figure 23.2 Surgical classification of cleft lip with or without cleft palate. Diagnostic findings are recorded in the grid.
Key:
A. Normal anatomy of nose, lip and alveolus and hard and soft palate with the lines of fusion emanating from the incisive foramen.
B. Differing extent of isolated cleft palate.
C. Cleft lip variants: Ci = left unilateral incomplete cleft lip; Cii = left unilateral complete cleft lip; Ciii = bilateral incomplete cleft lip; Civ = bilateral complete cleft lip.
D. Cleft palate variants: Di = left unilateral cleft of alveolus and primary hard palate; Dii = left unilateral cleft of alveolus, hard and soft palate; Diii = bilateral cleft of alveolus and primary hard palate; Div = bilateral cleft of alveolus, hard and soft palate.
Source: Reprinted from Demircioglu M, Kangesu L, Ismail A et al. Increasing accuracy of antenatal ultrasound diagnosis of cleft lip with or without cleft palate, in cases referred to the North Thames London Region. *Ultrasound in Obstetrics and Gynaecology* 2008; 31:649 copyright ISUOG, with permission from John Wiley and Sons.

long face, hypotonia in infancy, defective thymic development and learning disabilities.

Multidisciplinary Care: The Cleft Team

The care of a child with a cleft requires a multidisciplinary approach from a team ideally involving specialist cleft nurses; plastic, maxillofacial and ENT surgeons; orthodontists; speech therapists; dieticians; paediatricians; geneticists; and clinical psychologists. In the United Kingdom, resources have now been concentrated into 11 specialist cleft centres following a review of services in 1998 by the Clinical Standards Advisory Group

Table 23.1 Common syndromes associated with cleft lip and/or palate

Name of syndrome (affected chromosome)	Clinical features
22q11.2 deletion syndrome (velocardiofacial, Di George syndrome)	Isolated cleft palate or submucous cleft, cardiac defects, defective thymic development, learning disability, characteristic facial appearance
Pierre Robin sequence	Cleft palate, micrognathia and glossoptosis
Stickler (12q13.11–q13.2, 1p21, 6p21.3)	Collagen disorder, micrognathia, eye abnormalities, hearing loss, joint problems, characteristic facial appearance
Van der Woude (1q32–q41)	Cleft lip, lower lip pits, missing teeth. Autosomal dominant
Treacher Collins (5q32–q33.1)	Micrognathia, ear abnormalities, deafness, abnormal lower eyelids
Hemifacial microsomia (Goldenhar) (14q32)	Vertebral and cardiac abnormalities
Down (trisomy 21)	Macroglossia, cardiac defects, atlanto-axial instability, learning disability
Edwards (trisomy 18)	Cardiac defects, micrognathia, renal malformations. Often fatal in infancy
Patau (trisomy 13)	Developmental delay, microcephaly, micrognathia, cardiac defects. Often fatal in infancy
Ectodactyly ectodermal dysplasia and clefting syndrome (7q11.2–q21.3, 3q27)	Disordered temperature control with hypohidrosis, malnutrition, respiratory tract infections

(CSAG). The goals of treatment are to enable the baby to develop into a confident child who becomes a socially integrated adult with normal speech and hearing, minimal facial disfigurement and normal dentition.

Timely detection has steadily improved in the United Kingdom over last 10 years, and nearly 85% of babies born with clefts are diagnosed antenatally or within 24 hours of birth. Over 70% of cleft lips are picked up on the 20-week anomaly scan, and support from the cleft team can be arranged at this stage. In other cases, diagnosis and referral are made after birth. Paediatricians are advised to make a visual inspection of the palate (rather than digital examination) during routine postnatal screening to exclude cleft palate. After diagnosis, assessment of feeding, facial appearance, speech, hearing and dentition is necessary to plan treatment.

Feeding difficulties are common for babies with cleft palate as they will not be able to suck effectively; the baby will have to work hard to feed and will be slow to put on weight. Parents are advised to use a special feeding bottle. Nasogastric feeding is sometimes necessary if oral feeding is unsuccessful, or if the baby presents for surgery with severe failure to thrive. Babies may also have uncoordinated swallowing and problems with reflux of milk. Most infants with isolated cleft lip are able to breastfeed successfully.

Hearing assessment is important in older infants, as the Eustachian tube does not open properly. Secretory otitis media is common, and grommets may be required at the time of cleft palate surgery.

Expert surgical care is the cornerstone of successful management. Repair of cleft lip is undertaken after three months of age and involves correction of the lip and nasal deformity (flattened alar margin and loss of nasal tip prominence on the cleft side and a displaced septum). Closure of the palate before the age of 13 months is recommended for optimum speech development. Late repair in older children or adults produces poorer results. The aim of surgery is to close the cleft palate and ensure good function of the soft palate without affecting normal maxillary growth. If the palate is short or does not function properly, air escapes through the velopharyngeal orifice during speech (VPI), which impairs phonation and causes hypernasal speech. The function of the normal soft palate is shown in Figure 23.3.

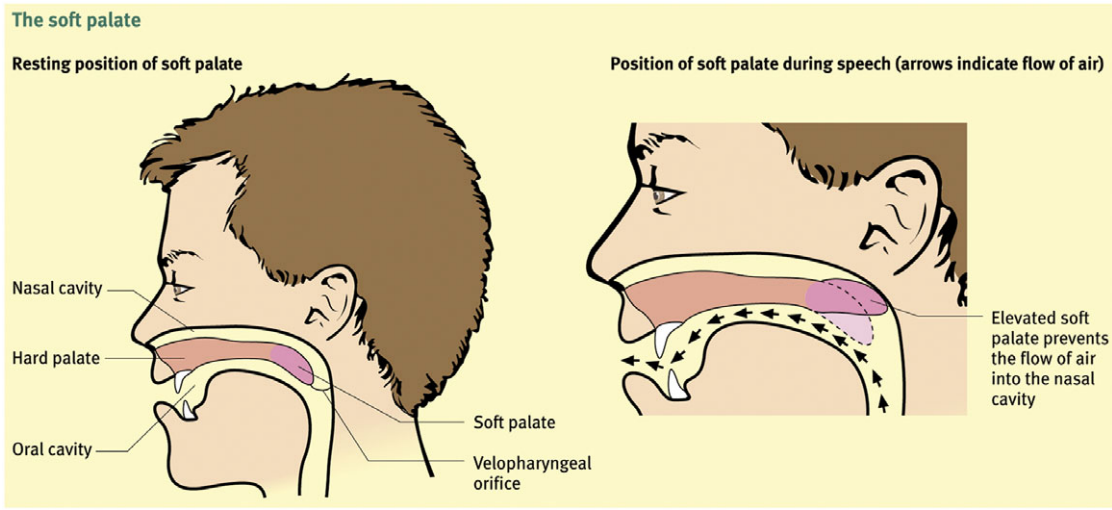


Figure 23.3 The position of the soft palate at rest and during speech.
Source: Reprinted from Mosahebi A, Kangesu L. Cleft lip and palate. *Surgery (Oxford)* 2006; 24:34–7, with permission from Elsevier.

Later orthodontic surgery may be required to repair an alveolar defect to facilitate normal eruption of secondary teeth. Towards early adulthood, when facial growth has ended, surgery may be undertaken to improve bite, improve the airway or change the appearance of the nose.

Depending on the type of cleft, the child may require one procedure or multiple operations from infancy to early adulthood. The surgical procedures and the typical age at surgery in the North Thames Cleft Network are described in Table 23.2. Ongoing support for the family by the cleft team is essential, and in the United Kingdom, there are also national groups such as the Cleft Lip and Palate Association (CLAPA) that have played an important role in the design of patient-centred services.

Table 23.2 Timing of surgery in the North Thames Cleft Service

Age at surgery	Potential surgical procedures
3–6 months	Primary cleft lip and nose repair and anterior palate
9–12 months	Primary cleft palate repair +/– grommets
1 year	Second-stage lip repair for bilateral cleft lip
3 years onwards	Lip +/– nose revision
3 years onwards	Secondary speech surgery – re-explore palate, buccinator flaps, pharyngoplasty, autologous fat transfer
Any age	Repair of palatal fistula
Any age	Primary repair of submucous palate
9–10 years	Alveolar bone graft
17–20 years	Maxillary osteotomy
17–20 years	Rhinoplasty

Primary Repair of Lip and Palate

The quality of primary surgery is thought to determine the outcome of speech and facial growth. The practice in the North Thames Cleft Service is to undertake primary repair of cleft lip, nose and anterior palate (if required) at 3–6 months and primary repair of the palate at 9–12 months. Closure of the lip and palate during the neonatal period has been advocated previously but adds additional risk and appears to confer no surgical benefit.

Repair of the lip involves repositioning of the orbicularis and perioral muscles and correction of the nasal deformity. The anterior palate, if involved, can be closed at the same time, mobilising the periosteum of the septum, called a vomerine flap, to cover the defect.

The aim of palate repair is to close the defect, ensuring good function of the soft palate and the

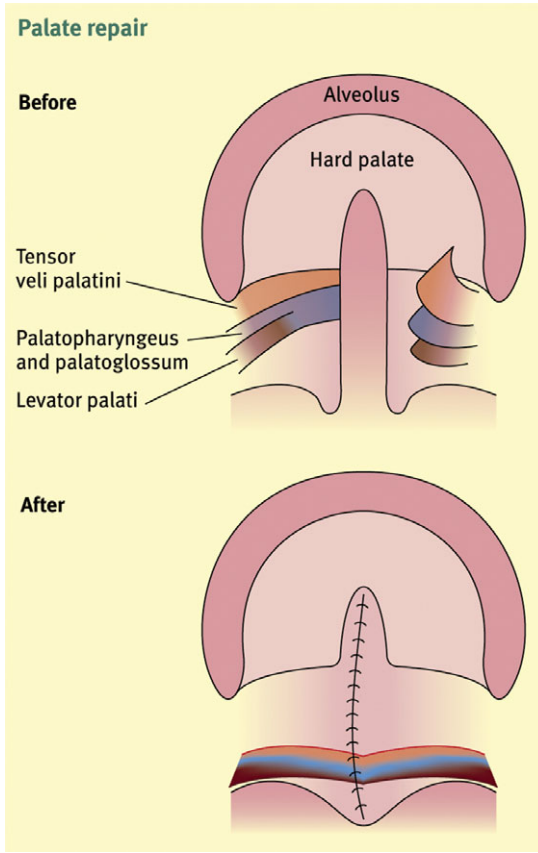


Figure 23.4 Primary cleft palate repair – muscles of soft palate are realigned transversely.
Source: Reprinted from Mosahebi A, Kangesu L. Cleft lip and palate. *Surgery (Oxford)* 2006; 24:34–7, with permission from Elsevier.

least effect on maxillary growth. Closure of the palate is performed using an operating microscope to dissect the soft palate muscles and realign them transversely to the back of the soft palate (Figure 23.4). Lateral releasing incisions through the mucosa may be performed to reduce tension on the midline repair. Grommets may be inserted at the same time as palatal surgery, if required.

Preoperative Assessment

Prior to surgery, infants should be well, thriving, gaining weight and following centile lines. Twelve per cent of babies with cleft are born prematurely compared to 7% in the general population. The historic guide of '10s' advocated by Wilhelmsen and Musgrave in 1966 was a useful guide for infants having lip repair. The infant should be:

Over 10 lb (4.5 kg)
Over 10 weeks old with a haemoglobin >10 g dl^{-1}
WCC $<10 \times 10^9$ dl^{-1}

Review of postoperative complications has identified that feeding difficulty, oesophageal reflux, poor weight gain and respiratory tract infections are associated with higher incidence of wound dehiscence and need for re-repair surgery. Our practice is to have multidisciplinary assessment led by general paediatricians to optimise these systemic concerns prior to surgery.

Infants with cleft palate often have chronic nasal discharge and reflux of milk through the nose, but this should be distinguished from an active upper respiratory tract infection. Infants who are unwell with mucopurulent nasal secretions or raised temperature are best postponed, as they are at much higher risk of airway complications perioperatively.

It is important to identify potential airway problems and other associated abnormalities (see Figure 23.5).

Airway assessment includes questioning regarding a history of obstructive symptoms and looking for micrognathia (best viewed in lateral profile). Severity of obstructive symptoms can be investigated with overnight oxygen saturation monitoring and polysomnography. Infants with severe micrognathia, most commonly associated with Pierre Robin sequence, may have a nasopharyngeal airway (NPA) in situ to relieve obstruction. They may have grown sufficiently so that the NPA is no longer needed but may still be required in the immediate postoperative period.

Infants with suspected cardiac abnormalities, such as those with low oxygen saturation readings or a heart murmur, require an echocardiogram and formal cardiology assessment. Healthy infants scheduled for simple lip repair do not require any routine blood tests. Those who are failing to thrive, are clinically anaemic or are having a bilateral lip repair require baseline haemoglobin measurement.

Anaesthesia and Airway Management

Premedication in infants is not generally required. Inhalational induction of anaesthesia with sevoflurane in 100% oxygen is recommended to ensure good oxygenation and allow assessment of the airway whilst maintaining spontaneous ventilation. Infants with micrognathia may develop airway obstruction as they lose pharyngeal tone;

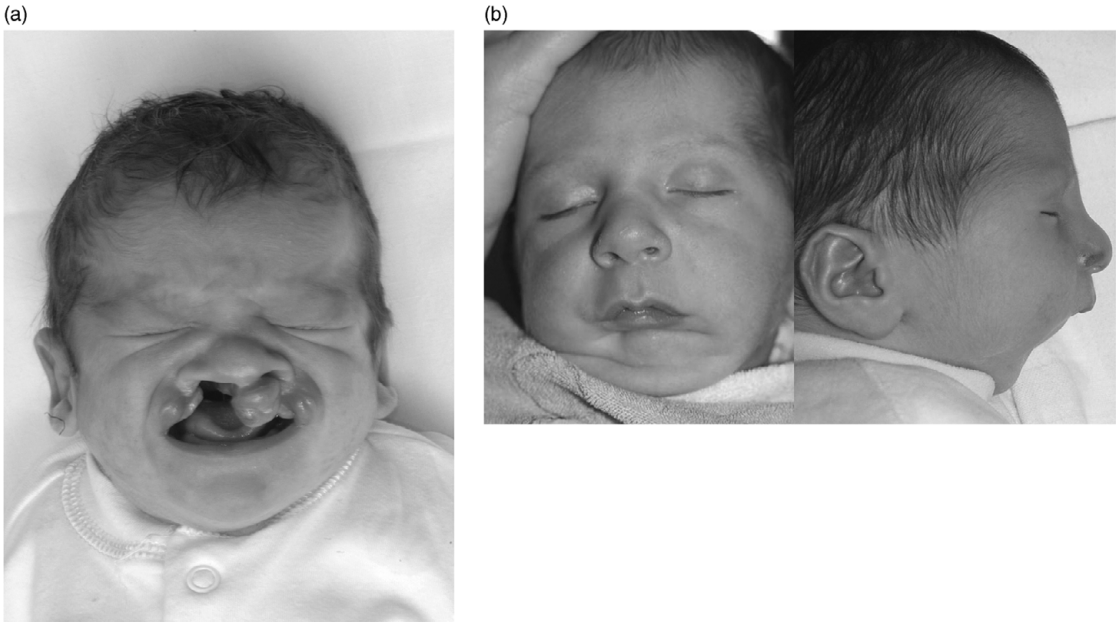


Figure 23.5 Factors predicting difficult intubation. (a) Wide bilateral cleft lip and palate with protruding premaxilla. (b) Micrognathia in an infant with Pierre Robin sequence.
Source: Reprinted with permission from Sesenna E, Magri AS, Magnani C et al. Mandibular distraction in neonates: indications, techniques and results. *Italian Journal of Pediatrics* 2012; 38:7, www.ijponline.net/content/pdf/1824-7288-38-7.pdf).

this can usually be relieved by repositioning, use of continuous positive airway pressure or insertion of a nasal or oral airway. It is an early indicator of the need for a nasopharyngeal airway postoperatively. After intravenous access is secured, a non-depolarising muscle relaxant can be administered after the ability to inflate the lungs is confirmed. If there is difficulty, spontaneous breathing should be maintained.

Most infants will be easy to intubate. A preformed south-facing cuffed tracheal tube fixed in the midline provides a stable airway and aids surgical access. Difficult laryngoscopy is seen in approximately 5–8% of cases. Factors that predict a grade 3 or 4 Cormack and Lehane view at laryngoscopy are a wide bilateral cleft with protruding premaxilla or micrognathia in an infant under six months of age (Figures 23.5a, b). It is helpful that these factors are all readily identifiable preoperatively. Children with Pierre Robin sequence generally become easier to intubate as they become older, but this is not the case for children with other syndromes such as Treacher Collins or hemifacial microsomia.

Should difficulty be encountered with intubation, a variety of aids can be employed. Simple manoeuvres include pressure over the larynx and use of a gum elastic bougie or a stylet. Videolaryngoscopy has the obvious advantage of clearer visualisation of anatomy and helps in shared situational awareness for the airway team. Hyperangulated blades facilitate viewing the anterior larynx, although intubation of the trachea often needs shaping the tube with a stylet. Care should be taken to avoid the flange of the laryngoscope blade slipping into the cleft or injuring the premaxilla (in BCLP) as it is supplied by an end artery. More advanced techniques depend on the equipment available and the experience of the anaesthetist. A fibreoptic scope can be used alone or using a laryngeal mask airway as a conduit. The tracheal tube may be railroaded over the scope or a guidewire placed via the suction channel. An airway exchange catheter is then railroaded over the guidewire and the tracheal tube passed over this (see Chapter 22 regarding difficult airways in children).

It is rare that intubation is not possible (less than 1% of cases). Surgery should be postponed in this situation, as the child may become easier to intubate with age, and access for the surgery will be improved. Plans for difficult airway management can be made, including tracheostomy if necessary.

Routine use of a throat pack to reduce soiling of the airway with blood is now questioned. With increasing use of cuffed tracheal tubes, careful suctioning and meticulous haemostasis, several

centres have stopped using throat packs, thereby avoiding the risk of mucosal injury, sore throat, oedema and retained throat pack.

Anaesthesia should be maintained with a volatile agent. Dexamethasone is administered to reduce airway oedema, and surgeons may request tranexamic acid to reduce operative blood loss and aid surgical view. Analgesia may be achieved with a variety of analgesic techniques such as intermittent boluses of fentanyl (1–3 mcg kg^{-1}) or remifentanil infusion (0.1–0.2 mcg kg^{-1} min^{-1}). Lateral releasing incisions are particularly painful. Effective analgesia will contribute to smooth anaesthesia and may avoid the need for further muscle relaxation.

During surgery, the infant is positioned with the head in a neutral position on a head ring. Ensuring correct tube length is crucial. Note that the tracheal tube tip may move up towards the larynx if the head is extended (conversely, down towards the carina if the head is flexed). Fixation of the tube in the midline is important to facilitate symmetric and aesthetic repair of cleft lip. Water-resistant transparent dressings aid tube fixation following skin preparation with cleaning solution. A mouth gag is inserted during cleft palate repair. The tracheal tube must be checked by hand ventilation or by assessing airway pressures when the gag is opened to make sure that the tube is not compressed. The gag time should be limited to two hours to avoid development of tongue oedema secondary to impaired perfusion. The anaesthetist must be constantly alert to possible disconnection of the circuit near the patient end, and to accidental extubation when the gag is adjusted or removed. Clear surgical drapes allow for easy visualisation of the patient, breathing circuit and tracheal tube position.

Total anaesthetic and surgical time ranges from 90 minutes for a simple lip or palate repair to up to three hours for a bilateral lip repair. Pressure areas should be protected, the infant warmed and temperature monitored to avoid overheating. Blood loss is usually not significant, although there is often more bleeding associated with a vomerine flap repair or if lateral release incisions have been performed.

Extubation and Postoperative Airway Management

Upper airway obstruction may occur in some infants after palate repair, particularly those with micrognathia or where the repaired palate is long. It is due to obstruction of the nasopharyngeal space by the oedematous palate and is particularly problematic in young infants who are still obligate nose breathers. The infant should be extubated when completely awake, after direct inspection and suction of the pharynx and documented removal of the throat pack. Signs of airway obstruction include sternal and subcostal recession, decreasing oxygen saturation and stertor. Residual anaesthetic effect, opioids and blood or secretions can contribute to upper airway obstruction. Other important causes to exclude are a swollen tongue, if the gag has been in place for a long time, or a retained throat pack.

The obstructed child should be managed with 100% oxygen, continuous positive airway pressure and/or careful insertion of an NPA or oral airway, taking care with the surgical repair. The NPA can be fashioned from a tracheal tube or a preformed nasal airway, usually 0.5 mm diameter smaller than that used to intubate the trachea. Tube length can be estimated by measuring from tip of the nostril to the tragus of the ear. If obstruction is not relieved by the NPA, reintubation may be required. In infants with micrognathia, particularly for those with Pierre Robin sequence who are likely to develop airway obstruction postoperatively, it is better for the surgeon to place the NPA electively at the end of surgery whilst the child is still anaesthetised. This minimises damage to the surgical repair and ensures the correct length of the NPA, that is, with the tip of the NPA just protruding from behind the soft palate. If the NPA is too short, it will be ineffective; if it is too long, it will irritate the larynx and the baby will not settle postoperatively due to coughing.

Any baby that is dependent on an NPA needs to be monitored closely postoperatively in a level 2 high-dependency area, and the NPA needs to be suctioned regularly to make sure that it remains patent. A spare NPA of the same dimensions should be immediately available at the bedside in case of accidental dislodgement or blockage of the existing one. Intravenous dexamethasone is useful to help to reduce surgical oedema (first dose 250 mcg kg^{-1} IV; three further doses 100 mcg kg^{-1} IV 8 hourly). Positioning the baby in the lateral or sitting position may help airway patency. Tongue sutures are seldom used. The NPA can usually be removed after 24 hours.

Analgesic Management

Multimodal analgesia should be used so that the infant is comfortable but not over-sedated at extubation. There are a wide variety of analgesic techniques available, and our current practice in the North Thames Cleft Network will be described.

Infants undergoing cleft lip repair are managed with infraorbital nerve blocks with bupivacaine, performed using an intraoral approach. Blocks are performed prior to the start of surgery by the anaesthetist or the surgeon. The surgeon will also infiltrate the surgical field with local anaesthetic containing adrenaline 1:200,000 to reduce blood loss and aid surgery. Intravenous paracetamol is given intraoperatively, with regular paracetamol and ibuprofen PO postoperatively. The anterior palate is not anaesthetised by the infraorbital nerve block, and children who undergo vomerine flap repair at the same time as lip repair receive a loading dose of morphine during surgery (50–100 mcg kg^{-1} IV). These children may also require oral morphine for breakthrough pain in the postoperative period, as for cleft palate repair.

Cleft palate repair is a more painful procedure. All children receive a combination of IV paracetamol, rectal diclofenac and morphine administered towards the end of surgery (100 mcg kg^{-1} IV). Our practice is for the surgeon to infiltrate the palate with bupivacaine 0.25% and adrenaline 1:200,000, maximum 0.8 ml kg^{-1}, to provide pain relief and to reduce blood loss. Bilateral suprazygomatic maxillary nerve blocks placed before the start of surgery have been shown to be effective in reducing opioid consumption. Anatomical landmarks are used to guide the injection, and confirmation of local anaesthetic spread can be obtained with ultrasound. Clonidine 1–2 mcg kg^{-1} IV is a useful adjunct to provide analgesia without respiratory depression for children older than six months undergoing cleft palate repair.

The use of a validated pain score such as FLACC allows objective assessment of pain in the postoperative period and guides analgesic use. If the infant does not settle with a feed or comforting in the recovery room, increments of morphine (20 mcg kg^{-1}) are given. On the postoperative ward, regular oral paracetamol and ibuprofen are given, with oral morphine for breakthrough pain as required (morphine 100 mcg kg^{-1} PO).

Perioperative Fluid Management

It is our practice to administer an intraoperative fluid bolus of 20 ml kg^{-1} of Hartmann's solution to replace deficit and for maintenance requirements. Additional Hartmann's is given as required, but blood loss is usually small, and transfusion is seldom required.

Infants are encouraged to feed postoperatively, and after lip repair most will manage well. After palate repair, some infants require short-term intravenous fluid until feeding is established. Alternatively, a nasogastric tube can be placed at the end of primary palate surgery to enable top-up feeding and to meet maintenance fluid requirements postoperatively via the enteral route. This avoids the need for monitoring urea and electrolytes and cannulation in the postoperative period and allows for the regular administration of oral analgesia.

Postoperative Care

Postoperative bleeding after palate repair is rare but may require blood transfusion and return to theatre. Anaesthesia management is similar to that of bleeding post tonsillectomy – the infant will need fluid resuscitation, there will be blood in the airway, intubation may be difficult, residual anaesthesia must be taken into account and there may be a full stomach due to swallowed blood.

Infants who undergo uncomplicated cleft lip surgery are usually discharged from hospital after one night, although term infants having lip repair without vomerine flap may be managed as a day case if there are no other concerns. A two-night stay is usual for infants having palate repair. Absorbable sutures are increasingly being used for lip repair, eliminating the need for removal of sutures under general anaesthesia or oral sedation.

Secondary Cleft Surgery

Children may present for a variety of secondary procedures in childhood (see Table 23.2). Children for secondary procedures may become increasingly anxious about anaesthesia. Discussion about their concerns, giving choice for method of induction and using different distraction and coping techniques can help. Sedative premedication may be useful.

Secondary procedures on the lip and nose are undertaken to improve cosmetic appearance or

improve nasal airway patency. Anaesthetic technique is generally similar for primary lip repair, although revisions in older children and adults can be performed using a laryngeal mask. Palate re-exploration may be undertaken to improve palatal length (and hence function) or to repair a palatal fistula. Principles are as for primary palate repair.

Alveolar bone graft is performed for children with a defect in the alveolus to provide a base for the eruption of the permanent teeth. Bone is taken from either the iliac crest or tibia. For details of anaesthesia for maxillofacial procedures, please refer to Chapter 24.

Secondary speech surgery may be carried out after investigation by videofluoroscopy and nasal endoscopy performed by speech therapists. Children are usually over three years old, and the procedure involves improving apposition of soft palate to posterior pharyngeal wall by using one or more of the following techniques:

- Repositioning muscles of soft palate – re-repair or Furlow Z-plasty
- Lengthening of the soft palate with buccinators myomucosal flaps
- Pharyngoplasty, of which there are three broad types: Hynes, sphincter (Orticochea) or midline superior posterior pharyngeal flaps

Buccinator Flaps

This operation increases the length of the palate by rotating buccinator myomucosal flaps from the inner cheek into the junction between the hard and soft palate. The donor sites on each cheek are closed directly. The trachea should be intubated with a pre-formed south-facing tracheal tube. Good pain relief is important with a combination of local anaesthetic infiltration, paracetamol, an NSAID and morphine. Intravenous clonidine is a useful adjunct. Postoperatively, regular simple analgesics are given, with rescue analgesia provided by oral morphine.

This is a painful procedure, and some children are particularly reluctant to take oral fluids postoperatively. It is useful to pass a nasogastric tube perioperatively to provide a route for analgesia administration and postoperative feeding and fluid intake until the child is eating and drinking. After three weeks, the muscle flaps have vascularised, and the flap pedicles are divided under general anaesthesia.

Pharyngoplasty

A Hynes pharyngoplasty involves mobilising local flaps from the posterior wall of the pharynx to produce a bulge at the junction of the nasopharynx and oropharynx that can then oppose the end of the soft palate to provide palatal competence. In the Orticochea pharyngoplasty, palatopharyngeus myomucosal flaps are positioned lower down on the posterior oropharyngeal wall, and the posterior tonsillar pillars are sutured to the posterior pharyngeal wall. This procedure purportedly utilises the sphincter action of the muscle in the flaps, helping to seal the velopharyngeal orifice during speech. Another form of pharyngoplasty consists of a posterior pharyngeal flap sutured onto the soft palate. Two lateral nasopharyngeal ports are thus created. The choice and exact nature of surgical procedure is individualised depending on the anatomy and the dynamic physiology of a patient.

Conduct of anaesthesia is similar to primary palate repair and requires tracheal intubation. The procedure is painful, and multimodal analgesia should be used with care so as not to cause oversedation with opioids. The operation, by its nature, produces a degree of upper airway obstruction, particularly in the immediate recovery period. Close monitoring is required, including monitoring of oxygen saturation. Dexamethasone may reduce local oedema. The passage of a nasopharyngeal airway is contraindicated, as this will cause damage to the surgical site. After Orticochea or posterior pharyngeal flaps, nasal intubation for future surgery is absolutely contraindicated.

Postoperative haemorrhage may occur as a complication of pharyngoplasty, owing to bleeding from the posterior pharynx. Particular care should be taken with children with 22q deletion who have more medial carotid arteries lying close to the pharyngeal flap donor site. Occasionally, bleeding may necessitate transfusion and/or return to theatre. The child will need fluid resuscitation and careful induction of anaesthesia to avoid aspiration.

Obstructive sleep apnoea occurs more frequently after posterior pharyngeal flap surgery than other techniques and may persist for up to three months postoperatively. Severe obstruction can be life-threatening, and death has been reported in the literature. The child may require reintubation and return to theatre to reverse the surgery.

Transoral Robotic Cleft Surgery (TORCS)

This technique shows some promise and may be carried out in the future in UK settings. The learning curve for this technique is steep, and all anaesthetic concerns of robotic surgery in a shared airway apply, in particular, the challenges of managing airway emergencies requiring rapid disengagement of robot arms. Infrastructure upgrade and further experience are needed before it becomes a mainstream technique.

Key Points

- There are two distinct cleft conditions: cleft lip with or without cleft palate (Cl+/–P) and isolated cleft palate (iCP).
- Many children with cleft will have associated abnormalities, more commonly in those with isolated cleft palate; of particular importance are airway problems and cardiac defects.
- Depending on the type of cleft, children may require only one operation for correction or several procedures throughout childhood. A multidisciplinary approach is essential.
- Most children will be easy to intubate for primary repair. Children under six months with micrognathia or a wide bilateral cleft may be more difficult to intubate.
- Cleft palate repair is managed with multimodal analgesia, including opioids, taking care to avoid over-sedation.
- Upper airway obstruction following cleft palate repair should be anticipated. It is more common in infants with micrognathia and can be managed with a nasopharyngeal airway.
- Pharyngoplasty surgery may be complicated by postoperative upper airway obstruction and bleeding.

Further Reading

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