

Anaesthesia for cleft lip and palate surgery

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Key points

A high index of suspicion for conditions associated with cleft lip and palate should be maintained.

A difficult view at laryngoscopy is a more frequent finding than a difficult airway; the anaesthetist should be prepared for either.

Patients should be extubated when fully awake with close observation for signs of airway obstruction.

Analgesia is an important part of the balanced anaesthetic technique.

Children with clefts should be managed by a multidisciplinary team of experts.

Cleft lip and palate

The presence of a cleft lip, cleft palate or both, has a huge impact on the life of an individual and their family. Modern management has much to offer these people, revolutionizing appearance and functional defects to a point where they may be difficult to detect. Surgery aims to correct the anatomically obvious cleft lip, augment normal dento-alveolar development and lead to effective palatal function.

Worldwide, cleft lip and palate (CLP) is one of the more common congenital malformations. In the UK, the incidence is around 1:700, amounting to ~1000 babies every year presenting with cleft lip and/or palate (Table 1).

CLP is more common in males; strangely, cleft lips are usually left-sided. The aetiology is often unknown, though likely to result from both environmental and genetic causes. The incidence is greater than expected amongst first-degree relatives of those with CLP. Both cleft lip and palate occur as a result of defects in palatal growth during the first trimester. The palate grows inwards to fuse in the midline in two stages. By 6 weeks the primary palate forming the alveolus and lip fuses, followed 8 weeks later by the secondary palate, which is posterior to the incisive foramen. The window of opportunity for fusion to occur is relatively short, and suggested theories for failure of fusion include mechanical obstruction by tongue position, structural hypoplasia or primary breakdown. Clefts of the lip and alveolus can be diagnosed reliably at the routine 18–20 week antenatal ultrasound scan, allowing for earlier preparation of support services and counselling of parents. Clefts of the palate are not easily seen by ultrasound and can only be excluded by examination of the palate after delivery.

Anaesthetic significance

All patients require anaesthesia for primary surgical repair; the anaesthetist may also be involved in airway management for some cases before surgery. Although the focus of this article is anaesthesia for primary CLP repair,

Table 1 Type of abnormality in babies born with cleft lip and palate

Unilateral cleft lip	25%
Unilateral cleft lip and palate	25%
Bilateral cleft lip and palate	10%
Cleft palate alone	40%

these patients may require surgery well into adulthood, for example to improve lip aesthetics, close residual palatal defects, bone graft and align alveolar and dental defects, realign the jaws and correct nasal deformity. Surgery for otitis media, or non-cleft related anomalies is not uncommon.

As with any patient group, the anaesthetist requires knowledge of the surgical problem. While most patients present with isolated CLP, a significant number have associated anomalies. The most obvious are other defects of branchial arch development (e.g. ear or upper airway defects). However, they also include heart, renal or skeletal anomalies. Associated anomalies may be part of a described pattern, providing guidance as to which defects can be expected;¹ the list of such conditions including CLP is becoming endless, ranging from the outwardly obvious such as Trisomy 21, to more subtle such as Velocardiofacial Syndrome (Table 2).

Preoperative care

General

Children with CLP usually present for correction in infancy, neonatal surgery is unusual unless it is for non-cleft operations. Waiting until 3 months of age gives time to detect most congenital abnormalities, and allows anatomical and physiological maturation. Upper respiratory tract infections are particularly common at this age and carry an increased risk of airway complications and impaired wound healing.² Routine or targeted treatment of low-grade nasal infections with antibiotics will reduce the incidence of postoperative pyrexial illness, though many of these children

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Table 2 Common syndromes associated with cleft lip and palate

Pierre Robin Sequence	80% associated with cleft palate Micrognathia Glossoptosis Underlying syndrome/anomalies Usually easier to intubate with age
Treacher Collins Syndrome	28% associated with cleft palate Micrognathia and maxillary hypoplasia Choanal atresia Eye and ear malformations Intubation may become more difficult with age
Hemifacial Microsomia	Hemifacial and mandibular hypoplasia Cervical spine abnormalities Ear and eye abnormalities Intubation may become more difficult with age
Velocardiofacial Syndrome	Microcephaly and microstomia Flat nasal bridge, small ears, short stature Immune deficiency, congenital cardiac disease, laryngeal and tracheal anomalies Velopharyngeal incompetence ± cleft palate 22q 11 deletion (FISH test)
Stickler Syndrome	Progressive connective tissue disorder Micrognathia and flat face Eye, ear and joint abnormalities Congenital cardiac disease
Down Syndrome	Microstomia and relative macroglossia Epicanthic folds, simian crease Congenital cardiac disease Atlantoaxial subluxation and instability
Fetal Alcohol Syndrome	Smooth philtrum, thin vermillion, small palpebral fissures, growth deficit CNS abnormalities
Klippel-Feil	15% associated with cleft palate Short, webbed neck and fused cervical vertebrae Congenital cardiac disease

have a continuous nasal discharge without overt infection. Anaemia may be seen from a combination of nutritional and physiological causes. Preoperative visits should allow time to assess general and specific problems, as well as provide information to the parents about the induction technique, provision of analgesia and possibility of additional measures such as postoperative nasopharyngeal airway insertion.

Specific

Clefts of the lip and palate *per se* do not lead to upper airway obstruction; this is generally a result of other problems (structural to neuromuscular) affecting the normal function of the upper airway. The most important group of CLP patients with airway problems are those with micrognathia, glossoptosis and cleft palate, termed Pierre Robin Sequence (Fig. 1). The primary deformity in these infants is believed to be micrognathia, resulting in a tongue position further back in the oropharyngeal cavity. The clinical effects of the sequence are twofold: (i) *in utero*, reduced pharyngeal space prevents palatal fusion; and (ii) at birth, or soon after, upper airway obstruction of varying degrees of severity is apparent.

A clear airway is obviously paramount and first line management consists of placing the infant prone, this may suffice for milder forms. The next step is to provide an artificial airway, usually nasopharyngeal. Very rarely, obstruction is severe or complicated by other problems such as neuromuscular disease or choanal atresia; under these circumstances tracheostomy is required. Any of these infants can be a challenge to intubate; the laryngeal mask may provide a very useful rescue airway.

The term 'sequence' is used for this condition to describe a clinical pattern resulting from a single initiating anomaly (in this case micrognathia) of multiple cause. For example, chin growth may be restricted mechanically *in utero* by oligohydramnios, or affected by abnormal development of the temporomandibular joint. A 'syndrome' describes a condition for which the aetiology is the same between individuals (e.g. Edwards Syndrome is always caused by trisomy 18). From this definition it will be apparent that Pierre Robin Sequence may occur in addition to an underlying syndrome. When managing any patient with CLP, even those labelled as 'Pierre Robin Syndrome or Sequence', a high index of suspicion for other underlying problems should be maintained.

Surgery is usually performed at 3 months for cleft lip repair and 6 months for cleft palate. It may be delayed by the investigation of other problems or on-going airway difficulties; in such cases, the timing of surgery is critical and is best decided by discussion between all specialities involved, including anaesthesia. No clear guidelines exist, particularly for the infant with airway complications; practice is inconsistent and based largely on individual experience.

Intraoperative and postoperative care

As usual, preparation is paramount and any infant with CLP requires all the normal facilities of paediatric care, together with those for dealing with a potentially difficult airway.

Our practice is to induce anaesthesia by inhalation with sevoflurane in oxygen 100%; i.v. access, if not previously established, is secured and routine monitoring (ECG, end-tidal carbon dioxide, non-invasive blood pressure, SpO₂, core temperature) is established. Once a suitable depth of anaesthesia is reached, face mask ventilation is confirmed and paralysis achieved with either a depolarizing or a longer acting neuromuscular blocking agent. Tracheal intubation is then attempted. Neuromuscular blocking agents are contraindicated if the ability to inflate the lungs is in doubt, spontaneous ventilation must be maintained for the safe induction of general anaesthesia in the difficult paediatric airway.

The difficult airway

Difficult face mask ventilation is thankfully very rare; however, if the airway becomes obstructed after loss of consciousness, treatment options include turning the patient lateral or semi-prone and the use of a nasal or oral pharyngeal airway. The laryngeal mask airway may help, and may enable the patient to achieve adequate



Fig. 1 An infant with Pierre Robin Sequence, note marked micrognathia in lateral view. There is a nasopharyngeal airway in one nostril and a nasogastric tube in the other to supplement feeding. (Reproduced with parental permission.)

depth of anaesthesia for direct laryngoscopy and intubation to be tolerated. In the event of being unable to secure the airway, consideration should be given to allowing the patient to wake up and deferring surgery to when they are older, have more structural and neuromuscular maturity, and when the airway may be easier to manage. Providing an emergency surgical airway (cricothyrotomy or tracheostomy) is a difficult option in this group, but may be needed to secure and subsequently maintain the airway.

A difficult view at laryngoscopy is a more frequent finding than difficult face mask ventilation.³ A large alveolar defect, especially on the right side, may make positioning of the laryngoscope more difficult as it tends to fall into the cleft. A piece of gauze packed into the cleft may help as may the use of a straight laryngoscope blade, the lateral or molar approach and external laryngeal manipulation. The laryngeal mask provides an excellent means to maintain the airway and ventilate the lungs; it can also be used to guide the fiberoptic bronchoscope to the larynx. When using any such conduit to the upper airway, care is needed to ensure that components easily fit through one another and avoid difficulty in threading the tube into the larynx. Techniques vary, for example with an adult bronchoscope, a wire is threaded through the suction channel and the bronchoscope and laryngeal mask removed over the wire, which is then used to guide the tracheal tube into place. Alternatively, a narrow diameter 'paediatric' bronchoscope such as the Olympus LF-P can be passed via the LMA into the trachea and used to directly guide a preloaded tracheal tube. The laryngeal mask has been successfully used alone to

maintain the airway for cleft palate repair; however, it is bulky and less secure than a tracheal tube.

Maintenance of anaesthesia

The ideal tracheal tube is the preformed RAE tube, which passes out over the lower lip, where it is fixed centrally, allowing for optimal surgical access. The shared airway presents opportunities for tube occlusion and inadvertent extubation at almost any stage. A throat pack is used for lip surgery; palate surgery is usually conducted without a pack. A head ring and a roll under the shoulders is frequently used to extend the neck. The eyes must be protected.

Anaesthesia is maintained by controlled ventilation with volatile anaesthetic agents, noting that MAC values are higher than those for adults. Intraoperative analgesia is provided with fentanyl $1\text{--}2\text{ }\mu\text{g kg}^{-1}$ i.v. in combination with local anaesthetic infiltration. For lip repair, infraorbital nerve blocks have been shown to be effective. Longer acting opioid analgesia is required for cleft palate surgery and cleft lip surgery in older infants or when the anterior portion of the palate is repaired as part of the lip repair (vomerine flap). Owing to fears of respiratory depression and excessive sedation, some recommend the use of codeine instead of morphine. However, recent evidence suggests that some individuals metabolize codeine poorly into its active constituents making it ineffective.⁴ Metabolism of morphine is more consistent and may be a better choice. Acetaminophen is commonly prescribed for

postoperative analgesia, often given as a loading dose pre- or intraoperatively. Most paediatric anaesthetists prescribe non-steroidal anti-inflammatory drugs for children from the age of 6 months.

The duration of surgery is usually 1–2 h. Infiltration of local anaesthetic with epinephrine may help reduce blood loss, which requires careful monitoring. Fluid losses (preoperative starvation, maintenance and on-going losses) are usually replaced with crystalloid; blood transfusion is unusual.

Antagonism, extubation and recovery

On completion of surgery, the oropharynx is inspected, the throat pack and any blood clots removed and haemostasis assessed. There is an association between slow recovery and postoperative airway obstruction and time should be allowed for adequate elimination of anaesthetic agents. The child is extubated after antagonism of neuromuscular block and when fully awake; supplementary oxygen is given. At this time, attention must be paid for signs of airway obstruction, most likely to be seen in infants with pre-existing airway problems. This may occur at any part of the upper respiratory tract and thought should be given to possibilities such as laryngospasm, upper airway narrowing, blood clot, retained throat pack, tongue swelling from retraction or inadequate mouth breathing. Having ruled out these causes, it may be sufficient to apply continuous positive airway pressure for a time or to turn the baby lateral or even prone. Further to this, careful insertion of a naso-pharyngeal airway relieves upper airway obstruction and should not damage the palate repair. It is usually only required until the following day, by which time operative swelling has partially resolved and nearly all infants will have mastered mouth-breathing. A few infants will require re-intubation and potentially a tracheostomy, possibly as a result of pharyngeal incoordination in addition to reduced pharyngeal space.

Close observation continues into the recovery period. Once the child is awake and no bleeding is seen, feeding with clear fluids can begin and is usually comforting. Parents are encouraged to join their child at this time. Postoperative analgesia is provided most rapidly by i.v. opioid (e.g. boluses of $20 \mu\text{g kg}^{-1}$ morphine). Other centres use opioid infusions or nurse-controlled analgesia very effectively. Oral analgesics (oramorph prn and regular acetaminophen and ibuprofen) will usually provide good extension of analgesia. In the majority of cases, it is not necessary to splint the arms to prevent the baby disrupting the sutures. Careful monitoring must be performed for the first 12 h after surgery for the early detection of airway obstruction or postoperative bleeding.

Future developments

Cleft lip has been treated surgically since ancient times but cleft palate surgery evolved about the time that anaesthesia became

available. Only comparatively minor refinements to anaesthetic technique have been necessary since the development of intratracheal insufflation in the 1920s, the use of a single large-bore tracheal tube in the 1930s and subsequently Ayre describing his valveless T-piece circuit for cleft lip and palate surgery in 1937. The early pioneers of anaesthesia developed the means to alleviate their patients suffering and make surgery both safer and more effective; their basic techniques are difficult to better.

Advances in the understanding of infant anaesthesia continue. Whilst difficult to prove objectively, improvements in monitoring, equipment and drugs probably result in improved safety and quality. The residual respiratory depressant and sedative effects of anaesthetic agents and opioids remain a major concern in infants scheduled for cleft surgery. Drugs such as desflurane with its rapid elimination characteristics may provide advantages, as may remifentanyl with its fast and predictable metabolism. Use of non-steroidal analgesia in this age group is common, though further trials are needed to elucidate the most effective and safe regimens; their effect on perioperative bleeding is still disputed. The timing and type of surgery have been debated over recent years. The advantages of foetal wound healing are being explored with experimental evidence suggesting *in utero* surgery for cleft lip and palate repair provides superior wound healing without scarring.

Changes have taken place in the organization of CLP services in the UK following the recommendations of the Clinical Standards Advisory Group that the number of hospitals providing CLP surgery be reduced to 8–15 units managing 40–50 cases of primary cleft lip and palate per annum.⁵ This has resulted in care being concentrated in fewer units with the potential advantages of improved resources, increased patient numbers, improved outcomes and better research and audit.

References

1. Shprintzen RJ, Siegal-Sadewitz VL, Amato J, Goldberg RB. Anomalies associated with cleft lip, cleft palate or both. *Am J Med Genet* 1985; **20**: 585–95
2. Takemura H, Yasumoto K, Toi T, Hosoyamada A. Correlation of cleft type with incidence of perioperative respiratory complications in infants with cleft lip and palate. *Paed Anaesth* 2002; **12**: 585–8
3. Gunawardana RH. Difficult laryngoscopy in cleft lip and palate surgery. *Br J Anaesth* 1996; **76**: 757–9
4. Williams DG, Hatch DJ, Howard RF. Codeine phosphate in paediatric medicine. *Br J Anaesth* 2001; **86**: 413–21
5. Clinical Standards Advisory Group—Report. *Cleft Lip and Palate*. London: The Stationery Office, 1998

See multiple choice questions 58–61.