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Review article

ANAESTHESIA FOR CLEFT LIP & PALATE

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Abstract
Background:
cleft lip & palate is congenital orofacial defects in paediatric patients.
Purpose:
Anaesthesia for cleft lip & palate is challenging as patients may have congenital other defects, cardiac problems & when not corrected in time there is a problem in dentition, speech, & looks of the patient also.so we have reviewed articles& how anaesthesia was conducted in cases, is discussed.
Methodology:
we have reviewed articles, also we have given anaesthesia for cleft lip & palate surgery. Right from preoperative assessment, NBM for paediatric
patients, pre oxygenation,
difficulty in bag mask ventilation, appropriate type & size of endotracheal tubes, fixation of tube to prevent accidental extubation, perioperative adequate depth of anaesthesia, muscle relaxation, haemostasis, postoperative analgesia was discussed in detail.
Results:
In all patients we have a secure airway with a south pole RAE tube of appropriate size secured in midline so actual measurement for surgical success can be achieved, i
In 1 patient endotracheal intubation was done with second attempt as patient has difficult intubation& Trachear Collin syndrome, in 1 patient unusual bleeding from surgical site was there which was corrected by giving fresh frozen plasma. In 1 patient delayed recovery from non-depolarise muscle relaxant, plasma cholinesterase was measured, recovery after fresh frozen plasma & ventilation till spontaneous ventilation.
Conclusion:
In nutshell cleft lip palate surgery is elective paediatric surgery, in which anaesthetist should have multitasking from through preoperative assessment for possibility of other congenital defects, cardiac defects, swift induction, maintain adequate depth of anaesthesia, vigilant monitoring, prevention of emergence extubation, prompt postoperative analgesia, so that early recovery & discharge can be possible for each patient.

Keywords: Anaesthesia, Cleft lip, palate

Introduction;

Orofacial clefts, encompassing cleft lip with or without cleft palate and cleft palate alone, are the most common craniofacial abnormalities. T

Definition and classification

Cleft lip refers to a fissure in the upper lip and may be incomplete or complete (through to the nasal cavity), unilateral, or bilateral.

Cleft palate describes a gap in the soft palate that may or may not involve the hard palate. They may be submucous (the mildest form with no visible cleft but a failure of the palatal muscles to unite), incomplete or complete, unilateral or bilateral.
*There are several complex classification systems that describe the extent of individual defects
Incidence
The overall incidence of orofacial clefts in the UK is around 1000 cases per year; approximately 1 in 700 live births. There is significant international variation and published figures depend on the accuracy of the data reported. There were regional differences; It is unclear why, but cleft lip with or without cleft palate is seen more in males, whilst cleft palate is seen predominantly in females.
Proportion of cleft types in patients born with clefts in the UK in 2018 were as follows:
Unilateral cleft lip 22%
Unilateral cleft lip& palate23%
Bilateral cleft lip & palate12%
Cleft palate alone 43%

Embryology

The embryological development of the lip and palate from neural crest cells is complex and requires coordinated cell migration, growth, and differentiation. Formation of the lip and primary palate involves the fusion of the lateral and median nasal processes and maxillary mesodermal processes. This is usually complete by the sixth week after conception. Failure at any of these sites results in unilateral, bilateral, or median cleft lip. Development of the secondary palate follows; the palatal shelves fuse in the midline and the position of the hard and soft palate is formed by the 10th week of gestation. As the lip and primary palate have an embryologically distinct developmental process to the secondary palate, cleft palate may occur with or without cleft lip.

Aetiology

Environmental factors may be contributory including maternal age, tobacco smoking, alcohol consumption, drug use, nutrition including folate deficiency, infection, and teratogen exposure.
However, patients with orofacial clefts may have additional congenital abnormalities (~30% in cleft lip with or without cleft palate and ~20% in cleft palate), including head and neck deformities, cardiac defects, and musculoskeletal and renal issues.
Usually infants or young pediatric patient
1)Associated comorbidities and syndromes

2)Nutritional status
3)Airway obstruction
4)Potential difficult airway
5)Anesthetic factors General anesthesia with tracheal tube
6)Shared airway
7)Surgical factors for Elective surgery (time to optimize)

***Velocardiofacial (DiGeorge) syndrome**

-Microcephaly and microstomia
-Flat nasal bridge, small ears, short stature
-Immune deficiency, congenital cardiac disease
-Velopharyngeal incompetence with or without cleft palate
-Laryngeal and tracheal anomalies
-22q 11 deletion (FISH test)
-Cleft palate in 30% of cases

***Downs syndrome**

-Microstomia and relative macroglossia
-Epicanthic folds, simian crease
-Congenital cardiac disease
-Atlantoaxial subluxation and instability

***Van der Woude syndrome**

-Lower lip pits
-Hypodontia
-Congenital cardiac disease
-Musculoskeletal issues
-Most common orofacial clefting syndrome

***Treacher-Collins syndrome**

-Eye and ear malformations
-Intubation may become more difficult with age
-Cleft palate in 30% of cases

***Hemifacial microsomia (Goldenhar syndrome)**

-Hemifacial and mandibular hypoplasia
-Cervical spine abnormalities
-Ear and eye abnormalities
I-ntubation may become more difficult with age
-Stickler syndrome
-Micrognathia/PRS
-Retinal detachment and early cataracts
-Deafness
-Hypermobility of joints

***Pierre Robin Sequence**

-Micrognathia
-Glossoptosis
-Underlyingsyndrome/anomay
-Usually easier to intubate with age
-Cleft palate in 80% of cases

Common syndromes associated with cleft lip and palate

Considerations for anesthesia for cleft lip and palate surgery

The anesthetic considerations include factors related to the patient, the anesthetic, and the surgery .

Patient-related factors

1-Usually infants or young pediatric patient

2-Associated comorbidities and syndromes
3-Nutritional status
4-Airway obstruction
5-Potential difficult airway

Anesthetic factors

1)General anesthesia with tracheal tube
2)Shared airway
3)Surgical factors
4)Elective surgery (time to optimize)
5)Timing of surgery

Surgical factors:

1)Primary repair of cleft lip may occur between 6 weeks and 6 months of age, allowing for growth of the patient, investigation of related anomalies before surgery, and avoidance of giving a general anesthetic in the neonatal period.
2)If cleft palate repair is required, this is generally completed between 6 and 12 months of age. The anesthesia team is involved from an early stage, for preoperative review and counseling, or in cases where there is significant airway obstruction.

Preoperative assessment

A comprehensive preoperative anesthetic assessment should be completed, being mindful of syndromes associated with orofacial clefts and comorbidities .
→ History and examination should include a systems review, specifically looking to identify anysigns of congenital heart disease (15% of patients); upper respiratory tract infection (URTI);
—> obstructive sleep apnoea (OSA); renal disorders; neuromuscular disorders or malnutrition.
For patients who have multiple comorbidities it may be prudent to delay cleft surgery until their health has been optimized and the balance between risks and benefit is more favorable.
Patients for primary cleft palate repair are generally admitted for 1–3 days as they are at higher risk of postoperative bleeding and airway obstruction.
URTIs, Chronic rhinorrhoea is common in patients with cleft lip with or without cleft palate and cleft palate secondary to reflux of feeds into the nasal passages. This should be distinguished from an active URTI. Patients' conditions should be optimised where possible, as these are elective procedures and may be postponed if necessary.
—>Nutrition and hydration
Feeding issues are common in these patients. Orofacial clefts hamper the infant's ability to feed effectively from either bottle or breast whilst associated abnormalities, such as Pierre Robin sequence (PRS) and OSA, may lead to apnoea during feeding or prolonged feeding. Any signs of malnourishment or dehydration should be investigated appropriately, and the procedure postponed until they are growing well. It should also be noted that, in some centres, infants may be weaned from the nipple/bottle to a cup before surgery to protect the repair after surgery.
—>Airway assessment
Airway assessment is mandatory, including appraisal of syndromic features and any history of chronic airway obstruction or OSA symptoms such as snoring and apnoea during feeds.

Patients with OSA are at higher risk of airway obstruction at induction, and after surgery, and are more sensitive to sedative drugs, especially opioids.
In the non-syndromic patient, evidence from a study of 800 patients suggests difficult direct laryngoscopy (Cormack and Lehane grade III or IV, 7.4%), difficult intubation (between two and four attempts, 8.4%), and failed intubation (1%) in ASA 1 cleft patients has, historically, been relatively uncommon. Difficult laryngoscopy was associated with patients <6 months of age, bilateral cleft lips, and retrognathia.
*** Certain syndromes are well known to be associated with difficult intubating conditions . Difficulty was increased in patients with cleft lip with or without cleft palate (10%) and cleft palate with PRS (23%)
Rule of 10
Patient should be at least of age 10 weeks,weight of patient atleast 10 pounds,Haemoglobin of 10,Total count should be <10,000,

Conduction of anesthesia

Ideally, patients with clefts should be managed by experienced cleft teams, in specialist paediatric centres, that are able to provide the required facilities and expertise.

Premedication

Sedative premedication can be considered in the patients who have separation anxiety.
Preoperative analgesia such as oral acetaminophen 15–20 mg /kg is recommended.
Basic noninvasive monitors applied like spo2,NIBP,ECG& temprature.
Preoxygenation with oxygen for 3-5 min done.
Induction : In patients whose airways are not anticipated to be difficult, any anesthetic induction technique can be considered. Our practice involves gaseous induction of anaesthesia using sevoflurane in 100% oxygen with standard routine monitoring (pulse oximetry, ECG, non-invasive arterial BP, capnography, and temperature), although i.v. induction is equally appropriate. Intravenous access, if not already established, is sited as soon as adequate depth of anesthesia is achieved. Additional medications may be given to facilitate tracheal intubation.
The ideal airway device for cleft repair, allowing for optimal surgical access, is a preformed 'south facing' ***Ring-Adair-Elwyn (RAE) tracheal tube (TT),*** inserted orally and secured in the midline below the lower lip .However, the preformed curve of these TTs may not perfectly fit the patient's features; careful placement of the tip, visually at intubation and with auscultation of the chest, is necessary to avoid endobronchial intubation. If the ETT protrudes too far from the bottom lip it is possible to use padding, such as dental rolls or gauze, to build a platform that the ETT can be secured to, to reduce the risk of inadvertent advancement of the tip. If this is still inadequate, a standard reinforced tube can be used as it is flexible enough to follow the contours of the chin but should not kink or obstruct. Infant whose trachea has been intubated with an RAE ETT. This patient has a complete cleft lip and palate, which may be associated with difficult laryngoscopy. A full view of the larynx is difficult to achieve on laryngoscopy and this image demonstrates migration of the tube into the cleft --->.
Cuffed or uncuffed ETTs may be used. Cuffed ETTs are becoming more popular in pediatric patients and confer benefits in shared airway procedures as they provide a truly secure airway and may negate the need for a throat pack. If an uncuffed tube is used, it would be slightly larger than the cuffed tube which allows for ventilation with lower resistance. It would also

be easier if suctioning via the ETT is required. Midline positioning, especially in cleft lip cases, is important for surgical access and ongoing assessment of landmarks. The TT should be secured so that it does not distort the tissues and for palate repair. Having the TT in the midline will prevent the mouth gag from obstructing the ETT when it is positioned.

The difficult airway

***If a difficult airway is anticipated, additional skilled assistants for airway management, optimisation of airway positioning, and a clear plan of approach including having the right equipment is essential. It would be very challenging to intubate these young patients' trachea when they are awake, but this may be the safest option. Most of the time, depending on the level of concern with the airway, gaseous induction with sevoflurane can be performed, or i.v. access can be established while the patient is awake to achieve an appropriate level of sedation before airway manipulation. It is ideal to maintain spontaneous ventilation and ensure ease with bag mask ventilation. Laryngoscopy should not be attempted until the patient is in a deep plane of anaesthesia, achieved by using deep inhalation anaesthesia, a propofol bolus, or spraying the vocal cords with local anesthetic.

There are pros and cons associated with use of neuromuscular blocking agents, especially where bag-mask ventilation may be difficult. Paralysis of the muscles and the cords may facilitate laryngoscopy, but respiratory efforts will be abolished. Direct laryngoscopy with techniques such as backwards, upwards, rightwards pressure (BURP) and the use of gum-elastic bougies are low-cost, simple, and effective techniques that may make the difference between a difficult and a failed intubation. Videolaryngoscopy is increasingly popular and offers a view of oral and laryngeal structures throughout the procedure, especially in patients with difficult airway.

The use of a supraglottic airway device (SAD) as a rescue device not only allows oxygenation of the patient but may also act as a conduit for ‘low skill’ flexible scope-assisted tracheal intubation. The use of an SAD throughout cleft repair has been described but it presents added challenges to the surgical team because of its bulk.Appropriate flexible bronchoscopes can be used for fibreoptic intubation. Finally, an **elective tracheostomy** may be considered in patients with **extremely difficult airways.**In patients with a particularly wide cleft palate, there may be a tendency for the laryngoscope to slide into the cleft offering no view of the laryngeal structures.

Solutions for this include using a straight laryngoscope blade with a lateral approach or **packing the cleft with gauze.**

When considering the ‘cannot intubate, cannot ventilate’ (CICV) scenario, the Paediatric Difficult Airway Guidelines group have written consensus guidelines which are available for further reference.

The shared airway poses many challenges for both surgical and anaesthetic teams. In general, considerations can be summarised with the acronym ‘BADD’—blood, access, debris,dislodgement.

In the case of cleft lip or palate repair, excessive bleeding is not anticipated and significant contamination of the airway is, rare. A throat pack should inserted to contain blood and debris.

There are potential risks of ***circuit disconnect and ETT dislodgement, including endobronchial intubation and unexpected extubation throughout the surgery. Surgical

preparation will include placing the patient on a head ring and a shoulder roll, or alternatively a head ring on a Mayfield device, to extend the neck. The position of the tube should be checked whenever the patient is moved. In Addition, neck extension may exacerbate the leak.figure show pt. of cleft lip& palate having difficult intubation with RAE tube.
For cleft palate surgery, the insertion and adjustment of the surgical (Dingman) mouth gag may displace the ETT or cause an increase in airway pressures. The surgeon may have to move the head or the position of the patient intraoperatively. Insertion or removal of the throat pack may lead to accidental extubation. The patient will be fully draped such that access to the airway, the chest (e.g. to listen to breath sounds), and the i.v. line is limited. Vigilance and open communication with the surgical team are necessary throughout the case.

Maintenance of anesthesia

Anesthesia may be maintained using either volatile agents or TIVA techniques. The addition of fentanyl reduces volatile and propofol anesthetic requirements and may allow controlled ventilation throughout, promoting a smooth, rapid emergence.
But there are 2 schools of theory. The use of muscle relaxation allows perfectly controlled ventilation which may reduce blood loss secondary to tighter control of Paco2.
Spontaneous ventilation is a safe and acceptable technique, especially if there were an inadvertent extubation or disconnection. Total operative time is between 1 and 4 h depending on the repair involved.
Multimodal analgesia for cleft procedures includes enteral and parenteral medication and local anesthetic techniques. Options include
1.perioperative acetaminophen (15–20 mg/ kg oral.),
2.NSAIDs (e.g. ibuprofen 5–10 mg /kgoral or ketorolac 0.5 mg /kg i.v.),
3.lntraoperative opioids (e.g. fentanyl 1–2 μg /kg i.v.), ketamine (0.1–0.2 mg /kg i.v.), and surgical infiltration of local anaesthesia with adrenaline (epinephrine), which has the added benefit of further reducing blood loss. A longer-acting agent such as morphine (20–100 μg/ kg i.v.) may be required and should be titrated to clinical effect, as the risk of perioperative respiratory adverse events (PRAE) is significant and potentially devastating.now a days not used routinely.
4Nerve blocks can be considered for analgesia. Infraorbital nerve blocks are effective for cleft lip repair, and multiple nerve blocks have been described for cleft palate repair including greater and lesser palatine nerve blocks, nasopalatine nerve blocks, and bilateral suprazygomatic maxillary nerve blocks.
5.Dexmedetomidine has several beneficial properties including an opioid-sparing analgesic effect, anxiolysis, and the potential to reduce incidence of emergence agitation with a low risk of respiratory depression. It may have a role when given intraoperatively, balanced with potential adverse effects such as bradycardia and hypotension.
6.Dexamethasone, a synthetic glucocorticoid, has both analgesic and anti-inflammatory effects (0.1–0.2 mg /kg i.v.) and may be useful in cleft surgery. Prevention of postoperative nausea and vomiting by using ondansetron (0.1 mg/ kg i.v.) can be considered.
Surgical blood loss is usually minimal and blood transfusion is rare. Infusion of IV fluids in form of a glucose-containing solution should be considered as patients are generally young with minimal glucose reserves, the surgical time may be relatively long, and they may not resume oral intake immediately after surgery. If there had been a relatively long fasting time,

one may consider replacing the deficit with balanced crystalloid solutions. In addition, strategies to minimize heat loss, including warm ambient temperature and use of forced air blankets, should be considered.

Emergence and recovery

Successful extubation and recovery relies on efficient teamwork and succinct communication between surgical, nursing, and anesthetic teams. The main concerns at the time of extubation, and during postoperative care, include airway obstruction, bleeding, and disruption of the suture lines.Once the surgical procedure is complete the oropharynx should be visualized to assess the **haemostasis**. Careful suction of the airway, including posterior to the palate, to remove any clots may be required. This should always be performed under direct visualization for proper examination of the oropharynx, taking care not to disrupt the delicate suture lines. Once this is done, further instrumentation of the airway should be avoided.

1)Airway obstruction may present at almost any stage during the emergence and **extubation** process. This may be related to pre-existing patient factors such as OSA or the surgical repair may lead to swelling in the oropharynx and nasopharynx. As obligate nasal breathers, infants may experience partial or complete airway obstruction.

2)Other factors to be aware of include the residual effects of the anaesthetic agents, traumatic intubation, blood clots, retained throat packs, tongue swelling, and laryngospasm. Depending on the repair, the surgical team may have already placed a nasopharyngeal airway or nasal stents before tracheal extubation, to be removed a few days later.

3)In selected cases a tongue suture may be used to relieve anticipated airway obstruction in the postoperative period. Arm restraints may be applied to prevent the child disturbing sutures by rubbing their face or exploring their mouths.

The ideal emergence from anesthesia involves

1.The correct timing of full antagonism of any residual neuromuscular block to facilitate a safe extubation, 2.whilst avoiding coughing and straining, which may lead to further bleeding.

3.Extubation may be performed deep or awake depending on the personnel involved and patient-specific risk factors.

4. At this critical moment, oxygen is provided via facemask and the patients' respiratory pattern and effort should be observed carefully for any signs of upper airway obstruction.

5.Although airway problems may occur up to 48 h after surgery, most present in the immediate postoperative period.

***Management of actual or suspected airway obstruction should be timely and appropriate and may include:

Simple airway maneuvers, such as repositioning the child in the lateral or prone position, or the application of CPAP while providing a jaw thrust.

•Insertion of a nasopharyngeal airway, often in discussion with the surgical team as to which nostril, depending on the repair. The use of oropharyngeal airways is not contraindicated but is likely to cause more damage to the suture lines than a nasal airway.

•Reintubation may be required if the above interventions are ineffective and this may be more difficult than the initial intubation because of potential swelling, bleeding, and the surgical repair. There may be tongue swelling secondary to prolonged use of the surgical mouth gag. If the patient is on the ward and reintubation is deemed necessary, if time allows,

***one should consider airway intervention after transfer back to the theatre where the necessary equipment and assistance are easily accessible in a familiar environment.

Postoperative care

Patients should be monitored in the PACU with supplementary oxygen until fully awake and settled, often in the lateral or semi-prone position to allow drainage of any secretions. Rescue analgesia may be required; the i.v. route is available for acetaminophen, NSAIDs (if not already given), and opioids (e.g. fentanyl 1 μg/kg) with appropriate monitoring. The patient's ultimate postoperative destination may vary from a standard ward to pediatric intensive care (PICU) depending on the surgery, preoperative and intraoperative status, and comorbidities. For **cleft palate** surgery, close monitoring is required for the first 12–24 h to identify potential airway obstruction, bleeding, or both.

Discharge time (Mean hospital stay)

Cleft lip patients may stay overnight and usually return home the next day,
cleft palate patients are usually discharged between Day 1 and Day 3 after surgery.

** The main determinant of discharge is to ensure patients are feeding well. Oral medications including acetaminophen, ibuprofen, and morphine may be prescribed for analgesia at home and surgical follow-up will depend on the procedure completed and where the patient is on their surgical pathway.

Follow-up and long-term expectations

Patients with cleft lip repair are brought back for suture removal within a week if non-absorbable sutures are used. This is an outpatient procedure that involves a brief general anesthetic. Once completely repaired, most children with an isolated cleft lip do not require further follow-up. It is interesting to note that once a cleft palate is successfully repaired, nasal intubation is not contraindicated provided a pharyngoplasty has not been performed. The soft tissue will have healed within approximately 6 weeks of the primary repair.

***Any patients with clefts involving the alveolus will need an alveolar bone graft at intermediate dentition (approximately 10 yrs) and those with continued velopharyngeal insufficiency may require a repeat palate procedure.

As patients may require multiple surgeries, from primary closure as an infant, to alveolar bone grafting and dental surgery into adulthood, it is imperative that we understand the anesthetic concerns relating to various development stages of the patient and the surgical considerations. The psychological impact of having to undergo repeat procedures should also be considered when interacting with these patients and their families.

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