Cleft lip and palate are the commonest craniofacial abnormalities. A cleft lip, with or without a cleft palate, occurs in 1 in 600 live births. A cleft palate alone, is a separate entity and occurs in 1 in 2000 live births. Many complex classifications have been devised but essentially the cleft can involve the lip, alveolus (gum), hard palate and/or soft palate and can be complete or incomplete, unilateral or bilateral.

Embryologically, clefts arise because of failure of fusion or breakdown of fusion between the nasal and maxillary processes and the palatine shelves that form these structures at around 8 weeks of life. Without repair these children suffer from facial disfigurement and potentially social isolation, feeding problems and abnormal speech. Surgical repair of a cleft lip is usually undertaken at around 3 months of age for cosmetic reasons, although there is now a trend to do the operation in the neonatal period in Western countries. Correcting the defect early is popular with parents and facilitates bonding and feeding. The timing of cleft palate repair is a balance between poor facial growth with an early repair and poor speech development with a repair after the age of 1 year. It is usually done at about 6 months of age in developed countries. Cleft lips and palates are often done much later in less affluent countries.

For surgical repair of clefts to be performed safely requires a team approach. A surgeon wrote in 1912 that ‘the difference to the surgeon, between doing a cleft palate operation with a thoroughly experienced anaesthetist and an inexperienced one, is the difference between pleasure and pain!’ The majority of anaesthetic morbidity related to these procedures relates to the airway: either difficulty with intubation, inadvertent extubation during the procedure or postoperative airway obstruction. The optimum anaesthetic management will depend on the age of the patient, the availability of intraoperative monitoring equipment, anaesthetic drugs and expertise, and the level of postoperative care that is available.

Table 1:

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Major features</th>
<th>Anaesthesia problems</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pierre Robin Syndrome</td>
<td>Cleft palate</td>
<td>Difficult intubation</td>
</tr>
<tr>
<td></td>
<td>Small jaw</td>
<td>Chronic airway obstruction</td>
</tr>
<tr>
<td></td>
<td>Small jaw and mouth</td>
<td>Airway and intubation difficulties (tend to get more difficult to intubate as they get older)</td>
</tr>
<tr>
<td></td>
<td>Glossoptosis</td>
<td></td>
</tr>
<tr>
<td>Treacher Collins Syndrome</td>
<td>Small jaw and mouth</td>
<td>Airway and intubation difficulties (tend to get more difficult to intubate as they get older)</td>
</tr>
<tr>
<td></td>
<td>Choanal atresia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ear and eye abnormalities</td>
<td></td>
</tr>
<tr>
<td>Goldenhar Syndrome</td>
<td>Hemifacial and mandibular hypoplasia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Abnormalities of the cervical spine</td>
<td></td>
</tr>
<tr>
<td></td>
<td>External ear and eye abnormalities</td>
<td></td>
</tr>
</tbody>
</table>
opioid analgesics. Where available an ECG, echocardiogram and overnight saturation monitoring preoperatively will quantify the problem. However surgery is the treatment and most teams operate observing the child closely postoperatively, if possible in ICU.

● **Anticipated difficult intubation.** A difficult intubation is especially common in patients less than 6 months of age with either retrognathia (receding lower jaw) or bilateral clefts.\(^3\)

● **Nutrition/hydration.** Because of potential difficulty with feeding, the state of hydration and overall growth needs to be assessed. A haemoglobin concentration should be checked and blood sent for cross matching although the need for transfusion is uncommon. There is a physiological decline in haemoglobin concentration after birth, which is at a maximum between 3 and 6 months of age. This is due to the change from fetal to adult haemoglobin. Nutritional anaemia is also common, especially in the developing world. Ideally all patients should have a haemoglobin concentration above 10g/dl. Clear fluids can be given up to two hours preoperatively and exclusively breast fed young infants can feed until four hours preoperatively.

● **Need for premedication.** Sedative premedication is not indicated in infants with cleft palates and should be avoided because of the risk of airway obstruction. Atropine may be prescribed to dry oral secretions and block vagal reflexes but the tachycardia produced makes it more difficult to assess anaesthetic depth and the intravascular volume status during the procedure. Anaesthetic techniques employing ether or ketamine or where particular difficulty with intubation is anticipated benefit from atropine premedication. A good rapport needs to be established with older children and parents.

**Intraoperative Management**

Induction of anaesthesia is most safely performed by inhalational anaesthesia with halothane or sevoflurane. Intravenous access is gained when an adequate depth of anaesthesia is achieved and endotracheal intubation performed either under deep volatile anaesthesia or facilitated by suxamethonium or a non-depolarising neuromuscular blocking agent. No neuromuscular blocking agents should be given until one is sure that the lungs can be ventilated with a mask.

Endotracheal intubation may be difficult, especially in children with a craniofacial syndrome, and a variety of techniques such as blind nasal intubation, fibreoptic intubation, the use of bougies or retrograde techniques may need to be employed. An oral, preformed RAE tube is usually chosen and is taped in the midline. For palatal surgery, a mouth gag that fits over the tube is used to keep the mouth open and the tongue out of the way. Problems with the endotracheal tube are common. It may be pulled out, pushed into the right main bronchus when the head is moved or kinked under the mouth gag. After the patient has been finally positioned for surgery, check the patency and position of the endotracheal tube by auscultation and by gentle positive pressure ventilation to assess airway resistance.

Maintenance of anaesthesia with an inhalational agent can be with spontaneous ventilation or controlled ventilation. A spontaneous breathing technique with halothane provides an element of safety in the event of accidental disconnection or extubation but is not suitable in very young infants.

Controlled ventilation with muscle paralysis allows for a lighter plane of anaesthesia and more rapid awakening with recovery of reflexes and the lower PaCO\(_2\) probably causes less bleeding. It is usual for the surgeon to inject local anaesthetic and adrenaline into the surgical field to reduce blood loss and improve the surgical field. It also provides some intraoperative analgesia. Limiting the dose of adrenaline to 5mcg/kg in the presence of normocapnia (can only be guaranteed if the patient is ventilated) and halothane is normally safe.\(^4\)

Both palates and lips should either receive paracetamol 20mg/kg orally as premedication or rectal paracetamol post induction (40mg/kg) so that adequate paracetamol levels are attained by the end of surgery. Local anaesthetic infiltration provides useful intraoperative analgesia but cleft palates benefit from careful use of intraoperative opioids. Morphine sulphate 0.1-0.2mg/kg intravenously is commonly used and provides good early postoperative analgesia. The use of opioids results in a smoother emergence and less crying on extubation. This reduces trauma to the airway and decreases the risk of postoperative bleeding. A small dose of intraoperative morphine or fentanyl may be used for cleft lips but the attraction of bilateral infraorbital nerve blocks in this population is that they produce excellent intra- and postoperative analgesia and no respiratory depression. These nerve blocks are especially useful if a spontaneously breathing technique is used to repair cleft lips in young infants. Intraoperative and postoperative opioids are then not required (see inset for description of technique). NSAIDS, although very effective analgesics, may increase the risk of early postoperative bleeding. Their use should probably be delayed until at least twelve hours postoperatively. Anaesthetising a briskly bleeding cleft palate that has had to return to theatre can be a real challenge!

Although there is the potential for the blood loss to be significant enough to require blood transfusion, a better awareness of the risks of blood transfusion, especially the risks of transmission of infectious diseases has meant that this practice is less common than it used to be. The risks of transfusion need to be weighed against the expected benefits in every case. Blood transfusion of cleft lip repairs should be extremely uncommon but cleft palates will occasionally require blood transfusion.

Appropriate intravenous fluids should be given, taking into account the period of preoperative starvation, intraoperative and postoperative maintenance requirements and blood loss. Most surgeons allow early postoperative oral intake. Attention to temperature control is always important in paediatric patients but because of the extensive draping and little exposure during this operation, heat loss is rarely a problem.

**Extubation**

Acute airway obstruction is a very real risk at the end of the procedure following extubation. The surgeon needs to remove
the throat packs and ensure that the surgical field is dry. Suctioning should be kept to a minimum to avoid disrupting the surgical repair. Oropharyngeal airways are best avoided, if possible. Extubation should be undertaken only after the return of consciousness with protective reflexes intact. A tongue stitch will often be placed in patients with preoperative airway obstruction. This pulls the tongue forward away from the posterior pharyngeal wall as a treatment for postoperative airway obstruction.

Postoperative Management

These patients need to be closely observed in recovery for evidence of blood loss or airway obstruction and only returned to the ward when fully awake. Supplemental oxygen should be given until the child is fully awake and additional analgesia (intravenous morphine) can be carefully titrated to effect.

Postoperative analgesic regimes need to take into account where the child will be nursed. Cleft lips (especially those who received infraorbital nerve blocks) will only require rectal or oral preparations of paracetamol or NSAID’s. Cleft palates should receive adequate doses of paracetamol and possibly oral codeine or NSAID’s after twelve hours. Ideally these patients should be returned to a high dependency area with experienced staff and oxygen saturation monitoring. Only then is the administration of postoperative morphine for analgesia safe. A low dose morphine infusion is the most predictable and titratable form of analgesia safe. Airway complications need to be closely observed in recovery for evidence of blood loss or airway obstruction and only returned to the ward when fully awake. Supplemental oxygen should be given until the child is fully awake and additional analgesia (intravenous morphine) can be carefully titrated to effect.

Infraorbital Nerve Block

The infraorbital nerve is a terminal branch of the trigeminal nerve. It supplies sensory innervation to the skin and mucous membrane of the upper lip and lower eyelid, the skin between them and to the side of the nose. It can easily be blocked as it emerges from the infraorbital foramen, just medial to the buttress of the zygoma (bony prominence immediately lateral to the nose). In adults, the infraorbital foramen is in line with the supraorbital notch and mental foramen or the second upper premolar tooth. In neonates these landmarks are difficult to palpate or absent. Bosenberg performed an anatomical study on neonates that showed that the infraorbital nerve lies halfway between the midpoint of the palpebral fissure and the angle of the mouth, approximately 7.5 mm from the side of the nose. The nerve is blocked by inserting a needle perpendicularly to the skin and advancing it until bony resistance is felt. The needle is then withdrawn slightly and 1-2mls of 0.5% bupivacaine and 1:200,000 adrenaline is injected after performing a negative aspiration test. The needle should not enter the infraorbital foramen.

In many countries, because of health service constraints, cleft lips and palates are not repaired as infants and many hospitals do not have oxygen and volatile anaesthetic agents. In these situations a very different approach to the anaesthetic management of these cases may be indicated. Airway management is of paramount importance and any surgical treatment requires an experienced surgeon and anaesthetist used to working in the local environment.

Cleft lips in older children and adults can be repaired under local anaesthesia alone with good preoperative explanation and an experienced surgeon. Intramuscular ketamine or intravenous boluses of ketamine with atropine can be used without intubation for a cleft lip repair in children over 12 months. This is only advisable if pulse oximetry is present and all equipment to intubate and ventilate is immediately available.

All patients undergoing cleft palate repair should be intubated. Techniques to maintain anaesthesia in this situation include volatile agents (halothane or ether) or total intravenous anaesthesia with ketamine and muscle relaxants. Either spontaneous or controlled ventilation have been used successfully, but it may be safer to ventilate or at least support ventilation in smaller children or those undergoing prolonged surgery.

Small children and patients for a palate repair require careful attention to detail. If halothane and oxygen are both available then a gas induction is the safest method in all children for a cleft palate repair. Following induction and intubation, small children, particularly infants, are best ventilated by hand. Since capnography is rarely available, manual ventilation provides an excellent means of detecting any change in respiratory resistance. This may occur if the endotracheal tube is occluded by the gag, displaced, disconnected, or blocked by sputum. Non-depolarising relaxants allow IPPV with a light plane of anaesthesia and rapid awakening. If necessary however, small children and infants can usually be ventilated without muscle relaxants. Intermittent boluses of suxamethonium can be used although care must be taken that the total dose does not exceed 8mg/kg body weight for the entire procedure.

If ether is the only volatile agent available, then an inhalational induction is extremely difficult and time consuming. Intramuscular ketamine and atropine provide an alternative. Airway reflexes are preserved and there is time to obtain intravenous access. If a difficult intubation is suspected or muscle relaxants are not available, ether can then be introduced and the patient deepened until the child can be intubated. This procedure requires time, expertise and patience. The alternative is to attempt to hand ventilate after ketamine anaesthesia is established and if this can be done easily, suxamethonium can be given to facilitate intubation.

Intraoperative and postoperative analgesia can be provided with infiltration of local anaesthetic and adrenaline directly into the surgical field or with the use of infraorbital blocks and regular paracetamol syrup on the ward postoperatively. If patients are nursed on large over-crowded, understaffed wards then opioids are best avoided. In view of the high incidence of postoperative airway complications it is safer to recover the patients in or adjacent to the operating theatre until they are fully awake.

Postoperative care facilities vary widely and it is crucial that adequate provision is made including trained staff, suction, airway equipment in a well lit environment.

With a knowledge of the potential pitfalls and careful case selection safe anaesthesia can usually be provided by those with experience of working in difficult circumstances for these challenging cases.

References


