Anaesthesia for a Child with Down syndrome Undergoing Cleft Palate Repair and Review of Anaesthesia Management of Other Associated Congenital Syndromes

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Abstract: Cleft lip/palate are the most common craniofacial anomalies in children, with an incidence of 1:800 live births. It occurs due to the failure of fusion or break in fusion of nasal and maxillary processes with the palatine shelves, which form during 8th week of the embryonic period. About 150 syndromes may be associated with cleft deformities. The most well-known are the Pierre Robin’s, Treacher Collins and Goldenhar syndrome. Congenital heart disease (CHD) occurs in 5-10% of these patients. Surgical repair of cleft lip is usually done at 1-3 months of age for cosmetic purpose and cleft palate at 6 months to 1 year of age to promote facial growth and the speech. The successful outcome following cleft repair depends on the age of the patient, associated morbidities, anaesthetic expertise and post-operative care. Infants with facial deformities are usually associated with abnormal dentition/hearing defect, recurrent ear/upper respiratory tract infection (URTI), pulmonary aspiration and poor nutrition. Until recently criteria for cleft repair in infants was 10 pounds of weight, 10 weeks of age and haemoglobin of 10gm%. Recent concepts of early repair in neonates are based on improvements in parent-infant bonding, feeding, growth and speech development.

Anaesthesia for cleft surgery in infant and children carries a higher risk with general anaesthesia and airway complications due to associated respiratory problems. Review of literature mentions higher incidence of perioperative respiratory complications when associated with the common cold symptoms in children for cleft repairs. Morbidity during general anaesthesia is associated with the difficult airway, endotracheal (ET) tube compression/disconnection and post-operative airway obstruction. Down syndrome or Trisomy 21 is the most common chromosomal abnormality, and similar to KFS it also results in anatomical changes of the airways, such as: cleft lip and palate, narrow nasopharynx, and relatively large and protruberating tongue. The larynx and the cricoid ring tend to be small predisposing to acquired subglottic stenosis. Patients might have atlantoaxial subluxation, making neck extension risky.

Here we are presenting a 5 yr old male child with weight of 12kg for cleft palate repair with Down syndrome done under general anaesthesia. He was having complete cleft palate was seen with a bifid uvula. The cardiovascular, respiratory and per abdominal examinations had no positive findings. However, CNS examination showed hypotonia in all four limbs with power 2/5 in upper limbs and lower limbs.

Keywords: Down syndrome, cleft palate, general anaesthesia

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I. Introduction

Cleft lip/palate are the most common craniofacial anomalies in children, with an incidence of 1:800 live births. Cleft palate alone occurs in 1:2000 live births. It occurs due to the failure of fusion or break in fusion of nasal and maxillary processes with the palatine shelves, which form during 8th week of the embryonic period. About 150 syndromes may be associated with cleft deformities. The most well-known are the Pierre Robin’s, Treacher Collins and Goldenhar syndrome. Congenital heart disease (CHD) occurs in 5-10% of these patients. Down’s syndrome is the commonest chromosomal abnormality and is named after John Langdon Down, who described the syndrome in 1866. Down’s syndrome is also referred to as Trisomy 21 due to the presence of an extra copy of chromosome number 21. It has an incidence of 1.5 per 1000 live births. Maternal age is a risk factor for Down’s syndrome and the risk increases with maternal age. A 20 year-old mother has a risk of about 1 in 2000 while a 35 year-old mother has an estimated risk of 1 in 400. The risk increases to 1 in 40 at 45 years of age. This syndrome affects many organ systems, and there is impaired global development. Many of these problems have significance for the anaesthetist.

II. Case Report

A 5 yr old male child with weight of 12kg presented to our hospital in the pediatric department with complaints of difficulty in swallowing since birth and speech defect. On taking history it was revealed that the patient was delivered at home at term and cried immediately after birth. Since birth the child was unable to take feeds orally, as whatever was fed to him regurgitated through his nose. He was taken to a local physician who diagnosed him with cleft palate and advised surgery at 5 years of age. Child also had history of recurrent upper respiratory tract infections. His gross and fine motor, language and social milestones were all delayed. On general physical examination showed his weight and height was below the third percentile. The child had a flat occiput with parietal and frontal bossing. He had an upward slant of eyes with epicanthal folds. He also had a flat nasal bridge. Oral examination showed an open mouth (fish shaped) with a large tongue. A complete cleft palate was seen with a bifid uvula. The cardiovascular, respiratory and per abdominal examinations had no positive findings. However, CNS examination showed hypotonia in all four limbs with power 2/5 in upper limbs and lower limbs.

Image 2-Cleft Palate

With the given history and the positive examination findings a diagnosis of down syndrome with cleft palate was made. Routine blood tests were ordered during pre anesthetic check up which were normal. Special tests included 2D echo to rule out associated congenital cardiac condition, thyroid profile test to rule out hypothyroidism both of which had a normal report. X ray neck was ordered with special emphasis on the atlanto axial joint to rule out atlanto axial ligament laxity which is commonly seen in patients with down syndrome. X ray revealed no abnormal findings. The child was accepted under ASA II and posted for surgery. Pre-operative fasting was observed for 4 h for milk, 6 h for solid food and 2 h for clear fluid. Baseline vital parameters like heart rate (HR), non-invasive blood pressure (NIBP), (ECG), pulse oximetry (SpO2) were noted inside the O.T. Pre cordial stethoscope fixed. All the anaesthesia equipments and drugs were checked. The ET tubes (Ring, Adair and Elwyn (RAE) south polar tube for cleft lip, Oxford/Armoured for cleft palate) and LMA's of appropriate sizes were kept ready. Premedication given was inj. glycopyrrolate 0.04mg, midazolam 0.12mg and inj. Fentanyl 25µg IV. Diclofenac suppository (25mg) was put per rectally as preemptive analgesia. Patient was induced with inj. Thiopentone 60mg and succinylcholine 25mg IV. After 30 secs of bag and mask ventilation with oxygen patient was
intubated with 4.5no. uncuffed Southpole RAE tube. Tube was fixed after checking bilateral air entry. Throat packing was done. Maintenance of anaesthesia was carried out with manual ventilation with bag and mask with oxygen, nitrous oxide, sevoflurane and atracurium. Inj. dexamethasone 2.5 mg was given to reduce post operative intraoral oedema. Patient received 300ml of isolytePand 100 ml of colloid(HAES– 3%). Patient had an urine output of 85ml and total 100ml of blood loss. The intraoperative period was uneventful.

Image 3-Intraoperative Image 4-Postoperative
At the end of surgery, patient was reversed with inj.neostigmine 0.6mg and inj.glycopyrrolate 0.08mg. Throat pack was removed and thorough ET and oral suctioning was done. Patient was extubated when the child was fully awake and his reflexes were intact. Child was put in left lateral position after extubation and was supplemented with oxygen at 4 liters/minutes with Hudson mask. After 30 minutes of observation the patient was shifted to PICU for post operative care. Patient had an uneventful post operative period and was shifted out of PICU to ward after 24 hours. The child was discharged after 7 days of hospital stay.

Image 5-Post operative image of the patient

III. Discussion
Twenty percent of all children with a cleft will have some other congenital abnormality, most of which are not part of a recognizable syndrome. If classified by the organ system involved, malformations of the upper and lower limbs and vertebrae are the commonest group, being present in 5% of all cases. Usually these are minor abnormalities (accessory digits or club foot deformities) and do not affect anaesthetic management. Abnormalities of the cardiovascular system are the second commonest group, affecting 4% of all cleft children, a 16-fold increase in prevalence over the general population, with ventricular septal defects being the commonest single abnormality seen.

More than three hundred syndromes have now been described involving the combination of a cleft with multiple other congenital abnormalities. The syndromes of the head and neck, including those involving abnormalities in branchial arch development, are particularly important. They are commonly difficult to intubate, and therefore will be individually described. Treacher Collins syndrome is a bilateral symmetrical abnormality of
the first and second branchial arches, resulting in hypoplasia of the maxilla, zygoma and mandible. Intubation is difficult due to the small jaw and retroposition of the tongue. Pierre Robin sequence (PRS) is not an individual syndrome, but a group of conditions involving abnormal development of the mandible (a derivative of the first branchial arch) in utero.

It consists of:
(a) micrognathia (a small symmetrical retro-positioned jaw)
(b) glossoptosis (tongue displaced high and posteriorly due to the small jaw, causing airway obstruction)
(c) mid-line U shaped cleft of the palate.

At birth, the facial features of PRS are often quite marked, and severe airway obstruction and feeding difficulties common. Soft nasopharyngeal airway sheathed in place by acrylic splints will often maintain an airway in hospital, allowing time for spontaneous improvement to occur as the infant grows. Many infants have PRS in isolation, with no other abnormalities. Here, the small mandible is felt to be due to external restriction to growth in utero, and significant catch up growth is possible during early childhood, if airway and feeding problems are treated. In other infants the small jaw is due to intrinsic abnormality of the mandible, as part of a syndrome, and catch up growth post-delivery will not be possible. These infants have other abnormalities suggestive of an underlying syndrome.

The commonest are:
(a) Stickler’s syndrome.
(b) Velocardiofacial syndrome.
(c) Foetal alcohol syndrome.

Stickler’s syndrome is a connective tissue dysplasia of autosomal dominant inheritance with variable penetrance. Ophthalmic problems including retinal detachments are common and thus all children with PRS should undergo ophthalmic review. Velocardiofacial syndrome (VCF) is a genetic condition characterized by structural and palatal abnormalities, cardiac defects, unique facial characteristics and mild learning difficulties. Although VCF syndrome is the most common clefired syndrome, associated with least 8% of all cleft palates, it was first described as recently as 1978 by Dr. Robert Shprintzen of the Montifio Medical Centre, Bronx, New York. It is the second commonest syndromic cause of congenital heart disease after Down’s syndrome. The resulting phenotype is very wide. The underlying chromosomal abnormality is a deletion in position 11 of the long arm of chromosome 22 (22q11 deletion), picked up by chromosomal fluorescence in situ hybridization (FISH test).

Hemifacial microsomia (Goldenhaar’s syndrome) is a relatively common birth defect involving poor development of the first and second branchial arches on one side. This leads to facial asymmetry with a small mandible, maxilla and zygoma on one side of the face. Most cases are sporadic. Children with Down’s syndrome are generally gentle, cheerful and outgoing, but they may have a tendency towards hyperactivity, and will have limited understanding compared to normal children of their age. It is best to conduct the pre-anesthetic visit when the parent or carer is with the child. The anaesthetist should try to explain (to the child) what is going to happen in a language appropriate to the child; the presence of the parent or carer will be greatly reassuring for the child. It is important that the parent has a clear explanation of the anaesthetic procedure and what to expect.

Play therapists may also have a role to ensure smooth induction of anaesthesia. Structural heart disease in children with Down’s syndrome is common and there should be a high index of suspicion. Symptoms suggestive of congenital heart disease include failure to thrive, breathlessness and fatigue on exertion, or unexplained ‘funny turns’. Important signs include central cyanosis, finger clubbing, respiratory distress, signs of cardiomegaly with displaced cardiac apex, hepatomegaly or the presence of a heart murmur, possibly with associated ‘thill’ (palpable murmur).

The characteristics of pathological murmurs are as follows:
• All murmurs associated with cardiac signs or symptoms
• All pansystolic and diastolic murmurs
• Late systolic murmurs
• Loud murmurs, those associated with a thrill, or continuous murmurs

A detailed cardiovascular examination, ECG, and ideally a cardiology opinion and echocardiography should be obtained in all children with Down’s syndrome before proceeding with surgery. A thorough examination of the respiratory system and airway is necessary to rule out a difficult airway or intubation. History of snoring during sleep associated with daytime lethargy and somnolence, behavioural changes, poor concentration and inattention at school may suggest the presence of severe obstructive sleep apnoea (OSA). Children with severe OSA are not suitable for day case surgery. Atlanto-axial instability is seen in 15% of children with Down’s syndrome. History of neck pain, limited neck mobility or head tilt, change in gait pattern, clumsiness, abnormal neurological reflexes, abnormal sensation or presence of bladder and bowel dysfunction suggest the possibility...
of neck problems. Atlanto-axial instability is diagnosed by identifying an increase in the distance between the posterior border of the arch of the atlas and the anterior border of the odontoid peg on lateral flexion and extension radiographs of the cervical spine (atlantodental interval, ADI) [see fig 1,2]. In children over eight years old, the ADI should be 3 mm or less while in younger children the ADI should be 4 mm or less (some consider up to 5 mm as normal)20.

![Figures 1 & 2. Line diagram of the atlas (C1) and axis (C2) vertebrae showing increased atlantodental interval (shaded area) in extension. The odontoid peg (dens) of C2 projects from the superior surface of the body of C2 and may move to cause spinal cord compression21.](image)

Currently, there is no consensus in the literature to suggest whether every patient with Down’s syndrome should be screened radiologically prior to an anaesthetic/surgical procedure. However, it is recommended that if there are signs and symptoms suggestive of cervical cord compression or a difficult laryngoscopy is anticipated or if the surgery requires that the neck is placed in a non-neutral position for a long time intraoperatively, then cervical spine radiography should be performed before an elective case22. If cervical spine radiography is not done in an asymptomatic Down’s syndrome patient, then the goal should be to keep the head and neck in an neutral position.

In a study looking into the safety of neck rotation for ear surgery, Todd et al have concluded that patients with Down’s syndrome who are neurologically intact with normal neck radiographs do not appear to be at high risk with neck rotation up to 60 degrees23. However, there has been a case report where the ADI has been normal prior to surgery but post operatively the ADI increased, requiring upper cervical fusion at a later date22.

Despite cleft repair being a relatively common operation worldwide, there are no prospectivestudies comparing different anaesthetic techniques. Many papers describe the use of specific drugs, usually reflecting the agents available locally, and use mortality as a measure of anaesthetic success. Despite this lack of information there is general agreement on the principles (if not the detail) of anaesthetic management of the infant presenting for primary cleft repair.

(a) Pre-operative assessment

Repair of a cleft involves sharing the airway with the surgeon. To provide the best possible access to the operative site the infant will need to be intubated with a south facing tube, and ventilation controlled in view of the age of the patient and likely duration of the surgery. Two key questions dominate the pre-operative assessment of any child presenting for surgery:

(a) Is another congenital abnormality or a syndrome present?

(b) Is intubation likely to be difficult?

In view of the high incidence of congenital heart disease, all infants require thorough examination of the cardiovascular system. The presence of a murmur warrants an echocardiogram to assess the structure of the heart. The multi-specialist nature of cleft care in the United Kingdom means that many infants with underlying syndromes will have been picked up prior to surgery and the extent of associated abnormalities identified. The methods of airway assessment used in adults to predict the degree of difficulty with intubation are not useful and cannot readily be applied to infants. No specific thyromental distance has been validated as being a cut off for a difficult intubation, and clearly infants cannot cooperate with Mallampati scoring. The majority of non-syndromic infants are easy to intubate and, even if the larynx is difficult to view, are usually straight forward to ventilate. Gunawardana has prospectively reported a series of 800 cleft patients managed over a 10-year period11. Difficult laryngoscopy (Cormack and Lehane Grade III or IV once cricoid pressure had been applied) was present in 7% of patients, and was associated with retrognathia, age less than 6 months and bilateral cleft with prominent premaxilla. None of these patients was difficult to ventilate. The incidence of failed intubation was 1%. However, two points are important when considering this series from Sri Lanka. Firstly, no child was known to have a syndrome, and secondly a straight laryngoscope blade was not tried in any patient.
In the developed world, more children with syndromes survive and present for cleft surgery. The physiological changes in any syndromemust be fully understood before an anaesthetic is given. Despite the lack of a definitive test for difficult intubation, children should be examined pre-operatively for signs of retrognathia. The history must also include questions to pick up any breathing problems or feeding difficulties since birth, how they were managed and whether they have resolved. A diagnosis of PRS, Treacher Collins syndrome or hemifacial microsomia must be assumed to predict difficult intubation until proved otherwise. Standard starvation policies are followed and this author does not routinely use an atropine premedication.

(b) Anaesthetic management

Either an intravenous or gaseous induction of anaesthesia may be undertaken. In the age group 3 months to 1 year, when venous access is difficult, gentle gaseous induction, using a cupped hand and T piece, with either halothane or sevoflurane in oxygen, usually allows smooth loss of consciousness and separation from the parent. If intubation is expected to be straightforward, and once it is confirmed that ventilation may be easily assisted, administration of a non-depolarizing muscle relaxant will facilitate the process. An adequate view of the larynx is usually easily achieved with gentle cricoid pressure and a choice of curved and straight laryngoscope blades. A paediatric gum elastic bougie is the most useful aid to intubation. Care must be taken not to bruise the lip or palate before repaired. The tendency of the laryngoscope to fall into a wide cleft may be overcome in two ways—packing the lip cleft with a small gauze roll or using the laryngoscope to approach from the side of the mouth rather than in the midline.

Where intubation is expected to be difficult, the same initial approach as above may be taken. The choice then is to either keep the infant spontaneously breathing and view the larynx under deep inhalational anaesthesia, or to confirm that ventilation may be easily assisted, and then view the larynx. In any child with severely abnormal facial anatomy, maintaining spontaneous ventilation is the safest approach. If the larynx remains difficult or impossible to view, a range of approaches is possible. Inserting a laryngeal mask airway (LMA) provides an alternate airway, a route to deliver anaesthetic agents, and a conduit through which a fibre-optic laryngoscope may be inserted to view the larynx. A guide wire may then be passed into the trachea. The fibre-optic laryngoscope and LMA are then removed, and the endotracheal tube passed over the wire into the trachea. The guide wire needs to be stiff enough or it will be displaced as the endotracheal tube is pushed over.

Alternatively, with spontaneous ventilation, an endotracheal tube may be cut to length, placed as a nasopharyngeal airway, and anaesthesia maintained via one nostril while intubation is performed with the fibre-optic laryngoscope via the other. Multiple other methods of management have been described. Once the airway has been secured, ventilation is maintained control to maintain normocapnia, thus avoiding vasodilatation and excess blood loss from the operative site.

For a lip repair, intra- and postoperative pain relief may be achieved with local anaesthetic block of the infraorbital nerves. The site of the infraorbital foramen varies with age, and concerns about the close proximity of the eye inn the neonate. Dr. Rosenberg has proposed inserting a local anaesthetic through the skin. This author prefers a more classical “dental approach”, inserting the needle between the lip and gum into the sulcus, with a finger placed anteriorly to confirm the extent of needle advancement, as local anaesthetic is administered. This block, in association with regular oral analgesia, provides effective pain relief. A loading dose of paracetamol 20 mg/kg rectally is recommended in infants and should be administered as early as possible. It takes 2 h for a rectal dose of paracetamol to reach peak serum concentration, and a further hour for it to maximally to pass across into the brain (effect site).

Clinical experience suggests that the addition of non-steroidal anti-inflammatory drug (NSAID), prescribed regularly, improves analgesia with no increased incidence of postoperative haemorrhage requiring a return to theatre. For a CP repair, a Dingman retractor or Dottmouth guard are used to keep the mouth open and hold the tongue and endotracheal tube away from the surgical field. As the gag is being opened, it is important to check that the endotracheal tube is not being crushed or occluded. Armoured endotracheal tubes are not recommended as they are readily displaced. A small gauze throat pack is placed by the surgeon to reduce aspiration of blood. Following induction, intravenous antibiotic prophylaxis, particularly against Staphylococcus, is important.

The nerve supply to the hard and soft palate is from the greater and lesser palatine nerves passing through the sphenopalatine ganglion. There is no simple, long-lasting block that provides effective postoperative analgesia after CP repair without risking interference with swallowing. Although surgical infiltration is undertaken with short acting local anaesthetic and vasoconstrictor, it rarely blocks the profound autonomic response caused by the operation, nor provides effective post-operative pain relief. Short acting opioids (fentanyl 3 mcg/kg for a 1 h procedure) in association with an inhalational agent effectively block a major part of the autonomic response. This means that intraoperative blood pressure is well controlled, providing a relatively bloodless surgical field and reducing the need for blood transfusion. Remifentanil by infusion in combination...
with an inhalational agent has been shown to provide intraoperative haemodynamic stability in infants undergoing cleft repair, but no specific advantage over other short-acting opioids has been demonstrated. With all long-acting opioids, intravenous morphine is required in the initial postoperative period. Regular administration of paracetamol and NSAIDs is also valuable.

When a Cleft palate has been repaired an infant’s airway is smaller than pre-operatively. Obstruction to the upper airway may occur at the time of extubation.

Causes include:

1. Critical reduction in size of a previous tenuous airway.
2. Excessive sedation from opioids, so that infant fails to adequately hold the tongue forward.
3. Laryngeal oedema due to a large endotracheal tube, resulting in stridor.

Ideally, extubation should only take place once the infant is fully awake and has full control of its airway reflexes. Few prospective series exist, but a 5% rate of immediate upper airway obstruction on extubation has been reported, and occurs particularly in children with an associated syndrome (especially PRS). Very gentle placement of nasopharyngeal airway is frequently effective, providing a clear airway, a route to undertake suction and, if inserted gently, not prejudicing therepair. It is usually needed for less than 24 h. Occasionally re-intubation and ventilation is required. Episodes of late upper airway obstruction, up to 24 – 48 h post-operatively, where re-intubation and ventilation was necessary, are also well described. Causes include swelling of the tongue due to ischaemia from excessive retraction by the gag. All at risk infants should therefore be observed closely in the initial postoperative period for the development of upper airway obstruction.

IV. Conclusion

Down’s syndrome is a common congenital anomaly which is associated with multi-system problems that need thorough assessment pre-operatively. Of particular importance is the assessment of airway, cervical spine (forsigns and symptoms of atlanto-axial instability), cardiovascular and respiratory system. Children may present with varying degrees of learning disabilities. Anaesthesia should be carefully planned and the risks discussed with the parents/caregivers. Peri-operative complications include airway obstruction, difficult intubation, bradycardia, post-extubation stridor, bronchospasm and rarely, neurological problems due to atlanto-axial subluxation. With adequate pre-operative preparation and good intra-operative care, children with Down’s syndrome can be anaesthetised safely. Post-operatively, the children should be closely observed in the recovery room and adequate analgesia and anti-emetics should be prescribed to ensure that the child is pain free and comfortable.

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