

Anesthetic Management of Pediatric Patients with Down Syndrome

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INTRODUCTION

Down syndrome, or trisomy 21, is a common congenital abnormality associated with characteristic morphological features, impaired intellectual development and disorders of multiple organ systems with a broad spectrum of severity. It is the most prevalent genetic disorder worldwide, affecting more than 1 in 800 live births [1], and remains the most commonly encountered congenital anomaly in medical practice. Patients with Down syndrome are of special concern to the anesthetic provider, primarily due to their unique set of medical conditions, including respiratory, cardiovascular, atlantoaxial instability, congenital heart disease and other systemic problems [2]. The risks of anesthesia are increased in these children and skillful management during the perioperative period is essential for a successful outcome. In this chapter, the anesthetic implications of the syndrome are reviewed and the principles of perioperative management will be discussed.

ANESTHETIC IMPLICATIONS

Airway

Tracheal stenosis is a more common finding in children with Down syndrome and poses a major concern related to intubation and increased airway narrowing after intubation or bronchoscopy. There is an absence of the membranous portion of the trachea with fusion of the posterior aspects of the cartilaginous rings. This entity may present as respiratory distress, stridor, croup, bronchiolitis, cyanosis, wheezing and failure to thrive. Subglottic narrowing is also more common in children with Down syndrome [3,4]. Other structural abnormalities include flattened nasal bridge and macroglossia.

Respiratory

Respiratory problems are not uncommon in children with Down syndrome. Both upper and lower airway concerns arise in this population namely due to their hypotonia, relative obesity, cardiac disease, small upper airway, stenotic anomalies, and a degree of pulmonary hypoplasia [5]. There is also a high incidence of Obstructive Sleep Apnea (**OSA**) in this patient population, placing them at greater risk for airway obstruction during anesthesia and sedation. Causes of Down Syndrome-associated OSA include central apnea, hypotonia of the mouth and upper airway, poor coordination of airway movements, narrowed air passages in the midface and throat, relative enlargement of the tongue, and hypertrophy of adenoid and tonsillar tissues [6].

Pulmonary vascular disease may also occur as a consequence of Eisenmenger's Syndrome and unrepaired cardiac lesions such as Atrioventricular Septal Defects (**AVSD**), where reversal of a left to right shunt leads to cyanosis. A high mortality is associated with such conditions while under anesthesia. Pulmonary vascular disease may also arise due to OSA, chronic hypoxemia due to repeated pulmonary infections, and hypoventilation due to muscle hypotonia [7].

Cardiac

Congenital heart defects are not uncommon in children affected by Down syndrome, with endocardial cushion defects being the most common. Of these lesions, **AVSD** (40%) are the most frequently identified. Clinically apparent cardiac lesions occur in about 12% of the pediatric Down syndrome population, however, on autopsy as many as 60% are found to have cardiac lesion [4,6]. The incidence of ventricular septal defects is approximately 30%, followed by patent ductus arteriosus (12%), atrial septal defects (10%) and tetralogy of Fallot (8%) [5,7]. With the exception of tetralogy of Fallot, uncorrected **AVSD** can result in pulmonary over circulation with subsequent pulmonary hypertension. Depending on the nature of the lesion, these patients will often need surgical repair. Following repair of congenital heart lesions, conduction disturbances, particularly atrial dysrhythmias, are not uncommon. Up to half of the children with Down syndrome may exhibit bradycardia during induction of anesthesia or sedation, which can quickly escalate to asystole and cardiac arrest. Abnormalities in the heart valves, such as mitral valve prolapse, are also concerns in the pediatric Down syndrome population [6].

Gastrointestinal

Gastroesophageal reflux is an important factor to consider during the anesthetic evaluation given the increased risks of aspiration while sedated under anesthesia. Children with Down syndrome are more likely to have gastroesophageal reflux than those who do not suffer from this congenital disorder. Duodenal atresia is 300 times more common than compared to the general population [5].

Musculoskeletal

Approximately 15% of patients with Down Syndrome present with ligamentous laxity of the atlantoaxial joint [8,9]. This condition predisposes children to C1-C2 subluxation and catastrophic spinal cord injury as a result of this instability. Several factors contribute to Atlantoaxial Instability (AAI) such as laxity in the transverse ligament, hypoplasia, malformation and absence of the odontoid process. This laxity can cause C1 to subluxate on C2, affecting flexion, extension, and possibly result in cord compression. There may also be laxity of other joints such as the fingers, thumb, elbow or knee and generalized poor muscle tone [10].

OTHER ORGAN SYSTEMS

Down syndrome is associated with higher occurrences of hypothyroidism, Hirschsprung disease, impaired cellular immunity (i.e. leukemias), developmental delay, microcephaly, moderate to severe mental retardation and epilepsy (5-10%) [5].

BEHAVIORAL AND COMMUNICATION CONSIDERATIONS

Children with Down syndrome are prone to developing more anxiety and agitation than children of the same age when placed in unfamiliar settings. Such heightened anxiety may be related to their limited understanding, which can lend itself to unsafe behaviors for the child, caregivers and medical personnel. Given this consideration, it is beneficial to conduct the pre-anesthetic visit with the child and caregivers present with the use of child appropriate language. Similarly, it is important to encourage the caregivers to display calm and encouraging emotional behaviors during the evaluation, as Down syndrome children tend to exhibit receptive communication skills elite to their expressive communication capabilities. Play therapists may also have a role in ensuring smooth induction. Preoperative sedation may also be warranted.

PRE-OPERATIVE AND INTRAOPERATIVE MANAGEMENT

Children with Down syndrome generally have a warm and pleasant demeanor; however, parental presence during the preoperative period is a valuable way to help provide them with reassurance. Child life specialists are also helpful in this regard.

Evaluation of the severity of obstructive sleep apnea by symptoms or polysomnography should be performed before anesthesia. Preparation for upper airway obstruction with an oral airway will decrease chances of hypoxia on induction. In patients with subglottic stenosis, an LMA

(for short cases) or smaller endotracheal tubes should be available during intubation to lessen the risk of airway trauma [2,11].

The high prevalence of congenital heart disease warrants a thorough cardiac history to assess severity and to mitigate the morbidity and mortality of any cardiac events that may occur during surgery. Symptoms suggestive of congenital heart disease include failure to thrive, breathlessness and fatigue on exertion, central cyanosis, finger clubbing, respiratory distress, signs of cardiomegaly with a displaced apex, hepatomegaly and heart murmur (possibly with an associated “thrill”). Ideally, an ECG, an echocardiogram and a cardiology consultation should be obtained in all children with Down syndrome. Collaboration with a pediatric cardiologist may be needed to better medically optimize the patient and assess surgical risk. Prophylactic antibiotics may be indicated based on whether the patient has undergone cardiac surgery, if there is a residual defect after surgery, if prosthetic devices were used, or if anesthesia will be administered shortly after surgery [2,12,13].

The diagnosis of atlanto-axial instability is difficult, requiring multiple films in neck flexion, extension and odontoid views. These films are not reliable, however, in predicting cervical spine compression and are difficult to reproduce. In addition, children under the age of three do not have sufficient epiphyseal development and vertebral mineralization for acceptable radiographic evaluation. The American Academy of Pediatrics guidelines recommend that asymptomatic children with down syndrome not have routine radiologic evaluation of the cervical spine. Instead, a thorough history and physical should be performed (Table1) to rule out atlanto-axial instability [2,10].

Table 1: Evaluation of Cervical Spine Instability [10].

Evaluation of Cervical Spine Instability [10]	Without Imaging
History:	Physical Exam:
Has behavior/ activity level changed?	Abnormal range of motion of the head and neck, and examine for neck tenderness
Is there decreased ambulation?	Abnormal gait
Has fine motor function decreased?	Weakness, spasticity, increased deep tendon reflexes, a positive Babinski reflex, or clonus of the lower extremities
Has there been any change in bladder or bowel function?	
Is there head or neck pain?	
Is there inability to turn/ rotate head?	
Have there been any episodes of dizziness or syncope?	

Should any signs or symptoms of possible cervical spine instability exist during the preoperative exam for an elective case, the surgery should be delayed for a complete cervical spine evaluation. If the operation is emergent, then cervical spine precautions should be used to decrease the risk of subluxation, including a discussion with the surgeon if the operative procedure requires neck extension [10]. Patients with Down syndrome have in increased incidence of anemia and neonatal thrombocytopenia, therefore potentially necessitating a preoperative **CBC** and platelet count.

A thorough discussion regarding the need for premedication and induction options (inhalation versus intravenous) should be had with the parents, bearing in mind the disposition of the child and the nature of the operation. If the child is anxious, or uncooperative, premedication with **PO** midazolam (0.5-1mg/kg) can be considered and ample time (15-20 minutes) should be allowed for the medication to take optimal effect. For patients with behavioral issues, intramuscular ketamine (1-2 mg/kg) may be useful. Before giving any premedications, careful consideration should be given to precluding factors such as severe **OSA** or other respiratory issues.

Routine monitors including electrocardiogram, blood pressure, pulse oximetry, end tidal capnography, and temperature, should be used. Due to the associated conduction abnormalities seen in children with Down syndrome, it is not uncommon to see bradycardia during induction, particularly following administration of succinylcholine. To this end, atropine and glycopyrrolate should be readily available. In addition, sevoflurane, which is the agent of choice for inhalation inductions, due to minimal airway irritability and favorable hemodynamics, is also associated with a higher incidence of bradycardia. Depending on patient hemodynamics, it may be necessary to decrease the concentration of sevoflurane. Narcotics should be titrated carefully so as to adequately treat pain, while not compromising the airway. Adjuvants such as acetaminophen and non-steroidals are also helpful in minimizing the amount of narcotic required.

POST-OPERATIVE MANAGEMENT

These patients should be monitored closely postoperatively until they have fully recovered from anesthesia. Airway maneuvers like jaw thrust and chin lift may be required to counteract the obstruction that can occur due to hypotonia.

CONCLUSION

Down syndrome is a common congenital abnormality characterized by multi-system involvement. For this reason, a thorough preoperative assessment, particularly of the airway, cervical spine, cardiovascular, and respiratory system, is essential to achieving optimal anesthesia conditions. Communication with the patient's medical providers, such as the pediatric cardiologist and surgeon, will help optimize the patient and decrease the incidence of morbidity and mortality. The most common perioperative complications including airway obstruction, postoperative stridor, bradycardia, and difficult intubation, can be safely managed if anticipated and an appropriate plan established. Continued vigilance in the post-operative setting for airway obstruction is warranted to decrease the risk of hypoxia.

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