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Review Article

Anesthesia for Patients with Down syndrome

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ABSTRACT

Down syndrome is the commonest chromosomal abnormality and involves multiple systems of the body. Patients with Down syndrome are posted for various cardiac and non-cardiac surgeries requiring general anesthesia. Multiple systemic involvements and congenital anomalies require meticulous preoperative assessment. Cardiac and respiratory examinations are particularly focused upon. Depressed immune function and presence of antibodies are quite common. Intraoperative management targets airway management and proper positioning of these patients. These patients also have raised propensity to cardiac and respiratory complications intraoperatively. A discussion focusing on anesthetic implications and management of patients with Down syndrome has been presented here.

Key Words: Down syndrome, Trisomy 21, anesthesia

INTRODUCTION

Down syndrome (DS), also called Trisomy 21, is a condition associated with extra genetic material causing delayed physical and mental development. DS, characterized by dysmorphic facies, is commonest of the congenital anomalies. The incidence of DS differs between populations and increases with maternal age.

Pathophysiology

DS causes wide spectrum of phenotypic changes and organ involvement as a result of extra chromosome 21(Hsa21). The genes on additional copy of chromosome Hsa21, in people with DS, get decoded and results in increased expression. It is hypothesized that phonotype characterizing DS is results due to imbalance in expression of Hsa21 and non-Has21 genes. [1]

General anesthesia in Down syndrome

The DS patient may require general anesthesia for various surgeries, some of it may be for the presenting complications of DS itself. Congenital anomalies like cardiac, gastro-intestine, esophagus and urinary tract may require early surgical intervention. Chronic health problems involving eyes, ears and joints can potentially require intervention in later part of life. A patient with DS for general anesthesia, challenges the knowledge and skills of an anesthesiologist.

Pre-anesthetic assessment:

The multiple manifestations of DS require that the overall physical and mental health should be recorded carefully. Cardiovascular system, respiratory system, airway and neck require particular attention during pre-anesthetic assessment.

Cardiovascular system:

Common cardiac anomalies in DS are tetralogy of fallot, atrioventricular canal (AV canal), ventricular septal defect, atrial septal defects, patent foramen ovale and defects of cardiac valves. Other specified cardiac anomalies are hypoplastic right heart, hypoplastic left ventricle, anomalies of the coronary artery or sinus, patent ductus arteriosus, coarctation of the aorta, hypoplasia of the aorta, persistent right

aortic arch, overriding aorta, anomalies of the pulmonary artery, anomalies of the great veins and single umbilical artery. [2]

The patients with suspected DS are routinely assessed by the cardiologist in postnatal period in first month and echocardiography is performed; moreover, echocardiogram should be performed in all patients with DS preoperatively. [3,4] Cardiac re-evaluation is warranted for repaired lesions and clinical examination is directed to rule out or correct cardiac failure, pulmonary hypertension, arrhythmia or any other disturbance.

Patients with DS are at risk of developing pulmonary hypertension, of respiratory or cardiac origin. Left-to-right intracardiac shunting and obstructive sleep apnea are major risk factors for pulmonary hypertension. [5] The patients with repaired congenital cardiac defects may have respiratory infection, pulmonary hypertension, or arrhythmias including atrio-ventricular block. Pacemaker is required for complete heart block in patients with repaired atrio-ventricular septal defect. [6,7]

The airway and respiratory system

The patients of DS may have significant airway obstruction like lymphoid hyperplasia, narrow nasopharynx, macroglossia, laryngomalacia, congenital subglottic stenosis, tracheal stenosis and, tracheobronchomalacia. The airway may have multiple sites of obstruction. [8] Concomitant pulmonary hypertension should be ruled out in these patients. [9,10]

Obstructive sleep apnea associated with airway obstruction may have clinical presentation like developmental delay, enuresis, daytime somnolence, behavioral problems, and poor school performance. There may be delay in diagnosis due to overlap of clinical presentation of obstructive sleep apnea with that of DS. [11]

Recurrent respiratory tract infection associated with DS may be due to anatomical and immunological defects. Associated anatomical abnormalities are congenital anomalies of upper and lower airway, congenital heart disease, gastro-esophageal reflux, and deglutition disorders. [12]

The cervical spine

Atlanto-axial instability may be due to bony abnormalities of C1 or C2, or both in about 15% of individuals with DS. [13] It may be asymptomatic initially with becoming symptomatic in 1-2% cases, which may be associated with odontoid impingement. [14] Cumulative trauma over a period of time may lead to development and progression of atlanto-axial instability, therefore period radiological evaluation is necessary. [15]

Central nervous system

DS, one of the major causative factors of mental retardation, can cause many neurological complications like structural changes, Alzheimer's disease, strokes, epilepsy and basal ganglia damage. [16] Cognitive and intellectual disability may be present along with deficits in adaptive behavior and impairment in learning and memory. [17,18] Majority of persons with DS suffer from generalized poor muscle tone. [19]

Immune deficiency

Patients with DS have raised susceptibility to infection, and are predisposed to develop malignancy and autoantibodies. [20] Immune system may be deficient and lead to depressed T cell function and neutrophil chemotactic responsiveness. [21,22] The person with DS may also have history of prolonged respiratory infection.

Other systems

Hypothyroidism, which has age related varied prevalence in DS, should be screened in all patients because the symptoms mimic that of DS. [23] Thyroid hyperfunction caused by TSH receptor stimulating antibodies may also be present in some of the cases. [24] Gastric motility disorder (gastroesophageal reflux, dysphagia and constipation) is common in persons with DS. [25,26]

Induction of anesthesia

An attentive approach should be taken in patients with DS during induction because of presence of a lot of anxiety. Parents of children may be helpful during induction. The presence of cognitive and intellectual disability may be troublous for both the patient and anesthesiologist. [27] Oral midazolam with oral ketamine, and oral atropine are generally prescribed as premedications in these children. [28,29,30] The use of atropine has been debatable in terms of increased chronicity of heart and mydriatic effects in DS, in connection to this varied results has been presented by different studies over period. [31,32]

Multiple cranio-facial abnormality may produce adverse situation in airway management, and difficult airway cart should be readily available. [33] Careful airway maneuver should be performed to avoid injury and subluxation of atlanto-axial and submandibular joints. A smaller size endotracheal tube may be required due to presence of subglottic stenosis, when intubation is indicated

Sevoflurane is the inhalational agent of choice in children; however it can cause bradycardia when given without anticholinergics. [34,35] Intravenous induction may be preferred in older children when a cannula is in place; moreover a cannula may be placed after application of local anesthetic cream. .

Maintenance of anesthesia

The patient should be positioned carefully to prevent any injury especially in head and neck surgeries. Neuromuscular blockade should be monitored and muscle relaxant should be used judiciously. Difficult airway requires planning and precaution during extubation.

Anesthesia-related complications

Airway related complications (postintubation croup, difficult intubation, bronchospasm) and bradycardia are common in patients with DS for non cardiac surgeries. [36,37] These patients may require longer hospital stay especially after adenoidectomy and tonsillectomy. [38]

CONCLUSION

The Down syndrome, being commonest chromosomal disorder, presents with multiple systemic organs involvement and congenital anomalies. With rising incidence of Down syndrome, an anesthesiologist has a high chance to encounter a patient of Down syndrome posted for interventions or surgeries. A systemic examination, focusing on airway, respiratory and cardiac systems, is necessary prior to surgery. Strict aseptic precautions are required during anesthesia and surgery. Intraoperative management requires special care during airway management and patient positioning. With prior knowledge about the disease process and anesthetic management, optimal perioperative care of these patients should not be difficult.

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