

Congenital Syndromes Affecting Heart and Airway Alike

With great interest, I read the article written by Dr. Yadav about a bifid epiglottis.^[1] He has presented a case with multiple congenital malformations that was taken for surgery many times. Dealing with a patient with syndrome is major concern for an anesthesiologist. Syndrome is a group of signs and symptoms that occur together and characterize a particular abnormality or a condition. Thirty percent of congenital heart diseases are related to genetic syndromes and are accompanied by extracardiac anomalies.^[2] Many of the patients with heart disease having these syndromes also have craniofacial abnormalities, and anesthesiologist may face difficulty in mask ventilation or intubation. While managing these patients, we have to deal with systemic effects of cardiac abnormalities and we often face problems in intubation. The American Society of Anesthesiologists Task Force on Management of the Difficult Airway has defined difficult airway as the clinical situation in which a conventionally trained anesthesiologist experiences difficulty with facemask ventilation of the upper airway, difficulty with tracheal intubation or both.

Most common condition related to this issue is Down syndrome. There is the addition of one extra chromosome linked to chromosome 21 resulting in its trisomy. It is occur in 1:600–800 live births.^[3] Approximately 60% of Down syndrome patients have congenital heart disease.^[4] Most common are endocardial cushion defects, ventricular septal defects, complete atrioventricular defects, patent ductus arteriosus (PDA), or tetralogy of Fallot.^[5] These patients have facial dysmorphism, short neck, relative macroglossia, microdontia, mandibular hypoplasia, congenital subglottic, and/or tracheal stenosis. Twenty percent of patients have ligamentous laxity of the atlantoaxial joints.^[6] Hypoplasia, malformation, and the absence of the odontoid process predispose the patients to C1–C2 instability. This can lead to spinal cord injury. Patient may develop tonsillar and adenoidal hypertrophy which further contributes to upper airway obstruction, including obstructive sleep apnea syndrome. The incidence of postextubation stridor is also common in these patients.

The mutation in PTPN11 gene on chromosome 12 results in Noonan Syndrome. It is an autosomal dominant disorder and the second most common genetic syndrome of congenital heart disease. Its incidence is 1 in 1000–2500 births. Congenital heart diseases occur in 50%–80% of individuals. Pulmonary valve stenosis is the most common heart defects and is found in 20%–50% of individuals. Hypertrophic cardiomyopathy is found in 20%–30% of the individuals. Other cardiac anomalies include atrial septal defects, ventricular septal defects, PDA, and tetralogy of Fallots.^[7] These patients have short stature, unusual face, flat nose, hypertelorism, down-slanting eyes, palpebral fissures ptosis,

low-set posteriorly rotated ears, webbed neck, cervical spine anomaly, and pectus excavatum. These features lead to difficulty in airway management.

The DiGeorge, velocardiofacial, and conotruncal face syndromes are phenotypic variability of 22q11.2 deletion syndrome.^[8] It is the most common microdeletion syndrome found in humans.^[9] The incidence is 1 in 4000 live births. Some of the congenital anomalies are conotruncal defects of outflow tract of the heart, tetralogy of Fallot, interrupted aortic arch Type B, truncus arteriosus, aortic arch anomalies, and ventricular septal defects. Such patients have palatal defects including cleft palate and bifid uvula and have feeding and swallowing problems. These patients can also have hypocalcemia, hearing loss, seizures, immunodeficiency, renal anomalies, skeletal anomalies, mental retardation, and psychiatric disorders.

The absence or malfunction of lysosomal enzymes required for glycosaminoglycan breakdown leads to lysosomal storage disorders. It involves accumulation of mucopolysaccharides throughout the body. These patients may suffer from cardiac dysfunction, cardiomyopathy, or coronary artery disease. Patients have coarse facial features, macroglossia, thickened nasal, oral, and pharyngeal mucosa, hypertrophy of adenoids and tonsils, hypoplastic mandible, reduced temporomandibular joint mobility, narrowed trachea and short, immobile and unstable neck. There are chances of atlantoaxial subluxation, which may lead to anterior dislocation and spinal cord compression. Mask ventilation and intubation may be difficult in these patients.^[10]

Patients with these syndromes have cardiac problems and various degrees of airway malformations. It is very important to look for systemic effects of cardiac disease and the adequacy of the airway. The predictors of difficult intubation are presence of dysmorphic features, limited mouth opening, limited neck extension, restricted mobility of temporomandibular joints, large tongue, limited submandibular space (retrognathia, micrognathia, mandibular hypoplasia, or dysplasia), and the presence of structural abnormalities in the laryngotracheal passage. Obstructive sleep apnea syndrome is often present in these patients. These features lead to potential difficulties with airway management in syndromic children.^[11]

During preanesthetic checkup, anesthesiologist may need to examine carefully for the facial anomalies, dentition, extent of mouth opening, head–neck mobility, anomalies of the palate and mandibular floor, and assess thyromental distance. If the patient has undergone surgery before, history can give us an idea about the airway management but the airway dimensions significantly change with growing age.

As there is an expected airway management difficulty, there can be a need of establishing a surgical airway. It is better to chart a plan of management. We should discuss the problems and overall management plan with patient or parent to avoid legal issues.

In patient with cardiac disease, a good airway management is very important, as even a short period of airway obstruction/hypoventilation may result in hypoxemia. Sudden change in ventilation pattern, PaO₂, PaCO₂, or pH affects pulmonary vascular resistance, which can lead to detrimental effects on shunt magnitude, cardiovascular function, and hemodynamic. Hence, a prompt control of airway ventilation is required for optimal pulmonary blood flow in these patients.

Few children have large occiput that causes the head to flex forward. Putting a small roll under the neck or shoulder helps us to maintain the head in neutral position.

High fraction of oxygen may reduce pulmonary vascular resistance and may comprise systemic perfusion, but preoxygenation is recommended for denitrogenation of the lungs.^[12] It is helpful at the time of apnea. Mask ventilation might be difficult. One should not insufflate stomach during mask ventilation. The use of an oral and/or nasal airway pulls the large tongue away from pharyngeal wall and is useful in mask ventilation. It is better to confirm the reliability of facemask ventilation before paralysis. Once satisfactory facemask ventilation has been confirmed, neuromuscular blockade is used for intubation. The adequate size laryngoscopy blade is useful to displace the large tongue. Such patients often have tracheal narrowing and it is better to use tracheal tube 0.5–1 mm smaller than the calculated size for the age of the child. Nasal airway has better stability, so endotracheal tube is commonly passed through nares in the child <10–12 years age. Few patients with cardiac disease may have high venous pressure, and the passage of nasal tube can injure mucosa and initiate nasal bleeding. One should be gentle enough while passing a nasal tube. Uncuffed tube with leak at or near 25–30 cm of H₂O is used in child <6–8 years age. However, few patients have poor lung compliance; so in these patients, a cuffed oral endotracheal tube may be used to allow higher pressure for optimal ventilation. The patient may have cervical ligamentous anomalies and chance of atlantoaxis instability. Neck flexion-extension and rotation movements should be kept to a minimum in all these patients. Full cervical spine precautions should be undertaken and care should be taken to maintain the neck in a neutral position. This can be maintained with the use of a soft cervical collar after induction to preserve the position of the neck.

Intraoperative management of these patients is a nightmare for anesthesiologists. We have to achieve optimum hemodynamic and a prompt airway in the patient. The management of each case is different. It is better to make management strategy of each case and explain it to all the medical staff involved in the case. Such patients require gentle care, and with detailed knowledge of a syndrome,

we can deal with the patient in a controlled manner. The optimal management enables successful airway control while avoiding airway complications and hemodynamic instability.

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