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Anaesthetic management in Congenital Diaphragmatic Hernia with Down's syndrome -A case report

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ABSTRACT *Down's syndrome constitutes to be the most common chromosomal disorder. It is found in 1/800 live births.[1]These babies have many associated congenital anomalies that affect almost all of their organ systems. Overall frequency of congenital diaphragmatic hernia (CDH) in Down's Syndrome seems to be low. Morgagni hernia is the most common type of CDH among these patients. Gastro-esophageal reflux disease (GERD),Pulmonary hypoplasia, susceptibility to respiratory infections, along with atlanto axial instability and sensitivity to anaesthetic agents modify the anaesthetic management in these cases. [2] Carefully designed anaesthesia is must for successful perioerative outcome.*

Introduction:
Down's Syndrome is a congenital anamoly occurring due to extra chromosome attached to Chromosome 21.This is also called as Trisomy 21**.** [3] Diaphragmatic hernia is a protrusion of abdominal viscera into the chest cavity through communication, which is congenital or acquired . [4] Sleep apnoea/ airway obstruction and respiratory tract infections are common with Down's Syndrome. Rapid sequence or awake endotracheal intubation and GERD profylaxis is the key to avoid further respiratory complications in both the conditions. [5] We represent the anaesthetic considerations in rare combination of Down's Syndrome and Morgagni CDH.

Case History:
A three month old baby born to a 35 yrs old mother at term, came with regurgitation on feeding with chest infection. There was no other significant birth history. On examination, Pulse rate- 120/min, Resp. Rate 26/min, with respiratory distress. Baby had flat nose and forehead, with low set ears, epicanthal fold of eyes and macrognothia. Other general physical examination was normal. On systemic examination, Heart sounds were normal and heard at the left border of the sternum. On respiratory examination, respiratory sounds were reduced on left side and gurgling sounds were heard over left side of the chest. Abdomen was scaphoid. On investigation, routine blood investigations were normal. Chest X ray showed intestinal loops in left side of chest, with collapsed ipsilateral lung. Coiled nasogastric tube was observed in left chest. Mediastinum was shifted to right. On right side, prominent broncho vascular markings were seen. Child was posted for CDH repair. On preoperative visit, iv line was in situ, flowing. Baby was kept NPO for 6 hours. On the day of surgery, premedication was given in the form of Inj. Glycopyrrolate 0.04mcg/kg, Inj. Ondencetrone 0.08mcg/kg. IV. Inj. Ranitidine 1mg/kg was administered as aspiration prophylaxis 45min prior. In Operating room (OR), SPO2, ECG, NIBP and precordial stethoscope monitoring attached. Baby was induced with Inj. Ketamine 0.5 mg/kg with O2, N2O and Sevoflurane mixture. Oral intubation done with size 3.5 Endo tracheal tube (ETT). Bilateral air entry checked and ETT secured. Analgesia was provided with Inj. Fentanyl 1mcg/kg. Muscle relaxation was provided with Inj. Atracurium. Anaesthesia was maintained on O2, N2O and Sevoflurane mixture on Ayres T piece. CDH was reduced by abdominal approach. Procedure was uneventful. Baby was shifted to PICU with ETT in situ. On second post operative day, baby was extubated and discharged on 10th post operative day in good health.

Discussion: Downs's syndrome is a congenital chromosomal anomaly which affects almost all the systems in the body. It includes macroglossia, microcephaly, endocardial cushion defects, ventricular septal defects, duodenal atresia, and atlantoaxial instability and supraglottic stenosis**.** [6] There is increased incidence of respiratory complications in children with Downs's syndrome. Upper and lower airway problems exist in this subset of the population which is attributed to hypotonia, relative obesity, cardiac disease, small upper airway, pulmonary hypoplasia, and congenital anomalies of airway.[4] All these results in unique sets of challenges to the anesthesiologists. Diaphragmatic hernia is a protrusion of abdominal viscera into the chest cavity through communication. The classic diagnostic triad includes respiratory distress, scaphoid abdomen, and signs of mediastinal shift.[8]

The prevalence rate for all types of CDH is approximately one in 3000 live births, although considerable variation has been reported with frequencies as low as one in 5000. [4] Physiologically, the hernia affects mainly one of two systems:

(A) the cardiorespiratory or (B) the gastrointestinal. It leads to pulmonary hypoplasia and aspiration pnumonitis; which is also evident in Downs Syndrome.

This was a delayed presentation of diaphragmatic hernia as it was diagnosed at the age of more than 2 months.[10] The congenital diaphragmatic hernia (CDH) is classified according to the location of the protrusion. It includes posterolateral (Bochdalek) hernias, Morgagni and other anterior hernias, and (rarely) central hernias. About 50%-60% of affected individuals have isolated CDH; the remainder have complex CDH – that is, CDH occurring with additional malformations or as part of a single gene disorder or chromosome abnormality. Infants with trisomy 21 can have either Bochdalek or Morgagni hernia, although overall frequency of CDH in trisomy 21 seems to be low. Morgagni hernias are more commonly reported than Bochdalek hernias, suggesting that it is the most common type of CDH among individuals with trisomy 21. [11] Diaphragmatic eventration is incomplete muscularization of the diaphragm resulting in a thin membranous sheet of tissue. Severe diaphragmatic eventration is associated with pulmonary hypoplasia and respiratory distress during infancy. Milder degrees of diaphragmatic eventration can present later in life with respiratory symptoms such as cough and pneumonias, or without symptoms so that the diagnosis is made incidentally on chest x-ray.[12] CDH occurs approximately 85% of the time on the left side, 10% on the right side, and less than 5% of the time bilaterally [13] The pathogenesis of the pulmonary hypoplasia so frequently associated with CDH is not fully known, but appears to have both a primary component, i.e., the hypoplasia occurs along with the diaphragm defect, and a secondary component, i.e., arising from competition for thoracic space particularly in the lung ipsilateral to the

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hernia. [14-15] Overall frequency of congenital diaphragmatic hernia (CDH) in Down's Syndrome seems to be low. Following table shows the incidence of Common Chromosomal Anomalies Associated with CDH.

Table 1. Common Chromosomal Anomalies Associated with CDH
View in own window

Chromosome Abnormality/Locus	Frequency of Congenital Diaphragmatic Hernia [1]	
	Found in This Disorder	Attributed to This Disorder
Pallister-Killian syndrome/ (isochromosome or tetrasomy 12p)	~30%	?<5% [2]
Trisomy 13	Rare	Very rare
Trisomy 18	?1%-2% [2]	Rare among all CDH; most common chromosome abnormality in prenatally diagnosed CDH
Trisomy 21	Rare (Morgagni hernias > Bochdalek hernias)	Very rare
Del (4)(p16) (Wolf-Hirschhorn syndrome)	Rare	Very rare
+der (22) t(11;22) (q23;q11)	5%-10%	Very rare
Del (15)(q26.2)	Unknown (?but possibly majority)	Unknown [1]
Del (1)(q41-q42)	Unknown	Unknown [1]
Del (8)(p23.1)	?30% [2]	Unknown [1]

1. Small chromosome deletions of these regions, or point mutations of genes mapping to these regions, may cause CDH. The frequency with which these occur is presently unknown.
2. ?=Number provided represents an educated estimate, derived from the medical literature and authors' personal experiences.

In our patient, anaesthesia was designed by keeping in mind both the conditions together. Gastro-esophageal reflux disease (GERD) is more prevalent in children with Downs's syndrome. The symptoms to be assessed preoperatively include vomiting, oesophagitis, respiratory symptoms like apnea, wheezing and aspiration pneumonia. Aspiration prophylaxis with modified rapid sequence induction may be used along with the agents to decrease the pH in the stomach [16] Hence endotracheal intubation should be performed either awake or by restoring spontaneous respiration. There is also increased incidence of pulmonary infectionsin both the conditions. This may also be due to thymus dependent immune system depression in children with Downs's syndrome. [17]Peripheral lines may be the source of infection so the lines are not to be kept in place for long periods of time. Downs babies are very sensitive to anaesthetic agents and carefully titrated dosages should be used. Sleep induced ventilatory dysfunction may be exaggerated by narcotic induced sedation and residual anesthetic concentration in the body. Volatile anesthetic agent requirements in these patients are less than normal patients.[18] Ligamenus laxity leads to atlanto axial joint instability in Down's babies. It poses a potential risk of C1-C2 subluxation. During induction and endotracheal intubation, great care must be taken to maintain the neck in neutral position. Placing a soft coller after induction of anaesthesia can serve as reminder to avoid neck movements intra operatively. Downs's syndrome should be intubated with an endotracheal tube 0.5–1.0 mm diameter smaller than the standard age-appropriate endotracheal tube size due to possible tracheal stenosis[19]These patients are also prone to have hypothermia during surgery. Proper covering of head and extremities is essential. Post-operative respiratory complications are also more common. In CDH, in addition to transport and installation of the newborn infant the dangerous periods of the anaesthesia are represented by abdominal closure because of the risk of compression. Patients are left intubated at the end of surgery since postoperative artificial ventilation is a necessity in such cases. Awake extubation should be considered after assessing spontaneous ventilatory efforts; to minimize post operative respiratory complications.

Summary:
Due to high prevalence of Down's Syndrome, anaesthetists may come across to these patients with need to operate for congenital defects. A full-term baby born with congenital diaphragmatic hernia unassociated with other major anomalies can have good prognosis with proper anesthetic and surgical management. Pertinent aspects of the embryology, pathology, and physiology involved should be considered in anaesthetizing these babies. Extra care should be taken to tackle combination of two different congenital problems for successful intra-operative and peri-operative management.

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