Discussion: Downs’s syndrome is a congenital chromosomal anomaly which affects almost all the systems in the body. It includes macroglia, microcephaly, endocardial cushion defects, ventricular septal defects, duodenal atresia, and atlantoaxial instability and supraglottic stenosis. There is an increased incidence of respiratory complications in children with Down’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. All these results in unique sets of challenges to the anaesthesiologists. Diaphragmatic hernia is a protrusion of abdominal viscosa into the chest cavity through communication. The classic diagnostic triad includes respiratory distress, scaphoid abdomen, and signs of mediastinal shift. 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. Physiologically, the hernia affects mainly one of two systems: (A) the cardiorespiratory or (B) the gastrointestinal. It leads to pulmonary hypoplasia and aspiration pneumonitis; which is also evident in Down’s Syndrome.
hernia. 19-21 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** | **Found in This Disorder** | **Attributed to This Disorder** |
| Pallister-Killian syndrome (isch-romosome or tetrasomy 12p) | ~30% | Rare | Very rare |
| Trisomy 13 | Rare | Very rare |
| Trisomy 18 | 1%-2% | Rare among most CDH; most common chromosome abnormality in prenatally diagnosed CDH |
| Trisomy 21 | Rare (Morgagni hernias > Bochdalek hernias) | Very rare |
| Del (4)p(16) (Wolf-Hirschhorn syndrome) | Rare | Very rare |
| Del (15)(q26.2) | Unknown (but possibly majority) | Unknown |
| Del (1)(q41-q42) | Unknown | Unknown |
| Del (8)(p23.1) | ?30% | Unknown |

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 Down’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 infections in both the conditions. This may also be due to thymus dependent immune system depression in children with Down’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 collar after induction of anaesthesia can serve as reminder to avoid neck movements intraoperatively. Down’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 anesthetizing 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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