**Challenging Airway Management in a Neonate Undergoing Truncus Arteriosus Repair – Report of Case and Review of Literature for Congenital Airway Anomalies Associated with Conotruncal Defects**

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**Background**

- Congenital airway anomalies (CAA) are present in about 4% of all children with congenital heart disease (CHD).
- The incidence of CAA in children with conotruncal heart defects may be as high as 56%.
- We will describe a case of challenging airway management in a neonate with truncus arteriosus undergoing surgical repair.
- We will review and discuss the literature on congenital airway anomalies in children with CDH, with a focus on CAA in children with conotruncal heart defects.

**Case Presentation**

- A 3 day-old 3 kg female with a postnatal diagnosis of truncus arteriosus type 1 with VSD and normal left sided aortic arch presents for repair.
- Nasal intubation was performed and choanal atresia of the right nare was noted. The left nare was stenotic and only able to accommodate a 2.5 uncuffed endotracheal tube (ETT).
- After serial dilatations, a 3.5 uncuffed ETT was passed.
- The laryngeal inlet was normal and intubation was accomplished with a Miller 3.5. A direct laryngoscopy was performed which confirmed the tip of the ETT was wedged anteriorly against the laryngeal inlet.
- The tip was in the trachea below the thoracic inlet.
- The area of obstruction was confirmed with ventilation.
- Bulge created by the abnormal takeoff of the left bronchus leading to trouble branching and anteriorly directed left bronchus was withdrawn immediately due to concern for airway compression versus conotruncal anomaly.
- An echocardiogram (TEE) probe was inserted. At this time, end tidal CO2 was recorded.
- After completion of surgical repair, a mini ETT was inserted. The laryngeal inlet was normal and intubation was accomplished with a Miller 3.5.

**DiGeorge Syndrome**

- Structural Airway Anomalies in DiGeorge Syndrome

**Table:**

<table>
<thead>
<tr>
<th>Chromosome Abnormality</th>
<th>Type of Conotruncal Defect</th>
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<tbody>
<tr>
<td>22q11.22 (DiGeorge)</td>
<td>TOF, Truncus Arteriosus, IAA Type B, DORV, Conoventricular VSD</td>
</tr>
<tr>
<td>Trisomy 13</td>
<td>Conoventricular VSD, DORV, TOF</td>
</tr>
<tr>
<td>Deleted 17p11.2</td>
<td>TOF, DORV, Truncus arteriosus</td>
</tr>
<tr>
<td>Partial trisomy 8q</td>
<td>TOF, DORV, Truncus arteriosus</td>
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**DiGeorge Syndrome**

- Congenital heart disease, particularly conotruncal malformations (40%)
- Cranofacial anomalies including microcephaly, micrognathia, choanal atresia, cleft palate (50%)
- Hypocalcemia due to hypoparathyroidism (50%)
- Immune deficiencies as a result of low T cells numbers due to absent thymus
- Renal anomalies (37%)
- Hearing loss, both conductive and sensorineural
- Cognitive deficits (80%)
- Structural airway anomalies, including laryngotracheoesophageal anomalies

**Conotruncal Heart Defects**

- Malformations of the great vessels arising from the embryological structures of the truncus arteriosus and trachea
- Accounts for about 30% of CHDs
- Greater than 80% of conotruncal heart defects are associated with microdeletion of chromosome 22q11.2
- Types of conotruncal defects:
  - Tetralogy of Falot (TOF)
  - Truncus Arteriosus
  - Double Outlet Right Ventricle (DORV)
  - Double Left Outlet Ventricles (DOLV)
  - D-Transposition of the Great Arteries (TOA)
  - Conoventricular septal defects
  - Interrupted aortic arch type B (IAA)

**Syndromes Associated with Conotruncal Defects**

**Figure 1.** Tracheobronchomalacia. Lumen area of trachea at expiration is significantly decreased, compared with inspiration. CT and bronchoscopy images in (a) inspiration and (b) expiration.

**Figure 2.** Bronchomalacia. Lumen area of trachea at expiration is significantly decreased, compared with inspiration. CT and bronchoscopy images in (a) inspiration and (b) expiration.

**Figure 3.** DiGeorge patient with choice radiographs, vocal nodules, choanal atresia, cleft palate, and anteriorly directed left bronchus.

**Figure 4.** Slit lamp bronchoscopy and CT images of a DiGeorge patient with choanal atresia, anteriorly directed left bronchus.

** References**