Congenital tracheal stenosis is a rare anomaly that is difficult to diagnose. The anesthesiologist can be the first to diagnose CTS usually due to an unexpected difficult airway management either upon intubation or in the immediate postoperative period. We present a series of patients with previously unrecognized complete tracheal rings diagnosed in the perioperative and immediate postoperative period.

**Case 1**
- 6 month female with VSD presented for surgical VSD closure
- Uneventful induction, intubated with 3.5 cuffed ETI nasally taped at 13.5cm
- Post-operative CXR showed tip of ETI at the level of clavicles (see figure 1). ICU team attempted to advance ETI but met resistance. After recovering from neuromuscular blockade, patient became hypoxic.
- DLBB and CT showed stenotic long segment complete tracheal rings extending from the subglottic area to above the carina (see figure 2).

**Case 2**
- 14 month male presented for TOF repair
- Uneventful induction, intubated with 4.0 ETI nasally at 14cm
- Intraoperative finding of LPA sling which was repaired. This finding prompted CT chest post operative which showed long segment CTR.
- Failed extubation POD 4 due to stridor, re-intubated with 2.5 ETI orally at 10cm
- Shortly after recovery from neuromuscular blockade developed significant desaturations prompting an emergent sliding tracheoplasty.

**Case 3**
- 5 month old Trisomy 21 male with complete AV canal presented for repair
- 3.5 cuffed ETI nasally at 15cm
- Post-operative CXR revealed high positioned ETI. Unable to advance ETI.
- DLBB with ETI which revealed a complete tracheal ring. (Figure 3)
- Extubated on POD 2. He was managed with non-invasive ventilation until repair with a slide tracheoplasty one month later.

**Case 4**
- 1 year male with Truncus Arteriosus type I, imperforate anus, other urogenital anomalies presented for diverting colostomy operation.
- 3.0 ETI orally at 8cm
- Multiple Failed intubation due to stiidor.
- DLBB showed mid trachea short segment tracheal ring (figure 4) and left main bronchus compression. A 2.5 uncuffed ETI was passed below the tracheal narrowing. CTA done which confirmed the diagnosis in addition to an LPA sling.
- Later on he had his tracheal repair and LPA sling division. Plan for a slide tracheoplasty in the future.

**Introduction**

Incidental Diagnosis of Congenital Tracheal Stenosis in Patients with Congenital Heart Disease
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**Discussion**

Congenital tracheal stenosis is a rare anomaly (incidence of 1/64000), it is a reduction of the tracheal diameter by more than 50% due to the formation of complete or near complete tracheal rings. It can be a neonatal life threatening respiratory anomaly. It is usually associated with congenital heart disease >50% which can make diagnosing these patients very challenging.

Congenital tracheal rings are frequently associated with cardiac anomalies including ASD, VSD, CAVC, PDA, vascular anomalies (pulmonary sling) and non-cardiac anomalies like tracheo-esophageal fistula, esophageal atresia, Trisomy 21, VATER/VACTERL syndrome, Pfeiffer’s syndrome. Patient’s symptoms range from being asymptomatic to having biphasic stridor with respiratory compromise. Patients with CTS and congenital heart disease can develop pulmonary edema and respiratory distress that can lead to undiagnosing CTS. Some of these patients present early in their neonatal life to the OR for cardiac surgery prior to having developed respiratory symptoms caused by CTS. In the Immediate postoperative period, difficulty ventilating the patient after recovery from the neuromuscular blockade or with attempting to advance the ETI should prompt the anesthesiologist to suspect tracheal narrowing.

Diagnosis is confirmed with a bronchoscopic evaluation, cardiac CT and or cardiac MRI may be necessary to evaluate for vascular anomalies. We presented a series of patients where the diagnosis of complete tracheal ring was made after intubation or postoperatively. In all these cases the trachea was successfully intubated with the tip of the ETI placed at what was thought to be an appropriate position. In the patients who were symptomatic in the immediate postoperative period, their symptoms occurred after recovery from neuromuscular blockade.

If you suspect complete tracheal rings:
1. Right mainstem ETI to help clinically confirm presence of a high stenotic lesion.
2. Fiberoptic examination of the trachea will confirm the narrowing.
3. Keep the patient paralysed and avoid further manoeuvring of the ETI to avoid edema that can further worsen the narrowing.
4. Scheduling desamethasone is recommended to minimise any possibility of airway edema.
5. Direct laryngoscopy and rigid trachobronchoscopy by ENT are essential.
6. Extubate to noninvasive ventilation (i.e. CPAP)

**REFERENCES**