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A Case of Larsen Syndrome
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CASE DESCRIPTION
A 37-year-old wheel chair bound female with a BMI 41, with Larsen syndrome presented for upper endoscopy and colonoscopy. Features consistent with syndrome in this patient were short stature, multiple skeletal abnormalities, cervical vertebral anomalies and high arched palate with an anticipated difficult airway with direct laryngoscopy. Other associated comorbidities were obstructive sleep apnea and non-insulin dependent diabetes mellitus.

The procedure was performed uneventfully with monitored anesthesia care with intermittent bolus of propofol.

CASE DISCUSSION
- Larsen Syndrome is a rare inherited genetic disorder of collagen formation with skeletal abnormalities involving large joints and axial skeleton.
- Most common mode of inheritance is autosomal dominant involving Filamin B Gene (FLMB) and less commonly inherited as autosomal recessive mode.
- Incidence is 1: 100, 000 in general population.
- Males and females are affected equally.

Characteristic Features of Larsen Syndrome
- Short stature (Height below the 10th percentile in 70% of cases)
- Congenital anterior dislocation of large joints (80% hip, 80% knee, and 65% elbow),
- Cervical kyphosis (50%),
- Thoracicolumbar scoliosis (84%),
- Conductive deafness (21%),
- Cleft palate (15%),
- Laryngotracheomalacia
- Cardiac anomalies similar to Marfan’s syndrome

Cardiovascular Abnormalities associated with Larsen Syndrome
- Elongation of aorta
- Bicuspid aortic valve
- Subaortic stenosis
- Mitral valve prolapse with mitral regurgitation
- Atrial septal defect
- Ventricular septal defect
- Patent ductus arteriosus
- Tricuspid valve prolapse
- Aortic dissection and aneurysm

Facial Features of Larsen Syndrome
- Frontal bossing
- Midplace hypoplasia
- Flattening of nasal bridge
- Hypertelorism

Causes for airway Difficulties in Larsen Syndrome
- Anterior Larynx
- Laryngotracheomalacia
- Cervical kyphosis
- Cervical vertebral hypoplasia
- Cervical subluxation
- Cervical cord compression

Anesthetic Implications of Larsen Syndrome
- Airway difficulty
- Perioperative respiratory compromise
- Cervical instability and cord compression
- CHD

CONCLUSION
Careful attention to minimize the neck movement is essential during difficult airway management due to the presence of cervical kyphosis and subluxation in patients with Larsen syndrome.

Airway should be secured with flexible fiberoptic/ rigid laryngoscopy or direct laryngoscopy with manual in line immobilization of the cervical spine.

Associated laryngomalacia may lead to perioperative respiratory compromise. Presence of cardiac anomalies may further complicate the perioperative care.

REFERENCES
1. Larsen syndrome, National organization for rare disorders (NORD)