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# A Case of Larsen Syndrome

Stanlies D'Souza

*Baystate Health*, dsouzastan@yahoo.com

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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<sup>1</sup>

- Short stature (Height below the 10<sup>th</sup> percentile in 70% of cases)
- Congenital anterior dislocation of large joints ((80% hip, 80% knee, and 65% elbow),
- Cervical kyphosis (50%)
- Thoracolumbar scoliosis (84%),
- Conductive deafness (21%)
- Cleft palate (15%)
- Laryngotracheomalacia
- Cardiac anomalies similar to Marfan's syndrome

## Facial Features of Larsen Syndrome <sup>2</sup>

- ✓ Frontal Bossing
- ✓ Mid place hypoplasia
- ✓ Flattening of nasal bridge
- ✓ Hypertelorism

## Cardiovascular Abnormalities associated with Larsen Syndrome<sup>3</sup>

Elongation of aorta  
 Bicuspid aortic valve  
 Subaortic stenosis  
 Mitral valve prolapse with mitral regurgitation  
 Atrial septal defect  
 Ventricular septal defect  
 Patent ductus arterioiosis  
 Tricuspid valve prolapse  
 Aortic dissection and aneurysm

## Causes for airway Difficulties in Larsen Syndrome<sup>4</sup>

Anterior Larynx  
 Laryngotracheomalacia  
 Cervical kyphosis  
 Cervical vertebral hypoplasia  
 Cervical subluxation  
 Cervical cord compression

## Anesthetic Implications of Larsen Syndrome<sup>4</sup>

- ▶ 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

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