

## INTRODUCTION

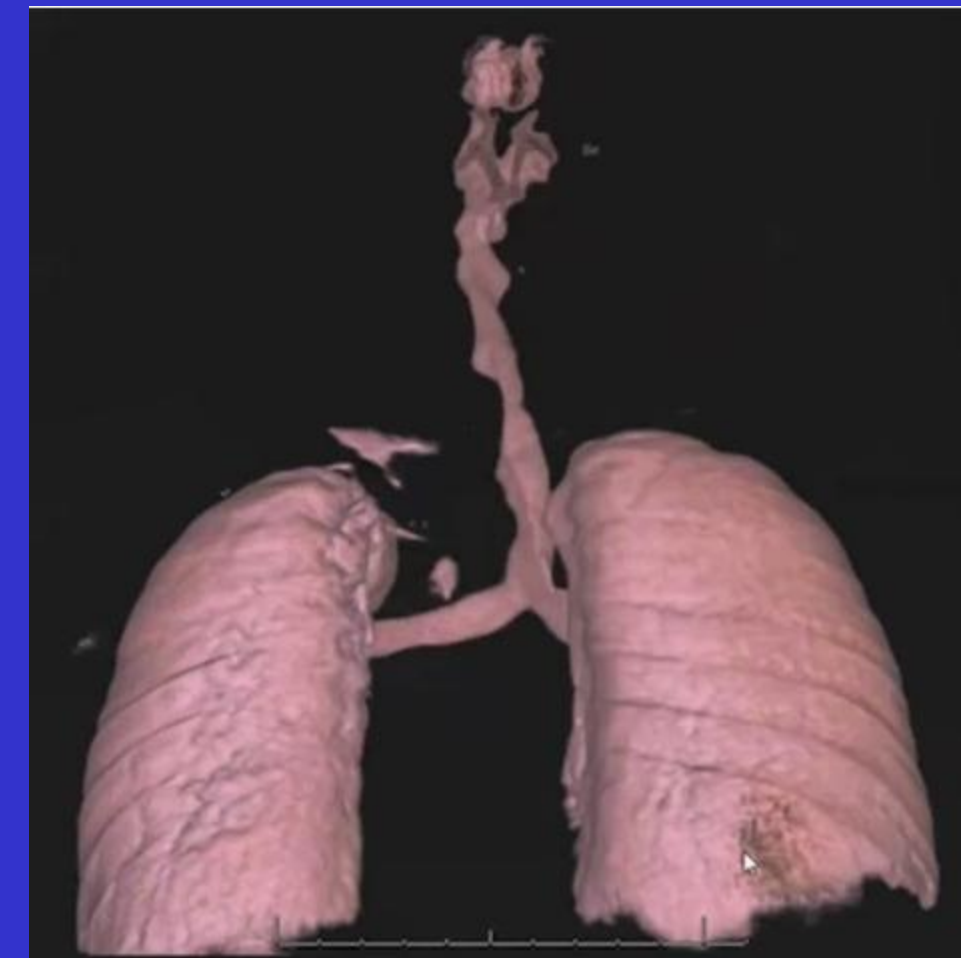
- Severe tracheal abnormalities have been reported in mucopolysaccharidosis (MPS) IVA patients (1,2).
- MRI images of the cervical spine report 67.9% of patients have at least 25% tracheal narrowing and this worsens with age (1).
- Tracheal obstruction along with difficulties in managing the upper airway of MPS IVA patients can lead to life-threatening complications during anesthesia (3).
- We present the results of MPS IVA patients who had CTA and report on tracheal abnormalities, with a plan to assess and manage the airway.

## METHODS

- IRB-approved, observational, longitudinal study.
- History and physical exam was performed.
- Important clinical symptoms included:
  - need to keep the head and neck extended
  - change in voice quality
  - difficulty in speaking due to the need to pause to inspire
- Data collected also included:
  - intraoperative airway management
  - CTA
  - pulmonary function testing

## RESULTS

- 66 patients have been enrolled, and 27 patients had CTA studies.
- Mean age: 18 years( range 5-36 years).
- 26 had tracheal abnormalities including:
  - narrowing
  - buckling
  - twisting (Figure 1 below)



<https://youtu.be/GLOtPHjGhN4>

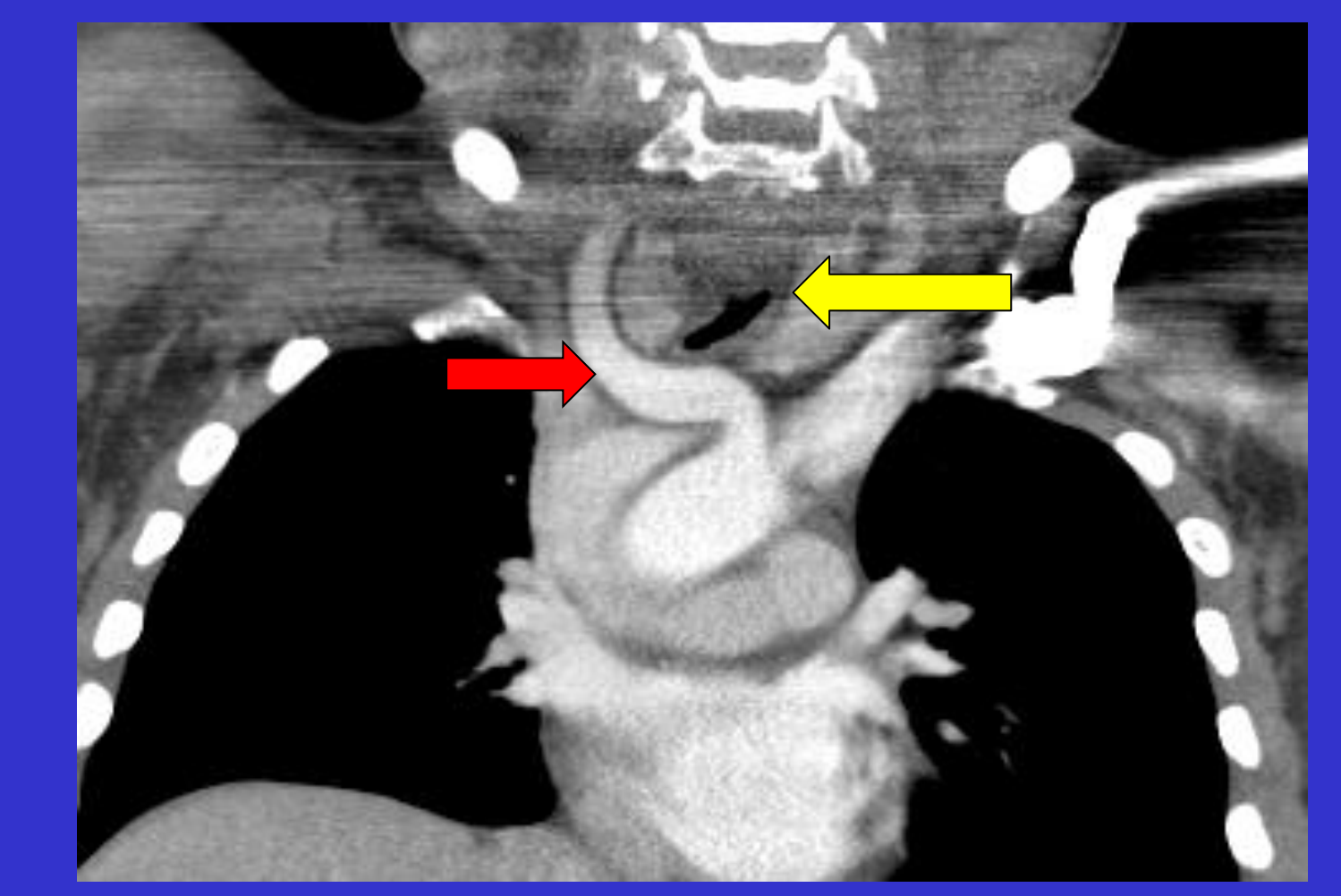


[https://youtu.be/BFSrnl2x\\_g](https://youtu.be/BFSrnl2x_g)

- Information regarding airway management was available for 21 patients (Table 1).

Airway Management	Number of Patients (n=27)
Tracheostomy	1
Direct Laryngoscopy	3
Unknown	6
Glidescope	15
Awake Fiberoptic	2

- Cardiothoracic surgery for tracheal stenosis: n=6.
- 6 patients had their trachea resected and anastomosed
- 5/6 had relocation of the innominate artery due to the crowded thoracic inlet and compression of the trachea by the innominate artery.
- 2 of the 6 patients were difficult to mask ventilate and intubate.



## DISCUSSION

- MPS patients have both upper and lower airway pathology.
- Tracheal pathology includes severe narrowing, tortuosity and compression.
- Tracheal and vascular reconstruction can aid in resolving airway stenosis as well as extrinsic compression caused by the innominate artery.