MPS IHS

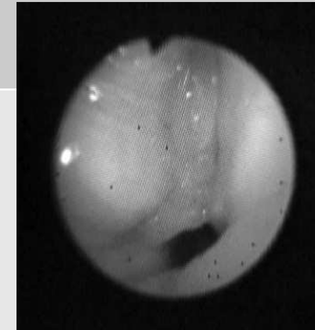
Clinical background

- 19-year old girl, small stature, 25 kg, presented for corneal graft surgery
- Pre-operative fiberoptic bronchoscopy on 3 occasions in previous 3 years showed thickened and narrowed glottic anatomy
- Recent decompression of spinal cord at craniocervical junction: patient remained intubated and ventilated for 3 days, followed by successful extubation
- Increasing hoarsness on presentation for corneal graft surgery

Procedure

- General anaesthesia using fiberoptic intubation and inhalation induction
  Airway deteriorated after loss of conciousness
  insertion of LMA size 2 first and then changed to size 2.5 resulting in good O2 saturation, but persistent degree of airway obstruction
  fiberoptic bronchoscopy: revealed extremely thickened larynx
  procedure abandoned to prevent further swelling

- Second trial of anaesthesia: same procedure, but airway less obstructed
  LMA size 2.5 placed when level of anaesthesia was sufficient and airway seemed relatively unobstructed (required some chin lift with LMA in place)
  again, bronchoscopy showed extremely narrowed airway
  glottis sprayed via suction channel of fiberoptic scope with lidocaine, followed by 3 mL of 1/10,000 epinephrine to shrink structures
  after a few minutes, trachea could be entered succesfully, a guide wire was placed, and a Cook airway exchanger railroaded over the guide wire after removal of the bronchoscope
  size 4.5 plain endotracheal tube railroaded over exchange catheter, but replaced by 4.5 mm microcuff endotracheal tube due to substantial leak
  after 2 mL of air cuff, successful ventilation of patient albeit with a small leak
  dexamethasone given during surgery to reduce any further glottic swelling
  successful extubation in anaesthetic room and discharged home 1 day later



Endoscopic image showing thickening of larynx (laryngeal opening 3-4 mm)