A Multidisciplinary Approach to Perioperative Management in a Neonate with Robinow Syndrome whose Parents Refuse Tracheostomy

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Background
• Robinow syndrome
  • Dwarfing syndrome
  • Facial features include hypertelorism with midfacial hypoplasia, small face, tented upper lip, ankyloglossia, and gum hyperplasia.
  • Congenital heart defects
  • Macrocephaly
  • Limb defects

Case
• A 2-week old 2.9 kg boy
  • Born at 37 week and 3 day gestational age
  • Diagnosed with Robinow Syndrome

Airway issues:
• Upper airway noises and desaturations particularly with feeding
• Fiberoptic laryngoscopy showed patent airway
• At 5 weeks of age, returned to the hospital for continued respiratory issues
• Patient then scheduled for G-tube
• Patient returned to hospital at 5 weeks of age for continued respiratory problems

Intraoperative gastrostomy tube course:
• Despite maintaining spontaneous ventilation, the intubation was difficult with video laryngoscopy. Ultimately, a 3.0 uncuffed ETT was placed.
• The rest of the intraoperative course was uneventful.
• Patient was taken to the PICU intubated.

• Patient failed extubation in the PICU and was emergently reintubated by an anesthesiologist. Patient returned to the operating room for surgical tongue lip adhesion and glossopexy in effort to avoid tracheostomy.

Summary
• Given the characteristic features of Robinow syndrome, careful planning needs to be considered due to potential difficult management of the airway.
• In this patient with continued respiratory failure, a tracheostomy should have been performed earlier, but lack of parental consent posed a challenge.
• This patient had other unnecessary procedures when he needed a tracheostomy from the beginning.

References

Figure 1: Surgical tongue lip adhesion with glossopexy to situate the tongue in a more anterior position in the neonate’s mouth.

Figure 2 (left): The suture stitches had dehisced. The tongue had receded and created an airway obstruction again.

Figure 3 (Right): Rigid bronchoscopy demonstrating airway edema.

Figure 4: Tongue recessed.