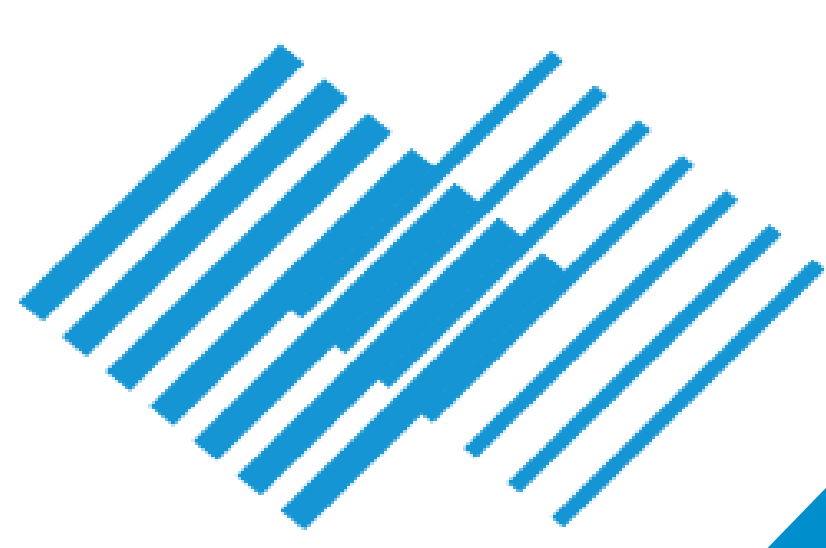




Anesthetic Considerations in Pediatric Patients with ZTTK Syndrome

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Introduction

Zhu-Tokita-Takenouchi-Kim (ZTTK) syndrome is a rare genetic disorder characterized by various facial dysmorphic features and congenital abnormalities. Caring for these patients involves unique diagnostic and perioperative challenges. We present the anesthetic management of a 1 year old boy requiring surgical repair of hypospadias.

Case Summary

- 1 year 7 months old boy, Weight: 7.8 kg
- Born 3 weeks early via emergency cesarean section, 20 days stay in NICU
- Medical history include congenital chordee, asthma, GERD and hypothyroidism
- No previous surgical history
- PE: patient appeared small for his age, no dysmorphic features noted
- Planned for staged repair of hypospadias; first stage being correction of the chordee and the second stage being urethroplasty

Preoperative Planning

- Anticipated difficult airway based on clinical history. Multimodal pain management given complexity of surgery
- Avoidance of inhalational anesthetic; possible underlying neuromuscular disorder.
 - Initial mask induction with cessation of inhalational agent following IV placement
 - TIVA with propofol and caudal anesthesia

Intraoperative Course

- Glidescope with S1 blade and CMAC with 0 blade standby
- Mask induction with sevoflurane briefly
- 22G peripheral IV placed in left lower extremity
- Caudal anesthesia with 6 mL 0.25% bupivacaine
- Direct laryngoscopy with MAC 2 blade → grade 1 view
- Endotracheal intubation with size 3.5 cuffed tube
- IV acetaminophen, fentanyl and morphine administered
- Successful chordee repair and extubation to room air

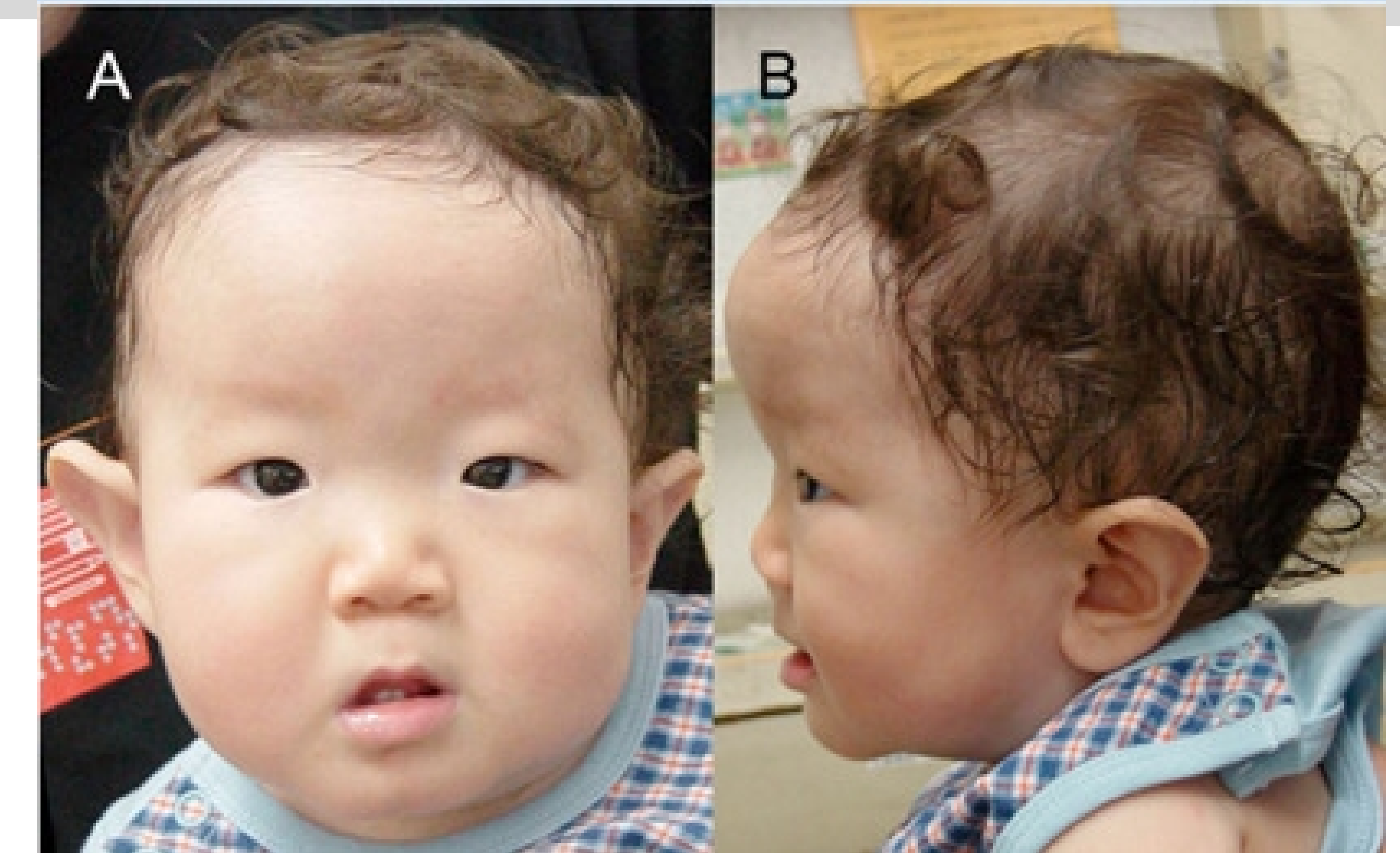


Figure 1: Facial features of the individual with Zhu-Tokita-Takenouchi-Kim syndrome reported in Takenouchi et al [2016]

Discussion

- ZTTK is a rare genetic disorder caused by a heterozygous mutation in the SON gene, first described in 2015, with around 50 cases reported to date.
- Facial dysmorphism may include facial asymmetry, macrocephaly, midface retrusion, and a short philtrum.
- Cases of rhabdomyolysis have been reported.
- Anesthesia teams should anticipate and prepare for difficult airway management.