

Bilateral Percutaneous Second Metatarsal Shortening Osteotomy in a Patient with Apert Syndrome

Matthew Colontonio², Ahmad Oseili², Sumaya Aboobacker², Edward Haupt¹, Jacob King²

¹Department of Orthopedic Surgery, Mayo Clinic, Jacksonville, FL; ²Department of Anesthesiology and Perioperative Medicine, Mayo Clinic, Jacksonville, FL

Background

Apert syndrome is a rare autosomal dominant congenital disorder with most cases arising from de novo mutations. It is characterized by craniosynostosis, syndactyly, and distinct facial anomalies. The patients have a mutation in the fibroblast growth factor receptor 2 (FGFR2) gene, which plays a critical role in skeletal development.

Individuals with Apert syndrome may also present with additional complications such as intellectual disability, midface hypoplasia, and cleft palate, though the severity of these symptoms can vary widely. Due to the facial abnormalities, difficult airway situations are often encountered in this patient population, necessitating extra vigilance before attempting intubation.

Early diagnosis and a multidisciplinary approach to treatment are essential for improving outcomes in individuals with Apert syndrome.

Case Presentation 1

A 16-year-old, 62 kg female with Apert syndrome sought a second opinion on foot surgery for bilateral foot metatarsalgia. Her extensive surgical history included 42 other surgeries, multiple craniofacial reconstructions, and tracheostomy secondary to tracheal stenosis which was later decannulated after tracheal reconstruction. She had an additional history of hearing loss, sleep apnea on CPAP, and asthma.

Anesthesia records from 5 years prior noted a difficult mask ventilation and airway requiring multiple attempts, with eventual success with a LMA, and a history of tracheostomy.

Physical examination on current admission showed decreased mouth opening, limited neck extension, hypersalivation, short neck with reduced motion, macroglossia, and micrognathia. Both the patient and family expressed a strong preference for avoiding general anesthesia due to concerns about the patient's difficult airway history. Joint decision-making was made to pursue bilateral ankle blocks with 0.5% ropivacaine.

The block was performed using a sterile technique and ultrasound guidance, targeting specific nerves with ropivacaine without complications. During the nerve block, she had anxiety requiring minimal sedation with incremental remimazolam. Her favorite music and her mother's presence were also implemented then. She was also given 1000 mg of Tylenol preoperatively for multimodal analgesia.

Case Presentation 2

Intraoperatively, the patient experienced no pain during the incision or osteotomy but remained anxious. Additional minimal to moderate sedation was provided with remimazolam, dexmedetomidine, and propofol. The procedure lasted 33 minutes, with 53 minutes total in the OR. The patient tolerated it well, with no additional IV analgesics intra-op. A supplemental block was required to obtain analgesia over an extended incision on the left foot.

In the PACU, she remained comfortable with a pain score of 2/10. She received Toradol, improving her pain score to 0. She was relieved to avoid general anesthesia and its associated risks due to her difficult airway and tracheal reconstruction.

Table

Timing	Anesthetic	Medications
Pre-operative	Bilateral ankle block	30 ml Ropivacaine 0.5%
Intra-operative	Supplemental left saphenous IV sedation	Bupivacaine 0.25% + Lidocaine 1% Remimazolam 8 mg, Dexmedetomidine 16 mcg Propofol 60 mg (3x 20 mg)
Post-operative	IV analgesia	Toradol 15 mg

Case Presentation 2

In conclusion, this case highlights the complexity of managing a patient with Apert syndrome, with a known difficult airway and significant anxiety during surgery. Using regional anesthesia and short acting sedation with remimazolam and music therapy allowed for a successful block and surgical outcome while minimizing the risks associated with general anesthesia.

References:

- Conrady CD, Patel BC, Sharma S. Apert Syndrome. [Updated 2023 Jun 1]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK518993/>
- Das S, Munshi A. Research advances in Apert syndrome. J Oral Biol Craniofac Res. 2018 Sep-Dec;8(3):194-199. doi: 10.1016/j.jobcr.2017.05.006. Epub 2017 May 25. PMID: 30191107; PMCID: PMC6107905.