TPM Vol. 32, No. S4, 2025
ISSN: 1972-6325
https://www.tpmap.org/

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ANESTHETIC MANAGEMENT OF A PEDIATRIC PATIENT WITH COCKAYNE SYNDROME FOR CORRECTIVE EQUINOCAVOVARUS DEFORMITY SURGERY

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Abstract
Background: Cockayne syndrome (CS) is a rare, autosomal recessive progeroid disorder presenting significant anesthetic challenges, including potential difficult airway, neurological sensitivity, and multiorgan dysfunction. We report the successful anesthetic management of a pediatric patient with CS undergoing corrective orthopedic surgery.
Case Presentation: A 13-year-old male, with a genetically confirmed diagnosis of Cockayne syndrome, presented with bilateral equinocavovarus deformity. His clinical features included microcephaly and developmental delay. He was scheduled for corrective surgery on the right foot. General anesthesia was induced with intravenous thiopentone, with the airway secured uneventfully using a 4.5 mm endotracheal tube. Anesthesia was maintained with sevoflurane and atracurium.
Discussion: The anesthetic plan was tailored to mitigate potential risks associated with CS. Atracurium was specifically chosen for neuromuscular blockade due to its organ-independent metabolism, a prudent choice in a syndrome with potential subclinical systemic dysfunction. A landmark-guided ankle block was utilized as part of a multimodal analgesic strategy to minimize systemic opioid requirements and the associated risk of postoperative respiratory depression in a neurologically vulnerable patient.
Conclusion: The intraoperative and postoperative periods were uneventful. This case demonstrates that with meticulous preoperative evaluation, a tailored anesthetic plan, and a multimodal approach to analgesia, patients with Cockayne syndrome can be safely managed for major orthopedic procedures.

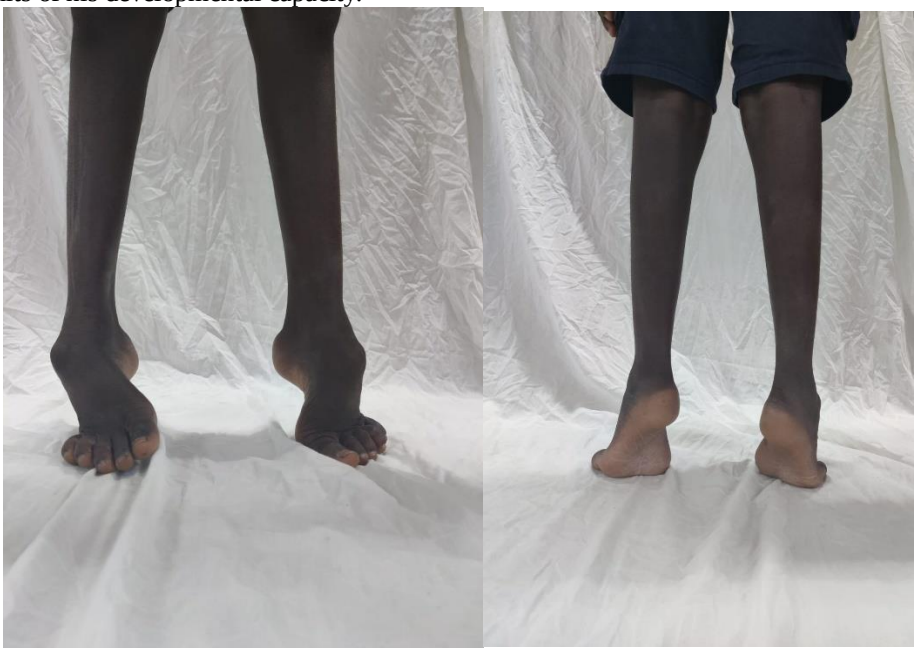
Keywords: Cockayne Syndrome, Anesthetic Management, Difficult Airway, Regional Anesthesia, PediatricAnesthesia.

INTRODUCTION

Cockayne syndrome (CS) is a rare, autosomal recessive disorder characterized by profound postnatal growth failure, premature aging (progeria), photosensitivity, and progressive neurological dysfunction, including microcephaly and developmental delay. The multisystem nature of CS presents a unique constellation of challenges for the anesthesiologist.
Key anesthetic considerations include a high risk of a **difficult airway** due to facial dysmorphism, micrognathia, and potential temporomandibular joint limitation. Neurological abnormalities may alter patient sensitivity to anesthetic agents and lower the seizure threshold. Furthermore, associated cardiac, renal, and thermoregulatory dysfunctions necessitate careful perioperative management. Given the rarity of the syndrome, literature on anesthetic management is limited primarily to case reports. We present the successful anesthetic management of a 13-year-old male with CS undergoing corrective surgery for an equinocavovarus foot deformity.

Case Presentation

A 13-year-old, 25 kg male with a genetically confirmed diagnosis of Cockayne syndrome was scheduled for a right-sided equinocavovarus deformity correction. His medical history was significant for profound developmental delay and microcephaly. On preoperative evaluation, the patient was calm and cooperative within the limits of his developmental capacity.



Preoperative Evaluation

His significant medical history included profound **developmental delay** and **microcephaly**, which are hallmark features of the syndrome. A thorough preoperative assessment was conducted to identify potential anesthetic risks.

- **Airway Assessment:** The physical examination revealed micrognathia (an undersized jaw) and a small mouth opening. However, the patient had adequate neck mobility, and there were no other indicators to suggest an exceptionally difficult intubation. Despite the reassuring findings, preparations were made for a potential difficult airway, with alternative airway devices readily available.
- **Systemic Evaluation:** The patient's cardiovascular and respiratory examinations were unremarkable, with no clinical signs of cardiac failure or respiratory compromise. Baseline laboratory investigations, including a complete blood count and electrolyte panel, were all within normal limits.
- **Consent:** After a detailed discussion of the anesthetic plan and associated risks with the patient's guardians, informed consent was obtained.

Intraoperative Management

Upon arrival in the operating room, standard American Society of Anesthesiologists (ASA) monitoring was applied, including electrocardiography, pulse oximetry, non-invasive blood pressure, and end-tidal capnography.

- **Induction and Airway:** General anesthesia was induced with an intravenous injection of **thiopentone (50 mg)**. Following confirmation of easy mask ventilation, neuromuscular blockade was facilitated with **atracurium (7.5mg)**. Direct laryngoscopy provided a clear view of the vocal cords (Cormack-Lehane Grade I), and the trachea was smoothly intubated with a 4.5 mm cuffed endotracheal tube.
- **Maintenance:**Anesthesia was maintained with **sevoflurane** in a 50% oxygen-air mixture to ensure adequate oxygenation and anesthetic depth. Intermittent boluses of atracurium were administered as needed, guided by clinical signs.
- **Regional Anesthesia:** To provide effective postoperative pain relief and minimize the need for systemic opioids, a landmark-guided **ankle block** was performed on the right foot. A total of 10 mL of 0.25% bupivacaine was administered to block the relevant peripheral nerves.
- **Surgical Course:** The surgical correction of the equinocavovarus deformity proceeded as planned and lasted 90 minutes. The patient remained hemodynamically stable throughout the procedure, with no significant fluctuations in heart rate or blood pressure. There were no intraoperative complications.

Postoperative Recovery

- **Emergence and Extubation:** At the conclusion of the surgery, the residual effects of the neuromuscular blockade were reversed. The patient began to breathe spontaneously and, once fully

awake and meeting all standard extubation criteria (e.g., following commands, adequate respiratory effort), was smoothly extubated in the operating room.

- **Analgesia and Monitoring:** The patient was transferred to the Post-Anesthesia Care Unit (PACU) for close monitoring. The ankle block provided excellent and effective analgesia, and the patient reported no significant pain. He did not require any opioid analgesics in the immediate postoperative period.
- **Outcome:** The postoperative course was uneventful. There were no signs of respiratory depression, prolonged muscle weakness, or any other anesthetic-related complications. After a standard period of observation in the PACU, the patient was transferred to the orthopedic ward in stable condition.

DISCUSSION

The anesthetic management of a patient with Cockayne syndrome requires a comprehensive understanding of its multisystem manifestations. The primary concern is often the potential for a difficult airway. While our patient exhibited micrognathia, intubation was fortunately straightforward. However, preparedness for a difficult airway, with appropriate equipment readily available, is mandatory.

Our choice of anesthetic agents was tailored to the patient's condition.

- **Thiopentone** provided a smooth induction.
- **Atracurium** was selected for neuromuscular blockade due to its organ-independent metabolism (Hofmann elimination), making it a safe choice in patients with potential, even if subclinical, renal or hepatic dysfunction.
- **Sevoflurane** is a suitable maintenance agent due to its low arrhythmogenic potential and rapid titratability.

The incorporation of a **regional anesthetic technique** (ankle block) was a key component of our management. This multimodal approach to analgesia minimized the need for systemic opioids, thereby reducing the risk of postoperative respiratory depression—a significant benefit in a patient with underlying neurological impairment. The uneventful intraoperative and postoperative course in our patient underscores the success of this tailored approach.

CONCLUSION

Cockayne syndrome presents a complex array of potential perioperative risks. This case report illustrates that with thorough preoperative assessment, careful planning for a potentially difficult airway, judicious selection of anesthetic agents, and the use of multimodal analgesia including regional blocks, patients with this rare syndrome can be managed safely and effectively through major surgical procedures.

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