

Anaesthetic management of a pediatric patient with progeria undergoing orchiectomy: A case report anaesthetic management in progeria

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Abstract

Background

There is a rare genetic condition called Hutchinson-Gilford progeria syndrome that causes premature aging. Perioperative management of such cases is challenging and needs meticulous evaluation and monitoring.

Clinical description

This report discusses the successful anaesthetic management of a 9-year-old male child with Hutchinson-Gilford Progeria syndrome (HGPS), who was scheduled for an elective left orchidectomy at our hospital.

Management and outcome

The case was successfully managed under general anaesthesia with meticulous planning and vigilant intraoperative monitoring. We secured the airway with a ProSeal laryngeal mask airway and monitored the electrocardiogram, heart rate, blood pressure, oxygen saturation using pulse oximetry, and end-tidal carbon dioxide intraoperatively. The child was successfully extubated at the end of surgery.

Conclusions

Children with progeria present particular anesthetic challenges because of anatomical variations and possible multisystem involvement. For successful results, titrated anesthetic dosage, careful preoperative evaluation, and attentive intraoperative monitoring are necessary.

Keywords: Anaesthesia, progeria, orchidectomy

Introduction

Hutchinson-Gilford progeria syndrome (HGPS) is a rare genetic disorder characterized by premature aging, with an estimated incidence of 1 in 4-8 million live births [1]. Affected children present with unique anesthetic challenges due to craniofacial abnormalities, cardiovascular risks, and metabolic considerations. Literature on anesthetic management in progeria remains limited, making case reports valuable for guiding clinical practice.

Case Report

A 9-year-old male, weighing 18 kg, diagnosed with Hutchinson-Gilford Progeria syndrome (HGPS), was scheduled for left orchidectomy at our hospital. The patient had no additional comorbidities. Preoperative workup, including echocardiography and computed tomography (per the pediatric surgeon's request), was normal. Family history revealed that the elder sister also suffered from progeria.

Anaesthesia was induced with intravenous midazolam 0.5 mg, fentanyl 45 µg, propofol 50 mg, cisatracurium 2 mg, and glycopyrrolate 80 µg. Airway management was achieved with a ProSeal laryngeal mask airway (LMA). Anaesthesia was maintained with sevoflurane in a mixture of medical air and oxygen. Standard American Society of Anesthesiologists (ASA) monitors were used (electrocardiogram, heart rate, blood pressure, oxygen saturation using pulse oximetry, and end-tidal carbon dioxide). Intraoperative analgesia included paracetamol 250 mg IV, and fluid management consisted of 150 mL of Ringer's lactate. The surgical site was infiltrated with 0.25% bupivacaine 10 ml by the surgeon. At the conclusion of

surgery, reversal of neuromuscular blockade was achieved with glycopyrrolate 200 µg and neostigmine 1 mg. The patient was smoothly awakened, the LMA was removed without complications, and he was transferred to the ward after an uneventful recovery. The child was discharged from the hospital uneventfully the next day.

Management and Outcome

Anaesthetic management in patients with progeria requires meticulous planning due to multisystem involvement [2]. Although our patient did not demonstrate cardiac or respiratory compromise, children with HGPS are at risk of coronary artery disease, hypertension, and cerebrovascular accidents. Airway management may be challenging due to craniofacial dysmorphism, micrognathia, and limited neck mobility. The use of ProSeal LMA provided a safe and effective alternative to tracheal intubation in this case.

Discussion

This report discusses the successful anaesthetic management of a 9-year-old male child with Hutchinson-Gilford Progeria syndrome (HGPS), who successfully underwent an elective left orchidectomy under general anaesthesia. Drug dosing in progeria requires caution, as altered body composition and increased sensitivity may predispose to exaggerated responses [3]. We chose lower induction doses and titrated carefully. Intraoperative hemodynamic stability and smooth recovery were achieved, consistent with a few similar case reports published in the literature. In these patients, the skin is fragile [4]. Therefore, gentle handling and use of extra-soft padding to prevent skin tears

and pressure sores is recommended. Osteopenia and pathologic fractures occur even with minor stress, and therefore, positioning should be done with extreme caution by avoiding overextension of joints.

This case contributes to the limited data available on perioperative anesthetic care in progeria and highlights the importance of individualized anesthetic planning, careful monitoring, and readiness for airway or cardiovascular complications.

Conclusion

To conclude, children with progeria pose unique anaesthetic challenges due to potential multisystem involvement and anatomical variations. Careful preoperative assessment, titrated anesthetic dosing, and vigilant intraoperative monitoring are essential for favorable outcomes. Our case demonstrates that safe anesthesia can be achieved with proper planning and multidisciplinary care.

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