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Anesthetic Management for Cesarean Section in a Patient with Achondroplasia: Case Report and an Updated Review

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ABSTRACT

Achondroplasia is a rare genetic disorder and the leading cause of dwarfism worldwide. Patients with achondroplasia often present with spinal abnormalities, respiratory and cardiac disorders, obstructive sleep apnea, and facial deformities. Cesarean section is commonly required due to cephalopelvic disproportion in these patients. However, the choice of anesthetic technique for cesarean sections in this population is controversial and should be based on a careful assessment of the risks and benefits of each approach. In this article, we present a case of a 39-year-old primiparous woman with achondroplasia who underwent a cesarean section under titrated epidural anesthesia. We also discuss other anesthetic techniques described in the literature, highlighting their advantages and limitations.

Keywords

Achondroplasia, Anesthesia, Cesarean section, Epidural anesthesia.

Introduction

Achondroplasia is an autosomal dominant genetic disorder characterized by dwarfism, resulting from mutations in the fibroblast growth factor receptor 3 (FGFR3) gene [1]. Cesarean section is often necessary for women with achondroplasia due to their small pelvis and increased risk of cephalopelvic disproportion [2]. However, choosing the appropriate anesthetic technique for cesarean sections in these patients is challenging due to potential difficulties in airway management, cervical spine instability, and uncertainty in predicting the level of neuraxial blockade [3]. Existing literature on anesthetic management of pregnant women with dwarfism is limited to case reports and case series, encompassing various techniques such as general, spinal, epidural, and combined anesthesia [4]. Here, we present a case report of cesarean section in a patient with achondroplasia performed under continuous epidural anesthesia with dose titration. We also discuss alternative anesthetic techniques to aid in selecting the safest and most effective approach based on individual patient characteristics.

Case Report

We describe the case of a 39-year-old primiparous woman with achondroplastic dwarfism, without comorbidities or allergies. Physical examination revealed 55 kg, height 138cm, lumbar hyperlordosis, macroglossia, limited mouth opening and cervical extension, and Mallampati class IV. Labor induction was initially planned at 38 weeks and 5 days of gestation, but a cesarean section was elected due to cephalopelvic disproportion. In the operating room, standard monitoring was applied, and the patient's baseline blood pressure, heart rate, and oxygen saturation were within normal ranges. After considering predictors of difficult airways and the challenges in predicting neuraxial blockade levels with spinal anesthesia, we opted for epidural anesthesia with dose titration via catheter. The patient was positioned seated, and after local anesthesia, an 18G Tuohy needle was inserted at the L3-L4 level. A catheter of the same caliber was inserted. Subsequently, lidocaine 2% with vasoconstrictor was injected in small increments until a satisfactory blockade was achieved (240

mg of anesthetic). Additional medications included morphine, dipyrone, dexamethasone and ondansetron. The patient remained stable throughout the procedure, and the infant had a favorable Apgar score. The patient was closely monitored postoperatively and discharged within two days.

Discussion

The anesthetic management of cesarean sections in patients with acondroplasia requires individualized consideration due to the limited scientific evidence available. Most of the literature consists of individual case reports or case series. Neuroaxial anesthesia, such as spinal anesthesia, can be technically challenging due to spinal stenosis, possible vertebral anomalies (e.g., kyphoscoliosis), and the need for higher doses to achieve sufficient blockade in pregnant women with acondroplasia. In contrast, epidural anesthesia with small, gradual boluses may be considered safer as it allows for dose titration. However, difficult catheter insertion and the time required for the technique make it less suitable for emergency cases [4]. Moreover, the inadvertent perforation of the dura mater and subsequent total spinal anesthesia are possible complications. General anesthesia also presents challenges, especially related to potential difficult airway management. Predictors of intubation difficulty, such as cervical instability, macroglossia, and limited cervical extension, are often present in these cases. Additionally, mechanical ventilation can be challenging due to restrictive lung disease, decreased functional residual capacity (FRC) caused by scoliosis, and obstructive sleep apnea.

In the table below, the possible advantages and disadvantages of each technique are listed.

	General anesthesia	Spinal anesthesia	Epidural anesthesia
Advantages	- Widely known technique.	- Simpler and faster technique when compared to epidural catheterization. - Fast recovery. - Fewer respiratory repercussions.	- Possibility of titration reducing the risk of insufficient or excessively high blocking level.
Disadvantages	Technical difficulties related to dwarfism: - Disproportionally large head. - Macroglossia. - Small mouth. - Short jaw. - Large jaw. - Atlantoaxial instability. - Limited neck extension. - Restrictive lung diseases with decreased functional residual capacity (FRC) due to scoliosis. - Obstructive sleep apnea. - Pulmonary hypertension. - Cor pulmonale. Difficulties inherent to pregnancy: - Mucosal edema. - CRF reduced by 20%. - Increased risk for bronchoaspiration.	Technical Difficulties: - Lumbar hyperlordosis. - Difficulty in identifying the intervertebral spaces. - Anatomical distortion. Factors that lead to unpredictability of the blocking level: - Short stature. - Spinal canal stenosis. - Pregnancy (increased abdominal pressure).	Technical Difficulties: - Narrow spinal canal. - Vertebral Deformities. - Difficulty in identifying landmarks. - Difficulty in inserting the catheter, with the possibility of inadvertent intrathecal insertion. A more difficult and time consuming technique, not indicated in cases of urgency.

Table 1: Advantages and disadvantages of general, epidural and spinal anesthesia.

Some studies have reported successful emergency cesarean sections using spinal anesthesia in acondroplastic patients, but there is no consensus regarding the ideal dose [5,6]. However, inadequate blockade has also been reported, requiring intravenous sedation after administration of hyperbaric bupivacaine and intrathecal morphine. These findings have led to recommendations for the use of epidural anesthesia, continuous spinal anesthesia, or combined (CSE) anesthesia with small injections of local anesthetics and incremental doses [7][8]. Other studies have reported complications and technical difficulties associated with neuraxial anesthesia, including multiple attempts at needle placement, paresthesia, and the need for conversion to general anesthesia [9]. Lower doses of local anesthetics, both subarachnoid and epidural, are generally required to achieve adequate blockade in acondroplastic patients. General anesthesia data is controversial, with reports of difficulties in ventilation and intubation [10,11].

Based on this review, it can be observed that acondroplastic patients usually require lower doses of neuraxial anesthetics (epidural or spinal) to achieve sufficient blockade. However, a safe dose cannot be definitively determined. The various anatomical changes associated with acondroplasia contribute to predictors of difficult ventilation and intubation, making general anesthesia less attractive as a primary choice but still an excellent backup method in case of failed neuraxial blockade [9].

Conclusion

The literature on the anesthetic management of cesarean sections in patients with acondroplasia is divergent and unclear, highlighting the importance of individualizing the choice of anesthesia technique. A thorough examination of the specific anatomical alterations in the pregnant patient is crucial, along with consideration of potential complications and preparedness for alternative plans in case the initial choice of anesthesia is unsuccessful.

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