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## Case Report

### A case of achondroplasia for emergency cesarean section

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#### Abstract

Patients with achondroplasia present with various anatomical and physiological features including skeletal, neurological, and cardiopulmonary abnormalities. These characteristics create a difficult scenario wherein the selection of general or regional anesthesia becomes a dilemma. Moreover, pregnancy-induced changes in gravid achondroplastic patients further complicate the anesthetic management. The unavailability of proper guidelines and controversies regarding proper anesthetic techniques, drug choice, dosage, and involvement of multiple organ systems challenges anesthesiologists. Here, we present a case of a gravid achondroplastic patient, with a height of 112 cm and mild thoracolumbar scoliosis with intrauterine fetal death in labor, scheduled for emergency lower segment cesarean section and who was managed successfully with spinal anesthesia using low dose anesthetic agents.

**Keywords:** Cesarean delivery; Dwarf; Short stature; Spinal anesthesia

#### Introduction

Achondroplasia is an autosomal dominant genetic disorder with a mutation in fibroblast growth receptor gene-3 (FGFR3).<sup>1</sup> The prevalence rate is 1:5000 to 1:40000 per live births.<sup>2</sup> These individuals present with typical craniofacial features and spinal deformities with cardiorespiratory and neurological issues leading to difficulty in anesthesia management. The preferred mode of delivery is the cesarean section due to congenitally small and contracted pelvis.<sup>2</sup> Our objectives are to highlight issues associated with gravid achondroplastic patients that challenge anesthesiologists and to extend knowledge regarding the use of spinal anesthesia in these patients. To the best of our knowledge, this is the first published report of such a case from Nepal.

#### Case Report

A 20-year-old primigravida with achondroplasia at 31-32 weeks of gestation (by ultrasound) in labor presented with absent fetal movements for 3 days and cephalopelvic disproportion. Ultrasound abdomen showed intrauterine fetal death and oligohydramnios.

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After an unsuccessful trial of the expulsion of the dead fetus vaginally, an emergency cesarean section was decided as the patient was in labor. On examination, her weight and height were 40 kg and 112 cm respectively. She had a large head, short limbs, short neck, protruded chin, and mild thoracolumbar scoliosis. Airway assessment showed adequate mouth opening with Mallampati grade I, a large tongue, and normal neck movements. Systemic examinations and laboratory investigations were within normal limits. Chest X-ray and cervical spine AP and lateral were normal. The X-ray of thoracolumbar spine AP and lateral detected thoracolumbar scoliosis with wedging at L2 vertebrae. The blood pressure was 140/68 mm Hg, pulse rate was 128 bpm, regular and SpO<sub>2</sub> was 98% on room air. Subsequently, emergency lower segment cesarean section under spinal anesthesia was planned. A difficult airway cart was arranged. A peripheral intravenous line was secured using 18G cannula in the right forearm and 500 ml of ringer lactate was commenced. Injection ranitidine 50 mg,

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metoclopramide 10 mg, and ceftriaxone 1gm were administered intravenously as per routine practice.

Spinal anesthesia was performed in the sitting position at L3-L4 interspace using a 25 gauge Quincke spinal needle with a median approach and subarachnoid space was encountered at the first attempt. After obtaining a free flow of cerebrospinal fluid, injection fentanyl 0.2ml (10 mcg) and 0.5% heavy bupivacaine 1 ml (5mg) were injected. The patient was positioned in a modified supine position with 15 degrees left lateral tilt. Single, female, dead baby with weight 2.1 kg was delivered and injection oxytocin 5 IU intravenously was given. The patient experienced slight discomfort during peritoneal traction, injection midazolam 1 mg, and fentanyl 40 mcg was hence given intravenously. The surgery lasted for approximately 40 minutes. The estimated blood loss was approximately 200 ml and a total of 500ml normal saline and 500 ml ringer lactate were infused. After an hour of observation in the recovery room, sensory block height for pain and temperature were at L1 and T8 respectively. Assessment of motor block showed an inability to lift legs but with the ability to move them while eliminating gravity. The patient was shifted to the ICU for overnight observation and was transferred to the ward the next morning. Finally, she was discharged on 4th postoperative day from the ward.

### **Discussion**

Pregnancy-induced physiological changes such as airway edema, decreased functional residual capacity, hypoxia, risk of aspiration of stomach contents and supine hypotension syndrome are well-known contributors for the difficulty in anesthesia management.<sup>2</sup> Moreover, the uterus remains an intrabdominal organ in achondroplastic parturients, resulting in a further reduction in their functional residual capacity, an increased risk of aspiration, and severe aortocaval compression.<sup>3</sup> Achondroplastic are prone to possess difficult airway features such as large head, large mandible, macroglossia, hypoplastic larynx, atlantoaxial instability, and limited neck extension. Spinal abnormalities include kyphoscoliosis, lumbar hyperlordosis, and spinal stenosis.<sup>3</sup> Cardiorespiratory and neurological issues are also present.<sup>2</sup> Preoperative MRI and CT scan images guide regarding difficult airway and spinal deformities.<sup>3</sup> Since our patient was diagnosed with IUFD, we took appropriate X rays to detect any skeletal and vertebral deformities, which are common in achondroplasia.

Spinal anesthesia with landmark technique was selected as we routinely practice it for cesarean delivery. In these patients, deformities of vertebrae

and spinal stenosis pose difficulty in identifying interspinous space and a dry tap.<sup>3</sup> Fortunately, lumbar interspinous spaces were easily palpable in our patient and CSF flow was obtained in the first attempt. We injected 1ml of 0.5% heavy bupivacaine (5 mg) and 0.2 ml (10 mcg) of fentanyl in the subarachnoid space, which were consistent with previous studies which suggest the minimum effective dose of intrathecal bupivacaine should not exceed 0.06 mg/cm of height for achieving desired spinal block to avoid hemodynamic instability due to exaggerated block height.<sup>4,5</sup> In our patient, we used minimum dosage, which was bupivacaine 1ml (5mg) and fentanyl 0.2ml (10 mcg) and we didn't exceed the dose of bupivacaine for more than 6.72mg (0.06mg/cm) to avoid high block height, which could have lead to unstable hemodynamics and respiratory complications. We used 0.045 mg/cm height as it was within the maximum limit of the dose. Our dosage was consistent with Sukanya et al, where for a height of 109cm, 1ml of 0.5% heavy bupivacaine and 0.2ml fentanyl was used to obtain T4-T6 level. There was no decline in BP and this low dose was concluded to be safe and effective for surgical procedures of 1-hour duration.<sup>6</sup> Similarly, Madanmohan et al used 1 ml (5 mg) of bupivacaine along with Inj fentanyl 0.3 ml (15 mcg) in a 110 cm height achondroplastic parturient and achieved a sensory blockade of T4 and motor blockade of B3.<sup>7</sup> Thus, as stated by previous studies, the recommended consensus is lowering the dose of local anesthetics with a combination of opioids.<sup>5,6,7</sup> In a case report, where the height of the achondroplastic patient was not mentioned, a subarachnoid dose of 1.3ml of 0.5% bupivacaine with 10mcg fentanyl was injected and a block height up to T3 was achieved with a transient decrease in BP, which was managed successfully using ephedrine. They proposed a 30 % reduction in the standard dose regimen.<sup>8</sup> It should be noted that the initiation of spinal anesthesia leads to severe hypotension and aortocaval compression in these patients.<sup>9</sup> The management includes adequate volume co-loading and the use of vasopressors when required.<sup>9</sup> Although there were no significant hemodynamic changes, our patient felt slight discomfort due to peritoneal traction during the procedure, for which we added midazolam 1 mg and 40 mg fentanyl intravenously to calm her as achondroplastic patients have pronounced anxiety than the general population.<sup>10</sup> Nevertheless, advantages of regional anesthesia and avoidance of exposure to the hazards of general anesthesia cannot be ignored.<sup>8</sup> USG guided approach is beneficial in increasing the likelihood of a successful block.<sup>9</sup>

Moreover, the unpredictable spread of drugs through a stenosed spinal canal may lead to the risk of a high block. Therefore, cautious use of spinal anesthesia and careful titration of drugs through the catheter to ascertain the proper height of the block during epidural anesthesia has been recommended.<sup>1,6</sup> Nonetheless, literature review reports cases successfully managed by both epidural and spinal anesthesia. We believe that spinal anesthesia can be appropriate in emergency cases, where epidural anesthesia is practiced less frequently. Besides that, a narrow epidural space leads to difficulties in placing epidural catheter and threading, epidural venous puncture, inadvertent subarachnoid injection, and subarachnoid tap with difficulty recognizing subarachnoid tap due to impaired CSF flow.<sup>1</sup>

Apart from managing a difficult airway and pregnancy-induced upper airway changes, associated thoracolumbar kyphoscoliosis, rib deformities, restrictive lung disease, recurrent respiratory tract

infections, and obesity complicates the administration of general anesthesia.<sup>1,2</sup> Besides, the size of the endotracheal tube should be smaller and predicted by weight and not by age.<sup>9</sup> The awake fiberoptic technique is preferred for intubation. Postoperative mechanical ventilation and intensive care should be planned due to an increased risk of postoperative complications due to impaired cardiopulmonary function.<sup>1,2</sup> For difficult peripheral venous access due to short stubby extremities, ultrasound devices are considered extremely helpful.<sup>1</sup>

### **Conclusion**

The pros and cons of the chosen anesthetic technique should be weighed based on the individual attributes of each achondroplastic patient. A low dose spinal anesthesia is an excellent and suitable technique. However, one should be ready to manage a difficult airway. An immense care is advocated to safely conduct cesarean section in an achondroplastic mother.

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