



Case Report

Achondroplastic Parturient and Regional Anaesthesia: Is Ultrasound A Panacea

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ABSTRACT

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Background: Achondroplasia, the most common form of disproportionate dwarfism, presents significant anesthetic challenges during pregnancy because of craniofacial abnormalities, obesity, altered spinal anatomy, and potential cardiorespiratory compromise. Neuraxial anesthesia is generally preferred for cesarean delivery to avoid airway manipulation; however, technical difficulties and unpredictable local anesthetic spread may complicate its administration. Pre-procedural Ultrasound can improve neuraxial block success by facilitating identification of anatomical landmarks and estimation of epidural depth.

Case: A 29-year-old primigravida with achondroplasia at 36 weeks of gestation underwent emergency lower segment cesarean section for cephalopelvic disproportion. She was 112 cm tall with a body mass index of 40.7 kg/m². Airway assessment revealed limited mouth opening and grade III tonsillar hypertrophy, suggesting potential airway difficulty. Considering the risks associated with general anesthesia, a combined spinal-epidural technique was planned. Ultrasound-guided assessment identified the epidural space at a depth of 3.93 cm. Following intrathecal administration of low-dose bupivacaine(H) (5 mg) with fentanyl (10 µg), the sensory block was gradually extended to the desired level using epidural top-ups. Surgery and recovery were uneventful, postoperative analgesia was maintained with continuous epidural ropivacaine infusion.

This case highlights the value of meticulous pre-anesthetic evaluation and individualized anesthetic planning in achondroplastic parturients. Ultrasound guidance improves neuraxial landmark identification and depth estimation; however, clinicians should recognize potential discrepancies between ultrasound-estimated and actual needle depth due to tissue compression, obesity, pregnancy-related changes, and needle angulation. Combining ultrasound guidance with clinical expertise can facilitate safe and effective neuraxial anesthesia while minimizing airway-related risks in this high-risk population.

Keywords: Achondroplasia; Cesarean section; Combined spinal-epidural anesthesia; Ultrasound-guided neuraxial block; Difficult airway.

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INTRODUCTION

Achondroplasia is the most common form of short-limbed dwarfism and is inherited in an autosomal dominant pattern. It exhibits a slight female preponderance and has an estimated incidence of 1 in 25,000 to 30,000 live births (1). Characteristic clinical features include a proportionate trunk, shortened limbs, limited elbow extension

and macrocephaly with a prominent forehead(2). The fingers are short and a separation between the middle and ring fingers can produce a distinctive "trident hand" appearance.

In achondroplastic patients anaesthetic complexities stem from craniofacial abnormalities, skeletal deformities, and associated systemic anomalies. In pregnant patients, the anatomical and physiological changes, makes airway management and neuraxial anaesthesia more challenging. This case report highlights the need for careful pre-anaesthetic assessment, and a tailored anaesthetic plan anticipating airway and neuraxial difficulties and the role of USG to ensure maternal and fetal safety during the peripartum period.

Case History

A 29-year-old primigravida with achondroplasia presented to the obstetrics casualty at 36 weeks of gestation in active labour. She was scheduled for an emergency lower segment caesarean section (LSCS) in view of with cephalopelvic disproportion (CPD). Her antenatal course was unremarkable with no obstetric complications. On examination, she was 112 cm tall, weighed 51 Kg, her BMI of 40.7 Kg/M² was consistent with morbid obesity. She had short limbs, a hallmark of achondroplasia.

Airway assessment revealed a mouth opening of 2.8cm, modified Mallampati (MMP) Class II (in supine position), normal neck mobility and grade III tonsillar hypertrophy (Figure 1). Systemic examination was unremarkable. Preoperative investigations, including complete hemogram, liver and renal function tests and coagulation profile were within normal limits.



Figure 1: Airway assessment showing Mallampati Class II (in supine position) and grade III tonsillar hypertrophy

She was administered aspiration prophylaxis and in the operating room (OR), standard monitors were attached and base line vitals were recorded which were within normal limits. An 18-G intravenous cannula (Figure 2) was placed under ultrasonography (USG) guidance in the left hand. A difficult airway cart was kept ready in view of potential airway challenges. The anaesthesia plan was a combined spinal epidural (CSE). In the sitting position, epidural and subarachnoid spaces were identified and depth (3.93 cm, Figure 3) by USG. Under all aseptic precautions, after local infiltration, an 18-G Tuohy needle was inserted at L3-L4 level, loss of resistance (LOR) was appreciated at 4.5 cm, the spinal needle was then inserted, free flow of CSF was observed and 5 mg (1ml) of hyperbaric bupivacaine and 10 mcg fentanyl was given intrathecally, followed by threading of epidural catheter which was fixed at 9 cm.

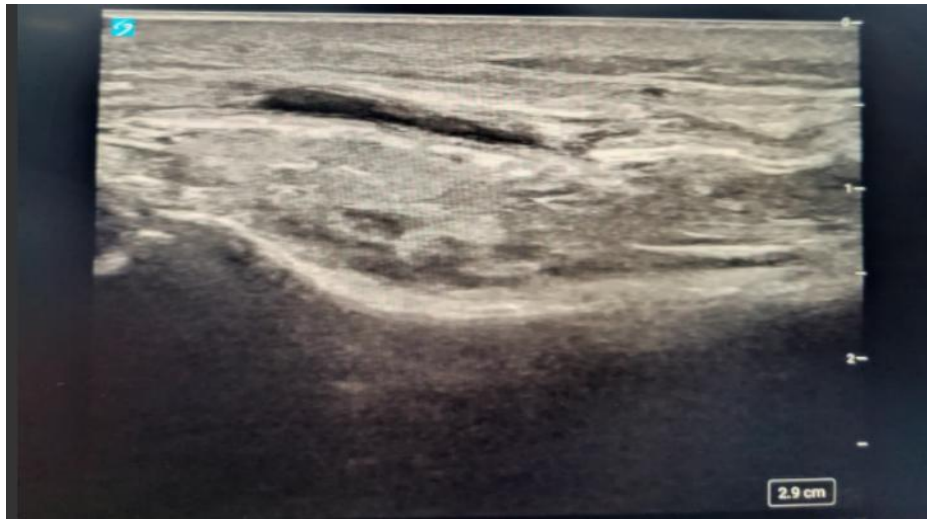


Figure 2: USG showing peripheral vein for venous access

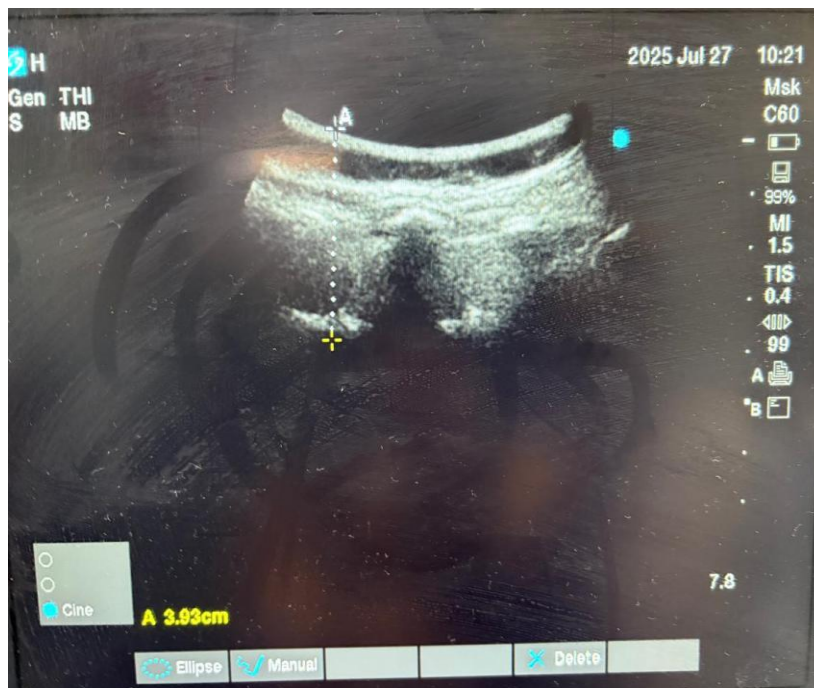


Figure 3: USG of epidural space taken at L3-L4 level showing epidural space at 3.93 cm from skin

A sensory level up to T10 was achieved. Sensory level of block was augmented to T6 with 0.5% bupivacaine (4+ 4 mL in aliquots). A pelvic wedge was placed, and oxygen was supplemented via nasal prongs. A healthy live male infant weighing 2.5 kg was delivered 5 minutes after skin incision. The intraoperative course was uneventful; and the patient was transferred to the recovery after surgery. Analgesia was managed via the epidural catheter, using an elastomeric pump delivering 0.2% Ropivacaine at 6 mL/hour, continued for 48 hours.

DISCUSSION

Achondroplasia is the most common cause of disproportionate dwarfism and results from a mutation in the fibroblast growth factor receptor-3 (FGFR-3), leading to impaired endochondral bone formation. This condition is associated with characteristic craniofacial features, spinal abnormalities and altered body proportions, all of which have important anesthetic implications (Table 1), especially during pregnancy and childbirth(1,2). Pregnancy in women with achondroplasia is considered high risk due to CPD, increased likelihood of caesarean delivery, and anesthetic challenges related to airway management and neuraxial techniques(3).

| | Anatomical variation/ abnormality | Anesthetic implications |
|--------------------|---|--|
| Airway | <ol style="list-style-type: none"> 1. Limited neck extension 2. Foramen magnum stenosis 3. Prominent mandible 4. Macroglossia 5. Atlantoaxial instability 6. Narrow nasal passage | <ul style="list-style-type: none"> • Difficult mask ventilation • Difficult laryngoscopy • Difficult intubation • May be difficult supraglottic • Risk of trauma during nasal instrumentation |
| Venous access | Excessive subcutaneous tissue Lax skin | Difficult venous cannulation |
| CNS | Kyphoscoliosis Shortened Vertebral column Reduced CSF | <ul style="list-style-type: none"> • Technically difficulty in giving spinal and epidural • Unpredictable and patchy spread of LA • Excessive cephalad spread of LA; High Spinal or total spinal |
| Respiratory system | Thoracic dysplasia, rib hypoplasia, kyphoscoliosis | Restrictive lung disease which leads to dec Cardiopulmonary reserve |
| CVS | Congenital cardiac malformations, cardiomyopathies | Poor cardiopulmonary reserve led to rapid desaturation |

Table 1: Different abnormality associated with Achondroplasia and their anesthetic implications.

Even fundamental tasks such as securing peripheral venous access may be difficult due to lax skin and excess subcutaneous tissue which can be troubleshooted with USG guidance. Airway management is a major concern in achondroplastic parturient. Craniofacial abnormalities such as mid-face hypoplasia, macroglossia, tonsillar hypertrophy, limited mouth opening and potential cervical spine instability increase the risk of difficult mask ventilation and tracheal intubation (1,4). Additionally, narrowing of the foramen magnum and atlanto-axial instability necessitate cautious neck manipulation (4). In the present case, the presence of reduced mouth opening and grade III tonsillar hypertrophy warranted preparedness for a difficult airway. Normal physiological changes of pregnancy such as airway oedema, upper airway capillary engorgement and increased oxygen demand further complicate airway management.

Neuraxial anesthesia is preferred in achondroplastic parturient as it avoids airway manipulation and other risks associated with administering GA (4). However, neuraxial blocks may be technically challenging due to exaggerated lumbar lordosis, kyphoscoliosis, spinal canal stenosis, reduced interlaminar spaces and vertebral body distortion commonly seen in achondroplasia (5). Before regional anaesthesia, it is essential to document any pre-existing neurological deficits. USG is strongly recommended (Grade A recommendation with level of evidence 1a) (6) in these patients as it enhances the accuracy of landmark identification and depth of the epidural space. The reduced cerebrospinal fluid volume and shortened vertebral column increases the risk of excessive cephalad spread of intrathecal local anaesthetics (LA), potentially resulting in a high or total spinal block (4). An unpredictable spread of LA due to kyphoscoliosis may necessitate supplemental analgesia through an epidural (4).

Many achondroplasia patients also develop restrictive lung disease early in life due to thoracic dysplasia, rib hypoplasia and kyphoscoliosis, which can further progress to pulmonary hypertension or cor-pulmonale (5). Morbid obesity, (BMI of 40.7 Kg/m²) further compounded the anesthetic challenges in our parturient. To mitigate the risks of GA, a CSE technique was chosen, also a reduced dose of intrathecal hyperbaric bupivacaine was administered. Desired sensory level was subsequently achieved using incremental epidural top-ups.

Postoperative analgesia facilitates early mobilization and reduces respiratory and thromboembolic complications. Continuous epidural analgesia using low-concentration LA was provided to our patient, no motor blockade, allowing early ambulation and maternal comfort.

Research verifies that epidural depth measured by USG closely correspond to the actual depth (6). Obesity significantly increases insertion depth across all techniques which can be accurately identified by USG (9). However, a factor of discrepancy may appear because the Tuohy needle touches skin in its relaxed state while the USG probe compresses the subcutaneous tissue during pre-procedural imaging, typically underestimating the real needle depth by a small margin, 0.52 cm (9,10,11). Also, due to geometric differences, the actual needle insertion depth will be greater than the perpendicular distance measured by USG if the Tuohy needle is inserted at an angle greater by 5 mm (12). This discrepancy will worsen if conditions like kyphoscoliosis, pregnancy and obesity coexists like it did in our patient. We wish to highlight that the Tuohy needle was inserted at very acute angle and LOR (4.5 cm) was appreciated at greater depth than measured by USG (3.93 cm).

In a nutshell, this case emphasises the anaesthetic challenges that are encountered in a parturient with achondroplasia due to craniofacial abnormalities, difficult airway anatomy, altered spinal anatomy. Pre-procedural USG significantly improves identification of neuraxial landmarks and estimation of epidural depth; however, clinicians should be aware that the actual needle depth may differ because of tissue compression by the probe and angulation of needle. Thus, a meticulous

pre-anaesthetic assessment, preparedness for difficult airway management, judicious reduction of intrathecal drug dosage, and the complementary use of ultrasound guidance with clinical expertise is required to achieve safe maternal and neonatal outcomes in achondroplastic parturient.

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