Case Reports

Journal of INTERNATIONAL MEDICAL RESEARCH

Journal of International Medical Research
49(6) 1–8

DOI: 10.1177/03000605211023701
journals.sagepub.com/home/imr
SAGE

Real-time ultrasound-guided epidural anesthesia for cesarean section in a parturient with achondroplasia

Xueqin Cao, Wenchao Yang and Wei Mei

Abstract

Achondroplasia is a type of disproportionate dwarfism with short limbs and a normal-sized torso. This condition results in a potential spinal abnormality and a difficult airway may increase the anesthetic risk, not only in neuraxial anesthesia, but also in general anesthesia. We report a 25-year-old primigravida with achondroplasia who underwent cesarean section under epidural anesthesia with the assistance of real-time ultrasound guidance. A total dose of 17 mL 2% lidocaine with 7.5 μg sufentanil was administered via epidural catheter intermittently. The level of anesthesia reached T4. No other anesthetic was administered during the operation and the procedure was uneventful. The mother and her newborn were routinely discharged without any adverse events. During the follow-up at 10 months postoperatively, the patient did not have any discomfort. We suggest that titrated epidural anesthesia at the time of real-time ultrasound-guidance is a safe and effective epidural anesthesia for patients with achondroplasia.

Keywords

Achondroplasia, caesarean section, epidural anesthesia, real-time ultrasound, airway, pregnancy, lumbar lordosis

Date received: 5 January 2021; accepted: 18 May 2021

Introduction

Achondroplasia is a type of disproportionate dwarfism with short limbs, but a normal trunk. Achondroplasia is the most common type of dwarfism and occurs in 1:15,000 to

Department of Anesthesiology and Pain Medicine, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan, China

Corresponding author:
Wei Mei, Department of Anesthesiology and Pain Medicine, Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology, 1095 Jiefang Road, Wuhan 430030, China.
Email: wmei@hust.edu.cn

40,000 live births.[1] This is an autosomal dominant disorder. However, 80% of cases occur sporadically. Anesthetic management of achondroplastic dwarfs presents a unique challenge to anesthesiologists because of their unique anatomical syndromes. General and regional anesthesia can pose problems.[2] Pregnancy complications associated with dwarfism also include an increased risk of cardiopulmonary compromise, and an unpredictable anesthetic level with neuraxial techniques.[3] However, there is still no standard anesthesia for patients with achondroplasia. We report the anesthetic management of a parturient with achondroplasia who had cesarean section. We also searched the literature published in PubMed on anesthetic management of achondroplasia, and summarized the perioperative management to provide a comprehensive and safe guidance for surgery. The reporting of this study followed the CARE guidelines.[4]

Case report

A 25-year-old primigravida with achondroplasia planned to have emergency cesarean section at 37^{+4} gestational weeks owing to maternal tachycardia, hypertension, and cephalopelvic disproportion. The patient had a history of untreated hypertension (blood pressure: 142/97 mmHg) found at 14 weeks' gestation. She had no known concurrent medical problem and no history of previous surgery. There was no family history of any skeletal disorders.

The patient was 120 cm tall, which largely comprised her trunk and head, and she weighed 63 kg. She had typical achondroplastic features with a protruded forehead, prominent eyes, flat nose, and large tongue. She had a short neck, good neck extension, and adequate mouth opening (>3 transverse fingers). The Mallampatti assessment was I and the thyromental distance was 6 cm, which suggested a difficult airway. On a spinal examination, we found that she had mild lumbar lordosis and her lumbar intervertebral spaces were narrow (Figure 1). Her routine blood investigation, blood chemistry, and urinalysis results were unremarkable. A transthoracic echocardiographic examination was also normal. An electrocardiogram (ECG) showed sinus tachycardia. The results of fetal Doppler color ultrasound were normal. After discussion with obstetricians and neonatologists, cesarean section was decided to be the best choice.

In consideration of a potentially difficult airway and narrow lumbar intervertebral spaces, we planned to administer neuraxial anesthesia under the guidance of ultrasound. Routine monitors were placed. Her heart rate was 120 beats/minute via an ECG, non-invasive blood pressure was 148/87 mmHg, and pulse oximetry saturation was 96% in room air. After intravenous access was obtained, the patient was placed in the right lateral decubitus position. We evaluated the spinal canal through ultrasound because there were no related results of magnetic resonance imaging or computed tomography. Her spinal canal appeared normal under ultrasound. Subcutaneous 2% lidocaine was administered, and a 25-gauge spinal needle was inserted into the subarachnoid space at the L3–4 interspace via a 21-gauge syringe needle with the assistance of real-time ultrasound. However, no outflow of cerebrospinal fluid was observed, even after two adjustments. After discussion with obstetricians and the patient, epidural anesthesia with a backup plan of general anesthesia was chosen. A 17-gauge Tuohy needle (Yixinda Medical Equipment Technology Co., Shenzhen, China) was placed at the T12–L1 interspace under ultrasound guidance and an epidural catheter was threaded into the epidural space with ease. This process was uneventful. The patient was placed in the supine position. A volume of 3 mL

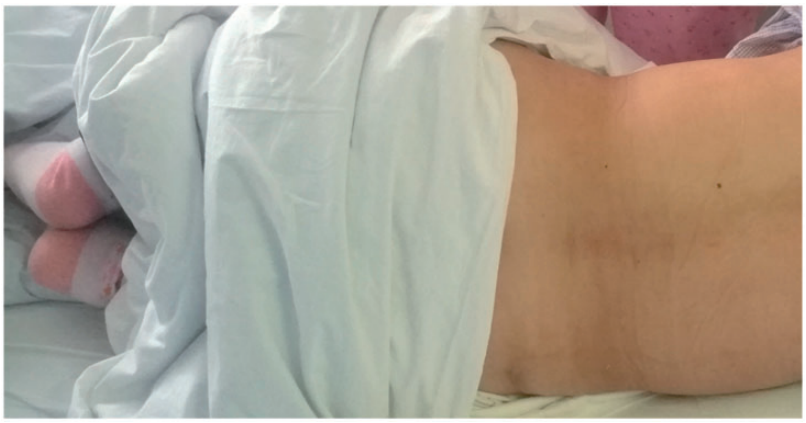

Figure 1. Photograph of the patient. She had a mild lumbar lordosis and her lumbar intervertebral spaces were narrow.

2% lidocaine was administered via the epidural catheter as a test dose. After 5 minutes, 2% lidocaine with 0.5 μg/mL sufentanil was administered via the epidural catheter intermittently over the next 15 minutes.[5] The anesthetic plane was measured by needle pricking intermittently until it reached the T4 level. The total dose of local anesthesia was 17 mL with approximately 7.5 μg sufentanil. Surgery was then started and a healthy neonate was delivered with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. No other anesthetic was administered during the operation and the procedure was uneventful. At the end of the operation, the epidural catheter was removed and intravenous patient-controlled intravenous analgesia was used for postoperative analgesia. The visual analog scale score was 2 points at rest and 3 points at movement on the second day postoperatively. The patient did not complain of paresthesia or muscle weakness of the lower limbs. The mother and her newborn were routinely discharged, without any adverse events. During the follow-up at 10 months postoperatively, the patient did not experience any discomfort.

Discussion

Achondroplasia is caused by fibroblast growth factor receptor 3 gene mutation, which disrupts bone formation and development, especially chondrogenesis and endochondral ossification. These patients have characteristic features, including typical facial features, rhizomelic shortening in the limbs, and a relatively normal trunk length. Severe lumbar deformity is common, which may lead to narrowing of the spinal canal from the upper to lower segments of the vertebral column.[6] Parturients with achondroplasia need to have cesarean section because of accompanying cephalopelvic disproportion.

The anesthetic management of parturients with achondroplasia presents a major challenge for anesthesiologists. The pathophysiological changes in pregnancy further increase the risk of managing anesthesia. Expansion of the circumference of the thorax during pregnancy is limited owing to rib deformities. Additionally, an enlarged uterus will affect the small thoracic cage, and severely decrease functional residual capacity. Cesarean section with either

general or regional anesthesia is associated with problems for this type of patient.

Regional anesthesia may be an attractive option for patients with achondroplasia because difficult intubation has been reported.[7] However, neuraxial anesthesia for patients with achondroplasia is also a large challenge. In adults with achondroplasia, lumbar distortion, poor landmarks, and spinal stenosis are common, and 38% of adult patients even have neurological consequences. These characteristic features contribute to increasing difficulty with neuraxial anesthesia.[7,8] In our patient, although she had mild lumbar lordosis with no neurological symptoms, there was difficulty in palpating her lumbar vertebral spaces. After checking intervertebral spaces using ultrasound, we initially attempted L3–4 for spinal anesthesia under real-time ultrasound guidance, but no cerebral spinal fluid was obtained.

We performed a review of the literature on anesthetic management of achondroplasia. Relevant research published in PubMed from inception to November 2019 was searched. The species was limited to humans and the language was limited to English. After reviewing the literature, we found that the majority of patients with achondroplasia had no cerebral spinal fluid outflow during attempted CSE combined spinal and epidural anesthesia or spinal anesthesia.[9] Dubiel et al. reported that magnetic resonance imaging showed a low-level conus medullaris terminating at mid-L3 in a patient with achondroplasia.[1] Therefore, spinal anesthesia or combined spinal-epidural anesthesia may not be an optimal anesthetic technique in these patients, and epidural anesthesia may be a better choice. However, repeated attempts could increase the risk of dural puncture. Moreover, narrowing of the epidural space may make insertion of an epidural catheter difficult, and lead to an unintentional intrathecal catheter. In consideration of these problems, we performed epidural puncture and catheterization at T12–L1 during real-time ultrasound-guidance. Several studies showed that ultrasound guidance increased the success rate of epidural puncture with the first attempt and decreased the rate of procedure-related complications.[10] To the best of our knowledge, we report the first case of an epidural catheter being inserted under real-time ultrasound guidance. Real-time ultrasound guidance can determine the needle insertion site and distance accurately under vision compared with the conventional landmark technique.[11]

Choosing the dosage of local anesthetics for neuraxial anesthesia in these patients can be difficult. Ten reports described parturients with achondroplasia who underwent cesarean section under neuraxial anesthesia (Table 1).[2,9,12–19] Three patients underwent cesarean section under epidural anesthesia, four underwent spinal anesthesia, and three were under combined spinal-epidural anesthesia. Most of these patients suffered from a spinal deformity. L3–4 was the most selected puncture point. There was no standard dose of local anesthetics in these reports. The doses for spinal anesthesia used in patients with achondroplasia are generally one third lower than those used in normal parturients.[3,9] Quantifying the optimal dosage of local anesthetics in patients with obesity and achondroplasia may be difficult. Sharma et al. found that the dose of spinal anesthesia in a patient with morbid obesity and achondroplasia was 50% that of normal.[8] A titrated epidural anesthetic dose has been proven to be safe.[20] In the present case, the total dose of epidural anesthetic was 17 mL, which was not less than that for a normal parturient. The reason for this similar dose may because the trunk in patients with achondroplasia is relatively normal. Overall, achondroplasia is the most common type of disproportionately short stature with

Table 1. Literature review of neuraxial anesthesia of parturients with achondroplasia.

Author/year	Age (years)	Height (cm)	Weight (kg)	Abnormal condition	Anesthetic technique	Puncture point	Anesthetics	Sensory block
Morrow and Black/1998[9]		120	64	Limited neck extension, difficult airway	Epidural anesthesia	L3–L4	12 mL 2% lidocaine with 1:200,000 epinephrine and 37.5 μg fentanil	Extended from S4 to T4 after 25 minutes
Ravenscroft and Rout/1999[12]	18	119	61	Marked lumbar lordosis and moderate thoracic kyphosis	Epidural anesthesia	T11/T12	8 mL 2% lidocaine with 1:200,000 epinephrine and 50 μg	Extended to T5
Waugaman et al./1986[13]	19	120	48	Lumbar spine normal with landmarks easily palpated	Epidural anesthesia	L2–L3	21 mL 0.75% bupivacaine	Extended from S4 to T4 after 25 minutes
Samra and Sharma/2010[14]	24	126	64	Lumbar lordosis	Spinal anesthesia	L3–L4	1.6 mL 0.5% hyperbaric bupivacaine	Extended to T4
DeRenzo et al./2005[15]	36	124	46.3	Spinal stenosis	Spinal anesthesia	L3–L4	10 mg 0.5% hyperbaric bupivacaine	Extended to T3
Mikhael et al./2011[16]	18	127	48.1	Obstructive sleep apnea syndrome, lumbar lordosis	Spinal anesthesia	L2–L3	11 mg 0.5% hyperbaric bupivacaine	Extended to T4
Palomero et al./2007[17]	37	127	51	Difficult airway, thoracolumbar kyphosis, lumbar lordosis	Spinal anesthesia	L3–L4	1.1 mL 0.5% hyperbaric bupivacaine	Extended to T2
Melekoglu et al./2017[18]	25	106	53	No severe kyphoscoliotic deformity	Combined spinal-epidural anesthesia	L3–L4	1.2 mL 0.5% hyperbaric bupivacaine with 10 μg fentanil	Not mentioned
Wight et al./2013[19]	30	130	52	Impalpable lumbar vertebrae	Combined spinal-epidural anesthesia under ultrasound-guidance	L3–L4	1.4 mL 0.5% hyperbaric bupivacaine with 300 μg morphine	Not mentioned
Li et al/2015[2]	27	120	63.5	Slight lumbar lordosis	Combined spinal-epidural anesthesia under ultrasound-guidance	L2–3	1.4 mL 0.5% hyperbaric bupivacaine	Extended to T4

pregnancy and can require dose adjustments.

Airway management of patients with achondroplasia can be risky when general anesthesia is chosen. Tracheal intubation may prove difficult because of the characteristic facial features, limited neck extension and cervical instability of patients with achondroplasia.[21] Therefore, these patients should be prepared with advanced airway equipment if a difficult airway is suggested by a physical examination. A smaller size of endotracheal tube should also be chosen because of the narrow nasopharynx in achondroplasia. The most appropriate size should be estimated on the basis of weight rather than age.[22] In consideration of a lower oxygen reserve in the mother and newborn, performing intubation via rapid sequence induction for parturients with dwarfism is not appropriate. Awake fiberoptic intubation may be a better choice in this situation. In our case, we avoided general anesthesia owing to the potential difficult intubation for this parturient. There have been four reported cases of parturients with achondroplasia who underwent cesarean section under general anesthesia (Table 2).[1,21,23] Two of these patients suffered from severe spinal scoliosis, and one of them even underwent cesarean section with spontaneous breathing because of a difficult airway. The third patient underwent cesarean section under general anesthesia after six failed attempts of puncture. The fourth patient suffered from a low terminal level of the conus at the mid-L3 level.

There are few data on the effect and pharmacokinetics of anesthetics in patients with dwarfism. There is only one review, which described that the requirement of propofol for these patients was not different from that of normal adults because the clearance rate of propofol in these patients did not change. To avoid overdose, measurement of the bispectral index or

Table 2. Literature review of general anesthesia in parturients with achondroplasia.

Author/year	Age (years)	Height (cm)	Weight (kg)	Abnormal condition	Anesthetic technique	Anesthetic induction
Mahbooheh Shirazi/2016[23]	26	98		Lumbar lordosis	General anesthesia	Rapid sequence intubation
Ekwere et al/2010[21]	22	129	55	Scoliosis involving the T8–S1 vertebrae, difficult airway	General anesthesia	Breathing spontaneously
Ekwere et al/2010[21]	25	120	42	Six failed attempts at L3/4 and L4/5	General anesthesia	Rapid sequence intubation
Dubiel et al/2014[1]	20	99	30	A low lying conus medullaris terminating at the mid-L3 level	General anesthesia	Modified rapid sequence intubation

electroencephalographic monitoring during general anesthesia for patients with dwarfism should be performed.[24]

Considering the high risk of these patients during delivery, multidisciplinary consultation in the preoperative and perioperative periods should be emphasized. Initially, comprehensive pre-pregnancy examinations and regular prenatal check-ups should be encouraged. Moreover, a cardiovascular specialist and anesthesiologist should be consulted early during pregnancy. The focus of the examination should be on the patient's airway, cardiac and pulmonary function, and the spine. An electronic laryngofiberscopic examination and a magnetic resonance imaging scan of the spine should be considered before the operation.

In conclusion, titrated epidural anesthesia with the assistance of real-time ultrasound-guidance can offer safe and effective epidural anesthesia for parturients with achondroplasia.

Ethics approval and consent to participate

This case report was approved by the Research and Ethics Committee of Tongji Hospital, Tongji Medical College, Huazhong University of Science and Technology (approval number TJ-IRB20200718). Written informed consent was obtained from this patient for publication of this case report and any accompanying images.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

ORCID iD

Wei Mei https://orcid.org/0000-0001-6556-6628

References

1. Dubiel L, Scott GA, Agaram R, et al. Achondroplasia: anaesthetic challenges for caesarean section. *Int J Obstet Anesth* 2014; 23: 274–278.
2. Li X, Duan H and Zuo M. Case report: Anesthesia management for emergency cesarean section in a patient with dwarfism. *BMC Anesthesiol* 2015; 15: 59.
3. Bakhshi RG and Jagtap SR. Combined spinal epidural anesthesia in achondroplastic dwarf for femur surgery. *Clin Pract* 2011; 1: e120.
4. Gagnier JJ, Kienle G, Altman DG, et al. The CARE guidelines: consensus-based clinical case reporting guideline development. *Headache* 2013; 53: 1541–1547.
5. Cheng Q, Bi X, Zhang W, et al. Dexmedetomidine versus sufentanil with high- or low-concentration ropivacaine for labor epidural analgesia: A randomized trial. *J Obstet Gynaecol Res* 2019; 45: 2193–2201.
6. Daugherty A. Achondroplasia: Etiology, Clinical Presentation, and Management. *Neonatal Netw* 2017; 36: 337–342.
7. Burgoyne LL, Laningham F, Zero JT, et al. Unsuccessful lumbar puncture in a paediatric patient with achondroplasia. *Anaesth Intensive Care* 2007; 35: 780–783.
8. Sharma R, Magoon R, Choudhary R, et al. Anaesthesia for emergency caesarean section in a morbidly obese achondroplastic patient with PIH: Feasibility of neuraxial anaesthesia? *Indian J Anaesth* 2017; 61: 77–78.
9. Morrow MJ and Black IH. Epidural anaesthesia for caesarean section in an achondroplastic dwarf. *Br J Anaesth* 1998; 81: 619–621.
10. Lacharite-Roberge AS, Raza F, Bashir R, et al. Case series of seven women with uterine fibroids associated with venous thromboembolism and chronic thromboembolic disease. *Pulm Circ* 2019; 9: 2045894018803873.
11. Weiniger CF and Sharoni L. The use of ultrasound in obstetric anesthesia. *Curr Opin Anaesthesiol* 2017; 30: 306–312.
12. Ravenscroft A and Rout C. Epidural anaesthesia for caesarean section in an achondroplastic dwarf. *Br J Anaesth* 1999; 82: 301–303.
13. Waugaman WR, Kryc JJ, Andrews MJ. Epidural anesthesia for cesarean section

and tubal ligation in an achondroplastic dwarf. *AANA J* 1986; 54: 436–437.
14. Samra T and Sharma S. Estimation of the dose of hyperbaric bupivacaine for spinal anaesthesia for emergency caesarean section in an achondroplastic dwarf. *Indian J Anaesth* 2010; 54: 481–482.
15. DeRenzo JS, Vallejo MC, Ramanathan S. Failed regional anesthesia with reduced spinal bupivacaine dosage in a parturient with achondroplasia presenting for urgent cesarean section. *Int J Obstet Anesth* 2005; 14: 175–178.
16. Mikhael H, Vadivelu N, Braveman F. Safety of spinal anesthesia in a patient with achondroplasia for cesarean section. *Curr Drug Saf* 2011; 6: 130–131.
17. Palomero MA, Vargas MC, Pelaez EM, et al. Spinal anaesthesia for emergency caesarean section in an achondroplastic patient. *Eur J Anaesthesiol* 2007; 24: 981–982.
18. Melekoglu R, Celik E, Eraslan S. Successful obstetric and anaesthetic management of a pregnant woman with achondroplasia. *BMJ Case Rep* 2017; 2017.
19. Wight JM, Male D, Combeer A. Ultrasound-guided combined spinal-epidural anaesthesia for elective caesarean section in a patient with achondroplasia. *Int J Obstet Anesth* 2013; 22: 168–169.
20. Vance CE, Desmond M, Robinson A, et al. Pregnancy in a woman with proportionate (primordial) dwarfism: a case report and literature review. *Obstet Med* 2012; 5: 124–129.
21. Ekwere IT, Edomwonyi NP and Imarengiaye CO. Anaesthetic challenges associated with achondroplasia: a case report. *Afr J Reprod Health* 2010; 14: 149–155.
22. Daugherty RJ, Nadkarni V and Brenn BR. Endotracheal tube size estimation for children with pathological short stature. *Pediatr Emerg Care* 2006; 22: 710–717.
23. Shirazi MF, Golshahi F, Teimoory N. Successful delivery in a woman with achondroplasia: A Case report. *Acta Med Iran* 2017; 55: 536–537.
24. Tsubokawa T, Yamamoto K, Komuro A, et al. Propofol pharmacokinetics in a dwarfism patient. *Acta Anaesthesiol Scand* 2003; 47: 488–490.