Anesthetic Management for Emergency Cesarean Delivery in Parturient with Achondroplasia – A Case Report and Review of the Literature

Abstract

Achondroplasia is a genetic disorder where there is underdevelopment and shortening of the long bones formed by endochondral ossification without abnormalities of cartilage formation. Patients with achondroplasia can exhibit various manifestations: spinal abnormalities; thoraco-lumbar kyphoscoliosis, respiratory and cardiac abnormalities; obstructive sleep apnea; and changes of the head and the midface. In women with achondroplasia, cesarean delivery is inevitable because the pelvis is invariably small. Anesthetic management for cesarean section in this population is controversial; the choice of anesthetic technique depends on the benefits and risks of each technique. Emergency cesarean is another factor to consider for the choice of anesthetic technique. We report the case of a 34-year-old Moroccan primipara with achondroplasia who had a cesarean delivery under Spinal anesthesia. Other possible anesthetic techniques used for cesarean sections in such patients are also discussed.

Keywords: Achondroplasia, anesthesia, cesarean section, spinal anesthesia

Introduction

Achondroplasia is a genetic disorder with an autosomal dominant inheritance. About 250,000 individuals are affected worldwide. The clinical manifestation of the mutation in this disorder is an underdevelopment and shortening of the long bones of the extremities formed by endochondral ossification; in fact, the cartilage formation is unaffected but the cartilage is converted to bone. Patients with achondroplasia have characteristic anatomical features including disproportionate short stature, trident-shaped hands, a relatively large head with frontal bossing, midface hypoplasia, muscular hypotonia during infancy, genu varum, and spinal abnormalities.[1] In parturients with achondroplasia, the maternal pelvis is small due to the described skeletal abnormalities, which results in cephalopelvic disproportion and thus makes the cesarean delivery necessary.

These parturients pose great challenge to the anesthesiologist: head abnormalities, spinal abnormalities with direct effect on the thoracic cage, and respiratory consequences with potential problems of airway management during general anesthesia. In addition to this, respiratory function is even more compromised by physiological changes in late pregnancy: rapid desaturation due to reduced functional residual capacity of the lungs, risk of aspiration, etc. Therefore, parturients with achondroplasia are high-risk candidates for general anesthesia. However, neuroaxial anesthesia has a high risk of failure and complications as well.

Anesthetic management of such patients is complicated because of their multisystem affection. Several observations have been reported in the literature describing different anesthetic techniques in achondroplastic parturients: general anesthesia,[2-4] spinal anesthesia,[5,8] epidural anesthesia alone[8,9] or in combination with spinal anesthesia,[10,11] and continuous spinal anesthesia.[12]

We report a successful anesthetic management of a parturient with achondroplasia for emergency cesarean under spinal anesthesia.

Case Report

A 34-year-old, gravid 1/para 0, achondroplastic dwarf parturient was admitted in labor at 37 weeks and 3 days of gestation; an elective cesarean had

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been planned at 38 weeks in virtue of cephalopelvic disproportion. In her past medical history, there were no neurological, respiratory, or musculoskeletal known disorders. Physical examination demonstrated a patient who was 130-cm tall and weighed 54 kg (body mass index 31.95 kg/m²). She had short extremities with a normal-sized face, thoracic kyphoscoliosis, and lumbar hyperlordosis. Her blood pressure was 113/63 mmHg and her heart rate was 95 beats/min. Chest examination revealed normal heart sounds and good bilateral air entry. On airway assessment, she had a Mallampati 2 score, with a thyromental distance <6 cm, a full range of neck movement, and a normal jaw protrusion.

Obstetric examination found a parturient in labor, cervix dilated at 3 cm, obstetric ultrasound was normal. However, cardiotocography showed signs of severe fetal distress; thus, cesarean section was indicated for immediate delivery of the compromised fetus. Given the urgent indication of cesarean and the absence of neurological and cardiovascular abnormalities, we opted for a spinal anesthetic as our anesthetic of choice. Laboratory investigations were normal including the coagulation tests.

The patient was admitted to the operating room where basic monitoring was instated including electrocardiogram, noninvasive blood pressure, and pulse oximetry. Her basal blood pressure was 128/72 mmHg, her heart rate was 105 beats/min and her oxygen saturation was 98%. She was given 10 mg of Metoclopramide, 50 mg of Ranitidine, and 2 g of Cefazolin intravenously, an infusion of 500-mL isotonic saline was started. The patient was positioned in a sitting position, and a spinal anesthetic was given at the level of L3-L4 interspace; 7.5-mg intrathecal hyperbaric bupivacaine 0.5% with 20 µg of Fentanyl, and 80 µg of morphine were administered. The spinal anesthetic was successful after a single attempt using a 25-ga pencil point spinal needle via a spinal introducer.

The parturient was immediately placed supine and the operating table given a 15° left lateral tilt, she had a urinary catheter inserted and the block height was assessed; a sensory level corresponding to T4 was achieved. Patient was started on 3 L/min oxygen via a nasal cannulae.

Intraoperatively, except one episode of hypotension at 85/50 mmHg that was treated with 6 mg of ephedrine along with 300 mL of isotonic saline, blood pressure (BP) and heart rate (HR) remained stable throughout surgery (BP varied between 91–135 and 60–85 mmHg and HR between 75 and 90 bpm). Six minutes later, the patient delivered a live male baby with an Apgar scores of 8 at 1 min and 10 at 5 min; 5 units of oxytocin were given intravenously; and then an infusion of 10 units/h was started. Patient received 750 mL of isotonic saline during surgery; the estimated blood loss was 500 mL.

Postoperatively, the patient was monitored in the recovery room until the motor block had completely worn off. The parturient remained well on follow-up visit and was discharged 5 days later.

Discussion

In achondroplastic pregnant women, the most common mode of delivery is elective cesarean section. Spinal anesthesia with a lower dose of local anesthetic can be used safely in emergency cesarean section in absence of absolute neurological and cardiovascular contraindications.

Achondroplasia is the most common form of dwarfism; a genetic disorder characterized by underdevelopment and shortening of the long bones formed by endochondral ossification; the articular cartilage formation is unaffected.[1] Patients with achondroplasia have several anatomical and physiological changes that can present challenges to the anesthesiologist.[13] Superimposed physiological changes during pregnancy can considerably complicate the management of labor.

Fertility in this population is normal,[4] but cephalopelvic disproportion due to small and contracted pelvis in these women make cesarean section inevitable.[13] Anesthetic management of cesarean section in achondroplastic parturients is controversial with an ongoing debate regarding general versus regional anesthesia in these patients. Many cases have been reported in literature describing variable techniques: general anesthesia, spinal anesthesia, epidural anesthesia or combined spinal epidural anesthesia, and continuous spinal anesthesia.

Achondroplasia is characterized by morphological and multisystemic abnormalities that have anesthetic implications in gravid women candidate for cesarean section: craniofacial modifications; large head, saddle nose, macroglossia, forehead protrusion, short maxilla, large mandible, atlantoaxial instability. Airway management can be challenging because of a big head, limited neck extension, narrow nasal passages and nasopharynx, and small trachea imposing the use of small tracheal tube.[1,4,13] Logistic factors should be considered prior to anesthesia; cephalopelvic disproportion being frequent and cesarean section, the way of delivery, a preoperative assessment of comorbidities should always happen before the date of surgery. Compatible anesthetic equipment should be inspected and operational; facemask and blood pressure cuff of an appropriate size should be available. In addition, many factors are to consider for patient positioning; neck hyperextension should be avoided because of the cervical instability, the reduced functional residual capacity during pregnancy is exaggerated by thoracolumbar kyphoscoliosis and the risks of flexion contractures of the elbow and radial head subluxation require careful positioning.

Patients with achondroplasia may develop multiple neurological symptoms: hydrocephalus due to changes of the bones at the skull base and a small craniocervical junction, imposing a cerebrospinal shunt in case of

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hydrocephalus. In another side, foramen magnum stenosis can be responsible for brainstem compression. In another hand, thoracolumbar kyphosis with indication for surgery, mostly unoperated in our context. Spinal stenosis due to vertebral anatomical abnormalities such as shortened vertebral bodies, shortened pedicles, reduced interpedicular distance in the lumbar spine, and thickened intervertebral disks and ligamentum flavum. All these modifications lead to the reduction of the sagittal and coronal diameters of the spinal canal which can sometimes cause nerve compression requiring surgical decompression.[1]

Vertebral bone abnormalities are frequent in achondroplastic patients; thus, technical difficulty can be encountered during spinal anesthetic owing to reasons such as lumbar lordosis, difficult to find landmarks, unpredictable spread of local anesthetic due to spinal stenosis, and nerve compression.[1,4,13] In addition to these factors, parturients have narrowed epidural and intrathecal spaces with expanded epidural veins.[6]

Respiratory difficulties and breathing disorders are common in achondroplasia; they include restrictive lung diseases with decreased functional residual capacity due to scoliosis and pregnancy. Obstructive sleep apnea is another cause of morbidity and is due to a cervicomedullary cord compression rather than a central cause; it can also be the result of the midface hypoplasia with relative adenotonsillar hypertrophy or upper airway obstruction.[1,4,6,13] Moreover, scoliosis along with chronic airway obstruction and sleep apnea may lead to a cor pulmonale with pulmonary hypertension. Anesthetic management is based on general principles of avoiding factors that increase pulmonary pressures, such as hypoxia, hypercarbia, acidosis, nitrous oxide, and stress.[4,13] Cardiac and respiratory abnormalities make airway and administration of general anesthesia challenging. For all these systemic abnormalities, anesthetic management of achondroplastic parturients for cesarean section is challenging and the choice of the anesthetic technique remains an ongoing debate.

Spinal anesthesia has been reported as a technique used for emergency cesarean section in achondroplastic parturient with successful outcome[5,7,9] and failure of spinal anesthesia in such patients has also been reported.[6] Some authors have used epidural anesthesia alone[8-10] or combined with spinal anesthesia[11,12] or continuous spinal anesthesia[14] for a more rational use of anesthetic drugs. Epidural anesthesia alone or combined with spinal anesthesia or continuous spinal anesthesia offer the possibility of titration of anesthesia and the block level. However, the catheter insertion takes usually longer due to anatomic abnormalities (spinal stenosis, lumbar hyperlordosis, and thoracolumbar kyphosis), which can increase the Decision Delivery Interval in emergency cesarean. To improve the safety and the quality of the anesthetic technique, some authors suggested the use of ultrasound guidance to overcome the difficulty of epidural catheter insertion.[12]

Neuraxial anesthesia was previously described for cesarean section in achondroplastic parturients, although it has relative merits over general anesthesia, but the neuraxial anesthesia has significant technical challenges due to short stature and spinal abnormalities like difficulty to insert catheters and the unpredictable block spread. Lange et al.[15] reported a series of 15 cesarean sections in parturients with dwarfism, 8 of them were achondroplastic parturients, all were operated under neuraxial anesthesia; combined spinal-epidural (CSEs), epidural, and spinal anesthesia. CSE was practiced in four elective cesarean sections; they reported complications such as technical difficulty, multiple attempts for needle placement, and paresthesia; only one patient did not have any complication and one patient required a conversion to general anesthesia for desaturation. In three parturients operated under epidural anesthesia only one was converted to general anesthesia due to inadequate anesthetic block. Spinal anesthesia was performed in one patient; it required multiple attempts, 8.25 mg of hyperbaric bupivacaine 0.5% with 25 µg of fentanyl and 100 µg of morphine were injected. Authors conclude that dwarf parturients require lower doses of local anesthetics in both spinal and epidural anesthesia to achieve adequate surgical blockade.[15] To our Knowledge, pharmacological data of local anesthetic and opioids doses in patients with achondroplasia has never been published. Qiu et al.[16] conducted a meta-analysis including 11 randomized controlled trials with 605 patients; they concluded that low dose bupivacaine combined with opioids (LBO) regimen was the preferred drug regimen of bupivacaine in cesarean section for reducing the risk of intraoperative hypotension and offering reliable analgesia compared to low-dose and high-dose bupivacaine. The fentanyl doses used in this meta-analysis varied between 20 and 25 µg[16]; the recommended dose of morphine for cesarean section in the literature is 100 µg, but lower doses were used combined with hyperbaric bupivacaine.[17]

For our patient, because of the need to perform an emergency cesarean section for fetal distress, in absence of neurological abnormalities (symptoms of nerve compression, cranial nerve dysfunction, difficulty of swallowing), respiratory symptoms and cardiac abnormalities and given the risks of general anesthetic during pregnancy, successful spinal anesthesia was performed using low-dose hyperbaric bupivacaine 0.5% (7.5 mg) and opioids (20 µg of fentanyl and 80 µg of morphine).  

**Conclusion**

Spinal anesthesia for cesarean section in achondroplastic parturients is a promising technique in absence of contraindications. Anesthetic assessment must be exhaustive to assess all comorbidities associated with achondroplasia. The use of ultrasound guidance may facilitate and reduce risks associated to puncture.
Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent. In the form, the patient has given her consent for clinical information to be reported in the journal but her image. The patient understands that her name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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