Anesthesia for Cesarean Delivery in an Achondroplastic Dwarf: A Case Report

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There are more than 100 different types of dwarfism. Achondroplasia is the most common form of this rare condition. The incidence of achondroplasia in the United States is about 15 per 1 million births. Although inherited as an autosomal dominant condition, 80% of cases result from spontaneous mutation. Underdevelopment and premature ossification of bones result in characteristic craniofacial and spinal abnormalities.

Limited neck extension, foramen magnum stenosis, a large tongue, large mandible, and atlanto-axial instability can lead to increased difficulty of airway management. Severe kyphosis, scoliosis, spinal stenosis, and unpredictable spread of local anesthetics in the epidural space and subarachnoid space lead to reluctance to apply regional anesthesia in this patient group. In addition, pregnancy in a person with achondroplasia poses more problems for anesthetic selection. These problems include potential hypoxia, severely decreased functional residual capacity, risk of gastric aspiration, and supine hypotension.

In this case report, we describe the anesthetic management of an achondroplastic dwarf who underwent cesarean delivery.

Keywords: Achondroplastic dwarf, difficult airway, general anesthesia, regional anesthesia.

A healthy 26-year-old achondroplastic dwarf was admitted to the hospital for an elective cesarean delivery following an uneventful pregnancy. The patient’s surgical history included multiple oral facial surgical procedures for cosmetic reasons and low limb operations for limb elongations under general anesthesia without anesthetic complications. Anesthesia records of these procedures were not available. The patient was allergic to penicillin and aspirin. She was 110 cm tall and weighed 62 kg. Physical examination revealed a small face, short neck, a Mallampati class III airway with full range of motion of her neck. She had a marked lumbar lordosis, impalpable lower lumbar vertebrae, and prominent lower back musculature. She was unable to eliminate the lumbar lordosis either in the lateral or sitting position. Because of maternal contracted pelvis and breech fetal presentation, the decision was made for a cesarean delivery.

The patient expressed a desire to be awake for birth. She had no preexisting neurological problems; therefore it was decided to perform the cesarean delivery under epidural anesthesia. The patient was made aware that a general anesthesia may be required in the event of failed epidural anesthesia.

In the operating room, the patient was placed in the sitting position and premedicated with oral sodium citrate, 30 mL. Using a sterile technique, her back was prepared with iodine. Using an 18-gauge Tuohy needle, epidural insertion was attempted at the L4 to L5 interspace. It was difficult to reach the epidural space. The patient refused to have another attempt at a different interspace. She requested general anesthesia.

It was decided to attempt awake oral intubation. Cetacaine spray, 2%, was applied to anesthetize the airway. A GlideScope video laryngoscope (Verathon, Inc, Bothell, Washington) was advanced into the airway; however, even the rearmost portion of the vocal cords could not be viewed. The GlideScope was removed, and then a flexible fiberoptic bronchoscope was used. The fiberoptic scope was then advanced under direct vision into the trachea through the glottic opening. Then a 6.0-mm endotracheal tube was advanced into the trachea over the bronchoscope. The fiberoptic scope was removed and the endotracheal tube was secured. The anesthesia circuit was attached to the endotracheal tube and carbon dioxide was detected by capnography. General anesthesia was induced with ketamine and rocuronium. Anesthesia was maintained with oxygen and sevoflurane. Left uterine displacement was applied. Cesarean delivery was performed without complications. A 2,373-g baby girl with APGAR scores of 4 at 1 minute and 6 at 5 minutes was delivered. The patient had no complications related to her delivery or anesthesia.

Discussion

Both general and regional anesthesia can pose problems for the gravid achondroplastic dwarf. The anesthetists must consider the advantages and disadvantages of each technique for patients on an individual basis.

Regional anesthesia may be technically difficult because of poor landmarks. Thoracic kyphoscoliosis,
severe lumbar lordosis, reduced interpedicular distance, shortening of pedicles, and osteophyte formation make identification of bony landmarks difficult. Under-development of the vertebral arch leads to narrowing of the subarachnoid and epidural space. A narrow epidural space may make epidural catheter insertion difficult and dural puncture more likely and may also limit spread of local anesthetics. Engorged epidural veins related to pregnancy increase the risk of venous puncture. Aortocaval compression may be severe because of the intra-abdominal position of the uterus, causing significant decompensation with the onset of central neural blockade. Recognition of a subarachnoid tap may be more difficult because free flow of cerebrospinal fluid may be difficult to obtain.

Epidual anesthesia is preferable to spinal anesthesia because it allows titration of the dose of local anesthetic according to the designed height of the block. Regional anesthesia (epidural and spinal) has been used successfully in dwarfs for cesarean delivery. Because our patient expressed a desire to be awake for birth and she had no preexisting neurological problems, an epidural anesthesia was chosen. However, technical difficulties forced us to abandon regional anesthesia.

Difficulties associated with intubation in achondroplastic dwarfs have been reported. Facial features of achondroplastic dwarfs include a large protruding forehead, short maxilla, large mandible, and large tongue, which contribute to difficulties in airway management. Hyperextension of the neck should be avoided during tracheal intubation to prevent cervical spinal cord compression, because foramen magnum stenosis is common. Cardiorespiratory-tory function may be reduced by a narrow rib cage and a variable degree of kyphoscoliosis. The addition of a totally abdominal uterus compressed within the short distance between symphysis pubis and xiphoid can compromise cardiorespiratory reserve. A low functional residual capacity reduces the safety margin provided by preoxygenation during induction. High abdominal pressure may put the mother at greater risk of aspiration. Our patient had multiple surgical procedures in her childhood without anesthesia problems, but the subsequent development of her head and neck might have made tracheal intubation difficult. Therefore, awake intubation was selected.

Management of the difficult airway remains one of the most relevant and challenging tasks for anesthesia providers. Many new airway management devices have been used. The Glidescope has been shown to be useful in patients with difficult airways, and it has several advantages: superb laryngeal visualization, similarity in technique to direct laryngoscopy making this technique very accessible, and ease of endotracheal intubation. Using the Glidescope for awake intubation has not been reported previously in the literature. The Glidescope may not be the right tool for awake intubation. The fiberoptic scope is generally accepted as the most reliable approach to difficult airway management. The indications of fiberoptic scope in this case included anticipated difficult intubation from physical examination, unstable cervical spine disease, and the need for awake intubation. Because of decreased cardiorespiratory reserve and supine hypotension in the gravid achondroplastic dwarf, general anesthesia was induced with ketamine and rocuronium.

Conclusion

The management of anesthesia for a gravid achondroplastic dwarf for cesarean delivery presents a significant challenge. Regional anesthesia can be successfully administered. Technically difficult problems are associated with regional anesthesia and unsuccessful attempts can be encountered. The abnormalities of the airway may make tracheal intubation difficult, especially in the presence of pregnancy-induced change in the upper airway. Awake fiberoptic intubation is an option when a difficult airway is anticipated. Adequate preparation and planning are fundamental for successful management of anesthesia for a gravid achondroplastic dwarf undergoing cesarean delivery.

REFERENCES


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