



Anaesthetic Management of Emergency Caesarean Section in an Achondroplastic Dwarf

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Abstract: There are more than 100 different types of dwarfism. Achondroplasia is the most common form of this rare condition. 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 difficulty in airway management. Severe kyphosis, scoliosis, spinal stenosis, and unpredictable spread of local anaesthetics in the epidural space and subarachnoid space may cause regional anaesthesia a difficult choice in this patient group. In addition, pregnancy in a person with achondroplasia poses many more problems for anaesthetic selection including risk of hypoxia, severely decreased functional residual capacity, risk of gastric aspiration, and supine hypotension. In this case report, we describe the anaesthetic management of an achondroplastic dwarf who successfully underwent caesarean delivery under general anaesthesia.

Keywords: Achondroplasia; caesarean section; general anaesthesia; spinal; epidural.

Introduction: Dwarfism is defined as failure to achieve height of 148 cm by adulthood.¹ There are more than 100 different types of dwarfism. Achondroplasia is most common form of this rare condition.² The term "achondroplasia" was proposed by Jules Parrott in 1878 to describe a condition of children and adults with disproportionate short stature.

Achondroplasia is caused by the mutation of the fibroblast growth factor receptor 3 gene (FGFR3).³ This mutation results in an inhibition of cartilage proliferation and a disorder of endochondral ossification. As a consequence, premature ossification of epiphyseal cartilage is observed. Achondroplastic patients have peculiar anatomical features like large head, saddle nose, short limbs, foramen magnum stenosis, limited neck extension and atlanto-axial instability. Other anomalies include an exaggerated lumbar lordosis, thoracic kyphoscoliosis, generalized spinal stenosis, and vertebral deformities. These features make the administration of anaesthesia in this

group of patients quite challenging. The physiologic and hormonal changes of pregnancy further complicate anaesthetic administration. Pregnant patient in last trimester have severely decreased functional residual capacity, increased risk of acid aspiration and of supine hypotension. We describe the anaesthetic management of an emergency caesarean section in an achondroplastic dwarf and discuss the inherent anaesthetic considerations in management of such patients.

Case Report: A 27-year-old primigravid achondroplastic dwarf at 37-weeks of pregnancy was admitted for an emergency Caesarean section for cephalopelvic disproportion with foetal distress. She was an unbooked patient. Patient came to our hospital for the first time when she was in labor. On examination, she was found to have foetal distress, so she was shifted to operating room for emergency caesarean section. Her previous medical history was unremarkable. Physical examination revealed a 35-kg, 104 cm and normal intelligent female with large head, short limbs and mild kyphoscoliosis. She had short neck, protruded chin and full set of teeth (Figure 1& 2).



Figure:1



Figure:2



Mouth opening was adequate with Mallampati II airway and neck extension was not limited. She had taken biscuits and half glass of orange juice about three hours before shifting to operation theatre. She received premedication of ranitidine 50 mg and metoclopramide 10 mg intravenously preoperatively. She was anxious and wanted to be asleep during procedure. So after discussion of potential risks and advantages, general anaesthesia was planned for her and written informed consent was taken.

On arrival in the operating room standard monitoring (ECG, SpO₂, EtCO₂ and NIBP) was instituted. Wedge was placed under her right hip and rapid sequence induction was done with intravenous thiopental 150 mg and neuromuscular blockade was achieved with succinylcholine 50 mg. Trachea was intubated at the first attempt with a 6.5mm ID endotracheal tube. Anaesthesia was maintained with halothane in 50% oxygen-nitrous oxide mixture. Muscle relaxation was achieved with vecuronium. Patient was given oxytocin and fentanyl after the umbilical cord was clamped. Lower segment caesarean section proceeded and a live baby was delivered weighing 3000 grams with APGAR scores of 7 and 9 at the first and the fifth minutes respectively. The total duration of anaesthesia was one hour and all monitored parameters remained stable. At the end of the operation, neuromuscular blockade was reversed with neostigmine 2 mg with glycopyrrolate 0.4 mg. Trachea was extubated when patient was fully awake and ventilatory efforts were adequate. The patient was shifted to post anaesthesia care unit and administered 4 L min⁻¹ of oxygen through a face mask. The postoperative period was uneventful and the patient and neonate were discharged on the fifth day.

Discussion: There are more than hundred different types of dwarfism. People with severe short statures can be divided into two categories:

1. Midgets, who have proportionate growth and normal ratio of trunk to limb length.
2. Dwarfs, having short stature and disproportionate development characterized either by short limbs, or short trunks.

Achondroplasia is the most common type of disproportionate short stature with an incidence of 0.5–1.5/10 000 live births.⁴ Eighty percent of all "little people" have achondroplasia. Spontaneous mutation accounts for 80% of cases, while the remaining 20% of cases are of autosomal dominant inheritance.⁴ Females are affected more frequently than males.⁵ The basic defect is thought to be a quantitative decrease in the rate of endochondral ossification, and this, coupled with normal periosteal bone formation, leads to the shorter tubular bones present in achondroplastic dwarfs. These patients have a number of anatomic and physiological abnormalities that contribute to problems with the administration of anaesthesia. Careful preoperative anaesthetic evaluation aims to document preexisting neurological deficits and co-morbidities to minimize the associated complications.

Achondroplastic dwarfs characteristically have low fertility rates, however when they do conceive, they often require delivery by caesarean section because the normal sized foetal head and smaller than normal maternal pelvic diameter result in cephalo-pelvic disproportion during the later stages of pregnancy.⁶ This leads to further problems as the uterus remains an entirely intra-



abdominal organ. By the 16th week, these patients appear to be in the 30th week of pregnancy.¹ Subsequent diaphragmatic splinting causes a greater than usual pregnancy induced reduction in functional residual capacity and there is severe aortocaval compression.⁷

Cardio-respiratory functions may be impaired by several factors specific to achondroplasia. Patient may suffer from mild restrictive lung disease from rib hypoplasia, thoracic lordosis and cor pulmonale from restrictive lung disease. Neurogenic effects of brainstem compression lead to reduced vital capacity, which leads to higher incidence of pneumonia, cyanotic spells and apnoea. Long term, patients with achondroplasia are prone to obstructive and central sleep apnoea and pulmonary complications.^{6,8}

Achondroplastic patients have facial features that alert the anaesthetist to potential problems in airway management.⁹ Short stature, enlarged head, saddle nose, maxillary hypoplasia, mandibular enlargement, megaloccephaly with protuberant forehead and narrow nasal passages are some features that contribute to airway difficulties in these patients.^{10,11} Hyperextension of patient's neck during laryngoscopy should be avoided considering the likely presence of foramen magnum stenosis. Despite these characteristics, clinical experience has not confirmed any difficulty with maintaining an upper airway or achieving tracheal intubation in most of these patients.¹¹ Weight rather than age is the best guide for predicting the proper size of the endotracheal tube.¹¹

Several anatomical abnormalities found in achondroplasia may complicate regional techniques. Vertebral deformities, shortening of pedicles, decreased interpedicular distance and osteophyte formation make regional anaesthesia difficult in these cases.¹² Under-development of the vertebral arch leads to narrowing of the subarachnoid and epidural space.⁷ A narrow epidural space makes dural puncture more likely, increases the difficulty in epidural catheter insertion and also limits the spread of local anaesthetics.¹³ Engorged epidural veins increase the risk of venous puncture either by the Tuohy needle or catheter, and result in unpredictable level of spread of local anaesthetic within the space. Spinal stenosis may impair free flow of CSF and make identification of dural puncture more difficult.¹⁴ There are no guidelines on use of epidural or spinal local anaesthetics in achondroplasia. Because of large inter individual variation in spinal column anatomy in these patients; it is difficult to predict appropriate doses of local anaesthetic.

There are several reports of parturient with achondroplasia undergoing caesarean section successfully under both general anaesthesia^{15,16} as well as under central neuraxial blockade.^{14,17,18} We opted for general anaesthesia in our patient for various reasons. First of all, our patient wanted to be asleep during procedure. Moreover, preoperative examination suggested no significant abnormality in airway. Further, there is no data confirming appropriate doses of local anaesthetic for spinal anaesthesia. Any given doses of local anaesthetic may result in either a high level of anaesthesia or inadequate level of spinal block due to unpredictable spread of local anaesthetic. Both these conditions are undesirable and potentially harmful. Epidural anaesthesia has been successfully used in many cases as it permits titration of local anaesthetic doses to achieve desired sensory level of block. We avoided epidural anaesthesia due to the urgency demanded by the situation.



Conclusion: General as well as regional anaesthesia, both pose potential hazards in patient with achondroplasia and choice should be individualized to each patient. A complete history, physical examination and development of a safe anaesthesia plan tailored to individual needs can help reduce the risk.

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