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CASE REPORT

EMERGENCY CAESARIAN SECTION IN A PATIENT WITH ACHONDROPLASIA: A CASE REPORT

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ABSTRACT: The word Achondroplasia literally means "Without Cartilage Formation**"**. It results from abnormal cartilage formation at epiphyseal growth plates. It is the most common form of short limbed dwarfism. Since it is a congenital short statured disease patient can have respiratory, neurological and cardiac problems associated with and poses many challenges to anaesthesiologists to choose best anaesthetic choice. **CASE REPORT:** A 27 years old parturient with short stature (3 feet 8 inches) weighing 45.5 Kgs came to the hospital in early labor with 37 weeks of gestation. Patient presenting cephalopelvic disproportion and foetal distress. She had mild lumbar lordosis and very mild scoliosis and the patient known hypothyroid since the beginning of the pregnancy and on treatment. There is no previous history of exposure to anaesthesia and no previous surgical history. Family history reveals, no person in the family has got Achondroplasia. We used low dose Bupivacaine 1.2ml (6mg) along with fentanyl 0.2ml of fentanyl (10 micrograms) for spinal anaesthesia. First attempt was a dry tap followed a successful spinal anaesthesia with adequate block achieved. **CONCLUSION:** In spite of not many case reports of spinal anaesthesia in Achondroplasia, spinal anaesthesia remains an appropriate option in emergency caesarian section.
KEYWORDS: Achondroplasia, dwarfism, spinal anaesthesia, emergency caesarian section.

INTRODUCTION: Achondroplasia is the most frequent of more than 100 described types of skeletal dysplasia, which lead to dwarfism. Achondroplasia is apparent at birth and has a birth prevalence of 1 in 20,000-30,000 live-born infants. Although it is inherited as an autosomal dominant condition, 80% of cases occur sporadically.[1] Incidence increases with paternal age.[2] It occurs because of abnormal cartilage formation at epiphyseal growth plates. People with Achondroplasia generally have normal intelligence levels, their abnormalities are only physical, and these patients have neurological, cardiopulmonary problems that pose great challenge to anaesthesiologists.

There are two main categories of dwarfism-disproportionate and proportionate. Disproportionate dwarfism is characterized by an average-size torso and shorter arms and legs or a shortened trunk with longer limbs. In proportionate dwarfism, the body parts are in proportion but shortened. Causes of proportionate dwarfism include metabolic and hormonal disorders such as growth hormone deficiency. The most common types of dwarfism, known as skeletal dysplasias, are genetic. Skeletal dysplasias are conditions of abnormal bone growth that cause disproportionate dwarfism.

During early foetal development much of skeleton is made up of cartilage. Normally most cartilage converts to bone however in dwarfism people a lot of cartilage does not convert in to bone. This is caused by mutations in the FGFR3 gene. In more than 80% cases Achondroplasia is not inheritant. According to the National Human Genome Research Institute (NHGRI), these cases are caused by spontaneous mutations in the FGFR3 gene; above 20% cases are inheritant. The mutation follows as an autosomal dominant inheritance pattern.

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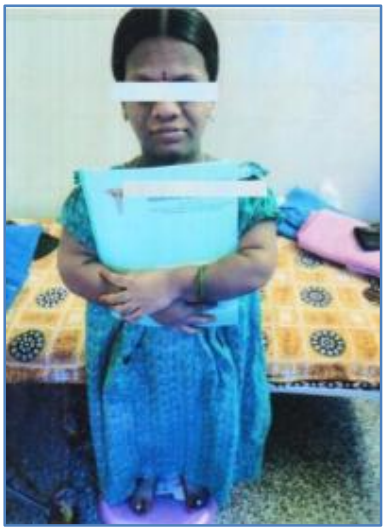

Fig. 1: Patient Showing Typical Features of Achondroplasia

Fig. 2: Patient before Discharge

Fig. 3: Patient with her Mother

A 27 years old primi parturient achondroplastic dwarf (Fig-1 & 2) with 37 weeks of gestation was admitted for an emergency lower segment caesarian section with cephalopelvic disproportion and foetal distress. There was no significant family history and no consanguineous history (All the family members are healthy). She is a known hypothyroid patient on treatment. Her blood pressure is 120/70mmHg. Pulse rate 88/min.

On examination she was seen short statured with height of 112centimetres, Weight 45.5Kgs. She had large head with frontal bossing (Fig. 1) and a saddle nose. She has short neck with limited movements and very short limbs. Her airway examination put her in mallampati grade-II.

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Examination of spine showed mild lordosis and very mild scoliosis. All the base line investigations are normal.

The patient is explained about the spinal anaesthetic technique and Informed consent was obtained. Premedication with Inj. Ranitidin. 50mg, Inj. Metaclopramide. 10mg and Inj. Ondonsetron. 4mg given for aspiration prophylaxis. All the precautions for anticipated difficult intubation management are taken in case of any complications or a failed spinal.

Non-invasive blood pressure, ECG, Pulse oxymeter monitoring were established. She was preloaded with 500ml of ringer lactate solution.

The patient was put on left lateral position (As she was more comfortable for spinal anaesthesia in this position). Literature says the medical sympathectomy following spinal anaesthesia with enhanced gravity-induced peripheral blood pooling, especially in the sitting position often results in significant hypotension.[3] Compared to the sitting position, the lateral position may cause less hypotension.[4]

L3/4 space was chosen and 25 gauge Quincke's spinal needle was used to introduce intrathecally. There was a dry tap in the first and second attempts. Then with another 25 gauge Quincke's spinal needle L4/5 space was chosen and was successful. Then 0.5% Bupivacaine, 1.2ml (6mg) with 0.2ml of fentanyl (10micrograms) was given intrathecally. There was wedge placement with 15^0 tilt.

In the first couple of minutes there was a fall in blood pressure to 70 systolic and 56 diastolic. It was combated with 6mg of Ephedrine IV. After waiting for 5-6 minutes and checking for sensory block till T5-T6. Surgery was started and a female baby weighing 2.10Kgs was delivered after 10 minutes. The APGAR score was 8 at 1min and 10 at 5mins. Except one low pressure episode the patient was stable during intra operative period without any much blood loss. The post-operative period was uneventful; all the fluid intake, Urine output (Input-Output chart) was normal and patient got discharged on the 7^{th} post-operative day.

DISCUSSION: Since achondroplastic patient presents with different physical characteristics, these influence the choice of anaesthetic procedures and complications. Though all the characteristics may not present in each case but when present can vary widely in severity. Since so many factors influence in deciding the best course of anaesthetic management, a proper pre anaesthetic evaluation is mandatory to prepare anaesthesiologist in anticipating potential complications.

In these patients all types of anaesthesia, whether general anaesthesia, Epidural anaesthesia or spinal anaesthesia pose great problems. So as an anaesthesiologist one needs to be well aware of all these before taking up patients for surgery and anaesthesia.

Although each particular disorder is relatively rare, the large number of dwarfs make that many practicing anaesthesiologists is likely to encounter these patients.[s]

Most of the literatures say general anaesthesia is a preferred technique.[6] But it is unclear that the recommendation is based on documentation of complication free anaesthesia technique, as these patients have several derangements presenting complications especially with general anaesthesia which are:

- Narrow nasal passages and Nasopharynx,
- Large mandible & tongue
- Craniofacial abnormalities: frontal bossing, macrocephaly and depressed nasal bridge, maxillary hypoplasia and occipitalisation of first cervical vertebra.

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All,[7] above things can lead to difficult:

a) Mask ventilation.
b) Glottic opening & visualization of vocal cords.
c) Difficult Intubation.

Since these patients have difficult airway, awake fiber optic intubation has been recommended.[8]

In these patients there will be ventilation-perfusion mismatching due to decreased FRC and increased closing volume. This will lead to hypoxia and potentiate severe post op atelectesis. Because of thoracic hypoplasia and kiphoscoliosis, restrictive lung disease common in this patients. So thorough pulmonary evaluation is required in these patients.

Because of craniofacial abnormalities and hipotonia of upper airway muscles,[5] there can be sleep apnea syndrome (SAS) which can lead to development of pulmonary hypertension. All these problems may lead to difficult in maintaining of oxygen saturation during GA and patient may require post-operative ventilator care. There are several reports of pregnant women with Achondroplasia undergoing caesarian section successfully under GA.[9] but the decision to give general anaesthesia guided by many factors as discussed above.

Several authors preferred epidural anaesthesia in pregnant women with Achondroplasia and all have used epidural catheters. This gives advantage of titrating the doses so that targeted levels (by giving graded doses) of analgesia and anesthesia is achieved.[10,11,12,13]

In spite of many reports of conduction of epidural anaesthesia there lie difficulties. They are:

1. In emergency situation technical difficulties
2. There will be inadvertent Dural puncture.[14]
3. The level of block will be high because of short stature of the patients.

The most important thing in epidural and spinal anaesthesia is there are no dosage guidelines unlike to normal patients. That's why the unpredictable spread of the drug through epidural space.

Though available reports say spinal anaesthesia is least preferred and rarely been used citing the causes of,

a) Spinal stenosis and dry tap.
b) Spinal cord ending at lower level.
c) The unpredictability of the spread of the drug.

However spinal anaesthesia has been successfully advocated without any complications in such patients with sensory blockade till T4 was obtained with 1.3ml of Bupivacaine 0.5%.[15,16]

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System	Characteristics	GA implications	EA implications	SA implications
Bones	Lumbar lordosis, Thoracolumbar kyphoscoliosis		Difficulty in positioning	Like in EA
Spine	Relatively narrow spinal canal Vertebral deformities (Shortening of pedicles, decreased interpedicular distance especially in lower lumbar spine, osteophyte formation)		Difficulty in finding landmarks Difficulty in catheter/ needle insertion	Difficulty in obtaining free flow of CSF due to spinal canal stenosis
Bones	Rib hypoplasia, with flattened rib cage	Difficulty of midline positioning of laryngoscope due to pectus carinatum		
Others	Pectus carinatum or excavates Genu varum			
Bones	Maxillary hypoplasia, large mandible, megalocephaly with protuberant	Difficulty in mouth opening and visualization of glottis		
Short neck	Limited neck extension	Difficulty in sealing facial mask due to peculiar facial features risks of forced extension,		
Neurologic	Foramen magnum stenosis	Risk of cervico-medullary compression		
Cardiopulmonary	Sleep apnoea Restrictive lung disease Pulmonary hypertension Cor pulmonale (from pulmonary hypertension, restrictive lung disease and apnea	Risk of hyperthermia Possible complications due to preexisting lung and/or heart disease		
Decreased height & weight		Unclear dose requirements for anesthetic and muscle relaxant	Dosage Guidelines Unclear	Risks of inadvertent high spinal block , e.g., profound hypotension
Disproportionate dwarfism Non-uniform spinal canal lumen		Unclear dose requirements for anesthetic and musclerelaxant	Dosage Guidelines Unclear	Risks of inadvertent high spinal block , e.g., profound hypotension

Table 1: Major Abnormalities Which May Cause Anaestehtic Implications in Achondroplastic Patient in all Types of Anaesthesia

CONCLUSION: In this case we have chosen spinal anaesthesia and successfully done the procedure without any sequele. The reasons being since it's an emergency LSCS so,

1. Lack of adequate preparation.
2. Risk of aspiration.
   Both will be detrimental for GA.

Patient also has got hypothyroidism (Anticipatory delayed recovery).

Since patient has got mild lordosis and very mild scoliosis, We decided to give spinal anesthesia with low dose Bupivacaine i.e. 1.2ml (6 mg)+0.2ml (10 micrograms) fentanyl combination with we achieved T4-T5 levels of analgesia and anaesthesia except for the brief episode of hypotension which can be manageable with Ephedrine the procedure was smooth and uneventful.

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