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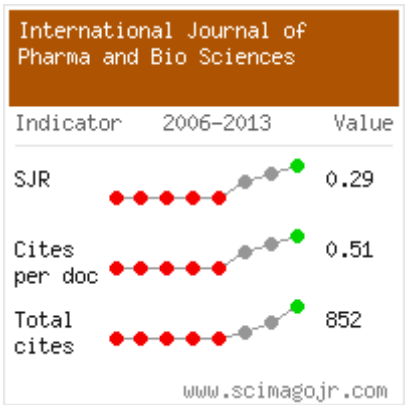

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CASE REPORT **ANAESTHESIA**

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ANAESTHETIC MANAGEMENT OF A PATIENT WITH ACHONDROPLASIA POSTED FOR BILATERAL URETEROSCOPIC LITHOTRIPSY- AN ANAESTHETIC CHALLENGE.

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ABSTRACT

Achondroplasia is the commonest form of dwarfism which results fromabnormal cartilage formation at epiphyseal growth plates.A forty year old male , short stature presented to the hospital with bilateral ureteric stone and posted for bilateral ureteroscopic lithotripsy.This is a case report of a successful Anaesthetic management of a patient with achondroplasia under spinal anaesthesia.

KEY WORDS: **Achondroplasia,Spinal anaesthesia,Scoliosis, Difficult airway.**

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INTRODUCTION

Achondroplasia is the commonest form of dwarfism which results from abnormal cartilage formation at epiphyseal growth plates.It occurs as a sporadic mutation in approximately 80% of cases or may be inherited as an autosomal dominant genetic disorder.this disorder is caused by a change in the gene for fibroblast growth factor receptor 3($FGFR_3$) Prevalence is approximately 1 in 25,000, It can be detected prenatally by use of ultrasound guided chorionic villi sampling for DNA testing. Achondroplastic adults are known to be as short as 62.8cms.It is also known as short limbed dwarfism. Anaesthetic management in these patients poses a significant challenge as airway management can be difficult due to cranio- facial abnormalities. Centrineuraxial blockade is technically difficult with unpredictable block even after successful technique.

CASE REPORT

A forty year old male with bilateral ureteric stone was posted for bilateral ureteroscopic lithotripsy.He was short statured with height of 120 cm & weight of 40 kgs.He had a large head with frontal bossing, saddle nose, short neck & short limbs. No history of breathlessness ,palpitaions,orthopnea and PND symptoms.Examination of spine showed right scoliosis in the lumbar area.Mallampatti Grade 3 on airway assessment, adequate neck movements and short neck.ECG was normal. ECHO showed mitral valve prolapse , trivial mitral regurgitation with mild pulmanary hypertension with LVEF 65% .Written informed consent was taken. Spinal anaesthesia was planned .Equipment for the management of difficult airway was kept ready.ECG , non invasive BP & pulse oximeter monitoring was instituted.Baseline Spo_2 88%. He was preloaded with 500ml Ringer's Lactate solution.He was premedicated with inj ondansetron 4mg & midazolam 0.5 mg iv.Under aseptic precautions spinal anaesthesia was tried in the sitting position using 25G Quincke spinal needle through midline approach.After two dry taps , left paramedian approach for subarachnoid block was tried & was successful.Subarachnoid block was given in L4-L5 space with 1.4cc of Inj.hyperbaric bupivacaine(0.5%) with 60 microgrms of Inj buprenorphine.The patient was placed supine with a slight head up tilt . Sensory block was achieved till T8 in 2mins.A significant delay in motor blockade was noticed (15mins).Complete motor blockade of lower limb was seen after 30mins.Pulse , BP& Respiration were maintained. O2 was supplemented by face mask. Intra & post op period were uneventful.Consent to publish this case report was taken from the patient.

DISCUSSION

Achondroplasia is an autosomal dominant disorder caused by mutation in the gene which codes for fibroblast growth factor receptor type 3.[1]Achondroplasia is characterised by narrow nasal passages in nasopharynx, large tongue ,large mandible, short neck, cranial and facial abnormalities such as large head, frontal bossing, nasal bridge will be deppressed, hypoplasia of maxilla and fusion of occipetal of 1st cervical vertebra(figure 1).[2]All these features lead to difficulty in mask ventilation, glottic opening visualisation and intubation.Thoracic dysplasia, thoracic lordosis and thoracic kyphoscoliosis which lead to restrictive lung disease.There is ventilation perfusion mismatch due to decreased FRC and increased closing volume promoting atelectasis.There is tendency for sleep apnoea which may be central or obstructive due to craniofacial abnormalities.All these complications can lead to pulmonary artery hypertension.These conditions may lead to difficulty in maintaining oxygenation during general anaesthesia and mechanical ventilation may be required post operatively.[3] There is constriction of spinal cord resulting in narrowing of subarachnoid and epidural spaces which may cause difficulty in establishment of centrineuraxial blockade.There may be difficulty in identifying interspinous spaces due to lumbar hyperlordosis, decreased interpeduncular

distance, osteophyte formation and malformed vertebra, spinal stenosis make spinal anaesthesia difficult.(fig -2) There is unpredictability of spread of drug as the spinal cord ends at a lower level in these patients.In our patients repeatedly dry tap was obtained on attempting lumbar puncture.However spinal anaesthesia was given successfully without any sequelae in our patient.

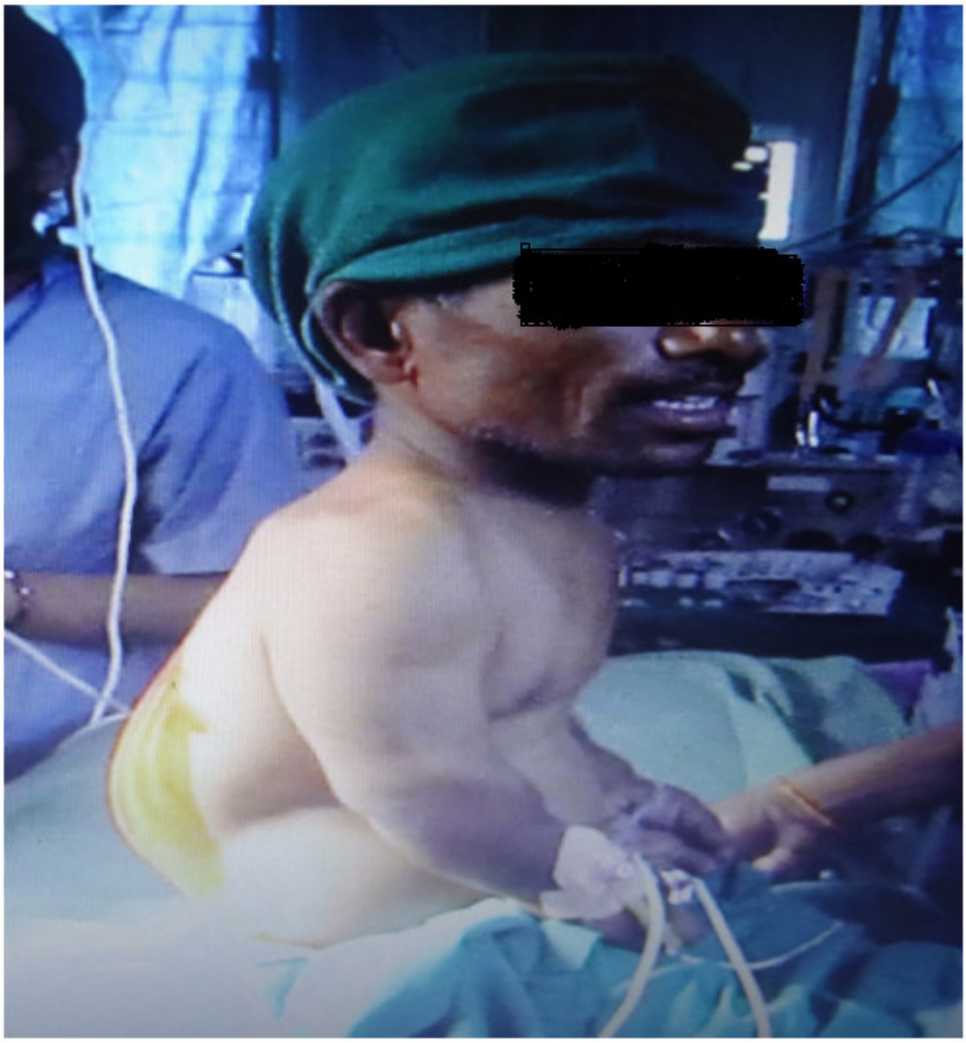

FIGURE 1
MORPHOLOGICAL FEATURES OF ACHONDROPLASIA

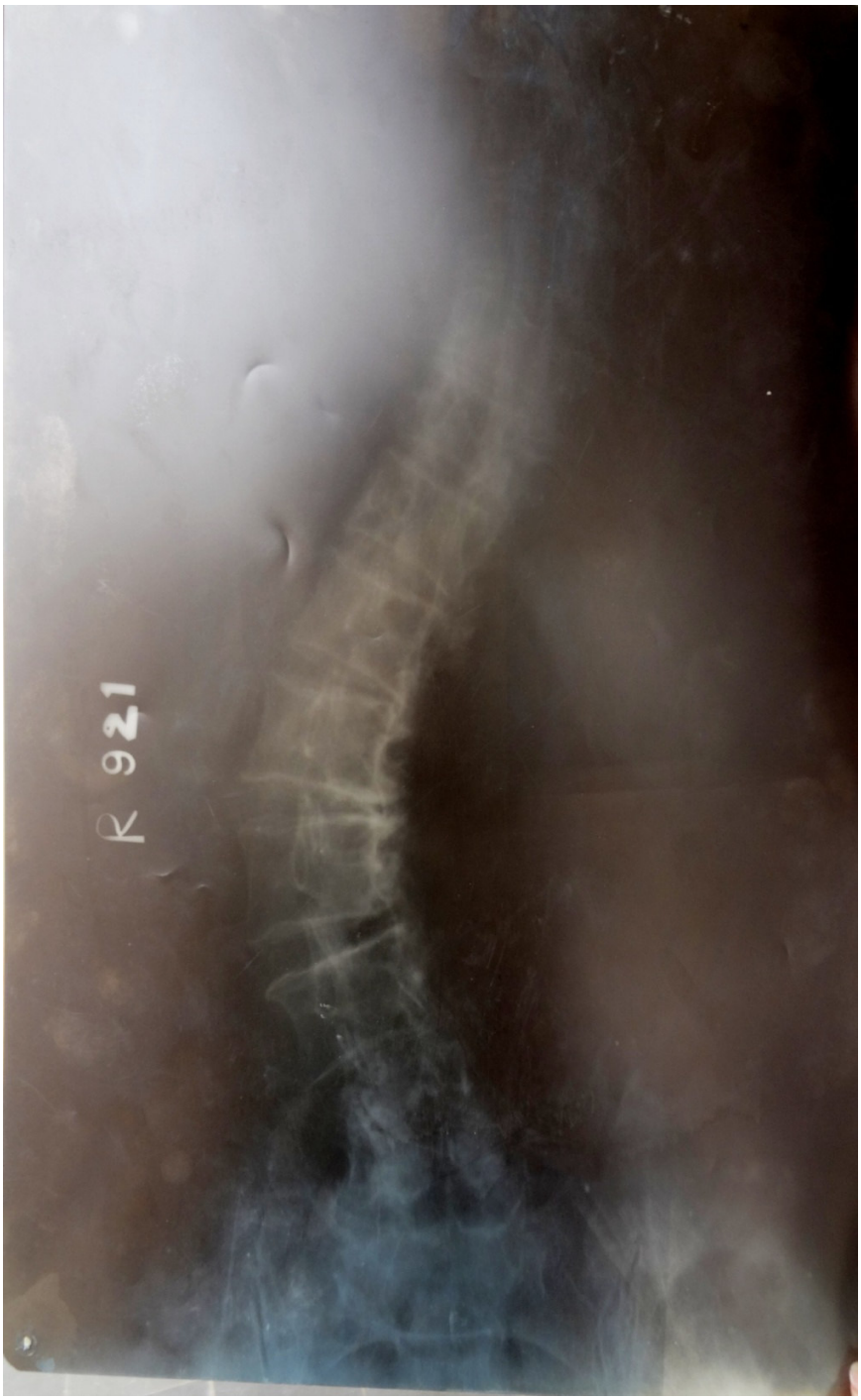

FIGURE 2
XRAY FEATURES OF CHEST AND SPINE .

CONCLUSION

In achondroplasia patients, due to effects on cardiovascular , respiratory and upper airway it may lead to various peri-operative mishaps.Hence, appropriate anaesthetic management and post-operative care is essential.Regional anaesthesia is the technique of choice even though it is technically difficult.

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