

## Anesthetic Management of Achondroplastic Dwarf with Bronchial Asthma and Hypothyroidism Posted for Abdominal Hysterectomy

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### Abstract

**Introduction:** Achondroplasia is most common cause of dwarfism caused by FGFR-3 gene mutation. It possess anesthetic challenges due to multiorgan system involvement, spine abnormality, difficulty in mask ventilation and endotracheal intubation.

**Case Report:** A 38yr old, female, case of achondroplastic dwarfism with history of bronchial asthma and hypothyroidism was posted for abdominal hysterectomy. She weighed 30 kgs and 43 cm in height.

Spine examination showed thoracolumbar kyphoscoliosis. Airway assessment revealed large tongue, receding mandible, Mallampatti class IV. Pulmonary function test suggested mild restrictive disease.

Difficult intubation trolley was kept ready. Routine monitoring, radial artery cannulation done for invasive blood pressure monitoring. Patient was given general anesthesia. On laryngoscopy, CL grade III view was obtained, with the aid of bougie endotracheal tube was placed. Delayed recovery time from general anesthesia was encountered, neuromuscular monitoring was done and reversed and extubated uneventfully.

**Conclusion:** we present a successful anesthetic management of a dwarf patient with bronchial asthma and hypothyroidism who underwent hysterectomy. We emphasize the risk of neurological injury while extending the neck during laryngoscopy for tracheal intubation due to anatomical abnormalities in these patients. A detailed pre anesthetic evaluation and planning is utmost important and the anesthetic technique has to be individualized based on the patients anatomical characteristics and associated co-morbidities.

**Keywords:** Dwarf, Asthma, Hypothyroidism.

**Key Messages:** A 38 yrs old female patient with achondroplastic dwarfism (43cm tall) with associated comorbidities like bronchial asthma and hypothyroidism, thoracolumbar kyphoscoliosis and mild restrictive lung disease posted for abdominal hysterectomy possesses various anesthetic challenges. Since multiple systems are involved preanesthetic evaluation, preoperative optimization of the patient and planning of anesthesia is important. As the spread of drug in regional anesthesia is unpredictable, we planned general anesthesia. A successful management of major abdominal surgery in such patients, with anesthetic challenges and implications in these patients are reviewed in this case report.

## Introduction

Dwarfism is defined as failure to achieve a height of 148cm by adulthood.<sup>2</sup> Achondroplasia is the most common cause of dwarfism with an autosomal dominant trait and caused by mutation in Fibroblast Growth Factor Receptor 3 (FGFR-3) gene.<sup>1</sup> It possesses multiple anesthetic challenges due to multiorgan system involvement, spine abnormality, difficulty in mask ventilation and endotracheal intubation.<sup>3</sup> We report a successful anesthetic management in a patient with achondroplastic dwarfism with hypothyroidism and bronchial asthma posted for abdominal hysterectomy.

## Case Report

A 38 year old, female patient, case of achondroplastic dwarfism came with complaints of mass per abdomen posted for total abdominal hysterectomy. She was a known case of bronchial asthma and hypothyroidism on medication since 3 years. She was 43cm in height and weighed 30kgs. On general physical examination, she was afebrile with pulse of 96/min, blood pressure of 130/90mm Hg and respiratory rate of 16/min. She had normal intelligence. Examination of spine showed a gross thoracolumbar kyphoscoliosis (Fig. 1). Airway assessment revealed large tongue with



Fig. 1: Kyphoscoliosis

receding mandible, buck teeth. Mouth opening was adequate with Mallampatti class IV. Her blood investigations were within normal limit. Pulmonary function test suggestive of mild restrictive disease. Electrocardiography was normal and transthoracic echocardiography showed Mitral valve prolapse with mild MR, normal chamber dimensions and ventricular ejection fraction of 60%. MRI pelvis showed a large pelvico-abdominal heterogeneous lobulated mass connected with uterine fundus. Patient was advised to continue Tab. Thyronorm 50mcg and salbutamol and theophylline, nebulization with duolin and budesonide on the day of surgery.

Written informed consent with high risk explained was obtained. 18G IV access was secured. Difficult intubation trolley including bougie, video laryngoscope was kept ready. Emergency resuscitation drugs, equipment and defibrillator were kept ready. After applying routine monitoring, left radial artery cannulation was done for invasive blood pressure monitoring because non-invasive blood pressure measurements were inconsistent due to shortness of the limbs. Preoxygenation done with 100% oxygen for 3 mins. Patient was premedicated and anesthesia was induced with Inj. Ketamine 60mg IV. On laryngoscopy, a Cormack and Lehane grade III view was obtained using bougie a 6.5mm cuffed endotracheal tube was successfully placed and fixed at 17cm after confirming bilateral air entry equal. Anesthesia was maintained with oxygen, nitrous oxide and isoflurane. Muscle relaxant Inj. Atracurium was given. Blood loss was significant, 1 unit of PRBC was transfused and patient was hemodynamically stable in the intraoperative period. Delayed recovery time from general anesthesia was seen and neuromuscular monitoring was done using train of four (TOF) stimulation when TOF ratio was > 0.7

patient was reversed with Inj. Neostigmine 1.5mg IV and Inj. Glycopyrolate 0.2mg IV and extubated uneventfully. She was shifted to post operative care unit. Postoperative analgesia was maintained with inj. Paracetamol 15mg/kg IV TID.

## Discussion

Achondroplasia is the most common cause of dwarfism caused by gain of function mutation in fibroblast growth factor receptor 3 (FGFR3) gene. 80% mutations are the result of sporadic mutation while 20% are autosomal dominant. Incidence is approximately 0.5-1.5 in 10,000 newborns. Females are more affected than the male.<sup>1,4</sup> A general

recommendation regarding the ideal anesthetic technique cannot be given, as both general and regional anesthesia present potential problems.<sup>1</sup> Short stature, enlarged head, saddle nose, maxillary hypoplasia, macroglossia, megaloccephaly with protrudent forehead and narrow nasal passages and nasopharynx are some features that contribute to difficult airway in these patients.<sup>2,4</sup> These patients have respiratory problems that may complicate general anesthesia, particularly restrictive lung disease due to thoracic kyphoscoliosis. There is ventilation perfusion mismatch due to decreased FRC and increased closing volume leading to atelectasis.<sup>3</sup> Kyphoscoliosis associated with narrowing of the vertebral canal, reduced distance between the pedicels and osteophyte formation can produce technical difficulty for spinal anesthesia.<sup>3</sup> A narrow epidural space increases the difficulty in epidural catheter insertion and also limits the spread of local anesthetics.<sup>2</sup> The consensus among anesthesiologists thus remains to avoid neuraxial anesthesia in patients with skeletal dysplasia.<sup>6</sup> There are no definite recommendations regarding anesthetic procedures, the decision has to be taken on individual basis after a detailed risk-benefit analysis.<sup>5</sup>

### Conclusion

In our case report, we present a successful anesthetic management of a dwarf patient with bronchial asthma and hypothyroidism who underwent hysterectomy. We emphasize the risk of neurological injury while extending the neck during laryngoscopy for tracheal intubation due to anatomical abnormalities in these patients. A

detailed pre anesthetic evaluation evaluation and planning is utmost important and the anesthetic technique has to be individualized based on the patients anatomical characteristics and associated co-morbidities. It provides instructive significance for anesthesia management in this rare condition.

**Conflict of Interest:** NIL

### References

1. Deshpande JP, Kale JH, Godbole MR, Phalke TL. Anesthetic management of an achondroplastic dwarf with difficult airway and kyphoscoliosis for total abdominal hysterectomy. *Indian J Clin Anaesth* 2021;8:475-478.
2. A Jain, K Jain, JK Makkar & K Mangal. Case study: Anesthetic management of an achondroplastic dwarf undergoing radical nephrectomy. *Southern African Journal of Anesthesia and Analgesia* 2010;16:2, 77-79.
3. Karnalkar AP, Deshpande A. Anesthesia management of patient with achondroplasia for abdominal hysterectomy. *Int J Sci Rep* 2015;1:264-6.
4. NeisevilieNisa, Puneet Khanna, Dhruv Jain. Anesthetic Management of an Achondroplastic Dwarf with Difficult Airway and Spine for Total Hip Replacement: A Case Report. *Gen Med (Los Angeles)* 2016;4:227.
5. TasnimN, Ghani M, Kumar S, Naeem S, Luqman S. Successful pregnancy and delivery in a women with achondroplasia. *J Pak Med Assoc.* 2021;71:146- 149.
6. Tyagi A, Dass C, Rao NT, Soni KD. Emergency anesthetic management of an achondroplastic elderly gravida with polytrauma. *Int J Crit Illn Inj Sci.* 2019;9:191-193.

