

Failed rapid sequence induction in an achondroplastic dwarf

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ABSTRACT

Achondroplasia, a common cause of short limbed type of dwarfism is due to quantitative decrease in rate of endochondral ossification. This abnormal bone growth leads to disproportionate body and head structure, thus placing them under high risk for anaesthetic management. There is paucity in literatures, regarding appropriate drug dosage selection in these patients. Use of drugs as per standard dosage recommendations based on body weight or body surface area, may not be adequate in these patients owing to discrepancies in overall body weight and lean body weight, especially during rapid sequence induction. Here, we report a case of failed rapid sequence induction due to abnormal response to administered drugs in an adult achondroplastic dwarf. Standard doses of thiopentone and rocuronium had to be repeated thrice to achieve adequate conditions for intubation.

Key words: Achondroplasia, rapid sequence induction, drug dosage, pharmacokinetics

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INTRODUCTION

Achondroplasia is the most common cause of the short-limbed type of dwarfism. 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 shorter tubular bones present in the achondroplastic dwarfs. This abnormal bone growth is responsible for several potential anaesthetic problems. The cardiorespiratory and airway abnormalities are well described in these patients.^[1] However, the selection of an appropriate dose of drugs is not elucidated in the literature. We report a failed rapid sequence induction (RSI) in an adult achondroplastic patient due to unpredictable response to the administered drugs.

CASE REPORT

A 37-year-old achondroplastic dwarf was admitted with acute gastric outlet obstruction due to gastric volvulus, and emergency laprotomy was planned.

On examination, he was a dwarf with a height of

117 cm and weight of 18 kg. There were signs of bony abnormalities consistent with the diagnosis of achondroplasia, such as shortening of the proximal segments of the upper and lower limbs, increased skull size, and mandibular and frontal protusion. He had pectus excavatum with bilaterally reduced chest expansion. His laboratory results were unremarkable. There was no history of substance abuse.

In view of gastric outlet obstruction, RSI was planned. The patient was pre-medicated with intravenous pantoprazole 40 mg. Routine monitors were attached in the operating room. He had a pulse rate of 70/min and blood pressure of 110/80 mmHg. The room air saturation was 94%. He was pre-oxygenated for 5 min and Sellick's manoeuvre was used to safeguard against possible regurgitation. Anaesthesia was induced with thiopentone 5 mg/kg and rocuronium 1.2 mg/kg. Intubation was attempted after 1 min, but it was observed that the patient was still awake. Because there was no evidence of slow circulatory status or other physiological causes to explain the delayed onset of drug action, the technical causes for inadequate drug effect, like loss of drug from the I.V. tubing, cannula, or

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extravasation, were excluded. Additional thiopentone and rocuronium in similar doses were administered. He still opened his eyes and moved on attempt to intubation. A third dose of thiopentone 5 mg/kg and rocuronium 1 mg/kg was given. Intubation could be accomplished with a 7.5-mm cuffed endotracheal tube without difficulty. Hyperextension of the neck was avoided. Anaesthesia was maintained with 1 MAC sevoflurane, air, and oxygen with positive-pressure ventilation in a circle system. He required a minute ventilation of 5.4 L to achieve normocarbida. Intraoperative analgesia was provided with fentanyl 100 µg. Atracurium was clinically titrated to provide adequate muscle relaxation. The surgery lasted for 2.5 h. The total atracurium administered was 25 mg. Antagonism of residual neuromuscular blockade was achieved satisfactorily with neostigmine 2.5 mg and glycopyrrolate 0.5 mg. Spontaneous ventilation and consciousness were quickly re-established. He remained clinically well and his oxygen saturation was satisfactory following extubation. Post-operative analgesia was provided with injection diclofenac 50 mg intramuscularly thrice a day. He had an uneventful hospital course.

DISCUSSION

RSI is designed for the expeditious intubation of the trachea. The success of the procedure depends on the administration of appropriate drugs in adequate doses that facilitate induction and intubation within the safe apnea period. Improper conductance of RSI or failure to intubate rapidly may result in pulmonary aspiration of the gastric contents, leading to acute airway obstruction, arterial hypoxaemia, hypercapnia, acidosis, pneumonitis, and even death. The literature on the anaesthetic management of achondroplastic patients is scanty. The cardiorespiratory and airway abnormalities that pose a challenge to anaesthetic management are well documented. However, there are no pharmacokinetic studies in these patients to determine the anaesthetic drug doses. Most recommendations on drug dosing in achondroplastic dwarfs are based on case reports in elective surgical settings.^[2-5] Titration of drug requirement is simpler in an elective setting as it can be adjusted based on response. However, in a situation of RSI, administering the appropriate dose is essential to facilitate rapid smooth intubation within 1 min.

The literature suggests that the anaesthetic drugs should be administered on a precise body weight

basis in achondroplastic patients.^[6] However, our observation of drug requirement was contrary to that of earlier reports. We had initially followed the standard recommendations and administered drugs based on body weight. But, it was observed that these doses were insufficient and required repeated administration of anaesthetic drugs. This resulted in an inability to provide RSI and intubation. Fortunately, although the patient had massive distention of stomach due to volvulus, he did not suffer from pulmonary aspiration.

We have attempted to explain this observation based on the possible physiological and pharmacokinetic changes described in achondroplastic individuals. It has been reported that they have distinctively greater caloric requirements per unit body weight or for an identical fat-free mass than do average height adults.^[7] A partial explanation for this may reside in the relative difference in the organ masses in these populations. For metabolic purposes, human organs can be arbitrarily classified into high, moderate, and low energy-requiring tissues, and the blood flow to these tissues varies accordingly. Large variances between normal adults and achondroplastic adults occur, not because of major differences in high energy-requiring well-perfused brain, heart and liver masses but because of large differences in the amount of moderate energy-requiring skeletal muscle mass and in the amount of low energy-requiring adipose tissue mass. Most of the dwarfs have disproportionately enlarged central nervous systems^[8] and probably enlarged hepatic masses compared with their relatively small extremities or small other total body parts. In the average-sized adult, the vital organs like brain and liver constitute about 2% of the body weight, but they consume about 20% of the resting metabolic requirements.^[7,9] The high energy requirements and blood flow of the brain and liver may be more evident in dwarfs with disproportionately small extremities or other body parts. Consistent with this contemplation, it was also observed in our patient that the minute volume required to maintain normocarbida was larger for the body weight. The anaesthetic drug dosage depends on the distribution volume, which is mostly contributed by the vessel-rich group, which, in an achondroplastic patient, is not different from a normal statured adult. Most achondroplastic adults are obese.^[10] When the influence of obesity on body weight is eliminated, the calculation of drug doses according to the lean body weight would result in underdosing, as observed in our patient. In obese achondroplasia, the precise body weight may be close to the lean body weight of a

normal statured person of that age. However, in non-obese achondroplastic patients, the drug doses may be similar to an average statured adult. Pharmacokinetic studies of anaesthetic drugs are required in these patients to substantiate our observations.

CONCLUSION

The anaesthetic management of the achondroplastic dwarf is a challenge. In emergency situations, where drug dose titration is not possible or when RSI is indicated, it is advisable to administer the drugs as required for an average statured adult of that age rather than weight of the achondroplastic patient.

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