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# Is Management of Anesthesia in Achondroplastic Dwarfs Really a Challenge?

**Pablo Monedero, MD, PhD,***
**Francisco Garcia-Pedrajas, MD, PhD,***
**Ines Coca, MD,† J. Ignacio Fernandez-Liesa, MD,†**
**Alfredo Panadero, MD, PhD,***
**Jose de los Rios, MD, PhD†**

Department of Anesthesiology and Critical Care, Clinica Universitaria, School of Medicine, University of Navarra, Pamplona, Spain.

Study Objective: *To review our eight-year anesthetic experience with achondroplastic patients.*
Design: *Retrospective study.*
Setting: *University hospital.*
Patients: *15 achondroplastic patients who underwent 53 surgical procedures of orthopedic surgery between 1987 and 1994.*
Interventions: *Anesthetic technique, drugs, number of incidents, and complications in the intraoperative and postoperative period were recorded.*
Measurements and Main Results: *Adequate premedication before the transfer to the operating room was very useful to reduce anxiety and increase cooperation. Inhalation induction was well tolerated and allowed easy peripheral venous cannulation. Only one patient presented difficulties during intubation (on two occasions). In the other patients, we found small difficulties only during ventilation with a face mask, which was easily corrected by modifying the position of the patient and/or inserting an oropharyngeal airway. No adverse effect was identified for any particular anesthetic drug or technique used.*
Conclusions: *Although the characteristic deformities of achondroplastic patients can impede the management of anesthesia, in our study we found no special difficulties. Airway complications did not occur. Thus, no specific optimal anesthetic regimen can be recommended.* © 1997 by Elsevier Science Inc.

**Keywords:** Achondroplasia; airway management; anesthesia; dwarfism.

## Introduction

Achondroplasia is the most common cause of dwarfism, with an estimated frequency between 1 in 15,000 and 1 in 40,000 live births.[1] A change in the genetic information for fibroblast growth factor receptor 3, which is located in nucleotide 1138, produces a decrease in the rate of endochondral ossification.[2] Achondroplasia is inherited as an autosomal dominant trait with complete penetrance, but approximately 80% of cases represent spontaneous mutation. The diagnosis can be made at birth based primarily on clinical and roentgenographic findings.[3] The characteristic changes that can impede the management of anesthesia are a large protruding forehead, depressed nasal bridge, short maxilla, large mandible, large tongue, tonsil enlargement, nasal, pharyngeal, and laryngeal hypoplasia, and a disproportionately large head; neurologic

*Associate Professor of Anesthesiology

†Assistant Professor of Anesthesiology

Address reprint requests to Dr. Monedero at Clinica Universitaria Apartado 192, 31080 Pamplona, Spain.

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655 Avenue of the Americas, New York, NY 10010

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**Table 1.** Demographic Data and Number of Surgical Interventions

| Patient | Gender | Number of interventions | Age (range) (yrs) |
|---|---|---|---|
| 1 | M | 5 | 15–17 |
| 2 | M | 1 | 4 |
| 3 | F | 3 | 20 |
| 4 | F | 1 | 10 |
| 5 | F | 4 | 4–9 |
| 6 | M | 1 | 13 |
| 7 | M | 4 | 7–11 |
| 8 | F | 4 | 8–10 |
| 9 | F | 1 | 15 |
| 10 | F | 12 | 8–13 |
| 11 | M | 3 | 5–6 |
| 12 | F | 6 | 4–10 |
| 13 | M | 5 | 8–11 |
| 14 | F | 1 | 23 |
| 15 | F | 2 | 15–16 |

dysfunction due to cervicomedullary compression or thoracolumbar spinal stenosis; and scoliosis, thoracolumbar kyphosis, or lumbar hyperlordosis.[4] Numerous respiratory and neurologic complications have been described.[5–8] The aim of our study was to evaluate, through our experience with a large number of orthopedic surgical patients, the technical difficulties and complications in the management of anesthesia in the achondroplastic patient.

## Materials and Methods

Between 1987 and 1994, 15 achondroplastic patients underwent orthopedic surgical procedures in our hospital. All patients fulfilled the radiologic criteria of achondroplasia according to the International Classification of Osteochondrodysplasia.[9] The differential diagnosis with other types of dysplastic diseases (eg, tanatophoric dysplasia, hypochondroplasia) was made by the surgeon based on specific anatomic abnormalities (eg, facial abnormalities, rizomielic micromelia, trident hand) and radiologic criteria. The homozygosis of achondroplasia was ruled out in our patients because all were older than 4 years and homozygotes never survive more than 1 year. Their hospital records were reviewed. We analyzed the demographic characteristics, surgical procedures, anesthetic techniques, medication, and intraoperative and immediate postoperative complications.

## Results

Fifteen patients, 6 men and 9 women, between the ages of 4 and 23 were studied (*Table 1*). They underwent 53 orthopedic surgical procedures with general anesthesia, mainly limb-lengthening orthopedic procedures, using a dynamic axial external fixation system (*Table 2*). The mean duration of the anesthesia was 102 minutes, with a large range depending on the type of surgery. Careful assessment of the airway and a neurologic examination was carried out in all the patients to screen for possible difficulties in intubation and signs of cord compression. No neurologic alteration or special airway difficulty (Mallampati class 1 or 2) could be detected. Patients were premedicated with nasal (0.4 mg/kg) or rectal (0.5 to 0.6 mg/kg) midazolam in the holding area before the transfer to the operating room (OR), with pulse oximeter monitoring and nursing care. We observed certain difficulties of peripheral venous access, with a 30% failure rate in the first attempt that decreased below 10% as our experience increased. Induction of anesthesia was achieved with increasing concentrations of halothane in a mixture 1:1 of nitrous oxide ($N_2O$) and oxygen ($O_2$), or intravenously with sodium thiopental (*Table 3*). Tracheal intubation, when required, was facilitated by a dose of neuromuscular blocker (succinylcholine 1 mg/kg in 1 case, pancuronium 0.1 mg/kg in 18 cases, and vecuronium 0.1 mg/kg in 14 cases) (*Table 3*).

**Table 2.** Surgical Procedures and Anesthesia Time (min)

| Type of surgery | Number of procedures | Anesthesia time* |
|---|---|---|
| Lengthenings tibia/femur/humerus (corticotomy + external fixation system) | 27 | 139 (75 to 210) |
| Replacement/correction of fixation system | 16 | 69 (15 to 110) |
| Plasters | 5 | 58 (23 to 95) |
| Tenotomies | 5 | 95 (60 to 125) |

*Data are given as means, with ranges in parentheses.

Anesthesia was maintained with isoflurane in 50% $N_2O/O_2$, or a combination of fentanyl (3 to 8 mcg/kg$^{-1}$), $N_2O$, and isoflurane (*Table 3*). The complications that arose were two difficulties of intubation in the same patient and two postextubation laryngospasms in the OR treated with $O_2$ 100% plus continuous positive airway pressure (CPAP) through facial mask (*Table 4*). The difficult intubation was due to impossible visualization of the epiglottis (laryngoscopy grade 4) in a 15-year-old female. On the first occasion, a nondepolarizing relaxant was used because there was no suspicion of any difficulty with intubation. After two failed attempts of tracheal intubation, a laryngeal mask airway (LMA) number 3 was inserted without difficulty. Once a correct placement of the LMA was assured, the surgical procedure was done

**Table 3.** Anesthetic Management [no. of Procedures (%)]

| | |
|---|---|
| *Induction* | |
| Inhalatory | 21 (40%) |
| Intravenous | 32 (60%) |
| *Airway management* | |
| Intubation | 33 (62%) |
| Mask ventilation | 20 (38%) |
| *Maintenance* | |
| Inhalatory | 39 (74%) |
| Balanced (inhalatory + morphinics) | 14 (26%) |

**Table 4.** Complications [no. of procedures (%)]

| | |
|---|---|
| Difficult intubation | 2 (3.8%) |
| Laryngospasm | 2 (3.8%) |

with mechanical ventilation until recovery of neuromuscular blockade. On the second occasion, once the hypnotic drug had been administered and manual ventilation with a face mask checked, 1 mg/kg of succinylcholine was administered, resulting in partial vision of the larynx (laryngoscopy grade 3), which permitted tracheal intubation with the aid of a stylet.

In the postoperative period, we found no cases of respiratory depression, one case of postoperative vomiting, and one case of cutaneous rash. In the recovery room, 11 patients required postoperative analgesics with paracetamol or magnesium metamizol.

## Discussion

We offer the largest published series of nonneurosurgical procedures on achondroplastic patients. In order to avoid the psychological consequences of their physical abnormalities and improve their quality of life, these patients may undergo numerous surgical procedures, such as lengthening of the shortened limbs, other orthopedic treatments, and correction of craniofacial deformities.

Management of anesthesia of achondroplastic dwarfs can be challenging due to the difficulty of manual ventilation and intubation. Difficulties appear when obtaining a good fit with the face mask during manual ventilation because of macroglossia, bad dental occlusion, a flattened nasal bridge, and a short mandible or relative prognathism.[4,7,10] In our series, the most common difficulty with ventilation was caused by macroglossia. When it was necessary, an oropharyngeal airway solved the problem, allowing spontaneous or assisted ventilation. We believe that it is possible to compare these problems to the typical ones found in the ventilation of children (eg, macroglossia, short neck) and those of elderly patients without teeth. The low functional residual capacity of these patients favors a rapid desaturation of $O_2$ with inadequate ventilation, warranting a good preoxygenation. As for tracheal intubation, we only had difficulties with one patient due to difficult visualization of the larynx. On the first occasion, a nondepolarizing relaxant was used because, even though an airway assessment was done, there was no suspicion of any difficulty with intubation. Careful assessment of the airway should be carried out in all achondroplastic patients to screen for possible difficulties in intubation, and it is usually possible to identify these cases.

Problems with intubation are due to craniofacial deformities and the premature fusion of the bones at the base of the skull, producing a limit to cervical extension that makes visualization of the larynx impossible.[7] In our patient, there was no clear limitation of extension or any other signs that might have indicated difficulty with intubation. Unfortunately, no cervical radiologic control was done to identify abnormalities that might have helped us foresee the resulting difficulty with intubation. The radiologic images of the head and neck can be used to predict the difficulty of intubation and some authors suggest that they should be compulsory in all achondroplasic patients.[11] Radiologic changes that can be associated with difficulty in intubation are an increase in the depth of the mandible, a decrease in the distance between the occiput and the spinous apophysis of $C_1$ (atlanto-occipital distance), a reduction in the $C_1$–$C_2$ intervertebral space, and a reduction of mobility of the mandible.[12] In our opinion, a cervical radiologic study is not essential in the absence of neurologic symptoms; a careful clinical examination of signs that may indicate a possible difficulty in intubation is usually sufficient. In our patient, a LMA was inserted without difficulty the first time, and tracheal intubation with the aid of a stylet was achieved after the administration of succinylcholine on the second occasion. We did not use a pediatric flexible fiberoptic laryngoscope for intubation because it was not available. Blind nasotracheal intubation was not done because of the risk of bleeding by lesion of hypertrophic adenoid growths. This event would make ventilation and intubation even more difficult. We did not consider an awake intubation for our patient because there was neither limitation of the buccal opening, nor a very short neck with limited mobility, nor notable thoracic deformity. Awake intubation would have been considered if risk of spinal cord compression were present. Furthermore, when there is a risk of spinal cord compression, it is necessary to use cervical stabilizing methods before any manipulation.

In the achondroplastic patient, it is necessary to use endotracheal tubes that are smaller than those used in normal patients because the larynx is smaller and the size correlates more closely with the weight of the patient than with age.[10] As a safety measure, materials for difficult intubation, including tubes of various sizes, should be available.

Although none of our patients presented with neurologic symptoms, hyperextension of the neck during intubation was avoided through a gentle intubation technique, using the least hyperextension necessary for vocal cords visualization. Injury to the cervical spinal cord[8] is feasible when there are signs of cord compression. Cervical musculature hypotonia in children increases the risk of cord injury. Radiologic examination of the atlantoaxial joint should be carried out in all patients with neurologic symptoms of compression of the cervical cord (eg, hypotonia, muscular weakness, spasticity, clonus, hypereflexia, paraplegia) or for cases in which physical examination leads to suspected difficulty in tracheal intubation.[13,14]

It may seem paradoxical that although numerous authors[4,7,10,11,15] warn of the neurologic and respiratory complications in the management of the achondroplastic patient, in our series it was present in only one patient. We believe that it may be due to the age of our patients. The greatest risks for the achondroplastic patient occur during the first year of life,[15] with a high mortality due to obstruction of the superior airway[6,15] and compression of the cervical spinal cord.[14] Once the period of greatest risk has passed, life expectancy is normal if no complications

arise. In our series, none of the patients was younger than 4 years of age. Furthermore, none of our patients presented an associated organic pathology or facial-cervical deformities that were abnormal in this type of dwarfism. Therefore, the difficulty of intubation was similar to that of a patient of normal size with a short neck and an anterior larynx.

The premedication of our patients proved useful in reducing anxiety and increasing cooperation with line placement or mask tolerance, more so because these were overprotected children and teenagers who had undergone repeated anesthetic-surgical procedures. Usually greater difficulty is experienced in venous line placement for such cases. Heavy premedication is contraindicated if there is any previous history of obstruction of the upper airway or there is suspicion of difficulty of intubation, especially if the venous line is to be secured before anesthesia.

Some authors have reported considerable difficulty with the venous access in achondroplastic patients (up to 50% failure rate), mainly in children, due to the laxity and excess of skin and subcutaneous tissue.[10] We did not experience the same failure rate. Our initial 30% failure rate was due more to the anxiety of the patients and lack of cooperation than to the excess of skin and subcutaneous tissue. As the experience of our group increased, the rate of failure fell below 10%, because (1) we waited for an adequate premedication effect, (2) our series contained a larger number of teenagers and young adults who were more cooperative than children at the time of the venous puncture, and (3) those patients younger than 10 years of age and teenagers who so desired received inhalation anesthetic induction, which, for several reasons (e.g., immobility, vasodilation) made venous puncture easier. Our experience showed that, except in the young adults, patients who had previously received anesthesia asked for inhalation induction and the venous puncture was carried out once they were asleep. Although venous cannulation has been recommended before induction to prevent any difficulty in ventilation or intubation,[16] we think that inhalation induction of the achondroplastic patient can be carried out prior to venous cannulation if (1) there is no suspected difficulty of intubation, (2) there is no history of sleep apnea and/or obstruction of the upper airway, (3) there are no symptoms of cervical spinal cord compression, and (4) the anesthesiologist relies on the help of qualified personnel who can carry out the venous puncture while the patient ventilates spontaneously.

For inhalation induction, we chose halothane because of its lower irritability on the airway; we used isoflurane for anesthesia maintenance due to its better pharmacokinetic profile. Sevoflurane was not available to us but it could be an alternative choice both for induction and maintenance. None of our patients required the insertion of a central venous line, and we have no data to compare with other published studies.[4,10]

Regional anesthesia was not used in our patients because of the technical difficulties, the potential for development of neurologic problems, patient refusal, and the unpredictability of anesthesia level. Regional anesthesia has been used successfully in adult achondroplastic patients for cesarean section without apparent neurologic sequelae.[17]

The complications found in the postoperative period are not specific to achondroplastic patients. Two cases of postextubation laryngospasms, which are frequent in pediatric population, had a fast recovery. The one case of a postoperative vomiting was not aspirated and the patient did not require treatment. The case of cutaneous rash was due to an analgesic drug and required no treatment. Characteristic aspects of the analgesic treatment in these patients are their lower tolerance (children or teenagers) and their special psychological profile (overprotected children with possible previous negative experiences). We think it is very useful to administer nonsteroidal antiinflammatory drugs before induction and to provide the patient with a greater psychological support.

No patient presented respiratory complications, even though there is a reported high incidence of sleep apnea in achondroplastic dwarfs.[15]

In conclusion, we report an anesthetic experience of 8 years in 15 achondroplastic patients older than 3 years, undergoing 53 orthopedic surgical procedures. Although the characteristic deformities of these patients can impede management of anesthesia, only one patient presented a difficult intubation. Several anesthetic drugs and techniques were used, and no adverse effects could be identified for any particular drug or technique. A preanesthetic study to avoid difficulties of the airway and/or compression of the cervical spinal cord is important. It is very useful to administer an adequate premedication to reduce anxiety and encourage cooperation during anesthetic induction in patients who undergo multiple interventions, if they are not contraindicated. No other specific optimal anesthetic regimen can be recommended.

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