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#### **Case report**

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### Walter Osorio Rudas<sup>a,\*</sup>, Nury Isabel Socha García<sup>a</sup>, Alejandro Upegui<sup>a</sup>, Ángela Ríos Medina<sup>b</sup>, Adrian Moran<sup>c</sup>, Oscar Aguirre Ospina<sup>b</sup>, Carlos Rivera<sup>d</sup>

<sup>a</sup> MD, Specialist in Anesthesiology and Resuscitation, Unidad Materno infantil, Clínica Universitaria Pontifica Bolivariana, Medellín, Colombia

<sup>b</sup> MD, Specialist in Anesthesiology and Resuscitation, Universidad de Caldas, Manizales, Colombia

<sup>c</sup> MD, Resident of Anesthesiology and Resuscitation, Universidad de la Sabana, Bogotá, Colombia

<sup>d</sup> MD, Resident of Anesthesiology and Resuscitation, Universidad Sur Colombiana, Huila, Colombia

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#### ABSTRACT

*Introduction:* Cesarean section under general anesthesia is recommended in achondroplastic pregnant patients; however, the use of conductive techniques has been recently reported, with acceptable results.

*Objective:* To describe the anesthesia management in an achondroplastic patient scheduled for C-section under combined spinal–epidural anesthesia.

Methods and results: We present the case of a first pregnancy in a patient with achondroplasia, height 110 cm and 37 weeks of gestation. The patient underwent cesarean section under ultrasound-guided conductive anesthesia, using a titrated mixture of local anesthetic and opiate, with good results for the mother and child.

Conclusions: Conductive anesthesia is an option in C-section in patients with achondroplasia. Although there are no clear recommendations to guide a safe access to the neuroaxis or to administer anesthetic agents at this level, ultrasound and the titrated administration of neuraxial drugs (epidural, epidural-spinal and continuous spinal) for improved safety and efficacy of the technique in this type of patients may be considered.

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#### Anestesia para cesárea en paciente con acondroplasia

#### RESUMEN

Introducción: En gestantes acondroplásicas se recomienda el parto por cesárea con anestesia general; sin embargo, recientemente se ha reportado el uso de técnicas conductivas con resultados adecuados.

*Objetivo:* Describir el manejo anestésico de una paciente con acondroplasia programada para cesárea utilizando anestesia combinada espinal-epidural.

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\* Corresponding author at: Ctra 82 A No. 32 B 147, Casa 110, Medellín, Colombia. E-mail address: walterosorio1@gmail.com (W. Osorio Rudas).

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Palabras clave:

Anestesia Anestesia obstétrica Cesárea Acondroplasia Métodos y resultados: Mostramos el caso de una primigestante acondroplásica con 110 cm de estatura y embarazo de 37 semanas, en quien se realizó cesárea con anestesia conductiva guiada por ecografía, empleando una mezcla titulada de anestésico local y opiáceo, con buenos resultados para la madre y el hijo.

Conclusiones: La anestesia conductiva es una alternativa en la cesárea de pacientes con acondroplasia. Aunque no existen recomendaciones claras para orientar el acceso seguro al neuroeje ni para administrar medicamentos anestésicos a este nivel, se puede considerar el uso de la ecografía y la administración titulada de fármacos neuroaxiales (epidural, epidural-espinal y espinal continua) para mejorar la seguridad y la eficacia de la técnica en este tipo de pacientes.

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#### **Case description**

First pregnancy in an achondroplastic 29-year old with 37 weeks of gestation was scheduled for Cesarean section. Personal history of acid-peptic disease and ultrasound finding of fetus with achondroplasia was given; the results of the physical examination were: height 110 cm (see Fig. 1), blood pressure (BP): 120/70, heart rate (HR): 75 min, weight 41 kg and BMI: 33,9: short limbs and thoracolumbar kyphoscoliosis. No



Fig. 1 - Patient with short stature.

predictors of difficult airway or cardiopulmonary/neurological findings.

The patient requested to be awake during the birth of the baby and then discuss the risks and benefits of the anesthetic strategy; a combined spinal-epidural technique is considered.

Prior to administering the anesthesia, the patient received 50 mg of ranitidine and 10 mg of metoclopramide IV under basic ASA monitoring, including preparation of a difficult airway kit.

With the patient in sitting position, following ultrasound measurement, the epidural space L2–L3 is identified at 3.5 cm from the skin; using a tuohy #18 needle and continuous loss of resistance with saline solution, the epidural space was accessed free of complications. A multi-orifice epidural #20 catheter was introduced and fixed to the skin. The catheter aspiration test was negative and the decision is made not to perform a test dose.

After three Whitacre<sup>TM</sup> 25-needle taps the subarachnoid space in L3–L4 was identified. 5 mg of hyperbaric bupivacaine with 64 µg of morphine and 16 µg of fentanyl were injected. The patient did not experience any numbness during the procedure. Sensory block was verified up to T4. BP at 5 min was 80/40 and HR 72/min. 8 mg of etilephrine were required to maintain the systolic BP > 90 mmHg and the mean BP > 50 mmHg. The patient experienced discomfort during peritoneal traction and hence 60 mg of epidural lidocaine without epinephrine were administered and a slow IV bolus of 48 µg of remifentanyl. The newborn had an apgar score of 8 at one minute and 10 at 5 min (see Fig. 2). In total, 1000 cc of crystalloids were administered and the procedure was completed free from complications.



Fig. 2 - Vigorous newborn.



Fig. 3 – Typical abnormalities of the thoracic and lumbar spine in achondroplasia.

#### Discussion

Dwarfism is defined as the failure to reach a stature over  $148 \,\mathrm{cm^1}$  and achondroplasia is the most frequent cause.<sup>2</sup> This is a hereditary bone metabolism disorder with a prevalence of 1/26,000 live births.<sup>3</sup> It is an autosomal dominant transmission<sup>4</sup> and in 80% of the cases is a spontaneous genetic mutation.<sup>5</sup>

Achondroplasia is characterized by short stature, macrocephalus with prominent forehead, depressed nasal bridge, protuberant buttocks and abdomen, short limbs, particularly the proximal segment, lumbar hyperlordosis and thoracic kyphoscoliosis<sup>6</sup> (see Fig. 3). Symptomatic spinal stenosis usually does not manifest itself before the fourth or fifth decade of life, when osteophytes, kyphosis, scoliosis and disc hernias cause additional narrowing of the spinal canal.<sup>7</sup>

Women are often affected and have low fertility rates.<sup>8</sup> Delivery is usually through Cesarean section because of the cephalopelvic disproportion.<sup>9</sup>

The choice of anesthesia for C-section in achondroplastic patients has been controversial,<sup>10</sup> but general anesthesia is usually preferred,<sup>11</sup> though it is particularly challenging since the anatomic alterations further compound the typical morphological changes during pregnancy.<sup>12</sup>

General anesthesia presents specific risks such as: tendency to obstruct the airway; cervical instability<sup>13</sup> and difficulties for direct laryngoscopy as a result of rigid temporomadibular joints, macroglossia and pharyngeal stenosis.<sup>14</sup> The passage of the endotracheal tube may be difficult and there is a need to choose a small tube, in accordance with the weight of the patient rather than age.<sup>13</sup> Odontoid process dysplasia is a frequent finding and is accompanied in some cases by atlanto-axial instability and bone marrow compression that may deteriorate with orotracheal intubation maneuvers.<sup>15</sup> Additionally, dystrophy and thoracic kyphoscoliosis predispose for the development of restrictive lung disease.<sup>16</sup> Heart abnormalities predispose to ischemic events.<sup>17</sup>

Subarachnoidal anesthesia may prove technically challenging and its distribution may be inappropriate due to lumbar hyperlordosis, marked thoracic kyphoscoliosis, progressive narrowing of the inter-peduncular space and generalized stenosis of the spinal or epidural space.<sup>18</sup> Some authors do not recommend this approach to prevent blaming the anesthetic technique for any neurological abnormality caused by the spinal deformity.<sup>19</sup>

There are no literature reports of neurologic injury in achondroplastic patients with conductive anesthesia; however, the risk is undeniable and any neurological disorders must be appropriately registered in the clinical record and post-block evaluations should be performed for early detection.

Whichever the technique, aorta-caval compression may be severe<sup>20</sup> resulting in hypotension that requires uterine displacement, fluids co-load and rapid administration of vasopressants.<sup>21</sup> Short limbs and obesity may hinder non-invasive BP measurement and hence occasionally intraarterial measurements are needed.<sup>22</sup>

Due to existing reports of failed subarachnoidal anesthesia for the management of Cesarean sections in patients with achondroplasia,<sup>23</sup> excellent results were achieved with the use of a combined spinal–epidural technique to enable epidural titration of the anesthetic agent if needed.<sup>22</sup>

In sum, we think that conductive anesthesia is an option for the management of achondroplastic patients undergoing Cesarean section; however, in the light of inter-individual anatomic variations of the spine, ultrasound guided lumbar tap should be considered to facilitate the approach and reduce the risk of neurological complications. Likewise, neuraxial titration of local anesthetic agents with techniques such as epidural, combined spinal–epidural or continuous spinal is needed. Finally, even when selecting regional techniques, do not ever rule out the possibility of general anesthesia and always be prepared for difficult airway management.

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Own resources.

#### **Conflict of interests**

None

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