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### Case report

## Anesthesia for bariatric surgery in a patient with Prader-Willi syndrome: Case report<sup>☆</sup>



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#### ABSTRACT

Prader-Willi syndrome is a genetic disorder characterized by hypotonia, obesity, short stature, mental retardation, hyperphagia, hypogonadism and low life expectancy.

We describe the case of a 31-year-old female patient with Prader-Willi syndrome scheduled for bariatric surgery. Anesthetic considerations are reviewed highlighting perioperative complications associated with this syndrome.

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### Anestesia para cirugía bariátrica en paciente con síndrome de Prader-Willy: reporte de un caso

#### RESUMEN

El síndrome de Prader-Willi es un desorden genético caracterizado por hipotonía, obesidad, baja estatura, retraso mental, hiperfagia, hipogonadismo y expectativa de vida reducida.

Describimos el caso de una paciente de 31 años con antecedente de síndrome de Prader-Willi, programada para realización de cirugía bariátrica. Se revisan las consideraciones anestésicas, haciendo énfasis en las complicaciones perioperatorias secundarias a este síndrome.

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## Introduction

Prader-Willi syndrome is a genetic disorder characterized by hypotonia, obesity, short stature, mental retardation, hyperphagia, hypogonadism and low life expectancy.<sup>1</sup>

It was described initially by JL Down in 1887 in a patient diagnosed with "polysarcia". In 1956, Prader, Labhart and Willi described nine other cases and gave the syndrome its name. In 1980, Ledbetter discovered the existence of a microdeletion of the 15q11-q13 region and three years later, Butler and Nichols observed this genomic imprinting in patients with Prader-Willi syndrome.<sup>2</sup>

There are few reported cases in the literature on the anesthetic management of adult patients with Prader-Willi syndrome with low life expectancy as a consequence of morbid obesity.<sup>3-5</sup> Furthermore, these patients are predisposed to sudden death from respiratory diseases or in the postoperative period.<sup>6-10</sup>

We present the anesthetic management of a 31-year-old patient diagnosed with Prader-Willi syndrome who was scheduled to undergo bariatric surgery.

## Case description

The patient was admitted to surgery at the Manuel Uribe Ángel Hospital in Envigado, Antioquia, Colombia. The patient was 31 years old and female with a background of Prader-Willi syndrome associated with hypertension, extreme obesity (BMI 54), mental retardation, hypothyroidism and obstructive sleep apnea syndrome. She was admitted to undergo a gastric bypass.

The patient is considered to be a NYHA functional class III, ASA 4.

The physical exam revealed central cyanosis and blood pressure of 140/80 mmHg, heart rate of 68 bpm, respiratory rate of 19 breaths/min, Mallampati score of II, oral aperture above 4 cm, cervical perimeter below 40 cm, 35° extension of the upper neck and thyromental distance above 6 cm.

Rhythmic cardiac sounds were auscultated, revealing P2 > A2 along with a protomesosystolic murmur at the pulmonary and tricuspid focuses, III/VI. There was a diminished bilateral vesicular murmur.

Oxygen saturation oscillated between 80 and 90% through the nasal canal at 3 l/min.

The patient did not resist any of the examiner's maneuvers and collaborated during the whole evaluation.

Presurgical exams revealed: hemoglobin 12.2 g/dl, hematocrite 41%, 371 000 platelets/mL, TSH 7.92 mIU/L, HA1G 6.75%, arterial blood gases (FiO<sub>2</sub>) 0.21: pH 7.38, PCO<sub>2</sub> 54 mmHg, PO<sub>2</sub> 25 mmHg, HCO<sub>3</sub> 29 mEq/l. Echocardiogram (TTE): FE: 60%, PSAP 52 mmHg. There were slightly insufficient tricuspid and pulmonary valves, but normal cavity size.

In the operating room, a 16G cannula was placed on 2 cephalic veins; a rapid sequence induction was performed with previous oxygenation during 5 min until a maximum saturation of 95% was reached, followed by a 100 mcg bolus intravenous administration of remifentanyl, 60 mg lidocaine, 150 mg propofol, 100 mg succinylcholine, laryngoscopy with

valve curve #3 (Cormack II visualization), introduction of 8.0 mm orotracheal tube, maintenance with 3% sevoflurane, flow of oxygen gases at 0.3 l/min and air at 0.5 l/min, and finally an intravenous infusion of remifentanyl at 0.1 mcg/kg/min.

The anesthesia machine was programmed for volume, with a tidal volume of 7 ml/kg, PEEP 5 cm H<sub>2</sub>O, showing peak pressures higher than 35 cm H<sub>2</sub>O with plateau pressures lower than 30 cm H<sub>2</sub>O. The bilateral position of the orotracheal tube was immediately verified through auscultation to rule out obstruction or kinking. The monitor showed a progressive increase in the level of CO<sub>2</sub> at the end of the expiration without presenting inclination in the curve of the capnogram in phase 2. 6 mg cisatracurium was administered intravenously, with significant improvement in the respective values.

During the intraoperative period the surgical team identified severe cardiomegaly associated with hepatomegaly, which led to the decision to perform gastric sleeve surgery to then proceed with a bypass in the second surgical period.

At the end of the procedure, 0.6 mg of hydromorphone was administered intravenously.

There were no episodes of desaturation, severe hypotension/hypertension or arrhythmias. She was extubated after finishing surgery and transferred to the special care unit and supplemented with 50% oxygen at 10 l/min with the Ventury system. No reintubation was required.

## Discussion

Prader-Willi syndrome is considered to be the leading cause of obesity associated with genetic syndromes and has a prevalence of approximately 1/25000.<sup>11</sup> The partial deletion of the long arm of the paternal chromosome 15 is a common marker of this disease,<sup>12</sup> though it can also present as a maternal uniparental disomy.<sup>13</sup>

The annual mortality rate was 3% for all ages, but the rate increases to 7% over the age of 30.<sup>11</sup> Patients generally suffer an early death due to secondary complications related to obesity, such as diabetes mellitus, hypertension, obstructive apnea-hypopnea, cardiovascular disease and respiratory failure.<sup>14</sup>

The clinical course for Prader-Willi syndrome is usually divided in two phases.<sup>15</sup> The first phase occurs during the neonatal and lactation periods and is characterized by marked hypotonia, difficulties with suction, persistent cough, crying and episodes of asphyxia.<sup>16,17</sup> The second phase, between the ages of 2-5, is characterized by hypogonadism, mental retardation and obesity related to hyperphagia, most likely due to a hypothalamic defect in the satiety center<sup>18</sup> or an innate failure to metabolize lipids and carbohydrates.<sup>19</sup>

As was previously mentioned, there are few available reports on the anesthetic management of patients with Prader-Willi syndrome, and in the case at hand the patient who was scheduled for a low-risk cardiovascular procedure was in her fourth decade of life with extreme obesity, obstructive sleep apnea syndrome and chronic hypoxia.

Alterations in body temperature (hyperthermia and hypothermia), intraoperative arrhythmias, cor pulmonale and clinical consequences of obesity (decrease in functional

residual capacity) are particular problems during the perioperative period.<sup>20-24</sup>

During the intraoperative period the only difficulty with our patient was the elevated peak pressure values accompanied by normal plateau pressure levels. An elevated peak pressure can be accompanied by a concomitant elevation of plateau pressure, but when this association is not present, we are faced with an increase in the resistance of the airways (decrease of dynamic pulmonary distensibility), causing us to rule out bronchospasms, presence of secretions, obstruction, kinking or biting of the orotracheal tube.<sup>25</sup>

It should be noted that excessive intra-abdominal pressure decreases the pulmonary volumes in morbidly obese patients under sedation or anesthesia, notably in terms of residual functional capacity, while the alveolo-arterial oxygenation gradient is increased along with respiratory resistance.<sup>26</sup> Our patient's condition improved with the administration of a neuromuscular blocker. This technique is similar to that performed in patients with intra-abdominal hypertension syndrome, in which neuromuscular blockers are used to lower intra-abdominal and airway pressure.<sup>27</sup>

There was also no difficulty in performing the laryngoscopy, despite reports of difficult airways in these patients.<sup>28</sup>

A transfer to the special care unit was considered since these patients have an increased risk of postoperative hypoxia attributed to the altered response in consciousness level due to changes in oxygen and CO<sub>2</sub> in blood pressure.<sup>29</sup>

Another issue is the documented presence of an oppositional and aggressive personality type associated with this syndrome,<sup>30</sup> although in our case the patient cooperated at all times during her hospital stay and did not require sedatives such as benzodiazepine which can possibly cause episodes of desaturation and apnea.

In conclusion, the approach that an anesthesiologist takes with a Prader-Willi patient should include all the associated comorbidities, especially secondary ones like morbid obesity, taking into account the appropriate postoperative monitoring to avoid the onset of respiratory complications.

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## Conflict of interest

None declared.

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