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CASE REPORT

## Anesthetic management for non-cardiac surgery in a patient with Fontan palliation: case report

### Manejo anestésico para cirugía no cardíaca en paciente con paliación de Fontán: Reporte de caso

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#### Abstract

Survival of patients with Fontan palliation has improved significantly over the years and it constitutes a condition frequently found nowadays in the perioperative setting for non-cardiac surgery. A history of this disease condition implies complex physiologic and multiorgan considerations for the anesthetist who may need to resort to special measures in order to maintain homeostasis and avoid complications. In this paper we present the case of a patient with a history of Fontan, and describe successful anesthetic management during laparoscopic surgery.

#### Resumen

En los últimos años la sobrevida de pacientes con paliación de Fontán ha aumentado significativamente, y hoy es una condición frecuente en el escenario perioperatorio para cirugía no cardíaca. Este antecedente supone complejas consideraciones fisiológicas y multiorgánicas para el anestesiólogo, quien puede requerir medidas especiales para mantener la homeostasia y evitar complicaciones. En este artículo presentamos el caso de un paciente con antecedente de Fontán y describimos el manejo anestésico exitoso en cirugía laparoscópica.

#### Introduction

Since its introduction in 1971, the Fontan procedure has been used as palliative surgery in children with complex congenital heart diseases with a single physiological ventricle, unsuitable for biventricular repair, as is the case with tricuspid atresia.<sup>1</sup> It initially consisted of right pulmonary artery anastomosis to the right atrium in order to enable blood to pass directly to the pulmonary circulation from systemic venous return following closure of the atrial septal defect.<sup>2-4</sup>

Since it was first applied, several modifications have been made over the past decades. In particular, the atrial-pulmonary anastomosis has been replaced with a total cava-pulmonary artery anastomosis, which is made by creating an atrial-pulmonary tunnel or an extracardiac tunnel in order to establish a direct connection between the inferior vena cava and the right pulmonary artery.<sup>3</sup> This has significantly improved prognosis and patient survival, with a lower occurrence of arrhythmias, probably delaying the onset of cardiac failure,<sup>5,6</sup> and allowing individuals with congenital heart diseases to reach adult life. As a result, these patients are found with increasing

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frequency in perioperative settings for non-cardiac procedures.<sup>1</sup>

### Case presentation

We present the case of a 24-year-old male patient scheduled for elective laparoscopic cholecystectomy due to stone disease. The patient was asymptomatic and had a history of Fontan palliation due to tricuspid atresia at birth, performed in 2 stages at 2 weeks and at 2 years of age, consisting in the end of a total cava-to-pulmonary artery anastomosis with extracardiac conduit. Since that time, the patient has been asymptomatic, practices sports with no limitation, is a functional class New York Heart Association I, American Society of Anesthesiologists (ASA) classification 3, and has no other surgical history beside the one associated with the underlying heart disease. However, he reports a significant history of intraoperative recall during the last surgery. He is receiving enalapril 10mg/día, spironolactone 25mg/día, and acetyl salicylic acid 100mg/día. He does not smoke, drinks alcohol occasionally, and has no known allergies.

On physical examination, the patient was in good general condition, weight 55kg and height 155cm, no difficult airway predictors, hydrated mucosas, blood pressure within normal ranges in the 2 arms, arterial oxygen saturation 88%, heart rate 65bpm, and respiratory rate 18bpm. Cardiopulmonary exploration was normal, with no murmurs, but distal cyanosis and clubbing of fingers, and normal capillary filling. There was mild tenderness in right-upper quadrant on palpation, but no other positive signs on physical examination.

The following were the findings during the pre-operative assessment: hemoglobin 20.6mg/dL; hematocrit 66.1%; red blood cell count 7.05 mill/ $\mu$ L; leukocytes 4970/ $\mu$ L; platelets 127,000/ $\mu$ L; creatinine 0.77mg/dL; total bilirubin 3.65mg/dL; direct bilirubin 0.6mg/dL; alkaline phosphatase: 91IU/L; International Normalized Ratio 1.29; blood gases pH 7.4; PCO<sub>2</sub> 40.30mm Hg; PO<sub>2</sub> 44.80mm Hg; HCO<sub>3</sub> 25.00mmol/L; excess base 0.3mmol/L, oxygen blood pressure / inspired oxygen fraction 312.33. The electrocardiogram showed sinus rhythm and left anterosuperior hemiblockade. Echocardiography showed ventricular ejection fraction of 69%, left ventricular diastolic dysfunction, inferior vena cava and hepatic vein dilatation, and normal flow through the cava-pulmonary artery shunt and pulmonary arteries. Spirometry showed mild obstructive pattern and significant response to bronchodilation, with 77% and 89% left ventricular ejection fraction before and after bronchodilation, respectively. The chest X-ray was normal, showing a small cardiac silhouette.

Anti-platelet aggregation was interrupted 15 days before. Mechanical anti-thrombotic measures were implemented during surgery, 400mL of blood were removed for hemodilution, and 200mL of Hartmann's solution were infused 2 hours before the initiation of anesthesia. Basic

ASA monitoring was instituted, including pulse oximetry, cardioscopy, capnography, and temperature, together with left radial catheter insertion connected to a Vigileo monitor. Bispectral analyzer (BIS) for depth of anesthesia and train-of four (TOF) for neuromuscular blockade monitoring were used.

Total intravenous general anesthesia was administered; 2mg of Midazolam were used for pre-medication, and 2 infusion pumps were prepared for target controlled infusion (TCI), one with propofol at a concentration of 10 $\mu$ g/mL and the second one with remifentanyl at 40 $\mu$ g/mL. Cisatracurium at a dose of 5mg was used for muscle relaxation. Complication-free orotracheal intubation Cormack I was accomplished readily on first attempt, with # 8 tube, 24cm from the labial commissure. Propofol and remifentanyl doses for central effect were 4 $\mu$ g/mL and 3ng/mL, respectively, during induction, followed by a constant dose of 2 $\mu$ g/mL of propofol, while the same dose of remifentanyl was maintained until the end of the surgery. Anesthetic depth was maintained at a BIS between 43 and 47.

In terms of ventilation parameters, volume controlled mechanical ventilation was maintained with maximum airway pressure at 19mm Hg and positive end-expiratory pressure at 3mm Hg. Hemodynamically, cardiac output was maintained at 2.4, 4.8, and 5.1L/min, systolic volume variability at 9%, 12%, and 8%, and systolic volume at 47, 93, and 87 mL at the start, during and at the end of surgery, respectively.

The surgical procedure was completed successfully with pneumoperitoneum time of less than 30 minutes and intra-abdominal pressure not higher than 10mm Hg, minimum blood loss, and no complications. Multimodal analgesia was administered using 40mg of paracoxib, 0.6mg of hydromorphone and 1mg of dipirone at the end of the procedure.

On leaving the operating room, the patient was alert, extubated, with ventilation patterns and hemodynamic status within normal ranges, and adequate pain control. He was transferred to the intensive care unit for 24-hour monitoring. Thromboprophylaxis with enoxaparin 40mg/day was initiated, and oral fluid tolerance was tested 6 hours after surgery, with no complications. The next day, the course was uneventful, with no fever or vomiting, and adequate urine output. The patient was then transferred to the special care unit and later to the ward to continue under follow-up by the surgical service and pain management, and was then discharged on the fifth postoperative day after full recovery, adequate pain control, and no complications.

### Discussion

In patients with Fontan, there are 2 fundamental physiological considerations: (1) the presence of a single physiological ventricle, and (2) the need to understand

that the entire systemic venous return reaches the pulmonary arterial circulation passively, depending on central venous pressure (CVP), which is in turn determined by pulmonary vascular resistance (PVR) and venous system capacitance.<sup>1,3</sup> Pulmonary blood flow (PBF) is a determining factor for cardiac output because the systemic circulation is connected in series with the pulmonary circulation, without right ventricular involvement.<sup>7</sup> Consequently, these patients are characterized by increased CVP, low cardiac output,<sup>8</sup> and a slight reduction in arterial oxygen saturation, for which they develop adaptations such as increased arterial vascular resistance, cardiac output redistribution to vital organs, and increase in hemoglobin.<sup>9</sup> In our patient, despite adequate ejection fraction, hemoglobin, and hematocrit hemoconcentration is present as a compensatory mechanism for chronic hypoxemia.

Other parameters to consider are systolic function status, diastolic compliance of the single ventricle, and adequate valve system function, given that the ability to increase cardiac output is limited as a result of the small reserve to increase oxygen uptake and ventricular preload.<sup>3</sup>

PVR is determined by mechanical factors and biochemical mediators that produce vasoconstriction and vasodilation; that is why excess positive pressure results in alveolar overdistension, reducing capillary radius and, consequently, lowering PBF. At the same time, atelectasis causes hypoxic capillary vasoconstriction, contributing to the increase in PVR and pulmonary hypertension.<sup>1</sup>

Ideally, optimal transpulmonary gradient should be maintained in order to preserve PBF, with low pulmonary resistance and adequate CVP. For this reason, we avoided the use of catecholamines as much as possible, administered 100% oxygen, and provided fluids in order to maintain preload and systemic pressure. Some authors recommend the use of colloids,<sup>3</sup> but we decided to use Hartmann's solution, with adequate response and no major postoperative complications, always taking into account ventricular ejection fraction. Preoperative hemodilution is an anti-thrombotic measure used to compensate for anti-aggregation interruption 15 days before surgery, as a protocol to reduce the risk of bleeding, trying to maintain hemoglobin above 10 g/dL, besides the use of mechanical compression of the lower limbs.

Positive pressure ventilation must be limited as much as possible, and surgery must be performed in the shortest time possible, considering that spontaneous ventilation creates lower intra-thoracic pressure, promoting PBF. The normal effects of pneumoperitoneum include an increase in intra-abdominal pressure and diaphragm elevation, resulting in lower lung compliance, increased airway resistance, and the risk of atelectasis. Added to hypercapnia due to CO<sub>2</sub> inflation, this creates an imbalance in ventilation/perfusion ratio, with secondary hypoxemia. Increased pressure in the abdominal cavity compresses

the large intra-abdominal vessels, resulting in a drop in venous return and lower preload and cardiac output. Aortic compression induces the release of neurohumoral factors and activates the renin-angiotensin-aldosterone system, resulting in increased systemic vascular resistance (SVR) and diminished myocardial contractility.<sup>10</sup> In this case, we believed it was appropriate to maintain pneumoperitoneum pressure under 10 mm Hg<sup>1,3</sup> together with Fowler's position in order to offset pulmonary restriction, and we used low tidal volume and high respiratory rate ventilation in order to maintain normocapnia. All these measures were designed to preserve adequate preload, given that patients with a single ventricle depend on it to maintain cardiac output.

Regarding intraoperative monitoring, basic ASA and cardioscopy are enough, and in patients with pneumoperitoneum, direct arterial pressure monitoring is ideal. In this case, we used a left radial catheter connected to a Vigileo monitoring system which uses the patient's own arterial pressure for continuous cardiac output measurements, taking into account variables such as weight, height, age, and gender.<sup>11</sup> This system has the advantage of being minimally invasive, measuring key flow parameters like systolic volume, SVR, SVR index, cardiac output and cardiac output index, and not requiring manual calibration. In terms of disadvantages, manipulation of the arterial line may induce changes, and the measurement is altered in cases of severe arrhythmias, spontaneous ventilation, and open chest.<sup>12</sup>

Central catheter insertion is not recommended due to the risk of thrombosis, injury or Fontan infection, which would be fatal in these patients.<sup>1</sup> We considered general anesthesia to be the ideal technique. Conductive techniques are not recommended due to preload reduction from venous vasodilation and potential cardioaccelerator fiber blockade. Total intravenous anesthesia was selected because it favors improved modulation of the sympathetic response to stress, better hemodynamic control, a lower incidence of postoperative nausea and vomiting<sup>13</sup> and tendency to low heart rate, all of which were desirable in our patient in order to maintain sufficient diastole to preserve ventricular cardiac output.<sup>14</sup>

BIS and TOF are useful for guiding the depth of anesthesia and neuromuscular relaxation, respectively. TCI infusions of remifentanyl and propofol are also advisable in order to use the minimum required dose of anesthetics, avoid sharp drops in SVR, and preserve adequate coronary perfusion to maintain sufficient ventricular systolic and diastolic function. Likewise, it is important to maintain sinus rhythm and identify any type of arrhythmia, to which these patients are prone.

Postoperatively, continuous monitoring is recommended for at least 24 hours in the intensive or special care unit; it is also recommended to maintain adequate ventilation and oxygen source. Adequate pain control is

crucial to avoid excess catecholamine release that may increase SVR and PVR and lead to hypoxia. Nausea and vomiting should be prevented, enteral nutrition should be reintroduced as soon as possible, anti-coagulation must be defined, and the patient must be ambulated.

## Conclusion

Fontan patients have a unique physiology, posing a challenge for anesthetists during any type of surgical procedure. Preserving PBF is a priority, which means avoiding hypoxemia, high airway pressures, the use of agents that increase PVR, and ensuring adequate analgesia. The choice of the anesthetic technique must be driven by the goal of maintaining normovolemia and preserving adequate cardiac output and sinus rhythm.

## Ethical responsibilities

**Human and animal protection.** The authors declare that the adopted procedures were in agreement with the ethical standards of the Committee for responsible human experimentation, the World Medical Association and the Declaration of Helsinki.

**Data confidentiality.** The authors declare having followed the protocols of their institution regarding patient data disclosure.

**Right to privacy and informed consent.** The authors obtained the informed consent of the patient and/or subject to whom this article makes reference. The document is in the hands of the corresponding author.

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## Conflicts of interest

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