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CLINICAL INFORMATION

Anesthesia for surgical repair of the pentalogy of Cantrell: case report☆

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KEYWORDS
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Abstract
Pentalogy of Cantrell is a congenital anomaly associated with defects in the abdominal wall, sternum, diaphragm, and diaphragmatic pericardium formation, in addition to the development of cardiac abnormalities. It is a rare disease with an estimated incidence of one case for every 65,000 births, being more common in males (60% of cases). It has a reserved prognosis with mortality around 63%, and a maximum of 9 months survival after surgery. There are few case reports addressing the pentalogy of Cantrell, which is justified by the rarity of this pathology. In this report our objective was to describe a surgical case of a female patient and make some anesthetic considerations about this rare congenital malformation.

PALAVRAS-CHAVE
Pentalogia de Cantrell;
Malformação congênita;
Doença rara

Anestesia em cirurgia para correção de pentalogia de Cantrell: relato de caso

Resumo
A pentalogia de Cantrell é uma anomalia congênita associada a defeitos na formação da parede abdominal, do esterno, diafragma e pericárdio diafragmático, além do desenvolvimento de anomalias cardíacas. É uma doença rara, com incidência estimada em um caso para cada 65.000 nascimentos, mais comum no sexo masculino (60% dos casos). Apresenta

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prognóstico reservado com mortalidade em torno de 63% e sobrevida após procedimento cirúrgico de no máximo nove meses. São escassos os relatos de casos referentes à pentalogia de Cantrell, o que se justifica pela raridade dessa patologia. Com este relato, os autores objetivam descrever um caso cirúrgico, em paciente do sexo feminino, e tecer algumas considerações anestésicas sobre essa malformação congênita rara.

Introduction

Pentalogy of Cantrell is a congenital malformation characterized by defects involving the abdominal wall, lower part of the sternum, anterior region of the diaphragm, and diaphragmatic pericardium, as well as cardiac abnormalities that vary in degree and severity, with ventricular septal defect being the most prevalent. In addition to ventricular septal defect, ventricular diverticula, pulmonary artery stenosis or atresia, dextrocardia, anomalous left superior vena cava, anomalous pulmonary venous return, truncus arteriosus, ventricular aneurysm, and tetralogy of Fallot.[1,2]

It was first described in 1958, due to the failure of lateral mesoderm development, which usually occurs between the 14th and 18th day of embryo development.[1] Consequently, there is failure in the development of the transverse septum and the contralateral mesoderm folds over the upper abdomen, does not migrate ventromedially, there are sternal and abdominal wall defects.[3] Some cases were associated with trisomy 18 and X-linked inheritance.[4]

It is a rare disease, with an estimated incidence of one case for every 65,000 births, more common in males (60%),[2] with only 90 cases recorded by 2004.[3] This malformation has a reserved prognosis, with mortality around 63%. Survival after cardiac, diaphragmatic and abdominal repair reaches a maximum of nine months.[2]

Surgical repair of this disease is complex due to the poorly developed structure of the rib cage and the difficulty of positioning the ectopic heart inside the chest. Surgical management should be as brief as possible, thus minimizing the risks of spontaneous rupture, arrhythmias, and thrombus formation in the present ventricular diverticulum.[5] Early correction of the abdominal defect reduces the risk of infection and fluid loss.[4]

Anesthesia for surgically treating the pentalogy of Cantrell is a major challenge, as the closure of each defect may severely compromise the function of the underlying organ. Hemodynamic and respiratory management requires attention and ability due to pulmonary immaturity, cardiac defects, and high abdominal pressure after the closure of abdominal and thoracic wall defects.[4]

There are few case reports concerning the pentalogy of Cantrell, which is justified by the rarity of this pathology. In an extensive literature review, no records of female patients undergoing anesthesia to repair this condition were found.

Thus, the purpose of this report was to describe a surgical case of a patient with the pentalogy of Cantrell and make some anesthetic considerations about this rare congenital malformation.

Case report

A 20-year-old pregnant woman, with two previous uneventful pregnancies, was admitted to the obstetrical service at 39 weeks and 4 days gestational age. At the 24th week of gestation, a fetus with omphalocele and ectopia cordis was diagnosed.

The newborn female weighed 2726 g, presented with 33 cm cephalic perimeter, 46 cm height, and Apgar 8 in both the first and fifth minutes. Ectoscopy revealed cyanosis, tachypnea, with presence of omphalocele, ectopia cordis, and lower sternum defect. She was admitted to the neonatal intensive care unit under invasive mechanical ventilation, received initial treatment with clinical and hemodynamic support, and was taken to the operating room after 10 h of birth for thoracoplasty and abdominoplasty.

The newborn was admitted to the operating room already intubated (3 mm non-cuffed tube) and with peripheral venous access in the right upper limb (Jelco® 24 G). Monitoring was performed with cardioscope, pulse oximetry, esophageal thermometer, and capnography. Induction of anesthesia was performed using midazolam 0.2 mg (0.1 $\mu g.kg^{-1}$), sufentanil 1.35 μg (0.5 $\mu g.kg^{-1}$), and cistracrium 0.4 mg (0.15 $mg.kg^{-1}$). Central venous access was obtained in the right femoral vein and invasive arterial pressure in the left radial artery with Arteriofix® 24 G.

Anesthesia was maintained with continuous infusion of 0.5 μg sufentanil (1 $\mu g.kg^{-1}.h^{-1}$) at an end-tidal sevoflurane concentration of 2%. Infusion of 50% glucose (5 $mg.kg^{-1}.min^{-1}$) and 0.9% saline solution: 12 $mL.kg^{-1}.h^{-1}$ (maintenance + surgical support). Mechanical ventilation was adjusted to a pressure-cycled mode with inspiratory pressure indexes of 14 cm H_2O, respiratory rate 30 breaths/min, PEEP 6 cm H_2O, FiO_2 80%, expiratory volume about 22 mL, inspiratory/expiratory time 1:1.5, and minute volume 0.7. Initially, expired fraction of carbonic gas was maintained between 40 and 50 mmHg and O_2 saturation at 80%–92%.

Surgical exploration confirmed the diagnostic hypothesis of pentalogy of Cantrell. Omphalocele contained the small intestine and liver. In addition, the following were observed:

anterior diaphragmatic defect, aplastic lower half of sternum, hypoplastic and defective pericardium, and caudal displacement of the heart.

During the surgical procedure, the heart was moved to the inside of the thorax, the pericardium was closed, and the ventral part of the diaphragm was rebuilt. The thoracic and abdominal cavities were separated.

The anterior abdominal wall defect was partially repaired by mobilization and minimal stretching of the abdominal muscles. A residual defect of about 10 cm transverse diameter was covered with a bovine pericardium patch (B 4012.5 × SJM Biocor® 10-pericardial patch).

Chest wall reconstruction was performed with metal rods (Kirschner wires) parallel to the edges of the incision, according to the Charles Bacellar's technique.

The procedure lasted 5 h. During the anesthetic approach, there were significant clinical changes in the hemodynamic parameters, with a drop in blood pressure, heart rate, oxygen saturation, and expired fraction of carbon dioxide. Continuous infusion of vasoactive drugs was used: dobutamine (15 $\mu g.kg^{-1}.min^{-1}$) and noradrenaline (1 $\mu g.kg^{-1}.min^{-1}$). Packed red blood cells (50 mL) were used for blood replacement. Mean blood pressure ranged from 40 to 55 mmHg, blood glucose from 70 to 125 $mg.dL^{-1}$, and hemoglobin from 8.9 to 12.1 $mg.dL^{-1}$. After such measurements there were improvements in the hemodynamic parameters of the patient, it was possible to reduce the doses of vasoactive drugs. Final fluid balance was negative in 18 mL, with total urine output of 6.75 mL.

The patient was taken intubated to Neonatal Intensive Care Unit, 90% saturated with 100% oxygen and hemodynamically stable at the expense of dobutamine (5 $\mu g.kg^{-1}.min^{-1}$) and noradrenaline (0.6 $\mu g.kg^{-1}.min^{-1}$). Despite high doses of noradrenaline, she evolved unsatisfactorily in the ICU with progressive hypotension, oliguria, and reduced saturation. Incremental doses of adrenaline were added to the infusion pump without adequate pressure improvement. She progressed with severe anuria and acidosis and had two cardiac arrest, the first one in ventricular fibrillation and the second in asystole. She died 48 h after the surgical procedure.

Discussion

Treating patients with pentalogy of Cantrell represents a great multidisciplinary challenge, involving anesthesiology, cardiovascular surgery, pediatric surgery, and neonatal intensive care. Survival is determined by the severity and complexity of cardiac malformations. In addition, we can state that the surgical procedure outcome and possible postoperative complications will define the patient's prognosis. A literature review showed that all patients who underwent surgery on the first day of life died.[2] Our patient showed similar results to other studies.[4]

Prenatal ultrasonography is an effective method to detect omphalocele and ectopia cordis; however, it does not allow an accurate description of the cardiac malformation.[2] Studies have shown that through fetal magnetic resonance along with prenatal echocardiography, it is possible to perform a detailed evaluation of the cardiac defect.[3] Therefore, a preoperative cardiovascular evaluation of these patients is recommended in order to plan the induction and maintenance of anesthesia.

Early postoperative mortality is more common in patients with complex cardiac malformations, or in those with postoperative complications, such as respiratory failure, intestinal dysmotility, pulmonary hypertension, and increased intra-abdominal pressure.[3,4] The congenital heart diseases more commonly found in the pentalogy of Cantrell are interventricular communication, atrial septal defect, pulmonary artery stenosis, and tetralogy of Fallot.[1]

Postoperative complications vary according to the duration of surgery and the material used to repair defects.[4,5] There are complications related to surgical repair with the use of screens, which was avoided in the diaphragmatic reconstruction in our patient.

Intraoperative hypoxia was probably linked to cardiac anatomical defects characteristic of the pentalogy of Cantrell. In addition, it has been reported that patients with this malformation commonly are born with pulmonary hypoplasia and that this fact is not related to compression by the abdominal organs, but to a preexisting genetic defect.[4] Therefore, the low oxygen saturation is a predictable finding in this pathology.

Patients with pentalogy of Cantrell have been shown to tolerate intra-abdominal pressure of up to 25–35 cm H_2O_2. On the other hand, they are vulnerable to hemodynamic instability after abdominal wall closure, as observed in our patient, and therefore require a differentiated attention regarding monitoring.

Abdominal malformation management should be planned according to its extension and type It has been reported that repair of the abdominal wall defect should be a late procedure due to the risks and difficulties of respiratory management. Patients undergoing early repair of omphalocele became more dependent on mechanical ventilation.[2]

In general, the prognosis is poor. In a case report with five patients, three survived but none had ectopia cordis classified as true.[1] Divkovic reported a 20% survival rate in his review.[2] In a study with 17 patients diagnosed during prenatal care there was no survivor.[3]

Pentalogy of Cantrell is a rare malformation. Anesthetic managements are decisive and will have repercussions on potential intraoperative complications, with special attention to hemodynamic instability associated with increased intra-abdominal pressure due to the primary closure of the abdominal wall defect. A comprehensive preoperative assessment, a multidisciplinary approach, and a special care with the anatomy and function of the involved organs are essential.

Intensive postoperative care is essential due to main postoperative complications, such as hypoxia resulting from pulmonary hypoplasia, hemodynamic changes resulting from congenital malformation surgical repair, and increased abdominal pressure after the wall closure. Although historically the result is discouraging, we should not allow the scarcity of studies to influence the negative prognosis associated with this disease.

Conflicts of interest

The authors declare no conflicts of interest.

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