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Anesthetic Management of an Omphalopagus Conjoined Twins Separation Surgery - A Case Report

Fernando Antônio da Silva Cifuentes[1*], Marina Ayres Delgado, Camila Biazussi Damasceno, Maíra Cota Torres, and Valéria Jorge Souza Castro

[1]Departamento de Anestesiologia do Hospital das Clínicas da Universidade Federal de Minas Gerais, Brazil

[*]Corresponding author: da Silva Cifuentes FA, Departamento de Anestesiologia do Hospital das Clínicas da Universidade Federal de Minas Gerais, Brazil; Tel: +5531993232016; E-mail: fascifuentes[AT]gmail.com

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Abstract

Conjoined twins are a rare congenital malformation associated to significant morbidity and mortality. In this article, we reported a successful anesthetic management and challenges related to the surgical separation of an omphalopagus conjoined twins. In this case, some aspects were fundamental to the positive outcome, such as a multidisciplinary approach and anticipation of complications: difficult intubation, massive blood loss, hemodynamic instability and hypothermia, for example.

Keywords: ***Anesthesia; Conjoined twins; Omphalopagus; Separation surgery.***

1. Introduction

Conjoined twins are monozygotic, monoamniotic, and monochorionic and their bodies are joined in utero [1]. The incidence of conjoined twins is around 1:200,000 live births, with higher incidence in Africa and South West Asia. The classification is based on the union site of their body. Thoracopagus is the most common type of conjoined twins and the site of union is the thorax [3]. Other types of conjoined twins are omphalopagus, pyopagus, and craniopagus [2]. Heart defects have been reported in one or both twins in approximately 66% of the cases [1].

The number of conjoined twins presenting for surgery is still low, and many infants do not survive very long [1]. Only 50% of the conjoined twins survive when undergoing separation surgery in the neonatal period [4].

In this article we describe a case report of the anesthetic management of an omphalopagus conjoined twins separation surgery.

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2. Case Report

Forty-one-day male conjoined twins were born from a 32 weeks primigravida by elective caesarean delivery. Their birth total weight was 3,740g. They were thoraco-omphalopagus twins, joined ventrally at epigastrium to umbilicus, facing each other (Fig. 1).

Fig. 1. Thoraco-omphalopagus twins. The ventral fusion ranges from thorax to umbilicus, making babies face each other.

Imaging evaluation of shared organ system were done with ultrasound and computerized tomography scan, revealing an isthmus-like part in the left and right lobe of each liver to be common between them with separate biliary and circulatory systems, along with a possible communication of pericardium.

A multidisciplinary team was formed and its members studied the case. The decision to perform the separation surgery and the necessary steps for the successful procedure took place, ranging from the preoperative preparation to the follow-up after hospital discharge. Intraoperative room was prepared with two anesthesia workstations and two multi-parameters monitors and a team of anesthesiologists was available for each baby (Fig. 2). A second fully equipped surgery room was promptly prepared to receive one twin after the separation to perform the closure of abdominal incision.

On the day of surgery, twins were admitted in the operating room after consent form has been signed by parents. Both patients had previous intravenous lines established. Blue and red identification were used to identify each one. Standard monitoring included pulse oximeter, cardioscopy and non-invasive blood pressure. Anesthesia was induced in one twin at the beginning with sevoflurane, fentanyl 1mcg/kg and rocuronium 0,6mg/kg; and oxygen was offered to the other via facemask. The same steps were made with the second twin and, when both airways were secured, capnography was established. The surgery team proceeded to guarantee invasive blood pressure and central venous line. Temperature probe and urinary catheter were also placed, and, to prevent hypothermia, warming mattresses and fluid warmers were used.

Fig. 2. Operating room layout. Two anesthesia workstations and two multi-parameters monitors are available. Red and blue tags are used to identify the equipment matching with patients, avoiding errors during anesthetic management.

An injection of atropine 0,10mg was administered in one of the babies and an increased heart rate was observed in the other baby, confirming the presence of cross-circulation. All drugs and intravenous fluids were calculated based on the total weight of the babies and half was given to each. Maintenance of anesthesia was carried out with sevoflurane, and both twins were ventilated on pressure control mode.

The separation surgery begun with the incision on the skin bridge connecting the babies, proceeding to the excision of common isthmus and livers separation. Then, the cardiovascular team explored the possible pericardium communication, with no shared important part found. The two babies were separated, and the abdominal walls were closed with synthetic material.

During surgery, continuous infusions of adrenaline were necessary to maintain hemodynamic control. At the end of the procedure, both of the babies went to the intensive care unit (ICU) intubated, with adrenaline infusion of 0,3μg/Kg/min. Extubation was performed in two days, and the babies discharged from ICU on the fifth day. They were kept in the hospital for more thirty days to proper surgical incision cares, leaving home with a designated team to maintain the following up at home.

3. Discussion

Most conjoined twins have multiple congenital abnormalities as well anatomic fusions [5]. Normally, if the new born twins are stable, gaining weight, it is usual and prudent to wait several months before planning separation, because the anatomy and physiology become more clearly defined with time [5]. Emergency separation may be necessary when one of the twins threatens the life of the other, when deterioration of both twins occurs due to hemodynamic instability and respiratory compromise, or when the condition of one of the twins is incompatible with life and the other has a good chance to survive [1].

Special ethical concerns exists for all of these cases of conjoined twins, especially when it involves unequal sharing of organs or limbs, or when separation induces death of one of the twins. In addition, surgical separation typically has high morbidity and mortality, and may lead to a lifelong disability of either twin due to the nature of the union [3].

To perform the anesthetic management, special resources are needed such as two separate anesthetic teams, two independent anesthetic workstations, ventilators, monitors, infusion pumps and temperature control devices [1].

Management of the airways of the conjoined twins can be a challenge to the anesthesiologist, as they can have distorted neck anatomy, limited cervical rotation and exaggerated cervical lordosis. Thoracopagus twins sometimes are facing each other, which may turn tracheal intubation difficult [4].

Vascular access may be complicated by the age and weight of the conjoined twins. Arterial access is indispensable due to the possibility of a significant cardiovascular instability and need for massive transfusion during the surgical procedure [5].

In general, omphalopagus have a variable degree of cross- circulation. The degree of cross-circulation is dynamic and highly dependent on both twins` relative systemic vascular resistance [1]. Due to the cross-circulation, the pharmacology of the anesthetic agents may be modified [5]. Administration of neuromuscular blockers should not be performed until the airway is appropriately secured in both infants since its administration to one of the babies may result in paralysis of the other one [1].

In that way, long-duration surgery, difficulty of vascular access and management of the airways, the possibility of massive blood and fluids loss, logistics of safe intraoperative repositioning during separation and the transfer of one twin to an adjacent operating theatre after separation makes the procedure quite complex [5].

In the intraoperative period, optimization of haemostasia and avoidance of hypoxemia, hypercarbia, acidosis, hypotension, hypothermia and electrolyte imbalance is mandatory [1]. The amount of intraoperative blood loss is dependent on the extensive and complexity of the fusion [2].

In the postoperative period, cardiac instability and respiratory compromise can occur due to alterations in postoperative anatomy, inflammatory response or infection [1].

A multidisciplinary approach is fundamental for the success of the conjoined twins separation surgery. The anesthetic management include a particular attention to safe airway management, adequate intravascular access, careful positioning, proper management of fluids, and prevention of hypothermia. All these meticulous perioperative efforts contribute significantly to the children survival.

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