Rev Bras Anestesiol. 2019;69(2):214–217

Sociedade Brasileira de Anestesiologia

REVISTA BRASILEIRA DE ANESTESIOLOGIA

Publicação Oficial da Sociedade Brasileira de Anestesiologia
www.sba.com.br

CLINICAL INFORMATION

Anesthesia and perioperative challenges for surgical separation of thoraco-omphalopagus twins: case report

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Received 16 November 2017; accepted 15 June 2018
Available online 13 November 2018

KEYWORDS
Conjoined twins;
Pediatric anesthesia;
Multidisciplinary care

Abstract
Background and objectives: Conjoined twins are monozygotic twins physically joined at some part of the body. This is a rare phenomenon, estimated between 1:50,000 and 1:200,000 births. The objective of this report is to present the anesthetic management and the perioperative challenges for a separation surgery.
Case report: Thoraco-omphalopagus twins were diagnosed by ultrasound and were followed by the fetal medicine team of the service. After 11 h of cesarean surgery, the pediatric surgical team chose to separate the twins. They were monitored with cardioscopy, oximetry, capnography, nasopharyngeal thermometer, urinary output, and non-invasive blood pressure. We chose inhaled induction with oxygen and 4% Sevoflurane. T1 patient was intubated with a 3.5 uncuffed endotracheal tube, and, after three unsuccessful intubation attempts of patient T2, a number 1 laryngeal mask was used. After securing the twins' airway, the induction was supplemented with fentanyl, propofol, and rocuronium. Mechanical ventilation in controlled pressure mode (6 mL.kg^{-1}) and lumbar epidural (L1–L2) with 0.2% ropivacaine (2.5 mg.kg^{-1}) were used. The pediatric surgical team initiated the separation of the twins via sternotomy, ligation of hepatic vessels. After 2 hours of procedure, the separation was completed, continuing the surgical treatment of T1 and the support of T2 until his death.
Conclusions: Conjoined twin separation surgery is a challenge, which requires planning and coordination of a multidisciplinary team during all stages.

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https://doi.org/10.1016/j.bjane.2018.10.004

PALAVRAS-CHAVE
Gêmeos conjugados;
Anestesia pediátrica;
Cuidados multidisciplinares

Anestesia e desafios perioperatórios para cirurgia de separação de gêmeos toraco-onfalópagos: relato de caso

Resumo
Justificativa e objetivos: Gêmeos conjugados são gêmeos monozigóticos conectados por alguma parte do corpo. Esse é um fenômeno raro, estimado entre 1:50.000 a 1:200.000 nascimentos. O objetivo deste relato é apresentar o manejo anestésico e os desafios perioperatórios para cirurgia de separação.
Relato de caso: Gêmeos toraco-onfalópagos foram diagnosticados por ultrassonografia e acompanhados pela equipe de medicina fetal do serviço. Após 11 horas da cesárea, a equipe cirúrgica pediátrica optou pela separação dos gêmeos. Foram monitorados com cardioscopia, oximetria, capnografia, termômetro nasofaríngeo, débito urinário e pressão arterial não invasiva. Optou-se por indução inalatória com oxigênio e sevoflurano a 4%. O G1 foi intubado com tubo orotraqueal 3,5 sem *cuff* e após três tentativas de intubação do G2 sem sucesso usou-se máscara laríngea número 1. Após obtenção da via aérea nos gêmeos, complementou-se indução com fentanil, propofol e rocurônio. Ventilação mecânica no modo pressão controlada 6 $ml.kg^{-1}$ e peridural lombar L1-L2 com ropivacaína 0,2% (2,5 $mg.kg^{-1}$). A equipe cirúrgica pediátrica iniciou a separação dos gêmeos através de esternotomia, ligadura de vasos hepáticos. Após duas horas de procedimento, a separação foi concluída, prosseguiram-se o tratamento cirúrgico de G1 e os cuidados de G2 até o óbito.
Conclusões: A cirurgia de separação de gêmeos conjugados é um desafio, requer planejamento e coordenação de uma equipe multidisciplinar durante todos os estágios.

Introduction

Separation of conjoined twins is an anesthetic-surgical challenge. A multidisciplinary team should be available with at least two anesthesiologists for each patient. Important factors to consider are: the presence of circulatory communication between the twins, shared organs, difficult airway, great volume loss, and extensively prolonged perioperative time.[1]

Ideally, separation surgery should be postponed for 4-12 weeks when the rate of survival reaches up to 90%.[2] This article reports the anesthetic care for thoraco-omphalopagus twins separation and reviews the literature.

Case report

A.R.G.S., 18 years old, primiparous, 38 weeks and 5 days of gestational age diagnosed by ultrasound (US) in the first trimester. She attended seven prenatal visits with negative serology for syphilis, HIV, hepatitis B, rubella, and toxoplasmosis. Female thoraco-omphalopagus twins were diagnosed by US in the first trimester, and then the mother was referred to the fetal medicine team of the Institute of Integral Medicine Prof. Fernando Figueira (IMIP) where they were monitored.

The patient was admitted to the service on July 10, 2017 for magnetic resonance imaging (MRI) and birth follow-up. On July 28 at 9:30 pm, an emergency cesarean section was indicated due to fetal bradycardia. The twins were born uneventfully, weighing together 2980 g, with Apgar 9/10; Twin 1 (T1) was identified as the healthy and Twin 2 (T2) as the malformed, both were taken to the Intensive Care Unit (ICU). T1 had no malformations, except for the hepatic (single liver) connection with T2, who had multiple malformations: kidney agenesis; hypoplastic lung; esophageal atresia; unichamber heart; agenesis of pelvis, bladder, and limbs.

The pediatric surgical team of the service opted to perform the separation procedure the next day, due to the risk of clinical deterioration of T2 and possible need for emergency surgery. Eleven hours after birth and mobilization of a multidisciplinary team, the separation surgery was started. The anesthetic team consisted of two pediatric anesthesiologists, one third year resident and two first year residents. The surgical team consisted of five pediatric surgeons, two with experience in pediatric oncology surgery, one with experience in separation of conjugated twins and one in renal transplantation; in addition to three pediatric surgery residents.

Venous access (22G) was punctured in T1 and without possibility of puncturing the other twin due to limb agenesis. Heated mattress and thermal blanket were used. Ambient temperature was maintained around 24 degrees Celsius. Monitoring with cardioscopy, oximetry, capnography, nasopharyngeal thermometer, and urinary output was installed in both and non-invasive blood pressure in T1. We opted for inhalation induction due to suspected difficult intubation and the twin's atypical position, with 100% oxygen and 4% sevoflurane for T1 and 100% oxygen supply for T2. T2 had to be positioned above T1 for better airway access; orotracheal intubation was performed with a 3.5 uncuffed

Figure 1 Twin 2 had to be positioned above Twin 1 for better airway access.

tube, without hemodynamic repercussions of autotransfusion (Fig. 1). Patient T2 was repositioned and induced in the same way as T1; laryngoscopy and three attempts of intubation with a 2.5 uncuffed orotracheal tube were unsuccessful because the tube did not progress to the glottic cleft. During the intubation attempts, there was no desaturation of T2; we chose to use a laryngeal mask number 1, with success.

After securing the twin's airway, fentanyl 5 $\mu g.kg^{-1}$, propofol 4 $mg.kg^{-1}$, rocuronium 0.6 $mg.kg^{-1}$, gentamicin 5 $mg.kg^{-1}$, and metronidazole 10 $mg.kg^{-1}$ were administered. Pressure-controlled ventilation was performed with 50% oxygen, aiming at 16 mL tidal volume, respiratory rate of 36 breaths/min, end-tidal CO_2 about 33 mmHg, and 93% peripheral oxygen saturation. Lumbar epidural (L1–L2) using the Dogliotti technique with glass syringe was performed in T1, with 0.2% ropivacaine (2.5 $mg.kg^{-1}$; volume 2.5 mL).

The surgical team positioned the patients for catheterization of the left internal jugular vein of T1 and subsequently repositioned them to initiate separation. Hydration was calculated using the Holiday Formula (4 $mL.kg^{-1}.h^{-1}$) and surgical loading (10 $mL.kg^{-1}.h^{-1}$) with Lactated Ringer and 1% dextrose. No vasoactive drugs or blood products were used. Heart rate ranged from 98 to 132 bpm, capnometry of 32-47 mmHg, glycemia 64-127 $mg.dL^{-1}$, and temperature from 33.1 °C after induction to 36.4 °C at the end of surgery.

The separation of twins started at 9:35 AM through sternotomy and ligation of hepatic vessels. After 2 h of procedure the separation was completed. The surgical treatment of T1 proceeded with correction of intestinal malrotation, hemostasis, and synthesis. At the end of surgery, T1 was taken intubated to the ICU at 13:05 PM (Fig. 2). Because of the non-viability due to the multiple malformations, T2 remained with the laryngeal mask on spontaneous ventilation, monitored, and sedated until death at 2:35 PM.

After 72 h of the procedure, T1 presented with hematemesis due to the orogastric tube. The institutional endoscopy service was activated, identified the bleeding vessel in great curvature and cauterized it. T1 progressed uneventfully to extubation on the 7th postoperative day, discharge from ICU to ward on the 10th postoperative day, and was discharged from hospital on the 15th day.

Figure 2 Twin 1 was taken intubated to the ICU after surgery.

Discussion

This case summarizes the anesthetic care and management of conjoined twins. Preoperative evaluation, intraoperative planning, and postoperative care are crucial to the success of the procedure.

Conjoined twins are monozygotic, monochorionic, and monoaminiotic twins. They are of the same sex, with a female ratio of 3:1. This is a rare phenomenon, estimated between 1:50,000 and 1:200,000 births. The first successful conjoined twins surgery dates back to 1869 and since then more than 1200 procedures have been performed around the world.[3]

The occurrence of conjoined twins is explained by the fission theory arising from a partial separation of the zygote around the 20th gestational day, or fusion theory secondary to the union of the embryos around the 4th gestational week.[4] Conjoined twins are classified according to the site of inter-connection: omphalopagus (abdomen), thoracopagus (thorax), ischiopagus (hips), craniopagus (head), cephalopagus (neck), spinal cord (spinal column).[1] The reported case was of thoraco-omphalopagus twins, which corresponds to the most common type in 74% of cases, ranging from multiple organ connections to hepatic union.

The diagnosis of conjugated twins begins ideally in prenatal care with ultrasound examination, which identifies the twins' inter-connection around the 9th to 12th weeks and, when visible, confirms fusion of limbs, abdomen, chest or

liver. More detailed assessment is done with a fetal MRI to identify which organs are shared and plan the plausible surgical approach.

Obstetric intervention is planned around the 36-38 weeks of gestation, once the pulmonary maturity has already been achieved, avoiding a complicated vaginal delivery or an emergency cesarean-section. Emergency procedures have a mortality rate of 70% versus 20% of the electives.[2] Emergency surgery is indicated in some situations, such as cardiorespiratory deterioration, necrotizing enterocolitis, intestinal obstruction or other threatening situation for one or both of the twins. In the case reported, the procedure was postponed until approximately 39 weeks due to the difficulty of obtaining a fetal MRI.

Neonatal care should be provided with one team for each twin. When one of the twins is a parasite, the ex-utero intrapartum (''Exit'') procedure should be performed, as decompensation of the malformed twin can compromise the healthy one. ''Exit'' is a strategy that aims to approach and control the airway of the newborn while the gas exchanges are still done through the umbilical cord.[5] In our case, considering the indication of emergency cesarean-section due to fetal bradycardia, the procedure was not performed with a view to neonatal resuscitation care.

One anesthetic team for each child is required with monitoring and duplicate anesthetic station. Assessment of circulatory communication between twins can be made by administering anticholinergic in one of them and assessing the presence or absence of response in the other. Sequential induction is performed when there is no circulatory communication.

Drugs administered to one of the twins may have an unexpected effect on the other. It is recommended that doses of anesthetic agents be calculated by the estimated weight of each twin and infused separately. Blood loss is especially extensive in thoracopagus or ischiopagus twins and requires blood transfusion between 10% and 450% of estimated blood volume.

Tracheal intubation is a challenge in the separation of twins due to the atypical position. Some authors report that tracheal intubation can be done in one twin while the other is supported on top, other authors warn about the risk of autotransfusion when this positioning is adopted. Awake intubation is recommended by some authors due to the usually presence of difficult airway. However, other complications such as burking, laryngospasm, and sympathetic discharge from airway manipulation are at increased risk. In the case reported, it was possible to intubate one twin and insert the laryngeal mask on the other.

Maintaining hemodynamic stability is essential during the surgical procedure. Blood loss assessment can be done by the aspirated volume of the operative field, hematocrit variation, and weighing of surgical gauzes. When there is circulatory communication there is no need to separately estimate the blood loss of each twin, as the volume and hematimetric variation are shared.

Normotermia is another challenge in large neonatal surgeries. Drop in central temperature is expected because of the thermal self-regulation breakdown caused by the anesthesia, especially in the first hours due to heat redistribution. To reduce losses by irradiation and convection, the room temperature should be regulated between 24 °C and 26 °C and active forms of heat transfer such as thermal blanket, mattress, and heated solutions should be used.

One aspect that should be remembered is the particular physiology of these newborn patients: frequency-dependent cardiac output; poor complacent lung; fatigability of the respiratory muscles; hepatic and renal immaturity; and more cephalic and more anterior glottis. At that age, the organ systems are not mature: the heart is not prepared for hypervolemia or hypovolemia. The kidney has difficulty retaining sodium under stress and there is tubuloglomerular imbalance, with urine concentration difficulty. The liver has not yet developed mechanisms for drug metabolism. The concentration of proteins that bind to drugs is small (albumin and α1-acid glycoprotein), predisposes to a higher percentage of free drugs.[2]

Cardiorespiratory insufficiency is the main cause of immediate death. Multiple reconstructive procedures are usually required. Ethical considerations are important in twin separation surgery, especially when there is need to sacrifice a twin due to multiple malformations. The religious view in the latter case is quite controversial.

Conclusion

The surgical separation of conjoined twins is a challenge, requires planning and coordination of a multidisciplinary team during all stages.

Conflicts of interest

The authors declare no conflicts of interest.

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