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Case report

Omphalopagus conjoined twins separation during coronavirus disease-19 pandemic era: A case report

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ABSTRACT

Introduction and importance: Conjoined twin is a rare congenital anomaly characterized by a fusion of certain anatomical structures. Coronavirus-19 (COVID-19) is a new emerging infectious respiratory disease affecting worldwide and potentially leads to acute respiratory distress (ARDS) in children. COVID-19 has reconstructed the healthcare system, including surgical care and decision-making.
Case presentation: Herein we describe a surgical separation of 2.5 months old omphalopagus conjoined twins, with one of them (Baby A) presenting COVID-19-associated respiratory distress, as well as the challenges faced during the preparation and the execution of the complex surgical procedure.
Clinical discussion: Baby A underwent antiviral therapy, oxygen supplementation, and ventilation in the ICU, while baby B remained stable and confirmed negative for SARS-CoV-2. The separation surgery was conducted after baby A had become clinically stable. Defect closure and reconstruction were accomplished. At one week follow-up, Baby A died of lung infection, while baby B remained well after one year.
Conclusion: The complexity of surgical separation requires careful planning by a multidisciplinary team. Surgical separation of conjoined twins during the pandemic era has not been reported much in the literature, more reports are required to provide further insight.

1. Introduction

Conjoined twins are monozygotic identical twins that share a physical fusion. Conjoined twinning is a rare anomaly, with an incidence of 1.5/100.000 to 1/500.000 births in western countries [1,2]. Fusion at the anterior thorax and abdomen, including omphalopagus being the most common (70%) [3].

Coronavirus disease-19 (COVID-19) is a global pandemic that had been declared by World Health Organization (WHO) on 11 March 2020. In situations where surgical intervention is required for COVID-19 patients with sequelae, a poor postoperative outcome is expected. In addition to that, the increased risk of COVID-19 transmission to healthcare providers (HCP) prompts prioritizing urgent cases and postponing the elective ones [4].

Herein, we report the first case of omphalopagus conjoined twins during the COVID-19 pandemic, with one of the twins diagnosed with a

Abbreviations: ARDS, acute respiratory distress syndrome; cm, centimeter; CDC, Centers for Disease Control and Prevention; CPAP, continuous positive airway pressure; CT-scan, computed tomography-scan; COVID-19, coronavirus disease-19; CUSA, cavitron ultrasonic surgical aspirator; ECMO, extracorporeal membrane oxygenation; HCP, healthcare providers; HFN, high flow nasal; HFOV, high-frequency oscillation ventilation; ICU, intensive care unit; IVC, inferior vena cava; lpm, litres per minute; MRI, magnetic resonance imaging; MRSA, methicillin-resistant *Staphylococcus aureus*; NPWT, negative pressure wound therapy; ORC, oxidized regenerated cellulose; PCR, polymerase chain reaction; PDA, patent ductus arteriosus; PDS, polydioxanone suture; PICU, pediatric intensive care unit; POD, postoperative day; PPE, personal protective equipment; SCARE, surgical case report; WHO, World Health Organization.

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COVID-19 infection, along with the difficulties encountered in the preoperative, intraoperative, postoperative management, and the preventive measures of HCP transmission at the beginning of the COVID-19 pandemic era.

This case report has been reported in line with the SCARE Criteria [5].

2. Case presentation

Omphalopagus female conjoined twins were referred to our hospital at the age of 2.5 months old (Fig. 1). One baby (baby A) presented with sepsis and acute respiratory dysfunction syndrome (ARDS) due to COVID-19, confirmed by bronco-alveolar lavage, manifesting shortness of breath. At the first week of hospitalization, baby A was lethargic, underwent recurrent desaturation, and had respiratory acidosis. Although a Computed tomography scan (CT-scan) showed ground-glass opacity and multiple consolidations in both babies, baby B's clinical appearance was well and tested negative with COVID-19 polymerase chain reaction (PCR). Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Imaging from CT scan showed pericardium, sternum, and liver fusion, whereas a non-ionic contrast meal evaluation showed no bowel fusion (Fig. 2). The fusion was 9 centimeter (cm) in latero-lateral, and 15 cm in cranio-caudal direction. Fusion and fenestration were not evident in the heart, but cardiomegaly was noted in both babies. Part of Baby A's heart herniated into Baby B's thoracic cage. Atelectasis was present in both babies, however, pulmonary hypertension did not occur. In the liver, a connection was identified between the left portal vein of baby A and the left portal vein of baby B, with an arterioportal shunt. Magnetic resonance imaging (MRI) showed no fusion in the biliary system.

Baby A received antivirals (Lopinavir, Ritonavir), hydroxychloroquine, and antibiotics (Amikacin, Vancomycin) as definitive therapy for *Methicillin-Resistant Staphylococcus Aureus* (MRSA) infection. Baby A received breathing support in the form of continuous positive airway pressure (CPAP). Eventually, a conventional ventilator and high-frequency oscillation ventilation (HFOV) were used due to ARDS. Baby B remained stable and did not require any oxygen therapy. On the 10th day of treatment, the PCR test of both showed negative.

During the two months in the ICU, Baby A developed recurrent pneumonia, ventilator weaning difficulty, and sepsis that escalated to septic shock. Three months post-hospitalization, the ventilator was weaned to non-invasive support for baby A and baby B received an oxygen nasal cannula 2 l per minute (lpm). There was no fever, the antibiotic treatment achieved infection control. Both babies were in stable condition and the plan to perform the separation surgery proceeded.

Fig. 1. Clinical appearance of the conjoined twin.

The operation was performed by a multidisciplinary team and was completed in 13 h by pediatric, cardiothoracic, and plastic surgeons. The intraoperative findings in the chest cavity were underdeveloped sternum and costae, with a defect in the diaphragm. Baby A did not have a sternum and the pericardial tissue required dissection for separation.

Hepatectomy was performed Using Cavitron Ultrasonic Surgical Aspirator (CUSA®), whereas bleeding control utilized monopolar and bipolar cautery, surgical clips, and suture. A portal vein crossing between both livers was carefully identified and ligated. We performed a hanging maneuver using a small Nelaton catheter placed behind the liver.

In baby B, Cardiothoracic surgeons performed reconstruction of the pericardium using a Gore-Tex® patch and plates were placed to attach the diaphragm to the costae to allow diaphragm movement for adequate respiration. The abdominal cavity was closed using a mesh, followed by the defect closure utilizing a flap.

The defect dimension on baby A was 15 × 25 cm after separation surgery (Fig. 3). Due to unstable hemodynamics, the skin closure in baby A was postponed. POD+3 of separation surgery, baby A was scheduled for closure of the defect. A defect on baby B was 9 × 15 cm (Fig. 4). Bilateral keystone design perforator island flap from the lateral side of the defect was designed around the perforator which was identified using hand-held Doppler ultrasound (Fig. 5). We used dynamic sutures for an expansion effect on the keystone flap. Baby A's general condition improved, hence we performed surgery to cover the remaining central defect by re-elevation of the lower limb of the right keystone perforator flap. A skin graft was performed on the lateral donor area. The remaining defect on Baby B's abdominal area was closed using the lower abdominal flap, which was advanced cranially. Defects on baby B were sufficiently covered (Fig. 6).

During a follow-up, baby A was extubated on POD+28, while baby B on POD+5. Baby A needed oxygen treatment until she was discharged. Both babies were hemodynamically stable with the support of milrinone and epinephrine in baby A, epinephrine, and norepinephrine in baby B. Treatments were discontinued on POD+4.

Baby A experienced bacterial septicemia, severe wound infection and underwent multiple debridement procedures while Baby B had no significant postoperative complication. Baby A was discharged on POD+66 after defect closure. Baby B was discharged one month after separation surgery.

One week following discharge, baby A developed a lung infection and high fever, which caused mortality while she was treated at another hospital. Baby B's condition remained well at a one-year follow-up.

3. Discussion

Conjoined twins were referred to our hospital due to sepsis with severe COVID-19-associated ARDS on one of the babies. The ethics committee was faced with whether or not to proceed with immediate versus delayed definitive separation of the twins. The benefits of delayed separation are reduced risks of anesthesia, establishing anatomic relationships with careful imaging studies, detecting previously unrecognized congenital anomalies, and setting up treatment plans to ensure adequate wound coverage. However, a delay may result in the death of both babies due to COVID-19 ARDS.

The issue was not on the technicality, as our team has experience in separation surgery of conjoined twins for more than thirty years. The presence of a pandemic was a contributing challenge. With minimal resources and infrastructure in our country, we struggle to offer the best treatment for COVID-19 patients.

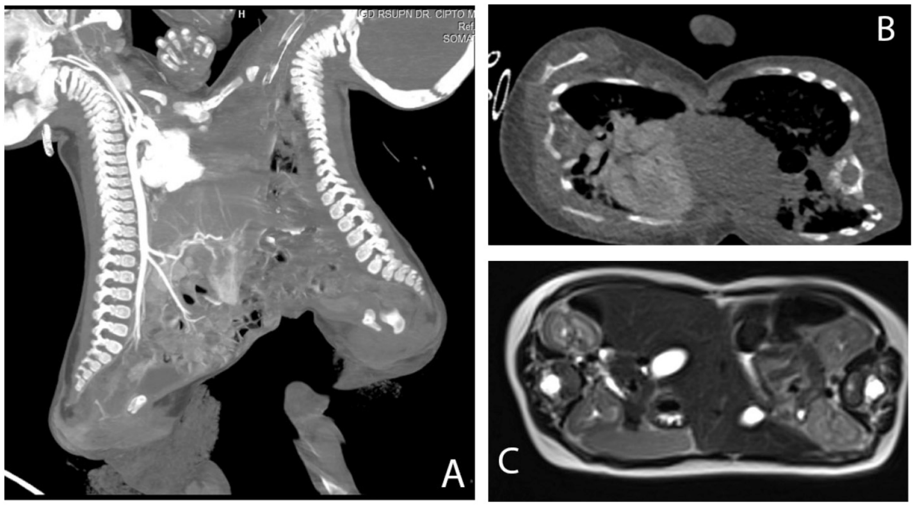

Fig. 2. A) Clinical presentation of the case. Baby A: left side. B: Abdomen and thoracic CT scan (baby A: right side). C: Thoracic CT scan (baby A: right side) D: Abdominal MRI showed separated biliary system (baby A: right side).

Fig. 3. Defect closure of baby A. A) Raw surface of the liver was lined by Surgicell®, reconstruction of the pericardium with Gore-Tex®, the thoracic cage was strengthened by a plate. B) Abdominal cavity was closed with Proceed® mesh. C) Lateral view of the defect. D) After delayed and staged closure (2 months after separation).

The vital points that affect the decision to perform surgical separation lie in the mortality and morbidity of the outcome. The surgeons agreed to perform the surgical separation when a stable condition in both babies was well-established and infants were aged around 3–4 months old with an accumulated weight over 10 kg, along with a negatively confirmed SARS-CoV-2 following 3 months from the initial infection.

In the absence of COVID-19 infection, the timing of surgery remains controversial and varies between institutions. A delay of a few months yields a higher chance of survival. Without an indication for early separation, the surgery is ideally performed at the age of 3 months, allowing detailed investigations and a careful wound closure plan [6]. A separation surgery that is delayed until the age of 10 months old and further instigates a psychosocial issue, as it interferes with the developmental aspect [1,7].

High-frequency oscillation ventilation (HFOV) was administered for one of the twins at the occurrence of ARDS and presented a good outcome, despite the finding of a systematic review and meta-analysis

Fig. 4. Defect closure of baby B. A) Thoracic and abdominal cavities were closed with a Proceed mesh. B) Lateral view of the defect closure. C) Defect closure at the time of separation surgery.

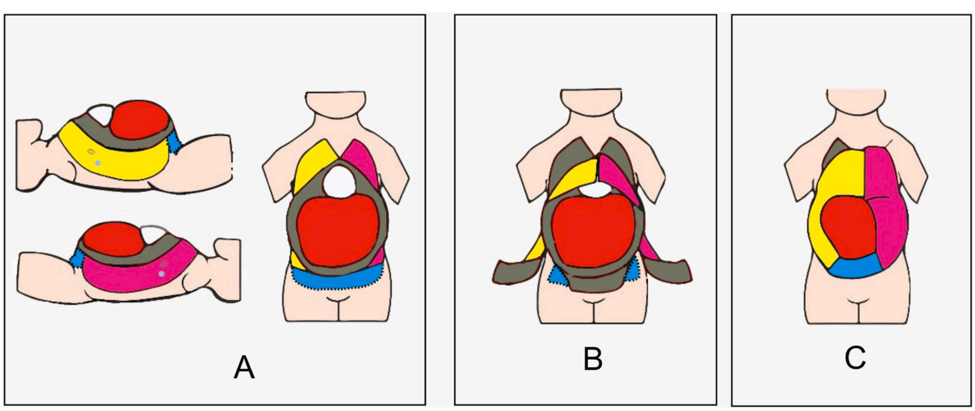

Fig. 5. Flap design of baby A. A) flap design, B) flap elevation, C) post-operative.

on the failure of HFOV to reduce hospital and 30-day mortality due to ARDS compared to mechanical ventilation [8]. Ventilation intervention recommended by Kache et al. [9] for children with COVID-19 is mechanical ventilation, as HFOV is not the choice for routine application but may be considered in selected cases, although these statements have insufficient evidence [9].

3.1. *Operative planning*

A twin separation requires accurate determination of anatomy and vascular supply. Evaluation of vascular shunts and cross-circulation is important for anesthetic management, as shared blood vessel ligation and separation may be necessary during surgery to prevent hypovolemic shock due to loss of blood volume [10]. Once the entire system has been evaluated and vascular territories have been established, the multidisciplinary team reviews the information to plan separation.

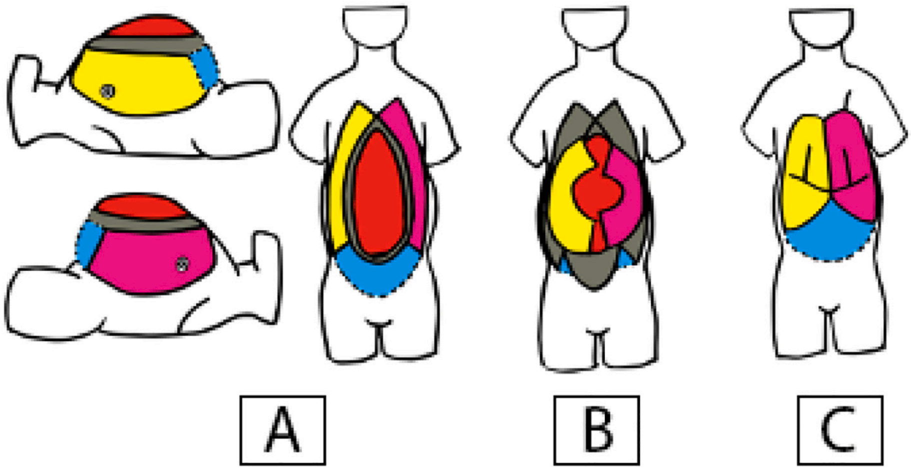

Fig. 6. Flap design of baby B. A) flap design, B) flap elevation, C) post-operative.

3.2. Infection prevention control of COVID-19

COVID-19 infection control and prevention were compliant with the guidelines set by the Centers for Disease Control and Prevention (CDC). As the surgery involved the opening of the thoracic cavity, we utilized aerosol PPE including an N95 mask, goggles, disposable gowns, and gloves. All operating theatre (OT) staff stepped up infection control measures and conducted N95 mask fitting sessions. Appropriate PPE utilization, maintenance of a pre-determined surgical team, and minimizing surgical traffic were vital in preserving equipment and limiting HCP exposure. Reducing OT staff and entering and exiting were strictly applied to avoid transmission [11].

3.3. Operative management

The separation of a conjoined liver is a highly complex operation carrying risks for hemorrhage, vascular thrombosis, and biliary complications. To minimize blood loss during hepatic parenchymal dissection, we incorporated conventional techniques with CUSA. Staying at the cleavage line facilitated both a safe and relatively dry dissection. Al-Rabeeah et al. [12] suggested that applying a large Penrose drain circumferentially helped identify the cleavage line.

Intrahepatic portal vein crossings joined the liver parenchyma. We performed a hanging maneuver circumferentially, as described by Belghitti et al. [13]; encircling the portal pedicle after carefully dissecting across the anterior surface of the intrahepatic IVC, elevating the liver from the anterior surface of the IVC. Vessel loop or Penrose drain used to elevate the hepatic parenchyma provided adequate exposure and effective hemostasis if required.

3.4. Reconstruction

Chest and abdominal wall closure present a challenge in omphalopagus twins. Body wall defect coverage post-separation was accomplished using prosthetic materials and preoperative tissue expansion. Preoperative tissue expansion requires time, but time for tissue expansion was inadequate. There was an increased risk of infection in the tissue expander. Protrusion of the heart predominantly posed closure difficulty, similar to the case of thoracic ectopia cordis. Autologous tissue coverage with various methods typically produces excessive compression of the heart, limiting cardiac output by impeding cardiac filling or kinking great vessels. The thoracic cavity of Baby B was bigger than baby A. Pericardium reconstruction used Gore-Tex® soft tissue patch. The diaphragm and the entire anterior costaes were fixed to the miniplate. The heart apex of Baby B and Baby A projected approximately 3 cm and 5 cm above the chest wall, respectively. The liver of Baby A projected 10 cm above the abdominal wall.

Chest and abdominal wall coverage of baby B were successful with Proceed® surgical mesh and bilateral keystone design perforator island flap, and coverage was left open. The closure of baby A was delayed due to inadequate soft tissue coverage and hemodynamic instability. Bioprosthetic materials are better than permanent synthetic materials for gradual remodeling into autologous tissue to adapt to body growth. Proceed® mesh had three distinct materials, providing a bioresorbable layer that physically separates polypropylene mesh from visceral organs absorbed within 6 months.

Rehabilitation was facilitated before surgical separation to increase overall health, strength, and endurance to withstand surgical demands. Respiratory rehabilitation for Baby A with post-ARDS due to COVID-19 infection was essential, as mentioned in the guidelines by WHO. Rehabilitation programs from post-acute to long-term are tailored according to patient's needs.

4. Conclusion

Omphalopagus conjoined twins were successfully separated with a multidisciplinary approach during the COVID-19 pandemic. A well-planned preparation, operation timing, and procedure were paramount. Postoperative management involving a multidisciplinary team to obtain the best outcome for patient survival.

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Author contribution

- Study conception and design: Tri Hening Rahayatri, Rizky Amaliah, Nandita Melati Putri, Mulya Rahma Karyanti
- Acquisition of data: Tri Hening Rahayatri, Rizky Amaliah, Nandita Melati Putri, Niken Wahyu Puspaningtyas, Mulya Rahma Karyanti, Aryono Hendarto
- Analysis and interpretation of data: Tri Hening Rahayatri, Rizky Amaliah, Nandita Melati Putri, Niken Wahyu Puspaningtyas, Mulya Rahma Karyanti, Aryono Hendarto
- Drafting of manuscript: Tri Hening Rahayatri, Rizky Amaliah, Nandita Melati Putri, Mulya Rahma Karyanti
- Critical revision: Tri Hening Rahayatri, Rizky Amaliah, Nandita Melati Putri, Niken Wahyu Puspaningtyas, Mulya Rahma Karyanti, Aryono Hendarto

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Declaration of competing interest

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