

Successful emergency separation of premature omphalopagus conjoined twins: an anaesthetic experience

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Abstract

Anaesthesia for early emergency separation of premature conjoined twins is extremely rare as surgery generally done electively between 2 to 4 months of age. However, urgent separation may be needed due to life-threatening complications. We report a case of successful early separation of premature omphalopagus twins at 36 weeks of gestational age with a combined weight of 2.7 kg. To the best of our knowledge, this was the lowest weight yet reported of successful surgical separation in Malaysia. Early separation was indicated as extrauterine twin-to-twin transfusion with unbalanced blood shunting through the porto-systemic anastomoses within the shared liver parenchyma potentially risked impending life-threatening organ failures. Anaesthesia for the separation of premature conjoined twins in the emergency setting requires extensive multidisciplinary discussion and planning. Factors predicting difficult anaesthesia in this case were the twins' size and age as well as duration of anaesthesia. Two separate anaesthetic teams were required

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with all team members well versed in the operative workflow. Simultaneous airway management, prevention of hypothermia, and vigilant haemodynamic monitoring are key to successful anaesthesia in premature conjoined twin separation.

Keywords: extrauterine twin-to twin-transfusion, premature conjoined twins, separation surgery

Introduction

Conjoined twins (CT) are a rare phenomenon of monochorionic, monoamniotic twins. The prevalence of CT varies geographically and socioeconomically. The incidence varies from 1 in 50,000 to 1 in 10,000 live births, with only 50% being liveborn.¹ CT are classified based on the site of attachment, where five types of CT are commonly identified: thoracopagus (thorax), omphalopagus (abdomen), pygopagus (sacrococcygeal junction), ischiopagus (pelvis), and craniopagus (skull). Omphalopagus CT have the best chances of survival if successfully separated. For a successful outcome, surgical separation of omphalopagus CT is often performed electively when the twins reach an acceptable age and weight, after extensive preparation and planning. Preparation includes examination of the twins with different imaging protocols and multidisciplinary team discussion. Here, we report the successful separation of premature omphalopagus CT weighing 2.7 kg at day 17 of life. The separation was done in an emergency setting due to life-threatening extrauterine twin-to-twin transfusion. Anaesthesia for premature CT separation surgery, especially in an emergency setting, imposes enormous challenges to the anaesthesiologist perioperatively.

Case presentation

A pair of premature symmetrical omphalopagus CT were born at 33 weeks and 5 days of gestation, via emergency caesarean section in a hospital distant from our centre. They were boys with a combined birth weight of 3 kg. The babies, designated as “twin A” (TA) and “twin B” (TB) (Fig. 1), were symmetrically conjoined from xiphisternum to below the umbilicus with normal head and neck, limbs, spine, and urogenital systems.

Both babies developed respiratory distress syndrome shortly after birth, received surfactant, and were mechanically ventilated for 72 hours. Initial echocardiography revealed two structurally normal and separated hearts, but dex-

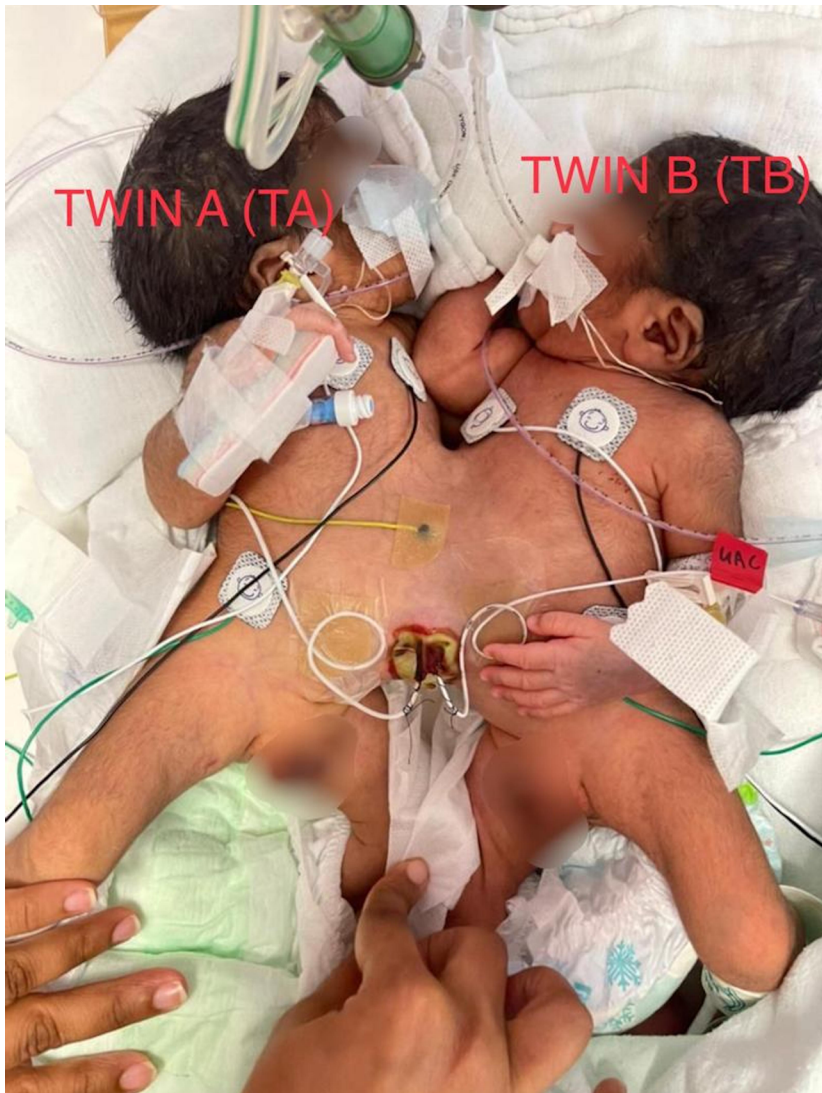


Fig. 1. Omphalopagus conjoined twins A and B.

trocardia and a patent foramen ovale in TB. At day 6 of life, TA developed oliguria and hypotension requiring inotropes, and TB was noted to be hypervolaemic and polyureic. Extruterine twin-to-twin transfusion was suspected and a contrast-enhanced computed tomography of the chest and abdomen was performed. The scan reported symmetrically conjoined twins, with shared liver parenchyma and small bowel, with possible complex biliary anatomy. The presence of cross-circu-

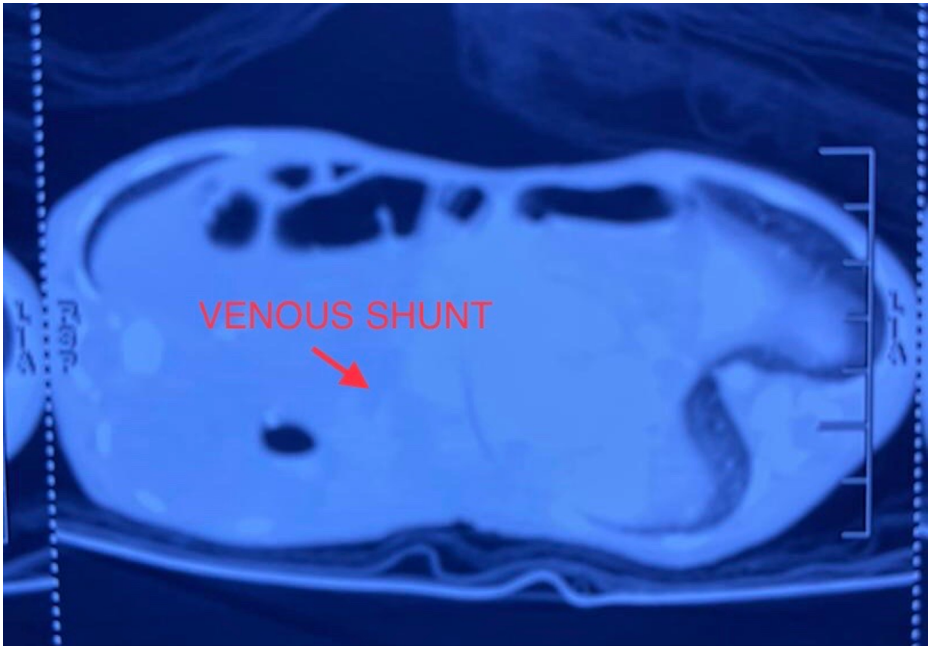


Fig. 2. Computed tomography of the abdomen showed venous vessels traversing the fused liver parenchyma.

lation between the twins was confirmed based on the presence of venous vessels traversing across the fused liver parenchyma (Fig. 2).

At day 13 of life, TA deteriorated, requiring bolus doses of fluids and increment in inotropic support, whereas TB became hypertensive and polyureic. The decision to transfer the babies to our centre for possible emergency separation was made after multidisciplinary discussion. A repeat echocardiography in TB revealed global hypokinesia and impaired systolic function, therefore intravenous (IV) adrenaline and IV milrinone were started preoperatively. The warning signs of imminent renal failure in TA and cardiac failure in TB prompted a multidisciplinary decision to proceed with the operation on day 17 of life, at a combined body weight of 2.7 kg, despite the high risk of morbidity and mortality. Written informed consent was obtained from the parents, specifying saving the baby with the better chance of survival should conditions arise where such a decision became necessary.

All team members involved were briefed on the flow of anaesthesia and surgery a day before surgery. The blood bank unit was alerted on the complexity of the case. Two anaesthetic teams were established and two operation rooms with an interconnecting door were used. Two sets of colour-coded equipment including the

anaesthetic machine, monitors, and induction carts were prepared. The identification tag of each team member was colour-coded to differentiate their roles.

The twins were transferred to the operation theatre in an incubator. Both babies faced each other in the semi lateral position. The drug dosage calculated for each baby was half of the combined body weight. They had femoral double lumen catheters inserted from the neonate intensive care unit (NICU), and were monitored with electrocardiogram, non-invasive blood pressure, and two sets of peripheral oxygen saturation.

Anaesthesia induction began in TB due to his cardiorespiratory instability. Following induction with sevoflurane 1% and IV fentanyl 1 mcg/kg, continuous positive airway pressure (CPAP) was applied with mask oxygenation and atracurium 1 mg administered for intubation. TB was intubated at the first attempt with a size 3 uncuffed endotracheal tube (ETT) using the C-MAC video laryngoscope (Karl Storz, Tuttlingen, Germany). During this time, CPAP and mask oxygenation with 1% sevoflurane in 100% oxygen was concurrently provided for TA. The same drugs and ETT size were used for TA. The ETTs were well secured with adhesive plasters to prevent ETT displacement during positioning or transfer. Anaesthesia was maintained with 0.2–1% sevoflurane in a 50% air-oxygen mixture, atracurium bolus doses, and IV fentanyl and morphine for analgesia.

IV and intraarterial line insertions were challenging. Catheterisation of the right internal jugular vein (IJV) for TA was achieved with ultrasound guidance. However, attempts of left IJV cannulation for TB were unsuccessful, hence achieved via venous cutdown by the surgeon. The twins' intraarterial blood pressure, central venous pressure (CVP), and temperature were continuously monitored

The total procedure time was approximately 7 hours, and they were separated 66 minutes after surgical incision. The surgery was performed with the twins in supine posture throughout the surgery. There were fluctuations in blood pressure and episodes of transient oxygen desaturation intraoperatively. The latter was managed by maintaining mean arterial pressure (MAP) above 35 mmHg for TA and above 40 mmHg for TB. The fused area of the liver was separated and the large abnormal vein ligated. TB was then transferred to the connecting operating room after separation. The diaphragmatic wall defect was repaired and abdominal wall defects were closed with Lyoplant® (B. Braun Melsungen AG, Germany). TA was infused with 5% dextrose and 0.45% normal saline solution as maintenance drip, and sterofundin 2% dextrose for TB. The twins' blood sugars were monitored. Estimated blood loss for TA was 30–50 ml; 20 ml of packed cells and 20 ml of fresh frozen plasma were transfused. The amount of packed cell transfusion for TB was equal to the amount of blood loss of 16 ml. Total fluids given for TA was 15 ml (10

ml/kg) of sterofundin while TB received 107 ml (approximately 70 ml/kg) of human albumin 5%.

Hypothermia was prevented intraoperatively by use of the radiant heat warmer, warming mattress, forced warmed air, fluid warming devices, and warm fluids. Core body temperature dropped to 35°C during line insertion, but remained above 35°C throughout surgery.

Postoperatively, both babies were transferred to the NICU ventilated. Urine output in TA normalised (1 ml/kg) within 12 hours postoperatively. TB developed anuric acute kidney injury within 18 hours postoperatively, which improved on postoperative day 2. Both babies were alive and well at the time of reporting.

Discussion

There is a recorded total of 25 CT separations performed in Malaysia since 1981. This is the third CT separation in our centre, and the first premature emergency separation due to extrauterine twin-to-twin transfusion. Each surgery involving CT has its unique circumstances, and its success depends on the site(s) of conjunction, organs shared, age and weight of patients, associated malformations, general condition of the twins, occurrence of serious illness in one twin or specific physiologic conditions, and the experience and skills of the attending team.² Our twin babies had liver fusion with venous shunt, and diaphragmatic defects.

Surgical separation is best done electively between 4 and 11 months of age.³ Operative survival is 50% in the neonatal period, as opposed to 90% if surgery is performed after 4 months of age.⁴

Emergency situations warranting immediate separation include the presence of a stillborn twin, intestinal obstruction, rupture of an omphalocele, heart failure, obstructive uropathy, and respiratory failure.² In this case, urgent separation was indicated to avoid complications from significant circulatory crossover, resulting from extrauterine twin-to-twin transfusion, which potentially threatened the lives of both twins. TA developed CT extrauterine transfusion syndrome, with anuria despite creatinine levels within normal limits.⁵ Despite the usual occurrence of vascular shunts and cross-circulation in CT, only a few cases of haemodynamically significant unbalanced circulatory shunting have been reported.⁵ The extent of shared vasculature affects drug pharmacokinetics and pharmacodynamics, as well as fluid and blood administration. Prior to completion of surgical separation,

CVP in TA was maintained at 5–6 mmHg to minimize cross-circulation to TB. TA required packed cell transfusion for hypotension secondary to volume depletion. The use of inotropes was individualized, where TB was started on milrinone and adrenaline for impaired cardiac contractility, whereas adrenaline and noradrenaline were commenced in TA.

The perioperative management of CT requires extensive planning and teamwork with a multidisciplinary approach. The anaesthetic management for this case of CT was challenging due to their size and age at surgery. The younger the baby, the more immature the organs, especially the liver and kidneys, and thus the greater the challenges of all aspects of anaesthesia.⁶ Their smaller size posed problems in terms of monitoring, vascular access, temperature control, and positioning.

Constant vigilance and monitoring were essential, and in this case experts of various fields including the neonatologist, nephrologist, and cardiologist were involved perioperatively. Advances in diagnostic techniques, meticulous anaesthetic management with careful intraoperative monitoring, improved surgical techniques with minimal blood loss, postoperative care with particular attention to potentially labile cardiovascular parameters,⁴ and most importantly, prior experience, all contribute to the improved survival for CT separation.

Conclusion

CT separation is best managed by an experienced multidisciplinary team, functioning in a tertiary referral centre where a full range of medical and surgical specialties are available. The timing and planning of CT separation should be individualized based on the need for emergent separation and the degree of organ fusion. Anaesthesia for premature CT separation is very challenging and requires meticulous attention to details.

Declarations

Informed consent for publication

The guardians provided written informed consent for the publication of the images and clinical data contained in this case report.

Competing interests

None to declare.

Funding

None to declare.

Acknowledgements

We thank the Director General of Health Malaysia for his permission to publish this article.

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