CASE REPORT

The First Successful Separation of Conjoined Twins – Thoracoomphalopagus in RSUP Prof R. D. Kandou Manado: A Case Report

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ABSTRACT

Conjoined twins, a rare congenital anomaly, occur in approximately 1 out of every 100,000 births, and not all cases result in survival. Only about 25% of conjoined twins who are born alive survive and then decide whether they can be separated or not. This separation of conjoined twins is the first case carried out in our institution. These thoraco-omphalopagus conjoined twins experience partial union of the sternal bone, diaphragm, liver, epigastric abdominal wall to infraumbilical. The imaging process showed the appearance of a small vascular shunt between the liver of twin A and the liver of twin B children, and vascularity and other organs that were visualized normally and were present in each of the two twins so that elective separation was decided. Hepatic union occurs in hepar segments 2 - 3 from the right direction (twin A) merging with segments 4a and 2 from the left direction (twin B). Hepatic resection between these segments is done without problems, no bleeding and bile leakage. The sternal bone and xyphoid process are surgically cut, followed by suturing the anterior diaphragm to the new lower end of the sternum. Closure of the abdominal wall, along with umbilicoplasty, is then carried out without tension. Intraoperative and postoperative respiratory and hemodynamic status is progressing well and signs of abdominal compartement syndrome also do not occur. Separation of conjoined twins cases can be done effectively with careful consideration as long as there is no emergency in one of the babies.
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INTRODUCTION

Conjoined twins are an uncommon occurrence, manifesting in approximately 1 in every 50,000 to 100,000 births. However, the actual incidence of conjoined twins in live births can be as low as 1 in 200,000 due to the fact that 60% of babies do not survive either during delivery or shortly afterward.[1,2] Conjoined twins are one of the congenital disorders that are quite grabbing the attention of the medical world both because of the complexity of the disorder and because of its treatment involving various medical disciplines so that it involves a large number of medical personnel. Of all conjoined twin cases, only about one-third of conjoined twin patients survive through the neonatal period and eventually successfully undergo separation.[3]

Siamese twins are categorized based on the areas where they are fused, including ventral union, lateral union, and dorsal union. In ventral union, thoracopagus (fused at the chest) accounts for 19%, omphalopagus (fused at the anterior abdominal wall) for 18%, ischiopagus (fused at the ischium) for 11%, and craniophagus (fused at the head) for 11%.[4]

This case report presents the first successful separation of thoraco-omphalopagus conjoined twins in RSUP Prof R. D. Kandou Manado, Indonesia. Thoraco-omphalopagus twins are a rare type of conjoined twins, and successful separations require meticulous preoperative planning, surgical expertise, and postoperative care. Documenting and sharing the experience of managing such a complex case in a resource-limited setting adds valuable information to the medical literature and can guide

other institutions facing similar challenges.

CASE REPORT

2-year-old female conjoined twins have been diagnosed with Thoracoomphalogus conjoined twins. The two children are designated as "twin A" and "twin B". Both children were born on 19/12/2019. Before separation, the child's weight reaches 20.1 kg with a height of 80cm (twin A) and 82cm (twin B). Both children are fused in the lower sternum - procesus xyphoideus up to the umbilicus. From the outside, a hard solid tissue bridge resembling bone in the lower sternal area with a thickness of 5 cm and the rest is thin soft tissue. The length of the area of joining the skin from the outside is 15 cm long. (Figure 1)

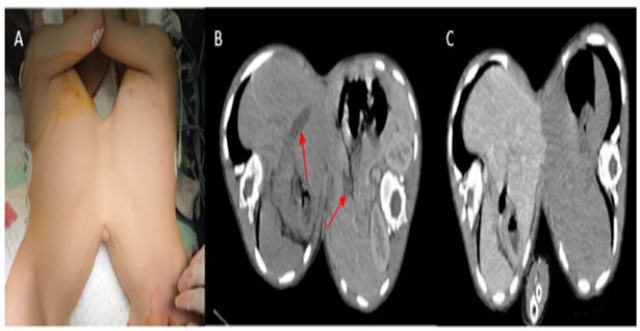

Figure 1: A. The external presentation of thoraco-omphalopagus conjoined twins. B. The CT-scan revealed liver fusion occurring between segments 2-3 in Twin A and segments 2-4A in Twin B, each possessing its own portal and biliary drainage system (indicated by the arrow). C. The portal vein phase displayed no notable shared biliary or portal pedicles within the fused liver.

Preoperative evaluation

The contrast-enhanced CT scan reveals liver fusion involving segments 2 and 3 in Baby A and segments 2, 3, and 4a in Baby B. The extrahepatic biliary system is separated by two gallbladders and two *common bile ducts*.(Figure 1) The other intra-abdominal organs do not fuse with each other. For the thoracic area, the sternal os appear separate with a bridge resembling bone in the fusion area. The heart and lungs are separate and not involved. The anterior side diaphragm is thought to have undergone fusion in its inertial in the proc. Xyphoideus. Echocardiography examination found no heart abnormalities. Blood laboratory tests obtained Hb results of 13.0 g / dL for twins A and 13.9 for child B, with other biochemical values normal for both children. Based on preoperative findings, these patients are assessed to be selectively separable. Written informed consent has been obtained from the parents provided that they accept any risks that may occur during surgery including death.

Timing of surgery

The patient was delivered via elective caesarean section at 39 weeks gestation at our institution, with the diagnosis of conjoined twins having been known since prenatal examinations. There are no complications in the neonatal period, the baby reaches 3 months of age and undergoes the *first imaging* examination.

Considering the uncertain condition of the COVID-19 pandemic, patients undergo the growth and development process with regular monitoring at the growth and development poly. There are no significant problems during the growth and development process until the child reaches the age of 2 years. As they grow, children sometimes experience conflicts where one child begins to grab / minor physical abuse to his own siblings in everyday life. One child's road mobilization sometimes pulls the body of another child. The body movement of one child can reach a rotation of about 1800. Compared to the neonatal period, the fusion areas of both patients are noticeably softer and thinner. The rest of the skin of the anterior thoraabdominal area is thought to cover the primary defect that will form. The second imaging was performed on children aged 2 years with results that were not significantly different from the first imaging. Based on the results of this evaluation and the improvement of the COVID-19 pandemic situation, the patient was decided to undergo separation.

While many institutions aim to separate conjoined twins around 3 months of age, the timing of surgery must be individualized based on the specific type of conjunction, the extent of organ sharing, the clinical stability of the twins, and the resources available at the treating facility. In this case, the 2-year delay did not appear to negatively impact the outcome, highlighting the importance of careful planning and monitoring in the management of these complex patients.

Interoperative finding

Surgical preparation, anesthesia planning, and the positioning of anesthesia and surgical equipment have been thoroughly discussed. Additionally, plans for patient positioning and repositioning post-separation have been established. Blood preparations, including fresh frozen plasma and platelet concentrate, have been arranged for the surgery. The conjoined twins were operated on a single operating table, with another table prepared for abdominal closure surgery post-separation. Anesthesiologists and surgical teams are prepared for each baby, with Twin A being induced first, followed by Twin B. Once anesthesia preparation is complete, both twins undergo aseptic and antiseptic measures before surgery commences on the anterior side. Subsequently, the twins are draped with sterile cloths in readiness for the surgical procedure. The incision is taken in the middle of the network connecting bridge. The skin is cut with a *multiple* Z-plasty incision design, then thoracotomy is performed in the attachment area, further identification appears the xypoideus process is fused to the distal sternum, the diaphragm is fused to release and separation of the xypoid to sternum, pericardium appears and continued liberation and separation of the

diaphragm. In the hepar section, segments 2 - 3 from the right direction (twin A) merge with segments 4a and 2 from the left direction (twin B) are carefully separated using a *harmonic* scalpel.

The surgical procedure progressed to address the back wall of the thoracoabdominal region using a similar approach as on the anterior side. After three hours of surgery, the twins were successfully separated, and Twin B was transferred to another operating table. Diaphragm repair continued using non-absorbable monofilament sutures, along with ongoing chest wall reconstruction for both children. The liver was affixed to the falciform ligament, and the hepatic bed was treated with fibrin glue. Abdominal skin advancement was achieved through multiple z-plasty flaps for both children, followed by umbilicus reconstruction and fixation to the peritoneum. The wound was covered with moist and dry gauze, and an octopus cloth was attached to both children. Both babies tolerated the procedure well, were extubated on the operating table, and subsequently transferred to the pediatric surgical intensive care unit.

During intensive care, both children were fully conscious on the second day and were generally active with stable vital signs. Intravenous antibiotics are given until the tenth day postoperatively. Oral feeding commenced on the fourth day following the operation. Baby B developed a wound infection postoperatively, which was managed conservatively. Both babies were discharged on the 12th day after surgery. They experienced weight gain and underwent follow-up appointments at the pediatric clinic for a week, during which wound care was conducted according to the established protocol.

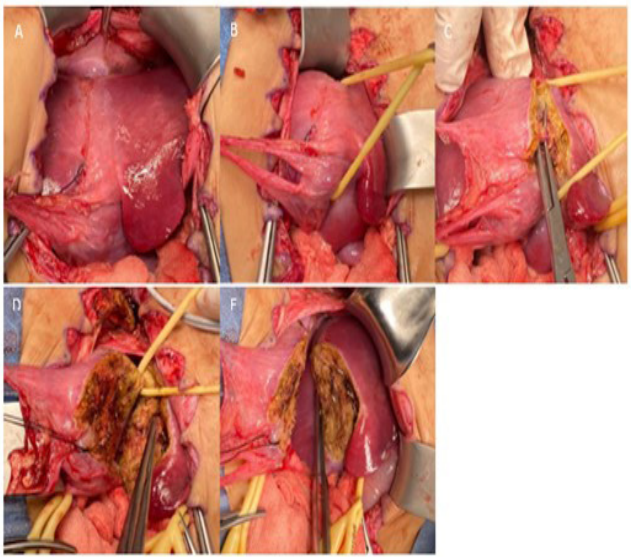

Figure 2: A. Liver fusion observed between segments 2-3 in Twin A and segments 2-4A in Twin B. B. Modified LHM (left hepatic mass) used to ascertain the liver transection plane. C. Intra-parenchymal separation of bile ducts and vessels. D. Control of bleeding from the raw surface after liver separation. E. Completion of liver separation.

DISCUSSION

Conjoined twins are a complex occurrence not tied to hereditary factors or maternal age. They represent a rare variant of twin pregnancies resulting from the delayed division of a single zygote. This phenomenon occurs at an estimated rate of about 1 in every 200 identical twin pregnancies, with an overall incidence ranging from 1 in 50,000 to 100,000 live births. Interestingly, this condition appears to affect females more frequently, with a ratio of 3 to 1. Conjoined twins typically share a single chorion, placenta, and amniotic sac, although these characteristics are not exclusive to them, as some non-conjoined monozygotic twins also share these structures. Like all monozygotic twins, conjoined twins are always of the same sex.[1,2]

The medical term used to describe conjoined twins ends with the suffix "pagus," derived from the Greek word meaning "fixed" or "attached." There are five described types of conjoined twins: [1]
1. Thoracopagus (attached at the thorax or chest).
2. Omphalopagus (attached at the anterior abdominal wall).
3. Craniopagus (attached at the cranium or head).
4. Syncephalus (twins joined at one head).
5. Ischiopagus (attached at the buttocks).

The most prevalent type of conjoined twins involves the fusion of the anterior thorax and/or abdomen, termed thoracopagus, omphalopagus, and thoraco-omphalopagus, accounting for approximately 70% of cases. Some cases, particularly thoracopagus conjoined twins, exhibit an increased thickness of nuchal folds. The cause of conjoined twins remains unknown, with two theories proposed to explain their origin:[5]
1. Fission theory suggests incomplete splitting of the fertilized egg, leading to delayed separation of the embryonic mass after the 12th day of fertilization.
2. Fusion theory proposes complete separation of the fertilized egg, followed by merging of stem cells from one twin with similar cells from the other twin, resulting in simultaneous fusion.

Diagnosis of conjoined twins can occur as early as the 10th week of gestation. Various imaging modalities such as two- and three-dimensional ultrasound, CT scan, or MRI are utilized to assess the type and severity of the abnormality. Survival rates for conjoined twins are low, with approximately 65% being stillbirths and 35% of those born alive dying within the first 24 hours. Only an estimated 25% survive to an age suitable for surgical separation.[1,4,5]

Surgical treatment for conjoined twins falls into three categories:
1. Category I: No surgical intervention is considered feasible if cardiac fusion precludes the composition of a

single, functioning heart.
2. Category II: Emergency separation is warranted if one twin has died, if one twin's life is at risk and threatens the other, or if untreated anomalies render survival impossible.
3. Category III: Planned separation is undertaken when the infant's condition is stable enough for necessary imaging evaluations to accurately map out the separation.

While thoraco-omphalopagus twins generally have the best survival prospects, meticulous preoperative planning and comprehensive team management are essential. Radiological investigations are crucial for assessing incorporated organs, anomalies, and the presence or absence of cross-circulation. Surgical separation is typically performed around three months of age, when physiological conditions and tissue flexibility are optimal. [3]

Factors considered during separation include joint organs, soft tissue integrity, potential bone defects post-separation, the child's age, and associated anomalies. Success rates heavily rely on accurate preoperative assessment, a multidisciplinary approach, surgical expertise, and meticulous postoperative care. Despite advancements, separation surgeries remain high-risk endeavors, with outcomes ranging from relatively straightforward to exceedingly complex, often posing life-threatening risks. Successful outcomes are less likely if cardiac fusion is present; however, separation success hinges on the condition of other organs. Post-separation, large areas of exposed tissue are common, making primary wound closure challenging and increasing the risk of postoperative complications such as sepsis.[3]

Comprehensive preoperative evaluation using advanced imaging modalities is crucial for assessing the extent of organ sharing and planning the surgical approach. In this case, CT scans revealed the twins shared segments of the liver, but had separate biliary and vascular systems, making separation feasible. A multidisciplinary team approach involving pediatric surgeons, anesthesiologists, radiologists, and intensive care specialists is essential for the successful management of conjoined twins before, during, and after separation surgery.

Meticulous surgical technique, including careful liver transection using a harmonic scalpel and precise closure of the abdominal wall, is critical for minimizing blood loss and preventing postoperative complications. The use of fibrin glue on the liver bed and multiple Z-plasty flaps for skin closure were effective strategies employed in this case. Close postoperative monitoring and wound care are important for detecting and managing complications. Despite the development of a wound infection in one twin, both children recovered well and were discharged home on postoperative day 12.

CONCLUSION

In this case, we successfully closed the abdominal wounds of both babies, despite Twin B experiencing a suture wound infection necessitating daily wound care. Research regarding survival and long-term outcomes, particularly in relation to educational and psychological outcomes, is ideally conducted on surgically separated conjoined twins.

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