"A CASE REPORT OF THORACO-OMPHALOPAGUS CONJOINED TWINS: THE DOWNFALL OF THE SEPARATED HEARTS"

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ABSTRACT

Background: Thoraco-omphalopagus conjoined twins is a rare occurrence of monozygotic pregnancies which involves fusion of the anterior thorax and abdomen. This type of conjoined twins are known to present a variety of cardiac anomalies, which contribute to their generally unfavorable prognosis.

Case Presentation: A 32-years-old multigravida with Gravida 6, Para 4, Abortus 1, was referred from a rural area in the eastern region of Indonesia at 28 weeks of gestational age due to suspicion of conjoined twins. Ultrasonography examination revealed the presence of thoraco-omphalopagus conjoined twins which the fetuses joined ventrally to each other. Prenatal MRI disclosed sharing of a single liver, omentum and diaphragm. Partial fusion was observed in the sternal bone, pericardium, and anterior wall of the hearts. However, with two separate heart chambers and distinct unsynchronized fetal heartbeats. Classical approach of cesarean section was performed due to sign of labor at the gestational age of 38 weeks, followed by a procedure of female sterilization. Pathological examination revealed a single placenta with one umbilical cord attached, suggestive of monochorionic monoamniotic twin pregnancy. Healthy female babies were born with a combined weight of 5400 grams with adequate APGAR score. Gross appearance of anterior fusion was seen on both babies at the level of chest and abdomen. Post-delivery echocardiography revealed the presence of a cardiac anomaly characterized by malposition of the great arteries in one of the twins. After 13 hours of close monitoring in the Neonatal Intensive Care Unit, the twins passed away due to cardiac complication.

Conclusion: The management of pregnancies involving thoraco-omphalopagus conjoined twins requires a comprehensive and multidisciplinary approach. This approach aims to provide holistic care, addressing the complex medical risks, complications, and ethical dilemmas associated with this condition.

Keywords: Conjoined Twins; Thoraco-omphalopagus; Monozygotic Twins; Congenital Cardiac Anomaly

BACKGROUND

Conjoined twins, also referred to as Siamese twins, is a highly uncommon occurrence in monoamniotic pregnancies. These twins are anatomically joined to one another, often sharing similar tissues, organs, or even larger body structures. The etiology of conjoined twins is believed to be associated with abnormal embryonic development, with a prevalence estimated at approximately 1.5 per 100,000 births. The thoraco-omphalopagus twins account for 70% of conjoined twins and are associated with the highest mortality rates of 51% due to complex cardiac anomalies.[1–4]

This phenomenon occur when the division of a zygote initiates after the 13th day of conception. This division results in two embryonic primordia, instead of one, within a single embryoblast, leading to the formation of two embryonic disks positioned closely to each other. Conjoined twins are categorized based on the site of attachment, indicated by the suffix "pagus" derived from the Greek term for "fixed". For instance, thoracopagus is a conjoined twins which fused at the thorax (chest). Each classification has a distinct embryological origin, contributing to the complex anatomical structures observed in conjoined twins.[2,3]

Aid of diagnostic imaging techniques, including ultrasonography and magnetic resonance imaging (MRI), enables physicians to achieve prenatal diagnosis in cases of conjoined twins. This allows for proactive anticipation and preparation for potential complications that might occur along the process of management.[3]

Caesarean sections have emerged as the preferred mode of delivery for conjoined twins. In the case of thoraco-omphalopagus twins, a classical incision is preferred during the cesarean section. This particular approach offers improved visualization and facilitates the delivery of twins, given the intricate anatomical structures and potential risk of obstruction. However, it is important to acknowledge that the classical approach is associated with certain drawbacks, including an elevated risk of maternal morbidity. [3,5]

This paper presents a case report of a pregnancy involving thoraco-omphalopagus conjoined twins. Additionally, a narrative review of the existing literature is provided, focusing on the diagnosis, procedure, and ethical consideration of this case. The aim of this paper is to contribute to the understanding of this condition and provide valuable insights for healthcare professionals involved in the management of conjoined twins.

CASE PRESENTATION

A 32-year-old multigravida, with a history of Gravida 6, Para 4, Abortus 1, was referred to our institution from a rural area in the eastern region of Indonesia due to suspected conjoined twins. The patient was transferred to our department at 28 weeks of gestational age (GA). There was no reported history of specific drug use, alternative medicine, radiation exposure, or fever during the first trimester of her pregnancy. Furthermore, the couple had no prior medical conditions or family history of congenital anomalies.

During the Leopold examination, multiple fetal parts were palpable, and the fetal heart rates of fetus 1 and fetus 2 were found to be 132 and 138 beats per minute, respectively. A comprehensive ultrasonography examination conducted at our institution (refer to Fig. 1) revealed the presence of thoraco-omphalopagus conjoined twins in a Monochorionic Monoamniotic (MCMA) twin pregnancy, with the fetuses joined ventrally to each other. To further elucidate the connected structures of both fetuses, a prenatal MRI was performed (refer to Fig. 2). In the conjoined twins, it was observed that they shared a single liver and diaphragm. Additionally, a shared thoracic and abdominal cavity was noted. Partial fusion was observed in the sternal bone, pericardium, and anterior wall of the hearts. However, the fetuses had two separate heart chambers with distinct unsynchronized fetal heartbeats. Another area of possible fusion was identified on the omentum, but no further details regarding the surrounding intestines could be discerned. Moreover, the two fetuses exhibited two heads, four extremities, two stomach bubbles, two spines, separate pelvises, and distinct urinary tracts.

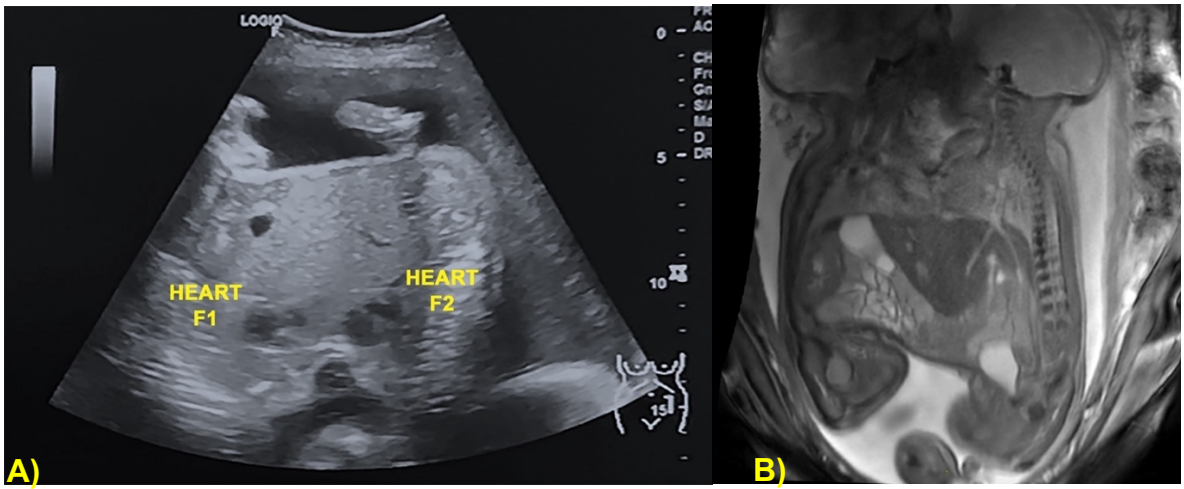

Figure 1. Radiological Evaluation of Thoraco-omphalopagus Conjoined Twins
A) Antenatal Ultrasonography Examination at 28 weeks of gestational age (GA) demonstrating two unsynchronous fetal heartbeats. B) Prenatal MRI at 32 weeks of GA revealing ventral fusion characterized by a shared sternum, diaphragm, and abdominal wall. The imaging also shows fusion of the hearts, diaphragm, liver, and possibly the omentum.

A multidisciplinary team comprising obstetricians, perinatologists, pediatric surgeons, thoracic surgeons, anesthesiologists, and members of the hospital's ethical committees was established to provide comprehensive management for the conjoined twin case. The team conducted thorough counseling sessions with the couple, during which they discussed the condition, potential outcomes, and prognosis of the conjoined twins. The malformations found were also explained, along with the likely outcome if the twins were to survive after delivery. Throughout the perinatal consultation, the parents did not express any intention to terminate the pregnancy and ultimately made a decision to continue the pregnancy to term.

At 33 weeks of GA, the patient presented with contractions and was subsequently admitted for the administration of antenatal steroids to promote lung maturation. A total of four doses of 6 mg of Dexamethasone were given intramuscularly, with each dose administered every 12 hours. Throughout the hospitalization period, cardiotocographic monitoring was

performed to ensure the well-being of the healthy fetuses. After three days of hospitalization, the patient was discharged without any further complaints and close follow-up was then provided as an outpatient in our fetomaternal clinic.

Cesarean section was performed due to sign of labor at the GA of 38 weeks. Classical approach of caesarean section was done with vertical incision in the midline of the abdomen, from just above the pubic bone to the level of the umbilicus, to allow better exposure and access to the uterus (See Fig. 2A). Another vertical incision is made on the uterus to better accommodate the delivery of both conjoined twins. (See Fig. 3A) The twins were delivered by breech extraction (See Fig. 3C). The obstetrician carefully delivered the twins' buttocks and legs first, followed by the remaining portions of their bodies. The procedure was completed efficiently without encountering any significant obstacles and the umbilical cords were promptly clamped and cut. Following that, a procedure for female sterilization was carried out by performing bilateral ligation of the fallopian tubes using the Pomeroy Modification technique. Subsequently, the surgical team proceeded to close the incisions using sutures and implemented measures to ensure proper hemostasis.

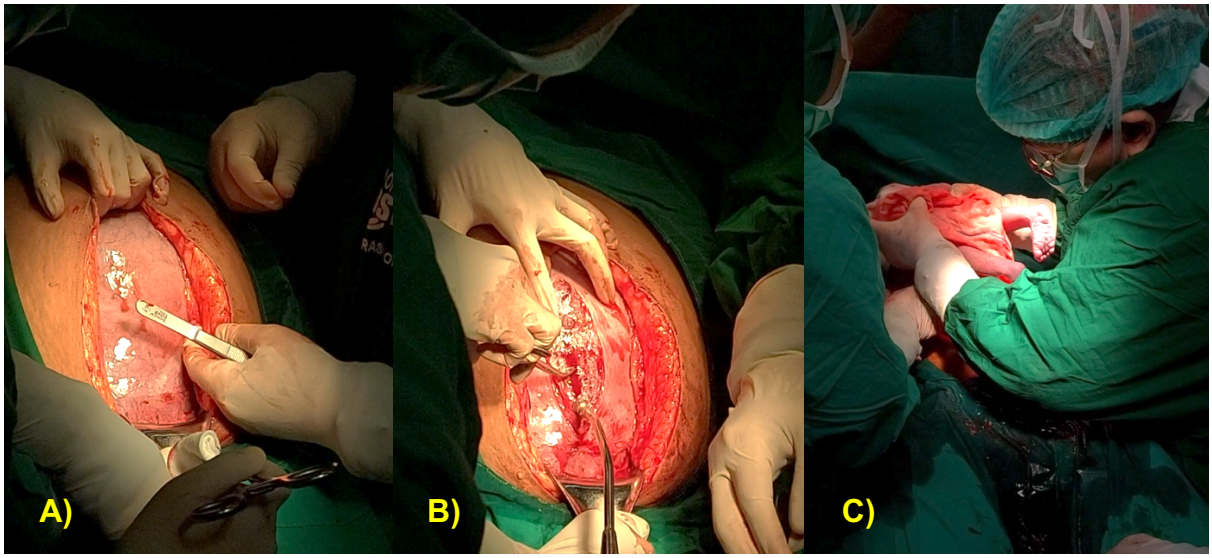

Figure 2. **Termination Process of Conjoined Twins with Classical Caesarean Section**
A) A vertical incision was made on the skin, extending through the peritoneum. B) Another vertical incision was performed on the uterus. C) The twins were in a breech presentation, and the operator successfully delivered them without encountering any significant obstacles.

A single placenta was delivered from the uterus, and upon gross examination, it displayed a single umbilical cord attached to the central part of the placenta. The gross appearance revealed a single layer of amniotic membranes without any notable amniotic septum. The placenta, along with a portion of the umbilical cord, was sent to the pathology lab for further analysis. Pathological examination revealed an umbilical cord with three vessels, including two arteries and one vein, surrounded by Wharton's jelly and chorionic villi. These findings are consistent with a MCMA twin pregnancy.

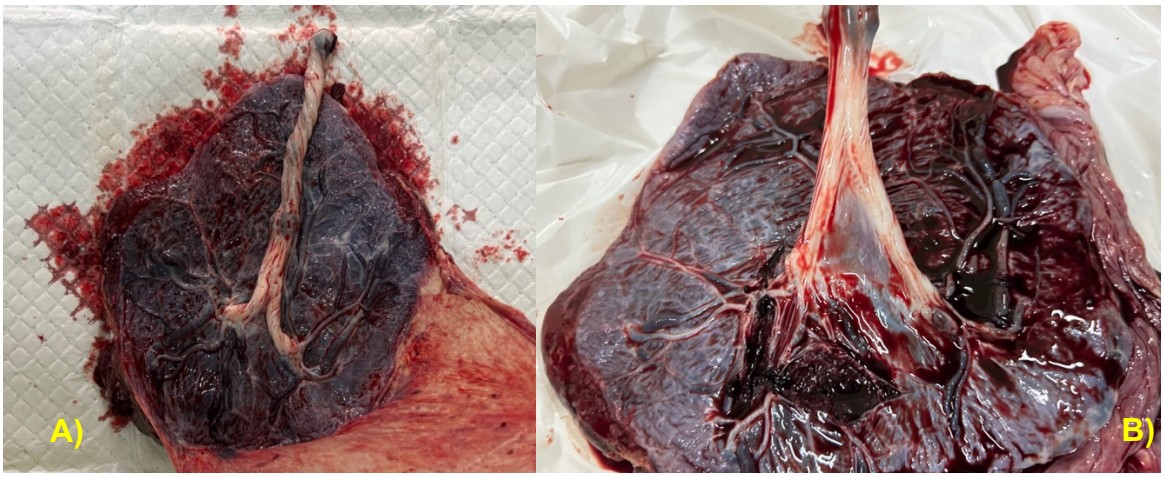

Figure 3. Placenta of Monochorionic Monoamniotic (MCMA) Twin Pregnancy
A) Gross appearance of a single placenta with a visible layer of amniotic membrane without septum. B) Depiction of one umbilical cord attached to the fetal portion of the placenta

Healthy female babies were born with a combined weight of 5400 grams. They showed Apgar scores of 8 and 10 at the first and fifth minute, respectively. Both babies cried immediately with adequate muscle tone. All vital signs were within normal limit with a notable oxygen saturation of 97% of both babies without any respiratory support, the babies scored 0 for DOWNE score which indicated no respiratory distress. According to the New Ballard score, the babies displayed characteristics consistent with a term gestational age of 38-40 weeks. Upon examination, Gross appearance of anterior fusion was seen on both babies at the level of chest and abdomen. Additionally, a single umbilical cord was attached to the central part of the fusion. Images of the conjoined twins following termination are presented in Figure 4. No other significant congenital anomalies were noted.

Figure 4. Thoraco-Omphalopagus Conjoined Twin in Stable Condition After Classical Caesarean Delivery. The image displays gross appearance of anterior fusion at the level of the chest and abdomen in a thoraco-omphalopagus conjoined twin. Only one umbilical cord is visible, attached to the central part of the fusion. The twins are shown to be in a stable condition, captured 10 minutes after delivery.

After delivery, both twins were transferred to the Neonatal Intensive Care Unit (NICU) under the care of the perinatologist for close monitoring. The multidisciplinary team promptly conducted a comprehensive assessment and initiated a thorough follow-up plan, which included laboratory examinations, echocardiography, and a scheduled for Computed Tomography Scan. Echocardiography performed within 30 minutes of birth revealed satisfactory contractility of both hearts. However, Fetus 1 was observed to have a malposition of the great arteries, while Fetus 2 did not exhibit any congenital heart anomalies. The laboratory results indicated moderate anemia in both babies, prompting the decision to proceed with a blood transfusion using Packed Red Cells.

For the first 10 hours, the babies remained stable. However, they subsequently developed bradycardia, resulting in the decision to initiate intubation and administer adrenergic support. Unfortunately, the twins did not respond adequately, as their pulses remained below the normal range. Bedside echocardiography revealed reduced contractility in both hearts. Despite close monitoring in the NICU, the conjoined twins experienced a progressive decline and passed away 13 hours after birth. The parents did not provide consent for an autopsy. As for the mother, no notable complications occurred during the postoperative period and she was discharged on the third day following the surgery.

DISCUSSION

Conjoined twins is a rare occurrence, with a prevalence of approximately 1.5 per 100,000 births. This condition is most commonly observed in MCMA twin pregnancies. Approximately half of conjoined twin pregnancies result in stillbirths; moreover, within the first 24 hours, 35% of conjoined twins do not survive, leaving only 15% to 18% of cases progressing from in utero ultrasonographic diagnosis to successful separation. [1,4–6]

Thoraco-omphalopagus twins are the most frequently encountered type of conjoined twins, accounting for approximately 70% of cases. Unfortunately, this subtype has the highest mortality rates, with a staggering 51% of cases resulting in death. This high mortality rate is primarily attributed to complex cardiac anomalies associated with thoraco-omphalopagus twins. The presence of these intricate heart defects significantly impacts the survival chances which results in the lowest overall survival rate. [1,4–6]

It is suggested that exposure to certain environmental factors during a critical period of pregnancy may disrupt the normal functioning of the spindle apparatus which may lead to abnormal process of cell cleavage. Additionally, some studies have proposed that low calcium levels in the mother can prolong the preimplantation phase and weaken intercellular bonds, thereby increasing the likelihood of monozygotic twinning. It has also been observed that pregnancies following assisted reproduction and infertility treatment have a higher rate of twinning.[1,3]

The etiology of conjoined twins is believed to be associated with abnormal embryogenesis. Conjoined twins are formed when zygote division occurs after the 13th day of conception. Instead of a single embryonic primordium, two embryonic disks develop in close proximity to each other. The degree of conjoinment and the timing of fusion determine the

extent of anatomical adaptations. In ventrally conjoined twins, the embryonic disks are fused at their anterior aspects. As the fusion progresses, there is sharing of umbilical, abdominal, thoracic, and even craniofacial structures. [1–3]

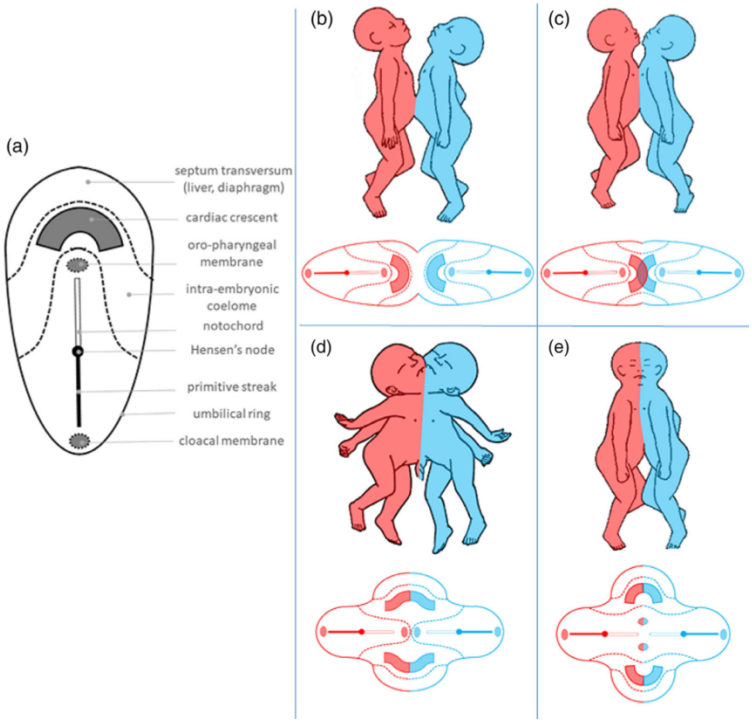

Figure 5. The Embryonic Disk Model in Ventral Conjoined Twins.
A) Schematic representation of the embryonic disks during gastrulation, prior to the folding process that forms the primordial embryonic body. B-E) In ventral conjunction, the initial separation between the antero-posterior opposing disk primordia determines the degree of sharing and involvement in the process of neo-axial orientation. This process gives rise to compound structures and organs. *(Image adapted from Oostra et al., 2022)*[2]

Conjoined twins are classified based on their site of attachment with a suffix "pagus," the Greek term for fixed. The main types are omphalopagus (abdomen), thoracopagus (thorax), cephalopagus (ventrally—head to umbilicus), ischipagus (pelvis), parapagus (laterally—body side), craniopagus (head), pygopagus (sacrum), and rachipagus (vertebral column). Each class has its own unique embryological basis of origin giving rise to their complicated anatomies.[3]

The most prevalent form of conjoined twins involves fusion of the anterior thorax and/or abdomen, which is referred to as thoracopagus, omphalopagus, and thoraco-omphalopagus, the latter which our case is presented with. This category encompasses approximately 70% of all conjoined twin cases. Omphalopagus twins are connected at the abdomen, extending from the lower thorax to the groin, while thoracopagus twins are joined ventrally at the upper chest, typically involving the sternum, diaphragm, and a portion of the abdominal wall. Even the presence of a single blood vessel or any sharing of cardiac tissue classifies twins as thoracopagus. In our case, there is fusion of both the thoracic and abdominal organs, but only partial fusion of the hearts, with connection limited to the anterior wall and the pericardium. The lungs are generally unaffected, which is consistent with our case. Although these twins may

share the same pericardial cavity, it is uncommon for this type to share vascular supply or cardiac musculature. The liver and diaphragm are almost always shared, with the extent of fusion varying based on the size of the fusion. Notably, our case also presented with fusion of the abdominal organs, including the liver and omentum.[3,7]

Ultrasonography serves as the primary diagnostic tool for conjoined twin pregnancies. During the first trimester, certain ultrasound features should raise suspicion, including the characteristic monochorionic pregnancy features (single yolk sac, absence of separating amniotic membrane, and single placenta). Additional features that warrant attention include hyper-reflexion of the spine and the absence of relative changes in the position of the twins towards each other during consecutive ultrasound assessments. Prenatal ultrasound can be performed as early as 12 weeks of gestation. Recent studies have shown that prenatal fetal MRI can provide valuable information to define precise anomalies and anatomical connections between the fetuses. In our case, the patient presented at 26 weeks of gestational age and already displaying prominent signs of thoraco-omphalopagus structures. Further investigation with MRI confirmed the initial suspicion.[1,3]

Proper diagnosis plays a crucial role in determining the obstetric management of conjoined twins, including decisions regarding the method and timing of delivery. The recommended approach in the third trimester is to perform a cesarean section, as this minimizes the potential damage to shared fetal tissues, reduces the risk of internal hemorrhage, and lowers the likelihood of intrapartum death. Specifically, in the case of delivering thoraco-omphalopagus twins via cesarean section, it is ideal to use a midline vertical incision. This incision technique provides improved visualization and facilitates the delivery of twins, considering the high risk of obstruction posed by complex anatomical structures. In our case, the extraction process was completed in less than a minute, reducing the risk of intrapartum pressure on the twins. Following delivery, the APGAR scores of the babies were satisfactory, indicating that this approach favour the safety of the twins.[3,5,8]

However, the classical approach of caesarean section is associated with certain drawbacks, including an increased risk of maternal morbidity. Complications such as puerperal infection, the need for blood transfusion or hysterectomy, and even maternal death can occur. Additionally, there is a higher likelihood of uterine rupture in subsequent pregnancies. Fortunately, in our case, the surgical procedure proceeded without significant complications, and the postoperative period was uneventful. Considering the potential risks of future pregnancies following this procedure, the patient was counseled and provided informed consent for a female sterilization procedure. This decision was based on factors such as having already four biological living children and her advancing age.[5]

The postnatal course of non-operative management is chosen when complex cardiac anomalies make separation and reconstruction of a functional heart unfeasible. Thoracopagus twins can exhibit a range of cardiac anomalies, classified into four groups: Group A with separate hearts and separate pericardium, Group B with separate hearts but a common pericardium, Group C with fused atria and separate ventricles, and Group D with atrial and ventricular fusion. Most thoraco-omphalopagus twins fall into Group A, while a majority of thoracopagus twins have Group C and D anomalies. In our case, the twins belong to Group B

in which the hearts share pericardium. The presence of complex cardiac anomalies contributes to the poor prognosis of thoracopagus twins. Consistent with our case where one of the fetuses had malposition of the great artery. Overall, the prognosis for thoracopagus twins is very poor. In a systematic review conducted by Saxena (2023), out of 158 reported sets of thoracopagus twins, 71 were deemed non-operable and all eventually passed away. Even with a comprehensive team and close monitoring in the NICU, the twins in our case developed cardiac complication and underwent a demise in less than 24 hours. This unfortunate outcome occurred despite the initial presence of sufficient cardiac contractility.[4]

The sudden regression in the twins' condition after initially being born in good health raises questions about the underlying factors contributing to this change. While it's challenging to determine one exact cause, a few potential factors that could be considered:

(a) **Cardiac decompensation:** The presence of a cardiac anomaly might have initially allowed the babies to maintain adequate cardiac function in utero and shortly after birth. However, as time progressed, the stress of transitioning to the outside environment and the demands on the compromised cardiac system could have exceeded its capacity, leading to decompensation and a decline in their condition.

(b) **Hemodynamic instability:** Conjoined twins with complex cardiac anomalies may experience unstable hemodynamics, where the balance between cardiac output and systemic circulation becomes compromised. Initially, the babies may have adapted to this instability, but over time, their condition could have deteriorated due to the challenges of maintaining adequate perfusion and oxygenation.

(c) **Impaired respiratory function:** Conjoined twins with certain cardiac anomalies might also experience respiratory challenges. The initial stability in their condition could have been supported by their innate respiratory efforts, but as the demands on their respiratory system increased, they may have experienced respiratory distress, leading to a decline in overall well-being. The twins encountered shortness of breath and were intubated.

(d) **Metabolic and physiological demands:** The newborn period is a critical time of transition, during which various physiological systems undergo significant changes. The presence of a cardiac insufficiency could have limited the babies' ability to meet the metabolic demands of the transition process, resulting in a delayed regression in their condition.[4]

In clinical management, it is important to consider the parents' social situation, religious beliefs, and psychological factors. Following ultrasonography, many expectant couple choose to terminate the pregnancy due to the low survival rate and the high risk of termination. Even if the mothers decide to continue the pregnancy, uncertainty concerning the neonates' survival chances creates a dilemma for the physicians. In some countries, legally permitted abortion may be considered for conjoined twins with poor prognoses, particularly for thoracopagus twins with low likelihood of successful separation and survival. In our case, it is crucial to ensure that the parents fully understand the risks and prognosis for both the mother and the offspring. This is necessary to adequately prepare them mentally and physically, as they had chosen to continue the pregnancy with the hope of viable babies.[5,6,9]

CONCLUSION

Conjoined twins is a rare complication of monoamniotic pregnancies that present several medical risks and ethical dilemmas. In this study, we report a case of successful follow-up and termination of thoraco-omphalopagus conjoined twins. The partial fusion of the hearts along with the congenital abnormalities ultimately led to the demise of both infants within 13 hours after delivery even after a thorough observation. A comprehensive approach is crucial in managing cases with conjoined twins, involving an arrangement of multidisciplinary team that focuses on holistic aspects in the prenatal, intrapartum, and postnatal period of both the twins and the mother.

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