

A Case Report of Thoraco-Omphalopagus Conjoined Twins: The Downfall of the Separated Hearts

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Abstract

Introduction: Thoraco-omphalopagus conjoined twins are a rare occurrence of monozygotic pregnancy that involves fusion of the anterior thorax and abdomen. This type presents a variety of cardiac anomalies, which contribute to its generally unfavorable prognosis.

Case Presentation: A 32-year-old multigravida with Gravida 6, Para 4, and Abortus 1 was referred at 28 weeks of gestation. Ultrasonography revealed thoracomphalopagus-conjoined twins in which the fetuses joined ventrally. Prenatal MRI revealed the sharing of a single liver, omentum, and diaphragm. Partial fusion was observed in the sternal bone, pericardium, and anterior wall of the hearts, but with separated heart chambers and unsynchronized heartbeats. Classical cesarean section was performed at 38 weeks of gestational age. Histopathology revealed a single placenta with one umbilical cord, suggesting monochorionic–monoamniotic pregnancy. Healthy female babies were born with a combined weight of 5400 g. Post-delivery echocardiography revealed a cardiac anomaly characterized by malposition of the great arteries in a twin. After 13 h of close monitoring in the NICU, the twins died due to cardiac complications.

Conclusion: The management of pregnancy involving thoraco-omphalopagus conjoined twins requires a comprehensive and multi-disciplinary approach aiming to provide holistic care, addressing complex medical risks, and ethical dilemmas associated with these twins.

Key words: Conjoined Twins; Monozygotic Twins; Thoraco-omphalopagus

Laporan Kasus Mengenai Kembar Siam Torako-Omfalopagus: Prognosis Buruk pada Jantung yang Terpisah

Abstrak

Pendahuluan: Kembar siam thoraco-omphalopagus adalah kejadian langka pada kehamilan monozigot yang melibatkan penyatuan antara toraks dan abdomen anterior. Tipe ini diketahui dapat disertai adanya kelainan jantung yang berkontribusi terhadap prognosis yang umumnya kurang baik.

Presentasi Kasus: Multigravida berusia 32 tahun dengan Gravida 6, Para 4, Abortus 1, dirujuk pada usia kehamilan 28 minggu. Ultrasonografi menunjukkan kembar siam torako-omfalopagus di mana janin menyatu secara ventral. MRI prenatal memperlihatkan adanya fusi organ hati, omentum dan diafragma. Di sisi lain, fusi parsial diamati pada sternum, perikardium, dan dinding anterior jantung, dengan ruang jantung terpisah dan detak jantung yang tidak bersifat sinkron.

Seksio sesarea klasik dilakukan pada usia kehamilan 38 minggu. Histopatologi menunjukkan plasenta tunggal dengan satu tali pusat sugestif kehamilan monokorionik-monoamniotik. Dua bayi perempuan sehat lahir dengan berat kombinasi 5.400 gram. Ekokardiografi pascasalin menunjukkan adanya anomali jantung berupa malposisi arteri besar pada salah satu bayi. Setelah 13 jam observasi ketat di NICU, kedua bayi kembar meninggal akibat komplikasi jantung.

Kesimpulan: Penatalaksanaan kehamilan kembar siam thoraco-omphalopagus memerlukan pendekatan komprehensif dan multidisiplin yang bertujuan untuk memberikan perawatan holistik, mengatasi risiko medis, dan dilema etika yang terkait.

Kata kunci: Kembar Siam; Kembar Monozigotik; Thoraco-omphalopagus

Introduction

Conjoined twins, also referred to as Siamese twins, are 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 1.5 per 100,000 birth. This abnormal 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. The thoraco-omphalopagus twins account for 70% of conjoined twins and are associated with the highest mortality rate of 51% due to complex cardiac anomalies.¹⁻⁴

Diagnostic imaging techniques, including ultrasonography and magnetic resonance imaging (MRI), enable physicians to achieve prenatal diagnosis in cases of conjoined twins. This allows for proactive anticipation and preparation for potential complications that might occur during the management process. In the case of thoraco-omphalopagus twins, a classical incision is preferred during cesarean section.^{3,5}

This is a case report of a pregnancy involving thoraco-omphalopagus conjoined twins that presents a narrative review of the existing literature, focusing on the diagnosis, procedure, and ethical considerations. We contribute to the understanding of this condition and provide valuable insights for health care professionals involved in the management of conjoined twins. The mother provided written informed consent to obtain permission to publish the case of her babies.

Case Presentation

A 32-year-old G6P4A1 multigravida was referred to our institution at 28 weeks of gestational age (GA). No history of drug use, alternative medicine, radiation exposure, or first-trimester fever. The couple had no medical conditions or family history of congenital anomalies. Ultrasonography (Fig. 1B) identified thoraco-omphalopagus conjoined twins in a monochorionic monoamniotic pregnancy. Prenatal MRI (see Fig. 1B) confirmed shared thoracic and abdominal cavities, a single liver and diaphragm, and partial fusion in the sternal bone, pericardium, and anterior wall of the heart, but with separated heart chambers and unsynchronized heartbeats.

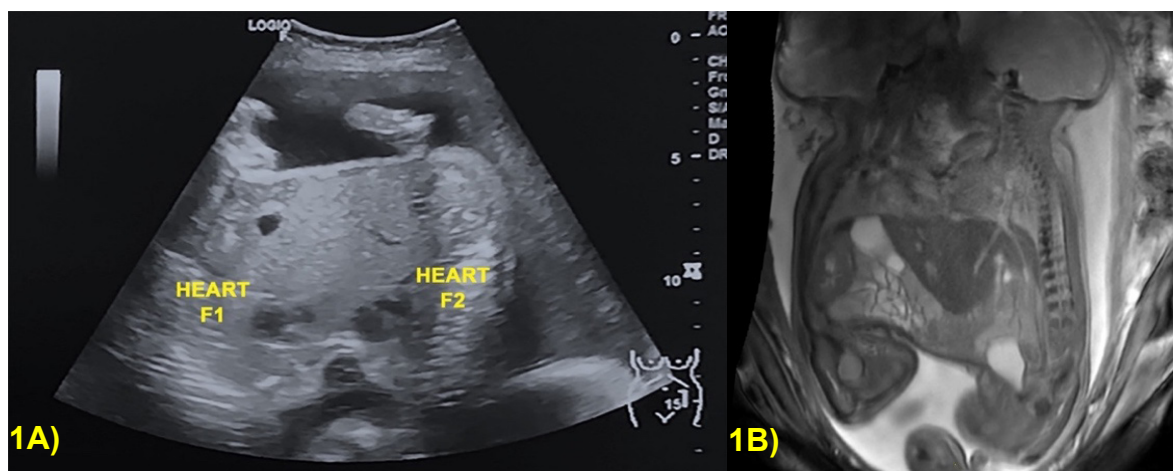


Figure 1 Radiological Evaluation of Thoraco-omphalopagus Conjoined Twins. A) Antenatal Ultrasonography at 28 weeks of gestation. B) Prenatal MRI at 32 weeks of gestation.



Figure 2 Thoraco-Omphalopagus Conjoined Twin in Stable Condition After Delivery.

A) Anterior chest and abdominal fusion in a thoraco-omphalopagus conjoined twin. B) A single placenta with an observable amniotic membrane without septum. C) A single umbilical cord attached to the fetal part of the placenta.

A multidisciplinary team, including obstetricians, perinatologist, pediatric surgeons, thoracic surgeons, anesthesiologists, and ethics committee members, was established to comprehensively manage this case. Extensive counseling sessions with the parents were conducted to discuss the maternal risk, malformations, and potential postnatal outcomes. Throughout the consultation, the parents did not express any intention to terminate the pregnancy. A classical cesarean section was performed at GA of 38 weeks with breech extraction. Subsequently, female sterilization was conducted using the Pomeroy Modification technique.

Healthy female babies weighing 5400 g were born with APGAR scores of 8/10 without notable respiratory distress. Figure

2A displays an image of the conjoined twins following termination. A single placenta (Fig. 2B-C), featuring a single umbilical cord and a layer of amniotic membrane, was sent for pathology analysis, which revealed three vessels (two arteries and one vein) surrounded by Wharton's jelly and chorionic villi, consistent with the findings of MCMA twin pregnancy.

After delivery, both twins were transferred to the NICU, and the multidisciplinary team promptly initiated comprehensive assessment, including laboratory tests, echocardiography, and scheduled CT scan. Echocardiography within 30 min of birth showed satisfactory contractility in both hearts; however, there was a finding of malposition of the great arteries in one of the

twins. For the first 10 h, both babies remained stable but later developed bradycardia, leading to intubation and adrenergic support. Subsequent bedside echocardiography revealed reduced contractility in both hearts. Despite close monitoring, the conjoined twins progressively deteriorated and died 13 h after birth. The mother experienced no significant postoperative complications and was discharged 3 days postoperatively.

Discussion

Conjoined twins are rare MCMA twin pregnancies, occurring at approximately 1.5 per 100,000 birth. Roughly half of these pregnancies result in stillbirths, and within the first 24 h, 35% of conjoined twins do not survive, leaving only 15-18% for potential separation. The high mortality rate is mainly due to complex cardiac anomalies. Conjoined twins are associated with abnormal embryogenesis when zygote division occurs after the 13th day of conception. Instead of a single embryonic primordium, two embryonic disks develop closely. In ventral conjunction, the initial separation between the anteroposterior opposing disk primordia influences sharing and neoaxial orientation. The most common form of conjoined twins involves fusion of the anterior thorax and abdomen, accounting for approximately 70% of cases. Although these twins share the same heart, it is uncommon for them to share vascular supply, whereas the liver and diaphragm are typically shared.¹⁻⁷

Ultrasonography can detect monochorionic pregnancy signs such as a single yolk sac, absence of an amniotic membrane septum, and a single placenta. In this case, at 28 weeks GA, prominent thoraco-omphalopagus features were observed and later confirmed by MRI. Accurate diagnosis is vital for determining the delivery method and timing. In the third trimester, a classical cesarean section was performed to minimize

harm to shared fetal tissues. A midline incision aid delivery in patients with complex anatomical structures. Swift extraction in this study led to satisfactory APGAR score post-delivery, affirming this approach's fetal safety. However, classical cesarean section has drawbacks, including increased maternal morbidity and risk of uterine rupture in subsequent pregnancies. Fortunately, in this study, the procedure and postoperative period proceeded without significant complications, and the patient consented for female sterilization.^{1,3,5,8,9}

The postnatal course of non-operative management is chosen when complex cardiac anomalies preclude separation and heart reconstruction. Thoracopagus twins may exhibit varied cardiac anomalies, which are grouped into four categories: Group A (separate hearts and separate pericardium), Group B (separate hearts but a common pericardium), Group C (fused atria and separate ventricles), and Group D (atrial and ventricular fusion). In this study, the twins were classified into Group B, also with a cardiac anomaly of malposition of the great arteries. In a systematic review by Saxena (2023), 71 of 158 thoracopagus twin cases were considered inoperable, resulting in eventual mortality.^{4,10}

The sudden regression in the twins' conditions after initially being in good condition prompt inquiries into the underlying factors. Although it is challenging to determine a single cause, potential factors include cardiac decompensation, hemodynamic instability, and increased metabolic demands. The newborn transition involves significant physiological changes, and the presence of a cardiac anomaly may have only initially supported adequate cardiac function in utero and shortly after birth. However, as time progressed and the demands of the external environment stressed the compromised cardiac system, it might have exceeded its capacity to maintain sufficient perfusion and

oxygenation, leading to decompensation and regression in condition.⁴

In clinical management, social factors, religious beliefs, and psychological considerations are essential. Many couples opt for pregnancy termination because of the low survival rate and high risk associated with thoraco-omphalopagus conjoined twins. Even when choosing to continue the pregnancy, uncertainty about neonatal survival poses a dilemma for physicians. In certain countries, legally permitted abortion may be an option for conjoined twins with poor prognoses. In our case, it was vital to ensure the parents' full comprehension of maternal and fetal risks and prognosis, preparing them for their chosen path of continuing the pregnancy.^{5,6,11}

In conclusion, conjoined twins, a rare complication of monoamniotic pregnancies, pose significant medical risks and ethical challenges. We have presented a case involving the successful management and termination of thoraco-omphalopagus conjoined twins, although with an unfavorable neonatal outcome. Comprehensive care requires a multidisciplinary team approach that addresses prenatal, intrapartum, and postnatal aspects for both the twins and the mother.

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