A Clinical Case of Thoracopagus Conjoined Twins

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Abstract
Introduction: Conjoined twins result from the incomplete division of a single fertilized egg, with an incidence of approximately 1 in 200,000 live births. This rare congenital anomaly presents significant anatomical and physiological challenges, necessitating a multidisciplinary approach for diagnosis, management, and ethical considerations.
Case Presentation: A pair of thoracopagus conjoined twins was delivered at Arifin Achmad General Hospital following preterm labor complicated by premature rupture of membranes (PROM). Prenatal ultrasound identified shared thoracic and abdominal structures, including cardiac and vascular abnormalities. The twins exhibited severe congenital anomalies, such as cardiomegaly and ventriculomegaly. Despite immediate resuscitation efforts, both infants succumbed due to extensive anatomical complications.
Conclusion: The prognosis of thoracopagus twins remains poor, particularly in cases with extensive organ fusion. Advances in prenatal imaging and fetal surgery may improve outcomes in select cases; however, a multidisciplinary and compassionate approach remains essential for optimal perinatal care.
Keywords: Conjoined twins, Thoracopagus, Prenatal diagnosis, Ethical challenges, Perinatal management

Kasus Klinis Bayi Kembar Siam Tipe Thoracopagus

Abstrak
Pendahuluan: Kembar siam terjadi akibat pembelahan zigot yang tidak sempurna, dengan insiden 1 dari 200.000 kelahiran hidup. Anomali kongenital langka ini menimbulkan tantangan kompleks dalam aspek anatomi, fisiologi, serta etika sehingga memerlukan pendekatan multidisiplin dalam diagnosis dan penatalaksanaan.
Laporan Kasus: Sepasang kembar siam thoracopagus lahir prematur di RSUD Arifin Achmad akibat persalinan preterm yang disertai ketuban pecah dini (KPD). Pemeriksaan ultrasonografi prenatal mendeteksi fusi struktur torako-abdominal, termasuk keterlibatan organ jantung dan pembuluh darah utama. Bayi mengalami kelainan kongenital berat kardiomegali dan ventrikulomegali. Meskipun dilakukan resusitasi segera, keduanya tidak bertahan hidup akibat komplikasi anatomi yang luas.
Kesimpulan: Prognosis kembar siam thoracopagus tetap buruk, terutama pada kasus dengan keterlibatan organ yang luas. Kemajuan dalam pencitraan prenatal dan pembedahan janin dapat meningkatkan harapan hidup dalam kondisi tertentu; namun, pendekatan multidisiplin yang komprehensif dan penuh empati tetap menjadi kunci dalam perawatan perinatal.

Kata kunci : Diagnosis prenatal, Kembar siam, Manajemen perinatal, Tantangan etis, Thoracopagus

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Introduction

Conjoined twins, or Siamese twins, result from the incomplete division of a single fertilized egg during early embryonic development. This rare condition occurs in approximately 1 in 200,000 live births and presents a unique array of anatomical and physiological challenges. The extent of connection varies, ranging from shared placentas and amniotic sacs to the fusion of vital organs, which significantly impacts clinical management and outcomes.[1,2] Conjoined twins are categorized based on their point of union, including thoracopagus (joined at the chest), omphalopagus (joined at the abdomen), and craniopagus (fused at the skull). Each classification presents distinct surgical, ethical, and psychological considerations. Prognosis depends on the extent of shared anatomy, potential for surgical separation, and the overall health of each twin.[3,4] Management involves a multidisciplinary approach, including obstetricians, pediatricians, and surgeons, with early ultrasound diagnosis aiding in planning for delivery and possible surgical intervention. Ethical dilemmas frequently arise regarding separation, as decisions must weigh the risks and benefits for both twins. Recent advancements in surgical techniques have improved outcomes in some cases, highlighting the complexities of caring for conjoined twins and the ongoing challenges faced by families and healthcare providers. Complications associated with conjoined twins vary depending on the extent of organ sharing and anatomical fusion. Common complications include cardiovascular abnormalities, respiratory difficulties, gastrointestinal malformations, and increased risk of intrauterine fetal demise. In thoracopagus twins, congenital heart defects are the most significant prognostic factor, often limiting the feasibility of surgical separation. The risk of preterm labor and perinatal complications is also significantly higher, requiring specialized obstetric care.[5,6]

Case Presentation

A 33-year-old woman, G3P2A0, presented to Arifin Achmad Hospital Pekanbaru at 27 weeks of pregnancy, complaining of increasingly frequent and intense uterine contractions. She was a referral patient from an outside hospital, where an ultrasound examination had suggested that the twin fetuses were suspected to be conjoined. Upon vaginal examination, the cervix was 4 cm dilated, 50% effaced, and anteriorly positioned, with the fetal head palpable at Hodge 2. The patient reported clear amniotic fluid leakage for the past five hours, indicating premature rupture of membranes (PROM). There were no signs of infection, such as maternal fever or foul-smelling discharge.

A fetomaternal ultrasound revealed twin fetuses with several abnormalities. The first fetus had a cardiothoracic ratio (CTR) of 76%, with an impression of cardiomegaly and a visible thoracic wall defect measuring 3.70 cm. An abdominal wall defect of 5.36 cm was also observed, with the stomach and liver protruding through the defect. The estimated fetal weight was 857 grams. The second fetus showed ventriculomegaly, with lateral ventricles measuring 2.01 cm, along with a thoracic wall defect of 3.70 cm. Similarly, an abdominal wall defect of 5.36 cm was observed, with the stomach and liver protruding, along with the presence of ascites. The estimated fetal weight was 721 grams. Given the condition, an emergency cesarean section was performed. The twins were delivered with their anterior chests fused, with a total combined weight of 1400 grams. The APGAR scores were 2/3 for the first twin and 2/4 for the second twin. Despite resuscitation efforts, both babies did not survive and were declared deceased after 30 minutes. The mother was monitored postoperatively

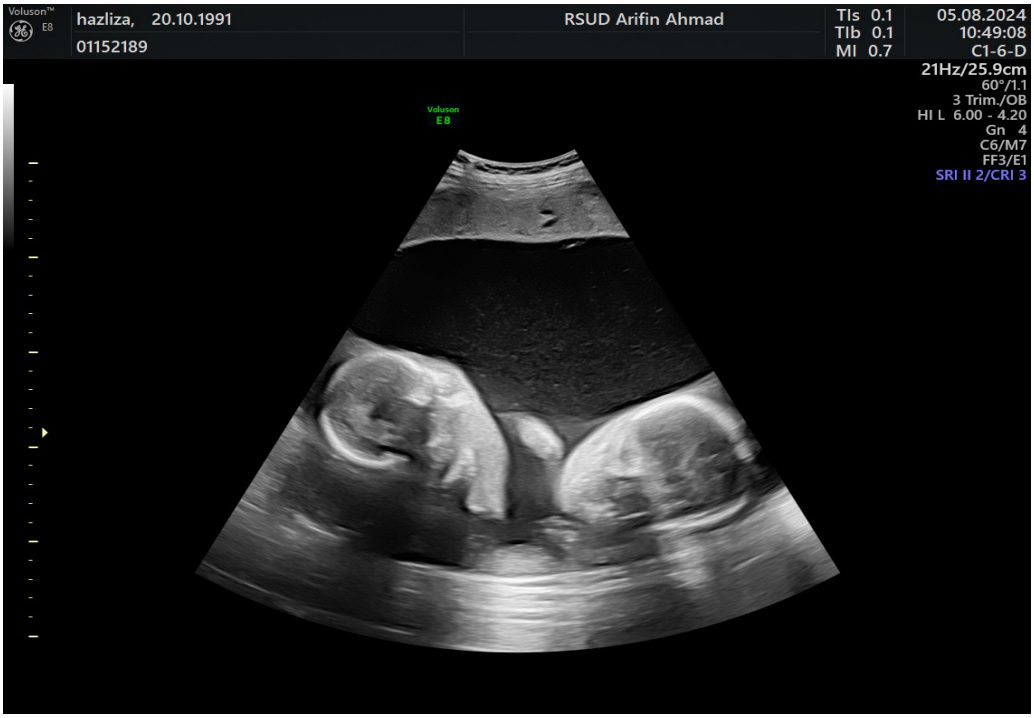

Figure 1 Ultrasound examination of the fetal head shows two separate ones

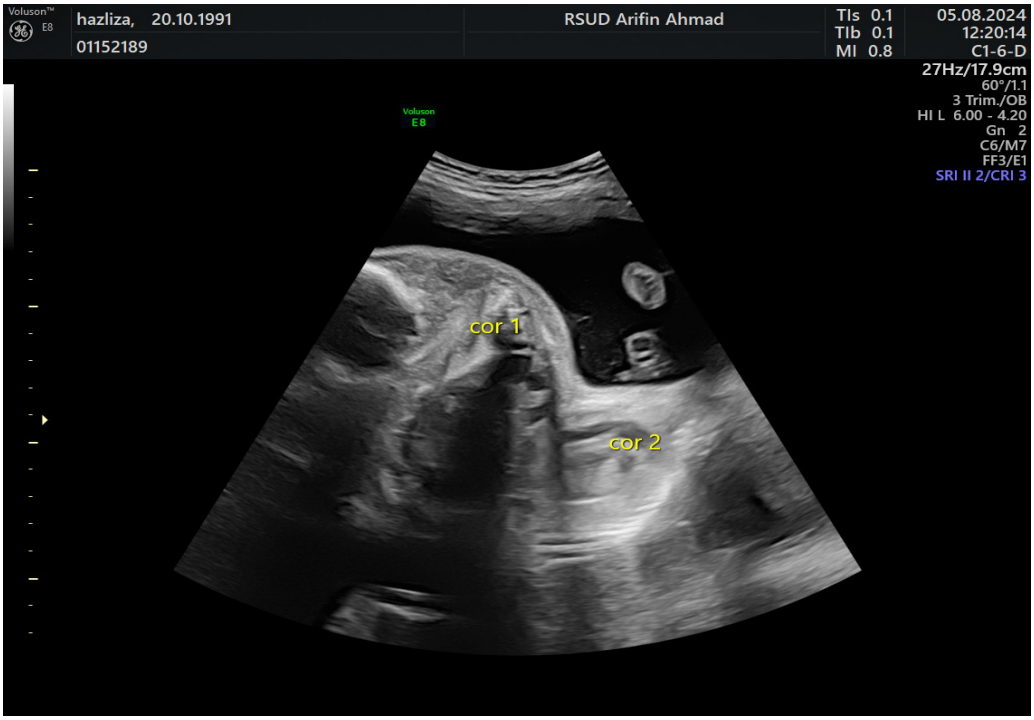

Figure 2 Ultrasound examination of the thorax shows that the fetus appears to be united and the two hearts are close together

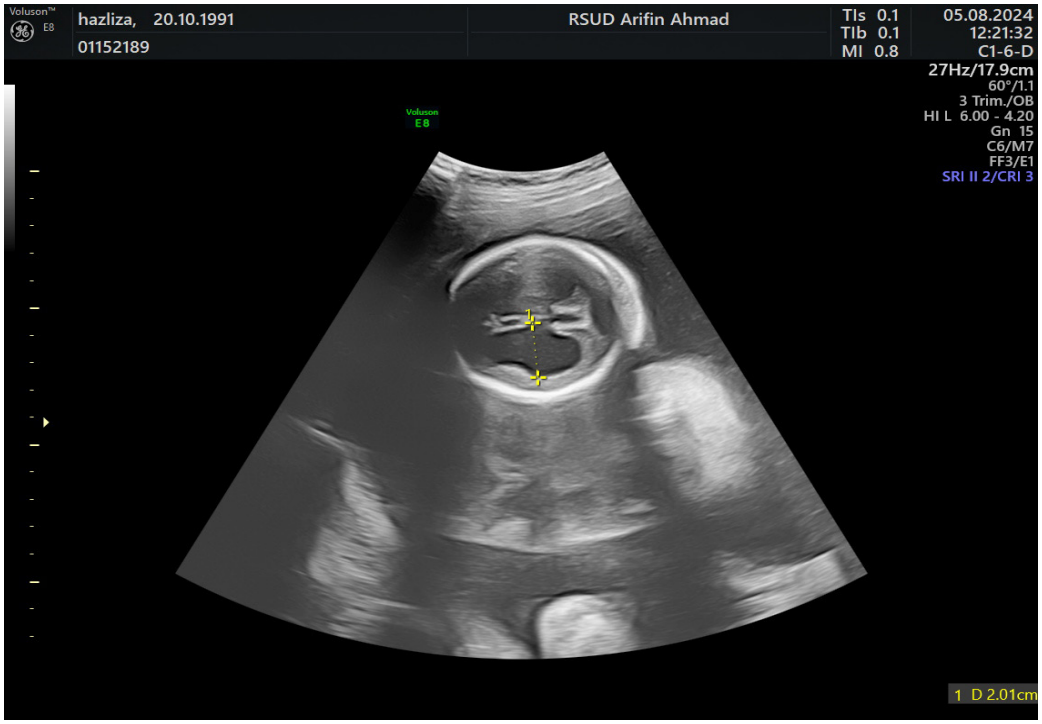

Figure 3 The first fetal ultrasound showed ventriculomegaly

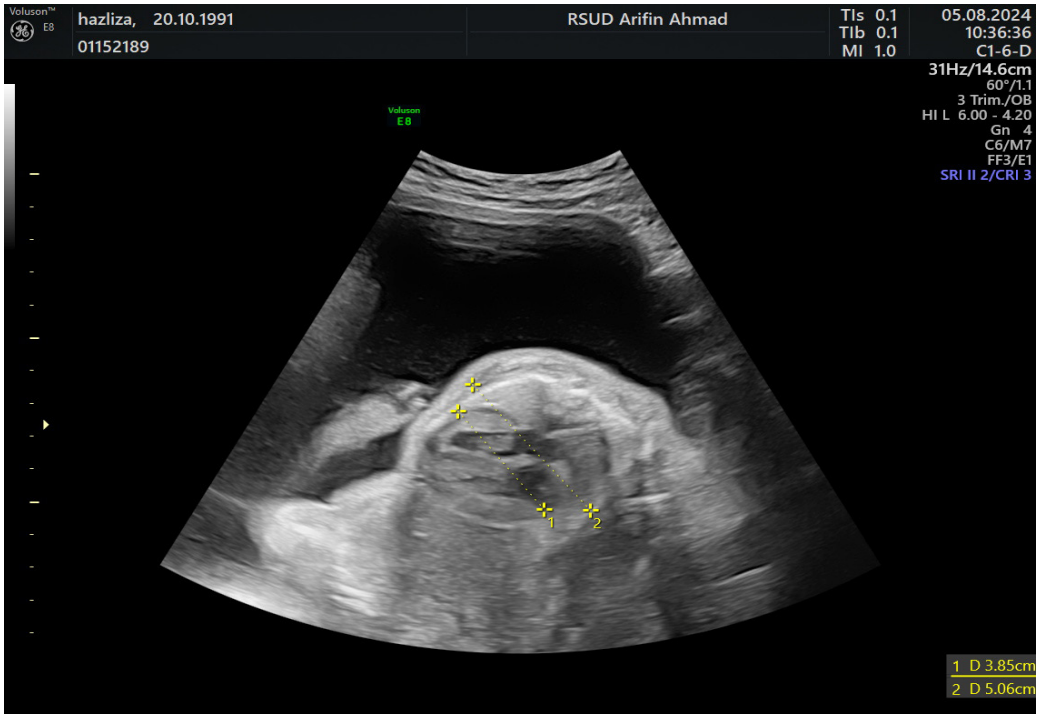

Figure 4 The second fetal ultrasound showed cardiomegaly with CTR >50%

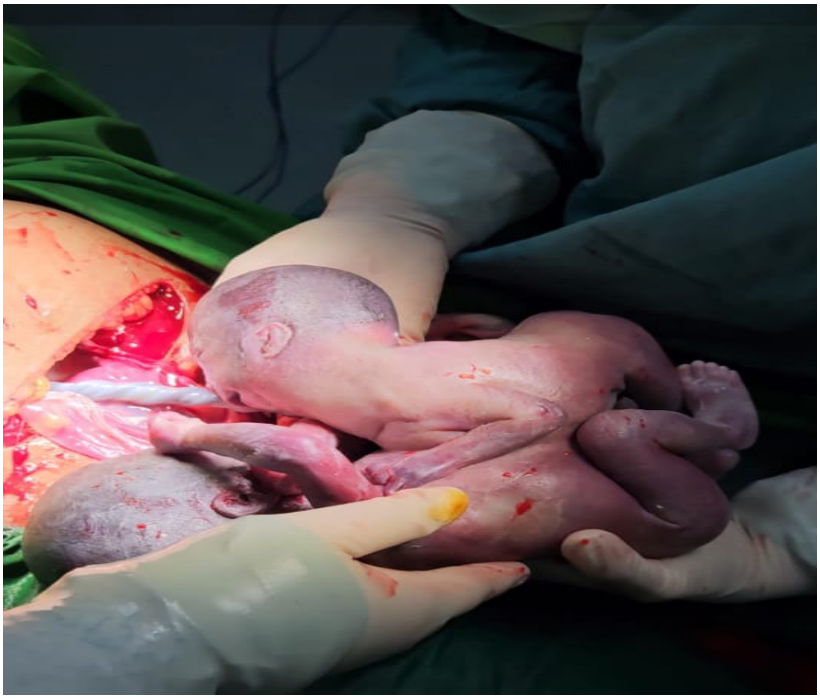

Figure 5 During operation the fetus shortly after birth

Figure 6 The fetus after failed resuscitation showed fused chest and abdomen

and was discharged in good condition on the third day of hospitalization. Tocolytic therapy with intravenous isoxsuprine (10 mg at 20 dpm) was administered to delay labor progression but was unsuccessful. A single dose of dexamethasone (10 mg) was given for fetal lung maturation, and intravenous cefotaxime (2x1 gram) was administered as prophylactic antibiotics.

Investigation

Fetomaternal ultrasound confirmed the presence of conjoined twins with multiple congenital anomalies, including thoracic and abdominal wall defects, cardiomegaly, ventriculomegaly, and ascites. The ultrasound findings suggested thoraco-omphalopagus conjoined twins, a condition characterized by fusion at the chest and abdominal wall. The diagnosis was confirmed intraoperatively during the cesarean section. Laboratory investigations included hemoglobin (Hb), leukocyte count, and platelet count, which were within normal ranges (Hb: 11.2 g/dL, leukocytes: 8,400/mm³, platelets: 264,000/mm³). Blood glucose levels were also normal (94 mg/dL). MRI was not performed due to logistical constraints and long waiting times.

Differential Diagnosis

Differential Diagnosis of Thoracophagus Conjoined Twins:

1. **Thoraco-omphalopagus Conjoined Twins**
   . The most likely diagnosis, characterized by fusion at the thorax and abdomen, often involving shared cardiac and hepatic structures. [7]
2. **Omphalocele with Twin Pregnancy**
   A differential diagnosis where both fetuses have large abdominal wall defects, though this would not typically result in fusion.[7]
3. **Pentalogy of Cantrell**
   . A rare condition involving midline defects, including a sternal cleft, diaphragmatic hernia, and abdominal wall defects, though it would not account for fetal fusion.[8]

Treatment

The patient was initially managed with tocolysis using isoxsuprine to delay preterm labor; however, due to progressive cervical dilation and active labor, an emergency cesarean section was performed. The neonates were immediately assessed, and resuscitation was attempted but was unsuccessful.

Outcome and Follow-Up

The twins were delivered via cesarean section, and despite resuscitation efforts, both neonates did not survive. The estimated blood loss during surgery was 500 mL. The mother was closely monitored postoperatively and was discharged in stable condition on the third day. Future pregnancy planning included a recommended interpregnancy interval of at least 18 months, along with preconception counseling, folic acid supplementation, and routine obstetric follow-ups.

Discussion

Thoraco-omphalopagus conjoined twins represent one of the most complex congenital anomalies, with a prevalence of approximately 1 in 200,000 live births. The prognosis largely depends on the extent of shared organs, particularly the heart and liver. In this case, prenatal ultrasound identified cardiomegaly, thoracic wall defects, and abdominal wall defects in both fetuses, strongly suggesting the fusion of vital thoracoabdominal structures, which significantly reduced the chances of survival.[9,10] The primary challenge

in this case was the delayed diagnosis, as the referring obstetrician was uncertain about the nature of the anomaly. MRI, which could have provided better anatomical details, was not performed due to scheduling constraints. Despite these limitations, prenatal ultrasound was instrumental in identifying the conjoined anatomy, aiding in early counseling and delivery planning.[11,12] Preterm labor remains a significant concern in conjoined twin pregnancies, often due to polyhydramnios, uterine overdistension, or premature rupture of membranes (PROM), as observed in this patient. PROM was a key contributing factor to preterm birth in this case, along with a history of leukorrhea suggesting an underlying infection risk. Although tocolytic therapy with isoxsuprine was initiated, it was unsuccessful in halting labor progression.[13,14] Ethical considerations in conjoined twin cases are profound, particularly regarding resuscitation and surgical separation decisions. Prenatal counseling helped the family understand the prognosis, and informed consent was obtained regarding neonatal care options. Despite resuscitation efforts, both twins did not survive due to severe congenital anomalies.[15] Psychosocial support is essential in such cases. This patient did not exhibit significant distress post-delivery, likely due to thorough prenatal counseling. Future pregnancy planning should include an 18-month interpregnancy interval, folic acid supplementation, and regular antenatal monitoring to optimize maternal and fetal outcomes. [16,17]

Conclusion

The management of thoracopagus twins presents significant medical, ethical, and psychosocial challenges that require a multidisciplinary approach. Surgical separation, when feasible, demands careful planning and consideration of shared anatomy, particularly regarding vital structures like the heart. Ethical dilemmas surrounding separation decisions necessitate comprehensive genetic counseling and psychological support for families. Long-term follow-up care is essential, as both twins may face ongoing health issues related to their shared physiology. Ultimately, prioritizing the well-being of both twins and their family through coordinated care can enhance outcomes and improve quality of life for these unique individuals.

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