

Case Report

Dicephalic Parapagus Conjoined Twins in a 37 Week Dichorionic Twin Pregnancy: A Case Report

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Received: October 21, 2025 | Accepted: July 6, 2026 | Published: July 10, 2026

Abstract

Introduction: Conjoined twins are a rare congenital anomaly resulting from incomplete embryonic division between days 13 and 15 post-fertilization. Dicephalic parapagus is exceptionally rare, characterized by two heads on a single trunk with variable degrees of organ sharing, and is associated with high perinatal morbidity and mortality.

Case presentation: A 20-year-old gravida 3 para 2 woman at 37 weeks was referred with suspected conjoined twins. Ultrasound revealed key clinical findings consistent with dicephalic parapagus, including two heads, a single thorax containing one heart, and shared abdominal organs (one liver, one kidney, one urinary bladder, and two gallbladders). After multidisciplinary planning, an elective cesarean section was performed, delivering live conjoined twins with ambiguous genitalia. The neonates were admitted to the NICU for stabilization and further imaging. The mother had an uncomplicated postoperative recovery and remained hemodynamically stable. The primary diagnosis was dicephalic parapagus with shared vital thoracoabdominal structures. The presence of a single functional heart, identified prenatally and confirmed postnatally, is the most decisive prognostic factor and significantly limits the possibility of surgical separation. This case highlights the importance of early prenatal detection and the role of coordinated multidisciplinary management.

Conclusions: As a rare and complex case, this report underscores the challenges of late diagnosis of severe congenital anomalies in resource-limited settings. It emphasizes the need for improved antenatal screening, comprehensive parental counseling, and ethical considerations when managing conjoined twins with non-separable anatomy.

Keywords: Prenatal diagnosis; conjoined twins; neonatal outcome; cesarean section; dicephalus parapagus; multidisciplinary management

Kembar Siam Parapagus Dicephalus pada Kehamilan Kembar Dikorionik 37 Minggu: Laporan Kasus

Abstrak

Pendahuluan: Kembar siam merupakan anomali kongenital yang sangat jarang terjadi akibat kegagalan pembelahan diskus embrionik antara hari ke-13 hingga ke-15 pascafertilisasi. Parapagus disefalus merupakan kasus yang sangat langka, ditandai dengan dua kepala pada satu batang tubuh dengan derajat berbagi organ yang bervariasi, serta memiliki morbiditas dan mortalitas perinatal yang tinggi.

Presentasi kasus: Seorang wanita berusia 20 tahun dengan gravida 3 para 2 pada usia kehamilan 37 minggu dirujuk dengan dugaan kehamilan kembar siam. Pemeriksaan ultrasonografi menunjukkan temuan klinis penting berupa parapagus disefalus, dengan dua kepala, satu toraks berisi satu jantung, serta organ abdomen yang berbagi, termasuk satu hati, dua kantong empedu, satu ginjal, dan satu kandung kemih. Setelah perencanaan multidisiplin, dilakukan seksio sesarea elektif dan berhasil melahirkan bayi kembar siam hidup dengan genitalia ambigu. Keduanya dirawat di NICU untuk stabilisasi dan pemeriksaan lanjutan, sementara ibu menjalani pemulihan pascaoperasi tanpa komplikasi. Diagnosis utama adalah parapagus disefalus dengan berbagai organ vital torakoabdominal yang digunakan bersama. Keberadaan satu jantung fungsional merupakan faktor prognostik paling kritis yang sangat membatasi kemungkinan dilakukan pemisahan bedah. Kasus ini menekankan pentingnya deteksi prenatal dini, konseling orang tua yang komprehensif, dan tata laksana perinatal yang terkoordinasi.

Kesimpulan: Sebagai kasus yang sangat langka dan kompleks, laporan ini menunjukkan tantangan diagnosis terlambat pada anomali kongenital berat, terutama di fasilitas kesehatan dengan sumber daya terbatas. Kasus ini menegaskan perlunya peningkatan kualitas skrining antenatal, edukasi keluarga, serta pertimbangan etis dalam menangani kembar siam dengan anatomi yang tidak dapat dipisahkan.

Kata kunci: Diagnosis prenatal; kembar siam; luaran neonatal; operasi caesar; parapagus dicephalus; tatalaksana multidisiplin

Introduction

Conjoined twins are among the rarest forms of twin pregnancy. They occur in monozygotic twins that fail to undergo complete embryonic division between the 13th and 15th day post-fertilization, resulting in varying degrees of fusion between the fetuses. The reported incidence ranges from 1 in 49,000 to 1 in 200,000 live births, with higher prevalence in Southeast Asia, Africa, and Brazil.^{1,2} Approximately 50% of cases are stillborn, and an additional one-third die within the first 24 hours of life. Among those who survive, the majority are female, with a female-to-male ratio of about 3:1. Classification of conjoined twins is based on the anatomical site of union, including thoracopagus, omphalopagus, craniopagus, ischiopagus, and parapagus, with thoracopagus being the most frequently reported type.³

Parapagus conjoined twins are characterized by lateral fusion of the body and account for less than 5% of all conjoined twin cases. A rare variant, dicephalic parapagus, presents with two heads on a single trunk and is often associated with severe malformations, including shared thoracic and abdominal organs. Prognosis is generally poor, depending on the extent of organ sharing, especially of cardiac structures, which greatly influence the possibility of surgical separation and survival.^{4,5}

Prenatal diagnosis is critical to the management of conjoined twin pregnancies. Advances in ultrasonography and fetal MRI enable detection as early as the first trimester, supporting multidisciplinary planning regarding continuation of pregnancy, timing, and mode of delivery, as well as neonatal care. Despite these advances, conjoined twins continue to pose complex medical, ethical, and psychosocial challenges for families and healthcare teams.⁶

Herein, we report a case of a 20-year-old gravida 3 para 2 woman at 37 weeks

of gestation with intrauterine live twin fetuses in cephalic presentation, suspected dicephalic parapagus conjoined twins, who underwent elective cesarean section. This case is uniquely significant because dicephalic parapagus twins with a single functional heart are exceedingly rare and almost uniformly fatal, making detailed perinatal documentation especially valuable. Recent literature indicates that fewer than a handful of such cases are reported each decade globally, and most are diagnosed late or identified only at delivery, particularly in low-resource settings.^{3,7} The combination of late-term diagnosis, shared vital organs, and delivery of live twins at term underscores the clinical and academic importance of reporting this rare presentation.

Case Presentation

A 20-year-old (G3P2A0L2) at 37 weeks of gestation with no history of spontaneous abortion or congenital anomalies was referred from Pasir Pangaraian General Hospital to our tertiary care center for further management of a suspected conjoined twin pregnancy. She presented without abdominal pain, vaginal bleeding, or leakage of amniotic fluid, and reported good fetal movements. Antenatal care had been received four times at the obstetric clinic, but no visits to a midwife were reported. There was no history of assisted reproductive technology, trauma, or chronic medical illnesses such as hypertension or diabetes mellitus. Family history of twinning and congenital malformations was negative. Her two previous pregnancies had ended in term vaginal deliveries of healthy male infants weighing 3200 g and 3600 g, respectively. Clinically, she was asymptomatic upon admission, with stable vital signs and no pain, bleeding, contractions, or signs of obstetric complications. Her past medical and surgical history was unremarkable, with no previous abdominal or pelvic surgeries,

no history of cesarean delivery, and no prior invasive or non-invasive diagnostic procedures related to congenital anomalies. Routine antenatal screening had been limited to basic ultrasound examinations without detailed anomaly scans, and no fetal MRI or echocardiography had been performed. Prior obstetric outcomes were uncomplicated, making the current pregnancy notable for the unexpected detection of a rare congenital malformation in the absence of risk factors.

On admission, her general condition was stable, with blood pressure of 129/78 mmHg, pulse rate of 90 bpm, respiratory rate of 20 breaths/min, and temperature of 36.7°C. Abdominal examination revealed a symmetrically distended uterus with a fundal height at the xiphoid level. Leopold's maneuvers detected two fetal heads, consistent with a cephalic–cephalic presentation. Fetal heart rate was 130 bpm, and no uterine contractions were noted.

A fetomaternal ultrasound at 36 weeks confirmed two intrauterine live twin fetuses with cephalic–cephalic presentation. Two separate heads were visualized with

identifiable craniofacial structures. A single thorax contained one heart, and the abdominal cavity showed one liver, two gallbladders, a single urinary bladder, and one kidney. Two upper and two lower limbs were noted. Estimated fetal weight could not be calculated because of the fused anatomy. Biometric measurements were consistent with 33–36 weeks of gestation (Figure 1). Cardiotocography showed a reactive tracing with a baseline of 140 bpm, moderate variability, and no decelerations (Figure 2). Routine laboratory investigations were unremarkable. Based on these findings, a diagnosis of dicephalic parapagus conjoined twins was made.

After multidisciplinary consultation and counseling, the patient underwent an elective cesarean section at 37 weeks of gestation. A live conjoined twin was delivered with ambiguous external genitalia. Baby 1 weighed 4150 g, measured 42 cm in length, with a head circumference of 34 cm, and had Apgar scores of 7 and 9 at 1 and 5 minutes, respectively. Baby 2 had a head circumference of 36 cm; however, other anthropometric

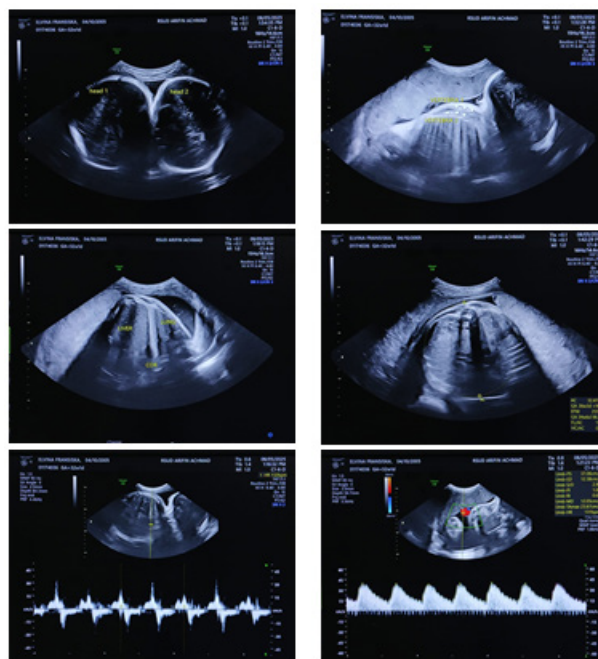


Figure 1 Fetomaternal Ultrasonography



Figure 2 Cardiotocography



Figure 3 Clinical and Intraoperative Findings of The Conjoined Twins

parameters were not obtained. The placenta was intact, weighing 650 g with a diameter of 25 cm, and the umbilical cord measured 25 cm in length (Figure 3).

The postoperative diagnosis was dicephalic parapagus conjoined twins in cephalic–cephalic presentation, delivered via cesarean section. The mother tolerated the procedure well and remained hemodynamically stable. She received intravenous fluid therapy with Ringer's Lactate 500 mL and oxytocin 10 IU at 20 drops/min for 24 hours. Prophylactic antibiotics were administered with intravenous ceftriaxone 1 g twice daily, along with analgesia using intravenous ketorolac 30 mg three times

daily. To minimize postoperative bleeding, tranexamic acid 500 mg intravenously three times daily was also administered. A urinary catheter was maintained for 24 hours, and a complete blood count was performed 6 hours postoperatively. Gradual mobilization was encouraged as tolerated.

The conjoined twins were admitted to the neonatal intensive care unit (NICU) for stabilization and multidisciplinary evaluation. Planned steps included advanced imaging (echocardiography, CT/MRI) to assess the extent of shared thoracoabdominal organs, followed by counseling of the parents on prognosis, potential surgical options, and ethical considerations.

Discussion

Dicephalic parapagus is an exceptionally rare form of lateral (nondorsal) conjoined twinning, characterized by two heads and necks on a single trunk, with variable sharing of thoracic and abdominal organs. The critical determinant of prognosis is the extent and complexity of shared vital structures, especially the cardiac, pulmonary, and vascular systems.^{7,8} In our case, ultrasound and intraoperative findings confirmed two heads, one thorax with a single heart, and a shared abdominal cavity containing one liver, two gallbladders, one kidney, and one urinary bladder, findings that align with classical representations of dicephalic parapagus.

Prenatal diagnosis is essential for optimal management, parental counseling, and delivery planning. Ultrasonography remains the first-line imaging modality; signs such as inseparable bodies, fixed relative positions, and the absence of a separating membrane raise suspicion. When sonographic visualization is limited, fetal MRI provides enhanced anatomical delineation and mapping of fusion planes. In low-resource settings, many conjoined twin cases go undetected until later in pregnancy or even at birth.^{9,10}

A 2025 case report by Arkorful et al. described the diagnosis of dicephalic parapagus in the third trimester via ultrasound, after earlier scans showed no anomaly, paralleling our experience of delayed diagnosis. Another recent report in the *Pan African Medical Journal* (2025) described a dicephalic parapagus dipus tribrachius twin pair diagnosed at 36 weeks, underscoring that late presentation remains common in many settings.¹¹

The overall prognosis for conjoined twins remains grim: approximately 50% are stillborn, and an additional one-third die within 24 hours of birth. Survival among dicephalus variants is even rarer. In one

recent non-peer-reviewed news report, a dicephalic parapagus infant survived for 16 days before succumbing, highlighting the extreme fragility even when early survival is achieved.^{1,12} In our case, although both babies had relatively good Apgar scores, the presence of a single thorax and heart suggests a guarded long-term prognosis.^{4,13}

Early prenatal diagnosis is crucial for optimal pregnancy counseling and perinatal management. Advances in high-resolution ultrasonography have enabled earlier detection of conjoined twins, often in the late first or early second trimester.¹⁴ Studies from the last decade emphasize the importance of targeted anomaly scanning at 11–14 weeks, as recommended, for early identification of severe congenital anomalies.¹⁵ However, late diagnosis remains common in low-resource settings because of limited ultrasonography expertise and delayed antenatal visits. Several case reports from 2018 to 2024 describe similar scenarios in which dicephalic parapagus twins were detected only in the third trimester, delaying multidisciplinary planning and limiting available clinical options.⁷

In recent years, imaging modalities such as fetal MRI, 3D reconstruction, and detailed fetal echocardiography have significantly improved the ability to map organ fusion and assess surgical feasibility. Modern literature underscores that cardiac anatomy is the most decisive prognostic factor: twins sharing a single heart or a common major cardiac outflow tract have a near-zero likelihood of successful separation, and survival beyond the neonatal period is exceedingly rare. The anatomy identified in our case, shared heart, shared abdominal organs, and a single thoracic cavity, is fully consistent with these high-risk features.¹⁶

Moreover, this case highlights the psychosocial and ethical complexities of managing pregnancies involving lethal fetal anomalies. Over the past decade, research

has consistently reported that parents experience significant emotional distress, anticipatory grief, and decisional conflict after the diagnosis of severe congenital anomalies such as conjoined twins. Best-practice guidelines now emphasize a family-centered, multidisciplinary approach that includes maternal–fetal medicine specialists, neonatologists, pediatric surgeons, psychologists, and ethics teams to support shared, informed decision-making. In our case, coordinated counseling helped the family understand the anatomic limitations and prognosis, as well as the expected course of neonatal care.¹⁷

Finally, this case underscores the need to strengthen antenatal screening systems in resource-limited regions. Updated ISUOG standards recommend systematic anomaly screening during early gestation as a global strategy to improve detection of severe structural abnormalities and enable timely referral to tertiary care centers. Improving sonographer training, ensuring access to high-quality imaging, and promoting early antenatal visits could reduce diagnostic delays and improve care pathways for similar high-risk pregnancies in the future.¹⁸

Conflict of Interest

The authors declare that they have no competing interests. The funding comes from the authors.

Acknowledgements

Not applicable

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