

CASE REPORT**Dicephalus Parapagus Conjoined Twins**

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Abstract

Background: Conjoined twins are monoamniotic monochorionic twins resulting from incomplete division of the embryonic disk and amniotic sac. This rare condition has an incidence of 1:33,000 to 1:165,000 pregnancies. Common types include thoracopagus and omphalopagus, with less common forms like thoracoomphalopagus, pyopagus, ischiopagus, and craniopagus. Parapagus, where twins are joined at the sides with a shared pelvis and organs, occurs in less than 0.5% of cases.

Case: A 27-year-old patient presented to the Fetomaternal Clinic at RSUP M. Djamil Padang, referred from Hermina Hospital, with a diagnosis of G2P1A0H1 gravid preterm and suspected conjoined twins. Ultrasound at 5 months showed two heads and one body. Initial assessment noted a family history of twins and palpable round, firm masses. Laboratory tests, Fetomaternal ultrasound, MRI, and 3D CT Scan confirmed conjoined twins (dicephalus, parapagus, dibrachius). A cesarean section was planned.

Discussion: Termination involves a multidisciplinary team to manage fetal anatomical abnormalities optimally. Emergency separation has a 70% mortality rate, higher than the 20% for elective procedures. Treatment depends on cardiovascular anatomy, with higher success if only the pericardium is divided. Cardiac anomalies' severity influences prognosis, survival, and separation feasibility.

Conclusion: Survival rates depend on the degree of union and cardiac anomalies. In cases like dicephalus, the anatomical structure often makes it unlikely for both twins to survive separation.

Keywords: Dicephalus, Parapagus, Conjoined Twins

INTRODUCTION

Conjoined twins are one of the rare and fascinating human congenital anomalies. The incidence is very rare, reported between 1:33,000 and 1:165,000 pregnancies.¹ Conjoined twins are monoamniotic monochorionic twins where the twinning starts after the embryonic disk and amniotic sac have incompletely formed and the division of the embryonic disk is incomplete.² Common types of conjoined twins include thoracopagus and omphalopagus. Less common forms of conjoined twins include thoraco-omphalopagus (joined at the chest and abdomen), pyopagus (joined at the buttocks), ischiopagus (joined at the ischium), and craniopagus (joined at the head).² Parapagus is a rare form of conjoined twins where the twins are joined at the sides of their bodies with a shared pelvis and organs. The incidence of parapagus is less than 0.5% of all conjoined twin cases.²

These twins are further divided into symmetric or asymmetric types, depending on whether there are 2 well-developed babies or a small part of the body is duplicated. The widely accepted mechanism for the development of conjoined twins is the incomplete separation of monozygotic twins after 12 days of embryogenesis.³ Due to the high morbidity and mortality associated with conjoined twins, prenatal diagnosis is crucial. All monozygotic twins should be screened for possible conjoinment. If conjoinment is present, associated congenital anomalies such as complex congenital heart disease, lower GI anomalies like imperforate anus, genitourinary anomalies, and CNS anomalies should be ruled out.⁴

Early diagnosis is the most important factor for pregnancy management. Prenatal diagnosis is usually done with ultrasonography, which is safe, accurate, reliable, and fast. Alternatively, obstetric MRI can provide better tissue contrast and further detail related congenital anomalies.⁵

With transvaginal ultrasound, conjoined twins can now be diagnosed at 8 weeks of gestation. Proposed diagnostic criteria include: absence of a separating membrane, joined body parts, inseparable body or head despite fetal position changes, bifid fetal pole appearance in the first trimester, more than three vessels in the umbilical cord, complex structural anomalies, heads or bodies at the same level, hyperextended spine, unusual proximity of extremities, and fixed relative positions after movement or on follow-up scans. Recent reports also present data on the contribution of 3D ultrasonography for prenatal diagnosis of conjoined twins.⁶

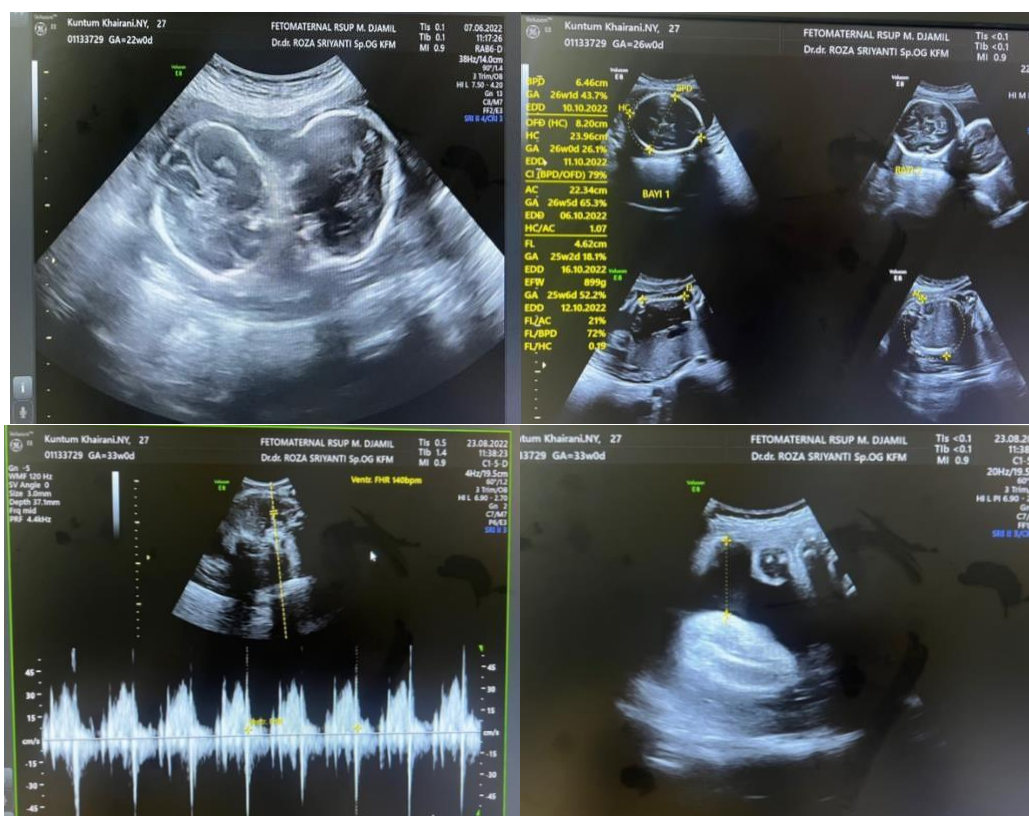
A rare case report of conjoined twins (Dicephalus Parapagus Conjoined Twins) diagnosed by ultrasound from a network hospital and then referred to RSUP M. Djamil Padang.

CASE REPORT

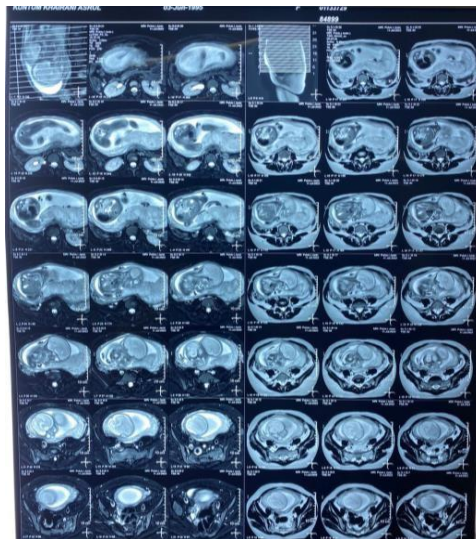
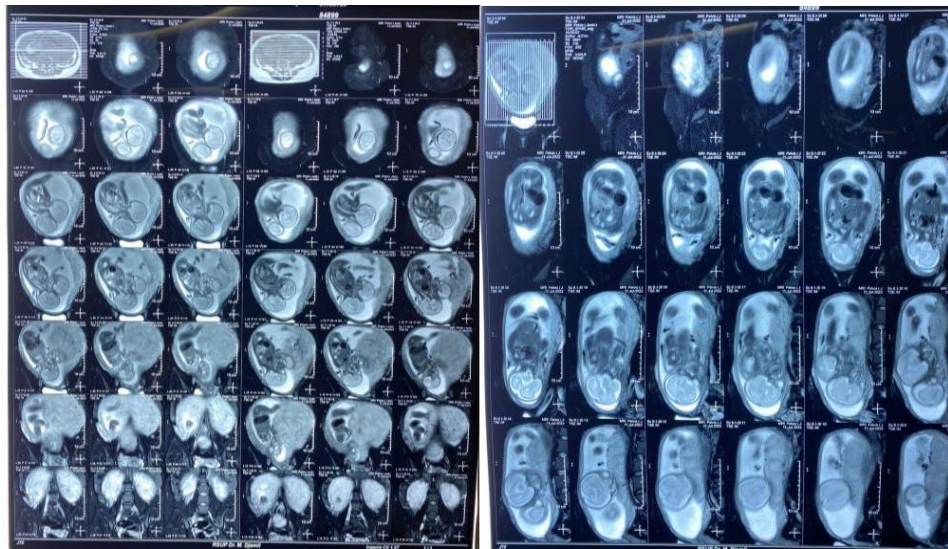
A 27-year-old woman presented to the Fetomaternal clinic with a diagnosis of G2P1A0H1 gravid preterm at 33-34 weeks, suspected of carrying dicephalus parapagus conjoined twins. The patient had been referred from Hermina Hospital, where the diagnosis of conjoined twins was suspected due to the presence of two heads and one body on ultrasound at five months gestation.

Upon examination, an ultrasound revealed a 33-34 week intrauterine pregnancy with monochorionic monoamniotic twins. The imaging showed two heads without abnormalities, a single shared thoracic cavity with one heart, a conjoined abdomen with one stomach and liver, and paired kidneys, arms, and legs, confirming the diagnosis of dicephalus parapagus dibrachius conjoined twins.

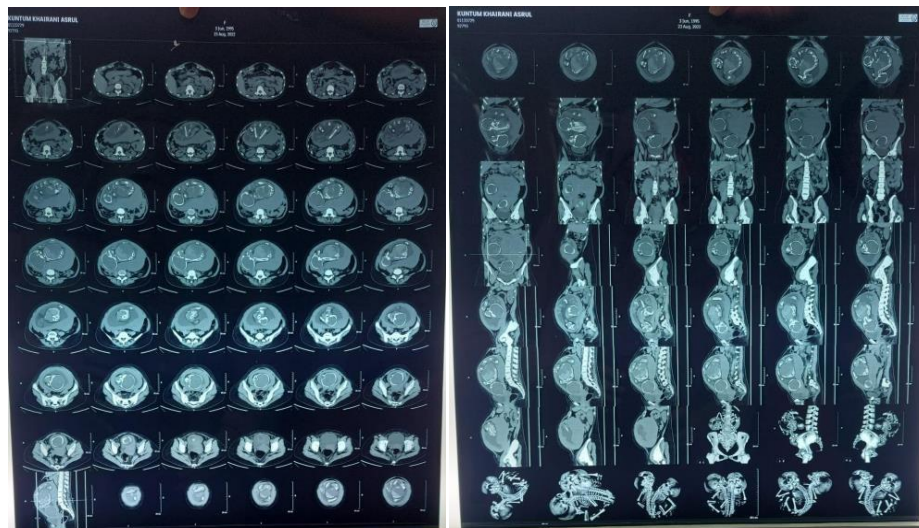
The patient was planned for a cesarean section, with comprehensive and functional management preparations for the anatomical abnormalities. The cesarean section plan involved a multidisciplinary team from the departments of Obstetrics and Gynecology, Pediatric Surgery, Thoracic Surgery, Neurosurgery, Digestive Surgery, and Perinatology to ensure optimal care for the anatomical anomalies present.



Ultrasound: The ultrasound at 33-34 weeks gestation revealed monochorionic monoamniotic twins with two heads, a single heart in a fused thoracic cavity, a conjoined abdomen with one stomach and liver, paired kidneys, arms, and legs, confirming dicephalus parapagus dibrachius conjoined twins.



MRI confirmed the ultrasound findings, detailing two well-formed heads, a single thoracic cavity with one heart, and a conjoined abdominal cavity with one stomach and liver, showing clear visualization of shared and separate structures.





CT Scan: The CT scan with 3D reconstruction corroborated the ultrasound and MRI results, illustrating the shared thoracic and abdominal cavities, single heart, stomach, liver, and detailed skeletal morphology, reinforcing the diagnosis of dicephalus parapagus dibrachius conjoined twins.

Preoperative assessment was crucial for determining the best surgical approach, reconstruction methods, and prognosis for the infants. Discussions with other specialties highlighted the importance of careful planning, as emergency separations have a 70% mortality rate compared to 20% for elective procedures.

DISCUSSION

In the case found, a 27-year-old woman was admitted to the Fetomaternal clinic with a diagnosis of G2P1A0H1 gravid preterm 33-34 weeks + gemelli letkep letkep + Dicephalus Parapagus Conjoined Twins. The patient had previously visited the fetomaternal clinic at RSUP M Djamil, referred from Hermina, with a diagnosis of G2P1A0H1 gravid 33-34 weeks with suspected conjoined twins. The suspicion of a twin pregnancy arose from the presence of two heads and one body.

Upon examination, ultrasound revealed a 33-34 week pregnancy, with a live intrauterine monochorionic monoamniotic twin pregnancy showing two heads without abnormalities, a single heart with fused chests, a single stomach and liver without abnormalities, paired kidneys, and paired hands and feet, indicating live conjoined twins (dicephalus, parapagus, dibrachius).

The patient was planned for a cesarean section with comprehensive and functional preparation for the anatomical abnormalities. The planned cesarean section surgery was prepared by the Obstetrics & Gynecology department. The termination procedure in this case will involve an experienced multidisciplinary team. The Obstetrics & Gynecology department will seek assistance from colleagues in Pediatric Surgery, Thoracic Surgery, Neurosurgery, Digestive Surgery, and Perinatology to provide optimal management for the fetal anatomical abnormalities. Detailed preoperative assessment is crucial to determine the best surgical approach, reconstruction method, and prognosis for the baby. Discussion of this case with other fields is essential because emergency separation has resulted in a 70% mortality rate, much higher than the 20% for elective procedures.

Ethical, legal, and religious questions often arise with conjoined twins. For example, will the surgery be successful? Is sacrificing one twin to save the other justified? The moral and ethical aspects of separation must be considered, especially in the following circumstances: A choice must be made regarding a single organ system -The twin receiving the organ system will live and thrive, while the other twin will suffer or die,

The twins have a shared heart - Complex heart separation surgery is mostly unsuccessful; in some cases, one twin is left alive with the entire heart complex; the availability of two heart transplants at the same time could improve the options. Craniopagus twins with a complete brain connection - These twins usually cannot be separated.

Previous studies reported that some parapagus cases were stillborn while others died shortly after birth. The survival rate of dicephalus conjoined twins depends on the degree of union and the presence of cardiac anomalies. Some dicephalus parapagus twins survive without being separated. Treatment options mainly depend on cardiovascular anatomy, with a higher success rate for separation where only the pericardium is divided. The severity of cardiac anomalies determines the prognosis, survival, and feasibility of separation.¹⁸

Dicephalic conjoined twins may have a long lifespan. The primary predictors of survival are the degree of conjunction and cardiac anomalies. Most stillborn infants have cardiopulmonary malformations incompatible with extrauterine life. In dicephalus, and other extensive forms of conjoined twins, the anatomical structure is such that it is unlikely that both twins will survive an attempt at separation.¹⁹

CONCLUSION

The case of Dicephalus Parapagus conjoined twins presented in this report underscores the complexity and rarity of such congenital anomalies. Comprehensive prenatal imaging revealed two distinct heads, a single thoracic cavity, a shared heart, and fused abdominal organs, necessitating a multidisciplinary approach for optimal management. The prenatal identification was crucial for planning the mode of delivery and preparing for immediate postnatal care, with cesarean section chosen to minimize risks.

The prognosis for conjoined twins, especially those with shared vital organs, depends on the extent of organ sharing and the presence of additional anomalies. This case highlights the importance of early and accurate prenatal diagnosis, detailed anatomical assessments, and coordinated care. Advances in imaging and surgical techniques will continue to improve outcomes and quality of life for these rare and complex cases.

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