

CASE REPORT

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Antenatal diagnosis of thoracopagus and thoraco-omphalopagus conjoined twins: case series

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Abstract

Background Conjoined twin pregnancy is a rare variety, which occurs due to incomplete separation of the embryonic plate after 13 days. Each type of this entity is linked to distinct levels of organ sharing, structural abnormalities, complications, and difficulties concerning the potential for separation. So a comprehensive multidisciplinary prenatal assessment of conjoined twins is crucial for providing accurate parental counseling, effectively managing the pregnancy, and formulating an appropriate delivery strategy. The purpose of our study was to early diagnosis of conjoined twins using ultrasound, describing the various types of conjoined twins and the role of recent advances like 3D ultrasound in aiding their management.

Case presentation Here, we are describing the case series of four conjoined twins, which were diagnosed antenatally by using ultrasound between the period of January 2019 and December 2019. Four cases of conjoined twins (two thoracopagus and two thoraco-omphalopagus) were accurately diagnosed on antenatal ultrasound. After detailed counseling of the parents regarding the risks and benefits of this complicated pregnancy and obtaining written consent, all four cases were terminated. Ultrasound plays a vital role in the early diagnosis and management of conjoined twins. Also, 3D ultrasound and MRI help in better characterization of fused body parts and connection between vital organs, thus it aids in the management.

Conclusions It is important to detect these cases early as termination of pregnancy is less risky in the early stages as compared to the late stages for proper management.

Keywords Conjoined twins, Thoracopagus, Antenatal, Pregnancy

Background

Conjoined twins are monozygotic monochorionic twins, formed when embryonic plate separation is incomplete from days 13 to 17. The incidence of conjoined twins is

1:50,000 in pregnancy and 1:250,000 total live births, as 60% die in utero or are stillborn [1] and almost 35% of cases die within a day after birth [2]. Conjoined twins are classified according to their prominent site of connection. The incidences of conjoined twins are in the following order as per Spencer [3] and McHugh et al. [4] Thoracopagus (20–40%), omphalopagus (18–33%), parapagus (28%), pygopagus (18–28%), cephalopagus (11%), ischiopagus (6–11%) and craniopagus (2%). In this article, we demonstrate the appearances of conjoined twins with different sites of connection using prenatal 2D and 3D ultrasound (USG) findings in four cases. We presented postnatal autopsy results for three cases, except for one case of a 13-week pregnancy where an autopsy

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photograph could not be obtained due to a miscarriage occurring outside the hospital. Timely antenatal screening, including ultrasound and fetal MRI, plays an important role in the early detection of conjoined and in counseling parents about potential surgical or medical termination options and outcomes.

Study method

All four cases were diagnosed antenatally by using an ultrasound machine—ALOKA with curved array transducer (1–5 MHz). Detailed parents' histories were taken regarding exposure to medications, use of assisted reproductive techniques, and family history of twins. Ultrasound examination was done by two radiologists on those who were suspected of conjoined twins.

Ultrasound criteria for diagnosing conjoined twins: Conjoined twinning is suspected when there is a single placenta with a single umbilical cord having more than three vessels, hyperextension of the cervical spine, fused body parts with shared organs and obvious fusion site, constant relative positions of the fetuses with heads, and other body parts persistently at the same level on repeat scans. The relative position of both fetuses is fixed without associated intervening amniotic membranes and limbs may be fewer than expected in some cases [5, 6].

Examination of each placenta was done for any abnormality and number. Each fetus of the four twin cases was

individually evaluated from the head to the foot. The site of fusion was noted for the classification of twins. The number of vital organs such as the heart and color Doppler was applied for identifying vascular connections between the twins. The liver, GIT, limbs, and pelvic organs were evaluated for fusion abnormalities.

After detailed counseling and obtaining written consent from all the parents, both the thoracic-omphalopagus and thoracopagus twin pregnancies were terminated by medication. No complications were observed in the mother in the postoperative period in all four cases.

Case presentation

Case 1

The first case was a pair of thoracopagus twins (Fig. 1) with 14 weeks of gestational age diagnosed in a 23-year primigravida. USG revealed a single placenta, single umbilical cord with conjoined twins from the thorax region till the upper part of the abdomen facing toward each other with two separate heads, fused thorax with shared heart. They had two unfused spines and two separate pelvis. Each of the twins had two pairs of limbs. One of the twins had club foot deformity in both lower limbs with occipital encephalocele. One of the twins had fused both lower limbs (mermaid deformity). After proper counseling and obtaining written consent from parents, medical termination of pregnancy was done.

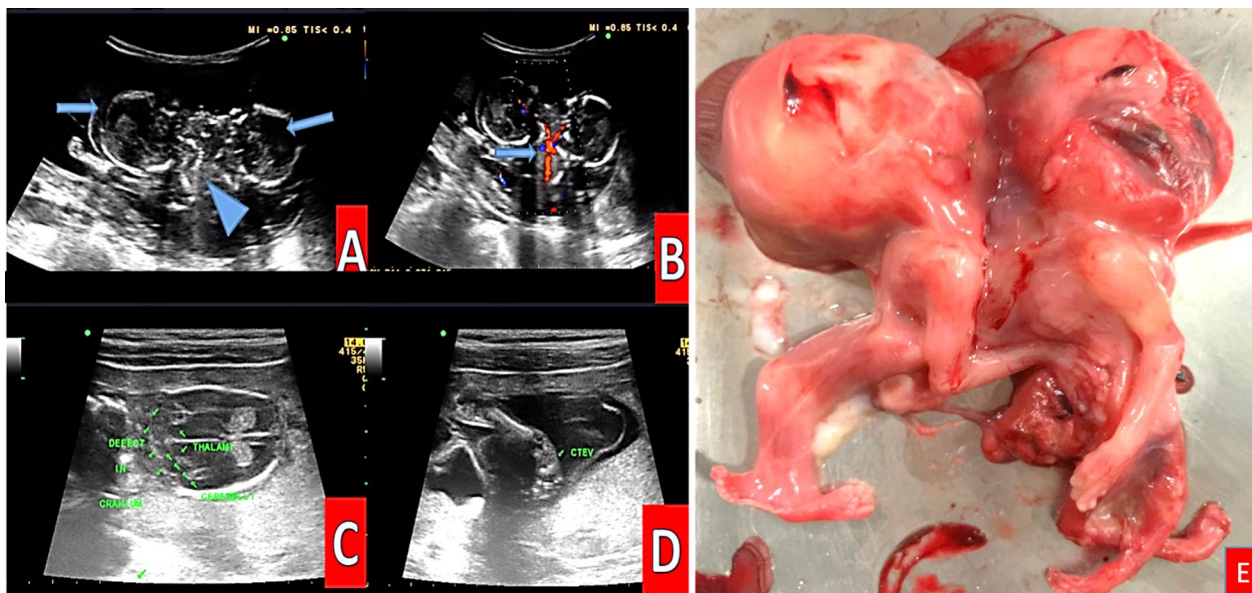


Fig. 1 Transabdominal ultrasound images of a thoracopagus conjoined twins shows **A** Two separate head (blue arrow) with fused thorax (triangle), **B** a common heart with color Doppler flow (arrow), **C** occipital encephalocele and **D** foot abnormality CTEV. **E** Autopsy specimen. Mermaid deformity or fused lower limbs are seen in one fetus (left-hand side)

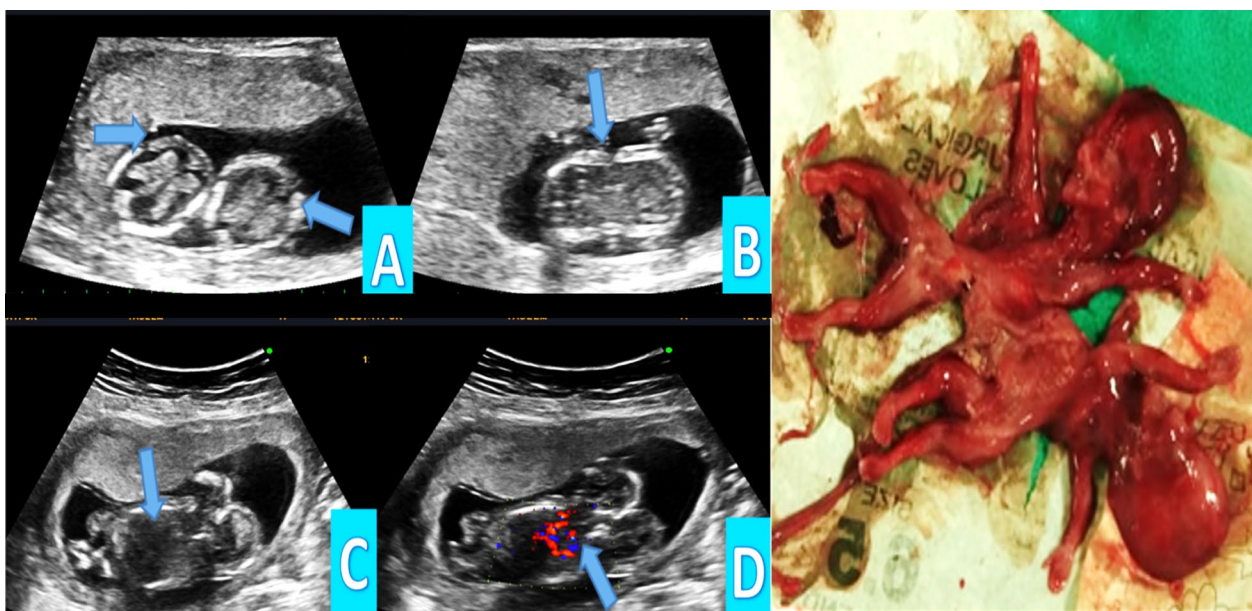


Fig. 2 Trans-abdominal ultrasound images of a thoraco-omphalopagus conjoined twins shows **A** Two separate head (blue arrow), **B** fused thorax and upper abdomen (arrow), **C** common abdominal organ (arrow indicated liver), and **D** common heart (arrow). **E** Autopsy specimen

Case 2

The second case was a pair of thoraco-omphalopagus twins (Fig. 2) with 15 weeks of gestational age diagnosed in a 24-year-old primigravida. USG with curved array probe revealed a single placenta with a single umbilical cord and conjoined twins fused from the thorax to the umbilical region. Twins had two separate heads with a shared sternum, diaphragm, and abdominal wall till the umbilicus. They were sharing a common heart and liver. The pelvis and urinary tract were separated. Each of the twins had two pairs of limbs.

Case 3

The third case was a pair of thoraco-omphalopagus twins (Fig. 3) with 20 weeks of gestational age diagnosed in a 27-year-old second gravida with past obstetric history of normal vaginal delivery of a normal-term child. USG revealed a single placenta with a single umbilical cord conjoined twin fused from the thorax to the umbilical region with the sharing of a common heart and liver. The pelvis and urinary tract were separated. Each of the twins had two pairs of limbs. After obtaining proper consent from the parents, a medical termination of pregnancy was performed while ensuring appropriate concealment.

Case 4

The fourth case was a pair of thoracopagus twins of 13 weeks of gestational age diagnosed in a 24-year-old primigravida. USG revealed a single placenta with

monochorionic monoamniotic, single umbilical cord conjoined twin, fused from thorax region till the upper part of the abdomen facing toward each other with two separate faces, fused thorax with shared heart. Two unfused spines and two separate pelvis. Each of the twins had two pairs of limbs. A 3D ultrasound was done for better visualization of the thoracopagus twin (Fig. 4).

Discussion

Conjoined twinning is a spontaneous occurrence and unrelated to genetic factors [7]. The embryonic categorization of conjoined twins encompasses two primary forms: ventral union, accounting for 87% of cases, and dorsal union, constituting the remaining 13%. These two categories are further subdivided into a total of eight types of conjoined twins, each differing in the scope and specific location of their union [7]. Fusion theory and fission theory are two theories behind the origin of conjoined twins. According to the fusion theory, two separate mono-ovulatory embryonic disks undergo a secondary association after which conjoined twins develop. According to the fission theory, about 13–15 days post-fertilization, the embryonic disk undergoes an incomplete separation leading to some parts of the twin remaining fused. The largest study on conjoined twins was conducted by Edmonds and Layde, a total of 7,803,000 births were analyzed, their study revealing 81 pairs of conjoined twins (an incidence of 10.25 per 1,000,000 births) during

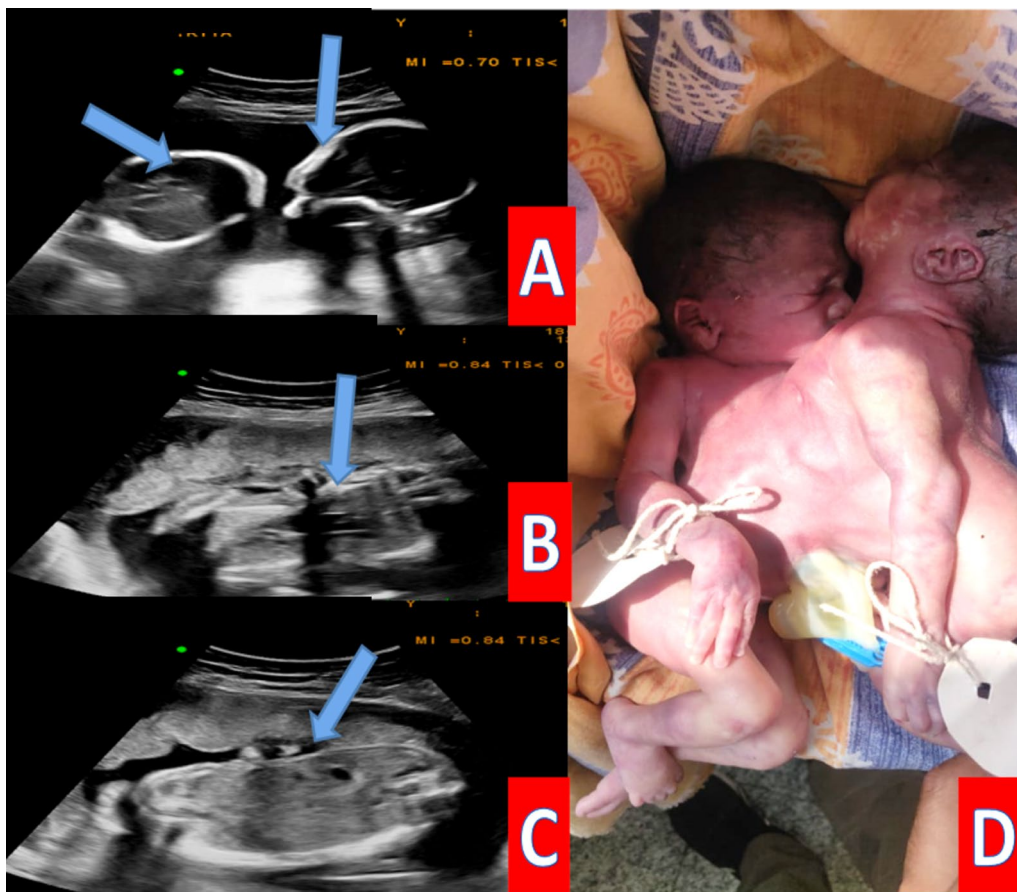


Fig. 3 Trans-abdominal ultrasound images of a thoraco-omphalopagus conjoined twins shows **A** Two separate head (blue arrow), **B** fused thorax and upper abdomen (arrow) with common abdominal organ (arrow indicated liver), and **C** single umbilical cord (arrow). **D** Autopsy specimen

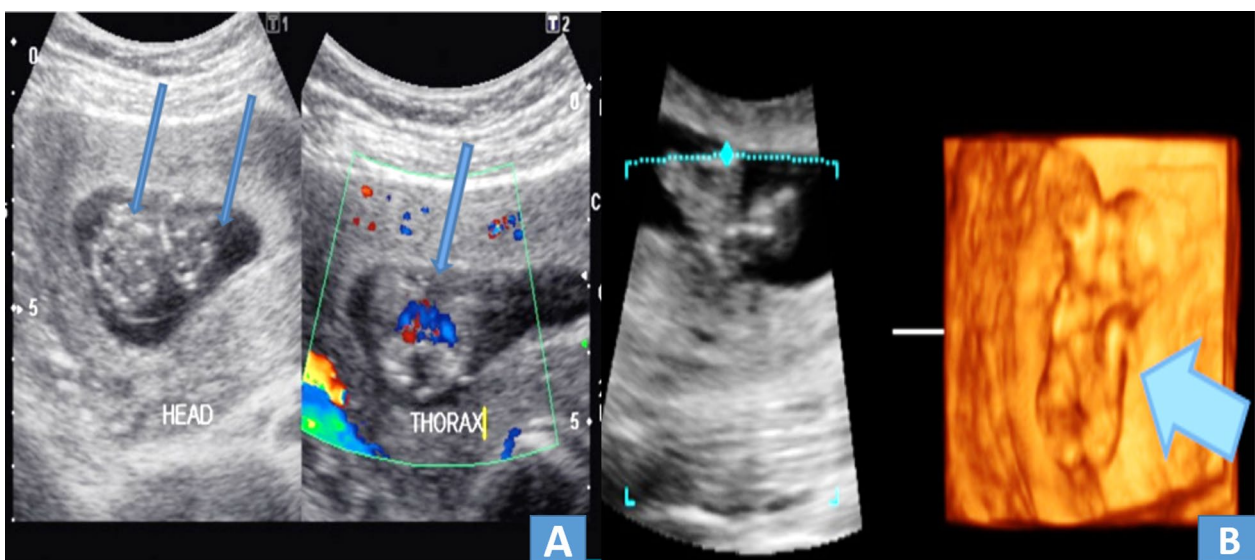


Fig. 4 Trans-abdominal 2D ultrasound images of a thoracopagus conjoined twins show **A** Two separate heads (double arrow in 1st image) with fused thorax and common heart (arrow in 2nd image). **B** 3D Ultrasound was performed for better illustration (arrow)

Table 1 Showing the types of twins from our study with the status of organs between twins

Case	Type of twins	Head	Heart	Liver and GIT	Pelvis organ	limbs	Vertebral column
Case 1	Thoracopagus	Separate	Common	Separate	Separate	Separate	Separate
Case 2	Thoraco-omphalopagus	Separate	Common	Common	Separate	Separate	Separate
Case 3	Thoraco-omphalopagus	Separate	Common	Common	Separate	Separate	Separate
Case 4	Thoracopagus	Separate	Common	separate	Separate	Separate	Separate

the period from 1970 to 1977. Among these cases, the most prevalent type was thoraco-omphalopagus (28%), followed by thoracopagus (18%), omphalopagus (10%), parasitic twin (10%), and craniopagus (6%) [8]. In our study, we observed two cases of each thoracopagus and thoraco-omphalopagus conjoined twins as described in Table 1.

Assessing complex congenital issues during pregnancy is a daunting task. A combination of ultrasound and MRI scans has proven beneficial. While ultrasound offers real-time scanning, color Doppler flow, accessibility, and safety advantages, MRI provides superior detail, improved tissue contrast, and consistent fetal anatomy representation. MRI, being nonionizing and safe, poses no known risks to fetuses [9].

The early detection of conjoined twins has been significantly improved by the integration of first-trimester transvaginal ultrasonography, complemented by color Doppler and 3D ultrasound techniques, and has proven prognostic value [10]. According to Hill [11], the earliest prenatal indication of conjoined twins may manifest at the 7th week of gestation and the degree of suspicion should heighten in cases where a solitary yolk sac is observed alongside two embryos or dividing membranes during very early pregnancies. However, accurate prenatal diagnosis of conjoined twins can be performed with USG as early as 10–11 weeks gestation [12]. USG images are easier for parents to understand, which can help in decision making and counseling. The antenatal USG and Fetal echocardiography should be done optimally at 18 weeks because the degree of cardiac fusion will determine the likelihood of successful separation and postnatal viability of the twins. During ultrasound, conjoined hearts are easier to examine in utero because the amniotic fluid acts as a buffer. Other US features that suggest the possible diagnosis of conjoined twins include constant relative positions of the fetuses with heads and other body parts persistently at the same level on repeat scans, fetus facing toward each other, fused body parts with shared organs, fewer limbs than expected, and a single umbilical cord with more than

three vessels. Fetal MRI is a complementary mode and provides excellent soft-tissue detail and anatomic demonstration of visceral conjunction; multiplanar capability allows accurate evaluation of organ positions and anomalies.

Surgical separation of conjoined twins carries a lot of risks including difficulty during anesthesia, pre-separation phase investigation (angiogramographic examinations), unexpected anatomic variations frequently identified during the surgery, and a high chance of post-operative wound infection. Surgery may be harmful to the body's function reserve and often presents a high mortality rate (up to 50% in 12 separations reported by Spitz [2]).

However, in our case series of thoracopagus and thoraco-omphalopagus twins, it was difficult to treat this form because of a shared heart in all four cases and were poor candidates for surgical separation. Conjoined twins pregnancy carries a high risk of premature delivery. The rate of stillbirth is high and is estimated to be around 60 percent [13]. The parents received information and written consent regarding the malformation and were informed about the potential outcomes if the twins were to survive after delivery. In India, the MTP (Medical Termination of Pregnancy) Amendment Act 2022 allows the termination of a pregnancy of up to 24 weeks. All four cases were below 20 wk of gestation.

Parents were counseled about the risk of medical termination of pregnancy such as excessive bleeding, retained product of conception, and need for dilation and curettage. After considering this information, they chose to terminate the pregnancy and declined any additional assessment or inquiry.

Conclusions

The antenatal diagnosis of conjoined twins can be made with high-resolution ultrasound as early as 12 weeks gestation and is important for optimal obstetric management because early termination of pregnancy has less risk of complications.

Abbreviations

USG Ultrasonography
MRI Magnetic resonance imaging

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Not applicable.

Author contributions

BDC and AG contributed to the acquisition, analysis, conception, design, and drafting of the work. SJ along with BDC and AG contributed to the final draft, revisions, upload, and submission of the final revised work. All authors have agreed both to be personally accountable for their own contributions and ensured that questions related to the accuracy or integrity of any part of the work, even ones in which one was not personally involved, are appropriately investigated, resolved, and the resolution documented in the literature.

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Declarations**Ethics approval and consent to participate**

This work has been approved by Institute Ethical Committee (IEC). Retrospective, consent waiver

Consent for publication

All the authors have approved the submission of the manuscript to your esteemed journal. On behalf of all the contributors, I will act as guarantor and will correspond with the journal from this point onward.

Competing interests

The authors declare that they have no competing interests.

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