DOI: 10.7860/JCDR/2025/75641.20970

Case Report

Obstetrics and Gynaecology Section

Antenatal Diagnosis of Thoracopagus and Parapagus Dicephalus Dibrachius Dipus Conjoined Twins: A Report of Two Cases

AASTHA GARG[1], SIMMI AGGARWAL[2], INDERJEET KAUR[3], RAVINDER GARG[4]

ABSTRACT

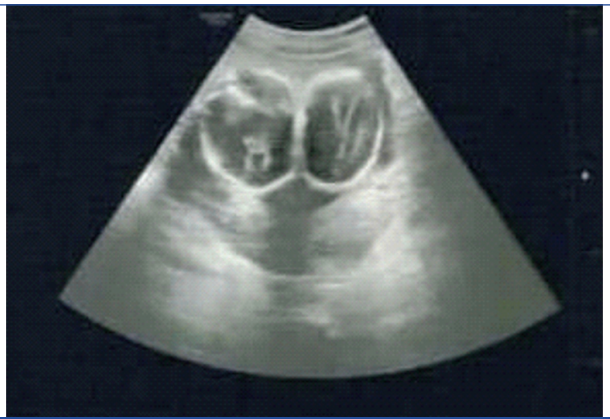
Conjoined twins are a rare obstetric phenomenon, occurring in approximately 1% of monochorionic twin pregnancies. Many present with life-incompatible anomalies; thus, early prenatal diagnosis is crucial for optimal management and delivery. We report two cases: a pair of 18-week-gestation thoracopagus twins (one with dorsolumbar meningocele, deformed limbs, and low-set ears), and a 21-week-gestation parapagus dicephalus dibrachius dipus conjoined twin pair with two separate heads, a common thorax and abdomen, and a single pair of upper and lower limbs. Both were diagnosed via routine ultrasonography and underwent medical termination. This case series highlights ultrasonography's potential in diagnosing and managing complex twin presentations, thereby reducing maternal morbidity and mortality.

Keywords: Birth defects, Congenital anomalies, Mortality, Ultrasonography

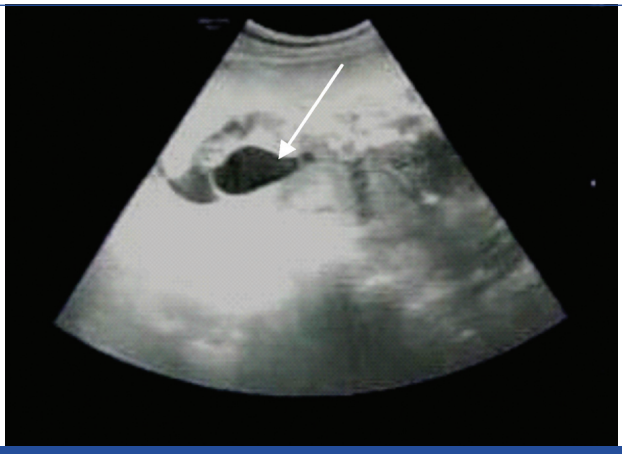
CASE REPORT

Case 1

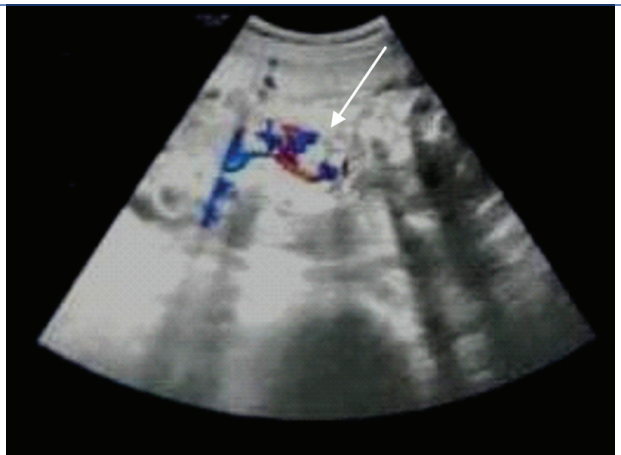
A 24-year-old primigravida presented at her first antenatal check-up at four and a half months' gestation with amenorrhoea. She reported no complaints and had no relevant medical or family history. Routine ultrasonography revealed a single placenta with conjoined twins fused in the thorax and upper abdomen, facing each other with separate heads. The thorax was fused, sharing a single heart. The spines were unfused, and the pelvises were separate. A single umbilical cord was present, and each foetus had two pairs of limbs. One foetus exhibited a cystic dorsolumbar mass with spinal deformity (suggestive of meningocele), deformed limbs, and low-set ears. A diagnosis of thoracopagus conjoined twins with meningocele was made [Table/Fig-1-4]. Following counselling, the parents opted for termination. Autopsy was not performed due to lack of parental consent.



[Table/Fig-1]: USG showing CT twin with separate heads.



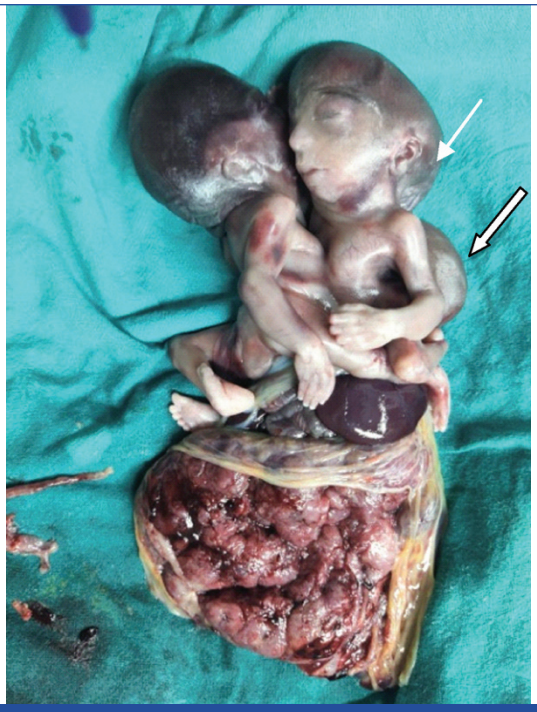
[Table/Fig-2]: USG showing cystic SOL in dorsolumbar region-meningocele (Arrow).



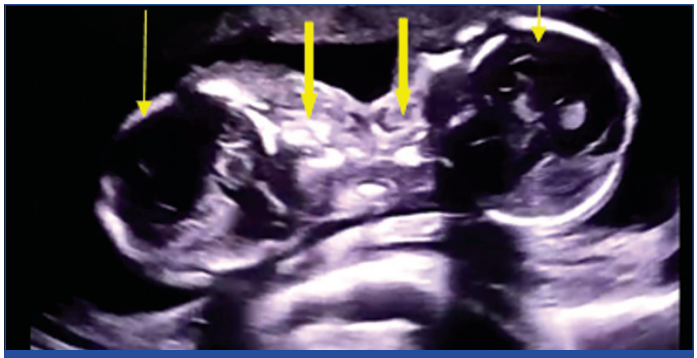
[Table/Fig-3]: USG showing single umbilical cord (arrow).

Case 2

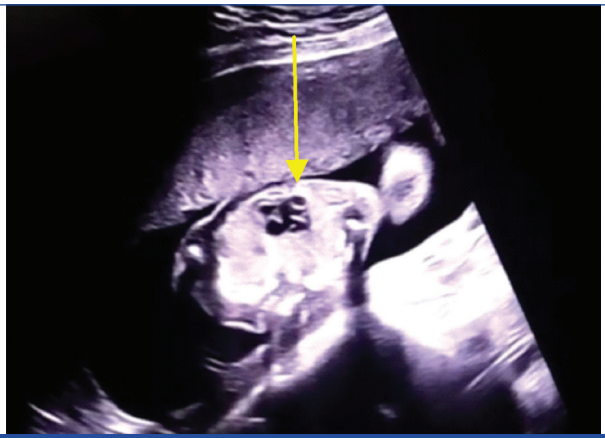
A 20-year-old woman (gravid 1, para 0) presented at her initial obstetric visit at 21 weeks' gestation. She had received no prior prenatal care or ultrasound. She reported infertility for 18 months and had used indigenous medications to conceive. There was no relevant medical or family history, and the marriage was nonconsanguineous. Ultrasonography showed conjoined twins with ventrolateral fusion, a common thorax and abdomen, and an apparently enlarged, fused heart. Two upper and two lower limbs were present. The placenta was anterior, with adequate amniotic fluid [Table/Fig-5,6]. A diagnosis of dicephalic parapagus conjoined twins was made. The patient received education and counselling regarding the congenital malformation, poor prognosis, and the option of pregnancy termination. She opted for medical termination,



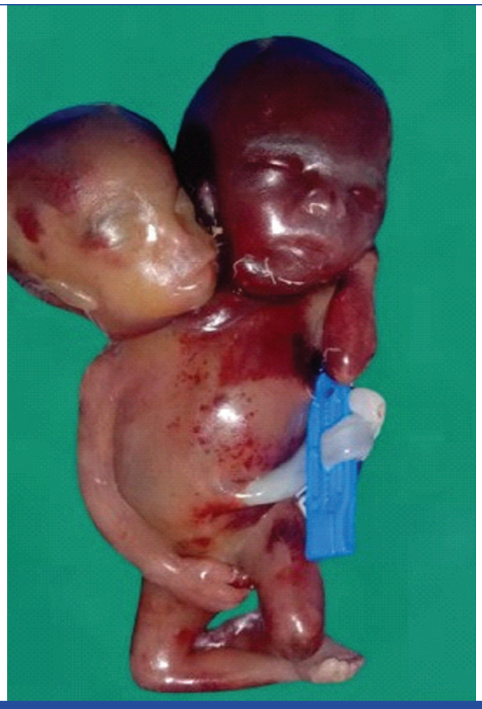
[Table/Fig-4]: Abortus showing conjoined twins with meningocele (thick arrow) with low set ears (thin arrow).



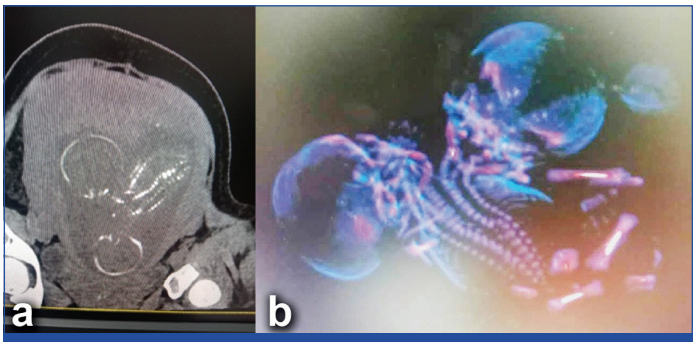
[Table/Fig-5]: Dicephalic parapagus dibrachis dipus conjoined twin separate heads (arrows) with separate cervical spines (open arrows).



[Table/Fig-6]: Dicephalic parapagus dibrachis dipus conjoined twin separate heads (arrows) with separate cervical spines (open arrows).



[Table/Fig-7]: Dicephalic parapagus dibrachis dipus abortus.



[Table/Fig-8]: a,b) CT scan axial MPR and MIP images showing ventrally fused spines with separate heads.

delivering a stillborn male conjoined twin [Table/Fig-7]. A CT scan (performed with informed consent for academic purposes) showed features consistent with conjoined twins, spinal fusion beyond the cervical region, two separate heads, and a single trunk and pelvis, with two upper limbs and two legs and a single umbilical cord and placenta [Table/Fig-8a,b]. Autopsy was not performed.

DISCUSSION

Conjoined twins are extremely rare, with a worldwide prevalence of 1:49,000-189,000 births and a reported 2:1 female predominance [1]. They are always monoamniotic monochorionic, with either fused or partially fused anatomy. Approximately 50% are stillborn, 25% are born with severe congenital anomalies, and 25% survive [1,2]. Thoracopagus twins have the highest incidence (19-56.3%), while parapagus dicephalus twins have a relatively low incidence (13.5-24.2%) [3]. Conjoined twins occur in approximately 1% of monoamniotic monochorionic monozygotic twins [4]. Fission and fusion theories explain conjoined twinning: incomplete division of the developing embryo (fission) or secondary fusion of two embryonic discs after the 13th day postconception (fusion) [5]. Based on embryonic disc union, conjoined twins are classified into ventral and dorsal groups, further subclassified into eight varieties depending on the fusion site(s): omphalopagus (umbilicus/abdomen), thoracopagus (thorax/upper abdomen), cephalopagus (maxillofacial), craniopagus (skull), ischiopagus (pelvis), rachipagus (spine), and pygopagus (sacrum) [3]. Additional terminology includes parapagus (lateral fusion), brachius (upper limb), pus (lower limb), and prosopus (face) [5]. Parapagus conjoined twins are further classified into parapagus dicephalus (two heads, single trunk/abdomen/pelvis, 4-7 limbs) and parapagus diprosopus (two faces on a single head, single trunk/abdomen/pelvis, four limbs). Dicephalus dibrachius dipus parapagus conjoined twins often have a single heart, congenital anomalies, two oesophagi emptying into a single stomach, and partial or complete spinal duplication [3].

CT anomalies are classified into those at the conjunction site ('shared' anomalies) and those beyond ('discordant' or 'concordant'). Concordance indicates the same anomaly with the same or differing severity in both twins; discordance indicates an anomaly present in only one twin, as seen in our thoracopagus case with meningocele in one twin [6]. These discordant anomalies are rare [6].

Banjarnohar DPP et al., reported two dicephalic parapagus cases at 28/29 and 20/21 weeks' gestation [7]. Case 1 showed two separate heads and necks, two vertebral columns, a fused heart (four ventricles, three atria), two stomachs, a fused liver, two pairs of kidneys, two urinary bladders, and two pairs of limbs. Case 2 showed a single large heart, talipes equinovarus in one foetus, an absent lower limb in one foetus, and an occipital lymphangioma colli [7]. Our dicephalic parapagus case also showed a single large heart but no limb anomalies.

On sonography, dichorionic diamniotic or monochorionic diamniotic twin pregnancies rule out conjoined twins. Early sonographic findings showing conjoined body parts despite changes in foetal position or a bifid foetal pole in a monochorionic monoamniotic twin pregnancy suggest conjoined twins [8]. Antenatal diagnosis of organ sharing is crucial for management and determining the feasibility of surgical separation [3]. Three-dimensional sonography, colour Doppler foetal echocardiography, and MRI can confirm the diagnosis, classify the CT type, and assess the extent of organ sharing. Multimodality imaging aids parental counselling by demonstrating disease severity and prognosis [9,10].

CONCLUSION(S)

High-resolution ultrasonography as early as 12 weeks is a valuable tool for diagnosing conjoined twins and guiding treatment. Early antenatal diagnosis enables timely pregnancy termination in cases of unsustainable foetal anomalies.

REFERENCES

[1] Annas GJ. Conjoined twins-the limits of law and limits of life. N Engl J Med. 2001;344:1104-08.
[2] Zubovic SV, Prevljak S, Behmen A, Bektesevic H, Zubovic D, Jusufbegobic M. Conjoinedtwins-Acasereportofprenataldiagnosisofcephalothoracoomphalopagus. Radiography.2020;26(2):e126-e128.Availablefrom:https://www.radiographyonline.com/article/S1078-8174(19)30145-2/abstract.
[3] Byers CD, Young JR, Connell M. Parapagus dicephalus dibrachius dipus conjoined twin case. Appl Radiol. 2018;47(5):29-32.
[4] Bega G, Wapner R, Lev-Toaff A, Kuhlman K. Diagnosis of conjoined twins at 10 weeks using three-dimensional ultrasound: A case report. Ultrasound Obstet Gynecol. 2000;16(4):388-90.
[5] Spencer R. Theoretical and analytical embryology of conjoined twins: Part II: Adjustments to union. Clin Anat. 2000;13(2):97-120.
[6] Boer LL, Winter E, Gorissen B, Oostra RJ. Phenotypically discordant anomalies in conjoined twins: Quirks of nature governed by molecular pathways? Diagnostics (Basel). 2023;13(22):3427. Doi: 10.3390/diagnostics13223427.
[7] Banjarnahor DPP, Wardhana MP, Sulistyono A. Case report: Dicephalic parapagus conjoined twin. Int J Health Scie. 2022:6(S7):2992-97.
[8] Tongsong T, Channprapaph P, Pongsatha S. First trimester diagnosis of conjoined twins: A report of three cases. Ultrasound Obstet Gynecol. 1999;14(6):434-37.
[9] Rama K, Jahagirdar V, Gouri TI, Vadagandla K, Rama A. A case report of Dicephalus Dipus Dibrachius- A rare variant of conjoined twins. Arch Clin Med Case Reports. 2020;4(5):913-19.
[10] Yang PY, Wu CH, Yeh GP, Hsieh CT. Prenatal diagnosis of parapagus diprosopus dibrachis dipus twin with spina bifida in first trimester using two- and three-dimensional ultrasound. Taiwan J Obst Gynecol. 2015;54(6):780-83.

PARTICULARS OF CONTRIBUTORS:
1. Postgraduate Resident, Department of Obstetrics and Gynaecology, PGIMS, Rohtak, Haryana, India.
2. Consultant Radiologist, Department of Radiodiagnosis, Garg Hospital and Diagnostic Centre, Faridkot, Punjab, India.
3. Medical Officer, Department of Radiodiagnosis, Garg Hospital and Diagnostic Centre, Faridkot, Punjab, India.
4. Professor, Department of Medicine, GGS Medical College and Hospital, Faridkot, Punjab, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Simmi Aggarwal,
Garg Hospital and Diagnostic Centre, Main Bazaar, Faridkot, Punjab, India.
E-mail: drsimmigarg@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Sep 20, 2024
- Manual Googling: Jan 14, 2025
- iThenticate Software: Jan 16, 2025 (10%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Sep 19, 2024**
Date of Peer Review: **Nov 20, 2024**
Date of Acceptance: **Jan 18, 2025**
Date of Publishing: **May 01, 2025**