DOI: 10.7860/IJARS/2024/69943.3002

Case Report

Radiology Section

# Antenatal Diagnosis of Non-Janiceps Type of Cephalophagus Conjoined Twins: A Case Report

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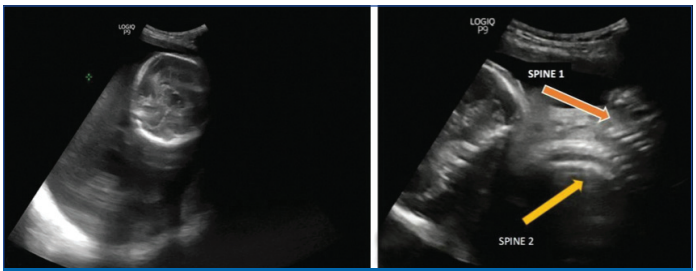
## ABSTRACT

Cephalophagus is the rarest type of conjoined twins. Given the exceedingly dismal prognosis, early prenatal detection is crucial because surgical separation is not an option in many situations. The present case report presents a rare non janiceps cephalophagus conjoined twin pregnancy in a 22-year-old primigravida, identified during the third-trimester antenatal ultrasound. Imaging revealed a single face, a shared thoracic cavity with one heart. There were two lungs, the upper abdomen was fused, and the stomach and liver were shared. Two vertebral columns, separate pelvises, and two pairs of limbs were seen. An omphalocele containing liver tissue was observed. An emergency C-section delivered the cephalophagus foetus, underscoring the vital role of early antenatal ultrasound in managing such cases.


**Keywords:** Cephalophagus twins, Omphalocele, Ultrasound, Vertebral columns

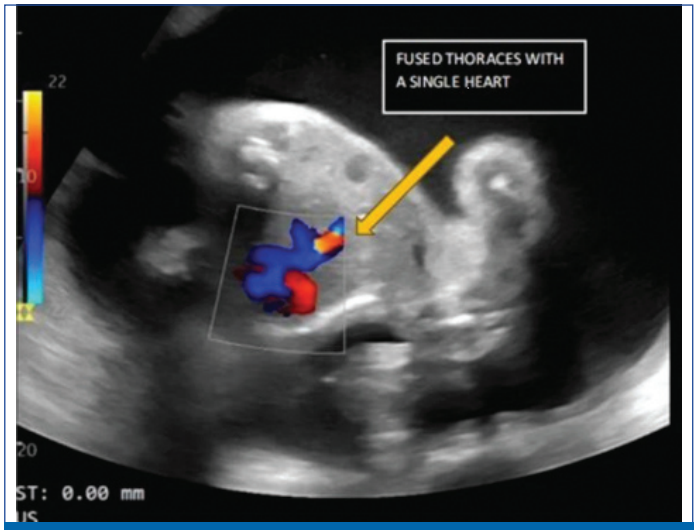
## CASE REPORT

A 22-year-old primigravida first presented to the Department of Radiodiagnosis at a Government Medical College and Hospital in her third trimester of pregnancy for antenatal ultrasound. She had a history of a non consanguineous marriage and had conceived naturally. She was not on any medications/drugs and had no co-morbidities. She came to the hospital for her first prenatal ultrasound. On antenatal Ultrasonography (USG), a live intrauterine conjoint twin pregnancy was noted. The placenta was seen on the fundo anterior wall grade 2 in maturity. Liquor was increased with the single deepest vertical pocket being 18 cm. USG revealed a conjoined twin baby with a single head [Table/Fig-1] and a single face (a pair of eyes, one mouth, one nose), two vertebral columns [Table/Fig-2], Fused thorax with a single heart [Table/Fig-3], two lungs, a fused upper abdomen with a common stomach and liver, two separate pelvises with two separate bladders and two pairs of upper and lower limbs. The femur length of Foetus 1 was 5.47 cm i.e., 28 weeks six days and the femur length of foetus two was 5.55 cm i.e., 29 week two days. In addition, a minor midline defect was observed at the umbilical region, through which a membrane-covered sac was herniating with part of the liver inside it. There was also an insertion of the umbilical cord onto the sac suggestive of an omphalocele.




**[Table/Fig-1]:** Large single foetal head.
**[Table/Fig-2]:** Two vertebral columns in foetus. (Images from left to right)

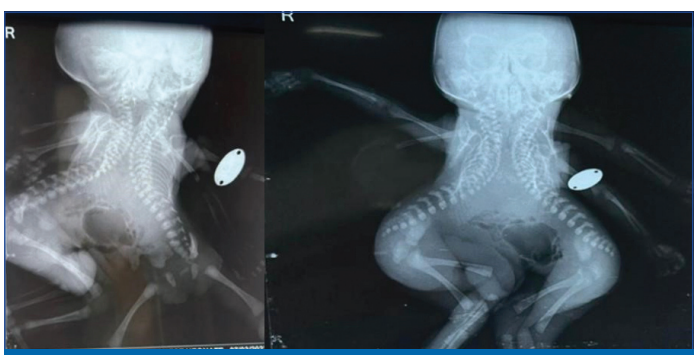
A cephalophagus foetus was delivered by emergency C-section once labour pain started two days after the ultrasound. A postnatal babygram was taken with due consent from the parents which showed a fused cranium, two vertebral columns [Table/Fig-4], and four upper limbs and four lower limbs, with an absent ulnar bone on the right upper limb [Table/Fig-5]. Postnatal ultrasound revealed



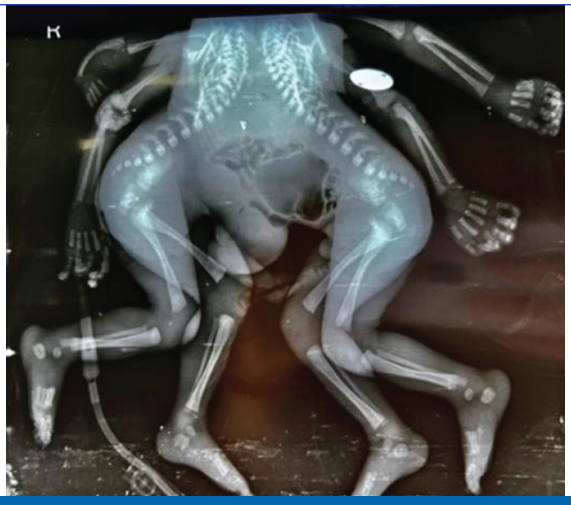

**[Table/Fig-3]:** Fused thoraces with a single heart.

a small omphalocele with gut and liver as contents [Table/Fig-6], with four kidneys noted on either side of two spines [Table/Fig-7]. An axial section of the brain showed a normal frontal horn of the lateral ventricles and a dilated third ventricle [Table/Fig-8], while a sagittal section of the brain showed normal midline structures (Corpus callosum, Cavum septum pellucidum, and cerebellar vermis) [Table/Fig-9].

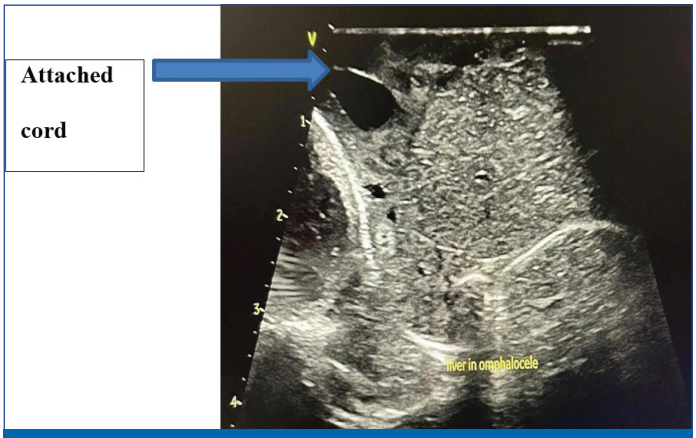
The baby could only survive for a brief period post-delivery even with support. The parents received proper prenatal counselling.



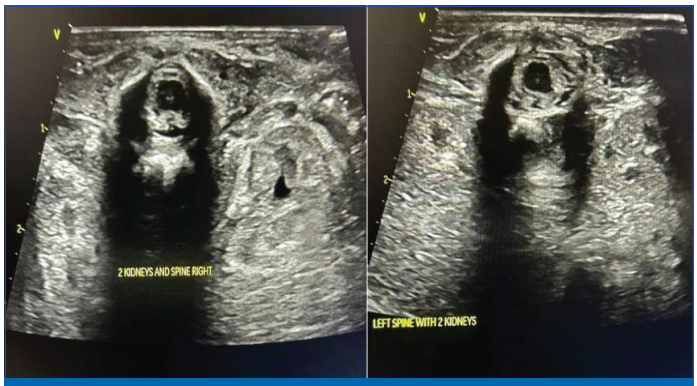
**[Table/Fig-4]:** Postnatal babygram shows fused cranium and b/l vertebral columns.



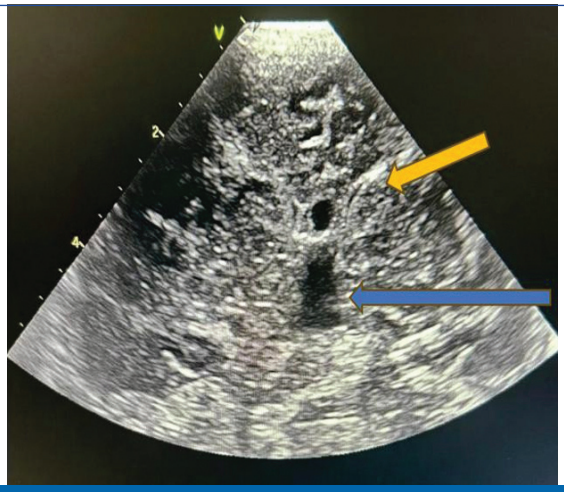
**[Table/Fig-5]:** Babygram showing 4 upper limbs and 4 lower limbs. Absent ulnar bone on right upper limb.



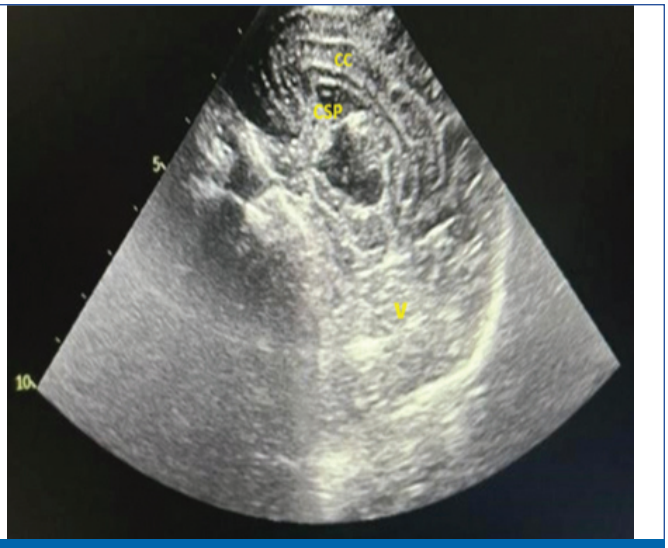

**[Table/Fig-6]:** Postnatal ultrasound showing omphalocele with gut and liver as contents along with attached cord.




**[Table/Fig-7]:** Four kidneys two on either side of spine.



**[Table/Fig-8]:** Axial section of brain showing frontal horn of lateral ventricle (yellow arrow) and dilated third ventricle (blue arrow).




**[Table/Fig-9]:** Sagittal section of brain shows normal midline structures (CC: Corpus Callosum; CSP: Cavum septum pellucidum; and V: Cerebellar vermis).

## DISCUSSION

Conjoined twin pregnancy is a very infrequent occurrence (1 in 100,000 pregnancies) [1], resulting from the failure of complete separation of the embryonic plate beyond 13 days [2].

Conjoined twins come in different varieties, as detailed in [Table/Fig-10] [1]. Out of these, Cephalophagus variants are the least common, with an incidence reported as 1 in 58 of all conjoined twins or 1 in 3 million births [3], where the head, thorax, and upper part of the abdominal cavity are merged. Janiceps (two faces on either side of the head) and non janiceps (one head and a single face) are the two types of cephalophagus twins. These fusions might be ventral or dorsal in embryology. The specific abnormal embryologic process that results in the development of conjoined twins from a single zygote is unknown, although two theories are currently proposed, as outlined by Wedberg R et al., (1979). The collision theory hypothesises that duplicated embryo axes fuse before tissue differentiation [3], while the fission theory hypothesises that embryonic tissue divides incompletely and remains fused locally [4,5].

| Varieties | Description |
|---|---|
| Cephalophagus | The twins are joined at the head and upper body |
| Craniopagus | Joined at the skull share meninges but rarely the brain surface |
| Thoracopagus | Joined face to face at the thorax. It always involves the heart. |
| Omphalophagus | Fusion occurs at the umbilical region also the lower thorax but the heart is never involved |
| Ischiopagus | Fusion includes lower abdomen and duplicated fused pelvic bones, external genitalia and anus |
| Pygopagus | Dorsally fused sharing the perineal and sacrococcygeal area, has only anus but two rectums |
| Rachipagus | Joined in the dorsal aspect with sharing of spinal cords. |
| Parapagus | Laterally joined regularly share the pelvis. Varieties include: |
| | Parapagus dithoracic: Separated thoraces |
| | Parapagus dicephalus: Separated heads |
| | • Parapagus diprosopus: One trunk one head and two faces |

**[Table/Fig-10]:** Classification of conjoined twins [1].

Srinath N et al., reported a similar case of non janiceps type of cephalophagus at 12 weeks of gestation [2]. In the present case, additionally, a small omphalocele was present, which was missed antenatally and could only be detected after the foetus was delivered. Likewise, other associated malformations should also be kept in mind while scanning. For example, Ozkur A et al., in 2006 reported a case of cephalophagus conjoined twins with encephalocele and omphalocele at 24 weeks of gestation. They used both ultrasonography and ultrafast Magnetic Resonance Imaging (MRI)

for diagnosing the same. Dual modalities and an earlier gestational age allowed for better assessment [6].

Inseparable foetal bodies, continuous skin contour at the same anatomical level, bi-breech or bi-cephalic presentations, a single umbilical cord with more than three vessels [1], and a fixed position of the foetus relative to one another [7], shared heart with complex anomalies [8] help in diagnosing the condition. However, these findings on ultrasound are not definitive as they have several drawbacks. Sabih D et al., published a similar case of non janiceps cephalophagus twin diagnosed at 29 weeks. The twin had a delta-shaped head with an absent cerebellum, a fused thorax showing two spines, a shared liver, four kidneys, two bladders, a separate lower abdomen with two male external genitalia and four legs, with multiple vessels in the cord. But unlike in the present case, this twin had a fused thorax with two separate beating hearts [9]. Shared vital organs like the heart (as in the present case) indicate severe conjoining with very little possibility of postnatal correction.

## CONCLUSION(S)

Both antenatal MRI and ultrasound (2D and 3D) play crucial roles in diagnosing conjoined twins. The present case was a rare variety of non janiceps type of conjoined twins with a fused single head and a single face. Early detection and accurate assessment are essential for managing these complex cases.

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**PLAGIARISM CHECKING METHODS:** [Jain H et al.]

- Plagiarism X-checker: Feb 28, 2024
- Manual Googling: May 24, 2024
- iThenticate Software: May 29, 2024 (21%)

**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: **Feb 28, 2024**
Date of Peer Review: **Apr 17, 2024**
Date of Acceptance: **May 30, 2024**
Date of Publishing: **Jul 01, 2024**