

CASE REPORT

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# Prenatal diagnosis of non-janiceps type of cephalopagus conjoined twins: a case report

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## Abstract

**Background** Conjoined twin pregnancy is a very rare variety of twin pregnancy which results from the failure of complete separation of a zygote after 13 days, 'cephalopagus' being the rarest variety of such twin pregnancies. Two forms of cephalopagus conjoined twins have been described in the literature: Janiceps (two faces are on the either side of the head) and non-janiceps (with one head and a single face); non-janiceps type being even rarer. The features that may hint towards the diagnosis on ultrasound include inseparable fetal bodies, unvarying relative positioning of the two fetuses, both heads persistently at the same level to each other, bi-breech or bicephalic presentations and a single umbilical cord with more than three vessels. 3-D ultrasound and Fetal MRI may help in confirmation of this rare entity.

**Case presentation** A 19-year-old primigravida, presented at 12 weeks of gestation for antenatal ultrasound. Ultrasonography showed evidence of a conjoined twin with a single head and single face, fused thoracic cavities with single heart, two lungs, fused upper abdomen with shared stomach and liver, separate pelvic cavities with two different urinary bladders and two pairs of upper and lower limbs. Fetal MRI corroborated these findings. The pregnancy was terminated vaginally, and cephalopagus fetus was delivered. The parents were appropriately counselled for future pregnancies.

**Conclusions** A very rare case of non-Janiceps variety of cephalopagus conjoined twin gestation which was detected at 12 weeks of gestation by antenatal ultrasound and confirmed on fetal MRI. Early antenatal diagnosis is extremely important for appropriate and timely management as the prognosis is very poor in such twins. Antenatal ultrasound and fetal MRI prove very beneficial in this aspect as well as for diagnosis of other associated fetal abnormalities.

**Keywords** Case report, Conjoined twins, Cephalopagus, Non-janiceps, Fetal MRI

## Background

Conjoined twins are a form of monozygotic monochorionic twins and occur when the embryonic plate fails to completely separate between 13 and 17 days. The exact prevalence of conjoined twins is not known. However, it varies between 1:50,000 and 1:200,000 total births [1]. It is more common in the black population and common in female fetuses [1], [1]. Conjoined twins have

been classified according to the site of fusion (cranium, face, thorax, abdomen, sacrum, pelvis or back) as further detailed in Table 1 [3].

Cephalopagus twinning is the rarest type of conjoined twinning accounting for about 1 per million births [4] and has a very poor prognosis. Because of extensively shared organs, surgical separation of such twins after delivery have been rarely successful. Hence, early antenatal detection using ultrasound and fetal MRI as a complementary, non invasive modality of investigation for appropriate and timely management followed by parental counselling is necessary.

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**Table 1** Classification of conjoined twins according to the site of fusion

Types	Definitions
Cephalopagus	Fused from head to umbilicus. One or two faces on either side of the fused head
Thoracopagus	Face to face fusion involving upper thorax to upper abdomen with usually a common heart
Omphalopagus	Fused at the umbilical level and share abdominal organs but never the heart
Ischiopagus	Fusion of the lower abdomen, duplicated fused pelvic bones, external genitalia and anus
Parapagus	Fused laterally with a common pelvis. Types of parapagus conjoined twins are <i>parapagus dithoracic</i> (separated thoraces), <i>parapagus dicephalus</i> (one trunk two separate heads), and <i>parapagus diprosopus</i> (one trunk, one head and two faces)
Craniopagus	Fused at the skull with shared meninges. However, brain surface, face and trunk are separate
Pygopagus	Type of dorsal fusion with shared perineum, sacrococcygeal area and anus. Rectum is separate
Rachipagus	Type of dorsal fusion involving the dorsolumbar vertebral column and rarely the cervical vertebrae and the occipital bone

If diagnosed prior to 24 weeks, vaginal delivery is preferred. Post 24 weeks of gestation, caesarean section is preferred to avoid any birth trauma. Delivery is always preferred in a tertiary care centre for want of immediate paediatric and paediatric surgical support. Surgical separation is not done in cephalopagus twins as the success rate is negligible [5–8]. Parents need to be counselled for further pregnancies.

### Case presentation

A 19-year-old female, primigravida, presented at 12 weeks of gestation for antenatal ultrasound. She had a non consanguineous marriage and had no co-morbidities. She had conceived naturally. There was no history of twinning in family. She reported for the first obstetric ultrasound to our centre. USG showed evidence of a conjoined twin with a single head with single face (a pair of eyes, one mouth, one nose), single heart within single thoracic cavity, two lungs, fused upper abdomen with shared stomach and liver, two vertebral columns, separate pelvises with two different bladders and two pairs of upper and lower limbs. The findings were better appreciated on oblique/parasagittal views than true axial sections. 3-D Ultrasound was not carried out due to technical limitations. However, the patient was taken up for fetal MRI study. Fetal MRI revealed a single head with two optic globes, two cerebral hemispheres, two spinal canals were seen to enter the base of skull, the presence of a single heart, a pair of lungs, shared liver, two urinary bladders and two pairs of upper and lower limbs were confirmed (Figs. 1,2,3).

A cephalopagus fetus was delivered vaginally.

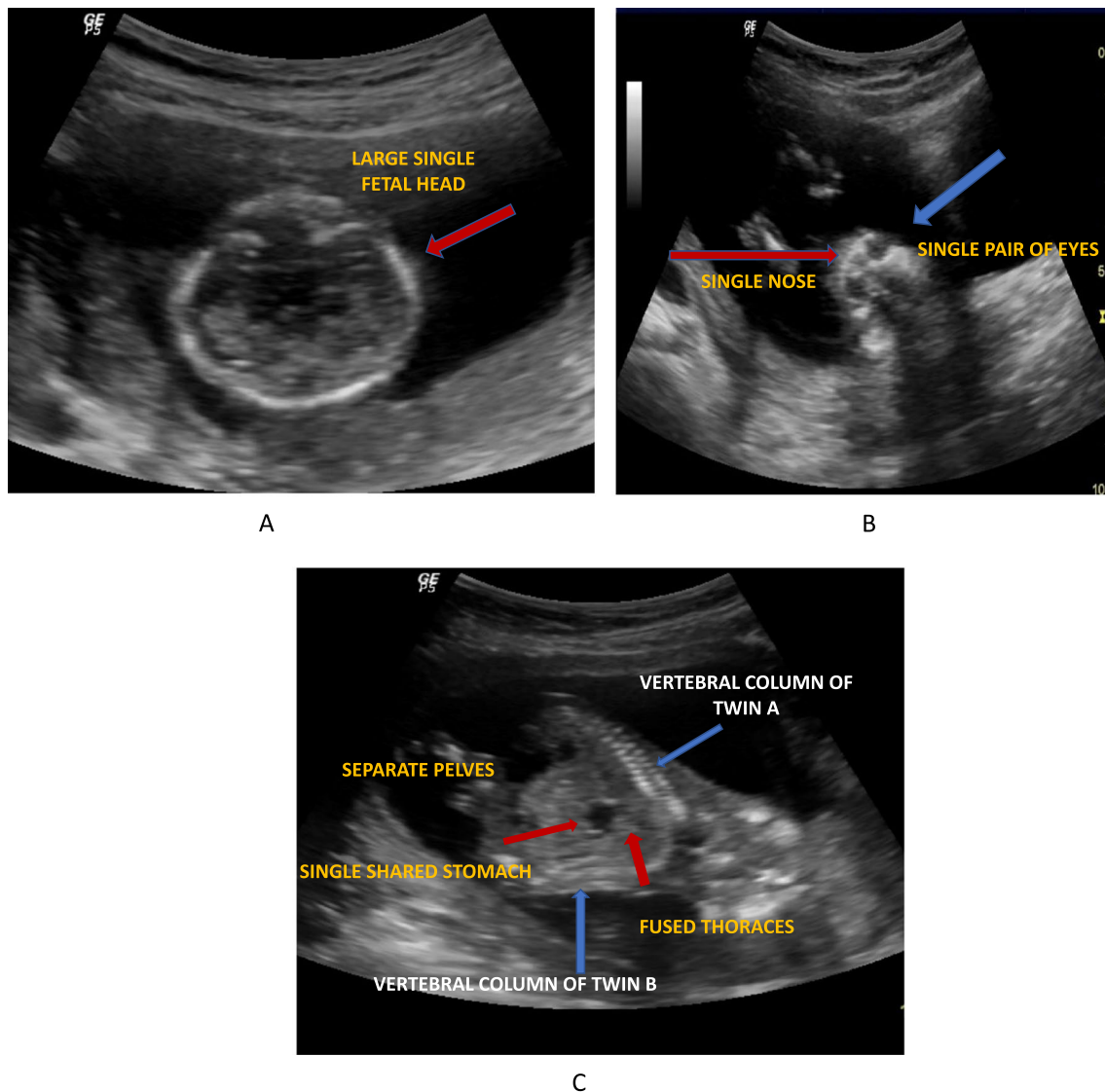
The parents were appropriately counselled for future pregnancies.

### Discussion

Conjoined twins are all monozygotic monochorionic twins and occur due to incomplete separation of the embryonic plate between 13 and 17 days [1]. Embryologically, these fusions can be ventral or dorsal. Various classifications are available of the conjoined twins. However, a commonly used classification is as described in Table 1 [2], 2].

Cephalopagus are among the least common types of ventral fusion variety of conjoined twins with an incidence of about 1 in a million births. These twins have fused head, thoracic cavity and upper part of abdominal cavity. Two forms of cephalopagus conjoined twins have been described in the literature: *Janiceps* which have two faces are on the either side of the head and *non-Janiceps* with one head and a single face; non-janiceps type being even rarer [4]. Usually, these twins die in utero or immediately after birth. The type that we encountered was a non-janiceps type of cephalopagus twins.

Among the organs, these twins usually have a conjoined heart and liver. The lower abdomen, pelvis and limbs are however not fused. High level of suspicion is required to diagnose such cases on an ultrasound. One can diagnose a conjoined twin pregnancy as early as at 12 weeks of gestation. Possibility of conjoined twin should be suspected in any monochorionic monoamniotic pregnancy (single placenta with no separating amniotic membrane) [5],5]. The findings on ultrasound that would help in diagnosing conjoined twins include inseparable fetal bodies, unvarying relative positioning of the two fetuses, both heads persistently at the same level to each other, bi-breech or bicephalic presentations and a single umbilical cord with more than three vessels [7],7]. Almost 50% of cases of conjoined twins are complicated with polyhydramnios [9]. 3-D ultrasound can help in determining the extent



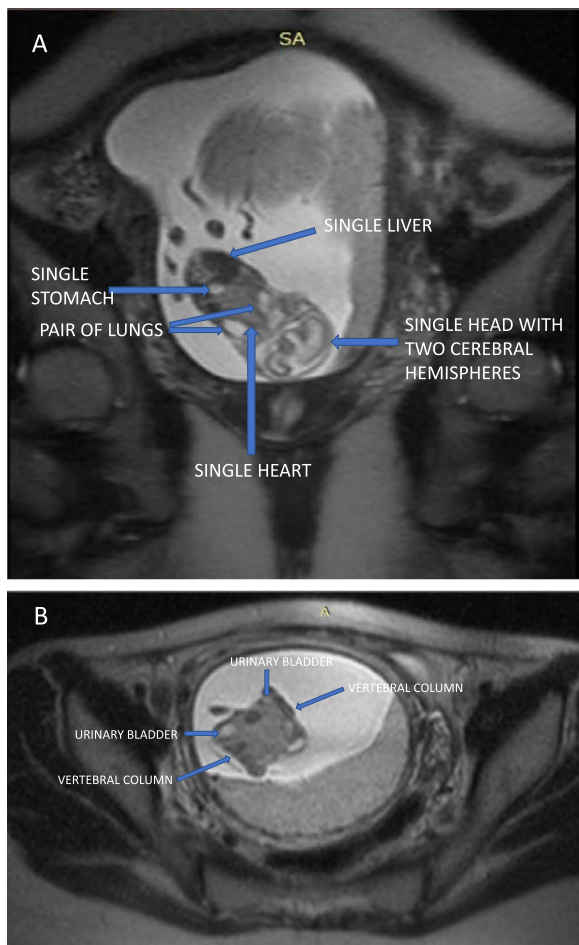
**Fig. 1** A USG showed a conjoined twin with a single head (red arrow); B: single face with a pair of eyes (blue arrow) and one nose (red arrow)

of fusion of conjoined twins and thus help in proper classification, which would further help in appropriate management.

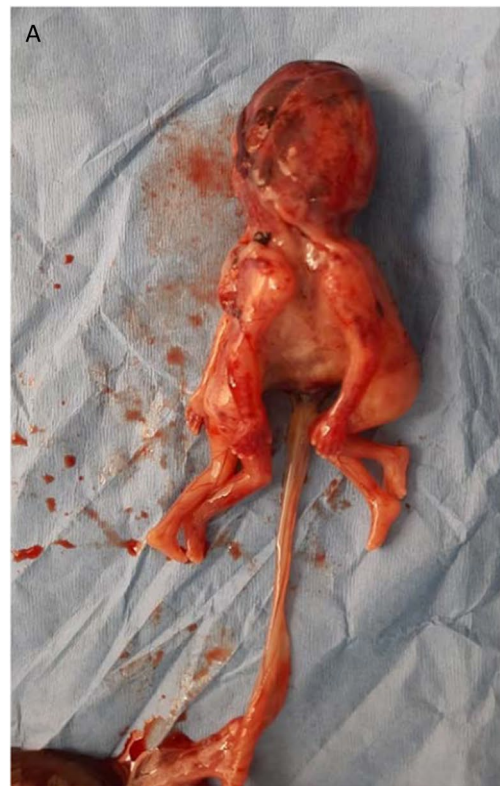
Fetal MRI can help as a complimentary mode of investigation to the ultrasound as it can give reproducible fetal anatomy with better and detailed delineation of the associated congenital fetal abnormalities. MRI provides better tissue contrast and has no known deleterious effects on the fetus. The newer faster MRI sequences like HASTE or SSFE sequences provide images with good

resolution, better T2-weighted contrast and lesser degradation of images due to fetal movement [10, 11]. Also, the larger field of view of MRI provides better delineation of anatomical abnormalities and relationship between normal structures.

Diagnosis of conjoined twins can be missed in case of a bicephalic fetal presentation where one of the fetal heads is engaged and another one is floating. Also in case of severe fusion, conjoined twin pregnancy may be misdiagnosed as a singleton pregnancy. In a few cases of



**Fig. 2** **A:** Fetal MRI coronal T2WI confirmed findings of a single head with two optic globes, two cerebral hemispheres, two spinal canals seen to enter the base of skull, a single heart, a pair of lungs & shared liver (blue arrows) **B:** Fetus showed two urinary bladders and 2 pairs of upper and lower limbs. (blue arrows)



**Fig. 3** A single head and face were seen with two eyes, two ears, single nose, and mouth. No cleft-palate/-lip was seen (A–B). No obvious other facial structures/skin tags were identified on the other side of the head

dichorionic diamniotic twins, the separating twin membrane is not easily visible giving rise to a false suspicion/diagnosis of a conjoined twin.

### Conclusions

Prior to the era of ultrasound, conjoined twins were rarely diagnosed antenatally. These usually presented with dystocia and such fetuses had to be delivered by fetal destructive manoeuvres associated with high incidence of birth injuries. Many radiological signs were described which have now become of historical importance [12, 13]. Now with the advent of 3-D ultrasound techniques and faster MRI sequences, early prenatal diagnosis and appropriate classification of conjoined twins has become easier for optimal and timely obstetric as well as post natal management.

**Abbreviations**

USG	Ultrasonography
3-D	Three dimensional
MRI	Magnetic resonance imaging

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**Author contributions**

NS, NR and PR were involved in manuscript formation and analysis. AB helped in manuscript analysis. NS, NR and PR helped in data collection. AB contributed to data collection. All authors have read and approved the manuscript.

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**Availability of data and material**

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

**Declarations****Ethics approval and consent to participate**

Approval and consent from Institutional Ethical Committee, Armed Forces Medical College has been taken for research purpose. Written informed consent was obtained from the patient for publication of this case report and accompanying images.

**Consent for publication**

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

**Competing interests**

There are no competing interests declared by the authors.

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