

A case report on prenatal diagnosis of Non Janiceps type of cephalopagus conjoined twins

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Case Report

Keywords: Conjoined twins, cephalopagus, non-Janiceps, fetal MRI

Posted Date: August 31st, 2022

DOI: <https://doi.org/10.21203/rs.3.rs-2008787/v1>

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Abstract

Background

Conjoined twin pregnancy is a rare occurrence resulting from the failure of a zygote to separate completely after 13 days. Cephalopagus type is the rarest type of conjoined twins. There are two types of cephalopagus twins: Janiceps (two faces are on the either side of the head) and non-janiceps (with one head and a single face). Non-janiceps type of cephalopagus are extremely rare variety of conjoined twins.

Case presentation:

A 19Y old female, primigravida, presented at 12 weeks of gestation for antenatal ultrasound. USG showed evidence of a conjoined twin with a single head with single face, fused thoraces with single heart, two lungs, fused upper abdomen with shared stomach and liver, separate pelvises with two different bladders and two pairs of upper and lower limbs. Fetal MRI corroborated these findings. The pregnancy was terminated and cephalopagus fetus was delivered vaginally. The parents were appropriately counselled for future pregnancies.

Conclusion

An extremely rare case of non-Janiceps type 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.

Background

Conjoined twins are a form of monozygotic monochorionic twins and occur when the embryonic plate fails to completely separate between 13–17 days. The exact prevalence of conjoined twins is not known. However, it varies between 1:50,000 to 1:200,000 total births [1]. It is more common in the black population and common in female fetuses [1][2]. Conjoined twins have been classified according to the site of fusion (cranium, face, thorax, abdomen, sacrum, pelvis or back. These fusions can be ventral or dorsal.

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.

Case Presentation

A 19Y 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), fused thoraces with single heart, 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. Figure 1(A-C):USG showed a conjoined twin with a single head, single face with a pair of eyes, one mouth & one nose (A), fused thoraces with single stomach & heart (B), fused upper abdomen with shared stomach and liver & two vertebral columns (C).

Fetal MRI showed: 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 2 pairs of upper and lower limbs were confirmed. Figure 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. (B): Fetus showed two urinary bladders and 2 pairs of upper and lower limbs.

A cephalopagus fetus was delivered vaginally. A single head and face were seen with two eyes, two ears, single nose, and mouth. No cleft-palate/-lip was seen (**Fig. 3A-B**). No obvious other facial structures/skin tags were identified on the other side of the head. 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 13–17 days. 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][3]}.

Types	Definitions
Cephalopagus	There are two faces and are joined from the top of the head to the umbilicus
Thoracopagus	Are joined face-to-face from the upper thorax to the upper part of the abdomen and always involve the heart
Omphalopagus	The fusion includes the umbilicus region frequently at the lower thorax, but never the heart
Ischiopagus	The union usually includes the lower abdomen and duplicated fused pelvic bones, and external genitalia and anus are always involved
Parapagus	Are laterally joined, regularly share the pelvis. Varieties of parapagus conjoined twins are parapagus dithoracic (separated thoraces), parapagus dicephalus (one trunk two separate heads), and parapagus diprosopus (one trunk, one head, and two faces)
Craniopagus	Joined by the skull, share meninges but rarely the brain surface and do not include the face and trunk
Pygopagus	Are dorsally fused sharing the perineal and sacrococcygeal areas, has only one anus but two rectums
Rachipagus	Dorsally fused, the defect may involve the dorsolumbar vertebral column and rarely the cervical vertebrae and the occipital bone

Cephalopagus are the rarest variety among the ventral union sub group of conjoined twins with an incidence of about 1 in a million births. The twins with this disorder have their head, thorax and upper part of their abdominal cavities fused. There are two types of cephalopagus twins: Janiceps (two faces are on the either side of the head) and non-janiceps (with one head and a single face). Non-janiceps type of cephalopagus are extremely rare variety of conjoined twins [4]. These twins usually 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). 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. Almost 50% of cases of conjoined twins are complicated with polyhydramnios [4][5]. 3-D ultrasound can help in determining the extent 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[6][7]. 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 dichorionic diamniotic twins, the separating twin membrane is not easily visible giving rise to a false suspicion / diagnosis of a conjoined twin.

Conclusion

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 [8][9] which have now become of historical importance. 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. 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 [10]. Parents need to be counselled for further pregnancies.

Abbreviations

USG

Ultrasonography

MRI

Magnetic resonance imaging

Declarations

Ethics approval and consent to participate: Ethical committee approval and consent has been taken for research purpose.

Consent for publication: The patient provided written informed consent for publication of the data of this study.

Availability of data and material: The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests: There are no competing interests declared by the authors.

Funding: This research work has not been funded.

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

Acknowledgements: Dept of Obs-Gyn, Armed Forces Medical College, Pune.

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Figures



A



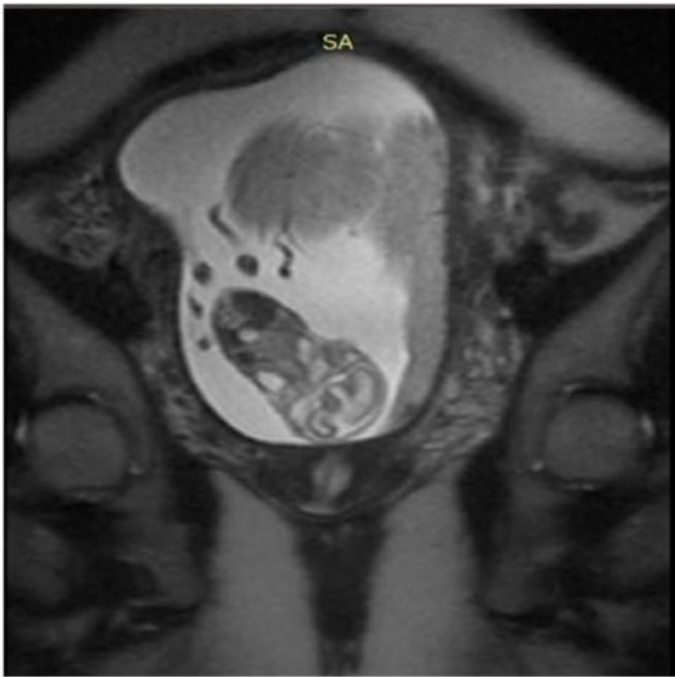
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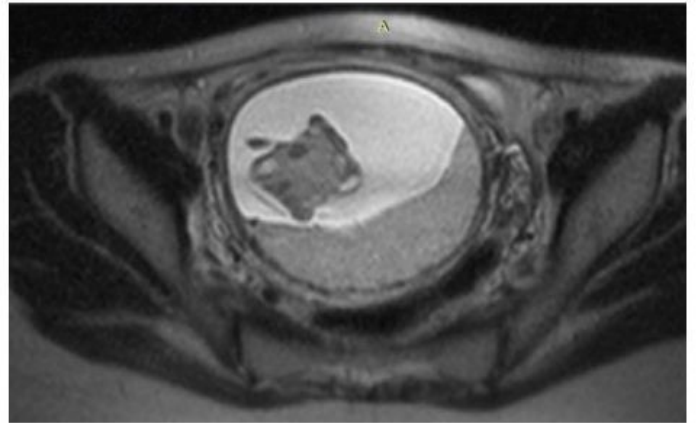
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Figure 1

A. USG showed a conjoined twin with a single head , single face with a pair of eyes, one mouth & one nose. B. The twin had fused thoraces with single stomach & heart. C. There was fused upper abdomen with shared stomach and liver & two vertebral columns noted.



A



B

Figure 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. B. Fetus showed two urinary bladders and 2 pairs of upper and lower limbs.



A



B

Figure 3

A. Post delivery photographs showing fetus with single head with facial features only on one side, two pairs of upper limbs. B. There were two pairs of lower limbs with fused thoraces & separate lower abdomens and pelvises were noted.