



Conjoined twins with a single heart: A rare case report

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CASE REPORT

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Abstract

A case of abdomino-thoracopagus twins with a single heart is described. The male twins were delivered in the 15th week of gestation following the parents' request to terminate the pregnancy. This case is of particular interest because of the rarity of the abdomino-thoracopagus twins with a single heart, in the literature.

Key Words: Conjoined twins, abdomino-thoracopagus, single heart

Background

Although rare, with an incidence of between 1 in 50,000 to 1 in 100,000 births¹⁻², the possibility of conjoined twins must be excluded in a pregnancy. Successful surgical separation of thoracopagus twins with separate hearts has been reported, but the success rate for separation of the same with a single heart is very low.³ Hence their antenatal diagnosis should be made very early in pregnancy. Colour Doppler imaging is useful for imaging the cardiovascular anatomy in suspected.

conjoined twins of having a fused hearts, even at an early gestational age.⁴

Conjoined twins who survive after separation are mostly females, at a ratio of three to one over males, the reason is not known. The other, and most commonly held, theory is that conjoined twins result from the incomplete division of the early embryo with two foetuses, but joined at various sites. Monozygotic identical twins, whether separate or conjoined, are identical in sex and genetic make-up and share a common placenta.

Incomplete division of the embryo is associated with incomplete formation of the organ systems and thus conjoined twins with fused organs usually have incompletely developed hearts and liver.⁵

Case Report

A young primigravida aged 25 years reported to the outpatient department with a history of 15 weeks of amenorrhea. She had been married for six months. Routine antenatal investigations and electrolytes, creatinine and sugar levels were normal. Urine analysis was normal. HIV, HBsAg, VDRL were all negative. Her blood group was O positive.

On routine screening ultrasound revealed conjoined twins, abdomino-thoracopagus with a single cardiac activity. There was no history of any drug intake or twins in the family. The two foetuses with a single cardiac activity were fused at chest and abdomen. Two separate spines were seen (Figure 1). Amniotic fluid was within normal limits and there was a single placenta. All confirmed the diagnosis of conjoined



twins, abdomino-thoracopagus, at 15 weeks of gestation. Colour Doppler studies further supported the diagnosis (Figure 2).



Figure 1: Conjoined twins with two separate spines

Pedigree analysis of both parents: revealed no similar cases in the family. Karyotyping of both parents revealed no chromosomal abnormality.

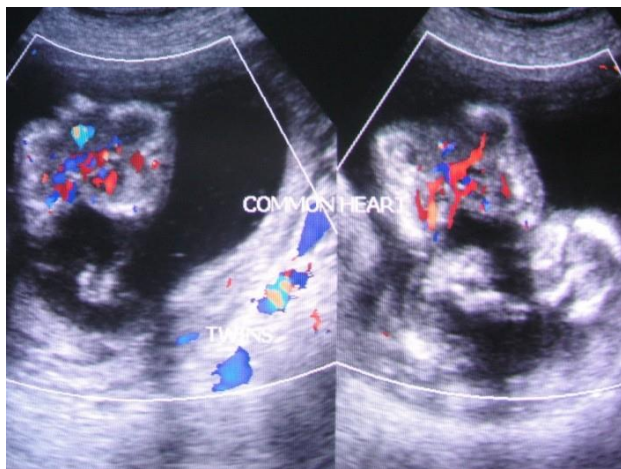


Figure 2: Colour Doppler showing single cardiac activity

Management: The parents were counselled and the pregnancy was terminated with the help of misoprostol, without any complications, after obtaining written consent.

On examination: Conjoined male twins of 15 weeks size abdomino-thoracopagus with monochorionic monoamniotic placenta measuring 12x10.5cm with complete cotyledons (Figure 3). Histopathology of placenta: revealed mature placental tissue.



Figure 3: Clinical photograph of conjoined twins

Discussion

Conjoined twins is a rare congenital abnormality that affects 1 in 200 mono-zygotic twins. Although the exact aetiology of conjoined twins is unknown, it arises as a result of unequal and incomplete fusion of the embryonic disk beyond the 12th day of fertilisation.⁴ Late embryonic splitting and haemodynamic imbalances due to the large and multiple anastomosis may account for the extremely high prevalence of intrauterine foetal deaths in conjoined twins, being as high as 60%.⁶

Preoperative evaluation of the cardiovascular system is essential in all conjoined twins to ascertain whether or not the hearts are conjoined as determined from prenatal studies. Echo-cardiogram and electrocardiography (EKG) are performed to determine the nature of anomalies in the case of separate hearts and to determine the sites where the hearts are joined and the presence of abnormalities in the case of conjoined hearts.

The presence of a single heartbeat on EKG usually indicates that successful separation would not be possible, but the presence of two separate beating systems necessarily indicates a better outcome.

Techniques such as MRI are used for specific purposes such as evaluating the heart. Prenatal ultrasound imaging and three-dimensional MRI of the heart and other organs usually provide sufficient information to help families decide whether or not to continue the pregnancy.



As the heart anomaly is a crucial factor in survival, its detection in very early pregnancy is important. There is a sex predilection of 70–90% being females.⁷ Cases are classified according to the anatomical area of conjunction with 75% being the thoracopagus.⁷ In our case there was a single heart in colour Doppler imaging.

Prenatal ultrasound is also important in planning the obstetric management of conjoined twins who frequently have a breech presentation. Caesarean section would appear to be the approach to delivery when the diagnosis is known ahead of time. Conjoined twins often cause premature labour, so efforts must be made to inhibit labour to permit the foetus to become as mature as possible for survival.

Determination of conjoined twins necessitates multidisciplinary discussion, as to the prognosis and possibility of surgical correction wherever feasible and with full involvement of parents. Many ethical considerations emerge, such as: is separation possible with any reasonable chance of success? What will the quality of the twins' life be following separation? Will one or both survive? Is it reasonable to attempt separation when it is known ahead of time that only one can survive? Should a pregnancy be terminated if conjoined twins are discovered in utero that they are inseparable?

Conclusion

Conjoined twins although rare is a well-known entity. It is very important to make a diagnosis in time to prevent complications. An ultrasound and colour Doppler proves to be of great help in making the early diagnosis.

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PEER REVIEW

Not commissioned. Externally peer reviewed.

CONFLICTS OF INTEREST

The authors declare that they have no conflict of interest.

CONSENT

The authors declare that:

1. They have obtained informed consent for the publication of the details relating to the patient in this report.
2. All possible steps have been taken to safeguard the identity of the patient.
3. This submission is compliant with the requirements of local research ethics committees.