

# Epigastric heteropagus conjoined twins combined with TRAP sequence

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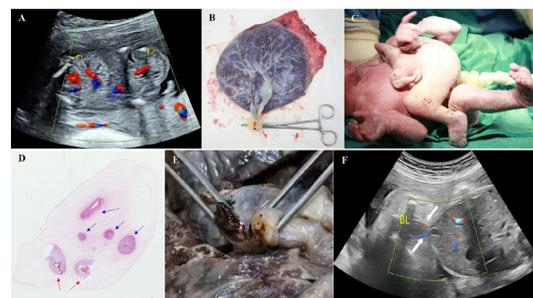
## DESCRIPTION

Epigastric heteropagus is a rare condition that refers to unequal and asymmetric conjoined twins in which the parasite is smaller and usually attached to the epigastrium of the autosite.<sup>1</sup> Twin reversed arterial perfusion (TRAP) sequence refers to a rare complication unique to monochorionic twin pregnancies in which a twin with an absent or rudimentary heart ('acardiac twin') is perfused by its co-twin ('pump twin') via aberrant arterioarterial anastomoses.<sup>2</sup> Here, we report an extremely rare case of epigastric heteropagus combined with TRAP sequence.

A primipara in her 30s was admitted to the hospital at 35 gestational weeks because of fetal abnormalities. At 12 gestational weeks, a singleton with a nuchal translucency of 4.9 mm and omphalocele were observed. She refused chorionic villous biopsy but underwent observation. In the 17th gestational week, a systematic ultrasound showed a monochorionic monoamniotic twin pregnancy with the absence of cardiac tissues in one of the twins and uncertain short bands between the twins. Amniocentesis was performed, and the karyotyping, chromosomal microarray analysis and whole-exome sequencing results of the amniotic fluid cells were normal. Follow-up antenatal ultrasound showed monochorionic monoamniotic conjoined twins ([figure 1A](#)). Informed consent was obtained, and the couple decided to continue the pregnancy. At the

35th gestational week, an increased cardiothoracic ratio, mild tricuspid valve regurgitation, an elevated peak flow velocity of the middle cerebral artery of the pump twin and a significantly increased abdominal circumference of the acardiac twin (over 70% when compared with that of the pump twin) were detected. Considering the progression of the disease and the risk of heart failure for the pump twin, an emergency caesarean section was performed.<sup>3</sup> A newborn boy with an omphalocele and a parasite in his abdomen, weighing 2900 g in total, was delivered. The Apgar score was 9 at 1 min and 10 at 5 min. A single, normal-appearing umbilical cord was attached from the autosite to the only placenta ([figure 1B](#)). The parasite, with only part of a trunk, double rudimentary upper limbs, a pelvis, double lower limbs and normal appearing male external genitalia, was seen ([figure 1C](#)). The parasite was connected to the autosite's upper abdomen, 3 cm above the omphalocele. The diagnosis was revised to be epigastric heteropagus conjoined twins.

The newborns underwent separation surgery 2 days after birth. Incredibly, two umbilical arteries and one umbilical vein were confirmed in the connective tissue, combined with a small blood vessel branch off the right subclavian artery (images not included). Where did the umbilical blood vessels come from? Three days later, the placental histopathological examination results were released



**Figure 1** Antenatal ultrasounds, physical features and pathological examinations of the appendages of the newborn. (A) Ultrasound at 17 gestational weeks. F1 shows the autosite and F2 shows the parasite. (B) The placenta showed one umbilical cord. (C) The autosite and parasite a few minutes after birth. (D) Histopathological examination showed four umbilical arteries (blue arrows) and two umbilical veins (red arrows). (E) The separation part of the umbilical cord 2 cm before the insertion into the placenta. (F) Ultrasound showing double reversed umbilical arteries separated on either side of the bladder of the parasite from the autosite (arrows).

## Patient's perspective

When my family and I were told that my baby was abnormal, I was stunned. However, when we got the professional advice from the doctor, we decided to keep the child and take follow-up examinations. Although there were some twists and turns in the follow-up examination, we persevered, we were lucky and now we have a healthy child, thanks to the efforts of all the physicians.

The mother of the newborn wrote the perspective. Translation was performed by the author.

## Learning points

- ▶ Ultrasound plays an important role in identifying twin reversed arterial perfusion (TRAP) sequence.
- ▶ The umbilical cords of pump twins and acardiac twins with TRAP sequence may differ from the classic pattern.



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and showed four umbilical arteries and two umbilical veins in the umbilical cord (figure 1D). The two umbilical cords of the twins were fused together but were separated from each other 2 cm before insertion into the placenta (figure 1E). We do not know if there was an arterial–arterial anastomosis between the two circulatory systems in the placenta; however, in the acardiac twin, reversal of blood flow of the two umbilical arteries was seen at an ultrasound performed at the 35th gestational week (figure 1F), which indicated an extremely rare case of epigastric heteropagus conjoined twins combined with TRAP sequence.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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