

Epigastric heteropagus conjoined twins combined with TRAP sequence

Zhang Y, Wu J, Zhang D, Mao J
Peking University Shenzhen Hospital, Shenzhen, China

Objective

We report an extremely rare case of epigastric heteropagus combined with TRAP sequence.

Methods

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 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. 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 to 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 cesarean section was performed. A newborn male with an omphalocele and a parasite on his abdomen, weighing 2900 g in total, was delivered. The Apgar score was 9' at 1 minute and 10' at 5 minutes. A single, normal-appearing umbilical cord was attached from the autosite to the only placenta. 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. 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.

Results

The newborns underwent separation surgery two 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. Where did the umbilical blood vessels come from? Three days later, the placental histopathological examination results were released and showed four umbilical arteries and two umbilical veins in the umbilical cord. The two umbilical cords of the twins were fused together but were separated from each other 2 cm before insertion into the placenta.

Conclusion

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, which indicated an extremely rare case of epigastric heteropagus conjoined twins combined with TRAP sequence. (Note: This case was published by BMJ Case Reports with the citation: Zhang Y, Wu J, Zhang D, Mao J. Epigastric heteropagus conjoined twins combined with TRAP sequence. *BMJ Case Rep.* 2023 Jan 30;16(1): e253159. doi: 10.1136/bcr-2022-253159.).

