Thoraco omphalopagus: Conjoined twins

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Section: Paediatric radiology
Area of Interest: Foetal imaging
Procedure: Education
Procedure: Intrauterine diagnosis
Imaging Technique: MR
Imaging Technique: Ultrasound
Special Focus: Foetus Obstetrics Case Type: Clinical Cases
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Patient: 27 years, female

Clinical History:

A 27-year-old multigravida with history of seven months of amenorrhoea with no previous imaging was referred to our department for antenatal ultrasound examination. There is past history of non-consanguineous marriage and previous death of first male child.

Imaging Findings:

Antenatal ultrasonography (Fig. 1) revealed monochorionic monoamniotic (MCMA) twin pregnancy corresponding to 27 weeks of gestation in breech presentation.

Ultrasound showed conjoined twins, fused face to face from upper thorax to abdomen with common sternum, diaphragm and abdominal wall.

The twins shared a common pericardial sac with fused cardia, fused liver and common intestinal loops with two heads, four extremities each, two stomach bubbles, two spines, separate pelvises and urinary tracts. There was single placenta, of anterior fundal insertion with single umbilical cord containing five vessels.

Foetal Magnetic Resonance Imaging (MRI) (Fig. 2) confirmed the ultrasound findings with no other congenital anomalies.

Gross specimen examination (Fig. 3) after elective caesarean section at 38wks of gestation confirmed the diagnosis of thoraco-omphalopagus conjoined twins. The conjoined twins died after 2 days of birth.

Discussion:

Conjoined twinning is a random event, unrelated to hereditary factors. They are rare forms of twin gestation. The incidence is 1 in 50,000 to 100,000 live births. More frequently found among females with a ratio of 3:1. [1]
Conjoined twins are classified based on site of fusion – thoracopagus (thorax), omphalopagus (abdomen), pyopagus (sacrum), ischiopagus (pelvis), craniopagus (skull), cephalopagus (face) and rachipagus (back). [2]

The commonest form is fusion of the anterior thorax and/or abdomen, that is thoracopagus, omphalopagus and thoraco-omphalopagus, altogether constitutes 70% of conjoined twins. [3]

Conjoined twins are monoamniotic, monochorionic, monozygotic twins. The fission theory suggests incomplete splitting of the embryo at the primitive streak stage. This explains increase in the incidence of situs inversus and mirror imaging in conjoined twins. The fusion theory suggests embryos re-join at vulnerable sites after initial separation. This theory accounts for fusion of conjoined twins at various angles. [4]

Thoracopagus twins are united face to face with common sternum, diaphragm, and upper abdominal wall. 90% have a common pericardial sac with complex cardiac fusion. The liver is invariably fused. 50% have a common small intestine. [5]

Omphalopagus twins are joined at umbilicus level, involving the lower thorax. Liver fusion occurs in 80%. The pericardium may be common, but the heart is never shared. In 33% the small intestines join at the level of Meckel diverticulum. [5]

Successful separation of conjoined twins is dependent on good prenatal imaging with ultrasound and MRI. [6]

Antenatal ultrasound suspicion of conjoined twins can arise as early as 7 weeks of gestation. Diagnosis may be suspected and confirmed if two foetuses cannot be visualized separately in a single gestational sac. Bifid appearance of the foetal pole, presence of more than three umbilical cord vessels, and persistency of heads at the same level and body plane. [7]

Foetal MRI is an important adjunct to ultrasound in evaluation of complex foetal anomalies and organ positions. [8]

Post-natal management falls into three categories. Non-operative management in cases with complex cardiac union, emergency separation in cases with cardiac instability or additional structural anomalies warranting immediate surgical intervention and planned elective separation. [9]

Conjoined twins are generally incompatible with life. 65% are stillborn, 35% die within the first 24 h. Only 25% survive to an age where surgical separation can be performed. [10]

The major role of imaging is to analyse the extent to which the organs are shared to assess surgical separability. Sharing of heart or brain virtually excludes separability. [11]

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Thoraco-omphalopagus conjoined twins, Thoracopagus, Omphalopagus

**Final Diagnosis:** Thoraco-omphalopagus conjoined twins

**References:**
imaging and recent advances. J Ultrason. (71):259-266. (PMID: 29375901)
**Figure 1**

Description: Antenatal ultrasonography of the abdomen. Grey scale and colour Doppler images showing complex cardiac fusion in conjoined twins. **Origin:** Department of Radiology, Krishna Rajendra Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.
Description: Antenatal ultrasonography of the abdomen. Axial image shows fused liver in conjoined twins. Origin: Department of Radiology, Krishna Rajendra Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.

Description: Antenatal axial ultrasound images in conjoined twins. Showing two stomach bubbles and bladder. Origin: Department of Radiology, Krishna Rajendra Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.
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Description: Grey scale and colour Doppler antenatal ultrasound images. Showing single umbilical cord with five vessels. Origin: Department of Radiology, Krishna Rajendra Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.
Description: Grey scale and colour Doppler antenatal ultrasound images. Showing single umbilical cord with five vessels. Origin: Department of Radiology, Krishna Rajendra Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.
Description: Foetal MRI SSFSE sequence Coronal sections of thoracoomphalopagus conjoined twins. Fused face to face with common sternum, diaphragm and abdominal wall with fused cardia, fused liver and common intestinal loops. Origin: Department of Radiology, Krishna Rajendra Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.
Description: Foetal MRI SSFSE sequence Axial sections. Showing fused cardia and fused liver.
Origin: Department of Radiology, Krishna Rajendra Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.
Description: Foetal MRI SSFSE sequence Axial sections. Showing fused cardia and fused liver.
Origin: Department of Radiology, Krishna Rajendra Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.
Figure 3

Description: Fused face to face from upper thorax to abdomen. Origin: Department of Obstetrics and Gynaecology, Cheluvamba Hospital, Mysore Medical College and Research Institute, Mysuru, Karnataka, India 2021.