Prenatal Sonographic Diagnosis of Thoracophagus Conjoined Twins - ACase Report

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ABSTRACT: Conjoined twins are a rare and extreme form of monozygotic twinning. Approximately half are stillborn, and a smaller fraction of pairs born alive have abnormalities incompatible with life. The overall survival rate for conjoined twins is approximately 25%. In recent years pre-natal diagnosis has become possible at a very early stage of gestation and detailed ultrasonography has provided an opportunity for the management of pregnancy and planning delivery. In this paper, we present a case of thoracophagus conjoined twins and the related ultrasonographic findings.

Keywords: conjoined twins, thoracophagus, twin pregnancy, monozygotictwins.

I. INTRODUCTION:

Conjoined twins are a rare form of monozygotic twinning with incomplete cell division between 13th and 15th days after fertilisation(1). This affects 1% of the monozygotic twins with the incidence being 1:30,000 to 1:200,000(2). Of them 40-60% are still born and about 35% of live birth do not survive beyond 24 hours(4). They are classified based on the site of attachment(5).

| Туре | Description | Frequency |
|-------------------------|---|-----------|
| Thoracopagus | Ventrally joined, thorax to upper abdomen, involves heart | 42% |
| Parapagus dicephalus | Laterally joined; one trunk, two heads | 11.6% |
| Cephalopagus | Laterally joined; top of head to umbilicus, two faces | 5.5% |
| Omphalopagus | Ventrally joined; abdomen to lower thorax, never the heart | 5.5% |
| Parasitic | Asymmetric, fetus in fetus | 3.9% |
| Craniopagus | Skulls joined; shared meninges, not face or trunk | 3.4% |
| Parapagus diprosopus | Laterally joined; one trunk, one head, two faces | 2.9% |
| Ischiopagus | Lower abdomen and pelvic bones joined | 1.8% |
| Rachipagus | Dorsally fused, dorsolumbar vertebrae area | 1.0% |
| Pygopagus | Dorsally fused, perineal and sacrococcygeal area | 1.0% |
| Unspecified | Rare types | 21.4% |

Data from Mutchinick OM, Luna-Munoz L, Amar E, et al: Conjoined twins: a worldwide collaborative epidemiological study of the International Clearinghouse for Birth Defects Surveillance and Research. Am J Med Genet 157:274-287, 2011.

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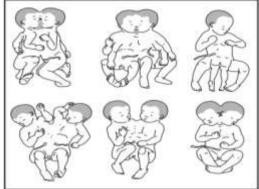


FIG 7-33 Different types of conjoined twins. Left, Examples of limited fusions in which heads and limbs are separate and retain their identity: A to C, craniopagus; D to G, thoracopagus; H and I, pyopagus. Right, Examples of more extensive fusion. (From Patten BM: Human Embryology, 4th ed. New York, McGraw Hill, 1976, Figs. VIII-5, VIII-6, pp 128-129.)

The location and extent of fusion between the twins determine the potential for surgical separation and postnatal survival.

II. **CASE REPORT:**

An 18 year old primi with 20+2 weeks gestational age was referred from the Department of Obstetrics and Gynaecology for a routine 2nd trimester ultrasonography. The following findings were noted:

- 1. Twin intrauterine gestation with evidence of fusion of the ventral thoracic and abdominal walls above the level of umbilicus.
- Fully developed heads facing each other with

- the gestational age according to Bi-parietal diameter being 19 weeks 2days.(Fig.1 and 2)
- Fused thoracic cavities ventrally with a single cardia seen in midline giving rise to 2 separate aortas, each supplying the corresponding twin. Cardiac chambers couldn't be well demarcated into atrium and ventricles, hence a diagnosis of partially fused heart was made. (Fig. 3)
- 4. Fused abdominal cavities ventrally upto the level of umbilicus with separate stomach bubbles visualised.(Fig. 4)
- 5. Abdominal cavities below umbilicus were separate with separate pelvisesand lower limbs.



Figure 1



Figure 2



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Figure 3

After ultrasound examination, the findings were properly charted, informed to the family and further referred back to the Department of Obstetrics and Gynaecology for further management. There the patient was provided with the management options available and she chose to terminate the pregnancy. The abortus was a conjoined thoracophagus conjoined twin, consistent with the sonographic findings.(Fig.5)



Figure 5

DISCUSSION:

The overall survival rate of conjoined twins is 5-25%. For better management and treatment planning early prenatal diagnosis is essential. Once the diagnosis of conjoined twins is made, the organs shared and vascular connections should be looked for. In thoracopagus the twins are positioned face-to-face, and very commonly sternum, diaphragm, upper abdominal wall and around 75% the heart are conjoined. Almost always there is an atrial joint and a mutual pericardium. Twins with omphalopagus are conjoined in the



Figure 4

umbilical region, often containing the lower thorax. Liver is around 80% mutual and this sharing is usually not equally balanced. In cases where duodenum is shared biliary anomalies are frequent. Heart is almost always separate, but a pericardial adhesion may be present. Congenital heart disease will be seen in 30% of the cases. In our report foetuses were found to share the heart and liver. Cardiac defects, congenital diaphragmatic hernia, neural tube defect, cystic hygroma, renal dysplasia, club foot, intestinal atresia are frequently seen in conjoined twins.(6) After diagnosis an accurate counselling to family is needed to define the possible therapeutic options including termination, selective foeticide if associated with multiple gestation and postpartum surgical separation of twins.

CONCLUSION: IV.

The present case highlights morphological features of an antenatally diagnosed thoracophagus conjoined twins. Although the gestational age at diagnosis in this cannot be truly termed early, picking up the findings as early as possible is very important so as to provide a wide range of therapeutic options and time to the family to make a well informed decision. Hence all cases of twin pregnancy should be thoroughly scanned especially when the relative position of foetuses remains unchanged. Although the diagnosis of conjoined twins is pretty straightforward, the presence of shared organs and vascular connections should be looked for in case the family opts for postpartum surgicalseparation.

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