

## THORACO-OMPHALOPAGUS CONJOINED TWINS ASSOCIATED WITH OMPHALOCELE AND AN UMBILICAL CORD CYST

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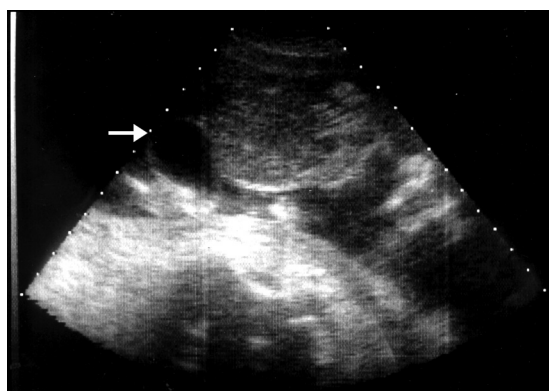
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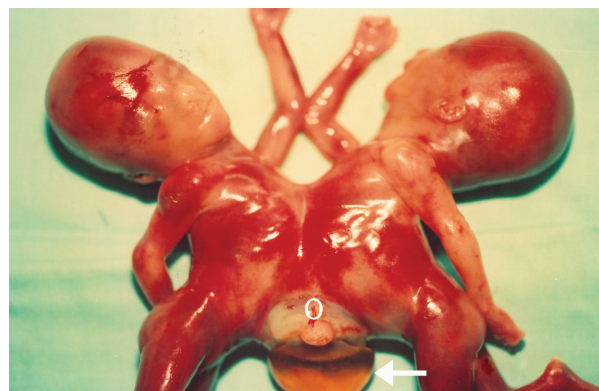
A 27-year-old primigravid woman was referred for sonographic examination at 19 gestational weeks because of suspected fetal abnormalities. The parents involved in this pregnancy were unrelated, and there was no history of diabetes mellitus, twinning or congenital malformations. The mother denied any exposure to drugs for ovulation prior to conception, and to teratogenic medications, irradiation or infectious diseases during the pregnancy. Sonographic examination performed at 19 gestational weeks revealed conjoined twins. The amniotic fluid volume was normal. The fetal biometry of both twins was consistent with the menstrual age. The twins were united face-to-face from the upper thorax down to the umbilicus, with associated omphalocele at the umbilical cord insertion. There was a 3 × 4 cm umbilical cord cyst around the omphalocele (Figure 1). The twins shared a common heart, liver

and the anterior chest wall. The parents opted to terminate the pregnancy. The thoraco-omphalopagus conjoined twins, weighing 460 g, were delivered. Chromosome analysis revealed a karyotype of 46,XX. Autopsy confirmed the diagnosis of thoraco-omphalopagus. The twin fetuses shared a common sternum, heart, spleen, liver, diaphragm, pericardium and upper abdominal wall, with associated omphalocele and an umbilical cord cyst (Figures 2 and 3).

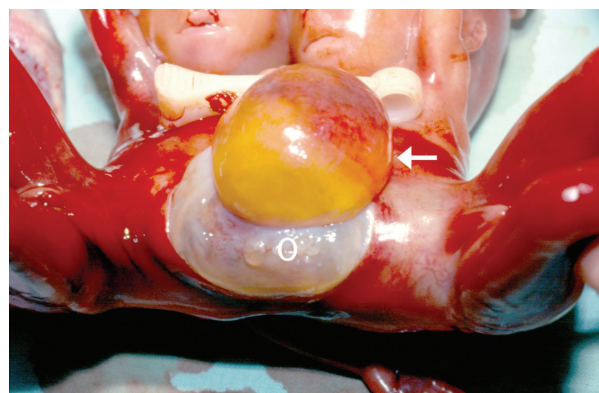
Conjoined twinning is a rare congenital malformation that occurs in 1 in every 100 sets of monozygotic



**Figure 1.** Prenatal ultrasound at 19 gestational weeks shows a 3 × 4 cm umbilical cord cyst (arrow) around the omphalocele and the fetal abdomen.



**Figure 2.** Thoraco-omphalopagus conjoined twins associated with omphalocele (O) and an umbilical cord cyst (arrow).



**Figure 3.** Omphalocele (O) and an umbilical cord cyst (arrow).

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twins [1] and 1 in 50,000–100,000 births [2]. Conjoined twinning occurs most likely at the stage of primitive streak formation in the embryonic plate (day 15–17) [1].

Conjoined twins have been classified into eight types: cephalopagus, thoracopagus, omphalopagus, ischiopagus, parapagus, craniopagus, pygopagus, and rachipagus [3,4]. Thoracopagus is the most common variety and, with omphalopagus, represents about 75% of the cases [5]. In cephalopagus, the twins are fused from the top of the head down to the umbilicus. In thoracopagus, the twins lie face-to-face from the upper thorax down to the umbilicus. The thoracopagus conjoined twins share a common sternum, diaphragm and the upper abdominal wall, and about 75% have conjoined hearts. In omphalopagus, the twins are united face-to-face and are joined in the area of the umbilicus, sometimes involving the lower thorax but always preserving two distinct hearts. The omphalopagus conjoined twins are often associated with omphalocele at the level of umbilical cord insertion. In ischiopagus, the twins are united ventrally from the umbilicus down to a conjoined pelvis with two sacrums and two symphyses pubis. In parapagus, the twins are united laterally and share a conjoined pelvis with one symphysis pubis and one or two sacrums. In craniopagus, the twins are united on any part of the skull except the face and the foramen magnum. The craniopagus conjoined twins share the cranial bones, the meninges and, occasionally, the brain surface. In pygopagus, the twins are united dorsally at the sacrococcygeal and perineal regions and may share the spinal cord. In rachipagus, the twins are joined dorsally above the sacrum, involving different segments of the vertebral column or even the occiput. In the present case, the twin fetuses shared a common sternum, heart, diaphragm, upper abdominal wall, liver and pericardium, with associated omphalocele and an umbilical cord cyst.

Tongsong et al reported the prenatal delineation of the complex cardiac anatomy in thoraco-omphalopagus twins using fetal echocardiography [6]. Basgul et al reported the first-trimester diagnosis of thoraco-omphalopagus conjoined twins by transvaginal ultrasound and color Doppler [7]. This report demonstrated the perinatal findings of an umbilical cord cyst around the omphalocele in thoraco-omphalopagus conjoined twins.

## References

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