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Case Report

Early prenatal diagnosis of conjoined twins: a case report

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ABSTRACT

Conjoined twins are a very rare complication of monochorionic monoamniotic pregnancies, associated with severe mortality and morbidity. Ultrasonography has made early diagnosis of conjoined twins possible, allowing prompt management and counselling of parents. The diagnosis should be suspected when monozygotic fetuses are shown to consistently hold the same fixed position relative to each other. The location and extent of fusion between the twins determine the potential for surgical separation and postnatal survival. The authors report a case of conjoined thoraco-omphalopagus twins, diagnosed by ultrasonography at 10 weeks of gestation. Due to high complexity of fetal fusion, termination of pregnancy was performed upon patient's request.

Keywords: Conjoined twins, Prenatal diagnosis, Thoraco-omphalopagus, Ultrasonography

INTRODUCTION

Conjoined twins represent one of the rarest forms of twin gestation, estimated to occur in 1.5 per 100,000 births worldwide.¹⁻⁴ In about 1% of monochorionic pregnancies, the twins are conjoined, probably resulting from relatively late attempts at cleavage on day 13 or later.¹⁻⁵ Therefore, when there is a case of monochorionic monoamniotic pregnancy, the possibility of conjoined twins should always be kept in mind.

Ultrasonography plays the most important role in the diagnosis. In fact, prenatal ultrasound diagnosis has been described since 1974, and lately most of the cases are diagnosed early in pregnancy.⁶ The earliest reported case of thoracopagus twins in literature was in the 7th week of pregnancy.⁴ Conjoined twins are classified based on the site of fusion. The most frequent type is thoracopagus, but mixed types are also possible.

Even though the degree and the location of fusion and the shared vital organs determine the prognosis of conjoined

twins, they are associated with a high perinatal mortality rate, with an overall survival rate of 25 %.² Early prenatal diagnosis and assessment is crucial to counsel parents for their informed decision, so that early termination can be performed, or to plan for prenatal and perinatal care if pregnancy continues.³ Authors present a case of a thoraco-omphalopagus twins diagnosed at 10 weeks of gestation, highlighting the nature of this condition, the ultrasonographic findings and the management of these cases.

CASE REPORT

A 33-year-old woman, gravida 2 para 1, was referred to our fetal medicine unit at 10 weeks of gestation for suspected conjoined twins after a routine ultrasound examination. Her past medical history was unremarkable, there was no family history of twins and the actual pregnancy was a spontaneous conception. Present ultrasound (US) revealed a monochorionic monoamniotic twin pregnancy with two fetuses in a fixed face-to-face position.

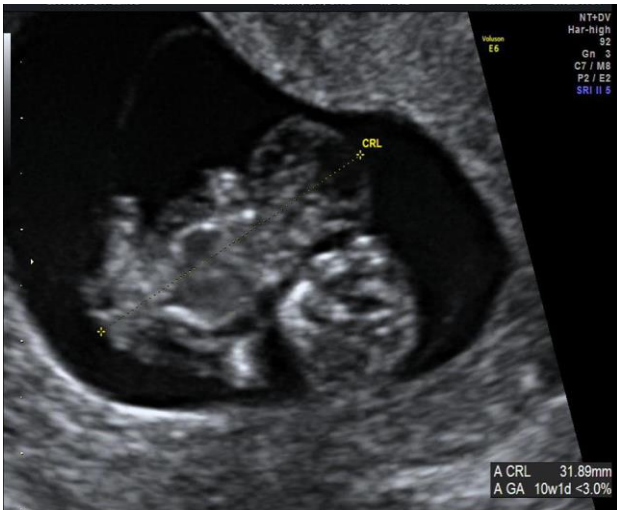


Figure 1: Ultrasound image of thoraco-omphalopagus twins at 10-week gestation.

Two heads were visualized, and two upper and two lower limbs were seen for each fetus. There was no change in the relative positions of the fetuses despite attempts. The twins were found to be joined at the thorax and upper abdomen, and only one fetal heart seemed to be observed. Both fetuses had cystic hygroma (Figure 1 and 2).

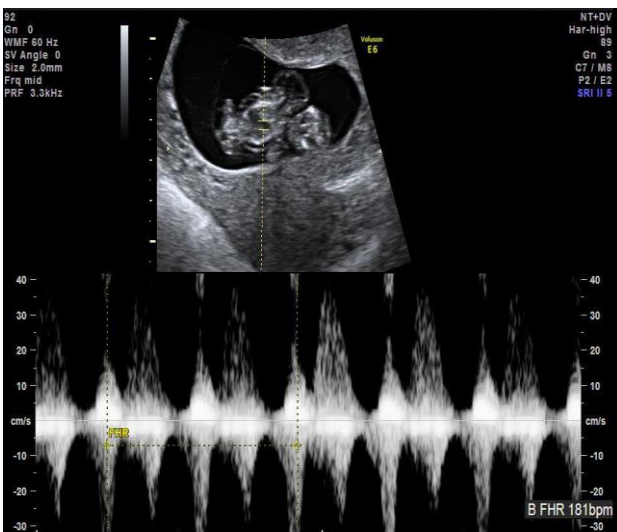


Figure 2: Ultrasound showing a single fetal heart rate.

On the base of these findings, the diagnosis of thoraco-omphalopagus twins was made. The couple was informed about US findings and counseled on the management options. They opted for termination of pregnancy, which was performed medically within one week. Autopsy confirm the diagnosis: two fetuses fused from the upper thorax to umbilicus. Two hearts were present, but they shared the stomach, liver, pancreas and bowel. Fetus 1 had esophageal atresia and absent right lung and fetus 2 had a complete atrioventricular canal defect. Gonads were not identified (Figure 3). Analysis showed a 46, XX karyotype.

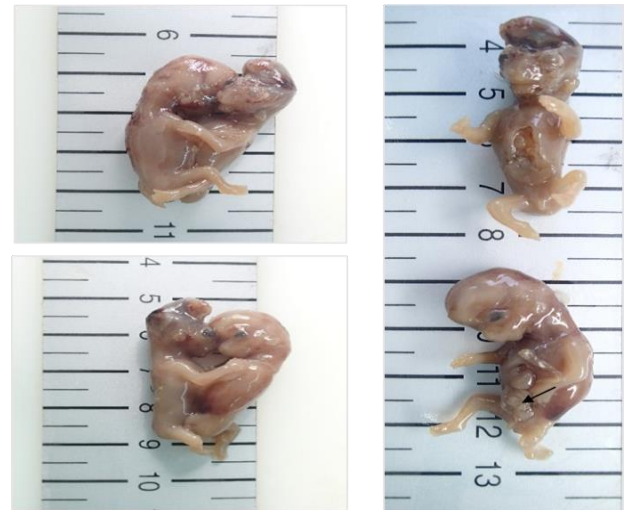


Figure 3: Gross features of thoraco-omphalopagus conjoined twins; notochordal axis as far apart as ventro-ventral, mid-torso, with shared intestinal tract (arrow); two spines and two heads, four upper limbs and four lower limbs and two pelvises.

DISCUSSION

Conjoined twins are a rare type of monozygotic twins, estimated to occur in 1.5 per 100,000 births worldwide.¹⁻⁴ Etiology is unknown, but there are two different theories that explain the formation of conjoined twins. According to the fission theory, 13-15 days after fertilization, the embryonic disc undergoes an incomplete separation, whereas in the “fusion theory” two separate monozygotic embryonic discs undergo a secondary association.^{1,4}

Female fetuses are more commonly affected with male to female ratio of 1:3, particularly in thoracopagus type.^{5,6} Based on the anatomical site of union, conjoined twins are classified as craniopagus (skull), thoracopagus (thorax), omphalopagus (abdomen) ischiopagus (ischia), rachipagus (vertebral column), pygopagus (sacrum) and parapagus (torso).⁷ The most frequent type of all conjoined twins is thoracopagus (40%) and the rarest is craniopagus.⁷ Mixed types are also found, like in this case, thoraco-omphalopagus, combining features of two groups, with a reported incidence of 28%.⁵

Early diagnosis by US is possible in modern day obstetrics.³ Suspicious US findings that may suggest the diagnosis include: both fetal heads in the same plane, no change in the relative position after maternal movement and manual manipulation, fetal scoliosis, contiguous skin, unusual limb position and more than three vessels in the cord.^{3,8-10}

Associated congenital defects unrelated to the area of fusion are common, as is stillbirth. Three-dimensional US examination, echocardiography, and magnetic resonance images (MRI), may be helpful to determine associated

anomalies, clarify the anatomy and determine the extent of deformity.⁶ Detailed evaluation is crucial to counsel the parents about prognosis and to prepare for possible postnatal surgical separation.

Surgery to separate conjoined twins may range from relatively simple to extremely complex, depending on the points of fusion and the shared organs. Most cases of separation are extremely risky and life-threatening.¹ Prognosis is very poor among conjoined twins in general. In all, 40-60% are stillbirth and among the live births, almost 35% do not survive beyond 24 hours of life.⁷ The present case focuses on morphological features of an early diagnosis of thoraco-omphalopagus conjoined twins, emphasizing the importance of sonographic assessment in all twin pregnancies.

A shared heart in fetuses is particularly difficult to diagnose, especially in such an early stage of pregnancy. Either a repeat US, echocardiography or MRI could be used to overlap this limitation. Also, fetal autopsy can contribute in understanding normal and abnormal embryogenesis and anatomic classification of conjoined twins according to axis orientation, symmetry and expression.

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