

A Very Rare Form of Conjoined Twinning: Diprosopus Twins Diagnosed in the First Trimester of Pregnancy by Transvaginal Ultrasonography: Case Report

Yapışık İkizin Çok Nadir Bir Şekli: İlk Trimesterde Transvajinal Ultrasonografi ile Tanı Konulan Diprosopus İkiz

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ABSTRACT Diprosopus twinning (two faces, one head, and one body) is extremely rare with a reported frequency of 1/180 000-1/15 000 000. We reported diprosopus twins diagnosed in the first trimester in a fetal demise case of a 21-year-old woman in her first pregnancy. Ultrasonography showed an in utero ex fetus with the bifid appearance of the cranium, two faces, two upper and two lower extremities and one body. The pregnancy was terminated using misoprostol. Macroscopic appearance confirmed the diagnosis with an additional finding of open spina bifida at the lumbosacral region. Therapeutic termination before 24 weeks of pregnancy can be offered following the early diagnosis of inseparable twins with severe conjoined anomalies. Diagnosis of conjoined twins in early gestation by ultrasonography with Doppler examination and magnetic resonance imaging (MRI) allows us to give adequate counselling to families to enable them to continue with or to terminate the pregnancy.

Key Words: Twins, conjoined; ultrasonography, prenatal; pregnancy trimester, first; neural tube defects; prenatal diagnosis; prognosis

ÖZET Diprosopus ikiz (iki yüz, tek kafa ve tek gövde) son derece nadir olup, bildirilen sıklığı 1/180 000-1/15 000 000 arasındadır. Burada, 21 yaşındaki bir hastanın ilk gebeliğinde, ilk trimesterde tanı koyduğumuz bir diprosopus ikiz olgusu bildirilmektedir. Ultrasonografi, in utero ex fetus olgusunda kraniumun ikili görünümü ile birlikte iki yüz, iki üst ve iki alt ekstremit ve bir gövde olduğunu gösterdi. Gebelik misoprostol kullanılarak sonlandırıldı. Makroskopik görünüm tanıyı doğruladı; ek olarak lumbosakral bölgede açık spina bifida mevcuttu. Ciddi yapışıklık anomalileri bulunan ve ayrılamayacak durumda olan ikizlerin erken tanısı ile 24. gebelik haftasından önce terapötik sonlandırma önerilebilir. Erken gebelik döneminde Doppler ultrasonografi ve manyetik rezonans görüntüleme (MRG) ile yapışık ikizlerin erken tanısı, ailelere gebeliğin devamı veya sonlandırılması açısından yeterli danışmanlık sunmamızda bize yardımcı olur.

Anahtar Kelimeler: İkizler, bitişik; ultrasonografi, prenatal; gebelik trimesteri, birinci; nöral tüp defektleri; prenatal tanı; prognoz

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Conjoined twins are monozygotic, monochorionic and monoamniotic, and occur in every 50 000-100 000 births. Site and extent of fusion are very variable with a classification system based on the anatomical regions, which are fused.¹ The prognosis for conjoined twins is usually poor and depends on the associated anomalies and the degree of fusion. In recent years, detailed ultrasonography facilitated the prenatal di-

agnosis at a very early stage of pregnancy and provided an opportunity for the management of conjoined twin gestations.

Here we reported an exceptional type of conjoined twins (a fetus with two faces=diprosopus) diagnosed in the first-trimester with transvaginal ultrasound and discuss the prevalence, clinical presentations, prognosis and management of conjoined twins.

CASE REPORT

The sonographic examination of a 21-year-old primigravid at 12 weeks of gestation showed an in utero ex fetus with the bifid appearance of the cranial pole, two faces, two upper limb buds, and two lower limb buds. Her history was unremarkable without any family history of twins. The measurement of the fetus was in accordance with 9 5/7 weeks of gestation. Ultrasound scans (at 8 MHz, with 4-9 MHz transvaginal probe, General Electric®, A5) confirmed that the fetus had two faces with a fused head, fused neck but only one set of other structures including placenta and umbilical cord and therefore, diprosopus conjoined twins was highly suspected (Figure 1). Therapeutic abortion was performed using misoprostol 100 mg with an initial intravaginal and sublingual administration, followed by 100 mg sublingual use every two hours for a maximum of 10 doses/day. The patient aborted, with intact membranes within 24 hours. The gestational sac included a fetus with two faces, one broad neck and a skull partially separated by a groove. There were two ears, two upper and lower extremities, one trunk, and an open spina bifida at the lumbar region of the fetus. There was physiologic herniation at the umbilicus and a cystic structure on the umbilical cord of the fetus (Figure 2,3).

Autopsy could not be performed as the family did not give consent; however, macroscopic findings confirmed the prenatal sonographic diagnosis. X-ray examination was performed to see whether there was separation in the cervical spine. Unfortunately, we could not demonstrate the skeletal structure due to the insufficient ossification at that gestational age.

DISCUSSION

Conjoined twins arise from monozygotic monoamniotic pregnancies in which separation of the embryo occurs after the 13th day of conception. Spontaneous twinning occurs in 1.6% of all pregnancies, of which 0.4% are monozygotic and of all monozygotic twins only 1% are monozygotic



FIGURE 1: Two lower and two upper extremities, two faces and bifid appearance of the cranium.

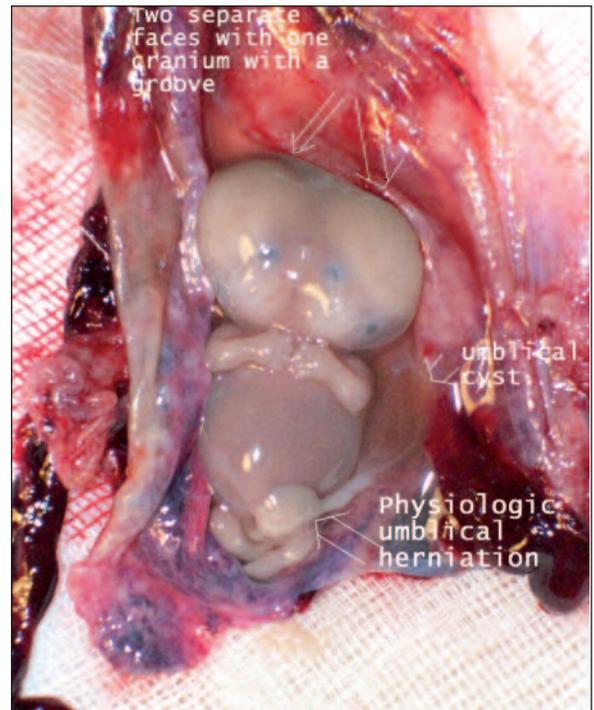


FIGURE 2: Two completely formed faces and one cranium with a groove, physiologic umbilical herniation and a cyst of the umbilical cord.

(See for colored form <http://tipbilimleri.turkiyeklinikleri.com/>)



FIGURE 3: Open neural tube defect in lumbosacral region.
(See for colored form <http://tipbilimleri.turkiyeklinikleri.com/>)

and monoamniotic.² The incidence of conjoined twins is reported to be as low as 1/58 000 births in the Caucasian population, and as high as 1/6454 births in some Asian populations. Diprosopus twinning (two faces, one head, and one body) is an extremely rare, symmetrical type of conjoined twinning, which occurs in less than 1% of cases.¹

The different types of fusion in conjoined twinning are classified into eight groups by Spencer: four types of a ventral union as cephalopagus, thoracopagus, omphalopagus and ischiopagus, the lateral union as parapagus, and the dorsal union as craniopagus, pygopagus and rachipagus.¹ However, each case has additional individual variations. The most common type of union is thoracopagus (anterior thoracoabdominal fusion) and is found in 75% of cases.¹

Diprosopus is a rare form of conjoined twins with a reported prevalence of 1/180 000-1/15 000 000, and the etiology is unknown. Partial or complete duplication of facial structures in one head with forking at the level of the cervical spine are the characteristic features. It is almost always associated with other anomalies such as neural tube defect, diaphragmatic hernia or cardiac malformations.³ Other commonly associated abnormalities include cleft lip and palate. A strong association between diprosopus and neural tube defects supports the hypothesis that conjoined twins and neural tube defects may share some pathological

mechanism secondary to a failure of the rostral neuropore to close.² Our case had an additional neural tube defect at the lumbosacral region.

Early diagnosis and precise delineation of the shared organs of conjoined twins are essential for the optimal obstetric and postnatal management. The sonographic detection of conjoined twins early in the first trimester is possible by transvaginal scan by showing a 'V-shaped' twin pregnancy which could not be separated by manipulation with the transducer.² However, if conjoined twins are extensively fused (i.e., diprosopus with double faces and single head and one body), the diagnosis would be difficult to make until later in gestation. In this case report, we emphasized the ability to diagnose diprosopus twins in the first trimester. However, a careful approach is necessary to avoid misdiagnosis. Where a definitive diagnosis in early pregnancy is uncertain, a follow-up study should be performed. Ultrasonographic findings for the diagnosis of conjoined twins are as follows: Visualization either of the two placentas or a separating membrane excludes conjoining of the twin pregnancy. The sonographic diagnosis of conjoined twins should be based on the lack of separating membrane, conjoined body parts, inseparable bodies or heads between the twins despite the changes in fetal position or a bifid appearance of the fetal pole in the first trimester. In addition, other signs suggesting conjoined twins include more than three vessels in a single umbilical cord, complex fetal structural anomalies, heads or bodies at the same level, hyperextended spine, unusual proximity of the extremities, and persistence of the position relative to one another after movement or during the follow-up scan. Search for duplication of any anatomical parts, including the brain, heart, liver, extremities and spine will confirm the correct diagnosis. The persistence of the same inseparable parts on repeated scans confirms the diagnosis.

In the recent years, 3D ultrasound is also used for prenatal diagnosis of conjoined twins.⁴ Prenatal magnetic resonance imaging (MRI) can be used as a complementary tool in the imaging work-up both for diagnosis and for correct prognostic assessment to plan postnatal surgery.⁴

The prognosis of conjoined twins depends on the degree of union. Conjoined twins commonly result with early intrauterine death, as in our case. One study from Spain showed that the frequency of conjoined twins among stillborns was 99 times higher than that observed among live births.⁵ Most of the conjoined twins are born prematurely, 40% are stillborn, and 35% survive only one day.⁵ Postnatal separation can be achieved in rare cases. Survival chances and mode of delivery depend on the degree of fusion of the organs.⁴ Therapeutic termination before 24 weeks of pregnancy can be offered following the early diagnosis of inseparable twins

with severe conjoined anomalies.⁵ However, if there are no associated anomalies, and if surgical separation is feasible as assessed by antenatal detailed ultrasonography and other methods such as MRI, then the parents may choose the continuation of pregnancy. In that case, the prognosis will be good in an experienced center with proper preparation and operation.

An appropriate imaging strategy is a fundamental part of prenatal diagnosis to allow correct prognostic assessment, counseling family about continuation or termination of pregnancy, and postnatal surgical treatment planning.

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