

A Case of Parapagus Dicephalus Conjoined Twins Diagnosed at 17th Weeks of Gestation

Yasemin Taşcı, Şadımın Altınbaş, Ömer Kandemir, Serdar Yalvaç

S.B. Etlik Zübeyde Hanım Kadın Hastalıkları Eğitim Hastanesi, Kadın Hastalıkları ve Doğum Kliniği, Ankara, TR

Abstract

Objective: Conjoined twins are a rarely seen anomaly with an incidence of 150.000-1100.000 gestations and together with severe mortality and morbidity. Early diagnosis and treatment seems to be very important in the followup.

Case: An 32 yearsold, nulliparous woman whom personal and family background were uneventful was referred to our clinic with a diagnosis of conjoined twins at 17th weeks of gestation. Ultrasonographic examination revealed a singleton fetus with measurements consistent with a gestational age of 17 weeks and the fetus with 2 heads with separate necks, 2 arms, 2 legs, only one medulla spinalis and vertebral column.

Conclusion: Conjoined twins are a rare form of twinning. Early diagnosis and management of the pregnancy regarding to social, ethical and economic problems seems to be very important.

Keywords: Conjoined twins, prenatal ultrasound.

Prenatal tanısı 17. Gebelik haftasında konan parapagus disefalus yapışık ikiz: olgu sunumu

Amaç: Yapışık ikiz, insidansı 150.000–1100.000 olan, ağır mortalite ve morbidite ile seyreden nadir bir anomalidir. Erken tanı ve tedavi gebelik takibinde önemlidir.

Olgu: 32 yaşında, nullipar, özgeçmiş ve soy geçmişinde özellik bulunmayan hasta yapışık ikiz ön tanısı ile, 17. gebelik haftasında başka bir merkezden hastanemize refere edildi. Yapılan USG incelemesinde intrauterin 17 hafta ile uyumlu, iki başı, tek gövdesi, iki kol ve iki bacağı, tek medulla spinalis ve vertebral kolonu bulunan fetus izlendi.

Sonuç: Yapışık ikiz, ikiz gebeliğin nadir görülen bir formudur. Erken gebelik döneminde tanı konulması ve gebeliğin mevcut duruma göre yönetilmesi, etik ve ekonomik sorunlar düşünüldüğünde oldukça önemli görünmektedir.

Anahtar Sözcükler: Yapışık ikiz, prenatal ultrasonografi.

Introduction

Conjoined twin is a quite rare congenital anomaly which can be seen in 1/50.000 – 1/100.000 pregnancies.¹ Considering that 60% of these fetuses die in a short time after delivery or born dead, real incidence of live birth is calculated as 1/200.000.² Though the exact etiology of conjoined twins are not know, two theo-

ries are suggested: According to the first theory, monovular embryo is divided incompletely at 13th-15th days of conception. In fusion theory, a secondary fusion occurs between two monovular embryonic discs.³ It has been argued in the literature that fusion theory can explain all conjoined twin cases; by spherical theory, it has been explained that conjoined twins are

attached to each other asymmetrically or through different body parts.^{4,5}

Classification of conjoined twins was widely accepted by the classification suggested by Spencer; according to this, ventral or dorsal fusion may occur as to fusion point of embryonic disc.⁴ Body part numbers are expressed as di- (two), tri- (three), tetra- (four) and body parts expressed by Latin words [for instance, brachius (arm), -pus (lower extremity), prosopus (face) etc.]. When different fusions are compared, conjoined twins with central fusion are seen more frequently since somite development begins from the central and proceeds towards caudal and cranial.⁶ Conjoined twins with ventral fusion cover 87% of all cases; 11% of them are cephalopagus cases, 19% of them are thoracopagus cases, 18% of them are omphalopagus cases, 11% of them are ischiopagus cases and 28% of them are parapagus cases.⁷

In the article that we present, the significance of early diagnosis and treatment for dicephalic parapagus case diagnosed at 17th gestational week will be emphasized.

Case

Thirty-two years old woman with gravida 1 who had eventless personal and family backgrounds without kin marriage was referred to our clinic by a pre-diagnosis of conjoined twins through external center at her 17th gestational week. Obstetric ultrasonographic examination (USG) performed by Voluson 730 PRO (General Electric, Healthcare, Milwaukee, WI) revealed a singleton fetus with measurements consistent with intrauterine gestational age of 17 weeks having 2 heads and 2 necks on one body, 2 arms and 2 legs, single or conjoined two medulla spinalis and vertebral column (Figure 1). It was observed in the transverse and longitudinal cross-sections that fetus had one heart, one stomach, one bladder, one placenta and umbilical cord.

The patient and her husband were informed in detail about the current status of pregnancy and that their babies would have a low chance to live when surgical separation process is performed after delivery. The case was discussed in the council of Perinatology Clinic of our hospital; and gestational termination was concluded.



Figure 1. Ultrasonographic view of conjoined twin case: the view of two heads and two necks of a case consistent with intrauterine gestational age of 17 weeks having 2 heads and 2 necks on one body, 2 arms and 2 legs, single or conjoined two medulla spinalis and vertebral column.



Figure 2. The postnatal view of terminated conjoined twin; two heads, two necks, two arms and two legs of the fetus.

ed after written consent of the patient and her husband was taken. Karyotype analysis was not planned as it is known that heredity is not seen in conjoined twins together with karyotype anomaly. In the macroscopic examination of fetus after termination, it was seen that fetus had two heads, two necks, two arms and two legs (Figure 2). One anus and one external genitalia existed on one body. Single or conjoined two vertebral colon(s) and one sacrum were observed in the x-ray film (Figure 3).

Discussion

Parapagus conjoined twin cases developed due to parallel duplication of two notochords at proximal level are a kind of ventrolateral conjoin twins; these fetuses share umbilicus, abdomen and pelvis. The fusion includes pelvis which has single or double sacrum(s) and single symphysis pubis; parapagus twin case with sacral agenesis was also reported in the literature.⁸ When thoraxes of twins are separate, they are called as dithoracic conjoined twins and when they have two heads on single body as the case that we presented, they are called as dicephalic conjoined twins. Their arm and leg numbers may vary between 2 and 4. Although all visceral organs are rarely double in parapagus cases, there are cases reported in the literature.³ In the case that we presented, there were two arms, two legs on a single body, the case was called as parapagus dicephalous dibrachius dipus; also visceral organs were displayed separately. The prognosis in conjoined twin cases depends on the scale of fusion and the decision for maintaining pregnancy should be taken into the cases which do not have other organ anomalies and on which surgical separation operations can be performed.⁹ In parapagus cases with single heart as in the presented case, this situation is a factor negatively affecting prognosis. Although each fusion type possibly may have cardiac anomaly, thoracopagus cases are at the top in terms of cardiac anomaly rate.



Figure 3. X-ray view of conjoined twin case: Single or conjoined two vertebral colon(s) and one sacrum.

Dibrachius dipus cases generally have one heart; however, the heart is duplicated or conjoined in most of the cases.¹⁰ Intracardiac pathology existence and cardiac fusion rates in conjoined twins are important due to the success for surgical procedures and their effects on long-term survival. Due to the existence of distorted heart in one of two thoracopagus cases presented in the study of Şen et al. in 2003, the case was terminated at 19th gestational week; the second thoracopagus case was diagnosed at 25th week and the pregnancy was followed up until 38th gestational week since conjoined liver with separate vascular structure and conjoined non-pericardium heart on the sides of ventricles facing each other were found and there was no accompanying anomaly, then the surgical separation operation was successfully performed at 10th month after delivery by cesarean.⁹ In another study, stillborn parapagus dicephalous conjoined twin case was presented which admitted to clinic at 38th gestational week without any follow-up and was delivered vaginally with difficulty.¹¹ This case is significant in terms of showing the significance of prenatal diagnosis and antenatal follow-up in conjoined twin cases. In the case that we present, visceral pathologies (cardiac fusion) were not confirmed by autopsy since the family did not

approve the autopsy though they were informed; however, no intracardiac pathology was seen in the prenatal echocardiography.

Prenatal diagnosis of conjoined twins is important when considered together with high mortality rate and postnatal ethic, social and economical problems. Conjoined twins can be diagnosed by transvaginal or transabdominal USG as from first trimester. If single yolk sac is followed up together with two fetuses at first trimester, or if there is the case of monaoamniotic twin, conjoined twins should be considered.³ After diagnosis, fusion existence should be checked in brain, liver, heart, extremities and spinal cord. Also revealing vascularization in conjoined vital organs is determinant in terms of the prognosis of surgical separation operations as well as revealing additional accompanying fetal anomalies. Especially three-dimensional USG can present complex fetal anatomy and case type in conjoined twin cases and may provide selective termination in this way.⁷

Conclusion

Fetal anomalies can be diagnosed at early period by ultrasonography and prenatal invasive initiatives and pregnancies with fetal anomaly can be terminated. Conjoined twin is a rare anomaly and may progress with high mortality and morbidity according to the level of organ share. Diagnosing this situation at early prenatal period would have a significant role in

advanced periods of pregnancy, informing family and terminating pregnancy when required.

References

1. Spitz L, Kiely EM. Conjoined twins. *JAMA* 2003; 289: 1307-10.
2. Spitz L, Kiely EM. Experience in the management of conjoined twins. *Br J Surg* 2002; 89: 1188-92.
3. Al RA, Altaş S, Çetinkaya K, Delibaş İ, Taştekin A, Kadanalı S. Bir disefalus parapagus birleşik ikiz olgusu ve literatürün gözden geçirilmesi. *TJOD Derg* 2010; 7: 55-60.
4. Spencer R. Theoretical and analytical embriyology of conjoined twins: part I: embryogenesis. *Clin Anat* 2000; 13: 36-53.
5. Spencer R. Theoretical and analytical embriyology of conjoined twins: part II: adjustment to union. *Clin Anat* 2000; 13: 97-120.
6. Erler A, Koyuncu F, Kurt S, Demir N, Kolday K, Turan E et al. Conjoined Twins (Case Repport) *Perinatology Journal* 1996; 4: 110-2.
7. Kökçü A, Çetinkaya MB, Aydın O, Tosun M. Conjoined twins: Historical perspective and report of a case. *J Matern Fetal Neonatal Med* 2007; 20: 349-56.
8. Hari S, Sharma R, Gupta AK, Das C, Singal AK. Magnetic resonance imaging of parapagus twins. *European Journal of Radiology Extra* 2004; 51: 25-9.
9. Şen C, Çelik E, Vural A, Kepkep K. Antenatal diagnosis and prognosis of conjoined twins-a case report. *J Perinat Med* 2003; 31: 427-30.
10. Andrews RE, McMahon CJ, Yates RWM, Cullen S, De Leval MR, Kiely EM ve ark. Echocardiographic assesment of conjoined twins. *Heart* 2006; 92: 382-7.
11. Harma M, Harma M, Mil Z, Öksüzler C. Vaginal delivery of dicephalic parapagus conjoined twins: case report and literature review. *Tohoku J Exp Med* 2005; 205: 179-85.