Fırat Tıp Derg/Firat Med J 2013; 18(3): 182-184

Case Report

www.firattipdergisi.com

Alive Borned Parapagus Dicephalus Conjoined Twins at Term: A Case Report

Tolgay Tuyan ILHAN[a1], Turkan ILHAN[2]

[1]*Ozalp State Hospital, Department of Obstetrics and Gynecology, Van, Turkey*

[2]*Van Obstetrics and Children's Hospital, Obstetrics and Gynecology, Van, Turkey*

ABSTRACT

Parapagus dicephalus conjoined twins are rare but severe complication of multigestational pregnancies. Many conjoined twin pregnancies are diagnosed before reaching viability. We present a case report of conjoined twins reached term. A 35-year-old pregnant woman, who had no prenatal follow up, delivered conjoined twins at Van Maternity Hospital. The total weight of the fetuses was 4750 g. and they could be alive without advance support. Conjoined fetuses can be detected on prenatal screening and ultrasound. This case reinforces the importance of prenatal follow up.

Key words: *Conjoined twin, Parapagus dicephalus, Prenatal screening.*

ÖZET

Miadında Canlı Doğan Parapagus Dicephalus Yapışık İkiz: Olgu Sunumu

Parapagus dicephalus yapışık ikizler çoğul gebeliklerin nadir fakat ciddi komplikasyonlarıdır. Yapışık ikizlerin birçoğuna yaşayabilir büyüklüğe erişmeden tanı konulmaktadır. Biz miadına kadar erişmiş bir vakayı sunmak istiyoruz. Otuzbeş yaşında prenatal takibi olmayan gebe Van Doğumevi' nde yapışık ikiz doğurmuştur. Fetusların toplam ağırlığı 4750 gramdır ve ciddi bir yaşam desteği sağlanmadan hayatta kalmışlardır. Yapışık ikizler prenatal tarama ve ultrasonla tespit edilebilmektedir. Bu olgu prenatal değerlendirmenin önemini desteklemektedir.

Anahtar Sözcükler: *Yapışık ikizler, Parapagus dicephalus, Prenatal tarama.*

Conjoined fetus is a rare but challenging congenital anomaly for physicians and families. This rare anomaly occurs in 1 in 50000 to 100000 pregnancies (1, 2). Many types of this anomaly are described according to side of union. Parapagus is the most common anomaly, occurring in 25% of conjoined cases (3). Routine ultrasound evaluation and other prenatal screening tests are very sensitive in the diagnosis of conjoined twins (CTs). Currently with the use of these techniques, CTs are generally diagnosed before reaching viability (4). We aim to describe a case that reached term.

CASE REPORT

A 35-year-old pregnant woman, gravida 8, parity 7, abortus 0 was evaluated in our emergency department with a chief complaint of abdominal pain. She was evaluated by a midwife first. It was noted that her cervix was dilated to seven centimeters and 80% effaced with vertex presentation and positive fetal heart beats. She had no prenatal follow up, and her previous pregnancies were uncomplicated. She did not have a family history of twinning. She was admitted to the hospital and observed by external fetal heart rate monitor. After

25 minutes of observation, the midwife detected early deep decelerations in the fetal heart rate and notified the obstetrician. Since the fetus was in acute distress and the fetal head was posterior and asynclytic with frontal bossing, the patient was taken to the operating room for cesarean delivery under general anesthesia. CTs were delivered through a low segment uterine incision. The Apgar scores at 1, 5 and 10 minutes were 8, 10, 10 respectively. Total weight of the newborns was 4750 g. The placenta was monochorionic and minimally enlarged. There was only one umbilical cord with three vessels and one umbilicus.

During first evaluation, both of newborns were active and crying. There were two heads, three upper limbs and three lower limbs. The twins could move right and left side limbs and the medial arm and foot appeared to be non-functional. The external genitalia was not clearly identical (Figure 1, 2). X-ray images revealed that there was one heart and fused lungs (Figure 3, X-ray). After initial evaluation, the patient was transferred to a multidisciplinary hospital. According to information received from the hospital,

[a] Corresponding Adress: Dr. Tolgay Tuyan ILHAN, Ozalp State Hospital, Department of Obstetrics and Gynecology, Van, Turkey
Phone: 0 532 7860591 e-mail: tolgaytuyan@yahoo.com
Received/Geliş Tarihi: 09.08.2012 Accepted/Kabul Tarihi: 31.01.2013

the twins could be feeding with nasogastric catheter. Surgery for separation was excluded because of visceral anatomy.

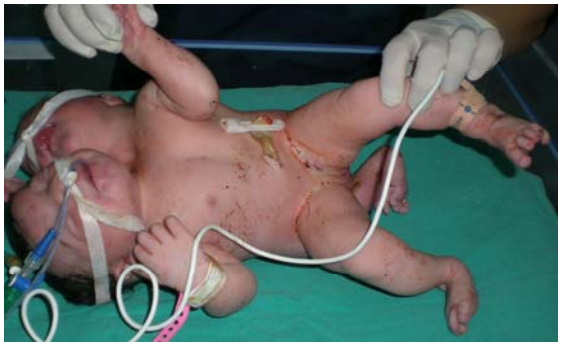

Figure 1. *Conjoined twins after delivery*

Figure 2. *Conjoined twins with one anal canal and ambigius genitalia*

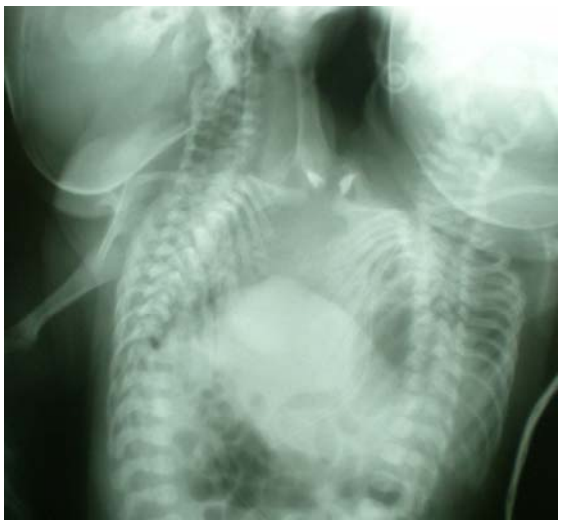

Figure 3. *X-ray image of conjoined twins with enlarged single heart*

DISCUSSION

Conjoined twinning is a severe and rare complication of multigestational pregnancies. The first scientific report was published in the 19th century (5). CTs are classified by side of union. The side of union is not only used for classification but can also be a prognostic factor. Although the etiology of conjoined pregnancy is not clear, some theories attempt to explain the pathogenesis. The most accepted etiology is incomplete division of embryos after conception (3, 6, 7). Some studies also suggest that secondary union of the embryonic disk may play a role. However, diamniotic CTs have been reported (8). The secondary fusion theory could explain diamniotic twinning (9, 10).

CTs are classified according to side of union and can be a major prognostic factor. Ventral unions (87%), which can occur rostrally (48%), laterally (28%) and caudally (11%), are the most frequent form. Dorsal unions (13%) are rare (3, 11). Depending on the severity of visceral malformation, survival of CTs is extremely low (4, 12). With the wide spread use of prenatal screening these anomalies can be detected before viability.

First trimester ultrasound screening is useful for early diagnosis of CTs. In our case, the patient did not have prenatal follow up. If two fetuses cannot be visualized clearly, conjoined twinning must be suspected. Umbilical cord anomalies, fetal motion abnormalities and increased nuchal translucency are ultrasound findings which could be helpful in the diagnosis of CTs (13-15). Polyhydramnios is seen in 50 to 76 % of cases. Pregnant women are recommended to have at least one ultrasound examination during pregnancy.

Depending on the severity of visceral anomalies, most CT pregnancies do not reachterm. Although vaginal deliveries have been reported at term, cesarean section is the preferred for prevent fetal and maternal complications (16-18).

After birth of live CTs, the most challenging period for the physicians starts. Most CTs live for minutes or hours. The fetal outcomes depend on visceral organs status. Fetal visceral anomalies can occur in many types. Immediate separation is a plausible option if appropriate resources are available. In our case, the twins were doing extremely well during the early postpartum period so the decision was made not to immediately separate them. In most parapagus twins, visceral organs can be shared with twins after thorough preoperative evaluation.

Cardiovascular anomalies are the most severe and life-threatening anomalies that CTs encounter. The type and severity of heart anomalies depends on the side of union. The reason for death is cardio-pulmonary insufficiency in most cases. Complex congenital heart disease, transposition of the great vessels, atrioventricular septal defects are the most common anomalies in dicephalus parapagus twins (19). Gastrointestinal system anomalies are another prognostic factor. Since there was no information regarding the gastrointestinal tract, the patient had an orogastric catheter placed for feeding. After insertion of catheters, x-ray images were used to confirm correct placement. X-ray images revealed that the catheters

were located in two individual stomachs (Figure 4). One patent anus was identified. The digestive systems therefore united after the stomach but the precise segment of union was not clear. These types of visceral organ sharing are the limiting factor of surgical repair.

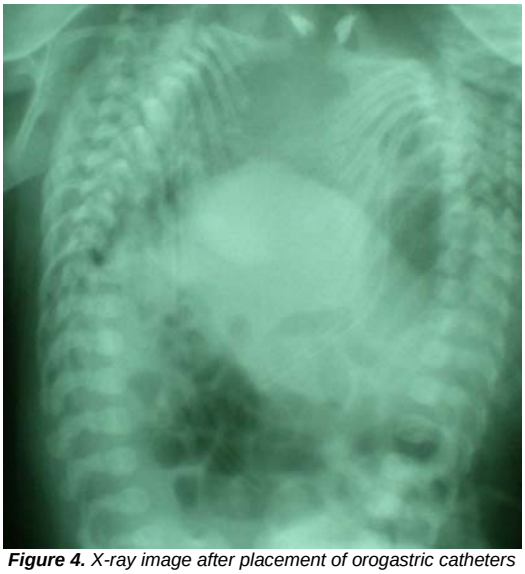

Figure 4. *X-ray image after placement of orogastric catheters*

In most parapagus twins, there is a single but shared genitourinary tract (3). Conjoined twinning is more common in the female gender, and nearly 70% of all CTs are female (20, 21). In our case, we could not clearly identify the gender.

Most CTs pregnancies do not result in live births, however viable CTs are possible if they are followed closely during the prenatal period. Management of living CTs is a complex issue for health professionals and the patient's families. Perinatal and ethical outcomes must be discussed in a multidisciplinary setting. Early diagnosis of this anomaly is the main stay for prevention. Prenatal screening tests and routine ultrasound are recommended for all pregnant women.

REFERENCES

1. Hanson JW. Incidence of conjoined twinning. Lancet 1975; 2: 1257.
2. Edmonds LD, Layde PM. Conjoined twins in the United States 1970-7. Teratology 1982; 25: 301-8.
3. Spencer R. Theoretical and analytical embryology of conjoined twins: part I: embryogenesis. Clin Anat 2000; 13: 36-53.
4. Machin GA, Keith LG, Bamforth F. An atlas of multiple pregnancy: biology and pathology. CRC Press, Parthenon Publishers, New York 1999.
5. Taruffi C. Storia della teratologica, vol. 2. Bologna: Regia Tipografia 1882; 572-8.
6. Levin M, Roberts DJ, Holmes LB, Tabin C. Laterality defects in conjoined twins. Nature 1996; 384: 321.
7. Crane JP. Sonographic evaluation of multiple pregnancy. Semin Ultrasound CT MR 1984; 5: 144.
8. Spencer R. Minimally united ischiopagus twins: infraumbilical union with cloacal anomalies. J Pediatr Surg 1996; 31: 1538-45.
9. Charles A, Dickinson JE, Watson S, Phillips N, Yovich J. Diamniotic conjoined fetuses in a triplet pregnancy: An insight in to embryonic topology. Pediatr Dev Pathol 2005; 8: 666-72.
10. Kapur RP, Jack RM, Siebert JR. Diamniotic placentation associated with omphalopagus conjoined twins: Implications for a contemporary model of conjoined twinning. Am J Med Genet 1994; 52: 188-95.
11. Spencer R. Anatomic description of conjoined twins: a plea for standardized terminology. J Pediatr Surg 1996; 31: 941-4.
12. Mackenzie TC, Crombleholme TM, Johnson MP, et al. The natural history of prenatally diagnosed conjoined twins. J Pediatr Surg 2002; 37: 303-9.
13. Van den Brand SF, Nijhuis JG, van Dongen PW. Prenatal ultrasound diagnosis of conjoined twins. Obstet Gynecol Surv 1994; 49: 656-62.
14. Sebire NJ, Souka A, Skentou H, Geerts L, Nicolaides KH. First trimester diagnosis of monoamniotic twin pregnancies. Ultrasound Obstet Gynecol 2000; 16: 223-5.
15. Pajkrt E, Jauniaux E. First-trimester diagnosis of conjoined twins. Prenat Diagn 2005; 25: 820-6.
16. Agarwal U, Dahiya P, Khosla A. Vaginal birth of conjoined thoracopagus- a rare event. Arch Gynecol Obstet 2003; 269: 66-77.
17. Creinin M. Conjoined twins. In: multiple pregnancies: Epidemiology, gestation and perinatal outcome. LG Keith, E Papiernik, DM Keith & B Luke (Editors). Parthenon, New York, 1995; 93-112.
18. Harman M. Vaginal delivery of dicephalic parapagus conjoined twins: Case report and literature review. Tohoku J Exp Med 2005; 205: 179-85.
19. Gilbert-Barness E, Debich-Spicer D, Opitz JM. Conjoined twins: Morphogenesis of the heart and a review. Am J Med Genet A 2003; 120: 568-82.
20. Wedberg R, Kaplan C, Leopold G, et al. Cephalothoracopagus (Janiceps) twinning. Obstet Gynecol 1979; 54: 392-6.
21. Turgut F, Turgut M, Basaloglu H, Basaloglu HK, Haberal A. Extremely rare type of conjoined twins: Cephalothoracopagus deradelphus. Eur J Obstet Gynecol Reprod Biol 1998; 80: 191-4.