

Conjoined twins: three cases in one tertiary medical center and literature review

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Summary

Conjoined twins are so rare that most obstetricians will not be personally exposed to such cases during their professional lifetimes. The authors report three cases including one of dicephalic parapagus conjoined twins and two thoracopagus conjoined twins in the present tertiary medical center, and discuss the diagnosis and management details. They also review the incidence, embryological, diagnostic, obstetrical, and prognostic aspects of conjoined twins. Regular antenatal visits and serial ultrasound scanning are crucial for early diagnosis of conjoined twins, optimal obstetric management and perinatal preparation still remain challenging, and multidisciplinary cooperation is urgently needed.

Key words: Conjoined twins; Dicephalic parapagus; Thoracopagus; Prenatal diagnosis.

Introduction

Conjoined twins are very rare and associated with high perinatal mortality. It occurs once in every 50,000 to 100,000 births [1]. Accurate prenatal diagnosis of conjoined twins is possible in early pregnancy if the patients have regular antenatal visits and serial ultrasound scanning. However, a few of conjoined twins were detected during the third trimester or just before labor, and this makes obstetric management more complicated. The authors report a case of dicephalic parapagus conjoined twins and another two cases of thoracopagus conjoined twins.

Case Report

Case 1: Dicephalic parapagus

A 19-year-old unbooked primigravida, presented for prenatal care at 37 weeks' gestation. Unlike other mothers, she had had no previous antenatal medical evaluation. Ultrasound examination during the first prenatal visit revealed conjoined twins. The abdominal ultrasonography showed a breech fetus with two heads, three arms, single thorax, two hearts (one of which with complete type endocardial cushion defect), single abdomen, single liver, double vertebra, and two legs. The amount of amniotic fluid was normal. According to these ultrasound findings, dicephalic parapagus conjoined twins was diagnosed. In view of the fact that the pregnancy was already full term, and given the poor prognosis, the decision was made to proceed with delivery of the babies via cesarean section. The authors performed cesarean section for the patient, because in this situation vaginal delivery is impossible for the full-term conjoined twins in breech presentation. An experienced obstetrician accomplished the operation and she chose the lower uterine segment incision. The whole process was completed without complication.

The Apgar scores of the neonates at one, five, and ten minutes were 5, 5, and 3. Physical examination revealed the twins' birth weight was 3,600 grams. The neonate had two heads, three upper limbs (one of which with two palms), and three lower limbs (one of which without foot) with fusion of the thoracic, abdominal, and pelvic regions. There was a single anal opening and male external genitalia. The placenta was monochorionic and weighed 1,100 grams. The umbilical cord was centrally located and included two vessels (one artery and one vein). The appearance was consistent with dicephalic parapagus (Figure 1).

The conjoined twins died 50 minutes following delivery. The parents declined an autopsy.

Case 2: Thoracopagus

A 30-year-old multigravid woman (G3P2) registered for antenatal care at 24 weeks' gestation. She had two healthy children, and denied the intake of any medication during pregnancy. Routine antenatal laboratory investigations were all normal. However, the routine screening of ultrasound revealed conjoined twins. The twins, facing each other, were joined at the thoracic level. Each had a separate set of structures except for a shared heart and anterior chest wall. A thoracopagus conjoined twin pregnancy was diagnosed.

Since the fetuses were thought previable and having poor prognosis, the parents opted for immediate termination of pregnancy. Ethacridine lactate was administered to the patient by amniocentesis. Two days later, the conjoined twins were delivered, which weighed 1,270 grams. The patient had no any severe genital tract laceration. It was seen that the twins were united from the upper thorax down to the umbilicus with the presence of cheilopalatognathus. The upper and the lower limbs with normal morphology were in appropriate locations. They had male external genitalia. The placenta was monochorionic, weighing 260 grams. There was a single umbilical cord with three vessels (two arteries and one vein) (Figure 1). No autopsy was done due to the objection of the family.

Case 3: Thoracopagus

A 24-year-old, G2P0 female presented to the department of obstetrics at 35 weeks' gestation. There was no history of exposure to teratogenic agents in pregnancy. She had never consulted any obstetrician before applying to the present department. Routine

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screening ultrasound revealed conjoined twins. The two fetuses with a single heart were fused at chest and abdomen. Two separate spines were seen. Amniotic fluid was normal and there was a single placenta. All confirmed the diagnosis of thoracopagus conjoined twins. Since premature rupture of membrane occurred, cesarean section was carried out to deliver the twins. The doctor chose lower uterine segment incision. The operation went very well.

The two female infants both had Apgar scores of 5, 7, and 9 at one, five, and ten minutes, and had a combined weight of 3,450 grams. The pediatrician intubated the twins and examined them carefully. They were fused from upper thorax down to the umbilicus with the presence of obvious gastroschisis. Each baby had respective arms and legs in the right positions, while only one heartbeat was palpable. There was a single placenta with two separate umbilical cords (one artery and one vein respectively), which coincided at a distance of ten centimeters from placenta (Figure 1).

The conjoined twins died six hours after delivery since the parents refused any advanced medical assistance. Autopsy was declined by the family.

Discussion

Incidence

Conjoined twins are monoamniotic, monochorionic, and monozygotic twins. They occur once in every 50,000-100,000 births [1], although it is speculated that many may result in a spontaneous abortion. Many authors reported a higher incidence of conjoined twinning in females, which is approaching 75-90% [2].

Etiopathogenesis

The exact etiology of conjoined twins is unknown. Zeng *et al.* reported that genetic or environmental factors other than abnormal X-inactivation must be involved in causing conjoined twins [3]. Steinman reported that a significant number of women with conjoined twins pregnancy were subjected to environmental triggers, such as preconceptual weight, use of oral contraceptives, and so forth [4].

There are two important theories proposed to account for the origin of conjoined twins. One involves the incomplete fission of a single embryonic disc, which occurs 13 to 15 days after the ovum is fertilized. The other is that two embryonic discs unusually unite. The latter theory is postulated on the fact that the ovum undergoes fission before the notochord develops, and there are separated notochords in most conjoined twins [5].

Classification

Conjoined twins are classified on the basis of site of union. Ventral unions account for 59% and are classified as: cephalopagus (11%), thoracopagus (19%), omphalopagus (18%), and ischiopagus (11%). Lateral unions, also described as parapagus, account for 28%. Dorsal unions occur in 13% of conjoined twins and are classified as: craniopagus (5%), rachiopagus (2%), and pygopagus (6%) (Table 1) [6].

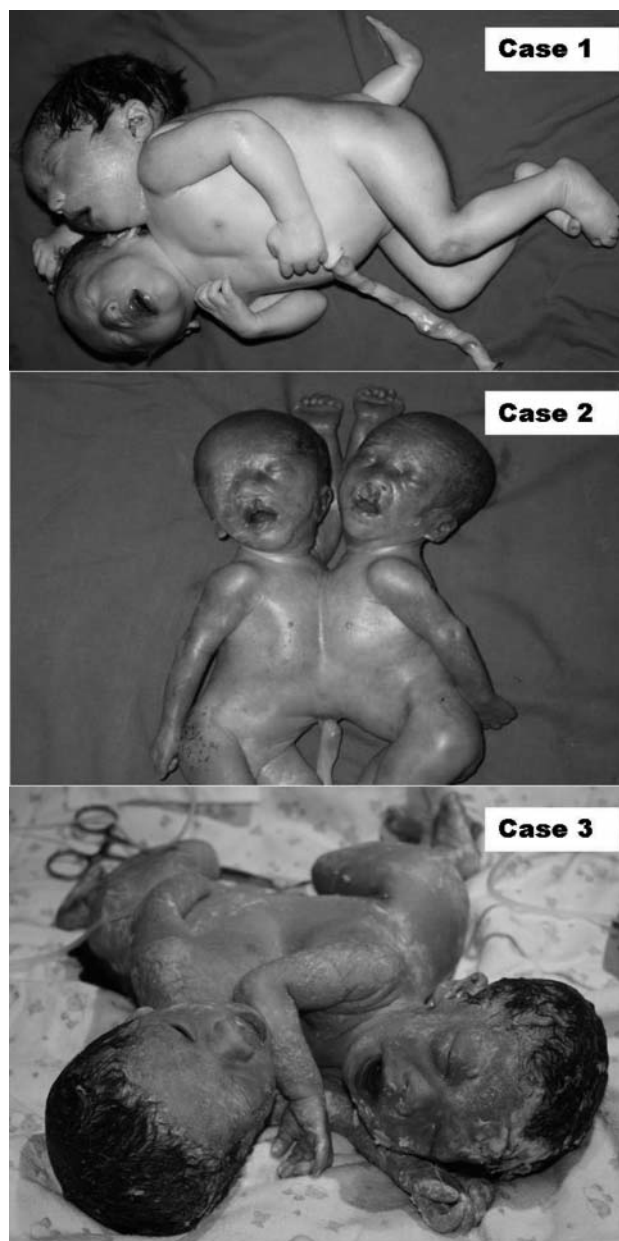


Figure 1. — Conjoined twins (Case 1 – Case 3).

Diagnosis

All of these three cases were revealed by ultrasound, but unfortunately the diagnoses were not made during the first trimester for some family and social factors. The diagnosis of conjoined twins by ultrasound was first reported by Wilson *et al.* in 1976 [7]. Serial ultrasound scan plays a crucial role in diagnosis of conjoined twins as a non-invasive approach. Conjoined twins can be diagnosed during the first trimester by both transabdominal and transvaginal sonography. High-resolution sonography makes diagnosis possible as early as the seventh week of gestation. Criteria for ultrasonographic diagnosis of conjoined twins include inseparable fetal bodies, lack of sep-

Table 1. — *Classification of conjoined twins.*

Classification	Characteristics	Separability
Ventral union		
Cephalopagus	Fused from top of head to umbilicus	None
Thoracopagus	Fused face-to-face from upper thorax to umbilicus, shared heart or interatrial vessel	Rare
Omphalopagus	Fused face-to-face from lower thorax to umbilicus, without shared heart or interatrial vessel	82% success
Ischiopagus	Fused end-to-end from umbilicus to a large conjoined pelvis, shared external genitalia, and anus	63% success
Lateral Union		
Parapagus		
• Dithoracic parapagus	Fused side-by-side, shared umbilicus,	Separate thoraxes and faces
• Dicephalic parapagus	abdomen, and pelvis	Separate heads with fused thoraxes
• Diprosopic parapagus		Two faces on the same side of single head
Dorsal Union		
Craniopagus	Fused on any portion of the skull except the face and foramen magnum, sharing bony cranium and meninges	None
Pygopagus	Fused dorsally, shared sacrococcygeal and perineal regions, even the spinal cord, sharing one anus with two rectums	Not reported
Rachipagus	Fused dorsally above the sacrum, may involve occiput and segments of vertebral column	68% success

arating amniotic membrane, constant relative positions, presence of fetal anomalies, and the identifications of more than three vessels in a single umbilical cord [8]. Polyhydramnios occurs in 50-76% of cases. Detailed scanning around 20 weeks' gestation will demonstrate the extent of the conjoined area and provide an assessment of prognosis [9].

In the present cases, because all the families had chosen delivery proceeding or pregnancy termination, further diagnostic intervention like magnetic resonance imaging (MRI) has not been considered. MRI is an important adjunct to ultrasound, particularly in the evaluation of complex fetal anomalies. With its ability to differentiate soft tissues, MRI can provide excellent detail, increased tissue contrast, and reproducible fetal anatomy [10]. Prenatal MRI has also recently been found to be of value in planning for an extrauterine intrapartum procedure and immediate separation [11].

Delivery

Once the antenatal diagnosis of conjoined twins is made, the mode of delivery must be considered thoroughly to avoid obstructed labor and morbidity for mother and fetus. If the twins are considered to have poor survival chances, termination of pregnancy should be offered prior to viability. Beyond viability, pregnancy termination may or may not be possible, depending on the laws of each country. Vaginal delivery is recommended when the twins are dead or pre-term, but cesarean section may be required if the twins are large [12]. Although there are many reported successful vaginal deliveries of conjoined twins, selective cesarean section is the preferred method of delivery with near-term-sized twins, even if the fetuses are dead [13]. To avoid maternal trauma, craniotomy, decapitation, evisceration or amputation may be needed in some circumstances.

Prognosis

Approximately 40% of conjoined twins are stillborn and 60% liveborn, only about 25% of those that survived can live long enough to be considered for surgical separation [14]. Survival of conjoined twins depends largely on the site of fusion and the organs involved. Surgical separation is one of the options, which may be successful if organs critical for life are not shared. The surgery may also cause death of one twin to allow independence alive for the other. However, no surgical separation should be considered in the presence of complex cardiac fusion or where there would be a severe unacceptable deformity following separation (Table 1) [9].

Once the decision to proceed with the pregnancy is made, delivery should be planned to take place at or close to the surgical unit where separation will be performed. Surgical separation of conjoined twins involves communication, coordination, and multidisciplinary planning by a specialized team. On the other hand, the postpartum psychosocial counseling for the parents should be provided [15].

The parents in first and third cases declined any resuscitative efforts for the conjoined twins, as the prenatal assessments were insufficient. The babies were believed to be unlikely to survive.

Conclusion

The authors have reported three cases of conjoined twins with pictures in the present tertiary medical center, which are very rare and valuable. Early diagnosis is critical for optimal obstetric and perinatal management. The delivery option is determined by the complexity of union and the size of twins. The prognosis depends on the extent of organ sharing, and surgical separation may lead to an independent life for twins.

References

- [1] Rees A.E.J., Vujanic G.M., Williams W.M.: "Epidemic of conjoined twins in Cardiff". *Br. J. Obstet. Gynaecol.*, 1993, 100, 388.
- [2] Ogutu D., Anastasakis E., Chi C., Kadir R.A.: "First trimester diagnosis of conjoint (pygopagus) twins: A case report of successful prenatal and postnatal management". *J. Obstet. Gynaecol.*, 2008, 28, 340.
- [3] Zeng S.M., Yankowitz J., Murray J.C.: "Conjoined twins in a monozygotic triplet pregnancy: prenatal diagnosis and X-inactivation". *Teratology*, 2002, 66, 278.
- [4] Steinman G.: "Mechanisms of twinning. V. Conjoined twins, stem cells and the calcium model". *J. Reprod. Med.*, 2002, 47, 313.
- [5] Spencer R.: "Theoretical and analytical embryology of conjoined twins: Part 1: Embryogenesis". *Clin. Anat.*, 2000, 13, 36.
- [6] Spencer R.: "Anatomic Description of conjoined twins: A plea for standardized terminology". *J. Pediatr. Surg.*, 1996, 31, 941.
- [7] Wilson R.L., Cetrulo C.L., Shaub M.S.: "The prepartum diagnosis of conjoined twins by the use of diagnostic ultrasound". *Am. J. Obstet. Gynecol.*, 1976, 126, 737.
- [8] Neilson J.P.: "Prenatal diagnosis in multiple pregnancies". *Cur. Opin. Obstet. Gynaecol.*, 1992, 4, 280.
- [9] Spitz L.: "Conjoined twins". *Prenat. Diagn.*, 2005, 25, 814.
- [10] Spielmann A.L., Freed K.S., Spritzer C.E.: "MRI of conjoined twins illustrating advances in fetal imaging". *J. Comput. Assist. Tomogr.*, 2001, 25, 88.
- [11] Mackenzie T.C., Crombleholme T.M., Johnson M.P., Schnaufer L., Flake A.W., Hedrick H.L. *et al.*: "The natural history of prenatally diagnosed conjoined twins". *J. Pediatr. Surg.*, 2002, 37, 303.
- [12] Agarwal U., Dahiya P., Khosla A.: "Vaginal birth of conjoined thoracopagus—a rare event". *Arch. Gynecol. Obstet.*, 2003, 269, 66.
- [13] Kokcu A., Cetinkaya M.B., Aydin O., Tosun M.: "Conjoined twins: historical perspective and report of a case". *J. Matern. Fetal. Neonatal. Med.*, 2007, 20, 349.
- [14] Kaufman M.H.: "The embryology of conjoined twins". *Childs. Nerv. Syst.*, 2004, 20, 508.
- [15] Athanasiadis A., Mikos T., Zafrakas M., Diamanti V., Papouli M., Assimakopoulos E. *et al.*: "Prenatal management and postnatal separation of omphalopagus and craniopagus conjoined twins". *Gynecol. Obstet. Invest.*, 2007, 64, 40.

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