Conjoined twins: Report of a case and review of the literature

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Conjoined twins, known to the layperson as Siamese twins, are a rare and catastrophic obstetric event. With the approximate incidence being 1 in 50,000 births, few physicians practicing obstetrics will have the unfortunate occasion to manage this biologic anomaly. Should this event occur, early diagnosis, close prenatal management, and the proper route of delivery will assure the best possible outcome for mother and both babies. The author describes the anatomic types of conjoined twins, the prenatal diagnosis and management, the route of delivery, and the overall statistical expectation. Under the best of circumstances, a good outcome is rarely achieved, but the obstetric goal should be a healthy mother and two salvageable babies.

Conjoined twins are the product of a single ovum (monozygotic) and thus have the same chromosomal composition and sex.¹ The etiology of conjoined twins remains obscure, but several possible causes have been suggested. One of these is that at a critical period of susceptibility, development is retarded and an abnormal budding or fission of the embryonic disc occurs.² Gore and colleagues³ stated that conjoined twins result from an arrest in the division of the inner cell mass that occurs in monozygotic twins before the third week of gestation. Another postulation is that the aging ovum is the major cause⁴; still another, that changes in oxygen concentration and temperature affect the metabolism of the embryo and upset the normal early developmental sequence, resulting in the formation of a monster or double monsters.⁵ Also, one could suspect that the incidence of conjoined twinning is related to the seasons, but in a study of 43 sets of conjoined twins, Bender⁶ found no significant seasonal differences.

The birth of conjoined twins is an extremely rare phenomenon.⁵ In this paper, I report one case of conjoined twins and describe the intrapartum di-

agnosis and management. From a review of the literature, I describe the morphopathologic types of conjoined twins and discuss concurrent congenital malformations, female sex bias, and incidence of stillbirths as well as the recommended modalities of antepartum diagnosis and the optimum type of delivery.

Report of case

A 25-year-old woman (gravida 2, para 1) with a twin gestation was admitted to the hospital for bed rest and observation. Two prenatal ultrasound examinations had confirmed twins at 38 weeks' gestation in the breech-breech presentation. The patient's cervix was 90% effaced and dilated 2 to 3 cm; and the presenting part was at a -2 station.

The patient's medical history included a tonsillectomy and adenoidectomy in childhood and the uncomplicated vaginal delivery of her first pregnancy 10 years previously. During the current pregnancy, she had taken iron and vitamins. She smoked six to seven cigarettes a day. She denied the use of alcohol, and she had no medical illnesses or known allergies. The family history was non-contributory.

Findings of the physical examination were normal. The fundal height was 43 cm; two distinct fetal heart tones were present, and the cervix was effaced and dilating.

Laboratory data indicated a hematocrit level of 39.4%; the blood type was O positive; the serology was nonreactive; and rubella immunity was confirmed. The results of the SMA-6 and urinalysis were normal; and the urine culture was negative for bacteria and glucose.

On the third hospital day, the patient had spontaneous rupture of the membranes, followed by uterine contractions. Because of the breech-breech presentation, a cesarean delivery was elected. A primary low transverse cervical cesarean section through a low midline abdominal incision was performed with the patient under general anesthesia. At delivery, a diagnosis was made of conjoined male twins of the thoracopagus morphopathologic type, with multiple anomalies (Fig 1). No heart rate was present at delivery, and resuscitation efforts by the pediatricians were not successful. The Apgar scores were 1 at 1 minute and 0 at 5 minutes. The surgical procedure was without complication, and the patient's postoperative hospital course was uncomplicated. The postoperative hematocrit level was 37%. The patient was

discharged from the hospital on the fourth postoperative day, and thereafter she was seen for follow-up by social work services.

The postmortem examination showed postmature white male thoracopagus-type twins, with fusion of sternum, heart, liver, duodenum, jejunum, and umbilical cord (Figs 1 and 2). Autopsy findings are listed in the Table.

Discussion

The birth of conjoined twins is a highly unusual obstetric event: Its incidence is generally considered 1 per 50,000 births.^{2,4} However, the rate has been reported to be from as high as 1 in 20,000 births⁷ to as low as 1 in 200,000.⁶ For the years of 1976 to 1987, Kaplan and Eidelman⁵ reported an incidence in Israel of 1 per 47,500 deliveries. In a New York study,⁸ the frequency was found to be 1 set of conjoined twins per 52,000 births, similar to the classically quoted figure.^{2,4} In their study, Edmonds and Layde⁸ found an incidence of 1 set per 100,000 births.

Conjoined twins are classified as follows, according to the anatomic region of union:^{2,3}

- *Craniopagus*: joined at some homologous portion of the cranial vault.
- *Syncephalus*: joined at the face.
- *Thoracopagus*: sharing part of the thoracic wall.
- *Thoracomonomphalus* (*thoracoomphalopagus*): having a common ophatocele in addition to sharing part of the thoracic wall.
- *Xiphopagus* or *monomphalus* (*omphalopagus*): joined from the umbilicus to the xiphoid cartilage.
- *Rachiopagus*: joined at the vertebral column at any point above the sacrum.
- *Pygopagus*: joined at the lateral and posterior surfaces of the coccyx and sacrum (almost back-to-back).
- *Ischiopagus*: joined at the inferior margins of the coccyx and sacrum, with the two completely separate spinal columns lying in the same axis.

The most common types of conjoined twins are thoracomonomphalus (28%), thoracopagus (18%), omphalopagus (10%), and craniopagus (6%).^{8,9} Padilla and colleagues¹ reported that approximately 75% of all reported cases are thoracopagus.

Studies indicate that conjoined twins are predominantly girls, tend to be premature with low birth weights, and in the majority of cases are stillborn.^{1-3,5,6,8-10}

Many authors have reported the higher incidence of girls in conjoined twinning: Padilla and colleagues¹ noted that girls are more common than boys. Harper and colleagues² reported that the

twins are girls in 69% of the cases; and Edmonds and Layde⁸ in 75%. Gore and colleagues³ reported that girls account for as many as 75% to 90% of the cases of conjoined twins.

In a review of cases, Harper and colleagues² reported an average gestational age of 37 weeks, with a range of 32 to 40 weeks, an average combined birth weight of 4340 g, and a stillbirth rate of 19%. Edmonds and Layne⁸ reported that 39.5% of all conjoined twins are stillborn and that an additional 34.6% die within 24 hours after birth.

Maternal complications associated with conjoined-twin pregnancies include polyhydramnios (11% to 75%), dystocia (36%), preeclampsia, and premature labor.²

A review of the literature shows no common etiology for conjoined twins. In no study has the mother's use of alcohol, tobacco, or drugs; exposure to radiation; or a family history of genetic diseases been implicated. Although an aging ovum has been implicated in conjoined twinning,⁴ Bender⁶ found the parents of conjoined twins to be younger than those in the control group: 91% of the mothers and 76% of the fathers were under the age of 35. Edmonds and Layde⁸ reported that the incidence of conjoined twinning is not affected by maternal age or race. Because all the conjoined twins in their study were born in the colder months, Kaplan and Eidelman⁵ suggested a possible seasonal effect. However, no seasonal variations are found in the data reported by Bender.⁶

Conjoined twins of the thoracopagus (and thoracomonomphalus) type are the most common and the most carefully studied.¹ Neonatal anomalies that Gore and colleagues³ associated with this type include a common pericardium (90%), joined hearts (75%), a common alimentary tract (50%), and a common liver (100%). Harper and colleagues² reported conjoined sternal cartilage (56%), conjoined diaphragm (17%), common liver (81%), and gastrointestinal anomalies (33%). Padilla and colleagues¹ reported on a female thoracopagus with conjoined hearts and livers and other multisystem anomalies similar to those seen in the case reported here (Fig 1). Similar anomalies were also found in two of the cases reported by Kaplan and Eidelman⁵; in all of the cases, the twins died.

Austin and colleagues¹¹ described male thoracopagus-type twins with a large omphalocele, imperforate anus in twin A, shared jejunum and ileum, and a conjoined liver. The case was very similar to the one reported here, with the exception that the twins survived.

Figure 1. *Postmortem radiograph of thoracopagus (thoracomonomphalus) twins.*

The antepartum diagnosis of conjoined twins is difficult but extremely important for the maximization of infant viability and reduction of maternal morbidity.³ Only two (6%) of the 36 sets of xiphopagus-type twins reported by Harper and colleagues² were diagnosed before birth. If the sternal fusion is not extensive and the absence of sternal ossification permits rotation and positional change, the diagnosis is not always possible with plain roentgenograms.^{2,11} In the case I report, two prenatal ultrasound examinations did not diagnose the thoracomonomphalus-type twins (Fig 1).

Austin and colleagues¹¹ reported that a success-

ful antepartum diagnosis of thoracopagus-type twins with a common omphalocele was made in the eighth month of pregnancy by means of ultrasonography. Gore and colleagues³ reported a similar case of sonographic antepartum diagnosis of thoracomonomphalus-type twins at 33 weeks' gestation; and Alkalay and colleagues⁹ reported one at 30 weeks. Sabbagha¹² discussed the use of ultrasound in the diagnosis of conjoined twins in his report on the ultrasonic evaluation of fetal congenital anomalies.

Sanders and colleagues¹³ reported the usefulness of echocardiography in the prenatal evaluation of

Figure 2. Conjoined twins with common omphalocele (thoracomonophalus).

thoracoabdominally conjoined twins. They stated that conjoined twins can be readily diagnosed by ultrasound examination, but that the extent of the cardiac conjunction is correctly determined by echocardiography. Four sets of thoracoabdominally conjoined twins were correctly diagnosed. When surgical separation is being considered, a postnatal evaluation including echocardiography and angiography is required.¹³

On the basis of a case load of three sets of conjoined twins, Sakala¹⁰ has outlined six steps that should take place in the ideal obstetric management of conjoined twins:

- Rule out conjoining whenever twins are diagnosed.
- Establish extent of conjoining.
- Determine postnatal viability.

- Determine route of delivery.
- Meet parental psychosocial needs.
- Provide postpartum counseling.

If the twins are at term, elective cesarean section is the preferred method of delivery.³ This view is supported by Sakala,¹⁰ who reported cesarean section as the delivery route of choice in most cases in which conjoined twins are diagnosed antenatally. A maneuver for the vaginal delivery of intact thoracopagus-type twins has been reported. In a 1964 report, Fine and colleagues¹⁴ described the intrapartum diagnosis of thoracopagus twins after the vaginal delivery of the head of twin A. After 20 minutes and demise of the twins, the operator delivered the legs, hips, and body of twin A by hyperextending the back. The second twin was then

Table
Congenital Anomalies Found at Autopsy of
Thoracopagus-Type Twins

Anatomic System	
Twin A	Twin B
Cardiovascular	
1. Single umbilical artery	1. Same as twin A
2. Persistent left superior vena cava forming larger coronary sinus in continuity with right atrium of twin B	2. Same as twin A
3. Common atrioventricular valve	3. Same as twin A
4. Anomalous left pulmonary vein to persistent left superior vena cava	4. ...
5. Common pericardium	5. Same as twin A
	6. Absent inferior vena cava
	7. Truncus arteriosus
Gastrointestinal	
1. Fusion of distal duodenum with that of twin B	1. Same as twin A
2. Fusion of jejunum with that of twin B	2. Same as twin A
3. Imperforate anus	3. ...
4. Common liver with twin B	4. Same as twin A
5. Common omphalocele	5. Same as twin A
Genitourinary	
1. Cystic renal dysplasia, bilateral, with left ureteral atresia	1. ...
2. Urethral agenesis	2. ...
Reticuloendothelial	
Accessory spleens, two	...
Respiratory	
...	Bilobed right lung
Endocrine	
...	Unilobed thyroid
Musculoskeletal	
Constriction band, left thumb	Absent right radius constriction band, right thumb
Cytogenetic	
No karyotypic abnormalities (normal male 46, XY)	Same as twin A

delivered as a double-footling breech extraction. Although this method may be suitable for the delivery of small nonviable thoracopagus-type twins, in this day and age, cesarean delivery should be considered the delivery route of preference.

Summary

Reported here is a rare case of thoracopagus-type twins with a common omphalocele (thoracomonophalus)—the most common and best-studied type of conjoined twins. Also discussed are the diagnosis, obstetric management, and autopsy findings. In a review of the literature, I outline the morphopathologic types of conjoined twins, the current methods of diagnosis and management, and the preferred route of delivery.

1. Padilla MM, Chin AJ, Padilla TMM: Cardiovascular abnormalities in thoracopagus twins. *Teratology* 1981;23:101-113.
2. Harper RG, Kenigsberg K, Sia CG, et al: Xiphopagus conjoined twins: A 300 year review of the obstetric, morphopathologic, neonatal, and surgical parameters. *Am J Obstet Gynecol* 1980;137:617-629.
3. Gore RM, Filly RA, Parker JT: Sonographic antepartum diagnosis of conjoined twins. *JAMA* 1982;247:3351-3353.
4. Potter EL: *Pathology of the Fetus and Infant*. Chicago, Year Book Medical Publishers Inc, 1961, p 218.
5. Kaplan M, Eidelman AI: Clustering of conjoined twins in Jerusalem, Israel: An epidemiologic survey. *Am J Obstet Gynecol* 1983;145:636-637.
6. Bender C: Studies on symmetrically conjoined twins; letter to the editor. *J Pediatr* 1967;70:1010-1011.
7. Hanson JW: Incidence of conjoined twinning. *Lancet* 1975;2:1257.
8. Edmonds LD, Layde PM: Conjoined twins in the United States, 1970-1977. *Teratology* 1982;25:301-308.
9. Alkalay AL, Gonzalez CL, Chou PJ, et al: Radiological case of the month. Thoracomonophalus twins with big omphalocele. *Am J Dis Child* 1987;141:89-91.
10. Sakala EP: Obstetric management of conjoined twins. *Obstet Gynecol* 1986;67(suppl to no. 3):21S-25S.
11. Austin E, Schiffrin BS, Pomerance JJ, et al: The antepartum diagnosis of conjoined twins. *J Pediatr Surg* 1980;15:332-334.
12. Sabbagha RE: Ultrasonic evaluation of fetal congenital anomalies. *Clin Obstet Gynecol* 1980;7:103-119.
13. Sanders SP, Chin AJ, Parness IA, et al: Prenatal diagnosis of congenital heart defects in thoracoabdominally conjoined twins. *N Engl J Med* 1985;313:370-374.
14. Fine E, Lewis PL, English TJ: Maneuver for vaginal delivery of intact conjoined twins. *Obstet Gynecol* 1964;24:554-557.

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