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Case Report

Vaginal Delivery of Dicephalic Parapagus Conjoined Twins: Case Report and Literature Review

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HARMA, M., HARMA, M., MIL, Z. and OKSUZLER, C. *Vaginal Delivery of Dicephalic Parapagus Conjoined Twins: Case Report and Literature Review.* Tohoku J. Exp. Med., 2005, **205** (2), 179-185 —— After an unsuccessful midwife-assisted delivery in which a head was born but delivery could not be advanced, episiotomy performed at Sanliurfa Maternity Hospital allowed vaginal delivery of female conjoined twins. Visual and x-ray examination showed two heads, two vertebral columns, two feet, two arms, and fusion at the level of the pelvis. The baby was born dead, but the mother made an uneventful recovery. Parapagus (anterolaterally joined) dicephalus (two-headed) twins account for only 11-13% of all conjoined twins, and they rarely survive. Complex malformations of hearts, lungs and abdominal organs, duplication of the tracheae, upper gastrointestinal tract and spinal column, and either double or single versions of other organs have been reported in parapagus dicephalus cases. The incidence, anatomical, embryological, diagnostic, prognostic, obstetrical, perinatal, and ethical aspects of conjoined twins are reviewed, with a focus on parapagus dicephalus conjoined twins. ———— conjoined twins; dicephalus; parapagus; vaginal delivery

Conjoined twins of the parapagus (anterolaterally fused) or dicephalus (two-headed) type are extremely rare (Castilla et al. 1988; Machin 1993; Yang et al. 1994; Groner et al. 1996; Bondeson 2001; Tansel and Yazicioglu 2004). Vaginal delivery of conjoined twins is also uncommon. We report a vaginal birth of parapagus dicephalus conjoined twins, unsuspected before delivery.

CASE REPORT

A 34-year-old pregnant woman, gravida 5, para 4, aborta 0, went into active labor at approximately 37 to 38 weeks of gestation. Her previous pregnancies had resulted in 4 live births at term. She had no personal or family history of twins. Like most mothers in her village, she had no prenatal medical evaluation. She was attended at home by a lay midwife for the delivery. The baby's head appeared to have been born normally, but the midwife was not able to advance the delivery further. Therefore, the woman was urgently admitted to Sanliurfa Maternity Hospital, two hours away.

At the time of presentation to the hospital, the baby's head was visible and was cyanotic. A mediolateral episiotomy, approximately 10 cm in

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length, was performed. With gentle traction, the remainder of the baby's body was delivered. The neonate was found on examination to have two heads, two arms, two feet, and one pelvis. The Apgar scores at 1, 5, and 10 minutes were 0, 0, and 0 respectively, and the weight was 3,500 g. The uterus was examined bimanually for rupture, and the genital tract was inspected for injury, with no detectable problems.

The placenta was monochorionic and weighed 550 g. The umbilical cord was centrally located and included three vessels. The family did not allow an autopsy, but photographs and x-ray films were taken. The radiographic examination showed two heads, two vertebral columns, and fusion of the lower half at the level of the pelvis (Fig. 1).

The mother had an uneventful postpartum course and was discharged on the second day after delivery.

Discussion

Incidence

Conjoined twins (CTs) are always derived from one fertilized ovum, and are monochorionic (shared placenta) and monoamniotic (shared amniotic sac) (Quiroz et al. 1989; Hammond et al. 1991). They occur once in every 2,800 to 250,000 births, with incidence usually cited as 1 per 50,000 to 100,000 deliveries (Fitzgerald et al. 1985; Quiroz et al. 1989; Barth et al. 1990; Hammond et al. 1991; Itoh et al. 1993; Shija et al. 1994; Creinin 1995; Bianchi et al. 2000; Bondeson 2001; Daskalakis et al. 2004; Tansel and Yazicioglu 2004). They may be more common in parts of Asia and Africa (Shija et al. 1994).

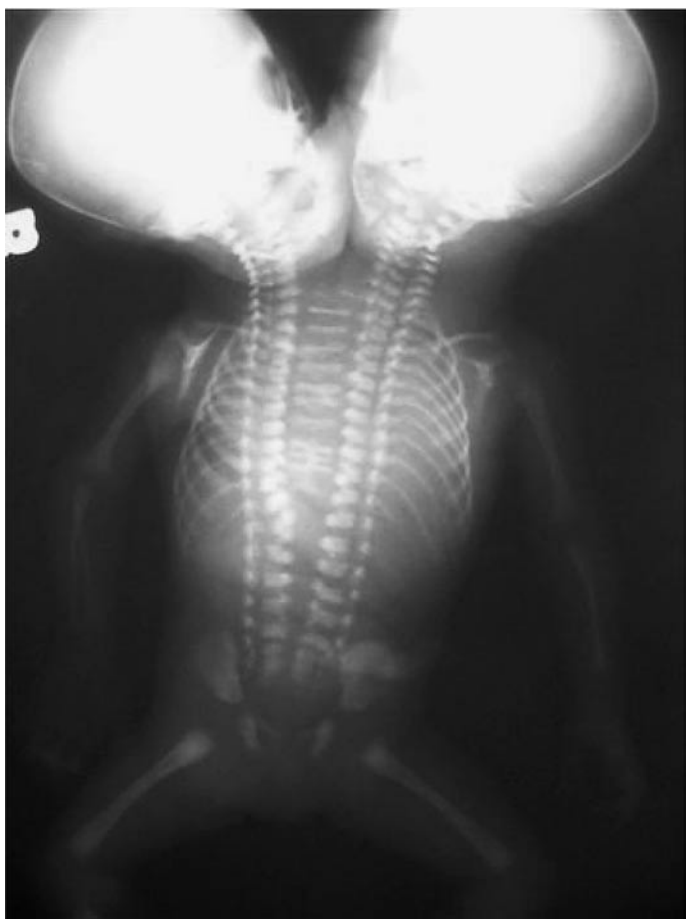

Fig. 1. x-ray of dicephalic parapagus twins. Note two heads, two vertebral columns, two arms, two feet, and fusion of the lower body at the level of the pelvis.

Incidence is 1:192,000 in southern Africa, 1:33,000 to 1:165,000 in North America, 1:100,000 in Japan, 1:75,000 in Sweden and Latin America, 1:68,000 in Hungary, 1:14,000 in Africa, and 1:6,500 in Taiwan (Castilla et al. 1988; Yang et al. 1994; Bianchi et al. 2000; Gilbert-Barness et al. 2003). Increased prenatal detection of CTs with ultrasound may explain an apparent rise in occurrence (Mackenzie et al. 2002). Most authors report that 70-95% of conjoined twins are female (Apuzzio et al. 1984; Barth et al. 1990; Gilbert-Barness et al. 2003), although one study cited a nearly equal male: female ratio (Castilla et al. 1988).

Spencer proposed a parapagus classification for anterolaterally conjoined twins, e.g. dicephalus (two-head) and diprosopus (two-face) cases (Spencer 1992). Dicephalus and diprosopus twins comprised 11.2% of conjoined twins in Machin's review, and 13% in the Latin American Collaborative Study (Castilla et al. 1988; Machin 1993). The authors' Medline search revealed only four references to human parapagus twins, and a search on "dicephalus conjoined twins" yielded only 48 human citations since 1959. Itoh noted fewer than 80 cases before 1987 (Itoh et al. 1993). Of note, one dicephalus set was associated with the 1986 Chernobyl nuclear accident (Hammond et al. 1991).

Anatomical aspects of parapagus dicephalus twins

Although autopsy was not permitted in our case, other dicephalus twins have had characteristic findings. Past studies describe fused hearts with complex anatomy, including right aortic arch and reversal or transposition of great vessels (Cunniff et al. 1988; Hammond et al. 1991; Spencer 1992; Itoh et al. 1993; Gilbert-Barness et al. 2003; Tansel and Yazicioglu 2004). When two hearts are present, the right twin's heart is usually more severely malformed (Spencer 1992). Defects of laterality, with absent spleen or situs inversus of the right twin's abdominal organs, often accompany heart abnormalities (Cunniff et al. 1988). If the hearts are not fused, abdominal viscera are situated normally (Cunniff et al. 1988). There are generally two sets of lungs (Spencer 1992), which may be underdeveloped or anomalous, especially in the right twin (Cunniff et al. 1988; Hammond et al. 1991; Yang et al. 1994; Gilbert-Barness et al. 2003). The liver, pancreas, gallbladder, genitourinary tracts, and rectum may be shared (Cunniff et al. 1988; Spencer 1992; Yang et al. 1994; Groner et al. 1996; Gilbert-Barness et al. 2003). Neural tube defects, cystic hygroma, clubfoot, and imperforate anus have occurred in parapagus twins (Itoh et al. 1993; Machin 1993; Bondeson 2001; Mackenzie et al. 2002).

Embryological theories of conjoined twinning

Two theories have been proposed to explain conjoined twinning. (Spencer 1992; Itoh et al. 1993; Machin 1993). The first asserts that incomplete fission of a single embryonic disc occurs 13 to 15 days after the ovum is fertilized (Quiroz et al. 1989; Barth et al. 1990; Hammond et al. 1991; Machin 1993; Creinin 1995; Sen et al. 2003; Tansel and Yazicioglu 2004). Spencer (1992) proposed a second theory: that a fertilized ovum divides completely into two embryonic discs whose unusual proximity results in secondary fusion into CTs as the embryos enlarge. A similar theory is that CTs arise from ectopic primitive streaks. (Gilbert-Barness et al. 2003).

Most CTs face each other (Barth et al. 1990; Machin 1993; Gilbert-Barness et al. 2003). In contrast, dicephalus twins' axes are side-by-side and nearly parallel (Spencer 1992; Machin 1993; Gilbert-Barness et al. 2003). Complex anomalies of the heart and abdominal laterality are seen more often in dicephalus and thoracopagus than in other types of CTs, and might result from disturbed cross-signaling between tissues in adjacent primitive streaks, as suggested in animal models (Gilbert-Barness et al. 2003). Cunniff et al. (1988) hypothesized that rotation of the heart at the atrial level initiates lateralization in the embryo, and that interference with heart rotation causes abnormalities in some right dicephalus twins.

Diagnostic challenges

Where access to technology is limited, as in

our case, antepartum diagnosis of CTs may be nearly impossible (Apuzzio et al. 1984; Grover et al. 1990; Agarwal et al. 2003). Prenatal identification of conjoined twins was rare before ultrasonography was available (Fitzgerald et al. 1985; Barth et al. 1990). Sometimes twins are unsuspected at delivery of CTs (Grover et al. 1990; Groner et al. 1996; Agarwal et al. 2003), while auscultation, palpation, and even ultrasound suggest separate twins at other times (Klug et al. 1978; Fitzgerald et al. 1985; Sakala 1986).

Diagnosis of CTs before birth allows practitioners to minimize injury by planning a suitable delivery (Fitzgerald et al. 1985; Sakala 1986; Quiroz et al. 1989; Barth et al. 1990; Hammond et al. 1991). Therefore, careful ultrasound examination is recommended for all suspected twins (Fitzgerald et al. 1985; Quiroz et al. 1989; Barth et al. 1990; Hammond et al. 1991). First or second trimester detection of CTs enables obstetricians to counsel parents about potential termination, or about delivery and treatment options if pregnancy is continued (Apuzzio et al. 1984; Fitzgerald et al. 1985; Quiroz et al. 1989; Barth et al. 1990; Yang et al. 1994; Mackenzie et al. 2002; Sen et al. 2003; Daskalakis et al. 2004).

Criteria for ultrasonographic diagnosis of CTs include absence of a separating amniotic membrane, inseparable fetal bodies, lack of change in relative positions of bodies and fetal heads on repeated examinations (Fitzgerald et al. 1985; Barth et al. 1990), heads at the same level and body plane, unusual proximity and/or hyperextension of spines, (Apuzzio et al. 1984; Fitzgerald et al. 1985; Daskalakis et al. 2004), unusual proximity of limbs, bifid appearance of the first-trimester fetal pole, complex anomalies, and more than three umbilical vessels (Daskalakis et al. 2004). Polyhydramnios occurs in 50-76% of cases. (Mackenzie et al. 2002; Daskalakis et al. 2004). False-positive and false-negative diagnoses have occurred due to rotation about a tissue bridge or misdiagnosing severe conjunction as a single fetus (Sakala 1986; Barth et al. 1990; Bianchi et al. 2000). Three-vessel cords have been noted in dicephalus twins (Hammond et al. 1991; Gilbert-Barness et al. 2003).

Three-dimensional ultrasound, magnetic resonance imaging, echocardiography, or computed tomography before birth, and angiography, cardiac catheterization, radionuclide scanning, cystography or urethrography, and gastrointestinal contrast studies after birth, may clarify the degree of conjoining, the potential for separation, and the ideal obstetrical and perinatal management (Quiroz et al. 1989; Creinin 1995; Bianchi et al. 2000; Mackenzie et al. 2002; Sen et al. 2003; Daskalakis et al. 2004). Fetal echocardiography may miss transposition of great vessels and abnormal atria or pulmonary vascular connections, but the good buffer for imaging provided by amniotic fluid may permit better visualization than postnatal echocardiography (Barth et al. 1990; Hammond et al. 1991; Bianchi et al. 2000; Mackenzie et al. 2002).

Prognosis

In general, few conjoined twins survive, due to heart, lung, abdominal, and neurological malformations often present even in unshared structures (Sakala 1986; Barth et al. 1990; Itoh et al. 1993; Groner et al. 1996; Bianchi et al. 2000; Mackenzie et al. 2002; Gilbert-Barness et al. 2003). Roughly 40% of CTs are stillborn, and 35% die in the first 24 hours of life (Sakala 1986; Barth et al. 1990). Moreover, only 60% of surgically treated CTs survive (Daskalakis et al. 2004). A retrospective tertiary center review found only 5 survivors out of 14 pairs of CTs, a survival rate of 18% (Mackenzie et al. 2002). Stillbirth and mortality rate are extremely high in dicephalus twins (Hammond et al. 1991; Yang et al. 1994; Groner et al. 1996; Bondeson 2001; Mackenzie et al. 2002). Groner et al. (1996)'s dibrachius (two-arm) dicephalus twins had a remarkable 11-day survival. Rare three- and four-arm dicephalus twins live to adulthood (Bondeson 2001).

Delivery

There are many reported vaginal deliveries of CTs (Pennings et al. 1982; Hoogeboom et al. 1983; Sakala 1986; Itoh et al. 1993; Shija et al. 1994; Creinin 1995; Agarwal et al. 2003). Although compressible fetal tissues may facilitate

vaginal birth (Agarwal et al. 2003), dystocia, uterine rupture, or maternal soft tissue injury can occur (Sakala 1986; Creinin 1995). When CTs are identified before birth, Cesarean section is preferred, to avoid maternal trauma and to facilitate treatment of viable neonates (Fitzgerald et al. 1985; Creinin 1995; Bianchi et al. 2000). However, vaginal delivery may be attempted with small, very premature, or nonviable fetuses, and with pregnancy termination before 24 weeks of gestation (Sakala 1986; Barth et al. 1995; Creinin 1995). Maneuvers to facilitate vaginal delivery have been described (Grover et al. 1990; Creinin 1995), but craniotomy, decapitation, evisceration, or amputation may be needed as a last resort (Grover et al. 1990; Creinin 1995).

Perinatal and ethical dilemmas in conjoined twins

When separation of CTs is not immediately required, surgery is usually delayed until late infancy, to permit careful preoperative evaluation and better tolerance of surgery (Creinin 1995; Bianchi et al. 2000; Mackenzie et al. 2002; Sen et al. 2003). Intervention is sometimes needed at birth or within 24 hours (Mackenzie et al. 2002). Separation of a pair of tribrachius (three-arm) dicephalus twins resulted in one infant's death (Bondeson 2001), and dicephalus twins generally are stillborn or only live a few hours, so comfort measures may be most appropriate for them (Groner et al. 1996).

Ethical issues are thus often urgent (Atkinson 2004; Bratton and Chetwynd 2004). Sanctity and quality of life for each twin are primary concerns (Atkinson 2004). Multidisciplinary committees are best suited to consider the medical, social, cultural, religious, and legal implications of management options (Sakala 1986; Atkinson 2004). Clinicians must involve parents in decisions soon after diagnosis, and can encourage them to consult counselors and clergy (Atkinson 2004). Courts may become involved (Atkinson 2004). Society may assume CTs are two people who wish to survive regardless of bodily form, with a preference for separate existence (Atkinson 2004). However, some adult CTs prefer to remain attached, rather than to risk death or an inferior quality of life (Atkinson 2004). Thus, separation is not always desirable (Atkinson 2004; Bratton and Chetwynd 2004).

There are three management options: 1) withholding or withdrawing life support to allow both twins to die; 2) separating the twins, with the possibility that one will die; or 3) allowing twins to remain conjoined (Creinin 1995). With the first option, one may question whether parents can justifiably refuse interventions that would permit one or both twins to live (Atkinson 2004). Absolute clinical indications for separation usually include removing a stillborn or critically ill twin's connection to the healthier twin, or repairing an anomaly that would require immediate surgery in a singleton (Creinin 1995; Mackenzie et al. 2002).

The second option, separation, may cause death of one twin to allow life or independence for the other (Atkinson 2004; Bratton and Chetwynd 2004). Sacrifice of one individual to save another is supported by most religions and by courts, although some ethicists disagree with assumptions made in legal analyses (Atkinson 2004; Bratton and Chetwynd 2004). Several opinions have been expressed: 1) one twin is a parasite on the other, 2) CTs are entangled singletons, meant to be physically separated, even if one dies and perhaps more reasonably, 3) CTs require a unique ethical approach, as psychologically separate individuals in a shared body (Atkinson 2004; Bratton and Chetwynd 2004).

The third option, leaving twins conjoined, must be considered when dicephalus twins who share organs extensively cannot be separated (Groner et al. 1996; Bondeson 2001). Attempts to separate them could be deemed unethical (Bondeson 2001).

Professionals must protect the confidentiality of families with CTs (Atkinson 2004). Societal condemnation, as feared by the family in our case, may occur either locally as a negative reaction to an anomalous birth, or worldwide as the media criticize a private decision (Atkinson 2004). Obstetricians must not abandon patients after delivery. Empathic counseling for decisions, similar

to that given to parents of anomalous singletons, can be offered, with bereavement support if one or both babies die (Sakala 1986).

Conclusions

Conjoined twins are rare, and parapagus dicephalus twins represent only a small proportion. Anomalies of the heart and abdominal organs occur in most dicephalus twins. Early diagnosis facilitates termination, or optimal obstetric and perinatal preparation. Practitioners may unexpectedly encounter conjoined twins at delivery. If no antenatal studies were performed in suspected twins, practitioners can obtain an ultrasound to rule out conjunction before delivery. Ethical issues surrounding separation do not apply to parapagus twins, whose extensively shared organs preclude survival or acceptable quality of life.

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