Separation of Conjoined Twins with the Twin Reversed-Arterial-Perfusion Sequence after Prenatal Planning with Three-Dimensional Modeling

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Brief Report

There are two congenital anomalies specific to multifetal pregnancies: twin reversed-arterial-perfusion sequence and conjoined twinning. The twin reversed-arterial-perfusion sequence is a rare complication of monzygous twinning in which one fetus (the "pump" twin) perfuses the other fetus (the "perfused" twin), resulting in reversed flow in the umbilical vessels and multiple structural anomalies, including acardia, in the perfused twin. It occurs in fewer than 1 percent of pregnancies with monzygous twins and in about 1 in 35,000 births overall. Conjoined twinning occurs as a result of incomplete duplication of a single blastocyst during the process of monzygotic twinning. It is a sporadic complication that occurs at rates ranging from 1 in 30,000 to 1 in 100,000 births.

Surgical separation of conjoined twins is commonly undertaken after birth, but surgery is usually delayed for weeks or months. With time, the infants become larger, the anatomical relations between them can be better delineated, other congenital anomalies may be identified, the risk associated with anesthesia usually decreases, and the separation procedure can be carefully planned. In contrast, a twin reversed-arterial-perfusion sequence may be treated by sacrificing the acardiac twin in utero. We report here a unique case of conjoined twins with reversed-arterial-perfusion sequence. Because the twins were joined, antenatal surgical intervention to treat the twin reversed-arterial-perfusion sequence was not possible, so immediate surgical separation at birth was necessary. Given this situation, we used three-dimensional computer modeling with prenatal magnetic resonance imaging (MRI) to assist planning for the surgical separation of the twins at birth. This approach made it possible for the pump twin to survive.

Case Report

Routine ultrasonic examination at 14 weeks' gestation in a 29-year-old woman (gravida 2, para 0) revealed thoracoamphalopagus twins (twins joined at the chest and abdomen), with acardia in one twin. Amniocentesis revealed a 46,XX fetal karyotype. The woman declined termination of the pregnancy. Fetal echocardiography at 19 weeks' gestation revealed a single heart with two functional ventricles and a functionally unipartite liver in one twin, with each part extending into the thorax of the other twin. The latter twin had a rudimentary cardiac structure consisting of a single, thick-walled, slowly contracting chamber that was fed and drained by a single large vessel. A single umbilical cord with four arteries was identified. Doppler studies showed that the arterial supply to the acardiac twin flowed from the umbilical arteries of the twin who had a heart and that blood drained back to the twin with a heart through a single large ductus venosus that coursed through the fused livers. No other large communicating vessels could be identified, and there were no other fetal anomalies. The head circumference of each twin was normal for gestational age. Follow-up fetal echocardiography at 27 weeks' gestation demonstrated retrograde flow up the aorta of the perfused twin, thereby confirming the diagnosis of the twin reversed-arterial-perfusion sequence (Fig. 1). A video of the echocardiogram can be viewed on the Internet at http://www.nejm.org/content/2000/0343/0306/0399.asp. It was not possible to determine whether the shared arterial circulation crossed in the placenta or in the umbilical cord.

Clinical instability immediately after birth was anticipated because division of the umbilical cord would impair perfusion to the acardiac twin. Cesarean delivery followed by immediate surgery was planned. Surgical separation of the twins was therefore recommended. This plan was approved by the ethics committee of Children's Hospital, Boston. The family was counseled that the acardiac twin could not survive, and that any delay in surgical separation might jeopardize the survival of the normal twin. Given the urgency of the separation procedure, it was necessary to obtain as much anatomic information as possible before delivery. Prenatal ultrasonographic examination had revealed that the twins' livers were fused, but the gallbladder and biliary drainage systems were not seen.

To delineate the anatomical features of the upper abdomen of the fetuses more clearly, MRI was performed at 28 and 32 weeks' gestation (Fig. 2). T1- and T2-weighted images were obtained in the axial, sagittal, and coronal planes with a 1.5-T magnet (General Electric Medical Systems, Milwaukee) and a pelvic phased-array coil. The imaging settings were as follows: repetition time, 4200 msec; echo time (effective), 108 msec; phase encoding steps, 128; field of view, 24 cm; and slice thickness, 3 mm with no gap and with two acquisitions. The sequence was repeated after the slice locations were adjusted to obtain contiguous images 1.5 mm in thickness. The images were then transferred electronically to an UltraSPARC-30 graphics computer (Sun Microsystems, Mountain View, Calif.). The data were segmented into anatomical components, and three-dimensional computer models were generated with the use of software (Slice) developed in the Surgical Planning Laboratory of Brigham and Women's Hospital, Boston. The three-dimensional surface models confirmed the cardiovascular anomalies described above. In contrast to the results of ultrasound examination, the MRI model revealed two biliary drainage systems within the fused livers (Fig. 3). A video of the MRI studies and...
of the three-dimensional model can be viewed on the Internet at http://www.nejm.org/content/2000/0343/0006/0399.asp.

At 38 weeks, severe preclampsia developed in the mother. An urgent cesarean delivery was performed, and conjoined female twins were delivered without complication. The umbilical cord was ligated at its junction with the placenta in an attempt to avoid interrupting any intraumbilical vascular shunts. Appar scores of 7 at one minute and 8 at five minutes were assigned to the twins. Within minutes after delivery, the acardiac infant was cooler than her twin. The infants were intubated. Postnatal echocardiography confirmed the prenatal findings, but there was only low-velocity, nonpulsatile retrograde flow in the aorta of the acardiac twin. She had no pulse, and her blood pressure could not be measured. Immediate surgical separation was initiated.

Since the acardiac twin would not survive, the incision for separation was performed far toward her side of the fusion plane so that her tissue could be used to achieve complete closure of the ventral defect in the surviving twin. Exploration revealed abutting, but not joined, pleural and peritoneal cavities. There were no great vessels communicating from the twin with a heart to the acardiac twin. A large venous structure draining from the acardiac twin’s vestigial cardiac chamber to the superior aspect of the fused livers was divided to facilitate hepatic separation. The presence of separate biliary tracts was confirmed, and the fused livers were divided bloodlessly in the central plane. The single large ductus venosus was encountered in the central hepatic parenchyma and was ligated. The procedure culminated in closure of the ventral defect in the surviving infant with use of her twin’s ribs to construct a cage-like structure that would protect her protonotar heart.

Hemodynamic instability developed in the surviving twin on the first postoperative day and necessitated further surgery to relieve a possible compartment syndrome or tamponade effect and to ligate a large patent ductus arteriosus. At three and a half months of age, progressive feeding intolerance in the infant led to the identification of a small-bowel stricture requiring surgical resection with primary anastomosis. The infant was discharged home at six months of age. The mother’s postoperative course was uneventful, and she was discharged home on day 4 after delivery.

DISCUSSION

Conjoined twins are classified according to the location of the tissue that links the twins. Twenty-eight percent of conjoined twins are classified as thoraco-omphalopagus.5,8 Anatomical abnormalities are common among conjoined twins. Congenital heart disease (most commonly, a ventricular septal defect or tetralogy of Fallot) is present in 75 percent of thoracopagus twins (those joined at the chest), and there is some degree of fusion of the pericardial sac in 90 percent8,10; the liver is shared in 81 percent of omphalopagus twins (those joined at the abdomen).8,11

Twin reversed-arterial-perfusion sequence is a condition that results from one or more vascular communications between twin fetuses, such that deoxygenated blood from the pump twin is diverted directly to the perfused twin before gas is exchanged in the placenta. The pump twin usually develops normally but is vulnerable to intratruine congestive heart failure and premature delivery because of polyhydramnios. The perfused twin usually has aplasia or hypoplasia of the heart, head, and arms and therefore dies immediately after birth. Mortality among pump twins ranges from 50 percent to 75 percent, with most deaths due to intratruine cardiac failure or complications of prematurity.2,12 In view of this poor prognosis, it is often recommended that the umbilical circulation between the twins be interrupted prenatally.
(thus sacrificing the acardiac twin) before heart failure develops in the pump twin. In the current case, however, antepartum surgery for twin reversed-arterial-perfusion sequence was not possible because the twins were joined. Paradoxically, the unique vascular configuration in this case may have resulted in improved flow of oxygenated blood to the cephalad region of the acardiac twin, which may explain the absence of aplasia or hypoplasia in the head and arms of that twin.

Surgical separation of conjoined twins must be planned on a case-by-case basis in accordance with the anatomical arrangement of the shared tissues. The surgical challenges include separation of important shared organs (such as the liver and the heart) and closure of defects in soft tissue and bone. Survival depends on the type and extent of joining and on the presence or absence of associated anomalies. In the absence of severe anomalies, thoracopelapagous twins have a reasonable chance of survival after surgical separation. Prognosis is usually determined by the extent of fusion of the heart, but as noted above, surgical separation is usually deferred for weeks or months. In a review of 47 pairs of surgically separated thoracopagus twins, Chiu et al. reported an overall survival rate of 48 percent. However, the survival rate was 38 percent among those separated in the neonatal period, as compared with 64 percent among those separated after the first month. Early separation is indicated when one twin threatens the life of the other. In the current case, the presence of the twin reversed-arterial-perfusion sequence and concern about potential adverse hemodynamic and metabolic changes after delivery necessitated immediate surgical separation at birth. To assist in planning the separation, a three-dimensional computer model of the conjoined twins was constructed on the basis of prenatal MRI studies. The most important information provided by MRI was related to the detailed anatomical structure of the biliary tracts and the spatial relation of the shared anatomical structures.

MRI has emerged as a useful imaging technique because of its high resolution, its ability to characterize tissue, and its capacity to generate information in three planes (axial, sagittal, and coronal). To date, the use of MRI during pregnancy has been limited to the evaluation of pelvic masses in the mother and of rare fetal signs that indicate a possible need for in utero surgical intervention. We describe a new use for MRI during pregnancy — namely, to construct three-dimensional models to assist in pre-
natal planning when it is anticipated that surgery will be required immediately after delivery. In this case of thoracocoophalopagus twins with the twin reversed-arterial-perfusion sequence, fetal echocardiography and three-dimensional computer modeling with MRI contributed to the success of the surgical separation of the twins at birth and to the survival of the pump twin.

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