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Classification and clinical evaluation

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ABSTRACT

Conjoined twins represent a great challenge for most pediatric specialists including pediatric surgeons, anesthetists, neonatologists, urologists, neurosurgeons, and orthopedic surgeons. This anomaly can be classified according to the type of twins' fusion. Various organs can be fused making the separation difficult. Conjoined twins are usually diagnosed antenatally by ultrasound. Detailed fetal echocardiography is necessary to counsel the parents during pregnancy. Postnatally, the majority of the conjoined twins can be thoroughly investigated using various imaging techniques. This allows careful planning of the operation. However, in approximately one-third of the patients an urgent operation is required at birth without a complete assessment of the joining. This is associated with a poorer outcome.

Introduction

Conjoined twins are rare. This anomaly is characterized by joining of two identical twins who share one or more organs. Conjoined twins have fascinated mankind from many centuries and the description of conjoined twins date back to the sixth millennium BC in Turkey and to 80 BC in Italy.

The incidence of conjoined twins ranges from 1:50,000 to 1:100,000 live births,[1] and it is reported to be higher in Africa and in South East Asia. This number could be higher, but most of these pregnancies result in miscarriages and stillbirths; 18% of all conjoined infants survive, and approximately 35% of live births die within the first 24 h, and only 18% of all conjoined twins survive longer than 24 h.[2]

The exact etiology of conjoined twins remains unknown and two theories have been proposed for the embryologic development, the fission and the fusion theories. Fusion seems to be the more likely explanation for all the various types of conjoined twinning.

Classification

Conjoined twins can be subdivided into (i) symmetric conjoined twins and (ii) heteropagus or parasitic twins. This article focuses on the symmetric twins.

Conjoined twins are always of the same sex and are joined homologously,[1] i.e., chest to chest, abdomen to abdomen, pelvis to pelvis.

Twins can be classified according to the most prominent site of connection.[3] The mostly accepted classification is the one proposed by Spencer,[4] which divides the anomaly in eight types (Figure 1).

Thoracopagus

Thoracopagus twins are united face to face from the upper thorax to the umbilicus with a common sternum, diaphragm, and upper abdominal wall (Figure 2). Overall, 90% of such twins have a common pericardial sac, and there is almost always a degree of cardiac fusion; in 75% of cases, the severity of cardiac fusion precludes successful surgical separation. They may have a common small intestine (50%) that joins at the duodenum and separates at ileum; the biliary tree can be joined in 25% of patients. There may be associated cardiac anomalies such as ventricular septal defect, atrial septal defect, and tetralogy of Fallot.

Omphalopagus

Omphalopagus twins are joined ventrally in the abdomen, often including the lower thorax (Figure 3). The heart is never fused, although the pericardium may be shared. Liver fusion occurs in approximately 80% and there is an omphalocele. The distal duodenum and small intestine to the level of the Meckel's diverticulum in the distal ileum is usually shared. There is usually no union of the genitourinary tract.

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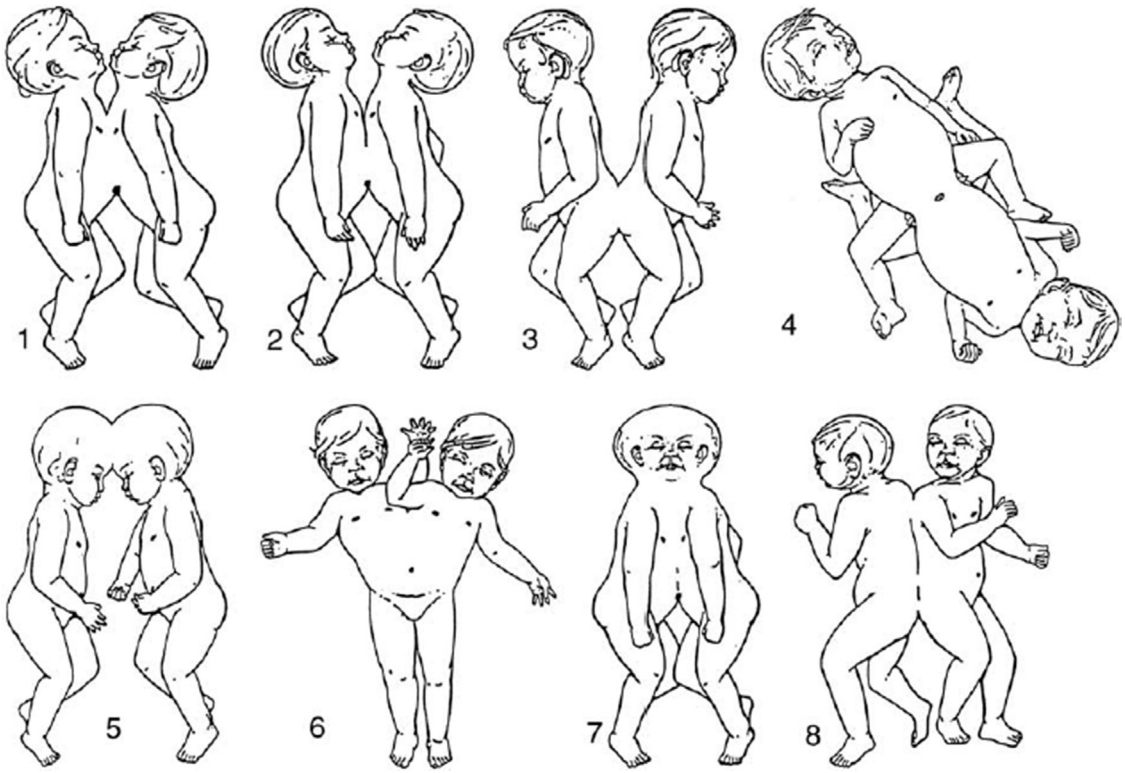

Fig. 1. Classification of conjoined twins according to Spencer.[4] 1 = thoracopagus, 2 = omphalopagus, 3 = pygopagus, 4 = ischiopagus, 5 = craniopagus, 6 = parapagus, 7 = cephalopagus, 8 = rachipagus. (Adapted with permission from Spencer.[4])

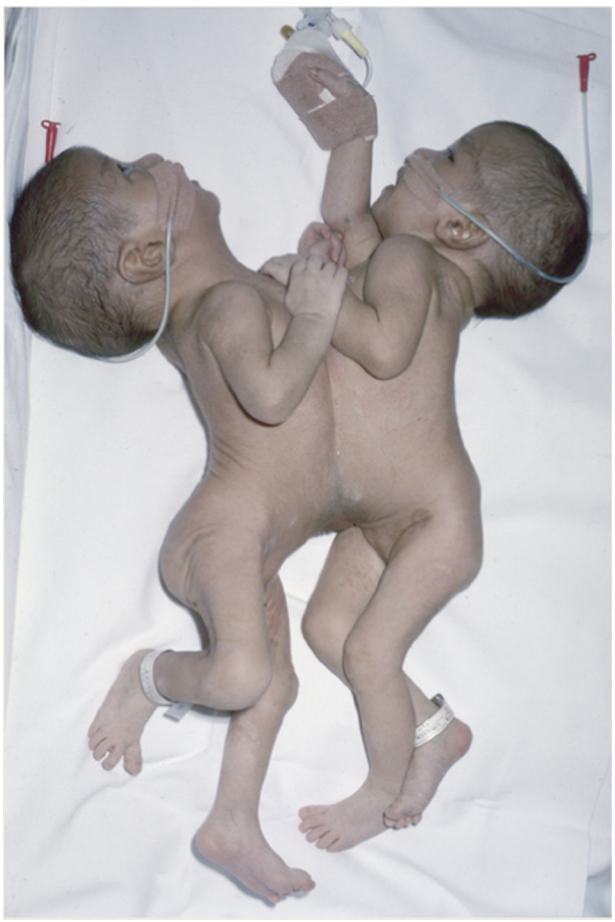

Fig. 2. Thoracopagus.

Pygopagus

Pygopagus twins are joined dorsally, facing away from each other and sharing the sacrococcygeal and perineal regions. Fusion of sacral vertebrae frequently occurs, but the spinal cords are usually joined at the filum. A total of 25% share the lower GI tract and have a single anus and one or two rectums. In 15% of cases there is a single bladder. There is an increased incidence of vertebral anomalies, including hemivertebrae, hemisacral agenesis, and thoracic anomalies. Numerous other associated orthopedic anomalies have been reported in association with pelvic conjunction, such as hip subluxation or dislocation, congenital vertical talus, talipes equinovarus, and scoliosis. Although there may be only one anus and rectum, the remainder of the intestines are usually separate. The upper bodies are not fused and there are four arms and four legs.

Ischiopagus

Ischiopagus twins are fused from the umbilicus to a large conjoined pelvis (Figure 4). The spinal columns are usually separate. They may lie face to face or end to end with the vertebral columns in a straight line. The components of the pelvis vary; usually, there are two sacra and one or two symphysis pubis. The twins are termed tetrapus (four), tripus (three), or bipus (two) according to the number of legs attached to the conjoined pelvis. Tetrapus twins are the most common. Pelvic conjunction gives rise to complex anatomy requiring thorough preoperative evaluation, especially from a urologic and orthopedic point of view. The intestine is shared from the distal ileum to the rectum/anus.

Craniopagus

Craniopagus twins may be joined at any part of the skull except the face or foramen magnum. The fusion is vertical and parietal in

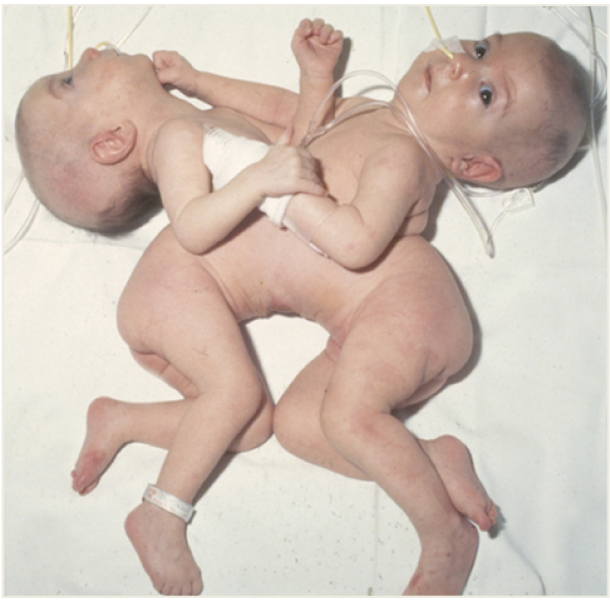

Fig. 3. Omphalopagus twins.

over 60% of cases, but frontal fusion, occipital fusion, and extensive temporo–parieto—occipital fusion have been reported. The conjoined twins share the skull, the meninges, and the venous sinuses. The brains are usually separate, although some cortical fusion can occur in 33% of cases (Figure 5).

Parapagus

This is a relatively new term denoting extensive side-to-side fusion (Figure 6). Parapagus twins lie side to side with ventrolateral fusion. The twins share the umbilicus, lower abdomen, pelvis (single symphysis pubis), and genitourinary tract. They can have anorectal anomaly and colovesical fistula and may be at risk of anencephaly. The conjoined pelvis usually has a single symphysis pubis and one or two sacra. The thorax may be involved. The twins can have (i) separate heads (*dicephalic*) but the entire trunk is conjoined and (ii) separate thoraces (*dithoracic*) with the fusion involving the abdomen and pelvis. They can have two, three, or four arms and two or three legs.

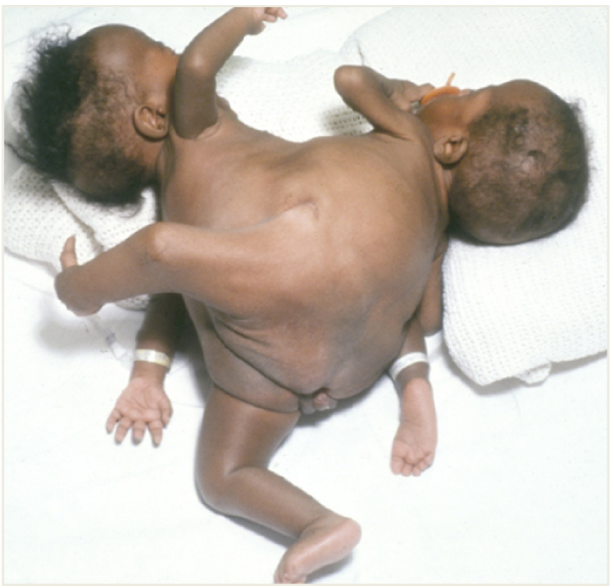

Fig. 4. Ischiopagus twins.

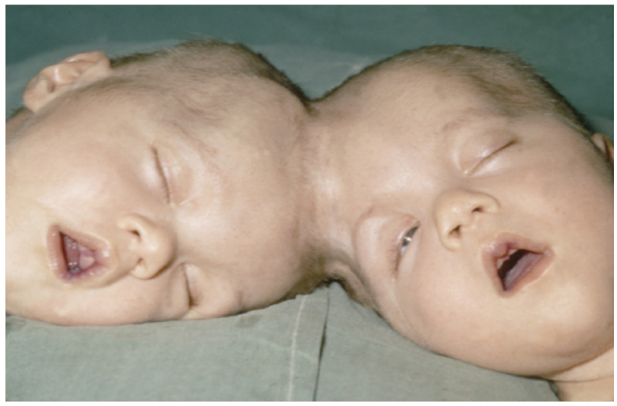

Fig. 5. Craniopagus.

Cephalopagus

The twins often have a fused thorax in addition to a fused head. They are fused from the vertex to the umbilicus. There are two faces on opposite sides of the conjoined head; one face is usually rudimentary. These twins are terminated or die in utero. They are nonviable.

Rachipagus

Rachipagus twins are extremely rare. They are joined dorsally and face away from each other. The occiput may be involved, along with varying segments of the vertebral column. The fusion terminates above the sacrum. The twins have generally vertebral anomalies and neural tube defects.

Fig. 6. Parapagus twins.

Clinical evaluation

Antenatal diagnosis and imaging

The diagnosis of conjoined twins should occur antenatally allowing adequate counseling of the parents. Prenatal ultrasonography (US) is capable of detecting fetal conjoined twins as early as 12 weeks' gestation. A repeated US scan at 20 weeks' gestation is usually able to confirm the diagnosis and provide an accurate definition of the site and extent of the joining. The diagnosis of conjoined twins can be straightforward when fusion of the fetuses is detected. Conjoined twins should be suspected when a single placenta and no visible separating amniotic membrane are seen at US. Conversely, the US visualization of two placentas or an amniotic membrane excludes the presence of conjoined twins. The US features that suggest the antenatal diagnosis of conjoined twins include constant relative positions of the fetuses over time, with heads and other body parts persistently at the same level; inseparable body and skin contours; fetuses facing each other with hyperflexion of the cervical spines; fewer limbs than expected; shared organs; and a single umbilical cord with more than three vessels.[5–7]

Most conjoined twins are fused ventrally[8] and present cervical hyperextension, a feature that is commonly observed postnatally. In addition, polyhydramnios is present in approximately 50% of the conjoined twins pregnancy compared to 10% of normal twins.

The most important evaluation prenatally is the fetal echocardiography. The degree of cardiac fusion and the severity of associated cardiac anomalies determine the postnatal viability of the twins and the likelihood of successful separation. The hearts of conjoined twins are easier to examine in utero because the amniotic fluid acts as a buffer during US. After birth, the lungs inflate with air and thoracic fusion prevents optimal access; hence, limited information may be obtained.[7] A detailed fetal echocardiography is important, as there is an increased incidence of cardiac anomaly in conjoined twins. In addition, joined hearts are usually incompatible with life. The information provided by fetal echocardiography is very important to predict the outcome of the twins and counsel the parents.

A CT scan is very useful to delineate the anatomical arrangement (Fig. 7).[9] In recent years magnetic resonance imaging (MRI) has been used more commonly in confirming the diagnosis and defining the type of joining. Ultrafast T2-weighted MRI sequences eliminate the need for maternal sedation and allow precise anatomical assessment of fetal organs avoiding the artifact due to fetal movement.

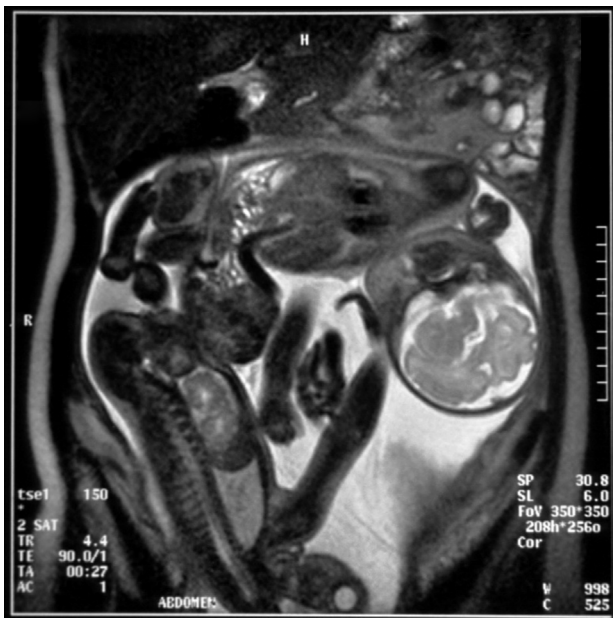

Fig. 7. MRI scan of omphalopagus twins in utero showing intestine in the connecting bridge.

The information obtained by US and MRI are essential to the pediatric surgeon to hold a multidisciplinary discussion with the other experts in the fields of fetal medicine, neonatology, cardiology, cardiac surgery, neurosurgery, urology, and orthopedics. The treatment plan is discussed with the parents to inform them about the chances of survival and the expected functional outcome. The ethical issues discussed include termination of pregnancy, time, location, and type of delivery. Termination of pregnancy is recommended in twins with complex cardiac fusion or extensive cerebral fusion. A detailed and objective description of the postnatal management is presented to the parents. The extent of the deformity following separation and the expected functional outcome are carefully explained to the parents so that they can make an informed decision about either termination of pregnancy or proceeding with the pregnancy. Delivery should take place in the same hospital or close to the center with pediatric surgical expertise where the twins' separation will be done. Delivery is planned between 36 and 38 weeks' gestation by cesarean section to avoid iatrogenic injuries to the twins and the mother.

Postnatal evaluation

After birth, the twins are fully assessed clinically to evaluate the type of joining. It is useful to take into account the antenatal imaging predominantly the fetal echocardiography. At birth, the twins can be intubated and ventilated, particularly during transport. Their physiological status is evaluated taking into account the respiratory need, the cardio-respiratory function, the urine output, and the passage of meconium. All conjoined twins should have chest and abdominal radiographs for general assessment.

Associated anomalies are common in conjoined twins and the presence of diaphragmatic hernia, intestinal obstruction, or vertebral anomalies can be detected early. In our series of conjoined twins, we noticed associated anomalies in 65% of the twins. These anomalies involve various systems with the most common being major cardiac defect, intestinal atresia, omphalocele, anorectal atresia, diaphragmatic hernia, and spinal cord anomaly.

The initial concern is whether or not emergency surgery is needed in the immediate postnatal period. Approximately one-third of the conjoined twins require an emergency operation at birth when:

- One twin is dead or dying and therefore threatening the survival of the remaining twin. In this event, an operation is required immediately to separate the twins. This usually does not leave time to perform the diagnostic tests and the imaging techniques discussed below.
- There is a life threatening but correctable congenital abnormality such as intestinal atresia, volvulus, ruptured omphalocele, or anorectal agenesis. In these scenarios, an emergency operation is performed. In twins with intestinal atresia or anorectal anomaly, the operation is limited to stoma formation postponing the twins' separation to a later date. This allows growth of the twins and performance of imaging aimed to detailing the anatomy and the type of joining.

In most circumstances, the twins' separation will not occur before 3–4 months of age leaving time for the performance of diagnostic imaging and carefully planning surgery. The choice of imaging will depend to some degree on the site of fusion. Due to the high incidence of associated anomalies, it is important to

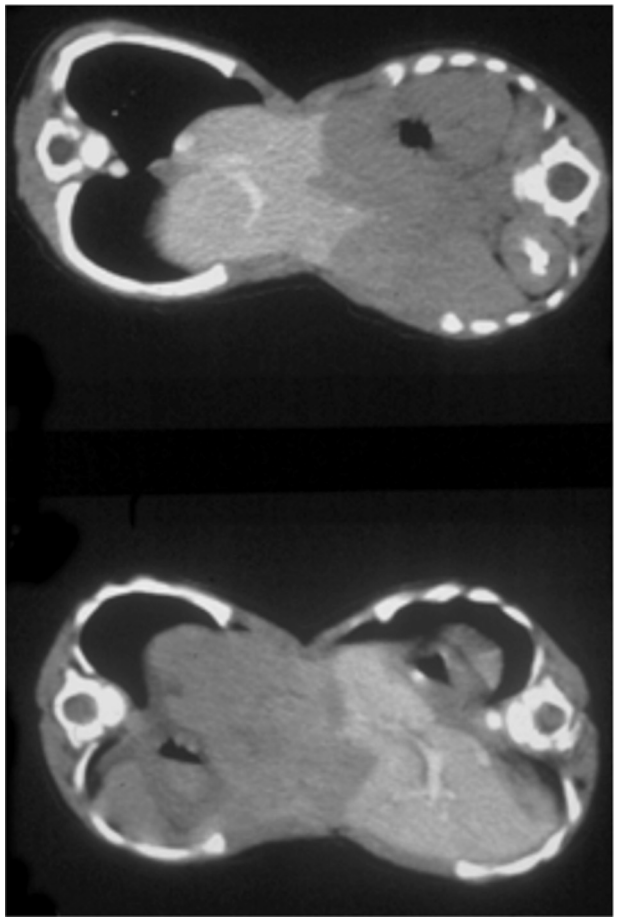

Fig. 8. CT scan with contrast to show line of liver union.

perform a detailed radiological assessment of the twins. All conjoined twins, irrespective of the site of connection, should undergo echocardiography, as there is a higher frequency of cardiac abnormalities in all forms of conjoined twinning. On arrival at the pediatric surgical center, twins who have not undergone imaging are initially assessed with a combination of plain radiography, US, and echocardiography. These techniques can be easily performed in the intensive care unit and do not require sedation or anesthesia. They are used to provide an overview of conjoined anatomy and identify potential problems such as diaphragmatic hernia or bowel obstruction that may require interim procedures. Twins in stable condition in whom detailed imaging is possible undergo a full range of investigations, which are mainly determined by the site of conjunction. MRI techniques provide the best overall anatomic detail, but computed tomography (CT) is more useful if bone detail is required. A CT scan with contrast may be of potential value to delineate the line of surgical separation of the fused liver (Fig. 8).[7]

There is often a discrepancy in the size of the twins in the period between birth and the separation at around 3 months. One twin is more active, feeds well, and is thin, while the other is less active, feeds less, and is fatter. Their personalities are also different matching their activity. Metabolic rate measurements have shown that the thin twin has a higher respiratory quotient than its twin, but the difference equals out following separation.[10] We have repeatedly seen that less active twin is physiologically less robust following separation and needs very careful monitoring of cardiac function.

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