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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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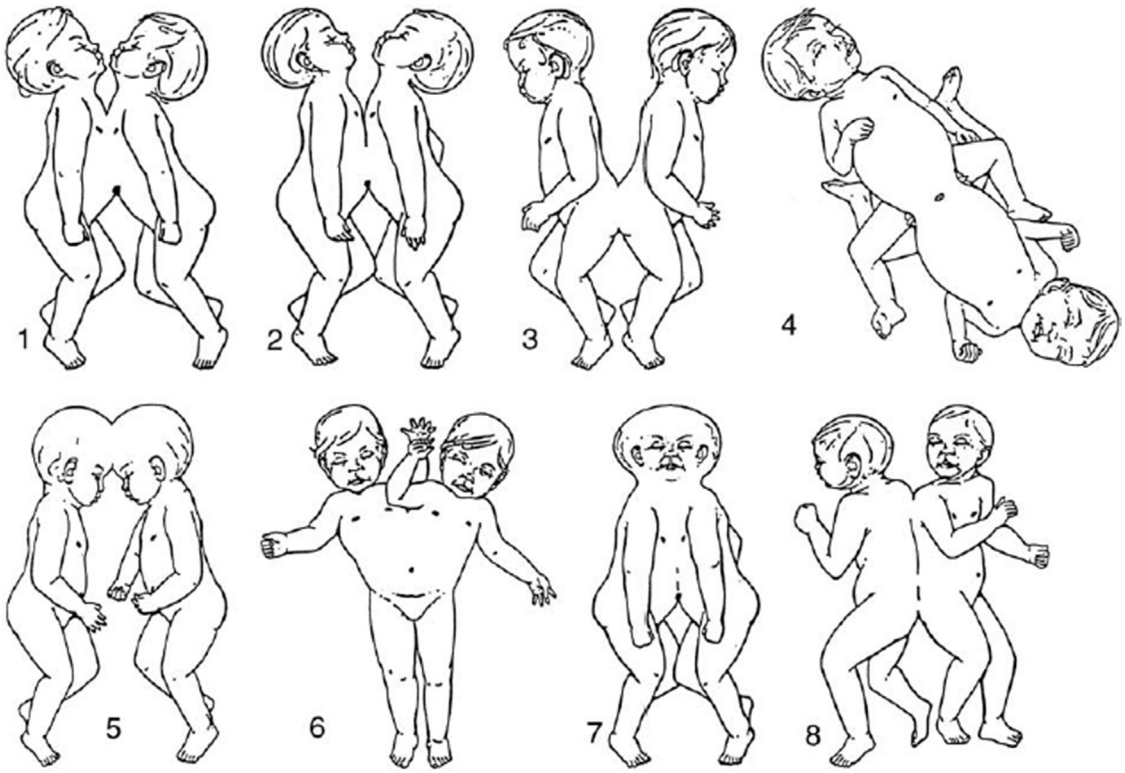

Fig. 1. Classification of conjoined twins according to Spencer.[4] 1 = thoracopagus, 2 = omphalopagus, 3 = pygopagus, 4 = ischiopagus, 5 = craniopagus, 6 = parapagus, 7 = cephalopagus, 8 = rachipagus. (Adapted with permission from Spencer.[4])

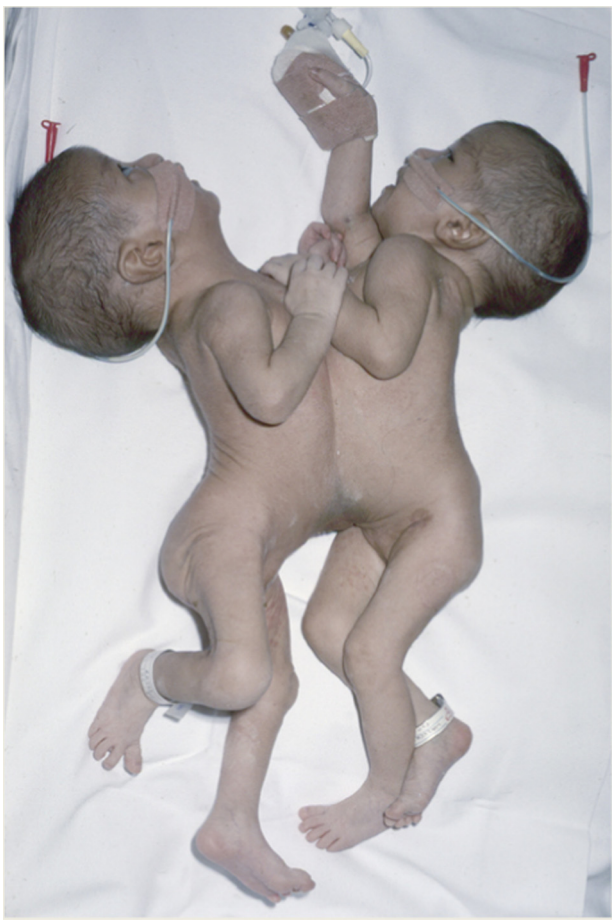

Fig. 2. Thoracopagus.

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