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Prenatal diagnosis and obstetric management

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ABSTRACT

Conjoined twins are rare, representing 1 in 50,000 to 1 in 200,000 live births, and the prognosis is generally poor. Accurate prenatal diagnosis by an experienced multidisciplinary team using a combination of imaging modalities allows parents to make fully informed choices. This may include termination of pregnancy, which is easier and safer at the earlier gestations at which diagnosis is now being made; continuing with the pregnancy but accepting that only palliative care is appropriate after birth; or planned intensive care and separation of the twins after birth. Delivery will invariably be by cesarean section in order to minimize the risk of peripartum harm to both mother and babies.

Introduction

Conjoined twins are so rare that it is difficult to be accurate about the incidence, but it probably lies somewhere between 1 in 50,000 and 1 in 200,000 live births.[1–5] Conjoined twins are always monochorionic monoamniotic and represent around 1% of monozygotic twins. It is thought to be due to splitting and incomplete separation of the inner cell mass at 13–15 days post-fertilization, but the underlying etiology of this anomaly of duplication is unknown. The female to male sex ratio is around 3 to 1.[6]

The incidence may well be falling because of more widespread prenatal diagnosis with good-quality ultrasound and termination of affected pregnancies. However, 40% of conjoined twins are stillborn and a further 30% die on the first day of life.[7] Consequently, worldwide only 6 – 8 sets of conjoined twins make it as far as neonatal surgery every year.

Conjoined twins are classified according to the site of fusion; they are always joined at corresponding anatomical points (Table)[8]. The commonest forms are thoracopagus (20–40%), omphalopagus (18–33%), and parapagus (28%), although combinations of the various types can occur.

Prenatal diagnosis

The first reported ultrasound diagnosis of conjoined twins was in 1976 by Wilson et al.[9] and in economically developed countries, most cases are now diagnosed prenatally, usually as early as the end of the first trimester.[10–12] In contrast, because most women in developing countries have little or no antenatal care, the diagnosis is often not made until the time of labor and birth, usually with traumatic consequences for both the mother and babies.[13,14] First trimester ultrasound clues that may raise the suspicion of conjoined twins are the following: only one yolk sac with two embryos in very early pregnancy, the embryos are juxtaposed with the focus of cardiac motion for each side appearing to merge, antero-posterior fusion with a single central motion and two separate cranial poles, motion of the probe moves both embryos at the same time, and no sign of separate movement of the twins.

Accurate prenatal diagnosis is of critical importance to optimize outcomes for both mother and babies. Once the diagnosis is made, the woman should be referred to a fetal medicine center with expertise in the management of these pregnancies. Assessment and counseling of the parents should be carried out not just by the fetal medicine scanning specialists, but also by a multidisciplinary team that includes the obstetrician who will deliver the twins, a senior midwife, senior neonatal nurse, neonatologist, pediatric surgeon, and pediatric cardiologist (not necessarily all at the same time, of course!). Assessment will be primarily by ultrasound; in recent years, the addition of color Doppler and 3-D ultrasound has aided earlier and more accurate diagnosis. These imaging techniques have also made it easier for parents to visualize the abnormalities affecting their twins, which facilitates counseling and decision making.[15,16] Associated cardiac anomalies are common, being present in up to 90% of cases, and have an adverse effect on prognosis.[17] Fetal echocardiography is therefore an essential part of the prenatal assessment. Other associated fetal anomalies are common (61.8% have one or more), including limb abnormalities (36.1%), abdominal wall defects (25%), cleft lip and/or palate (13.9%),

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Table
Classification of conjoined twins with incidence.

Classification	Incidence (%)
Ventral union	
Cephalopagus: fusion from top of head to umbilicus. Each twin has two extremities and lower abdomen and pelvis are separated.	11
Thoracopagus: twins are located face to face, with fused thoraces and shared heart or single interatrial vessel.	20–40
Omphalopagus: twins have similar fusion to thoracopagus without shared heart or interatrial vessel.	18–33
Ischiopagus: twins share a large conjoined pelvis and are more commonly joined end to end. External genitalia and anus are always shared.	6–11
Dorsal union	
Craniopagus: twins are joined by any portion of the skull except the face and foramen magnum. The bony cranium, meninges and brain are shared.	2
Pygopagus: twins have fused sacrococcygeal and perineal regions, typically with shared anus but separate rectums. The spinal cord may be shared.	18–28
Rachipagus: twins have dorsal fusion above the sacrum.	Rare
Lateral union	
Parapagus: twins have side-by-side connection with shared pelvis and variable cephalad sharing defined as follows: – Dithoracic parapagus: separate thoraces and heads. – Dicephalic parapagus: separate heads with fused thoraces. – Diprosopus parapagus: two faces on the same side of single head.	28

Adapted with permission from Winkler et al.[8]

and diaphragmatic hernia (5.5%),[17] so expert ultrasound assessment is mandatory, and will usually be carried out every 2 weeks or so. Amniocentesis or chorionic villus sampling (CVS) is usually offered to exclude chromosome abnormality. Magnetic resonance imaging (MRI) does not involve radiation and is safe in pregnancy. Prenatal fetal MRI can add information that helps define the precise anomalies and the connections between the fetuses. It is useful in the first trimester to help define the anatomy; later in pregnancy, the additional information it provides can help in deciding the mode of delivery and planning postnatal surgery. The combined use of these imaging modalities in expert hands will usually accurately diagnose the anatomical anomalies and connections between the twins before the birth (Figure).

While all of this medical assessment is taking place, the pastoral care of the parents and extended family must not be forgotten. This is obviously a very stressful time for the parents and they should be involved in every aspect of the planning of the pregnancy and birth. Continuity of care from named team members providing kind professional support is rewarding for everyone, particularly on the day of delivery.

Early referral to a named neonatal counselor for antenatal and postnatal support is required, and honesty around the uncertain outcomes after birth is essential. At all times, the parents' religious and cultural views should be respected. Clear communication and documentation is essential. Confidentiality must be meticulously maintained in the face of the intense media attention such pregnancies usually attract.

Although the prognosis for all types of conjoined twins is very poor, accurate definition of the anatomy, vascular connections, and position of the twins is very important.[11,18,19] Those with craniopagus, a sole cardiac mass or complex cardiac connections have the worst prognosis, but all parents with a diagnosis of conjoined twins should be offered termination of pregnancy, where this is legally permitted. However, even if termination of pregnancy is not permitted or desired by the parents, the accurate prenatal diagnosis of a lethal abnormality allows the parents time to prepare for the birth and demise of their twins, preparation for a delivery that minimizes the risk of trauma to the mother, and planning of palliative postnatal care. Accurate prenatal anatomical description of conjoined twins that are potentially amenable to postnatal separation will facilitate delivery in the appropriate setting, with the necessary obstetric, neonatal, and pediatric surgery expertise on hand. It is clear that such preparation can make a huge difference to the outcome. A paper from Poland describes two similar sets of conjoined twins; one set delivered in a regional hospital had a disastrous delivery and both died. The other set, delivered at a center of excellence, underwent successful postnatal surgical separation, and both survived.[14] In another

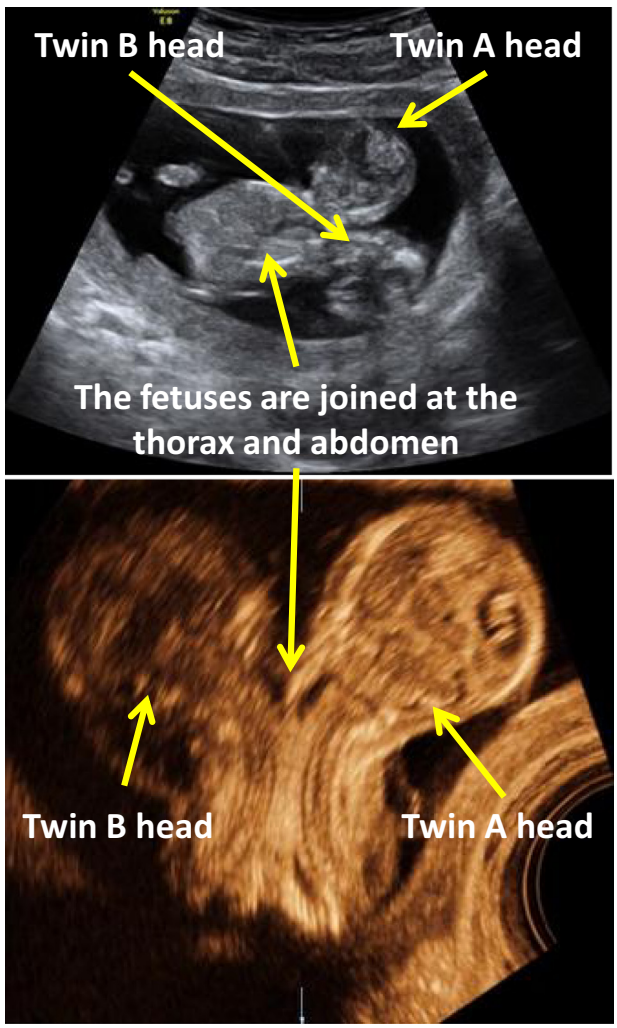

Fig. Ultrasound (2D upper image; 3D lower image) of conjoined twins at 12+5 weeks (Thanks to Professor Rabih Chaoui, Berlin).