

SUMMARY & CONCLUSION

Conjoined twinning is a rare sporadic occurrence with no Genetic predisposition. Females predominate with a ratio of 3:1. Conjoined twins develop from a single fertilized ovum. The most widely held theory of their occurrence is that of failure to undergo complete separation of the embryonic disc at around the 15th to 17th day of gestation (fissure theory). An alternative postulation, by Spencer, is that secondary fusion occurs between two Originally separate monovular embryonic discs (fusion Theory). The twins are monozygotic, monoamniotic, and monochorionic.

The twins are classified according to the most prominent site of fusion, plus 'pagus', the Greek term for fixed: the thorax (thoracopagus), abdomen (omphalopagus), sacrum (pygopagus), pelvis (ischiopagus), skull (craniopagus), face (cephalopagus), or back (rachipagus).

Fetal US is the main prenatal screening imaging modality. Antenatal US is capable of diagnosing Conjoined twins as early as 12 weeks of gestation. The Characteristic imaging features, apart from direct identification of a tissue bridge or fused anatomical structures, is the fact that the twins move in a fixed position in relation to each other. Exact and detailed prenatal diagnosis of conjoined twins is essential for optimal obstetric management and parental counseling.

Fetal ECHO assessment of the heart needs to be detailed in all patients as there is an increased incidence of congenital heart disease in conjoined twins overall. A particularly detailed cardiac assessment is vital in thoracopagus twins as a shared heart is seldom compatible with life and usually an indication for termination of the pregnancy.

Recently 3D US imaging has been advocated as a new tool to demonstrate the extent of fusion in conjoined twins. 3D US may add anatomical information and thus improve the accuracy of classification of individual twins. In some cases the category of conjoined twins is suspected by 2D scanning, but can be better appreciated on 3D US. 3D sonography also provides images that are easier for parents to understand, which can help them with decision making

MRI may be an essential tool in the prenatal and postnatal diagnostic workup of the exact fetal anatomy in order to plan the surgical approach. The high spatial resolution and different tissue contrasts that can be generated by fetal MRI can give important and detailed anatomical information regarding the fetal anatomy. The recent introduction of fast imaging techniques in fetal MRI, allow minimal image degradation by fetal motion, thereby producing high-quality images. The high spatial resolution of the fetal MRI is sufficient to confirm the diagnosis made on prenatal US and is able to rule out additional fetal malformations. There are no reported adverse biological effects of MRI, which is thus far considered a safe imaging modality in pregnancy.

Accurate antenatal assessment allows the parents to be counseled in depth as to the likely outcome of the pregnancy and the chances of postnatal separation and survival. Most conjoined twins are delivered at 36–38 weeks gestation by elective cesarean section, often at centers where appropriate obstetric and pediatric surgical facilities are readily available.

The successful completion of a conjoined twin pregnancy, with the birth and successful subsequent separation of the conjoined twins, requires an extensive interdisciplinary team effort involving obstetricians,

anesthesiologists, pediatric surgeons, neonatologists and pediatric radiologists.

The choice of postnatal imaging depends to some degree on the site of fusion. Nevertheless, all twins inevitably have chest and abdominal radiography for an overall general Assessment. Unexpected Diaphragmatic hernia or vertebral anomalies can thus be detected early.

All neonates should have routine cerebral US, and probably spinal US also, as baseline investigations. In addition, abdominal US to assess the liver, to document the presence of two spleens, gallbladders, biliary systems, kidneys and bladders, is necessary. Detailed Doppler studies to evaluate the great vessels in the abdomen and hepatic venous drainage should also be performed, ECHO is mandatory for every twin due to the high frequency of congenital heart disease in all types of conjoined twins. 3D ECHO has been advocated postnatally as it may facilitate an easier understanding of the cardiac connections.

Radiologists avoid CT where possible in infancy due to the high radiation burden. Conjoined twins, even when stable and asymptomatic, are a well-justified exception. Due to the high spatial resolution and speed in particular of multidetector CT (MDCT).

MRI has an increasing role in the postnatal evaluation of conjoined twins, particularly those joined at the head or thorax. MR has the capability of producing 3D reconstructed images in any direction with much improved resolution and tissue characterization than 3D US. MR is the optimum examination to assess for any cortical fusion in craniopagus twins. Conventional images after IV gadolinium administration is utilized in the postnatal imaging of conjoined twins.

Intracardiac anatomy, great vessel anatomy and blood flow, and ventricular wall motion can all be accurately assessed in thoracopagus cases. Many complex fusions will inevitably get both MRI and MDCT. These modalities are often complementary, with MRI showing soft tissue anatomy to best effect and CT detailing complex bony anatomy in pelvic conjunction.

The management of conjoined twins can be divided into Four separate time frames.

1. ***Prenatal:*** In particular, termination would be recommended in the event of complex cardiac fusion in thoracopagus twins or extensive cerebral fusion in craniopagus twins. Therefore, detailed echocardiography and accurate ultrasonography complemented as necessary with MRI scanning is essential. An informed decision can be made either to terminate or to proceed with the pregnancy. Where the decision is to proceed with the pregnancy, delivery should be planned to take place at or close to the surgical unit where separation will be performed. Delivery is always by caesarean section at 36–38 weeks' gestation.
2. **Non-operative treatment:** should be considered in the presence of complex cardiac fusion or where there would be a severe unacceptable deformity following separation.
3. ***Emergency separation:*** is undertaken when one twin is dead or dying, threatening the survival of the remaining twin, or where a life-threatening correctable congenital abnormality (e.g. intestinal atresia, malrotation with or without volvulus, ruptured omphalocele, or anorectal agenesis) is present in one or both twins.

4. ***Elective separation:*** It allows the twins to stabilize and thrive and provides time to carry out detailed investigations to define the nature and the extent of union. Detailed planning of the operative procedure with all members of the operating team should take place before the separation.