

MRI of omphalopagus conjoined twins with a Dandy-Walker malformation: prenatal true FISP and HASTE sequences

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ABSTRACT

Conjoined twins are an extremely rare congenital malformation without any known genetic predisposition. Omphalopagus twins are the second most common variety of conjoined twins and usually are joined at the umbilicus. We present omphalopagus conjoined twins demonstrated with true FISP (fast imaging with steady-state procession) and HASTE (half-Fourier acquisition single-shot turbo spin-echo) magnetic resonance imaging (MRI) sequences, which showed Dandy-Walker malformation in one of the pair. To our knowledge, this is the first case of conjoined twins with this malformation, which was diagnosed with ultrafast MRI.

Key words: • twins, conjoined • magnetic resonance imaging • Dandy-Walker malformation

Conjoined twins (formerly known as Siamese twins) are the result of a very rare congenital anomaly, the incidence of which is estimated to be around one per 250,000 live births (1). The classification is based on the most prominent site of anatomical connection. The anatomic location is followed by the designation “pagus”, from the Greek word meaning something fixed or solid. Thoracopagus is the most common variety of these twins (2). Omphalopagus twins are the second most common variety, usually joined at the umbilical region (3). Pelvic bones are usually separate in this type. Although ultrasound (US) is widely used for the diagnosis of conjoined twins, magnetic resonance imaging (MRI) is the best modality to provide detailed imaging of fetuses with complex anomalies, especially in late pregnancy. Ultrafast imaging sequences have revolutionized fetal MRI, especially in neuroradiological examination (4). We present a case of omphalopagus conjoined twins with a Dandy-Walker malformation evaluated by ultrafast MRI sequences.

Case report

A 26 year-old woman, gravida 2, para 1, at 24 weeks of gestation was referred to our radiology department with a diagnosis of conjoined twins. Her history was unremarkable. A pair of twins was diagnosed antenatally by fetal ultrasound, which revealed that they were joined at the abdomen and pelvis. A single posterior placenta with polyhydramnios was identified. A single artery and a single vein in the umbilical cord were noted on color Doppler sonography. At 24 and 32 weeks of gestation, MRI of the fetuses was performed using 1.5 T MRI system (Symphony, Siemens, Erlangen, Germany) with coronal and axial images that were obtained using T2-weighted fast imaging with steady-state procession (true FISP) and half-Fourier acquisition single-shot turbo spin-echo (HASTE) sequences to establish the diagnosis, and to better delineate the anatomic features.

The study protocols were: TR/TE/flip angle/matrix/slice number/acquisition time, 81 ms/5.5 ms/80°/256x156/40/25/24 s for true FISP; and 1100 ms/120 ms/150°/256x156/40/44 s for HASTE.

Conjoined twins were visualized successfully, and were observed to be joined from the abdomen to the pelvic area (Fig. 1). The liver was shared, and the pelvic bones were conjoined. The four legs were connected with the conjoined pelvis. Gallbladder drainage and hepatic venous drainage were separate. The fetuses had two separate chests, urinary tracts, and gastrointestinal tracts. Although polyhydramnios was visualized by the first MRI, at the second examination the amniotic fluid was diminished.

In Fetus 1, the posterior fossa was filled with cerebrospinal fluid, as visualized on MRI examination. The cerebellum, vermis, and corpus callosum were not seen. The third and lateral ventricles were enlarged. The

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Received 10 March 2008; accepted 7 April 2008.

Published online 21 October 2009
DOI 10.4261/1305-3825.DIR.1747-08.0



Figure 1. Coronal true FISP MR image of conjoined omphalopagus twins at 24 weeks of gestation shows fusion of the abdomen at the level of the umbilicus. Dandy-Walker malformation (*star*) in Fetus 1, shared liver (L), polyhydramnios, and single umbilical artery (*arrow*) are seen. Increased nuchal thickness is also demonstrated next to the umbilical artery.

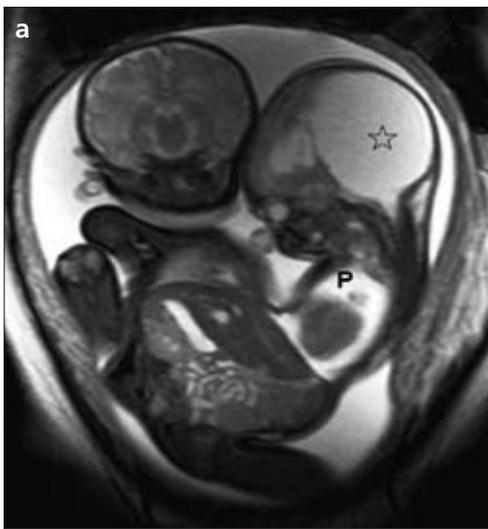


Figure 2. a, b. Coronal true FISP (a) and HASTE (b) MR images at 32 weeks of gestation. Cystic dilatation of the posterior fossa (*star*) is larger than before. Pleural fluid (P) is seen around the heart.



tentorium was elevated. Dandy-Walker malformation was diagnosed with these findings. Pleural fluid and total atelectasis were clearly demonstrated

with US and MRI. Nuchal thickness was 1 cm. The head and thorax of the second fetus were completely normal (Figs. 2, 3).

At 35 weeks of gestation, when the patient began active labor, caesarean section was performed. The twins were joined from the level of the umbilicus to the pelvic bones. One penis and scrotum, and bilaterally undescended testes were noted at birth (Fig. 4). Club foot deformity was seen in both fetuses. Surgical separation was performed under emergency conditions by a pediatric surgeon. Fetus 1 died during this operation and Fetus 2 died from cardiac failure after the operation.

Discussion

Conjoined twins result from a separation defect in the embryonic plaque between the 13th and 17th days of gestation. They are monozygotic, monoamniotic, and monochorionic, and are classified according to the site of fusion: thoracopagus (thorax), omphalopagus (abdomen), pygopagus (sacrum), ischiopagus (pelvis), cranio-pagus (face) or rachipagus (back). In addition, conjoined twins may be classified as asymmetric (heteropagus) or symmetric.

Omphalopagus twins are usually joined in the front at the level of the umbilicus, commonly involving the lower thorax (3). Omphalopagus twins account for 18–33% of all conjoined twins. Liver fusion occurs in 80% of cases. The pericardium may be common, but the heart is never shared. Although ischiopagus twins are usually joined from the level of the umbilicus to the pelvis, the lower gastrointestinal tract is affected in 70%, and the genitourinary tract is affected in 50% of twins with this anomaly. In addition to these findings, end-to-end union of the infants joined at the pelvis may be seen in ischiopagus twins (1).

Most conjoined twins are delivered in the 36th–38th weeks by caesarean section after the lungs have matured; however, many are born prematurely, as in our case. After birth, fetuses are evaluated for cardiovascular problems, fusion of thoracoabdominal organs, and organ anomalies by imaging modalities such as echocardiography, US, computed tomography, MRI and barium studies. Scintigraphy can be performed for the biliary tree. Morbidity and mortality rates are high in conjoined twins even if they are admitted to a neonatal intensive care unit. In our case, caesarean section was performed

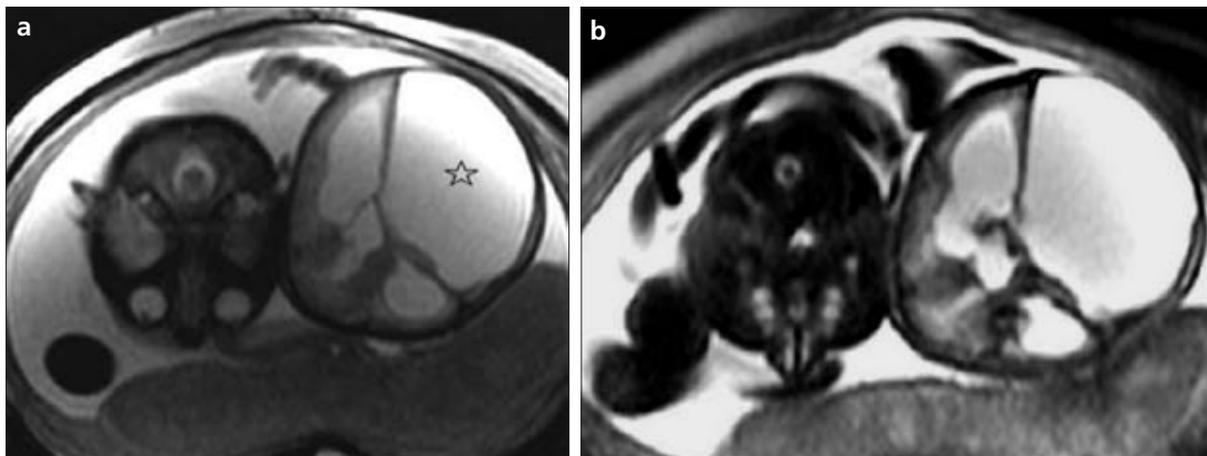


Figure 3. a, b. Axial true FISP (a) and HASTE (b) MR images demonstrate the Dandy-Walker malformation.



Figure 4. Postnatal image of omphalopagus twins shows genital malformation (curved arrow) and club foot deformities (straight arrow).

at 35 weeks. Prior to the delivery, we were not able to perform any roentgenographic studies or nuclear medicine examinations.

Prenatal sonography is the primary imaging modality in pregnancy. It usually permits accurate diagnosis of congenital anomalies. Ultrasonography has a wide range of uses, and provides good results in detection of cranial anomalies, despite some limitations. The limitations of US are mostly due to an inability to visualize fetal intracranial anomalies secondary to reverberation artifacts of the calvarium, and low sensitivity for the detection of cerebral cortical malformations, as well as small destructive lesions of the cerebrum and cerebellum.

In later pregnancy, especially if there is maternal obesity or oligohydramnios, MRI is superior to US for overall fetal assessment. Recently, prenatal MRI also has been advocated when one twin is very unlikely to survive (5) such as in our case. Ultrafast T2-weighted sequences of short duration, such as single-shot fast spin-echo MR sequence (HASTE) allows minimal image degradation by fetal motion and high quality images of fetal organs without the need for fetal or maternal sedation (6). It has been used in most fetal imaging studies (1, 3, 7). However, the true FISP imaging method, based on gradient echo, offers an excellent signal-to-noise ratio with short acquisition times, which makes it appealing for fetal MRI

applications (8, 9). T2-weighted true FISP imaging methods are very useful in ultrafast fetal imaging. Usually, each slice is performed in 1 second, and the full sequence in each imaging plane requires 15–25 s, without breath-holding technique. Manganaro et al. suggested that true FISP has unique advantages over HASTE in fast fetal imaging (9). We used both sequences in our case. Contrast resolution of true FISP was slightly better than HASTE images, especially in the detection of the fetal body and the umbilical cord. Dandy-Walker malformation was seen clearly on both sequences.

Cardiac defects, abnormal pulmonary and hepatic venous drainage, congenital diaphragmatic hernias, intestinal atresia, neural tube defects, cystic hygroma, urologic anomalies such as renal dysplasia and double collecting system, and orthopedic anomalies such as hip dislocation, club foot, vertical talus, and scoliosis can be seen in conjoined twins (10). Cerebrospinal anomalies such as dysgenesis of the corpus callosum, encephalocele, and syringomyelia have been reported in conjoined twins (4, 11). In our case, genital anomalies (single penis, undescended testes), orthopedic anomaly (club foot), umbilical cord anomaly (single artery) and Dandy-Walker malformation were seen. Dandy-Walker malformation was demonstrated in the posterior fossa of Fetus 1.

Dandy-Walker malformation, first described in 1914, is a rare congenital malformation involving the cerebellum and fourth ventricle. This condition is characterized by agenesis or hypoplasia of the cerebellar vermis, cystic dilatation of the fourth ven-

tricle, and enlargement of the posterior fossa. Approximately 70–90% of patients have hydrocephalus, which often develops postnatally. Dandy-Walker malformation may be associated with atresia of the foramen of Magendie and, possibly, the foramen of Luschka. Rarely has this syndrome been reported using fetal MRI examination (12). The occurrence of a Dandy-Walker malformation in one of the conjoined twins is probably coincidental and has not been reported previously in the literature.

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