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Omphalopagus conjoined twins: ultrafast MR imaging findings

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ABSTRACT
Conjoined twins are a rare and often catastrophic obstetrical event. Although ultrasonography is widely used in the diagnosis of conjoined twins, it may fail to demonstrate the details in fetuses with complex anomalies, especially during late pregnancy. We present an omphalopagus conjoined twins case evaluated by HASTE magnetic resonance imaging, which showed the conjunction site and cranial anomalies, and aided antenatal counseling and neonatal surgical planning.

Key words: • *twins, conjoined* • *magnetic resonance imaging*

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Conjoined twins are a rare anomaly accompanied by severe complications. Parts of the cardiovascular and gastrointestinal systems are usually shared between the twins (1). Classification is made according to Latin names of joined regions. Omphalopagus twins are usually joined at the umbilical region involving the lower thorax (2). Although ultrasound (US) is widely used for the diagnosis of joined twins, it can be unsuccessful in providing detailed imaging of fetuses with complex anomalies in the late pregnancy period. Magnetic resonance (MR) imaging is an imaging modality that has no known harmful or ionizing effect on fetuses. Before ultrafast scanning methods, MR imaging had limited benefit for antenatal diagnosis due to artifacts secondary to fetal movements (3–5). Fetal movements can be slowed down by sedation for a fetal MR imaging examination. Ultrafast MR imaging provides image acquisition within seconds and does not require sedation. In this study, we present an omphalopagus case examined with half-Fourier acquisition single-shot turbo spin-echo (HASTE) MR imaging. Conjoined parts and cranial anomalies of the fetuses were imaged by HASTE MR imaging for antenatal counseling and surgical planning.

Case report

A 25-year-old woman (gravida II, para I, living 0), who had previously given birth to a boy with anomalies that included tracheoesophageal fistula, esophageal atresia, tetralogy of Fallot, polydactyly, renal agenesis, and outer ear agenesis (i.e., VACTERL) was admitted to our hospital. The present pregnancy of the patient was uneventful until the 32nd week. At that time, the patient was referred to our hospital with a presumed diagnosis of encephalocele by an obstetrician and for fetal echocardiography, the patient was sent to a pediatric cardiologist. Since two separate fetal heart activities were found, the patient was referred to the radiology department for obstetric US due to a possible diagnosis of twin pregnancy.

Obstetric US revealed the presence of a conjoined twin pregnancy; the fetuses were joined at abdomen and they were sharing the same liver. A cystic lesion was detected in one of the fetuses (Fetus 1), but differentiation of the cystic hygroma or encephalocele could not be made. Because of this situation, and in order to detect other possible accompanying anomalies, a fetal MR imaging examination was performed with a four-element phased array surface coil in a 1.5 T superconductive system (Siemens, Erlangen, Germany). The whole-body specific absorption rate was lower than 3.0 W/kg. HASTE MR imaging was applied at axial, coronal, and sagittal planes (TE, 60 msec; slice thickness, 4 mm; FOV, 26 x 35 cm; matrix, 192 x 256). In order to avoid radiofrequency loading, a repeat focusing pulse of 130° was used. A single study of thirteen slices was performed in only 17 seconds.

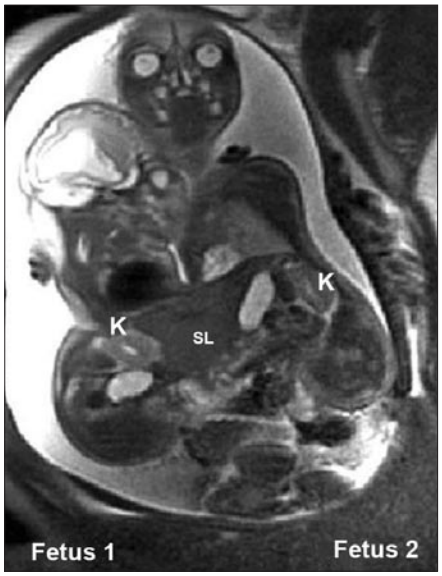

Figure 1. Coronal HASTE MR image of conjoined twins shows shared liver. K: kidney; SL: shared liver.

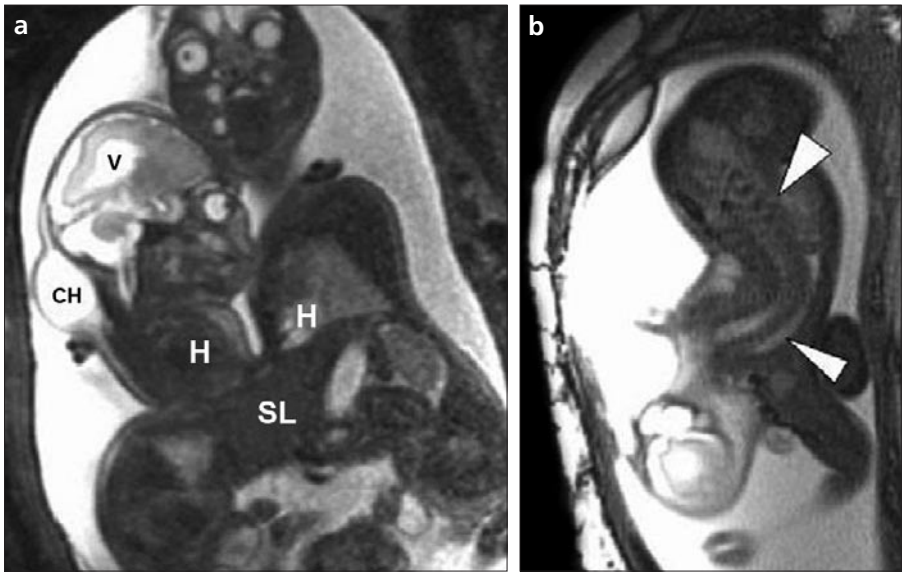

Figure 2. a, b. Coronal HASTE MR image **(a)** shows ventricular dilatation and cystic hygroma in Fetus 1; two separate chests and intrathoracic structures; and shared liver. H: heart; V: ventricle; CH: cystic hygroma; SL: shared liver. Sagittal HASTE MR image **(b)** shows kyphoscoliosis in Fetus 1 *(arrows)*.

The conjoined twins were visualized successfully and seen as joined at their abdomen on midline with a common liver (Fig. 1). Although the liver was shared, there were separate gallbladders and hepatic venous drainages. The fetuses had 2 separate chests, kidneys, and urinary bladders. Stomach and colonic segments were separate, but intestinal loops were directed from one twin to the other. In Fetus 1, corpus callosum dysgenesis, cystic hygroma on the posterior side of the neck, ventricular dilatation, and significant kyphoscoliosis were detected (Fig. 2). Fetus 2 was completely normal, except for the common liver.

Caesarian section was performed with spinal anesthesia at 37.5 weeks of gestation. The lungs of fetus 1 were not aereted, and both a single ventricle and a big atrial septal defect were detected in echocardiography. Separate intestinal systems were demonstrated on barium studies. Liver anatomy-vasculature and the biliary system were evaluated by MR imaging and MR angiography. On the second day after birth, as vital findings of the twins were stable, surgical separation was performed. Fetus 1 died due to cardiac arrest during the operation, Fetus 2 was lost due to intestinal ischemia 12 hours after the operation.

Discussion

Conjoined twin cases are rare, but the actual prevalence is not known. The prevalence in the literature has a broad range: 1/30,000–1/200,000 (1, 6–8). Those twins are monozygotic, monoamniotic, and monochorionic. Between the 13th and 17th days of gestation, separation defect occurs in the embryonic plaque (7). Conjoined twins are classified as thoracopagus (thorax), omphalopagus (abdomen), pygopagus (sacrum), ischiopagus (pelvis), craniopagus (cranium), cephalopagus (face), or rachipagus (back), according to fusion site.

Cardiac defects, abnormal pulmonary and hepatic venous drainage, congenital diaphragm hernia, intestinal atresia, neural tube defects, cystic hygroma, urologic anomalies like renal dysplasia and double collecting system, orthopedic anomalies like hip dislocation, club foot, vertical talus, and scoliosis can be seen in conjoined twins (8, 9).

Omphalopagus twins are joined at the front and umbilicus level, commonly involving the lower thorax. Liver fusion occurs in 80% of cases (6). The pericardium may be common, but the heart is never shared (2). Prenatal diagnosis of omphalopagus can be established in the 8th gestational week by transvaginal US. The suggested time for evaluation is between the 11th and 12th weeks (3). Ultrasonography has a wide use and it 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 and small destructive lesions of the cerebrum and cerebellum. In addition to these, imaging quality of US can diminish due to obesity of the mother, olygohydramnios, and engagement of fetal head in late pregnancy (10).

In fetal MR imaging, patients are not exposed to ionizing radiation and there is no evidence for any teratogenic side effect of MR imaging; but, because of significantly low image quality due to motion artifacts during long examination times, usage of routine intrauterine MR imaging for fetuses is limited (11). Technical developments in ultrafast MR imaging, especially HASTE and single-shot fast spin-echo sequences led to the increased the use of MR imaging in prenatal diagnosis. HASTE MR imaging sequences are now used for the detection of central nervous system anomalies as they provide perfect contrast between cerebral spinal fluid, brain, and spinal cord. Additionally, some investigators successfully used ultrafast MR imaging for the evaluation of herniation of the liver into the thorax in fetuses with congenital diaphragm hernias. These researchers also used ultrafast MR imaging for the differentiation of cystic adenomatoid malformations from bronchopulmo-

nary sequestration and for the evaluation of potential airway obstruction in fetuses with large neck masses (12, 13). As in our case, it is difficult to determine structure and organs of conjoined twins with US in the second and third trimesters of gestation; therefore, we performed MR imaging examination with a HASTE sequence. In Fetus 1, MR imaging clearly detected cranial anomalies (corpus callosum dysgenesis and ventricular dilatation) that were not determined by US. Additionally, HASTE MR imaging provided differential diagnosis of cystic hygroma and encephalocele by showing the lack of their relationship with intracranial structures. Moreover, scoliosis, absence of thoracic fusion, width of abdominal fusion of Fetus 1, and abdominal structures of both fetuses were determined.

Most conjoined twins are delivered in the 36–38th weeks by caesarian section after the lungs have matured (8, 9). After birth, fetuses are evaluated for cardiovascular problems, fusion of thoracoabdominal organs, and organ anomalies by US, echocardiography, barium studies, angiography, computed tomography, and MR imaging. Tc 99m-HIDA scintigraphy can be performed to obtain dynamic information about the biliary tree (9, 14).

Morbidity and mortality rates are high in conjoined twins despite developments in techniques of radiological imaging, anesthesia, and surgery. Prognosis depends on fusion site, complexity, and spread of shared organs, and accompanying anomalies. Only 40%–60% of conjoined twins are delivered alive, and of those, 35% are lost in first 24 hours (1, 9).

In conclusion, HASTE MR imaging can be a perfect complementary of US in complex anomalies like omphalopagus, as it was in the presented case. The detailed anatomic information obtained by the method can guide the surgical planning and can aid antenatal counseling.

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