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Heteropagus twins: report of two cases

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Title figure:
Parasitic twin 1686 (source: www.fineartamerica.com)

Heteropagus twins are an extremely rare form of asymmetrical conjoined monochorial monoamniotic twins with an estimated incidence of less than one per one million live births (1). An often-used synonym is «parasitic twins». The term heteropagus describes twins in which one of them has a mostly intact body that is able to survive and which is referred to as «autosite», while the counterpart, referred to as «parasite», is only rudimentarily developed being physically attached to and nourished by the other twin.

CASE REPORT 1

This male infant was born at 37 6/7 weeks of gestation to a 37-year-old G4/P4 by Cesarean section because of known fetal malformations. The family had three healthy children. The pregnancy following spontaneous conception was uneventful. At 32 weeks of gestation, a prenatal ultrasound scan revealed an omphalocele with herniation of liver and bowel. The diagnosis was confirmed by fetal magnetic resonance imaging. The parents rejected invasive genetic investigation.

After delivery, the baby adapted well. Apgar scores were 9, 9 and 9 at 1, 5 and 10 minutes, respectively, and the arterial umbilical cord blood pH was 7.37. Birth weight was 3130 g (P25-50), length 46 cm (<P3) and head circumference 34.5 cm (P25-50). Physical examination showed an omphalocele and, in addition, a thoracoabdominal mass with rudimentary limbs and male genitalia (Fig. 1 – 2). The baby was transferred to a pediatric surgery unit for further treatment. Resection of the heteropagus, reduction of the omphalocele as well as closure of the abdominal and thoracic wall, were performed on the first day of life. The baby recovered well and has since shown normal growth and development.

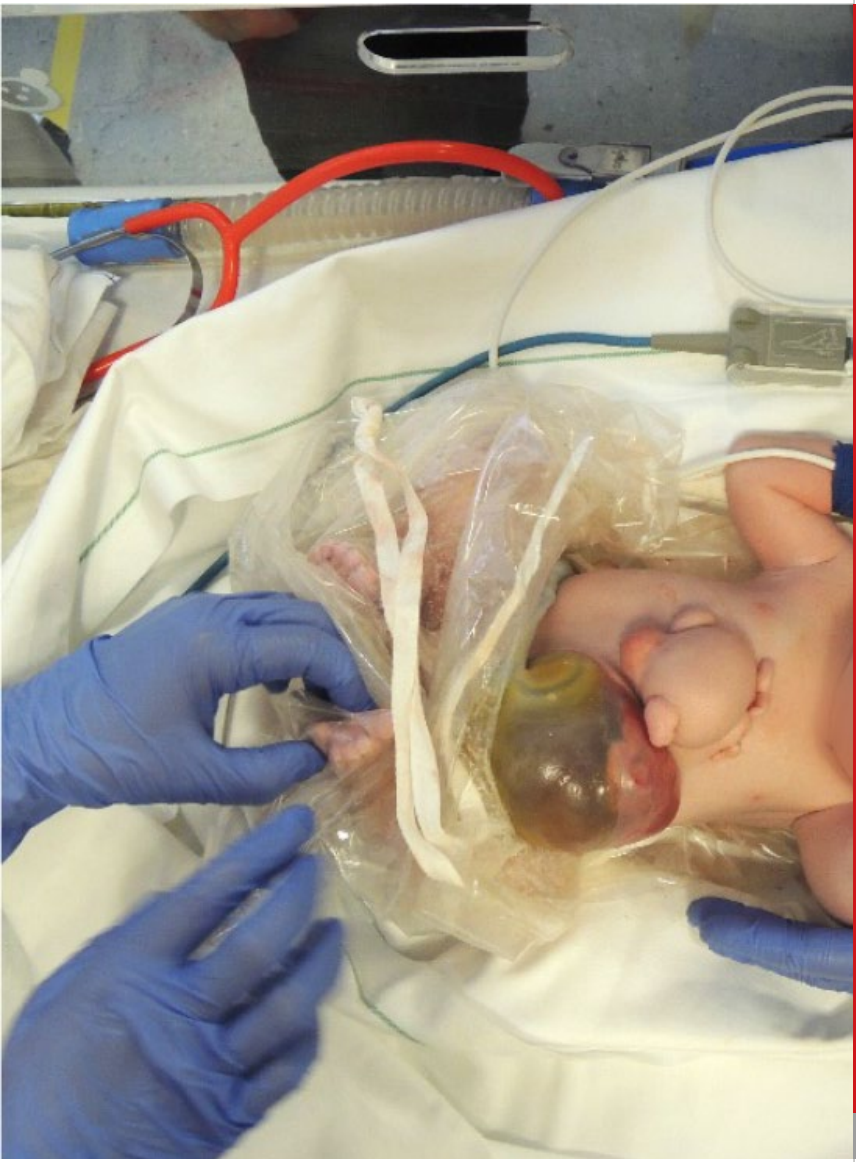

Fig. 1

Patient 1 shortly after delivery: omphalocele, heteropagus twin with rudimentary limbs protruding from the epigastric area.

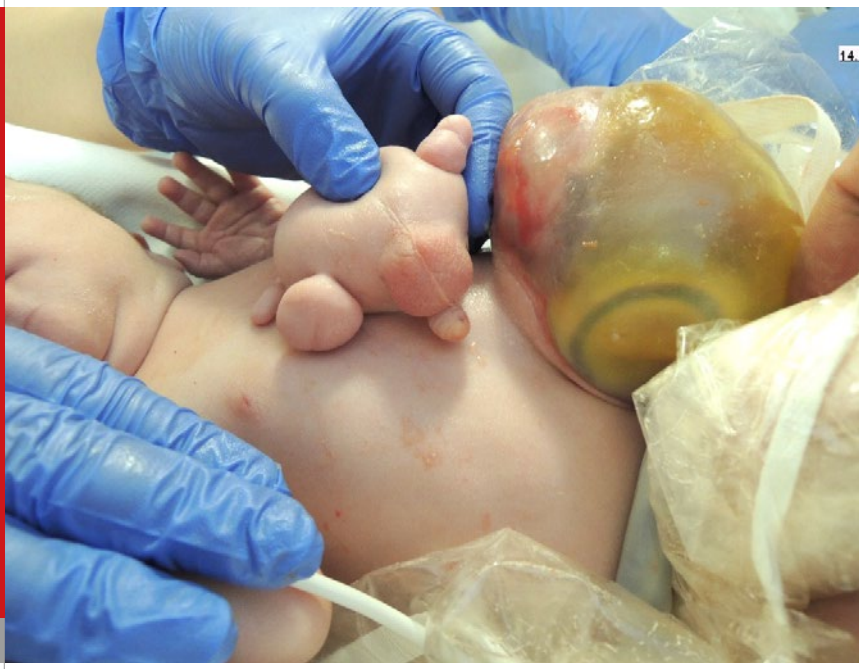

Fig. 2

Patient 1 shortly after delivery: omphalocele, heteropagus twin with rudimentary limbs and male genitalia.

This male infant was born at 37 2/7 weeks of gestation to a 28-year-old G2/P2 by Cesarean section. The family has one healthy daughter and there was no history of congenital anomalies. Except for a bicornuate uterus, the mother was in good health. At 22 weeks of gestation, an ultrasound examination at a gynecology practice revealed a suspected omphalocele. Findings on another ultrasound examination performed at the Department of Obstetrics at the University Hospital of Zurich were felt to consistent with a hernia into the umbilical cord. Amniocentesis revealed a normal male karyotype. Fetal MRI showed polymelia with four legs, two of them hypoplastic, without evidence of a spinal defect.

After birth, the boy adapted well with a Apgar scores of 8, 9 and 9 at 1, 5 and 10 minutes, respectively. The birth weight was 3750 g (P 90-95), length 48 cm (P 10-25) and head circumference 36.5 cm (P 90-95). On physical examination (Fig. 3 – 5), there were a small omphalocele with a few intestinal loops, polymelia with four legs with a hypoplastic ventral pair of legs without any spontaneous movements and a normally developed dorsal pair of legs with spontaneous movements, as well as a pes equinovarus of the right foot. There were two sets of male genitalia located lateral to the legs with a hypoplastic medial scrotum on each side and anal atresia. Micturition was observed from both penises.

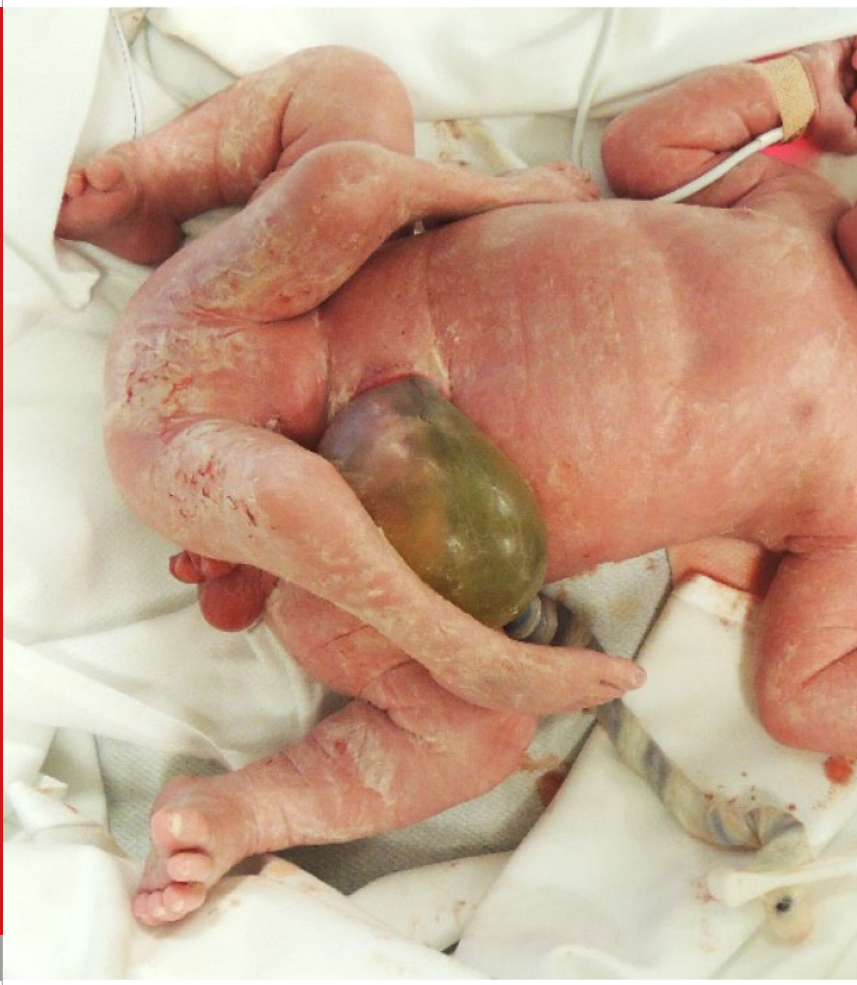

Fig. 3

Patient 2 shortly after delivery: small omphalocele, polymelia with four legs; the ventral pair is hypoplastic without spontaneous movements.

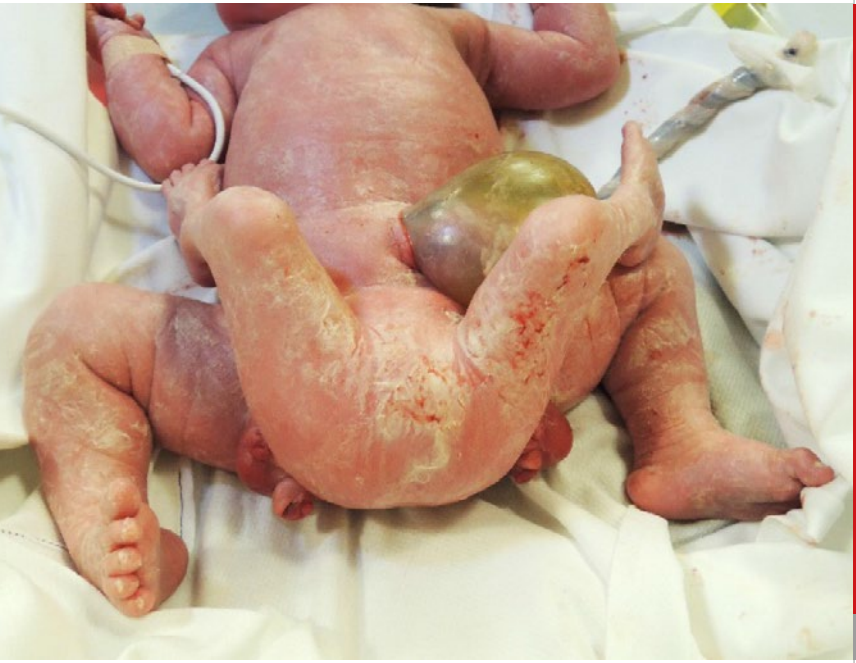

Fig. 4

Patient 2 shortly after delivery: polymelia with four legs, clubfoot deformity of the right foot of the dorsal pair of legs.

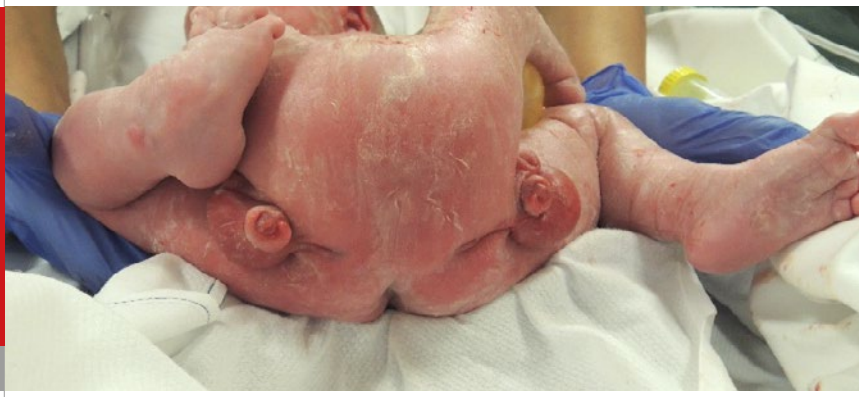

Fig. 5

Patient 2 shortly after delivery: polymelia with four legs, two sets of male genitalia and anal atresia.

For further treatment the baby was transferred to a pediatric surgery unit. Imaging studies (Fig. 6 – 8) showed a fusion of two pelvises, a third kidney that belonged to the parasite (which concentrated contrast agent), along with two other kidneys that drained urine into one common urinary bladder. Furthermore there were two urethrae. A branch of the internal iliac artery provided the vascular supply to the parasite. The content of the omphalocele was found to be intestinal loops belonging to the parasite.

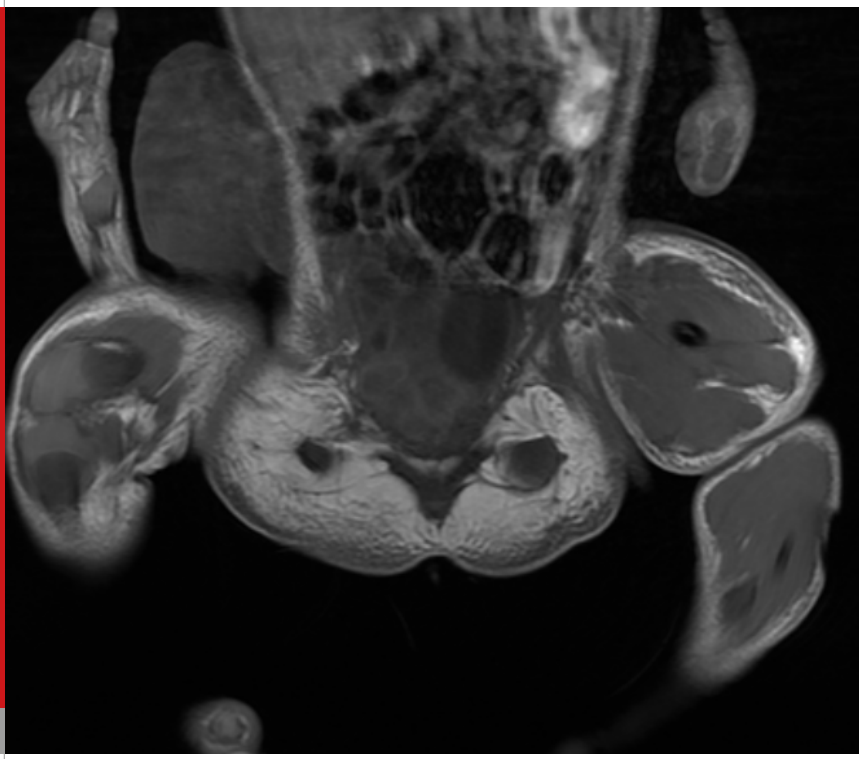

Fig. 6

MRI of the pelvic region of patient 2: fat and bone is the predominant tissue of the parasite.

Fig. 7

MRI of the pelvic region of patient 2:
3D reconstruction.

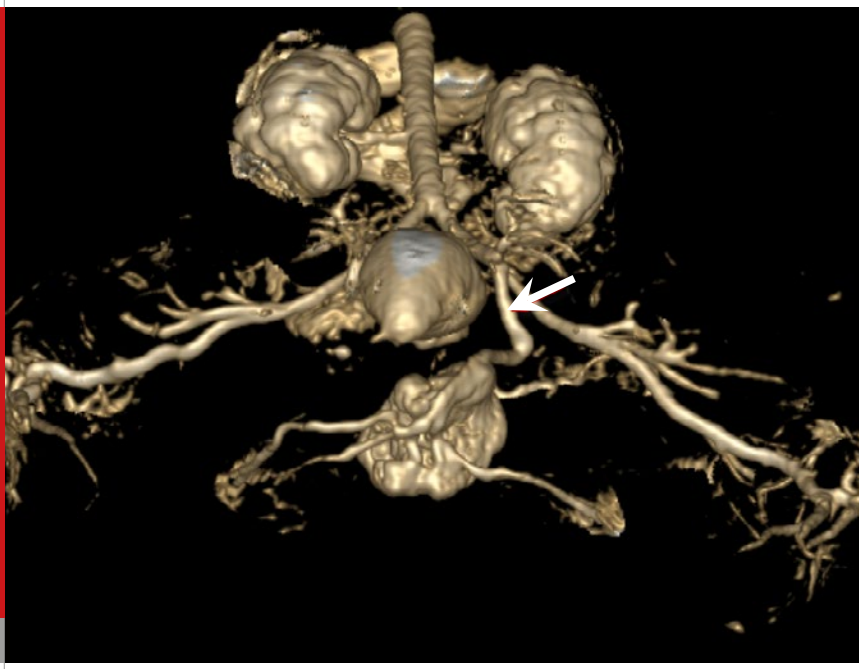

Fig. 8

MR angiography of patient 2: a branch of the internal iliac artery (arrow) supplies the parasite.

The first surgery was performed on the second day of life with full resection of the omphalocele, closure of the abdominal wall and construction of a dismembered colostomy. The pes equinovarus was redressed and placed in a cast. In additional surgical interventions, the hypoplastic legs and the parasitic kidney were removed and the external genitalia repositioned. The patient's anal atresia has not yet been corrected.

DISCUSSION

Heteropagus twins are extremely rare and represent only 1 to 2 percent of all conjoined twins (2). Most authors mention a clear predominance of the male sex with about 78% being males (2, 4). Until now, no particular risk factors for developing heteropagus twins have been reported. The most frequent form observed is the epigastric heteropagus, also called omphalopagus. Less common are ischiopagus, rachipagus and craniopagus (1).

The cause of conjoined twins and therefore of heteropagus twins is not fully clarified. A widely accepted theory suggests an incomplete fission of one zygote, which occurs about 14 days after fertilization. Logroño et al. suggest the alternate hypothesis that heteropagus twins originate from the fusion of two separate zygotes at a later stage. In a similar way, Ratan et al. mention a fusion theory, also called Ratan's theory. It is postulated that an unbalanced distribution of the placental blood leads to an ischemic insult in the parasite twin with selective atrophy of this twin as a consequence (1, 2).

There are only few reports of cardiac tissue found in the parasitic twin who is usually acephalic (1). A differential diagnosis for this medical finding is the TRAP (twin reversed arterial perfusion) sequence, which also occurs in monochorionic twins. In this case, the twins are separated from each other, but have an abnormal arterioarterial anastomosis in the placenta that sup-

ports the development of an abnormal (acardiac) twin with severe and lethal developmental abnormalities. The abnormal twin is reversely perfused with deoxygenated blood through this anastomosis by the apparently normal (pump) twin (6-9).

In heteropagus twins, the parasite's limbs normally do not show any spontaneous movements. This is explained by the absence of neural innervation of the parasite myoblasts, which leads to incomplete differentiation and consecutively to skeletal muscular atrophy. Therefore the histology shows predominantly fat and bone but no muscle tissue (1, 2, 4, 5),

The autosites often have malformations themselves. Most common among these are congenital heart defects, especially ventricular septal defects (1). Furthermore, the pumping twin can suffer from congestive heart failure due to the additional load on the cardiovascular system that has to supply the parasite twin as well. There is also a high incidence of associated omphalocele, especially in epigastric heteropagus twins (2, 4). Spinal cord defects are less commonly seen (1).

Bony and visceral connections between the heteropagus twins can occur but are rarely observed. More frequently residues of parasite organs within the body of the autosite have been described. Vascular communications seem to be less marked than in symmetrical

conjoined twins, making surgical separation less complicated (1, 2).

The prognosis regarding survival of the autosite is good. In a review of cases by Sharma et al., the mortality rate was calculated to be 31 percent (1). In order to reduce morbidity and for a good esthetic and functional results, a pregnant woman with the diagnosis of heteropagus twins should be referred to a perinatal center with experience in treating this disorder and be managed by a multidisciplinary team. Due to the rarity of this malformation, there is only limited literature available, mainly consisting of case reports.

1. Sharma G, Mobin SS, Lypka M, Urata M. Heteropagus (parasitic) twins: a review. J Pediatr Surg 2010;45:2454-2463 *(Abstract)*

2. Abubakar AM, Ahidjo A, Chinda JY, et al. The epigastric heteropagus conjoined twins. J Pediatr Surg 2011;46:417-420 *(Abstract)*

3. Logroño R, Garcia-Lithgow C, Harris C, Kent M, Meisner L. Heteropagus conjoined twins due to fusion of two embryos: report and review. Am J Med Genet 1997;73:239-243 *(Abstract)*

4. Xie JT, Zhou L, Yang ZL, Sun HY. Epigastric heteropagus conjoined twins: two case studies and associated DNA analysis. Clinics (Sao Paulo) 2012;67:527-529 *(Full Text)*

5. Hager J, Sanal M, Trawöger, et al. Conjoined epigastric heteropagus twins: excision of a parasitic twin from the anterior abdominal wall of her sibling. Eur J Pediatr Surg 2007;17:66-71 *(Abstract)*

6. Berg C, Holst D, Mallmann MR, Gottschalk I, Gembruch U, Geipel A. Early vs. late intervention in twin reversed arterial perfusion sequence. Ultrasound Obstet Gynecol 2014;43:60-64 *(Abstract)*

7. Prasad RH, Prasad TR, Kumar KD. TRAP sequence – an interesting entity in twins. J Clin Imaging Sci 2012;2:56 *(Abstract)*

8. Chalouhi GE, Stirnemann JJ, Salomon LJ, Essaoui M, Quibel T, Ville Y. Specific complications of monochorionic twin pregnancies: twin-twin transfusion syndrome and twin reversed arterial perfusion sequence. Semin Fetal Neonatal Med 2010;15:349-356 *(Abstract)*

9. Malhotra N, Sinha A, Deka D, Roy KK. Twin reversed arterial perfusion: report of four cases. J Clin Ultrasound 2004;32:411-414 *(Abstract)*

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