Parasitic conjoined twins are extremely rare. The true incidence is not known but is reported as 1-2% of all conjoined twins (1, 2). In parasitic twining one baby is fully formed (autosite) whereas the other baby is incomplete (parasite) and is dependent upon the autosite for its nutrition. Management of babies with parasitic twining may look easy but the autosite usually has high incidence of associated cardiac anomalies and it may alter the outcome of surgery in these babies. We reviewed three pairs of parasitic epigastric heteropagus twins (EHT). First pair of twins had transpositioning of great vessels along with VSD and ASD. Parasite had fully formed pelvis and lower limbs but the upper limbs were partially formed with rudimentary spine from where it was attached to the autosite in the epigastrium. Babies had successful separation of the parasite but died after 8 months due to cardiac complications. Second pair of EHT had very similar appearance and had a soft systolic murmur suggesting congenital cardiac anomalies. Baby was taken abroad by volunteers and ultimate fate is not known. Third pair of twin was brought in sepsis and died within two days before full evaluation and any surgical intervention. Early referral and availability of pediatric cardiac services may improve ultimate outcome in many such babies.

Key words: conjoined, parasitic, heteropagus, twins.

Introduction
Parasitic conjoined twins are extremely rare. The true incidence is not known but is reported as 1-2% of all conjoined twins (1, 2). In parasitic twining one of the twins is incompletely formed (parasite) and is dependent upon it nutrition to the normal twin (autosite). In epigastric parasitic twining the parasite is attached to the autosite in the epigastric region mostly to the xiphi-sternal area (3). The parasites usually have the limbs well formed but are usually cardiac and acephalic (4, 5). The autosite is usually a normal looking baby but may have associated anomalies especially cardiac anomalies. Technically separation of epigastric parasitic twining is not difficult but the outcome is dependent upon associated anomalies in the autosite. We are reporting three epigastric parasitic twins having similar presentation with a view to understand their anatomy, associated anomalies and management issues.

Cases report
Case 1:
A 6 days old pair of parasitic twins was born at home after a full term normal delivery. The parents initially kept the twins at home with a fear of having a monstrous baby. The twins developed episodes of cyanosis and were thus brought to the hospital. On arrival the autosite was active and alert and was on mother’s feed. He was passing urine and stools normally. Autosite had a large omphalocele and a soft systolic cardiac murmur. The parasite had fully formed hind limbs and pelvis but had incompletely formed upper limbs with no motor activity in any limbs (Fig. 1). Parasite was attached to the epigastric region. Parasite had bilateral mild talipes equinovarus. Head, neck and chest were not formed. Upper limbs were incompletely formed and attached separately to the epigastrium. Parasite had a normal looking phallus from where he was attached to the autosite in the epigastric region.
passing urine. Scrotum was fully developed but testes were not palpable. Anal canal was absent. Ultrasound suggested normal internal anatomy of the autosite. Echocardiography of the autosite showed transpositioning of great vessels, large ventriculo-septal defect and a large atrial septal defect with cardiomegaly. Sonographic evaluation of the parasite showed a single kidney and bladder like structures with some cystic areas. Plain x-ray showed partially developed pelvis with both hips in position (Fig. 2). Few scattered vertebrae were noticed on right side of pelvis. Intravenous urogram showed normal excretion in both kidneys in the autosite and one fully formed kidney and bladder in the parasite (Fig. 3). Barium meal and follow-through showed grade-II Gas-

Figure 2. X-rays showing partially developed pelvis and hind limbs of the parasite with scattered vertebral bodies of the parasite.

Figure 3. IVU outlining solitary kidney and bladder in the parasite.

tro-esophageal reflux in the autosite with free passage of contrast through the gut with no communication with the parasite. MRI confirmed the findings of other studies (Fig. 4). MRA showed that parasite was getting its blood supply from the liver and also a large vessel was seen going in to the omphalocoele sac and sharing the blood supply with the gut of the autosite (Fig. 5).

Initial management included resuscitation and managing cardiac failure. A multidisciplinary team involving paediatric cardiologist, cardiac surgeon, Paediatricians, paediatric cardiac anaesthetist, paediatric surgeon and intensivists were involved in the management of the baby. After three weeks of stabilization, surgery was performed. Separation of the parasite was rather easy. It had a narrow attachment to the xiphisternum with cartilaginous tissue. A large feeding vessel was entering the parasite from the falciform ligament of autosite. A single loop of intestine was present which was entering into the omphalocele in the autosite (Fig. 6). It had blood supply coming from the omphalocele which was also seen on MRA. A patent urachus was also entering in to the omphalocele. There was no communication of these structures with the peritoneal cavity of the autosite. Both the intestinal loop and urachal remnants were ending blindly and were easily dissected out after ligation of the feeding vessel. Postoperatively the

Figure 4. MRI showing cystic (intestine) and solid areas in the parasite.

Figure 5. MRA showing dual blood supply of the parasite from the liver and also from the omphalocele through the superior mesenteric vessels.

Figure 6. Resected specimen of the parasite showing loops of intestine. Area of union was xiphisternum.
Case 1

A baby needed cardiac care but had an eventful recovery and was discharged in stable condition. He remained under regular follow-up by the cardiac team. We learnt that the baby developed serious gastroenteritis and cardiac failure at the age of 8 months and died at home.

Case 2

A ten days old baby was referred with epigastric parasitic twining. The baby was initially treated in another hospital but developed cellulites of left forearm and then was referred to us. The autosite was taking feeds normally. The parasite had well developed lower limbs and pelvis with contractures, bilateral talipes equinovarus and dislocated hips. Upper limbs were partially developed. No head, neck or thoracic structures present. It had well formed scrotum, gonads not palpable and had penile hypospadias with chordee. He never passed urine from this urethral opening. The autosite looked normal except that he had cellulitis of the left forearm from an intravenous cannula. No omphalocele present in this case but a ventral hernia was present in the autosite at the attachment of parasite. Ultrasonography of the autosite showed normal renal system with no other obvious pathology. Sonographic studies of parasite showed cystic areas in the pelvis suggesting rudimentary intestine. No renal tissue visualized. Detail evaluation of the twins was planned but the family was offered treatment abroad by some donors and were discharged on request for treatment abroad.

Case 3

A 7 days old male baby was referred with an epigastric mass present since birth. The baby was dehydrated and in serious sepsis. There was a cystic area in the upper part and mass of skin covered tissue in the lower part with tuft of hairs. The upper cystic mass looked like an incompletely developed brain tissue. Small rudimentary tags of skin were present which may suggest rudimentary limb buds. Diagnosis of an epigastric parasitic twining was made. The autosite looked normal on external appearance but was in sepsis and needed aggressive resuscitation. Due to the precarious condition of the baby detailed investigations could not be performed. The autosite had a systolic cardiac murmur suggesting congenital heart disease. Ultrasonography of the autosite suggested normal genito-urinary system. Evaluation of the parasite showed cystic areas without well formed structures. The baby died after two days of admission due to overwhelming sepsis. The parents refused for a post-mortem examination and thus detail evaluation of internal organs was not possible.

Discussion

Heteropagus or parasitic twining is a rare form of conjoined twining where one of the twins the autosite is fully formed and other so called the parasite is incomplete. The parasite as the name implies is dependent upon its nutrition on the autosite. Heteropagus twining is rare and is seen in one in 2 million live births (2). In most cases the parasitic twin is attached to the epigastric area or hypogastric region of the autosite. It
may have formed limbs and trunk but is accephalic and acardiatic. Some cases have now been reported where rudimentary heart, thorax and head has been reported (4). Parasitic twins are mostly male which is in contrast to complete conjoined twins with a female preponderance (7, 8, 9). All the three twins in our series were male confirming the above fact. Hind limbs are better developed when present. Internal structures of the parasite include variable length of the intestine and genitourinary system (10). Absent heart is a consistent feature except for few reports when there was a rudimentary heart present in the autosite (4,5). Blood supply to the parasite is from the falciform ligament and the liver in most reported cases (11,12). In our first case the parasite had dual blood supply, from the falciform ligament and also from the superior mesenteric vessel of the autosite. Omphalocele is a consistent feature in parasitic twins. In a review by M Bhansali et al it was present in more than 50% cases (13). Two of our babies had omphalocele and one had ventral hernia which has not been reported before. There is high association of cardiac anomalies in the autosite. These included VSD, ASD, PDA and Coarctation of aorta. Transpositioning of great vessels is a serious anomaly and has been reported previously. In our patients serious cardiac anomalies were the main factors which delayed the separation of the twins. Long term survival also depends upon the severity of associated anomalies and their optimal management. Our baby died after 8 months of cardiac complications.

Birth of a baby with anomalies like heteropagus twinning is still considered a stigma in the developing countries. There was significant delay in bringing these babies to the hospital resulting in death of one baby due to overwhelming sepsis. Early referral and appropriate treatment can help in achieving better survival in these babies. The ultimate outcome however will depend upon the associated anomalies especially cardiac. A team approach in the management of these babies and improvement in paediatric cardiac care services can improve survival in these babies and many other babies having cardiac anomalies.

References


Figure 9. Third pair of twins having incomplete and disorganized formation of body organs.