

## CASE REPORT

# Omphalocele and epigastric heteropagus: implications and treatment

MuneerAbas Malik, Umesh Bahadur Singh\*, Shalini Hedge, J. K. Mahajan and Ram Samujh

Department of Pediatric Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

\*Correspondence address. Department of Pediatric Surgery, Advanced Pediatric Centre, PGIMER, Chandigarh, India. Tel: +91-89685567891, +91-8728830791; E-mail: singhub.2008@gmail.com

## Abstract

In heteropagus twins, the parasitic twin is incompletely formed which is attached to the autosite. We report a case of epigastric heteropagus twins with omphalocele. The parasite had two lower limbs, a rudimentary upper limb, genitalia with developed phallus and scrotum but absent testis. An omphalocele was present just below the attachment of the parasitic twin. A single kidney with ureter and a bladder filled with urine seen during exploration. The parasite bowel was attached to a sleeve of liver tissue from the autosite within the omphalocele sac and this connection was also the main source of its blood supply. Less than 20 cases of omphalocele with EH have been reported previously. The thin sac with underlying adherent bowel led us to go ahead with surgery in the neonatal period for both, excision of epigastric heteropagus and omphalocele repair. The rectus sheath around the omphalocele sac helped in the final wound closure.

## INTRODUCTION

Conjoined twins can be symmetrical or asymmetrical (heteropagus). Heteropagus twins consist of the anatomically normal autosite and a parasitic twin that is incompletely formed which is attached to the autosite and dependant on it for survival. It has been noted that omphalocele is the most common anomaly associated with heteropagus twins. We report a case of epigastric heteropagus twins with omphalocele and likely pathogenesis.

## CASE REPORT

The child had an epigastric heteropagus twin (Fig. 1). The parasite had two lower limbs, a rudimentary upper limb and male genitalia with developed phallus and scrotum but absent testis.

The parasite was passing urine but did not have an anus. The limbs did not move or respond to external stimulus. An omphalocele was present just below the attachment of the parasitic twin. Contrast enhanced CT scan picture showed common peritoneal space with joined liver and other viscera along with bony structures (Fig. 2).

On laparotomy, the pelvis of the heteropagus contained a single kidney with ureter and a bladder filled with urine (Fig. 3). There was around 25 cm of the small intestine that abruptly ended with dense adhesions to the sac of the omphalocele. A tongue of the liver of the autosite was also part of the omphalocele sac and continuous with the parasitic bowel (Figs 4 and 5). The rectus sheath around the common omphalocele sac and extra skin tissues harvested after removing parasite, helped in final wound closure of autosite.

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**DISCUSSION**

The incidence of conjoined twins is considered to be 1 in 50 000–100 000 births and is more commonly seen in females (70%). Heteropagus twins, however, are even rarer with an incidence of 0.05 to 0.1 in 100 000 births [1].

Epigastric heteropagus (EH) is the term used when the parasite is joined to the epigastrium of the autosite and <75 cases have been reported in the literature. Due to its rare occurrence, limited data on this entity is available but the following points have been understood.

There is a male predominance in EH of 78% in contrast to the female predominance of conjoined twins [2]. All cases for which DNA analysis has been done showed the twins to be monozygotic except for one case which was dizygotic [3].

The orientation of the parasite is along the longitudinal axis of the autosite. Only the caudal portion of the parasite develops. Rudimentary limbs, pelvis, external genitalia, genitourinary system and part of bowel are common anatomical structures developed in the parasite. Liver and rudimentary heart are other rare contents in the parasite that have been identified [4].

The limbs on the parasite do not move and are not sensate. There is absence of skeletal muscle in the limbs and this is thought to be due to the failure of myoblast differentiation due to lack of proper innervation. The parasite blood supply is derived from the autosite and this connection is evident during surgery. It can also be identified on preoperative CT or MRI angiography. The most common blood supply to the parasite is derived from the falciform ligament. Other vessels that have been identified are left internal mammary, left subclavian, epigastric artery and left intrathoracic artery [1].

The peritoneal cavities of the parasite and autosite are separate. Bowel communication has been seen in only three cases till now. Omphalocele is the most common anomaly associated, followed by cardiac anomalies in the autosite. It has been suggested that cartilaginous attachments of the parasite to the xiphoid of the autosite interferes with the abdominal wall closure of the autosite that results in the formation of an omphalocele. In our case, the omphalocele was present as the parasite bowel was attached to a sleeve of liver from the autosite within the sac and this connection was also the source of its main blood supply.

In the review of cases by Manish *et al.* omphalocele was associated with EH in 50% of the cases [5]. Overall in literature, <20 cases have been reported of omphalocele with EH. According to our institutional protocol, in view of the higher incidence of congenital heart disease associated with omphalocele, we routinely



Figure 1: Image of heteropagus twins having omphalocele

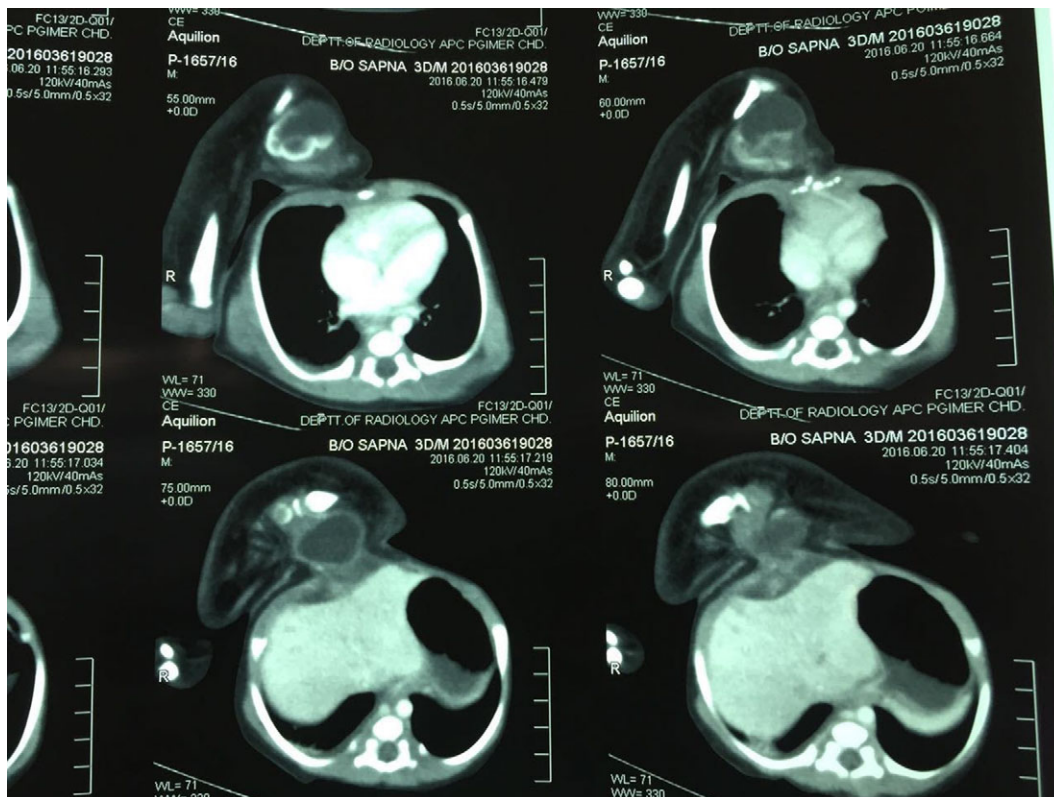


Figure 2: Contrast enhanced CT scan image showing parasite's viscera and bony parts



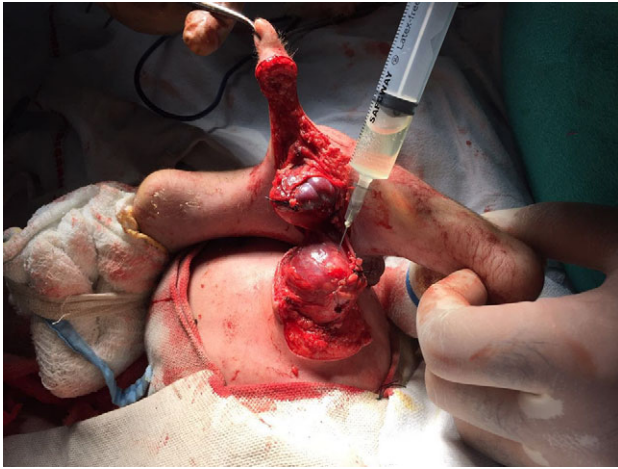


Figure 3: Operative picture showing single kidney, ureter and bladder (filled with urine) of parasite

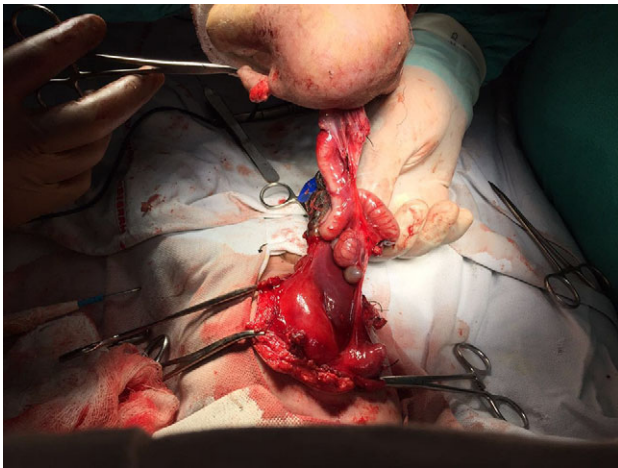


Figure 4: Operative picture showing rudimentary gut attached to part of liver



Figure 5: Excised specimen of parasite twin

screen these cases with 2D ECHO, being a non-invasive and feasible diagnostic modality at this place. Although, congenital heart disease seen independently associated with EH. The thin sac



Figure 6: Follow up pic of patient at 6 months of follow up.

with underlying adherent bowel led us to go ahead with surgery in the neonatal period for both excision of EH and omphalocele repair. The learning point was the skin and soft tissue management for large defect closure after parasite removal by harvesting rectus sheath and skin from parasite. There are very few reported cases of emergency surgery or early separation and indication was either rupture of omphalocele or death of parasite twin. Early separation in neonatal period appears more beneficial as the tissues are more pliable and available for defect closure as in some of the reported cases, the death or devitalization of parasite occurs after birth. Moreover the late separation of EH with omphalocele results in loss of domain of peritoneal cavity as gut tends to occupy more space towards the omphalocele sac so chances of ventral hernia formation are increased. However, in this index case, we did not face the problem of a ventral hernia, probably due to early separation and defect repair using rectus sheath (Fig. 6). Early separation also reduces the chances of intestinal obstruction or other bowel complications, if the parasite twin has bowel communication or adhesion with the autpagus bowel.

The neonate withstood early surgery of EH with omphalocele well with only minor wound infection. The rectus sheath around the omphalocele sac can help in final wound closure. Immediate exploration of an EH with a ruptured omphalocele has also been reported with no adverse complication.

#### CONFLICT OF INTEREST STATEMENT

None.

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**ETHICAL APPROVAL**

Not required.

**CONSENT**

Written consent from parents taken and submitted.

**GUARANTOR**

Corresponding author—Dr Umesh Bahadur Singh.

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