

# External and Internal Parasitic Conjoined Twinning: Diverse Presentation and Different Surgical Challenges

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## Abstract

**Background:** Parasitic conjoined twin is a rare but well-known entity with unclear embryopathogenesis. Abnormal conjoined twinning can result in an externally attached parasitic twin (PT), an enclosed foetus in foetu, or a mature teratoma. The treatment requires complete excision and reconstruction of local anatomy which is not always straightforward. **Materials and Methods:** PT cases presenting over 12 years were analysed. Patients with complete data, histopathological diagnosis and follow-up were included in the study. During follow-up, specific complications and related investigations were considered. **Results:** A total of five patients at four different sites were identified: two retroperitoneal foetus in foetu and three externally attached PTs which were located in the lumbar region, sacrococcygeal area and on the lower anterior abdominal wall. All patients underwent complete surgical excision. In foetus in foetu cases, the blood supply was directly from the aorta with a short stump while the three externally located ones required meticulous and careful dissection with the reconstruction of local anatomy. **Conclusion:** Parasitic conjoined twinning can present at different sites and surgical challenges vary accordingly. For surface lesions, reconstruction may be as complicated as excision. Prognosis can be affected by the excellence of anatomical restoration. Long-term follow-up is essential to address problems specific to the site of lesion and method of surgical reconstruction.

**Keywords:** Embryogenesis, foetus in foetu, mature teratoma, parasitic twin, surgical challenges

## INTRODUCTION

Parasitic twin (PT) is an extremely rare condition, with an estimated incidence of <0.1 in 100,000 births.<sup>[1]</sup> Historically, the terminology used in reports of PT has been inconsistent. Spencer defined PT as twins conjoined in one of the same anatomical locations as the more common 'complete' conjoined twins, one twin of the pair is severely defective (the 'parasite') and its counterpart ('autosite') must be mostly intact. The PT spectrum includes the externally attached parasite twin, foetus-in-foetu (FIF) or endo PT and mature teratoma.<sup>[2]</sup> PT can be detected during antenatal ultrasound (USG) screening. Postnatally, they may have varied presentations: asymptomatic, incidentally detected, symptomatic or obvious visible mass lesion.<sup>[3]</sup> Although treatment of choice is complete excision, as the site varies, the challenges met during surgeries are different. Here, we are describing PT at four different sites along with their management, surgical challenges, outcome and follow-up.

## MATERIALS AND METHODS

This study was conducted in the department of paediatric surgery of a tertiary care centre. Patients were included from March 2008 to January 2020, and data were collected from record registers maintained by the department. Ethical clearance was obtained from the institute ethics committee (IEC/2020/SPL-821). All patients of PT (external and internal, i.e. FIF) who presented during this time duration were included in the study. Patients with complete clinical details including presentation, pre-operative investigations, histopathological diagnosis and follow-up were included in the study. Patients up to the age of 12 years were included irrespective of gender. Demographic data, clinical presentation, pre-operative investigations including imaging and

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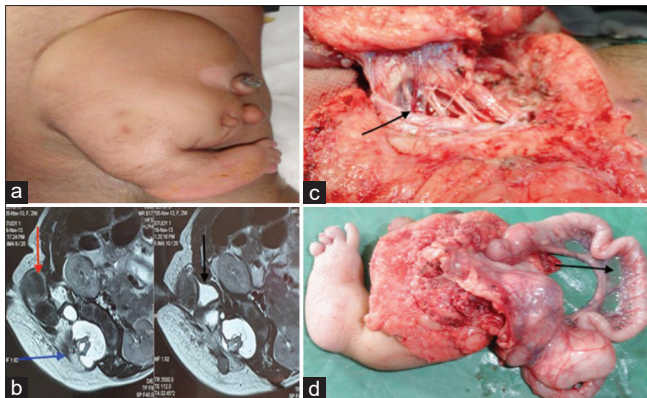
tumour markers, operative details, post-operative complications, outcome and follow-up were analysed. Patients presenting in the emergency room or routine outpatient department were examined thoroughly and assessed for any immediate intervention. They underwent routine blood investigations such as complete blood count, renal function test and liver function test along with tumour markers including alpha-fetoprotein and human chorionic gonadotropin ( $\beta$ HCG). Radiological investigations were done according to the site of the mass lesion and clinical presentation. In general, plain radiographs were done for all the children as per the location. For intra-abdominal masses, USG was performed initially. Either contrast-enhanced computed tomography (CECT) scan or magnetic resonance imaging (MRI)

was done for all the children before undergoing surgery. All patients were kept under routine follow-up, and along with clinical examination, timely radiological investigations and tumour markers were also assessed.

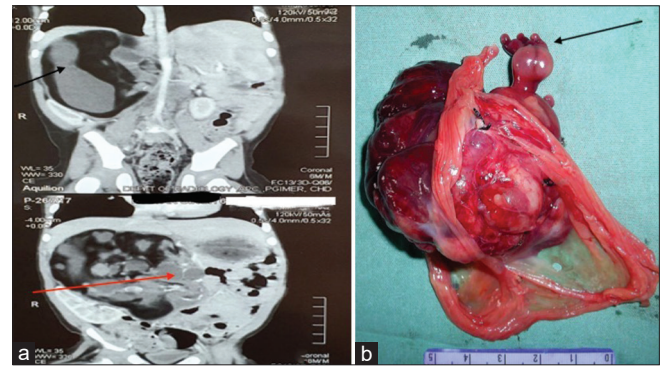
## RESULTS

A total of five patients were enrolled in the study. The patients' details and their management are mentioned in Table 1. Preoperatively, AFP and  $\beta$ HCG levels were in the normal range in all the cases. In all patients, complete surgical excision of the mass could be achieved without injuring the neighbouring structures. All excised specimens were sent for histopathological examination which was suggestive of PT with the presence of different mature organs. In the last follow-up, none of the patients had any major issues related to the primary lesion or surgery.

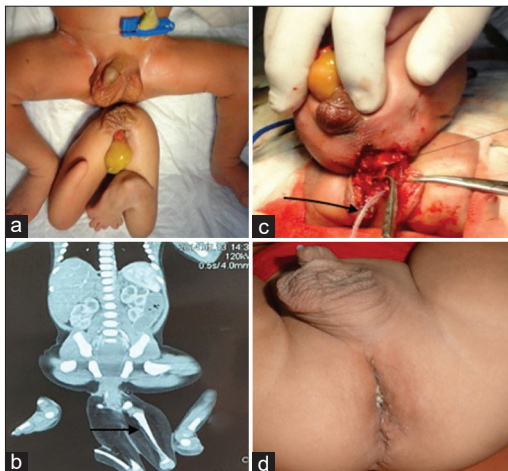
In our first case [S. no. 1, Table 1 and Figure 1], during surgery, the mass was meticulously separated from the neural structures



**Figure 1:** (a) Pre-operative photograph showing a skin-covered mass lesion with limb formation at the back. The magnetic resonance imaging (b) showing solid (red arrow) and cystic component (black arrow) of the mass lesion and open spinal canal (blue arrow) with the adhesion of nerves with mass. Intraoperative image (c) showing open spinal canal and nerve fibres with mass lesion and excised specimen (d) containing limb, bowel, and mesentery along with soft tissue



**Figure 2:** The contrast-enhanced computed tomography (a) shows solid and cystic mass (black arrow) in the right retroperitoneum, reaching up to midline (red arrow) and displacing major vessels. Excised specimen of the left retroperitoneal foetus in foetu (b) shows the budding limb-like structure (black arrow) and bowel with mesentery



**Figure 3:** Pre-operative image (a) shows an attached mass lesion at the sacrococcygeal region. The contrast-enhanced computed tomography (b) shows attached pedunculated mass from the anal canal region containing long bones. Intraoperative image (c) showing close adherence with the anal canal and stenotic anal opening (black arrow showing feeding tube in the anal opening). Follow-up image (d) of the patient showing healed wound



**Figure 4:** Pre-operative image (a) showing exomphalos (black arrow) and distorted perineum (b) and abnormal vestibule (black arrow). Intraoperative image (c) after the closure of rectus sheath and immediate post-operative image (d) after local reconstruction

**Table 1: Patients' detail and their management**

Age/sex	Location/presentation	Investigations	Intra-operative findings	HPE	Post-operative complications	Follow-up
21 days/male	Back (rachipagus) Visible mass lesion with LL like structure [Figure 1a]	MRI – solid, cystic and calcified lesion over the lumbar region, non-fusion of spinal arch element, herniation of spinal cord and neural elements adhered to a mass lesion [Figure 1b]	The lesion was attached to the spinal cord from deep inside, vertebral defects were present, back muscles were splayed due to lesion [Figure 1c] Complete excision	Gross: Intestine with mesentery and foot with a bone [Figure 1d] HPE: All three germ layer components consistent with PT	Nil	5 years old Fully continent for stools and urine No distal neurological deficit
6 months/male	RP (FIF) Incidentally detected lump	CECT-lesion with cystic, fatty, calcified and ossified areas in right RP, displacing right kidney and closely adherent to the aorta [Figure 2a]	Mass of 10×10 cm size, adherent to colon and right renal vessels Complete excision Cut section showed bony elements (vertebra), bowel loops and multiple cystic areas	HPE: Disorganized mature elements, fibrous tissue, smooth muscle, neural tissue and tubular structure resembling colon, consistent with FIF	Nil	1 year old No complaints
11 days/female	RP (FIF) Antenatally diagnosed	CECT-solid, cystic lesion 6×5×3 cm with an area of calcification in left RP	A solid cystic lesion in the left side of the abdomen extending between mesocolon and retroperitoneum, pushing left kidney upwards Blood supply from aorta and 2 veins separately draining into the left renal vein Cut section shows cartilaginous and bony structure like limb (2B)	HPE: Tissues of cerebellum, cerebrum, skin, bone and pleural lining, consistent with FIF	Nil	10 years old No complaints
1 days/male	Sacrocoecal region (pygopagus) Mass with LL like structure and external male genitalia (phallus and scrotum without the testis) A firm stalk was seen attached in the region measuring 2.5×1 cm and obliterating the anal canal [Figure 3a]	Plain CT-pedunculated mass lesion 11×7.5×6 cm over the sacrocoecal region, closely adherent to the anal verge Shows fatty attenuation area, soft-tissue component and well-formed long bones [Figure 3b]	Closely adherent to the anal canal [Figure 3c] Complete excision Anorectoplasty Muscle complex reconstruction	Gross: Mass with two rudimentary LLs, the right had seven toes while the left was thinner with no foot HPE showed fibrocartilage, wall of urinary bladder and colon. No embryonic elements or immature tissue identified Features compatible with PT	Stool frequency Perineal excoriation	5 years old Continent with 1-2 bowel movements per day [Figure 3d]
3 days/female	Anterior abdominal wall (omphalopagus) Large omphalocele and protruding mass with the appearance of nipple and breast tissue along with hair, extending inferiorly till labial region and distorting perineal anatomy but not involving vaginal or urethral orifice [Figure 4a and b]	CECT: Anterior abdominal wall defect in the infra-umbilical region with herniation of bowel loops and urinary bladder Ill-defined soft tissue mass with exophytic component seen with a bony structure within, posteriorly lesion was in close contact with bladder	Central infra-umbilical abdominal muscle defect Complete excision Abdominal wall closure Genital reconstruction	Cut section showed a cartilaginous and bony structure Tumour composed of all three germ layers, skin, bone, cartilage, pancreatic acini and hepatocytes seen, no immature neuroepithelium identified	Postoperatively elective ventilation for 48 h Skin dehiscence at the lower end of the suture line	1 year old No issue Wound healed well Perineal anatomy: cosmetically acceptable

CT: Computed tomography, CECT: Contrast-enhanced computed tomography, FIF: Foetus in foetu, HPE: Histopathological examination, IVC: Inferior vena cava, LL: Lower limb, MRI: Magnetic resonance imaging, PT: Parasitic twin, RP: Retroperitoneum

and required a lengthy dissection as it was not easy to distinguish between normal structures and abnormal tumour elements. After complete excision of the large mass in the presence of an open neural canal, closure of the defect was also a challenge. The dura was initially closed followed by reinforcement with lumbosacral fascia. The skin and subcutaneous tissues were also extensively mobilised and closure was achieved. The child is under regular follow-up and doing well.

Two patients had retroperitoneal (RP) FIF [S. no. 2 and 3, Table 1 and Figure 2]; in both cases, the mass was very close to the major vessels and was receiving its blood supply directly from the aorta. The supplying vessels were also very short and dissection had to be done close to the aorta.

Our fourth case [S. no. 4, Table 1 and Figure 3] was a pygopagus PT and the mass lesion was at the sacrococcygeal region, close to the anal verge and obstructing the anal opening. The muscle complex was stretched out due to the pedicle of the tumour, which was coursing through it. The anal canal had a common wall with the tumour base and had to be carefully dissected from it. After the excision of the mass, the remaining anal opening was found to be very narrow, the surrounding muscles were very thin and abnormally oriented. The patient required anorectoplasty and muscular reconstruction in the presence of flabby, stretched and imprecise anatomy. Our patient had frequent stools during the 1<sup>st</sup> year of his life but at present is fully continent at 5 years of age.

The last case [S. no. 5, Table 1 and Figure 4] had very complex anatomy (omphalopagus PT) because of the presence of a large omphalocoele along with the mass and distorted perineal structure. Hence here, the main challenge was the closure of the abdominal muscle defect as well as reconstruction of perineal anatomy. For abdominal wall closure, both rectus muscles were mobilised from the pubic bone and closed in the midline under tension. For genital reconstruction, labial folds were created from the remaining surrounding tissue, maintaining the appropriate position of the urethral and vaginal orifices.

## DISCUSSION

The embryopathogenesis of the PT spectrum forms a continuum and they are all variations of abnormal conjoined twinning, with the site of union and the extent of damage (or defect) of one embryo resulting in one of the pathologies.<sup>[2]</sup>

There are several theories on its origin, with the two major theories being labelled as ‘fission’ and ‘fusion.’. Proponents of the former suggest that incomplete fission of the blastocyst inner cell mass during the primitive streak stage, which occurs 13–15 days post-fertilisation, results in two centres of axial growth that retain a connection at some point.<sup>[1]</sup> ‘Fusion’, in contrast, refers to two originally distinct inner cell masses that coalesce secondarily at a later stage.<sup>[1]</sup> The theory for the development of asymmetry between autosite and parasite after these events was first postulated by Donitz, who proposed that vascular compromise causes the tissue of the PT to become

dependent on collaterals derived from the autosite and selective ischemic atrophy of the deprived portion of the parasite’s body follows.<sup>[1,4]</sup>

In a previous report of two similar cases, the authors have mentioned the attachments of the rachipagus PT to the spinal cord and highlighted the difficulty as well as the importance of untethering and proper dissection of neural elements.<sup>[5]</sup> Similarly, two cases of RP FIF were reported, wherein the blood supply was arising directly from the aorta as seen in our cases also.<sup>[6]</sup> One should therefore be very careful during dissection as injury to major vessels can be catastrophic. Second, before ligation of a vessel, one must ensure that it is supplying only the mass and not any nearby organ like the kidney. The latter problem is more likely if FIF is receiving its blood supply from the same area as normal aortic branches such as the celiac trunk, superior mesenteric artery and renal artery.

One report of PT at the sacrococcygeal region was reported in a very low birth weight baby, who was haemodynamically unstable due to the large mass and required emergency surgery as a life-saving procedure.<sup>[7]</sup> In another case, the mass was not apparent from the outside but presented with a lump over the buttock noticed by the mother. In both cases, a good outcome was reported during the follow-up.<sup>[8]</sup>

A case of omphalopagus PT in an 11-year-old female child has been reported. The patient had an abdominal mass that was noticed since birth. A computed tomography (CT) scan showed that there was no sharing of any organ with the parasite. The mass was excised completely, and primary closure was achieved. The mass was composed mainly of bowel along with skin, hair, skeletal muscle and cartilage.<sup>[9]</sup> Another reported case described in a newborn with omphalopagus PT had large omphalocoele and multiple cardiac anomalies, i.e., transposition of great arteries, coarctation of aorta and ventricular septal defect. The patient underwent multiple surgeries. Initially, the rudimentary limb was excised then cardiac surgery was performed followed by laparotomy for gastric perforation repair and at last excision of mass was done. The child had a stormy post-operative course with a long hospital stay (117 days) but eventually was discharged on nasojejunal feeds.<sup>[10]</sup> In large omphalocoeles, abdominal wall closure is always a challenge. In our case, it was under tension even after extensive mobilisation of muscles, and therefore, the child was kept on a ventilator along with deep sedation. In reported literature, staged closure, use of tissue expander (subcutaneous as well as intraperitoneal) and mesh have been described to overcome tissue deficiency.<sup>[11]</sup>

Due to overlapping embryopathogenesis, and common histopathological features, there have been many attempts to define external PT, FIF and mature teratoma separately. Spencer suggested that the external PT is a grossly defective foetus, or foetal parts, attached externally to a relatively normal twin in one of the same areas in which intact conjoined twins are united. In FIF, there is a mass enclosed within a distinct sac, covered by skin, recognisable anatomic features and attached

to the host by a pedicle containing a few large blood vessels. A teratoma consists of a chaotic mixture of microscopically identifiable tissues, rarely with more differentiation than a well-formed tooth or a portion of the intestine. It is also found within the body of the autosite and is immediately adjacent to the original location of the neural tube or alimentary tract but enclosed in connective tissue with a broad attachment to the surrounding tissue. It is capable of independent growth and may become malignant.<sup>[2]</sup>

During clinical examination and investigations, often it is not possible to differentiate between teratoma and PT preoperatively. Therefore, in every patient, serum AFP and  $\beta$ HCG should be done. These tumour markers can be normal or high in cases of immature teratoma while cases of PT and mature teratoma always show a normal level. In our study, all five cases had normal tumour markers. As most cases present in the early neonatal period or infancy, it is easier and quicker to perform a CECT. However, as seen in our rachipagus PT [S. no. 1, Table 1] where there was the involvement of the spinal cord, MRI was preferred.

In general, most cases have a good outcome after surgery. Few site-specific complications such as tracheostomy in the case of neck PT and ventriculoperitoneal shunt in two cases of intracranial PT have been reported.<sup>[12]</sup> Malignant transformation of PT has also been reported.<sup>[13]</sup> We have previously reported a case of partial FIF over the mandible region although that patient did not survive because of associated complex congenital heart anomalies.<sup>[14]</sup>

These children require long-term follow-up as problems can arise later in life. In our patients, during follow-up, along with clinical examination, radiological investigation in the form of USG was performed for the RP lesions. In both cases, USG was normal at the last follow-up. No CT/MRI was performed for surface lesions as there was no indication during the clinical examination. The problems that could appear related to anatomical reconstruction were also addressed during follow-up such as size of the anal opening and continence status in the case of anorectoplasty, signs of tethered cord in the case of spinal dissection and urethral/vaginal stenosis in the cases where perineal reconstruction was done.

## CONCLUSION

Parasitic conjoined twinning can present at different sites and surgical challenges vary accordingly. For surface lesions, reconstruction may be as complicated as excision. Prognosis can be affected by the excellence of anatomical restoration. Long-term follow-up is essential to address problems specific to the site of lesion and method of surgical reconstruction.

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## Conflicts of interest

There are no conflicts of interest.

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