

# Asymmetrical Conjoined Twins or Saints? - A Case Report

R. SIVAKAMI, D. KARTHIKEYAN

## ABSTRACT

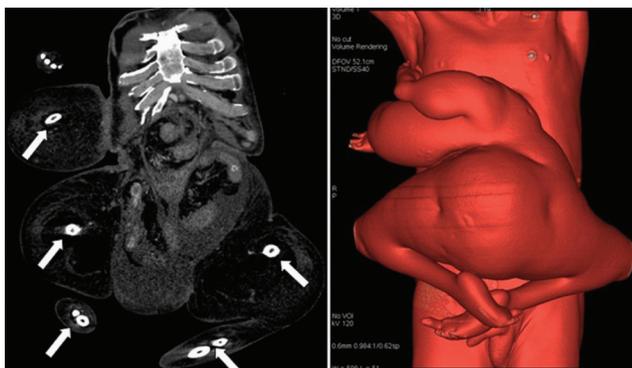
Conjoined twins are one of the rare developmental anomalies, where twins do not separate completely. They are of two types, symmetrical the common type and asymmetrical the rarest form. Asymmetrical twins are of

two types, one is heteropagus or parasitic twin and the other is fetus in fetu. Two cases, one each of these two rare forms are presented here with a short discussion about pre operative role of radiology in clearly delineating the level and nature of connection between the twins.

**Keywords:** Autosite, Fetus in fetu, Omphalo-Heteropagus Twin, Parasitic twin, Pre operative investigation

## CASE-1

A 54-year-old male who was considered a saint in his place came to the Outpatient Department (OPD) with complaints of mass attached to his upper abdomen since birth. On inspection it was a parasitic twin with small body and four rudimentary extremities and referred to the Radiology Department, for MDCT-chest and abdomen contrast with angiography. 128 slice MDCT angiography was done with MPR and 3D post processing and revealed the parasite of the twins attached to the autosite at the level of umbilicus with a large abdominal wall defect. It showed two small upper limbs pointing towards right side, small abdomen and pelvic region and two small lower limbs on either side of pelvis with dysplastic bone and soft tissue [Table/Fig-1a,b]. The parasite did not show head, chest, heart or any well-formed solid organs in the abdominal cavity.



**[Table/Fig-1a,b]:** (a) Plain CT coronal image showing right upper and 2 lower extremities of the parasitic twin (arrows). (b) 3D volume rendered image showing autosite and parasite.



**[Table/Fig-2a,b]:** a&b – Plain CT-axial images showing upper, lower extremity bones and pelvic bone of parasitic twin (arrows).

The parasitic twin and the host were connected at bony level through autosite xiphisternum with upper extremity of parasitic twin, at vascular level, right internal mammary artery and vein of autosite are seen supplying and draining the parasitic twin [Table/Fig-3].

Enhancing soft tissue structure supplied by a branch of autosite left hepatic artery with adhered falciform ligament of the host liver is seen [Table/Fig-4] communicating to the parasitic twin via a narrow pedicle. Few loops of bowel signature structures [Table/Fig-5] with surrounding fat and vasculature are seen in the parasitic twin probably representing remnants of hepatic diverticulum/omphalomesenteric complex. Surgical separation of the parasitic twin was suggested for the patient.

Orally informed consent of the patient was obtained for future publications purpose.

## CASE-2

A 12-year-old male child presented to the OPD with history of long standing abdominal protuberance and abdominal



**[Table/Fig-3]:** Post processed angiographic images showing prominent internal mammary artery supplying parasitic twin.

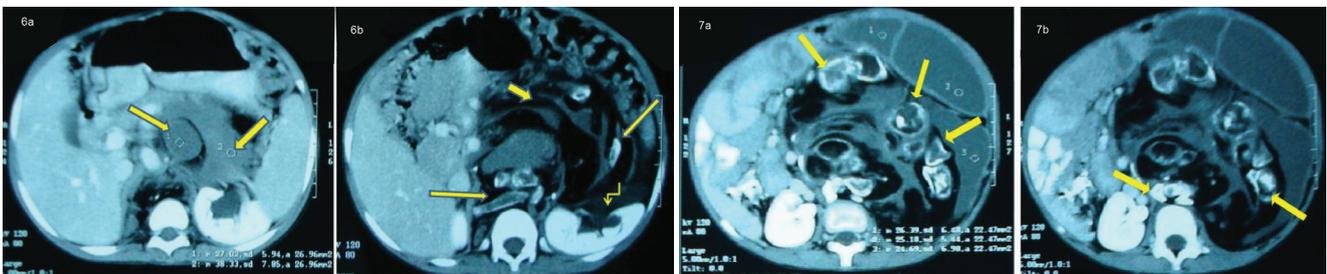
**[Table/Fig-4]:** Contrast CT-sagittal image showing soft tissue structure attached to autosite liver (arrow) entering in to parasite.

**[Table/Fig-5]:** Contrast CT-axial image showing tubular structures with bowel signature (arrows) in the parasite.

pain. Patient referred to Radiology Department for evaluation. Dual slice helical contrast CT-abdomen was performed which revealed a well encapsulated mixed density mass in the abdominal cavity with thick well-defined wall. Multiple thick septations are seen in the lesion. The mass showed solid component with multiple coarse calcifications. Cystic and fat components are also seen with probable malformed fetal vertebral column (axial) and extremity bones (appendicular

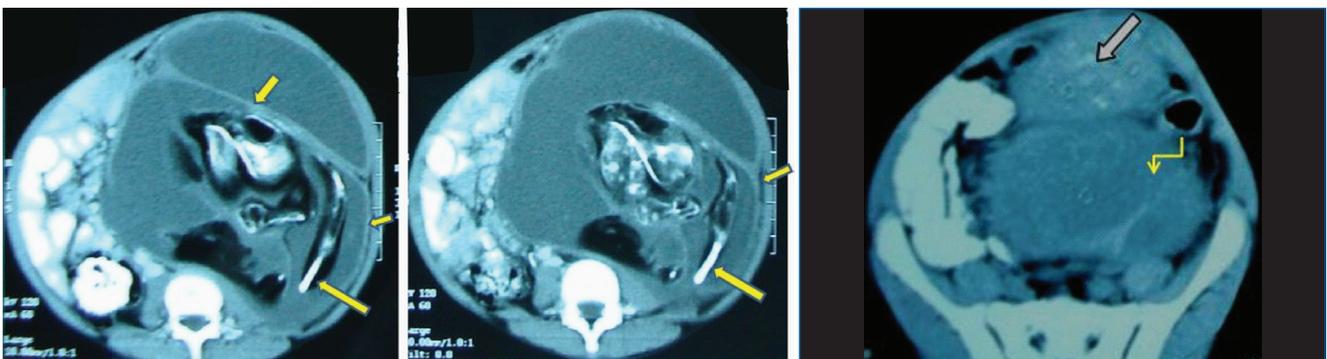
skeleton) [Table/Fig-6-9]. There was no demonstrable vascular communication between the mass and the host.

Differential diagnosis considered was teratoma and fetus in fetu. The presence of the poorly formed vertebral column was favoring the diagnosis of fetus in fetu in our case. Separation surgery was suggested for the patient, however patient was lost for follow-up.



**[Table/Fig-6a,b]:** (a,b) Contrast axial CT abdomen showing mixed density mass with fluid component (6a arrows), fat (6b short arrow) and malformed fetal appendicular skeleton (6b long arrows) with left kidney being displaced postero-inferiorly by the lesion (6b elbow arrow).

**[Table/Fig-7a,b]:** (a,b) Contrast axial CT-abdomen showing mixed density mass with fat component and malformed fetal axial skeleton (arrows).



**[Table/Fig-8a,b]:** (a,b) Contrast axial CT-abdomen showing fetiform mass with well defined thick capsule and septa (short arrows) and malformed fetal appendicular skeleton (long arrows). **[Table/Fig-9]:** Contrast axial CT-abdomen showing fetiform mass with solid component (yellow arrow) and multiple focal coarse calcifications (white arrow).

Orally informed consent was obtained from the patient's parents for future publications purpose.

## DISCUSSION

Twinning occurs in approximately 1 of every 87 live births. Monozygotic twins account for one third of twin births. Conjoined twins account for 1% of monozygotic twins with total prevalence of 1.47 per 100000 births [1]. An increased incidence of 1:14 000 to 1:25 000 is described in various parts of Southeast Asia and Africa [2]. When born live, females are affected more often than males, with a female-to-male ratio of 3:1 [2].

Conjoined twins are of two types, symmetrical the common type and asymmetrical i.e. heteropagus/parasitic twins and fetus in fetu the rarest forms. Twins that are identical to each other are called symmetrical twins which are classified according to the site of attachment with the Greek suffix pagus which means fixed, union at the level of thorax (thoracopagus; 30-40%), abdomen (omphalopagus; 25-30%), sacrum (pygopagus; 10-20%), pelvis (ischiopagus; 6-20%), skull (craniopagus; 2-16%), face (cephalopagus) and back (rachipagus) [3-5].

Parasitic or heteropagus twin is a grossly defective fetus, or fetal parts, attached externally, with or without internal connections, to an autosite i.e. relatively normal twin in one of the same eight areas of union as described with symmetrical twins. The defective twin is usually composed of extremities but may also contain viscera and only rarely a normal brain and beating heart. The heteropagus twin's incidence is much less than for symmetrical twins, i.e 1 per 1 million births or less, accounting for 1-2% of conjoined twins. Until 2003, only 157 cases of heteropagus twins have appeared in the literature [6]. Heteropagus twins are equally distributed among both the sexes, not like the symmetrical twin where female preponderance is noted. Omphalopagus type accounts for the majority of case of parasitic twin, however presentation in later years of life is rare as in our case.

Fetus in fetu is another rarest form of asymmetrical conjoined twin with incidence is 1 per 5,00,000 births [7], with total of 57 reported cases, in which fetus like mass with poorly identified fetal parts is seen within the body of the host twin, commonly the abdominal cavity in encapsulated sac with or without vascular communication with the host twin. Some authors suggest that fetus in fetu and fetiform teratomas could be part of a same spectrum of abnormalities; however, a most important finding which points to the diagnosis of fetus in fetu is the presence of vertebral column in the lesion. The presence of vertebral column suggests that the development of the included twin has reached up to the formation of notochord [8]. Surgical separation of parasite/fetal mass is the treatment in these types of twins.

Two cases of these rare forms of conjoined twins are presented here for their rarity of presentation in later years of life and to discuss the role of pre operative radiological investigations for clearly delineating the level and nature of connection between the twins in terms of vascular, visceral, bowel and bone for appropriate surgical planning and prognostic information.

**Embryology:** Etiology of different types of twins is complicated and the mechanism considered is error in blastogenesis resulting in division of single zygote in to two in varying periods of post conception. When the embryonic disc divides 13 days after conception, the division is commonly incomplete resulting in formation of conjoined twins [9,10]. Most accepted theory for heteropagus twin pathogenesis suggested by Dönitz in 1866 says that heteropagus twin originates from symmetrical twin as a result of vascular deficiency of one the twins (parasite) causing ischemic damage and hence dependence on the autosite for vascular supply. This theory has been supported by finding vascular connection from autosite supplying parasite surgically, however all the abnormalities of heteropagus twin especially acephalic status could not be explained by this theory. Generally, the heteropagus twins are of identical sex and this has been supported by DNA analysis [11]. In fetus in fetu the totipotent inner cell mass of developing blastocyst of monozygotic, diamniotic twin divides unequally resulting in incomplete smaller cell mass inclusion in maturing sister embryo.

Radiology plays a role in diagnosing conjoined twins through prenatal sonography when fusion of fetal parts is noted. For more detailed evaluation of degree, levels of fusion and associated anomalies fast sequence MRI can be used [12]. Proper prenatal evaluation helps in planning the labor. Postnatally radiology plays a role in clearly delineating the level and nature of connection between the twins. Contrast enhanced MDCT provides excellent anatomic and bone detail, demonstrating presence and sharing of viscera and evaluation of the gastrointestinal and urogenital tracts. Vascular connections can be delineated by CT angiography which helps in planning of proper surgical procedure.

## CONCLUSION

Conjoined twins are a challenge to the multidisciplinary team consisting of pediatric surgeon, gastro surgeon and radiologist. Sonography and MRI plays a significant role in diagnosis and delivery planning of conjoined twins in prenatal period. Postnatally contrast enhanced MDCT provides excellent anatomic details, clearly delineating the level and nature of connection between the twins, and helps in proper surgical planning. Most of these patients present to the hospital in neonatal or infant period for separation surgery. Our patients presented in later years of life, which is rarely reported in literatures.

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