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## Fetus in Fetu: Case Report and Brief Review of Literature on Embryologic Origin, Clinical Presentation, Imaging and Differential Diagnosis

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**Background:**

Fetus in fetu (FIF) is a rare entity in which a malformed diamniotic monochorionic parasitic fetal twin develops inside a normal co-twin's body, most commonly in the abdominal cavity. FIF is differentiated from the teratoma by the presence of vertebral column often with an appropriate arrangement of other organs or limbs around it.

**Case Report:**

A two-and-a-half-year-old girl presented with a painless abdominal swelling in the right hypochondrium. On imaging, a heterogenous soft tissue mass with internal calcific densities was noted in the retroperitoneum. The mass had vertebral organization, limb and pelvic bones. The presence of a fetiform teratoma was suspected and surgery revealed an encapsulated mass with an anencephalic head, spine, upper and lower limb buds. Histopathology confirmed the presence of a fetus in fetu. The postoperative period was uneventful with no evidence of recurrence.

**Conclusions:**

FIF is a pediatric rarity. Cross-sectional imaging helps in differentiating it from a teratoma, meconium peritonitis and abdominal ectopic pregnancy. Surgical excision is the treatment of choice for this benign condition, which requires a follow-up only in certain cases. This case report describes a retroperitoneal fetus in fetu and discusses its clinical presentation, differential diagnosis and embryologic origin.

**MeSH Keywords:**

**Diagnostic Imaging • Retroperitoneal Neoplasms • Teratoma • Twinning, Monozygotic**

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<http://www.polradiol.com/abstract/index/idArt/899956>

### Background

Fetus in fetu (FIF) is a rare congenital anomaly with an incidence of 1 in 500,000 live births. It was first described by Meckel, a German anatomist. FIF is a malformed parasitic twin found inside the body of its host co-twin, usually in the abdominal cavity. It is formed as a consequence of an unequal division of the totipotent inner cell mass of a developing blastocyst, which results in an inclusion of a small cell mass within a maturing sister embryo. Thus, a vestigial remnant representing the co-twin in a diamniotic monochorionic pregnancy gets incorporated within the normal co-twin's body. FIF is commonly described in the

retroperitoneum; however, it has been reported in other sites as well, from the scrotal sac to the cranium. It is differentiated from teratomas by the presence of vertebral organization. Unlike teratomas, it is a completely benign condition.

### Case Report

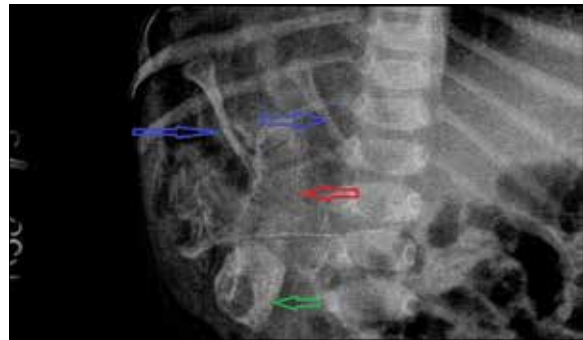
A 2½ year-old girl presented with an asymptomatic painless mass in the right upper abdomen. On physical examination, a 7×6 cm, nontender, firm and well-defined mass with smooth surface was noted in the right hypochondrium extending to the epigastric and umbilical regions as



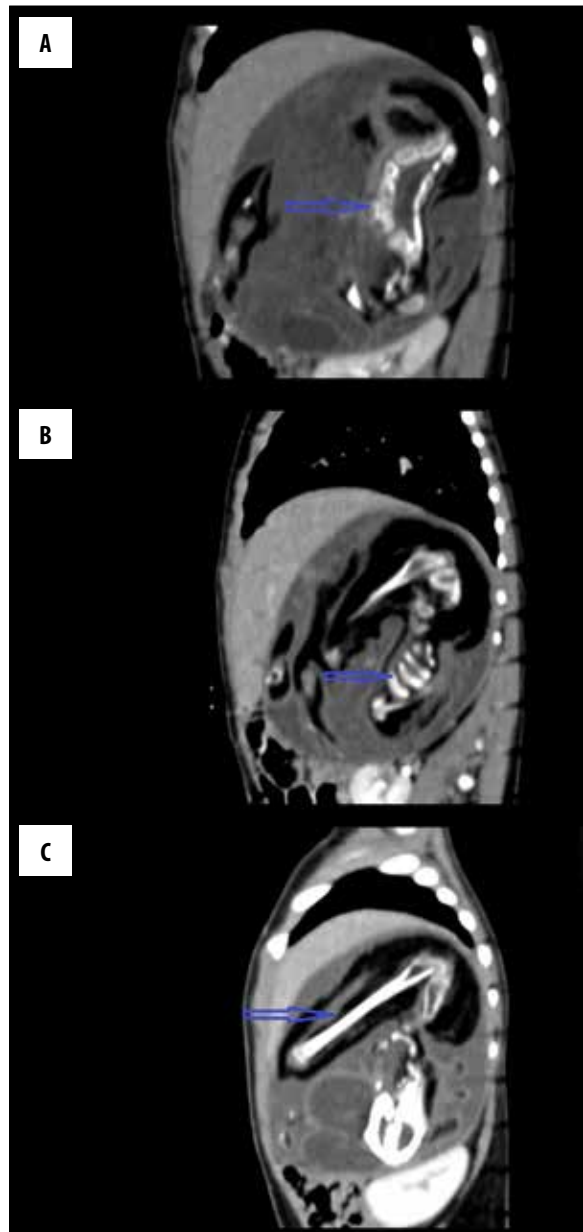
**Figure 1.** Frontal radiograph showing multiple dense radio-opacities in the right upper quadrant of the abdomen, resembling well-formed limb bones and spine.

well as to the right iliac fossa. Family history of paternal twinning was present. On imaging, a frontal radiograph of the abdomen showed multiple dense radio-opacities in the right upper quadrant of the abdomen resembling well-formed limb bones and spine (Figures 1, 2). On abdominal ultrasound, there was evidence of a heterogeneous right suprarenal mass with coarse calcifications and solid areas. On contrast-enhanced computed tomography of the abdomen (Figures 3, 4) with a 3D-volume-rendering technique (Figure 5), a heterogenous, well-circumscribed soft tissue mass was noted in the right anterior pararenal space in a suprarenal location. It showed multiple bony densities resembling a spine, limb bones, ribs and pelvic bones. These bony densities were surrounded by soft tissue and fat. No obvious contrast enhancement in the soft tissues was noted. Radiological diagnosis of FIF was considered and the child was referred to a surgical ward.

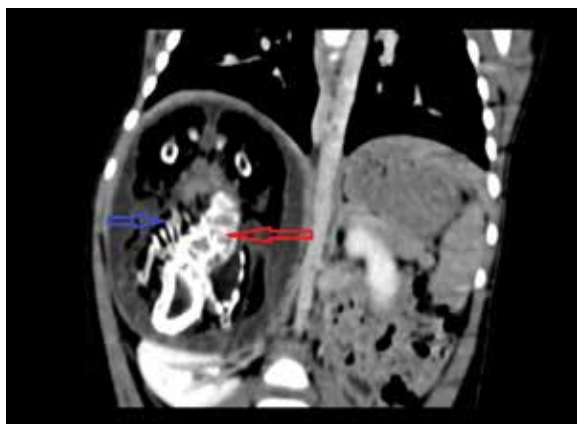
On laparotomy, a right upper quadrant retroperitoneal mass (Figure 6A), ~10×12 cm in size, covered with a sac was noted displacing the liver towards the left side and the right kidney anteriorly, stretching the right renal vessels. Venous drainage from the mass was enabled by a direct tributary of the inferior vena cava (IVC). On opening the



**Figure 2.** Magnified view of the radiograph from Figure 1; Magnified view: blue arrow – limb bones; red arrow – spine; green arrow – pelvic bones.



**Figure 3.** Sagittal multiplanar reformats of contrast-enhanced computed tomography of the abdomen. Blue arrows are showing. (A) Spine; (B) Rib; (C) Limb bone.



**Figure 4.** Coronal multiplanar reformat of contrast-enhanced computed tomography of the abdomen. Blue arrow point to the ribs and red arrow to the spine.

sac (Figure 6B, 6C), 50 ml of vernix caseosa, along with a malformed fetus with an anencephalic head, spine, upper and lower limb buds with nails and partially differentiated digits were noted.

On the histopathological examination, the gross specimen showed a partially developed fetus with partial differentiation of the four limbs, skull and spine with palpable vertebrae, bones, scapula and ribs. Microscopic sections showed mature derivatives of the ectoderm, mesoderm and endoderm. No immature components were identified. There was no evidence of somatic or germ cell malignancy. A final diagnosis of fetiform teratoma was made. The child is on a regular follow-up and at 1½ years following surgery there is no evidence of recurrence.

**Discussion**

FIF presents in various age groups with a predominance in infancy and in the majority of cases it is diagnosed in patients younger than 18 months of age [1,2], with very few case reports in adults [3,-5]. It commonly presents as an asymptomatic abdominal mass. In our case, the child was brought to hospital at 2½ years of age when the mother noticed a swelling in the right upper quadrant. It is commonly located in the retroperitoneum; however, it has been reported in other sites including the cranial cavity [6], oral cavity,



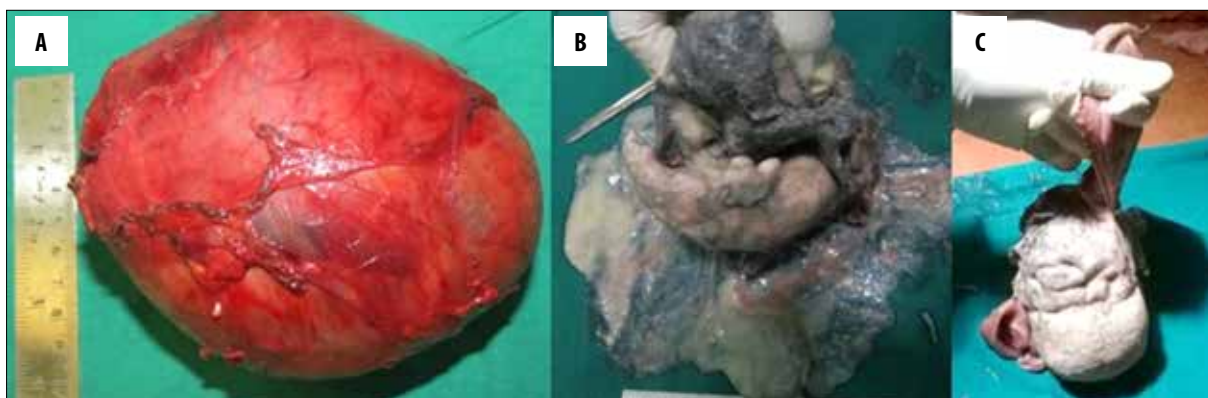
**Figure 5.** Volume rendered CT image showing Fetus in fetu.

neck [7], mediastinum [8], back [9], sacrococcygeal region [10] and within the scrotum. Most commonly, it presents with a single parasitic fetus as in our case; however, multiple fetuses ranging from 2 to 5 have also been reported [11].

Different organ systems can be found in these fetuses. Commonly noticed organs are the vertebral column and limbs. However, other organs such as the ribs, central nervous systems, gastrointestinal tract, vessels and occasionally thymic tissues can also be seen. In most cases, FIFs are anencephalic as noticed in our case; however, there is a single report of rudimentary brain tissue with ventricles reported by Reddy et al. [12].

The blood supply of the FIF is derived most commonly from the abdominal wall plexus, as the mass is attached to the abdominal wall. The size and weight of FIF varies depending on the blood supply. Those FIFs having vascular connections with the host are larger with better-developed features.

In our case, the FIF was located in the upper retroperitoneum and weighing 480 gram. The sac was closely adherent



**Figure 6.** Photos of Intraoperative gross specimens. (A) Retroperitoneal mass covered with sac; (B) Specimen showing hair and limb buds; (C) Partial differentiation of vertebrae.

to the undersurface of the liver and having its venous drainage through a direct tributary of the IVC.

There is a controversy whether FIF is a separate entity or a well-organized teratoma. A teratoma is a neoplasm containing multiple tissues from all 3 germ layers, which has a malignant potential. Hence, FIF is described by some authors as a highly organized teratoma. Some authors [13] think that FIF consists of a spectrum of malformations that result from abnormal embryogenesis in a monochorionic pregnancy, which includes conjoined twins at one end to fetiform teratomas at the other end, with parasitic twins and embryonic vestigial fetal inclusions in between.

However, a lot of other authors feel that FIF is a distinct entity, separate from the teratoma, for several reasons. FIF occurs mostly in the upper abdomen, while teratoma occurs in the lower abdomen and pelvis. The presence of vertebral bodies indicates that fetal development advanced to the stage of primitive notochord stage (the notochord is a precursor of vertebral body) [14]. However, it is important to differentiate between FIF and teratoma because of a slight risk of malignancy associated with retroperitoneal teratomas. In contrast, FIF is almost always benign with only one reported case of malignancy [15]. Clinically, FIF can be differentiated from a teratoma by the presence of vertebrae and limbs.

The presence of vertebrae signifies that the stage of gastrulation is already passed and reflects FIF's derivation from the primitive streak. Therefore, FIF usually arises from a zygote at the primitive streak stage and the fetiform mass develops to a degree similar to a normal fetal development [16].

FIF should also be differentiated from meconium peritonitis with pseudocyst formation in the neonate and from ectopic pregnancy in adults. A clinical history of an elevated  $\beta$ -human chorionic gonadotrophin level and the

documentation of chorionic tissue can further differentiate FIF from ectopic pregnancy.

Imaging plays a very important role in the diagnosis of FIF. Abdominal radiographs are sufficient when vertebral columns and bony structures in the form of limb bones are identified. Nocera et al. [17] was the first to describe CT findings in FIF. CT images in FIF show a mass that consists of a round or tubular collection of fat surrounding a central bony structure. Moreover, the identification of vertebrae or long bones is essential for a tentative diagnosis of FIF. CT also helps in establishing the exact relationship between FIF and the surrounding structures and their displacement due to mass effect. Three-dimensional CT with volume rendering techniques depicts bony structures thoroughly, which was done in our case. MRI can be used as a problem-solving tool.

## Conclusions

FIF is a rare pediatric entity which is being recognized more frequently nowadays because of routine imaging. The most common location is the retroperitoneum but FIF can be seen in other sites as well. It must be differentiated from a teratoma, meconium peritonitis and abdominal ectopic pregnancy, which is quite easy with the availability of cross-sectional imaging techniques. Although it is essentially a benign condition treated by surgical excision, follow-up in certain cases may be necessary.

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## Competing interests

None.

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