

Revisiting fetus-in-fetu

Dilip Gude,^a Batta Ramasubba Rayudu,^b Dharam Bansal,^c Chennamsetty Sashidhar^d

From the ^aDepartment of Internal Medicine, ^bDepartment of Radiology, ^cDepartment of Pulmonology and Critical Care, ^dDepartment of Nephrology, Medwin Hospital, Nampally, Hyderabad, Andhra Pradesh, India

Correspondence: Dilip Gude · Acute Medical Care, 3rd Floor, Medwin Hospital, Chirag Ali Lane, Nampally, Hyderabad, Andhra Pradesh 500 001, India · T: 091-9985445947, F: 091-40-23201120 · letsgo.dilip@gmail.com

Ann Saudi Med 2012; 32(4): 427-429

DOI: 10.5144/0256-4947.2012.427

Fetus-in-fetu is a rare congenital anomaly in which a malformed parasitic twin is found within the body of its partner. Less than 100 cases have been reported in published studies. Although it is a relatively benign condition, clinicians need to have a high index of suspicion for the associated complications that may arise. We report the case of an infant presenting with jaundice and steadily growing abdominal mass, who was diagnosed with fetus-in-fetu syndrome. We review the published studies and discuss the pathophysiology, complexities, and management options.

Fetus-in-fetu is an extremely uncommon cause of abdominal mass in the neonate, presenting as an encapsulated and pedunculated vertebrate fetoid mass in the abdomen usually from birth.¹ Less than 100 cases have been reported in the published studies. There is a male preponderance (male: female 2:1),² and about two-thirds of the cases present before 2 years of age. The presence of such an entity may lead to various manifestations secondary to mass effect on the adjacent viscera (obstructive jaundice in our case). We report a case of an infant presenting with mass abdomen and jaundice, and on evaluation was found to be fetus-in-fetu. We review the published studies and attempt to throw light on the possible pathophysiologic mechanisms, the manifestations, and management.

CASE

We report a case, in retrospect (1974), of an 11-month-old infant who presented with mass abdomen, failure to thrive, recurrent vomiting, and jaundice from about 4 months of age. He was a product of a non-consanguineous marriage and was born full term by normal spontaneous vaginal delivery. The abdominal mass had been steadily increasing in size with progressive worsening of the associated symptoms. The weight of the baby was 7.8 kg. The examination revealed icterus and mild pallor. Vital signs were unremarkable. The abdominal exam revealed an irregular mass on palpation, spanning from the left hypochondrium to the iliac fossa, and multiple irregular, firm to bony masses palpable all over the ab-

domen. A plain abdominal x-ray taken in 1974 showed multiple calcific/ossific densities on either side of the abdomen with evidence of long bones of limbs (**Figure 1**). (Further investigations like computed tomography (CT) and ultrasonography (USG) were not available at that time). The intravenous pyelogram depicted good excretion on both sides, showing a normal collecting system on the left side with evidence of hydronephrosis

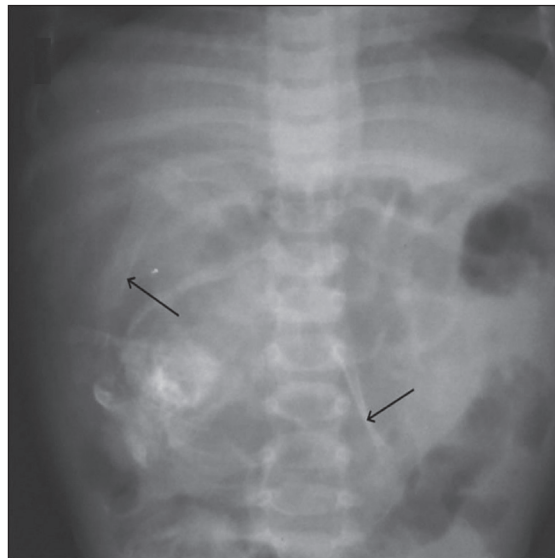


Figure 1. Plain abdominal x-ray showing multiple calcific/ossific densities on either side of the abdomen with evidence of long bones of limbs.



Figure 2. Radiograph of the specimen (post operative) delineating the fetus's spine (broken arrow), upper and lower limbs (thin arrows) with evidence of anencephaly (thick arrow).

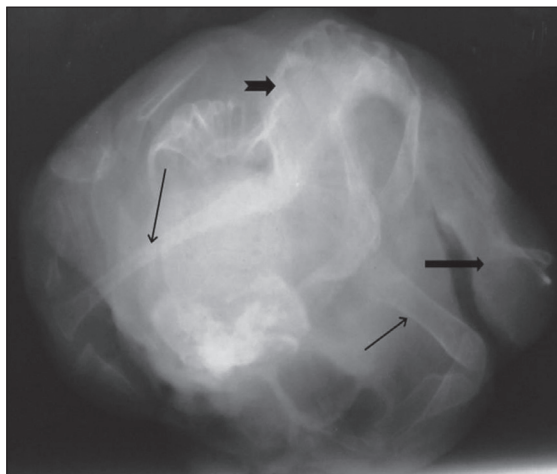


Figure 3. Post-operative radiograph of the specimen confirming Fetus-in-fetu disorder with evidence of spine (broken arrow), upper and lower limbs (thin arrows) and anencephaly (thick arrow).

and upward and lateral displacement of the right kidney by the calcific/ossific density mass. The patient's liver function tests were deranged (total bilirubin 5.3 mg/dL [normal 0.3 to 1.9 mg/dL], direct bilirubin 3.8 mg/dL [normal 0 to 0.3 mg/dL], alkaline phosphatase 385 U/L [normal 45-125 U/L]). Other lab tests were essentially normal. The surgical evacuation of the mass grossly showed features of a fetus having upper and lower limbs with anencephaly. Radiographs of the specimen further delineated the fetus's spine (broken arrow in Figures 2 and 3), upper and lower limbs (thin arrows in Figures 2 and 3), and evidence of anencephaly (thick arrow in Figures 2 and 3). On follow-up after 2

weeks, the patient's jaundice significantly improved and he was doing well.

DISCUSSION

Fetus-in-fetu is hypothesized to result from an unequal division of the totipotential inner cell mass of the developing blastocyst, leading to the inclusion of a smaller cell mass within a maturing sister embryo.³ The aborted or suppressed monozygotic twin (monoamniotic, monochorionic) later develops vitelline circulation anastomoses⁴ ultimately resulting in partial or complete external morphologic characteristics of a human form. Although a single fetus-in-fetu is the usual presentation, there have been reports of 2,⁵ 3,⁶ 5⁷ or more fetuses in the published studies. The vascular dominance of the host twin or, at times, an inherent defect in the parasitic twin will hamper the growth of the fetus-in-fetu which, initially in utero, parallels that of the host twin. Although an accurate preoperative diagnosis is not difficult most of the time, in some cases only a laparotomy can clinch the diagnosis. The diagnosis in our case was especially difficult, given the paucity of investigations at that time (1974). It is mostly found in the retroperitoneum, but has also been reported to occur in the cranial cavity,⁸ lateral ventricles, scrotum,⁸ testicles,⁹ pelvis,⁶ iliac mesentery,¹⁰ adrenal, coccyx,¹¹ and inguinal region.¹² The symptoms (from the mass effect) usually are abdominal distention, difficulty in feeding, vomiting, jaundice, and urinary retention. Differentiating from the much more frequent, highly organized, and well-differentiated teratoma, which is of paramount importance as the prognostic implications vary drastically. Grossly, the amniotic-like sac appearance, fine hair-bearing skin covering the whole specimen, and vestigial limbs or organs appropriately arranged with respect to the vertebral axis help in distinguishing it from a teratoma. Most authors opine that the presence of a vertebral column/neural tube, an amnion or umbilical cord, tissue indigenous to an embryo/fetus, and a high degree of structural organization (organoaxial orientation) differentiates the entity from teratomas. There are reports of "pseudofetu" or a sacrococcygeal teratoma without a vertebral column, but with rudimentary organs and other such presentations that argue the possibility of an overlap or entities representing the same pathology at different stages of maturation (about 9% of the cases do not have a well-defined vertebral axis). There have been case reports of a fetus-in-fetu and a teratoma occurring in the same patient reinforcing such theories. Both these entities may present with different degrees of spinal dysgenesis or residual posterior enteric remnants secondary to early focal disturbance of endodermal-ectodermal dif-

ferentiation.¹³ The other differentials include Wilm tumor, neuroblastoma, and hydronephrotic kidney.

Almost every organ in fetus-in fetu is deformed due to hypoxia, with anencephaly being common. Although the body is closed most of the time, ventral defects, like omphalocele, are not uncommon.⁷ Usually, the host does not manifest with any anomalies other than those related to the presence of a space-occupying lesion. Malignant degeneration has never been reported and appears highly unlikely. Saponification and skeletonization of fetuses, as occurs in a longstanding abdominal pregnancy, have not been described in cases of fetus-in-fetu for mechanisms that are unclear.¹⁴ USG commonly shows a well-delineated capsule, with an echogenic mass suspended/partially surrounded in fluid and oc-

asionally a rudimentary spine.¹⁵ CT and other imaging modalities may better delineate the long bones, spine, and appendages of the mass.

Treatment includes excision of the fetus with its capsule, and usually the mass is easily removed from the loosely adherent adjacent structures. The complete excision curbs the problems associated with increasing size, local pressure, hemorrhage into cystic mass, and uncertainty about diagnosis. The outcome is generally excellent. The child needs to be adequately followed up. Fetus-in-fetu is a very rare entity, but warrants a place in the differential diagnoses of abdominal mass in a child. Although mostly benign, the possible co-occurrence of a teratoma mandates surgical removal of the mass and the prognosis is mostly good.

REFERENCES

1. Mohta A, Shrivastava UK, Sodhi P, Upreti L. Fetus in fetu: A case report. *Pediatr Surg Int* 2003;19:499-500.
2. Hoeffel CC, Nguyen KQ, Phan HT, Truong NH, Nguyen TS, Tran TT, et al. Fetus in fetu: A case report and literature review. *Pediatrics* 2000;105:1335-44.
3. Boyce MJ, Lockyer JW, Wood CB. Foetus in foetu. Serological assessment of monozygotic origin by automated analysis. *J Clin Pathol* 1972;25:793-8.
4. Kim OH, Shinn KS. Postnatal growth of fetus in fetu. *Pediatr Radiol* 1993;23:411-2.
5. Gross RE, Clatworthy HW Jr. Twin fetuses in fetu. *J Pediatr* 1951;38:502-8.
6. Lee EY. Fetus-in-fetu. *Arch Dis Child* 1965;40:689-93.
7. Kimmel DL, Moyer EK, [1]et al. A cerebral tumor containing five human fetuses: A case of fetus in fetu. *Anat Rec* 1950;106:141-58.
8. Kakizoe T, Tahara M. Fetus-in-fetu located in the scrotal sac of a newborn infant: a case report. *J Urol* 1972;107:506-8.
9. Alpers CE, Harrison MR. Fetus in fetu associated with an undescended testis. *Pediatr Pathol* 1985;4:37-46.
10. Numanoglu I, Gokdemir A, Oztop F. Fetus in fetu. *J Pediatr Surg* 1970;5:472-3.
11. Narasimharao KL, Mitra SK, Pathak IC. Sacrococcygeal fetus-in-fetu. *Indian Pediatr* 1984;21:820-2.
12. Knox AJ, Webb AJ. The clinical features and treatment of fetus in fetu: Two case reports and a review of the literature. *J Pediatr Surg* 1975;10:483-9.
13. Bentley JF, Smith JR. Developmental posterior enteric remnants and spinal malformations: the split notochord syndrome. *Arch Dis Child* 1960;35:76-86.
14. Heifetz SA, Alrabeeah A, Brown BS, Lau H. Fetus in fetu: A fetiform teratoma. *Pediatr Pathol* 1988;8:215-26.
15. Daga BV, Chaudhary VA, Ingle AS, Dhamangokar VB, Jadhav DP, Kulkarni PA. Double fetus-in-fetu: CT scan diagnosis in an adult. *Indian J Radiol Imaging* 2009;19:216-8.