

# Fetus-In-Fetu: Unique Re- Case Report from Kingdom of Saudi Arabia and Review of the Literature

A Halabi<sup>1</sup>, U Khan<sup>\*2</sup>, Kitar.M<sup>2</sup>, Khelifi M<sup>2</sup>, Al Oteibi.R<sup>2</sup>, Al Thobaiti.F<sup>2</sup>, Krichen.I<sup>2</sup>, A Amin<sup>1</sup>, S

Kaabi<sup>3</sup>, T M Alghamdi<sup>3</sup>, T H Elkhoully<sup>3</sup>, Moutaz H<sup>4</sup>, Abdelmoneim A<sup>4</sup>, Maazoun K<sup>2</sup>

<sup>1</sup> Department of Neonatal Intensive Care, Alahda Armed Forces Hospital, Taif, Kingdom of Saudi Arabia (KSA)

<sup>2</sup> Department of Paediatric Surgery, Alahda Armed Forces Hospital, Taif, Kingdom of Saudi Arabia (KSA)

<sup>2</sup> Department of Radiology, Alahda Armed Forces Hospital, Taif, Kingdom of Saudi Arabia (KSA)

<sup>4</sup> Department of Histopathology, Alahda Armed Forces Hospital, Taif, Kingdom of Saudi Arabia (KSA)

**\*Corresponding Author:** Dr. Ubaidullah Khan, Pediatric Surgery, Department of Surgery, Alhada Armed Forces Hospital, Taif, Kingdom of Saudi Arabia (KSA), Tel: 00966547005971, Email: ubaidafr@gmail.com

**Received:** January 31, 2019; **Published:** March 06, 2019

## Abstract

Fetus in Fetu (FIF) is an extremely rare congenital anomaly in which a malformed parasitic fetus is found within the body of his living twin. We RE-report case of FIF from Saudi Arabia, a newborn male who was discovered to have a large abdominal mass born in our hospital and then seen in Nursery unit. Imaging studies confirmed the presence of a large retroperitoneal fetus in fetu with significant mass effect of the adjacent structures. Surgical removal was performed and pathology confirmed the diagnosis. This unique pathology is rarely reported in literature with only 200 reported cases. Our department is one of others authors to report, describe the demographics, updated genetic findings, pathology and outcomes of this unusual tumor and discuss off fail parasitic twinning and teratomas that may warrant a longer follow-up.

**Keywords:** Fetus in fetu, Retroperitoneal tumor, Abdominal mass, Parasitic twinning, Mature teratoma

## INTRODUCTION

Fetus-in-fetu (FIF), is a term for a still not yet clearly defined rare pathology with overlap with mature teratomas [1,3,4]. It has an estimated incidence of around 1/500,000 births [13]. This is the unique such case that we have reported from Kingdom of Saudi Arabia. In 2015, Romeo published his review for 95 FIF patients published in between 1999 and 2015 while the totally reported cases including the mature teratomas are less than 200 cases [2].

Many authors used both terms alternatively but indeed FIF can be differentiated from a mature teratoma by the presence of axial skeleton and limb buds which are fulfilled in our report [3-16]. Embryologically, FIF can be explained by an incompletely developed diamniotic monochorionic monozygotic twin in the body of his normally developed sibling [28, 29, 30, 31].

## CASE REPORT

A full-term male baby delivered at 38-weeks gestational age to un-booked mother, with birth weight of 3.3 kg, was discovered soon after birth in our nursery unit to have an abdominal mass. Only one antenatal ultrasound was done at 22 weeks gestational age which showed no significant abnormalities.

On examination, mildly distended abdomen with a soft mass in the upper middle part of abdomen, 10x10 cm, was palpated. Other body system review revealed no abnormalities apart from bilateral undescended testis.

X-ray showed; intra-abdominal calcifications and pushed gut shadow more to the left hypochondrium [fig.1]. Ultrasonography showed an ill-define hypoechoic mass in left upper and middle abdominal region, measuring 8x5cm, heterogeneous with cystic component and focal areas of calcifications, mostly boney [fig.2].

Computed tomography (CT) scan showed apparently large encapsulated heterogenous retroperitoneal mass,

8.2x5.5x6.1cm in transverse, anteroposterior and coronal measurement [fig.3]. The mass was displacing the left kidney posterior and gut to side, superior mesenteric vessel traversing and stretching out around, vascular pedicle from celiac vessel was assumed. The mass consisted of multiple bones and soft tissue suggestive of limbs bones, pelvic bones, vertebral column, and well-defined axial skeleton. The 3-dimensional reformats showed fetus pelvis, lumbar spine and clear limbs bone structure which in favor of fetus-in-feto [fig.4].

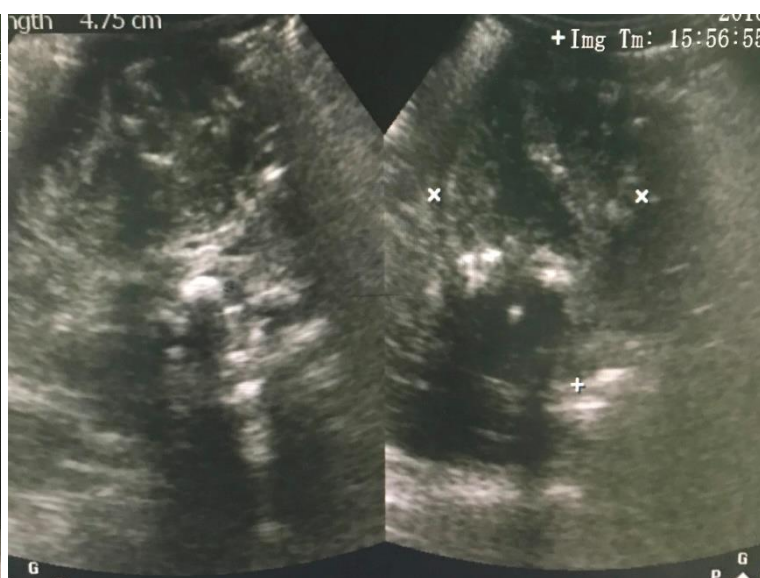
Further laboratory testing was done including alpha fetoprotein, carcino-embryonic antigen, neuron specific antigen,  $\beta$ -human chorionic gonadotropin and vanillylmandilic acid-creatinine ratio which all came out to be normal.

An elective laparotomy was performed with a transverse incision where a well-encapsulated, partially cystic retroperitoneal mass was found occupying most of the abdomen and displacing all abdominal structures including;

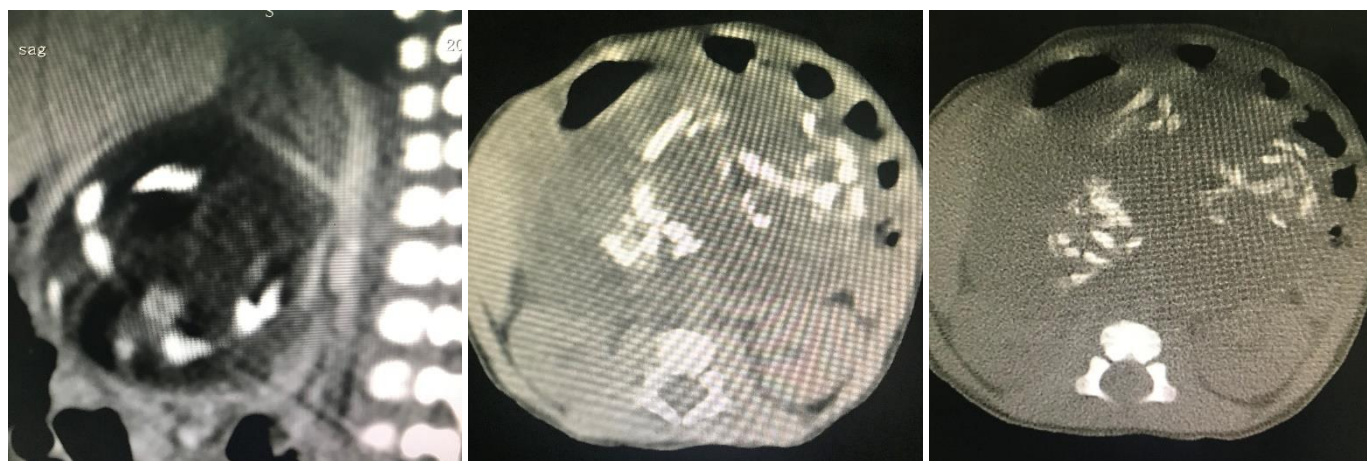
the left kidney, duodenum and liver laterally and teraservers mesentery structures anteriorly resulting in fixed to underlying structures like main vessels and pancreases. A well-defined vascular pedicle, origin of artery from superior mesenteric and venous from superior vena cava was identified supplying the mass, and the mass was removed en bloc [fig.5]. The gross and histological features were consistent with fetus in fetu, with the mass corresponding to an incompletely developed twin fetus. Both lower limbs had pincer like digital extensions, buttock and gentile mark present, whereas the upper limbs on the left side had axial as well as digital extension while small bud only on the right [fig.6,8]. Head with axial skeleton, facial marks and umbilical cord was also seen. Cross section showed tissue and gut structures inside the body with axial bones [fig.7]. The postoperative period was uneventful and the patient was discharged at 7 days post operatively with follow up appointment. The follow up time is 3 months since surgery, but symptoms free and need to monitor further.



**Fig.1:** X-ray shows calcification

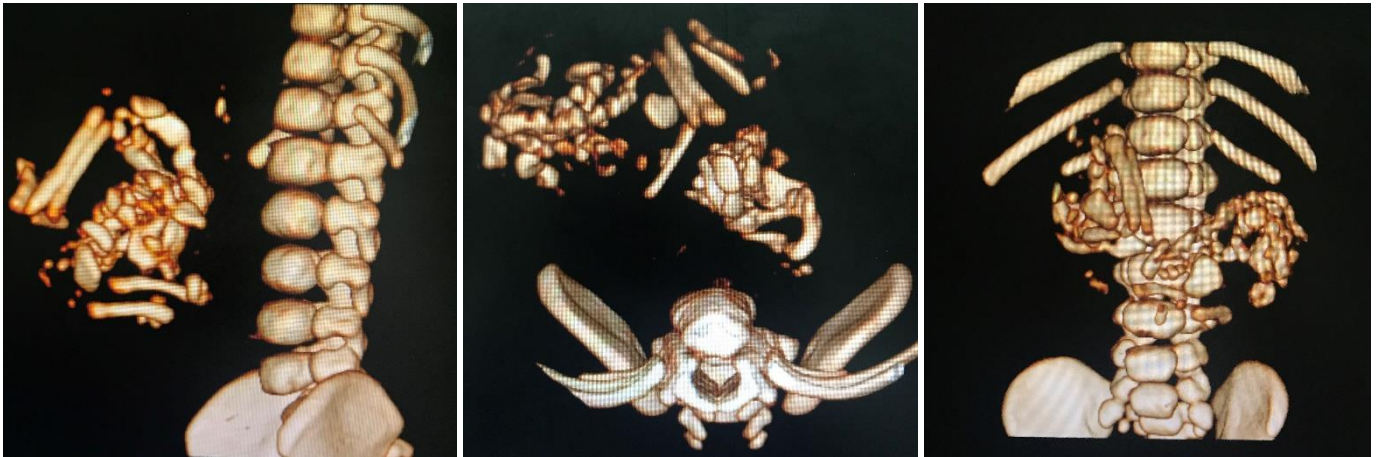


**Fig.2:** Ultrasonography of Fetus-in-feto, shows bone and mass

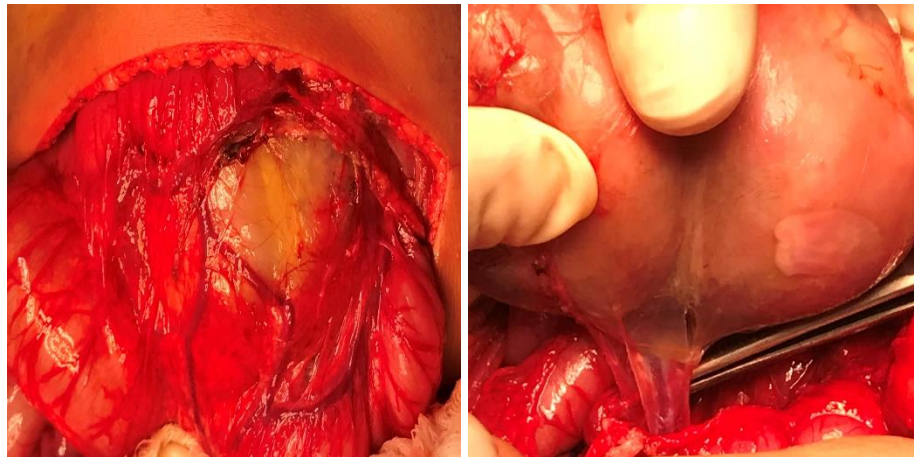


**Fig.3:** Axial images of CT scan show well structure consist of limbs bones and spine, fetus-in-fute

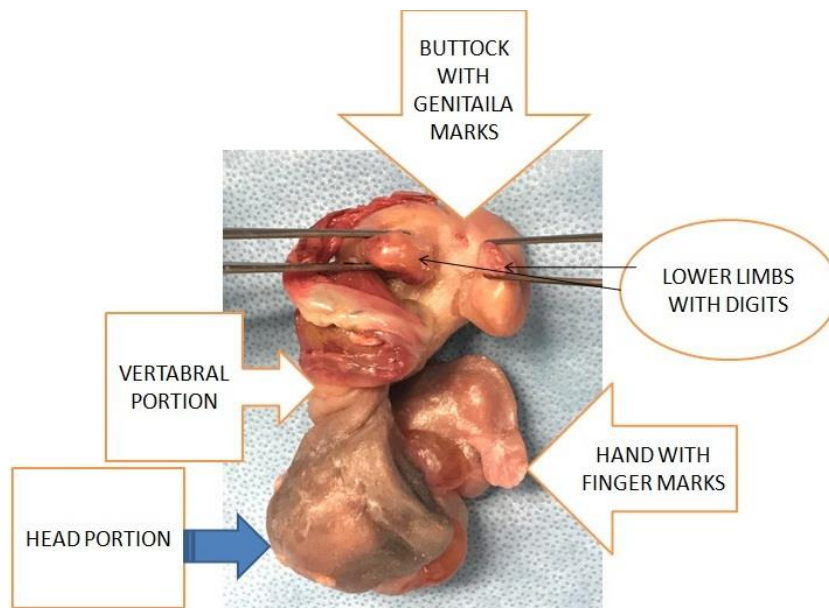




**Fig.4:** Three-dimensional reformats showed spatial relationship between child's lumbar spine and with visualized portion of the axial skeleton of the fetus in fetu



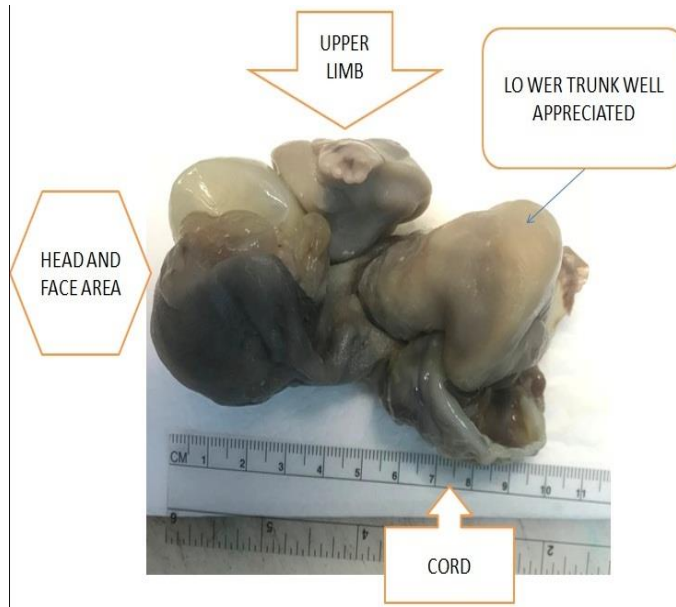
**Fig.5:** Retroperitoneal adherent to posterior structures, crossing vessel of mesentery which was saved and feeding vessel was lighted



**Fig.6:** Fetu-in-fetus



**Fig.7:** Ttransected portion, showing inside pathology



**Fig.8:** Complete gross pathology of our case

## DISCUSSION

In our case, radiological and surgical findings along with histopathology confirmed the diagnosis of fetus in fetu. The findings in favor of the fetus in fetu were the following:

a) There was heterogenous mass in retroperitoneal region, common area as reported before [2,20]. This mass was having complete axial skeleton, vertebral part and large formed long bone of the limbs. There was well defined long bone with attached finger, buttock with gentile and scalp area with facial portion. Umbilical cord attachment can be appreciated clearly [fig.6,7,8].

(b) There was also evidence of cartilage, hairy areas on head, covering skin. On cross section gut and other tissue is present. Microscopically, components of all three germ layers and under develop-twins were present [fig.7].

In the presence of above finding, we can say that the vertebra with an appropriate arrangement of limbs relative to the vertebral column identifies this specimen as a fetus-in-fetu, not features of a teratoma [1,2,3,4]. The requirement of an axial skeleton in fetus-in-fetu is because of a fact of embryogenesis which formed during the primitive streak phase of development. For this reason, it would have to progress through the primitive streak stage of embryogenesis to have axial skeleton [32,33].

FIF is a rare condition in which a malformed vertebrate fetus with organogenesis is found inside the body of its partner, usually in the abdominal cavity. It represents an aberration of

diamniotic, monochorionic, monozygotic twinning in which unequal division of the totipotent inner cell mass of the developing blastocyst leads to the inclusion of a smaller cell mass within a maturing sister embryo. Some authors believe that FIF is a type of mature teratomas.

On other hand, teratomas arise from the uncontrolled growth of pluripotent stem cells and develop without the organization imparted by embryogenesis [13,18]. Therefore, an axial skeleton within a mass represents a fetus-in-fetu not congenital teratoma. Upon review of the literature, the gross picture of presentation of this case is typical of fetus-in- fetu [1-6]. As compared to our case, fetus-in-fetu occurs predominantly in the upper retroperitoneum [2,20].

Second time Romeo C et al in 2015 published his reviewed of cases from 1999 to 2015, total 95 cases, before in 2000 also similar reviewed done by Hoeffel et al, which differentiate FIF from teratomas quit well [2,29]. The pathogenesis remains unknown, different theories but happened because of a diamniotic, monochorionic, monozygotic twin that becomes included in its host during the process. The inclusion in the sister embryo is speculated to be because of a persistent anastomosis of the vitelline circulation during development. The vitelline circulation develops into the superior mesenteric artery later in development, thereby explaining these lesions frequent appearance in the upper retroperitoneum [15,21].

This vascular disruption then results in incomplete morphogenesis and/or necrosis of the incorporated twin and leads into ill-formed mass with various developed structures [1]. Some reported cases of fetus-in-fetu does not exclusively

present in the abdomen, sites have included the cerebral ventricles [22], liver [9,19], sacrum [23], pelvis [2,3,13,24], oral cavity [7], skull [6], chest [1] and scrotum [25]. There has been no proposed etiologic explanation for these unusual cases.

Currently, it appears that FIF and teratoma are not considered as two distinct entities; pathology is identical but at different stages of maturation and features is not similar. Gonzalez C in 1982 proposed another definition: "Fetus in fetu is applied to any structure in which the fetal form is in a very high development of organogenesis and to the presence of vertebral axis" [34]. In contrast, Spencer in 2001, had mentioned that the fetus in fetu must have one or more of the following: (a) enclosed within a distinct sac, (b) covered by normal skin, (c) have grossly recognizable anatomic parts, (d) attached to the host by blood vessels, and (e) either be located immediately adjacent to one of the sites of attachment of conjoint twins or be associated with the neural tube or the gastrointestinal system [35].

Treatment for fetus-in-fetu is surgical. In contrast, teratomas, 6% to 10% of which are malignant [26], fetus-in-fetu is a benign disease. The most frequent symptoms reported are distension, palpable mass, emesis, poor feeding, and dyspnea [13]. One isolated case of malignancy 4 months after resection of a fetus-in-fetu [27] has been reported, stress on complete resection followed by postoperative surveillance of tumor markers. Although this rare entity still need more accuracy in diagnosis, However, analyzing the pathological controversy behind FIF, Willis in 1935, the 'features distinguishing a fetus-in-fetu from a highly differentiated teratoma are as follows: (1) There must be a separate spinal column, which demonstrates that the fetus has passed through a primary stage after gastrulation, involving formation of the neural tube, metamerization, and symmetrical development around this axis; and (2) the organs must have developed in a synchronized manner so that all have achieved the same degree of maturation [32,33].

Fetus-in-fetu present as an asymptomatic abdominal mass that is found in an infant during a scheduled physical examination [1]. The case presented above is typical of this presentation, although there have been several exceptions found in the literature, limitation of investigation at time, in this new era way not diagnosed on antenatal scan, still hurdles exists [2]. There are cases which present or diagnosed too late, 36-year old with mass abdomen [36], in contrast our case was spotted soon after birth. Second in our case the imaging which was done reached to final diagnosis before plan to proceed for surgery [fig.3,4], as compared to some reported cases where teratomas could not be separated [3,4]. The 3-D reconstruction of CT images was very helpful to well differentiate between FIF and teratomas, which help in pre-operated preparation and also family to take on board [fig.4].

Further to clarify FIF from teratomas, well vascular supply and

histopathology in our case helps to differentiate both quite clear. The artery supply from superior mesenteric artery (SMA) and venous to superior vena cava, one of silent feature of FIF, not describe in many cases [fig.5]. Gross cut section and tissue analysis was of high valuable in our case of FIF [fig.6,7,8]. Hens after all through analysis we can say our case was unique, with well vascular pedicle, artery from SMA and venous from superior vena cava, this feature are not described in many cases which are reported intra-abdominal FIF.

## CONCLUSION

In summary, accepted criteria of an intraabdominal fetus-in-fetu are clearly defined [1-16]. These criteria include presence of an axial skeleton, limb buds appropriately arranged relative to the axial skeleton, and karyotyping consistent with that of an included twin. The past literature after reviewed on fetoin-fetus and teratoma, show mix reporting between fetus-in-fetu and teratomas. This discussion clarified great difference among the two pathologies in detail. Treatment consists of complete resection.

**Funding:** The authors have not received any funding for this study.

### Ethics Approval and Consent to Participate:

The study was approved by the Alhada Armed Forces Hospital research and ethical committee.

**Conflicts of Interest:** authors declare no conflicts of interest

**Acknowledgements:** Thankful to family of child willing to publish these pictures and their kind response in this regard as well Operating and Neonatal nursing staff who work at best. This case report is presented in many forms and sends for short publication by other not concern with this publication.

## REFERENCES

1. Mauricio A. Escobar, Jon E. Rossman, Michael G. Caty. Fetus-in-fetu: report of a case and a review of the literature. *Journal of Pediatric Surgery* (2008) 43, 943–946.
2. Lindsey M. Prescher, William J. Butler, Tyler A. Vachon, Marion C. Henry, Thomas Latendresse, Romeo C. Ignacio Jr. Fetus in fetu: Review of the literature over the past 15 years. *J Ped Surg Case Reports* 3 (2015) 554-562.
3. Bindev Kumar, Amit Kumar Sinha, Tribhuvan Kumar, Prem Kumar, Chandni Sinha, Anil Kumar. Definition of "fetus in fetu" needs revision?: A case report and review of literature. *J Ped Surg Case Reports* 12 (2016) 24-26.
4. A. Landmann, J.L. Calisto, M. Reyes-Múgica, D. Thomas, M.M. Malek. Fetus-in-fetu presenting as a cryptorchid testis and abdominal mass: A report of a case and review of the literature. *J Ped Surg Case Reports* 13 (2016) 38-40.
5. Chowdhury Goutam, Francis Msumbe Banda. Fetus-in-fetu in Botswana. *Journal of Pediatric Surgery Case Reports* 24 (2017)



- 46-48.
6. L. Fieveta, N. Serratriceb, N. Degardina, G. PechGourgc, G. Lenac, D. Scavardac. Multisurgical approach for recurrent fetus-in-fetu of the skull. *Journal of Pediatric Surgery Case Reports* 30 (2018) 28–31.
7. NurudeenToyinAbdulraheem, Abdulrasheed A. Nasir, Lukman O. Abdur-Rahman, Oluwaseun R. Akanbi, Moses O. Olanrewaju, Musliamat A. Alada, OlanrewajuO. Oyedepo, TemitopeObasa, James O. Adeniran. Oral fetus-in-fetu: A case report. *J PedSurg Case Reports* 3 (2015) 171-173.
8. Ghulam Mustafa, Bilal Mirza, Shahid Iqbal, Afzal Sheikh. A Case of Fetus in Fetu. *APSP J Case Rep* 2012; 3: 9.
9. Al-Baghdadi R. Fetus in fetu in the liver: case report and review of the literature. *J PediatrSurg* 1992;27(12):1491-2.
10. Ariane Brand, Maria Cecilia Alves, Carla Saraiva, Pedro Loi'o, Joa'õGoula'o, Joa'õ Malta, J. Martins Palminha, and Mena Martins. Fetus in Fetu—Diagnostic Criteria and Differential Diagnosis—A Case Report and Literature Review. *Journal of Pediatric Surgery*, Vol 39, No 4 (April), 2004: pp 616-618.
11. MithatGunaydin , FatmaCakmakCelik , BurakTander , AsudanTugceBozkurter ,YurdanurSullu , SancarBaris , RizaRizalar , SukruKucukoduk. Two cases of fetus in fetu. *Journal of Pediatric Surgery* (2011) 46, E9–E12.
12. Jeyanthi N. Narayanasamy, Mohan ArunasalamNallusamy and NurDalizaBaharuddin. Fetus-in-fetu: a pediatric rarity. *JSCR* 2014; (3 pages).
13. Thakral CL, Maji DC, Sajwani MJ. Fetus-in-fetu: a case report and review of the literature. *J PediatrSurg* 1998;33(9):1432-4.
14. Kumar AN, Chandak GR, Rajasekhar A, et al. Fetus-in-fetu: a case report with molecular analysis. *J PediatrSurg* 1999;34(4):641-4.
15. Iftikhar Ahmad Jan, Basmah Al Hamoudi, MurthiShiniwasFetus in Fetu or Teratoma: A Report of Four Cases and Literature Review. *Journal of Pediatric Surgical Specialties*. 2015, Volume 9 No. 1.
16. Iftikhar A Jan, Anwar-ulHag, Ahmad Sharif, Saifullah Khan, Ubaidullah Khan. EpigastricHeteropagus Parasitic twins: A Report of three cases. *journal of Pediatric Surgical Specialties*, 2008, vol. 2. no. 1.
17. Knox AJS, Webb AJ. The clinical features and treatment of fetus in fetu: two case reports and a review of the literature. *J PediatrSurg* 1975;10(4):483-9.
18. Eng HL, Chuang JH, Lee TY, et al. Fetus in fetu: a case report and review of the literature. *J PediatrSurg* 1989;24(3):296-9.
19. Magnus KG, Millar AJW, Sinclair-Smith CC, et al. Intrahepatic fetus-in-fetu: a case report and review of the literature. *J PediatrSurg* 1999; 34(12):1861-4.
20. Grosfeld JL, Stepita DS, Nance WE, et al. Fetus-in-fetu. *Ann Surg* 1974;180(1):80-4. (21) George V, Khanna M, Dutta T. Fetus in fetu. *J PediatrSurg* 1983;18(3): 288-9.
21. Kimmel DL, Moyer EK, Peale AR, et al. A cerebral tumor containing five human fetuses. *Anat Rec* 1950;106:141-66.
22. Ouimet A, Russo P. Fetus in fetu or not. *J PediatrSurg* 1989;24(9):926-7.
23. Lee EYC. Foetus in foetu. *Arch Dis Child* 1965;40:689-93.
24. 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.
25. Fowler CL. Intraabdominal leg: unique variant of split notochord syndrome. *J PediatrSurg* 1998;33(3):522-4.
26. Hopkins KL, Dickson PK, Ball TI, et al. Fetus-in-fetu with malignant recurrence. *J PediatrSurg* 1997;32(10):1476-9.
27. Arlikar JD, Mane SB, Dhende NP, Sanghavi Y, Valand AG, Butale PR. Fetus in fetu: two case reports and review of literature. *PediatrSurgInt* 2009;25:289-92.
28. Hoeffel CC, Nguyen KQ, Phan HT, Truong NH, Sao Nguyen T, Tran TT, et al. Fetus in fetu: a case report and literature review. *Pediatrics* 2000;105(6): 1335-44.
29. Ji Y, Song B, Chen S, Jiang X, Yang G, Gao X, et al. Fetus in fetu in the scrotal sac: case report and literature review. *Medicine* 2015;94(32):1-7.
30. Khandaroo RG, Evan MG, Honore LH. Fetus-in-fetu presenting as cystic meconium peritonitis: diagnosis, pathology and surgical management. *J PediatrSurg* 2000;35(5):721-3.
31. Willis RA: The border land of embryology and pathology. 2nd edition ed. London, Dutterworths, 1962, pp442-462.
32. de Lagausie P, de Napoli Cocci S, Stempfle N, Truong QD, Vuillard E, Ferkadji L, et al. Highly differentiated teratoma and fetus-in-fetu: a single pathology? *JPediatrSurg* 1997;32:115-6.
33. Spencer R. Parasitic conjoined twins: external, internal (fetuses in fetu and teratomas), and detached (acardiacs). *ClinAnat* 2001;14:428e44.
34. Gonzalez-Crussi F. Extragonadal teratomas. *Atlas of tumor Pathology*. 2nd ser, fasc. 18. Washington DC: Armed Forces Institute of Pathology; 1982.
35. Sharma N, Verma P, Pathak P. 27 cm fetus in fetu presenting in a 36-year-old man: report of a rare case and brief review of literature. *Indian J Surg* 2007;69: 155-7

