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ORIGINAL ARTICLE

# Prenatal diagnostic clue for fetus in fetu

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**Abstract** The purpose of this paper is to define a prenatal diagnostic clue for fetus in fetu (FIF) based on the characteristic findings of four FIF cases studied by the authors. A retrospective analyses of prenatal ultrasonography (US), postnatal US, computed tomography (CT) scans and operative findings were carried out on each of the four FIF cases collected from a multi-center. Prenatal US findings for each of the four cases showed a fluid-filled sac with a solid portion "floating" within it. In addition, bony structures were found in the solid portion. In each of the four cases, the postpartum imaging studies were consistent with the prenatal US findings. Operative findings revealed a solid mass within a fluid-filled sac surrounded by a transparent membrane. The solid mass was connected to the membrane by a stalk. Histopathologically, the cells making up the sac were of the same type as those of the amnion, while the solid mass had the general characteristics of FIF. In conclusion, FIF can be suspected when prenatal US shows a solid mass with bony structures within a fluid-filled sac in a newborn.

**Keywords** Fetus in fetu · Prenatal ultrasonography · Preoperative diagnosis

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## Introduction

A monozygotic diamniotic twin internalized within the other is defined as fetus in fetu (FIF) [1]. In the past, the level of preoperative FIF diagnosis was low, but this has improved recently due to improvements in the technical skill of ultrasonography (US) and increased interest in prenatal examinations. In particular, computed tomography (CT) scans have greatly assisted accurate diagnosis. However, it remains difficult to differentiate between diagnoses of FIF, teratoma and cystic meconium peritonitis from radiologic findings, due to the fact that they all result in a calcified mass. Confirmative diagnosis can only be established by operative and histopathologic findings. FIF has often been misdiagnosed as a teratoma or a cystic meconium peritonitis due to its rarity.

Differential diagnosis is an important issue because FIF, teratoma and cystic meconium peritonitis are very different in terms of their respective disease courses. While teratoma has the possibility of malignant change, and cystic meconium peritonitis often has other combined abnormalities, FIF is almost always a benign disease. Therefore, accurate preoperative differential diagnosis can help to determine an accurate treatment plan. The authors in this paper describe the common characteristic of FIF as revealed by prenatal US, postnatal US, and by postnatal CT scans, and the operative gross findings.

## Materials and methods

The authors studied four cases of FIF confirmed histopathologically from a multi-center from 2001 to 2003. They were all born healthy without other problems. A retrospective analysis of prenatal US findings, postnatal US findings, postnatal CT scan findings, three dimensional (3-D) CT scan findings, and operative findings was carried out, and their common features are described.

The brief history of each case is shown in Table 1.

The study was approved by the Institutional Review Board at Yonsei University College of Medicine.

## Results

In prenatal US, the cystic mass of each case seemed to have a clear boundary and a fluid-filled cyst with a solid portion. The fluid-filled cystic portion was not lobulated and the floating solid portion contained multiple calcifications (Fig. 1a). The calcifications looked like multiple bony structures. Postnatal US were carried out for case 1 and case 3. Postnatal US findings did not differ from the prenatal US in both case 1 and case 3 (Fig. 1b). A postnatal CT scan was carried out in all four cases. CT scans were better able to reveal multiple long bony calcifications within the mass (Fig. 2a, b). In point of the bony structural characteristics, the calcifications were better seen in the 3-D CT scan than in regular CT scan (Fig. 2c). In the operative findings, the cystic mass of all four cases was covered with a transparent membrane (Fig. 3a) which was connected to the solid portion by a stalk (Fig. 3b, c). Rudimentary limb buds were observed on the solid portion (Fig. 3c). These findings matched the prenatal US findings. In cases 1, 2 and 3, the surface of the solid portion was covered by a skin-like layer (Fig. 3c). Histopathologically, the vertebral column, skin, cranium, osteochondral junction, enteric mucosa, respiratory mucosa, umbilicus, liver, ovary, amnion like membrane, and so forth were confirmed.

## Discussion

Willis [2] considered that the arrangement of limb structures around a vertebral axis is a step too far along in embryological development to be considered a teratoma. Until now, this definition was controversial.

FIF is a rare congenital anomaly whose incidence is 1 in 500,000 births [3] and a fewer than 100 cases have been reported [4]. The histopathologic diagnosis of FIF is confirmed by the presence of a vertebral axis, well-differentiated organs, and limb buds. Moreover, an amnion-like capsule, vascular bundle and umbilical cord are also included as diagnostic criteria [5].

A calcified intra-abdominal mass in the newborn may be a teratoma or cystic meconium peritonitis, but FIF should also be considered in differential diagnosis. FIF is almost always a benign disease except for one case where a malignant recurrence was observed after complete excision of FIF [6]. Because of FIF's benign nature, the preoperative distinction of FIF from teratoma and cystic meconium

**Table 1** Summary of four cases

| | IUP (wks) | kg | Type/delivery | Prenatal US (wks) | Postnatal US | CT scan | 3D CT scan | Figure |
|---|---|---|---|---|---|---|---|---|
| Case1 | $39^{+2}$ | 3.65 | NSVD | At 32, 36 | Done | Done | None | 1a, 2b, 3a |
| Case2 | $40^{+4}$ | 3.4 | NSVD | At $37^{+3}$ | None | Done | Done | 2a, 2c |
| Case3 | $37^{+5}$ | 3.5 | C/S | At 33 | Done | Done | Done | 1b, 3c |
| Case4 | 39 | 3.27 | NSVD | At $32^{+4}$ | None | Done | None | 3b |

*Wks* weeks, *NSVD* normal spontaneous vaginal delivery, *C/S* Caesarian section

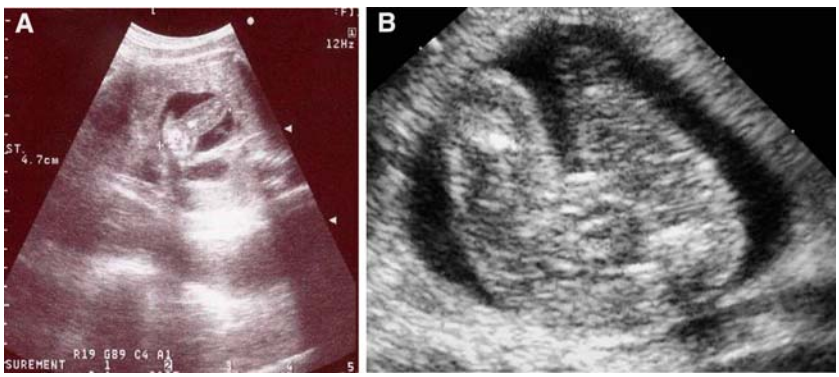


**Fig. 1** Prenatal US (**a**) and postnatal US (**b**) commonly show a calcified solid mass surrounded by a fluid-filled sac

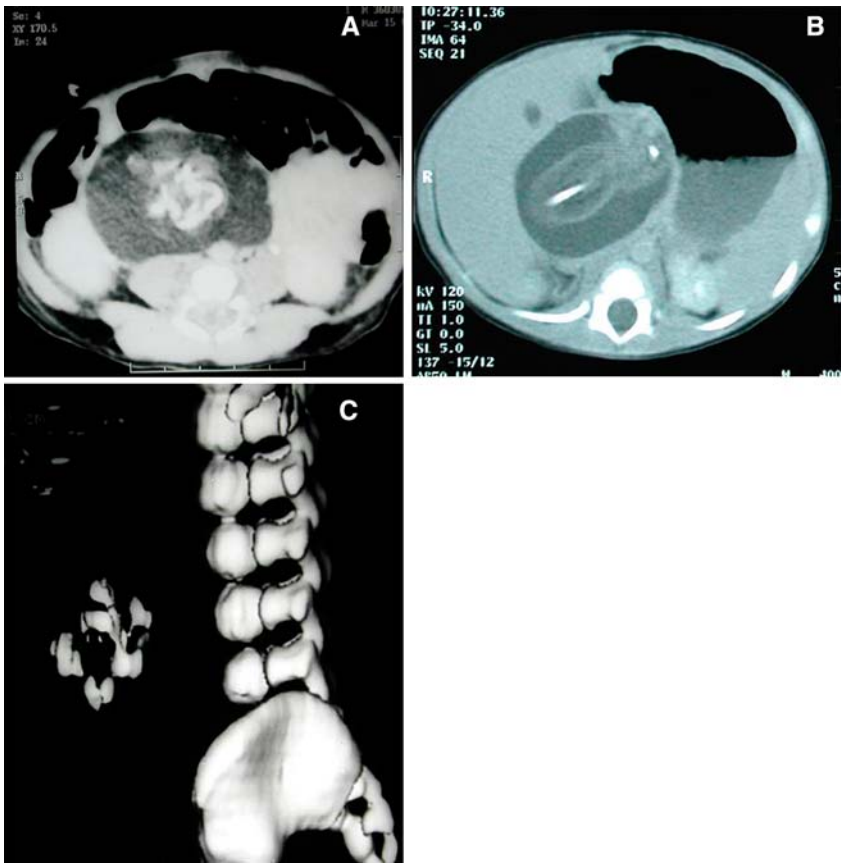


**Fig. 2** Postnatal CT scan (**a**) shows the same feature of US. The calcification of the cystic mass looked like a vertebra or long bone (**b**). Beside the maternal vertebrae, the bony structural characteristic of the calcification is better seen in the 3-D CT scan (**c**)

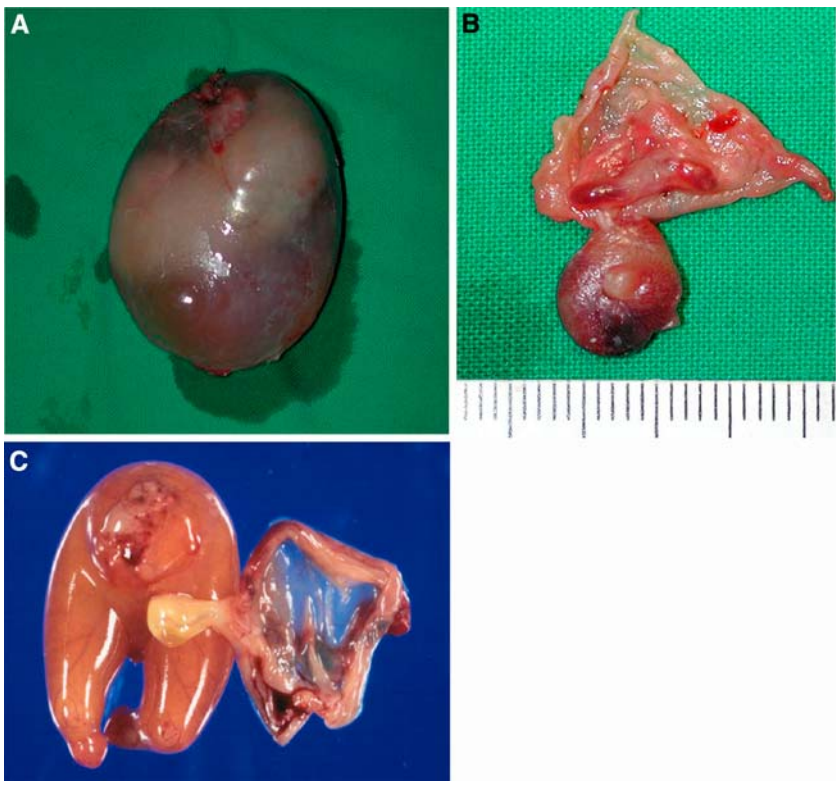


**Fig. 3** In the operative findings, the specimen indicates the transparent membrane (**a**) connected to the solid portion by a stalk (**b**). The solid portion has a number of rudimentary limb buds and vascular structures, which are covered with a skin-like surface (**c**)

peritonitis can help to determine an accurate treatment plan.

In the author's experience, there are several key radiologic and operative findings for each disease. The first clue is the overall composite feature of the solid portion and the fluid-filled cystic portion. In FIF, the mass was divided into two parts, the peripheral fluid-filled cystic portion and the central solid portion "floating" within. The cystic portion was not divided by a septum. A teratoma is different in that it is often a multi-loculated cystic mass, or a mixed mass of solid and cystic portion without clear border. The findings in cases of cystic meconium peritonitis are varied, but free air or ascites may be observed. The second clue is the type of calcifications found in the solid portion. Different opinions exist on this point, but CT and 3-D CT scans may distinguish the calcification of cases of FIF from those of a teratoma or cystic meconium peritonitis. In contrast with the bony calcification of FIF, the calcified features of teratoma have more of a tooth-like appearance, but in some cases of teratoma, separated bony structural calcifications were also seen. At this point, FIF is not always distinguished from a teratoma. Whereas those of cystic meconium peritonitis are amorphous, spiculated or deposited in a "peripheralized" capsule. The third clue is the site of occurrence. Eighty percent of FIF cases occur in the upper retroperitoneal space [7]. Other sites include the oral cavity, the scrotum, sacrum, adrenal gland and cranium [7–9]. In contrast, a teratoma is most commonly found in the sacrococcygeal, gonadal, mediastinal, and CNS. Only 5% of teratoma cases are found in the retroperitoneal region [10]. In cystic meconium peritonitis, the most common site of occurrence is intra-abdominal followed by the hollow viscus. The fourth clue is the operative findings of the specimen. The whole mass of FIF is surrounded by a transparent membrane which is connected to a solid portion by a stalk. The solid portion floats within a fluid-filled sac and may be covered with a skin-like structure. Histopathologically, the cells making up the membrane in the four FIF cases of this study were of the same type as those of the amnion.

In the four cases of this study, the common radiologic and operative findings were used to distinguish FIF from a teratoma or cystic meconium peritonitis. If more cases, including unusual cases, were observed, our consideration could have more objective validity. Nevertheless, given the rarity of FIF cases, an attempt has been made here with the means available to contribute to the body of academic research in this field, and it is submitted that these findings can reduce the misreading of a calcified mass found in a newborn, considering the possibility of such a rare diagnosis.

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