Fetus in fetu (FIF) is a rare congenital anomaly. Anomalies of inferior vena cava (IVC) in the host are rare and not reported in the literature. In this case report, the surgical management and the findings of a rare vascular anomaly of IVC in a 10-month-old male child with FIF are discussed. This case highlights the importance of antenatal ultrasonogram in the detection of FIF and to diligently look for structural anomalies of IVC in the host in such cases.

**Keywords:** Fetus in fetu, Inferior vena cava anomaly, infrarenal inferior vena cava thrombosis

## Case Report

A 10-month-old male child presented with right flank mass of 10 days duration, which was noticed by the mother. Antenatal ultrasonogram (USG) in the third trimester did not detect any abnormalities. On examination, the child was thriving well. A retroperitoneal mass of size 10 cm × 8 cm was palpable in the right hypochondrium, right lumbar, epigastric, and umbilical region. X-ray abdomen revealed soft-tissue shadow involving the right side of the abdomen with areas of calcification. USG abdomen revealed 10 cm × 8 cm hetero echoic mass with the displacement of the right kidney. Routine hematological investigations such as Hemoglobin, Complete blood count, ESR were normal. Tumor markers alfa-fetoprotein and beta-human chorionic gonadotrophin were within the normal limits. A diagnosis of retroperitoneal teratoma was suspected.

Contrast-enhanced computed tomography (CECT) abdomen and pelvis showed well-defined hetero-dense lesions arising from the retroperitoneum adjacent to the right kidney having formed axial skeleton and fat components. The diagnosis of FIF was established. Further, three-dimensional (3D) reconstruction CECT abdomen study revealed altered vascular anatomy of inferior vena cava (IVC).

During the laparotomy, FIF was found in the retroperitoneum. On the surface of FIF, a cord-like structure was found to be straddling across in the midline [Figure 1]. On dissection, the cord-like structure was found to be continuous with the confluence of both renal veins and coursing cranially as IVC (suprarenal IVC). Further dissection of the structure caudally revealed both common iliac veins coursing into the paravertebral area and disappearing. Based on the anatomical course, it was concluded that the cord-like structure was the hypoplastic IVC. The infrarenal IVC was devoid of blood and was found on top of the dumbbell-shaped FIF. This operative finding was in concordant with the preoperative 3D reconstruction image of the CECT abdomen. After opening the sac, FIF was excised after ligating the retroperitoneal feeding vessels. Histopathology revealed the following structures including the axial skeleton, rudimentary limbs, intestines, and skin lining. No lung, liver, and heart tissue were found.

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**DISCUSSION**

FIF results from unequal division of totipotent inner cell mass of the developing blastocyst leading to the inclusion of a smaller cell mass within a mature sister embryo. In 2001, Spencer redefined the criteria for FIF.\(^2\) It should have one or more of the following components: (a) be enclosed within a distinct sac, (b) partially or completely covered by normal skin, (c) have grossly recognizable anatomic parts, (d) to be attached to the host by only a few relatively large blood vessels e) either be located immediately adjacent to one of the sites of attachment of conjoint twins or be associated with the neural tube or gastrointestinal system. About 25% of cases reported of FIF have no axial skeleton.\(^3\) Our case satisfies all the above criteria for FIF.

This patient would be the first reported case of a host having an anomaly of the IVC due to FIF as a literature search did not reveal any similar report. During normal development of IVC, at 4 weeks of life, three distinct venous systems forms. The vitelline system draining the gut, the umbilical system draining the placenta, and the cardinal system draining the rest of the embryo. IVC develops from different segments of vitelline and cardinal veins. There are several anomalies of IVC, among which the development of infrarenal IVC is of particular interest in this case. The posterior cardinal vein along with the suprarenal part of the right and left supra cardinal veins forms the azygos and hemiazygos venous system, respectively. The infrarenal part of the supra cardinal vein disappears on the left side. The right-sided infrarenal part of the supra cardinal vein forms the infrarenal IVC [Figure 2]. Failure of development of infrarenal part of supra cardinal veins results in the absence or poor development of infrarenal IVC with preservation of suprarenal segment. This could be due to intrauterine or perinatal thrombosis of developing IVC.\(^4\) The developing FIF is the possible cause for the infrarenal IVC thrombosis in this case.

When infrarenal IVC is absent, the lower limb and pelvis venous return is through the iliac veins which, drains into the ascending lumbar veins, which convey blood from lower extremities to azygos and hemiazygos vein via anterior paravertebral collateral veins.\(^5\) 3D reconstruction images revealed these anomalies in our case [Figure 2]. These patients can present later in life with venous insufficiency or idiopathic deep venous thrombosis.

To conclude, FIF developing inside the abdomen of the host might have caused occlusion of the right supra cardinal veins in the early weeks of gestation. As a result, infrarenal IVC was replaced with a hypoplastic cord. This has not been described so far in the literature. Venous anomalies should be anticipated during the surgery of FIF to avoid major catastrophic events. Accidental ligation of any venous structure that is encountered during surgery of FIF should be avoided. Preoperative evaluation with CECT abdomen with 3D reconstruction is essential to detect this anomaly.

As a long-term follow-up, patients who have agenesis or hypoplasia of infrarenal IVC have a higher risk of developing deep venous thrombosis in lower limbs. This child is on close follow-up and possibilities of venous complications were explained to parents.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their
images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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