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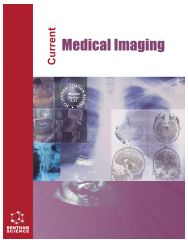

CASE REPORT

Analysis of Ileal Atresia from Prenatal Ultrasound to Postoperative Follow-up: Two Case Reports

Zimeng Lv[1,2], Hongyi Qu[3], Jingyuan Hu[1,2], Yue Dong[2] and Wei Liu[2,*]

[1]*Shandong First Medical University (Shandong Academy of Medical Sciences), Jinan, Shandong Province, China*
[2]*Department of Medical Ultrasound, The First Affiliated Hospital of Shandong First Medical University & Shandong Provincial Qianfoshan Hospital, Shandong Medicine and Health Key Laboratory of Abdominal Medical Imaging, Jinan, Shandong Province, China*
[3]*Department of Pediatric Surgery, The First Affiliated Hospital of Shandong First Medical University & Shandong Provincial Qianfoshan Hospital, Shandong Engineering and Technology Research Center for Pediatric Drug Development, Jinan, Shandong Province, China*

Abstract:

Background:

Congenital ileal atresia is a rare neonatal disease, the most common type of intestinal malformation in newborns, and one of the most common causes of congenital intestinal obstruction. It can cause various digestive system symptoms, including abdominal distension, vomiting, abnormal bowel movements, *etc*. In severe cases, it can be life-threatening. A prenatal ultrasound examination can assist clinical diagnosis of congenital ileal atresia, and those with a clear prenatal diagnosis should undergo surgical treatment after birth.

Case Presentation:

We have, herein, reported two cases of congenital ileal atresia, both of which showed fetal intestinal dilation (>7mm) and excessive amniotic fluid on prenatal ultrasound. Both newborns underwent surgical treatment after delivery and were confirmed to have congenital ileal atresia during surgery. Due to the different prenatal ultrasound manifestations of the two patients, they were divided into two different subtypes based on intraoperative manifestations. We observed significant differences in the prognosis of the two patients after surgery.

Conclusion:

Accurately locating and classifying ileal atresia using prenatal ultrasound is challenging; however, it plays an effective role in disease progression, gestational assessment, and prognosis. Accurately identifying intestinal diseases and/or the location of lesion sites through direct and indirect ultrasound findings in the fetal abdominal cavity is an important research direction for prenatal ultrasound.

Keywords: Ileal atresia, Prenatal diagnosis, Prognosis, Ultrasound, Anomaly, Prenatal ultrasound.

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1. INTRODUCTION

Ileal atresia is a rare congenital developmental anomaly. Ileal atresia is often associated with fetal intestinal perforation, and can also be associated with intestinal abnormalities, such as omphalocele, meconium peritonitis, volvulus, *etc*. Prenatal ultrasound examination can assist in the neonatal ultrasound characteristics, but there are significant difficulties in its classification and localization. However, prenatal ultrasound examination has significant diagnostic value for neonatal ileal atresia, as it can also assist in clinical evaluation of the gestational age of pregnant women and the prognosis of newborns.

2. CASE PRESENTATION

2.1. Case 1

A 25-year-old pregnant woman (G1P0), 29 weeks and 4 days into her pregnancy, visited our hospital for her first prenatal examination. The ultrasound revealed localized intestinal dilation in the fetus, with a width of about 2.1 cm. Additionally, there was pelvic and abdominal fluid accumulation, enhanced echoes in the local intestinal area, no

* Address correspondence to this author at the The First Affiliated Hospital of Shandong First Medical University, 16766 Jingshi Road, Jinan City, Shandong Province, China, 250000; Tel: 008615128990971; E-mail:lw_ivy@163.com

obvious calcification, and an abundance of amniotic fluid (Fig. **1A**). Four weeks later, a follow-up ultrasound showed the following changes: the abdominal cavity of the fetus was filled with dilated intestinal tubes, which were about 2.5 cm wide. There was a significant enhancement in the local echoes between the intestines (Fig. **1B**).

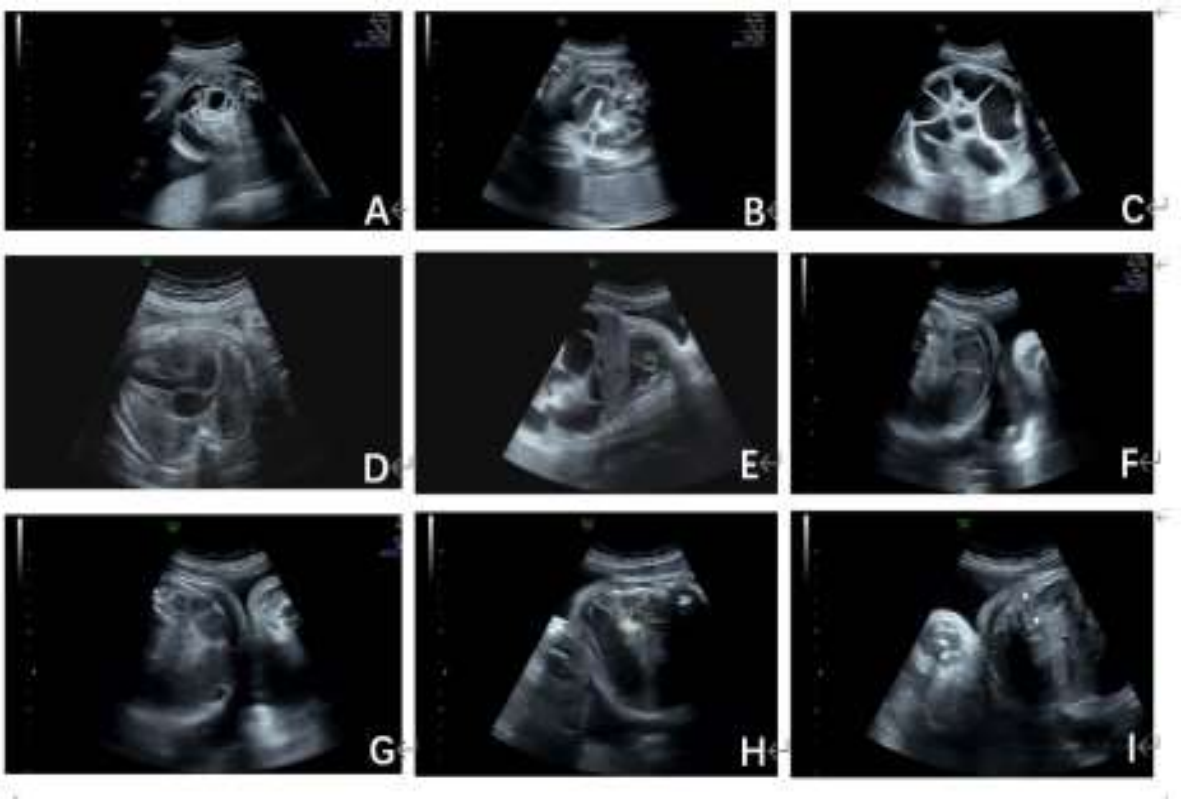

Fig. (1). (**A**) Fetal intestinal dilation, enhanced echo, and abdominal fluid accumulation at 29w+4 gestational age in case 1. (**B**) Fetal intestinal dilation at 34w+0 gestational age in case 1. (**C**) Fetal intestinal dilation at 35w+6 gestational age in case 1. (**D**) Increased cardiothoracic ratio in fetuses at 35w+6 gestational age in case 1. Sagittal view of pregnancy 35w+6 shows increased fetal abdominal pressure, elevated diaphragm, and chest compression in case 1. (**F**) and (**G**) Significant dilation of the local intestinal tract in the fetus of case 2. (**H**) and (**I**) A "bilateral" thickening of the local intestinal wall in the right lower abdomen of the fetus, with enhanced echogenicity in case 2.

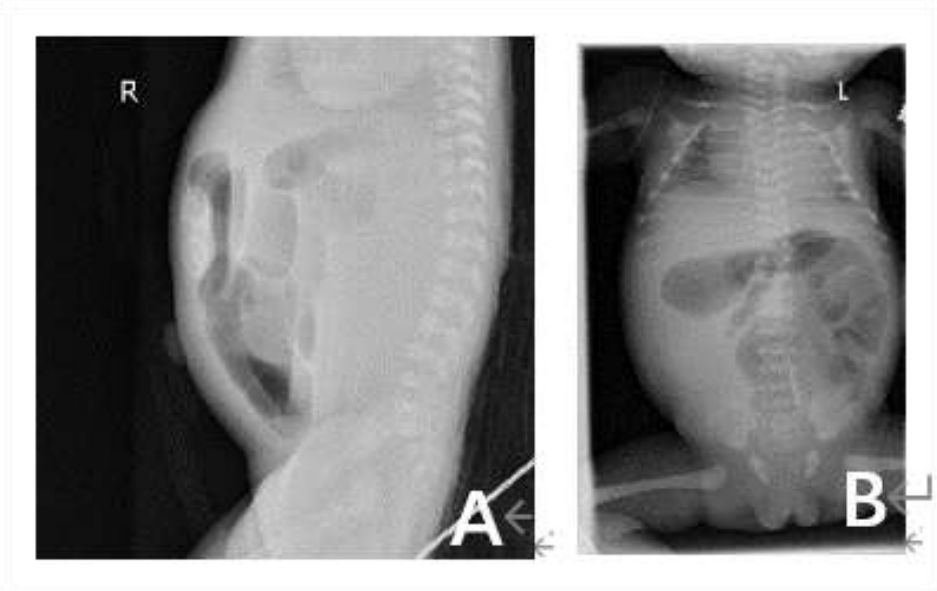

Fig. (2). (**A** and **B**) The anteroposterior and anteroposterior plain films of the newborn's abdomen in case 1 show dilation and accumulation of gas in the abdominal intestines, with visible gas-liquid level. Intestinal obstruction has been considered.

At 35 weeks and 6 days, another ultrasound examination revealed further developments: the abdomen of the fetus was obviously distended, with the intestines further expanded to a width of about 3.8 cm. The right side of the fetal heart was enlarged, exhibiting tricuspid valve regurgitation. The Pulsatility Index (PI) value of the fetal middle cerebral artery was reduced, while the Peak Systolic Velocity (PSV) was slightly higher. The amniotic fluid volume was increased (Fig. **1C - E**). Two days after this examination, the pregnant woman underwent a cesarean section. The newborn's Apgar score was as follows: 8 points in the first minute, with one point deducted for breathing and one point deducted for the skin; 9 points in the 5th and 10th minutes, with one point deducted for breathing.

During the neonatal postpartum examination, the following observations were made: the abdomen was swollen, and there were no gastrointestinal or peristaltic waves. The veins in the abdominal wall were visible, the abdomen was flexible, and crying was elicited upon touch. Bowel sounds were heard 1–2 times per minute, with a high pitch and poor continuity. Imaging examinations suggested intestinal obstruction (Fig. **2A - B**). The newborn underwent surgery 15 hours after the birth, revealing the following intraoperative findings: severe intra-abdominal intestinal adhesions, intestinal atresia occurring 50 cm from the Qurji ligament, evident proximal intestinal expansion with a diameter of approximately 6 cm, and distal intestinal collapse of approximately 1 cm. Initially, necrosis, torsion, and local calcification were observed. The mesentery remained intact at the severed end. The diagnosis confirmed a type IIIb ileal atresia. The distal intestinal canal was approximately 35 cm from the ileocecal portion and filled with hard fetal stools. The mesentery was poorly developed, and the distal intestinal canal was spirally arranged (Fig. **3A - D**). In the follow-up period, the newborn spontaneously defecated on the 17th day after the surgery.

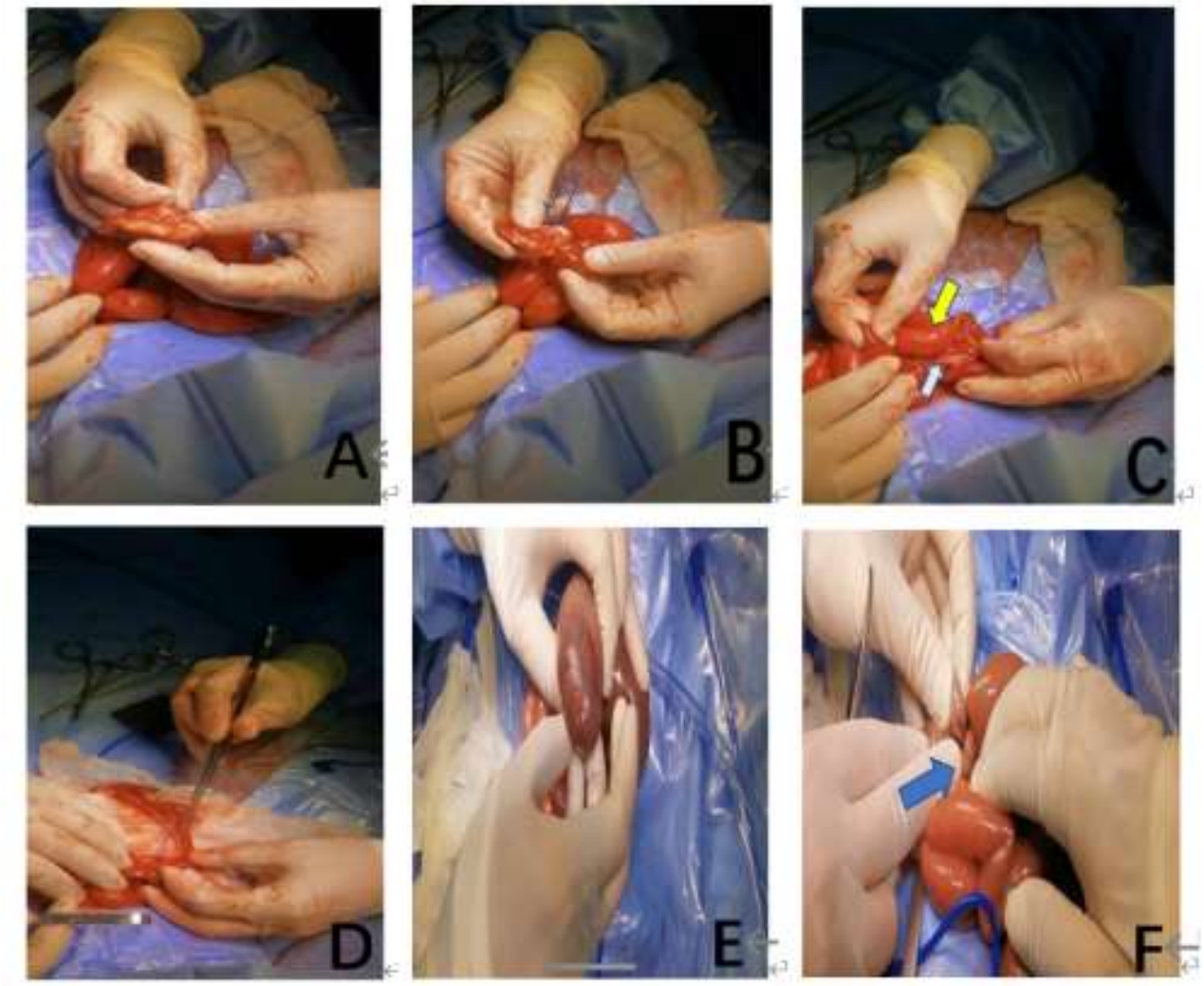

Fig. (3). (**A**) The twisted necrotic intestinal loop of the blind end in case 1. (**B**) The locking disconnection point in case 1. The yellow arrow in Fig. (**3**). Fig. (**3C**) shows the proximal dilated intestine in case 1, while the white arrow shows the distal collapsed intestine. The "apple peel" like lock is shown in Fig. (**3D**). Fig. (**3E**): Dilated intestinal blind end in case (2). Fig. (**3F**): Distal atresia of the intestinal canal, with blind end visible in the intestinal canal (case 2).

2.2. Case 2

A 40-year-old pregnant woman (G4P1) underwent an ultrasound examination at an external hospital at 26 weeks of pregnancy. The ultrasound indicated the fetal intestinal canal to be approximately 1.05 cm wide. Subsequently, amniocentesis was performed, but no abnormalities were found. At 38 weeks and 2 days, the pregnant woman came to our hospital for examination for the first time. The ultrasound revealed a significant dilation of the fetal intestinal tract (approximately 3.0 cm in width). The fetus' right lower abdominal intestinal wall exhibited localized thickening, presenting a "bilateral" pattern with enhanced echogenicity (Fig. **1F - I**).

Additionally, there was excess amniotic fluid. The pregnant woman underwent a cesarean section 2 days after the examination. The newborn had Apgar scores of 10 at 1 and 5 minutes; however, the newborn experienced frequent nausea and vomiting after birth. These symptoms gradually led to abdominal distension. Even after 21 hours post-birth, the newborn had not defecated. Abdominal ultrasound showed widespread dilation of the small intestinal canal, with the ascending and descending colon remaining unfilled. The newborn underwent resection and anastomosis for intestinal atresia on the second day after the delivery. During the operation, ileal atresia was discovered 100 cm from the Qurji ligament. The blind intestinal canal was significantly expanded, reaching a diameter of approximately 5 cm. A V-shaped defect was observed in the local mesentery. The distal blind end of the atresia was approximately 25 cm away from the ileocecal part, and the intestinal canal shrank to a diameter of approximately 0.8 cm. A section of the small intestine was embedded in a broken intestinal canal, approximately 4 cm in length. This confirmed the diagnosis of ileal atresia (type IIIa) (Fig. **3E - F**). In the follow-up period, the newborn autonomously defecated one day after the surgery.

3. RESULTS AND DISCUSSION

Ileal atresia is a type of intestinal atresia, a rare congenital malformation with an incidence rate of 1/10000~1/7000 [1]. Any interruption of local blood circulation in the intestinal tract caused by any reason can lead to small intestinal ischemia, necrosis, and atresia. Obstruction of the intestinal cavity is also a cause of intestinal atresia. There are few accompanying deformities (7%), generally limited to intestinal-related deformities (such as omphalocele, meconium peritonitis, and volvulus), which can occur simultaneously with jejunal obstruction [2].

The Grosfeld pathological classification of intestinal atresia includes the following types: type I: membranous atresia, where a diaphragm within the intestinal tract separates the intestinal cavity and forms atresia while maintaining continuity in appearance; type II: the two blind ends are connected by fibrous cords, and the mesentery maintains continuity; type IIIa: blind end closure, complete separation of the two closed blind ends, and a V-shaped defect in the mesentery; type IIIb: "apple skin-like" atresia, with completely separated blind ends of the two atresias, and a distal intestinal canal resembling "apple skin", with more missing mesentery; type IV, multiple intestinal atresias, like "sausage string" [3]. The first case of fetal atresia reviewed in this article presented a typical "apple skin"-like appearance in the distal intestine; therefore, it belonged to type IIIb, mostly caused by intestinal malrotation. Case 2 showed a complete separation of the two closed blind ends of the fetus, with a V-shaped defect in the mesentery, belonging to type IIIa caused by intussusception.

Direct signs of ileal atresia in fetal ultrasound include 1. intestinal dilation: when the inner diameter of the small intestine is greater than 7 mm, intestinal obstruction may occur, and the inner diameter of the dilated intestine gradually increases with increasing gestational age; 2. a dilated intestinal tract located in the middle abdomen of the fetus and appearing as multiple anechoic areas without echogenic colon bands; 3. "swirl sign": When intestinal malrotation is accompanied by a midgut volvulus, the dilated intestines may spiral around the superior mesenteric artery [4]. Indirect signs include 1. common signs of calcification in the fetal abdominal cavity; 2. the possible presence of fetal ascites; 3. excessive amniotic fluid; 4. formation of pseudocysts: complex cystic masses in the abdominal cavity with thick pseudocyst walls and high echogenicity, multiple septations within the cyst, poor sound transmission, and multiple inflammatory substances and calcifications. These indirect signs are mostly manifestations of meconium peritonitis, which often accompanies ileal atresia and intestinal perforation.

Meconium Peritonitis (MP) is often a sterile chemical inflammation caused by intestinal perforation secondary to intestinal torsion and atresia, resulting in meconium flowing into the abdominal cavity. Its different ultrasound manifestations may appear based on changes in the course of the primary lesion. MP can either worsen or gradually disappear due to adhesion and encapsulation [5]. When case 1 first visited our hospital for treatment, the ultrasonographic manifestations were small intestinal dilation and meconium peritonitis. The ascites disappeared, and calcification was not obvious in the subsequent two examinations. After surgery, a small tear was confirmed in the torsion of the ileal atresia. Later, due to severe fibrous adhesions, the perforation was closed, and calcification could only be observed on the left abdominal wall during surgery. This was consistent with the ultrasound examination results. In case 2, it was confirmed during surgery that intestinal atresia was caused by intussusception, and there were no secondary MP manifestations.

Accurately locating ileal dilation with or without MP using prenatal ultrasonography is difficult. During the second trimester of pregnancy, intestinal and colon dilatation can be distinguished by the location of the dilated bowel in the middle or around the abdominal cavity, the degree of dilatation, and the presence of *Taenia coli* [6 - 8]. In case 1, the dilated bowel was limited to the middle of the abdomen, and there was no Taenia coli. Small intestine expansion is considered during late pregnancy, and the degree of intestinal dilation continues to worsen, with dilated intestines visible in the middle and surrounding areas of the abdominal cavity. Based on the observation of these two cases, the dilated bowel is distributed around the abdominal cavity, and the torsion course is not easy to distinguish from the colon, making it more difficult to locate

the location of bowel obstruction or atresia in late pregnancy. If MP and intra-abdominal adhesions are combined, it becomes even more difficult for prenatal ultrasonography to accurately locate the obstruction. Therefore, it is particularly important to determine the lesions' location based on the intestine's local dilation during mid-pregnancy. In addition, ultrasound physicians need to pay close attention to the details and appearance around the two ends of the dilated intestine along its course [9, 10]. In case 2, we found a thickened “bilateral” intestinal wall with enhanced echogenicity at the distal end of the dilated intestine. This location was in the abdominal cavity's middle and lower part, suggesting ileal atresia. Whether the “bilateral” ultrasound manifestation of echo enhancement can be used as a characteristic manifestation of prenatal intestinal collapse and atresia requires further observation and confirmation in more cases.

When should pregnancy termination be considered if prenatal ultrasound detects intestinal dilation? Ileal atresia can often be detected in late or early pregnancy. The authors believe that if the fetus does not have MP that worsens with time and does not indirectly affect the development of other organs and fetal blood circulation due to increased abdominal pressure, the pregnancy can be monitored every 2–4 weeks until full-term delivery. In case 1, after 36 weeks of pregnancy and examination, the fetal abdominal cavity pressure significantly increased, leading to an elevation of the diaphragm, compression of the chest, increased cardiothoracic ratio, increased right heart size, tricuspid valve regurgitation, decreased PI value of the middle cerebral artery, and hypoxia. Therefore, the pregnancy was terminated. However, in case 2, the fetus did not exhibit the above symptoms except for intestinal dilation; therefore, regular follow-up was required until full-term delivery.

Ileal atresia requires postpartum surgical treatment, and except for cases with multiple atresias or widespread “apple skin” atresia, the prognosis is generally good. Additionally, the postoperative prognosis depends on the length and condition of the affected intestine, and short bowel syndrome is the main cause of long-term complications. Some domestic scholars grade meconium peritonitis using prenatal ultrasound (0–3 levels) based on one or more abnormal ultrasound images, such as intra-abdominal calcification, ascites, pseudocysts, intestinal dilatation, and polyhydramnios. This grading can predict newborns' clinical treatment and outcome after birth [11 - 13].

CONCLUSION

Accurately locating and classifying ileal atresia using prenatal ultrasound is challenging; however, it plays an effective role in disease progression, gestational assessment, and prognosis. Accurately identifying intestinal diseases and/or the location of lesion sites through direct and indirect ultrasound findings in the fetal abdominal cavity is an important research direction for prenatal ultrasound.

LIST OF ABBREVIATIONS

PI	=	Pulsatility index
PSV	=	Peak systolic velocity
MP	=	Meconium peritonitis

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Not applicable.

HUMAN AND ANIMAL RIGHTS

No animals/humans were used for studies that are the basis of this study.

CONSENT FOR PUBLICATION

A written informed consent was obtained from the patient for the publication of this report and any accompanying images.

STANDARDS OF REPORTING

CARE guidelines were followed.

AVAILABILITY OF DATA AND MATERIALS

Not applicable.

FUNDING

None.

CONFLICT OF INTEREST

The authors declare no conflict of interest, financial or otherwise.

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Declared none.

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