Ref: Ro Med J. 2023;70(2)
Doi: 10.37897/RMJ.2023.2.4

Diagnosis of a rare case of neonatal intestinal duplication cyst isolated from the gastrointestinal tract - Case report

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ABSTRACT

Duplication cysts are congenital anomalies that typically have some connection to the gastrointestinal (GI) tract, but in rare cases the cysts can be completed isolated from the GI tract. We present the case of an appropriate for gestational age male infant born at 40 weeks gestational age with neonatal intestinal duplication cyst isolated from the GI tract. Postnatal diagnostic methods included ultrasound (US) and magnetic resonance imaging (MRI) based on abnormal antenatal findings. The positive diagnosis was based on the imaging results. Duplication cysts may remain asymptomatic and that is the reason why antenatal and postnatal US screening plays an important key role in the diagnosis. Series of imaging may confirm the diagnosis, prevent future complication and help the clinician to determine the optimal operative moment.

Keywords: intestinal duplication, cyst, infant, gastrointestinal tract

INTRODUCTION

Duplication cysts are rare congenital anomalies that can occur throughout the gastrointestinal (GI) tract; usually, they are detected in infancy and childhood [1,2]. Intestinal duplication cysts are believed to occur between the 4th and 8th weeks of embryonic development and their aetiology is still unknown [3].

Duplication cysts typically have some connection to the GI tract as well as the local blood supply to that region; however, in rare cases the cysts can be completed isolated from the GI tract [2]. Pathologic events can be preceded by torsion or some vascular accident at the proximal end of the diverticulum, which results in detachment of the intestinal wall and ultimately in a completely isolated duplication cyst [4]. Completed isolated intestinal duplication cysts have no attachments to the intestinal wall and usually have their own vascular supply [2].

The clinical presentation is vague, variable and non-specific; it greatly depends on the size and site of the duplication [1]. Intestinal duplication cysts can be detected prenatally on screening ultrasound (US) and, manifests early postnatally as small bowel obstruction, perforation, haemorrhage, volvulus or intussusception [2,5]. Presenting symptoms among older children include abdominal pain, emesis, acute pancreatitis and intestinal obstruction [1]. Other malformations may be associated with it as spinal defects, intestinal malrotation, intestinal atresia, and other abnormalities of the urinary tract [5].

Antenatal US may demonstrate an intra-abdominal mass during 2nd or 3rd trimester of pregnancy [5]. Prenatal detection of the intestinal duplication cyst allows close neonatal surveillance [6]. Postnatal US and contrast computed tomography (CT) of the abdomen or magnetic resonance imaging (MRI) may

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Article History:
Received: 7 May 2023
Accepted: 16 May 2023

differentiate it from other intra-abdominal cystic lesions [5]. Postnatal US is often successful in determining the aetiology of cystic lesions and only occasionally is supplemented by MRI [7].

In some cases, CT or MRI can provide some additional information and demonstrate the relationship between the cyst and the adjacent bowel [8]. Intestinal duplications isolated from the GI tract are even more difficult to diagnose – both prenatally and postnatally – than classic duplications, most likely as a result of how difficult it is to establish a clear relationship with the neighbouring structures [9].

There is some debate regarding the preferred management in asymptomatic duplications because surgery always carries a certain amount of risk and there has been some success with observation with serial USs [8]. Surgical treatment could be deferred in some asymptomatic patients, in order to avoid general anaesthesia and surgery, thus limiting the potential neurocognitive damage induced by general anaesthesia at an early stage of neural development [9].

The aim of this article is to emphasize the fact that the imaging methods used postnatally are useful in order to establish whether the neonatal intestinal duplication cyst communicates with the GI tract or not. Written informed consent was obtained from the patient's mother, the minor's legal guardian, for the publication of this case report.

CASE REPORT

Presenting concerns

We present the case of an appropriate for gestational age male infant born at 40 weeks gestational age. His 27-year-old mother was a G1P1 with intrauterine diagnosis of pyelocaliceal dilatation at the level of the right kidney of the fetus established in the 2nd trimester. The birth was achieved by caesarean section due to the lack of progression of the labor. The amniotic membranes ruptured 2 hours before birth and the amniotic fluid was clear. The infant's birth weight was 3740 g (60th centile), length 56cm (94th centile) and cranial perimeter 33 cm (2nd centile). His Apgar scores were 10 and 10 at 1 and 5 minutes. The early postpartum adaptation was good.

Clinical findings

On examination the infant was alert and vigorous and appeared well. The temperature was 36.6 C, the heart rate 130 beats per minute, the blood pressure 77/44 (55) mm Hg, the respiratory rate 45 breaths per minute and the oxygen saturation 98% while he was breathing ambient air. The lungs were clear on auscultation. The abdomen was distended by volume, but soft without tenderness, or palpable masses. No hepatomegaly was detected, and the liver had a smooth surface and sharp edge. The male genitalia appeared normal. The anterior fontanelle was flat and soft. There were no dysmorphic features and neurologic examination revealed normal primitives reflexes.

Diagnostic focus and assessment

Antenatal US done in the 2nd and then in the 3rd trimester had revealed right fetal kidney with moderate dilatation of the calyx, of the pelvis and apparently with a 5 cm portion of the ureter in tension; bladder with normal dimensions; left kidney with normal echostructure.

Soon after birth a sepsis evaluation was performed and a summary of urine and uroculture were sent and they were negative.

Abdominal US showed a well-defined, oval, cystic, anechoic mass lesion of approximately size of 4×4 cm at the level of the lower pole of the right kidney; the upper calyx of the right kidney dilated by 1.2/1.6 cm; the left kidney of normal size with appropriate echostructure and echogenicity; liver with appropriate echostructure and echogenicity; transonic gall bladder; spleen of normal size with corresponding echostructure (figures 1 and 2).

The MRI was made at 2 months of life and it pointed out: the right kidney was located in the renal lodge, but it was rotated with the hilum directed anteriorly and showed slight pyelocaliceal dilatation; the renal parenchyma was preserved with good corticomedullary differentiation; anterior to the right kidney, in contact with it and the proximal portion of the ureter appeared a cystic structure of large dimensions (68/40/58 mm), with its own, thin wall; the formation had contact posteriorly with perirenal fat and the kidney, without invading it, laterally with the abdominal wall with which it had a cleavage plane and anteriorly with the intestinal loops that it moved without compressing them; no communication with the duodenum or other intestinal structures was evident; the content of this cyst was liquid and the cyst was homogeneous; left kidney with normal size and morphology, normally positioned; intestinal loops with peristalsis present; liver, gallbladder, pancreas, spleen, adrenal glands without obvious changes in the native MRI examination; no collections at the abdominal-pelvic level; bladder with homogeneous content, wall of normal appearance (figures 3 and 4).

The conclusion of MRI was: large abdominal cystic formation with its own wall that imprints and displaces the right kidney with malrotation without invading it; right pelvicalyceal dilatation, more likely secondary to compression. The imaging aspect suggested a cyst originating from the primitive intestine - intestinal duplication - without obvious communication with the intestinal structures. The relationship with the adjacent organs suggested a retroperitoneal extension.

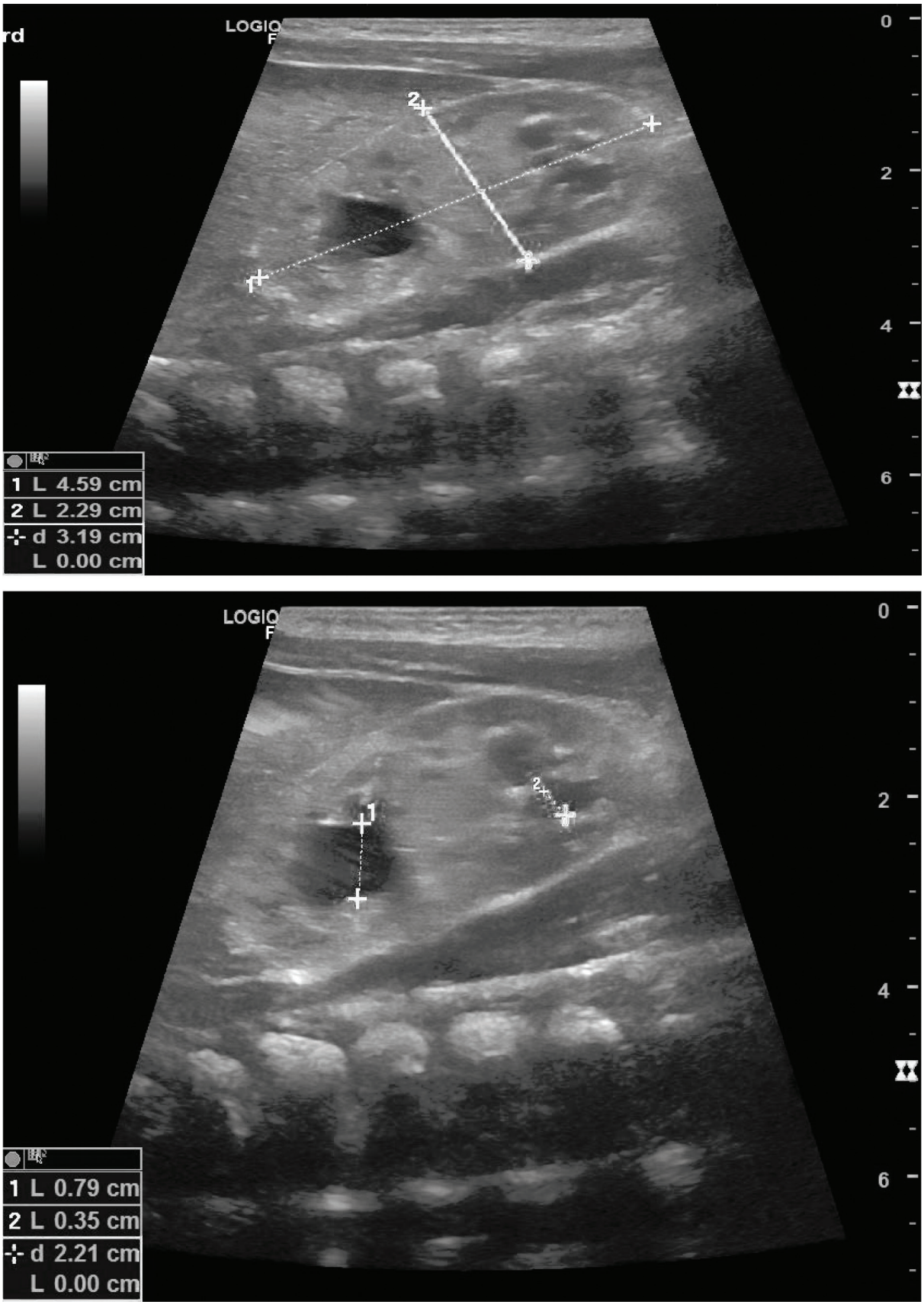

FIGURE 1. Echographic images of the right kidney

Postnatal diagnostic methods in this case included US imaging and MRI based on abnormal antenatal findings. US imaging was used for prenatal diagnosis and it also allowed neonatal surveillance. The positive diagnosis was based on the imaging results.

Therapeutic focus and assessment

Considering the US image, a bladder catheter was mounted and antibiotic prophylaxis with third generation cephalosporin was carried out.

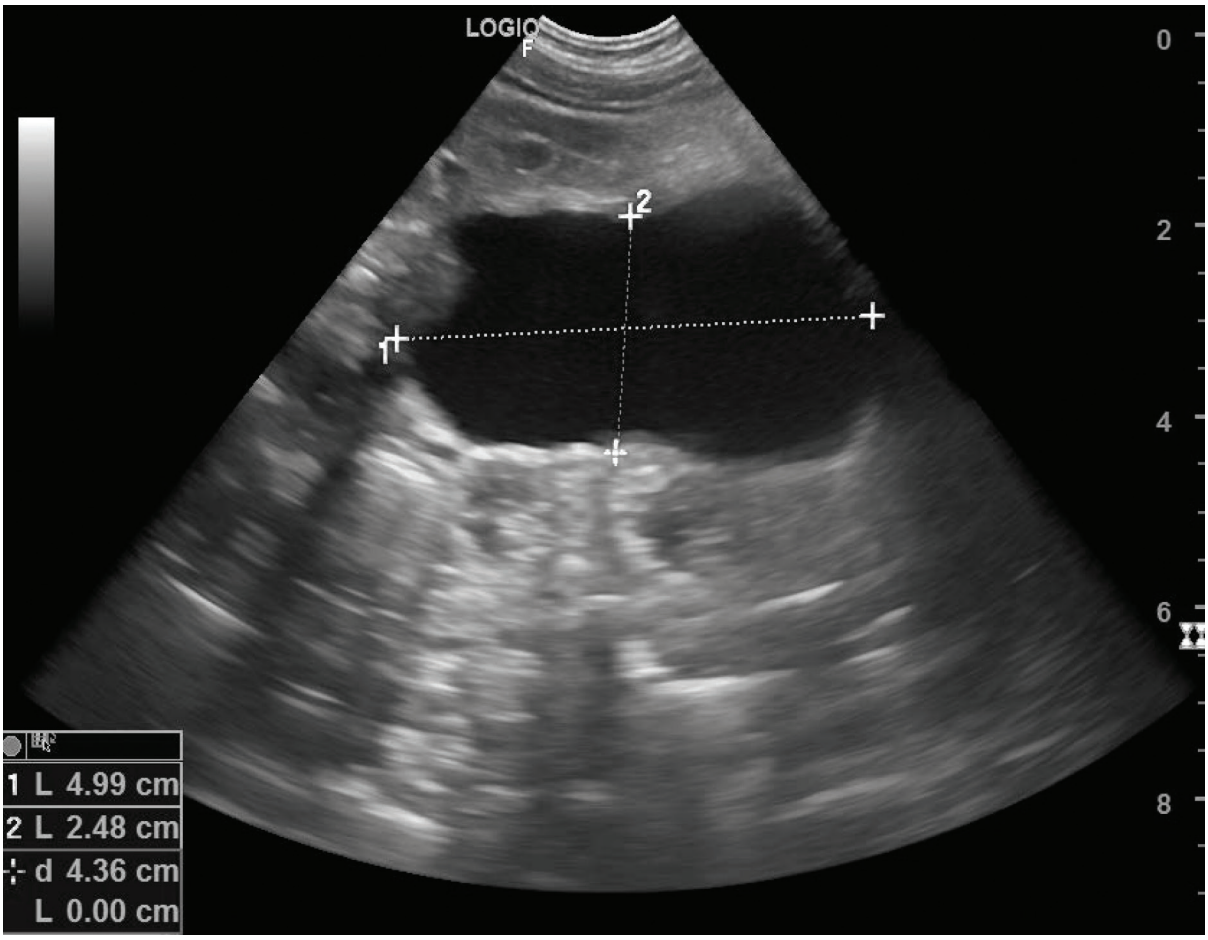

FIGURE 2. Echographic image of the cyst

The surgical treatment of this neonatal intestinal duplication cyst isolated from the intestinal tract was postponed because the clinical condition of the infant remained stable.

Follow-up and monitoring

The infant was discharged from the maternity unit at the ninth day of life in order to be admitted to a pediatric nephrology service. He remained stable, with an upward growth curve and with blood and urine tests with normal values. After the diagnosis was clear, he was discharged at home without bladder catheter and antibiotic prophylaxis and with the recommendation of a close monitoring of the cyst by US. Long term follow up is required in this case in order to establish when will be the ideal time for surgical treatment of the cyst.

DISCUSSIONS

Intestinal duplication isolated from the GI tract represents an extremely rare malformation; this type of duplication is not in close contact with any segment of the intestinal tract [9,10].

Intestinal duplications cysts that are isolated from the GI tract have been characterized in only a few case reports. Here we report a case of neonatal intestinal duplication cyst isolated from the GI tract. Like in our case, Seydafkan et al. described the case of a female with enteric duplication cysts that did not communicate with the GI tract [11].

Our case of neonatal intestinal duplication isolated from the GI tract was initially interpreted as a right pyelocaliceal dilatation, but in fact the cyst was compressing the right kidney and for this reason the kidney appeared with dilatation. Like in our case, Hakda et al reported the case of a newborn with duodenal duplication cyst presented as an abdominal mass that compressed the right kidney [12].

With the increasing use of prenatal US scan, many cases are being identified in utero. The mode of presentation in our case was unusual due to the mass effect performed on the right kidney. Similar to our case, Deng et al presented a case of an isolated retroperitoneal enteric duplication cyst, but differently to our case, it was associated with an accessory pancreatic lobe [13].

Like in our case, Luque-González et al revealed the case of an asymptomatic male newborn diagnosed by abdominal US and IRM in the early neonatal period [14]. Contrary to our case, Sampaio et al. described the case of a female newborn with two abdominal cysts found by abdominal US and because of

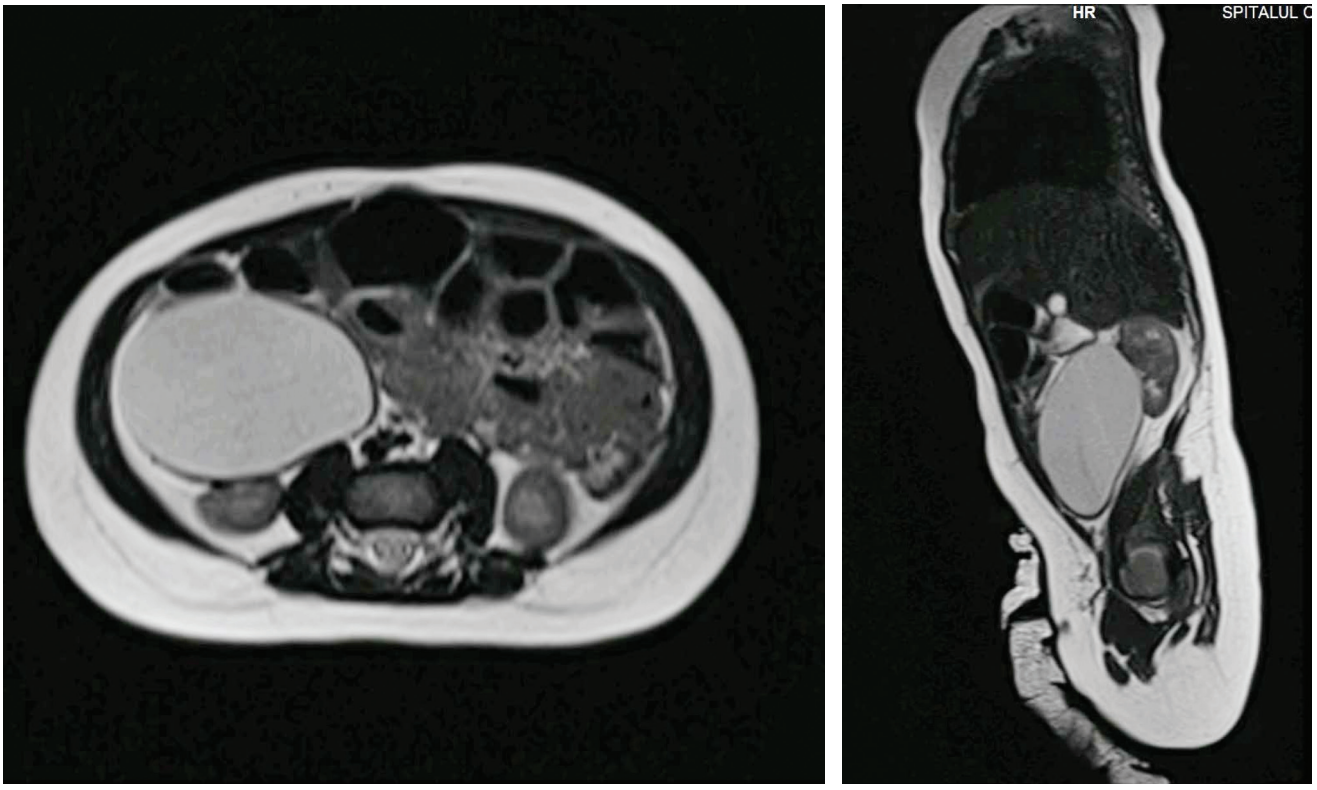

FIGURE 3. Native MRI images

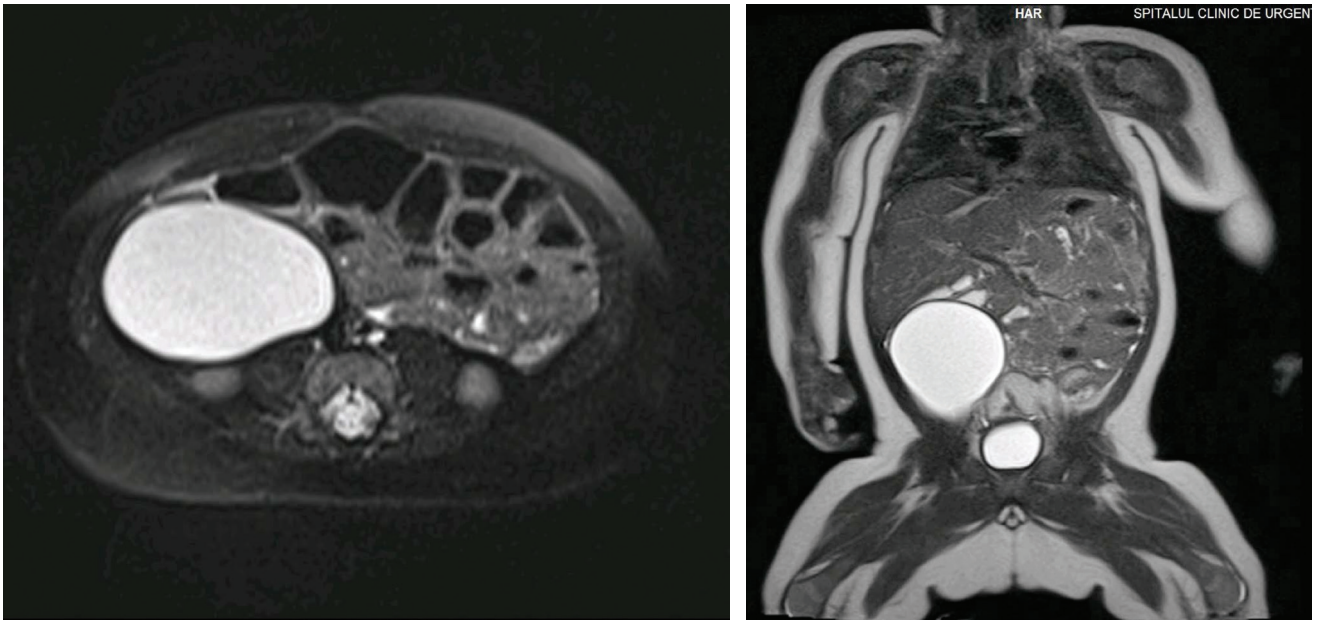

FIGURE 4. MRI contrast images

the enlargement of the abdominal cysts and the alterations on physical examination, the team prescribed surgery in this case [15].

There is no clear consensus on how to treat intestinal duplication cysts. In contrast to our case, the male neonate's case described by Dilawar et al presented with bilious vomiting and he need surgical treatment to relieve the symptoms [1]. Beura et al. stated that treatment of asymptomatic case is controversial, but early intervention prevents complications [5].

Despite the serial imaging investigations that were carried out in our infant's case, until the operative moment it cannot be said with certainty if this intestinal duplication cyst does not actually mask a mesenteric cyst.

CONCLUSIONS

Duplication cysts may remain asymptomatic and that is the reason why antenatal and postnatal US screening plays an important key role in the diagno-

sis. While detecting a cystic mass can be relatively simple, identifying its origin is less and in some cases CT or MRI completes the diagnosis. Lack of specific signs and symptoms makes diagnosis a challenge. Series of imaging may confirm the diagnosis, prevent future complication and help the clinician to determine the optimal operative moment. The choice of the best therapeutic technique depends on the size, location of the cyst and its relationship with the nearby anatomical structures. New advances still have to be made to decide the best moment for surgical treatment.

Conflict of interest: none declared
Financial support: none declared

REFERENCES

1. Dilawar B, Hamid LR, Pirzada AN, Arshad M. Duodenal duplication cyst presenting as bilious vomiting in a neonate. *J Pediatr Surg Case Rep.* 2020; 61:101617. doi:10.1016/j.epsc.2020.101617.
2. Weitman E, Al Diffalha S, Centeno B, Hodul P. An isolated intestinal duplication cyst masquerading as a mucinous cystic neoplasm of the pancreas A case report and review of the literature. *Int J Surg Case Rep.* 2017; 39:208–211. doi:10.1016/j.ijscr.2017.08.030.
3. Nebot CS, Salvador RL, Palacios EC, Aliaga SP, Pradas VI. Enteric duplication cysts in children: varied presentations, varied imaging findings. *Insights Imaging*. 2018; 9(6):1097–1106. doi:10.1007/s13244-018-0660-z.
4. Kim HY, Kim SH, Park KW, Jung SE. Noncommunicating isolated enteric duplication cyst in the abdomen in children: report of one case and review of the literature. *An Pediatr Surg.* 2013. 2013; 9(3):124–6. doi:10.1097/01.XPS.0000430647.23203.52.
5. Beura S, Mishra BN, Joshi RK, Pahi PP. Enteric duplication cyst in a neonate: A case report. *Int J Surg Sci.* 2020; 4(2): 303-5. doi: 10.33545/surgery.2020.v4.i2e.434.
6. Sheik S, Mathew M, Abdellatif M, Qureshi A, Mandhan P. Multiple Enteric Duplication Cysts in a Twin Fetus Diagnosis and management. *Sultan Qaboos Univ Med J.* 2013; 13(4):593-596. doi: 10.12816/0003323.
7. Muthee BW, Bray HJ. Approach to the postnatal sonographic evaluation of prenatally detected abdominopelvic cysts. *Ultrasonography.* 2022; 41(1):53-73. doi: 10.14366/usg.21070.
8. Losefsky Q. A comprehensive review of enteric duplication cysts, their pathophysiology, presentation, and treatment. *J Clin Exp Gastroenterol.* 2022;1(1):1-4. doi: 10.46439/gastro.1.001.
9. Moreillo CG, Mallol JM, Carretero SF. Intestinal duplication isolated from the digestive tract: an entity to be considered. *Cir Pediatr.* 2022; 35:36-41. doi: 10.54847/cp.2022.01.17.
10. Lee JU, Kim JO, Kim SJ, Sul HJ. Completely Isolated Enteric Duplication Cyst Presenting as an Inguinal Hernia. *Korean J Pathol.* 2010; 44:204-6. doi: 10.4132/KoreanJPathol.2010.44.2.204.
11. Seydafkan S, Shibata D, Sanchez J, Tran ND, Leon M, Coppola D. Presacral Noncommunicating Enteric Duplication Cyst. *Cancer Control.* 2016; 23(2):170-4. doi: 10.1177/107327481602300213.
12. Hakda RY, Makwana DV, Shukla R, Parikh U, Chandna SB. Isolated Duodenal Duplication Cyst in a Neonate. *Afr J Paediatr Surg.* 2022; 19(4):257-60. doi: 10.4103/ajps.ajps_176_21.
13. Deng Y, Hu Z, Liao J, Hao W, Hu G. Isolated retroperitoneal enteric duplication cyst associated with an accessory pancreatic lobe. *Int J Clin Exp Pathol.* 2019; 12(8):3089-95. PMID:31934150.
14. Luque-González P, Lozano-Vidal M, Gómez-Ortiz C, Gutiérrez IC, Limones-Jiménez S. Intestinal duplication and prenatal differential diagnosis of intra-abdominal cysts. *Ginecol Obstet Mex*. 2023; 91(3):218-25. doi: 10.24245/gom.v91i3.7868.
15. Sampaio RS, Colombiano CK, Fiorio IO, Do Carmo MD, Pinheiro PL, Bastos JMC, et al. A newborn with an enteric duplication cyst. *Residência Pediátrica.* 2022; doi: 10.25060/residpediatr-2022.v12n2-319.