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Duodenal atresia with apple peel jejunoileal syndrome

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ABSTRACT

Embryologically, duodenal atresia results from inadequate recanalization and proliferation of the solid cord in the 8–10th week of gestation, while apple-peel atresia is a consequence of a vascular accident. The presence of both malformations is rather rare. We report a case with both associations; to our knowledge, this is the twelfth such case reported in the English literature.

1. Introduction

Duodenal atresia is a common cause of neonatal small bowel obstruction, and in approximately 50% of cases, it is associated with other malformations. Duodenal atresia is believed to occur due to failure of recanalization, whereas jejunal atresia is due to an intrauterine vascular accident [1]. We report a rare case of duodenal atresia with apple-peel configuration of the remaining small intestine in a preterm child, which indicates that in rare circumstances, vascular accidents may be the underlying cause of duodenal atresia [2].

2. Patient presentation

A 1.7 kg baby girl was born by spontaneous vaginal delivery at 34.4 weeks of gestation to a 23-year-old mother, G1P0A0, whose pregnancy was complicated by two recurrent urinary infections. Prenatal ultrasonography diagnosed polyhydramnios with no double bubble, and fetal echocardiogram showed 4.5-mm interventricular communication, dual right ventricular outflow tract, mitral valve hypoplasia and a persistent left superior vena cava with no signs of Down syndrome. At birth, the baby was admitted to the neonatal intensive care unit in stable condition and without respiratory distress. An abdominal X-ray revealed a double bubble sign with no air distally (Fig. 1), and echocardiogram confirmed interventricular communication and a dual right ventricular outflow tract and showed patent ductus arteriosus.

The baby was taken up for surgery on her sixth day of life. On laparotomy, she was found to have atresia in the second and third parts of the duodenum, and the proximal part was dilated and elongated (Fig. 2). The proximal jejunum had an apple-peel configuration (Fig. 3A). This part of the bowel was spiraling around a retrograde vascular arcade, and the rest of the small and large bowel was normal. As only 20 cm of the jejunum had the apple peel configuration with precarious blood supply (Fig. 3A, arrow), we decided to resect this part of the bowel. An end-to-end duodenojejunal anastomosis was performed (Fig. 3B).

Postoperatively, the patient was kept on ventilator support and needed vasoactive drugs only in the first 48 hours after surgery. Thirteen days after surgery, the patient developed pulmonary focus infection, and treatment with cefepime was carried out for 14 days. It was necessary to complete the antibiotic therapy to undergo cardiac surgery: on her forty-third day of life, she was submitted to pulmonary artery banding and closing of the ductus arterius. The baby was started on gradual enteral feeds and was weaned from parenteral nutrition in the course of her sixth postoperative cardiac surgery. She was discharged on day of life 71. The last follow-up was performed at 3 months and 21 days of life.

3. Discussion

Small intestinal atresia is a congenital anomaly characterized by abnormal closure, discontinuity or narrowing of the duodenum, jejunum or ileum [3]. Congenital intestinal atresias and stenoses have an incidence of 0.7–0.8 per 10,000 live births [4], and duodenal atresia occurs in 1 in 5000 to 10,000 live births [5].

The etiology of duodenal atresia and stenosis is probably related to a failure of recanalization of the duodenal lumen from its solid cord stage, as proposed by Tandler in 1902 [6]. During the 4th to 8th weeks of gestation, obliteration of the lumen occurs, and this period is referred to

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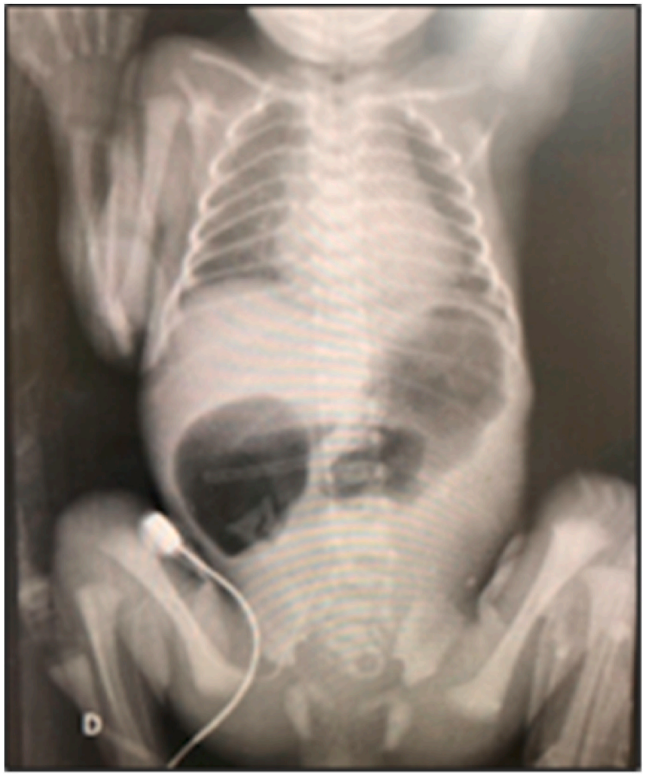

Fig. 1. The double bubble sign.

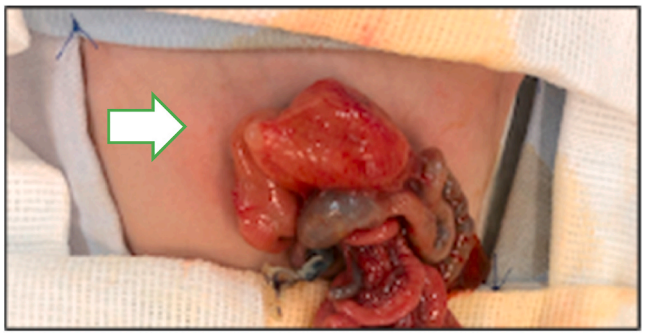

Fig. 2. Complete atresia of the second and third parts of the duodenum with dilation of the proximal part.

as the solid cord stage. Vacuolation of the solid cord begins during the 8th to 10th weeks of gestation, resulting in recanalization and development of a lumen. Failure of vacuolation may result in a variety of intrinsic abnormalities, including atresia (blind end), stenosis (narrowing), or formation of an atretic or stenotic mucosal web [7].

Apple-peel atresia, or Type-IIIb intestinal atresia, is the rarest variant of jejunoileal atresia, with a prevalence of less than 5% among all intestinal atresia [8]. It was first described in 1961 by Santulli and Blanc. Apple-peel atresia or "Christmas-tree deformity" consists of high jejunal atresia with discontinuity of the small bowel and a wide gap in the mesentery. The distal segment of the ileum is shortened and assumes a helical configuration around a retrograde perfusing vessel, which compensates for the partially absent superior mesenteric artery. The most accepted theory is that apple-peel deformity results from an intrauterine vascular accident in late gestation [9,10].

The present patient had atresia in the second and third parts of the duodenum and proximal jejunum with a typical apple-peel configuration of the small intestine. There was no family history of atresia; however, she had a cardiac malformation. Duodenal atresia, apple-peel configuration and cardiac malformation are even rarer. Kirtane, 2019 [11] is the only author who has reported this same association.

In our patient, as well as in most reported cases, the correction of atresia was performed by duodenojejunostomy. The outcomes of isolated duodenal atresia and isolated jejunal atresia are generally excellent, while the outcome of apple-peel atresia is poor [1]. The association of both malformations may reduce the survival rate; however, there is no study to date that statistically proves this theory. The outcomes of the reported similar eleven cases are as follows: four mortalities, two of whom were operated on at 8 days of life, one was considered inoperable because of the extent of multiple intestinal atresias that made the attempt of multiple primary anastomoses difficult, and the fourth patient succumbed on the third postoperative day due to cardiorespiratory failure. Our patient did well postoperatively and started on feeds gradually.

Type IIIb jejunoileal atresia is considered rare, and its association with duodenal atresia is even more uncommon; thus, it appears that both malformations are caused by a common intrauterine event. The mortality rate found in this literature review was 36%. To date, we have found only 11 reported patients, with 7 survivors. Our patient is the twelfth reported case and the eighth survivor (see Table 1).

Author contribution

Taís Gavira Wong – written, edited.
Lourenço Sbragia Neto – edited.
Wellen Canesin – surgeon during case.
Maurício André Pereira da Silva – surgeon during case.

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I declare that there is no funding source for this case report.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

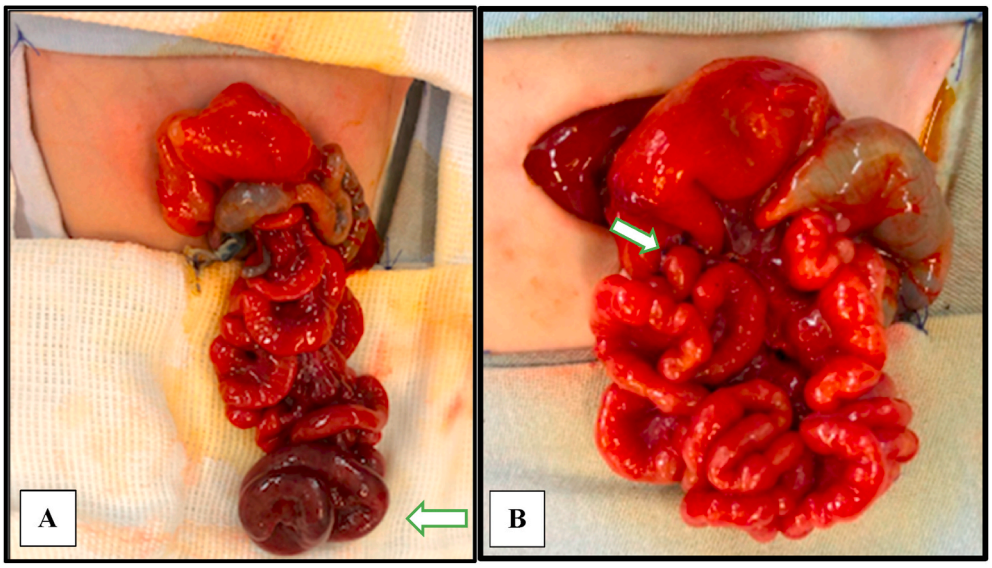

Fig. 3. A. Apple-peel configuration. Arrow: precarious blood supply and 3B end duodenojejunal anastomosis.

Table 1
Cases of duodenal atresia with apple-peel configuration published up to this point.

Author	Sex	Age of gestation	Age and weight at OR	Anomalies	Procedure	Outcome
Weber and Freeman (1999) [2]	Girl	36	3 days; 2.1 kg	- Loss of third and fourth parts of the duodenum and the proximal jejunum - Apple peel atresia - Absent superior mesenteric artery - Duodenal stenosis - Common hepatic duct	Duodenojejunostomy	Survived
Arbell et al. (2006) [12]	Boy	32	2 days; 1.5 kg	- Duodenal Atresia - Apple peel atresia - Choledochal cyst	- Duodenojejunostomy - Appenedectomy - Excised choledochal cyst and cholecystoduodenostomy	Survived
Tatekawa et al. (2007) [13]	Girl	36	2.1 kg	- Duodenal membranous atresia - Apple peel atresia - Multiple intestinal atresia	Duodenojejunostomy	Survived
Ahmad et al. (2009) [14]	Boy	34	8 days; 1.2 kg	- Apple peel atresia - Absence of superior mesenteric artery	Duodenojejunostomy	Died
Patil et al. (2011) [15]	Girl	33	8 days; 1.6 kg	- Duodenal atresia - Malrotation - Apple peel atresia	- Release of Ladd's bands - Appenedectomy - Duodenojejunostomy	Died
Alnosair (2014) [16]	Girl	31	1.4 kg	- Duodenal atresia just distal to the insertion of the biliary and pancreatic ducts	- Duodenojejunostomy - Appenedectomy	Survived
Altokhais (2014) [1]	Boy	33	2 days; 1.9 kg	- Duodenal atresia - Apple peel atresia - Malrotation	- Release of Ladd's bands - Duodenojejunostomy	Survived
Pathak and Narula (2014) [17]	?	33	2 days; 1.3 kg	- Duodenal atresia, the proximal 15 cm jejunum was apple peel	- Resection anastomosis done	Survived
Bem Hamida et al. (2016) [18]	Boy	34	3 kg	- Duodenal atresia - Apple peel atresia - Mesenteric defect - Agenesis of the superior mesenteric artery - Situs inversus abdominus	Considered inoperable because of the extent of multiple intestinal atresias that made the attempt of multiple primary anastomoses difficult	Died
Sasa et al. (2016) [19]	Boy	29	2 days; 1.24 kg	- Duodenal atresia - Apple peel atresia - Absent of superior mesenteric artery	End-to-end duodenoileal anastomosis	Died
Kirtane et al. (2019) [11]	Boy	36	2 days; 2.25 kg	- Apple peel atresia - Duodenal atresia - Malrotation - Patent ductus arteriosus - 3-mm atrial septal defect. With left-to-right shunt	- Duodenojejunostomy - Release of Ladd's bands - Jejunostomy	Survived

References

[1] Altokhais TI. Duodenal atresia with apple-peel jujenoilial deformity: case report and review of the literature. J Pediatr Surg Case Rep 2014;2(3):156–8. https://doi.org/10.1016/j.epsc.2014.03.001.

[2] Weber DM, Freeman NV. Duodenojejunal atresia with apple peel configuration of the ileum and absent superior mesenteric artery: observations on pathogenesis. *J Pediatr Surg* 1999;34:1427–9.

[3] Walker W. Pediatric gastrointestinal disease: pathophysiology, diagnosis, management. second ed. London: Mosby; 1996.

[4] Best KE, Tennant PW, Addor MC, et al. Epidemiology of small intestinal atresia in Europe: a register-based study. Arch Dis Child Fetal Neonatal 2012;97:F353–8.

[5] Sigmon DF, Eovaldi BJ, Cohen HL. Duodenal atresia and stenosis [Updated 2020 Jun 27]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 Jan. https://www.ncbi.nlm.nih.gov/books/NBK470548/.

[6] Tandler J. Zur entwicklunggeschicte des menschlichen duodenums. Morphol Jb 1902;29:187–216.

[7] Grosfeld JL, Rescorla FJ. Duodenal atresia and stenosis: reassessment of treatment and outcome based on antenatal diagnosis, pathologic variance, and long-term follow-up. World J Surg 1993;17(3):301–9. https://doi.org/10.1007/bf01658696.

[8] Zhu H, Gao R, Alganabi M, et al. Long-term surgical outcomes of apple-peel atresia. J Pediatr Surg 2019;54(12):2503–8. https://doi.org/10.1016/j.jpedsurg.2019.08.045.

[9] Louw JH, Barnard CN. Congenital intestinal atresia; observations on its origin. Lancet 1955;269(6899):1065–7. https://doi.org/10.1016/s0140-6736(55)92852-x.

[10] Santulli TV, Blanc WA. Congenital atresia of the intestine: pathogenesis and treatment. Ann Surg 1961;154:939e48.

[11] Kirtane JM, Bhange SA, Nabi F, Shah V. Duodenal atresia with familial apple peel syndrome: case study with review of literature. BMJ Case Rep 2019;12(8). https://doi.org/10.1136/bcr-2019-230160. *e230160*.

[12] Arbell D, Orkin B, Naveh Y, et al. Duodenojejunal atresia with absent dorsal mesentery, choledochal cyst, and malrotation in a premature newborn-a case report. J Pediatr Surg 2006;41:e11–13.

[13] Tatekawa Y, Kanehiro H, Nakajima Y. Duodenal atresia associated with "apple peel" small bowel without deletion of fibroblast growth factor-10 or fibroblast growth factor receptor 2IIIb: report of a case. Surg Today 2007;37:430–3.

[14] Ahmad A, Sarda D, Joshi P, et al. Duodenal atresia with 'apple-peel configuration' of the ileum and absent superior mesenteric artery: a rare presentation. Afr J Paediatr Surg 2009;6:120–1.

[15] Patil RT, Gupta R, Parelkar SV, et al. A rare case of duodenal atresia with apple-peel configuration of the small intestine and malrotation. Eur J Pediatr Surg 2011;21:340–2.

[16] Alnosair AA, Naga MI, Abdulla MR, et al. Congenital duodenal atresia with 'apple-peel configuration' of the small intestines and absent superior mesenteric artery: a case report and review of literature. J Pediatr Surg Case Rep 2014;2:215–8.

[17] Pathak M, Narula D. A case of duodenal atresia with apple peel appearance: challenging the current embryology. J Clin Neonatol 2014;3:112–4.

[18] Ben Hamida H, Hadj Salem R, Ben Ameur K, Rassas A, Chioukh FZ, Sakka R, et al. Duodenal atresia associated with apple peel atresia and situs inversus abdominus: a case report. J Neonatal Surg 2016;5:60.

[19] Saša RV, Ranko L, Snezana C, Lidija B, Djordje S. Duodenal atresia with apple-peel configuration of the ileum and absent superior mesenteric artery. BMC Pediatr 2016;16(1). https://doi.org/10.1186/s12887-016-0690-y.