Scientific Research Publishing

Open Journal of Pediatrics, 2025, 15(5), 734-740
https://www.scirp.org/journal/ojped
ISSN Online: 2160-8776
ISSN Print: 2160-8741

A Rare Case of Apple-Peel Configuration of the Small Intestine Associated with Intestinal Malrotation

Cheikh Tidiane Mbaye[1*], Christ Momo Tsague[1], Florent Tshibwid A. Zeng[2], Cheikh Diouf[1]

[1]Unit of Pediatric Surgery, Ziguinchor Regional Hospital Center, Ziguinchor, Senegal
[2]Department of Surgery, Faculty of Medicine, University of Lubumbashi, Lubumbashi, Democratic Republic of the Congo
Email: *mbayecheikhtig@hotmail.fr

How to cite this paper: Mbaye, C.T., Tsague, C.M., Zeng, F.T.A. and Diouf, C. (2025) A Rare Case of Apple-Peel Configuration of the Small Intestine Associated with Intestinal Malrotation. *Open Journal of Pediatrics*, **15**, 734-740.
https://doi.org/10.4236/ojped.2025.155070

Received: August 9, 2025
Accepted: September 5, 2025
Published: September 8, 2025

Abstract

The combination of jejunal atresia with an apple-peel configuration and intestinal malrotation is extremely rare. This malformative association poses significant surgical and prognostic challenges, particularly in low- and middle-income countries (LMICs), where prenatal diagnosis is rarely established and postoperative resources remain limited. We report the case of a newborn with this association, complicated by extensive small bowel necrosis. Detorsion of the volvulus was performed, followed by resection of the entire necrotic small intestine. This led to short bowel syndrome, the severity of which was exacerbated by the unavailability of parenteral nutrition. The postoperative course was complicated by severe malnutrition and electrolyte disturbances. This report aims to describe a rare case and to highlight the challenges faced in a resource-limited setting.

Keywords

Apple-Peel Configuration, Jejunal Atresia, Intestinal Malrotation

1. Introduction

Apple-peel syndrome refers to jejunal atresia in which the remaining jejunoileal parts are spirally placed around a single vascular branch, typically a branch of the ileocolic artery, due to the absence of the superior mesenteric artery. This configuration resembles the peeling of an apple [1]. Apple-peel small bowel is a variant of jejunoileal atresia (type IIIB) [1] [2]. Also known as "pigtail-like syndrome", it is extremely rare and reported to occur in less than 5% of intestinal atresia. Apple-peel atresia may occur in isolation but can also be associated with other malfor-

mations, such as intestinal malrotation [3]. Malrotation may lead to volvulus of the mid-gut around a narrow-based mesentery [4]. The simultaneous occurrence of these conditions in an individual is exceptionally rare. Through this case report, we aim to highlight this unusual condition.

2. Case Report

A 4-day-old male newborn, born at 35 weeks of pregnancy, has been admitted to neonatology since birth for a neonatal infection. He was born to a 24-year-old mother, gravida 4, para 4, with other children appearing to be in good health. Pregnancy was monitored with four prenatal visits and a normal second-trimester ultrasound. Delivery was spontaneous and vaginal, with a cephalic presentation and no need for perinatal resuscitation. Clear amniotic fluid, birth weight 1.7 kg (Z-score: −1.6). There was no passage of meconium. On the fourth day of life, after initiating nasogastric tube feeding, the newborn experienced episodes of bilious vomiting. On examination, the general condition was good, with normal neurological behavior and stable hemodynamics. Abdominal examination revealed a soft, nontender, and non-distended abdomen. On digital rectal examination, the anus was patent, and the rectum was empty. A nasogastric tube drained around 100 ml of bilious secretions. An abdominal X-ray revealed a double bubble sign with no air distally (**Figure 1**). The echocardiogram revealed no abnormalities. Duodenal atresia was diagnosed, and exploratory laparotomy was performed on the fifth day of life.

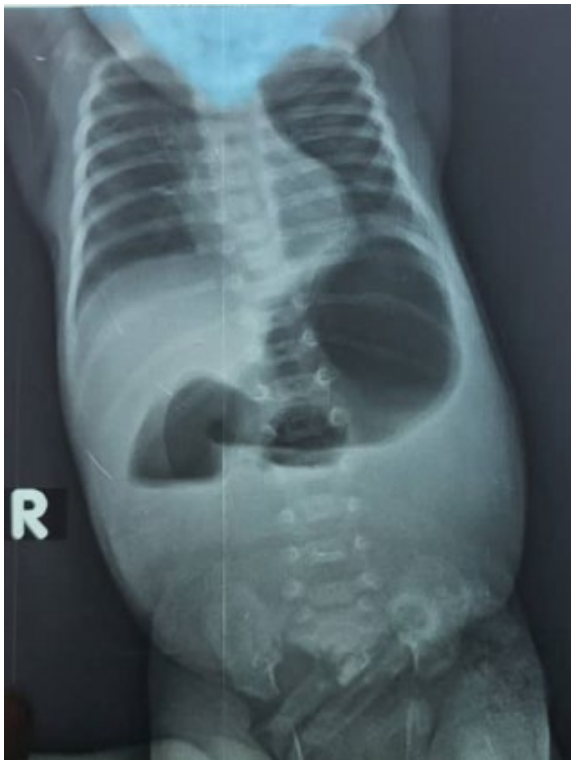

Figure 1. Abdominal radiograph showing a "double-bubble" sign.

Laparotomy revealed proximal jejunal atresia with a dilated and elongated duodenal segment (Figure 2). The ileal segment of the small intestine was wrapped around the atretic jejunal segment, forming a spiral configuration resembling an apple-peel (Figure 3). There was intestinal malrotation with a Ladd's band crossing in front of the duodenum and attaching to the cecum, which was located medially in the hypochondrium, along with a short common mesentery (Figure 3). Due to the malrotation, the root of the primitive intestinal loop was narrow, leading to a two-turn volvulus and extensive small bowel necrosis extending up to the ileocecal junction (Figure 4).

We performed detorsion of the volvulus, followed by resection of the entire necrotic small intestine, which measured approximately 180 cm. Ladd's bands were released and the cecum was placed in the left hypochondriac region. An end-to-side duodeno-colic anastomosis was performed. The immediate postoperative course was uneventful. Due to the unavailability of parenteral nutrition in our setting, basic nutritional and metabolic requirements were provided with glucose-infused saline, electrolytes, multivitamin complexes, and a postoperative blood transfusion. Enteral feeding via a nasogastric tube was initiated on postoperative day 3 but was not tolerated. Parenteral fluid and electrolyte support was maintained. By day 7, the patient had developed severe malnutrition and significant electrolyte imbalances. He passed away in the neonatal unit on postoperative day 26, as a result of severe sepsis and profound electrolyte imbalance.

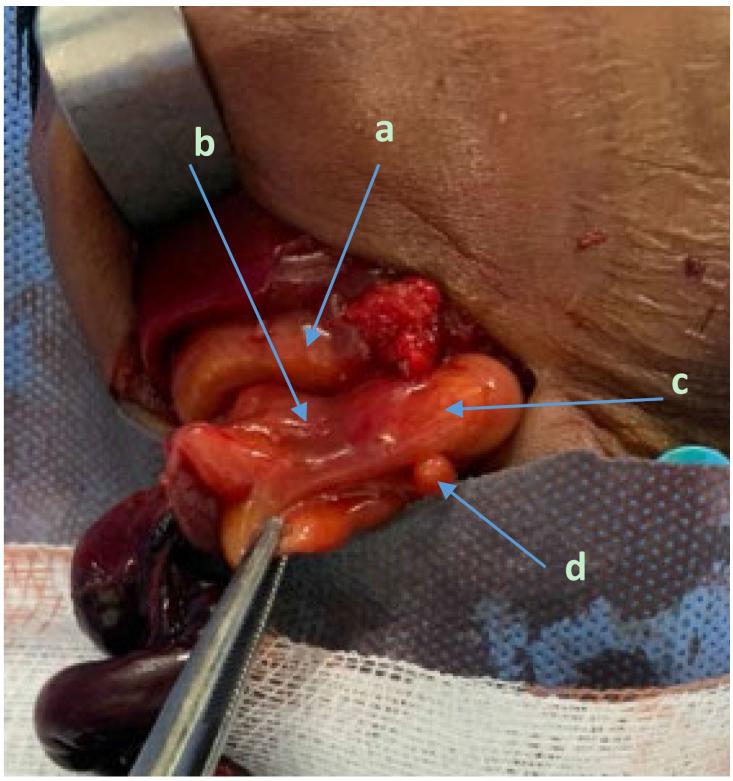

Figure 2. Proximal part of the duodenum dilated and elongated. a = duodenum; b = ileocaecal junction; c = cæcum; d = appendix.

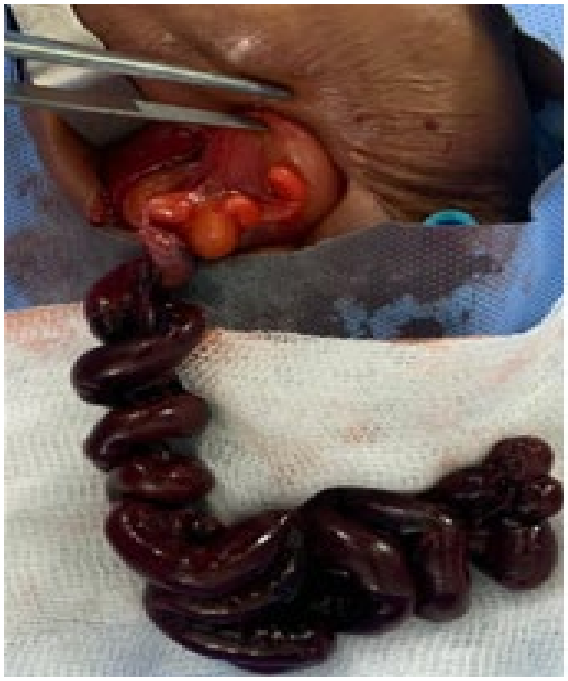

Figure 3. Apple-peel configuration.

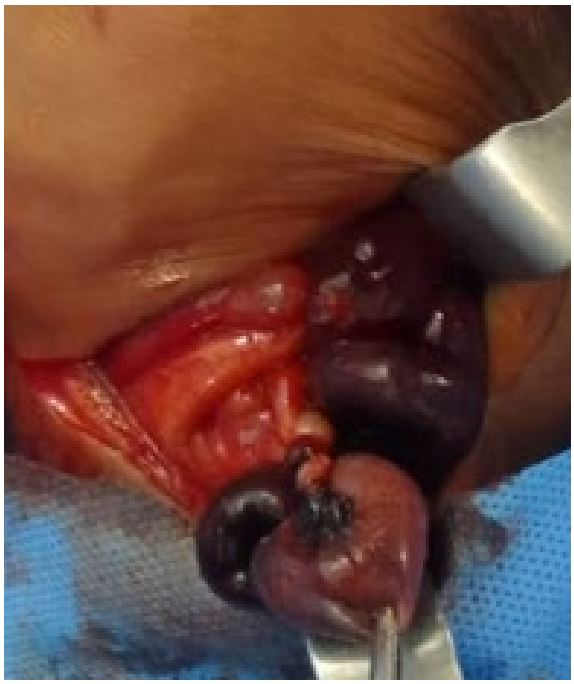

Figure 4. Volvulus due to malrotation with two spiral turns.

3. Discussion

Apple-peel syndrome is a rare form of jejunoileal atresia due to superior mesenteric artery obstruction, giving the aspect of proximal atresia with a pigtail-like form of the twisted distal segment around its vascular supply [5]. Apple-peel atresia can occur in isolation, but it may also be associated with other intestinal mal-

formations such as intestinal malrotation or duodenal atresia [1] [4] [6] [7]. Our patient has a very rare association with apple-peel syndrome associated with intestinal malrotation. The pathogenesis of intestinal atresia varies according to the site of the lesion. Embryologically, the endoderm gives rise to the gut tube, beginning in the fourth week of gestation. In the sixth week, gut epithelium proliferates rapidly, resulting in obliteration of the intestinal lumen. The intestine is then gradually recanalized over the next several weeks of development. Duodenal atresia is thought to result from a failure of recanalization during the solid stage of foregut development, whereas atresia of the remaining small intestine is believed to result from an interruption of blood supply to the atretic segment [8]-[10]. Apple-peel atresia or "Christmas-tree deformity" consists of a 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 [11]-[13]. An intrauterine vascular accident in late gestation has been accepted as the cause of apple-peel atresia, which presents with a spectrum of occlusions of one or more branches of the superior mesenteric artery [14] [15]. It should be noted that classic intestinal malrotation results from a failure of embryonic gut rotation between the 5th and 10th weeks of gestation and is therefore congenital. It is generally not caused by an intrauterine vascular accident. In contrast, certain intestinal atresias, such as apple-peel atresia, result from an intrauterine vascular insult, leading to this characteristic configuration. In these cases, secondary malrotation may occur, predisposing to volvulus around a shortened mesenteric root [4]. The cecum was located on the left side, with the first and last intestinal loops in close proximity. The patient exhibited extensive necrosis of the small intestine with an "apple-peel" configuration, attributed to volvulus of the recurrent ileal artery, which compromised retrograde vascularization. Small intestine necrosis is a factor of very poor prognosis; however, it is not always observed [11]-[13]. In our patient, detorsion of the volvulus was performed, followed by resection of approximately 180 cm of necrotic small intestine. Ladd's bands were divided, the cecum positioned in the left hypochondrium, and an end-to-side duodeno-colic anastomosis constructed. This approach was chosen due to the patient's hemodynamic stability after resection and the limited availability of resources for stoma care. Patients with apple-peel syndrome exhibit a high incidence of prematurity (70%) and mortality (54%) [1]. The patient was born prematurely at 35 weeks of gestation. He passed away on day 26 of life due to sepsis and metabolic disturbances.

In settings where parenteral nutrition is unavailable, the prognosis of short bowel syndrome remains particularly poor. Supportive strategies can nonetheless improve outcomes, including early trophic feeding, which promotes intestinal adaptation, and specialized enteral formulas. Careful management of fluid and electrolyte intake is also essential to maintain hydration, acid-base balance, and homeostasis. Regular monitoring and appropriate electrolyte supplementation help

prevent complications and support recovery.

The association between apple-peel syndrome and intestinal malrotation is extremely rare. This malformative combination presents significant diagnostic and therapeutic challenges, particularly in low- and middle-income countries (LMICs). In Africa, diagnosis is often delayed due to the lack of prenatal imaging, late referral of neonates, and a shortage of neonatal surgical expertise in some areas. Management is especially complex because of the high risk of short bowel syndrome, compounded by the frequent unavailability of parenteral nutrition in these settings. This situation results in very high mortality rates. Improving care requires strengthening neonatal care capacities, ensuring access to parenteral nutrition, and better organizing specialized services.

4. Conclusion

The association of apple-peel syndrome with intestinal malrotation represents a rare but severe congenital anomaly, which is particularly challenging in resource-limited settings. Delayed diagnosis and limited therapeutic options largely contribute to the high mortality rates reported. Strengthening neonatal care infrastructure and ensuring broader access to parenteral nutrition are critical. Future efforts should focus on creating regional neonatal surgery centres and implementing low-cost nutritional support protocols to improve outcomes in low- and middle-income countries.

Funding

This study did not receive any funding.

Ethics Approval and Consent to Participate

Formal ethics committee approval was waived for this retrospective case report, which was conducted in accordance with the principles of the Helsinki Declaration.

Availability of Data and Materials

The datasets used and analyzed during the current study are available from the corresponding author upon reasonable request.

Authors' Contributions

All authors contributed to the manuscript.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Seashore, J.H., Collins, F.S., Markowitz, R.I. and Seashore, M.R. (1987) Familial Apple Peel Jejunal Atresia: Surgical, Genetic, and Radiographic Aspects. *Pediatrics*, **80**,

540-544. https://doi.org/10.1542/peds.80.4.540

[2] Lambrecht, W. and Kluth, D. (1998) Hereditary Multiple Atresias of the Gastrointestinal Tract: Report of a Case and Review of the Literature. *Journal of Pediatric Surgery*, **33**, 794-797. https://doi.org/10.1016/s0022-3468(98)90225-1

[3] Chinya, A., Naranje, K. and Mandelia, A. (2019) Situs Inversus Abdominalis, Polysplenia, Complex Jejunal Atresia and Malrotation in a Neonate: A Rare Association. *International Journal of Surgery Case Reports*, **56**, 93-95. https://doi.org/10.1016/j.ijscr.2019.02.016

[4] Adams, S.D. and Stanton, M.P. (2014) Malrotation and Intestinal Atresias. *Early Human Development*, **90**, 921-925. https://doi.org/10.1016/j.earlhumdev.2014.09.017

[5] Gueye, D., Fall, M.B., Ndour, D., Ahye, D., Ndoye, A., Mbaye, P.A., *et al.* (2019) A Very Rare Case of Intestinal Atresia, the Apple Peel Syndrome. *Annals of Clinical Case Reports*, **4**, Article 1700.

[6] Gross, E., Armon, Y., Abu-Dalu, K., Gale, R. and Schiller, M. (1996) Familial Combined Duodenal and Jejunal Atresia. *Journal of Pediatric Surgery*, **31**, 1573. https://doi.org/10.1016/s0022-3468(96)90182-7

[7] Herman, T.E. and Siegel, M.J. (2008) Apple Peel Small Bowel. *Journal of Perinatology*, **28**, 380-382. https://doi.org/10.1038/sj.jp.7211922

[8] Tandler, J. (1900) Zur Entwicklungsgeschichte des menschlichen Duodenums in frühen Embryonalstadien. *Morphologisches Jahrbuch*, **29**, Article 187.

[9] Sadler, T. (2003) Digestive System. In: *Langman's Medical Embryology*, Lippincott Williams & Wilkins, 285-319.

[10] Kothari, P., Ahmad, A., Sarda, D. and Joshi, P. (2009) Duodenal Atresia with "Apple-Peel Configuration" of the Ileum and Absent Superior Mesenteric Artery: A Rare Presentation. *African Journal of Paediatric Surgery*, **6**, 120-131. https://doi.org/10.4103/0189-6725.54780

[11] Patil, R.T., Gupta, R., Parelkar, S.V., Oak, S., Sanghvi, B., Prakash, A., *et al.* (2011) A Rare Case of Duodenal Atresia with Apple-Peel Configuration of the Small Intestine and Malrotation. *European Journal of Pediatric Surgery*, **21**, 340-342. https://doi.org/10.1055/s-0031-1277134

[12] Altokhais, T.I. (2014) Duodenal Atresia with Apple-Peel Jujenoilial Deformity: Case Report and Review of the Literature. *Journal of Pediatric Surgery Case Reports*, **2**, 156-158. https://doi.org/10.1016/j.epsc.2014.03.001

[13] Wong, T., Volpe, F., Tazima, M.F.G., Canesin, W.C. and Sbragia, L. (2021) Duodenal Atresia with Apple Peel Jejunoileal Syndrome. *Journal of Pediatric Surgery Case Reports*, **64**, Article ID: 101709. https://doi.org/10.1016/j.epsc.2020.101709

[14] Lister, J. (1990) Intestinal Atresia and Stenosis, Excluding the Duodenum. In: Lister, J., Irving, I., Rickham, P.P., Eds., *Neonatal Surgery*, Butterworth & Co., 453-473.

[15] Chouikh, T., Ghorbel, S., Charieg, A., Benkhalifa, S. and Chaouachi, B. (2011) Antenatal Diagnosis of Intestinal Obstruction. An Unusual Association: Duodenal Atresia, "Apple Peel" Small Bowel Atresia, and Agenesis of the Superior Mesenteric Artery. *Revue de Médecine Périnatale*, **3**, 34-36.