

LETTER TO THE EDITOR

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Apple peel atresia: a rare case of a small premature infant

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Type IIIb intestinal atresia, also known as apple peel atresia, is a rare form of bowel atresia associating proximal jejunal atresia with wrapping of the distal small intestine in a spiral pattern around a thin vascular pedicle [1]. Even though mortality has declined steadily over the years, apple peel atresia remains a life-threatening malformation, which still leads to substantial morbidity and mortality [2]. The current opinion consists of preservation of as much bowel as possible (by doing end-to-end anastomosis) and of maximal peristaltic function (by doing resection, enteroplasty, or enterostomy of the dilated proximal segment), and an early enteral feeding [1,3,4]. Here, we report a case of a small premature infant with diagnosed apple peel atresia and prolonged the hospital stay.

A male baby was born with a birth weight of 1150 g at 29 weeks' gestation. He was intubated due to respiratory distress at delivery room, and then transferred to neonatal intensive care unit. On examination, he was intubated, pink, and had no abdominal distention or obvious external morphological congenital anomalies. Investigations revealed normal blood counts and biochemical values. Because of orogastric tube drained bilious aspirate, enteral feeding could not be started. Plain abdominal radiograph with contrast enema showed the classic "double-bubble appearance" of duodenal atresia with total absence of distal bowel gas (Figure 1). The other imaging studies were unremarkable for possible associated congenital anomalies.

He was taken for exploratory laparotomy at the end of the first week. The intraoperative findings showed atresia of the proximal bowel at the duodenojejunal junction, massively dilated duodenum and gall bladder and tracts, and that the distal small bowel was hypoplastic, and wrapped around its vascular supply in a spiral (Figure 2).



Figure 1. Upper gastrointestinal contrast study demonstrates dilated stomach and duodenum, with lack of passage of contrast agent to distal small bowel.

The distal, massively dilated duodenum was partially resected and end-to-oblique duodenojejunal anastomosis was done in a single extramucosal layer. The postoperative period was uneventful, but enteral feeding could not be begun due to the continuation of bile drainage through orogastric tube. Serial upper gastrointestinal contrast studies demonstrated the lack of passage of contrast agent to distal small bowel (Figure 3), and subsequently he required two additional exploratory laparotomies performing duodenojejunal re-anastomosis during the hospital stay (at the fourth week and the third month of admission, with weight of 1180 g and 1740 g, respectively) (Figure 4). Enteral feeds were started by the fourth month, but full feeds were not established until the sixth month due to recurrent vomiting and abdominal distension. He received total parenteral nutrition for a total of six months.

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The hospital course was complicated with nosocomial infection for three times and bronchopulmonary dysplasia requiring ventilator support for about three months. He was discharged with a weight of 3250 g (below the third percentile for corrected age) at the eighth month (Figure 5), and was doing well at the follow-up of three months, except several vomiting attacks.

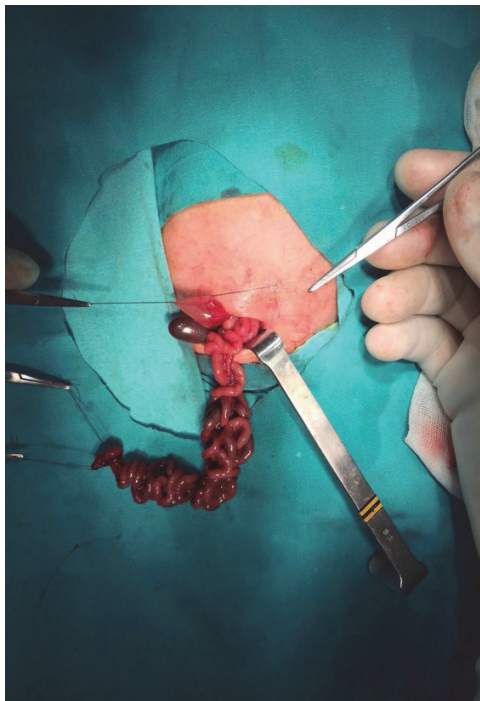


Figure 2. Typical 'apple peel' appearance of small bowel at operation.



Figure 3. Abdominal radiograph performed 2 weeks after the first operation shows the lack of passage of contrast agent to distal small bowel.

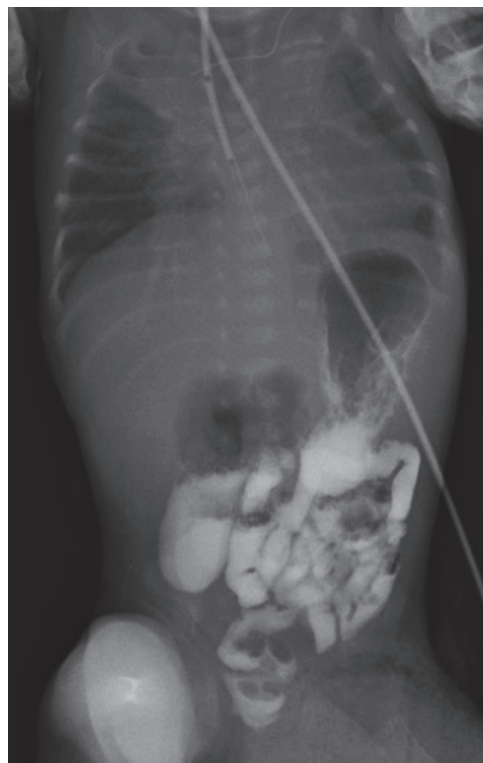


Figure 4. Abdominal radiograph performed 2 weeks after the third operation shows the passage of contrast agent to distal small bowel.



Figure 5. General appearance of the patient at discharge.

Apple peel atresia is a well-recognized though rare variant of jejunoileal atresia. Despite many surgical options have historically been used, the current surgery performed is the excision of the dilated proximal intestine and primary anastomosis [1,4]. The most frequent causes of death are associated anomalies, sepsis, anastomotic dysfunction, prematurity, intestinal gangrene, malnutrition and respiratory complications [1]. However, recent reports suggest an improvement in prognosis, which is generally attributed to improvements in surgical technique and neonatal intensive care facilities, and the advent of parenteral nutrition [1,4]. If the newborns survived the operative and postoperative periods, they are faced with the morbidity associated with malnutrition and prolonged parenteral nutrition [2,4].

The case presented here had very low birth weight, and this condition made him with poor prognosis. Due to anastomotic obstruction, which has been reported with a frequency ranging from 17% to 27% [2], two reoperations were required for achieving substantially intestinal passage. Although it initially seemed desperate, once the patient's weight gain seems to have increased the success rate of subsequent surgery.

Despite very low birth weight, initial complications, late morbidities and prolonged hospital stay, the long-term outlook for this patient is favorable. Although the vast majority of such patients are likely to weigh less than many of their peers, they can develop normally and achieve a normal growth curve [2]. Strategies that allow to early initial enteral feeding should be developed, so that the complicated hospital course for small premature babies can be reduced.

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