RARE JEJUNOILEAL ATRESIA IN TWO SINGLETON BROTHERS BORN THREE YEARS APART

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ABSTRACT

Small intestinal atresia is a common congenital malformation and it is a well-known cause of intestinal obstruction in neonates. Familial occurrence of duodenal atresia is extremely rare and has been attributed to chromosomal aberrations and parental consanguinity suggesting autosomal recessive inheritance. Distal intestinal anomalies, such as jejunoileal atresia, are a rare congenital malformation of the small bowel and have been related to vascular occlusion in the earlier or later stages in pregnancy and genetic causes. Familial jejunoileal atresia in twins is an extremely rare occurrence that is attributed to the use of some chemicals and other dyes instilled during diagnostic amniocentesis and has been described as a cause of jejunoileal atresia in twin-brothers. Authors of two different research institutes stated that jejunoileal atresia is significantly more frequent in twins than in singletons. In the present article we describe an extremely rare occurrence in two singleton infants, who were born three years apart, with similar jejunoileal atresia with no other associated malformations or chromosomal anomalies, who were treated in 2004 and 2007, respectively. Despite investigation, we did not find any reason for this particular occurrence; however they will be closely followed in order to detect any development alterations that could indicate an associated malformation.

Key words. Jejunoileal atresia; familial intestinal atresia; familial inheritance; intestinal obstruction.

INTRODUCTION

Small intestinal atresia is a common congenital malformation and it is a well-known cause of intestinal obstruction in neonates.1,2 Its prevalence has been described as of 1-1.8 per 10,000-14,000 live births.1,3 Proximal intestinal obstruction such as duodenal atresia seems to be related to polyhydramnios during pregnancy.4 Other associated malformations occur in almost 65% of the cases. They may be Down syndrome (30%),4 isolated cardiac
defects (34%), esophageal atresia (7-10%) and other gastrointestinal anomalies, as anterior portal vein, second distal web, anorectal, cloacal and urinary tract, intestinal malrotation, annular pancreas, and rarely choledochal cyst. Its cause is attributed to lack of recanalization by vascular coalescence in the solid developing duodenum. Familial occurrence of duodenal atresia is extremely rare, and has been attributed to chromosomal abnormalities and parental consanguinity suggesting autosomal recessive inheritance.

On the other hand, distal intestinal anomaly such as jejunoileal atresia is a rare congenital malformation of the small bowel. It has been related to superior mesenteric artery occlusion that leads to apple peel like atresia in the earlier stages of gestation or due to other mesenteric-vascular lesions that occur late in the gestational period. More recently it was published that more than 25% of cases of jejunoileal atresia were associated to chromosomal anomalies.

The occurrence of familial jejunoileal atresia in twins is very rare, but it was observed in cases in which some drugs, such as methylene blue, were instilled during diagnostic amniocentesis. It has been described as a cause of jejunoileal atresia only in twin pregnancies. Cragan et al. stated that the rate of jejunoileal atresia per 10,000 livebirths among twins is significantly higher than that among singletons.

Due to the interesting aspect of the present occurrence, two premature male singletons are presented, born three years apart, they were submitted to treatment in 2004 and 2007 of a very similar jejunoileal atresia. There was no consanguinity among parents and no other associated malformation.

CASES

Case 1. One white boy was born at 34 weeks, weighing 2,200g, delivered by cesarean section due to fetal mispositioning in uterus and early delivery. There was no prenatal diagnostic by ultrasound. Both parents were 28 years old at that time and had no history of consanguinity. No polyhydramnios was reported during late stages of pregnancy and gestation was uneventful. In the day after delivery, two bile-stained vomiting episodes and a moderate abdominal distention was noted. After the third bile-stained vomiting a plain abdominal radiograph was taken about thirty hours after birth. The radiograph showed large air-bubbles inside the intestinal loops and no air in the remaining abdomen (Figure 1), and the diagnosis of intestinal atresia was given. At surgery, a unique type III-A jejunoileal atresia was diagnosed about 40 cm ahead the Treitz angle (Figure 2). It was corrected with the resection of the proximal dilated portion and an end-to-end primary anastomosis. Genetic studies were normal. The baby thrived very well and was discharged 21 days after surgery.
Case 2. The second patient reported was born 3 years after his brother, at 34 weeks, weighing 2,100g, also delivered by cesarian section. He was diagnosed by a prenatal ultrasound at the 29th week. No polyhydramnios and no other events during gestation were reported. This baby had a large amount of secretion through the gastric tube and a plain thoraco-abdominal radiography taken few hours after birth showed poor air distribution only in the upper third of the abdomen (Figure 3). He was not fed before surgery. In the operating room, a type I jejunoileal atresia was diagnosed about 35 cm ahead the Treitz angle (Figure 4) and it was corrected with a short resection of proximal dilated portion and an end-to-end primary anastomosis, as the procedure to which his brother was submitted. Genetic studies were also normal. The baby thrived and was discharged 45 days after surgery due to infection problems.

DISCUSSION

Distal intestinal anomaly such as jejunoileal atresia is a very rare congenital malformation of the intestine. According to the literature, late intrauterine mesenteric vascular accident is related to the jejunoileal atresia origen. The low number of jejunoileal atresia cases is thought to be related to familial occurrence such as “apple peel atresia”, multiple atresias and duodenal atresia.

Familial jejunoileal atresia occurrence in twins is extremely rare, but it had been described in cases in which methylene blue and other dyes were instillated during diagnostic amniocentesis and may be a cause of jejunoileal atresia only in twin pregnancies.

The McDonnell Douglas Health Information System (MDHIS) published a study of approximately 2.8 million live-born infants between 1982 and 1988, of which more than 51,000 were twin infants. The findings of this research institute showed that in 569 singleton infants the rate of jejunoileal atresia was of 2 per 10,000 singleton live births. On the other hand, there were 30 twin infants with jejunoileal atresia at a rate of 5.5 per 10,000 live-born twin infants, which was significantly higher than the findings in singleton infants. In 1994, MDHIS reported that among all the studied strata, such as rate of jejunoileal atresia between twin and singleton; male twins and male singletons; female twins and female singletons and white twins and white singletons, twin infants presented significantly higher rates than those compared to singleton infants in all cases. Among singleton infants, black infants had a significantly higher rate of jejunoileal atresia than white infants.

The same results were obtained by The Metropolitan Atlanta Congenital Defects Program (MACDP). This research institute presented results similar to MDHIS findings during a 21-year period (1968-1989). In approximately 619 thousand live born infants, 12,300 were twins. The rate of

Figure 3. Abdominal radiography in supine position, about five hours after birth, showing three small dilatation of the intestinal loops in a moderate extension, without images of obstruction. Gastric tube in a good position. Poor distribution of gases in the remnant abdominal areas.

Figure 4. A large dilatation of the proximal jejunal loop in a continuing distal portion atresia of the intestinal loop as type I according to Louw and Barnard classification. There is no lesion of the mesenterium.
jejunoileal atresia was 2.5 per 10,000 singleton live births and 7.3 per 10,000 live-born twin infants.\textsuperscript{17}

In the present report, it is clear that these two cases are extremely rare, and present some characteristics that oppose to most of the current published cases, such as white and male singleton infants, given that those reported in the literature with a high jejunoileal atresia rate were male twin infants.\textsuperscript{17,20} Even among singleton infants and in the findings of both institutes (MDHIS and MACDP), black infants have a significantly higher jejunoileal atresia rate when compared to white infants. The same is observed in female singleton infants, that have higher jejunoileal atresia rate than male infants.\textsuperscript{17}

No other associated malformations were found in the babies reported here and no consanguinity of the parents was detected.\textsuperscript{10}

Concluding, the reported cases are extremely rare malformations in singleton infants and despite the undetected chromosomal or structural abnormalities, environmental factors may be involved. A close follow-up will be conducted in order to detect any alterations that might indicate an associated chromosomal malformation.

DISCLOSURE
No potential conflict of interests relevant to this article was reported.

ACKNOWLEDGEMENT
We acknowledge Ms. Chris Paiva for her worthy translation service.

REFERENCES