

Preduodenal Portal Vein (PDPV): A Very Rare Cause of Duodenum Obstruction

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Abstract

Congenital duodenal obstruction (CDO) is one of the most common anomalies presented in the neonatal period of intestinal obstruction, with an incidence of 1:10,000, and Preduodenal Portal Vein (PDPV) is a rare extrinsic cause of CDO. PDPV leads to CDO with a persistent vomiting, impairment of the oral feeding and weight loss, but other publications state an asymptomatic disease that may requires late surgical correction. Prenatal diagnosis of CDO, including PDPV, is evidenced by association of polyhydramnios with a dilated fluid-filled gastric and duodenum and a double-bubble image in the ultrasonography and permits a proper management of associated malformation, delivery, surgery and allows family counseling. PDPV is commonly associated to other intestinal obstruction such as extrinsic (*annular pancreas, adhesive bands, malrotation, peritoneal bands, intestinal duplication*), and intrinsic (*atresia, web and stenosis*), and also associated with cardiovascular anomalies, situs inversus, Down's syndrome and polysplenia. This is full term 5 days old newborn, 2865 g girl, without prenatal diagnosis of CDO. It was not still established PDPV until surgery procedure such as Duodenoduodenostomy (*Diamond-Shape*) and can guide Paediatric Surgeons that face high intestinal obstruction in the neonate with or without prenatal diagnosis. In the ambulatory follow up after 4 months, baby has developed very well, in breast-feeding associated with formula-feeding with a normal weight gain (average of 550 g/month).

Keywords

Duodenal Obstruction, Preduodenal Portal Vein, Polysplenia, Ultrasonography

1. Introduction

Congenital duodenal obstruction (CDO) is one of the most common anomalies

that is presented in the neonatal period in almost 50% of all cases of the newborn intestinal obstruction, with an incidence of 1:10,000 [1] [2]. Preduodenal Portal Vein (PDPV) is reported such as a very rare anomaly that causes CDO [3] [4]. In 25-year retrospective study Kouwenberg M *et al.*, 2008, reported 5 patients with PDPV [5]. Chen QJ *et al.*, 2014, reported only 2 cases of PDPV in 287 newborns (0.7%) in 10 years (2003-12) [6].

PDPV is described as a rare condition that happens in the neonatal period that leads to CDO causing persistent vomiting, impairment of the oral feeding and weight loss [6]. On the other hand, other publications state a generally asymptomatic disease and may lead to intestinal obstruction as well, requiring late surgical correction [7] [8] [9].

Prenatal diagnosis of CDO, including PDPV, is evidenced by association of polyhydramnios with a dilated fluid-filled gastric and duodenum and appearing as double-bobble image on ultrasonography [10] and provides important data for a proper management such as: search of associated malformation, plan of the delivery and neonatal surgery and allowing for better family counseling [11]. Other studies present that CDO can affect pre-operative complication, time for the surgery, delay of oral feeding and time of hospital stay, but mortality or morbidity are not affected [12].

CDO is commonly associated with other mechanisms of intestinal obstruction such as extrinsic (*annular pancreas, adhesive bands, malrotation, peritoneal bands, intestinal duplication, including PDPV*), and intrinsic (*atresia, web and stenosis*) [2] [13]. PDPV is also associated to cardiovascular anomalies, *situs inversus*, Down's syndrome and polysplenia [4] [5] [14].

This is a 5 days old newborn without prenatal diagnosis of CDO. Diagnosis was not still established PDPV until surgery and can guide specially Paediatric Surgeons that face high intestinal obstruction in the neonate with or without prenatal diagnosis [5]. An informed consent from the family was signed in the second month of follow-up and it is in the baby's chart at the hospital.

2. Case Report

A full term (38th week of gestation) 5 days old newborn with a 2865 g girl. At the delivery room Neonatologist has sucked up to 55 ml from gastric tube suggesting CDO. On the first day of life, the patient eliminated meconium and proceeds with vomiting after feeding with bile stained content.

On the second day of life, there was a persistent vomiting even with an open gastric tube and the baby was submitted to a radiologic procedure that showed an enlarging of the gastric image and a second at the duodenum site, strongly suggesting a double bobble image of CDO (Figure 1).

The newborn was submitted to a laparotomy in the 5th day of life, through a right upper transversal incision and confirmed CDO with a large Gastric and Duodenum dilatation. At the beginning it was supposed to be an intrinsic cause of obstruction, because it was not identified an annular pancreas or even a large

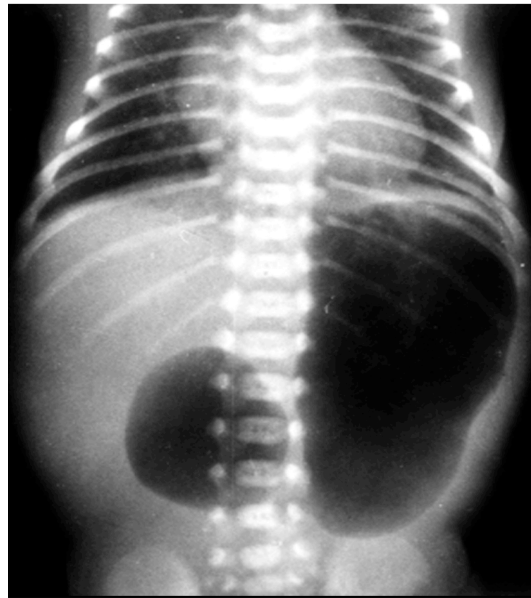


Figure 1. Showed an enlarging of the gastric image and a second one at the duodenum site, strongly suggesting a double-bubble image of CDO.

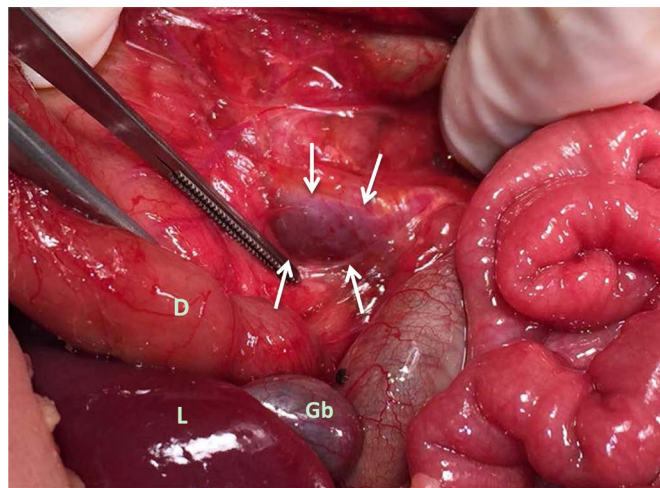


Figure 2. White arrows show PDPV obstructing Duodenum (D). Below D it is the Liver (L), Gallbladder (Gb). Tweezer is over the duodenum mesentery close to the duodenum narrowing behind PDPV.

Ladd's band. But following duodenum narrowing and obstruction towards its 3rd portion it was identified a PDPV causing complete obstruction (**Figure 2**) [14] [15]. It was associated with malrotation of the Colon but no polysplenia.

Correction was performed with a duodenoduodenostomy anastomosis through a Diamond-Shape suture with 5-0 polipropilene string [1] [14].

The baby was discharge from hospital in the 16th day after surgery, in breast-feeding associated with formula-feeding (50% - 50%), and she had a weight gain of 160 g in almost 2 weeks. Four months of age, she's developed very well, scar with a very good aspect, keeping a mix of breast and formula-feeding (30% - 70% in volume of each, respectively). She has got 2.030 g in this period.

3. Discussion

CDO is one of the most common anomalies in the neonatal period that causes high intestinal obstruction [1] [6]. Despite CDO frequently leads to persistent vomiting, impairment of the oral feeding and weight loss [6], other studies state asymptomatic or symptomless CDO requiring late surgical correction [2] [7]. Despite no consensus has yet been reached regarding its exact mechanism [16], extrinsic and intrinsic presentations of CDO have been reported in the literature [2] [13], other mechanisms such as isolated familial annular pancreas, suggesting a genetic basis for the CDO development [16], intrinsic obstruction such as food impact in a congenital membranous duodenal stenosis was recognized upon endoscopy have been published recently [17].

Predoduodenal Portal Vein (PDPV) is reported such as a very rare anomaly that causes CDO and associated with duodenal stenosis and Down syndrome, differently of the presented case that didn't present any malformations associated [3] [4]. Kouwenberg M *et al.*, 2008, reported 5 patients with PDPV [5] in a 25 years retrospective study. Chen QJ *et al.*, 2014, reported only 2 cases of PDPV in 287 newborns (0.7%) in 10 years (2003-12) [6]. Surgical correction was similar to the literature that is duodenoduodenostomy anastomosis through a Diamond-Shape [3] [6] [8] [18].

In this report, a term neonate had a good birth weight and presented high intestinal obstruction symptoms since he was born, while literature states 20% of premature and delayed presentation [6]. Literature states malrotation in 31% - 60% of the cases, such as in this neonate [1] [6]. There were no other associated anomalies.

Prenatal diagnosis of CDO provides important data for a proper management for the neonatal team and helps to prepare a better surgery conditions, besides permit family counseling [11]. Other study states that complication is significantly small in the prenatal diagnosis group of CDO when compared with the group without prenatal diagnosis, but it didn't affect mortality or morbidity in both of them [12]. Meanwhile, some authors state that "timely diagnosis and aggressive surgery are key" to improve prognosis [6].

In conclusion, a complex entity with various etiologies and concomitante disorders [6], we strongly recommend that in a presence of a newborn with CDO, with prenatal polyhydramnios and with the described symptoms, double-bubble image at the X-ray, Pediatric Surgeons investigate PDPV if intrinsic or extrinsic cause for CDO isn't identified. A prenatal detailed evaluation should be done in all suspected cases with those signs of CDO, searching for associated cardiovascular and other anomalies, due to preparing all the medical team and program an adequate family counseling.

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