



*Olgu Sunumu*

**Nonbilious Kusma ile Yenidoğan Döneminde Konjenital Duodenal  
Obstrüksiyonun Nadir Bir Nedeni: Annüler Pankreas**

Zühal Örnek <sup>a</sup>, Duygu Tatlı Uçarcı <sup>b</sup>

<sup>a</sup> Çocuk Sağlığı ve Hastalıkları Anabilim Dalı, Tıp Fakültesi, Bülent Ecevit Üniversitesi, Zonguldak, Türkiye,

<sup>b</sup> Çocuk Cerrahisi Anabilim Dalı, Tıp Fakültesi, Bülent Ecevit Üniversitesi, Zonguldak, Türkiye.

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Zühal Örnek

zuhalornek@gmail.com

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**ÖZET**

Yenidoğan bebeklerde konjenital duodenal obstrüksiyonun (CDO) çeşitli nedenleri iyi bilinmektedir, ancak spesifik bir radyolojik tanı koymak zordur. CDO'lar ekstrasik ve intrinik olarak sınıflandırılır. Annuler pankreas (AP) ekstrasik nedenlere karışır ve operasyon sırasında sıklıkla fark edilir. Anulus genellikle duodenumun ikinci bölümünü tamamen çevreleyen bir pankreatik doku bandından oluşur. Yenidoğan döneminde duodenumun ilk bölümünü tamamen çevreleyen, noninvaziv kusma ve sarılık ile birlikte CDO'nun nadir bir nedeni olarak AP'yi, tanımlayacağız ve tanı stratejilerini ile yönetimini tartışacağız.

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*Case Report*

**A Rare Cause of a Congenital Duodenal Obstruction in The Newborn Period with Nonbilious Vomiting: Annular Pancreas**

Zühal Örnek <sup>a</sup>, Duygu Tatlı Uçarcı <sup>b</sup>

<sup>a</sup> *Pediatric, Medicine Faculty, Bülent Ecevit University, Zonguldak, Turkey,*

<sup>b</sup> *Pediatric Surgery, Medicine Faculty, Bülent Ecevit University, Zonguldak, Turkey.*

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Zühal Örnek

zuhalornek@gmail.com

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**ABSTRACT**

The various causes of congenital duodenal obstruction (CDO) in the newborn infants are well known but a specific radiologic diagnosis is difficult to establish. CDOs are classified as extrinsic and intrinsic. The annular pancreas (AP) is involved in extrinsic causes, and is frequently recognised during operation. The annulus is usually consists of a band of pancreatic tissue that completely surrounds the second part of the duodenum. We will describe AP, completely encircled the first portion of the duodenum in the newborn period as a rare cause of CDO with nonbilious vomiting, jaundice and discuss the diagnostic strategies and management.

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## Introduction

Annular pancreas as a cause CDO was first described by Tieddeman in 1818 and first repaired by Vidal with a gastrojejunostomy in 1905. Annular pancreas is a rare congenital anomaly occurring in 1 of every 12,000 to 15,000 live births (1). During weeks 4 to 8 of normal fetal development, the dorsal and ventral pancreatic buds merge to form the pancreas during gut rotation (2,3). The tip of the ventral pancreas may become fixed to the duodenal wall during this rotation and, in turn, can be pulled around the right side of the duodenum forming an annulus (4). We want to emphasize that patients who are followed up with non-bilious vomiting should also be careful in terms of surgical pathology and that they can be shown with radiography.

## Case

A preterm baby weighing 2150 grams was born at 33 week gestation to a 30 year old second gravida mother. The pregnancy was complicated by polyhydramnios; and delivery was by Caesarian section. Both parents have no adverse genetic history and no known family history of congenital anomalies or gastrointestinal pathology. Physical examination revealed hypotonia, impaired sucking and signs of dehydration. Laboratory data was within normal limits except plasma glucose and bilirubin. Because of hypoglycemia intravenous fluid was began. Nonbilious vomiting was noted on the second day of life. Although she received phototherapy, total bilirubin levels increased. Following vomiting continued and revealed slight upper abdominal distention. Plain x-ray films of the abdomen demonstrated apparent high intestinal obstruction (see picture 1).

Roentgenograms revealed two large radiolucent intestinal outlines in the upper abdomen; the characteristic "double bubble" shadow of duodenal obstruction. Upper gastrointestinal barium graphy was then performed, showing overextension of the bulb and the first duodenal portion with minimum passage of contrast in the first duodenal portion (see picture 2).

A laparotomy was performed seventh day after delivery due to signs of gastrointestinal obstruction. At operation pancreatic tissue completely encircled the first portion of the duodenum (see picture 3).

Surgical gastrostomy was opened and the probe did not advanced from duodenum, then the case was accepted as AP. A side-to-side duodeno-duodenal anastomosis was performed.

Picture 1: Plain x-ray films of the abdomen demonstrated apparent high intestinal obstruction



Picture 2: Showing overextension of the bulb and the first duodenal portion with minimum passage of contrast in the first duodenal portion



The postoperative course was uncomplicated, and the baby was fed from gastrostomy. Then oral feeding was tolerated from the tenth day. The infant

was discharged from the hospital on the twenty sixth day following operation weighing 2600 grams.

Picture 3: The pancreatic tissue completely encircled the first portion of the duodenum



## Discussion

Congenital duodenal obstruction can present in two forms: intrinsic or extrinsic. The annular pancreas (AP) is involved in extrinsic causes, and is frequently recognised during operation (5). Annular pancreas is a congenital anomaly in which a ring of pancreatic tissue partially or completely encircles the proximal duodenum. The annulus is usually comprised of a band of pancreatic tissue that completely encircles the second portion of the duodenum.

Symptoms from AP can occur at any age from the neonatal period to adulthood, although it is estimated that almost two thirds of patients with AP remain asymptomatic for life. The age of onset depends upon the severity of obstruction at birth (6). The earliest and most common finding is bilious vomiting immediately after birth. Vomiting is bilious because, in the eightyfive percent of CDO cases, the bile duct penetrates the duodenum from the distal part of the obstruction. Although bilious vomiting is a surgical emergency and prompt investigation is the key in the management, nonbilious vomiting is also a reminder in the CDO cases. Generally nonbilious vomiting does not alert the physician to surgical problems. Most of the cases of AP are asymptomatic and a systematic screening for this anomaly does not exist in postnatal or prenatal standard practice (7). Obstruction of the intrapancreatic portion of the common bile duct from edema and fibrosis of the pancreatic head can also lead to jaundice (8,9).

The diagnosis is established by radiologic imaging. In symptomatic neonates a plain abdominal X-ray will show the classic 'double bubble' sign suggestive of duodenal obstruction. But this sign is

not specific for AP since it can also be seen in children other conditions including duodenal atresia, intestinal malrotation, and congenital duodenal or pyloric atresia. The diagnosis of AP used to be based on duodenography which showed duodenal overextension and restriction of passage of contrast material (10). On ultrasound, the fluid-filled dilated descending duodenum may also be seen encircled by pancreatic tissue in infants (11-15). Other patients will present beyond neonatal period with nonspecific symptoms or some will be asymptomatic during their entire life and AP will be discovered incidentally on Computed Tomography or Magnetic Resonance (16). In recent years, it has been reported that noninvasive methods such as Magnetic Resonance Cholangiopancreatography (MRCP) and Multidetector Computed Tomography (MDCT) may show congenital pancreatic abnormalities (17). However, some authors say that no further investigation is necessary, because all patients in newborn period with complete or partial duodenal obstruction require surgical correction and a definitive diagnosis can be made during laparotomy (18). Because of this, an emergency laparotomy was done without further examination.

The delay in the diagnosis of AP can cause dehydration, acid-base disorders and weight loss. For this reason, neonates with CDO are initially managed with nasogastric or orogastric tube decompression and intravenous fluids. Gastrointestinal losses are replaced appropriately, and placement of peripherally inserted central catheter line for parenteral nutrition is recommended, because feeding is commonly delayed for up to several weeks after repair. Surgery remains the main procedure of choice in patients in whom symptoms can be attributed to AP. The goal of surgery is to relieve duodenal or gastric outlet obstruction (19,20). The preferred surgical approach is bypass surgery of the annulus, which can be achieved via a duodenoduodenostomy, gastrojejunostomy, or a duodenojejunostomy. Resection of the annulus should be avoided since it is associated with numerous complications such as pancreatitis, pancreatic fistula formation, and incomplete relief of obstruction leading to unacceptably high morbidity (21). Although the optimal operation has been a matter of debate, in neonates, bypass using a duodenoduodenostomy is the preferred surgical modality, whereas in adults a duodenojejunostomy or gastrojejunostomy is recommended (22).

Annular pancreas is associated with other congenital anomalies in up to 70% of infants (for eg duodenal stenosis or atresia, Down's syndrome, tracheoesophageal fistula, and congenital heart defects) (23,24). Infants with AP associated with

duodenal obstruction were often born prematurely and/or had low birth weights. In all cases of polyhydramnios, the possibility of high intestinal obstruction should be entertained (25). Maternal polyhydramnios, noted in %30 to %65 of cases, is an early clue that should lead to further investigation. (25,26).

The differential diagnosis of AP causing neonatal duodenal obstruction include duodenal atresia, duodenal stenosis, congenital hypertrophic pyloric stenosis, malrotation of the colon, volvulus, periduodenal band, aganglionic megacolon, meconium ileus, web-like membrane across the duodenal lumen, jejunal or ileal atresia, paraduodenal hernia and intestinal neoplasm.

The prognosis of patients with AP depends largely upon the age of onset of symptoms and severity of the obstruction. The highest mortality, up to 40 percent, has been reported in infants and is possibly due to associated congenital abnormalities (27,28).

In conclusion; discovering AP as a cause of neonatal or pediatric emergency is a rare incident in any physician's career. Nonbilious vomiting in a newborn is generally considered due to medical reasons and does not alert the physician to a surgical cause and sometimes leads to under delay is taking a surgical opinion. Early diagnosis, combined with prompt surgical treatment and postoperative support, can improve these patients' outcomes as well as reduce costs.

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