CASE REPORT
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Partial annular pancreas in a 12-year-old girl

Parcijalni anularni pankreas kod devojčice od 12 godina

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Abstract

Introduction. Annular pancreas is a rare congenital anomaly in which a band of the pancreatic tissue, in continuity with the pancreatic head, completely or incompletely surrounds the descending part of the duodenum. An abnormal pancreatic development can cause complete annular pancreas, partial annular pancreas, and pancreas divisum. Complete annular pancreas is diagnosed in newborns, while the diagnosis of the partial annular pancreas is more frequently established in adults. The most reliable diagnostic methods are computed tomography and magnetic resonance cholangiopancreatography. The anomaly is treated surgically, using bypass procedures. **Case report.** A 12-year-old girl presented malnourished, with occasional feeding problems, vomiting, heartburn, and pain from infancy. The upper gastrointestinal series showed an extremely dilated stomach, the first and the second part of the duodenum. An endoscopic exam revealed the dilated stomach, pylorus, and the first and the second part of the duodenum with retained contrast, while the entrance of the endoscope into the third part of the duodenum was not possible. Computed tomography showed pancreatic tissue encircling the second part of the duodenum and the characteristic "crocodile jaw" sign. Roux-en-Y duodenojejunostomy was performed as a bypass procedure. **Conclusion.** The complete annular pancreas is a well-known and easily diagnosed anomaly in newborns. The partial annular pancreas is often poorly recognized, especially in patients who do not present with marked duodenal obstruction. Unrevealed, it causes chronic problems in food intake, with possible serious complications. Although a very rare condition in the pediatric population, partial annular pancreas should be taken into consideration in unclear cases of chronic poor oral food intake and vomiting.

Key words:
annular pancreas; child; diagnosis; endoscopy, digestive system; tomography, x-ray computed.

Apstrakt

Uvod. Anularni pankreas je retka kongenitalna anomalija kod koje deo tkiva pankreasa, u kontinuitetu sa glavom pankreasa, potpuno ili nepotpuno okružuje nishodni deo dvanaestopalačnog creva. Kao rezultat nepravilnog razvoja pankreasa mogu nastati kompletni anularni pankreas, parcijalni anularni pankreas i pankreas *divisum*. Kompletni anularni pankreas se dijagnostikuje kod novorođenčadi, dok se dijagnoza parcijalnog anularnog pankreasa češće postavlja kod odraslih osoba. Najpouzdanije dijagnostičke metode za otkrivanje ovog oboljenja su kompjuterizovana tomografija i magnetna holangiopankreatografija. Ova anomalija se leči hirurški, korišćenjem jedne od *bypass* procedura. **Prikaz bolesnika.** Prikazana je neuhranjena devojčica od 12 godina, koja je imala povremene probleme sa hranjenjem, povraćanjem, gorušicom i grčevitim bolovima od odojačkog uzrasta. Radiografskim pregledom otkriven je izuzetno proširen želudac, prvi i drugi deo dvanaestopalačnog creva. Endoskopskim pregledom viđen je proširen želudac, pilorus, prvi i drugi deo dvanaestopalačnog creva, sa zadržanim kontrastom, dok ulaz endoskopa u treći deo dvanaestopalačnog creva nije bio moguć. Kompjuterizovanom tomografijom otkriveno je tkivo pankreasa koje je okruživalo nishodni deo dvanaestopalačnog creva i karakterističan znak „čeljusti krokodila". *Roux-en-Y* duodenojejunostomija je učinjena kao *bypass* procedura. **Zaključak.** Kompletni anularni pankreas je dobro poznata anomalija, koja se lako dijagnostikuje kod novorođenčadi. Parcijalni anularni pankreas je često slabo prepoznatljiv, naročito kod bolesnika koji nemaju izraženu opstrukciju dvanaestopalačnog creva. Neotkriven, on izaziva hronične probleme u unosu hrane, sa mogućim ozbiljnim komplikacijama. Iako je veoma redak u pedijatrijskoj populaciji, parcijalni anularni pankreas treba uzimati u obzir kod nejasnih slučajeva hronično lošeg oralnog unosa hrane i povraćanja.

Ključne reči:
pankreas, prstenasti; deca; dijagnoza; endoskopija, gastrointestinalna; tomografija, kompjuterizovana, rendgenska.

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Introduction

Annular pancreas (AP) is a rare congenital anomaly in which a band of pancreatic tissue, in continuity with the pancreatic head, completely or incompletely surrounds the second part of the duodenum (D2). The malformation was first described by Tiedemann in 1818 and named as annular pancreas by Ecker in 1862 [1]. The first operation on the annular pancreas was performed by Vidal in 1905. The anomaly is a result of abnormal pancreatic development. The pancreas develops from a dorsal bud and two ventral buds that appear in the 5th gestational week. By the 7th week, the duodenal rotation causes the clockwise rotation of the fused ventral buds, passing behind the duodenum from right to left to fuse with the dorsal bud [1]. The ventral bud forms the posterior part of the pancreatic head and uncinate process, while the dorsal bud forms the anterior part of the pancreatic head, body, and tail [2]. There are several theories explaining the development of AP. There is an opinion that the ventral pancreatic bud fuses with the duodenal wall and rotates incorrectly around the duodenum [2]. Some authors stated that the reason for the development of AP is the abnormal movement of the ventral pancreatic bud [2]. On the other hand, some suggested that the reason for the condition is a primary duodenal abnormality, and the pancreas only fills the space around a narrowed duodenum. The hypertrophy of both dorsal and ventral pancreatic buds may be the cause of this condition [3]. However, most authors consider the ventral pancreatic bud as the cause of AP [1, 2, 4]. As a result of abnormal pancreatic development, complete annular pancreas (CAP), partial annular pancreas (PAP), and pancreas divisum (PD) can arise.

The prevalence of AP is about 5–15 cases per 100,000 adults according to autopsy studies [5], and 400 cases per 100,000 adults according to endoscopic retrograde cholangiopancreatography (ERCP) studies [6]. A review of 103 cases with AP suggested the same prevalence in children and adults [7]. PAP has a lower incidence than CAP. Moreover, according to recorded data, PAP is detected less frequently in children than adults. In the adult population, PAP is most frequently detected between 20 and 50 years of age, presenting with upper abdominal pain (70%), nausea, and vomiting (47%) as a result of incomplete duodenal obstruction [8]. Peptic ulcer, acute, chronic, or recurrent pancreatitis, and jaundice are complications of this anomaly [9–11]. Symptoms of duodenal obstruction predominate in children, whereas PAP most commonly presents as pancreatitis in adults [12]. The diagnosis is made using ultrasonography, upper gastrointestinal (GI) series, endoscopic procedures, computed tomography (CT), and magnetic resonance cholangiopancreatography (MRCP). The treatment of PAP is surgical and mainly directed to the relief of duodenal obstruction. The surgical options include duodenoduodenostomy, duodenojejunostomy, gastrojejunostomy, and Roux-en-Y duodenojejunostomy [8]. The long-term postoperative results are excellent in the absence of severe associated anomalies such as congenital heart diseases and anomalies of the alimentary tract.

Case report

A 12-year-old girl presented with occasional feeding problems, such as sporadic vomiting mostly occurring after dinner, which started from her infancy and worsened in the last 6 months. She was complaining of night heartburn and short-term spasmodic abdominal pain. She had a poor appetite, chewing food for a long time. During the last three months, before her admission to the local hospital, she lost 2.5 kg of body weight and menarche had not appeared. Upper GI series, performed in another hospital, had shown food retention in the distal esophagus, dilation, and ptosis of the stomach, while the endoscopic procedure had shown inflammatory changes in the gastric mucosa. The girl had only received antiulcer therapy and nutrition advice. She had also been reviewed by the endocrinologist and psychiatrist.

On admission to our hospital, the girl was very malnourished, aging 12 years and 8 months, with 145 cm of body height and 24.5 kg of body weight. Secondary gender characteristics were poorly developed.

An ultrasound exam revealed the distended stomach and D1 and D2 portions, with a larger amount of denser content, with peristaltic and antiperistaltic waves. At the level of mesenteric forceps, the duodenum was about 4.5 mm in diameter in the transverse plane.

The upper GI series showed extreme dilation and ptosis of the stomach down to the bispinal line. D1 and D2 were markedly dilated, with a transverse diameter of 4.5 cm (Figure 1).



Fig. 1 – Preoperative radiogram showing dilated stomach and first two duodenal portions.

Endoscopic findings revealed hyperemia of the distal part of the esophagus and elongated stomach with retained food and contrast medium for more than 16 hours. Pylorus was dilated, D1 and D2 portions strongly dilated and edematous, while the entrance into D3 was impossible. Upon the suspicion of upper mesenteric artery syndrome, a CT scan was performed and revealed the presence of barium contrast in the stomach, which was given 10 days earlier. The dilation of the

stomach, D1 and D2 portions of the duodenum, and collapsed D3 portion were evident. In the area of the major duodenal papilla, the pancreatic tissue was seen encircling D2 on the posterior and lateral sides ("crocodile jaw" sign). There were no signs of intestinal malrotation, dilation of the choledochus, or pancreatitis. Intraoperatively, a strong dilation of D1 and D2 was revealed, caused by the head of the pancreas, which surrounded 3/4 of the D2 circumference (Figure 2).

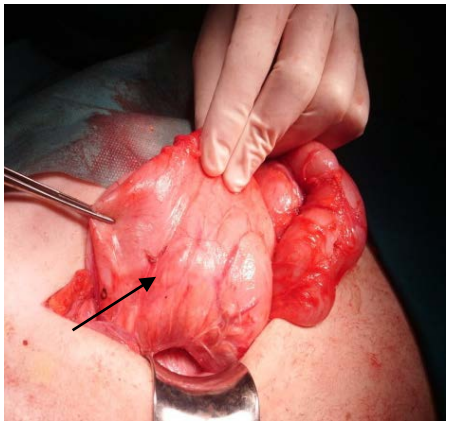


Fig. 2 – Intraoperative finding of pancreatic tissue on lateral duodenal wall (arrow).

The incision of the D2 showed the absence of a duodenal membrane. A Roux-en-Y duodenojejunostomy was performed as a bypass procedure. At the level of 20 cm from the ligament of Treitz, the jejunum was transected, and its distal portion anastomosed termino-laterally (T-L) with the D2. The proximal portion of the transected jejunum is T-L anastomosed with the distal portion of the jejunum, thus the conduit was about 25 cm long (Figure 3).

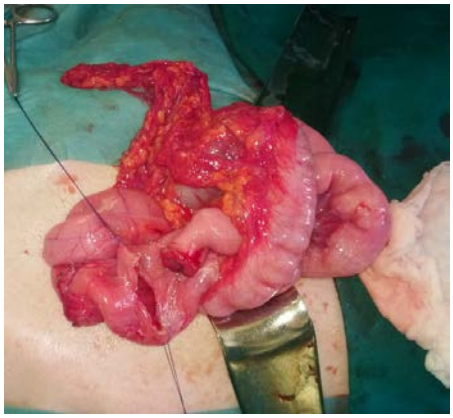


Fig. 3 – Duodenojejunal anastomosis.

The oral intake was started on the 4th postoperative day and was well-tolerated. The only postoperative problem was prolonged serious abdominal drainage due to hypoalbuminemia. During a 6-month follow-up period, the child had normal food intake, gaining 1 kg of body weight *per* month without any medical problem, so control radiographic and endoscopic exams were not indicated. Psychological and gynecological problems were resolved due to better food intake and weight gain.

Discussion

CAP is a well-known entity, presenting in newborns in the first days of life. Nowadays, the diagnosis of CAP is made in the early fetal period, according to an ultrasound exam. The time of onset of symptoms, clinical features, diagnostic methods, and treatment protocols are clearly explained in a huge number of literature data and are well-known because of clear and explicit symptoms of duodenal obstruction. On the contrary, PAP is a less frequent anomaly, and there is not enough literature data, especially considering the pediatric population [7]. The previous studies state that PAP, although a congenital anomaly, is most frequently detected between 20 and 50 years of age [5, 8]. The most prominent entities in the differential diagnosis of PAP are intestinal malrotation, perforated duodenal web, and upper mesenteric artery syndrome. There is a great delay in establishing the diagnosis of PAP. Therefore, patients suffer from duodenal obstruction symptoms, such as anorexia, vomiting, failure to thrive, and weight loss for a long time. A peptic ulcer can complicate the disease due to duodenogastric reflux caused by duodenal obstruction. The decreased outflow of pancreatic juice may give rise to acute or chronic pancreatitis. Jaundice can be present as a result of choledochal stenosis caused by the annular pancreas or as a result of biliary calculosis due to impaired biliary outflow [8]. In our patient, anorexia, vomiting, heartburn, and spasmodic abdominal pain were present sporadically 12 years prior to diagnosis. She was very malnourished, and her body weight was 24.5 kg at admission. A low body height of 145 cm and the absence of menarche were the results of chronic illness and poor food intake and were not caused by primary endocrinological disturbances. Our patient's symptoms were caused by duodenal obstruction, not by complications, which is the fact in accordance with literature data that children, unlike adults, seldom have complications of PAP [7]. Ultrasound exam should be indicative for PAP [10], although in our patient was not significant, suggesting superior mesenteric artery syndrome. Endoscopic ultrasonography has higher accuracy and is preferred [13]. The endoscopic exam can detect only complications of PAP but not the anomaly itself. CT scan and MRCP are reliable diagnostic tools in establishing the diagnosis, showing the characteristic "crocodile jaw" sign [14–16]. Two decades ago, in more than 40% of cases, the correct diagnosis was made only at laparotomy [17]. In our patient, a CT scan revealed the exact cause of the patient's problems and indicated operative treatment. Duodenoplasty, duodenoduodenostomy, duodenojejunostomy, gastrojejunostomy, and Roux-en-Y duodenojejunostomy are surgical options in the treatment of this anomaly [18, 19]. All these procedures can also be performed laparoscopically [20]. Although reported in the literature, the method of pancreatic resection has a higher incidence of complications, such as pancreatitis, pancreatic fistulae, and pancreatic insufficiency, and should be avoided [15]. Literature data do not suggest the preferable surgical procedure, so we

decided to perform Roux-en-Y duodenojejunostomy on our patient. In our opinion, duodenoplasty is not efficient for solving the problem. We consider that duodenoduodenostomy and duodenojejunostomy, the preferred methods in newborns, are inadequate in elder children because of a great dilation and disturbed motoric activity of the proximal duodenum and the stomach. Direct duodenojejunostomy and gastrojejunostomy can also lead to the postoperative reflux of jejunal content in dilated and hypoperistaltic duodenum and stomach [21]. Roux-en-Y duodenojejunostomy enables adequate duodenal emptying and prevents reflux of jejunal content; hence we consider this procedure an appropriate method for the treatment of PAP in elder children.

Conclusion

PAP is a rare congenital anomaly, unlike the well-known entity of CAP. The small incidence of this anomaly causes a great delay in establishing the correct diagnosis. It is very important to consider this anomaly as a potential cause of chronic feeding problems and vomiting. Operative treatment enables total cure and prompt recovery.

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