**Pancreas divisum: Analysis and therapeutic alternatives with a case report**

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**Abstract**

**Introduction.** Pancreas divisum is a relatively common pancreatic duct anatomic variant, firstly described in the 17th century. Case report. We reported a 2-year-old child admitted to the Pediatric Clinic with breathing difficulties and abdominal pains. Examination and X-ray image, showed a vast right hydrothorax containing rusty coloured solution with a high degree of amylase. Ultrasound and computed tomography examination revealed pancreatic polycyclic pseudocysts; following magnetic resonance cholangiopancreatography (MRCP), the diagnosis of pancreas divisum was confirmed. The general condition of the patient worsened, requiring an urgent operation. External drainage of the perforated pancreatic pseudocyst was performed. Following external fistula maturation, a change from external to internal drainage was performed using Roux-en-Y fistulojejunostomy. A 3-year postoperative period was uneventful. Conclusion. Pancreas divisum cases are unique requiring clinical experience, rational approach, and complex multimodal management. MRCP is a valuable diagnostic method. Amongst therapeutic options, outer and internal drainage can be seen as reliable methods. Further investigations are absolutely required to determine practical and appropriate conclusions.

**Key words:** pancreatic pseudocyst; diagnosis; digestive system surgical procedures; treatment outcome.

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tography (MRCP) is preferably used as a new and reliable non-invasive imaging technique. Many cases of PD may be asymptomatic, but an association with symptoms of acute or chronic is not rare. The anomaly is explained by the failure of the dorsal (Santorini) and the ventral (Wirsung) pancreatic buds to fuse during the gestational week 6–8, whereby the predominant drainage of the gland is through the duct of Santorini. The actual hypothesis is that there is an insufficient drainage of the entire dorsal gland through the minor papilla, meaning that there is a relative stenosis resulting in ductal hypertension. Other anomalies of the dorsal and ventral drainage of pancreatic buds include: the absence of the duct of Wirsung (PD type 2), the presence of a filamentous or a very small caliber communication between the dominant dorsal duct of Santorini and the duct of Wirsung (PD type 3, incomplete pancreas divisum). The predominant drainage of the gland is through the duct of Santorini channel and the duodenum was confirmed (Figure 2). The child had complications from acute pancreatitis to the point of acute abdomen necrosis or alike. Furthermore, the patient deteriorated from acute pancreatitis to the point of acute abdomen which required another urgent operation. External drainage of the perforated pancreatic pseudocyst was conducted by using a silicon catheter. The progression of the illness was stopped and in this manner the first signs of recovery were seen. Subsequently, pancreas divisum without communication of the Santorini channel and the duodenum was confirmed by MRCP (Figure 2). The child had complications of parenteral nutrition, as well. Afterward, a naso-jejunal sonde was placed to provide adequate enteral nutrition with minimal stimulation of pancreatic secretion. After the fistula's maturation of five weeks, a change of external to internal drainage was performed by using Roux-en-Y fistulojejunostomy (Figures 3 and 4). For this reconstruction, a silicone catheter was used as an endoprosthesis. During the postoperative period of 3 years, the child showed signs of continued clinical improvement and was symptom free.

**Case report**

A 2-year old child was admitted to the Pediatric Clinic with difficulties in breathing, mild abdominal pain, and refusal to eat. These symptoms began approximately two months prior to admission. They were mild at first but had worsened over time. Within the period of the onsetting symptoms, a bilateral inguinal hernia was also presented and the child was planned for herniectomy. After the admission to the Pediatric Clinic, chest X-rays confirmed vast hydrothorax on the right side (Figure 1). A thoracic drainage was made with a surprised discovery of a rusty coloured solution, with a high degree of amylase. Following an ultrasound and computed tomography examination, the diagnostics revealed polycyclic pseudocysts of the pancreas which ranged from 1 to 4 cm in size. The general condition of the child worsened and presented with a complete clinical picture of severe acute pancreatitis. Standard conservative treatments were initiated (continuous suction with a nasogastric sonde, antibiotics, H2 antagonists, parenteral rehydration and nutrition, stoppage of the autodigestion with octreotide, and pain control). The abdomen continued to gradually distend, and at one moment clinical signs of incarcerated inguinal hernia were presented. During herniectomy, diagnostic laparoscopy was performed through the inguinal hernial sac. The peritoneal solution was withdrawn containing a high level of amylase. However, we were unable to find other intra-abdominal signs or confirmations for pancreatitis (steatonecrosis or alike). Furthermore, the patient deteriorated from acute pancreatitis to the point of acute abdomen which required another urgent operation. External drainage of the perforated pancreatic pseudocyst was conducted by using a silicon catheter. The progression of the illness was stopped and in this manner the first signs of recovery were seen. Subsequently, pancreas divisum without communication of the Santorini channel and the duodenum was confirmed by MRCP (Figure 2). The child had complications of parenteral nutrition, as well. Afterward, a naso-jejunal sonde was placed to provide adequate enteral nutrition with minimal stimulation of pancreatic secretion. After the fistula’s maturation of five weeks, a change of external to internal drainage was performed by using Roux-en-Y fistulojejunostomy (Figures 3 and 4). For this reconstruction, a silicone catheter was used as an endoprosthesis. During the postoperative period of 3 years, the child showed signs of continued clinical improvement and was symptom free. Table 1 shows the results of the oral glucose tolerance test (OGTT) with the obvious preservation of physiological functions of the gland.

**Table 1**

<table>
<thead>
<tr>
<th>Time (hour)</th>
<th>Glycemia (mmol/L)</th>
<th>Insulinemia (mU/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>4.5</td>
<td>12</td>
</tr>
<tr>
<td>1</td>
<td>5.5</td>
<td>70</td>
</tr>
<tr>
<td>2</td>
<td>5.0</td>
<td>35</td>
</tr>
<tr>
<td>3</td>
<td>4.0</td>
<td>12</td>
</tr>
</tbody>
</table>

**Discussion**

The incidence of PD is estimated to be between 3–10% and an additional 0.13–0.9% of patients have incomplete form. Usually, patients have an early onset of recurrent episodic epigastric pain and vomiting at a mean age of 6 years. Many investigators have reported cases of pancreas divisum symptoms arising in late adulthood, as well. It is believed that the earlier onset of the disease represents a greater diagnostic problem even in cases where

Fig. 1 – X-ray of hydrothorax

MRCP, ERCP or other modern diagnostics are used. Usually, dominant abdominal symptoms such as pain, nausea, vomiting and some others are typical signs of acute or chronic pancreatitis. The difficulties in determining the diagnosis is further unfolded as patients may be asymptomatic.

During hospital admission, a number of diagnostics may be performed (standard ultrasound, CT, ERCP, endoscopic ultrasonography, Secretin test, Serum trypsin test, Fecal elastase test, and MRI/MRCP). Some of these diagnostic tests are not widely available (time and labour intensive, risk of pancreatitis, expensive, operator dependent, inaccurate, and may produce false-positive or false-negative results) 7. A main difficulty in the diagnosis of PD is due to its dimensions especially in younger patients. In our case, PD was discovered by MRCP. The radiologist must be experienced in order to understand and find the anomalies, and must be able
to recognize the Santorini duct which usually has an extremely small diameter as a probable typical sign of PD.

When discussing on therapeutic options, the problem may lie in choosing the appropriate procedures. Many studies have shown that these patients have benefits from the decompression of the minor papilla using sphincterotomy, sphincteroplasty or other surgical drainage procedures. In the absence of chronic pancreatitis, PD may be dealt with transpapillary pancreatic duct stent placement through the minor papilla and/or endoscopic sphincterotomy of the minor papilla. These procedures should be taken into consideration that they provide less invasive alternatives. Sherman et al. suggested the use of needle-knife pancreatic sphincterotomy over a pancreatic stent. The rate of pancreatitis following pancreatic sphincterotomy appears to be approximately 10–12%. Prolonged stinting, however, should be avoided due to the risk of inducing pancreatic damage and/or sepsis. It is recommended, thus, to remove a stent within 2–4 weeks. Dorsal pancreatic ductography should be applied to those patients who are suspected to have PD. Minor papilla cannulation can be achieved using catheters with tapered or 25-gauge needle tips. In patients with PD, if acute pancreatitis evolves into chronic pancreatitis, there may be an advantage to operate early in the disease by sphincteroplasty. Direct comparison of surgical and endoscopic series is difficult, although complication rates from surgery seem to be less frequent. If the results of sphincteroplasty are not better than endoscopic papillotomy, surgeons are usually recommended to perform pancreaticojejunostomy (Puestow procedure). A long-term success in the treatment of PD has been reported by performing percutaneous drainage for chronic pseudocysts. However, a persistent communication with the pancreatic duct might prevent its permanent and complete evacuation. It is suggested that only small cyst volumes and low amylase values might be associated with achieving percutaneous drainage. Currently, the experience in the majority of pediatric surgical clinics is limited to relatively few patients. Even with careful patient selection and meticulous surgical techniques to accomplish relief of ductal obstruction, the response to surgery is nevertheless inconsistent.

In our case, external drainage as a therapeutic method was shown to be successful. The patient from a near terminal stadium had lastly become stable, even without any signs of multiple-organ dysfunction. This was the reason to plan a permanent therapeutic solution. The patient’s recovery from a severe pancreatitis following a perforation of the pseudocyst due to external drainage inspired us to create an inner derivation as a definite therapeutic solution. A silicone catheter which was used as an endo-prosthesis in the external fistula prevented stenosis of the newly created fistulajejunostomy anastomosis, allowing its normal function.

The symptom-free period of three years has confirmed this approach, but the experience of other authors made us cautious and to continue with a long term follow-up of this patient. We were aware of alternative solutions whereby a formed external fistula can be completely resected, and that the standard pancreaticojejunal anastomosis can be created. This option comes to mind in the case of the realized derivation (fistulajejunostomy) that could be compromised.

**Conclusion**

All cases of pancreas divisum are unique and require clinical experience, rational approach, and even a complex multimodal management strategy. For these kinds of patients, a long-term observation is required with an active utilization of a wide range of diagnostic and therapeutic possibilities. Outer drainage showed to be a beneficial therapeutic option Altering outer drainage by permanent inner drainage, as in our case, seems to be successful. Puestow-pancreaticojejunostomy still represents a reliable solution in the management algorithm. A practical and appropriate conclusion requires, a longer series with a careful selection of patients.

**REFERENCES**


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