Pancreas divisum — peculiarity or anomaly?

N. B. Gubergrits, N. V. Byelyayeva, P. G. Fomenko

Donetsk National Medical University n. a. M. Gorky

Any strangeness can be regarded as an anomal,
if it attracts too much attention.

Unknown author

Key words: pancreas divisum, acute recurrent pancreatitis, chronic pancreatitis, diagnostics, treatment

Pancreas divisum is one of the most common abnormalities of the pancreas. Its diagnosis data are different in the general population. According to the results of the autopsy, this anomaly is found in 4-14% of cases, endoscopic retrograde cholangiopancreatography (ERCP) detects it in 3-8% and magnetic resonance cholangiopancreatography (MRCP) — in 9% of cases [10]. It is interesting that in Caucasians pancreas divisum occurs in 7% of cases, while the Africans and Asians, at least — in 1-2% of cases [8].

Normally, the dorsal and ventral ducts during embryogenesis are combined, the diameter of the ventral (Wirsung's) duct is substantially greater and drainage of pancreatic secretion is carried out mainly through it (Fig. 1A). Sometimes in the literature used the term "pancreatic anomaly with a dominant dorsal duct system" [8]. The first official description of pancreas divisum is made in 1865 by Joseph Hyrtl, although anatomists pointed out this anomaly earlier [16].

Development of pancreas divisum is associated with the fact that during the embryonic period ventral and dorsal pancreatic ductal systems are not merged, so the outflow of secretion occurs mainly through the extension (Santorini) duct. There are 3 variants of pancreas divisum (Figure 1.):

• complete pancreas divisum (Figure 1B, 2A), when the dorsal and ventral pancreatic ducts are completely isolated from each other, with the dominant dorsal duct;

• incomplete pancreas divisum (Figure 1B, 2B), when the dorsal and ventral ducts are interconnected at least a small sprig; the frequency of such an option — 15% of all cases of pancreas divisum [8];

• «reverse» pancreas divisum, when isolated dorsal duct is drained through a small duodenal papilla, and is not associated with the ventral duct (Figure 1E); in these cases, the pathology of small papillae, or break-it calculus cause more severe pancreatitis.

The clinical significance of pancreas divisum is determined by the fact that due to the small diameter and length of the main (Wirsung's) duct secret of the pancreas is drained primarily through the extension (Santorini) duct. Such drainage could not be complete and free. As a result, recurrent acute pancreatitis (AP) or obstructive chronic pancreatitis (CP) develop. Since pancreas divisum is the cause of idiopathic AP in 9,5-26,0% of cases [9]. In 25.6% of patients with idiopathic CP during ERCP reveal pancreas divisum [5]. CP development is associated with long-term increase in pressure in the extension channel, i.e. CP develops according to obstructive type. This explains the progressive atrophy of parenchyma of the dorsal part of the gland with the formation of its functional insufficiency (Fig. 3). However, pancreas divisum is the cause of CP is much less than the cause of AP — only 0.1% of all CPs [2].

In the vast majority of people with pancreas divisum there are no clinical manifestations. Indeed, the analysis of 6324 patients with ERCP protocols with biliopancreatic complaints shows that frequency of pancreas divisum was low and similar in CP (6.4%), AP (7.5%) and in the absence of pancreatic disease [6]. Similar results have been published by other authors [13, 14]. These findings contradict the results of P. B. Cotton [5]. Although, of course, the authors examined the different groups of patients with different purpose, which makes it difficult to compare the results.

On the contrary, in favor of a certain value in the development of pancreas divisum pancreatitis demonstrates the effectiveness of endoscopic or operative decompression, ductal pancreatic stenting in patients with abdominal pain and the presence of this anomaly [13, 14].

Perhaps a more equitable is an intermediate point of view that the risk of pancreatitis in the presence of pancreas divisum is promoted by the action of provoking factors, such as alcohol. In addition, the likelihood of developing pancreatitis is defined by the relationship between the small diameter of the small duodenal papilla and the degree of ductal hypertension Santorini duct [15].

Noting that biliary pancreatitis in the presence of pancreas divisum develops much less frequently than alcohol, which is associated with a low probability of blockade of additional duct by gallstones on the basis of the anatomical relations of the common bile duct and Santorini duct.

Diagnosis of pancreas divisum is based on the results of ERCP (Fig. 2), MRCP. Certain important data are also obtained by computer tomography (CT) and magnetic resonance (MR) imaging (Fig. 4). Attention is drawn to the possibility of developing santorinitsele — the bag-like expansion of additional duct in its terminal part (Fig. 5). To identify santorinitsele and stenosis of small papilla it is particularly informative to conduct MRCP with secretin.

When the presence of pancreas divisum is suspected and special methods should be applied (e.g., ERCP, MRCP with secretin) for its verification? If there is no difficulty in selective cannulation of the common bile duct, coupled with the inability of cannulation of the pancreatic duct; with a loss of contrast revenue entered through a large papilla, on the head of the pancreas; if you can’t put the conductor through a large papilla.

Treatment is prescribed only in the presence of clinical manifestations: with frequent attacks of light AP, severe AP, santorinitsele, in cases of absence of CP, alcohol abuse, the CFTR mutation. The principle of treatment is endoscopic or operative decompression of pancreatic ductal system. The first endoscopic methods are: sphincterotomy (usually small papilla), ductal stenting [11]. Papillectomy,

dilation of small holes papilla are also performed. In particular, in the endoscopic treatment of patients with frequent recurrences AP against the backdrop of pancreas divisum after sphincterotomy or stenting of small papillae of the dorsal duct with prolonged observation, a significant reduction in the frequency of relapses is observed. It should be noted that the number of patients with chronic abdominal pain decreased insignificantly [12].With the lack of effectiveness of endoscopic treatment doctors resort to surgery, in particular, perform transduodenal sphincteroplasty of small papilla, cholecystectomy and sphincteroplasty of large papillae. In a prospective study A. L. Warshaw et al. examined 88 patients who underwent sphincteroplasty. The period of follow-up was 29 months. The treatment was effective in 74% of cases. The frequency of restenosis small papilla was 8% [7].

An example of the successful surgical treatment of CP in patients with pancreas divisum is shown in Fig. 6.So after all, pancreas divisum is oddity or anomaly? It seems we still can’t clearly answer the question. First of all, we need to understand what is normal. This concept is also indefinite. The end of the article is finished by words of Sigmund Freud: "Everyone is normal only partly" This is worth considering...

References

1. Blair III A. J. Resection for pancreatitis in patients with pancreas divisum / A. J. Blair III, C. G. Russell, P. B. Cotton // Ann. Surg. — 1984. — Vol. 200. — P. 590–594.
2. Bradley E. L. 3rd. Accessory duct sphincteroplasty is preffered for long-term prevention of recurrent acute pancreatitis in patients with pancreas divisum / E. L. Bradley 3rd., R. N. Stephan // J. Am. Coll. Surg. — 1996. — Vol. 183. — P. 65–70.
3. Chronic pancreatitis: novel concepts in biology and therapy / Ed. M. W. Buchler [et al.]. — Berlin ; Wien : Wissenschafts–Verlag ; A Blackwell Publishing Company, 2002. — 614 p.
4. Clinical pancreatology for practicing gastroenterologists and surgeons / Ed. : J. E. Dominguez-Munoz. — Magdeburg : A Blackwell Publ. Co, 2005. — 535 p.
5. Cotton P. B. Congenital anomaly of pancreas divisum as cause of obstructive pain and pancreatitis / P. B. Cotton // Gut. — 1980. — Vol. 21. — P. 105–114.
6. Delhaye M. Pancreas divisum: congenital anatomic variant or anomaly? Contribution of endoscopic retrograde dorsal pancreatography / M. Delhaye, L. Engelholm, M. Cremer // Gastroenterology. — 1985. — Vol. 89. — P. 951–958.
7. Evaluation and treatment of the dominant dorsal duct syndrome (pancreas divisum redefined) / A. L. Warshaw, J. F. Simeone, R. H. Schapiro, B. Flavin-Warshaw // Am. J. Surg. — 1990. — Vol. 159, No 1. — P. 59–64.
8. Gastroenterology and hepatology: the comprehensive visual reference — pancreas, with MEDLINE-linked references / Eds. M. B. Feldman, P. P. Toskes. — Electronic Press Ltd., 1998.
9. Idiopathic recurrent pancreatitis: an approach to diagnosis and treatment / R. P. Venu, J. E. Greenen, W. Hogan [et al.] // Dig. Dis. Sci. — 1989. — Vol. 234. — P. 56–60.

10. Lehman G. A. Diagnosis and therapy of pancreas divisum / G. A. Lehman, S. Sherman // Gastrointest. Endosc. Clin. N. Am. — 1998. — Vol. 8, No 1. — P. 55–77.
11. Lehman G. A. Endoscopic management of recurrent and chronic pancreatitis / G. A. Lehman, S. Sherman, R. H. Hawes // Scand. J. Gastroenterol. — 1995. — Vol. 208. — P. 81–89.
12. Long-term results of endoscopic management of pancreas divisum with recurrent acute pancreatitis / L. Heyries, M. Barthet, C. Delvasto [et al.] // Gastrointest. Endosc. — 2002. — Vol. 55. — P. 376–381.
13. Pancreas divisum: is it a normal anatomic variant? / C. Sugawa, A. J. Walt, D. C. Nunez, H. Masuyama // Am. J. Surg. — 1987. — Vol. 153. — P. 62–67.
14. Pancreas divisum and pancreatitis: a coincidental association? / P. Burtin, B. Person, J. Charneau, J. Boyer // Endoscopy. — 1991. — Vol. 23. — P. 55–58.
15. Quest L. Pancreas divisum: opinio divisa / L. Quest, M. Lombard // Gut. — 2000. — Vol. 47, No 3. — P. 317–319.
16. Stern C. A historical perspective on the discovery of the accessory duct of the pancreas, the ampula “of Vater” and pancreas divisum / C. Stern // Gut. — 1986. — Vol. 27. — P. 203–212.

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The article analyzes the literature on embryology, prevalence, clinical manifestations of pancreas divisum. Special attention is paid to the diagnostics and treatment of pancreatitis in patients with pancreas divisum.

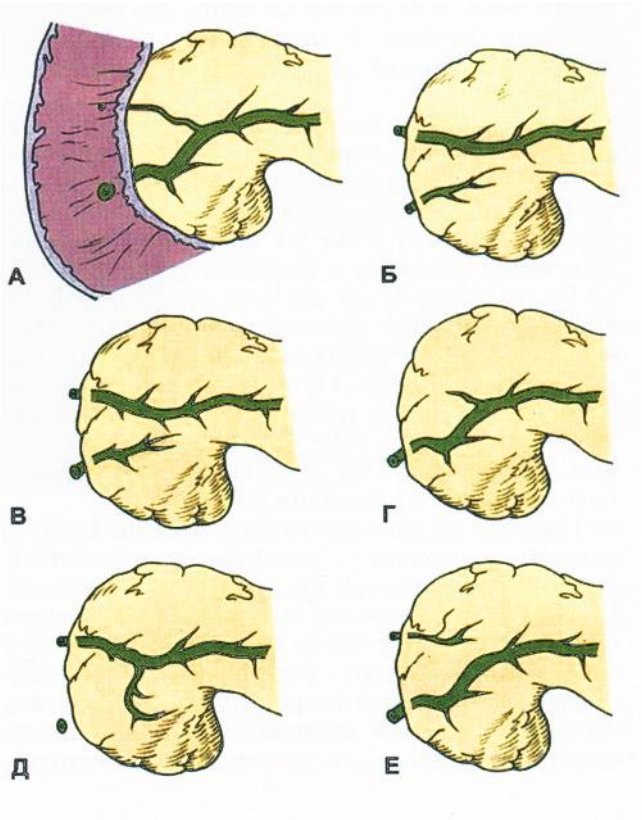

Fig. 1. Variants of pancreatic ductal anatomy (by M. B. Feldman et al., 1998 [8].)

А — the most common (normal) functioning anatomical variant with the main (Wirsung's, ventral) and extension (Santorini, dorsal) ducts, with the presence of large and small duodenal papilla;

Б — complete pancreas divisum;

В — incomplete pancreas divisum;

Г — no additional duct and small duodenal papilla;

Д — all pancreatic ductal system is drained through a small duodenal papilla;

Е — reverse pancreas divisum, when the dorsal and ventral ducts are isolated from each other, but in contrast to the complete pancreas divisum the ventral duct dominates.

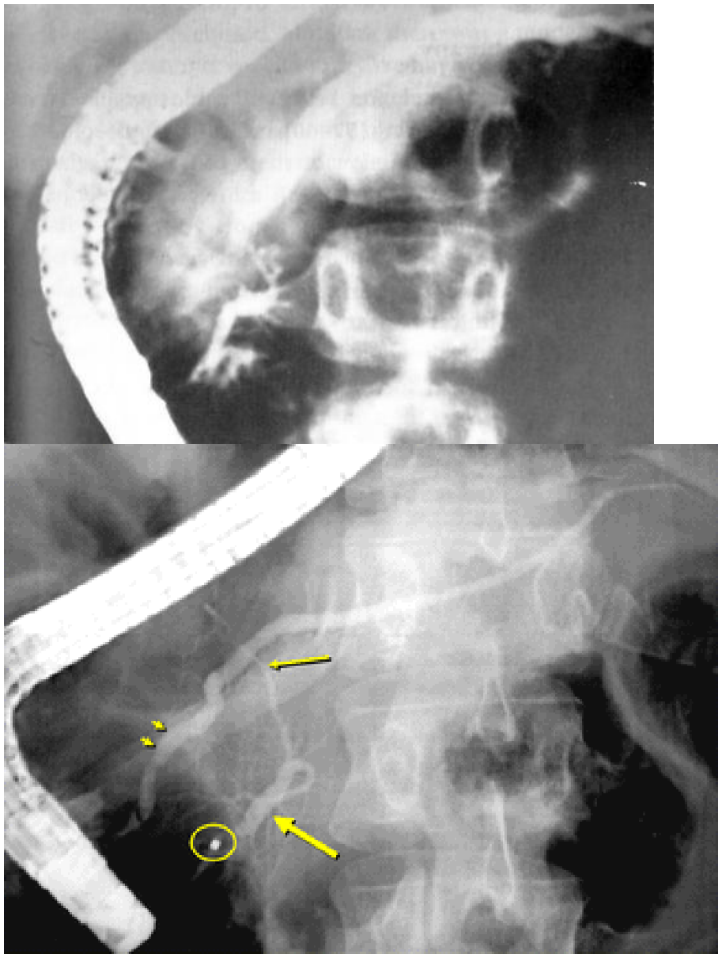

Fig. 2. ERCP:

А — complete pancreas divisum;

Б — incomplete pancreas divisum.

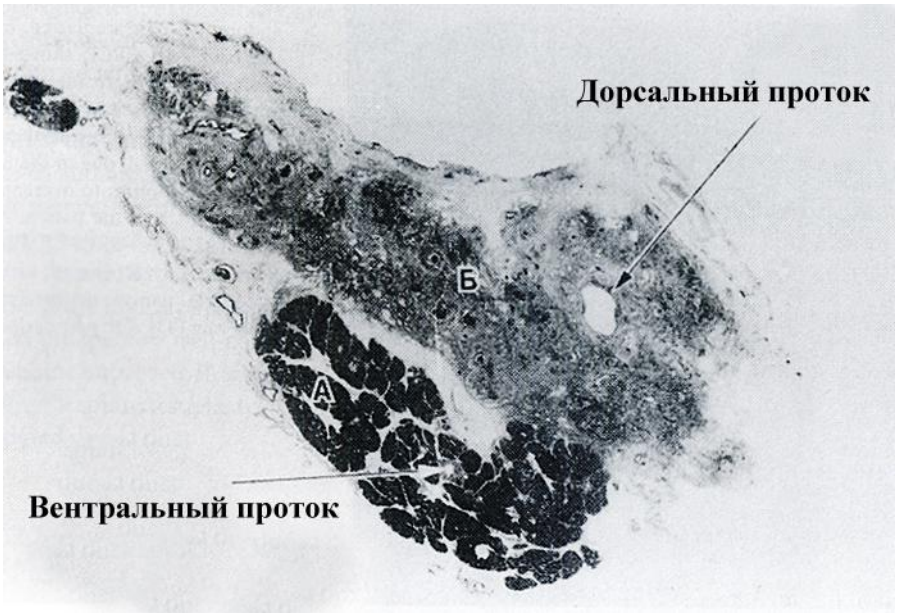

Fig. 3. Cross-section in the area of resected pancreatic head (pancreatoduodeneectomy). In the ventral part of pancreas (A) parenchyma has

normal structure, duct — normal diameter. In the dorsal part of pancreas (Б) — CP atrophy, fibrosis of the parenchyma, considerably extended duct (by A. J. Blair III et al., 1984 [1]).

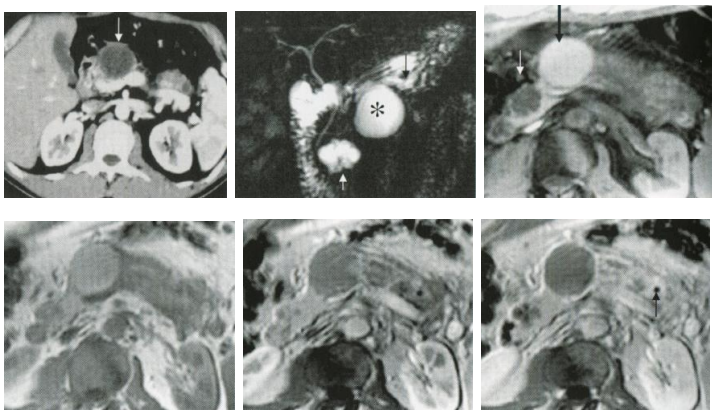

Fig. 4. CT, MRCP, dynamic MRI in a 34-years-old patient with pancreas divisum and severe CP (by J.E. Dominguez-Munoz, 2005 [4]):

А — venous phase of CT with intravenous contrast — pseudocyst in the neck of the pancreas (arrow);

Б — MRCP. The dorsal part of the small duct duodenal papilla in the body of the pancreas pseudocyst (asterisk), the expansion of the flow in the tail of the pancreas (black arrow), pseudocyst in hamate appendix (white arrow);

В — MRI. Axial T1-weighted image with fat suppression. Focal increase and a significant decrease in pancreatic parenchyma signal in the body and tail than the normal size, and the signal intensity of the parenchyma of the head of the pancreas. High signal intensity in the area of pancreatic pseudocyst in the neck (black arrow) due to the presence of detritus and protein content in it (confirmed by surgery) compared with hypointense signal in the pseudocyst in the hook-shaped appendix (white arrow);

Г–Е — dynamic MRI:

Г — axial T1-weighted image (without contrast) shows the difference in signal intensity between the head and the body, the tail of the pancreas;

Д — arterial phase of dynamic MRI with contrast. The dorsal part of the pancreas, and heterogeneous contrast less intense than the unmodified head of the pancreas;

Е — in the delayed phase dynamic MRI the dorsal part of the pancreas is contrasted more intensely, heterogeneous due to inflammation and fibrosis. Small cystic formation is detected (arrow).

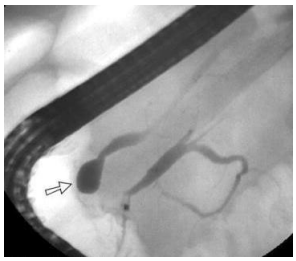

Fig. 5. ERCP. Santorinitsele (arrow).

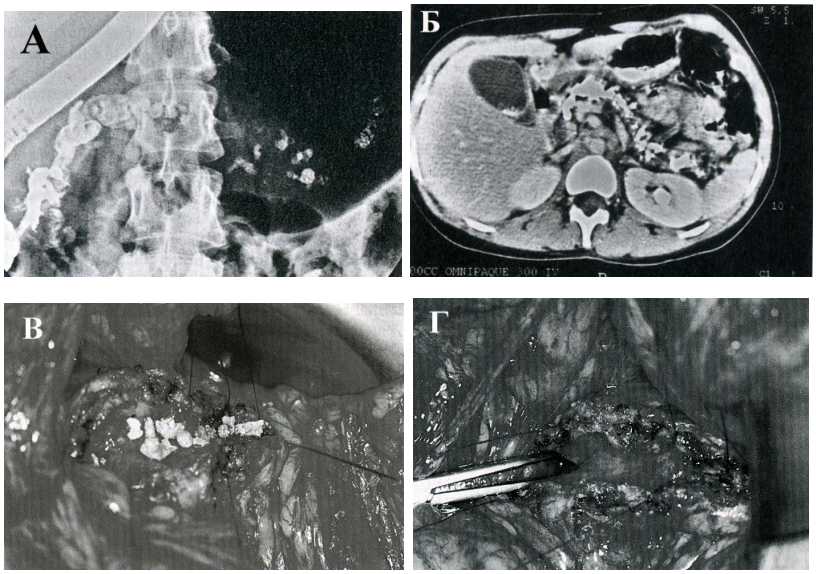

Fig. 6. 34-years-old patient with CP and pancreas divisum (by M.W. Buchler et al., 2002 [3].)

А — first patient was performed endoscopic drainage of the pancreatic duct. After the 8-fold replacement of stents (through a small papilla) for 2 years, she underwent surgical treatment, as multiple stones in pancreatic duct secretion hindered outflow of secret;

Б — for the determination of the surgical strategy CT was performed;

В, Г — after removing all the stones pancreatojejunostomy was made. The patient was discharged after 7 days in the absence of abdominal pain.