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LETTERS TO THE EDITOR

Agenesis of the dorsal pancreas

Wolfgang J Schnedl, Claudia Piswanger-Soelkner, Sandra J Wallner, Robert Krause, Rainer W Lipp

Wolfgang J Schnedl, Practice for General Internal Medicine, Hauptstrasse 5, A-8940 Liezen, Austria
Wolfgang J Schnedl, Claudia Piswanger-Soelkner, Robert Krause, Rainer W Lipp, Department of Internal Medicine, Medical University, Auenbruggerplatz 15, A-8036 Graz, Austria
Sandra J Wallner, Institute of Pathophysiology, Center for Molecular Medicine, Heinrichstrasse 31a, A-8010 Graz, Austria
Author contributions: Schnedl WJ, Lipp RW gave the conception and designed the letter; Schnedl WJ, Piswanger-Soelkner C analyzed and interpreted the data; Schnedl WJ drafted and critically revised the article for important intellectual content; Schnedl WJ, Wallner SJ, Lipp RW finally approved the article; Schnedl WJ, Piswanger-Soelkner C, Wallner SJ, Krause R, Lipp RW gave the administrative, technical, or logistic support; Wallner SJ, Krause R, Lipp RW collected and assembled the data.
Correspondence to: Wolfgang J Schnedl, MD, Associate Professor of Medicine, Practice for General Internal Medicine, Hauptstrasse 5, A-8940 Liezen,
Austria. w.schnedl@dr-schnedl.at
Telephone: +43-3612-22833 Fax: +43-3612-22833-22
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Abstract

During the last 100 years in medical literature, there are only 54 reports, including the report of Pasaoglu *et al* (*World J Gastroenterol* 2008; 14: 2915-2916), with clinical descriptions of agenesis of the dorsal pancreas in humans. Agenesis of the dorsal pancreas, a rare congenital pancreatic malformation, is associated with some other medical conditions such as hyperglycemia, abdominal pain, pancreatitis and a few other diseases. In approximately 50% of reported patients with this congenital malformation, hyperglycemia was demonstrated. Evaluation of hyperglycemia and diabetes mellitus in all patients with agenesis of the dorsal pancreas including description of fasting blood glucose, oral glucose tolerance test, glycated hemoglobin and medical treatment would be a future goal. Since autosomal dominant transmission has been suggested in single families, more family studies including imaging technologies with demonstration of the pancreatic duct system are needed for evaluation of this disease. With this letter to the editor, we aim to increase available information for the better understanding of this rare disease.

Key words: Agenesis of dorsal pancreas; Diabetes mellitus; Glucose intolerance; Abdominal pain; Pancreatitis

Peer reviewer: Sakhawat Rahman, Mr, Consultant in HPB & Minimally Invasive Surgery, Royal Free Hampstead NHS Trust, 133 King Henrys Road, Primrose Hill, London, NW3 3RD, United Kingdom

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TO THE EDITOR

We read with interest the published case report of Pasaoglu *et al* on agenesis of the dorsal pancreas[1]. Agenesis of the dorsal pancreas is a rare congenital pancreatic malformation and may be associated with some other medical conditions and diseases. During the last 100 years in medical literature, we know of 54 reports with clinical descriptions of agenesis of the dorsal pancreas in humans. We recently summarized systematically all reported patients with agenesis of the dorsal pancreas and discussed the associated medical conditions and diseases[2]. In 1911, the first description of agenesis of the dorsal pancreas was published as an autopsy finding. Now, as new imaging technologies have been developed and improved, the number of patients reported to show agenesis of the dorsal pancreas has increased rapidly over the last years. So far, the findings in autopsy and the radiological descriptions of anatomic pancreatic structures are highly variable and in most cases there is a diagnosis made without description of the pancreatic duct system[2]. In some patients, an enlarged or prominent or compensatory hypertrophy of the pancreatic head is described, whereas other descriptions include normal sized pancreatic head, as well as mildly atrophic and small head of the pancreas. We support the description of Pasaoglu *et al*[1] and confirm that the diagnosis of agenesis of the dorsal pancreas is inconclusive without demonstration of the absence of the dorsal pancreatic duct, either with endoscopic retrograde or magnetic resonance pancreatography.

Diabetes mellitus comprises a group of metabolic diseases characterized by hyperglycemia resulting from

defects in insulin secretion and/or insulin action. Criterion for diagnosis of diabetes mellitus is a fasting blood glucose > 126 mg/dL. Another diagnostic test to differentiate impaired glucose tolerance and diabetes mellitus is the oral glucose tolerance test[3]. Single familial observations of agenesis of the dorsal pancreas suggest autosomal dominant transmission. In one family with agenesis of the dorsal pancreas, a marked defect in hepatic glycogen metabolism, even in non-diabetic offspring, is demonstrated. An impaired index of the first phase insulin secretion in the diabetic and in both non-diabetic family members is described. Since the root cause of most common diabetes mellitus, type 1 and type 2 diabetes, is a decrease in-cell mass, this can be related to reduced-cell mass and might contribute to the development of glucose intolerance which ultimately leads to diabetes mellitus[4].

We suggest that more family studies including imaging technologies with demonstration of the pancreatic duct system are needed. Evaluation of hyperglycemia in all patients with agenesis of the dorsal pancreas including description of fasting blood glucose, oral glucose tolerance test, glycated hemoglobin and medical treatment would be a future goal.

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