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Review article

Disconnected Pancreatic Duct Syndrome☆

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ABSTRACT

Disconnected pancreatic duct syndrome (DPDS) is characterised by disruption of the main pancreatic duct with a loss of continuity between the pancreatic duct and the gastrointestinal tract caused by ductal necrosis after severe acute necrotising pancreatitis treated medically, by percutaneous drainage, or necrosectomy.

There are no clear epidemiological data on the real incidence of DPDS; approximately 10%–30% of patients with severe acute pancreatitis could develop DPDS. The existing literature is scarce, the terminology is confusing and therapeutic algorithms are not clearly defined. Both endoscopic management and surgical management have been described.

We have performed a systematic review of the literature on DPDS.

Síndrome del ducto pancreático desconectado

Palabras clave:
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RESUMEN

La desconexión del ducto pancreático, o síndrome del ducto pancreático desconectado (SDPD), es una entidad clínica que consiste en la existencia de una situación anatómica en la que hay ausencia de la continuidad del conducto pancreático entre el tejido pancreático viable y el tracto gastrointestinal, causada por necrosis ductal tras pancreatitis aguda grave tratada mediante necrosectomía, drenaje percutáneo o médicamente.

No hay datos epidemiológicos claros sobre la incidencia real de SDPD. Se ha postulado que entre un 10 y un 30% de los pacientes con pancreatitis aguda grave desarrollan un SDPD.

La literatura existente sobre este tema es escasa, los términos empleados son confusos y los algoritmos terapéuticos son poco claros. Las opciones terapéuticas son endoscópicas y quirúrgicas.

Hemos efectuado una revisión sistemática de la literatura sobre SDPD.

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A clinical entity, which had already been referred to in medical literature, consisting of an anatomical situation where there is no continuity of the pancreatic duct between viable pancreatic tissue and the gastrointestinal tract, caused by duct necrosis after severe acute pancreatitis and treated medically, by percutaneous drainage or by necrosectomy,[1–8] was named for the first time by Kozarek et al. as disconnection of the pancreatic duct or disconnected pancreatic duct syndrome in 1991. The isolated viable pancreatic segment continues its exocrine function, causing inflammatory intra or peripancreatic collections or an external pancreatic fistula.[5–11]

In addition to acute pancreatitis, other possible aetiologies of DPDS could be chronic pancreatitis, pancreatic trauma, pancreas divisum and other causes.[2,9,12,13] Other terms used to define this syndrome are: disconnected pancreatic tail syndrome and disconnected left pancreatic remnant.[2]

There are no clear epidemiological data on the real incidence of DPDS; approximately 10% and 30% of patients with severe acute pancreatitis could develop DPDS. The incidence of patients diagnosed with DPDS is increasing.[11] Existing literature is scarce; the terminology is confusing and therapeutic algorithms are not clearly defined.[2,4,7,9] We have performed a systematic review of the literature on DPDS.

Search

We carried out a search on Pubmed (1966–2012) for articles in English and Spanish using the terms «disconnected pancreatic duct syndrome» (17) and «disconnected pancreas» (29) and only 15 of the articles we revised were relevant. Given the few citations, the references of these articles were revised for more information on this subject which had not been included in the search terms. Finally, we revised a total of 23 articles. There are no randomised trials, clinical guides or meta-analysis of DPDS.

Definition

In acute pancreatitis, pancreatic glandular necrosis has traditionally been considered a determinant of severity. However, in some patients, necrosis of the ductal epithelium is more severe and significant than glandular necrosis.[6] DPDS occurs after a variable percentage of pancreatic parenchyma has necrosed, usually in a central location, which causes a ductal lesion that results in the distal remnant becoming disconnected from the pancreatic duct and its exocrine production being unable to drain into the gastrointestinal tract. All of this results in the formation of an intra-abdominal collection or external pancreatic fistula (EPF).[5,6,14] When there is an EPF there is usually no communication between the fistula and the proximal duct and the fistula is exclusively fed by the distal remnant.[14] In Howard's series of 27 patients with DPDS, 70% present with EPF and 30% with intra-abdominal collection.[5]

DPDS usually occurs after surgical necrosectomy due to acute pancreatitis or walled-off pancreatic necrosis (WOPN) (50%–75% of patients with DPDS).[2] 15%–25% of patients who have undergone necrosectomy present EPF.[14,15] If these data are real, the question is why do we not find more patients with DPDS,[2,9] since it can occur in patients treated with percutaneous drainage as well as in operated patients. It is likely that paucisymptomatic or wrongly diagnosed patients are the reason for the low number of patients diagnosed with DPDS.[2] The existence of unoperated cases indicates that the cause of DPDS is the ductal damage caused by pancreatitis per se and not surgical or percutaneous intervention.

Diagnosis

Correctly diagnosing DPDS is essential, as its treatment is different from that used in other post-pancreatitis intra or peripancreatic collections, pseudocysts, for example or post-necrosectomy EPF necrosectomies caused by ductal obstruction which can be resolved with a transpapillary prosthesis placed using ERCP,[7,15] and from so-called partial disruptions of the duct, which should not be considered DPDS as they are not really ductal disconnections.[13] This difficulty in diagnosis makes it enormously complicated to reach conclusions when results are compared, as it is likely that wrongly diagnosed patients have been included in the series.[8]

The existence on computerised axial tomography (CT) of a thin and small bridge of viable pancreatic tissue, compressed on the lower or posterior side, can suggest the possibility of a misdiagnosis of DPDS as this is showing us a partial disruption.[5] Fluid collections which compress the gland usually displace the duct that enters them at an oblique, not a straight, angle to the collection wall, as occurs in DPDS.[6]

Methods used for diagnosing DPDS are: CT, nuclear magnetic resonance (MRI) and endoscopic retrograde cholangio-pancreatography (ERCP).[2,6,16] It has been suggested that ERCP be replaced with nuclear magnetic cholangio resonance (MRC) with secretin stimulation, but the former is more sensitive in demonstrating ductal leakage, although it is more invasive.[5,6,9,10,13,14,16] Fistulography can be useful in some cases where there is EPF to differentiate between a terminal and lateral fistula.[5]

The traditional diagnostic criteria for DPDS are: discontinuity of the main pancreatic duct with evidence of viable distal pancreatic tissue and presence of a persistent fluid collection in the imaging methods, or discontinuity of the main pancreatic duct on ERCP and the impossibility of accessing or cannulating the distal duct.[4,7–9,11,14] A priori and traditionally, when the endoscopist was unable to cross the disconnected area with a guide or drain it was evidence that the duct was completely disconnected and not merely disrupted, although nowadays technical sophistication sometimes makes it possible for the disconnected distal remnant to be cannulated.[8,9,11]

More specific criteria have been proposed so that we have a DPDS if:

- The CT shows necrosis or a collection in the neck or body of the pancreas of at least 2 cm of pancreas and viable distal pancreatic tissue from the area of necrosis, or a pancreatic duct entering the collection at an angle of 90°.[2,6,14]
- Extravasation of contrast material injected into the pancreatic duct in the pancreatography obtained by ERCP,