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Review Article

Surgery Section

Disconnected Pancreatic Duct Syndrome: A Narrative Review of Pathophysiology, Classification, Emerging Diagnosis and Treatment Approaches

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ABSTRACT

Disconnected Pancreatic Duct Syndrome (DPDS) is a complicated outcome of pancreatic injury which is characterised by persistent secretory leakage from an isolated upstream pancreatic segment. Clinically, DPDS is associated with recurrent or non resolving Peripancreatic Fluid Collections (PFC), external pancreatic fistulae, chronic abdominal pain and metabolic derangements, which in most cases result in exocrine and endocrine dysfunction. Different radiological, endoscopic and anatomical-based classification systems have been designed to guide clinical decision-making. Diagnosis also has been progressing to a large extent due to the increasing utility of secretin-enhanced Magnetic Resonance Cholangiopancreatography (MRCP), Endoscopic Retrograde Cholangiopancreatography (ERCP), 3D-MRCP, peroral pancreatoscopy and Endoscopic Ultrasound (EUS)-assisted interventions, which enhance the detection of complete ductal disconnections. Management is based on the anatomy of the duct, endoscopic transmural drainage is preferable in cases of partial disruptions, and complete ductal disconnection often requires surgery, such as distal pancreatectomy or pancreaticojejunostomy. The current advances, which include Lumen-Apposing Metal Stents (LAMS), EUS-guided Pancreatic Duct Interventions (EUS-PDI), hybrid endoscopic-percutaneous techniques, and bioengineered anastomotic methods, are changing therapeutic practices to minimally invasive organ-preserving solutions. Early identification and multidisciplinary treatment should also be utilised to decrease morbidity and avoid recurrence of DPDS. Further research on the functional imaging and ductal reconstructive technologies holds promise to advance the long-term outcomes in DPDS. The article critically reviews clinical manifestation of the DPDS, its evolution of classification, diagnostic advancements, and modern management strategies in order to maximise patient outcome.

Keywords: Endoscopic ultrasound, Magnetic resonance cholangiopancreatography, Pancreatic fistula, Pancreatic neoplasms, Stents

INTRODUCTION

The DPDS is a condition which frequently arises after Acute Necrotising Pancreatitis (ANP), in which a segment of the main pancreatic duct becomes necrotic and separates from the viable upstream pancreatic tissue, typically in the body or tail of the gland [1,2]. This prevents pancreatic secretions from flowing into duodenum, leading to persistent fluid collections, external fistulae, chronic pancreatitis of the isolated segment, and recurrent abdominal pain too [1]. Although early descriptions of post-necrotic pancreatic fistulae and pseudocysts in the late 20th century have mentioned pancreatic-duct disruption as a discrete clinical entity, the specific notion of a so-called “disconnected” viable upstream gland separated from the gastrointestinal tract was formalised in the 1990s, and became popularised after that [3]. The term “disconnected duct/pancreas” is generally credited to Kozarek (with Traverso) in 1996, and later reports and review articles made use of the term DPDS [3]. The cut-off between destroyed and viable parenchyma causes continued enzyme secretion into a blind-ending ductal segment, often resulting in pseudocysts or Walled-Off Necrosis (WON) [4]. A study involving 647 patients with necrotising pancreatitis found that 299 (46%), with 68% of those needing surgery; outcomes showed a durable 90% success rate after proper operative strategies [5,6]. Complications include recurrent PFCs, external pancreatic fistulas, new-onset diabetes mellitus, exocrine and endocrine insufficiency, and chronic abdominal pain. Early recognition is crucial due to its potential for significant morbidity [2].

Clinical Features of Disconnected Pancreatic Duct Syndrome (DPDS)

The DPDS arises following ANP, though it can be also found in chronic pancreatitis, abdominal trauma, or surgery [7]. Patients with recurrent or persistent PFCs, which manifests from weeks to several months after the initial presentation [1,8]. Patient can also develop persistent external pancreatic fistulae, where pancreatic secretions continuously drain through a tract or catheter rather than into the gastrointestinal tract [9,10]. Recurrent or chronic abdominal pain can be generalised as a result of widespread inflammation, predominantly located in the epigastric region, radiating to the back along the nerve routes of the pancreas, and referred in other distant parts such as the chest or the shoulder, is typical with episodes of recurrent pancreatitis [9,11]. Other clinical features of DPDS are pleural effusions with high amylase content, persistent ascites, and rarely, sepsis and haemorrhagic complications due to enzymatic autodigestion [12].

Physical examination of patient shows abdominal tenderness in epigastric region [12]. Complications of DPDS include pseudoaneurysm formation with haemorrhage, resulting from enzymatic erosion of peripancreatic vessels [7,12]. Prolonged secretory loss from disconnected pancreatic tissue causes further metabolic and nutritional derangements, including protein-electrolyte depletion and hypoalbuminaemia [1,2]. Over time, the isolated pancreatic remnant may undergo atrophy or chronic pancreatitis, which in about 16% of cases leads to new-onset diabetes mellitus [13]. Clinical features and presentations of DPDS are depicted in [Table/Fig-1].

Feature	Description
Aetiology	Acute Necrotising Pancreatitis (ANP) (most common), chronic pancreatitis, abdominal trauma, or surgery
Peripancreatic Fluid Collections (PFC)	Recurrent or persistent; manifesting weeks to several months post initial event
Persistent external pancreatic fistula	Continuous drainage of pancreatic secretions via tract or catheter
Abdominal pain	Chronic or recurrent epigastric pain, radiating to the back; associated with recurrent pancreatitis
Pleural effusions	May contain high amylase levels
Ascites	Persistent fluid accumulation in the abdomen
Sepsis/haemorrhage	Rare, due to enzymatic autodigestion
Pseudoaneurysm and haemorrhage	Due to enzymatic erosion of peripancreatic vessels
Metabolic and nutritional derangements	Due to prolonged secretory loss; includes protein-electrolyte depletion and hypoalbuminaemia
Pancreatic remnant changes	May progress to atrophy or chronic pancreatitis
New-onset diabetes mellitus	Occurs in ~16% cases due to endocrine insufficiency
Physical examination findings	Epigastric tenderness

[Table/Fig-1]: Clinical features and presentations of Disconnected Pancreatic Duct Syndrome (DPDS).

Evolution of Classification Systems for Diagnosis of Disconnected Pancreatic Duct Syndrome (DPDS)

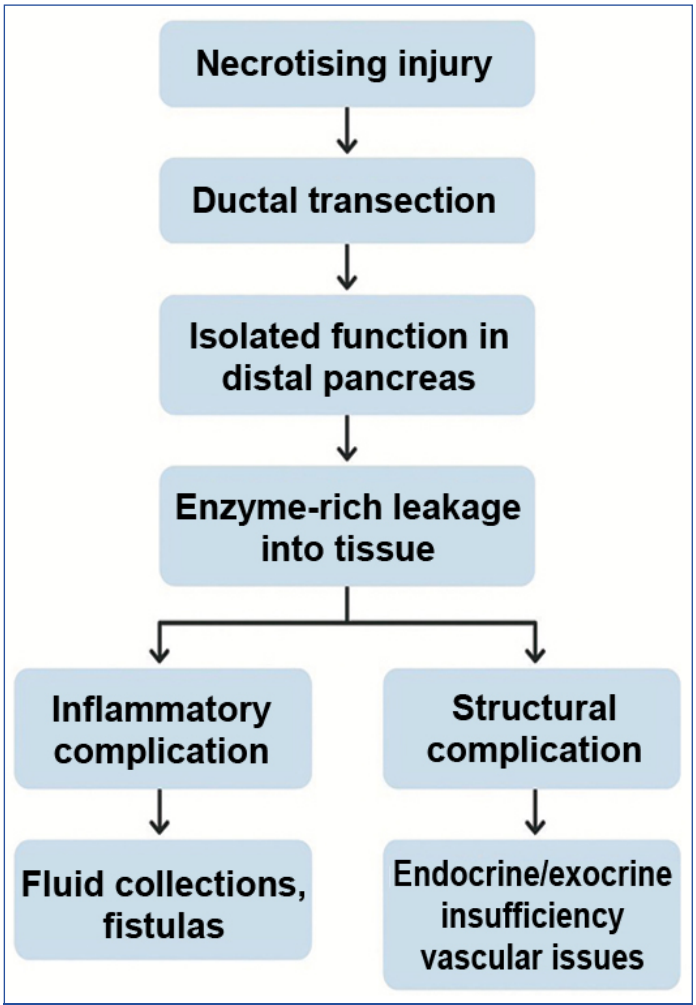
The DPDS has been classified based on anatomical, radiological, and clinical features, which is derived from ERCP, MRCP, and contrast-enhanced Computed Tomography (CT) [9]. Different classification systems for DPDS have been discussed and compared in [Table/Fig-2] [10,14,15].

Aspect	Fischer TD et al., [10]	Sandrasegaran K et al., [14]	Proença IM et al., [15]
Basis of classification	Clinical presentation following ANP or chronic pancreatitis	Anatomical, radiological, and clinical features on CT, MRCP, ERCP	Endoscopic pancreatography and therapeutic implications
Type I	DPDS with ANP, ductal disruption during acute necrosis	Complete ductal disconnection with viable upstream parenchyma → persistent fluid collection/fistula	Normal MPD, no stricture or disruption → no intervention required
Type II	Delayed pseudocyst/mature PFC formation after necrosis; underlying ductal disconnection identified	Disconnected duct with necrotic upstream parenchyma	Ductal stricture without disruption → suitable for endoscopic stenting
Type III	DPDS from chronic pancreatitis; progressive fibrosis leading to ductal obstruction	Partial ductal disconnection; segmental disruption with partial drainage	Partial disruption with contrast passage beyond disruption → bridged with transpapillary stent
Additional subtypes	-	-	Type IV: Complete disruption of MPD → IV-A: With contrast extravasation → IV-B: Abrupt ductal cut-off, no extravasation
Radiological criteria (Sandrasegaran)	Not specified	≥2 cm necrosis, viable upstream pancreas, non opacified/abruptly ending duct	Not applicable
Clinical utility	Identifies clinical scenarios to correlate with imaging and progression	Correlates radiologic patterns with ductal integrity and prognosis	Offers therapeutic decision-making guidance based on ductal anatomy
Main imaging tools	Surgical and clinical correlation	CT, MRCP, ERCP	ERCP

[Table/Fig-2]: Different classification systems for Disconnected Pancreatic Duct Syndrome (DPDS) [10,14,15].
ANP: Acute necrotising pancreatitis; CT: Computed tomography; MRCP: Magnetic resonance cholangiopancreatography; ERCP: Endoscopic retrograde cholangiopancreatography; PFC: Peripancreatic fluid collections; MPD: Main pancreatic duct

Pathogenesis of Disconnected Pancreatic Duct Syndrome (DPDS)

DPDS is caused due to severe ANP, although it can also occur after trauma and surgical injury. The essential pathogenesis involves complete disruption of the Main Pancreatic Duct (MPD) usually at the neck or body, secondary to necrosis of ductal epithelial cells within necrotic parenchyma. This disruption or transection causes isolation of viable distal segment, commonly the tail, from the remainder of the ductal system, preventing pancreatic secretions from draining into duodenum. Functionally, the disconnected distal segment continues production of exocrine secretions and is unable to drain through the ductal-anatomical pathway, these secretions leak into adjacent tissues, leading to persistent PFC, pseudocysts, external fistulas, pancreatic ascites, or even pancreaticopleural fistulae. The leakage further incite inflammation in the retroperitoneum, adding to para-pancreatic infiltrates and enzymatic erosion of surrounding tissues. An important anatomical predisposition relates with precarious blood supply at the pancreatic neck, supplied predominantly by the dorsal pancreatic artery making this region especially vulnerable to necrosis and ductal disruption. Flowchart illustrating pathogenesis of DPDS is shown in [Table/Fig-3].



[Table/Fig-3]: Flowchart illustrating pathogenesis of Disconnected Pancreatic Duct Syndrome (DPDS).

Emerging and Current Diagnostic Methods for Disconnected Pancreatic Duct Syndrome (DPDS)

The accurate diagnosis of DPDS is based upon clinical features, imaging findings, and endoscopic assessment [16]. MRCP and ERCP are considered as primary gold standard diagnostic methods for DPDS [9]. ERCP allows, direct ductal visualisation and contrast injection, which enables confirmation of a complete ductal disruption, particularly when contrast extravasates or fails to opacify the upstream duct [17,18]. Secretin-enhanced MRCP (S-MRCP) has also gained importance as a non invasive alternative, which provides functional imaging helping in identification of ductal leaks and assessing continuity by stimulating pancreatic juice flow [19,20]. Contrast-enhanced CT can help in identification of necrotic segments and PFCs but it lacks ductal resolution [16].

Peroral pancreatoscopy with usage of digital cholangioscopes is another novel diagnostic tool, allowing direct intraductal visualisation and guidewire passage across the site of ductal disruption [21]. Advanced magnetic resonance-based techniques are also useful in DPDS [12]. Delineation of the MPD is improved by S-MRCP which also facilitates functional assessment with increased pancreatic juice flow, thus helps identifying ductal disconnections [19].

The recent developments in the diagnosis of DPDS include improvement in the anatomical and functional evaluation of the pancreatic duct [7,22]. New endoscopic strategies, which includes EUS-assisted rendezvous along with EUS-guided drainage of the pancreatic ducts, enable the access to the upstream duct in cases where the conventional ERCP method fails, enabling a careful localisation of the ductal disruption [22]. Digital single-operator pancreatoscopy provides intraductal visualisation and passage of a guidewire through the disconnection site and is more accurate in facilitating diagnosis [23]. In the imaging sector, techniques such as quantitative diffusion-weighted MRI, three-dimensional MRCP, and deep-learning-reconstructed MR sequences are more suitable than the other imaging options regarding resolution and functional evaluation, which can be used to detect small ductal abnormalities and distinguish between sterile and infected collections [24-26]. The current practice shift highlights multimodal and functionally integrated imaging, where advanced methods of MR are integrated with endoscopic ones to promote the prompt and accurate diagnosis, plan treatment, and improve patient outcomes [24].

Differential Diagnosis of Disconnected Pancreatic Duct Syndrome (DPDS)

The differential diagnosis of DPDS includes few conditions which mimic its clinical presentation or imaging findings, notably pancreatic pseudocysts, WON, chronic pancreatitis with ductal strictures, Pancreatic Ductal Adenocarcinoma (PDAC), pancreatic fistulas, and incomplete duct disruption [17,27,28].

Pancreatic pseudocysts, which are caused due to ANP, shows similarities with DPDS on imaging, as both involve fluid collections [27]. However, pancreatic pseudocysts usually have intact duct and it resolves spontaneously with drainage, while DPDS involves complete ductal disconnection and persistent fluid collections due to isolated viable upstream parenchyma [27]. Another complication, which develops following ANP, is called WON, which may manifest itself as an intricate fluid accumulation with necrotic debris [28]. The distinction between WON and DPDS is based on the presence of non liquefied necrotic tissue and the absence of complete ductal transection and DPDS also has the characteristic of the disconnected upstream ductal segment on MRCP or ERCP [28].

Chronic pancreatitis with ductal strictures shows ductal irregularity or segmental obstruction, but unlike DPDS, there is no transection of duct, thus communication between ductal segments is preserved [11,29]. This is opposed to DPDS, and the ductal segments are not interrupted; the upstream parenchyma does not become isolated, which can be confirmed on imaging or endoscopic assessment [30]. Parenchymal atrophy in PDAC is usually associated with upstream ductal dilatation and ductal obstruction caused by tumours [31]. However, contrast-enhanced imaging along with cytology helps to identify malignancy, and a history of necrotising pancreatitis favours DPDS [31]. The criteria used to differentiate depend on the mass lesion on contrast-enhanced imaging, cytological confirmation of malignancy, and absence of necrotising pancreatitis history, which favours for DPDS [31].

Pancreatic fistulas, which can be both external or internal, resemble the persistent drainage as seen in DPDS [32]. Fistulas are result of surgical and procedural injury which do not necessarily indicate a transected duct with a viable distal segment [32]. In addition, in incomplete pancreatic duct disruption, there is partial continuity or intermittent flow across the disrupted site, is different to DPDS as it allows some drainage to the gastrointestinal tract, which is commonly seen on secretin-enhanced MRCP or ERCP [20,33]. Thus, differentiating DPDS from such similar conditions depends upon clinical features, imaging, and endoscopic evaluation [17]. Differential diagnosis of DPDS is described in [Table/Fig-4].

Condition	Key features	How it differs from DPDS	Diagnostic clues
Pancreatic pseudocyst	Fluid collection post-acute pancreatitis with an intact duct	Duct is intact; typically resolves with drainage	No ductal disconnection on imaging
Walled-Off Necrosis (WON)	Encapsulated necrotic collection following Acute Necrotising Pancreatitis (ANP)	May resemble DPDS radiologically, but lacks complete ductal disconnection	Imaging shows necrotic debris; not persistent like DPDS
Chronic pancreatitis with strictures	Ductal irregularity or obstruction without transection	No ductal transection; ductal continuity preserved	ERCP/ MRCP shows narrowed duct, not complete disconnection
Pancreatic Ductal Adenocarcinoma (PDAC)	Tumour causes ductal obstruction with upstream ductal dilation and parenchymal atrophy	Malignant cause; not post-necrotising event; no viable upstream isolation	Contrast imaging, cytology, and history help distinguish
Pancreatic fistula (internal/external)	Persistent pancreatic drainage due to surgical/ procedural injury	No viable upstream segment; not always associated with duct transection	Clinical history of surgery; imaging may show fistulous tract
Incomplete pancreatic duct disruption	Partial ductal continuity; some intermittent drainage	Allows partial drainage into GI tract; not a complete disconnection like DPDS	Seen on secretin-enhanced MRCP or ERCP

[Table/Fig-4]: Differential diagnosis of Disconnected Pancreatic Duct Syndrome (DPDS).

Management of Disconnected Pancreatic Duct Syndrome (DPDS)

The treatment of DPDS remains a clinical challenge and is mainly based on the patient's presentation, anatomical disruption, and associated complications [7]. According to several studies, endoscopic and surgical approaches are the mainstay of therapy [7]. Endoscopic drainage is utmost useful in pancreatic disconnections involving communication with a mature PFC or partial ductal disruption [18]. Endoscopic transmural drainage using LAMS is presented with good outcomes in similar cases

Category	Therapeutic approach	Indications/ use cases	Advantages	Limitations
Conventional endoscopic	Endoscopic transmural drainage with LAMS	Partial ductal disruption with PFC communication	Minimally invasive, reduces external fistulae and recurrence	Not suitable for complete disconnection
	Endoscopic transpapillary stenting	Partial disruption or communication with MPD	Maintains ductal flow, avoids surgery	Ineffective in complete ductal disruption
Surgical	Distal pancreatectomy	Symptomatic, resectable disconnected pancreatic segment	Removes diseased tissue, definitive solution	Surgical morbidity, loss of pancreatic tissue
	Roux-en-Y pancreaticojejunostomy	Disconnected segment with preserved function	Preserves pancreatic tissue and function	Technically challenging, risk of anastomotic leak
Emerging endoscopic	EUS-guided drainage	Non communicating collections or inaccessible via conventional drainage	Minimally invasive, alternative to surgery	Limited role in complete duct disruption
	EUS-guided Pancreatic Duct Intervention (EUS-PDI)	Failed ERCP or inaccessible MPD	Minimally invasive ductal access	Requires expertise, limited long-term data
	EUS-guided gastro-pancreatic ductal anastomosis	Experimental technique for ductal reconnection	Restores ductal continuity	Experimental, limited clinical application
Novel/ Hybrid approaches	Fully Covered Self-Expandable Metal Stents (FCSEMS)	Bridging of disconnected ductal segments	Long patency, minimally invasive	Limited to select cases, needs further validation
	Combined endoscopic-percutaneous approaches	External pancreatic fistulas or inaccessible collections	Facilitates internal drainage, reduces external catheter burden	Requires coordination between specialties
	Magnetic compression anastomosis, bioengineered ductal reconstruction	Re-establishment of ductal continuity in experimental settings	Potential for future non surgical reconnection	Still in experimental stages
Long-term management focus	Management of pancreatic fistulas, exocrine insufficiency, recurrent collections	Post-treatment complications	Symptom control, quality of life improvement	Requires ongoing monitoring and multidisciplinary care

[Table/Fig-5]: Therapeutic approaches for Disconnected Pancreatic Duct Syndrome (DPDS).

by facilitating internal drainage and reducing external fistulae or recurrence rates [34,35].

However, in cases of complete ductal disconnection, endoscopic treatment fails due to lack of continuity in gastrointestinal tract, necessitating further surgical intervention [36]. Surgical options in such cases are distal pancreatectomy when the disconnected segment is symptomatic and resectable, or Roux-en-Y pancreaticojejunostomy when pancreatic tissue is meant to be preserved [36,37]. Long-term management in such cases are focusd on controlling pancreatic fistulae, exocrine insufficiency, and recurrent fluid collections [38]. In recent few years, EUS-guided drainage and endoscopic transpapillary stenting have gained attention, especially in selected cases, although their role in complete DPDS remains limited [39,40].

Emerging and novel approaches focus mainly on minimally invasive, endoscopic innovations and tailored hybrid techniques which bridge the gap between traditional surgery and endoscopy [19]. One such promising technique is EUS-PDI, including transduodenal pancreaticogastrostomy, helps managing DPDS [41,42]. EUS-guided gastro-pancreatic ductal anastomosis is also been explored as an experimental method for re-establishment of ductal continuity and drainage [43]. One of the studies emphasised that EUS-guided Pancreatic Duct Drainage (EUS-PDD) and Peripancreatic Fluid Drainage (EUS-PFD) is increasingly utilised, particularly when the known cases are the results of ANP [44]. These procedures promote internal drainage and are linked to good results amongst few patients [44].

Another innovating technique is Fully Covered Self-Expandable Metal Stents (FCSEMS) which are specifically designed for the pancreatic duct, to provide longer patency and allow bridging of disconnected segments in selected cases [30,45]. Furthermore, combined endoscopic-percutaneous approaches are useful in DPDS, in which endoscopy and interventional radiology are used together to facilitate internalisation of external pancreatic fistulas or drain inaccessible collections [46,47]. Lastly, endoscopic pancreatic ductal reconstruction with usage of magnetic compression anastomosis and other bioengineering strategies is an area of active research, though still experimental [16,48]. All these approaches aim to reduce the need for surgery with preservation of pancreatic function, for improving quality of life of patient, thus a significant shift towards less invasive, organ-preserving treatments in DPDS [7]. MRCP has been useful in detecting DPDS and in the process of formulating the right management approach [48,49]. One of the studies has suggested that MRCP is capable of detecting DPDS in the presence of WON and aids in the planning of the management to avoid recurrence of PFCs [48,49]. Combination of endoscopic and percutaneous has proved useful in the management of complicated cases of DPDS [49,50]. This is an all-inclusive drainage and control of PFCs, particularly when the endoscopic approach is inappropriate [50]. Therapeutic approaches for DPDS are depicted in [Table/Fig-5].

Emerging Approaches in Disconnected Pancreatic Duct Syndrome (DPDS)

The future of DPDS study has the potential to provide a greater level of accuracy on diagnosis, treatment, and patient outcome. The new researches must incorporate new imaging modalities that are advanced e.g., S-MRCP to enhance the definition of ductal anatomy and functionality, and thus, aid in the more accurate detection of ductal disruptions [17,20]. There has also been the introduction of new endoscopic interventions such as two-scope-guided tractogastrostomy to offer minimally invasive treatment of the external pancreatic fistulas related to DPDS [51]. The consequences of interventions such as LAMS and double pigtail stents are still in their long-term research to identify optimal stent removal timing and to monitor the possible occurrences of complications such as chronic pancreatitis or new-onset diabetes [52]. Moreover, the interdisciplinary management integrating the endoscopic, radiological, and surgical approaches are under investigation to customise personalised treatment procedures that would preserve pancreatic functions and improve the quality of life of the patients [53].

CONCLUSION(S)

The DPDS is a complex result of pancreatic injury, mostly after necrotising pancreatitis, which is characterised by ductal disruption and persistent fluid collections. Proper diagnosis is done using MRCP, ERCP, and evolving modalities like EUS and pancreatoscopy is vital. Management strategies depend upon ductal integrity and clinical features, which ranges from endoscopic drainage and

stenting and further surgical interventions. Emerging minimally invasive techniques are also crucial for DPDS, which are EUS-guided interventions and hybrid approaches, offer promise for better outcomes. Early recognition of condition along with individualised treatment is essential for prevention of any complications and preserving pancreatic function, reflecting a shift toward organ-preserving therapies.

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