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### Case Report

# Type III Choledochal Cyst Presenting as Recurrent Pancreatitis in Young Child, Magnetic Resonance Findings Suggesting Etiology Prior to Endoscopy

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### Abstract

A type III choledochal cyst (choledochoceles) is a rare lesion affecting the distal common bile duct and pancreato-biliary junction, which is consistently described in the literature as a cystic lesion involving the duodenal wall at the ampulla and involving these structures. The exact etiology is unknown with some hypothesizing that this is a congenital anomaly while others supporting that a choledochoceles is acquired. This case presents a 7 year-old male child with recurrent pancreatitis and a choledochoceles having a varied appearance over time on magnetic resonance cholangiopancreatography (MRCP). Patient was amply imaged, a choledochoceles due to an ampullary obstruction was suspected and he underwent a therapeutic endoscopic intervention (ERCP) with resulting complete resolution of his symptoms. This case supports the hypothesis that these lesions are actually acquired and related to obstruction within the distal ampulla. Lastly, this case highlights the diagnostic utility of MRCP in the pediatric patient and ERCP as a therapeutic option for these lesions.

## ABBREVIATIONS

CBD: Common Bile Duct; PD: Pancreatic Duct; MRCP: Magnetic Resonance Cholangiopancreatography; ERCP: Endoscopic Retrograde Cholangiopancreatography.

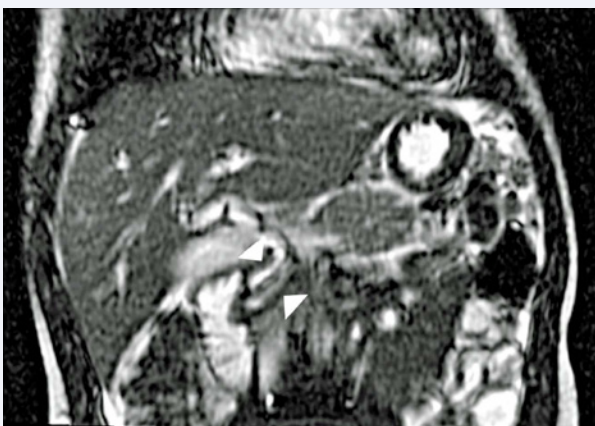
## INTRODUCTION

A type III choledochal cyst (choledochocele) is an unusual abnormality of the biliary system whereby the common bile duct (CBD) directly communicates with a cystic mass at the ampulla of Vater; the rarest of all choledochal cysts [1,2]. Due to its rarity and despite reports within the literature spanning over 70 years [3], the exact pathogenesis of this lesion remains elusive. Some hypothesize that a choledochocele represents a congenital anomaly [2]. Others proclaim that this represents an acquired lesion from obstructive ballooning of the common bile duct [4]. Irrespective of etiology, the accepted dogma is that a choledochocele, like other types of choledochal cysts, is a "fixed" lesion. The following report demonstrates that this is not always the case and supports the hypothesis that at least some choledochoceles are acquired.

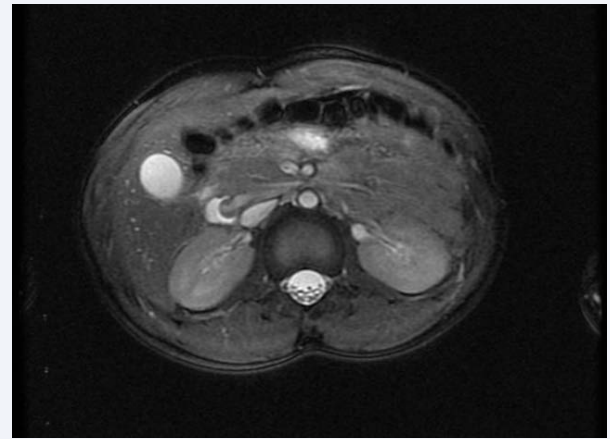
## CASE PRESENTATION

A 7-year-old male initially presented to an outside institution complaining of mesogastric, persistent abdominal pain. An abdominal ultrasound showed mildly diffuse dilatation of the CBD, a mildly hypoechoic pancreas, a prominent pancreatic duct (PD), and a normal gallbladder without stones. Elevated amylase and lipase levels yielded the diagnosis of pancreatitis.

Given the unexplained CBD and PD enlargement, a magnetic resonance cholangiopancreatography (MRCP) examination was performed, demonstrating a well-defined nodular soft tissue attenuating mass within the duodenal lumen at the level of the major papilla. The common channel appeared to course through or directly behind this lesion. The CBD and PD were diffusely dilated. The pancreas was diffusely enlarged and edematous consistent with pancreatitis. On more cephalad images, within the posteromedial liver, there were multifocal areas of T2 hyperintensity in a faint wedge-shaped configuration consistent with acute cholangitis. Given these findings, endoscopic



**Figure 1** Coronal MRI imaging showing dilated common bile duct and pancreatic duct (arrows) with cystic changes in common channel as it enters duodenal wall.



**Figure 2** Transverse MRI showing protuberant ampulla with soft tissue thickening and dilated/cystic changes of common channel as it courses through duodenal wall.



**Figure 3** (A) – Endoscopic image of bulbous ampulla with pinpoint opening prior to cannulation. (B) – Ampulla post needle-knife sphincterotomy with guidewire placed in pancreatic duct to mark opening and void accidental injury to the pancreatic sphincter.

retrograde cholangiopancreatography (ERCP) was attempted but failed at outside institution.

Repeat MRCP demonstrated increased changes of chronic cholangitis involving the posterior right lobe of the liver. There was also increasing biliary dilatation with a markedly tortuous CBD. Instead of the previously demonstrated nodular soft tissue mass, there was a cystic structure protruding from the papilla into the duodenal lumen. The patient was sent to our institution for urgent evaluation and started on an oral course of antibiotics (ciprofloxacin) for cholangitis.

After initial evaluation at our institution, MRCP was performed which showed improvement of cholangitis prior to ERCP. There was marked improvement in the CBD dilatation. Furthermore, the cystic lesion at the ampulla had resolved to a remnant of irregular soft tissue thickening at the ampulla without a discrete mass.

Given the dynamic appearance of the ampulla, which appeared to have a cystic lesion when the CBD and PD were most dilated and appeared as only soft tissue when decompressed, it was hypothesized that this likely represented changes of severe ampullary stenosis with an associated choledochoceles. The ERCP confirmed this hypothesis. On endoscopic examination, a "pin-point" opening was noted on a protuberant ampulla which did not even allow a 4F sphincterotome (Minitome, Wilson-Cook, USA) passage so sphincterotomy was performed using a needle knife after a 021 (0.51mm) guidewire was placed through the ampullary opening to mark the pancreatic sphincter and allow for a more targeted procedure. The pancreatic duct was noted to come off this dilated or "cystic" common channel and bile duct dilation also noted with no stones or sludge in the ducts. After the procedure, there was resolution of the marked dilatation. The patient was discharged to home and has had no further symptoms or evidence of pancreatitis since the procedure (18 months follow-up).

## DISCUSSION

A choledochocoele, is a rare anomaly of the distal bile duct representing 1.4-5% of all choledochal cysts [2]. Described over seventy years ago, the underlying etiology is uncertain with one competing theory proclaiming that a choledochocoele is the result of congenital, sphincter of Oddi dysfunction [2]. A competing theory championed by Vitellas, et al., theorizes that a choledochocoele is actually the result of ballooning of the intramural portion of the CBD secondary to inflammation and obstruction of the ampulla [4]. Whether a choledochocoele represents a congenital anomaly or an acquired lesion remains unresolved, likely due to the relative rarity of reports and studies on this entity within the literature. This case report provides further information on the imaging characteristics of this disorder as well as supports the hypothesis that a choledochocoele is an acquired lesion rather than a fixed, developmental anomaly.

Patients may initially present with abdominal pain, jaundice, gallstone disease, biliary colic, or with pancreatitis [2]. Laboratory evaluation consists of a liver panel, pancreatic enzyme analysis, and often an ultrasound. Though not usually diagnostic, ultrasound may show secondary signs, namely CBD and PD dilatation, as seen with our patient. The combination of symptomatology and

ultrasound findings suggestive of distal biliary obstruction often leads to biliary tract imaging, predominately ERCP. Only one case report has been published to date demonstrating MRCP's ability to demonstrate a choledochocoele.

De Backer, et al., reported MRCP findings in a 22-year-old female with a 10-day history of abdominal pain. Abdominal ultrasound and CT failed to achieve diagnosis. Due to elevated bilirubin levels and liver transaminases, an MRI and MRCP were performed demonstrating cystic dilatation of the CBD protruding into the duodenum, diagnostic of a choledochocoele. As our case demonstrates, choledochoceles can be readily diagnosed by MRCP even in a very small child.

The choledochocoele was initially described by Wheeler in 1940, resembling an ureterocele [5]. Todani, in a modification of an earlier congenital choledochal cyst classification [6], designated the choledochocoele as a type-III choledochal cyst, thus solidifying the idea that this is a congenital anomaly. A handful of papers within the literature question this theory.

In a very cogent argument, Elton, et al., support that these lesions represent an acquired cystic mass, differentiating from other congenital choledochal cysts in terms of origin and malignant transformation risk [5].

The authors firmly conclude that the underlying etiology lies in obstruction within the very distal tip of the common channel from either papillary stenosis or sphincter of Oddi dysfunction. Furthermore, after a thorough literature review, the authors only found three reported cases of malignant transformation within a choledochocoele as opposed to a 28% risk of carcinoma arising in other choledochal cysts. The authors conclude that prophylactic cyst resection is inappropriate and that the lesion can be unroofed through a therapeutic endoscopic procedure (ERCP).

Since the patient's clinical course was complicated by cholangitis and the need for transfer here for further care, we had the distinct advantage of being able to see the progression of this choledochal cyst over time with serial imaging. As the degree of obstruction worsened evidenced by increased CBD and PD dilatation along with increased areas of cholangitis, MRCP captured a more protuberant, cystic lesion, consistent with a choledochocoele. Once the obstruction and inflammation resolved, the lesion nearly disappeared completely with only soft tissue thickening noted along the duodenal wall at the papilla, most likely to representing the decompressed walls of the choledochocoele. Not only does this support the acquired theory of pathogenesis, this case, for the first time in the literature, shows the varied appearance over time of these lesions, contradicting the commonly supported dogma that these lesions are fixed cystic abnormalities.

In conclusion, this case demonstrates that at least a subset of choledochoceles is acquired. Depending on the degree of stenosis and obstruction at the ampulla, timing for ERCP needs to be measured and only trained and experienced providers should undertake it. Lastly, this case demonstrates the utility of MRCP in the detection of choledochoceles in very small children, with this case being the youngest reported to date.

## REFERENCES

1. Cory DA, Don S, West KW. CT cholangiography of a choledochocele. *Pediatr Radiol.* 1990; 21: 73-74.
2. De Backer AI, Van den Abbeele K, De Schepper AM, Van Baarle A. Choledochocele: diagnosis by magnetic resonance imaging. *Abdom Imaging.* 2000; 25: 508-510.
3. Scholz FJ, Carrera GF, Larsen CR. The choledochocele: correlation of radiological, clinical and pathological findings. *Radiology.* 1976; 118: 25-28.
4. Vitellas KM, Keogan MT, Spritzer CE, Nelson RC. MR Cholangiopancreatography of bile and pancreatic duct abnormalities with emphasis on the single-shot fast spin-echo technique. *Radiographics.* 2000; 20: 939-957.
5. Elton E, Hanson BL, Biber BP, Howell DA. Dilated common channel syndrome: endoscopic diagnosis, treatment, and relationship to choledochocele formation. *Gastrointest Endosc.* 1998; 47: 471-478.
6. Alonso-Lej F, Rever WB Jr, Pessagno DJ. Congenital choledochal cyst, with a report of 2, and an analysis of 94, cases. *Int Abstr Surg.* 1959; 108: 1-30.

### Cite this article

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