

## Imaging of the Choledocal Cystic Dilatation in Infants and Children

M. Ouha<sup>1\*</sup>, K. Zitouni<sup>1</sup>, B. Zouita<sup>1</sup>, D. Basraoui H. Jalal<sup>1</sup>

<sup>1</sup>Department of Pediatric Radiology, Mother and Child Hospital, University Hospital Mohammed VI, Marrakech, Morocco

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\*Corresponding author: M. Ouha

### Abstract

### Original Research Article

**Purpose:** The choledochal cyst is a rare surgical entity due to an anomaly of the bilio-pancreatic junction, which manifests itself mainly during childhood and often revealed by a complication. Imaging plays an important role in the positive diagnosis, allows to analyze the malformation of the main bile duct and to classify it according to the classification of Todani. The aim of our work is to show the contribution of imaging in the diagnosis of this disease in Pediatric Population and its complications. **Methods and Materials:** This is a retrospective descriptive study of 19 case reports carried out between 2013 and 2020 in the Pediatric radiology and Child Surgery departments at the mother and child hospital at the University Hospital MOHAMED VI. **Results:** The average age of our patients was 5 years, predominantly female. Abdominal pain was the most common clinical sign. All patients were examined by ultrasound, 11 cases had abdominal CT with injection of contrast agent, while a bili-MRI was performed in 7 patients. These examinations allowed us to objectify a cystic dilatation of the bile duct classified type I according to Todani in 80% of cases, and a type IVa in 20% of patients. Treatment consisted of a resection of the cyst with hepaticojejunal anastomosis associated with cholecystectomy, the postoperative outcome was good. **Conclusion:** Medical imaging plays an essential role in the diagnosis of cystic dilatations of the bile duct. Bili-MRI is the exam of choice that allows accurate biliary mapping, avoiding the use of invasive opacifications.

**Keywords:** The choledochal cyst, Child Surgery, Medical imaging.

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## INTRODUCTION

Cystic dilations of the bile ducts are congenital malformations, described for the first time in 1723 by VATER, characterized by a cystic dilatation which connects with biliary tracts. This disease includes several types, depending on the localization, the shape and distribution of the cyst. This pathology manifests itself mainly during childhood and often revealed by a complication. The diagnosis is essentially based on ultrasound and Magnetic Resonance Cholangiopancreatography.

By reporting a series of 19 children, the aim of this work is to show the contribution of imaging in the diagnosis of this condition and its complications in children and infants as well.

## MATERIAL AND METHODS

We conducted a retrospective descriptive study of 19 case reports carried out between 2013 and 2020 in the Pediatric radiology and Child Surgery departments at the mother and child hospital at the University Hospital MOHAMED VI. The average age of our

patients was 5 years, mostly female (n = 12). Abdominal pain was the most common clinical sign. All patients were scanned by ultrasound, 11 cases underwent abdominal CT with contrast agent injection, while a Magnetic Resonance Cholangiopancreatography (MRCP) was performed in 7 patients. These examinations allowed us to objectify a cystic dilatation of the bile duct classified type I according to Todani in 80% of cases, and a type IVa in 20% of the patients. Treatment consisted of resection of the cyst with hepato-jejunal anastomosis associated with cholecystectomy with good postoperative outcome. The anathomopathological study of the cysts showed no evidence of malignancy in all children, an aspect of hepatic cirrhosis was observed in one case.

## DISCUSSION

Bile duct cyst is a rare condition. Its incidence is 1/100,000 to 1/200,000 births, with a clear predominance in the female sex. Usually this pathology is diagnosed before the age of 10 years. Nevertheless, 20-50% of cases are discovered in adulthood in Western countries [1].

From an etiopathogenic point of view, it results from an abnormality of the bilio-pancreatic junction where the Wirsung unites with the bile duct upstream of the sphincter thus allowing the reflux of pancreatic secretions into the bile duct, which leads to an incomplete and intermittent obstruction of the low bile duct, explaining the progressive and late character of clinical signs [2].

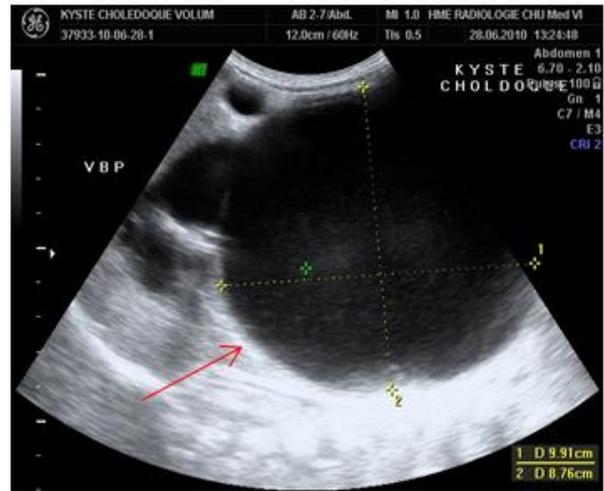
The clinical presentation is characterized by the classic triad: abdominal pain, jaundice, and a mass of the right hypochondrium. This symptomatology is observed in only one third of cases. Choledocal cysts can be complicated by pancreatitis, cholangitis or peritonitis by spontaneous rupture, but it can also remain asymptomatic, so the discovery is fortuitous during a medical imaging examination [3].

From the anatomical point of view, the first description of ductal dilatation of the biliary tree was made by Vater in 1723. In 1959, Alonso- Lej proposed the first classification of these anomalies, currently; however, Todani's classification is the most used, which describe five types according to the seat, extent and type of dilatation of the bile ducts [4].

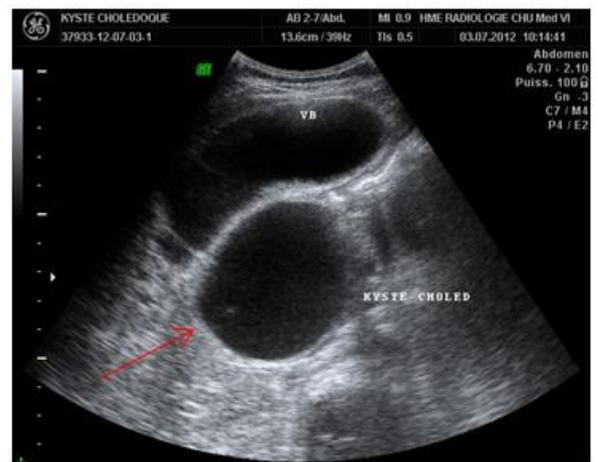
The diagnosis of cystic dilatation of biliary ducts is currently based on non-invasive imaging methods: ultrasound, CT and MR cholangiography.

Diagnosis can be done prenatally by ultrasound or MRI which allows the detection of the formation of the hepatic hilum suggestive of a biliary atresia or a choledochal cyst. Thus a small cyst with anechoic content is highly suggestive of biliary atresia, whereas a cystic formation that increases in size is in favour of choledochal cyst.

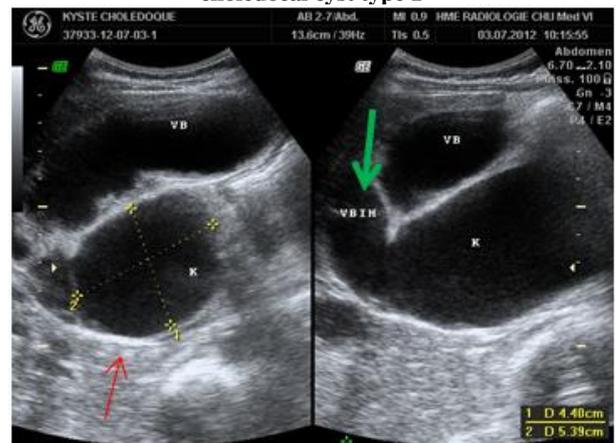
Postnatally, ultrasound is the first-line examination with a specificity that varies between 71% and 97% [5]. The choledocal cyst appears as a cystic formation well- defined thin-walled (fig.1, 2, 3) sometimes containing an echogenic material corresponding to the Biliary Sludge (fig.4). It sits in contact with the portal vein and the gallbladder, communicating with the bile ducts which can be dilated [5].



**Fig-1: Hepato-biliary ultrasound showing a cystic formation in the Hepatic hilum continuing with the main bile duct: choledocal cyst type 1**



**Fig-2: Abdominal ultrasound showing a cystic formation between the head of the pancreas and the gallbladder: choledocal cyst type 1**



**Fig-3: Hepato-biliary ultrasound showing the presence of cystic dilatation of the main biliary tract (red arrow) associated with dilation of the intrahepatic biliary ducts (green arrow)**



**Fig-4: Hepatobiliary ultrasonography demonstrating a cystic formation of the hepatic hilum containing Sludge (red arrow) and compressing the Gallbladder (green arrow)**

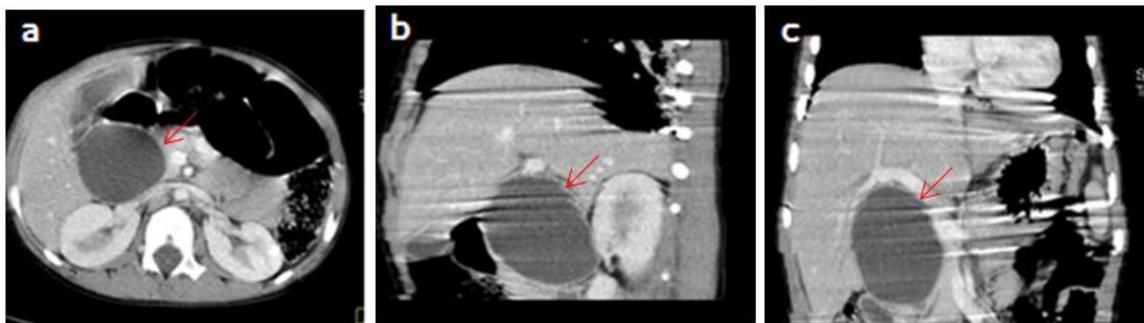
Ultrasound also allows the study of the state of the intrahepatic bile ducts and possibly can detect signs suggestive of a complication.

In ultrasonography, the differential diagnosis arises in the neonatal period with biliary atresia (cystic form), and in children and the young adult with a dilation of the bile ducts upstream of a choledocholithiasis or a cystic lesion of the head of the pancreas.

CT scan allows a good characterization of the cyst which appears as a mass of the hepatic hilum, in

contact with the head of the pancreas, iso or hypo dense thin wall, unmodified after the injection of the contrast product. The CT also allows the study of the distal portion of the common biliary duct, the head of the pancreas, the gallbladder, and finally allows the study of possible complications (fig. 5).

The CT cholangiography allows visualizing the accumulation of the contrast medium, excreted in the bile duct, in the cyst and allows a precise mapping of the biliary tree [6].



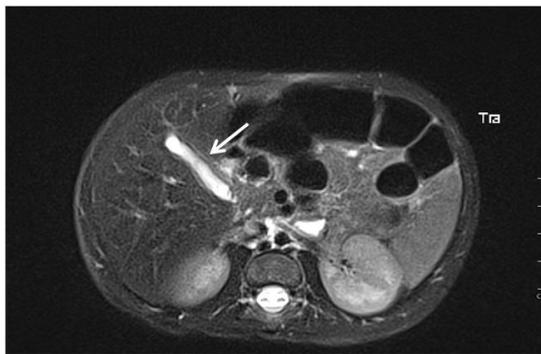
**Fig-5: Abdominal CT with injection of contrast product: Axial view (a) -Sagittal view (b)- Coronal view (c). Supra-pancreatic Hilar hepatic mass, fluid content, with thin walls, not enhanced by contrast (red arrow). Choledochal cyst type I of Todani**

Magnetic Resonance Cholangiopancreatography (MRCP), using heavily T2-weighted sequences (HASTE sequences), is the best test to diagnose a choledochal cyst (fig. 6, 7, 8). It is a

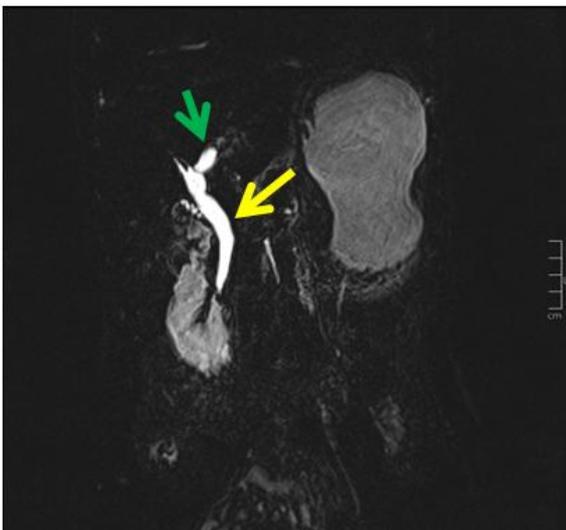
recent, non-invasive technique that allows a better analysis of the biliary and pancreatic ducts and bilio-pancreatic junction, as it makes it possible to rule out differential diagnoses [7, 8].



**Fig-6: Coronal T2-weighted MRCP shows dilation of common bile duct (Arrow). Choledochal cyst type IA**

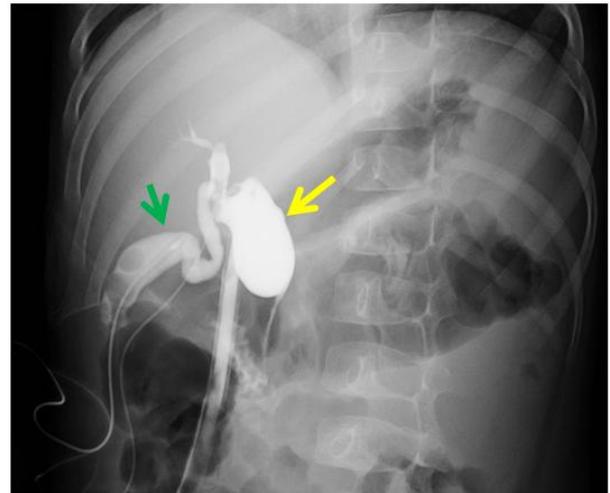


**Fig-7: Axial T2-weighted MRCP shows a fusiform dilation of common bile duct( arrow). Choledochal cyst type Ic**



**Fig-8: Coronal T2-weighted MRCP shows a cystic dilatation of the main biliary tract (yellow arrow) associated with dilation of the intrahepatic biliary ducts (green arrow). Bile duct cyst type IV?**

The direct opacifications of the biliary system find their interest mainly peroperatively



**Fig-9: Intraoperative cholangiography showing a sacciform cystic dilatation of the common bile duct type 1b according to TODANI (yellow arrow) and the dilation of the cystic duct (green arrow)**

The usual complications of choledochal cysts are stones that can block low bile duct, spontaneous rupture, pancreatitis, primary biliary cirrhosis, portal hypertension, cholangitis and malignant degeneration to a cholangiocarcinoma.

The treatment of this pathology is surgical and consists of a complete resection of the cyst with y shaped hepatico- jejunal anastomosis and disconnection of the bile duct from the wirsung. This wide resection is indicated because of the risk of secondary degeneration of the dysplastic biliary wall.

## CONCLUSION

The choledochal cyst is a malformation that can be diagnosed pre- and postnatally. Medical imaging, including initially ultrasound, and sectional imaging play a vital role in their diagnosis. The MRCP is the examination of choice that allows accurate biliary mapping, thus avoiding the use of invasive opacifications.

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