

## Imaging findings in – Choledochal cysts in infancy: A case report

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### Abstract

Choledochal cysts are rare and it is known that it manifests more in infancy and childhood.

They are in most cases considered congenital due to its occurrence in the fetuses and neonates.

Their origin has been the subject of many studies. However it was described that one of the origins may be related to an abnormal connection between the common bile duct and pancreatic appear in different forms, most often as fusiform dilatation or cystic of the common bile duct at radiography duct and chronic pancreatic juice reflux into the common bile duct, resulting in irritation of the duct and consequently dilatation.

Imaging diagnostic tools means a lot in diagnosis such as ultrasonography (US) is one of the best initial methods of evaluating dilatation of the intra- and extrahepatic bile ducts. Computed tomography (CT) is also an important in analyzing the intrahepatic biliary tree.

Magnetic resonance imaging (MRI) findings are similar to ultrasound, with a greater ability to demonstrate intrahepatic disease. It does, however, have the advantage that it conclusively demonstrates communication with the biliary tree.

**Key Word:** Choledochal cyst · abdominal ultrasonography (US) and magnetic resonance imaging (MRI), Pediatric.

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### I. Introduction

Choledochal cyst (CC), also known as a cystic dilatation of the common bile duct, considered as rare or an uncommon dilatation of the bile duct. This alteration of the anatomy of the bile ducts has provided serious condition for its surgical treatment (1). Today the CC has been widely published in the world and is generally considered to be a childhood disorder.

Although they may be discovered at any age, including adults, a considered number around 80% are diagnosed before the age of 10 years (1). There is a strong female predilection with M:F ratio of 1:4 with an incidence of 1:100,000-150,000. There is a greater prevalence in East Asia (2,3, 4).

The precise etiology of choledochal cysts is unclear. It has been a matter of considerable investigation and debate. The theory proposed by Babbitt and al about common channel (5) is the most widely accepted theory, and making one realize that one of the existences of anomalies in the pancreaticobiliary ductal union has aroused interest as a cause of dilatation of the common bile duct.

In this article, we reported a case of cystic dilatation of the common bile duct with repercussions in the cystic duct, common hepatic duct, bringing a description of radiology by US, CT and MRI.

### II. Case Report

A 6 years old child, 17Kg, lives in rural areas who had complained abdominal pain, intermittent fever and yellow coloration of the mucous membranes for 1 year in a context of alteration of his general condition. Physical examination demonstrated an abdominal distention with palpation of a voluminous abdominal mass.

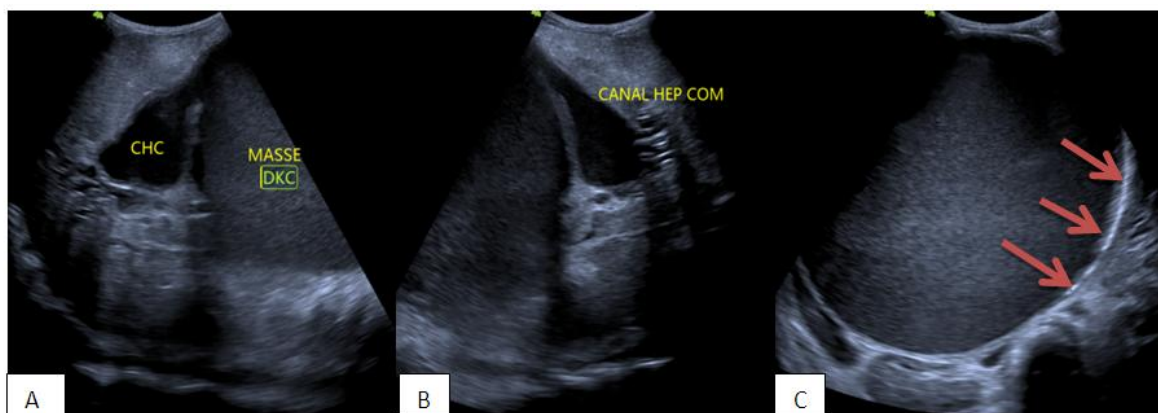
Laboratory analysis found a slight elevation of aspartate aminotransferase (ASAT) and Alanine (Amino) Transaminase (ALAT) 133 IU/L for reference range understood between 10 and 40 IU/L, hemoglobin level was lower than normal (9 g/dl). White blood cells (WBCs) 11000. However, other biologic values were normal.

An abdominal ultrasonography (US) was performed in our service showing anechoic cystic lesions under the liver, proving the presence of an important dilatation of the common bile duct and the cystic duct figure 1.

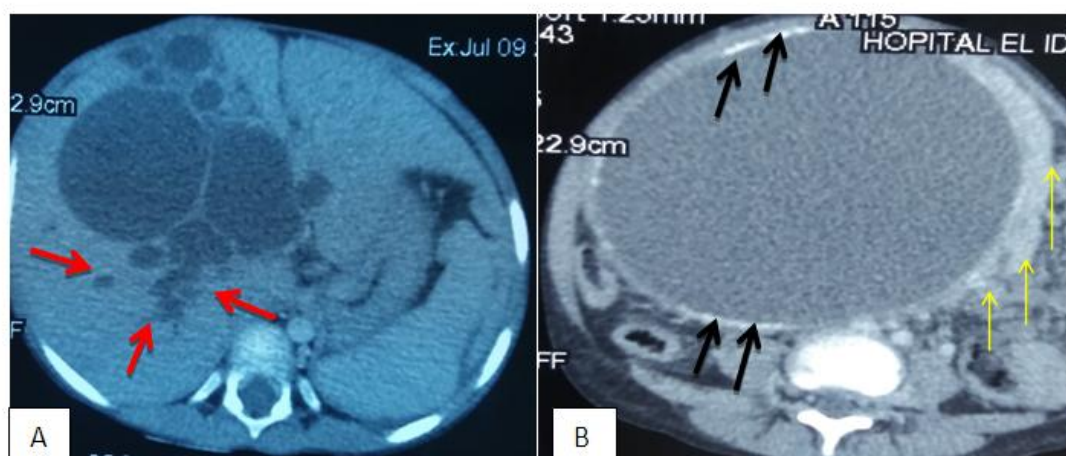
A CT and MRI image shows eccentric cystic dilatation of the common bile duct with dilatation of the common hepatic and the intrahepatic bile ducts (fig 2 and 3).

MRIs revealed hyperintense on T2-weighted images (Figure 3) and hypointense on T1-weighted images without intraluminal membrane or enhancement after administration of gadolinium (Figure 3A, B,C).

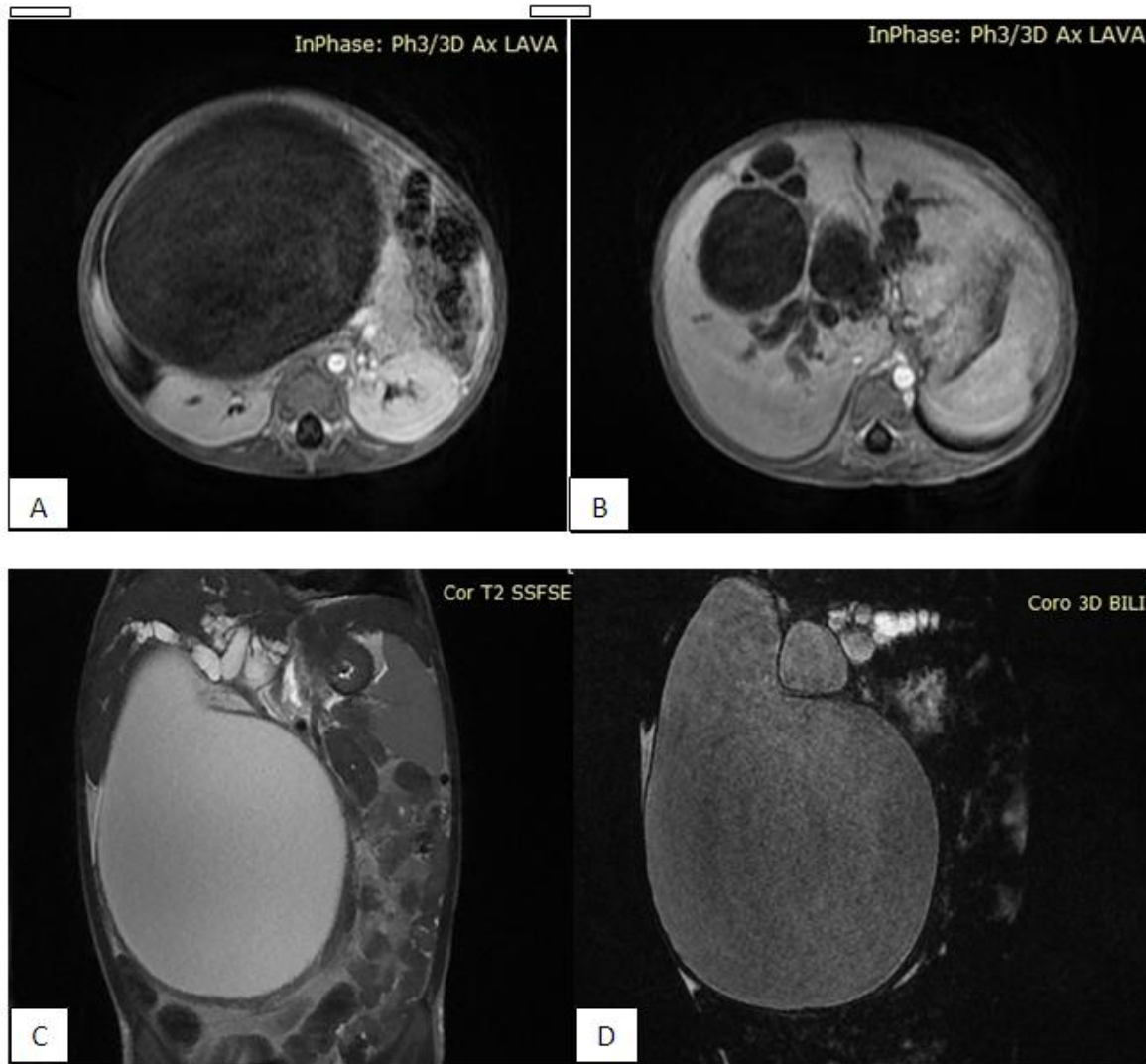
The recommended treatment is the surgical excision of the cyst with reconstruction of the extra hepatic biliary tree extends into the hepatic duct convergence.



**Fig 1:** Abdominal ultrasonography (US): Show a large Cystic dilation of common bile duct (A and B) with homogeneous hypoechoic content with (C) calcification of the wall (red arrow).



**Fig 2:** axial CT - Kystic mass of the hiliary plaque with well-defined limits. **A:** detection of dilatation of bile ducts intra hepatica (red arrow) and, **B:** the presence of calcifications in the walls (black arrow). It also has a masse effect on pancreas (yellow arrow) and abdominal vessels which are however permeable.



**Fig 3:**

Kystic mass of the hiliary plaque with well-defined limits. A and B figures ( InPhase: Ph3/3D axial LAVA):Hypointense on T1-weighted andC figure (Cor T2 SSFSE): hyperintense on T2-weighted imageswithout intraluminal membrane or enhancement after administration of gadolinium showing in the figures A and B.D (coro 3D BILI): There is also an enlargement of the intra hepatic bile ducts.

### III. Discussion

In the last few years the reports of bile duct cysts have been gone up, with more incidences in Asian continent, particularly in Japan with more than half of the cases. In a study done in Finland estimates that the incidence of bile duct cysts has been increased from 1:128.000 to 1:38.000 for the past 50 years; most of these cases were reported in children, although some studies have registered similar data in adults and children [6].

Despite various theories created, the exact etiology remains incompletely understood. The most accepted one is the proposal by Babbitts in 1969 [7], in which states that choledochal cysts are caused by an anomalous junction of the choledochal duct and pancreatic outside the duodenal wall, and proximal to the Ampulla of Vater or Oddi's sphincter while the embryonic period is underway; with a formation of a common duct about 15 mm and that generates reflux from pancreatic bile duct secretions [8 - 14].

It also describes that the activation of pancreatic enzymes in the biliary drainage system, can in a way generate an increased intra-ductal pressure and inducing inflammation, leading to a dilatation with cyst formation [13, 15, 16,].

In a retrospective study of 80 children pediatric patients who underwent choledochal cyst excision, the authors Jung et al. [18] noticed that patients with high biliary amylase levels were more likely to be diagnosed

later, present with abdominal pain, and had predominantly portal inflammation on histological examination after excision.

On the other side, embryological and motility disorders have been postulated after some patients with biliary cysts were noted to have fewer ganglion cells than expected [19, 20].

There is a huge variety in the interpretation of a possible association between CC and congenital anomalies.

There are also previous reports have demonstrated an association between pediatric CC and congenital cardiac anomalies.

In the United States, an analysis of 1646 patients with choledochal cysts, cardiac anomalies were detected in 44.9% of infants younger than those of 12 months old who were diagnosed with CC, so suggesting that screening for cardiac anomalies may be prudent in this population [21]. Many other cases have been reported showing the association of CC with annular pancreas, pancreatic cysts, colonic atresia, gastroschisis and duodenal atresia [22 - 24].

According to the classification of Alonso-Lej and colleagues formulated in 1959, which encompassed 4 types of biliary cysts (type I to IV) [25]. 18 years later, Todani and colleagues made some modifications where they added a fifth category characterized by the presence of intrahepatic cysts (Type V or Caroli Disease) [26]. In 2003 it was also recognized that the presence of anomalous pancreaticobiliary junction must be added to a classification modified by Todani, in which is currently the most utilized one [6].

Concerning the clinical presentation, opinions in different literature have been divergent, without consensus about the most common one. Some authors affirm that jaundice is the most observed symptom [27], whereas others consider being an abdominal pain [28]. However both symptoms added up to the presence of palpable abdominal mass in the right upper quadrant make up the classic triad seen in a minority of adult patients, but with higher expression in the pediatric group [9, 16, 17, 30].

The identification of bile duct dilation confirms the diagnosis of bile duct cysts after exclusion from other etiologies such as inflammatory, calculus or neoplastic [9]. For diagnostic accuracy, this condition can be identified by using specific imaging exams, namely ultrasound (US), computed tomography (CT), endoscopic retrograde cholangiopancreatography (ERCP), magnetic resonance cholangio-pancreatography (MRCP) and endoscopic ultrasound [6, 8, 10, 11, 16, 29]. For the diagnosis of biliary cyst in case of suspicion a US, a CT or MRCP is requested; The last one, however, would be considered gold standard these days and the main choice for diagnostic evaluation, as it is a non-invasive technique which does not use radiation ionizing agent [31, 17, 29, 16, 10, 29]. It should be noted that in most cases ultrasound is the first examination performed to investigate the condition, being effective to evaluate the presence of dilatations and gallstones in the biliary duct [9, 30].

Surgery is standard treatment for bile duct cysts with specific approaches for each type according to the classification.

#### **IV. Conclusions**

Bile duct cysts are rare pathology that can be found more in children than adults, especially in Asian population.

The classic triad signs and symptoms are characterized by jaundice, abdominal pain, with palpation of abdominal mass in the right upper quadrant.

Ultrasound is the most used in the initial approach followed by magnetic resonance cholangio-pancreatography (MRCP). The main treatment is consisted in surgical resection from cyst(s) when feasible, taking into account the risk of malignancy.

#### **AUTHOR'S CONTRIBUTIONS**

**ARTHUR SEMEDO INSUMBO** – Resident, Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

**KAUKONE RAISSA** – Resident in pediatric radiology service

Collection of patient clinical information, analysis and interpretation of data, revising the work critically for important intellectual content, final approval of the version to be published, agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

N.ALLALI– Analysis and interpretation of data, revising the work critically for important intellectual content, final approval of the version to be published, agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

S. EL HADDAD– Analysis and interpretation of data, revising the work critically for important intellectual content, final approval of the version to be published, agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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#### Conflict of Interest

The authors declare no conflict of interest.

#### Data Availability

All relevant data are within the paper and its Supporting Information files

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