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Short Communication Open Access

# The Curiosity, The Genealogy, The Pouch: Choledochal Cyst

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## Short Communication

Congenital deviations of the biliary tree displaying dilatation of intra and extrahepatic bile ducts are designated Choledochal cysts. The condition was initially depicted by Vater in 1723 as a dilatation of the common bile duct [1] and further narrated clinically in 1852 [2]. The Choledochal cyst primarily is categorized into: i) Cystic dilatation of the common bile duct, ii) Diverticulum of the common bile duct and iii) Choledochocoele. Two more denominations are identified [3] as Type I and Type IV-A. Choledochal cysts elucidate a female predominance, proportioned as 3.1 to 4.1:1. Majority of choledochal cysts (85%) are encountered in the first decade of life and 20% cases arise in adults [4]. Choledochal cysts are known to predispose to biliary tract malignancies in 6% to 20% of the occasions and the risk is enhanced in adults and patients of the Far East. Choledochal cysts detected below 10 years of age usually evolve into biliary tract cancer in 0.7% cases. Predominance of gall bladder cancer is 67.8% and bile duct cancer in 32.1% in encountered choledochal cysts (Figure 1).

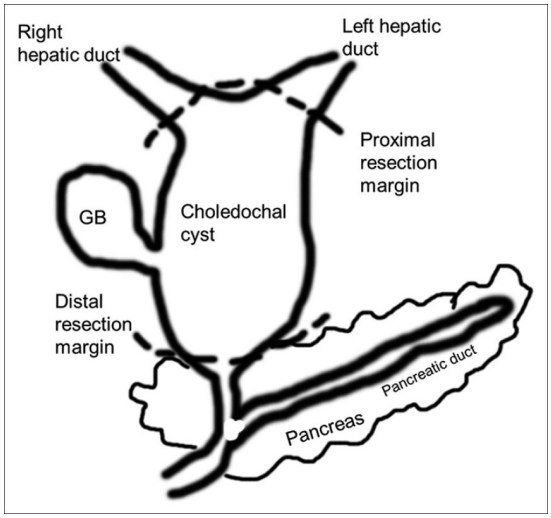


**Figure 1:** Representation of choledochal cyst.

### Categorization of Choledochal Cyst

Todani et al. [3] divided choledochal cysts into Type I: Saccular or fusiform dilatation of the extrahepatic bile ducts with 80% to 90% case frequency. Type I is further subdivided into Type I (A) elucidating diffuse cysts with a saccular dilatation of the common bile duct, Type I (B) describing focal cysts with a segmental dilatation of the common bile duct and Type I (C) depicting fusiform cysts and dilatation of the common hepatic and common bile duct. Type II represents a diverticulum of the common bile duct. Type III details Choledochocoeles (1.4% to 4.5%) with a dilatation of intraduodenal portion of the common bile duct, the pathogenesis of which entails duodenal duplication. Choledochocoeles are subdivided into two or more classes [5]. Type IV with the second periodicity (10% to 15%), denotes the dilatation of the intra or extra hepatic bile ducts or both. It is sorted into Type IV A comprising of multiple intra or extra hepatic ducts and Type IV B affecting only multiple extrahepatic ducts. Type V cysts (Caroli's disease) enunciates a segmental cystic dilatation of the intrahepatic bile ducts. Deformities of the embryonal ductal plate at varying levels of the biliary tract beget this autosomal recessive condition.

### Anomalous Pancreaticobiliary Junction

Type I and Type II cysts are associated with an Anomalous Pancreatico Biliary Junction APBJ. Type I and IV cysts need embryologic demarcation from cystic subsidiaries such as diverticulum of the extrahepatic bile duct (Type II), choledochocoele (Type III) and Caroli's disease (Type V). Anomalous Pancreatico Biliary Junction (APBJ) is the designated junction between the pancreatic and the common bile duct localized at the external duodenal wall [6,7]. The complex Type I and Type IV choledochal cysts specify up to an estimated 90%, majority of which are obstructed by APBJ (Figure 2).

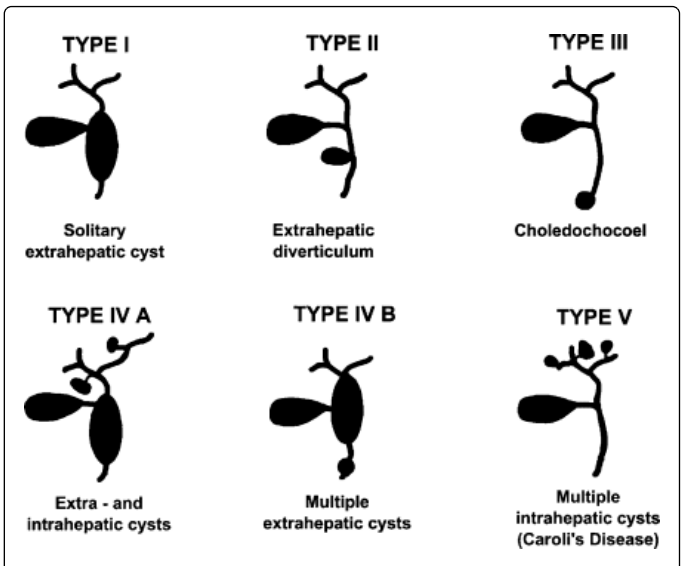


**Figure 2:** Corollaries of choledochal cysts.

The nature of concurrence between the terminal choledochus and the pancreatic duct delineates the APBJ. APBJ is categorized into 2

forms depending on the fusion prototypes of pancreatic and bile duct. The P-C Type visualizes the main pancreatic duct conjoined to the common bile duct, whilst the common bile duct compounds with the main pancreatic duct in the C-P type [8]. The fusion comprising of pancreatic and bile duct in choledochal cysts is categorized into four types-integrating a type a, type b, type c and the miscellaneous type. Type A denotes a C-P type and Type b designates a P-C type of connection. The segregation is in accordance with the position of the accessory pancreatic duct and the occurrence of dilatation in the common channel and the common bile duct. The confluence of the pancreatic and the bile ducts in choledochal cysts is collated into Type I which is exemplified as C-P type (35.3%), Type II anointed as P-C Type (21.6%) and Type III expounds a complex Type (43.1%) of the detected choledochal cysts [9]. The JSPBM (Japanese study group on pancreatico biliary maljunction) annotates the APBJ into three subgroups i.e. type a (right angle, the P-C type and 57.9% of designated choledochal cysts), Type B (acute angle, the C-P Type and 32.4%) and Type C (complex type with 5.6% cases), thus conforming the essence of convergence of the main pancreatic duct and the common bile duct. Non dilated variety of APBJ decodes type A in 29.4%, type b in 60.8% and type c in 7.2 % in the choledochal cyst disorder. Reflux of pancreatic juice into the bile duct is visualized in the elongated common conduit configured by APBJ. The common conduit generally is of 22 mm to 28.1 mm (13 mm to 67 mm) dimension in APBJ, although it is 4.6 mm to 5 mm (2 mm to 20 mm) in normal adults. Retroflux of pancreatic juice into the common bile duct can occur in individuals devoid of APBJ, in conditions where the pancreatic duct conjoins to the common bile duct as visualized on the cholangiopancreatogram, except when the contraction of the sphincter of Oddi occurs. Consequently, APBJ is assigned when the lengthy common conduit arises without any connection to the retracting sphincter of Oddi.

## Clinical Conferral

Choledochal cyst expounds a classic triad of abdominal pain, jaundice and an abdominal mass. Preponderantly 80% of the cases present clinically at a variable age, though prior to 10 years of age. Adults are generally asymptomatic (23%), however children can be clinically symptomatic (82%). Subdivision into the infant group (<1-year old) and the classic paediatric or adult group (>1-year old) of the condition is acknowledged. The infant group usually exhibits jaundice (64%), hepatomegaly and an abdominal lump (82%). Nevertheless abdominal pain is not manifested. Only a minority of the infants (0% to 17%) constitute the classic triad of clinical expressivity. Adult or paediatric patients perpetually demonstrate abdominal pain (78%-90%), fever, nausea, vomiting, cholangitis and jaundice (40% to 50%) [10]. Pancreatitis with cholangitis is characteristically illustrated in adults. Dilatation or stricture of the bile duct creates bile stasis, stone or sludge formation and evolves in to ascending cholangitis or pancreatitis.

## Aetiologic Propoundments

Recommended hypothesis are i) choledochal cyst commences from divergent proliferation of the epithelium of the common bile duct. ii) choledochal cysts originate from APBJ. Refluence of the pancreatic juice in to the bile duct accompanies the inflammation, epithelial denudation and the defective bile duct wall ultimately incites cyst formation. Scrutinizing cholangiopancreatograms depicts that APBJ is configured by an aberrant fusion of the common bile duct and the ventral pancreatic duct, thus the distal end of the common bile duct is in coherence with the ventral pancreatic duct. Besides Type I and Type IV-A choledochal cysts and cases of APBJ without choledochal cysts (non-dilated variants of APBJ) are distinguished as congenital anatomical deviations of the pancreas. Pathological conclusions such as review of the lobular structure, immunohistochemistry for the Pancreatic Polypeptides (PP) and islet cell delineation are utilized to discern the ventral/dorsal pancreas and the Type I and Type IVA choledochal cysts. Choledochal cysts and the head of the pancreas evince an inordinate amount of inessential pancreatic tissue, comprising of diminutive, compressed, lobular and PP poor architecture. The left ventricular analage can persist, the intra or extrahepatic ducts can recanalize and dilatation of the bile ducts can occur, thus inducing the formation of a choledochal cysts. The head of the pancreas with choledochal cyst may depict anatomically deviant structures such as an abnormally shaped head of the pancreas or atypical anatomical localization of the major papillae. In 70% patients with choledochal cysts it is located in the distal portion of the duodenum (Figure 3).

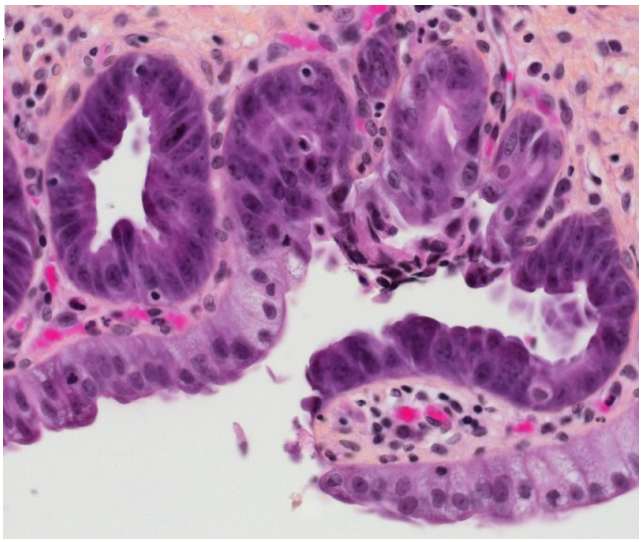

**Figure 3:** Biliary epithelial dysplasia in choledochal cyst.

## Interpretation

The preliminary imaging modality for detecting choledochal cyst is ultrasonography which delineates a cystic mass at the right upper quadrant abutting the gall bladder. Choledochal cyst detected by ultrasonography evokes a specificity of 71% to 97%. A Choledochal cyst depicts a continuum with the bile duct in 93% of the patients, as elicited by ultrasound. Alternative diagnostic mechanisms such as Computerized Tomography (CT), Magnetic Resonance Imaging (MRI) and Magnetic Cholangiopancreatography (MRCP) are imperative for distinguishing various intraabdominal cysts and displaying the connection with the biliary tree. CT is beneficial in exposing a choledochal cyst; however it is challenging to depict the confluence of the pancreatic and the bile duct. Multidetector computerized tomography (MDCT) allows minimal compilation with the Multiplanar reformation (MPR), which delivers a comprehensive guidance on the pancreatic and the bile duct. MPR images allow a diagnosis of APBJ with an accuracy of 80% [11]. Curved planar

reformation (CPR) is helpful in detecting pancreatic and bile duct. Computerized Tomographic Cholangiography (CTC) is utilized for the appraisal of the anatomical and atypical configuration of the bile ducts, especially prior to a laproscopic cholecystectomy and this diagnostic procedure demonstrates a sensitivity of 93%. Choledochal cyst expounds a sensitivity of 100% with MR cholangiography and exemplifies a detection rate of 91% with CT cholangiography. APBJ is delineated with CT cholangiography with a sensitivity of 84% though the detection of APBJ by MR cholangiography is a mere 46%. MR cholangiography is a non-invasive procedure which refrains from the usage of ionizing radiation and a contrast agent. Since a procedural breath holding is not recommended, it is a strategy of choice in the paediatric population suspected of a choledochal cyst. Endoscopic Retrograde Cholangio Pancreatography (ERCP) is the gold standard, a precise practice for distinguishing the APBJ. A sub-category of invasive, direct cholangiography, it is accompanied by considerable morbidity and mortality. In contrast, MR cholangiopancreatography is a noninvasive method of examination that displays the aberrations of the biliary tree. It is able to determine the choledochal cysts and anomalous pancreatobiliary junction (APBJ) in paediatric patients with a precision of 82% to 100% [12]. The execution and explication of biliary strictures, dilatation and filling defects <3 mm or smaller by the agendum of MR cholangiography is outstanding. Acute cholecystitis and neonatal jaundice is regularly investigated with Hepato-biliary Scintigraphy as is cyst rupture in the patients with choledochal cysts and the display of the continuity of the bile duct. Scintigraphy also aids alternative diagnostic measures in paediatric choledochal cysts.

## Therapeutic Regimen

Choledochal cysts are ideally managed by surgical extermination of the cyst. Cystoduodenostomy or Cystojejunostomy employed for internal drainage demonstrates a minimal mortality and appears technically effortless. Cystoenterostomy exhibits postsurgical complexities such as reoccurring cholangitis, intrahepatic calcification and carcinomatous transformation of the associated choledochal cysts. Coexistent ramifications are stone formation, pancreatitis, portal hypertension and hepatic abcess secondary to external drainage. Malignant conversions of the cyst wall and cyst fragments are predicted. Integrated excision of the cyst with cholecytectomy, Rous en Y hepatico-jejunostomy and reconstruction (RYHJ) is the contemporary, standardized therapeutic option for type I and type IVA. The development and aggravation of conditions such as recurrent cholangitis, (2.3% to 10%) intrahepatic calculi (10% to 16.7%) and postoperative anastomotic stricture (4.1%) has improved significantly with this therapeutic recommendation [13]. Postoperative dilemmas generally vary with age, the decreed surgical methods and establishments chosen for treatment. Adults represent 42.5% and paediatric population a mere 9% of the post-operative obstacles [14]. RYHJ as a therapeutic agendum is efficacious in 92% individuals with a minimalistic complication rate up to 7% in contrast to the Hepatico-duodenostomy. Optimal, definitive treatment for Type I and Type IVA cysts is extensive surgical excision, which diminishes the hazard of malignant transformation. Extended evaluation and follow-up is necessitated for evaluation of the neoplastic metamorphoses.

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