Bilio-Pancreatic Malformations in Adult Patients

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TABLE OF CONTENTS

LIST OF ILLUSTRATIONS

Part 1: Cystic Dilation of Bile Ducts

Part 2: Pancreatic Ducts Anomalies

LIST OF TABLES

Part 1: Cystic Dilation of Bile Ducts

Part 2: Pancreatic Ducts Anomalies

PREFACE

Ductal anomalies of the biliary and pancreatic ducts are congenital malformations that can remain asymptomatic for a long time and are thus diagnosed in adulthood particularly in case of complication.

The biliary atresia is the only biliary malformation that appears at birth and will not be dealt with in this manuscript. On the other hand, we will focus in this book only on bilio-pancreatic malformations in adults that are:

- Cystic dilation of the bile ducts
- Bilio-pancreatic maljunction
- Pancreas divisum: the most common pancreatic ductal anomaly
- The annular pancreas
- Dorsal agenesis of the pancreas and
- Ansa pancreatica (the least frequent malformation)

It is worth noting that the advent of MRCP has made the diagnosis of these anomalies relatively easy and has undoubtedly enabled clinicians to familiarize themselves with these pathologies. This has led to a considerable improvement in the therapeutic management of these malformations.

This work aims to review all medical aspects (the epidemiological, anatomopathological, clinical, morphological and therapeutic aspects) of the bilio-pancreatic malformations in adults.

List of Abbreviations

CBD: Common Bile Duct
CDBD: Cystic Dilation of Bile Ducts
ERCP: Endoscopic Retrograde Cholangio-Pancreatography
ESM: Endoscopic Sphincterotomy of the Major Papilla
ESm: Endoscopic Sphincterotomy of the Minor Papilla
EUS: Endoscopic Ultrasound
IPMNP: Intra Papillary Mucinous Neoplasm of the Pancreas
MRCP: Magnetic Resonance Cholangio-Pancreatography
PBMJ: Pancreatico-biliary Maljunction
PDiv: Pancreas Divisum
S-MRCP: MRCP with Secretine Injection
SPPNP: Solid Pseudo Papillary Neoplasm of the Pancrea
US: Ultrasound

PART 1

CYSTIC DILATION OF BILE DUCTS & PANCREATICO-BILIARY MALJUNCTION

CHAPTER 1

INTRODUCTION

Cystic dilatations of the bile ducts (CDBD) are a rare congenital biliary malformation. It is defined as an ectasia of a part or all of the intra and/or extra hepatic bile ducts (1,2). It is worth noting that it must not be secondary to a tumor, a lithiasis or a biliary inflammation.

CDBD is frequently associated with a pancreatico-biliary maljunction (PBMJ). The incidence of CDBD varies from 1/100,000 to 1/150,000 in Western countries and is about 1/13,000 in some parts of Asia (3).

The most commonly used classification is that of Todani (3), which groups CDBD into five types according to the location, extent and type of dilatation of the bile ducts. The type I is the most common (80%) (6).

CDBD can be discovered during childhood but can remain asymptomatic for a very long time (4). Patients with CDBD may also develop several complications such as biliary lithiasis, acute cholangitis, acute pancreatitis, cyst rupture and the redoubtable cholangiocarcinoma of which risk is significantly higher than that of the general population (7).

The diagnosis of CDBD has been revolutionized by the advent of Magnetic Resonance Cholangio-pancreatography (MRCP) as it not only determines the type of CDBD according to the Todani classification but also allows to search for a PBMJ. Furthermore, this imaging may reveal a possible biliary degeneration. That is why MRCP is currently essential to guide the surgical attitude.

CHAPTER 2

HISTORY

In 1723, Vater conducted a study that described the normal anatomy and the anomalies of the bile ducts (8). Nevertheless, the first complete description of CDBD was reported by Gouglas in 1852 (9).

From the year 1920 onwards, numerous works were focused on this subject in order to propose etiopathogenic and/or therapeutic concepts: these are the studies of Zinniger and Cash (1932), Poate (1936), Tsoudakas and Robinette (1956) and Yotuyanagi (1936). The latter suggested the congenital origin of CDBD based on the theory of the inequality of epithelial proliferation (10-12).

In 1959, Caroli considered that CDBD is secondary to congenital atresia most often located at the bilio duodenal junction (13). This was subsequently refuted by Babitt in 1973, as he incriminated a biliopancreatic junction anomaly (anomaly of the biliary and pancreatic ducts fusion) defined by a combination of 3 criteria (5,14,15):

- A common channel over 15mm long.
- An extra duodenal junction of the two channels away from the sphincter.
- A connection angle greater than 30°.

Alonso-Lej proposed in 1959 the first classification of the choledochal cyst. It solely takes into account the cystic dilatation of extra hepatic bile ducts, and it was only in 1975 that the cystic dilatation of intra hepatic bile ducts was included in the classification by Flangian (16,17).

Finally, it was in 1977 that Todani drew up a complete classification of CDBD into 5 groups according to their morphological aspects (18,19).

CHAPTER 3

EPIDEMIOLOGY

1. Frequency

The CDBD is a rare congenital biliary pathology and still less frequent than biliary atresia (20). Its incidence is 1/13500 births in the United States, 1/15000 births in Australia, and Western countries (in general). It is remarkably more frequent in Asian populations with an incidence of 1/1000 births: it is worth noting that the two thirds of cases occur in Japan and only 0.3% are observed in South Korea (21).

2. Sex-Ratio

A clear female predominance is reported by all authors (22). The CDBD is encountered three times more often among women than men, with a sex ratio ranging from 1/4 to 1/3 (23,24).

3. Age

The CDBD affects all age ranges, but it is generally discovered in children, teenagers and young adults (25,26):

- Twenty-five percent (25%) of cases are infants under one-year-old.
- Sixty percent (60%) of cases are children under 10-year-old.
- Twenty percent (20%) of cases are diagnosed in adulthood (27, 28).
- Less than 1% of CDBD are diagnosed in patients over 70 years. To date, the extreme age of diagnosis is 87 years (29).

Currently, it is widely accepted that the CDBD is of congenital origin as many cases were discovered on antenatal ultrasound (from the 15th week of pregnancy).

Adult forms are generally CDBD that already exists since childhood but remained unnoticed because there were no symptoms (30).

CHAPTER 4

CLASSIFICATION OF CDBD

Alonso-Lej and his colleagues proposed the first CDBD classification in 1959. Then, Todani et al. reported in 1977 the most widely accepted classification, derived from Alonso-Lej's original classification and based on the site of cystic dilatation (31).

1. Todani classification

In 1977, Todani and his colleagues extended the 1st classification of Alonso-Lej et al. made in 1959 (it included 3 types of CDBD).

The Todani's classification is typically morphological and takes into account both intra and extra hepatic bile ducts dilatations. It is currently, the most widely used classification (32).

1.1. Type I

It accounts for almost 90% of CDBD. The anomaly develops on the extra-hepatic bile duct below the intra-hepatic bile ducts and above the intra pancreatic portion of the choledocus.

A type I cyst may be associated with a slight dilation (or hypertrophy) of the intra-hepatic bile ducts secondary to biliary stasis. In addition, the cyst volume is variable and may hold more than one liter of bile.

There are 3 subtypes of Todani Type I CDBD. This differentiation is based on the location of the gallbladder in relation to the cyst on the one hand and on the dilation shape on the other hand (25) (Figure 4-1):

- *Type Ia:* it is a sacciform cystic dilation of the entire extra-hepatic common bile duct without intra-hepatic bile ducts involvement. The cystic duct (of the gallbladder) fits into the dilated duct.
- *Type Ib:* it is an isolated segmental dilation of the distal part of the common bile duct. The rest of the biliary tree is generally very normal.
- *Type Ic:* it is a smooth fusiform dilation of the common bile duct associated with a PBMJ (33).

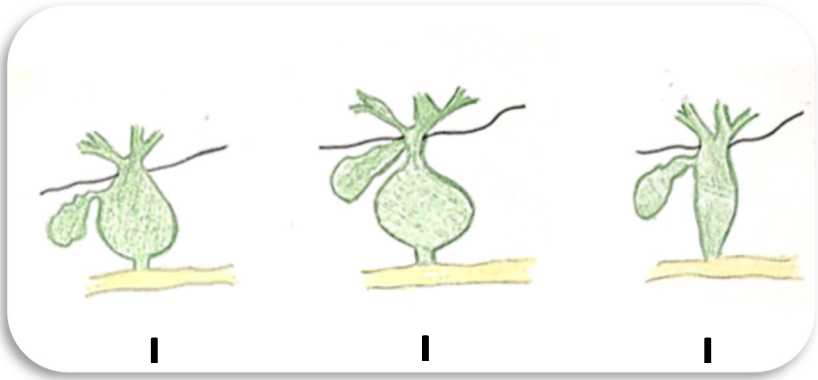

Figure 4-1: The 3 subgroups of CDBD type I according to the Todani classification. Type Ia: sacciform dilation. Type Ib: segmental dilatation. Type Ic: fusiform dilatation.

1.2. Type II

The type II of CDBD (2%) is a real diverticulum of the common bile duct. In general, it is a lateral sacciform dilation attached by a usually short and narrow pedicle to a non-cystic common bile duct (20). Such a diverticulum may resemble a duplication of the gallbladder.

Figure 4-2: The type II of CDBD according to the Todani classification

1.3. Type III

The type III of CDBD, also called choledochocele, was reported for the first time by Wheeler in 1940. It is a cystic dilatation of the terminal portion of the common bile duct protruding into the duodenal lumen. The overlying

portion of the common bile duct is usually normal or dilated but not cystic (34-37).

Figure 4-3: The type III of CDBD according to the Todani classification

1.4. Type IV

The type IV of CDBD may involve dilatation of both intra- and extra-hepatic bile ducts. They are classified into two subgroups, namely types IVA and IVB (38-41) (Figure 4-4):

- The type IVA: it is a cystic dilatation of the common bile duct, whatever its type, associated with intra-hepatic bile ducts cystic dilatation. The Intra-hepatic biliary dilatation is most often bi-lobar. When it is an associated uni-lobar dilation of intra-hepatic bile ducts, it is more frequently located in the left liver lobe.
- The type IVB: it is a multiple cystic dilatation of the extra-hepatic bile ducts, which may combine several of the previous types of CDBD. So, this type consists of multiple dilations of the common bile duct, classically described as a "string of pearls".

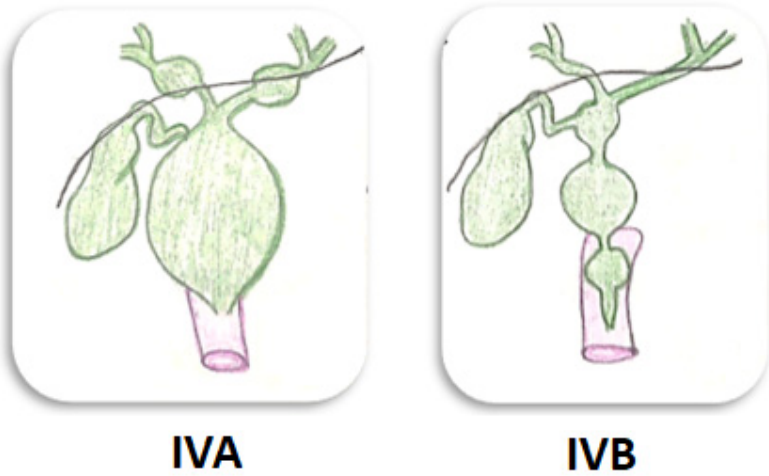

Figure 4-4: The type IV of CDBD according to the Todani classification

1.5. Type V

The type V of CDBD, also called Caroli disease, is a congenital, saccular or fusiform, non-obstructive dilatation of the intra hepatic bile ducts (42, 43).

In this case there may be intra hepatic biliary dilations interspersed with healthy ductular areas. This is called a rosary-like aspect of the intra hepatic biliary tree. They can be diffuse or localized and are most common in the left liver lobe. The extra-hepatic bile ducts can be normal or pathological. There are 2 subgroups of this kind (type V) of CDBD *(44)*:

- *The 1st subgroup or type I or Caroli disease:* it is a pure form of the CDBD type IV. The liver is apparently normal and there is no congenital hepatic fibrosis. This form concerns young adults (< 30 years) and the occurrence of acute cholangitis is common.
- *The 2nd subgroup or type II or Caroli syndrome:* it is a mixed form of the CDBD type IV. There is a congenital hepatic fibrosis with a bulky and harsh liver. The size and morphology of the dilated intra-hepatic bile ducts are variable (44).