

STUDY OF STRUCTURAL AND FUNCTIONAL DEFECTS IN CHOLEDOCHAL CYST

Rajib Khastagir¹ Md Akbar Husain Bhuiyan² Md Abdullah Al Farooq² Mohammed Saiful Islam³

Summary

Although many research works have been undertaken over decades to find out the etiology and mechanism of the structural defects resulting in formation of choledochal cyst and the consequent physiological dysfunction, the exact cause remained obscure. This study was undertaken to explore what structural changes in the choledochal cyst leads to its dilatation, at the same time it was also decided to find out how the choledochal cyst behaves with volume change. The study was conducted on 10 children with choledochal cyst in the Department of Paediatric Surgery, Bangabandhu Sheikh Mujib Medical University, Shahbag, Dhaka during the period of March 2007 to October 2008 by infusing a volume of contrast diluted with normal saline with the help of peroperative cholangiogram and a pressure measuring device. Biopsy was taken from resected choledochal cyst wall and wall of the CBD unaffected by the cyst. We hypothesized that, choledochal cyst is plastic in nature and there is increase of collagen fibers than elastic fibers in the wall of choledochal cyst that causes its dilatation. Out of 10 patient 2 were male and 8 were female. In quantitative estimation of collagen and elastic fiber, in control quantity of collagen fiber mean 106.40 ± 49.50 , quantity of elastic fiber mean 79.80 ± 15.98 , proportion of collagen and elastic fiber mean 1.35 ± 0.59 and in case (from choledochal cyst wall) quantity of collagen fiber mean $199.60 \pm$ quantity of elastic

fiber mean 46.10 ± 1.50 , proportion of collagen and elastic fiber mean 4.76 ± 1.96 . All this findings are statistically significant (p -value < 0.001). From the study it concluded that, choledochal cyst is plastic in nature showing almost a sigmoid shaped curve and increase in the collagen fiber over the elastic fiber in its wall causing dilatation with resultant stasis.

Key words

Choledochal cyst; Collagen; Elastic; Cystic pressure.

Introduction

At fully developed embryo, bile duct opens into the second part of duodenum through the ampulla of Vater [1]. Choledochal cyst is a dilatation of the common bile duct (CBD) with onward drainage problem resulting in poor emptying and significant bile stasis [2]. This dilatation of CBD is not a post obstructive consequential dilatation as hyperbilirubinaemia is not a presenting feature of uncomplicated choledochal cyst. However intermittent jaundice or hyperbilirubinaemia may occur as a result of obstruction due to fleck, sludge, gravel and stone formation and also during cholangitis and pancreatitis [3,4]. It signifies that although the dilatation is not post obstructive, stasis in choledochal cyst occur and may be a result of structural as well as physiological dysfunction [4,5]. This study was undertaken to explore structural changes in the choledochal cyst leads to its dilatation. At the same time it was also decided to find volume changes in choledochal cyst and these two phenomenon are structurally and functionally dependent. We hypothesized that choledochal cyst is plastic in nature, there are increase of collagen fibres than elastic fibres in the wall of choledochal cyst that causes its dilatation. From the study it concluded that choledochal cyst is plastic in nature showing almost a sigmoid shaped curve and increase in the collagen fibre over the elastic fibre in its wall causing dilatation with resultant stasis.

1. Assistant Professor of Pediatric Surgery
Chittagong Medical College, Chittagong
2. Associate Professor of Pediatric Surgery
Chittagong Medical College, Chittagong
3. Professor & Chairman of Pediatric Surgery
Bangabandhu Sheikh Mujib Medical University, Dhaka

Correspondance: Dr. Rajib Khastagir
Email: rajibkhastagirkhastagir@gmail.com
Cell: 01712 090980

Materials & methods

The study was conducted on children with choledochal cyst operated as routine cases. With the help of preoperative cholangiogram and by pressure device intracholedochal cystic pressure was measured in consequence of sequential volume infusion to observe the volume- pressure relationship of the cyst and thereby determine its plasticity. Biopsy was taken from resected choledochal cyst wall and wall of the CBD unaffected by the cyst. It was done to observe constitutional difference between affected and unaffected parts of the biliary tree as regards the proportion of collagen and elastic tissue contents by quantification of that collagen and elastic tissue contents. The study was carried out in the Department of pediatric surgery, Bangabandhu Sheikh Mujib Medical University, Dhaka from March 2007 to October 2008. Patients were of both sexes within age range of 1 to 15 years diagnosed with choledochal cyst type I. The subjects were selected after detailed clinical history, physical examination liver function tests and USG of whole abdomen. Information about the patients was obtained in a structured data sheet.

Results

Total 10 patients were included in the study. All were diagnosed as choledochal cyst and operated as routine cases. With the help of preparative cholangiogram and by pressure device intracholedochal cystic pressure was measured in consequence of sequential volume infusion. Biopsy was taken from resected choledochal cyst wall and wall of the CBD unaffected by the cyst. Each specimen was tested by Verhoeff's iodine iron hematoxylin (Verhoeff's) for elastic fiber and Masson's trichrome for collagen fiber [6]. Age ranges was 4 to 13 years with a mean of 7.50 ± 3.23 . Out of 10 patients 2 were male and 8 were female. In control group, mean quantity of collagen fiber was 106.40 ± 49.50 , mean quantity of elastic fiber was 79.80 ± 15.98 and mean of the proportions of collagen and elastic fiber was 1.35 ± 0.59 . In case group (from choledochal cyst wall) mean quantity of collagen fiber was 199.60 ± 64.40 , mean quantity of elastic fiber was 46.10 ± 1.50 and mean of the proportions of collagen and elastic fiber was 4.76 ± 1.96 (Table I).

All this findings were statistically significant ($p < 0.001$). With the help of preoperative cholangiogram and by pressure device intracholedochal cystic pressure was measured in consequence of sequential volume infusion to observe the volume-pressure relationship of the cyst. Mean volume of normal saline with contrast was 305.00 ± 64.33 ml and mean intracholedochal cystic pressure was 36.00 ± 8.81 (Table II).

Table I : Comparison of Intra choledochal cystic pressure and relative quantification of collagen and elastic tissue contents of choledochal cyst wall and normal CBD

Variables	Biopsy taken		P value
	From normal CBD (Sample 1)	From choledochal cyst (Sample 2)	
Quantity of collagen fiber	106.40+49.50	199.60+64.40	0.001
Quantity of elastic fiber	79.80+15.98	46.10+19.50	0.001
Proportion of Collagen and elastic fiber	1.35+0.59	4.76+1.96	0.001

Table II : Distribution of Contrast with normal saline volume and intra choledochal cystic pressure

Contrast with normal saline volume(ml)	Intra choledochal cystic pressure(mean+SD)
50	32.00±4.19
100	36.10±3.64
150	39.80±3.29
200	41.30±3.53
250	45.20±3.85
300	45.50±3.69

Discussion

In the study, the quantity of collagen fiber was increased significantly than elastic fiber in the wall of the choledochal cyst. Collagen fibers are inelastic and because of their molecular configuration have a tensile strength greater than that of steel [7]. Consequently, collagen imparts a unique combination of flexibility and extent to the tissues in which it lies. In 'the wonderful world of fascia'

by Heidi Lauber published that it is the most commonly found fiber in fascia. It is tensile in nature and this is reflected by the largely plastic nature of the fascia [8]. Elastic fibers rich in the protein elastin, stretch easily in response to tension. Elastin contains unusual amino acids desmosine and isodesmosine, and are thought to account for the rubber like qualities of this protein which forms fibers at least 5 times more extensible than rubber [7].

From this study it was found that, the increase in the collagen fiber in the wall of the choledochal cyst and its plastic nature is responsible for the dilatation of the cyst due to physiological dysfunction. Due to the resultant stasis fleck, sludge, gravel, stone and also bile acid are likely to accumulate in the cyst. This may induce intermittent jaundice and after a long period of time, malignant change as bile acid is a strong carcinogen. There are a few such study in literature done so far.

Conclusion

A practical implication of the present study is that, although the choledochal cyst is usually asymptomatic, there is a malignant potential resulting from stasis and due to this, it should be operated. Absolute rather than relative quantification, as well as computer controlled volume infusion system to study the pressure-volume relationship is very much important.

Disclosure

All the authors declared no competing interest.

References

1. Adkins, RB, Chapman WC and Reddy, VS, 2000, 'Embryology, Anatomy, and Surgical Applications of the Extra Hepatic Biliary System', *Surgical Clinics of North America*. 2000;80(1):363-379.
2. Grosfeld, JL 'Pediatric Surgery', in *Text Book of Surgery the Biological Basis of Modern Surgical Practice*, in DC, Sabiston and HK Lyerly (eds). Prism Books (Pvt.) Ltd. Bangalor, India. 1997;1234-1274.
3. Kaneko K, Ono Y, Tainaka T, Sumida W and Ando H, 'Fatty acid calcium stones in patients with pancreaticobiliary maljunction/choledochal cyst as another cause of obstructive symptoms besides protein plugs', *Journal Paediatric Surgery*. 2008;43(3):564-567.
4. O'Neill, JA 'Choledochal Cyst', in *Paediatric surgery*, in JL, Grosfeld, JA, O'Neill, EW, Fonkalsreed and AG Coran (eds), Mosby Elsevier, Philadelphia. 2006;1620-1634.
5. Besner, GE 2008, 'Excerpt From Choledochal Cyst: Surgical Perspective'. Retrieved on 4 May, 2008 from <http://www.emedicine.com/ped/byname/choledochal-cyst-surgical-perspective.htm>.
6. Rodrigues AJ, Rodrigues CJ, Cunha ACP and Yoo J, 2002, 'Quantitative analysis of collagen and elastic fibers in the transversalis fascia in direct and indirect inguinal hernia', *Revista do Hospital das Clinicas*, vol.57, no.6, pp.11-14 retrieved from <file:///C:/DOCUME-1/KSU/LOCALS-1/Temp/CHBAZW8P.htm> on 4/28/2008.
7. Junqueira LC, Carneiro, J and Kelly RO, 1998, *Basic Histology*, Appleton & Lange. 1998; 89-120.
8. Lauber H, 'The Wonderful World of Fascia', Retrieved on 8 April, 2009, From <http://www.vancouveryoga.com/fascia.html>.