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A Study of Impact of Early Diagnosis in the Management of Choledochal Cysts of Infancy and Childhood -Experience and Analysis of 205 Cases

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Abstract

Introduction: Choledochal cyst not an uncommon encountered pediatric surgical practices. Advances in technology have impacted timing of diagnosis. Advances in instrumentation and surgical access have added yet another way of excion. But the exact impact of early diagnosis on surgery of choledochal cysts have not been analysed and reported. Hence this attempt to analyse the three periods of choledochal cyst, namely 1. PTC (Percutaneous Trans-hepatic Cholangiography) and ERCP (Endoscopic Retrograde Cholangio â??" pancreatography), 2. USG (Ultrasonography) and CT Scan (Computerised Tomography era, and 3 .Period of MRCP (Magnetic Resonnace Cholangio â??"Pancreaticography) with reagrds to impact of early diagnosis in the management of Choledochal cysts. Materials and Methods: A total of 205 cases of choledochal cysts treated by the team were analysed. The data retrieval was from aaself developed Microsoft Access based soft ware used by senior pediatric surgeon. Materials and Methods:A total of 205 cases of choledochal cysts treated by the team were analysed. The data retrieval was from aaself developed Microsoft Access based soft ware used by senior pediatric surgeon.

Index terms— choledochal cyst, roux en y hepaticodochojejunostomy, anomalous pancreatico biliary portal junction, long channel.

1 I. Introduction

holedochal cyst is cystic dilatation of extra and/or intra hepatic biliary dilatation. Choledocal cyst is not uncommon particularly after the invention of high resolution USG and now MRCP. They continue to perplex surgeons regarding etiology particularly anomalous Pancreatico-biliary ductal junctions. Purpose to question if any advances in diagnosis changed the management of Choledochal Cyst.

2 II. Aims and Objectives

3 PTC: Percutaneous trans hepatic cholangiogram ERCP: Endoscopic retrograde cholangiogram C T Sca : Computerized tomogram

To present experience of 205 cases of Choledocal cyst across 1980's, an era of P.T.C -ERCP. 1990"s, an era of USG-CT scan and late part of 1990's & 2000 era dominated by MRCP difficulties in diagnosis in USG-era and accurate anatomical delineation in MRCP era targeted yet in this presentation.

4 III. Materials and Method

A total 205 cases were seen and treated from 1983 till 2014. This period is divided this period is divided in 3 periods. Materials and Methods: A total of 205 cases of choledochal cysts treated by the team were analysed. The data retrieval was from aaself developed Microsoft Access based soft ware used by senior pediatric surgeon. The parameter studied was actual impact on surgical aspects of the three main components of surgery of choledochal cysts, namely Pain abdomen was seen in majority of cases, while jaundice was seen less frequently. Palpable mass seen in only 5 patients in the series. 3 patients were presented with acute pancreatitis picked up on USG and MRCP. 12 babies presented with biliary peritonitis. 72 were picked up incidentally from an USG-abdomen.

For other symptoms see table 3 in all the three eras. Type l out of 4 was commonest Choledochal cyst seen. Type 2 choledochal cyst was seen in four children. Type 3 choledochal cyst was seen in one child. No Caroli's disease was seen in this series.

5 V. Discussion

Choledochal cyst is a rare disease of the biliary tract in children. There are five main types of Choledochal cyst described by TODANI et al., ??l) The estimated incidence in western countries varies between 1 in 100000 and 1 in 150000. The incidence is higher in Asia and occurs more in women with a male to female ratio of 1: 3 to 1: 4. (2) In our series of study of 205 cases 168 cases were of age group of less than 10 years.

The etiology of Choledochal cyst still remains unclear. One of the most accepted hypothesis is proposed by Bobbit et al..<3) is the presence of an anomalous Pancreatico-Biliary ductal confluence proximal to the regulatory control of sphincter mechanism within the duodenal wall. This predisposes reflux of pancreatic enzymes with deconiugated bile. This induces chronic inflammation predisposing dilation of the biliary tree wall. (3,4) Although Anomalous Pancreatico Biliary Portal Junction is not that frequently reported in other series. The present series also had only 15 cases with long common channel.

The clinical triad of jaundice, mass and pain is considered as the most common and significant findings in the diagnosis of the Choledocal cyst. Surprisingly no patient showed this triad in our series. We found pain abdomen 45.8% as the most common presenting finding of Choledochal cyst as described in a study by Bukukyavuz et al. (5) Rajeev Dhupar et al., (6) reported 26% cases presented without any symptoms. But, rather as an incidental finding at CT Scan, Cholangigram or at laparoscopic surgery for other reasons. Our studies noted 36.1% of cases as an incidental diagnosis most of them being in period 3. The incidental finding of Choledochal cyst is on rise due to advanced diagnostic techniques.

Diagnosis of Choledochal cyst was made by different techniques in different periods of study as per the most reliable diagnostic tool of that time including xray, PTC, ERCP, USG, CT Scan and ERCP.

The PTC and ERCP were definitive test with 80-90% diagnostic accuracy. The advantage of this is they delineate the anatomy of biliary tree. As both of them are invasive procedures they were associated with complications. It was considered that intra-op PTC as mandatory for knowing anatomy during this period. (7,8) The present series PTC -ERCP era 18/21 underwent PTC.

Anwaza et al., first used USG for the diagnosis of Choledochal cyst when it is small. USG Is the first non-invasive imaging modality of choice in evaluation of patients suspected to have bile duct dilatation. (9) In a study by J.S. devries et al., USG has been 93% diagnostic as primary imaging. But, for surgical procedure of Choledochal cyst the detailed anatomy made possible only by aid of improved diagnostic techniques like CT & MRCP, CT Scan can clearly visualize the location and relationship with surrounding structures. (9,10) USG has picked up one incidental Choledochal cyst. In the present series too larger number of choledocal cysts were diagnosed and treated in USG and MRCP era.

Past decade has seen MRCP replaced all other investigations as it is non-invasive and gives clear cut anatomy of biliary tree for good surgical plan. 11 Antenatal diagnosis by Antenatal high resolution ultrasound & fetal MR have reported Choledochal cyst with Biliary Atresia. (12).

The present serieaparticlualrystress that advances in imaging have lead to early diagnosis AND THRE BY decreasing late diagnosis , presentation with complications like stones, pancreatitis, recurrent cholangitis , biliary cirrhosis Choledochal cyst should be treated by Surgery as it is highly associated with the risk of malignancy(17) spontaneous or traumatic rupture have been reported by Suneel Chauhan et al.,

In our series out of 205 cases studied 204 presented with type 1 /4 & type 2, all of them underwent complete excision of cyst with Roux-en-y jejunostomy as treatment of choice as proposed by Kasai et al., and shown good results. One case presented with type 3 and underwent excision with repair as treatment of choice. All the patients who underwent excision and repair and are alive but, one expired due to biliary sepsis / peritonitis and associated complications Despite the advances in accurate anatomical delineation by advanced technology of imaging the surgical approach to the three main components of operation namely Choledochal cyst per se, distal end & biliary enteric anastomosis remains the goals of treatment. As of now open Choledochal cyst excision in toto, complete excision of distal end not injuring pancreatic duct Roux en Y common Hepaticodocho Jejunostomy is the standard care. . Laproscopic Choledochal cystectomy (14) and Robotic assisted Choledochal cyst excision (15) is being tried elsewhere. Although advances in surgical instrumentation, endo surgery and Robot assisted are gathering momentum for regular use, still they cannot be termed as standard in the presence of repeated infections. Portal vein is surely safer in open choledochocystectomy as compared to Laparoscopic or Robot assisted excision. Laparoscopic or Robot assisted excsiuon related accidental injury to portal vein makes

immediate laparotomy more time consuming and cumbersome , with consequences of blood loss and biliary injuries in attempting hemostasis.

The time taken and not so effective and adequate addressing of the distal end continue to be not accepted by purists. Addressing Distal end by a magnified view of laparoscope, although is an advantage, still what is practiced and is shown in many surgical workshops by experienced endosurgeonsis convenient ligation rather than complete distal end excision. This may be due to the apprehension of injuring pancreas and or duodenum. The open conventional approach particularly aided by an operating loop, one can safely and surely deal exactly the distal end.

Common Hepaticodochoduodenostomy (16) although claims as effective as Roux en Y Common Hepaticodocho Jejunostomy, long term results of Common Hepaticodocho duodenostomy are still awaited to be accepted as a part of standard care of Choledochal cyst excision.

As of now open Choledochal cyst excision with complete excision of distal end and atleast 25cms of Roux en Y Common Hepaticodocho Jejunostomy seems to be the gold standard.

Laparoscopic & Robotic approach to inflamed, adherent Choledochal cyst may not be acceptabl and safe e (15, **??**6)

6 VI. Conclusion

The present series clearly show the increase in the incidence of Choledochal cyst due to advances in imageology. Also has impacted early diagnosis is early surgical removal and there by delayed presentation , and with complications like stone, recurrent cholangitis, pancreatitis, biliary cirrhosis This technological anatomical detaining has not reflected any significant change in the surgical management of Choledochal cyst. The advances in instrumentation and minimal access surgery and Robot assisted surgery still needs to validated as safe and can be used as standard surgical option for excision of choldochal cyst.

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Figure 1: Period 1 :C

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Figure 2: Table 1 :

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Year	O/ month	%	<1 year	%	1-5 years	%	5-10 years	%	10-18 years	%
1983-1990	0	0	0	0	03	1.5%	07	3.4%	11	5.4%
1991-1998	0	0	08	3.9%	11	5.4%	15	7.3%	04	1.9%
1999-2014	08	3.9%	37	18%	58	28.3%	21	10.2%	22	10.7%
Total	08	3.9%	45	21.9%	72	35.2%	43	20.9%	37	18%

Figure 3: Table 2 :

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10 Volume XV Issue II Version I ( ) Medical Research Global Journal of	Clinical presentation Jaundice Pain abdomen Mass Acute appendicitis Acute peritonitis	Period 1 02(0.9%) 18(8.8%) 01(0.5%) 0 0	Period 2 04(1.9%) 22(10.7%) 0 0 04(1.9%
	Incidental	0	08(3.9%)
	All the children underwent excision of the cyst and hepatico-jujenostomy, Lilly's technique used in 5, 1 child in type 2 diverticulum had diverticulectomy + repair of CBD. 1 child born with biliary peritonitis died, remaining patients were alive. Follow up ranged from 1 year to 25 years.		and at hepaticodochojejunostomy were the end surgery. Choledochoscopy of distal end cystoscope in two but later not felt to h extra information.

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Figure 4: Table 3 :

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Year 2 015

Figure 5: Table 1 :

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Type of Choledochal cyst	Period 1	Period 2	Period 3
Type 1/4	21(10.2%)	38(18.5%)	141(68.8%)
Type 2	0	0	04(1.9%)
Type 3	0	0	01(0.5%)

Figure 6: Table 4 :

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