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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

By Dr. G Raghavendra Prasad, Dr. Kasha Aishwarya & Dr. J V Subbarao

Deccan College of Medical Sciences and Princess Esra Hospital, India

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 excision. 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 – pancreatography), 2. USG (Ultrasonography) and CT Scan (Computerised Tomography era, and 3. Period of MRCP (Magnetic Resonance Cholangio –Pancreaticography) with regards 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 a self developed Microsoft Access based software used by senior pediatric surgeon.

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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

Dr. G Raghavendra Prasad ^α, Dr. Kasha Aishwarya ^σ & Dr. J V Subbarao ^ρ

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 excision. 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 – pancreatography), 2. USG (Ultrasonography) and CT Scan (Computerised Tomography era, and 3 .Period of MRCP (Magnetic Resonance Cholangio –Pancreaticography) with regards 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 a self developed Microsoft Access based software 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 1. Approach to cyst excision per se, 2. Management of distal end, 3. Restoration biliary drainage.

Results: The advances in imageology have led to early diagnosis and early surgery before complications develop. This has impacted in disappearance of delayed presentation with complications as seen by the number of cases diagnosed in neonatal period. Neonatal, perinatal, rarely antenatal detection of choledochal cysts was possible due to advances in imaging choledochal cysts. All children underwent excision of cyst and common hepaticoduodenostomy. The safety of excision particularly when dealing inflamed, adherent choledochal cysts was better with open conventional excision. Similarly the confidence of handling the distal end was more with open surgery. Laparoscopy and Robot assisted have added another surgical access to choledochal cysts. Minimal access and magnification added to better visual appreciation, but ergonomics, cost, and availability, approach to distal end remain still to be validated.

Conclusions: 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 thereby delayed presentation, and with complications like stone, recurrent cholangitis, pancreatitis, biliary cirrhosis. This technological anatomical detailing 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 be validated as safe and can be used as standard surgical option for excision of choledochal cyst.

Keywords: choledochal cyst, roux en y hepaticoduodenostomy, anomalous pancreaticobiliary portal junction, long channel.

I. INTRODUCTION

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

II. AIMS AND OBJECTIVES

PTC: Percutaneous trans hepatic cholangiogram

ERCP: Endoscopic retrograde cholangiogram

CT Scan: Computerized tomogram

To present experience of 205 cases of Choledochal 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.

III. MATERIALS AND METHOD

A total 205 cases were seen and treated from 1983 till 2014. This period is divided into 3 periods.

Period 1: from 1983 - 1990 i.e., period of P.T.C and ERCP n=21

Author α: Affiliation: 1-Professor of Pediatric Surgery, Deccan College of Medical Sciences, Hyderabad. e-mail: grprasad22@gmail.com

Author σ: Junior Resident, Department of Pediatric Surgery, Deccan College of Medical Sciences, Hyderabad. e-mail: ash_kasha@yahoo.in

Author ρ: Associate Professor of Anesthesia, Deccan College of Medical Sciences, Hyderabad. e-mail: usha_subbarao@yahoo.com

Period 2: from 1991 -1998 i.e, period of USG and CT scan n=38

Period 3: from 1992 – 2014 i.e, period of MRCP dominance n=146

A particular emphasis during analysis was made to know if USG and MRCP have influenced the diagnosis and treatment of choledochal cyst, particularly three components in management of Cyst per se, distal end and Biliary drainage were parameters of study.

Period 1 had 21 cases, period 2 had 38 cases, period 3 had 146 cases, age of presentation, clinical presentation, type of Choledochal cyst, surgical treatment offered and outcome was analyzed.

IV. RESULTS

Table 1: shows the number of cases in each period. Period 1 had 21 cases, period 2 had 38 cases, period 3 had 146 cases.

Table 1 : Case distribution in relation to period of study

Year	Number of cases	%
1983 - 1990	21	10.2%
1991 - 1998	38	18.5%
1999-2014	146	71.2%

It is obvious from table 2 during the era of P.T.C and ERCP, most were diagnosed between 5-15 years, only 3 out of 21 cases (1.5%) were diagnosed in 1- 5 years, none were diagnosed below 1 year of age. Period 2 shows 19 out of 38 cases (9.3%) were diagnosed

during infancy. In period 3, Antenatal diagnosis was made in 8 cases (3.9%) and 103 out of 146 cases (50.2%) during these period were diagnosed and treated before 5 years of age.

Table 2 : Distribution according to age at diagnosis.

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%

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 I 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.

Table 3 : Clinical presentation in relation to period of study

Clinical presentation	Period 1	Period 2	Period 3
Jaundice	02(0.9%)	04(1.9%)	08(3.9%)
Pain abdomen	18(8.8%)	22(10.7%)	54(26.3%)
Mass	01(0.5%)	0	04(1.9%)
Acute appendicitis	0	0	06(2.9%)
Acute peritonitis	0	04(1.9%)	08(3.9%)
Incidental	0	08(3.9%)	66(32.2%)

All the children underwent excision of the cyst and hepatico-juenostomy, 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.

Complete excision of the cyst, complete excision of distal end without damaging pancreatic duct

and at least 25 cms of Roux en Y hepaticodochojunostomy were the end points of surgery. Choledochoscopy of distal end was done using cystoscope in two but later not felt to have added any extra information.

Table 4 : Frequency of type of Choledochal cyst in relation to period of study.

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%)

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.,⁽¹⁾ 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 deconjugated 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 Choledochal 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, Cholangiogram 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 x-ray, 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 choledochal 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 series aparticular stress that advances in imaging have lead to early diagnosis AND THERE 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 Hepaticoducho 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 care. The cyst excision itself cannot be as safely addressed in laroscopic or Robot assisted, particularly



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 excision 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 endosurgeons is 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 Hepaticoduodenostomy⁽¹⁶⁾ although claims as effective as Roux en Y Common Hepaticoduodenostomy, long term results of Common Hepaticoduodenostomy 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 Hepaticoduodenostomy seems to be the gold standard. Laparoscopic & Robotic approach to inflamed, adherent Choledochal cyst may not be acceptable and safe^(15,16)

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 be validated as safe and can be used as standard surgical option for excision of choledochal cyst.

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