Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

Treatment of Choledochal Cyst in a Pediatric Population: A Case Series of Nine Patients from a Tertiary Care Teaching Medical Institution in Odisha, India

AntaryamiPradhan, BhabagrahiMallick*, SudhanshuSekharpatra
Department of Paediatric, IMS and Sum Hospital, SOA deemed to be University, Bhubaneswar, Odisha, India
Corresponding author
Dr. BhabahrahiMallick, Department of Paediatric, IMS and Sum Hospital, SOA deemed to be University, Bhubaneswar, Odisha, India
Mail Id- bhabagrahimallick@soa.ac.in

Abstract
Choledochal cyst (CD Cysts) are congenital bile duct anomalies wherein the cystic dilatations of the bile duct radicles could be intra or extrahepatic or both. Though rare, about 75% of these are detected in in childhood. If untreated, complications ensue. Definitive treatment includes completed surgical excision of the cyst and reconstruction of the bile duct but there is a lack of consensus on the optimal technique. In this paper, we present a summary of patients with CD cysts operated at our tertiary care centre in Odisha, India, that caters to a relatively under-resourced clientele, over the past six years.We analyzed the clinical profile (diagnosis, treatment and outcomes) of the patients.Wehad operated on nine patients – six males and three females, aged 15 months to 13 years. Eight children had presented with only pain.One child had pain, jaundice and a palpable mass in the right upper quadrant (classic triad). All had Type I biliary tract pathology. We managed seven of the children with hepaticojejunal (HJ) and Jejuno-jejunal (JJ) anastomoses in Roux-en-Y with an average length of stay in the hospital of 11.0±1.4 days, and the remaining two children with isolated jejunal loop (HJ, Jejuno-Duodenal (JD), and JJ) anastomosis with an average length of stay of 13.0±1.0 days. We did not have any complications reported immediately or during follow-up period ranging from month to 1 year. In our series,we recommend that HJ and JJ anastomoses in Roux-en-Y as the most preferred treatment for Type I choledochal cysts.
Key words: Choledochal cyst, hepaticojejunal , cystic dilatations, jejunal loop, pediatric,

Introduction

Choledochal cyst (CD Cyst) is arare congenital anomaly of the biliary tree.(1,2)These are disproportionate intrahepatic and/ or extrahepatic cystic dilatations of the bile duct (BD). CD Cysts are rare in adults and detected more often in children.(3) In Western populations, the incidence of CD cysts ranges from 1 in 1,00,000 to 1 in 1,50,000. Reportedly, the incidence is higher in Asians. CD Cysts are more common in females than in males (ratio: 3-4:1).(4) If untreated, these can lead to bile stasis, and subsequent complications e.g. BD obstructionascending cholangitis, intrahepatic bile duct stones, intrapancreatic terminal choledochal calculi, pancreatic duct calculus, bowel obstruction, pancreatitis, and cholangiocarcinoma(in about 20-30% of the patients by the second decade of life).(1)CD

Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

cysts, in about one in six cases presenting to tertiary care centres, present with a classical triad of jaundice, right upper quadrant mass and abdominal pain.(5) About four of six cases (two-thirds) manage to get diagnosed before 10 year age and five of six by puberty.(3)

CD cysts are of five types as per Todani's classification – Type I: Cystic or fusiform dilatation of hepatic and common bile duct (40%–85%); Type II: Diverticulum of the common bile duct (2%–3%); Type III: Choledochocele (Intraduodenal dilatation of the Common Bile Duct) (1.4%–5.6%); Type IVa: Intra- and extrahepatic bile duct dilatation (18%–20%); and Type V: Intrathepatic bile duct dilatation (Caroli's disease).(6)The most common types i.e., Type I and IV While surgical excision is a definitive treatment there is no consensus on the optimal surgical technique for reconstruction of the BD.(6)The two common reconstruction techniques for the BD include hepaticojejunal (HJ) anastomosis in Roux-en-Y and hepatico-duodenal anastomosis with isolated jejunal loops but neither has an established advantage over the other.(7)

In the pediatric surgery department of our hospital, a tertiary care centre in Odisha, India that caters to a relatively under-resourced clientele, over the past six years, we have had the opportunity of operating on nine children with CD cysts. In this paper, we present our a summary of these cases.

Methods

Our hospital is a 15 year old 1500-bedded tertiary care hospital that provides specialty and super-specialty healthcare services and training. The hospital is located in Bhubaneswar, the capital city of the State of Odisha in India, and caters to a relatively under-resourced population. The hospital has been using a consistent case record format and maintains electronic patient health records with due quality check protocols for completeness and accuracy. ThePediatricSurgery department has a well-trained team of surgeons and undertakes some of the most advanced surgeries in this part of the country. The department was established in 2014.

Using a case series design, we retrospectively retrieved and reviewed records of all the CD cases of CD Cyst operated by the lead author (AP), a mid-career faculty in the Pediatric Surgery Department, between 1st January 2015 and 31st December 2020.We obtained records of nine patients from which we extracted information on age at the time of presentation, sex, clinical features, pathology, type of CD Cyst, surgical technique undertaken, complications, length of hospital stay and duration of follow-up.

The statistical analysis was done using MS Excel (Office 365 package). Categorical variables were summarized as frequency and proportions.The continuous variables were summarized as frequency and mean with standard deviation and/ or median with range, and group comparisons were made using Mann Whitney U test. Statistical significance was considered if p was less than 0.05.

Results

We retrieved andreviewed records ofnine patients– six males (age: 55±53.4 months; median-35 months; minimum-15 months; maximum-156 months i.e., 13 years) and three

Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

females (age: 53±6.2 months; median-48 months; minimum-48 months i.e. 4 years; maximum-60 months i.e., 5 years). While eight children had presented with only pain, there was onlyone child who had pain, jaundice and a palpable mass in the right upper quadrant (classic triad). All had Type I biliary tract pathology. We had conducted open laparotomy with right subcostal roof top incision on six children with an average duration of surgery of approximately 3 hours. On the remaining three children (aged 5 year, 6 year and 13 years), laparoscopic dissection and excision of the CD Cyst but the anastomosis was done externally through a mini-laparotomy incision; each such surgery lasted for about 4 hours (Fig 1,2).

Fig 1: Choledochal cyst operational site

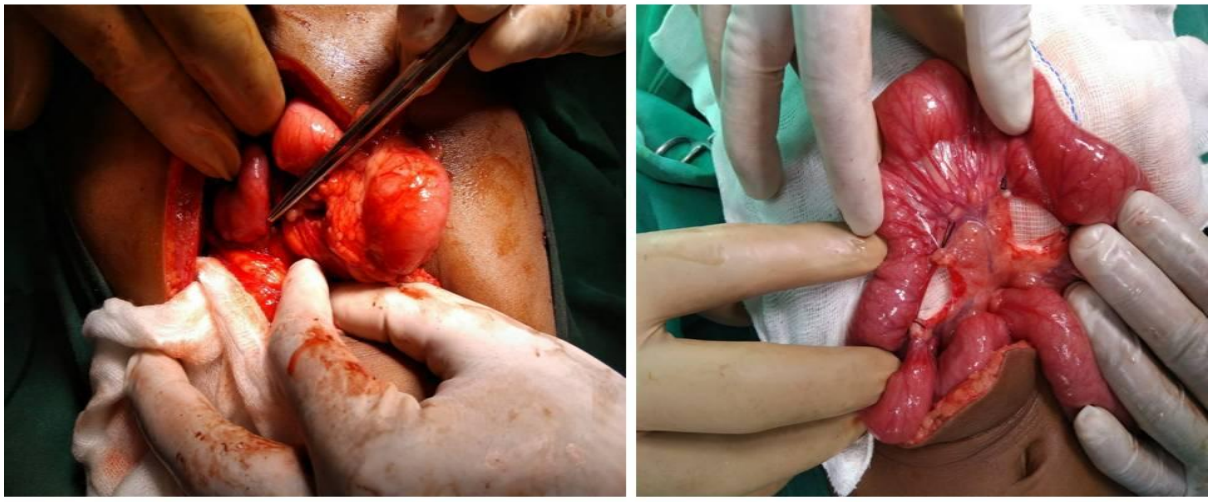

Fig 2: Choledochal cyst with multiple stones visible in the cyst

We managed seven of the children with hepaticojejunal (HJ) and Jejuno-jejunal (JJ) anastomoses in Roux-en-Y with an average length of stay in the hospital of 11.0±1.4 days, and the remaining two children with isolated jejunal loop (HJ, Jejuno-Duodenal (JD), and JJ) anastomosis with an average length of stay of 13.0±1.0 days. The difference in the length of hospital stay was statistically significant (p=0.00042). We did not have any complications reported immediately or during follow-up period ranging from one month to 1 year. (Table 1)

Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

Table 1. Profile of the patients operated for choledochal cyst at the study hospital 1st January 2015 and 31st December 2020

No	Age	Sex	Clinical symptoms at presentation							Pathology		Todani Pre-op	Surgical Technique	Complic-ations	Length of Hospital Stay (in days)	Duration of follow-up (in months)
			Pain	Jaundice	Vomiting	Fever	Hepato-megaly	Palpable mass	Triad	Biliary Tract	Liver					
1	1 year 3 months	Male	Yes	No	No	No	No	No	No	Type I	NR	Type I	Roux-en-Y HJ+JJ (laparotomy)	Nil	10	06
2	4 years 3 months	Female	Yes	Yes	Yes	Yes	No	Yes	Yes	Type I	NR	Roux-en-Y HJ+JJ (laparotomy)	Nil	12	12	Roux-en-Y HJ+JJ (laparotomy)

Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

											roto my)				
3	1 year 6 months	Male	Yes	No	No	No	No	No	No	Type I	NR	Isolated Jejunal loop HJ+ JD+ JJ (laparotomy)	Nil	14	06	Isolated Jejunal loop HJ+JD+JJ (laparotomy)
4	3 years	Male	Yes	No	No	No	No	No	No	Type I	NR	Isolated Jejunal loop HJ+ JD+ JJ (laparotomy)	Nil	12	06	Isolated Jejunal loop HJ+JD+JJ (laparotomy)
5	4 years	Female	Yes	No	No	No	No	No	No	Type I	NR	Roux-en-Y	Nil	10	06	Roux-en-Y HJ+JJ

Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

											HJ+ JJ (laparotomy)				(laparotomy)
6	2 year 10 months	Male	Yes	No	No	No	No	No	No	Type I	NR	Roux-en-Y HJ+ JJ (laparoscopy+)	Nil	10	12	Roux-en-Y HJ+JJ (laparoscopy+)
7	13 years	Male	Yes	No	No	No	No	No	No	Type I	NR	Roux-en-Y HJ+ JJ (laparoscopy+)	Nil	11	03	Roux-en-Y HJ+JJ (laparoscopy+)
8	6 years	Male	Yes	No	No	No	No	No	No	Type I	NR	Roux-en-Y HJ+ JJ	Nil	10	01	Roux-en-Y HJ+JJ (laparoscop

Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

												(laparoscopy+)				y+)
9	5 years	Female	Yes	No	No	No	No	No	No	Type I	NR	Roux-en-Y HJ+JJ (laparotomy)	Nil	14	06	Roux-en-Y HJ+JJ (laparotomy)

Abbreviations: HJ: Hepatico-jejunostomy; JJ: Jejuno-jejunostomy; JD: Jejuno-duodenostomy; laparoscopy+: laparoscopic dissection and excision of the CD Cyst with anastomosis done externally through a mini-laparotomy incision

Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

Discussion

Between 2015 and 2020, the lead author has conducted about 500surgeries and 14500 out-patient consultations in the PediatricSurgery department of our hospital. However, we noted that only nine children with CD Cyst had presented to usthus confirming the rarity of this condition. As established in literature,(6–8) we noted that Todani Type I CD Cysts were perhaps the most common type; this was the only type our patients had and we could manage all of them successfully with HJ and JJ anastomoses in Roux-en-Y in most cases. The length of stay in the hospital was mostly less than 2 weeks. Even as CD cysts have been reported to be more common in females by previous research,(4) we found them more common in our study. This could be a chance finding due to the small sample size of our study. Alternatively, this could be due to gender-based differences in careseeking patterns.

The classical clinical triad of jaundice, pain, and a palpable mass is usually found in less than 17-20% of patients with CD Cyst, and majorly in children.(3,5)In our case series, only one child i.e., 11.1% (95% CI: 0.3-48.2) presented to us with the triad. Pain abdomen was the most common presenting feature; all nine children had this. In our cases series, we did sevenHJ and JJ in Roux-en-Y, and two Isolated Jejunal loop HJ+JD+JJ.None of the patients had any complications during the study follow-up which mostly extended to at least 6 months. The length of hospital stay was lesser for those undergoing HJ and JJ in Roux-en-Y suggesting that this could be a favourable procedure as compared to isolated jenjunal loop HJ+JD+JJ.

We conducted laparotomy on most of the patients. Laparoscopic approaches may reduce the length of hospital stay even further.(8) However, undertaking laparoscopic procedures in pediatric patients requires both infrastructure and expertise failing which one may experience intraoperative as well as postoperative complications.(9,10)

The key limitations of our study are its retrospective design for which we had to limit our analysis to only a limited number of variables which were available with high quality. Our study reports findings from just nine patients and with limited duration of follow-up. However, being a synthesis of cases operated by a single surgeon in a single institution, we do not anticipate any differences in the evaluations and surgical care of the patients, and hence, do not foresee any bias in the study findings due to these.

Conclusion

In patients with CD Cyst, complete excision of the cyst is recommended but it has not been established which surgical technique is optimal for biliary tract reconstruction.Based on this study, we recommend that HJ and JJ anastomoses in Roux-en-Y as the most preferred treatment for Type I choledochal cysts. We also recommend for larger cohort of these patients be built and followed-up for more general inferences.

****

Figure Legends

1. Choledochal cyst operational site

Annals of R.S.C.B., ISSN:1583-6258, Vol. 25, Issue 4, 2021, Pages. 14391 - 14399
Received 05 March 2021; Accepted 01 April 2021.

2. Choledochal cyst with multiple stones visible in the cyst

Reference

1. Joseph V, Prema Raj J. A review of choledochal cyst in pediatric and adult patients. J Hep Bil Pancr Surg. 1996;3:396–404.
2. Banks JS, Saigal G, D'Alonzo JM, Bastos MD, Nguyen NV. Choledochal Malformations: Surgical Implications of Radiologic Findings. Am J Roentgenol. 2018 Feb 15;210(4):748–60.
3. Silva-Baez H, Coello-Ramírez P, Ixtabalán-Escalante EM, Sotelo-Anaya E, Gallo-Morales M, Cordero-Estrada E, et al. Treatment of choledochal cyst in a pediatric population. A single institution experience of 15-years.Case series. Ann Med Surg. 2015 Dec 21;5:81–5.
4. Baison GN, Bonds MM, Helton WS, Kozarek RA. Choledochal cysts: Similarities and differences between Asian and Western countries. World J Gastroenterol. 2019 Jul 14;25(26):3334–43.
5. Machado NO, Chopra PJ, Al-Zadjali A, Younas S. Choledochal Cyst in Adults: Etiopathogenesis, Presentation, Management, and Outcome—Case Series and Review. Gastroenterol Res Pract. 2015 Jul 15;2015:e602591.
6. Dumitrascu T, Lupescu I, Ionescu M. The Todani classification for bile duct cysts: an overview. Acta Chir Belg. 2012 Oct;112(5):340–5.
7. Kumar S, Chandra A, Chopra N, Rahul, Gupta V, Joshi P, et al. A Single-Center Experience with 73 Patients of Adult Choledochal Cyst: An Indian Perspective. Trop Gastroenterol. 2019 May 19;39(3):124–34.
8. Tang S-T, Yang Y, Wang Y, Mao Y-Z, Li S-W, Tong Q-S, et al. Laparoscopic choledochal cyst excision, hepaticojejunostomy, and extracorporeal Roux-en-Y anastomosis: a technical skill and intermediate-term report in 62 cases. Surg Endosc. 2011 Feb;25(2):416–22.
9. Perugini RA, Callery MP. Complications of laparoscopic surgery [Internet]. Surgical Treatment: Evidence-Based and Problem-Oriented. Zuckschwerdt; 2001 [cited 2021 Apr 11]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK6923/
10. Qu X, Cui L, Xu J. Laparoscopic Surgery in the treatment of children with Choledochal Cyst. Pak J Med Sci. 2019;35(3):807–11.