IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 24, Issue 12 Ser. 2 (December. 2025), PP 73-76
www.iosrjournals.org

Innovative Porto-Cysto-Enterostomy For Portal Cyst In Cystic Biliary Atresia As An Effective Modification During Kasai's Operation

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Abstract:
Background*: Biliary atresia (BA) is a common surgical cause of jaundice in infancy, characterized by progressive obliterative cholangiopathy. Cystic biliary atresia (CBA) a variant of BA, with relatively good prognosis. The cyst in CBA can be located anywhere along the atretic biliary tree, treated with excision of atretic biliary tree along with cyst and porto enterostomy.*
Clinical Description*: There were earlier reports of cysto enterostomy, where they found cyst with patent proximal biliary tree. We report a rare case of CBA in an infant, where a cyst was found at the porta hepatis adjacent to the atretic portal plate.*
Management & Outcome*: Intra operatively the cyst was found to have bile suggesting cysto biliary communication. Hence the cyst was de-roofed and portal plate was dissected. Enteric anastomosis was done by including the de-roofed cyst and the opened liver parenchyma beneath the portal plate in continuum, which can be termed as Cysto porto enterostomy. The child had uneventful recovery.*
Conclusion: *Whenever we encounter a cyst at porta with bile as content, which indicative of an occult cysto biliary communication, preservation and inclusion of cyst in continuity with porto enterostomy can be a viable option for additional bile drainage apart from the dissected portal plate*
Key Word: *Cystic Biliary Atresia, Portal cyst, Portocysto enterostomy.*

Date of Submission: 02-12-2025 Date of Acceptance: 12-12-2025

I. Introduction

Biliary atresia (BA) is a progressive obliterative disease of biliary tree with progressive jaundice in infancy, which if untreated, will lead to chronic hepatic dysfunction.[1] About 5-10 percent of biliary atresia will have cystic lesion anywhere along the atretic biliary tree and they are termed as cystic biliary atresia (CBA) which tends to have a better prognosis.[2] CBA needs to be differentiated from choledochal cyst (CDC) since the treatment of the CBA needs to done at the earliest possible to minimize the liver dysfunction. Surgical treatment for cystic biliary atresia is no different from traditional Kasai's porto enterostomy (KPE) for Biliary atresia and it involves excision of atretic biliary tree along with the cyst irrespective of its position. We report a case of cystic biliary atresia in which the cyst was used as a part of bilio-enteric anastomosis which can be a viable option in select cases.

II. Clinical Description

A 4 months old male infant presented with features of progressive jaundice from newborn period with failed medical management in the outside hospital. On evaluation he was found to have direct hyperbilirubinemia with ultrasonography (USG) suggestive of a 1.0x0.8x0.5cm choledochal cyst or simple hepatic cyst at the porta hepatis (Figure 1A) whereas magnetic resonance cholangio pancreato gram (MRCP) was suggesting a cystic biliary atresia (Figure 1B) which was further confirmed by a Hydroxy imino diacetic acid (HIDA) scan (Figure 1C).

III. Management And Outcome

The child was taken up for laparotomy in which there was atretic gall bladder with minimal white bile and intra-operative cholangiogram delineated only gall bladder without further flow distally (Figure 1D). Liver surface showed features of biliary stasis. Hence, we proceeded with KPE. On dissection of porta, we came across the cystic lesion which was mentioned in the pre-operative imaging (Figure 2). The cyst had dark green bile as content which indicates there is a cysto-biliary communication (CBC), though we were not able to visualize it macroscopically. Hence the decision was made not to excise the cyst, but to use the CBC as a

conduit for post-operative biliary drainage. Extrahepatic-component of the cyst was de-roofed and portal plate was excised as in traditional Kasai's procedure. Since the residual doom of cyst wall and the porta are in continuum as depicted in hand drawn image in figure 3, they were taken in toto as a part of porto-enterostomy, which can be termed as porto-cysto-enterostomy. Peri-operative medications were given as per our routine protocol for KPE. The child passed Bilious stools on 3rd post-operative day and recovered well. He is thriving well with normal liver functions on 6th month follow up.

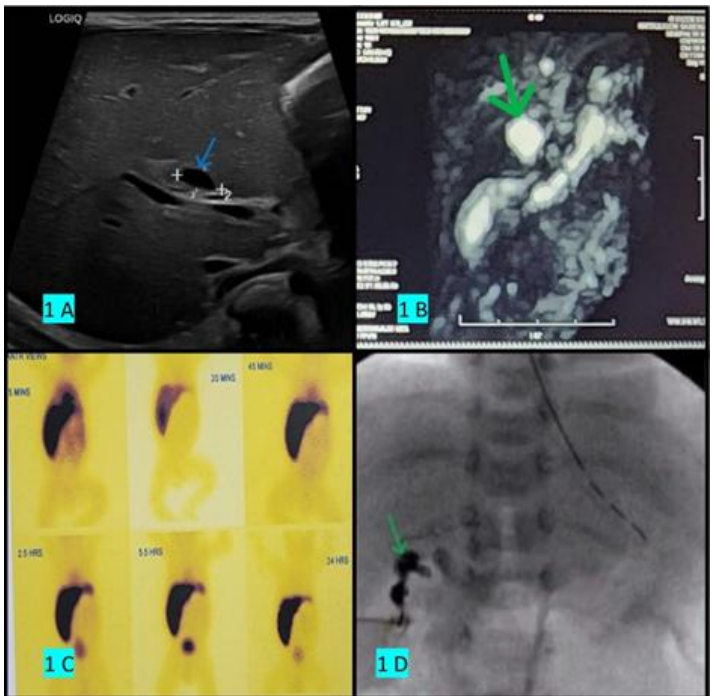

Figure 1A: Ultrasonogram showing the portal cyst (blue arrow)
Figure 1B: Magnetic Resonance Imaging showing the portal cyst (green arrow)
Figure 1C: HIDA scan showing absent tracer in duodenum
Figure 1D: Intra operative cholangiogram showing only gall bladder (green arrow)

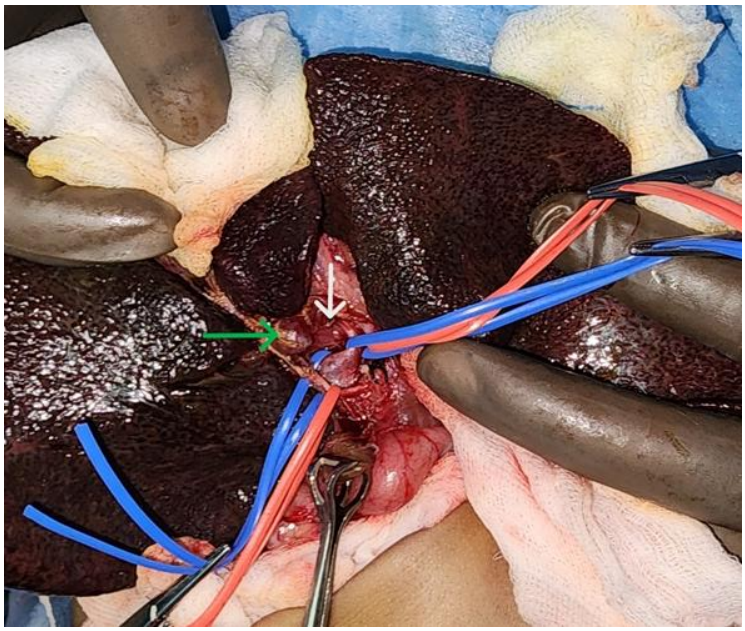

Figure 2: Intra operative image showing atretic portal plate (white arrow) and the portal cyst (green arrow)

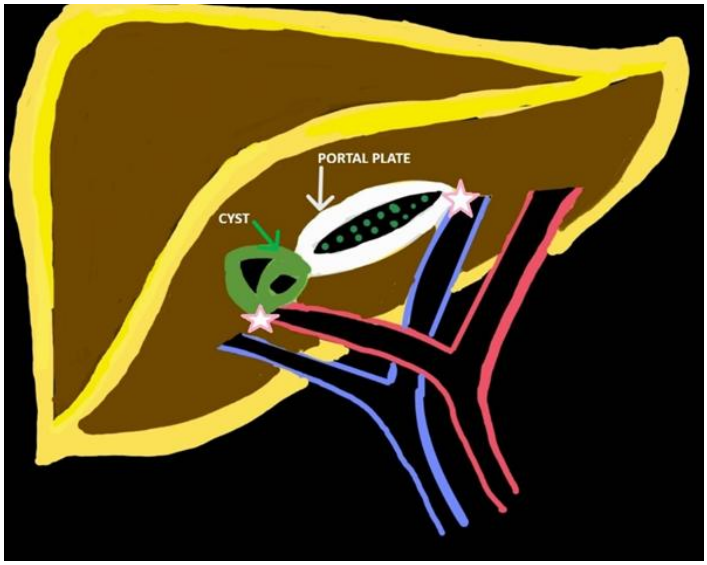

Figure 3: Hand drawn diagram showing the intra operative finding. The area marked with star depicts the right and left extent of anastomosis with the bowel

IV. Discussion

Biliary atresia (BA) is the most common cause of obstructive jaundice in first 12 weeks of life, which if left untreated, leads to progressive liver dysfunction in early childhood.[2] Prolonged neonatal jaundice with clay-colored stools and high colored urine should always raise the suspicion of BA, since earlier the intervention, better is the prognosis. CBA being a variant of BA hold better prognosis with timely surgical management.[3] BA is diagnosed in a cholestatic jaundice by absent or atretic gallbladder in USG with a triangular cord sign. MRCP has limited role in BA unlike in CDC where the stagnant bile in the dilated system will delineate the anatomy.[4] However MRCP can demonstrate the anatomy of cyst in CBA, hence it can be used for diagnosis of CBA in conjunction with HIDA scan. Since MRCP is not routinely indicated in suspected biliary atresia, the exceptional indication should be considered whenever USG shows a cyst in hilar region with a triangular cord sign and/or atretic gallbladder.[3] Calinescu et al reported antenatal diagnosis of hilar cysts which were either CBC or CBA in post-natal evaluation and the later was treated with excision of cyst and KPE since there was no continuity established with liver.[5] There has been older reports of Hepaticojejunostomy(HJ) being done for CBA with significant proportion of them becoming jaundice free post procedure. Some of them needed KPE in the follow up. All those with CBA underwent HJ since there was definite luminal availability at and above the level of common hepatic duct (CHD) though the tract was atretic below that (Table 1), which was not the scenario in our case where there was complete atretic tree with fibrosed portal plate.[2,5,6,7,8,9,10,11,12,13]

Reported by	Sample size	Procedure	Any further surgery (YES/NO/NOT AVAILABLE-NA)	RESULT
Tsuchida Y et al	1	HJ	NA	Anicteric
Matsubara H et al	1	HJ	NA	Anicteric
Komuro H et al	4	HJ	1-LIVER TRANSPLANT (LT)	3-Anicteric
Nio M et al	22	HJ	3- KPE	19- Anicteric
Caponcelli E et al	1	HJ	NA	Anicteric
Takahashi Y et al	12	HJ	1-KPE 2-LT	9- Anicteric
Faure A et al	3	Laparoscopic HJ	NA	2- Anicteric
Lal R et al	2	HJ	NA	2- Anicteric
Ji Y et al	27	Laparoscopic HJ	NA	25- Anicteric
Calinescu AM et al	1	HJ	NA	Anicteric
Sundaram J et al (Index case)	1	PORTO CYSTO JEJUNOSTOMY	NO	Anicteric

Table 1: Previous reports of Cystic Biliary Atresia with surgical techniques (HJ- Hepaticojejunostomy, KPE- Kasai's porto enterostomy)

V. Conclusion

Authors suggest that, it is always imperative to look for the content of portal cysts in case of cystic biliary atresia to rule out cystobiliary communication, before attempting to excise it from liver parenchyma. If the cyst contains bile, it is advisable to include the cyst as a part of bilio- enteric anastomosis, to increase the

bile flow after the Kasai's procedure. This can be a viable option in such a clinical scenario rather than doing excision of cyst with only porto enterostomy or only cysto enterostomy without dissection of portal plate.

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