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ASSOCIATION OF BILIARY ATRESIA WITH SMALL BOWEL ATRESIA AND SHORT BOWEL SYNDROME

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ABSTRACT

We are going to discuss A case report and review articles for developing treatment algorithm, options and complications in patient who have both anomalies simultaneously. Itsbidirectional effect on surgical and medical options in the treatment.

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INTRODUCTION

Biliary Atresia (BA) is a disorder of infancy characterized by the progressive obliteration of the extra- and intra hepatic biliary duct system leading to the obstruction of bile flow, but the causes remain in doubt. However, several associated factors, such as developmental malformations, perinatal viremia, toxicity of bile constituents, and anatomic abnormalities in the hepatobiliary system, have been proposed as causative factors (Davenport, 1993; Morecki, 1982; Jenner, 1978 and Miyano, 1979). Biliary atresia (Fig-1) has been implicated to be the result of a perinatal event, caused by some injurious agent, which results in fibrosis of extrahepatic bile ducts (Bill, 1977 and Landing, 1974).

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Etiology and Pathogenesis

The underlying cause or trigger of BA remains unknown. Several mechanisms have been hypothesized and detailed in several recent excellent reviews. These include viral, toxin-induced, vascular, genetic predisposition, abnormal morphogenesis/development, and autoimmune/ immune dysregulation. The association between biliary atresia and small bowel atresia has been reported by several investigators (Ohi, 1983 CLR Fête, 1983 and Kishida, 1988). *Oh et al* reported that 1 patient (0.5%) in 214 with biliary atresia was operated on because of meconium peritonitis from a perforation associated with jejunal atresia. *According to the Japanese Biliary Atresia Registry* between 1989 and 1999, 6 patients (0.5%) in 1,198 with biliary atresia were associated with small bowel atresia perforation complex (Ohi, 1983).

Case: A 2 days /old boy weighing 2100kg was delivered after 37 weeks' gestation by Normal Delivery in rural hospital. Brought to our Clinic because of bilious vomiting and

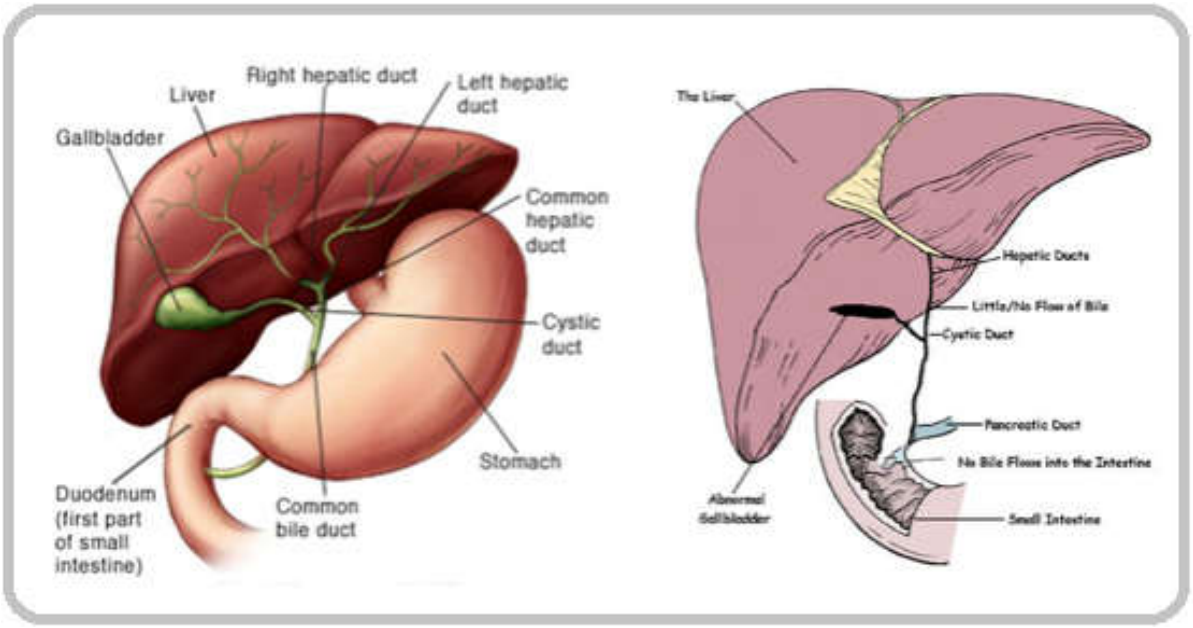

Fig. 1. Normal and biliary atresia

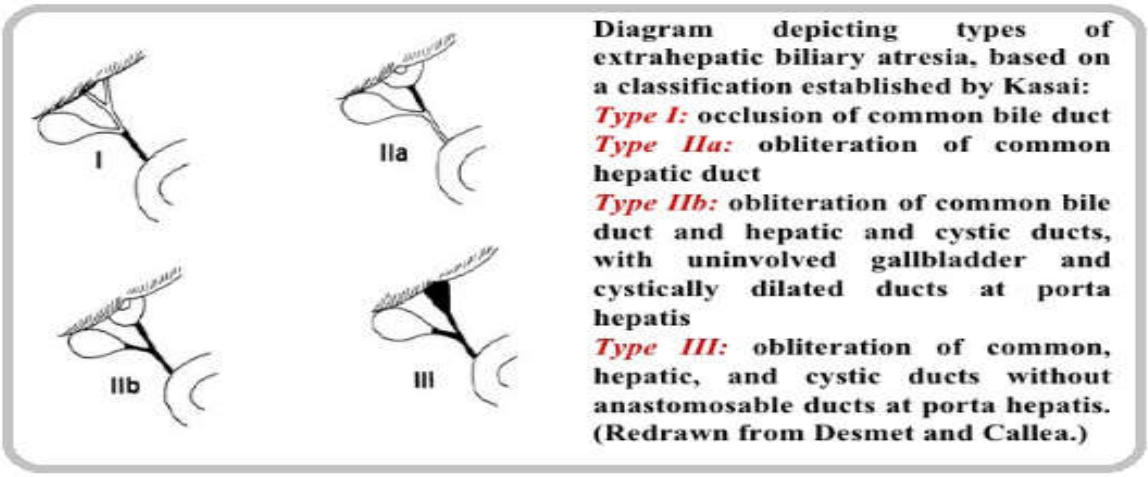

Fig. 2. Kasai classification BA

abdominal distension. The X-rays (up right and flat) showed thumb sign (large air fluid level in upper abdomen and B/E (shoed disused colon) both in favor of small bowel atresia. Laparotomy was performed on the second day of birth during which adhesion and 10 cm proximal jejunal pouch and a matted mass of small bowel with intact remaining 10 cm of terminal ileum was found. Irrigation and peritoneal toilet with anastomosis between proximal and distal bowel segments were performed. After the first operation, his stools were at first normally colored, but his stools became progressively colorless, and jaundice deepened, primarily caused by conjugated bilirubin, and this component remained high despite the cessation of parenteral alimentation. The patient underwent workup with suspicion to biliary atresia. Abdominal ultrasonography did not show any of an extrahepatic bile duct, and a HIDA scan did not show any bile excretion into the bowel. A second laparotomy was performed at 26 days of age. Operative cholangiography confirmed the diagnosis of biliary atresia (Type B). Fig. 2. Biliary appendico-duodenostomy were performed because of inadequate length of bowel for performing roux en y anastomosis. The baby showed transient bile flow after biliary appendico-duodenostomy, which stopped suddenly at 30 days of age which managed by blast corticosteroid therapy. Parenteral alimentation was necessary because of short bowel syndrome. The baby suffered from wound infection and short bowel syndrome manifestations. The symptoms related to short bowel syndrome progressively resolved, and for over 2 months,he kept on oral antibiotic and prednisone 2mg/kg with no dietary restrictions. He is under follow up and recent tests indicated that his bilirubin level is normal but, his liver enzymes are mildly elevated.

DISCUSSION

There are arguments that seemingly contradict this early maldevelopment hypothesis. More recently, the consensus opinion is that small bowel atresia is caused by an ischemic lesion that occurs during the later stage of pregnancy (De Lorimier, 1969). Biliary atresia may be related to a dynamic, acquired inflammatory process associated with meconium peritonitis that starts late in utero, and progresses post-Nataly. The meconium peritonitis-induced inflammation of adjacent periductal tissue which lead to a protracted inflammatory reactions and fibrosis, causing secondary obliteration of the extrahepatic biliary system.

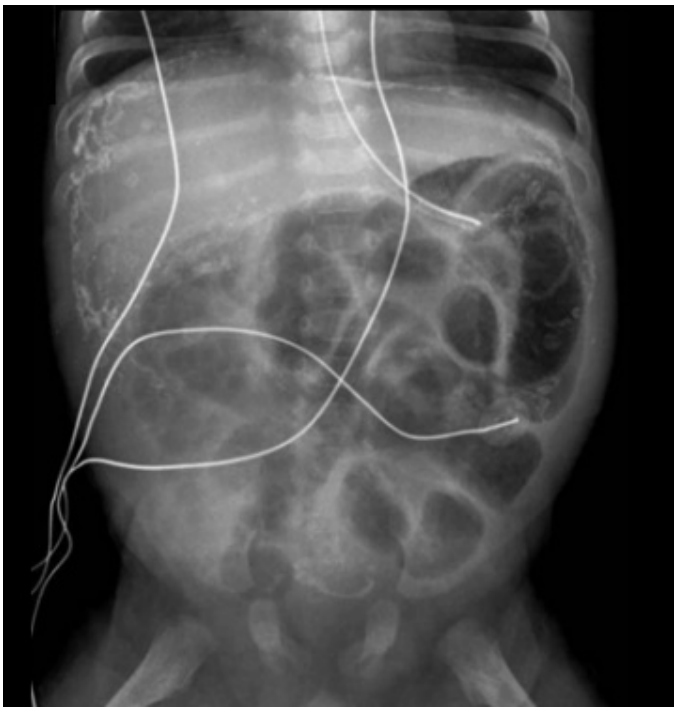

Fig. 3. The meconium peritonitis

Jaundice in small bowel atresia is not unusual because 20% to 30% of small bowel atresia cases also are associated with nonhemolytic jaundice (De Lorimier, 1986). The diagnosis of meconium peritonitis and perforation of small bowel atresia usually made in all cases at the first glance, but this is not the same thing for accompanying biliary atresia. The gallbladder was not noticed in all cases during the first operation and preoperative ultrasonography did not include details of the gallbladder. Because of the high incidence of biliary atresia in cases of meconium peritonitis, we recommended that the gallbladder should be examined by preoperative ultrasonography and during the operation for small bowel atresia. The etiology of persistent jaundice and acholic stool after the first operation for meconium peritonitis also is difficult to diagnose due to the duration of hyperalimentation. Central hyperalimentation usually is required for nutritional support in most cases of meconium peritonitis with intestinal atresia because of the combined effects of short bowel syndrome and the delayed return of adequate motility and absorptive capacity. The ascending cholangitis is more common in these cases of combined anomalies (De Lorimier, 1969). The option for biliary drainage in these patients are also problematic because of accompanying inadequate length of bowel for reconstruction. In 1971, *Grosfeld et al* first reported the use of appendiceal graft for biliary reconstruction. Since then, various techniques which include utilizing the appendix as a biliaryconduit have been introduced with different results. (Greenholz, 1989 and Crombleholme, 1989). *Crombleholme et al*,[12] described use of biliary appendico-duodenostomy as a non-refluxing conduit for biliary reconstruction. Appendico-duodenostomy (Crombleholme, 1989), was used to prevent reflux cholangitis and save the entire small bowel.

Infants with BA can be grouped into 3 categories (Schwarz, 2013)

BA without major malformations – sometimes referred to as isolated, nonsyndromic or perinatal BA (without major malformations), this pattern occurs in ~80-85% of infants with BA.

BA in association with laterality malformations – also known as syndromic biliary atresia, fetal-embryonal biliary atresia, or Biliary Atresia Splenic Malformation (BASM), this pattern occurs in about 10% of infants with BA and includes the clinical presence of laterality defects.

BA in association with at least one major malformation – this pattern occurs in about 5-10% of infants with BA who have associated anomalies most commonly manifested in the cardiovascular (71%), genitourinary (47%) and gastrointestinal (24%) systems and without laterality defects. Despite a lack of experience with thisprocedure, we think that it can be used in this special situation. We believe that bacterial overgrowth in the small bowel, a well-recognized problem in children with short bowel syndrome, may be another risk factor of ascending cholangitis. Another area of concern involves the management of short bowel syndrome in biliary atresia, which requires extended TPN, and has been shown to increase the risk of cholestasis leading to liver cirrhosis, sepsis, and increased risk of mortality it is difficult to manage the nutritional requirements of patients and to prevent ascending cholangitis. The combination of these major problems is associated with poor outcome in these patients. Although we are not able to detail the ideal management but Appendico-duodenostomy is the last bridge to transplantation. We recommend this procedure simultaneous with hyperalimentation to make child ready for liver or possibly multiple visceral transplantation.

Disclosure: The authors declare no conflicts of interest.

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REFERENCES

Bill, A.H., Haas, J.E. and Foster, G.L. 1977. Biliary atresia: Histopathologic observations and reflections upon its natural history. *J Pediatr Surg., 12.,* pp. 977–982.

CLR Fête, A Cuendet and J Berclaz, 1983. An unusual association of small bowel atresia and biliary atresia: A case report. *J Pediatr Surg.,* 18, pp. 136–137.

Crombleholme T.M., Harrison, M.R., Langer, J.C., *et al*: Biliaryappendico-duodenostomy: A nonrefluxing conduit for biliary reconstruction. *J Pediatr Surg*., 24:665-667, 1989

Davenport, M., Savage, M., Mowat, A.P., *et al.* 1993. The biliary atresia splenic malformation syndrome. *Surgery*113 pp. 662–668.

De Lorimier, E.W. Fonkalsrud and D.M. Hays, Congenital atresia and stenosis of the jejunum and ileum. Surgery65 (1969), pp. 819–827.

Delarue, A., Chappuis, J.P., Esposito, C. *et al*. 2000. Is the appendix graftsuitable for routine biliary surgery in children? *J Pediatr Surg.,* 35:1312-1316.

Greenholz, S.K., Lilly, J.R., Shikes, R.H., *et al*: 1986. Biliary atresia in thenewborn. *J Pediatr Surg.,* 21:1147-1148.

Jenner, R.E. 1978. New perspectives on biliary atresia. Ann R *Coll Surg Eng.,* 60., pp. 367–374.

Kishida, Y, Ito, T., Nagaya, M., *et al*., 1988. Three cases of biliary atresia associated with small bowel atresia. Ann Rep Nagoya Univ Br Hosp22., pp. 17–23.

Landing, B. H. 1974. Consideration of the pathogenesis of neonatal hepatitis, biliary atresia and choledochal cyst. The concept of infantile obstructive cholangiopathy. *Prog PediatrSurg6.,* pp. 113–139.

Miyano, T., Suruga, K., and Suda, K, 1979. Abnormal choledochopancreaticoduodenal junction related to etiology of infantile obstructive jaundice diseases. *J PediatrSurg.,* , pp. 16–26.

Ohi, R., Koike, N., Hanamatsu, M, *et al*., 1983. Associated anomalies in infants with hepatobiliary diseases. *Jpn J PediatrSurg*, pp. 595–601.

R Morecki, JH Glaser, WF Balisteri*et al*., Biliary atresia and reovirus type 3 infection. N Engl J Med307 (1982), pp. 481–484.

Schwarz KB, Haber BH, Rosenthal P, Mack CL, Moore J, Bove K *et al*. 2013. Extrahepatic anomalies in infants with biliary atresia: results of a large prospective North American multicenter study. *Hepatology.,* 58(5):1724-1731

TM Crombleholme, MR Harrison, JC Langer *et al*. 1986. Biliary appendico-duodenostomy: A nonrefluxing conduit for biliary reconstruction. *J PediatrSurg*., 24, pp. 665–667.

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