Indian Journal of Pediatrics (July 2021) 88(7):639–640
https://doi.org/10.1007/s12098-021-03814-5

EDITORIAL COMMENTARY

"Cyst at Porta" in Infants with Cholestatic Jaundice: The Time to Act Is Now

Rishi Bolia[1] · Gautham Pai[2]

Received: 6 May 2021 / Accepted: 12 May 2021 / Published online: 26 May 2021

Biliary atresia (BA) is a fibro-obliterative disease of the intrahepatic and extrahepatic bile ducts. Diverse presentations of BA including those that present with syndromic features associated with splenic malformation, cystic dilatation of the biliary tree, CMV IgM positive–associated BA, and isolated BA have been recognized. However, despite this diversity, all BA forms are united by a key feature that they have obliterative cholangiopathy that affects varying lengths of both intrahepatic and extrahepatic bile ducts.

Cystic BA (cBA) is defined as a cystic expansion of obliterated biliary tract remnants and has been reported in ~5%–22.4% BA [1, 2]. The cyst is a sequestration cyst which typically does not have a communication with the biliary tree. It lacks an epithelial layer and has little or no inflammation. Its inner wall has a cicatricial layer with a zone of myofibroblastic hyperplasia. The cyst itself may contain bile implying that it is formed after the continuity between intrahepatic and extrahepatic bile ducts has been established at around 10–12-wk gestation [3]. Lobeck et al. examined sections of proximal biliary remnants in children with cBA and found that the bile duct injury mimicked that of children with BA without a cyst (noncBA) suggesting that the histogenesis of noncBA and cBA is similar [4]. So it is conceivable that children with BA with or without a cyst should have similar outcomes. This notion is supported by the study by Shan et al. published in this issue of the Journal. The authors demonstrated that children with cBA fared similar to the propensity score matched noncBA controls [5].

In their paper, the authors have also described a cohort (n = 8) where the cyst communicates with the rudimentary gall bladder (GB) and has a poorer prognosis. The cyst–GB communication is an aberrant manifestation of stromal proliferation and there does not seem to be a pathophysiological explanation as to why these children should fare any differently. As the numbers are small this may represent a type-II error.

Cystic BA is the only type of BA that can be detected by an antenatal scan. Caponcelli et al. found that 41% children with cBA in their cohort were identified on antenatal scan, which translated into early portoenterostomy (median 36 d) and better outcomes than those with noncBA [6]. However, when compared to those operated at a similar age as in the study by Shan et al., the outcomes between cBA and noncBA are comparable [5]. So in a child with cBA, it is important not to be complacent and proceed for portoenterostomy at the earliest as one would do for noncBA.

Cystic BA is often confused with choledochal cysts (CC) leading to a delay in their diagnosis and management. Certain parameters on imaging—a small cyst size, triangular cord sign, gallbladder mucosal irregularity etc. have been suggested to favor the diagnosis of cBA [1]. However, there are a number of exceptions to these findings, often precluding a conclusive diagnosis. Cystic BA can be definitively differentiated from a CC only by an intraoperative cholangiogram with well-formed (often dilated) intrahepatic bile ducts corroborating the diagnosis of CC. Being a pan-ductal disease, cBA will have hypoplastic and irregular intrahepatic bile ducts [2].

The bottomline is that the prognosis of children with cBA and noncBA are comparable and it is important to differentiate cBA from a CC when evaluating a cholestatic child with a hilar cyst. A "cyst at porta" in a child with cholestatic jaundice should prompt an urgent intraoperative cholangiogram followed by timely and appropriate surgery. Any delay will lead to poor outcomes.

✉ Rishi Bolia
rishibolia@yahoo.co.in

[1] Division of Pediatric Gastroenterology, Department of Pediatrics, All India Institute of Medical Sciences, Virbhadra Road, Rishikesh, Uttarakhand 249 203, India

[2] Department of Pediatrics, Yenepoya Medical College, Mangalore, Karnataka, India

Declarations

Conflict of Interest None.

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