Biliary Atresia: Current Trends in Outcome and Management

KRISHNA KUMAR GOVINDARAJAN
From Department of Pediatric Surgery, JIPMER, Puducherry, India.
sasisang@rediffmail.com

Biliary atresia ranks the foremost surgical condition among the various disorders causing neonatal cholestasis. Although its etiology is not entirely clear, various offending agents have been described. Its presentation can be of two types: perinatal and embryonic. The perinatal type – believed to be due to altered interactions between viral agents and immune responses – is commoner (80%), whereas the embryonic type is associated with gene mutations concerning the development of the biliary tree such as Biliary Atresia Splenic Malformation (BASM) [1].

Detailed workup is essential to rule out the nonsurgical conditions, and as such biliary atresia is a disease of exclusion. Sonography is a useful initial investigation, followed by nuclear scintigraphy and intra-operative cholangiography. The fibrotic extrahepatic bile ductal apparatus, seen as echogenic triangular cord cranial to the portal vein bifurcation (triangular cord sign) along with a small (<1.5 cm), non-contractile gall bladder (confirmed on a 4 hour fasting scan) with visualization of hepatic subcapsular flow constitute the sonographic signs of biliary atresia [1]. Histological parameters such as ductal diameter, fibrosis, cirrhosis and presence of ductal plate malformation have been noted to be useful in the prediction of long-term outcome [2].

Early surgery (within 60 days of age) has been proposed as a good prognostic feature in several studies. However, surgery as late as 4 months of age has also been proven to lead to jaundice clearance [3]. Age as a factor has been linked to the etiology such as BASM and cystic type biliary atresia, whereas in the perinatal form, age does not seem to be associated with the outcome [4]. Scoring systems have been developed to predict the outcome after surgery, but lack universal application and wide usage [5]. Utility of post-operative scintigraphic clearance and serum bilirubin level at six weeks as an early predictor for outcome has been demonstrated in the literature [6]. Serum alanine transferase and serum direct bilirubin at 2 months after surgery have also been shown to predict the long-term outcome [7].

The research paper by Redkar, *et al*. [8], published in the current issue of *Indian Pediatrics*, assumes importance as it is based on a single surgeon's experience over a long duration of 14 years. The study lends support to the proposition that the jaundice clearance rate at 3 months is important as an early marker in the long-term outcome evaluation of children operated for biliary atresia. The unpredictability of the surgical outcome is evident, as the authors have acknowledged marginal success of long-term survival with native liver, even after a successful Kasai procedure. However, primary liver transplant is not a practically feasible option, as yet. At present, Kasai procedure is recommended as the initial surgical management of biliary atresia, even when the presentation is late. The scarcity of organs, financial constraints, and life-long requirement of immunosuppressant as part of transplant protocol are limiting factors, in addition to the challenging surgical technicalities behind undertaking transplant in a younger infant.

Funding: None; *Competing interest*: None stated.

REFERENCES

1. Govindarajan KK. Biliary atresia: Where do we stand now? World J Hepatol. 2016;8:1593-1601.
2. Mukhopadhyay SG, Roy P, Chatterjee U, Datta C, Banerjee M, Banerjee S, *et al*. A histopathological study of liver and biliary remnants in the long-term survivors (>10 years) of cases of biliary atresia. Indian J Pathol Microbiol. 2014;57:380-5.
3. Schoen BT, Lee H, Sullivan K, Ricketts RR. The Kasai portoenterostomy: When is it too late? J Pediatr Surg. 2001;36:97-9.
4. Davenport M, Caponcelli E, Livesey E, Hadzic N, Howard E. Surgical outcome in biliary atresia: Etiology affects the influence of age at surgery. Ann Surg. 2008;247:694-8.
5. Zhen C, Guoliang Q, Lishuang M, Zhen Z, Chen W, Jun Z, *et al*. Design and validation of an early scoring system for predicting early outcomes of type III biliary atresia after Kasai's operation. Pediatr Surg Int. 2015;31:535-42.
6. Rodeck B, Becker AC, Gratz KF, Petersen C. Early predictors of success of Kasai operation in children with biliary atresia. Eur J Pediatr Surg. 2007;17:308-12.
7. Goda T, Kawahara H, Kubota A, Hirano K, Umeda S, Tani G, *et al*. The most reliable early predictors of outcome in patients with biliary atresia after Kasai's operation. J Pediatr Surg. 2013;48:2373-7.
8. Redkar R, Karkera PJ, Raj V, Bangar A, Hathiramani V, Krishnan J. Outcome of biliary atresia after Kasai's portoenterostomy: A 15-year experience. Indian Pediatr. 2017;54:291-4.