

such as Progressive familial intrahepatic cholestasis (PFIC). A combination of PFIC and biliary atresia is unusual, but worthwhile considering as a possibility.

The most important question in both the cases is about timing of portoenterostomy. Surgery was undertaken when the children were more than 6 months old. Chardot, *et al.* showed that, the success of portoenterostomy is practically nil after 141 days [4]. In the absence of liver transplantation facility surgery one could argue for late surgery, as it might work in the absence of severe cirrhosis with stable synthetic liver function and give patients some survival advantage. The author could put forward the above mentioned argument for the second case, but for the first case, we are intrigued by the fact that the child underwent portoenterostomy when he/she was in frank liver failure, where Kasai's procedure is contraindicated [5].

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REFERENCES

1. Mohanty S, Shah I, Bhatnagar S. Evolving biliary atresia with cytomegalovirus. *Indian Pediatr.* 2011;48:644-6.
2. Rauschenfels S, Krassmann M, Al-Masri AN, Verhagen W, Leonhardt J, Kuebler JF, *et al.* Incidence of hepatotropic viruses in biliary atresia. *Eur J Pediatr.* 2009;4:469-76.
3. Hartley JL, Davenport M, Kelly DA. Biliary atresia. *Lancet.* 2009;14:1704-13.
4. Chardot C, Carton M, Spire-Bendelac N, Le Pommelet C, Golmard J, Reding R, *et al.* Is the Kasai operation still indicated in children older than 3 months diagnosed with biliary atresia? *J Pediatr.* 2001;2:224-8.
5. Engelmann G, Schmidt J, Oh J, Lenhartz H, Wenning D, Teufel U, *et al.* Indications for pediatric liver transplantation. Data from the Heidelberg pediatric liver transplantation program. *Nephrol Dial Transplant.* 2007;22:viii23-8.

REPLY

We thank Shanmugam and Jayanthi for raising questions that are important while managing children with biliary atresia. In both our patients, we had children who progressed to biliary atresia and had simultaneous active CMV infection. Whether CMV was the cause of this progression remains unknown as we have not been able to depict CMV in the liver tissue. CMV infection has been

found in a large number of extra-hepatic biliary atresia (EHBA) cases [1,2]. However, etiopathogenesis of infection and EHBA still remains little understood and till then it cannot be established that CMV can cause EHBA. However the association is too frequent to be ignored.

Regarding age of surgery of both patients, it is known that the prognosis of the Kasai operation worsens when the age of the child at surgery increases [3]. Liver transplant in patients with biliary atresia is expensive and not easily accessible and available in our country. Thus, portoenterostomy may remain the only option in most of these patients. In both our patients, parents refused the option of liver transplant. Chardot, *et al.* [4] also reported that the five year survival rate in patients who got operated after 90 days of age was 25%±6.1% whereas in those in whom surgery was not done, only one patient survived till five years [4]. Survival was 100% with those having biliary atresia limited to common bile duct and those who did not have cystic biliary atresia or splenic malformations. Similarly, Davenport, *et al.*, [5] showed that age of surgery had no effect on isolated biliary atresia as compared to those with associated cysts or splenic malformation [5]. These data suggest that although everything should be done to perform the Kasai operation as early as possible, it still has a chance of success when performed after the age of 3 months, especially in patients without embryonal biliary atresia. In our patients, biliary atresia evolved over a period of time and thus a Kasai surgery may not even have been feasible earlier.

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REFERENCES

1. Fischler B, Ehrnst A, Forsgren M, Orvell C, Nemeth A. The viral association of neonatal cholestasis in Sweden: a possible link between cytomegalovirus infection and extrahepatic biliary atresia. *J Pediatr Gastroenterol Nutr.* 1998;27:57-64.
2. Tarr PI, Haas JE, Christie DL. Biliary atresia, cytomegalovirus, and age at referral. *Pediatrics.* 1996;97:828-31.
3. Schneider BL, Brown M, Haber B, *et al.* A multicenter study of the outcome of biliary atresia in the United States, 1997 to 2000. *J Pediatr.* 2006;148:467-74.
4. Chardot C, Carton M, Spire-Bendelac N, Le Pommelet C, Golmard J, Reding R, *et al.* Is the Kasai operation still indicated in children older than 3 months diagnosed with biliary atresia? *J Pediatr.* 2001;2:224-8.
5. 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.