Extrahepatic Biliary Atresia : The Importance of Early Referral

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Extrahepatic biliary atresia (EHBA), characterized by obliteration or discontinuity of extrahepatic bile ducts, is still the major cause for liver transplantation among children nowadays¹. All untreated children eventually die due to complications resulting from portal hypertension and liver cirrhosis. The exchange and diffusion of information that can make the diagnosis of EHBA easier is of utmost importance, since prognosis is improved when patients are surgically treated by portoenterostomy in the first 2 months of life.

The clinical picture that characterize EHBA are jaundice, acholic stools, choloria and hepatomegaly. It should be noted that jaundice may be mild, despite bile duct obstruction. The change in skin colour might not be so evident in dark-skinned patients, with only mild sclera jaundice. Since at the time of symptom onset, the child usually has a good health status and adequate weight, mild jaundice is often missed and the diagnosis is established later. Other signs may be present, such as steatorrhea. In the most advanced stages of the disease, one can observe splenomegaly, collateral circulation, ascites, upper gastrointestinal bleeding due to the rupture of esophagogastric varices and other signs and symptoms resulting from portal hypertension and from liver cirrhosis².

Long-term survival and quality of life depend on early treatment. Therefore, neonatal cholestasis can be seen as a paediatric gastroenterological emergency.

Improved education leading to early referral and diagnosis

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to allow surgery before 8 weeks in specialised centres has led to improved management and outcome of biliary atresia³. Best results are achieved if surgery is performed before 8 weeks of age. If portoenterostomy is not performed, fibrosis will implacably progress to end-stage cirrhosis and death in the first year of life in 50 to 80% of children and up to the third year of life in 90 to 100% of patients^{4,5}.

Patients submitted to the Kasai procedure at an early age (< 60 days) have a better prognosis^{6,7} as shown in Table-1. After the third month of life, Kasai portoenterostomy is still indicated, because even with a lower success rate, the need for liver transplantation can be postponed.

The aspects described here highlight the importance of early referral of these children to specialized centers, but this does not usually happen. The possibility of physiologic or breast milk jaundice may hinder and delay diagnosis, except if bilirubin levels are measured in order to detect direct hyperbilirubinemia. Even in industrialized countries, 14 to 29% of patients with biliary atresia are referred for assessment only after the third month of life. Every patient with jaundice should be assessed after 14 days of life, since at this age, the diagnosis of physiologic jaundice can be totally ruled out. It should be underscored that the colour of stools and urine must be considered when examining a jaundiced infant.

We have operated around 45 infants having EHBA since the last three years. Patients who present early within two months of age did well in post-operative period. Their jaundice regressed, bilirubin levels return to normal, stools became cholic, gained weight. Patients, who present late after three months of age, with cirrhosis and ascites, did not

well. We stress on early referral of any neonate with cholestasis, so that EHBA can be early diagnosed and treated and prevent the morbidity and mortality of late presentation.

Table -1

<i>Patient's age at portoenterostomy (days)</i>	<i>Biliary drainage after portoenterostomy (%)</i>
<i>< 60</i>	<i>70 – 80</i>
<i>60 – 90</i>	<i>40 – 50</i>
<i>90 – 120</i>	<i>25</i>
<i>> 120</i>	<i>10 – 20</i>

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