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Ultrasound Findings in Biliary Atresia: The Role of Triangular Cord Sign

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Abstract: In this prospective study, 49 infants with prolonged jaundice, were studied between January 2005 to April 2007 in Tabriz Children's Hospital. All of these infants were evaluated with ultrasonography and isotopscan and finally biopsy was done in all of them under guide of sonography. In their sonographic evaluation, absence or presence of gall bladder, its size and also its evacuation after feeding was checked and beside this triangular cord sign was controlled. The PPV and specificity of triangular cord sign in diagnosis of extrahepatic biliary atresia (EHBA) was 100% but its NPV was only 78.5%. The overall accuracy, PPV, NPV, specificity and sensitivity of ultrasonographic findings in diagnosis of EHBA was about 94, 92, 94, 97 and 86%. Ultrasonography is a reliable screening method in early diagnosis of EHBA and triangular cord sign increases its accuracy especially its specificity.

Key words: Extrahepatic biliary atresia, ultrasonography, triangular cord sign

INTRODUCTION

Cholestatic jaundice in early infancy continues to represent a clinical and diagnostic intricate.

Prolonged conjugated hyperbilirubinemia can be caused by extrahepatic biliary atresia (22% of cases) or it may be the result of inflammatory, metabolic and miscellaneous causes including: idiopathic neonatal hepatitis (44.4%), alpha-1 antitrypsin deficiency (8.1%), Allagille syndrome (4.5%), choledochal cyst (2.4%), related to total parentral nutrition (2.3%) and Byler syndrome (2%) (Cauduro, 2003).

The estimated prevalence of EHBA is about 1 in 10,000-15,000 live born infants with a slight predominance among the female sex (1.4:1) and it is more prevalent among Asians (Bates *et al*., 1998). EHBA is subcategorized in three groups: in type I, the atresia is limited to Common Bile Duct (CBD) while more proximal ducts continue to be patent, whereas in type II, it is the Common Hepatic Duct (CHD) which has atresia while the proximal ducts continue to be patent, these two types correspond to 12-15% of EHBA cases. In type III which is found in 85-88% of cases, atresia occurs in right and left hepatic ducts (RHD, LHD) with or without involvement of CHD and CBD (Utterson *et al*., 2005).

Since immediate portoentrostomy may prevent further potentially fatal consequences of EHBA, it is very critical to differentiate EHBA from other causes of cholestatic jaundice (Davenport *et al*., 2007; Rafeey *et al*., 2008; Otte *et al*., 1994). Ultrasonography, hepatobiliary scintigraphy, ERCP, intraoperative cholangiography and liver biopsy are diagnostic modalities, but sonography and isotopscanning including HIDA, DISIDA and BRIDA are more commonly used.

Ultrasonography has play a role in screening patients with infantile cholestasis, mainly focusing on the size, shape and contractility of gall bladder. Ultrasonographic triangular cord sign which represents a cone-shaped fibrotic mass cranial to the bifurcation of the portal vein is also a useful diagnostic criterion (Kotb *et al*., 2001; Choi *et al*., 1999; Park *et al*., 1997, 1999).

The accuracy of ultrasound in diagnosis of EHBA is controversial and it was reported as low as 38% by Zheng *et al*. (2002) and as high as 95% in Park *et al*. (1997). The accuracy of ultrasound was between these limits in other studies. The aim of this study was to investigated the accuracy of the TC sign in the diagnosis of BA.

MATERIALS AND METHODS

In this prospective study, 49 infants with prolonged jaundice were studied between January 2005

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to April 2007 in Tabriz Children's Hospital. All of these infants were evaluated with ultrasonography and isotopscan and finally, liver biopsy under sonographic guide was done in all of patients.

The sonographic exam was performed by a multifrequency linear probe with frequency range of 8 to 14 MHZ.

All of the patients were studied after 4 h fasting. The presence or absence of gall bladder and when gall bladder was detectable, its size was measured. In the second stage, gall bladder size, one hour after feeding was measured.

Beside these measurements, the Triangular Cord (TC) sign was checked. The TC sign was defined as presence of triangular or tubular echogenic density just cranial to the portal vein bifurcation or thickness of the echogenic anterior wall of the right portal vein (EARPV) of more than 4 mm on longitudinal scan.

RESULTS

The mean age of patients was 56.1±17.8 days and mean weight of them was 4054±930 g. The male to female ratio was 1/1.

In diagnosis of EHBA, the sensitively and specificity of absence or small size of gall bladder (smaller than 15×5 mm in longitudinal plane or volume less than 0.2 mL) was 64.2 and 88%, respectively and its accuracy was 79.4% using the volume reduction of gall bladder one hour after feeding (more than 50% of primary volume) the sensitivity, specificity and accuracy of ultrasonography in diagnosis of EHBA was 83.3, 93.3 and 90.4%, respectively.

The triangular cord sign was completely specific in diagnosis of EHBA and its specificity and Positive Predictive Value (PPV) was 100% but its negative results was not reliable (negative predicative value was 78.5%).

Using all above mentioned criteria, sensitivity, specificity and accuracy of ultrasonography in diagnosis of EHBA was 96.9, 85.7 and 93.6%, respectively (Table 1).

Table 1: Comparison of ultrasonographic findings in diagnosis if EHBA

Ultrasonography finding	Sensitivity (%)	Specificity (%)	PPV (%)	NPV (%)	Accuracy (%)
Absence or small size of gall bladder	64.2	88.0	75.0	81.4	79.4
Gall bladder depletion after feeding	83.3	93.3	83.3	93.3	90.4
Triangular cord sign	25.0	100.0	100.0	78.5	80.0
Ultrasonography as a whole	85.7	96.9	92.3	94.1	93.6

Table 2: Comparison of ultrasonography and HIDA scan in diagnosis if EHBA

Diagnostic methods	Sensitivity (%)	Specificity (%)	PPV (%)	NPV (%)	Accuracy (%)
Ultrasonography	85.7	96.9	92.3	94.1	93.6
HIDA scan	100.0	51.5	46.6	100.0	65.9

In this study, HIDA scan was completely sensitive in diagnosis of EHBA and its sensitivity and NPV were 100% but its specificity was very low (only 51.5%) (Table 2).

DISCUSSION

In the evaluation and management of conjugated hyperbilirubinemia in infants, diagnosis of surgically correctable situation and disorder which specific treatment is available was important and timely manner. This study confirms the finding of Choi *et al*. (1996), that reported the sonographic feature of a fibrous cone at the porta hepatis as triangular-shaped ecogenic density (TC sign), just cranial to the bifurcation of the portal vein on transverse or longitudinal scan. Other study had suggested that hepatobiliary scintigraphy should be the next step if TC sign is not visualized (Park *et al*., 1999).

Although the accuracy of ultrasonography in diagnosis of EHBA is controversial but based on this study, when ultrasonographic study perform in a complete manner including studying the patient in fasting state and one hour after feeding, especially when using triangular cord sign as a criterion beside other ultrasonographic criteria, it is a reliable screening method in early diagnosis of EHBA and its PPV and NPV are more than 90%. The specificity and PPV of triangular cord sign by itself in diagnosis of EHBA is 100%.

HIDA scan although is very sensitive in diagnosis of EHBA (100%) but its positive findings are not reliable and its PPV is only 46.6%. Thus TC sign and ultrasonography was simple and reliable diagnostic modality test for evaluating of neonate with cholestasis.

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