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Ultrasonographic Diagnosis of Biliary Atresia in Infants

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〈국문초록〉

영아에서 담도 폐쇄증의 초음파 진단

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담도 폐쇄증은 수술로서 치료 가능한 신생아 황달의 중요한 원인중의 하나이다. 담도 폐쇄증의 감별 진단에 여러가지 검사방법이 이용되고 있으나 이중 초음파 검사가 가장 유용하게 쓰이고 있다. 29명의 수술로서 확진된 담도 폐쇄증의 초음파 소견을 분석한 결과 25명 (86.2 %)에서 담낭이 없거나 심한 위축 소견을 보였고 5명에서 담관 확장 소견을 보였고 24명(82.8 %)에서 정상 간 에코를 나타냈다. 가장 중요한 초음파 소견은 담낭의 위축(장축이 1.5 cm 이하) 또는 형성 부전이며 불규칙한 담관 확장 소견도 도움이 된다.

Index Words : Bile ducts, abnormality
Bile ducts, US studies
Infants, gastrointestinal tracts

Introduction

Biliary atresia composed the prime cause of neonatal and infantile jaundice with neonatal hepatitis and the differential diagnosis between them is important in patient management because the former can be surgically corrected (Kasai's biliary-enterostomy) and the latter is a medical disease.

The diagnostic modality of the biliary atresia is stil not conclusive. Ultrasonography, radioisotope scan, computed tomography, or even simple radiography all play some role in diagnosis.

Of all the modalities, the ultrasonography is easy to applicate, noninvasive, free of X-ray hazard, so is thought to be the most convenient modality of choice. But still no definitive diagnostic sign of the ultrasonography defined. We analyzed 29 patients of surgically proven biliary atresia, about the main ultrasonographic findings on diagnosis.

Materials and Methods

The 29 patients with surgically proven biliary atresia from July, 1985 through March, 1990

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were retrospectively analyzed by three radiologists.

The machine used was Diasonics DRF–400 and the transducers were 5 or 10 MHz, sector or linear probes.

The age distribution was from 1 month to 9 months (mean 3.1 months). There were 15 males and 14 females. The patients divided into two groups by 3 months of age, because the most appropriate time for surgery is known to be around 3 months of age[1,2,3].

Analyzing points were 1) gallbladder size, 2) intrahepatic ductal dilatation, 3) hepatic parenchymal echogenecity and 4) associated other findings including complications from secondary biliary cirrhosis and congenital malformation. The size of the gallbladder was roughly divided into 3 groups, 1) nonvisualization, 2) atrophic-(long diameter below 1.5 cm), 3) normal(long diameter above 1.5 cm). The results of the operative cholangiography, radioisotope scan, follow-up ultrasonography and other clinical findings were also included in available patients.

Clinical review showed that all 29 patients were admitted for evaluation of the jaundice. Total bilirubin level ranges from 6.3 to 17.2 mg/dl and direct bilirubin level were from 1.8 to 14.0 mg/dl.

Most of the patients were the first baby in their family and had no significant family history in 26. Among other 3, one was whose mother suffered hepatitis and became hepatitis B virus antigen carrier, one was whose father carried hepatitis history and one was whose elder brother had hepatitis history.

Operative findings, pathologic proof and final outcome of the patients were reviewed.

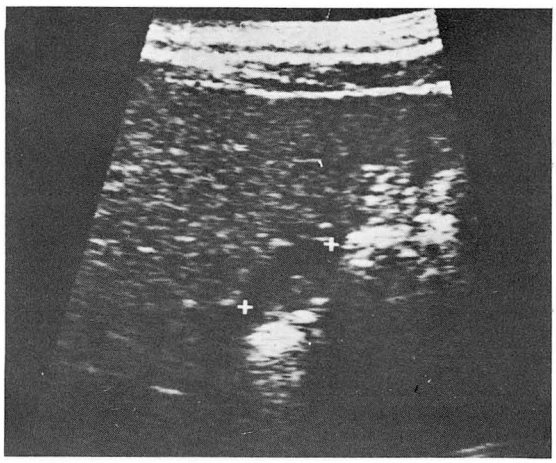
Results

About the size of the gallbladder, most patients showed nonvisualization or atrophy in both age groups (Table 1). In 86.2 % of the patients, could not find or could find only very small atrophic gallbladder (Fig. 1). But 4 patients(13.8 %) showed normal sized gallbladder(Fig. 2).

Intrahepatic ductal dilatation was found in only 3 patients and 2 patients showed cystic dialtation of the common bile ducts(Table 2).

Table 1. Gallbladder Size on Ultrasonography

Findinge/Age	<3mo.	≥3mo.	Total
Nonvisualization	12	10	22(75.9 %)
Atrophic(〈 1.5 cm)	2	1	3(10.3 %)
Normal(〉 1.5 cm)	1	3	4(13.8 %)
Total	15	14	29(100.0 %)



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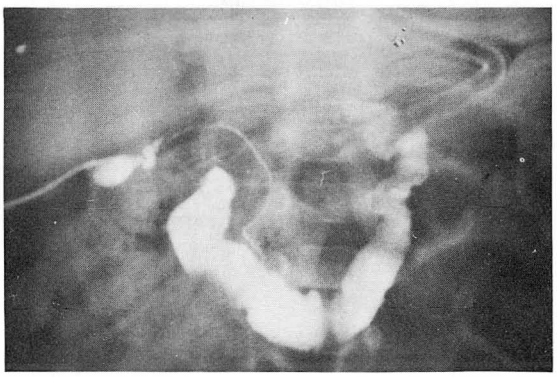

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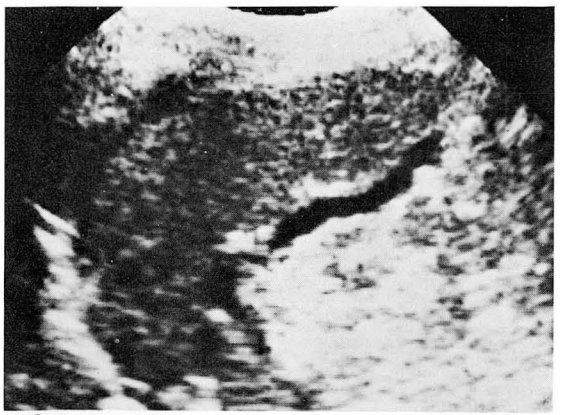
Fig. 1. a. This 2–months–old boy shows very small, atrophic gallbladder(less than 1 cm of long axis)
b. Operative cholangiography shows atrophic gallbladder and atretic extrahepatic bile ducts.

Most patients showed no significant hepatic parenchymal echo change, but many patients showed some degrees of cirrhotic changes by operation. Again, there was no significant difference between the two age groups(Table 3). Associated anomalies are one situs inversus and one case of ectopic pancreas in jejunum.

Associated findings on diagnosis are pneumonia, rickets, various stigmata of liver cirrhosis, such as ascites, splenomegaly, but in a small percentage.

Table 2. Changes of Biliary Tree (n=29)

Findinge/Age	<3mo.	≥3mo.	Total
IHD dilatation	1	2	3
Cystic dilatation of CBD	1	1	2



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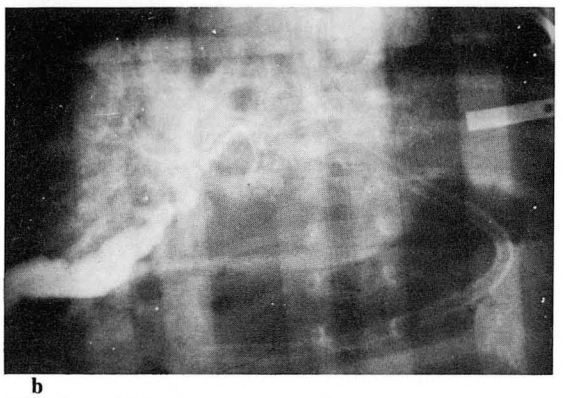

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Fig. 2. a. This is a case of the normal sized gallbladder. The longist diameter is about 5 cm on ultrasonography.
b. On operative cholangiography, the abnormally elongated, slender appearance is noted. No patent extrahepatic ducts visualized and duodenum is not opatified.

Table 3. Hepatic Parenchymal Echogenecity

Findinge/Age	<3mo.	≥3mo.	Total
Normal	12	12	24(82.8 %)
Slightly coarse or inhomogeneous	3	2	5(17.2 %)
Total	15	14	29(100.0 %)

RI scanning was done in 21 patients, using 99 m Tc–DISIDA scan in 16 patients, and definitely diagnosed as biliary atresia in 16 of 21 patients.

Operative cholangiography was done in 9 patients. Among these, 6 patients showed the same finding with ultrasonography, but 3 patients showed difference, one larger gallbadder(probably due to distension by contrast media) and two small visible gallbladders in sonographically non-visualized group. Other patients were not able to do operative cholangiography due to severely atrophic gallbladder.

Pathologic findings all showed intraparenchymal cholestasis, bile ductular proliferation, giant cell transformation, chronic periportal inflammation, fibrosis and extramedullary hemopoiesis.

Post–operative follow–up is completed in 9 patients. Of these, 3 patients are rather improved state, though have very many complications and 6 patients were expired. Residual 20 patients were followed up incompletely, probably due to grave prognosis at home, because 14 of the 23 patients(except expired 6) showed some complications from secondary biliary cirrhosis during management.

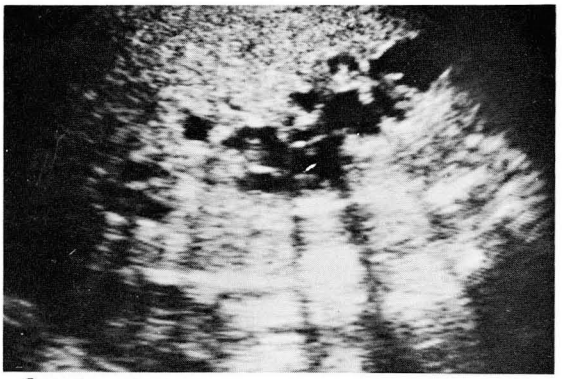
Discussion

From the earlier period, the differential diagnosis of the neonatal and infantile jaundice is a

very important issue for the pediatricians and pediatric surgeons[1,4] After introduction of the Kasai's biliary-enterostomy, the biliary atresia is thought to be a surgically correctable disease, though the final outcome is still grave[5]. Radiologists can use the ultrasonography as the first and essential diagnostic modality to differentiate biliary atresia from other causes, but still the definitive diagnostic point is not so well established.

In this study, 86.2 % of the patients showed not definable gallbladder, so it will be a valuable criteria of diagnosis of biliary atresia by ultrasonography. This percentage is higher than 60 % reported by Brun[6]. But also note that 4 patients(13.8 %) showed rather normal size of the gallbladder as the results of Weinberger[7], Daneman[8], and Brun[6]. In my opinion, these patients showed somewhat slender, elongated appearance of the gallbladder rather than normal ovoid, well-distended morphology(Fig. 2), these suggests that the size in itself cannot completely exclude the possibility of biliary atresia[6,7,8]. Fatty meal test or phenobarbital ingestion can give some valuable information about gallbladder function[2], but Weinberger[7] reported a case of normal gallbladder size and function in biliary atresia and suggested that such test revealed only about the patency between the cystic duct and duodenum and gives no information about biliary trees above the gallbladder.

Intrahepatic ductal dilatation was shown in 3 patients and one showed very irregular, noncommunicating. cystic "bile lakes"(Fig. 3). as mentioned by Ito[4] and Fonkalsrud[3]. I think that biliary atresia can show variable degrees of intrahepatic ductal dilatation but of low incidence, Green[2] said that intrahepatic ductal dilatation was more likely to suggest other obstructive anomaly, but our results are compatible with Ito's report[4]. Ishii[9] and Werlin[5] reported cystic dilatation of intrahepatic ducts after hepatic por-



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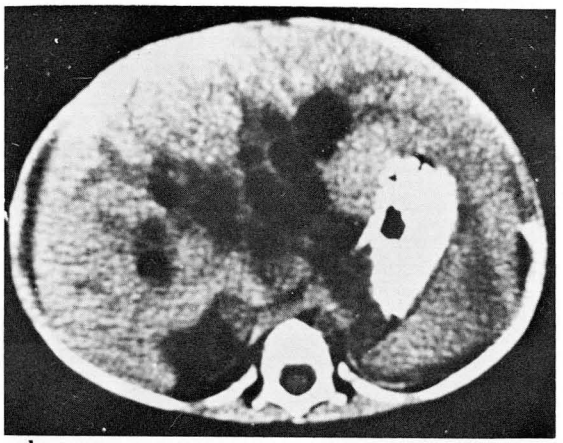

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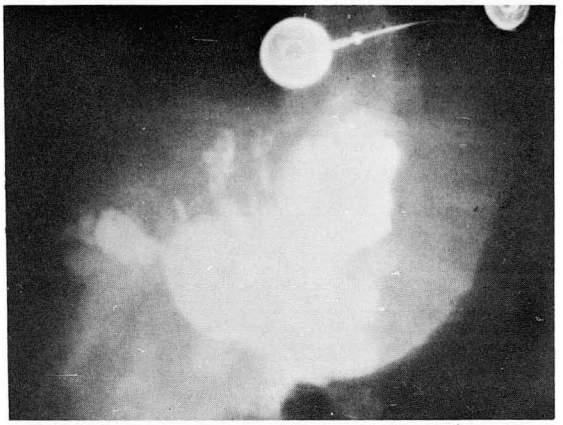

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Fig. 3. a. This 7–months–old boy shows massively dilated, non–communicating irregular cystic intrahepatic ducts in the central portion.
b. Abdominal CT shows the same finding. So obstructive Jaundice must be differentiated.
c. Percutaneous transhepatic cholangiography confirms that the dilated cystic spaces are intrahepatic biliary trees and completely occluded extrahepatic duct.

to-enterostomy, but we have no such case on follow-up study. Other findings such as pneumonia, rickets or stigmata of liver cirrhosis all can be explained to be some phenomena caused from secondary biliary cirrhosis and hepatic failure.

I hoped to find out some difference in the hepatic parenchymal echogenecity between the two age groups, expecting different levels of cirrhotic changes[2], but only 5 patients showed slightly coarse or inhomogeneous parenchymal echogenecity suggesting cirrhosis and no difference between the two age groups, So hepatic parenchymal echogenecity is not so helpful in diagnosing and characterizing the status of the patient, though helpful on follow-up study.

Daneman[8] reported abnormal band of increased echoes around the intrahepatic portal radicles and ducts as a useful diagnostic sign, but our cases show no definite such chagnse.

Some authors reported that 10 or 22 % of biliary atresia patients show congenital anomalies such as polysplenia, azygos continuation of the inferior vena cava, preduodenal portal vein, hepatic arterial anomalies, bilateral bilobed lungs or bowel stenosis, etc[8,10,11,12]. But our series show only two patients of situs inversus and ectopic pancreas. It can be explained by the more likely essentially inflammatory origin of the biliary atresia.

Still the above mentioned findings are not sufficient for the diagnosis of the disease and the combination of various modalities will be necessary, but the findings mentioned above will do help to distinguish the biliary atresia form many other disease entities causing neonatal and infantile jaundice.

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