ORIGINAL ARTICLE

Current Management On Neonatal Obstructive Jaundice

(The differential diagnosis between Neonatal Hepatitis and Biliary Atresia)

by

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Abstract

It is known in this country that surgical obstructive jaundice cases used to be sent to the surgeon in the terminal state.

This communication is a brief explanation made by the author about the current management on neonatal obstructive jaundice, which can easily be applied in this country.

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Introduction

Jaundice in neonates is easily recognized by parents or physicians; and if it does not subside within a few days it might raise a lot of questions.

The condition which may be responsible for prolonged jaundice in the neonate could be catagorized into two main groups :

A. Medical jaundice : hemolytic, enzymatic, metabolic or infections.
B. Surgical jaundice known as obstructive jaundice.

Diagnostic tests and treatments for group A are definite. However, there is a long list of controversial and sophisticated tests. Chiba and Kasai (1975), Hays and Kimura (1980) used to differentiate mechanical obstructive jaundice (extra-hepatic and sometimes plus intra-hepatic cholestasis) known as Biliary Atresia (BA) from the hepatocellular (intra-hepatic cholestasis) known as Neonatal Hepatitis Syndrome (NH).

The following communication is a review of the author's experience on neonatal cholestasis, and a proposal for a simple diagnostic approach which could easily be applied in Indonesia.

Diagnosis

It is difficult to make the diagnosis of prolonged obstructive jaundice in many infants due to the gradation of clinical pathological findings.

At present the known diagnostic procedures are as follows :

History :

- Previous siblings with prolonged jaundice and known diagnosis and outcome.
- Viral infection in the mother such as hepatitis, genital herpes, rubella or other infections such as syphilis which may be transmitted to the fetus.

A possible association between dextroamphetamine ingestion by the mother and occurrence of biliary atresia in her infant has been suggested by Levin (1971).

Physical examination :

Jaundice is easily recognized and it means that there is a disturbance between the production and excretion of the bile. Palpation of an enlarged liver with a firm blunt edge suggests biliary atresia. An enlarged liver with a sharp edge which feels rather soft suggests hepatitis. Hepatosplenomegaly suggests congestion due to the liver disease.

Determination of relevant components in cholestatic jaundice has been summarized by Hays and Kimura (1981) as listed in Table 1.

TABLE 1 : *Studies used in the evaluation of the infant with Neonatal Cholestatic Jaundice.*

Urine Studies :

a). Urine amino acid analysis
b). Culture for cytomegalovirus
c). Urobilinogen

Emaging Studies :

a). Upper GI series
b). Ultrasonography
c). Hepatic scintigraphy (HIDA, PIPIDA)

Feces Studies :

a). Bilirubin
b). ^{131}I rose bengal excretion

Serum Studies :

a). Coagulation screen
b). Plasma protein fraction, alpha-fetoprotein.
c). Serum enzymes : LDH, GOT, GPT, Alk. phosphatase
d). Bile acid profile
e). Hepatitis B surface antigen
f). Infant serologic studies
g). Serum colloidal reactions : lipoprotein-X, alpha-l-anti trypsin activity
h). Miscellaneous : parent blood type, Coombs test, Bilirubin, Cholesterol, Ureum, blood culture

Bile excretion in the intestinal studies :

The excretion of ^{131}I labeled rose bengal or BSP has been used in many hospitals in Indonesia. However, the isotope content in the 1 – 3 days collected stool has not yet been examined here. Actually the examination of the total isotope excretion in the stool is a more relevant method for diagnosing cholestatic jaundice than BSP test. Recently there is a growing interest in duodenal aspiration to obtain intraluminal content for bile analysis (Hashimoto et al., 1978; Yamada, 1983) which is now widely used. Under fluoroscopic controle the duodenal tube is inserted and a small amount of 25% Magnesium Sulfate is introduced via the tube. The aspirated fluid is examined by Ictotest for bilirubin. A blue colour means positive for bilirubin, which excludes the diagnosis of biliary atresia. Red means negative and if such is the case the test should be repeated again to confirm the result. The result of this examination can be readily obtained and establishes the diagnosis in more than 90% of the patients (Halimun et al. 1983).

Studies of stool for bilirubin and urine for urobilinogen are not conclusive. Slight bile colour in the stool may come from sloughed intestinal mucosal cells.

Radiographic Study :

Percutaneous transhepatic cholangiography (PTC), a technique widely used in adults with obstructive jaundice, has been used for neonates in some institutions. Guided by fluoroscopy, a fine long needle is introduced up to the main intrahepatic bile duct, through which 5 – 10 ml of urographine is injected to visualize the intrahepatic bile trees and the outflow to the duodenum. PTC is also used as a drainage procedure in inoperable obstructive jaundice cases known as PTD.

Percutaneous liver biopsy :

Histologic study of liver specimen cannot differentiate BA from NH in 20% of cases (De Lorimier 1973). "Giant Cells" a characteristic feature of neonatal hepatitis are found in 40% of the patients with biliary atresia. This more valuable procedure is taken intra operative to confirm the diagnosis and predict the ultimate prognosis.

Operative cholangiography and wedge liver biopsy will establish the correct diagnosis in 98 per cent of the patients (Hays and Kimura, 1981). Thaler and Gellis (1968) reported that the incidence of cirrhosis will increase three times in NH cases after surgery. However, Halimun, et al. (1983) stated that surgical intervention does not give any unfavourable effect on the liver regeneration.

Treatment of Bile duct obstruction.

Inspissated bile syndrome is a term that should be confined to infants who have bilious sludge obstructing the extrahepatic bile ducts. Usually the obstruction follows a severe hemolytic or severe dehydration episode. Irrigation of the bile ducts is the treatment of choice. The prednison therapy test which is widely used in Indonesia is not recommended before the diagnosis is established by duodenal aspiration test.

The results of treatment of biliary atresia have been dreadfully in the past, but today there is no doubt that the 'Kasai hepatoenterostomy' provides hope of survival. Successful bile drainage is the first step towards complete cure of the disease, however ascending cholangitis, irreversible process of the cirrhosis and the portal hypertension will influence the ultimate outcome of the infants.

Discussion

Biliary atresia is not a congenital malformation but a process that develops after birth, since it is ready found in routine autopsies in newborn infants (De Lorimer, 1973).

It is known that the effort for biliary reconstruction has to be done before irreversible liver damage occurs. Timing for surgery is between 6 to 12 weeks of age. In patients younger than 6 weeks the fibrous tissue around the porta hepatis is not strong enough and difficult for anastomosis. If they are older than 12 weeks one should be concerned about the progressivity and irreversibility of cirrhosis of the liver. Time consuming examinations before surgical intervention has to be avoided. The management and attitude suggested in this communication is as follows :

In every infant with prolonged jaundice and acholic stool, the routine liver function tests are examined (serum bilirubin, total protein, LDH, GOT, GPT and Alkaline phosphatase). If direct bilirubin and alkaline phosphatase are dominantly high, then the following step to do is the duodenal aspiration for bile excretion. If the bile is positive there is no indication for surgery. However, if repeated aspirations show negative findings then the case is due for surgical intervention.

At exploratory laparotomy the evaluation starts from the appearance of the gall bladder and the presence of lumen in it. If there is no lumen the next step is dissection of the porta hepatis for corrective surgery in biliary atresia. If a lumen is found then cholangiography is performed, and it might visualize the intra and extrahepatic biliary system. If no bile duct is found then this case belongs to BA. With this management the diagnosis of surgical jaundice or NH will be established on the 4th or 5th day after admission. A preoperative misdiagnosis in 7 out of 87 cases (8%) has been reported by the author in 1983.

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