and III (73.0±5.1 for Group I vs 58.4±8.9 for II, and 54.1±15.2% for III, P<0.02).
The data show that the synthesis of cholic acid was increased in hypertriglyceridemics, so that the relative concentration of total bile acids in bile was kept in normal range. The present results are in good agreement with the results of Einarsson et al (J Clin Invest 54: 1301, 1974), which showed an increased pool size of cholic acid in hypertriglyceridemia. The cause of the frequent occurrence of cholelithiasis in Type IV hyperlipidemia (European type of cholelithiasis) must be different from the mechanism working for normotriglyceridemic patients (American Indian type cholelithiasis).

(7) Hepatolithiasis—Clinical Findings and Cholangiographic Features

Masao Ohto, M D
The First Department of Internal Medicine, Chiba University, School of Medicine, Chiba

By using percutaneous transhepatic cholangiography (PTC) and endoscopic retrograde cholangiography (ERC) in practice, hepatolithiasis has been exactly diagnosed without surgical help. Surgical procedures have not yet shown a sufficient effectiveness to cure the disease.

Clinical features and pathogenesis of hepatolithiasis have been studied on the basis of cholangiographic findings by PTC and ERC.

Gross findings of hepatic calculi and their chemical composition

All hepatic calculi of 14 patients showed a brown coloured amorphous structure. They belonged to pigment- calcium stones from chemical composition.

Incidence of hepatolithiasis

Hepatic calculi were found by cholangiography in 5.5% of the patients of gallstone diseases. When the patients previously underwent cholecystectomy were excluded from the group, the rate of incidence became 3.2%.

Clinical symptoms and laboratory findings

Fever and jaundice in hepatolithiasis appeared more frequently and intensively than those in cholecystolithiasis and choledocholithiasis. Disturbed liver function and inflammatory findings were also evidenced more frequently by various clinical laboratory examinations.

Clinical features of hepatolithiasis

(1) Causes of postcholecystectomy symptoms

Hepatic calculi were found in a high incidence, 21.5% of the patients with postcholecystectomy symptoms who underwent roentgenologic examinations by PTC and ERC.

(2) Mortality due to gallstone disease

Mortality rate of hepatolithiasis was 23.5% and far higher than 0.3% of cholecystolithiasis and 2.2% of choledocholithiasis.

(3) Repeated surgical procedures of the biliary system

Most of the mortal patients due to hepatolithiasis had undergone repeated surgical laparotomies of the biliary system before the death.

Classification of hepatolithiasis

According to cholangiographic findings by PTC and ERC, hepatolithiasis is divided into two types, primary and secondary (Figs. 1 and 2).

The primary type of hepatolithiasis has a saccular or cylindrical segmental dilatation of main intrahepatic bile ducts where
Hepatolithiasis—Clinical Findings and Cholangiographic Features

Fig. 1. Cholangiographic features in 38 patients of the primary type of hepatolithiasis. Distribution of gallstones in the liver and junction of major intrahepatic ducts are diagrammatically shown.

a. Hepatic calculi are located in segmentally dilated main intrahepatic ducts of the left lobe.
b. Hepatic calculi are located in segmentally dilated main intrahepatic ducts of the right lobe.
c. Hepatic calculi are located in segmentally dilated main intrahepatic ducts of the right and left lobes. The diseased branches of each lobes join respectively to the right hepatic and the left hepatic duct.
d. Hepatic calculi are located in segmentally dilated main intrahepatic duct of the right and left lobes. The diseased branches of each lobes join together to the left hepatic duct.

Type a was seen in 10 patients (5), type b in 3 (2), type c in 9 (2) and type d in 16 (7). ( ) : Number of the patients having no stones in the extrahepatic bile ducts and the gallbladder.

Fig. 2. Cholangiographic features in 52 patients of the secondary type of hepatolithiasis. Obstructive changes of the extrahepatic bile ducts and distribution of gallstones are diagrammatically shown. Gallstones are mainly located in the extrahepatic bile ducts and concomitantly in the intrahepatic bile ducts.

a and b. Gallstones are distributed extra- and intrahepatically above acquired or congenital stenosis of the common hepatic or common bile duct. a : congenital, b : acquired.
c and d. Gallstones are distributed extra- and intrahepatically above a large stone fixed at the end of the common bile duct (c) or incarcerated in its' proximal portion (d).

Type a was seen in one patient, type b in 12, type c in 13 and type d in 26.
Table 1. Clinical findings and cholangiographic features of the primary type and the secondary type of hepatolithiasis.

<table>
<thead>
<tr>
<th>Pathogenesis</th>
<th>Primary type of hepatolithiasis</th>
<th>Secondary type of hepatolithiasis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Congenital, due to anomaly of intrahepatic bile ducts</td>
<td>acquired, due to lesions of extrahepatic bile ducts</td>
</tr>
<tr>
<td>Age of onset in symptoms and signs</td>
<td>Younger age (many in 20-30 years old)</td>
<td>Older age (many in 60-70 years old)</td>
</tr>
<tr>
<td>History of cholelithiasis</td>
<td>Various</td>
<td>Long</td>
</tr>
<tr>
<td>Dilatation of intrahepatic bile ducts</td>
<td>Segmental dilatation of a main hepatic duct</td>
<td>Diffusely spread uniform dilatation</td>
</tr>
<tr>
<td>Stenosis of a major intrahepatic duct</td>
<td>Remarked</td>
<td>Not remarked</td>
</tr>
<tr>
<td>Junction of main intrahepatic ducts</td>
<td>Sometimes, the aberrant type</td>
<td>The usual type</td>
</tr>
<tr>
<td>Location of gallstones in liver</td>
<td>In left or right intrahepatic ducts, occasionally in the both</td>
<td>In right and left intrahepatic ducts, occasionally in the either</td>
</tr>
<tr>
<td>Features of extrahepatic bile ducts</td>
<td>Slight to moderate dilatation with no or a few gallstones</td>
<td>Moderate to extensive dilatation with many gallstones including a fixed or casted one, or with stenosis</td>
</tr>
</tbody>
</table>

Calculi are located. It is often associated with an aberrant junction of main intrahepatic ducts. These findings are based on cholangiograms in the patients with hepatolithiasis having no gallstones in the extrahepatic bile ducts and the gallbladder. A congenital origin is considered to be causative for the primary type.

The secondary type of hepatolithiasis has a diffusely spread uniform dilatation of the intrahepatic bile ducts and also has an extensive dilatation of the extrahepatic bile ducts. Gallstones are mainly distributed in the extrahepatic bile ducts and concomitantly in the intrahepatic bile ducts. Hepatic calculi are usually located in both the right and left lobes.

These findings are based on cholangiograms in the patients of hepatolithiasis caused by strictures of the common hepatic or common bile duct after surgical procedures for the biliary system.

The patients of hepatolithiasis associated with a large fixed or incarcerated gallstone in the common bile duct also have the same findings. Acquired changes of the common hepatic or common bile duct are considered to be causative for the secondary type.

Clinical findings and cholangiographic features of the primary and secondary type of hepatolithiasis are concisely summarized in Table 1.

Comparative incidence of hepatolithiasis in different places and periods

Incidence of the secondary type of hepatolithiasis was higher in a rural than a urban district, and was lower in the later than in the former. Whereas, incidence of the primary type was almost indifferent to them.

Accurate diagnosis and early surgical treatment of gallstone diseases due to development of cholangiography and improvement of medical circumstances in society were thought to result in a remarkable decrease of incidence in the secondary type of hepatolithiasis.

72 Jap J Med Vol 17, No 1 (January 1978)