Scintigraphic Features of Choledochal Cyst

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The scintigraphic appearances of 12 surgically proven cases of choledochal cyst were retrospectively reviewed. In seven of 12 cases, radionuclide accumulated in the choledochal cyst (i.e., the dilated common bile duct) in less than 1 hr. In three additional cases, delayed accumulation (1–24 hr) within the cyst was seen. In two of the 12 cases, no ductal activity appeared and the diagnosis of choledochal cyst could not be made, although in one of these two cases delayed images were not obtained. Other frequent findings included delayed or nonvisualization of the gallbladder (11 of 12) and the appearance of prominent intrahepatic ducts (five of 12). We conclude that hepatobiliary scintigraphy is a noninvasive test useful in the diagnosis of choledochal cyst.

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Choledochal cyst is an uncommon disease, usually felt to be congenital in origin, represented by extrahepatic biliary tract dilatation involving principally the common bile duct. Until the last decade, the diagnosis was only occasionally made preoperatively as a result of the variability of the presentations of affected patients (*1–10*). The failure to consider the diagnosis preoperatively has been associated with poor surgical results and increased morbidity and mortality (*11*).

The gold standard for choledochal cyst diagnosis is direct cholangiography, but such tests are, unfortunately, invasive. The advent of computed tomography (CT) and ultrasound has dramatically increased the ability of radiologists to detect the cystic abnormality by noninvasive means. However, unless the lesion can be proven with these techniques to be part of the biliary tract, a definitive diagnosis of choledochal cyst cannot be made and additional tests are still required (*3,12–20*). In this situation, biliary scintigraphy will usually provide a definitive and noninvasive diagnosis.

In this review the scintigraphic appearances of 12 cases of choledochal cyst are described and compared to those cases that have been previously reported.

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MATERIALS AND METHODS

We reviewed the files of the Department of Radiologic Pathology at the Armed Forces Institute of Pathology and found 12 cases of pathologically proven choledochal cyst that had been evaluated preoperatively with technetium-99m (^{99m}Tc) hepatobiliary scans. The cases dated from 1980 to 1987 and were collected from 12 different institutions. All scans were performed using technetium-99m (^{99m}Tc) iminodiacetic acid derivatives, including N-(2,6-diethylphenylcarbamoylmethyl) iminodiacetic acid (DEIDA), N-(2,6-dimethylphenycarbamoylmethyl) iminodiacetic acid (HIDA), p-isopropylacetanilido iminodiacetic acid (PIPIDA), and diisopropyl iminodiacetic acid (DISIDA). From the clinical summary available in each case, the patient's age, sex, and degree of jaundice, if present, was recorded. The scintigrams were reviewed and the time of appearance, if any, of activity within the choledochal cyst, the gallbladder and the gut was recorded. In one case, images were obtained up to only 1 hr after injection despite the fact that the bile ducts were not yet visualized. Follow-up images obtained up to 24 hr after injection were obtained in all other cases of delayed or nonvisualization of the ducts. Flow and/or blood-pool images, when available, were analyzed for the presence of a photopenic region corresponding to the choledochal cyst. The scintigraphic appearance of intrahepatic ductal prominence was also recorded, if present. Additional images including ultrasonographs, cholangiographs, computed tomographs, and gross photographs of the surgical specimen were reviewed when available, but not specifically analyzed in this study.

RESULTS

Our results are tabulated in Table 1. Note that 11 of 12 patients were female with an average age of 7.1 yr.

TABLE 1
Clinical and Scintigraphic Findings in 12 Cases of Choledochal Cyst

Case no.	Agent‡	Age*	Sex	Jaundice†	Earliest visualization			Dilated ducts
					Cyst	Gallbladder	Gut	
1	HIDA	12	M	No (0.8)	<1 hr	<1 hr	<1 hr	No
2	DEIDA	16	F	Yes	Nonvis.‡ @ 1 hr	Nonvis.‡ @ 1 hr	Nonvis.‡ @ 1 hr	Yes
3	PIPIDA	1	F	No (0.9)	24 hr	24 hr	24 hr	No
4	HIDA	14	F	Yes (10.)	4 hr	<24 hr	<24 hr	Yes
5	HIDA	0.1	F	Yes (7.7)	24 hr (minimal)	Nonvis. @ 24 hr	Nonvis. @ 24 hr	No
6	DISIDA	21	F	Yes	<1 hr	Nonvis.‡ @ 1 hr	<1 hr	No
7	HIDA	2	F	No (0.8)	<1 hr	Nonvis.‡ @ 2.5 hr	Nonvis.‡ @ 2.5 hr	Yes
8	PIPIDA	2.3	F	No (0.2)	<1 hr	Nonvis. @ 24 hr	24 hr	Yes
9	DISIDA	2.2	F	No	<1 hr	Nonvis.‡ @ 1 hr	<1 hr	No
10	DISIDA	0.1	F	Yes (11.7)	Nonvis. @ 24 hr	Nonvis. @ 24 hr	Nonvis. @ 24 hr	No
11	DISIDA	12	F	No	<1 hr	Nonvis.‡ @ 1 hr	<1 hr	Yes
12	DISIDA	2.3	F	No	<1 hr	Nonvis.‡ @ 1 hr	<1 hr	Yes

* The age is expressed in years.
† When known, the total bilirubin levels expressed as mg/100 ml is included in parentheses.
‡ Indicates that additional delayed films up to 24 hr were not obtained.

Clinical jaundice was present at the time of the study in five of 12 cases, and had been present previously in one additional case (Case 1).

Seven of our 12 cases demonstrated visualization of the dilated common bile duct within 1 hr after injection (Figs. 1 and 2). One case showed cyst activity between 1 and 12 hr, and two more between 12 and 24 hr (Fig. 3). There was one case of total nonvisualization of the cyst at 24 hr (Fig. 4). In the final case, no ductal activity was visible at 1 hr, but images beyond 1 hr were not available. Whether in this case subsequent delayed films would have demonstrated extrahepatic biliary activity cannot be determined.

Normal visualization of the gallbladder was uncommon. Only one case demonstrated gallbladder visualization in less than 1 hr (Fig. 2B). Two cases visualized at greater than 3 hr, and nine cases did not demonstrate gallbladder visualization at all (Fig. 1C), although in six of these cases 24-hr delayed films were not obtained. The time of visualization of gut activity also varied. Five cases showed early (<1 hr) gut activity (Fig. 2B), three demonstrated activity at >1 hr, and in four cases no gut activity was visible (Fig. 1C), although in two of these cases 24-hr delayed films were not obtained.

In four cases, flow studies or blood-pool images were available for review and a photopenic defect corresponding to the choledochal cyst was demonstrated in each case. In six instances, intrahepatic duct prominence was visible on the scintigrams (Fig. 1B).

DISCUSSION

Choledochal cyst is an uncommon anomaly defined by congenital dilatation of the extrahepatic biliary tract and represents one of the conditions included in the spectrum of hepatobiliary fibrocystic disease. Other associated conditions include Carolis' disease, congenital hepatic fibrosis, infantile polycystic liver disease, simple hepatic cyst(s), and biliary atresia (*1–3,5–8,21*). The most widely accepted classification system of choledochal cyst is that of Alonso-Lej, with type I, representing cystic dilatation of the main portion of the common bile duct, being by far the most common (*7*). All of our cases were type I. Neither common bile duct diverticulum (type II choledochal cyst) nor choledochocele (type III choledochal cyst) was studied in this series. The etiology of choledochal cyst is debated, but there are two predominant theories. One of these holds that there is prenatal biliary obstruction associated with a congenital weakness of the common bile duct producing dilatation (*6*). The other maintains that there is an anomalous development of the distal common bile duct and pancreatic duct such that there is a long common channel allowing reflux of pancreatic juice into the common bile duct resulting in weakening and dilatation of the duct wall (*22*).

Choledochal cyst is much more commonly seen in females and usually presents in the first two decades of life (*1,2,4,8,10,23–25*), though patients may present in the third and fourth decade (*26*). It has also been diagnosed prenatally (*27*). The age and sex of the patients in our series matches those of previous studies. There is a predilection for Orientals with approximately one-third of the ~1,000 reported cases having occurred in Japan, China, or Korea (*1,6,8,10,15,28,29*). However, to our knowledge, none of our patients were Oriental.

The presenting signs and symptoms can include almost any combination of biliary obstruction, gastroin-

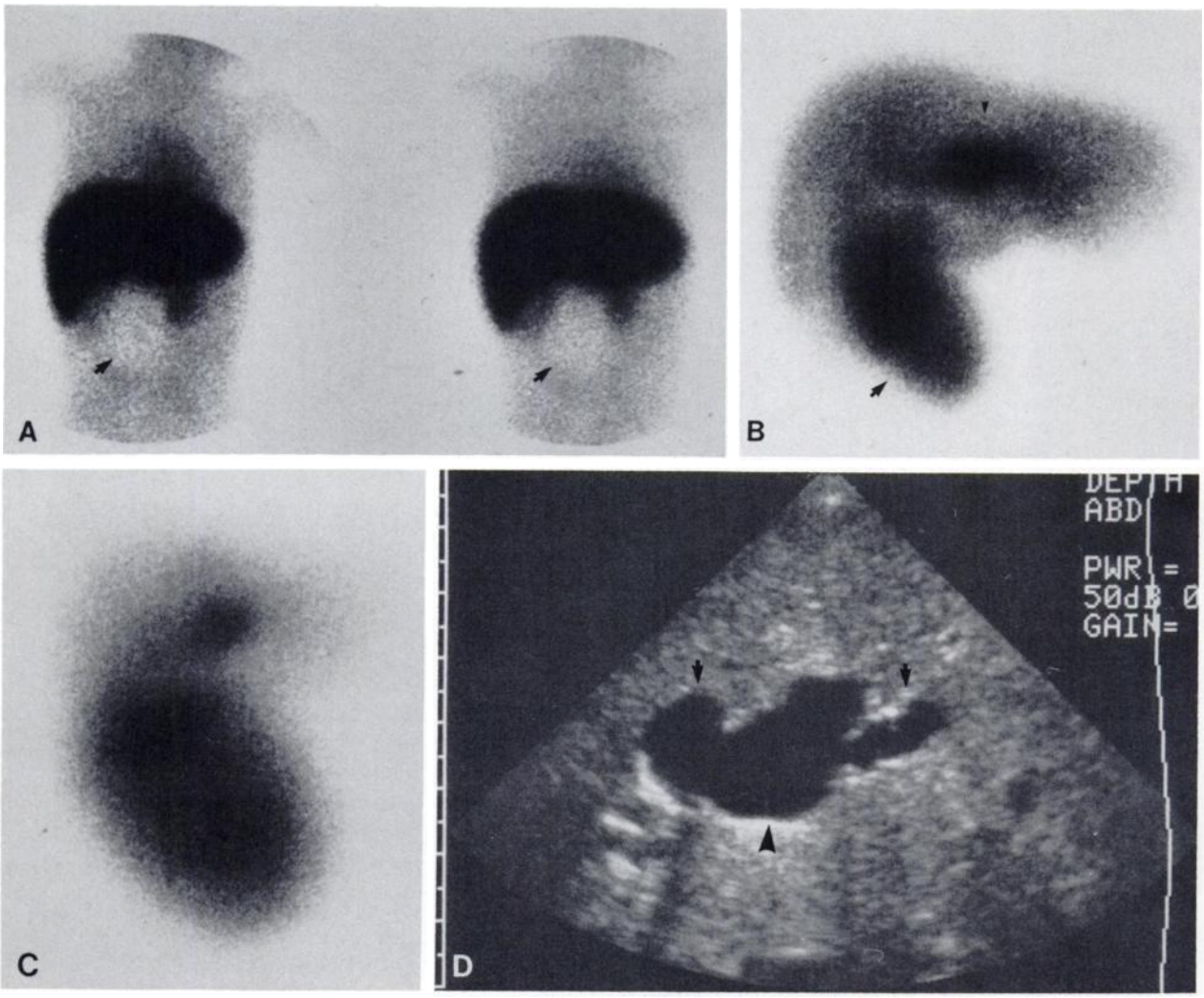

FIGURE 1
Case 8. Prompt visualization of choledochal cyst without gallbladder or gut visualization. A: Blood-pool phase images reveal a photopenic area (arrow) below the liver corresponding to the choledochal cyst. B: At 1 hr after injection, activity within the choledochal cyst (arrow) is clearly visible. Activity within dilated left hepatic ducts (arrowhead) is also noted. C: At 2.5 hr after injection, the amount of activity within the choledochal cyst has increased, but no gallbladder or gut activity has appeared. This nonvisualization persisted up to 24 hr. D: A transverse sonogram through the porta hepatis reveals the superior extent of the choledochal cyst (arrowhead) communicating with dilated intrahepatic ducts (arrows).

testinal obstruction, jaundice, abdominal mass, pain, cholangitis, or bile peritonitis. The classic clinical triad of jaundice, abdominal mass and pain is seen in only 20–30% of cases. This variable presentation of choledochal cyst along with its relative rarity, has made preoperative consideration of this condition uncommon (*1–4,8,9,12,13,15,19,23–26,28,30–31*). There is evidence that failure to consider choledochal cyst in the preoperative assessment can increase surgical morbidity and mortality (*11*). Additionally, failure to diagnose or adequately treat the condition has been associated with an increased incidence of complications, including cholangitis, choledocholithiasis cirrhosis, bile duct perforation and, especially, cholangiocarcinoma (*2,4,8,13,25,32*).

In the past, methods for the diagnosis of choledochal cyst were limited to plain film findings of an abdominal mass, indirect signs of abdominal mass on UGI, or contrast delineation of the cyst by oral, intravenous, or percutaneous cholangiography (*4,8,9,12,33*). Oral and intravenous cholangiography, though occasionally diagnostic in patients with no or minimal jaundice, are nondiagnostic with bilirubin levels >3 mg %, and intravenous cholangiography is associated with a significant risk of severe allergic reaction. Percutaneous cholangiography is sensitive and specific, but invasive and expensive.

More recently, the diagnosis of choledochal cyst has been made with CT and ultrasound. These methods are typically used when biliary obstruction or an abdominal mass is suspected, and will almost always identify the cystic mass (*12,14–17*). However, although a specific

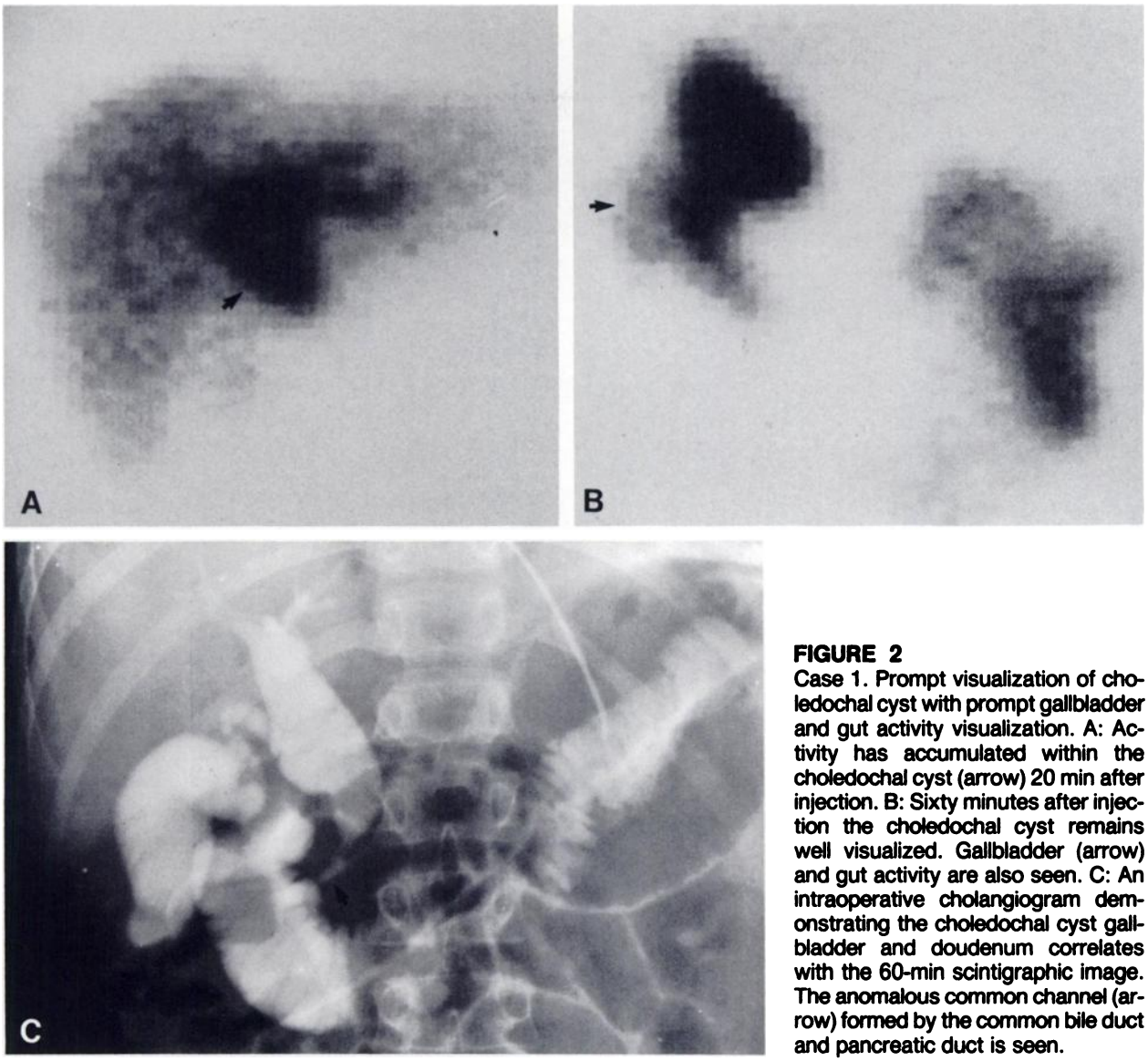

FIGURE 2
Case 1. Prompt visualization of choledochal cyst with prompt gallbladder and gut activity visualization. A: Activity has accumulated within the choledochal cyst (arrow) 20 min after injection. B: Sixty minutes after injection the choledochal cyst remains well visualized. Gallbladder (arrow) and gut activity are also seen. C: An intraoperative cholangiogram demonstrating the choledochal cyst gallbladder and doudenum correlates with the 60-min scintigraphic image. The anomalous common channel (arrow) formed by the common bile duct and pancreatic duct is seen.

diagnosis of choledochal cyst may be made by computed tomography (CT) or ultrasound in some cases (*20,23,31*), in other cases these anatomically oriented tests may be unable to differentiate choledochal cysts from other cystic lesions such as simple hepatic cysts, pancreatic pseudocysts, duodenal duplication cysts, and hepatic artery aneurysms, all of which may present with symptoms of pain, jaundice, and abdominal mass.

Since 1970, nuclear medicine methods have been used to diagnose choledochal cyst, first with iodine-131 Rose Bengal, and later with [^{99m}Tc]IDA compounds (*12,13,15,17,25,28–32*). The imino-diacetic acid family of ^{99m}Tc ligands provide good imaging statistics and allow functional assessment of the biliary tract even with bilirubin levels >20 mg % (*34–37*). These radiopharmaceuticals allow a definitive diagnosis of choledochal cyst, and frequently define other anatomic or physiologic abnormalities of the hepatobiliary system.

In the past, the "classic" scintigraphic appearance of choledochal cyst was that of an early photon deficient area that "filled-in" on late (>2 hr) images (*12,25,28,30–32*). This appearance of late cyst activity was seen in only three of our 12 cases (Fig. 1C), although had delayed images been obtained in Case 2, visualization might have been seen. The pattern seen more frequently in our experience, occurring in seven of our 12 cases, was that of early appearance of activity within grossly dilated extrahepatic ducts (Figs. 3B, 2A). Why this pattern was found much more frequently in our series is not clear. Regardless, the prompt visualization of the extrahepatic ducts reflects the fact that the obstruction that may have contributed to the formation of the choledochal cyst in utero does not necessarily persist. Note that there was not a close correlation between the time of visualization of the extrahepatic activity and the presence of jaundice. One of the four

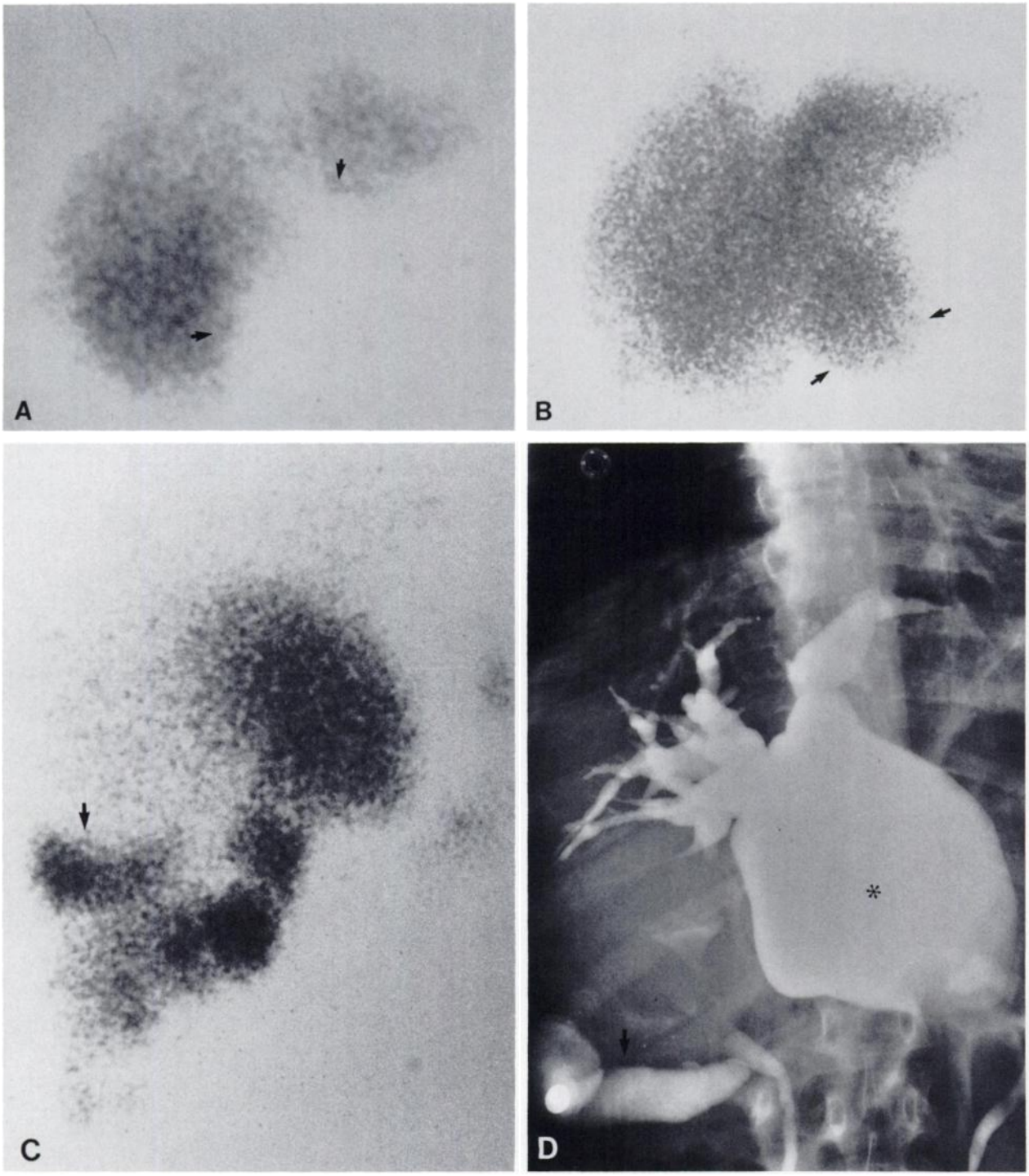

FIGURE 3
Case 4. Delayed visualization of choledochal cyst. A: Initially, there is a suggestion of photopenic mass (arrows) in the porta hepatis, but no evidence of extrahepatic activity. B: A subsequent image, obtained 4 hr after injection, demonstrates activity within the choledochal cyst (arrows). Gut and gallbladder activity are not yet visible. C: At 24 hr after injection, both gut and gallbladder activity (arrow) are visible. D: A percutaneous transhepatic cholangiogram demonstrates the choledochal cyst (asterisk) and the gallbladder (arrow). Contrast within the renal collecting system and ureters is also seen.

patients with jaundice demonstrated prompt extrahepatic ductal visualization while delayed visualization was seen in one of the five patients without jaundice.

In the final pattern, the cyst never accumulates radionuclide because of persistent or recurrent high grade biliary obstruction. In this situation, a definitive diagnosis of choledochal cyst cannot be made scintigraphically. Unfortunately, this pattern seems particularly

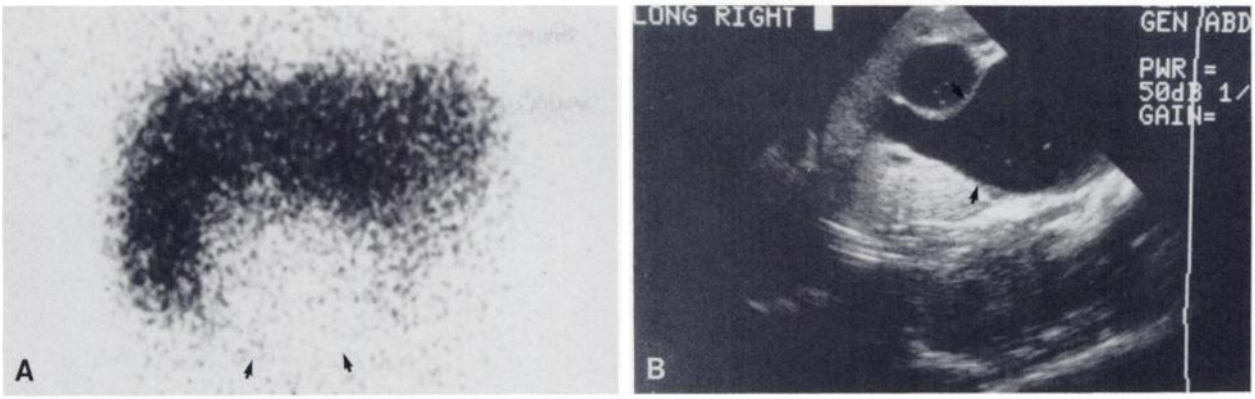

FIGURE 4
Case 10. Persistent nonvisualization of choledochal cyst. A: Twenty-four hours after injection, no bile duct activity is seen. The diagnosis of choledochal cyst cannot be established, although the relatively photopenic area in the porta hepatis (arrows) suggests the diagnosis. B: A longitudinally oriented sonogram of the porta hepatis identifies the superior aspect of the choledochal cyst communicating with the remainder of the biliary tract (arrows). The separate cystic structure anterior to the choledochal cyst is the gallbladder.

likely to occur in infants who present with jaundice. Our case of complete nonvisualization of the choledochal cyst at 24 hr occurred in a jaundiced infant 6 wk of age. In a second case, also involving a 6-wk-old girl, only minimal activity was seen in the choledochal cyst at 24 hr. In the former case, phenobarbital was not administered prior to the scintigram and it is possible that extrahepatic activity would have been seen had the hepatocellular function been improved with this medication. Nevertheless, it seems likely that in infants with choledochal cyst an incorrect diagnosis of biliary atresia can be made if the interpreter is unaware of the presence of a cystic or avascular mass in the porta hepatis based on ultrasound or blood-pool phase scintigraphy.

In our series, as in a previously reported series (*28*), delayed or nonvisualization of the gallbladder was very common. Cholecystectomy is frequently performed at the time of choledochal cystectomy. Of the six patients in our series with delayed visualization of the gallbladder, three were found to have chronic cholecystitis, and three were found to have normal gallbladders. Therefore, although in some cases cholecystitis may have contributed to the delayed gallbladder visualization, in others this may have simply represented an alteration in the pattern of biliary flow due to the reservoir effect of the choledochal cyst. The same phenomenon may also explain the delayed gut visualization in two patients without jaundice and with prompt visualization of the choledochal cyst.

In five of our cases the intrahepatic ducts appeared prominent and in four of these cases dilated ducts were confirmed by ultrasound examination. The lack of jaundice in one of these cases suggests that the intrahepatic dilatation is part of the congenital anomaly and not just a manifestation of extrahepatic biliary obstruction. The association between congenital intrahepatic ductal dilatation (i.e., Carolis' disease) and congenital extrahepatic ductal dilatation (i.e. choledochal cyst) has been previously described (*12,34*). Han et al. described this same scintigraphic appearance in two cases previously. However, in the last of our cases, the prominent ducts were actually proven to be normal by sonography, emphasizing scintigraphy's lack of specificity in assessing mild to moderate changes in ductal diameter.

We believe that hepatobiliary scintigraphy with [^{99m}Tc]IDA agents provides important information in choledochal cyst in the majority of cases and should be utilized whenever CT and sonography are unable to establish a definitive diagnosis. In our experience, prompt appearance of radionuclide within the dilated extrahepatic bile duct representing the choledochal cyst is the most common scintigraphic appearance. In other cases, delayed images may be required to demonstrate this accumulation of activity and establish the diagnosis. In some cases, however, particularly in infants, no extrahepatic activity will be seen even on 24-hr delayed images. In these cases either ultrasound or flow/blood-pool imaging is required to distinguish choledochal cyst from biliary atresia.

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