


# Biliary Atresia


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## Learning Overview

- 01 Introduction**  
What is biliary atresia?
- 02 Sonographic features**  
How do we diagnose biliary atresia on ultrasound?
- 03 Case Studies**  
Positive diagnoses of biliary atresia
- 04 Differential diagnoses**  
What else could it be if it's not biliary atresia?



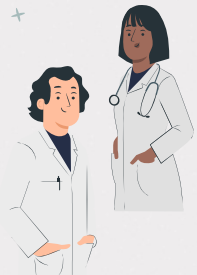
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## Clinical Presentation

Neonate (2 weeks – 2 months old) presenting with:

- Jaundice
- Clay coloured stools
- Conjugated hyperbilirubinemia
- Elevated GGT

**What could cause the above findings?**



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## Clinical Presentation

Causes of conjugated hyperbilirubinemia

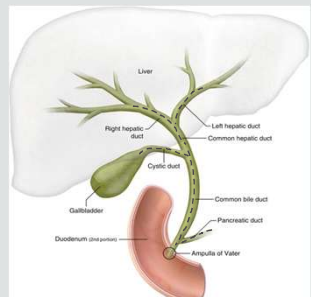
Genetic	Infections	Structural	Immune	Others
<ul style="list-style-type: none"> <li>• Progressive familial intrahepatic cholestasis</li> <li>• Alagille syndrome</li> <li>• Alpha-1-antitrypsin deficiency</li> <li>• Bile acid synthesis defects</li> <li>• Neonatal sclerosing cholangitis (CLDN1 and DCDC2 defects)</li> <li>• ARC syndrome (arthrogryposis, renal dysfunction and cholestasis)</li> <li>• Congenital hepatic fibrosis</li> <li>• Other</li> </ul>	<ul style="list-style-type: none"> <li>• CMV</li> <li>• Sepsis</li> <li>• Congenital syphilis</li> <li>• E coli UTI</li> <li>• Rubella</li> <li>• Herpes simplex virus</li> </ul>	<ul style="list-style-type: none"> <li>• Choledochal cyst</li> <li>• Caroli disease</li> </ul>	<ul style="list-style-type: none"> <li>• Gestational alloimmune liver disease</li> </ul>	<ul style="list-style-type: none"> <li>• Idiopathic neonatal hepatitis</li> <li>• Prematurity</li> <li>• Total parenteral nutrition</li> <li>• Acute liver failure</li> <li>• Drugs</li> <li>• Toxins</li> </ul>

**Biliary Atresia**

Patel 2022

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## Biliary Tree Anatomy

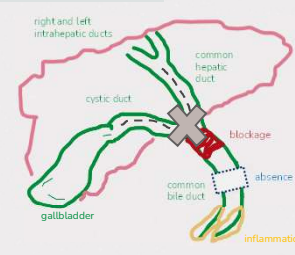


- The biliary tree is a branching ductal system that transports bile from the liver to the duodenum
- Many variations of the biliary tree but this anatomy is present in approximately 60% of the population

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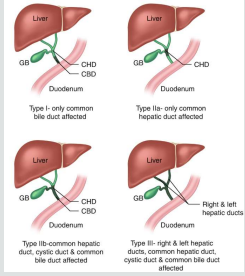
## What is Biliary Atresia (BA)?

- BA is a fibro-inflammatory and obliterative disease of the biliary tract in neonates
- This is characterised by a blockage, absence, or deformity of the biliary tree
- This causes cholestasis which leads to scarring and can progress to liver cirrhosis if prolonged



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## Biliary Atresia types



3 different types of biliary atresia

Type I: Obliteration of the CBD (proximal bile duct is patent)

Type IIa: Obliteration of CHD (cystic and CBD are patent)

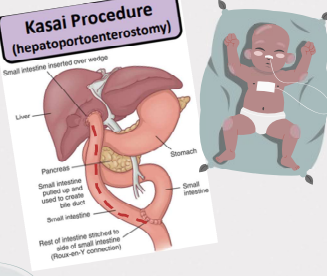
Type IIb: Obliteration of cystic duct, CHD and CBD

Type III: Obliteration of left and right IHD, CHD + CBD

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## Why is the diagnoses of BA important?

- BA could lead to progressive liver cirrhosis and even death by 1-2 years of age if left untreated
- Early diagnosis and timely surgical intervention is crucial to facilitate bile drainage and prolong liver survival
- Main treatment includes Kasai portoenterostomy
- Other treatment include liver transplantation



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## Ultrasound features of BA



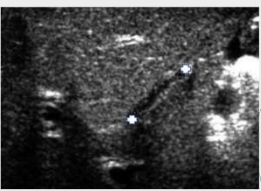
Sonographic feature	Definition	Positive criteria	Negative criteria
Gallbladder abnormalities	Abnormalities pertaining the gallbladder particularly the length, lumen size, integrity of the mucosal lining, degree of its contraction after feeding	<ul style="list-style-type: none"> <li>&lt;1.5cm in length of a fully filled gallbladder lumen</li> <li>Lack of smooth and echogenic mucosal lining of the gallbladder</li> <li>No contraction observed post feeding</li> </ul>	<ul style="list-style-type: none"> <li>&gt;1.5cm in length of a fully filled gallbladder lumen with a smooth and echogenic mucosal lining</li> <li>Incompletely filled gallbladder lumen with smooth and complete echogenic mucosal lining</li> </ul>
Triangular cord sign	The thickness of the echogenic anterior of the right portal vein with or without hepatic artery	<ul style="list-style-type: none"> <li>Thickness of &gt;2mm not including the hepatic artery OR</li> <li>Thickness of &gt;4mm including the hepatic artery</li> </ul>	<ul style="list-style-type: none"> <li>Thickness of &lt;2mm not including the hepatic artery</li> <li>Thickness of &lt;4mm including the hepatic artery</li> </ul>
Porta hepatis macro- or microcyst	Cysts located in front of the right portal vein at the porta hepatis	Presence of macro- or microcysts	Absence of macro- or microcysts
Hepatic hilar lymph node	Lymph nodes located at the porta hepatis, around the hepatoduodenal ligament	Presence of hepatic hilar lymph node	Absence of hepatic hilar lymph node
Hepatic artery diameter	Measurement of the right hepatic artery diameter at its proximal segment, running parallel to the right portal vein	A hepatic artery diameter of 2.1mm-2.5mm	A hepatic artery diameter of 1.5mm-1.9mm
Hepatic subcapsular flow	Vascular structures extending to the capsular surface on colour Doppler US	Presence of hepatic subcapsular flow	Absence of hepatic subcapsular flow

Zhou & Zhou 2022

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## Gallbladder abnormalities

Sonographic feature	Definition	Positive criteria
Gallbladder abnormalities	Abnormalities pertaining the gallbladder particularly the length, lumen size, integrity of the mucosal lining, degree of its contraction after feeding	<ul style="list-style-type: none"> <li>&lt;1.5cm in length of a fully filled gallbladder lumen</li> <li>Lack of smooth and echogenic mucosal lining</li> <li>No contraction observed post feeding</li> </ul>

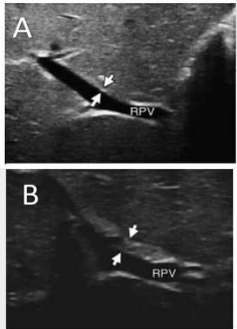


Zhou & Zhou 2022      Kendrick et al. 2003

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## Triangular cord sign

Sonographic feature	Definition	Positive criteria
Triangular cord sign	Thick echogenic tissue anterior of the right portal vein with or without hepatic artery	<ul style="list-style-type: none"> <li>Thickness of &gt;2mm not including the hepatic artery OR</li> <li>Thickness of &gt;4mm including the hepatic artery</li> </ul>




Zhou & Zhou 2022      Zhou et al. 2015

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## Macro- or microcysts

Sonographic feature	Definition	Positive criteria
Porta hepatis macro- or microcysts	Cysts located in front of the right portal vein at the porta hepatis	<ul style="list-style-type: none"> <li>Presence of macro- or microcysts</li> <li>≤2.8cm in size</li> </ul>

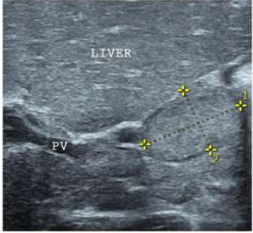


Zhou & Zhou 2022      Zhou et al. 2012

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### Enlarged hepatic hilar lymph nodes

Sonographic feature	Definition	Positive criteria
Hepatic hilar lymph node	Lymph nodes located at the porta hepatis, around the hepatoduodenal ligament	<ul style="list-style-type: none"> <li>Presence of hepatic hilar lymph node</li> <li>6mm LN length</li> </ul>



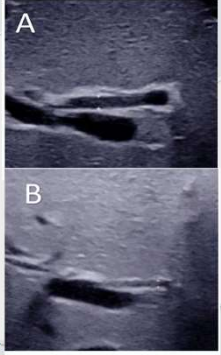
Zhou & Zhou 2022

Weng et al. 2019

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### Hepatic artery diameter

Sonographic feature	Definition	Positive criteria
Hepatic artery diameter	Measurement of the right hepatic artery diameter at its proximal segment running parallel to the right portal vein	<ul style="list-style-type: none"> <li>A hepatic artery diameter of 2.1mm-2.5mm</li> </ul>

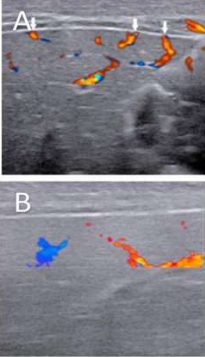


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### Hepatic subcapsular flow

Sonographic feature	Definition	Positive criteria
Hepatic subcapsular flow	Vascular structures extending to the capsular surface on colour Doppler US.	<ul style="list-style-type: none"> <li>Presence of hepatic subcapsular flow</li> </ul>




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### Which signs are most indicative of BA?

Sonographic feature	Definition	Positive criteria	Negative criteria	Diagnostic value of BA when positive
Gallbladder abnormalities	Abnormalities pertaining the gallbladder particularly the lumen, lumen size, integrity of the muscular lining, shape of its contraction after feeding	<ul style="list-style-type: none"> <li>&lt;1.5cm in length of a fully filled gallbladder lumen</li> <li>Lack of smooth and echogenic muscular lining of the gallbladder</li> <li>No contraction observed post feeding</li> </ul>	<ul style="list-style-type: none"> <li>&gt;1.5cm in length of a fully filled gallbladder lumen with a smooth and echogenic muscular lining</li> <li>Incompletely filled gallbladder lumen with smooth and complete echogenic muscular lining</li> </ul>	Strongly suggestive of BA
Triangular cord sign	The thickness of the echogenic anterior of the right portal vein with or without hepatic artery	<ul style="list-style-type: none"> <li>Thickness of &gt;2mm not including the hepatic artery duct</li> <li>Thickness of &lt;2mm including the hepatic artery</li> </ul>	<ul style="list-style-type: none"> <li>Thickness of &lt;2mm not including the hepatic artery</li> <li>Thickness of &gt;2mm including the hepatic artery</li> </ul>	Strongly suggestive of BA
Porta hepatis macro- or microcystic hepatic hilar lymph node	Cysts located in front of the right portal vein at the porta hepatis, around the hepatoduodenal ligament	<ul style="list-style-type: none"> <li>Presence of macro- or microcysts</li> </ul>	Absence of macro- or microcysts	Strongly suggestive of BA
Hepatic artery diameter	Measurement of the right hepatic artery diameter at its proximal segment, running parallel to the right portal vein	<ul style="list-style-type: none"> <li>Presence of hepatic hilar lymph node</li> <li>&amp; hepatic artery diameter of 2.1mm-2.5mm</li> </ul>	Absence of hepatic hilar lymph node	Possible BA
Hepatic subcapsular flow	Vascular structures extending to the capsular surface on colour Doppler US.	<ul style="list-style-type: none"> <li>Presence of hepatic subcapsular flow</li> </ul>	Absence of hepatic subcapsular flow	Not recommended for diagnosis alone



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### The literature

Napolitano et al. 2020

Ultrasound findings	Sensitivity	Specificity	Diagnostic odds ratio	Positive likelihood ratio	Negative likelihood ratio
Triangular cord sign	0.68 (0.57-0.78)	0.95 (0.92-0.97)	40.2 (20.8-70.8)	13.0 (8.3-19.7)	0.34 (0.23-0.46)
Gallbladder abnormality	0.79 (0.70-0.86)	0.81 (0.73-0.86)	16.5 (8.1-30.1)	4.2 (2.9-5.9)	0.27 (0.17-0.38)
Gallbladder absence	0.20 (0.13-0.29)	0.98 (0.93-1.00)	17.5 (4.0-50.1)	14.2 (3.4-40.5)	0.81 (0.73-0.88)
Gallbladder length	0.72 (0.59-0.81)	0.79 (0.70-0.86)	10.1 (5.5-16.9)	3.5 (2.5-5.0)	0.36 (0.24-0.50)
Triangular cord sign and gallbladder abnormality	0.87 (0.51-0.97)	0.90 (0.68-0.86)	99.0 (7.8-441.0)	10.0 (2.7-28.8)	0.19 (0.03-0.54)
Hepatic artery enlargement	0.82 (0.71-0.89)	0.70 (0.68-0.86)	12.8 (3.1-36.1)	2.9 (1.5-5.3)	0.28 (0.14-0.51)

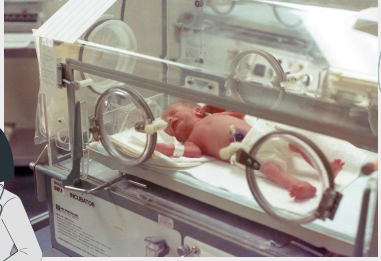

**Coexisting sonographic signs lead to confidence in BA diagnosis**



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### Case study 1

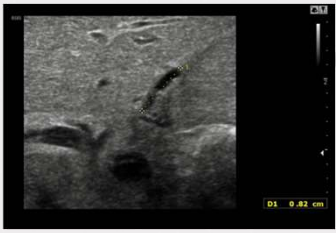
5-week-old male with conjugated hyperbilirubinemia, poor weight gain, deranged LFTs for investigation. ? To look at the liver and gallbladder

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### Case study 1

5-week-old male with conjugated hyperbilirubinemia, poor weight gain, deranged LFTs for investigation. ? To look at the liver and gallbladder




- Gallbladder measures 8mm in length
- The gallbladder wall appears irregular
- Gallbladder lumen size appears small

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### Case study 1

5-week-old male with conjugated hyperbilirubinemia, poor weight gain, deranged LFTs for investigation. ? To look at the liver and gallbladder

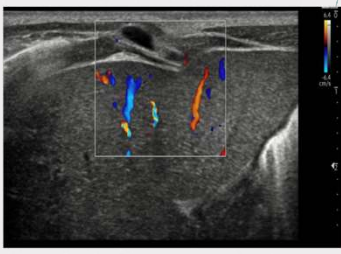


- Echogenic thick band of tissue anterior to the main portal vein measuring 2.4mm
- Triangular cord sign

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### Case study 1

5-week-old male with conjugated hyperbilirubinemia, poor weight gain, deranged LFTs for investigation. ? To look at the liver and gallbladder




- Vascular structures extending to the capsular surface on colour Doppler US

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### Case study 1

5-week-old male with conjugated hyperbilirubinemia, poor weight gain, deranged LFTs for investigation. ? To look at the liver and gallbladder




- **Biopsy results still pending**

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### Case study 2

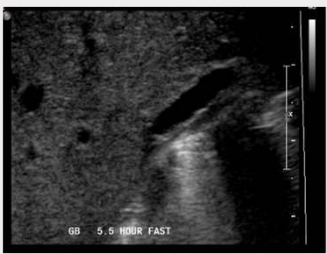
2-month-old female with conjugated bilirubinaemia ? Biliary atresia



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### Case study 2

2-month-old female with conjugated bilirubinaemia ? Biliary atresia



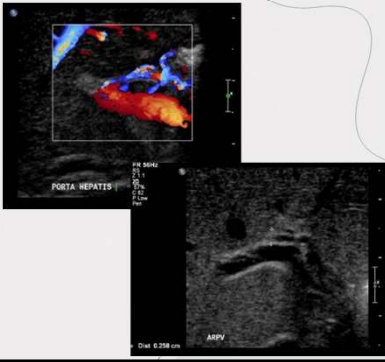
- Small gallbladder lumen, irregular gallbladder wall/contour despite adequate fasting

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### Case study 2

2-month-old female with conjugated bilirubinaemia ? Biliary atresia

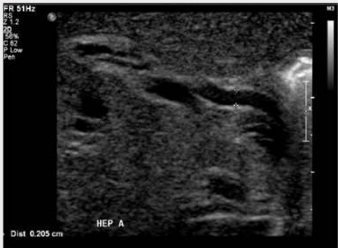


- Possible triangular cord sign anterior to the portal vein measuring 2.6mm

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### Case study 2

2-month-old female with conjugated bilirubinaemia ? Biliary atresia

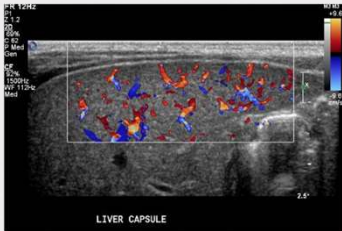


- Hepatic artery measuring 2mm in AP diameter
- Possible hepatic artery hypertrophy

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### Case study 2

2-month-old female with conjugated bilirubinaemia ? Biliary atresia

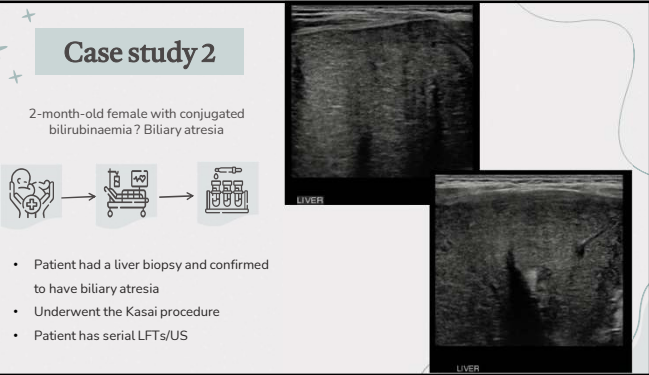


- Hepatic subcapsular flow
- Vascular structures extending to the capsular surface on colour Doppler US

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### Case study 2

2-month-old female with conjugated bilirubinaemia ? Biliary atresia




- Patient had a liver biopsy and confirmed to have biliary atresia
- Underwent the Kasai procedure
- Patient has serial LFTs/US

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### Case study 3

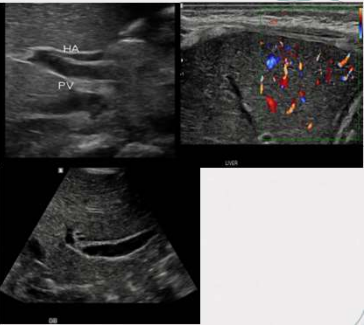
Ex-34+4 week baby in NICU with Hx of multiple bowel perforations. Jejunostomy. Hyperbilirubinemia ? Biliary atresia / obstruction



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### Case study 3

Ex-34+4 week baby in NICU with Hx of multiple bowel perforations. Jejunostomy. Hyperbilirubinemia ? Biliary atresia / obstruction


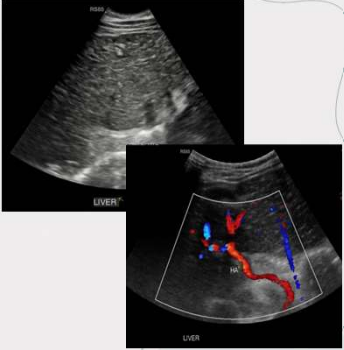


- No obvious triangular cord sign
- No obvious HA hypertrophy
- GB appears normal
- Potential hepatic subcapsular flow
- Possible absent CBD

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### Case study 3

Ex-34+4 week baby in NICU with Hx of multiple bowel perforations. Jejunostomy. Hyperbilirubinemia? Biliary atresia / obstruction

- Patient had a liver biopsy and confirmed to have biliary atresia
- Underwent the Kasai procedure
- Patient has serial LFTs/US

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### Cystic biliary atresia (CBA) Vs Choledochal cysts (CC)

What's the difference?

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### Cystic biliary atresia (CBA) Vs Choledochal cysts (CC)

- CBA is a relatively uncommon but a notable variant of biliary atresia
- Cystic changes along biliary tree
- This accounts for approximately 5-10% of biliary atresia cases

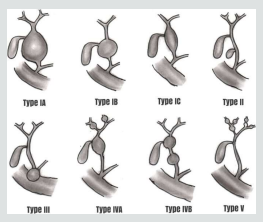


Sonographic feature	Definition	Positive criteria
Porta hepatis macro- or microcysts	Cysts located in front of the right portal vein at the porta hepatis	<ul style="list-style-type: none"> <li>• Presence of macro- or microcysts</li> <li>• ≤2.8cm in size</li> </ul>

Zhou et al. 2012

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### Cystic biliary atresia (CBA) Vs Choledochal cysts (CC)

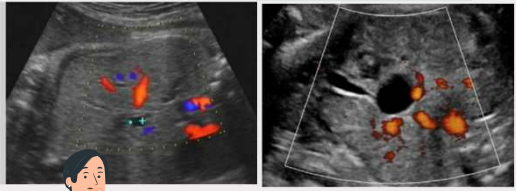


- CC represent congenital cystic dilations of the biliary tree
- There are different types characterised by the Todani classification

Hando et al. 2022

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### Cystic biliary atresia (CBA) Vs Choledochal cysts (CC)

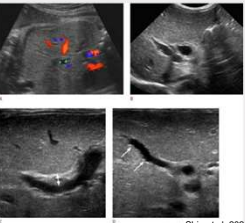


**How do we tell the difference on Ultrasound?**

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### Cystic biliary atresia (CBA) Vs Choledochal cysts (CC)

- CC will present as larger cysts size typically >2.8cm with absence of BA signs: triangular cord sign >4mm + abnormal GB morphology
- 0.6cm cyst at the region of the porta hepatis
- Periportal thickening with echogenic triangular band of fibrous tissue measuring 4.8mm
- Irregular gallbladder wall and small lumen size



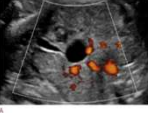
**Patient was confirmed to have CBA and underwent the Kasai procedure**

Shin et al. 2021


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## Cystic biliary atresia (CBA) Vs Choledochal cysts (CC)


- CC will present as larger cysts size typically >2.8cm with absence of BA signs: triangular cord sign >4mm + abnormal GB morphology



Shin et al. 2021



- 3.4 cm cyst at the region of the porta hepatis
- No periportal echogenicity with a triangular cord thickness of 1.9mm
- Distended gallbladder with internal sludge

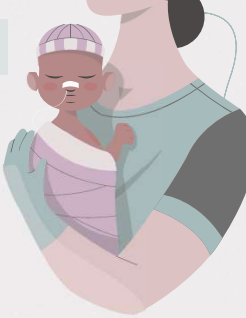


**Patient was confirmed to have CC**

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## Learning outcomes

- Gain understanding of the presentation and pathophysiology of biliary atresia
- Be able to identify and become familiar with sonographic findings of biliary atresia
- Know the key imaging features to differentiate between biliary atresia and choledochal cysts



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## References

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