Biliary Atresia

SUCHY Chapter 11 Section 2

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Early recognition these babies is particularly critical for optimal intervention.

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Portoenterostomy before 60 days of age

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Ideally B.A should be identified

by the time of the first well-baby visit after discharge from the hospital.

Biliary Atresia

Nelson: Noncystic Obstructive Cholangiopathy

Suchy: The end result of a destructive , idiopathic , inflammatory process that affects intra and extrahepatic bile ducts leading to fibrosis and obliteration of the biliary tract and eventual development of biliary cirrhosis = Rapidly progressive fibro-oblitrative process

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- Is the most common cause of chronic cholestasis in infants and children.
- IS the most frequent indication for liver transplantation.

Infant with jaundice

- Indirect hyperbilirubinemia = Ind.Bil >80% of total

  Physiologic or Breast-feeding jaundice

- Direct hyperbilirubinemia = D.Bil > 20% of total or >1.5

Infant with cholestasis
General evaluation

- Clinical evaluation (Family Hx / Gestational Hx / Feeding Hx / Physical Ex / Stool color)
- Hepatic synthetic function (PT / INR)
- Cultures (B/C , U/C , CSF/C)
- Serum bile acid levels (Followed by qualitative analysis of urinary bile acid profile if abnormal)
- GGT
- Serum electrolytes (To exclude acidosis)

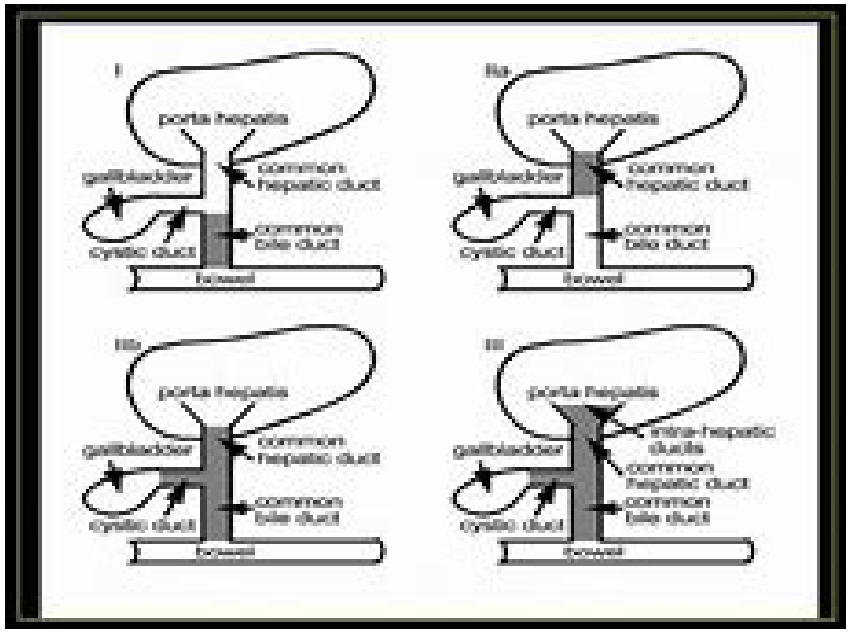
Specific evaluation (To R/O or confirm a specific Dx)

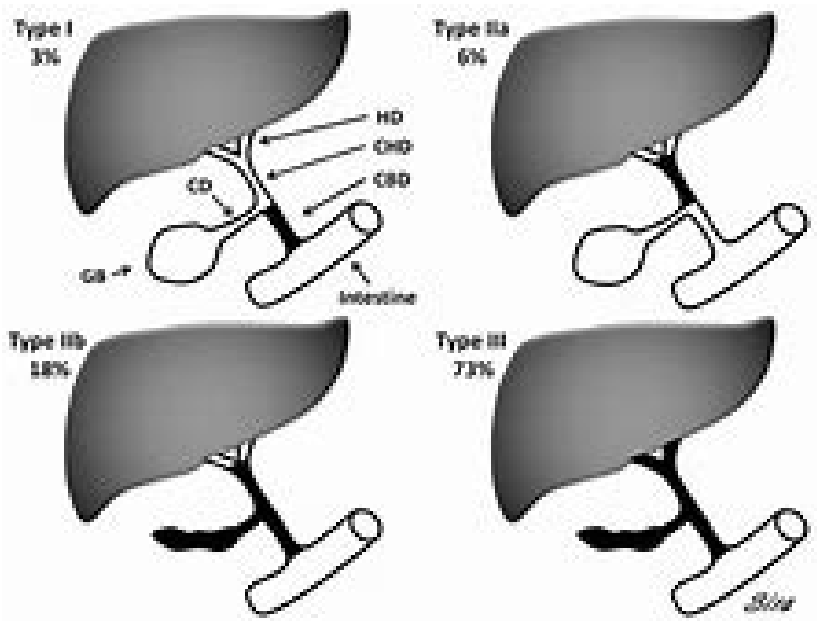
- α-1 antitrypsin phenotype
- T4 / TSH
- Sweat chloride-mutational analysis
- Ferritin-transferin concentration and saturation
- Metabolic screen (Urine-reducing substances , urine/serum aminoacids , UOA , succinyl acetone)
- HBS Ag , anti-HIV , VDRL (+/-)
- Abdominal US
- Liver Bx

The most common type

Peri / postnatal form:

Complete obstruction of extrahepatic bile ducts (EHBD) until porta hepatis





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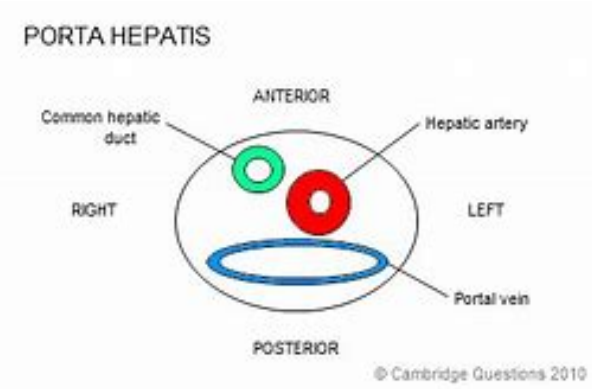
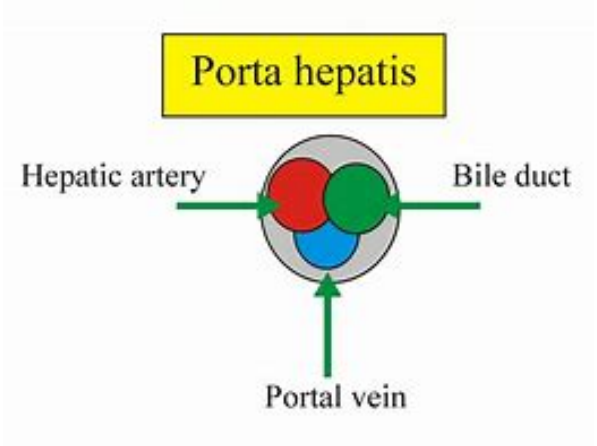
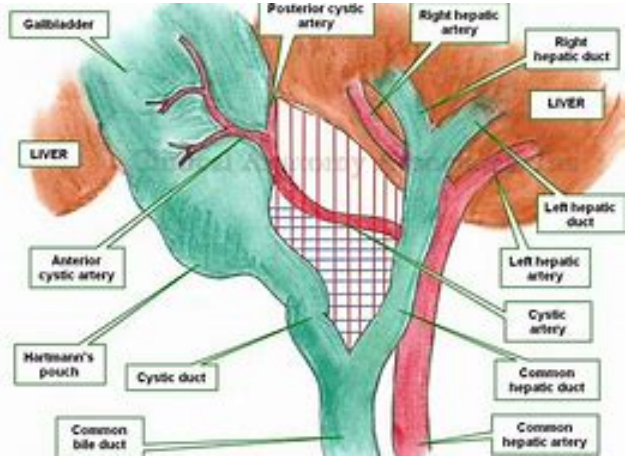
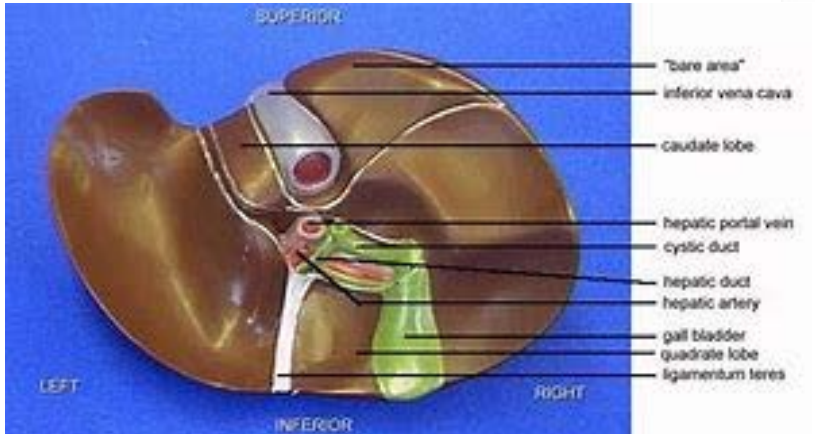
Porta hepatis









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90% of patients are normal at birth ----------> A progressive obstruction of ducts

o

↑

An immune or infective process

These 2 forms have not been distinguished on the basis of histology of porta hepatis

Fetal or embryonic form (10-20%)	Peri/postnatal form (80-90%)
1. Cholestasis is present from birth With no "jaundice-free intervals" 2. Bile duct remnants may not be detectable in the hepatic hilum. 3. There is 10-20% of associated malformations. 4. Has the greatest contribution from genetic factors in its pathogenesis.	May be result of an acquired obliteration.

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Extrahepatic anomalies reported in B.A

System	Anomalies
Spleen anomalies	*Polysplenia, double spleen, asplenia*
Portal vein anomalies	*Preduodenal position, absence, cavernomatous transformation*
Abdominal abnormalities	*Situs inversus, annular pancreas, duodenal/esophageal/jejunal atresia*
Cardiac anomalies	*Dextrocardia, atrial situs ambigus, ventricular inversion*
Immotile cilia syndrome(Kartagener)	
Renal anomalies	*Polycystic kidney, renal agenesis, hypoplastic kidneys*
Cleft palate	

When you thinck about B.A

In each neonate/ infant with

new icter or prolonged icter (after 8 weeks)

you should think about B.A

Clinical features

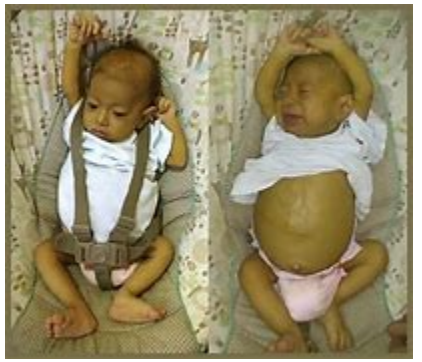
Symptomes:

- Patient usually is born at term , with normal BW , and appropriate weight gain early in the course.
- Jaundice (conjugated hyperbilirubinemia)
- Acholic stools
- Hepatomegaly
- FTT
- Pruritus
- Coagulopathy (+/-bleeding as a result of vit.K ↓)

Clinical manifestation

PH/Ex:

- **Hepatomegaly (with hard consistency because of fibrosis)**
- **splenomegaly**
- **If biliary cirrhosis > Ascites , wasting …**



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A differential diagnosis

Idiopathic Neonatal Intrahepatic Cholestasis

Must be differentiated from biliary atresia because the prognosis and Tx differ significantly.

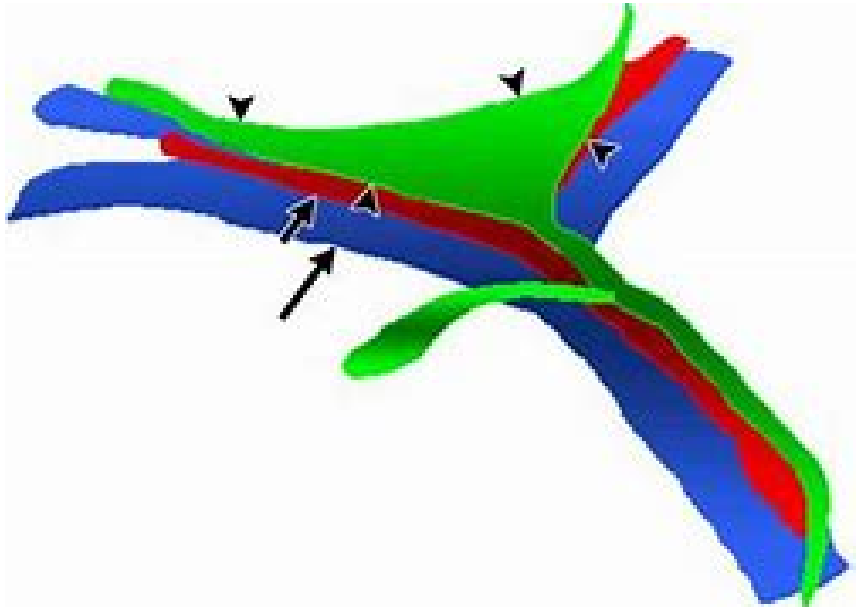
A differential diagnosis

Biliary Atresia	INH
Repeat in family : (-)	Familial occurrence : 20%
B.A = BW عالی	Prematurity/SGA is more common
Acholic stool : (+) مدفوع همیشه رنگی ، آترزی صفراوی را رد می کند. یافتن مایع آغشته به صفرا در انتوباسیون دوازدهه هم آترزی صفراوی را رد می کند.	Acholic stool can be seen in severe cases
Size/consistency of liver: abnormal	P/Ex of liver is normal (overall)
US	US
HIDA scan	HIDA
Liver Bx	Liver Bx

Us in B.A vs INH

B.A	INH
GB is absent or small **Triangular Cord Sign** Polysplenia +/- Vascular malformations +/-	Other Dx can be detected Findings in US are similar in: 1- INH 2- CF 3- TPN **GB is absent or small**

Triangular cord sign



HIDA scan in B.A (Scintigraphy)

HIDA scan = Hepatobiliary Iminodiacetic Acid scan = Cholescintigraphy = Hepatobiliary scintigraphy

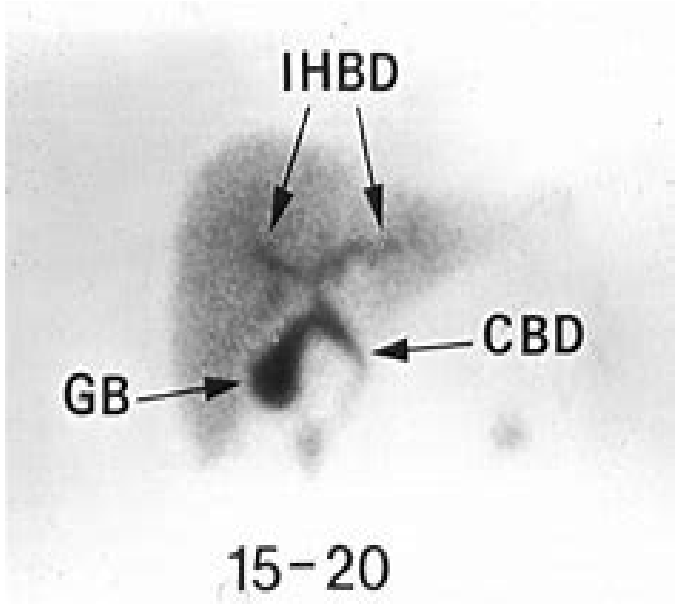
1- Is sensitive

2- But no specific

Scintigraphy = Sensitive

3- Cant detect vascular malformations

4- Need to 5 days waiting



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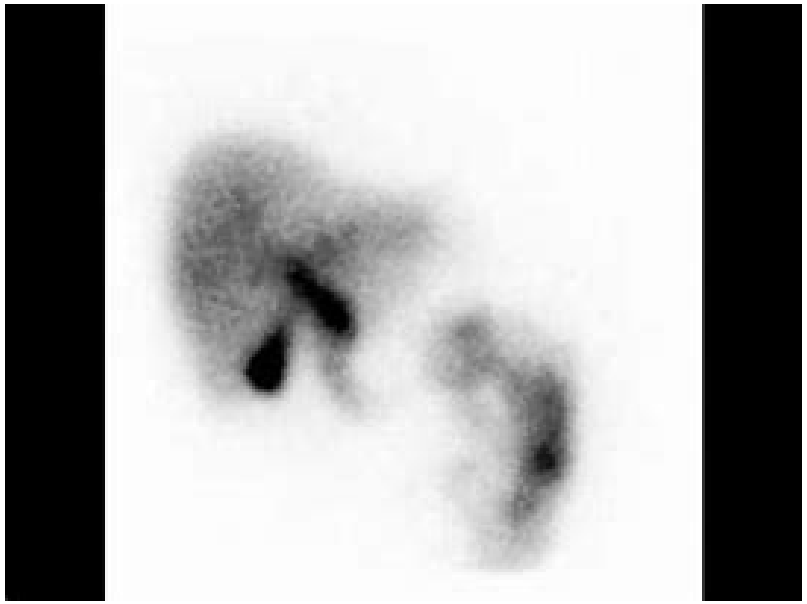
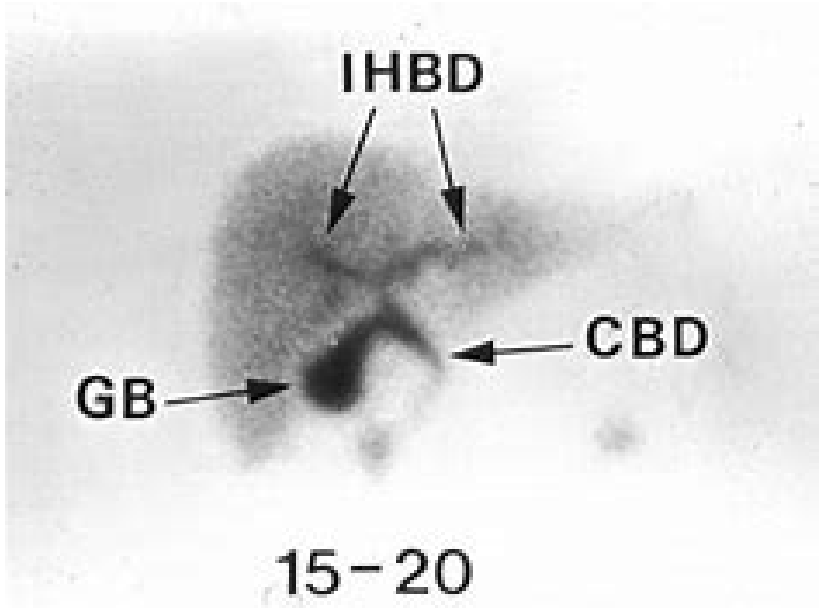
Normal vs B.A





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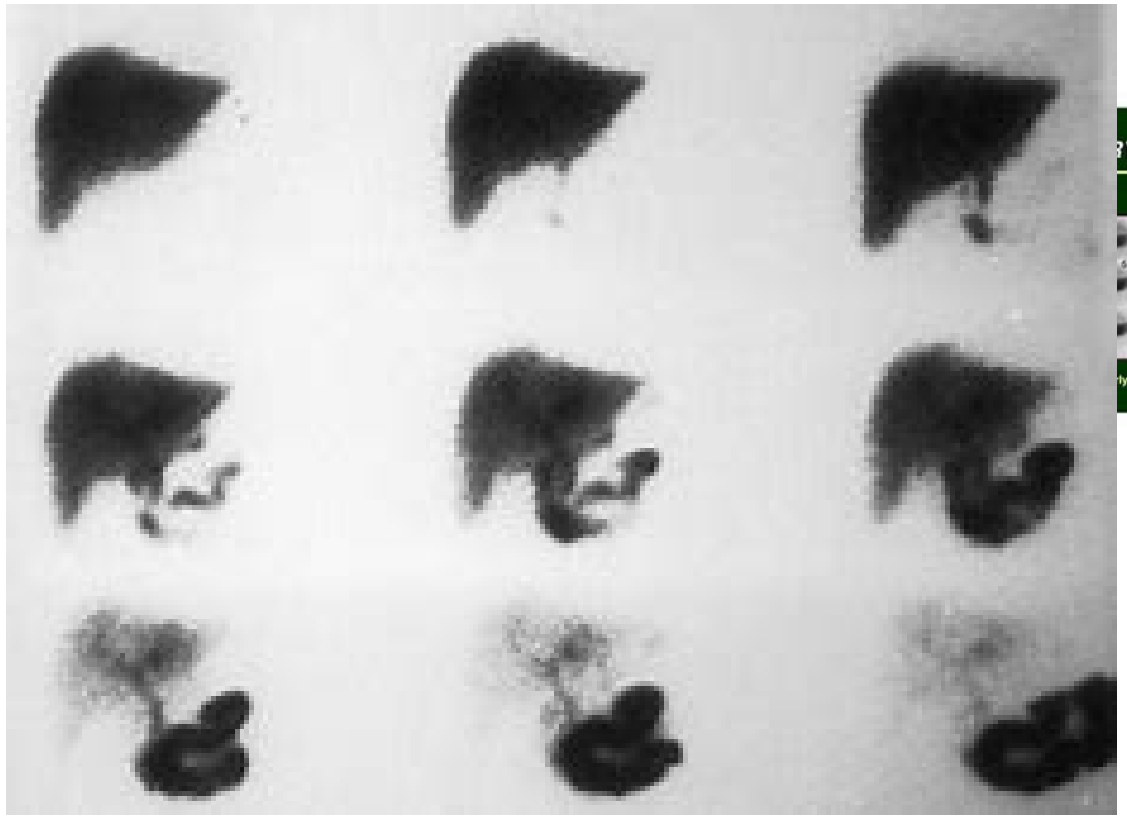

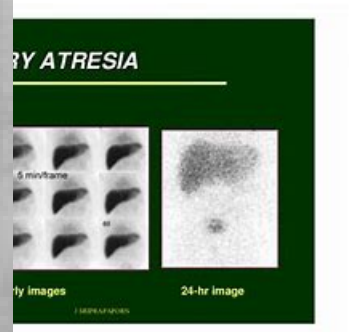

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Liver Bx

B.A	INH
1. Proliferation of biliary ductules 2. Biliary plugs 3. Fibrosis/Edema of portal space(triad) 4. ***Gient cell***	1. Severe and diffused hepatocellular disease = Focal necrosis of hepatocytes 2. Architecture o lobules is disorganized 3. Severe inflammatory infiltration 4. Minimal changes in ductules 5. ***Gient cell*** α1-AT ↓ , Galactosemia , Intrahepatic Cholestasis are similar to INH in histopathology.

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Treatment