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Approach to Management of Cholestatic Jaundice

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ABSTRACT

Cholestatic jaundice is a common presenting feature of neonatal hepatobiliary and metabolic dysfunction. Any infant who remains jaundiced beyond age 2 to 3 weeks should have the serum bilirubin level fractionated into a conjugated (direct) and unconjugated (indirect) portion in neonates mainly undirect bilirubin.

In cholestasis, the primary failure is of bilirubin excretion, resulting in excess conjugated bilirubin in the bloodstream and decreased bile salts in the gastrointestinal (GI) tract. Because of inadequate bile in the GI tract, there is malabsorption of fat and fat-soluble vitamins (A, D, E, and K), leading to vitamin deficiency, inadequate nutrition, and growth failure.

Our case is interesting present in first day of life with conjugate bilirubin which made us look for differential diagnosis, no family history of metabolic disease, 1st consequently, no dysmorphic feature, admitted in NICU as case of cholecystic jaundice for farther investigation, all investigations were done, and patient sent to higher center for farther management.

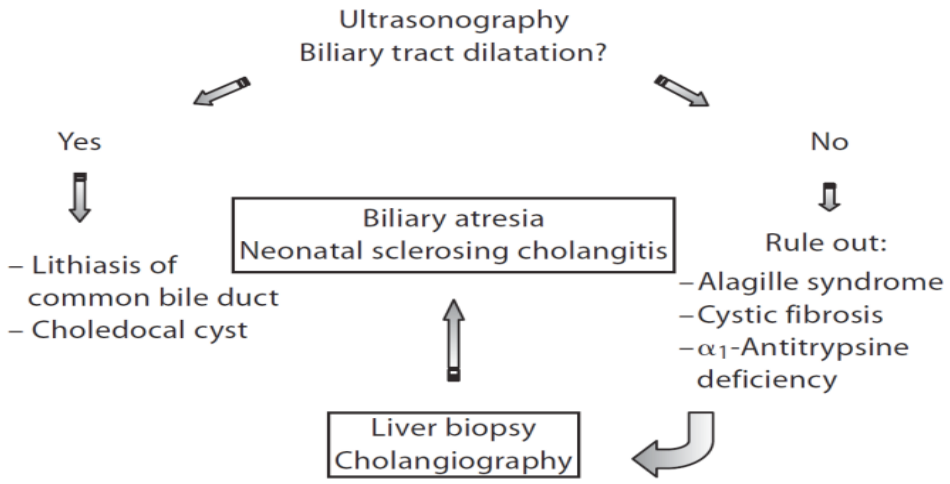
Keywords
Cholestatic jaundice, Parenchymal cells, Liver.

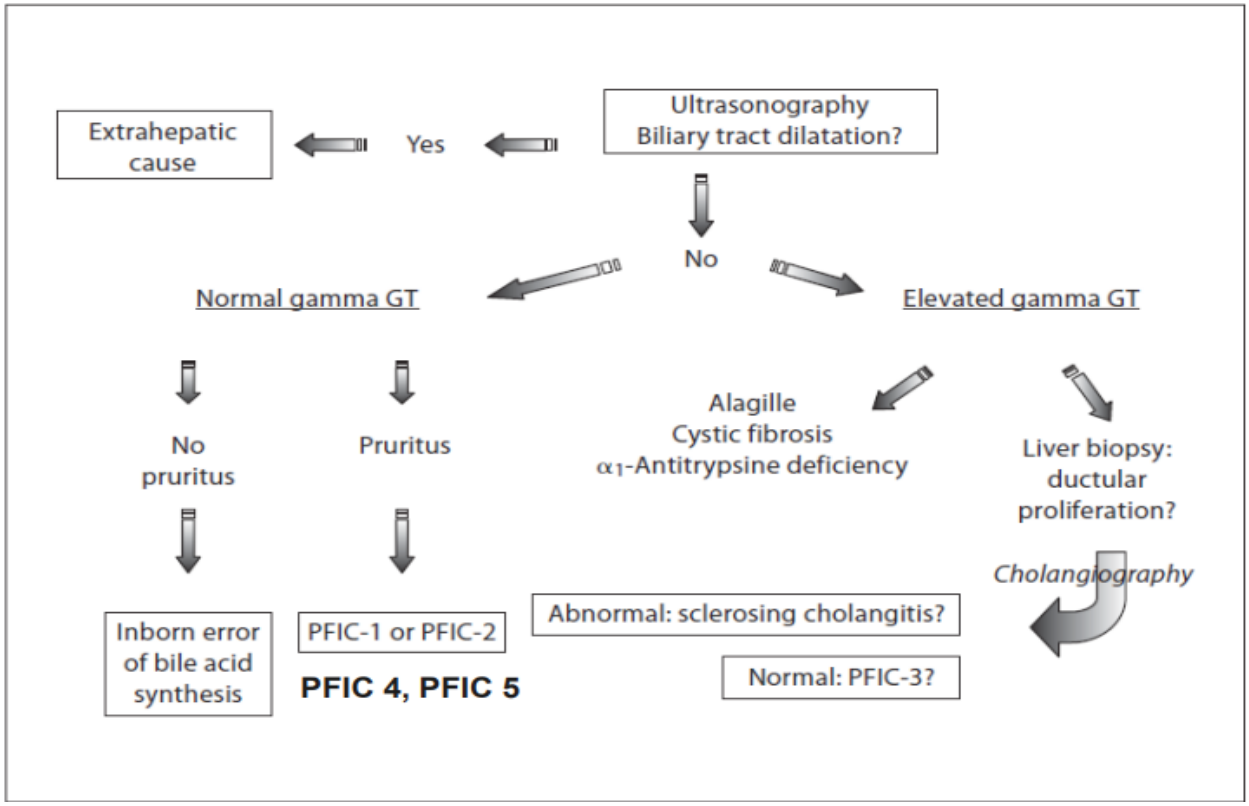
Introduction

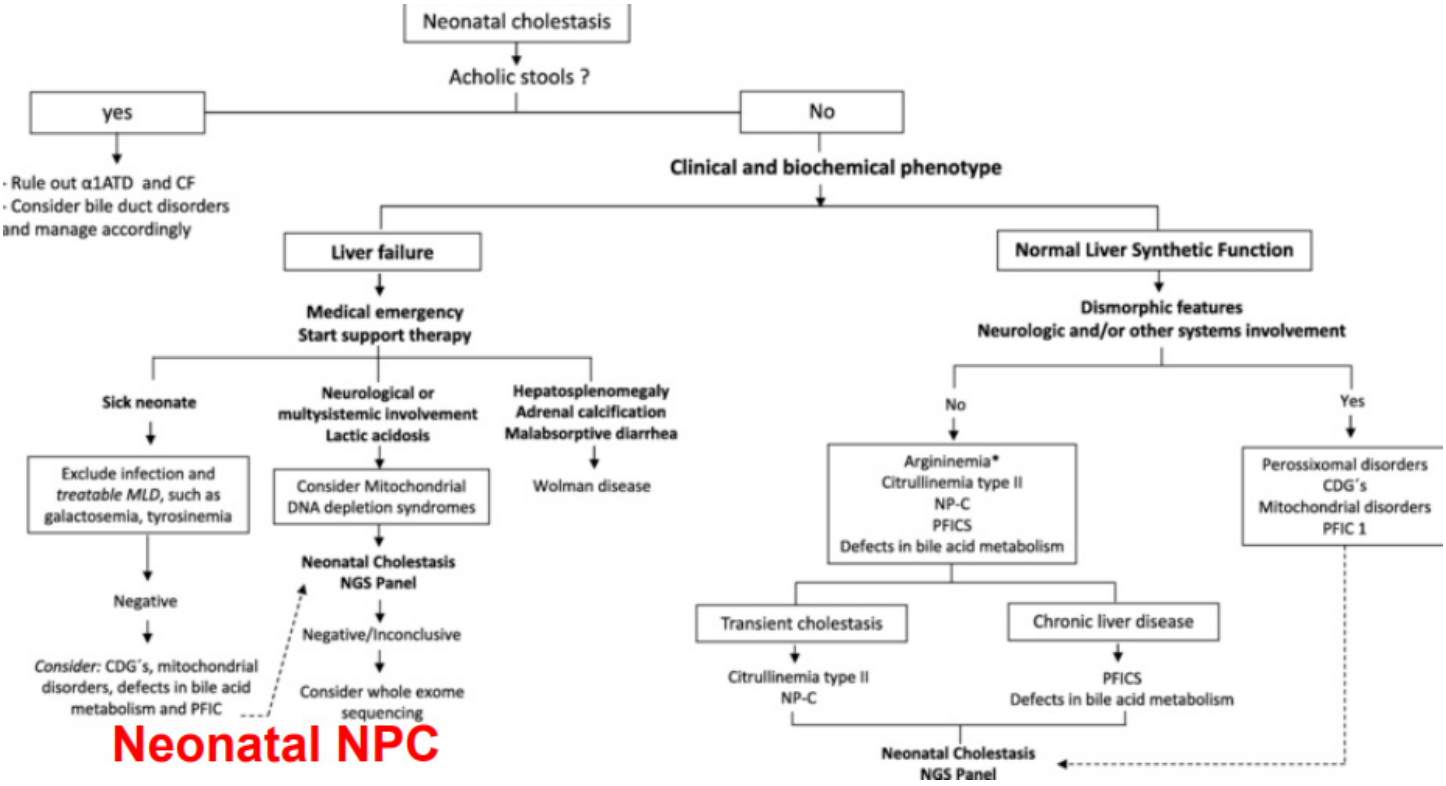
Cholestatic jaundice can thus be classified into intrahepatic or extrahepatic cholestasis, depending upon the level of obstruction to bile flow. Intrahepatic cholestasis or functional cholestasis can be due to a disease involving the liver c and/or the intrahepatic bile ducts. Intrahepatic cholestasis can be further sub classified as interlobular (disease of liver parenchymal cells and transporter molecules) and extra lobular (disease involving intrahepatic bile ducts) cholestasis. Extrahepatic cholestasis or obstructive cholestasis is due to excretory block outside of the liver, along with the extrahepatic bile ducts [1].

Pathogenesis of Cholecystic Jaundice

Differential Diagnosis	Organomegally	Liver Failure	Liver Enzyme	Jaundice stool color	Clinical Condition
Sepsis -Metabolic -Endocrine	+/-	+++	Mild-high ALT GGT	Variable ++	Ill looking infant
Syndromes -Storage diseases -Endocrine	++	+	Mild-high ALT GGT	++ Variable	Dysmorphic Infant
*Extrahepatic -BA -Choledochal cyst	- -/+	- -/+	Mild-high High GGT Bile acids	+++ Pale	Normal Infant Irritable
*Intrahepatic -Bile acid synthesis or transport	-	-/+	Mild-high NL GGT	++ Normal	Normal Infant







Our Case

Full term, LSCS to prim mother 20-year-old Saudi, no consequently, baby cried immediately shift beside the mother. Mother blood group O+(Positive) and the baby B+(Positive) and cord bilirubin was very high and combs test negative so admitted in NICU for phototherapy and consequent serum bilirubin.

Physical examination

Baby looks jaundice, no dysmorphic feature no rashes
Vital Sign HR=130 RR=30/M SPO_2= 98% on room air
Weight 5th percentile height 75th percentile HC=38CM 50th percentile
Good breath sounds no distress, equal air entry bilateral 1st and 2nd heart sound were normal no murmur and no gallop.

She had hepatomegaly 3cm below costal margin and spleen was palpable positive bowel sound and passed colored stool. She is conscious, crying no sign of encephalopathy.

Differential Diagnosis

Etiology of Neonatal Cholestasis

Extrahepatic causes of cholestasis

- Biliary Atresia
- Infection -Cytomegalovirus, Retrovirus type 3
- BILUARY Cyst associated with autosomal recessive polycystic kidney disease
- Cystic fibrosis
- Endocrine causes

Intrahepatic Causes of Cholestasis

- Infectious like Torch infection a-cytomegalovirus, herpes simplex, syphilis
- Metabolic like galactosemia, tyrosinemia, alpha 1-antitrypcine, Fatty acid oxidation, Mitochondria disease
- Alagille syndrome
- Maternal causes like autoimmune disease
- Total parenteral nutrition, short bowel syndrome
- Idiopathic causes

Investigation

Laboratory

Complete blood count to roll out sepsis and ascending cholangitis
WBC Normal, Hgb normal
Peripheral blood smear RBC=Normocytic normochromic with prominent polychromatic cell and some NRBC
WBC=Mild left shift granulocytic cell with prominent toxic granulation no blast or abnormal cell
Platelets = Mild to moderate thrombocytosis
Liver function test = Albumin very low, GGT (gamma glutamyl transferase) very high LDH HIGH AST high ALT Moderate high uncogitated and conjugated both were very high, repeated conjugated very high.
Coagulation profile within normal range
Lactate Normal
Neonatal screening test Normal TANDAM
Blood group of baby B+ Mother O+ Direct combs test negative

Ferritin very high with our hospital unite repeated decrease
TORCH Screening Reactive to CMV IGG and Rubella IGG
Alpha-fetoprotein not available in our hospital sent to outside
Thyroid function profile Normal
Ophthalmology examined the eyes normal red reflex, no any congenital anomalies

Radiology

US abdomen normal study, normal gall bladder and liver no choledochal cyst. Doppler US Abdomen normal
ECHOGRAM DONE: Small PDA with VSD
Spinal x-ray normal vertebra to rollout Alagille syndrome
Skeletal Survey normal.

Conclusion

Neonatal cholestasis is serious disease you must be evaluate the child carefully physically and biochemically, physically to roll out any dysmorphic feature any family history of jaundice in previous child, any exchange transfusion was done in previous child.

Then examination of child to rollout any dysmorphic feature and to rollout Alagille syndrome, any rashes and hepatosplenomegaly to rollout Torch infection and if the baby looks well or unwell due to sepsis.

Growth Parameters and put on charts to rollout intrauterine growth restriction duo to any maternal and fetal congenital infection. Then examen the stool if stool was acholic pale stool no pigmentation you must consider alpha antitrypsin deficiency, choledochal cyst and biliary atresia.

Our case was interested admitted as case of jaundice due to ABO Incompatibility, Mother had blood group O+ and the baby was B+ and coombes test negative admitted in NICU, Received intensive phototherapy and exchange transfusion was done as protocol, Then we noticed direct bilirubin was very high and same time albumin was very low so we went to investigate more by asking the nurse to observe the color of stool which was normal pigmented stool, Ultrasound abdomen done to rollout contacted gallbladder and to roll choledochal cyst report was normal so we consult the metabolic to come and see the baby because he had hepatosplenomegaly and no dysmorphic feature order for echocardiogram and skeletal survey to roll out Alagille syndrome which was not fit with our case, echocardiogram was normal and skeletal survey was normal as well as the TANDAM was normal.

Gastroenterology advises to go more in the investigation and repeat ultrasound abdomen, observe stool carefully, sent alpha antitrypsin level, and repeated enzymes of liver, which was very high from beginning and put the baby on the lactose free diet and send referral to higher center for liver biopsy. Parent were counselling from the 1st day about the general condition of their son and he needs more investigation especially if the liver functions and enzymes were still high and they accepted baby was doing well on lactose free diet and the direct bilirubin and liver enzymes decreased dramatically and patient discharge on vitamin D3 and lactose free formula to follow up in OPD.

Reference

1. Hasan MS, Karim AB, Rukunuzzaman M, et al. Role of Liver Biopsy in the Diagnosis of Neonatal Cholestasis due to Biliary Atresia. Mymensingh Med J. 2018; 27: 826-833.