

Prolonged Neonatal Jaundice

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Afya ya watoto wetu!

Prolonged Jaundice?

- >6 months
- >3 months
- >2 weeks
- >4 weeks

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Case Presentation

- 6 wk old born in UK via c/section to a primi-gravida
- Jaundice from week 2 of life; reassured by midwife
- Returned to Kenya and brought for well baby visit/vaccination at 6 weeks
- Noted to have jaundice; otherwise well
- What's your next step?



Case Presentation (2)

- Stool colour: pale
- Differential diagnosis?
- Investigations?
- Treatment: ?expose to sunlight
- Final diagnosis: Biliary Atresia

Prolonged Neonatal Jaundice

- Jaundice persisting for >2 weeks in a term infant, and >3 weeks in a preterm infant
- Needs urgent evaluation
- About 2.4-9% of exclusively breast-fed babies may have jaundice beyond 2 weeks of age (unconjugated!)

Winfield CR. Clinical study of jaundice in breast and bottle fed babies. *Arch Dis Child* 1978

Neonatal Cholestasis

- Defined as ***conjugated*** hyperbilirubinaemia developing within 60 days of birth
- Conjugated bilirubin >20% of total bilirubin
- Cholestatic jaundice affects 1 in 2500 infants
- Guidelines recommend evaluation of every neonate with jaundice > 2 weeks of age

Naspghan Guidelines JPGHN 2004

Extra-hepatic Causes

- *Extra-hepatic biliary atresia*
- *Choledochal cyst*
- Inspissated bile/mucus plug
- Cholelithiasis
- Cystic fibrosis
- Congenital hepatic fibrosis

Intra-hepatic Causes

- Infectious
 - *Idiopathic neonatal hepatitis syndrome*
 - TORCHES
 - Hepatitis B,C
 - Sepsis
 - *E.coli UTI*
- Endocrine
 - *hypothyroidism*
- Metabolic
 - *Galactosaemia*
 - Tyrosinaemia
 - Alpha1antitrypsin def.
 - Haemachromatosis
- Congenital
 - Alagille's syndrome
- Toxic
 - TPN associated
- Idiopathic

Initial Evaluation of Jaundiced Infant

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Is the infant **ill looking**?

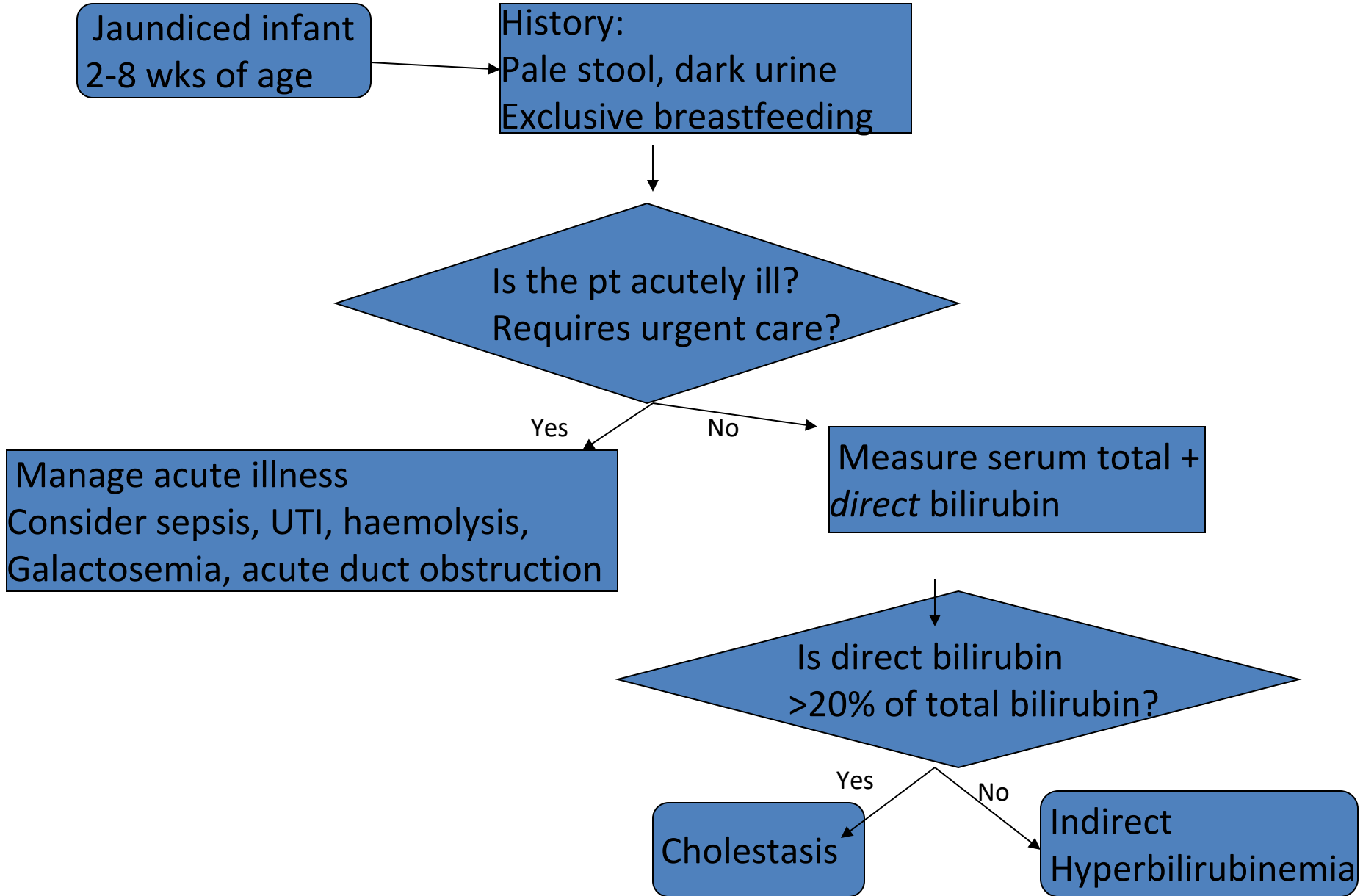
- Sepsis
- Galactosaemia
- Haemolysis



ADMIT

If infant **stable**:

- Bilirubin: total/direct
 - If cholestatic, refer!
- LFT's, PTI, Albumin, RBS
- FBC, Urine mcs, TSH
- *Abdominal US*
- Liver biopsy
- Others



CHOLESTASIS

Consider biliary atresia, choledochal cyst, galactosaemia, hypothyroidism, UTI

Investigations:
FBC, LFT, PTI, Albumin, Glucose
Urine reducing substances + m/c/s
Thyroid profile, ferritin levels
Abdominal Ultrasound, TORCH
Liver biopsy, Others

Consult appropriately:
Paediatric GI
Paediatric Surgeon
Paediatric Metabolic Expert

INDIRECT HYPERBILIRUBIN

Consider breast milk jaundice, UTI, ABO incompatibility, hypothyroidism

Investigations:
FBC, LFT, PTI, Albumin, Glucose
PBF + Blood Group + Coomb's test
Septic screen, Urine m/c/s
Thyroid profile, Others

Consult appropriately

Goals of Timely Evaluation

- Diagnose and treat known medical and/or life-threatening conditions
 - *Hypothyroidism*
 - Galactosaemia
 - *UTI*
 - Sepsis
- Identify disorders amenable to surgical therapy within an appropriate time-frame
 - Choledochal cyst
 - *Biliary atresia*

Biliary Atresia

- Progressive inflammation of bile ducts, causing biliary tract fibrosis, obstruction and eventually liver cirrhosis
- Incidence: 1 in 8,000; most common cause of chronic cholestasis in infants
- Aetiology: unknown
- Well looking, jaundiced infant in the first 3 months
- Early diagnosis and treatment before 60 days improves outcome

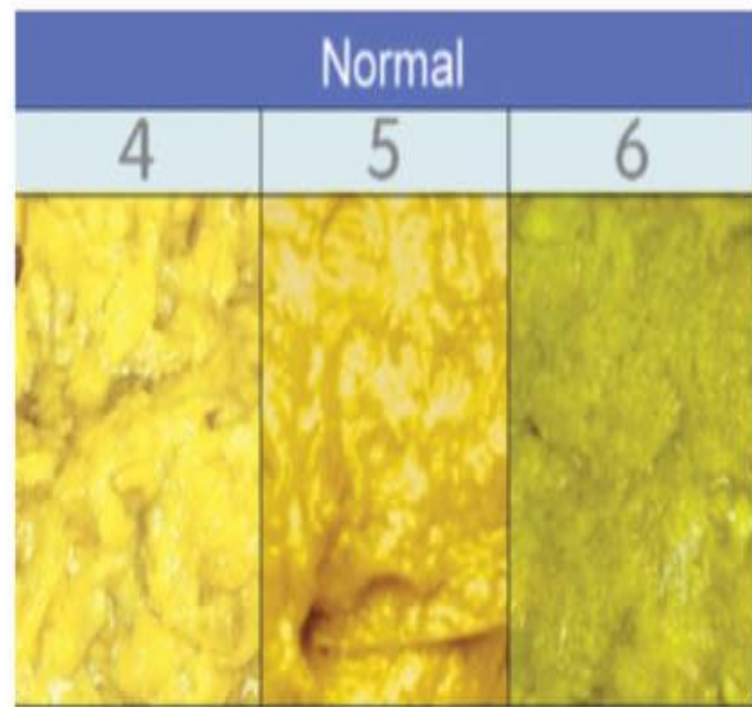
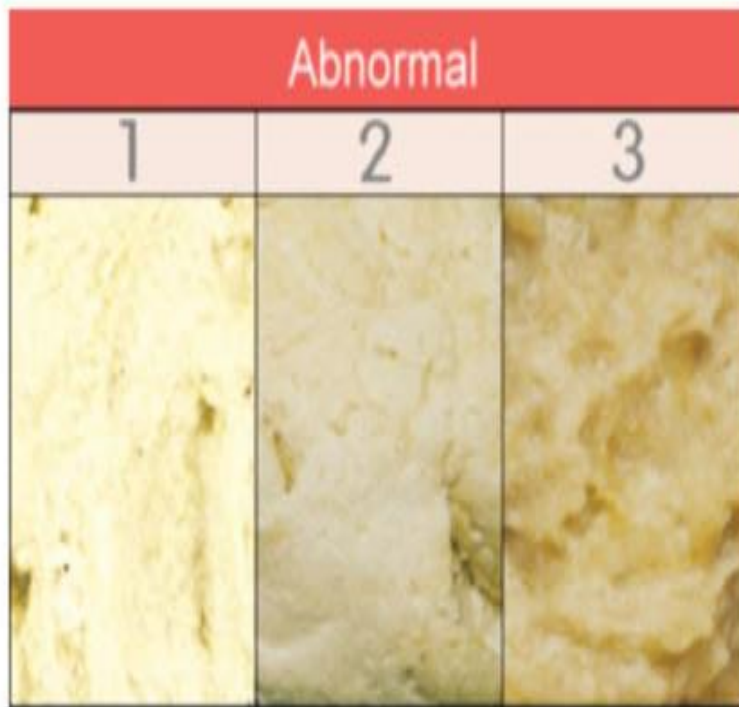
Balistreri WF. Liver disease in children 2007

Stool colour

- Parents not reliable observers of stool colour
- Clinicians better at detecting acholic stool
 - Look at the POO!
- Stool colour card improves early case finding

Crofts DJ. Assessment of stool colour in community management of prolonged jaundice in infancy. *Acta Paediatr* 1999;88

Infant Stool Color Card



Abdominal Ultrasound

- Fasting (3-4 hours)
- Biliary atresia likely if *gall bladder* contracted or not visualized (73-100% sensitivity, 67-100% specificity)
- “Triangular cord” sign: fibrous cone of tissue at bifurcation of portal vein (83-100% sensitivity, 98-100% specificity to detect biliary atresia)
- Detects choledochal cysts

Naspghan Guidelines JPGHN 2004

BA: Kasai Procedure

- Roux-en-Y portoenterostomy to re-establish bile flow
- Bile flow re-established in 80-85% if performed prior to 8 weeks-old
- Bile flow re-established in less than 20% if performed after 12 weeks-old
- Liver transplant for late diagnosis and “failed” Kasai

Chardot C. J Pediatr 2001, McClement JW. Br Med J. 1985

Summary

- Cholestatic jaundice should be excluded in all infants with prolonged jaundice >14 days
- Infantile cholestasis needs early recognition and diagnostic evaluation:
 - Early treatment of medical conditions
 - short window period for surgical treatment
- No role of “sunlight phototherapy” beyond 2 weeks of age

Thank You!

Infant Stool Color Card

