Prolonged Neonatal Jaundice

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Prolonged Jaundice?

- >6 months
- >3 months
- >2 weeks
- >4 weeks
Prolonged Jaundice?

- >6 months
- >3 months
- >2 weeks
- >4 weeks
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
Initial Evaluation of Jaundiced Infant

Is the infant **ill looking?**
- Sepsis
- Galactosaemia
- Haemolysis

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

ADMIT
Jaundiced infant 2-8 wks of age

History:
Pale stool, dark urine
Exclusive breastfeeding

Is the pt acutely ill?
Requires urgent care?

Manage acute illness
Consider sepsis, UTI, haemolysis, Galactosemia, acute duct obstruction

Yes

No

Measure serum total + direct bilirubin

Is direct bilirubin >20% of total bilirubin?

Yes

Cholestasis

No

Indirect Hyperbilirubinemia
**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*
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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

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

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[Image of stool samples]