Case: Portal Hypertension in Pregnancy

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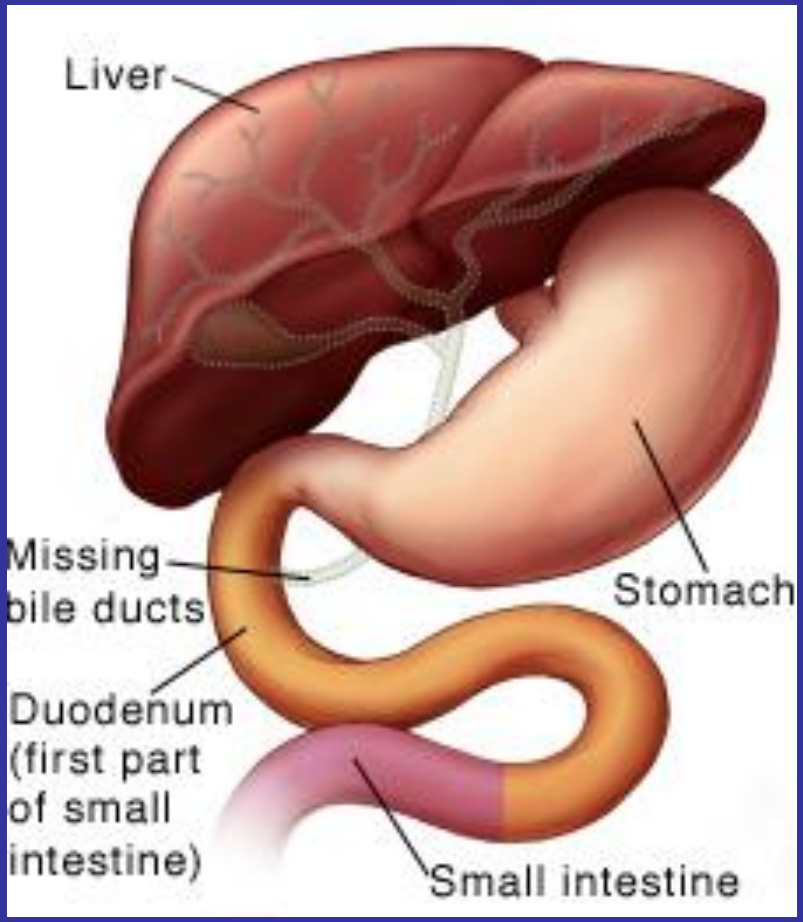
Case Presentation

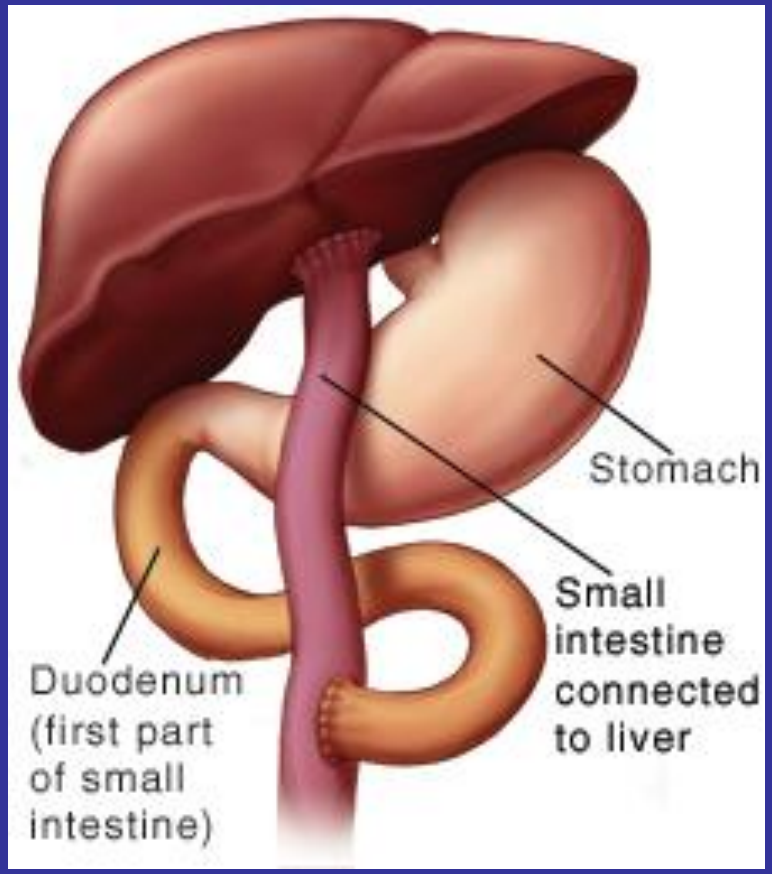
- 21 year old woman G1P0 at 14 weeks on liver transplant list with unplanned pregnancy
- Congenital biliary atresia s/p Kasai procedure at 1 month of age
- Portal hypertension
  - Splenomegaly
  - Thrombocytopenia
  - Esophageal varices

Past Medical History

- Never had decompensated liver disease
  - (ascites, GI bleed, hepatic encephalopathy)
- No functional disability
  - avoids contact sports because of splenomegaly
- Meds
  - Propranolol 80mg/40mg
  - vitamins A, D, E
  - Calcium
- No other medical issues
- MELD score – 10 prior to pregnancy

Kasai Procedure for Biliary Atresia





http://www.fairview.org/healthlibrary/Article/88701

Exam

- Exam: HR 63; BP 114/63
- Splenomegaly, scar on abdomen.
- No stigmata of chronic liver disease:
  - No asterixis
- Counseled extensively about pregnancy and decided to continue
- Gastroscopy at 14 weeks:
  - Small grade 1 esophageal varices
  - No high risk stigmata

Labs

Test	16 Wks	26 Wks	33 Wks
Hgb **g/L** ***(g/dL)***	**110** ***(11.0)***	**115** ***(11.5)***	**117** ***(11.7)***
Platelets **x 10^9/L**	**27**	**22**	**21**
Creatinine **umol/L** (***mg/dL)***	**41** ***(0.46)***	**46** ***(0.52)***	**43** ***(0.49)***
ALT **U/L**	**13**	**10**	**14**
Bilirubin **umol/L** ***(mg/dL)***	**28** ***(1.64)***	**37*(2.16)***	**38** ***(2.22)***
PT INR **s**	**1.4**	**1.1**	**1.1**
Bile acids **umol/L** ***(mg/dL)***	**24** ***(9.8)***	**46** ***(18.8)***	**16** ***(6.5)***
Vitamin A **(1.5-3.5 umol/L)**	**0.8**	**0.9**	**0.8**
Vitamin E **(12-45 umol/L)**	**21**	**26**	**22**
Vitamin D **(80-200 nmol/L)**	**95**		**105**
Albumin **g/L** ***(g/dL)***	**35** ***(3.5)***	**34** ***(3.4)***	**30** ***(3.1)***
MELD score	**12**	**10**	**11**

MELD score predicts outcome in cirrhotic patients during pregnancy

- Retrospective review of single center in UK – all cirrhotic patients with pregnancy 1984-2009
- 62 pregnancies in 29 women with cirrhosis
- 34% pregnancies unplanned
- Median MELD at conception was 7
- Fetal outcomes
  - Live birth rate 58%; miscarriage 19%; stillbirth 6%
  - Elective termination 15% (half medically advised)
  - Preterm delivery: 64% <37 weeks; 18% <30 weeks
- Higher MELD score was associated with risk for preterm delivery (p=0.01)

Westbrook et al. Clin Gastro Hepatol. 2011;9:694-699

MELD score predicts outcome in cirrhotic patients during pregnancy

- Maternal complications:
  - Maternal morbidity 10%
    - Variceal bleeding (3), Decompensation with ascites (2), encephalopathy(1)
  - 1 woman died; 2 women died 6 and 16 months post delivery
  - Variceal bleeding occurred in 2nd or 3rd trimester (esophageal and splenic)
  - Women with varices more likely to deliver by C-section
- MELD score $\geq$ 10 prior to conception had 83% sensitivity and specificity for predicting liver-related complication ante- or postpartum
- MELD score $\leq$ 6 had low risk of maternal complications

Westbrook et al. Clin Gastro Hepatol. 2011;9:694-699

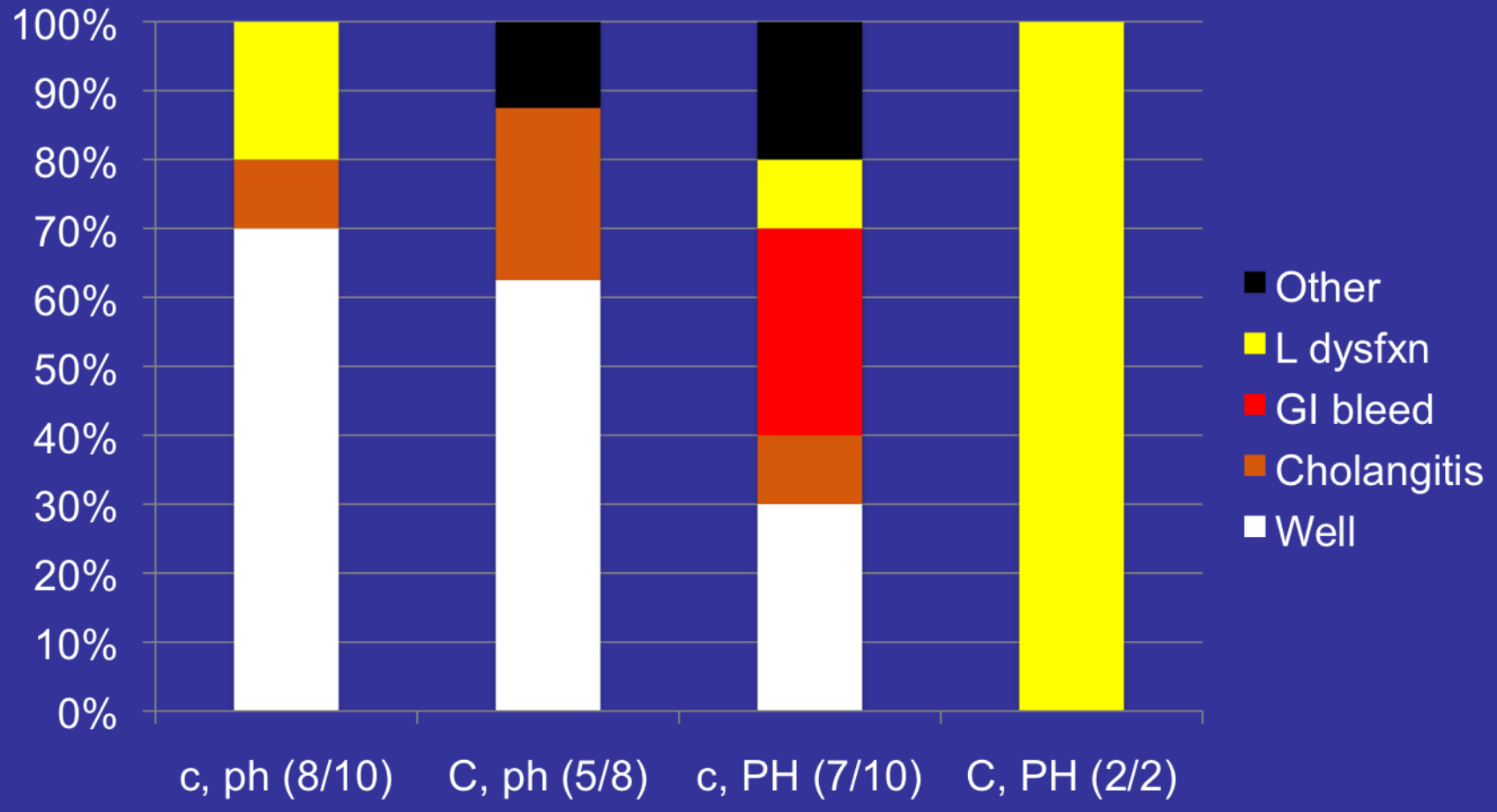
Outcomes Of Pregnancy In Patients With Cirrhosis

- Population based cohort of cirrhotic women with obstetric hospitalizations to US hospitals 1993-2005
- Using this sample, ~ 1100 deliveries in US of women with cirrhosis

Complication	Rate	OR (95% CI)
Cesarean delivery	42%	1.41(1.06-1.88)
Preterm delivery	38.7%	5.51(4.16-7.3)
Placental abruption	7.1%	4.41 (2.55-7.6)
Maternal death	1.8%	None in controls
PPH	13.3%	5.0(3.31-7.55)
Hypertension	14.5%	1.63 (1.19-2.23)
Variceal bleeding**	5.3%	** during all
Ascites**	10.9%	admissions
Hepatic decompensation**	15%	

Shaheen et al. Liver Int. 2009

Pregnancy in women with biliary atresia



Sasaki et al. J Ped Surg 2007

Management

- Continue propranolol
- Monitor and supply fat soluble vitamins
- Monthly labwork
- Fetal echocardiogram
  - (association between some types of biliary atresia and cardiac defects)
- Start Ursodeoxycholic acid for increased bile acids
- Surveillance for varices

28 Weeks:

- Gastroscopy
  - no significant varices; remains on propranolol
- No plan for further endoscopy unless bleeding as has reached maximal blood volume
- MRI Abdomen / Pelvis
  - Assess for splenic artery aneurysm, abdominal wall varices and portal varices as could present with complications, particularly if operative delivery required
- Multidisciplinary team meeting to discuss planning for delivery

Multidisciplinary meeting at 29+ weeks

- Vaginal delivery unless obstetric indication for C-section
- No further endoscopy planned
- Await MRI
- Continue propranolol
- Steroids for fetal lung maturity now
- Discussed anaesthesia issues and management of bleeding

MRI Result

- MRI report:
  - Cirrhotic liver with evidence of previous Kasai procedure
  - Evidence of portal hypertension with massive gastroesophageal and splenic variceal formation
  - No large abdominal wall collaterals are seen
  - Small / moderate intra abdominal ascites
  - Massive splenomegaly

Quandry!

1. Should she have repeat endoscopy in light of MRI findings?
2. What is correlation between MRI and endoscopy in detecting varices at risk for bleeding?
3. Should mode of delivery be changed to Cesarean section?
    - What type of anaesthesia?

MRA vs endoscopy for the assessment of GE varices in biliary atresia

- Pediatric population
- 34 children with biliary atresia had MRA and endoscopy post-operatively
- Sensitivity of MRA 75% (picked up 12/16)
- Specificity 88.9%
- 2 children had varices on MRA that couldn't be confirmed on endoscopy
- When broken down by age and liver function, MRA inaccurate in children <10years age and in those with moderate liver dysfunction

Takahashi et al. Pediatr Surg Int. 2007

Delivery plan: Round 2

- Summary:
  - Congenital biliary atresia, post-Kasai
  - Cirrhosis with portal hypertension and severe thrombocytopenia
  - No episodes of decompensation
  - Reasonable synthetic function
  - Poor absorption of fat soluble vitamins
  - Elevated bile acids on UDCA

MDM Attendees

- ObMed
- MFM, Ob
- Anesthesia
- GI, hepatology
- NICU
- Hematology, Blood bank physician (by phone before to get input)

Delivery plan: Round 2

- Elective C-section under GA
- Notify: ObMed, Blood bank, GI, ICU when admitted
- SBP prophylaxis for at least 5 days postpartum
- Decision made to not repeat endoscopy

Delivery plan: Round 2: Plan for Bleeding

- Blood bank notified to have several platelet pools available
- Platelets to be given prior to C-section and if any concern about bleeding
- Vitamin K IV pre-op for 2 days
- Wide surgical exposure with foceps for C-section to avoid pressure on spleen
- Tranexamic acid if concern about excess bleeding
- Cryoprecipitate early if bleeding
- Can use oxytocin, hemabate, cytotec; avoid ergots

Delivery plan: Round 2
What about variceal bleed?

- Continue propranolol
- Have octreotide available on ward
- Minimize fluid administration
- Notify GI when admitted and if any signs bleeding
- Notify GI/hepatology if any signs liver decompensation
- Have large bore IV's available
- Admit to observation bed post-partum
- Pneumatic compression stockings postpartum

Delivery plan: Round 2
Neonatal Care

- Routine vitamin K without delay
- NICU/pediatric dietician to assess need for vitamin A supplementation
- Vitamin E and D levels to be checked

Outcome

- C-section at 35+5 days
- 2450g male infant
- Baby did not require NICU
- Oxytocin, cytotec used
- Received 2 pools of platelets and 8 units cryo
- Discharged postpartum day 4 without complications