Case Based Severe GU Anomalies
Judy Estroff, MD

1. First line treatment for neonates in renal failure starting renal replacement therapy (RRT) in the newborn period:
   A. Peritoneal dialysis
   B. Hemodialysis
   C. Renal transplant
   D. Oral diuretics and fluid restriction

Correct Answer: A

Rationale:
92% of newborns in renal failure are first treated with peritoneal dialysis. Only 8% begin with hemodialysis in less than 1% have a renal transplant in the first few months of life.

Reference:

2. The most common cause for renal failure in newborns is:
   A. Cystic kidney disease, i.e., ARPCKD
   B. Obstruction, i.e posterior urethral valves or UPJO
   C. Bilateral renal agenesis
   D. Renal vein thrombosis

Correct Answer: B

Rationale:
About 55% of newborn renal failure is due to obstruction secondary to posterior urethral valves or UPJO. Only 13% of newborns are in renal failure because of genetic cystic kidney disease. Bilateral renal agenesis is rare, as is renal vein thrombosis.

Reference:
   i. Survival and clinical outcome of children starting renal replacement therapy in the neonatal period.
Fetal Intervention for GU Anomalies
Holly A. Hedrick, MD

1. Which of the following is a candidate for vesico-amniotic shunting?
   A. 17 week gestation with PUV, cystic renal disease, anhydramnios, Urine sodium > 100mmol/L and beta - 2 microglobulin > 6 mg/L
   B. 37 week gestation with PUV, cystic renal disease and AFI of 2
   C. 32 week gestation with PUV, no renal cysts and AFI of 12
   D. 20 week gestation with trisomy 18, PUV, AFI of 4, and a favorable urine sample including urine sodium < 100 mmol/L, chloride < 90 mmol/L, osmolarity < 210 mOsm/L, and a beta-2 microglobulin < 6 mg/L.
   E. None of the above

Correct Answer: E

Rationale:
The management of PUV has evolved to include prenatal surgical intervention but remains investigational, with most affected fetuses treated soon after birth. Data on outcome of fetal surgery for PUV suggest that it is associated with a risk of fetal and maternal morbidity without proven benefit for long-term renal outcome.

Fetal surgery for PUV should be considered only for fetuses who have a high risk of in utero / neonatal death due to midtrimester severe oligohydramnios, with normal karyotype and evidence of good renal function based upon fetal urinary evaluation.

Fetal urine from the first drainage is discarded and a urine sample obtained after bladder refilling is sent for analysis. A favorable urine sample indicating good renal function has urine sodium of less than 100 mmol/L, chloride less than 90 mmol/L, osmolarity less than 210 mOsm/L, and a beta-2 microglobulin less than 6 mg/L.

Option A is incorrect as urine electrolytes are abnormal.

Option B is incorrect as fetus close to term and should be delivered as soon as possible.

Option C is incorrect as the fetus is stable with normal AFI.

Option D is incorrect as the karyotype is abnormal.

References:
I. Up to Date Nicholas Holmes http://www.uptodate.com/contents/management-of-posterior-urethral-valves
Advanced Fetal GI Evaluation: When Is MR Useful?
Eva I. Rubio, MD

1. By what GA should meconium be seen in the distal rectum?
   A. 12 weeks
   B. 16 weeks
   C. 20 weeks
   D. 30 weeks

Correct Answer: C

Rationale:
The first trimester is too early to expect to see meconium in the intestines.

At 16 weeks, although some early meconium may be seen in the small bowel, it is too early to expect to see meconium in the rectum in all cases.

By 30 weeks, if meconium is not seen in the distal colon and rectum, we expect an abnormality, usually an obstruction.

Reference:

2. A fetus presents with findings of a distal bowel obstruction. Which of these findings are expected to be seen?
   A. Oligohydramnios
   B. Normal amniotic fluid
   C. Polyhydramnios
   D. Ascites

Correct Answer: B

Rationale:
In a distal bowel obstruction, such as the distal ileum or colon, swallowed amniotic fluid is usually resorbed in the pre-obstructed intestinal segments through the wall, whereas in a proximal obstruction swallowed amniotic fluid accumulates in the proximal dilated bowel loops or remains in the amniotic fluid around the fetus resulting in polyhydramnios. Typically bowel obstruction does not result in oligohydramnios, unless there are additional urinary tract or renal abnormalities. We do not expect to see ascites unless there is perforation, which may occur at different levels of obstruction.

Reference:
Case Based Ventral Wall Defects
Teresa Victoria, MD

1. A fetus presents with an infraumbilical abdominal wall defect. The defect is compatible with a:
   A. Omphalocele
   B. Gastrochisis
   C. Limb body wall defect
   D. Exstrophy

Correct Answer: D

Rationale:
In omphalocele the cord inserts at apex/base of defect, in gastrochisis and LBWX the defect is paramedian.

Reference:

2. Prenatal examination of a fetus fails to demonstrate a normal bladder in the presence of normal amniotic fluid and lung volumes. One may find all of the below, except:
   A. infraumbilical soft tissue “mass”
   B. possible hindgut abnormalities
   C. abnormal genitalia
   D. Cord insertion below the abdominal wall defect

Correct Answer: D

Rationale:
The findings suggest a diagnosis of exstrophy, be it cloaca or bladder exstrophy. The soft tissue “mass” represents the everted bladder, hindgut abnormalities accompany a dx of cloacal exstrophy (not bladder exstrophy), and abnormal genitalia may be found in both cloaca and bladder exstrophy. The wall defect is infraumbilical.

References:

3. The main clinical concern when delivering a child with an umbilical cord defect is:
   A. Inadvertently excising the bowel while cutting the cord at birth
   B. Bowel ischemia
   C. Rupture of the abdominal wall defect
   D. Cord compromise

Correct Answer: A
**Rationale:**
The main concern if the obstetrician or surgeon is not aware of this complication is that the bowel may be inadvertently cut while freeing the umbilical cord.

**Reference:**

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**Management of Lung Hypoplasia- Giant Omphalocele**
*Holly A. Hedrick, MD*

1. Which of the following is a risk of primary vs staged repair of a giant omphalocele
   A. Delayed enteral feeding
   B. Higher infection rate
   C. Abdominal compartment syndrome
   D. Longer NICU course

**Correct Answer: C**

**Rationale:**
Primary repair includes reposition of the herniated viscera and repair of the abdominal layers including the fascia. Advantages are a low infection rate and good chances of early enteral feeding-A major disadvantage is the high risk of ACS. The postoperative monitoring should include regular measurements of intra-abdominal pressure. Intravesical pressure can be used as a substitute for direct measurement Clinical signs of ACS, such as increasing respiratory insufficiency and renal failure, should be searched for.

Option A is incorrect. Successful primary repair allows for faster enteral feeding initiation.

Option B is incorrect. There is a lower infection rate with primary repair.

Option D is incorrect. Staged repair typically requires a prolonged course in the NICU.

**References:**


2. What criteria has been used to declare an omphalocele “giant” and lungs hypoplastic?
   A. Abdominal wall defect 2 cm in width
   B. Sac containing 25% of the liver
   C. O/E Total Lung volume < 25%
   D. Omphalocele / abdominal circumference ratio of >.26
Correct Answer: D

Rationale:
There have been attempts in predicting feasibility of primary closure based on ratios of omphalocele diameter to biometric measures. Even though ratios between volume of herniated organs and size of abdominal cavity might be helpful, other authors suggest that only an attempt of primary closure will show if this is possible or not. In a retrospective analysis, Fawley et al calculated omphalocele ratio defined as omphalocele /abdominal circumference in 30 neonates with omphalocele and available prenatal ultrasound scans. In 60% (12/20) of the patients with a ratio <0.26, primary surgical closure was achieved, compared to none in the group with a ratio >0.26.

Option A is incorrect. Size of defect may not help assess whether primary or staged repair is possible. Measures of 5-6 cm have been used but a small defect may have a large amount of liver and intestine herniated and may not be a candidate for primary reposition.

Option B is incorrect. All or majority of liver have been used as criterial for GO

Option C is incorrect. O/E TFLV < 50% has been suggested as the cut off to be used for giant omphalocele lung hypoplasia

References: