

American Pediatric Surgical Association

## Prenatal Counseling Series

### Bladder Outlet Obstruction



**APSA**  
American Pediatric  
Surgical Association  
*Saving Lifetimes™*

from the  
**Fetal Diagnosis and Treatment Committee**

of the  
American Pediatric Surgical Association

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## Prenatal Counseling Series Bladder Outlet Obstruction

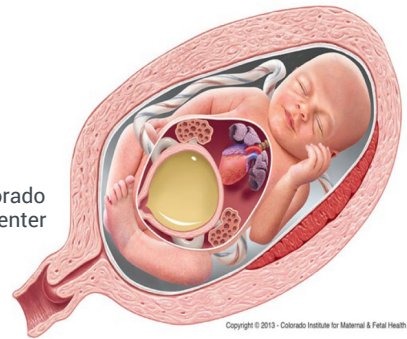
### Fetal Urologic Malformation

- ▶ Incidence: Approximately 1 in 500 pregnancies is complicated by significant fetal urologic malformations.
- ▶ Obstruction Disorders: any point along the GU tract
  - Upper tract obstruction
    - Prolapsing ureterocele
    - Ureteropelvic junction (UPJ), duplication abnormality, ureterovesical junction (UVJ)
  - Lower tract obstruction (Lower Urinary Tract Obstruction – LUTO)
    - Bladder outlet obstruction
      - Posterior urethral valves (PUV)
      - Urethral atresia
      - Meatal stenosis
    - Cloacal abnormalities

### Bladder Outlet Obstruction

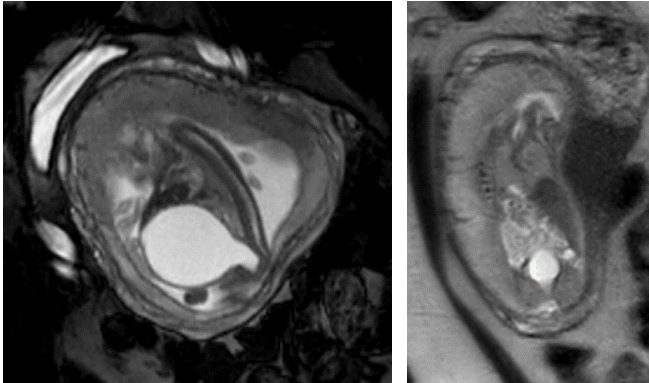
- ▶ Incidence: Estimated to be 1:8000 live births
- ▶ Etiology:
  - Male = PUV Prune belly syndrome with urethral atresia
  - Female = urethral atresia (uniformly lethal), persistent cloaca, caudal regression
  - Megacystis Microcolon Intestinal Hypoperistalsis (MMIH) syndrome
  - Rare causes = prolapsing ureterocele
- ▶ Diagnosis: Prenatal bladder outlet obstruction (BOO) leads to significant and persistent distension of the bladder
- ▶ Natural History: Highly variable: depends on gender, severity, duration and GA of onset of the obstruction.
  - High grade obstruction:
    - Severe oligohydramnios or anhydramnios
    - Massive bladder (megacystis)
    - Unilateral and/or bilateral and is not a good predictor of renal function
    - Hydroureter and hydronephrosis
    - Renal cystic dysplasia

Source: Colorado Fetal Care Center



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- Pulmonary hypoplasia & respiratory insufficiency leading to neonatal death



Sagittal and coronal MR images of a fetus with oligohydramnios demonstrate dilation of the urinary bladder, posterior urethra and ureters. The fetal thorax is subjectively small and the lung parenchyma shows diffusely decreased signal intensity as can be seen in pulmonary hypoplasia. Fetal lung volumes measure low for gestational age by MR. Images courtesy of Jill Stein, MD.

- Pulmonary dysfunction more critical than renal dysfunction for initial neonatal survival
- Incomplete obstruction:
  - Variable amniotic fluid volumes
  - Variable pulmonary hypoplasia
  - Compatible with neonatal survival
  - Variable degrees of renal dysplasia
  - 30% lifetime risk of ESRD
  - Poor in utero bladder cycling

### Initial Evaluation

- ▶ Obstetrical ultrasound with a possible diagnostic amnioinfusion
- ▶ Fetal echo
- ▶ Fetal karyotype (10-25% aneuploidy): chorionic villous sampling (CVS), amniocentesis or vesicocentesis
- ▶ Fetal Magnetic Resonance Imaging (MRI): Evaluate lung volumes – please refer to the CDH brochure
- ▶ Renal function - possible bladder tap

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### Obstetrical Ultrasound

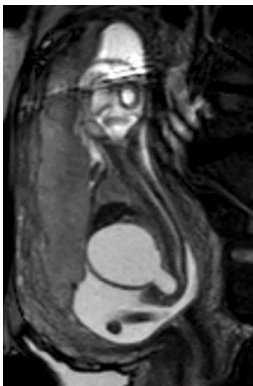
Low and high grade obstruction:

Presence of some or normal amniotic fluid (AF)

(AF) - low grade. Absence of AF - high grade

If complete, a diagnostic amnioinfusion of warmed normal saline may be necessary for accurate imaging

- ▶ Detailed survey to assess for other anomalies
- ▶ Evaluate urinary tract from kidneys to urethra to find obstruction
  - Presence of a ureterocele
  - Evidence of reflux
- ▶ Echogenicity and size of kidneys
- ▶ Cysts versus dilated calyces
- ▶ Bladder characteristics
  - Keyhole sign

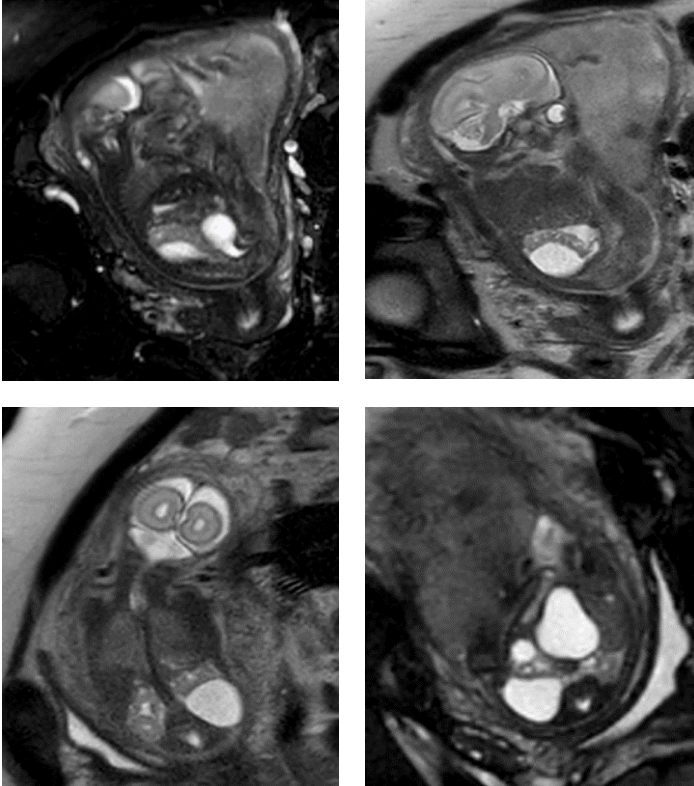


Sagittal and coronal MR images show dilation of the fetal urinary bladder and posterior urethra resulting in a characteristic 'keyhole' morphology. The urinary bladder wall is diffusely thickened. The ureters are dilated and tortuous, and there is mild bilateral renal pelvis and calyceal dilation. The renal parenchyma appears normal.

- Bladder wall hypertrophy
- ▶ Ascites

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► Peri-nephric urinoma



Sagittal, coronal and axial MR images of a fetus with anydramnios and small fetal thorax. There is relatively mild dilation of the urinary bladder and posterior urethra with a perinephric cystic collection indicative of urinoma. Images courtesy of Jill Stein, MD.

### **Serial Bladder taps (vesicocentesis)**

Indicated in patients with complete or near complete obstruction and questionable renal function

Ultrasound guided aspiration of fetal urine

Complete bladder drainage at 24-48-hour intervals

- Measure electrolytes and  $\beta$ 2-microglobulin
- Ensure bladder refilling

Initial bladder drainage evaluates urine present for undetermined period and therefore is not useful

Second drainage likely obtains fluid from upper urinary tract

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Third drainage procedure collects recently created urine reflective of renal function

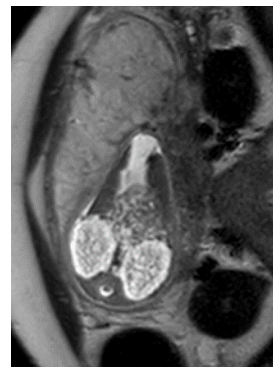
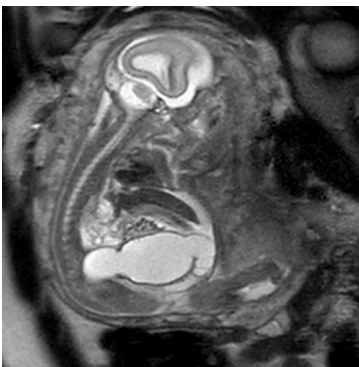
- ▶ Prognostication of current or later renal function
  - identify favorable prognosis
    - select good candidates for fetal intervention
  - Identify poor prognosis
    - option of interruption
    - continued pregnancy with neonatal palliative care

If renal function is present vesicoamniotic shunting should be considered

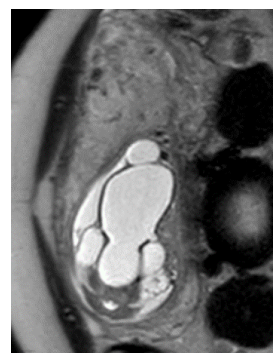
### Fetal Therapy

Not all fetuses with BOO meet criteria for intervention

- ▶ The fetus might be too healthy (where the risk of intervention outweighs any possible benefit)
  - Normal AFI
  - Non-obstructive dilation
  - Unilateral involvement
- ▶ The fetus might be too sick (where intervention has minimal possibility of benefit and might harm the mother)
  - Renal cystic dysplasia



Sagittal, coronal and axial MR images of a fetus with anydramnios and small fetal thorax. There is marked diffuse dilation of the urinary collecting system from the calyces through the posterior urethra; a dilated anterior urinary bladder diverticulum represents a urachal remnant. Numerous tiny cysts throughout bilateral kidneys are concerning for dysplasia. Images courtesy of Jill Stein, MD



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- Abnormal urinary parameters
- Abnormal karyotype or multiple associated anomalies

### Vesicoamniotic Shunting

Allows drainage of fetal urine into amniotic space

Promotes lung growth by allowing re-accumulation of amniotic fluid

Not curative but has the potential to lead to a pulmonary survival

Usually requires amnioinfusion

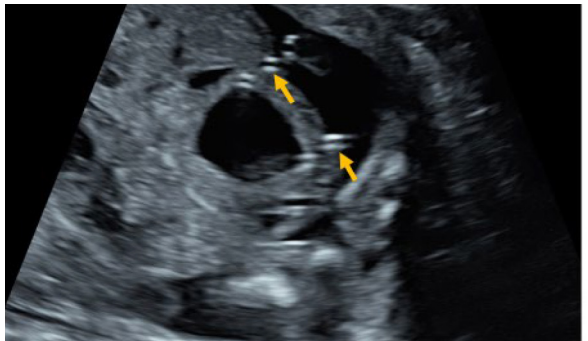
Double pigtail shunt (Harrison shunt) or rocket shunt is placed. One limb in fetal bladder and one limb in amniotic fluid.

It is possible that the baby will pull out the shunt

The average number of shunts/pregnancy is about 2.5.



Pre shunt



Post shunt

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- ▶ Low grade BOO with normal AF volume: good prognosis with likely functional kidneys and lungs
- ▶ Low grade BOO with low AF volume: indeterminate prognosis; may benefit from vesico-amniotic shunting
- ▶ High grade BOO with favorable urine markers and bladder refilling: poor prognosis but may benefit from vesico-amniotic shunting
- ▶ High grade BOO with no bladder refilling: extremely poor prognosis and unlikely to benefit from vesico-amniotic shunting, may be candidates for Renal Anhydramnios Fetal Therapy (RAFT) trial

### Postnatal Considerations

Respiratory function is very hard to predict but does have significant correlation with degree and duration of low or no AF; outcome varies from no respiratory support to non-survival.

Renal replacement therapy with dialysis may be required.

Patients on dialysis are very prone to infections and require an enormous amount of medical care.

Neonates, infants and children with renal failure need extra nutrition and may require a feeding tube.

The best form of renal replacement therapy is a kidney transplant.

Generally babies need to be about 10 kg/ 2 years old to be a candidate for transplant so infants with poor renal function may need PD as a bridge to transplant.

These babies may need prolonged hospitalization after birth to address determination of renal function, potential surgical intervention to treat the obstruction, as well as feeding/dialysis access issues.