Updated management of congenital hydronephrosis

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**Definition of Hydronephrosis:**

The most generally accepted definition is the maximum antero-posterior diameter of renal pelvis. No universally accepted value defining this abnormality till now.

**Timing of detection:**

90% after eighteen weeks of gestation.

95% by 22 weeks.
**Grading:**

The Society of Fetal Urology (SFU) has developed the following classification system:

- **SFU Grade 0**  Intact central renal complex (renal pelvis).
- **SFU Grade I**  Mild splitting of central renal complex.
- **SFU Grade II**  Pelviectasis but no calycectasis*.
- **SFU Grade III**  A markedly split pelvis with uniformly dilated calyces, but normal renal parenchyma.
- **SFU Grade IV**  Characteristics of grade III with thinning of renal parenchyma.

*Pelviectasis : Dilatation of pelvis  Calycectasis: Dilatation of calyx
Another system of grading looked at the maximum length of renal pelvis diameter (RPD). (2004)

- **Mild hydronephrosis** 5-8 mm
- **Moderate hydronephrosis** >8-12 mm
- **Severe hydronephrosis** >12 mm

Marked hydronephrosis is frequently seen in pelvic ureteric junction obstruction whereas the mild hydronephrosis is associated with vesicoureteric reflux.
In 2005 AAP updated the system of grading hydronephrosis according to renal pelvis diameter (RPD).

- **Mild hydronephrosis** 4-10 mm
- **Moderate hydronephrosis** >10-15 mm
- **Severe hydronephrosis** >15 mm

Now this is the most widely used grading system.
Still there is a lack of consensus on the threshold RPD that defines antenatal hydronephrosis.

Although most cases of mild renal pelvic dilatation, also referred to as “pyelectasia”, will resolve and not have a clinical impact on neonatal renal development, still there are reports of persistent cases that require postnatal intervention.

Fetuses with RPD >15 mm during the third trimester are at the greatest risk for CAKUT.
**Frequency:**

The reported incidence of antenatal hydronephrosis ranges from 0.6 to 4.5 % of pregnancies.

Increased frequency of up to 8% with positive family history of renal agenesis, multicystic kidney, reflux nephropathy and polycystic kidneys.

**Epidemiology:**

Male to female ratio is 2:1.

Bilateral in 20 to 40 %.

Differences in reported data may be due to different criteria used to define the disorder and the level of attention to the urinary system by the ultrasonographer.
Common causes of antenatal hydronephrosis:

- Transient 48%
- Physiologic 15%
- Pelvic ureteric junction (PUJ) obstruction 11%
- Vesicoureteric reflux (VUR) 4%
- Megaureter, obstructed or non-obstructed 4%
- Multicystic kidneys 2%
- Ureteroceles 2%
- Posterior urethral valves (PUV) 1%
Please remember:

Antenatal hydronephrosis without associated urinary tract anomaly has been termed isolated antenatal hydronephrosis (IAHN).

IAHN is believed to be caused by a physiologic dilatation of the developing ureter.

Also, could be due to the fact that the ureter begins normal development as a solid cord of tissue that canalizes to allow unobstructed passage of urine.

Metanephric urine production begins at approximately 8 weeks' gestation, potentially before completion of ureteral canalization. This results in transient obstruction with hydronephrosis. Once canalization is complete, this obstruction is relieved, and hydronephrosis should resolve.
Transient and physiologic hydronephrosis (IAHN) contribute to 60% of antenatal hydronephrosis.

In 97% of cases RPD do not exceed 10 mm.

This will resolve before end of pregnancy or within first year of life.
Routine Antenatal Ultrasound examination:

During the ultrasound examination, the appearance of the fetal renal system can vary in both normal fetuses and those with hydrenephrosis.

Therefore, this diagnosis should not be based upon a single measurement. An increase of maternal hydration can also increase the RPD in both normal fetuses and those with hydrenephrosis.
The following parameters need to be evaluated by ultrasonography as they guide further need for evaluation, intervention, and can be helpful in determining the cause of hydronephrosis:

- **Severity of hydronephrosis**

- **Unilateral versus bilateral involvement**: Bilateral involvement increases the risk of a significant abnormality and the risk of impaired postnatal renal function.

- **Ureter**: Dilatation of the ureter can be consistent with VUR or obstructive uropathy distal to the ureteropelvic junction [eg, ureterocoele megaureter, or posterior urethral valves (PUV)].

- **Renal parenchyma**: Thinning of the parenchyma indicates injury or impaired development of the renal cortex. An echogenic renal cortex may indicate abnormal renal parenchymal development (dysplasia), which may be associated with vesicoureteral reflux.

- **Bladder**: Abnormalities of the bladder such as increased thickness and trabeculation of the bladder wall are consistent with obstructive uropathy distal to the bladder (eg, PUV). In addition, dilatation of the proximal urethra (key-hole sign) may indicate PUV in male fetuses patients with a thickened bladder wall and hydronephrosis.

- **Amniotic fluid**: Oligohydramnios is consistent with impaired renal function resulting in a decrease production of fetal urine (amniotic fluid). It is a consistent feature of severe renal disease affecting both kidneys or a solitary kidney.
Grade 4 hydronephrosis (Severe hydronephrosis)

Dilated posterior urethra and bilateral hydroureter
Antenatal Management:

(1) Fetal karyotype:

(2) Vesicocentesis:

(3) Fetal surgery:

(4) Early delivery:

(5) Parents education:
(1) **Fetal karyotype:** In fetuses with mild hydronephrosis and normal amniotic fluid, a careful examination for genitourinary and extrarenal abnormalities should be performed. Clinical risk factors and ultrasound markers of Down syndrome should be evaluated. Assessment of the fetal karyotype should be offered if additional fetal anomalies are detected, in women of advanced maternal age, and women with abnormal maternal serum screening tests for Down syndrome.

(2) **Vesicocentesis:** Elevation of urinary sodium, chloride, calcium, alpha2-microglobulin, and osmolality indicates renal injury and potentially irreversible dysplasia. Urinary calcium is currently thought to be the most sensitive predictor of renal dysplasia. Currently, evaluation of urinary components is an extremely valuable tool in determining which fetuses are candidates for in utero intervention (3 samples should be obtained with 24-48 hours gap).

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Normal Range</th>
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<tbody>
<tr>
<td>Sodium</td>
<td>&lt;100 mg/dl</td>
</tr>
<tr>
<td>Chloride</td>
<td>&lt;90 mg/dl</td>
</tr>
<tr>
<td>Osmolality</td>
<td>&lt;200 mOsm/l</td>
</tr>
<tr>
<td>Calcium</td>
<td>&lt;8 mg/dl</td>
</tr>
<tr>
<td>β-2 Microglobulin</td>
<td>&lt;6 mg/dl</td>
</tr>
<tr>
<td>Total protein</td>
<td>&lt;20 mg/ml</td>
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</tbody>
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*Figure 3: Urine parameters indicative of renal function on antenatal bladder tap for determination of suitability for intervention*
**Fetal surgery:** Although there have been case series of antenatal surgery in fetuses with severe hydronephrosis and oligohydramnios, this intervention has not been shown to improve renal outcome.

These procedures may increase the amount of amniotic fluid, thus potentially improving lung development and survival rate. In these rare cases, the procedure should only be performed in selected centers with expertise.

**Early delivery:** has been suggested for fetuses with severe oligohydramnios and documented lung maturation. Although there are no data documenting improved renal outcome with early delivery, early delivery may be indicated to reduce the risk of other adverse outcomes from oligohydramnios such as umbilical cord compression.

**Parents education:** should be explained of all of the ultrasound findings and their potential significance (e.g., transient insignificant finding, urinary tract obstruction, VUR, risk of renal insufficiency, association with Down syndrome).

Prenatal and postnatal management options should also be explained.
Figure 2: Algorithm for antenatal management of fetal hydrenephrosis associated with reduction in amniotic fluid
Fetal surgery for antenatal hydronephrosis

- **Candidates:** Fetuses with bilateral obstructions (usually males with posterior urethral valves) who lose amniotic fluid volume and develop signs of renal compromise before 24 weeks.

- **Types:**
  
  - **Vesicoameniotic shunt:** By placement of a specially designed tube, the Harrison Catheter, into the fetal bladder. A small hollow needle is placed through the mother’s abdomen into the fetal bladder, using the sonogram for visualization. The problem with these catheters is that they become plugged or dislodged, sometimes requiring repeated procedures.
  
  - **FETENDO fetal surgery:** by putting a very small (3-mm) fetoscope directly into the fetal bladder and disrupting the valves. This can now be done through a very small scope placed through a nick in the mother’s skin rather than through open surgery.
Repeat antenatal ultrasound examinations are performed to help guide management decisions. The timing is dependent on findings on the initial examination (the presence and nature of associated renal and extrarenal anomalies, severity of hydronephrosis, unilateral versus bilateral involvement, gestational age, and the amniotic fluid volume.

In general, repetition of the ultrasound two to three weeks later in fetuses with bilateral involvement or an affected solitary kidney and at 32 to 34 weeks gestation in those with unilateral involvement.
Postnatal management:

Physical examination

Abdominal mass: Enlarged kidney due to pelvirecteral junction obstruction OR Multicystic dysplastic kidneys.

Palpable bladder: Posterior urethral valves in a male infant.

Deficient abdominal wall with undescended testes: Prune Belly syndrome.

Poor stream and dribbling: Posterior urethral valves in a male infant.

Abnormalities in spine and lower limb: Neurogenic bladder.

Patulous anus: Neurogenic bladder.

Examination for other anomalies should also be carried out.
Blood and urine investigations:

All patients with bilateral hydronephrosis should have the following done within 48 hours of life:

- **Renal profile:** (If the first creatinine is elevated serial monitoring is advised.)

- **Liver function test, calcium and phosphate.**

- **Full blood count.**

- **Blood gas, Urea & electrolytes.**

- **Urine C&S.**
Postnatal Imaging studies:

**Ultrasound:**
- Perform at 1 & 6 weeks of life (physiologic volume depletion and relative oliguria).
- Do earlier in male infants suspected of having bilateral hydronephrosis or those with hydronephrotic single kidneys.

**Micturating Cystourethrogram (MCU):**
- All patients with postnatal hydronephrosis should get an MCU done.
- Needs to be done on an urgent basis in boys with bilateral hydronephrosis to exclude posterior urethral valves.
- Non urgent appointments can be taken for other patients.
Isotope Scan:

Should be done after MCU with Persistent HN in the absence of VUR, or RPD > 10 mm even in the presence of VUR.

MAG3 study can be done within 3-5 days of birth (ideal at 4 weeks) whereas DTPA is best delayed to 6 weeks of age when GFR is maximally mature.

It will assess:
• Total and individual renal function
• Drainage time from renal pelvis
(a) In a dilated system, washout occurs rapidly after intravenous frusemide.
(b) The system is considered obstructed if washout is delayed beyond 20 minutes after intravenous frusemide

N.B. Dynamic MRI.
1. Intrauterine hydronephrosis is evident on a scan obtained at 37 weeks’ gestational age.

2. Longitudinal scan over kidney obtained during first day of life fails to confirm hydronephrosis.

3. Longitudinal scan obtained on the third day of life reveals dilatation of the renal pelvis.
Antibiotics:

Antibiotic prophylaxis is started in infants with antenatal hydronephrosis until reflux has been excluded. Commonly used prophylactic antibiotics:

- **Trimethoprim** 1-2 mg/kg at night
- **Nitrofurantoin** 1 mg/kg at night. Recommended after 3 months old. May cause haemolytic anaemia in G6PD deficient babies.
- **Cephalexin** 5 mg/kg at night
- **Amoxicillin** 25 mg/kg at night
Posterior urethral valves:

- Insert urinary catheter for continuous bladder drainage.
- Send urine routine and cultures.
- Urgent MCUG to confirm diagnosis of posterior urethral valves (under coverage).
- Empirical second-generation cephalosporin is recommended if urine is turbid.
- Assess hydration status.
- Monitor renal function, electrolytes, acid base status and blood gas.
- Refer to Pediatric urologist / nephrologists for primary valve ablation or vesicostomy as soon as possible.
Algorithm for postnatal management of congenital hydronephrosis:

1. **Antenatal hydronephrosis** (chemoprophylaxis)

2. **Ultrasound kidneys & urinary tract**
   - **No Hydronephrosis**: Repeat Ultrasound in three months
   - **Confirmed hydronephrosis**: MCU and DTPA
     - PUJ obstruction confirmed, refer to Pediatric urologist
     - VUR confirmed, continue prophylactic antibiotic
       (DMSA Scan)

3. **Male infants with bilateral hydronephrosis and/or palpable bladder and poor urinary stream**: Urgent MCU to exclude PUV
Important statements:

- Obstructive lesions and lesions that affect both kidneys are uniformly more threatening than nonobstructive and unilateral lesions.

- The survival rate with unilateral renal obstruction approaches 100%. In the presence of a bilateral obstructive process, oligohydramnios is the best predictor of an adverse outcome.

- Fetal urine is a significant component of the amniotic fluid volume, and maintenance of adequate volumes is essential for normal lung development. If oligohydramnios is present (the second trimester), pulmonary hypoplasia and compression deformities of the skeletal system can result and significantly influence quality of life and survival.