

8. UROLOGIC ANOMALIES

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A) **ROLE OF ULTRASOUND IN URINARY TRACT SURVEILLANCE**

- Identify fetus with genitourinary tract anomalies
- Monitor effects of the anomaly on fetus
- Variables:
 - Gestational age at diagnosis
 - Site of genitourinary tract abnormality
 - Degree of dilation of urinary tract
 - Evidence of urinary tract obstruction
 - Other system associated anomalies

B) **NORMAL ULTRASOUND FINDINGS**

- **Bladder:** visualized at 10 weeks
 - Fills and empties cyclically
 - Maximum capacity 10cc at 30 weeks/ 50cc at term
- **Kidneys:** visualized at 12-13 weeks
 - Collecting system not seen
 - Renal pelvis AP diameter >10mm significant hydronephrosis
 - Renal calyces: not normally seen
 - Parenchyma: echogenicity similar or < liver/spleen
- **Adrenals:** visualized after 13 weeks
 - Hypoechoic triangles
- **Fetal sex**
 - Male genitalia / labia majora
 - 3% misdiagnosis
 - Nonvisualization: prone position, full breech, oligohydramnios, maternal obesity

C) PRENATALLY DIAGNOSED UROLOGICAL ANOMALIES

- Genital anomalies
- Bladder anomalies
- Prenatally diagnosed hydronephrosis
 - Hydrocele, hypospadias, ambiguous genitalia
 - Exstrophy, bladder agenesis
 - Hydronephrosis
 - Cystic kidney disease
 - Ovarian cyst
- Most common prenatally diagnosed anomaly
 - 30% prenatal US anomalies
 - 1/100 pregnancies
 - 1/500 (0.2%) significant uropathy

D) RENAL EMBRYOLOGY

- 3 phases which are interdependent
- **Pronephrosis**: nonfunctional / involutes by 5 weeks
- **Mesonephrosis**: secretes urine
 - Duct (Wolffian) contributes to ureteral bud development
- **Metanephrosis**: requires ureteral bud for differentiation (7th week)
 - Nephrons develop until 36th week gestation

E) GENITOURINARY ANOMALIES

- Bilateral renal agenesis (1/4000)
 - Oligohydramnios and pulmonary hypoplasia cause demise
- Hydronephrosis = collecting system dilation
 - Physiologic / non-obstructive
 - Obstruction - upper / lower urinary tract
 - Vesicoureteral reflux
- Prenatal hydronephrosis: What is significant?
 - Early 2nd trimester
 - 5-8 mm AP diameter
 - Third trimester (>24 weeks)

- AP diameter >10 mm
- AP pelvic-to-renal cortex ration 0.5
- Caliectasis / ureteral dilatation
- Physiologic hydronephrosis (mild)
 - Maternal hydration – little influence
 - Transient obstruction / VUR
 - Matured kidneys / ureteral folds
 - Bladder distention
 - Hormonal factors (similar to maternal)
- Prenatal hydronephrosis: Categorization
 - Obstructive:
 - Ureteropelvic junction (UPJ)
 - Primary megaureter (UVJ)
 - Posterior urethral valves (PUV)
 - Prune Belly Syndrome (PBS)
 - [Multicystic dysplastic kidney (MCDK)]
 - Ectopic ureter
 - Ureterocele
 - Non-obstructive:
 - Vesicoureteral reflux (VUR)
 - Prune Belly Syndrome (PBS)
 - Renal Parenchyma Anomalies
 - Multicystic dysplastic kidney (MCDK)
 - Autosomal Recessive Polycystic Kidney Disease (Infantile)
 - Autosomal Dominant Polycystic Kidney Disease (Adult)
- Factors prediction of renal outcome
 - Amniotic fluid volume
 - Parenchyma echogenicity
 - Degree of hydronephrosis
 - Renal function
 - Urinary chemistries
 - Other system anomalies
 - Renal Functional Assessment:

- Indirect:
 - Bladder cycling / diuretic stimulation
- Direct:
 - Urinary biochemistries – requires multiple samplings
 - Urine normally ultrafiltrate of serum
 - “Poor urine” – salt wasting
- Oligohydramnios: 4-5% pregnancies
 - Amniotic fluid leak
 - Amnion nodosum
 - Urinary tract anomalies
 - Bilateral hydronephrosis
 - Abnormal renal parenchyma development
 - Bilateral renal agenesis / hypoplasia / dysplasia
 - Pulmonary hypoplasia – mechanical

F) PURPOSE OF PRENATAL INTERVENTION

- Preserve renal function
 - Never demonstrated clinically / experimentally
 - ? able to intervene early enough
- Prevent pulmonary hypoplasia:
 - Mechanical restriction of lung growth / chest expansion due to oligohydramnios
 - Insufficient AF inhibits lung branching
- Indications for prenatal intervention:
 - Obstructive hydronephrosis – lower tract
 - Progressive
 - Bilateral / solitary kidney
 - Progressive oligohydramnios
 - Favorable urine biochemistries
 - Minimal renal dysplasia
 - No other system abnormalities
- Timing of intervention
 - <20 weeks: ? irreversible renal dysplasia
 - >32 weeks: consider early delivery

- Assess pulmonary maturity (L/S ratio)
- Methods of intervention
 - Bladder aspiration: diagnostic / therapeutic / sequential
 - Vesicoamniotic shunt: multiple placements / dislodging
 - Open / fetoscopic surgery
 - Complications/ risks significant (>50% early series)
- Unfavorable prognosis renal function:
 - Early / sustained oligohydramnios (<20 weeks)
 - Renal cortical cysts / marked renal echogenicity
 - Urinary electrolytes: “poor urine”
 - Na >100 mEq /L
 - Cl >90 mEq/L
 - Osmolarity >210 mOsm/L
 - β microglobulin >2mg/L
 - Calcium >8mg / dl

G) PRENATAL HYDRONEPHROSIS SURVEILLANCE

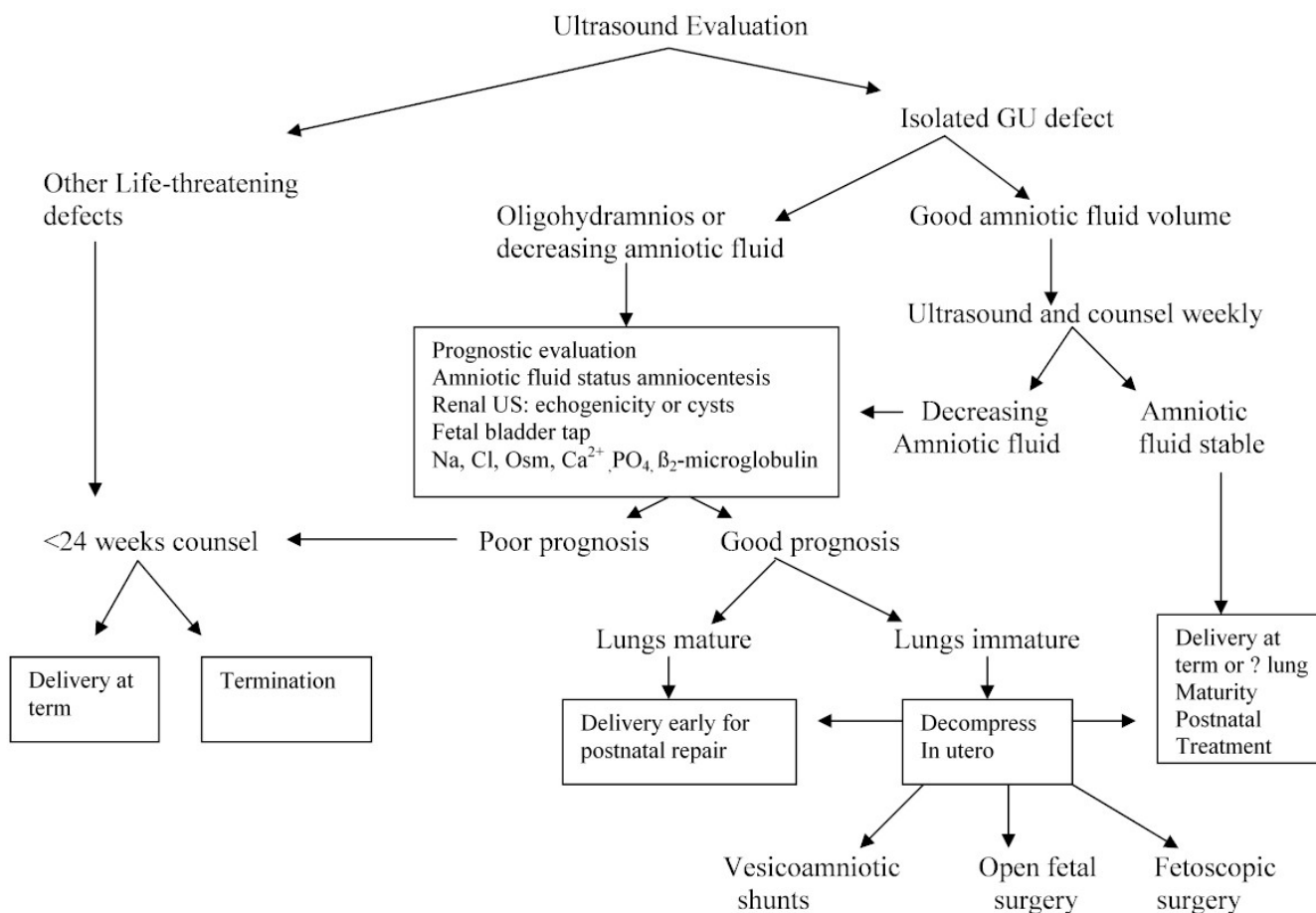
- Unilateral hydronephrosis <30 weeks
 - AF normal: US at 34-36 weeks
- Bilateral hydronephrosis
 - Complete diagnostic evaluation (Level II)
 - Complete ultrasound survey (GU / other systems)
 - Genetic amniocentesis (10% abnormalities)
 - Bilateral hydronephrosis and decreased AF
 - Bladder aspiration for biochemistries
 - Ultrasound every 2-4 weeks

H) POSTNATAL MANAGEMENT

- Evaluation for prenatal hydronephrosis
 - AP diameter >8-10 mm third trimester
 - Caliectasis
 - Ureteral dilatation
 - Abnormal renal parenchyma
 - Thick-walled / distended bladder
 - Urethral dilatation

- Is the child who presents with prenatal hydronephrosis the same child who would have presented with symptomatic postnatal hydronephrosis?
 - Evidence indicated not necessarily
 - Depends on etiology

Algorithm for Evaluation of Prenatal Hydronephrosis



Algorithm for the prenatal management of the fetus with bilateral hydronephrosis. GU, genitourinary; US, ultrasound. (Modified from Cendron, M D’Alton MD, Cromblie-holme M. Prenatal diagnosis and management of the fetus with hydronephrosis. Semin Perinatol 1994;18:163, with permission)

I) POSTNATAL RADIOGRAPHIC STUDIES

- Ultrasound: kidneys and bladder
 - Timing is critical to interpretation. False negative is possible within first few days (especially <48 hours old)
 - If initial US normal - repeat in 4-6 weeks
 - 50% will have a significant anomaly

SOCIETY FOR FETAL UROLOGY GRADING OF HYDRONEPHROSIS

Grade of Hydronephrosis	Central Renal Complex	Renal Parenchymal Thickness
0	Intact	Normal
1	Slight splitting	Normal
2 ^a	Evident splitting, complex confined within renal border	Normal
3	Wide-splitting pelvis dilated outside renal border; calices dilated	Normal
4	Further dilation of renal pelvis and calices (calices may appear convex)	Thin

^a An extrarenal pelvis extends outside the renal border, yet because the calices are not dilated hydronephrosis is grade 2. When the major calices are imaged but not dilated, hydronephrosis is also grade 2. From Maizels M. Grading nephroureteral dilatation detected in the first year of life: correlation with obstruction. J Urol 1992;148:1809, with permission.

- Voiding cystourethrogram (VCUG)
 - Even if ultrasound is normal
 - VUR/ PUV/ bladder wall thickness/bladder emptying
- Nuclear medicine studies
 - Technetium (Tc)- 99 m MAG-3 (mercaptoacetyl triglyceride) or Tc-99m diethylenetriamine pentacetic acid (DTPA)
 - Function and excretion
 - Tc-99m dimercaptosuccinic acid (DSMA) or Tc-99m glucoheptonate (GHA)
 - Function and renal cortical scarring (VUR)

Upper tract obstruction	Diuretic renogram DTPA / MAG-3
Function of MCDK	DMSA / MAG-3 / DTPA
Upper pole function duplex system	MAG-3 / DMSA / GHA
Renal agenesis confirmation	MAG-3 / DMSA / DTPA
Fusion anomaly (horseshoe/crossed ectopic)	DMSA / MAG-3 / DTPA
Acute pyelonephritis	DMSA
Renal scarring (congenital/VUR)	DMSA / GHA

- Antibiotic prophylaxis
 - Reduces risk of UTI / renal scarring
 - Newborn: penicillin based 1/3 – 1/4 therapeutic dose once daily until 3 months of age, then cotrimoxazole / nitrofuantoin / sulfa
- Nonrefluxing postnatal hydronephrosis
 - Grade 1-2
 - Follow up US 6 months
 - <3% risk obstruction
 - Nonobstructive UPJ / UVJ / ureteral fold

- Grade 3-4
 - MAG-3 lasix renogram- function/obstruction
 - UPJ / UVJ obstruction
 - Treatment based on function, symptoms, bilaterally
- Common causes of postnatal hydronephrosis
 - Ureteropelvic junction obstruction (UPJ)
 - Ureterovesical junction obstruction (primary megaureter / UVJ)
 - Duplication anomalies
 - VUR
 - Posterior urethral valves
 - MCDK

J) CONGENITAL ADRENAL HYPOPLASIA

- Autosomal recessive disorder, in female infants masculinized external genitalia due to elevated androgens. Enzyme defect in cholesterol-> cortisol pathway
- Masculization 10-16 weeks gestation
- Biochemical detection (17OH progesterone / adrenal androgens AF or HLA typing cultured AF cells) – 16-17 weeks gestation – too late for prevention
- Suppression of fetal pituitary adrenal axis could prevent masculinization of female fetus
 - Pregnancy with prior history of CAH infant
 - Dexamethasone / hydrocortisone treatment in 1st trimester
 - Reduces external virilization / no change in postnatal steroid requirements

K) MYELOYDYSPLASIA

- 1/1000 births, decreasing incidence
- Detection
 - Increased AFP (mother's serum)
 - US: open spinal defect
- Prevention
 - Folic acid early first trimester (pre-conception)
- In utero closure of spinal canal
 - No improvement in postnatal bladder function reported to date (urodynamic testing)