Neonatal Renal Review

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Session Summary

Homeostasis of the newborn is dependent upon a functioning renal system. The immature renal system of the newborn responds slowly and erratically to physiologic changes and demands that are placed on it. Knowledge of renal physiology and embryological development is essential in caring for these newborns, as renal disease, specifically renal failure, is common in ill neonates.

Acute renal failure (ARF) in the neonatal period has been recognized with increasing frequency and may occur in as many as 8% of infants in neonatal intensive care units. This lecture presents a practical overview of ARF in the newborn, reviewing practical considerations regarding diagnosis, recognition, and management of the infant with acute renal failure to help prepare the participant for certification exams.

Session Objectives

Upon completion of this presentation, the participant will be able to:

- differentiate between pre-renal, intrinsic, and post-renal causes of renal injury in the newborn;
- discuss the differences in neonatal vs. adult renal function;
- indicate the practical laboratory/radiology tests used to evaluate renal injury;
- discuss the evaluation and management of the newborn with renal injury.

Test Questions

1. Nephrogenesis is complete by ______ weeks EGA.
   a. 30
   b. 32
   c. 34
   d. 36

2. Prostaglandins produced by the kidneys have (a) ____________ effect on renal vascular system.
   a. vasoconstrictor
   b. vasodilation
   c. synergistic
   d. no
3. Newborns with spina bifida are at risk for ________:  
   a. intrinsic renal failure  
   b. post-renal failure  
   c. pre-renal failure  
   d. transcellular failure

4. The most common risk factor for renal failure in the newborn period is:  
   a. placental abruption  
   b. Apgar  
   c. prematurity  
   d. neonatal sepsis

References


Session Outline

See presentation handout on the following pages.
Neonatal Renal Function

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Non-Excretory Kidney Functions
- Produces renin
  - Regulates blood pressure
- Produces erythropoietin
  - Stimulates RBC production
- Metabolizes vitamin D
  - To calciferol (active form)
- Degrades insulin
- Produces prostaglandins
  - Potent vasodilator
  - Blood pressure regulation

Excretory Kidney Functions
- Maintain plasma osmolarity
- Maintain electrolyte balance
- Maintain water balance
- Excretes nitrogenous end products
- Excretes toxins
- Maintain acid/base balance
  - Regulating plasma bicarb level
  - Production of ammonia
  - Regulation of hydrogen ions

Anatomy
- Three major parts of the kidney
  - Renal pelvis
  - Medulla
  - Cortex

Renal Pelvis
- Innermost part
- Divide into major calyces
- Further divide into minor calyces
- Collect urine before it flows into ureter

Renal Medulla
- Middle layer
- Renal pyramids
- A portion of the tubules
- Loop of Henle
- Terminal collecting ducts
Renal Cortex
- Outer layer
- Glomeruli
- Proximal and distal tubules
- Collecting duct

Nephron
- Functional unit
- Clears plasma of unwanted substances
- Nephrogenesis
  - Begins @ 7-8 weeks
  - Completed by 34 wks
- Three major functions
  - Filtration
  - Reabsorption
  - Secretion

Filtration
- Blood is filtered under pressure
- Moves from glomerular vessel into nephron
- Filters through the walls of the glomerular capillaries and Bowman's capsule
  - Bowman's capsule prevents entry of RBCs and proteins
- Plasma entering nephron is the filtrate
- Filtrate is composed of water, ions (sodium, potassium glucose, small proteins

Functions of the Nephron
- Reabsorption
  - Movement of substances from filtrate into blood
  - Salts, nutrients, water
- Secretion
  - Active transport of substances into the nephrons

Glomerulus
The vascular component
- Afferent arterioles
- Efferent arterioles

The Tubular Component
Reabsorbs ions and water
Secretes unwanted substances
- Distal tubule
- Proximal tubule
- Loop of Henle
- Collecting ducts
**Glomerular Filtration Rate**

- Rate of fluid filtering from blood into Bowman’s capsule
- GFR dependent on
  - Blood pressure and renal blood flow
  - Gestational age
  - Postnatal age

**Renin–Angiotensin–Aldosterone System**

- Renin
  - Regulates ECF by regulating sodium content
  - Released when renal perfusion is decreased
  - Retention of sodium + water → Expansion of ECF
  - Regulates blood pressure

- Angiotensinogen
  - Produced in the liver
  - Renin converts it to Angiotensin I
  - Angiotensin I → Angiotensin II in the lung by ACE
- Antiotensin II
  - Potent Vasoconstrictor
  - Stimulates adrenals to release aldosterone
- Aldosterone
  - Stimulates reabsorption of Na and water are reabsorbed in the distal tubule
**Antidiuretic Hormone**

- Secreted by the posterior pituitary gland
- Increases reabsorption of water in collecting ducts
- Decreases urinary output
- Increases blood volume and blood pressure

**Syndrome of Inappropriate Antidiuretic Hormone**

- Inappropriate release of ADH
- Inappropriate retention of free water
- Severe dilutional hyponatremia
- Continued renal excretion of sodium (20 mEq/L)
- Serum Hypo-osmolality (<275 mMol/L)
- Urine hyper-osmolality (>100 mMol/L)

**Etiology of SIADH**

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<th>Other</th>
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<td>RSV</td>
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<tr>
<td>Hypoxia</td>
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**Treatment of SIADH**

- Water restriction is key!
  - Gestational age
  - Insensible water loss
  - Additional fluid sources of fluid loss
  - Sodium Replacement

Desired Na = Actual Na X (0.6) X weight in Kg

**Diabetes Insipidius**

- Abnormally high UOP
- Decreased levels of ADH
- Severe hypernatremia
- Dehydration
- Serum osmolality (less than 200 mMol/L)
### Diabetes Insipidus
- **Acute treatment**
  - Increase free water
  - UOP + IWL + water for growth
  - Limit sodium intake
- **Chronic therapy**
  - Desmopressin (DDAVP)
  - Synthetic ADH

### Kidney Maturation
- **Formation complete at 35 weeks**
- **Changes in renal blood flow**
  - Initially low due to high renal vascular resistance
  - Rate of decrease depends on degree of prematurity
- **Renal blood flow**
  - 6% of cardiac output during first week
  - 1st month of life (15–20%)
  - Adult (20–25%)
- **GFR**
  - Begins at 9–12 weeks
  - Triples from 28–40 weeks
  - Doubles by 3–7 days
  - Mature by 1–2 years

### Sodium regulation
- Infants have a poor ability to conserve sodium
- Exaggerated in premature infants
- Decreased proximal tubular reabsorption
- Decreased response to aldosterone

### Acid/base regulation
- Infants have very limited regulation ability
- Exaggerated in premature infants
- Potassium regulation
  - Decreased ability to excrete a potassium load

### Assessment of the Renal System
- **History**
  - Family history
  - Oligohydramnios
  - Maternal/infant drugs
  - Voiding history
  - Prenatal history
  - Ultrasound
- **Physical**
  - Palpitation
  - Enlarged kidneys
  - Masses
  - Distended bladder
  - Ascites
  - HTN
- **Laboratory**
  - Creatinine
  - Best clinical measure of GFR
  - Five days to reflect neonatal values
  - >1.5 mg/dl suggests renal impairment
    - Influenced by gestation and postnatal age
    - ↑ 0.3–0.5 mg/day abnormal
      - Doubling the value, decreases function by 50%
      - 0.5 have 100% of renal function
      - 1.0 have 50%
      - 2.0 have 25%
      - 4.0 have 12.5%
  - Isolated ear anomalies
    - Tags
    - Abnormalities of the lower GI system
    - Abnormal external genitalia
    - Chromosomal abnormalities

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**Assessment of the Renal System Laboratory**

- Blood Urea Nitrogen (BUN) (5–10)
- Nonspecific
  - Hydration status
  - Blood in GI tract
  - Catabolism (steroids)
  - Excessive protein administration
  - Total protein and protein metabolism

**Assessment of the Renal System Laboratory—Urinalysis**

- Specific gravity (1.001–1.020)
  - Kidney’s ability to concentrate or dilute urine.
  - False elevated by protein and glucose
- pH (4.5–8.0)
  - Kidney’s ability of the kidneys to maintain a normal acid base by excreting endogenously produced acids
- Protein
  - Can represent injury
  - Minimal amount should pass through glomerulus

**Assessment of the Renal System Laboratory—Urinalysis**

- Blood
  - ARF, congenital malformations, infection, RVT
- Leukocyturia (pyuria)
  - Most common cause is UTI

**Assessment of the Renal System Laboratory**

- Fractionated excretion of sodium (FeNa)
  - Reflects functional ability of the renal tubules
  - Proportion of sodium in GF excreted in urine
  - Distinguishes prerenal from renal failure
  
  \[ \text{FeNa} (\%) = \left( \frac{U \text{Na}}{U \text{Cr}} \right) \times 100 \]
  
  FeNa (\%) = \frac{U \text{Na}}{U \text{Cr}} \times 100

- Values (increased = renal disease)
  - 28 wk GA: 5–6%
  - 33 wk GA: 3–5%
  - 40 wk GA: 1–3%

**VCUG**

Voiding cystourethrogram

Outlines the bladder and urethra
Radionuclide Renal Imaging
- Visualize kidney mass, ureter and bladder outline
- Determines amount of functioning kidney
- CT/MRI – rarely indicated

Acute Renal Failure
- Loss of water & electrolyte homeostasis
- Acid/base disturbances
- Secondary to abrupt decrease in GFR
- UOP abnormalities
- Progressive rise in BUN & Cr

Often reversible if recognized and treated early!

Risk Factors for ARF
- 8–24% of NICU admissions
- Sepsis (39%)
- Perinatal asphyxia (17%)
- Hypotension not associated with sepsis (10%)
- “Just being premature” (32%)

Types of Acute Renal Failure
1. Prerenal ARF (80%)
2. Intrinsic ARF (11%)
3. Postrenal ARF (3%)

Pre-Renal Failure
- State of relative hypoperfusion
- Predisposing conditions
  - Hypotension
  - Hypovolemia
  - Hypoxemia
- Risk Factors
  - Dehydration
  - RDS
  - CHF
  - Asphyxia
  - Septic shock
  - Hemorrhagic conditions
  - Cardiac surgery

B12: NEONATAL RENAL REVIEW
Uncorrected Prerenal ARF Can Progress to Frank Renal Damage Leading to Intrinsic ARF!

Intrinsic Renal Failure

- Loss of function due to structural damage
- Acute tubular necrosis
  - Hypoxemia
  - Perinatal asphyxia
  - Prolonged hypoperfusion
  - Direct nephrotoxic injury
- Renal artery/vein thrombosis
- Congenital renal abnormalities

Post Renal Failure

- Secondary to obstruction of urine flow
  - Generally considered reversible
- Associated conditions
  - bilateral ureteropelvic junction obstruction (UPJ)
  - posterior urethral valves
  - neurogenic bladder

Symptoms of ARF

- Oliguria/Anuria
  - Less than 1cc/kg/hr urine output
- Accumulation of waste products
  - Electrolyte imbalance
  - Acidosis
- Polyuria (>5mL/kg/d)
  - 60% of infants with HI have non-oliguric ARF
- Hematuria
- Proteinuria

Distinguishing Types of ARF

- Fluid challenge
  - Rules out prerenal
  - 20 cc/kg NS.
  - If no urine produced then administer Lasix
- Urine Na
  - <30 meq/l in prerenal
  - >30 meq/l in renal
- Catheterize
- Continue with intrinsic renal workup

Prerenal ARF

- The most common cause
- Hypoperfusion of a normal kidney
- Rapidly reversible if treated
- If not treated, intrinsic damage may occur
**Intrinsic ARF**

*It's a Boy!*

**Acute Tubular Necrosis (ATN)**

- Damage to renal parenchyma
- Due to primary ischemic injury
- Risk factors
  - Hypoxia
  - Asphyxia – most common cause
  - Hypoperfusion of kidneys
  - Hypotension
  - Decreased cardiac output
  - Nephrotoxins
  - Infection

**Congenital Abnormalities**

- 1% of live births have renal anomalies

*The kidney shape was a cool idea. Hannens us what you said to pay for it.*

**Potter’s Syndrome**

- Bilateral agenesis
  - 1/3000
  - Incompatible with life
- Etiology
  - Metanephric diverticulum
  - Fails to develop or degenerates
- Diagnosis
- Consequences
  - Stillbirth – 40%
  - Premature delivery

**Potter’s Syndrome**

- Wide set eyes
- Parrot beak nose
- Receding chin
- Low set ears
- Contractures
- Pulmonary hypoplasia

**Multicystic Kidney**
- Usually unilateral
- Large parenchymal cysts
- No continuity between glomerulus and calyces
- No functional kidney
- Occluded/atretic ureter

**Multicystic Kidney**

- Symptoms
  - Flank mass
- Diagnosis
  - Ultrasound.
  - May be in utero
- Treatment
  - Removal of kidney
  - HTN, infection and cancer

**Symptoms**

- Abdominal mass  
- Pulmonary hypoplasia  
- Can be normal at birth

**Diagnosis**

- Ultrasound

**Treatment**

- Supportive
- Dialysis/Transplant
- Treat hepatic disease
Hydronephrosis

- Dilation of the renal pelvis and calyces due to obstruction of urine flow

### Degrees of Hydronephrosis

- Mild
- Moderate
- Severe

Hydronephrosis

- Obstruction
  - Pelvic–ureter junction
  - Vesico–ureter valve
  - Post-urethral valves
- Nonobstructive
  - Vesicourethral reflux
  - Prune belly syndrome

Hydronephrosis

- Symptoms
  - UTI
  - Abdominal mass
  - If severe signs of ARF
- Diagnosis
  - Renal Ultrasound
    - Delay for 24-72 hours
  - VCUG
  - Nuclear scan

Hydronephrosis

- Treatment
  - Most will spontaneously resolve
  - Mild to moderate – monitor
  - Prophylactic antibiotics

Prune Belly Syndrome (Eagle–Barrett Syndrome)

- A triad of defects
  - Deficient abdominal wall muscles
  - Genital abnormalities
    - More common in males
    - Intra–abdominal testes.
  - Urinary tract and kidney abnormalities
    - Dilated ureters, dilated urethra, reflux, patent urachus

Prune Belly Syndrome

- Other possible anomalies
  - Effects of oligohydramnios
  - CHD (ASD, VSD, TET)
  - GI abnormalities
**Prune Belly Syndrome: Treatment**
- Renal and urinary tract evaluation
- Delayed
  - Surgical plication of the abdominal muscles
  - Correction of urinary anomalies unless critically obstructed

**Vascular Disorders of the Kidney**
- Newborns are at increased risk
  - Decreased RBF
  - Increased with disorders such as hypoxia
  - Polycythemia
  - Lines
  - Hypovolemia

**Renal Vein Thrombosis**
- Usually unilateral
- Symptoms
  - Renal enlargement (60%)
  - Sudden flank mass
  - Gross hematuria
  - Thrombocytopenia (90%)
  - Decreased renal function
- Diagnosis
  - Ultrasound
  - Inferior vena cavogram

**Renal Artery Thrombosis**
- Risk factors
  - UAC
- Symptoms
  - No renal enlargement
  - Hematuria
  - Decreased renal function
  - HTN
- Diagnosis
  - Ultrasound
  - UAC cardiography
- Treatment
  - Treat HTN and renal failure
  - Thrombolytic therapy

**Postrenal Failure**
Post Urethral Valves

- Occurs exclusively in males
- Obstruction of flow from the bladder into the urethra

Treatment of Intrinsic Renal Failure

- Goals
  - Limit further injury
  - Enhance recovery
  - Treat underlying cause
- Treat complications
  - Hypervolemia
  - Electrolyte disturbances
  - Hypertension
  - Acid/base abnormalities

Electrolyte Abnormalities

- Hyponatremia
  - Fluid restriction
  - Sodium replacement
- Hyperkalemia
  - Limit potassium
  - May need to medically reduce
- Elevated phosphate
  - Kidney excretes excess
  - Calcium carbonate – binds and inactivate
  - Treat hypocalcemia
  - Hypocalcemia
  - Provide replacement

Hypervolemia

- Associated with HTN, CHF, hyponatremia
  - IWL + UOP = (25–40 cc/kg + UOP) – term
  - IWL + UOP = (70 cc/kg + UOP) = VLBW
  - Overhydration leads to edema, HTN, and CHF
- Calculate I&O and weight every 12 hours
- Frequent electrolyte monitoring
- Lasix
- Dopamine

Hypertension

- Due to volume overload and renal damage
- Fluid and sodium restriction
- Antihypertensive medications
**Overall Treatment of Intrinsic Renal Failure**

- Limit drugs and monitor levels
- Treat metabolic acidosis with bicarb
- Limit protein administration
- High degree of suspicion for sepsis
- Optimize nutrition
- Family support

**Peritoneal Dialysis**

- Easy and uncomplicated
- Treatment of choice in chronic management
- Slow removal of solutes and fluids
- Avoids hemodynamic instability

**Continuous AV Hemofiltration**

- Blood flow is generated by the patient’s blood pressure gradient
- Access to large vein and artery is necessary
- Difficult in small infants
- Systemic heparinization required
- Continuous venovenous hemofiltration

**ARF Prognosis**

- Depends on
  - Cause and extent of injury
  - Rapidity of diagnosis and treatment
- 50% mortality
- 40–50% have residual renal damage
  - Reduced GFR
  - Chronic renal failure (even ESRD)
  - Reduced concentrating abilities
  - Impaired renal growth
  - Chronic HTN
Fluid in the scrotal sac
- Originates in the peritoneal cavity
- Communicates with the scrotum through the processus vaginalis

**Diagnosis**
- Painless scrotal swelling
- Transillumination

**Treatment**
- 90% resolve by 1 year
- Surgery
  - Indicated if no resolution by 12 months
  - Drain fluid and close processus vaginalis

**Inguinal Hernia**
- Failure of the proximal part of the processus vaginalis to close
- Incidence increases as gestational age decreases
- More common in males
- May be unilateral or bilateral

**Signs and Symptoms**
- Mass in inguinal area
- May increase with crying
**Inguinal Hernia**

- Incarcerated Hernia (irreducible)
  - Firm, tense, won’t reduce
  - Pain
  - Vomiting, abd distension
  - Scrotal erythema

**Treatment**

- Immediate surgery consult
- Gentle reduction with ice, sedation and Trendelenburg position
- Surgery

**Incarcerated Inguinal Hernia**

- Strangulation
  - The constricted intestine becomes edematous and the blood supply is compressed
  - Ischemia and necrosis can result

**Extrrophy of the Bladder**

- 1/10,000–1/40,000
- More prevalent in males

**Three Variations**

- Epispadius
- Classic
  - An opening from the dorsal surface of the penis to the umbilicus
- Cloacal extrophy
  - Epispadius
  - Classic bladder extrophy
  - Extrophy of the bowel
  - Anus and anal canal fail to develop
  - Penis is divided into 2 halves
Epispadius

Cloacal Exrophy

Treatment

- Delivery room
- Wrap with saran wrap to protect from injury
- Correction
  - Close bladder during the 1st 48 hours to minimize trauma and chance of infection
  - Reconstruct external genitalia
  - Correction of bladder continence at 2.5 to 5 years with bladder neck reconstruction

Hypospadius

Hypospadius
Hypospadius

- Etiology
  - Failure/delay in midline fusion of the urethral folds
  - Due to inadequate production of testosterone
  - Types depends on timing and degree of decreased exposure to testosterone
- Treatment
  - Treat chordee if present
  - Use preputial skin to construct new urethra
  - Delay circumcision
  - 1 staged operation at 3-6 months

Ambiguous Genitalia

- A deviation in the normal steps of sexual differentiation
- A psychological and medical emergency
- Rapid diagnosis and management is critical
  - Ensure future psychological, social, and sexual health

Classification of Intersexuality

- True hermaphrodite (ovary and testes)
- Female pseudohermaphrodite (46,XX)
  - Congenital adrenal hyperplasia
  - Exogenous androgen exposure
- Male pseudohermaphrodite (46,XY)
  - Defects in testosterone synthesis
  - Inability to convert testosterone to DHT
  - Androgen receptor defects
- Gonadal dysgenesis

True Hermaphrodite

- Both ovarian and testicular tissue present
  - One on each side,
  - Bilat ovotestes
  - Combination of an ovotestes with either a ovary or a testes
- Phenotype
  - Normal male or female
  - Generally some ambiguity exists
- Diagnosis is through laparotomy

Female Pseudohermaphrodite

- 60–70% of all cases of disorders of sexual differentiation
- Always a normal female karyotype (46, XX)
- Normal uterus, fallopian tubes, ovaries, and upper vagina
- No Wolffian remnants
- External genitalia is virialized due to intrauterine exposure to androgens

Congenital Adrenal Hyperplasia (CAH)

- Most common disorder of sexual differentiation
- Due to endogenous exposure of androgens
- 21 hydroxylase deficiency
  - Most common (90%)
  - Necessary for the formation of cortisol
  - Maternal androgen exposure
  - Other missing enzymes
21 Hydroxylase Deficiency

Lack of 21 hydroxylase (an enzyme necessary for cortisol formation) =
- Decreased cortisol level
- Increased ACTH (adrenocorticotropin hormone)
  - Adrenal hyperplasia
  - And
- Accumulation of precursors which have androgenic actions =
  - Virilization of external genitalia

21 Hydroxylase Deficiency

- Elevated serum levels of 17-hydroxyprogesterone
  - A precursor of cortisol
- Phenotype (wide range)
  - Slightly enlarged clitoris
  - Normal appearing male without palpable gonads
- Phenotype based on magnitude, duration, and timing of exposure
- 21-hydroxylase required for the synthesis of aldosterone and the regulation of sodium
  - 30–50% develop hyponatremia

Treatment of CAH

- Rehydration
- Correct sodium imbalances
- Cortisol administration
- Florinef administration
  - Mineral corticoid to prevent hyponatremia
- Surgical repair
- Can be diagnosed through routine neonatal screening

Male Pseudohermaphrodite

- Normal male karyotype (46,XY)
  - Phenotype ranges from normal female with a blind vagina to a normal male with hypospadias
- Normally functioning testes
- Defect in the biosynthesis of testosterone
  - Deficiency of one of 5 enzymes necessary for synthesis
  - Inability to convert testosterone to dihydrotestosterone (DHT)
  - Five alpha reductase deficiency
  - Androgen receptor defects

Gonadal Dysgenesis

- Varying degrees of ambiguity
- Dysgenic male pseudohermaphrodite
  - Bilateral dysgenic testes
  - Testes do not secrete MIS so a uterus, vagina and at least one fallopian tube will be present
- Mixed gonadal dysgenesis (MGD)
  - Normal testes on one side
  - Gonadal streak on the other
**Evaluation and Differential Diagnosis**

- **Male**
  - Palpate bilateral testes
  - Most important element
  - Gonad below the inguinal ring are always testes
  - Stretched penis > 2 cm
  - Midline fusion line of the scrotum
  - No hypospadius/chordee
  - One perineal opening
- **Female**
  - Clitoris ≤ 1 cm
  - Two separate perineal openings
  - No fusion of the labia

**Ambiguous Genitalia**

- **Evaluation and Differential Diagnosis**
  - Ambiguous genitalia
    - Hypospadius with undescended testes
    - Hypospadius with separation of scrotal sacs
    - Clitoral hypertrophy of any degree
    - Partial fusion of the labioscrotal folds
    - Bilateral nonpalpable testes in a FT male

**Evaluation and Differential Diagnosis**

- **Family history**
- **Chromosomal analysis**
- **Endocrine studies**
  - 17-hydroxyprogesterone
  - Assess for rare forms of CAH
  - 5 alpha reductase
  - Testosterone level
- **Radiologic studies**
  - Pelvic ultrasound to assess for uterus
  - Genitogram to assess for vagina
  - Laparotomy and laparoscopy

**Ambiguous Genitalia**

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Management

- Main considerations in gender assignment
  - Future unambiguous appearance of the genitalia
  - Adequate sexual functioning
  - Fertility
- Extremely controversial

Family Support

- Considered a psychological emergency
- Care begins in the delivery room
- Naming the child/completion of the birth certificate
- Admit as a neutral name with a neutral name cared
- Discuss “touchy” subject with parents
- Examine with parents to demonstrate abnormalities
- Emotional tone established by team makes lasting impressions