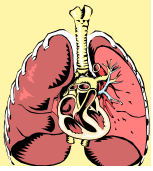
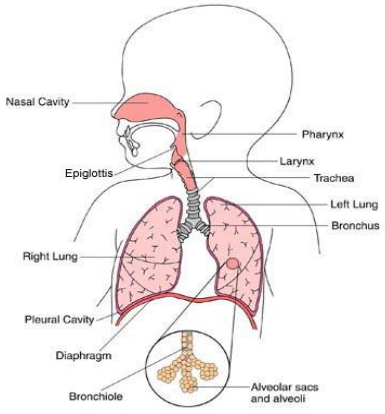
Problems of Development
Respiratory: Pediatrics

- BRONCHIOLITIS
- BRONCHOPULMONARY DYSPLASIA (BPD)
- CYSTIC FIBROSIS



L. Thompson MSN, RN
N124 - Pediatrics



BRONCHIOLITIS

- **Acute inflammation and obstruction** in:
  - Bronchioles
  - Small bronchi
- **Pathogens** (usually viral)
  - **RSV**-Respiratory syncytial virus **(50%)**
  - Parainfluenza virus
  - Adenovirus
- **Incidence**
  - < 2 yrs. (peak @ 6 months)
  - Winter and spring
  - One of the most frequent cause of hospitalization
- **Without Rx, may cause death in at-risk patients**

Bronchiolitis
Assessment

Assessment

- Fussy, malaise
- URI for 1-2 days (w/ serous nasal discharge)
- Low grade fever
- Elevated ESR or CRP
- Skin color
- S/S of lower respiratory obstructions
- Nasal flaring/retractions
- Breath sounds:
  - prolonged expiration, wheezes & crackles/rales

Vital Signs

Bronchiolitis
Diagnosis

- **Chest Xray**
- **Throat culture**
- **Nasopharyngeal Swab**
  - identify RSV & other viruses

Bronchiolitis
Therapeutic Management

- **Symptomatic Rx**
  - 95% can be managed at home with:
    - high humidity
    - adequate fluid intake
    - rest
    - manage fever

- **Indications for Hospitalization**
  - tachypnea/retractions/apnea
  - poor fluid intake

Bronchiolitis
Therapeutic Management

- **Medical Tx**
  - Nebulized bronchodilators/steroids
  - IV fluids, O_2, antipyretics
  - Antibiotics for secondary bacterial infection
  - Mechanical ventilation for respiratory failure

Bronchiolitis
Therapeutic Management

- **Medications** (controversial)
  - ***Ribovirin*** (Virazole) – aerosol antiviral medication
  - ***Caffiene Citrate*** - for apnea
- **Prevention of RSV**
  - **Contact Precautions while hopsitalized**
  - **High Risk Infants** (premature, BPD, CF)
    - ***IGIV*** - immune globulin intravenous (RespiGam)
    - ***Palivizumab*** **(Synagis)**
      - Antibodies to prevent serious lower resp. tract diseases

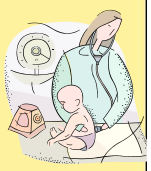
Bronchiolitis
Nursing Management

Assessment

- Vigilant observation

Nursing Diagnosis and Interventions

- ***Ineffective airway clearance r/t increased mucous production.***
  - SaO2 >94%
  - Elevate HOB
  - Ensure all nebulizer and CPT tx are given
  - Nasopharyngeal suctioning
- ***Fluid Volume Deficit r/t poor intake***
  - Strict I&Os
  - IV hydration, oral fluids
  - Monitor Labs

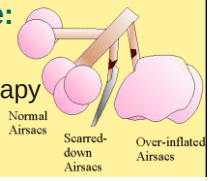


Knowledge deficit, caregivers r/t unfamiliarity w/disease

BRONCHOPULMONARY DYSPLASIA

BRONCHOPULMONARY DYSPLASIA (BPD)

- **AKA: CLD (chronic lung disease of infancy)**
- **Chronic obstructive lung disease**
- **Risk Factors**
  - Newborns requiring mechanical ventilation
- **Reduced incidence since:**
  - Antenatal steroids
  - Surfactant replacement therapy
- **Mortality 10%-25%**



BRONCHOPULMONARY DYSPLASIA

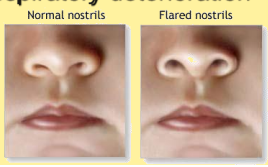
Pathophysiology

- Tx of Respiratory Distress Syndrome (RDS)
  - O2 + positive-pressure ventilation
- Damages bronchial epithelium
- Airway edema occurs
- Alveolar walls thicken
- Fibrotic changes occur in airways and alveoli
- Loss of ciliated cells
- Continued O_2 use decreases developing alveoli

BRONCHOPULMONARY DYSPLASIA

Clinical Manifestations

- Signs & Symptoms of respiratory distress
- Adventitious breath sounds
- Failure to thrive (FTT)
- Episodes of sudden respiratory deterioration r/t:
  - Bronchospasms
  - Mucous plugging
  - Air trapping



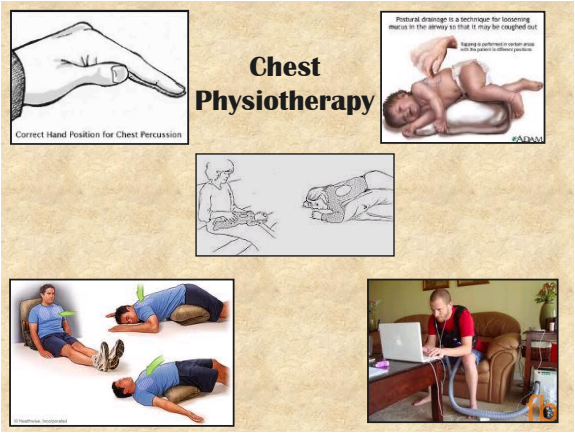
BRONCHOPULMONARY DYSPLASIA

Diagnostic Evaluation

- Xrays
- Clinical manifestations
- Required mechanical ventilation at age 1 wk
- O_2 dependence (*beyond 28 days of age*)
  - ***Mild***: room air by 36 weeks GA
  - ***Moderate***: <30% FiO_2 at 36 wk GA
  - ***Severe***: >30% FiO_2 at 36 wk GA.

BRONCHOPULMONARY DYSPLASIA
Therapeutic Management

- O_2 (possibly intubation or CPAP)
- Medications such as:
  - bronchodilators
  - corticosteroid
  - diuretics
  - electrolyte supplements
  - antibiotics, ribavirin
- Limiting fluids
- Nutrition – ex. Similac 27cal/oz
- Chest physiotherapy - CPT video
- Immunization – RSV - *Palivizumab* (Synagis)



BRONCHOPULMONARY DYSPLASIA

Therapeutic Management

- Ongoing therapy at home
  - Chest physiotherapy
  - Tracheostomy if necessary
  - Awareness of S/S of distress
- Potential outcomes:
  - Developmental delays
  - Growth retardation
  - Pulmonary dysfunction in adolescents

BRONCHOPULMONARY DYSPLASIA

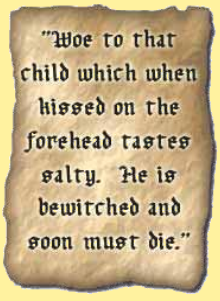
Nursing Management

- Ensure *all* therapeutic management
- Position infant to facilitate breathing
- Suction & tracheostomy care
- Prepare family for home care needs
- Discuss limitations
- Teach
- Teach
- Teach

Chris
1.2 lbs

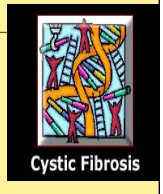
Problems of Development
Respiratory: Pediatrics

CYSTIC FIBROSIS



-Northern European folklore

CYSTIC FIBROSIS
Incidence



- **Gender is not a factor**
- **Incidence varies by race**
  - Whites 1: 3,200
  - Blacks 1:15,000
  - Asian 1:31,000
- **Median Life Expectancy**
  - 1930 1 year
  - 1960 7.5 years
  - 1990 27.5 years
  - 2004 30 years
  - 2014 37 years

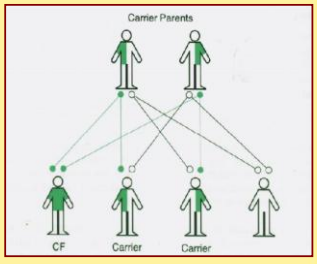
CYSTIC FIBROSIS
Inheritance Risks

- An inherited **autosomal recessive** disorder
- Genotype: ▪Homozygous ▪Heterozygous

Father

Mother	H	d
H	HH	dH
d	dH	dd

H="healthy" gene
d= disease gene
dd= disease
Hd or dH= carrier



CYSTIC FIBROSIS
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Father

Mother	H	H
d		
d		

(A)

	d	H
d		
d		

(B)

H="healthy" gene
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CYSTIC FIBROSIS
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Father

Mother	H	H
d	dH	dH
d	dH	dH

(A)

	d	H
d	dd	dH
d	dd	dH

(B)

H="healthy" gene
d= disease gene

dd= disease
Hd or dH= carrier

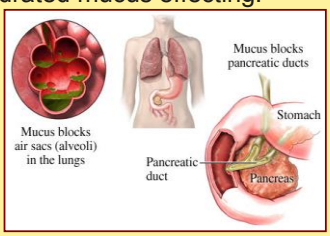
CYSTIC FIBROSIS
Pathophysiology and Etiology

- An inherited autosomal recessive disorder
- A mutation of chromosome 7
- The severity depends on form inherited
- **Dysfunction of the *exocrine* glands causing alterations in:**
  - Mucous secretions } leads to multiple
  - Sweat electrolytes } system damage

CYSTIC FIBROSIS
Pathophysiology

- Abnormal accumulation of viscous, dehydrated mucus effecting:

Respiratory System



GI System

Reproductive System

- Ultimately all body organs with mucous ducts become obstructed and damaged.

CYSTIC FIBROSIS
Clinical Manifestations

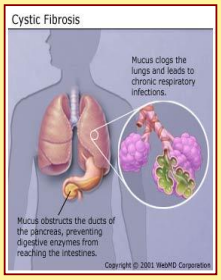
GI: Pancreatic Insufficiency

- *Earliest Signs*
  - **Newborn:**
    - Meconium ileus (distended abdomen)
  - **Infants & Toddlers**:
    - Fecal Impaction & Intussusception
    - FTT- from malnutrition (despite voracious appetite)
    - Steatorrhea (fatty stool)
    - Rectal Prolapse
    - Insulin insufficiency (DM)
    - Weight loss

CYSTIC FIBROSIS
Clinical Manifestations

- **Pulmonary manifestations**

(Major Cause of morbidity and mortality)
  - Coughing, sputum production
  - Chronic respiratory infections
  - Hemoptysis
  - Atelectasis
  - Pneumothorax
  - Digital clubbing
- **Metabolic manifestations**
  - "Salty Taste" (Sweat gland involvement)
  - Level of NaCl is 2-5 times above normal perspiration

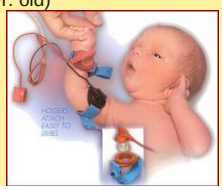


CYSTIC FIBROSIS
Clinical Manifestations

- **Reproduction**
  - ***Males*:**
    - Nearly all males are sterile due to the blockage of the vasdeferens.
  - ***Females*:**
    - Puberty delayed due to *chronic ventilation* and *nutritional deficiencies*.
    - Difficulty conceiving due increased mucus secretions in the reproductive tract interfering with passage of sperm.

CYSTIC FIBROSIS
Diagnostic tests

- Presence of classic symptoms
- Sweat Test (chloride concentration)
- Family history
- Genetic testing (DNA analysis)
- Pulmonary function test(>6 yr. old)
- Chest x-ray
- Stool analysis for fat and enzymes



Disease Management

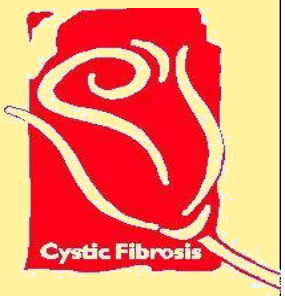
- **Maintaining respiratory function**
  - Bronchodilators, mucolytics
  - Chest Physiotherapy (CPT)
  - Steroids, NSAIDS
- **Managing infection**
  - Antibiotics
- **Promoting nutrition, hydration & exercise**
  - Pancreatic enzyme supplements
  - Diet: 120%-150% of RDA calories
    200% of RDA recommended protein
    Moderate Fat, salt supplements
  - Multi-vitamins & vitamin E in **water soluble** form (2x/day)
- **Preventing GI blockage** →Laxatives, increase fluids

Chest physiotherapy (CPT)

Airway Clearance

CYSTIC FIBROSIS
Home Care Teaching

- **Parents will need support and encouragement!**
  - Financial burden (medication, supplies & follow-up)
  - Obtaining necessary equipment (CPT 3-4x/day)
  - How to mix enzymes, what vitamins to give
  - Foods to encourage & avoid
  - Signs & symptoms of complications
- **Immunizations**
- **Family Support**
  - Genetic counseling
  - Support Groups
  - Cystic Fibrosis Foundation

Questions?

"65 Roses"