

## 5.3.) RESTRICTIVE LUNG DISEASES

Restrictive lung disease is due to low compliance (stretchability) of the lung tissue.  
As a result, lung volumes are low.

<p><b>pneumoconiosis</b></p>	<ul style="list-style-type: none"> <li>• <b>silicosis</b> (quartz)</li> <li>• <b>anthracosis</b> (coal)</li> <li>• <b>asbestosis</b> (fibrous mineral)</li> <li>• <b>berylliosis</b> (metal)</li> </ul>
<p><b>drug induced pulmonary fibrosis</b></p>	<ul style="list-style-type: none"> <li>➤ bleomycin</li> <li>➤ alkylating agents</li> <li>➤ oxygen therapy</li> </ul>
<p><b>hypersensitivity pneumonitis</b></p>	<ul style="list-style-type: none"> <li>• <b>farmer's lung</b> (actinomyces)</li> <li>• <b>cotton worker's lung</b> (byssinosis)</li> <li>• <b>pigeon breeder's lung</b> (animal protein)</li>   <li>➤ isocyanide</li> <li>➤ vinyl chloride</li> </ul>
<p><b>Löffler's syndrome</b></p>	<ul style="list-style-type: none"> <li>○ pulmonary infiltrate</li> <li>○ unknown cause</li> <li>○ eosinophilia</li> </ul>



*Asbestos related diseases :* - **asbestosis**: interstitial fibrosis  
 - **mesothelioma**: long latency, lethal  
 - **bronchogenic carcinoma**: 5-100 x risk



*“Honeycomb lung” = late stage interstitial fibrosis (any cause).*

## 6.14.) GALLBLADDER

Cholesterol is water insoluble and may crystallize in the bile depending on the balance of cholesterol and bile salt micelles. More than 20% of elderly persons in the US carry gallstones (usually asymptomatic).

<b>gallstones</b>	<p><b>75% cholesterol stones</b>  <b>25% pigment stones</b></p> <p>maybe asymptomatic (70%)          may cause biliary colic (20%)          may cause cholecystitis (10%)</p>
<b>biliary colic</b>	<ul style="list-style-type: none"> <li>• steady, cramplike (non-colicky!) pain in epigastrium</li> </ul> <p>pain <u>subsides over 30-60 min.</u></p>
<b>cholecystitis</b>	<ul style="list-style-type: none"> <li>• steady, cramplike pain in epigastrium</li> <li>• Murphy's sign (inspiratory arrest during palpation)</li> </ul> <p>pain <u>does not subside spontaneously</u></p>
<b>cholangitis</b>	<p><b>Charcot's triad</b> (1.) biliary pain          (2.) jaundice          (3.) fever</p>
<b>sclerosing cholangitis</b>	<ul style="list-style-type: none"> <li>• autoimmune inflammation of the bile ducts</li> <li>• rare complication of ulcerative colitis</li> </ul>

## 10.14.) AUTOIMMUNE HEMOLYTIC ANEMIAS

Caused by antibodies that cross-react with RBC antigens. RBCs covered with antibodies are then removed by phagocytic cells in the spleen (common) or lysed due to complement activation (rare).

<b>WARM TYPE - IgG</b>	<b>COLD TYPE - IgM</b>
<ul style="list-style-type: none"><li>• lymphoma, CLL</li><li>• SLE</li><li>• viral infections</li> <li>○ drugs: penicillin           quinidine</li></ul>	<ul style="list-style-type: none"><li>• mononucleosis</li><li>• mycoplasma infection</li><li>• lymphoma</li></ul>

### TWO UNUSUAL BUT INTERESTING DISEASES:

#### **1.) PAROXYSMAL NOCTURNAL HEMOLYTIC DISEASE:**

- RBCs, granulocytes and platelets are unusually sensitive to complement.
- Coombs' test negative.
- RBC lysis occurs in hypotonic solution ("sugar water test").

#### **2.) PAROXYSMAL COLD HEMOGLOBINURIA:**

- Cold-type IgG against red blood cell P-antigen (Donath-Landsteiner antibodies)
- complement mediated RBC lysis occurs after rewarming blood.

## 17.1.) BIRTH TRAUMA - 1

Head trauma during delivery is more likely when the mother has a small pelvis or if the infant is large for gestational age. Consider C-section rather than attempting difficult versions or vacuum or high-forceps deliveries!

Here is the range of trauma, from mild to severe:

<p><b>caput succedaneum</b></p>	<ul style="list-style-type: none"> <li>• diffuse edema of scalp soft tissue</li> <li>• <b>not limited to area of bones</b></li> </ul>
<p><b>cephalhematoma</b></p>	<ul style="list-style-type: none"> <li>• subperiosteal hemorrhage</li> <li>• <b>limited to area of affected bone</b></li> </ul>
<p><b>subdural hemorrhage</b></p>	<ul style="list-style-type: none"> <li>• <b>mechanical trauma, forceps</b> (cephalopelvic disproportion)</li> <li>• <i>more common in <u>large</u> infants</i></li> </ul>
<p><b>periventricular hemorrhage intraventricular hemorrhage</b></p>	<ul style="list-style-type: none"> <li>• <b>often occurs without obvious trauma</b></li> <li>• vulnerability to cerebral blood flow and pressure changes</li> <li>• <i>occurs in <u>small or preterm</u> infants</i></li> <li>• hypotension, bradycardia, apnea</li> <li>• lethargy, seizures, coma</li> </ul>

## **CRYPTORCHISM**

(if uncorrected: failure of spermatogenesis, increased risk for malignancy, but normal androgen production)

If present at birth:

- Observe: 80% of undescended testes are in the scrotum by age of 1 year
- Surgical repair (orchiopexy) should be performed at age of 1 years
- If bilateral: rule out that patient isn't a fully virilized female

If detected at puberty:

- Testis should be removed because of cancer risk

## **POLYCYSTIC KIDNEYS**

- Diagnosis: ultrasound
- Monitor renal function
- Manage complications of renal failure
- Strict blood pressure control
- Genetic counseling

## **WILM'S TUMOR**

- Resection plus multi-agent adjuvant chemotherapy
- Cure rates up to 90%

## **HENOCH SCHÖNLEIN PURPURA**

- Usually self limited: supportive care only
- Corticosteroids if gastrointestinal hemorrhage is present

## **CONGENITAL ADRENAL HYPERPLASIA**

- Goal: suppression of endogenous ACTH

Lifelong daily oral hydrocortisone:

- Increase dose 3 to 5fold during periods of stress (fever, surgery etc.)

Lifelong daily mineralocorticoid if salt wasting:

- Does not need to be adjusted for stress

## **CONGENITAL HYPOTHYROIDISM**

- Primary prevention: iodine supplementation prevents endemic hypothyroidism (cretinism)
- Secondary prevention: mandatory screening programs (T4, TSH)
- Levothyroxine for maintenance therapy

## **WILSON'S DISEASE**

(hepatolenticular degeneration)

- Lifelong penicillamine for symptomatic and asymptomatic cases
- Dietary copper restriction not practical
- Daily vit. B6 to prevent optical neuritis