**Cardiovascular Disorders**

**1-Acute rheumatic fever**

1-Acuterheumatic fever remains an important preventable cause of cardiac disease (1). Acute rheumatic fever **usually affects children** (most commonly between 5 and 15 years) or young adults.

2-The condition is triggered by **an immune-mediated response to infection with specific strains of group A streptococci**, which have antigens that may cross-react with cardiac myosin and membrane protein. Antibodies produced against the streptococcal antigens cause inflammation in **the heart as well as the joints and skin** .

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| **Table 1: criteria for the diagnosis of rheumatic fever** (2). |

**Clinical features**

1-Acute rheumatic fever is a multisystem disorder that usually presents with **fever**, and **joint** **pain**, **2–6 weeks after an episode of streptococcal pharyngitis**.

2-The presence of either two **major criteria** or one major and two **minor criteria**, along with evidence of preceding **streptococcal infection**, confirm a diagnosis of acute rheumatic fever.

[Streptococcal antibody tests, such as the antistreptolysin O (**ASO**) titer, are the most reliable laboratory evidence of prior infection].

**Management of the acute attack**

1-A single dose of benzyl penicillin 1.2 million U i.m. or oral phenoxymethyl penicillin for 10 days should be given on diagnosis to eliminate any residual streptococcal infection. If the patient is penicillin-allergic, erythromycin or a cephalosporin can be used.

2-**Bed rest is important**, as it lessens joint pain and reduces cardiac workload.

3-**Aspirin:** This will usually relieve the symptoms of arthritis rapidly. The usual dose of aspirin is 100 mg/kg/24 hr divided qid PO for 3-5 days, followed by 75 mg/kg/24 hr divided qid PO for 4 wk.

4-Patients with carditis and cardiomegaly or congestive heart failure should receive **corticosteroids**.

The usual dose of prednisone is 2 mg/kg/24 hr in 4 divided doses for 2-3 wk followed by a tapering of the dose that reduces the dose by 5 mg/24 hr every 2-3 days.

5-Supportive therapies for patients with moderate-to-severe carditis include **digoxin**, **fluid** and **salt restriction**, **diuretics**, and **oxygen**.

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| **2-Viral Hepatitis****Etiology**1-There are six primary hepatitis viruses: Hepatitis A virus (HAV), Hepatitis B virus (HBV), Hepatitis C virus (HCV), Hepatitis D virus (HDV), Hepatitis E virus (HEV) and Hepatitis G virus (HGV).2-They differ in their transmission, severity, likelihood of persistence, and subsequent risk of hepatocellular carcinoma.

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|  | **HAV** | **HBV** | **HCV** | **HDV** | **HEV** | **HGV** |
| **Transmission** | Fecal-oral | Transfusion, sexual,perinatal | Parenteral,transfusion,perinatal | Similar to HBV | Fecal-oral | Parenteraltransfusion |

**Note** - The most important risk factor for acquisition of **HBV in children is perinatal exposure to infected mother**. In most cases, transmission **occurred at the time of delivery**; virus contained in amniotic fluid or in maternal blood may be the source. However less commonly intrauterine infection occurred . 3-**HBV and HCV cause chronic infection**, which may lead to **cirrhosis** and is a significant risk factor for **hepatocellular** **carcinoma** .**Clinical Manifestations**1-**Asymptomatic or mild**, nonspecific illness without icterus (jaundice) **is common** with HAV, HBV, and HCV, **especially in young children**. 2-The **preicteric phase**, which lasts approximately 1 week, is characterized by headache, anorexia, malaise, abdominal discomfort, nausea, and vomiting and usually precedes the onset of clinically detectable disease.3-**Jaundice** and tender **hepatomegaly** are the most common physical findings and are characteristic of the **icteric phase**. Hepatic enzymes may increase 15- to 20-fold.4-Resolution of the hyperbilirubinemia and normalization of the transaminases may take 6 to 8 weeks .**Complications** 1-Most cases of acute viral hepatitis resolve without specific therapy, with less than 0.1% of cases progressing to **fulminant hepatic necrosis** which is associated with a high mortality rate.2-**HAV and HEV cause acute infection only**. HBV, HCV, and HDV may persist as **chronic infection** with chronic inflammation, fibrosis, and **cirrhosis and the associated risk of hepatocellular carcinoma** .**Diagnosis** The diagnosis of viral hepatitis is confirmed by **serologic testing**.**Treatment**1-The treatment of acute hepatitis (**except HCV** ) is largely supportive and involves rest, hydration, and adequate nutrition. Hospitalization is indicated for severe cases .2-**Chronic HBV** infection may be treated with **interferon alfa-2b or lamivudine**, and HCV may be treated with interferon alfa usually in combination with Ribavirin.**Respiratory Disorders** **1-Cystic Fibrosis****Background**1-Cystic fibrosis (CF) is an autosomal recessive multisystem disorder caused by mutations in the *cystic fibrosis transmembrane regulator* **(CFTR**) gene. 2- CFTR is important for the proper movement of salt and water across epithelial cell membranes especially in the airways, liver, and pancreas . The term *cystic fibrosis* arises from the fibrotic scar tissue that replaces the destroyed pancreas .**Pathophysiology****A-Pulmonary System**1-In CF, there are reduced chloride secretion with excessive sodium resorption which lead to dehydration of the airway lining leading to airway obstruction. This, in turn, leads to colonization with bacteria especially *Staphylococcus aureus* and *Pseudomonas aeruginosa* . 2-Chronic lung disease is a hallmark of CF, leading to death in 90% of patients. CF patients will usually experience **chronic respiratory infections**. **B-Gastrointestinal Involvement**1- Approximately 10% of patients with CF are born with intestinal obstruction caused by inspissated meconium (**meconium ileus**). In older patients, intestinal obstruction may result from thick inspissated mucus in the intestinal lumen .**C-Hepatic Involvement**1-In patients with CF, there is reduction in water and sodium movement into the bile. **The resulting decrease in the volume and flow of bile leads to stasis and obstruction of the biliary tree**. With chronic obstruction, this leads to **biliary cirrhosis** .**D-Pancreatic Involvement**1-The obstruction of the pancreatic ducts result in the inability to excrete pancreatic enzymes into the intestine. This leads to malabsorption of proteins, sugars (to a lesser extent), and **especially fat**. Fat malabsorption manifests clinically as **steatorrhea** (large foul-smelling stools), **deficiencies of fat-soluble vitamins** (A, D, E, and K), and **failure to thrive**.**E-Sweat Gland**In the sweat duct, CFTR reabsorbs chloride from sweat. Dysfunctional CFTR results in a **nearly fivefold elevation in sweat chloride concentrations**. This is the principal laboratory criterion for diagnosis of CF ( **sweat chloride test**) .**Diagnosis**CF is most commonly diagnosed on the basis of typical **signs and symptoms** and an abnormal sweat chloride concentration (>60 mEq/L) ( **sweat chloride test**).**Treatment** The treatment of CF is multifactorial, but it is primarily directed toward the gastrointestinal and pulmonary complications.**A-Gastrointestinal System**1-**Pancreatic enzyme replacement** (lipase, protease, and amylase) is the mainstay of gastrointestinal therapy .2-**Fat-soluble vitamins (A, D, E, and K) supplementation** is usually required in pancreatic insufficiency.3-The use **of ursodeoxycholic acid (UDCA)** may improves bile flow, prevent obstruction and slow progression of liver disease. **B-Treatment of Cystic Fibrosis Airway Disease**Treatment of CF airway disease involves the use of medications and techniques to mobilize pulmonary secretions, and antibiotics to manage infection. **1-Mucociliary Clearance****A-Physical Therapy:** Airway clearance can be performed using various techniques. These techniques are recommended **on a daily basis to help mobilize secretions** .**B-Mucolytic Therapy:** Sputum viscosity is increased by the large quantities of extracellular DNA that result from chronic airway inflammation and degradation of neutrophils. **Inhaled recombinant human deoxyribonuclease** (rhDNase, dornase alpha) cleaves extracellular DNA in sputum. **C-Airway Hydration Therapies :** Inhalation of hypertonic saline rehydrates the airways through osmotic flow of water.**D-Bronchodilators:** β-Agonists keep airways open and facilitate airway clearance **2-Antibiotics**1-Antibiotics are used to treat lung infection. *Typical regimens for severe infections include an antipseudomonal β-lactam plus an aminoglycoside for added synergy and delay of resistance development* 2-**Fluoroquinolone** use is common among CF patients infected with ***P. aeruginosa*,** even in children.3-**Chronic maintenance antibiotic therapy may be used in patients with *Pseudomonas*****colonization** in an attempt to prevent bacterial overgrowth. **Inhaled tobramycin** has been studied the most extensively **Pharmacokinetic Considerations**CF patients have **larger** **volumes of distribution of many antibiotics** and also have an enhanced total body clearance. As a result of these pharmacokinetic changes, **higher doses of antibiotics** (e.g. aminoglycosides, and β-lactam antibiotics) are needed **Lung transplantation** **Lung transplantation** is currently the only definitive treatment for advanced cystic fibrosis **Prognosis**The longevity of patients with cystic fibrosis is increasing, and the median survival age is over 35 years. Death occurs mostly from pulmonary complications **Endocrinology****1-Diabetic ketoacidosis (DKA)**1-**Definition**: Arterial pH <7.30, bicarbonate <15 meq/L, glucose >250 mg/dL, and Urinary ketones 2-DKA is a major **medical emergency** and remains a serious cause of morbidity, principally in people **with type 1 diabetes** (**More common in type 1 DM** but can occur in type 2 DM)  3-A significant number of **newly diagnosed diabetic children** present with DKA. In children with known diabetes, DKA occurs in patient who omit insulin doses or who do not successfully manage an intercurrent illness **Risk Factors**1-**Omission of insulin** is the most common precipitant of DKA 2-**Infections**, acute medical illnesses, and stress of recent surgical procedures can contribute to the development of DKA **Pathophysiology**1-The **hyperglycaemia** causes a profound **osmotic diuresis** leading to **dehydration**, hyperosmolarity, and **electrolyte loss**, particularly of sodium and potassium2-Owing to increased lipolysis and decreased lipogenesis, free fatty acids are converted to ketone bodies and lead to **metabolic acidosis** 3- **Electrolyte abnormalities** occur through a loss of electrolytes in the urineIn addition, The resulting metabolic acidosis causes efflux of potassium from cells, results in intracellular potassium depletion **Clinical Presentation**1-Patients with DKA present initially with **polyuria**, **polydipsia**, **nausea**, and **vomiting**. **Abdominal pain** occurs frequently 2-Respiratory compensation for acidosis results in **tachypnea with deep (Küssmaul) respirations**. The **fruity odor of acetone** frequently can be detected on the patient’s breath3-An altered mental status can occur, ranging from disorientation to coma **Management**1-DKA is a **medical emergency** which should be treated in hospital. The principal components of treatment are :• The administration of **short-acting** (soluble) insulin , rout and dose?• **Fluid** replacement • **Potassium** replacement, dose and rout of administration? Role of K+ administration• The administration of **antibiotics** if infection is present **Complications???** |