Rheumatic fever.
Peculiarities of clinical manifestations, diagnosis, treatment and prevention principles.

Definition. Acute rheumatic fever (FRA) is the most common rheumatic pathology and the main cause of acquired heart disease in children and adolescents.

Epidemiology and etiology
Acute rheumatic fever is aseptic complication arising after a latency period of 1-3 weeks of infection of the oropharynx, caused by β-hemolytic Streptococcus group A, which often affected children and adolescents aged 5 and 15 years. Only 0.3-3% of patients with streptococcal angina develop this complication, as there may be a predisposition genetics, that is not yet fully studied. Very often, patients with Acute rheumatic fever are from socially vulnerable. The pathogenic mechanism of the disease is the development of reaction of anti-streptococcal antibody and structures of individual's affected (molecular mimetics) that triggers the inflammatory.

Favoring causes of streptococcal infections:
• overcrowding and unsanitary conditions;
• promiscuity favoring contagiousness;
• lack of medical education - decreases addressability.

Clinical diagnosis
Diagnosis of Acute rheumatic fever is based on clinical. Tests, which serve to confirm the presence of pro-inflammatory ccess are positive in most cases. They are useful for assessing the existence of infection streptococcus. In 1992, Committee Rheumatic Fever, Endocarditis and disease Kawasaki the American Academy of Cardiology has updated Jones criteria to facilitate the diagnosis of Acute rheumatic fever. The presence of two major criteria or one major criteria and two minor indicates a high probability for disease provided demonstrating oropharyngeal streptococcal infection in recent medical history. There are two situations which included in this case: chorea delayed and subclinical carditis. In these cases clinical signs and laboratory data may be missing, may be in the normal range.

Criteria based on review criteria Jones
Primary or recurrent flare spurt in a patient with heart rheumatic - applied as such currently identified
Relapse in a patient with recurrent heart - two major + field see recent infection.
Core or aortic or mitral valve disease - no other criteria further rheumatoid

Major criteria: Carditis, Poliarthrite, Sydenham chorea, Erythema marginatum, Subcutaneous nodules
**Minor criteria:** fever, pain of joints; leukocytosis, high C-reactive protein and erythrocyte sedimentation rate; EKG: P-R high

**Polyarthrite** is usually migratory, affects big joints, each for a period of 4-5 days. Effect of joints lasts for 3-4 weeks. Arthritis is present over 75% of patients but rarely leave sequelae, answer quickly to anti-inflammatory treatment, usually after 48 hours its initiation.

**Carditis**

Carditis (usually pancarditis) is the only manifestation can cause death or late sequelae. It occurs usually in the first three weeks of disease. Patient blames fatigue, anorexia, chest pain and dyspnea. Myocarditis is manifested by tachycardia without fever, symptoms may progress to heart congestion and associations usually with the valvulite. Valvulite is manifested by the appearance of regurgitation systolic murmur at the apex with or without breath mezodiastolic the apex and / or diastolic murmur of the aortic regurgitation. Mitral valve is most commonly affected and may be associated with impaired aortic valve.

**Sydenham chorea**

Sydenham chorea is manifested by involuntary movements of trunk, face and limbs. These symptoms disappear during sleep, but it exacerbates the situation of stress, pana
tical and exercise. They may be unilateral or bilateral and permanent are associated with muscle pain and emotional lability. Emotional lability may precede chorea, sick child show decreased learning ability, laughing or crying without reason. Chorea may occur first 6 months of illness and must not be associated with other conditions clinical and laboratory of Acute rheumatic fever . It often affects teenagers.

**Erythema marginatum**

It manifests as a rash, with pale center and winding edges. It is caused by pression clothes and usually occurs on the trunk and proximal . Often it is associated with damage to the heart.

**Subcutaneous nodules**

They are rare, but often are associated with clinical signs of carditis. Nodules are hard, mobile, painless, localized, usually in the region of the bone and the path of the projections tendons, the spinous processes of the vertebrae in the region occipital snow.

**Diagnosis**

**Laboratory Tests**

Hemoleukogramme is usually not specific, are characterized by neutrophilic leukocytosis, normochromic anemia.

Particular importance is the inflammatory activity tests:
C-reactive protein, erythrocyte sedimentation rate, the mucoproteins, electrophoresis of proteins increase α2 and γ fractions are useful in the daily practice.
Standardized "gold" is smear oropharyngeal but streptococcal detection of streptococcus β-hemolytic group A in the acute phase of the disease may be less than 25% of the cases - due to influence previous antibiotic therapy.
Anti-streptococcal antibody titer is used extensively. These tests determine the antibodies developed against extracellular products of streptococcus and streptolysin O, deoxiribonuclease B, hyaluronidase, nicotinamide, adenine nucleotidase and streptokinase, but only anti-streptococcal O lysine can be easily determined.

EKG: P-R high

EcoKG: Valvulite of mitral valve and aortic valve

Radiografí cardiothoracic: cardiomegalie

**Treatment**

Symptomatic treatment includes NSAIDs are used in depending of the degree of joint damage. Aspirin 80-100 mg / kg / day.
In heart disease, it is recommended Glucocorticorticosterone. Sydenham chorea is treated with Haloperidol 1-2 mg/kg/ day
Also, the use of Valproic acid, Phenobarbital, Benzodiazepine and Sulpiride.

Primary prevention to prevent the development of Acute rheumatic fever after an episode of angina streptococcal, and the second prevention of new episodes in patients who have already Acute rheumatic fever in the past.

**Primary prophylaxis:**
Benzathine penicillin 600 000 U in children weighing less than 27 kg and those weighing more than 27 kg - 1.2 million U
Peroral use of Penicillin, Amoxicillin and Cephalosporins may be effective but it depends on duration of use, which must be at least 10 days.
Patients allergic to Penicillin will be write macrolide (Erythromycin or Azithromycin 10 mg /kg/ day - 5 days).

**Secondary prophylaxis:**
Benzatipenicilnine administered every 21 days in doses recommendeted for primary prevention. In patients with allergic reactions to Penicillins - Sulfadiazine is used - of 500 mg / day for children up to 12 years and 1 g / day for those older than 12 years.
Patients with heart disease are prophylaxis to those aged stay of 18 or for 5 years after initiation of treatment.
MORPHOLOGICAL AND FUNCTIONAL PARTICULARITIES OF THE CARDIOVASCULAR SYSTEM IN CHILDREN.

Cardiovascular system, with the complex functions of transport of nutrients, oxygen, and other metabolites by support immune defense and the humoral regulation of multiple physiological processes.

Ontogenesis cardiovascular system begins the second week follow from conception, the mesoderm, the formation, tube heart "primitive, located in the region" neck "of the embryo.

A series of changes, the tube will result in cardiac five segments:
• common arterial trunk, which will develop vessels main (aorta and pulmonary artery);
• cardiac bulb, which will develop the right ventricle;
• primitive ventricle - left ventricular predecessor;
• primitive atrium, which will develop the two atria;
• venous sinus, which will develop large veins.

Characteristics of fetal circulation
1 The existence of communications (fetal) between the right and left, and between ships lines - and two right-left shunts.
2 Significant increase in minute-volume circuit (higher) due to the absence of pulmonary function.
3 Providing preferential oxygen rich blood to organic vital (brain, heart, liver, upper limbs) by ascending artery and its bow.
4 Pressure equivalent practical aorta and pulmonary artery

Until puberty, there is a predominance of the right ventricle in terms of thickness and weight, after left ventricle triples the weight.

Heart rate decreases to adulthood, of 140 beats per minute to 70-72 beats per minute.
Minute-fluid volume is 120 ml / kg at the child to 60 ml / kg in adults.

The pericardium that surrounds the heart muscle on the outside, is constituent of conjunctive tissue elastic and composed of two skins with a few liquid between them.

Morphological cardiovascular system
• In neonates, the position of the heart in the chest is cross occupies about half of the width chest. By the age of 2-3 years heart position comes skewed.
• In neonates, the heart is oval with predominance of transverse dimensions.
• In children, atrial size relative to the size the ventricles are larger in adults.
• Weight heart relative to body mass in children is higher compared with adult, representing 0.8-0.9% and 0.4-0.5%.
• ventricular wall thickness at birth is almost equal, the ratio is 1: 1.4, the mass of the same, and by the age of 14 year left ventricular mass increases 17 times and mass of right ventricle, respectively 10.
• atria and vessels in neonates bus sizes relatively high compared to the respective dimensions of the adult.
• Histological structure of the myocardium in newborns: the myofibers very thin, poorly developed connective tissue, very good vascularization of the myocardium.
• Children myocardium has a cellular structure (syncytial) cardiomiocite with small nuclei, poorly differentiated.
• By age 7 children have morphological characteristic heart of adult. Myocardium grows particularly intense at the age of 12-14 years.
• Nervous system and conductivity in children is not perfect.

The functionality of the cardiovascular system
• The heart benefits from a more favorable compared to adults because of the absence of chronic poisoning in children (alcohol, nicotine, various noxious etc.).
• Cardiovascular system in children have opportunities repairing greater than in adults.
• Cardiac nervous activity in children is shown by the predominance of the sympathetic system.
• Children physiological tachycardia is characteristic with high frequency of cardiac contractions (120-140 beats / minute).
• Respiratory influence on the frequency of cardiac contractions of the children is manifested by respiratory arrhythmia.
• One of the most important functional parameters is the blood pressure (BP), which children's direct dependence of age - first is low, then increases with age. That's because the blood pressure is influenced by the ability of the left ventricle, volum of vascular circuit, blood vessel tone, etc. Can use the following formulas for assessing the value of blood pressure:
  
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  \text{BP (s) = 70 + 2n (n – represents the number of months);}
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  \[
  \text{after 1 year BP (s) = 80 + 2 n (n - age in years);}
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  \[
  \text{BP (s) = 102 + 0.6 from years of children;}
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  \[
  \text{BP diastolic by age}
  \]
  
  \[
  \text{BP (d) = 1/2 or 2/3 of the BP (s);}
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  \[
  \text{BP (d) = 63 + 0.4 from years of children.}
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Shortness of breath (dyspnoea)
The terms ‘shortness of breath’ and ‘dyspnoea’ are inter-changeable. They imply that the patient has difficulty getting their breath and that this difficulty in getting breathis in some way unpleasant. This symptom must be distinguished from hyperpnoea which is increased breathing and from angina.

Paroxysmal nocturnal dyspnoea
This dramatic symptom must be sought in taking the cardiovascular history and distinguished, if possible, from orthopnoea. When paroxysmal nocturnal dyspnoea (PND) occurs the patient wakes up with severe breathlessness, has to sit up and usually gets out of bed and stands up. They frequently go to the window because they believe that they can improve the situation by inhaling fresh air which they believe contains ‘more oxygen’.

Cheyne–Stokes respiration
The intermittent breathing that is usually the result of a low cardiac output is really a physical sign. It is often noticed by the patient’s partner during the night when breathing slows down and stops for some seconds before starting again. The patient may also be aware of it when they drift off to sleep, as the breathing slows and stops and then they wake up with a start as they begin to breathe again.
Sleep apnoea
The patient is usually obese and may be plethoric and their main complaint is of somnolence during the day and they may develop heart failure as a complication. However, in common with Cheyne–Stokes respiration, the patient’s partner is the best source for the history.

Cough
Although cough is usually regarded as a symptom denoting a respiratory origin to the patient’s breathing problems, there are circumstances where coughing may point to a cardiac problem. Coughing, particularly at night, may be caused by the early stages of pulmonary congestion and be a sign of impending left heart failure.

Palpitation(s) (cardiac arrhythmias)
Patients usually perceive cardiac arrhythmias as an awareness of their heartbeat and they may feel thumping in the chest but this may also be described as the heart racing or pounding or fluttering in the chest.

Presyncope and syncope
Occasionally patients may give a history of collapsing with syncope and then coming round after a short period and being aware of a rapid tachycardia. This may be because whatever caused the syncope also produced a tachycardia. However, more often it is because the sudden drop in cardiac output caused by the tachycardia and an unprepared dilated peripheral circulation causes a severe fall in blood pressure and syncope.

Fatigue
Fatigue is probably very common but hard to define as it has so many causes. It is most striking when a patient has a successful treatment and suddenly realizes how tired they were before. If it occurs intermittently it may have a definite underlying cause although this may be difficult to track down