ACQUIRED VALVULAR HEART DISEASE
IN CHILDREN: OUR EXAMPLES

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Abstract

The most prevalent congenital heart defects for paediatric cardiologists to deal with have traditionally been cardiomyopathy (primary disease of the heart muscle), arrhythmia (irregular heartbeat), and acquired heart disease. More recently, most developed countries are also engaged in combatting fetal echocardiography.

Rheumatic heart disease and Kawasaki disease are the two most common types of acquired disease in children. Rheumatic heart disease occurs in children aged 5-15 years, and Kawasaki disease is most common in those younger than 5 years. Both diseases represent inflammatory conditions. Kawasaki disease affects the lymph nodes, and blood vessels of the heart. Rheumatic disease affects the connective tissue, and causes inflammation of the joints, and especially inflammation of the heart.

Bacterial infections can affect not only the valves of the heart, but also the heart muscle, and the lining of the heart. Cardiac valves can be affected by systemic diseases, rheumatoid arthritis, and lupus.

Key words: heart valve, rheumatic heart disease, heart muscle, child

Introduction

Heart valves are elements of the cardiac apparatus, which enable it to work as a pump, supplying every cell with blood. When the valves are open, they allow blood to flow, and when they are closed they prevent it from returning. There are four heart valves: aortic, mitral, pulmonary and tricuspid. Heart valves do not work always properly. Congenital valvular disease is mainly evident immediately after birth, whereas more minor defects (such as bicuspid aortic valve disease), which are well-tolerated in childhood, are usually discovered later.
Acquired valvular heart diseases occur later in childhood. Almost all acquired heart diseases are rheumatic. Involvement of the mitral valve occurs in ¾, and aortic in ¼ of all cases with rheumatic heart disease. Stenosis and regurgitation of the same valve usually occur together. Isolated aortic stenosis of rheumatic genesis is extremely rare. Involvement of the tricuspid valve varies, and that of pulmonary valves almost never occurs.

Mitral regurgitation is the most common valve involvement in children with rheumatic diseases. Children do not usually show symptoms; fatigue and palpitations are rarely present. The main symptom is a systolic murmur at the apex.

Mitral stenosis, although rare in children, is the most common valve involvement in adult rheumatic patients. Most patients have no visible symptoms, but the main symptom is a diastolic murmur over the apex.

Aortic regurgitation is less common than mitral regurgitation, as most patients have a disease associated with the mitral valve. Patients with mild regurgitation have no symptoms. Tolerance to this disease can be reduced with severe regurgitation or heart failure. The patient can be without symptoms for a long time, but if symptoms begin to develop, the condition of most patients will rapidly deteriorate. Anginal pain, premature ventricular contractions, and congestive heart failure are danger signs.

In adult patients, degenerative diseases – such as senile aortic calcification – are the main cause of changes to the valves.
Rheumatic heart disease is a result of idling rheumatic fever, and occurs after untreated or improperly treated infection of the upper respiratory tracts, caused by the bacteria *streptococcus*. The main symptoms are manifested in the joints and the heart, but rarely in the skin and nervous system. This disease can lead to permanent damage of the heart. Since the introduction of penicillin to treat inflammation of the pharynx, there has been a sharp drop in the frequency of occurrence of rheumatic fever.

Rheumatic fever occurs in people who have a congenital tendency to the disease. After contact with bacteria, the immune system reacts not only against the bacteria, but also against the patient’s own tissues and organs. The most common sign — arthritis — leaves no lasting damage, but carditis (the most serious sign of rheumatic fever) if left untreated can have lasting effects on the heart. Rheumatic disease can affect any heart tissue, manifesting as pericarditis, myocarditis, or endocarditis.

Symptoms that indicate carditis are: tachycardia; quiet tones; cardiac murmur.

The doctor must assess the condition of the heart by listening to the patient’s heartbeat with a stethoscope. Discovery of a heart murmur is evidence that a rheumatic inflammation of the heart muscle is present.

![Figure 2. Clinical examination of a child’s heart](image)

ECG is also used to detect heartbeat irregularities.

A more accurate method for the diagnosis of carditis is ultrasound scanning, which shows the structure of the heart, and blood movement. If there is damage to the valves, ultrasound examination helps to detect the damage on time.

These methods are totally painless, and ultrasound scanning is harmless. There is no reason for the child not to be calm.
Rheumatic fever is treated with antibiotics. Streptococcus may remain hidden in the tonsils, and can stimulate the immune system.

In patients who have already had rheumatic fever, the use of the drug Extencillin is required every 3 weeks to prevent a relapse of the disease, which is common in the first 3-5 years. With each relapse the risk of damage to the heart increases. Antibiotics can be used for at least 5 years (or until 18 years of age), but no longer than this by those patients without heart damage. For patients with a damaged heart, it is recommended that antibiotic protection is taken until 40 years of age.

The prognosis of the disease depends on the severity of the clinical picture. If carditis was present at the first attack of the disease, the risk of later damage is increased. The most severe forms of heart failure require surgical intervention to replace the heart valves.

Case Study

Example 1:
A girl born in 1997, and with the initials Č.A., comes to the Department of Paediatrics for the first time at the age of 14. She was referred from the Health Centre with Dg: Cor in obs.

Anamnesis: Denies having frequent sore throats. Present illness began about a month previously, with fatigue and exhaustion. A heart murmur, which was not previously present, is auscultated by a pediatrician.

During a clinical examination, an asthenic material (afebrile, eupnoic) was found. The pharynx was calm. In the heart: hyperdynamic ictus; clear sounds; heartbeat accelerated to 120/min; holosystolic murmur over the ictus with an intensity III/6; quiet diastolic murmur over III ICPO left; sat O2 97%; RR 110/55 mmHg. Stomach at chest level, soft; liver not palpably enlarged; no aedema; pulse a.radialis et Femoral +.

Laboratory findings: Se 17, CRP 5.6.L:6.9.E: 4.7 Hb: 130, hct 0.39 tromb.299. ASTO 1180. RF neg. Thyroid hormones normal.

RTG pulmo et cor: The heart shadow was not enlarged, with a straightened left contour of the heart.

EKG: Sinus rhythm, frequency 110/min, normogram, PR 0.18 sec, high R tines in left precordial drains, T wave positive from V2-V6.

Heart ECHO: Situs solitus, segmental structure of the heart tidy. Left cardiac chambers enlarged. LVDd 60 mm (up to 49 mm), LVSD 40 mm (up to 37 mm), S and PW 8 mm, FS 34.6%, ardila output normal. The mitral valve morphology had changed, and old post-inflammatory changes were visible, especially on the front cusp, which slightly protruded with significant mitral regurgitation v=3.98 m/sec, PG
63.5 mmHg. Tricuspid regurgitation was not significant. The aorta was trivalar with a diameter of 27mm, LA enlarged, LA/AO 1.81. **Significant aortic regurgitation** $v = 3.80$ m/sec, PG 57.7 mmHg. Pericardial separation 2 mm, myocardial contractility tidy. Septum continuous.

![Figure 3. Mitral regurgitation](image1) ![Figure 4. Aortic regurgitation](image2)

Treatment includes Extencillin for 21 days, non-steroidal and anti-rheumatic drugs, and an ACE inhibitor. After a month the non-steroidal anti-rheumatic treatment is excluded, and other therapies are included.

**Dg: Vitium cordis aorto – mitrale rheumatica.**

Extencillin and ACE inhibitor continued.

The patient now visits a cardiologist every 6 months.

She has recently begun to complain of fatigue, and her needs tend towards referral to a larger clinical centre. The patient has been examined by pediatric and adult cardiologists. In addition to an ultrasound of the heart, a Holter ECG was ordered (there were no rhythm disorders) and ergometry was undertaken, although it was subsequently discontinued due to fatigue.

**Dg:** Insuffitientio valvulae aortalis et mitralis gradus medii/gravis.

Cor compensatum.

Regular monitoring with ultrasound is recommended, every 6 months.

This patient is on the list for surgery for her heart defect, but it has not yet occurred.

**When should surgery on valves be done, and which method should be applied?**

The treatment options for valvular heart disease are well-known: medicamentosa; balloon valvuloplasty; surgical replacement therapy; and surgical valve reconstruction.
**Balloon valvuloplasty,** or percutaneous mitral commissurotomy, is a method for treating mitral stenosis without mitral regurgitation and calcification.

In cases of mitral regurgitation, **surgical correction of the mitral valve** is preferable to its replacement. Anticoagulant therapy is not required, and the mortality rate is less than 1%. Replacing the valve is necessary if the valve is thickened, or significantly deformed.

Another possibility for surgical treatment is to **install an artificial valve,** which may be mechanical, biological or homograft. After installation of the mechanical valve, the patient must receive lifelong anticoagulation treatment, with regular control of coagulation parameters to determine the optimum dose of the drug. After reconstruction of the valve or biological valve installation (i.e. bioprosthesis) such therapy is needed in the first three months after surgery, while maintaining a target value of INR 2.5.

The goals in the treatment of valvular heart disease are: a reduction or elimination of the symptoms of the disease; avoiding damage to the left ventricular function; improvement to quality of life; prevention of complications; and mortality reduction.

**Example 2:**

H. E. came to the department of Paediatrics in 2003 at the age of 7, with Dg: Tachycardia. Carditis susp.

**Anamnesis:** He had been ill for about a month, with fever, poor appetite, exhaustion, and sweating.

**Clinical examination:** well-developed for his age, subfebrile, distressed, sweaty, tachypneic.

His pharynx was calm, but breathing was weakened in the left basal lung. Heart action was rhythmic, tones quieter, no sign of a murmur, frequency 124/min, RR 121/71 mmHg. The liver was not palpably enlarged. No aedema.

**Laboratory findings:** SE 90/125, CRP 118 gr. 72%.L. 11.8. E: 3.95 Hb:196, hct 0,32. tromb.476. ASTO 1382, fibrinogen 7.6. CPK 15, CPK Mb8. Urea 3,7 GUK 6,6 Rf neg,

**ECG:** Normogram, sinus rhythm, frequency 120/min, voltage QRS of complex tidy, T flattened U II, AVF, a neg from V1 to V6.

**RTG pulmo et cor:** Significantly increased whole cardiac shadow without infiltrative opacities in the lung parenchyma.

**Heart ECHO:** Situs solitus, segmental structure of the heart tidy. Heart holes not enlarged. LVDd 38 mm, LVSD 24 mm, S 7 mm, PW 6 mm, FS 36.7%. Pulmonary and aortic flow rates tidy. Minor tricuspid regurgitation. Weaker kinetics of interventricular septum, smaller pericardial effusion.

Abdominal ultrasound tidy.
Upon receipt of results, the treatment included: penicillin, a non-steroidal anti-rheumatic, a diuretic, and corticosteroids.

The general condition of the patient is gradually improving, with less dyspnoea. Tachycardia is reduced. On the fourth day, he showed a systolic murmur and auscultated with quieter tones. Pericardial effusion is gradually reducing.
Control findings: SE 24/38, CRP 38,6 ASTO 1023, fibrinogen 2,3. L. 20,28 (corticosteroid therapy).

On the tenth day, penicillin was excluded, and Extencilin included. Corticosteroids are being reduced, as are diuretic doses.

Heart tones are clearer, their frequency decreased. In the ultrasound, pericardial effusion has retreated, mobility of interventricular septum is better, and there are no changes to the heart valves, FS 38.1%. Control RTG of lungs and heart: cardiac shadow is smaller.

Discussion

Thanks to the primary and secondary prophylaxis of streptococcal infections, rheumatic heart disease is rare today.

If a child has a mild streptococcal infection, usually inflammation of the pharynx, and it is not treated, there is a chance of 1:1000 that rheumatic fever will develop. This figure increases to 3:100 if the infection is stronger.

Keeping in mind that a very small number of children suffer from rheumatic diseases, it is believed that other factors, such as a weakened immune system, participate in the development of the disease.

Clinical picture: Clinical symptoms and signs vary depending on the organ or system affected by the disease.

Some general symptoms are typical for an inflammatory process in the organism, and these are: fever, rapid heartbeat, fatigue and general malaise, loss of appetite accompanied by weight loss, sweating, headaches, bleeding from the nose, and vomiting.

Localised rheumatic symptoms include: migratory polyarthritis, rheumatoid carditis, chorea minor, subcutaneous rheumatoid nodules, and erythema marginatum, or anulare.

Diagnosis is based on the anamnesis of the idled streptococcal infection, clinical examination, laboratory findings, raised SE and CRP values, fibrinogen, and AST0 titre.

According to the Jones criteria, if there are two major or one major and two minor diagnostic criteria present, it can be assumed with high probability that the patient has acute rheumatic fever. If these criteria are accompanied with any of the above-mentioned symptoms, it is likely that an existing streptococcal infection preceded the appearance of the clinical picture of acute rheumatic fever, but without signs of carditis.

Often the clinical picture of rheumatic diseases pass asymptomatic, without arthritis. Carditis symptoms are often mild and can be anticipated, and the child is not treated.
The first symptom of idled rheumatic diseases can be a heart murmur that had not been present earlier. At this point, changes to the valves are already permanent, and over time they will require surgery.

*When to operate on valves is a contentious question among cardiologists and cardiovascular surgeons*

Many cardiologists believe that a need to think about surgery exists at the moment when symptoms of heart failure develop. Cardiologists and cardiovascular surgeons calculate whether and when to intervene. This is crucial for a successful surgery and further RCA work. Rarely is a patient sent to surgery immediately after the discovery of a defect. Sometimes two or more years will pass until the intervention, depending on the individual case.

Another issue for cardiovascular surgeons is which surgical approach to apply. There are typically two methods to choose from. One is the so-called “plastic surgery of heart surgery”, where the valve is repaired rather than replaced, as is usually the case with mitral valves. This is the right method for a significant percentage of diseases, although all surgical interventions are very demanding. However, valve replacement surgery has the advantage of the patient not requiring long-term post-operative anticoagulant therapy. **The second option is replacement of the valve.** This is done with two types of artificial prosthesis. One type is a mechanical valve, made from titanium, which effectively lasts for a lifetime (at least 20-30 years). Another type, biological prosthesis, is made from porcine valves or bovine pericardium. These valves have the advantage of not requiring anticoagulant therapy, but there is also a drawback – they have a shorter lifetime, of only about ten years.

If the patient is treated surgically at the right time, he will have a normal life expectancy, provided he undergoes permanent anticoagulation treatment, and takes regular prothrombic time tests.

In another case, a patient is in the acute phase of heart disease, with symptoms of myocarditis and pericarditis.

Treatment can begin immediately, using antibiotics, a non-steroidal antirheumatic, and corticosteroids. Clinical symptoms of carditis will quickly retreat, and changes to the valves did not develop.

It is important to establish the aetiology of carditis. Is there arthritis? Is the disease infectious? Bacterial? Rheumatic? Idiopathic?

In long-term monitoring the prognosis is good, without congenital changes to the valves.

**Conclusion**

Although heart defects in paediatric cardiac medicine are predominantly congenital, paediatric cardiologists are also faced with cases of acquired heart disease. Almost all acquired heart disease is rheumatic. Thanks to the primary and secondary
prophylaxis of streptococcal infections, rheumatic heart diseases are becoming less frequent. The disease is often asymptomatic, and as a result is not treated. In this situation, heart disease can develop many years later.

When a heart defect forms, it becomes a question of when is the best time to perform surgery, and which method to use: valve replacement or valve correction.

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