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CLINICAL COURSE AND APPROACHES TO TREATMENT OF CONGENITAL COMPLETE ATRIOVENTRICULAR BLOCK IN CHILDREN

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ABSTRACT

A description of the clinical picture of several patients with congenital complete atrioventricular block (CCAVB) is given: isolated conduction disturbance in 1 patient and in 3 children a combination of CCAVB with congenital heart defects such as ASD, PDA. One girl with congenital heart disease (ASD) was diagnosed with Frederick's syndrome. Characteristic complaints, objective changes, as well as signs of retarded physical development were identified. All children underwent implantation of a single-chamber cardiac pacemaker. Along with pacemaker implantation, surgical correction of congenital heart disease was performed. After the treatment, there was a positive trend in the condition of the patients, normalization of physical development indicators, restoration of ECG and EchoCG parameters.

KEYWORDS

Childrenn, complete atrioventricular block, congenital heart disease, ASD, PDA.

Introduction

Conduction disorders represent a complex problem in pediatric cardiology [1-9]. A particularly severe disorder should be considered complete atrioventricular block (CAVB), which is divided into congenital, acquired and hereditary. The frequency of congenital CCAVB is, according to various authors, from 1:15,000 to 1:25,000 [1,10,11]. Its causes in 70% of cases are considered to be an autoimmune conflict, in 25% of cases - structural pathology of the heart, as well as some hereditary syndromes.

The pathogenesis of autoimmune congenital AVB is based on the passage of maternal anti-Ro/SS-A and anti-La/SS-B autoantibodies through the placenta and their damage to the cardiac conduction system [12,13]. The mother is diagnosed with diseases such as systemic lupus erythematosus (SLE), Sjogren's syndrome, and other systemic connective tissue diseases [12].

The literature also describes cases of a number of orphan syndromes, the signs of which are a combination of congenital heart disease, such as atrial septal defect (ASD), Tetralogy of Fallot, patent ductus arteriosus (PDA), transposition of the great vessels, etc., with complete AVB; there may be other anomalies development. These syndromes are associated with mutations mainly in the NKX2.5, GATA4 and TBX5j genes. The type of inheritance is presumably autosomal dominant [14,15]:
Risk factors for unfavorable outcome in such children after birth are considered [1]:
• ventricular contraction frequency (HR) less than 55 per minute;
• replacement rhythm with wide ventricular complexes;
• presence of ventricular ectopy;
• high frequency of atrial contractions (more than 140 per minute). Dyspnea and episodes of freezing may occur [13]. An extreme manifestation is considered to be a Morgagni–Adams–Stokes attack. The attack is manifested by a sudden loss of consciousness, cyanosis, areflexia, and tonic-clonic convulsions [13].
In the natural course of congenital CCAVB, 3 critical periods for the occurrence of decompensation of the disease have been established: 0-1 year, 2-4 years, 12-14 years [10], which is associated with low compensatory capabilities and a high percentage of complications during these periods of time in children.
Treatment of children with asymptomatic AVB is not required [1,12,13]. If the cause is known (autoimmune AVB, CCAVB with congenital heart disease), etiotropic, pathogenetic and symptomatic therapy of the disease is carried out. Nootropic, anti-inflammatory drugs, antioxidants, cardiotropics, drugs that have a stimulating effect and improve the conduction of nerve impulses are used [2]. Treatment tactics are determined by the characteristics of rhythm disturbances, hemodynamic disorders, the presence of syncope, and the anatomical state of the chambers and valves of the heart [1]. The absolute indications for artificial pacemaker implantation in children are [2,12]:
• any form of AV block in combination with congestive heart failure occurring in utero or after birth;
• any form of AV block in combination with congenital heart disease;
• cardiomegaly caused by myogenic dilatation of the left ventricle (LV);
• average daily ventricular rate less than 55 beats. in a minute;
• distal form of blockade (wide QRS complex more than 0.1 s);
• prolongation of the QT interval by 50 ms or more from normal values;
• frequent multiform ventricular ectopia;
• episodes of asystole for more than 2 s on a resting ECG or Holter monitoring (HM).
For children with PAVB without pacemaker implantation, the prognosis is unfavorable [2].

Purpose of the Study: to provide clinical characteristics of cases of congenital PAVB, as well as approaches to their treatment.

Material and Methods:
Patient medical histories, ECG data, echocardiography before and after surgical treatment. We observed 4 clinical cases of congenital CCAVB: boy T, 2 years 11 months with congenital CCAVB; boy S., 2 years old with a PDA, girl T., 3 years old, and boy A., 14 years old, who had an ASD in

combination with complete atrioventricular block. All patients were observed and treated in the cardiac surgery department of the Samarkand Regional Medical Medical Center.

Results

All patients had general complaints of fatigue, periodic sudden attacks of weakness, cyanosis and coldness of the extremities, sweating, retardation in physical, and in one case, psychomotor development.

The diagnosis of the defect and block in patients with CCAVB in combination with ASD was established at birth, in patients with congenital CCAVB and PDA with PAVB at one year of age. According to the anamnesis, it was not possible to identify any autoimmune diseases in mothers and family members.

Delays in physical development were observed in all patients; in addition, girl T. had multiple stigmata of dysembryogenesis and neurological disorders, in particular, at 3 years old she could not walk or talk. An objective examination revealed characteristic clinical signs of existing CHD and CCAVB. These are: pallor, poor development of subcutaneous fat, deformation of the chest, extended apical impulse, percussion expansion of the borders of the heart to the right or left, with the exception of patient T. with isolated CCAVB. Auscultation heard characteristic murmurs and bradycardia from 52 in a boy with isolated CCAVB to 33 in a girl with an ASD with pulmonary stenosis and Frederick's syndrome. On the ECG we recorded signs of complete atrioventricular block, and in patient T - Frederick's syndrome.

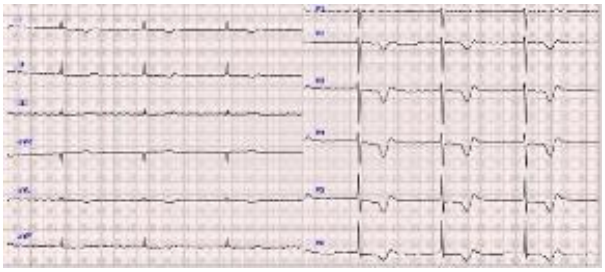

Fig 1. ECG of patient T. before surgery (from Frederica)

EchoCG revealed signs of congenital heart disease (Fig. 2A-C). In patient T., only signs of CCAVB were observed; structural changes in the heart were not detected.

Fig.2A EchoCG in a patient with PDA

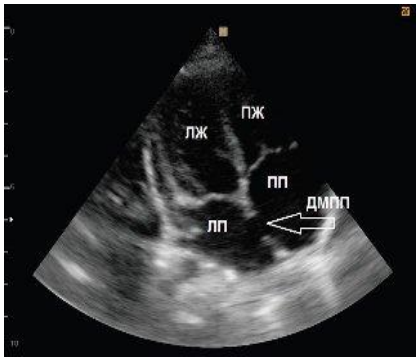

Fig. in patient with ASD with ASD, SLA

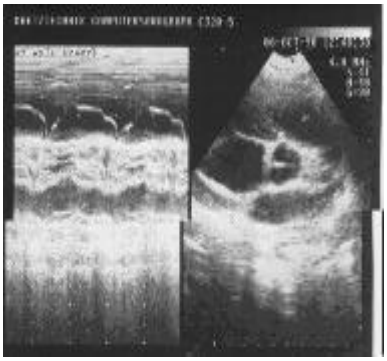

Fig. 2C in patient

Patients with ASD underwent surgical correction of the defect - suturing, and patients with PDA - ligation of the PDA. Taking into account the congenital nature of the block and the presence of clinical symptoms of CCAVB, all patients underwent pacemaker implantation. For patients with PDA, it was performed simultaneously with ligation of the duct, and for patients with ASD – on the 5th - 6th day after correction of the defect.

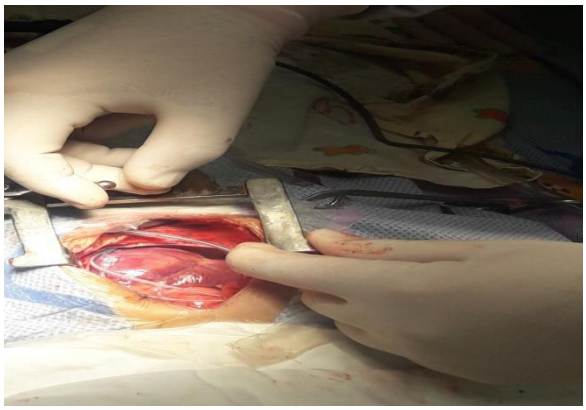

Fig.3 Implantable single-chamber pacemaker Fig.4. Implantation of pacemaker in patient A.

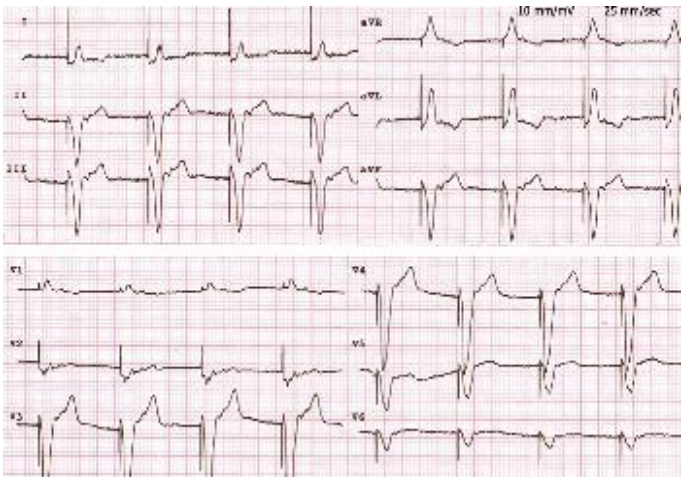

Fig.5 ECG of patient A..after pacemaker implantation

By the time of discharge, the condition of all patients was satisfactory, there was no shunt on the interatrial septum (pulmonary artery after ligation of the PDA), on the ECG: idioventricular rhythm, imposed with a certain frequency depending on age. When re-examined a year later, the indicators of the children's physical development did not differ from normal. The psychomotor development of the girl with ASD and Frederick's syndrome has noticeably improved: she began to walk and talk. EchoCG indicators are unremarkable; ECG shows that the pacemaker is functioning.

Discussion

All clinical cases had congenital AVB. Diagnosis of this condition is usually timely already in the maternity hospital, when auscultation reveals bradycardia [1,8,11]. In the 2 cases we described, cardiac

arrhythmias in patients were detected later: in 1 (in a boy with isolated CCAVB) and 2 years (in a child with CCAVB and PDA). Further, identifying the causes of congenital AVB becomes important: autoimmune or hereditary forms associated with mutations of certain genes [1,10,12,13]. Unfortunately, using the methods available to us, we were unable to determine the cause. However, according to the mother's medical history, she denies any autoimmune diseases in herself and her relatives. In all patients with a combination of congenital CCAVB and CHD, we, based on literature data [12,13], assume the presence of gene mutations. In the future, perhaps such patients will receive a more complete examination. As for physical development disorders, there is a lag, which is consistent with literature data [1,8]. Signs of cerebral palsy and retardation in psychomotor development most likely did not depend on the presence of congenital CCAVB, but were concomitant, however, after implantation of the pacemaker, the girl's neurological status improved significantly. Therefore, pacemaker implantation is necessary in the treatment of such patients.

Conclusions: Taking into account the anamnesis and clinical indicators, in the described cases of CCAVB and CHD, most likely, there was a hereditary syndrome of combination of CHD and CHD. To confirm the diagnosis, patients require further genetic testing.

Late presentation leads to a delay in the physical and psychomotor development of children.

All patients with congenital CCAVB require surgical correction of CHD and pacemaker implantation.

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