CLINICAL PICTURE, INTRACARDIAC HEMODYNAMICS AND DIAGNOSTICS OF SEPTAL HEART DISEASES IN CHILDREN AND ADULTS

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The high prevalence of congenital heart diseases in adults in the conditions of the Far North is a regional feature. In connection with the above-said, a comparative analysis of 258 adult patients and 344 pediatric patients was carried out in terms of clinical progression, intracardial hemodynamics and diagnostics of septal defects. Progression features of the given pathology in the North were found out.

Keywords: septal defects, intracardiac hemodynamics, pulmonary hypertension, bacterial endocarditis.

Purpose: To study clinical progression features, intracardiac hemodynamics and diagnostics of congenital septal heart diseases in children and adults in conditions of the Far North.

Materials and methods

Within the period of 2000-2010, total of 602 patients with septal defects had been treated in Cardiac Surgery Department of Clinical Center of Republican Hospital #1 – National Center of Medicine (CC of RH#1-NCM), including 344 (57,1%) children up to 17 y.o. Among these patients 409 (67,9%) were operated due to atrial septal defect (ASD) and 193 (32,1%) due to ventricular septal defect (VSD).

Out of 409 patients surgically operated due to ASD, 210 (51,3%) were children under 17 y.o. and 199 (48,7%) were adults over 17 y.o. Age of the patients who underwent the surgery was 4 days to 58 y.o. (see Table 1).

Table 1

Age distribution of patients surgically operated due to ASD

Children					Adults				
0-1 y.o.	2-3 y.o.	4-6 y.o.	7-10 y.o.	11-17 y.o.	18-20 y.o.	21-30 y.o.	31-40 y.o.	41-50 y.o.	>50 y.o.
39	42	58	45	26	26	61	58	37	17

The majority of patients with ASD among children were patients between 2 to 10 y.o. (145 – 69,0%), among adults – patients between 21 to 40 y.o. (119 – 59,8%). As for sex, there were more surgically operated females – total of 304 (74,3%), including 141 (67,1% among children) girls and 163 (81,9% among adults) women. Complaints of fast fatigue and shortness of breath during physical activities, pulse beat and precordialgia were registered in 112 (53,3%) children and 179 (89,9%) adults. Frequent respiratory diseases, pneumonia were noted in 18 (8,6%) children and 25 (12,6%) adult patients; pulmonary hypertension of the II hemodynamic group in children was diagnosed in 15 (7,1%) cases; pulmonary hypertension of the II and IIIa hemodynamic group in adults was diagnosed in 24 (12,0%) cases.

Adult patients with ASD had comorbid conditions more often – in total of 121 (60,8%) cases: chronic illnesses of lungs and bronchial tubes – 78 (39,2%), chronic pyelonephritis – 39 (19,6%), chronic hepatitis – 35 (17,6%), chronic gynecopathies – 24 (14,7% - among women), cholelithiasis – 18 (9,0%), arterial hypertension – 17 (8,5%), diabetes – 9 (4,5%). Children had suffered comorbid conditions less often – in total of 49 (23,3%) cases: chronic genyantritis and

sinusitis – 28 (13,3%), chronic tonsillitis – 23 (10,9%), chronic bronchopulmonary diseases – 15 (7,1%), neuropathies – 7 (3,3%), chronic pyelonephritis – 5 (2,4%). Clinic picture of chronic bacterial endocarditis was observed in 23 (11,5%) adult patients and 12 (5,7%) children.

During auscultation, all patients with ASD had systolic murmur of moderate intensity in 2-3 intercostal space to the left of the breastbone; heart sounds were clear. Sinus rhythm was registered on the electrocardiogram (ECG) of all pediatric patients; incomplete blockage of right crus of atrioventricular bundle was registered in 152 (72,4%) children; atrioventricular blockage of I degree was registered in 18 (8,6%) pediatric patients. Deviation of heart's electrical axis to the right with signs of hypertrophy of the right ventricle was registered in 186 (88,6%) children; vertical position of heart's electrical axis was registered in 24 (11,4%) children. Sinus rhythm was registered in 184 (92,5%) adults; atrial fibrillation of normal systolic character was diagnosed in 3 (1,5%) patients; episodes of auricular flutter were noted in 7 (3,5%) cases; atrial fibrillation strokes – in 6 (3,0%) patients; sinus arrhythmia with frequent premature atrial contraction – in 8 (4,0%) cases; sinus tachycardia strokes – in 5 (2,5%) adult patients. Among adults, incomplete blockage of right crus of atrioventricular bundle was registered in 156 (78,4%) cases; atrioventricular blockage of I degree was registered in 25 (12,6%) examined patients. Hypertrophy of the right ventricle (RV) was observed in all adult patients; combination with signs of hypertrophy of the left ventricle (LV) was registered in 7 (3,5%) cases. There were 187 (94,0%) adult patients with the deviation of heart's electrical axis to the right; the rest 12 (6,0%) patients had vertical position of heart's electrical axis.

Majority of patients with ASD had intensification of vascular pattern of lungs on X-Rays. Signs of hypervolemia of pulmonary circulation with prominence of the pulmonary artery (PA) and with distension of trunk of PA were found in 15 (7,1%) pediatric patients and 26 (13,1%) adult patients; moderate hypervolemia was noted in 84 (40,0%) children and 86 (43,2%) adults. On the average, Moore's index was 32,9 (P – 3,8)% in 172 (81,9%) children and 36,4 (P – 3,6)% in 183 (92,0%) adults. The rest 38 (18,1%) children and 16 (8,0%) adults had normal value of Moore's index. On the average, cardiothoracic ratio was 0,55 (P – 0,05) in 176 (83,8%) children and 0,56 (P – 0,06) in 185 (93,0%) adults; the rest 34 (16,2%) children and 14 (7,0%) adults had normal value of this index.

Our research has shown that the increase of heart volume in patients with ASD generally occurs at the expense of hypertrophy of the right chambers of heart, and these data coincide with the published data. The 1st degree of RV hypertrophy before surgery was found in 70 (33,3%) pediatric patients, and 65 (32,7%) in adults; the 2nd degree of RV hypertrophy was diagnosed in 82 (39,0%) children and 76 (38,2%) adult patients; the 3rd degree of RV hypertrophy was registered in 28 (13,4%) children and 46 (23,1%) adult patients. The rest 30 (14,3%) children and 12 (6,0%) adults had normal size of RV.

Size and position of ASD, intracardiac hemodynamics were identified by echocardiography (ECHO) and catheterization of heart cavities, angiocardiography. ECHO was carried out in all patients, the catheterization of heart cavities was performed on 57 (27,1%) children and 72 (36,2%) adults with ASD diagnoses. According to results of this research, secondary defects were found in 207 (98,6%) children and 197 (99,0%) adults; all patients had shunt through ASD from the left atrium (LA) into the right atrium (RA). According to angiocardiography before surgical treatment, pulmonary hypertension (PH) of II hemodynamic group was diagnosed in 16 (7,6%) pediatric and 27 (13,6%) adult patients. In all patients tests for oxygen saturation in RA cavity were higher than tests taken in venae cavae; the relation of systolic pressure in PA to system arterial pressure (SAP) fluctuated from 32,0 to 72,0% (45,8±8,4% on the average); systolic pressure in PA fluctuated from 30,0 to 78 mmHg (52,1±9,8 mmHg on the average); the relation of total pulmonary resistance to total peripheral resistance fluctuated from 8 to 32% (21,2±6,7% on the average); shunt from the left to the right in relation to minute volume of pulmonary circulation (PC) fluctuated from 38,0 to 85,0% (62,1±12,8% on the average).

Out of 193 patients surgically treated concerning VSD, there were 134 (69,4%) children and 59 (30,6%) adults. Among children 71 (53,0%) were girls and 63 (47,0%) were boys; among adult patients there were 17 (28,8%) women and 42 (71,2%) men. Age of the patients ranged from 7 days to 55 years (see Table 2).

Table 2

Age distribution of patients surgically operated due to VSD

Children					Adults				
0-1 y.o.	2-3 y.o.	4-6 y.o.	7-10 y.o.	11-17 y.o.	18-20 y.o.	21-30 y.o.	31-40 y.o.	41-50 y.o.	>50 y.o.
22	29	35	29	19	11	23	16	7	2

In children's group 32 (23,9%) patients did not have any complaints; the rest 102 (76,1%) complained about shortness of breath and fast fatigue during physical activity. It should be noted that children of younger age cannot identify pulse beat and precordialgia, nevertheless, it was possible to diagnose (besides shortness of breath and fast fatigue) precordialgia in 35 (26,1%) and complaints about unpleasant feelings in chest in 42 (31,3%) pediatric patients. All patients of adult group with VSD made complaints about fast fatigue and shortness of breath during physical activity; also 19 (32,2%) of them complained about periodic precordialgia. 12 (20,3%) adult patients complained about heart malfunctions. Frequent respiratory diseases were noted in 39 (29,1%) children; frequent respiratory diseases and pneumonia were found in 23 (38,9%) adult patients. Pulmonary hypertension of II hemodynamic group was registered in 21 (15,7%) children with VSD; pulmonary hypertension of IIIa hemodynamic group – in 15 (11,2%) patients. Among adults, pulmonary hypertension of II hemodynamic group was found in 12 (20,3%) patients with VSD; pulmonary hypertension of IIIa hemodynamic group – in 9 (15,3%) patients. Among patients with VSD, clinical picture of bacterial endocarditis was observed in 9 (6,7%) pediatric patients and 8 (13,5%) adult patients.

Adult patients with VSD had comorbid conditions more often – in total of 34 (57,6%) cases: chronic illnesses of lungs and bronchial tubes – 22 (37,3%), chronic pyelonephritis – 8 (13,6%), chronic hepatitis – 7 (11,9%), chronic gynecopathies – 3 (17,6% - among women), cholelithiasis – 6 (10,2%), arterial hypertension – 5 (8,5%), diabetes – 3 (5,1%). Pediatric patients suffered comorbid conditions less often – in total of 29 (21,6%) cases: chronic genyantritis and sinusitis – 18 (13,4%), chronic tonsillitis – 15 (11,2%), chronic bronchopulmonary diseases – 7 (5,2%), neuropathies – 5 (3,7%), chronic pyelonephritis – 4 (3,0%). In addition, it should be noted that among adults with VSD 2 (3,4%) had patent foramen ovale; 3 (5,1%) patients were diagnosed with ASD. Comorbid patent ductus arteriosus (PDA) was found in 5 (8,4%) adult patients. Among children with VSD, patent foramen ovale was diagnosed as comorbid CHD in 17 (12,7%) cases; PDA was diagnosed in 8 (6,0%) cases.

During auscultation, all patients with VSD had strong systolic murmur of various intensity to the left of the breastbone; splitting of the 2nd tone was found more often in adult patients (23 (39,0%)) than in children (35 (26,1%)).

According to ECG, sinus rhythm was registered in all pediatric patients with VSD; heart rate was from 82 to 132 beats per minute (99±12,0 on the average); incomplete blockage of right crus of atrioventricular bundle was registered in 21 (15,7%), atrioventricular blockage of I degree was registered in 15 (11,2%) children. Sinus rhythm was registered in 57 (96,6%) adult patients' ECG; heart rate was from 58 to 88 beats per minute (72,0±7,0 on the average). According to ECG, hypertrophy of LV and LA was diagnosed in 48 (35,8%) children and 12 (20,3%) adult patients; signs of hypertrophy of RV and RA were diagnosed in 58 (43,3%) children and 37 (62,7%) adults.

There was no deviation of heart's electrical axis found in 15 (11,2%) pediatric patients; 39 (29,1%) children had heart's electrical axis dislocated to the left; vertical position of heart's electrical axis was registered in 26 (19,4%) children; and 54 (40,3%) pediatric patients had heart's electrical axis dislocated to the right. Among adult patients with the same diagnosis, 4 (6,8%) were diagnosed with normal position of heart's electrical axis; 9 (15,3%) had horizontal

position of heart's electrical axis; and 28 (47,5%) had deviation of heart's electrical axis to the right.

All patients with VSD had intensification of vascular pattern of lungs on X-Rays. Nipple of pulmonary artery (PA) and distension of trunk and branches of PA were found in 37 (27,6%) pediatric patients and 21 (35,6%) adult patients. Enlargement of heart size due to hypertrophy of the left chambers of heart was noted in 47 (35,1%) children and 25 (42,4%) adults; enlargement of heart size due to hypertrophy of the right chambers of heart was found in 52 (38,8%) children and 31 (52,5%) adults.

According to ECHO, among pediatric patients, perimembranous VSD was diagnosed in 102 (76,1%), subarterial VSD – in 7 (5,2%), and intramuscular VSD – in 25 (18,7%) cases. Among adult patients, perimembranous VSD was found in 48 (81,3%), subarterial VSD – in 5 (8,5%), and intramuscular VSD – in 6 (10,2%) cases.

Catheterization of heart cavities and angiocardiography was performed on 42 (31,3%) children and 21 (35,6%) adults with VSD. Furthermore, all patients of both groups had increased saturation of blood by oxygen in RV in comparison with blood tests in RA. Pulmonary hypertension (PH) of II hemodynamic group was diagnosed in 19 (14,2%) children and 12 (20,3%) adults; PH of IIIa hemodynamic group – in 15 (11,2%) children and 11 (18,6%) adults. Classification of pulmonary hypertension by V.I. Burakovsky with coauthors (1975) was used for identification of degree of PH. In children with PH of II hemodynamic group systolic pressure in PA fluctuated from 38,0 to 78 mmHg (58,2±12,4 mmHg on the average); in adults with the same type of PH the pressure in PA fluctuated from 45,0 to 80,0 mmHg (60,2±12,3 mmHg on the average). The relation of total pulmonary resistance to total peripheral resistance in these patients of pediatric and adult groups averaged 18,9±3,7% and 19,9±3,8% respectively. In children of this group, the relation of systolic pressure in PA to system arterial pressure fluctuated from 33,9 to 62,0% (48,2±9,9% on the average); in adults this indicator fluctuated from 37,5 to 61,0% (49,1±9,6% on the average). In pediatric patients with VSD and PH of IIIa hemodynamic group, systolic pressure in the trunk of PA fluctuated from 62,0 to 94,0 mmHg (78±10,8 mmHg on the average); in adults with VSD and PH of the same hemodynamic group, this indicator fluctuated from 64,0 to 97 mmHg (80,7±9,3 mmHg on the average). The relation of systolic pressure in PA to system arterial pressure fluctuated from 66,0 to 100,0% (83,1±12,1% on the average) in children; this indicator fluctuated from 64,0 to 100,0% (82,4±12,3% on the average) in adults.

Discussion

Thus, the results of our research and observation allow us to note a number of features of clinical progression and intracardiac hemodynamics in pediatric and adult patients with septal defects:

1. Patients attaining adult age without surgery concerning correction of septal defects have comorbid conditions of adult age more often: chronic illnesses of lungs and bronchial tubes – 78 (39,2%) and 22 (37,3%), chronic pyelonephritis – 39 (19,6%) and 8 (13,6%), chronic hepatitis – 35 (17,6%) and 7 (11,9%), chronic gynecopathies – 24 (14,7%) and 3 (17,6% among women), cholelithiasis – 18 (9,0%) and 6 (10,2%), arterial hypertension – 17 (8,5%) and 5 (8,5%), diabetes – 9 (4,5%) and 3 (5,1%) patients with ASD and VSD respectively. Children less often have comorbid conditions (29 - 21,6% cases): chronic genyantritis and sinusitis – 18 (13,4%), chronic tonsillitis – 15 (11,2%), chronic bronchopulmonary diseases – 7 (5,2%), neuropathies – 5 (3,7%), chronic pyelonephritis – 4 (3,0%). Our observations correspond with the data received by L. Kidd et al. (1993), U. Neumayer et al., (1998) and B.D. Amirkulov (2004).

Furthermore, it is necessary to pay attention to the fact that, according to the majority of researchers [1,2,3,4,5,6,10,11,12], there are no data on occurrence of bacterial endocarditis. As a result of our research, the clinical picture of chronic bacterial endocarditis was observed in 23 (11,5%) adult patients and 12 (5,7%) pediatric patients with ASD. Children with VSD had the clinical picture of bacterial endocarditis in 9 (6,7%) cases; adults - in 8 (13,5%) cases. Our data

of frequency of infectious endocarditis development in patients with VSD exceed the results of research of the majority of authors [11,12].

2. According to the methods of functional research, it has been found that signs of hypertrophy of RA and RV were more evident in adult patients with ASD than in pediatric patients (100% and 3,5% in adult patients, 88,6% and 0,0% in children respectively). Our results coincide with the data of many authors [1,2,3,5,6,7,8].

Among patients with VSD signs of hypertrophy of the right and left heart chambers were more often observed in adults. According to ECG, hypertrophy of LV and LA was found in 48 (35,8%) children and 12 (20,3%) adults; signs of hypertrophy of RV and RA were diagnosed in 58 (43,3%) children and 37 (62,7%) adults.

3. Adult patients with ASD more often have heart rhythm disorders: sinus rhythm was registered in 184 (92,5%), atrial fibrillation of normal systolic character was diagnosed in 3 (1,5%), episodes of auricular flutter were noted in 7 (3,5%), atrial fibrillation strokes – in 6 (3,0%), sinus arrhythmia with frequent premature atrial contraction – in 8 (4,0%), sinus tachycardia strokes – in 5 (2,5%) patients of adult group. All pediatric patients had sinus rhythm. Results of our research confirm the published data [5,6,7,8,9].

4. Our data have shown more frequent increase in pulmonary blood stream in patients with ASD (7,1% of children and 13,1% of adults), more frequent increase in Moore's index (81,9% of children and 92,0% of adults), and increase in volume of the right chambers of heart (increase of the III degree in 13,4% of children and 23,1% of adults) in patients of adult group. These data are confirmed by the data of the majority of authors [5,6,7,8,9].

5. The observation data of intracardiac hemodynamics in patients with septal defects have revealed the development of pulmonary hypertension which is more frequently found in adult patients: pulmonary hypertension of II hemodynamic group in children was found in 15 (7,1%) cases, while pulmonary hypertension of II and IIIa hemodynamic group in adults was found in 24 (12,0%) cases among patients with ASD. Among patients with VSD, we diagnosed PH of II hemodynamic group in 19 (14,2%) children and 12 (20,3%) adults; PH of IIIa hemodynamic group – in 15 (11,2%) children and 11 (18,6%) adults.

Conclusion

The clinical progression and clinical picture of septal defects have the following specific characteristics:

a) frequent comorbid condition;

b) cardiac rhythm disturbance;

c) more frequent cases of pulmonary hypertension of II and IIIa hemodynamic groups;

d) most frequent cases of earlier or current bacterial endocarditis.

A frequent comorbid condition and bacterial endocarditis are characteristic to septal defect progression in conditions of the Far North and are caused by remoteness of settlements which are sparsely populated and dispersed on a vast territory with lack of medical care and transportation infrastructure in extreme geo-climatic conditions of the North.

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