Features of the course of comorbid pathology with congenital septal heart defects

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Abstract: Congenital heart defects are an important problem in pediatrics due to their high prevalence and the need for early surgical correction due to significant health problems and disability in children. According to WHO, congenital heart disease occurs in 0.7-1.7% of newborns. In some cases, the rapid progression of the stages of the course of congenital heart disease is determined not only by its severity, but also by the influence of concomitant diseases: hypoxic-ischemic encephalopathy, nutritional deficiencies, deficiency anemia, etc. Knowledge of the mechanisms of development of these disorders and the possibility of their timely detection with the help of modern research methods make it possible to diagnose and correct developing pathological conditions as early as possible. In this regard, the issue of studying clinical diagnostic criteria for improving the provision of timely medical care to children with congenital heart disease of an early age remains relevant and requires scientific research in this direction.

Keywords: congenital heart disease, comorbid conditions, developmental anomaly, extracardiac pathology

Currently, diagnostics of associated extracardiac congenital defects and extracardiac pathology is important for clinical practice in determining the timing and stages of surgery on the heart or other organs, preventing complications, and predicting the patient's clinical condition in follow-up.

In cardiac surgery, the presence of comorbid pathology has a significant impact on the course of the postoperative period, the development of complications, and also affects the prognosis. Current standards and approaches in cardiac surgery should include an analysis of comorbid diseases when choosing methods of anesthesia, intensive care, and tactics of surgical intervention.

Comorbidity is the presence of another disease or medical condition simultaneously with the current disease.

As is known, the presence of comorbid diseases contributes to an increase in hospital bed-days, leads to the development of disability, hinders rehabilitation, and

increases the number of complications after surgical interventions. This problem has attracted the attention of specialists working with children in the first years of life. In particular, the scientific literature contains works devoted to comorbid pathology in young children with septal congenital heart defects.

One of the domestic studies noted that hospital mortality among cardiac surgery patients with one or more comorbid diseases was 3.3%, and among patients without comorbidity - 0.4%.

Modern medical capabilities and a multidisciplinary approach have shown that the prevalence of extracardiac pathology in children with cardiac surgery pathology is 12-30% of cases, in adults - 4.5-60%.

In the works of J.H. Gonzalez et al. It was described that 7–50% of patients with CHD had extracardiac malformations (EM) with a high risk of comorbidity and mortality, with a high rate of complications after surgery.

H.H. Kramer et al. showed that in 13.3% of cases, patients with CHD had hereditary and teratogenic syndromes, in 7.7% of cases - congenital defects of the kidneys, lungs, musculoskeletal and central nervous systems. Moreover, the anomaly of the upper urinary tract was accidentally detected during angiocardiography and in 99.5% of cases, patients did not have clinical manifestations of urinary tract pathology. The most common anomalies of the urinary tract were: complete duplication of the kidneys and ureter or hydronephrosis. There was no correlation in the frequency of development of urinary tract defects with any specific heart defect. In similar studies, congenital malformations of the kidneys and ureters were diagnosed in 11.9–25.8% of children without clinical urinary symptoms, genetic syndromes were encountered in 38%; Congenital malformations of the digestive system - in 19.6%. Italian researchers found concomitant congenital malformations in 26% of cases in children with CHD. A correlation was noted between the frequency of interventricular defects, atrial septal defects and complex heart defects with other malformations. The most common EM were congenital malformations of the musculoskeletal system (25.3%), congenital malformations of the genitourinary (22.9%) and gastrointestinal (11.5%) systems. Karyotyping was performed in 19.4% of cases, and chromosomopathy was detected in 152 patients.

In 2014, L. Eskedal et al., based on a large sample of over 3,000 children with CHD, noted that 20% of children have concomitant CHD of the gastrointestinal tract (GIT) (intestinal anomalies and esophageal atresia) [11]. Similar work by foreign authors showed that 12% of children with CHD have combinations of CHD of the GIT and urinary system (US), chromosomal diseases in 4.9% of cases, and a correlation between the severity of CHD and a combination with other CHD. Works have been described where other CHDs were combined with CHD much more often (45.9%). Of these, the leading ones were facial anomalies (15%), genitourinary system (12%), and

CHD of the GIT (11.1%). Interventricular and interatrial septal defects, coarctation of the aorta, single ventricle, pulmonary artery stenosis, hypoplastic right heart syndrome, double outlet of the right ventricle, common atrioventricular communication, and aortic anomalies were often accompanied by EM (more than 50%). It was shown that spleen anomalies were more often combined with a single ventricle ($p < 0.002$). In 2015, M. Wojtalik et al. retrospectively studied 1856 pediatric cardiac surgery patients. Concomitant pathology was detected in 84 (4.5%) children. The structure of frequent anomalies included: anomalies of the digestive (35.7%) and urinary systems (22.4%). The authors did not find any correlation between CHD and concomitant congenital malformations. Multivariate logistic regression analysis revealed that comorbidity has a significant impact on the timing of surgery, course and mortality rate in children with CHD. The mortality rate in children with CHD without EM was 8.9%, the mortality rate in children with EM was 19% of cases. Of these, up to 50% of cases are neonatal patients. EM were more often observed in children with ventricular septal defects (7.6%, $p = 0.0012$).

In a prospective study by A. Meberg et al. among 662 patients with CHD, 22% of children were found to have concomitant congenital malformations. Atrioventricular septal deficiency, ASD, tetralogy of Fallot and single ventricle were the most frequently associated with EM. Concomitant congenital malformations were found in 31% of patients with VSD. The mortality rate was significantly higher among patients with CHD and comorbid disorders (29%) compared to patients with isolated heart defects (6%).

In Russian medicine, there is a research paper stating that in the structure of congenital malformations in stillborn and deceased children in the neonatal period, 1/3 were multiple malformations (28.6%) and that in the structure of multiple congenital malformations, more than 50% were cases of combination with CHD.

In the study by A. Amorim et al. Extracardiac anomalies were found in 31.4% of newborns and 48% of stillborns. EM were part of an unclassifiable syndrome in 23.1% of newborns and 32% of stillborns. Frequently encountered in the group of newborns were anomalies of the genitourinary system in boys (48.3%); in the group of stillborns, CHD was often combined with anomalies of the kidneys and ureter and defects of the musculoskeletal system (52.8%).

In 2017, researchers Gonzalez J.H. et al. conducted a retrospective analysis of medical records of 223 newborns with prenatal diagnosis of CHD. According to the analysis, prenatal abdominal ultrasound (US) was performed only in 58.7% of cases and associated anomalies were detected in 41.2% of cases. Among the detected CHDs, 36.6% of cases included clinically significant renal malformations or heterotaxic anomalies. Patients with cardiac septal defects were 3.7 times more likely to have abnormal findings on abdominal ultrasound than those without CHD. Almost 50% of

patients had one or more extracardiac or genetic disorders detected during ultrasound diagnostics, neurosonography or karyotyping. Based on the calculation of financial costs for additional examination of children with concomitant pathology, the authors propose a strategy for diagnosing EM in neonatal cardiac surgery patients, including three screenings: ultrasound of the brain, abdominal cavity, and karyotyping. In addition to cardiac surgery, newborns with CHD required additional surgical procedures or intensive care to correct defects in other organs and systems. In 2018, K.A. Tokmakova showed in her work that extracardiac pathology occurs in children with a frequency of 14.8% of cases. The condition of children with syndromic pathology is associated not only with the severity of heart damage, but also with various hereditary diseases that can lead to dysfunction of other organs and systems, thereby complicating the therapeutic and surgical treatment of the heart defect itself.

In the study by Yu. V. Petrenko and co-authors, the following were found in children with CHD: diaphragmatic hernia in 5–18% of cases, esophageal atresia in 15–39%, renal agenesis in 17–43%, anorectal anomalies in 22%, and central nervous system pathology in 15% of cases. The work noted a direct correlation between diaphragmatic hernia and hypoplastic left heart syndrome with VSD and coarctation of the aorta, and with atresia of the anus - a connection between such CHDs as tetralogy of Fallot and VSD [5]. In 2015, D. Dilber and I. Malcić retrospectively analyzed the charts of 1,480 newborns with CHD. The authors found that the most common anomalies were gastrointestinal tract anomalies (8.4%), chromosomal defects and syndromes, and multiple congenital malformations were detected in a total of 14.5% of patients.

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