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Original article

Adult congenital heart diseases - 10-years study in patients treated at the Institute for Cardiovascular Diseases of Vojvodina

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Abstract

Introduction. The number of adult patients with a congenital heart defect (adult congenital heart disease - ACHD) is higher than children born with the defect. The study aimed to examine the characteristics of patients with ACHD treated over a 10-year period at the Institute for Cardiovascular Diseases of Vojvodina (ICVDV).

Methods. A retrospective cohort study included patients examined and treated under the diagnosis of congenital heart defect at IKVBV from 2011 to 2021. Patients with ACHD were identified by searching the electronic database based on the ICD classification. In the examined group, we evaluated demographic characteristics, the presence of comorbidities, and complications that developed during follow-up in a predefined time period.

Results. Out of 106123 patients treated during the 10-year period, 0.8% were diagnosed with ACHD. The most frequently registered ACHD were atrial septal defect (ASD) with 49.1%, ventricular septal defect with 13.1%, congenital anomalies of the aortic valve 10%, and coarctation with aorta narrowing 6.2%. In comparison, complex defects like tetralogy of Fallot with 2.7% and transpositions of great arteries (1.2%) had lower prevalence. The most prevalent complications were heart rhythm disturbances in 44% and pulmonary hypertension in 15.2% of ACHD patients. The most common comorbidities were arterial hypertension (46.7%) and ischemic heart disease (20.5%).

Conclusion. The prevalence of ACHD was 0.8%. The most prevalent defects were from the group of septal defects. The most prevalent complications were heart rhythm disorders and pulmonary hypertension.

Keywords adult congenital heart diseases, complications, comorbidities

Introduction

Congenital heart defects (CHDs) represent the most common anomalies, occurring in 0.8% to 1% of cases. Advances in diagnostics and therapeutics enable timely detection, often as early as the prenatal period, offering the potential for prompt intervention. Despite a trend toward primary surgical correction, in some complex cases, only palliative surgical procedures may be feasible.[1,2] Conversely, percutaneous techniques for addressing CHDs are widely applied when applicable, such as using occluders for atrial septal defects (ASDs). With improved treatment, a growing number of these patients reach adulthood (more than 90%), and it is now acknowledged that the number of adults with CHDs far exceeds the number of newborns with these defects. According to the 2020 recommendations of the European Society of Cardiology, the term "adults with CHD" has been replaced with "Adult Congenital Heart Disease (ACHD)".[2]

It is estimated that there are over 1.2 million ACHD patients in Europe[3] and over 1.6 million in the United States.[4] Many ACHD patients have experienced frequent hospitalizations, repeated surgeries and interventions, lifelong medication regimens, physical intolerance, reduced quality of life, and a shortened lifespan since a young age.[2] Additionally, in adulthood, these patients face a higher incidence of complications related to CHDs. They are at greater risk of arrhythmias and sudden cardiac death, with a threefold higher prevalence of atrial arrhythmias (AA) compared to the general population.[5] Infective endocarditis (IE) is also more common in ACHD patients, with a 30-140 times greater incidence than in the general population.[6]

Heart failure (HF) develops in 20-50% of ACHD patients and is the leading cause of death.[2] The true incidence may be even higher, as signs and symptoms of HF are often subtle. The pathophysiology of cardiorespiratory dysfunction is complex, and it involves elements seen in acquired HF, with the primary pathophysiological mech-

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anism being chronic pressure and/or volume overload.[7] Myocardial lesions (such as those resulting from bypass surgery, ventriculotomy, chronic hypoxia, etc.) also play a role in ADCH HF. These patients may also develop coronary artery disease (CAD) associated with aging or congenital coronary anomalies. They are also prone to developing other acquired heart diseases during their lifetime, including myocarditis, rheumatic or non-rheumatic valvopathies, and persistent tachyarrhythmias.

Among complications related to CHDs, pulmonary hypertension (PH), especially post-capillary PH, is common. In severe and inadequately treated cases of left-to-right shunt defects, pre-capillary PH (PAH) and pulmonary vascular disease (Eisenmenger syndrome) can develop. The development of aortic aneurysms and/or dissections is more often associated with arthropathies or post-interventional (formation of a "neo-aorta").[2] Patients with cyanotic CHD experience symptoms of hyperviscosity and numerous complications specific to different types of cyanotic and non-cyanotic CHDs.

Intensive monitoring and regular follow-up visits for all ACHD patients are needed to prevent complications and deterioration and to determine the appropriate timing for possible catheter-based or surgical procedures to improve long-term prognosis. Echocardiography plays a crucial role, along with magnetic resonance imaging (MRI) and/or computed tomography (CT) of the heart.[8,9]

In addition to complications stemming from the underlying disease and prolonged lifespan, ACHD patients are at greater risk of developing comorbidities such as hypertension, diabetes mellitus, hyperlipidemia, obesity, and atherosclerotic disease. Paradoxically, some studies report a higher prevalence of obesity and metabolic syndrome in this population.[10]

Beyond traditional risk factors for ischemic heart disease, ACHD patients are subject to other contributing factors for premature coronary disease, including obstructive left heart lesions, aortic stenosis, reperfusion injuries during surgery, turbulent blood flow across residual defects, coronary anomalies, and more.[2]

The incidence of cerebrovascular events (CVEs) is higher in ACHD patients compared to the general population, primarily attributed to the presence of interatrial shunts, leads, artificial materials in the heart, arrhythmias, and HF.(11) These patients also have an increased likelihood of developing chronic kidney disease (CKD) due to various factors, including blood hyperviscosity, alterations in renal blood flow, neurohormonal activation, and the impact of postoperative intensive care.[12]

The aim of our study was to investigate the characteristics of the ACHD patient population treated at the Institute for Cardiovascular Diseases of Vojvodina (ICVDV) and to explore the presence of specific complications and comorbidities.

Methods

We conducted a retrospective cohort study. The study included patients with a congenital heart defect selected by diagnosis code for specific CHDs based on the International Classification of Diseases, 10th revision (ICD-10), and through notes associated with the given diagnosis containing words related to one of the CHDs. The following diagnosis codes for CHDs were specified: Q20.0-Q20.9; Q21.0-Q21.9; Q22.0-Q22.9; Q23.0-Q23.9; Q24.0-Q24.9, except Q24.6; Q25.0-Q25.9; Q26.0-Q26.9, except Q26.5 and Q26.6) and the codes for Marfan syndrome (Q87.4) and Turner syndrome (Q96), which are considered aortopathies, among others. For the search within diagnosis notes, the following words and abbreviations were provided: ASD, VSD, PDA, Eisenmenger, congenital, bicuspid valve, univalvular aortic valve, BAV, Fontan, univentricular heart, UVH, coronary anomalies, ALCAPA, ARCAPA, AAOCA, and coronary fistulas. Patients treated with the diagnoses above were categorized into the study group of ACHD patients. Exclusion criteria included patients diagnosed with persistent foramen ovale and mitral valve prolapse. A total of 926 patients meeting the inclusion criteria were identified. However, due to incomplete data in 90 patients, not all parameters could be analyzed. Demographic parameters were analyzed, along with complications and comorbidities that developed within a predefined time frame. Interventional procedures such as percutaneous coronary intervention (PCI) and the percutaneous placement of ASD occluders, pacemaker implantation, or implantable cardioverter-defibrillators (ICD) were also analyzed, as well as cardiothoracic surgical interventions.

The parameters were extracted from the patient's electronic records for predefined time frames. The data were finally checked by one investigator for missing or contradictory entries and for values beyond the normal range. The study was conducted according to the principles of the Declaration of Helsinki. The ethics committee approved the study of the ICVDV.

Statistical analysis

Data were statistically processed using the Statistical Package for Social Sciences (SPSS) version 21. Standard descriptive statistical methods were employed, continuous variables were expressed as means and standard deviations, and categorical variables were expressed as absolute numbers and percentages. Values with a significance level of $p < 0.05$ were considered statistically significant.

Results

Over the predefined 10-year period, a total of 106123 patients were examined and treated at the Institute for Cardiovascular Diseases of Vojvodina (ICDV). The prevalence of Adult Congenital Heart Disease (ACHD) was 0.8%, with an average patient age of 47.9 ± 36.4 years. Among the ACHD patients, 55% were females.

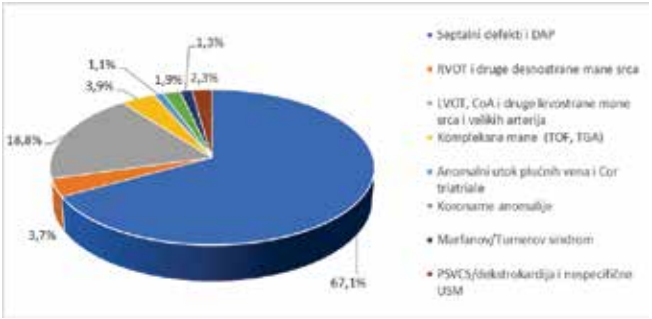
The atrial septal defect had the highest prevalence of 49.1%, VSD 13.1%, congenital aortic valve anomalies 10%, followed by coarctation of the aorta and other aortic stenoses 6.2%, while complex defects such as Tetralogy of Fallot (TOF) accounted for 2.7% and complete transposition of the great arteries (TGA) for 1.2% of cases (Table 1). The most prevalent ACHDs were within

Table 1. Different ACHDs with mean age±SD

Different ACHDs	N	%	Age	SD
ASD	455	49.1	52.6	16.98
VSD	121	13.1	40.6	17.82
Aortic valve defects	93	10	38.6	13.46
Coarctation and other aorta narrowings	57	6.2	45.6	18.16
Pulmonary valve stenosis	25	2.7	42.3	18.96
T. Fallot	25	2.7	38.6	15.11
Persistent arterial duct	23	2.5	44.9	19.57
Aortic stenosis	20	2.2	47.2	25.40
Coronary anomalies	18	1.9	50.6	19.73
AV canal	12	1.3	41.3	19.92
Marfan/Turner Sy	12	1.3	38.6	11.89
Dextrocardia	11	1.2	60.3	15.54
Transposition	11	1.2	32.0	15.46
Other congenital septal defects	9	1	45.2	12.02
Cor triatriale	6	0.6	58.0	18.85
Ebstein's anomaly	6	0.6	41.0	19.60
Unspecific heart defects	5	0.5	30.3	12.04
Persistent left superior vena cava	5	0.5	.	.
Anomalous pulmonary vein drainage	4	0.4	36.5	.71
Other aortic and mitral valve defects	4	0.4	48.5	12.02
TV/right heart malformation	3	0.3	24.3	6.66
Aortopulmonary window	1	0.1	21.0	.

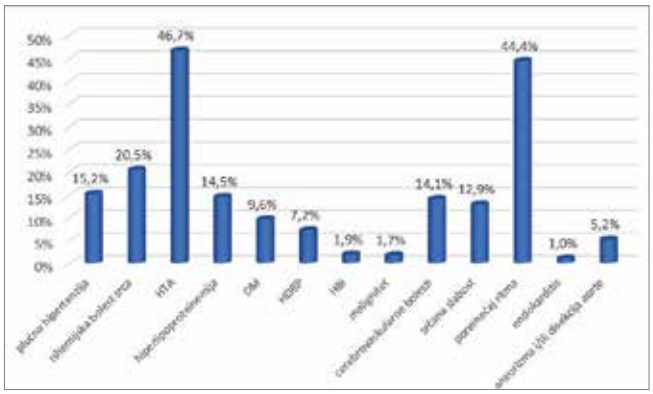
AV, atrioventricular; TV, tricuspid valve; N, number

Figure 1. Different comorbidities/complications in ACHD population



Legend: HTA, arterial hypertension; DM, diabetes mellitus; COPD, chronic obstructive pulmonary disease; CKD, chronic kidney disease; HF, heart failure; CVD, cerebrovascular disease; CAD, coronary artery disease; HLP, hyperlipoproteinemia; IE, infective endocarditis

Figure 2. Prevalence of complications and co-morbidities in ACHD



the septal defect group, including persistent arterial duct (PDA), which is an example of simple CHDs with left-to-right shunt. Next were anomalies associated with the left ventricular outflow tract (LVOT), including valvular, subvalvular, and supravalvular aortic stenoses, along with coarctation of the aorta (CoA) and malformations designated as other aortic abnormalities and left-sided heart defects.

Arrhythmia was the most common complication (44%) followed by pulmonary hypertension (PH) 15.2%, and heart failure (12.9%). Aneurysm and/or aortic dissection were less common (5.2%), and infective endocarditis (IE) was rare (1.1%). Regarding comorbidities among ACHD patients, hypertension (46.7%) and ischemic heart disease (IHD) (20.5%) were the most prevalent, followed by hyperlipoproteinemia (14.5%) and cerebrovascular diseases (14.1%). Diabetes mellitus (DM) was present in 9.6% of cases, followed by chronic obstructive pulmonary disease (COPD) (7.2%), CKD (1.9%), and malignancy (1.7%) (Figure 1).

Table 2 illustrates the distribution of these complications and comorbidities according to ACHD type. The most common complication was arrhythmia in the septal defect and PDA group (33.7%), and this same group had the highest prevalence of hypertension (34.9%). In the overall population, atrial rhythm disturbances were present in 19%, with atrial fibrillation (AF) occurring in 16.1% of cases. Chronic coronary syndrome was observed in 17.1% of ACHD patients (Table 3).

Cardiothoracic surgery was performed on a total of 141 patients (16.9%) during the 10-year treatment of ACHD patients, while septal ASD occluder implantation was required in 7.1% of cases. Pacemaker implantation was

Table 2. Complications by different ACHD

	Septal defects and DAP		RVOT		LVOT, CoA		Complex ACHD (TOF, TGA)		APVD and cor triatriale		Coronary anomalies		Marfan/ Turner syndrome		PSVCS/dextrocardia and unspecified	
Complications	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Arrhythmias	282	33.7	13	1.6	35	4.2	20	2.4	4	0.5	7	0.8	4	0.5	6	0.7
IE	4	0.5	2	0.2	1	0.1	0	0	0	0	0	0.0	1	0.1	0	0
HF	81	9.7	5	0.6	12	1.4	3	0.4	1	0.1	2	0.2	2	0.2	2	0.2
Aneurism and/ or dissection	15	1.8	0	0	29	3.5	0	0	0	0	1	0.1	2	0.2	1	0.1
HTA	292	34.9	8	1.0	61	7.3	8	1.0	2	0.2	10	1.2	2	0.2	7	0.8
HLP	91	10.9	3	0.4	17	2	1	0.1	2	0.2	5	0.6	0	0	2	0.2
DM	61	7.3	1	0.1	5	0.6	2	0.2	3	0.4	2	0.2	0	0	6	0.7
CAD	138	16.5	1	0.1	18	2.2	1	0.1	3	0.4	5	0.6	1	0.1	4	0.5
COPD	42	5.0	3	0.4	7	0.8	3	0.4	0	0	0	0	0	0	5	0.6
CKD	14	1.7	0	0	1	0.1	0	0	1	0.1	0	0	0	0	0	0
CVD	100	12.0	2	0.2	7	0.8	2	0.2	0	0	1	0.1	0	0	6	0.7
Malignitet	11	1.3	0	0	2	0.2	0	0	0	0	0	0.0	1	0.1	0	0

HTA, arterial hypertension; DM, diabetes mellitus; COPD, chronic obstructive pulmonary disease; CKD, chronic kidney disease; DAP, persistent arterial duct; LVOT, left-sided heart obstructive defects; CoA, aortic coarctation; RVOT, right-sided heart obstructive defects; TOF, tetralogy of Fallot; TGA, transposition of the great arteries; PSVCS, persistent left superior vena cava; HF, heart failure; CVD, cerebrovascular disease; CAD, coronary artery disease; HLP, hyperlipoproteinemia; APVD, anomalous pulmonary vein drainage

Table 3. Specific complications and comorbidities in ACHD

Specific complications	N	%
AV block, bundle branch block and rhythm disturbances	118	14.1
Atrial rhythm disturbances	167	19.9
Ventricular rhythm disturbances	57	6.8
Sinus node disfunction	6	0.7
Unspecific rhythm disturbances	39	4.7
AF	135	16.1
Aneurism and/or dissection with rupture	4	0.4
Aneurism and/or dissection without rupture	44	5.2
precerebral arteries occlusion	62	7.4
Cerebrovascular events	56	6.7
Acute coronary syndrome	28	3.3
Chronic coronary syndrome	143	17.1

AV, atrioventricular; AF, atrial fibrillation

necessary in 2.9% of patients, and implantable cardioverter-defibrillator (ICD) placement in 0.6%. Coronary angiography was performed in 23.7% of patients for the diagnosis and treatment of chronic or acute coronary syndrome, with percutaneous coronary intervention (PCI) performed in 5.1%.

Discussion

Patients with ACHD are becoming increasingly prevalent in adult cardiology as more children and young adults with complex, palliatively managed CHDs survive into adulthood. In Japan, the number of adults with ACHD equaled that of children with CHDs in 1997, with an estimated annual growth rate of 5%.[13] Our study found a 0.8% prevalence of ACHD during a ten-year follow-up, corresponding to the estimated prevalence in children (0.8-1%). A Canadian study reported a prevalence of CHDs of 4.09 per 1000 adults, or 5.78 per 1000 in the general population in 2000.[14] However, it should be noted that the registered number of ACHD patients in our study is not representative of the general population, as some adult ACHD patients gravitate towards other specialized cardiology centers with experience in managing CHDs.

Studies have demonstrated an increasing number of adults with ACHD and a higher proportion of severe defects. A Canadian study between 1983 and 2000 reported a prevalence of severe CHDs of 1.45 per 1000 children and 0.38 per 1000 adults, constituting 12% and 9% of all defects, respectively. The average age of all patients with severe CHDs increased from 11 years in 1985 to 17 years in 2000.[10] From 1996 to 2007, the number of simple CHDs steadily increased, while the number of severe CHDs significantly increased from 2008/2009[15]. In our studied ACHD group, simple defects were more prevalent compared to severe ACHD (TOF, TGA, atrioventricular canal). Authors report a prevalence of complex ADCH being twice more compared to our population.[14]

Among our ACHD patients, the most common conditions were ASD (49.1%), VSD (13.1%), congenital aortic valve anomalies (10%), followed by coarctation of the aorta and other aortic stenosis (6.2%). Other studies reported different prevalence rates, with VSD (19.2%), ASD (13.0%), TOF (9.3%), univentricular heart (9.4%), and CoA (7.0%).[15] A study in the same country but on newborns and infants up to 3 months reported the following prevalences: VSD (48.9%), ASD (17.0%), valvular pulmonary stenosis (6.1%), PDA (4.3%), and CoA (3.6%)[16]. Although many muscular VSDs spontaneously close in early childhood, this defect remains the most common in later years.[17)]

Countries with lower living standards, especially in Asian regions, exhibit a heightened occurrence of ASDs. This may be correlated with environmental factors such as

air pollution.[18] ASD is considered a simple CHD, and considerable defects have a good prognosis if identified and treated timely. The chosen treatment method is percutaneous septal occluder implantation, which was performed in 7.1% of our ACHD patients.

The third most prevalent defect in our group were aortic valve anomalies (most commonly BAV), presented in 10% of cases. This percentage is likely even higher, as BAV was initially reported as congenital aortic stenosis (2.2%) or as part of anomalies involving the aortic and mitral valves (0.4%). BAV occurs in 2% of the general population and does not always manifest as hemodynamic abnormalities. Valve dysfunction is reported in 16%-68% of cases, with dominant stenosis in 19.5% and regurgitation in 26.2%. Aortic dilation occurs in 84.8% of cases, more commonly in the ascending portion (81.3%) than in the aortic root (3.5%). BAV, aortopathies, and defects that require surgical correction in the aortic root present the risk factor for aortic dissection and/or aneurysms.[19]

Among other complications that can result from CHDs, stroke and arrhythmias are most frequently mentioned[2]. United States study from 1998 to 2011 reported a 91% increase in hospitalizations for ACHD patients compared to a 21% increase in patients without ACHD.[20] ACHD patients in our study had a high incidence of stroke 12.9%. ACHD patients are at higher risk of developing all types of arrhythmias, particularly atrial arrhythmias related to cardiac surgery and cardiac remodeling, while left atrial enlargement promotes atrial fibrillation (AF).[2] In a large study involving 38,428 ACHD patients, 15% had atrial arrhythmias, and the risk increased with age. The appearance of atrial arrhythmias increased the risk of any adverse event, with a 50% increase in mortality, doubling morbidity for cerebrovascular events and heart failure, and a threefold increase the risk of cardiac interventions.[5] In our study population, rhythm disturbances were common, with atrial arrhythmias being the most prevalent. The occurrence of atrial arrhythmias in our ACHD patients were higher when compared to other studies, likely due to the higher prevalence of septal defects[2]. ACHD patients are more likely to develop AF, even at a younger age.[5]

Individuals with CHDs are 15-100 times more likely to develop infective endocarditis (IE), with its occurrence more frequently associated with defects affecting the valves[6]. Pulmonary arterial hypertension (PAH) represents a complication of CHDs (PAH-CHD) that is sought to be avoided through early recognition of congenital defects and timely treatment. PAH-CHD is characterized by elevated pulmonary vascular resistance (PVR).[2,21] Post-capillary PH is more common and results from pressure "transmission" to the pulmonary circulation due to increased left ventricular filling pressure in cases of LV dysfunction or AV valve regurgitation when PVR is normal. Our study group had a 15.2% prevalence of PH, with over 95% of cases being secondary. We identified six cases of Eisenmenger syndrome, two of which had ASD and four had VSD.

Considering the older age of ACHD patients, it is expected that, in addition to their primary condition, they would have a higher risk of developing acquired diseases due to exposure to conventional risk factors for cardiovascular disease. In a cohort analysis of 250 patients[22] with ACHD, with an average age of 51±15 years and 53% of them being male, selective coronary angiography was performed for reasons unrelated to suspected CAD. Significant CAD was found in 9.2% of the participants, and its frequency was similar to that in the general population. Analysis of traditional risk factors for CAD concluded that they hold equal importance in prevention as in the general population. The strongest predictors of coronary disease were hypertension (HTA) and hyperlipidemia, with a prevalence of 29.3% for HTA and 19.1% for hyperlipidemia. None of the included cyanotic ACHD patients (7.2%) in the study had significant CAD. The occurrence of HTA and CAD in our study group was higher compared to data in other studies, while the prevalence of diabetes mellitus (DM) and hyperlipidemia was in line with previously published results in other studies. ACHD patients more frequently presented with chronic coronary syndrome. Approximately 2% of ACHD patients have congenital anomalies of coronary vessels, and it remains unexplored to what extent they contribute to the clinical manifestations of CAD.

Current epidemiological data indicate that CKD occurs more frequently among ACHD patients (in 30-50% of cases), especially in cyanotic defects and at an earlier age. Mild renal dysfunction was present in 41% of patients, while moderate to severe dysfunction was observed in 9%.[23] They are at 10-100 times greater risk of CVE despite the absence of classical cardiovascular risk factors. A large Danish study encompassing all adult ACHD patients from 1963 to 2017 revealed that the risk of myocardial infarction (MI) at the age of 30 was 0.7% in ACHD patients, compared to 0.1% in the general population cohort. At the age of 60, the respective risks were 7.4% versus 2.9%.[11] In this study the occurrence of stroke is associated with the following conditions: ASD (4%); closed ASD or VSD (1.4%); corrected TOF (2.4%); Eisenmenger's physiology (5.1%); other cyanotic ACHD (23.3%); mechanical valves (3.3%); absence of sinus rhythm (25%); implanted pacemakers (7%); endocarditis (2%); cardiac surgical interventions (11%); and percutaneous interventions (2%).(24) In our cohort, 6.78% of the total number of ACHD patients experienced a stroke.

Conclusion

The prevalence of congenital heart diseases in our adult population is 0.8%. The most common defects in adulthood are septal defects, while anomalies of the aortic valve rank second. The most frequent complications are cardiac rhythm disorders and pulmonary hypertension, with the most common comorbidities being hypertension and CAD.

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Sažetak

Adultne kongenitalne bolesti srca – desetogodišnje lečenje pacijenata u Institutu za kardiovaskularne bolesti Vojvodine

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Uvod. *Broj odraslih bolesnika sa urođenom srčanom manom (adultnom kongenitalnom srčanom bolešću - ACHD) je danas veći od broja dece rođenih sa manom. Cilj rada bio je ispitivanje karakteristika bolesnika sa ACHD lečenih u 10 godišnjem periodu na Institutu za kardiovaskularne bolesti Vojvodine (IKVBV).*

Metodologija. *Retrospektivna kohortna studija uključila je bolesnike pregledane i lečene pod dijagnozom urođene srčane mane na IKVBV od 2011–2021. godine. Pretraživanjem elektronske baze na osnovu MKB klasifikacije izdvojene su dijagnoze prema kojima su identifikovani bolesnici sa ACHD. Kod ispitivane grupe evaluirali smo demografske karakteristike, prisustvo komorbiditeta i komplikacija koje su se razvile tokom praćenja u predefinisanom vremenskom periodu.*

Rezultati. *Od ukupno 106 123 bolesnika lečenih u toku 10 godišnjeg perioda 0.8% je imalo dijagnostikovanu ACHD. Najčešće registrovane ACHD su: defekt atrijalnog septuma (ASD) 49.1%, defekt ventrikularnog septuma13.1%, urođene anomalije aortne valvule 10%, koarktacija aorte i druga suženja aorte 6.2%, dok je udeo kompleksnih mana, tetralogije Fallot 2.7% i transpozicije velikih arterija 1.2%. Od mogućih komplikacija najzastupljenije su: poremećaj srčanog ritma 44% i plućna hipertenzija 15.2%. Od komorbiditeta najzastupljenije su arterijska hipertenzija (46.7%) i ishemijska bolest srca (20.5%).*

Zaključak. *Prevalenca ACHD je 0.8%, najzastupljenije su mane iz grupe septalnih defekata. Najčešće komplikcije kod ACHD bile su poremećaj srčanog ritma i plućna hipertenzija, dok su najčešći komorbiditeti bili ishemijska bolest srca i arterijska hipertenzija.*

Ključne reči: *adultne kongenitalne srčane bolesti, ACHD, urođene srčane mane, komplikacije, komorbiditeti*