Kirurški rezultati bolesnika s prirođenim srčanim greškama u odraslih u Kliničkom bolničkom centru Zagreb

Surgical Outcomes for Patients with Adult Congenital Heart Disease in the University Hospital Centre Zagreb

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SAŽETAK: Broj bolesnika s prirođenim srčanim greškama u odraslih bolesnika (PSGO) stalno se povećava. U Kliničkom bolničkom centru Zagreb od 2015. godine ustanovljena je služba za liječenje takvih bolesnika, a ono uključuje i kirurško zbrinjavanje. Pregledali smo bazu podataka kirurških bolesnika s PSGO-om od 2009. do 2015. godine da bismo dobili uvid u broj bolesnika, najčešće dijagnoze i ishode liječenja te usporedili naše rezultate s inozemnim centrima, koji već imaju veće iskustvo u liječenju bolesnika s PSGO-om. Izvedeno je 111 operativnih zahvata u 108 bolesnika, jednakog udjela po spolu, prosječne dobi 35 godina. Četrdeset četiri posto operativnih zahvata bile su reoperacije. Mortalitet i morbiditet bili su niski (0,9 %). Činjenica je da se, otkako je ustrojena Radna skupina za PSGO Hrvatskoga kardiološkog društva, počeo povećavati i broj kirurški liječenih bolesnika.

SUMMARY: The number of patients with adult congenital heart disease (ACHD) is constantly increasing. In the University Hospital Centre Zagreb, a service was established in 2015 to treat such patients, including surgical treatment. We analyzed the database of patients with ACHD in the period between 2009 and 2015 to determine the number of patients as well as the most common diagnoses and outcomes, which we then compared to centers outside Croatia that are more experienced in treating ACHD. At our center, 111 surgical procedures were performed on 108 patients in that period, with an even ratio between women and men and a mean age of 35. Reoperations accounted for 44% of the procedures. Mortality and morbidity were low (0.9%). Since the establishment of the Working Group for ACHD of the Croatian Cardiac Society, the number of surgically treated patients has grown.

KLJUČNE RIJEČI: prirođene srčane greške, odrasli bolesnici, kirurško liječenje.

KEYWORDS: congenital heart disease, adult patients, surgical treatment.

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Uvod

Odrasli bolesnici s prirođenom srčanom greškom (PSGO) postaju skupina koja se stalno povećava, čemu je pridonijelo i uspješno kirurško liječenje tih bolesti u djetinjstvu[1]. Većina novorođenčadi nakon operativnog liječenja danas doživi odraslu dob. Mala grupa bolesnika nije dijagnosticirana u djetinjstvu te se sa zdravstvenim problemima prvi put javljaju u odrasloj dobi. Najčešće dijagnoze u takvih bolesnika jesu bikuspidni aortni zalistak, atrijski septalni defekt, obično tipa venskog sinusa, s parcijalnim anomalnim utokom plućnih vena u desni atrij ili bez njega, nadalje, koarktacija aorte, kongenitalno korigirana transpozicija velikih krvnih žila te Ebsteinova anomalija. Međutim, velika je većina već u ranome djetinjstvu bila podvrgnuta nekom intervencijom

Introduction

Adult congenital heart disease (ACHD) affects a growing patient group, partly because of improved treatment of these diseases in childhood[1]. Today, most newborns with congenital heart disease reach adulthood due to surgical treatment. A small group of patients is diagnosed only in adulthood, however. The most common diagnoses in this smaller group are bicuspid aortic valve disease, atrial septal defect, usually the sinus venosus type with or without partial anomalous pulmonary venous return to the right atrium, coarctation of the aorta, congenitally corrected transposition of the great vessels, and Ebstein's anomaly. However, most of these patients underwent some form of interventional treatment, either percutaneous or,

tipu liječenja, bilo perkutanom bilo, češće, kirurškom. Kirurško liječenje bolesnika s PSGO-om poseban je problem zbog velikoga broja srčanih grešaka, različita patoanatomskog supstrata te komplikacija koje nastaju zbog rezidualnih defekata, kao i činjenice da srčana bolest uzrokuje promjene i na drugim organskim sustavima.

Bolesnici i metode

Svi bolesnici s PSGO-om stariji od 15 godina koji su kirurški liječeni u razdoblju od 1. rujna 2009. do 8. listopada 2015. u Klinici za kardijalnu kirurgiju Kliničkoga bolničkog centra Zagreb uključeni su u ovu studiju. Analizirani su dob i spol bolesnika, dijagnoze, prethodni operativni zahvati, kao i sadašnje stanje te ishod liječenja. Vrste operativnih zahvata grupirane su na dva načina: prema anatomskoj lokalizaciji i prema kompleksnosti zahvata.

Rezultati

U razdoblju od pet godina u ustanovi je bilo podvrgnuto kirurškom zahvatu 108 bolesnika s PSGO-om, u potpuno jednakom omjeru, po 54 muškarca i žena. U troje bolesnika učinjena su po dva zahvata, u različitim vremenima, tako da je ukupan broj operacija 111.

Jedan od primjera jest bolesnica koja je u djetinjstvu operirana zbog Fallotove tetralogije s atrezijom pulmonalnog zalistka te je učinjeno zatvaranje ventrikulskoga septalnog defekta, a umjesto atretičnog zalistka ugrađen je provodnik s biološkom protezom. Bolesnica je još imala kongenitalno korigiranu transpoziciju velikih krvnih žila, uz dekstrokardiju. U ustanovu se javila nakon trideset godina, sa stenoziranim provodnikom i dilatiranom lijevom klijetkom, koja obavlja funkciju desne. Funkcija sistemske klijetke, kao i pripadajućih zalistaka bila je uredna. Nakon uspješno promijenjenog provodnika, bolesnica je uz dobro opće stanje otpuštena iz bolnice, no nekoliko mjeseci poslije počinju se pojavljivati zaduhe. Na ultrazvučnom se nalazu sada vidi mnogo manja lijeva, odnosno pulmonalna klijetka, no sada se proširila sistemska te se javlja insuficijencija sistemske atrio-ventikularne valvule, koja se prije nije vidjela zbog znatno dilatirane pulmonalne klijetke. Oboljeli je zalistak zamijenjen biološkom protezom. Ovo je i dobar primjer kompleksnosti bolesnika s PSGO-om, kao i poteškoća pri klasifikaciji takvih zahvata. Ta je bolesnica uvrštena u dvije skupine kirurških zahvata, kao zamjena biološke proteze u provodniku te kao zamjena oboljelog trikuspidalnog zalistka pri kongenitalno korigiranoj transpoziciji, koji ima ulogu mitralnog, odnosno sistemskog AV zalistka.

Dob bolesnika kretala se od 15 do 62 godine (prosječna dob 35,14 ± 14,69 godina). Raspodjela po dobi prikazana je na **slici 1**. Broj je operacija s godinama rastao, a vidljiv je znatan skok nakon što je 2015. godine organizirano započela s radom Radna skupina za PSGO Hrvatskoga kardiološkog društva (**slika 2**).

Od ukupnoga broja zahvata, 61 (56 %) bio je prvi operativni zahvat na bolesniku, a 47 zahvata (44 %) je bilo ponovljeno (drugi ili više). Jednoj bolesnici s Fallotovom tetralogijom u našoj je ustanovi učinjen šesti zahvat na srcu.

Kardiokirurški su zahvati, radi jednostavnosti, podijeljeni u velike grupe, složene po anatomskoj lokalizaciji (**slika 3**). Najveću skupinu činili su bolesnici koji su podvrgnuti zahvamore commonly, surgical. Surgical treatment of ACHD is a special problem due to the large number of heart defects, different pathoanatomical substrates, and complications caused by residual defects, as well as the fact that heart diseases also cause changes in other organ systems.

Patients and methods

We assessed the number of surgical procedures performed on patients with ACHD at the Cardiac Surgery Clinic of the University Hospital Centre Zagreb in the period between September 1, 2009 and October 8, 2015. Patients aged 15 and above were included in the analysis. The age and sex of the patients were analyzed, as well as diagnoses, previous surgical procedures, and current outcomes. The types of surgical procedures were classified by anatomical location and by complexity.

Results

Over the 5-year period, 108 patients with ACHD underwent surgical procedures in our institution, 54 men and an equal number of women. Three patients underwent two procedures each, bringing the total number of procedures to 111.

One example is a female patient who underwent a surgical procedure in her childhood for tetralogy of Fallot with pulmonary atresia during which the ventricular septal defect was closed and a shunt with a biological prosthesis was implanted in place of the atretic valve. The patient also had a congenitally corrected transposition of the great vessels with dextrocardia. She presented at our institution 30 years after the shunt implantation with shunt stenosis and a dilated left ventricle performing the function of the right. Systemic ventricular function and the associated valve was competent. After shunt replacement, the patient was discharged from the hospital in good condition; however, dyspnea began to manifest some months later. Ultrasound revealed a significantly smaller left, i.e. pulmonary, ventricle and an increase in the size of the systemic ventricle, causing insufficiency of the systemic atrioventricular valve, which was not visible earlier due to the significant dilation of the pulmonary ventricle. The dysfunctional valve was replaced with a bioprosthetic valve. This is good example of the complexity of cases of ACHD and the difficulty in classifying such procedures. The patient was included in two groups of surgical procedures: among biological valve replacements in the shunt and as a replacement procedure for dysfunctional tricuspid valves in congenitally corrected transposition with a mitral, i.e. systemic atrioventricular valve.

Among all patients, patient age was between 15 and 62 years of age (average ± standard deviation = 35.14±14.69). Age distribution is shown in **Figure 1**. The number of surgical procedures increased with time, and a significant increase is evident after the Working Group for ACHD of the Croatian Cardiac Society began its work in 2015 (**Figure 2**).

Out of all the procedures, 61 (56%) were the first surgical procedure performed on the patient and 47 (44%) were at least the second procedure performed on the patient. One female patient with tetralogy of Fallot had her sixth cardiac procedure performed at our institution.

For simplicity's sake, we divided the cardiac surgery procedures into large groups based on anatomical location (**Figure**

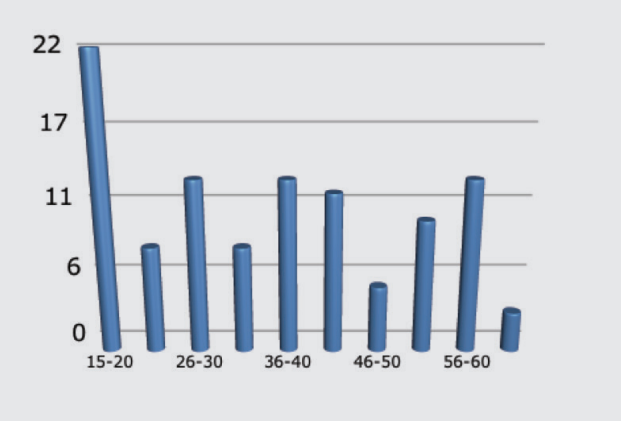


FIGURE 1. Age distribution (in years) of patients operated from 2009 to 2015.

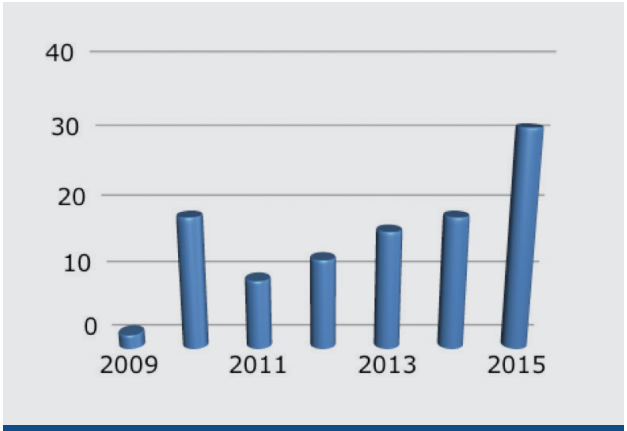
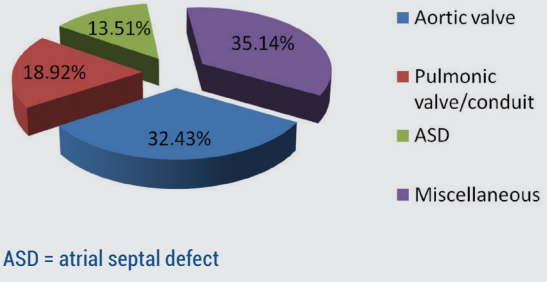


FIGURE 2. Number of patients operated from 2009 to 2015.

tu na aortnom zalistku (36 pacijenata; 32,43%). U svim je slučajevima riječ bila o bikuspidnom aortnom zalistku. U osam bolesnika učinjena je jednostavna zamjena bolesnog zalistka umjetnom protezom. Jedan je od njih imao i dodatni atrijski septalni defekt. Kod devet je bolesnika, osim zamjene zalistka, učinjeno i dodatno proširivanje anulusa aortnog zalistka. U jednog je bolesnika učinjena plastika zalistka, a kod 14 njih trebalo je, zbog dodatne aneurizme, zamijeniti i ascendentnu aortu. Jedan je bolesnik dobio plastiku aortnog zalistka i zamjenu ascendentne aorte umjetnom protezom, a kod još jednog, uz zamjenu aortnog zalistka, učinjena je i plastika pulmonalnog zalistka. Jednom je bolesniku zbog supravalvularne aortne stenoze učinjena plastika suženoga mjesta po Dotyju. Bolesniku s perzistentnom lijevom gornjom šupljom venom, koja se ulijevala u lijevi atrij, učinjena je translokacija lijeve vene u desnu. Još je učinjeno vađenje eletroda elektrostimulatora u bolesnika s transpozicijom velikih krvnih žila, nakon operacije po Jatenu.

Druga skupina, po brojnosti, bili su zahvati na pulmonalnom zalistku (21 bolesnik, 18,92%). U četiri bolesnika trebalo je ugraditi biološki zalistak nakon korekcije Fallotove tetralogije, a u još pet zbog drugih osnovnih dijagnoza. U šest bolesnika rađena je plastika izlaznoga trakta desne klijetke. Ostatak su bili bolesnici nakon operacije po Rossu. U troje bolesnika trebalo je zamijeniti pulmonalni homograft, a kod još troje,



ASD = atrial septal defect

FIGURE 3. The distribution of patients categorized by anatomic lesions.

3). The most common location was the aortic valve (36 patients; 32.43%), all of which were bicuspid aortic valve cases. A simple prosthetic valve replacement procedure was performed in 8 patients. One of them also had an additional atrial septal defect. In addition to valve replacement, 9 patients also underwent a procedure for additional aortic annular expansion. One patient had a valvuloplasty, and 14 required replacement of the ascending aorta due to an additional aneurysm. One patient underwent a vavuloplasty procedure and had his ascending aorta replaced with a prosthetic, and another patient underwent pulmonary valve repair in addition to aortic valve replacement. A patient had Doty repair performed due to supravalvular aortic stenosis. A translocation of the left caval vein into the right was performed in a patient with a persistent left superior vena cava draining into the left atrium. Additionally, lead removal was performed on a patient with transposition of the great arteries after a Jatene procedure.

The second most numerous group were procedures on the pulmonary valve (21 patients, 18.92%). A biological implant was necessary in 4 patients after tetralogy of Fallot repair, and in another 5 as well due to other basic diagnoses. In 6 patients, valvuloplasty was performed on the right ventricular outflow tract, and the rest were patients after a Ross procedure. Three of them required pulmonary homograft replacement, and a further 3 also required replacement of a dilated ascending aorta.

The third large group consisted of patients with an atrial septal defect (15 patients, 13.51%). All underwent valve repair with autologous pericardium.

The rest of the patients, 39 of them (35.14%), were classified in a miscellaneous group due to very diverse procedures.

A more detailed breakdown of surgical procedures are shown in **Table 1**, **Table 2**, and **Table 3**. The diagnosis, type, and number of procedures are shown, as well as the complexity of the procedures according to the Canadian model for ACHD patients[2,3]. Among the simple procedures (**Table 1**), most common were atrial septal defects (ASD) and aortic valve replacement. Of 14 patients with ASD, 3 cases were reoperations after a residual defect was found.

In the moderately complex procedure group (**Table 2**), 5 patients diagnosed with Ebstein's anomaly are shown in the late

osim homografta, trebalo je zamijeniti i proširenu ascendentnu aortu.

Treću veliku skupinu činili su bolesnici s atrijskim septalnim defektom (15 bolesnika, 13,51%). Kod svih je učinjena plastika defekta s autolognim perikardom.

Ostali su bolesnici, njih 39 (35,14%), zbog vrlo raznolikih zahvata svrstani u posebnu skupinu.

Detaljnija podjela operativnih zahvata prikazana je u **tablicama 1**, **2** i **3**. Navedene su dijagnoze, vrsta i broj operativnih zahvata, kao i stupanj složenosti zahvata, prema kanadskom modelu za bolesnike s PSGO-om[2,3]. Najveći broj procedura u skupini jednostavnih zahvata (**tablica 1**) bili su atrijski septalni defekt (ASD) i zamjena aortalnog zalistka. Među 14 bolesnika s dijagnozom ASD-a izvedene su tri reoperacije nakon nađenoga rezidualnog defekta.

U skupini umjereno teških zahvata (**tablica 2**), pet bolesnika s dijagnozom Ebsteinove bolesti prikazano je u kasnom stadiju bolesti te je kod svih učinjena zamjena trikuspidalnog zalistka biološkom protezom. Jednom je bolesniku, zbog smetnji provođenja, ugrađen trajni elektrostimulator. U skupini parcijalnog atrio-ventrikularnog kanala, dva su bolesnika imala primarnu operaciju, dok je u četiriju bolesnika, nakon prethodno učinjene korekcije, zbog teško promijenjenog mitralnog zalistka, ugrađena umjetna proteza. Sedmero je bolesnika, zbog parcijalnoga anomalnog utoka plućnih vena u desni atrij, podvrgnuto kirurškoj korekciji intraatrijskim tunelom, a još je jednom, zbog dodatne trikuspidalne insuficijencije, učinjena i plastika zalistka. Na isti je način liječen i jedan bolesnik koji se u odrasloj dobi javio s dijagnozom scimitar-sindroma. Tri su bolesnika imala ventrikularni septalni defekt i stenozu izlaznoga trakta desne klijetke. Jednom bolesniku nakon preboljenog karcinoida zaostala je teška insuficijencija pulmonalnog i trikuspidalnog zalistka, koji su zamijenjeni biološkim protezama. U bolesnika s dvostrukim desnim ventrikulom učinjena je resekcija mišićnoga snopa.

U skupini složenih kardiokirurških zahvata (**tablica 3**) bila su dva bolesnika s transpozicijom velikih krvnih žila, kojima je u djetinjstvu učinjena operacija po Senningu. Oba su se javila s teškim insuficijencijama atrio-ventrikularnih zalistaka te im je učinjena zamjena trikuspidalnog, kao i plastika mitralnog zalistka. Jednoj bolesnici s kongenitalno korigiranom transpozicijom velikih krvnih žila, o kojoj je bilo riječi prije u tekstu, učinjena su dva odvojena zahvata: ugradnja biološkog zalistka u pulmonalni provodnik te zamjena trikuspidalnog zalistka biološkom protezom. U dvije bolesnice učinjena je transplantacija srca zbog različitih dijagnoza. Jedna je bila mlada bolesnica koja je nakon kirurške korekcije Fallotove tetralogije zadobila kardiomiopatiju te je u nje uspješno provedena transplantacija. Druga je bila 38-godišnja bolesnica s kompleksnom srčanom greškom. Posrijedi su bili heterotaksija, potpuni anomalni utok plućnih vena u desni atrij, zajednički tip atrio-ventrikularnog kanala i pulmonalna stenoza. U djetinjstvu je palijativno liječena Blalock-Taussigovim šantom te se otada nije kontrolirala. U KBC Zagreb javila se u dekompenziranom stanju, a zbog kompleksnosti greške postavljena je indikacija za hitnu transplantaciju srca. Kiruški je zahvat izveden bez komplikacija, no 33. postoperativnog dana dolazi do letalnog ishoda pod kliničkom slikom multi-

TABLE 1. Number of operations with detailed diagnoses and surgical procedures classified as simple complexity category

Diagnosis	Surgical procedure	Number of operations
ASD	Patch closure	14
ASD+Aortic valve stenosis	Patch closure+AVR	1
Bicuspid aortic valve	AVR	7
	AV repair	1
BAV+AAA	Op. sec. Bentall	2
	AVR+aortic replacement	14
	AV repair+aortic replacement	1
BAV+Pulmonary valve stenosis	AVR+PV repair	1
CoA	Op. sec. Vossshulte	1
Supravalvular aortic stenosis	Op. sec. Doty	1
LVCS to LA connection	LVCS translocation	1
S/p Jatene	PM electrode extraction	1

ASD = atrial septal defect; AVR = aortic valve replacement; AAA = ascending aorta aneurysm; BAV = bicuspid aortic valve; CoA = coartation of aorta; LVCS = left vena cava superior; LA = left atrium; PM = pacemaker.

stage of the disease, and all five underwent tricuspid valve replacement with a bioprosthetic valve. One patient had a permanent pacemaker implanted due to conduction disorders. In the partial atrioventricular septal defect group, 2 patients underwent a primary procedure while 4 had a prosthesis implanted after a previous repair procedure due to severe changes on the mitral valve. Due to partial anomalous pulmonary venous return, 7 patients underwent an intra-atrial conduit procedure, and one patient also underwent plastic valve repair due to additional tricuspid insufficiency. A patient presenting with scimitar syndrome at an adult age was treated in the same way. Three patients had a ventricular septal defect and right ventricular outflow tract stenosis. After surviving a carcinoid, one patient was left with severe pulmonary and tricuspid valve regurgitation, which were then replaced with bioprosthetic valves. A muscle resection was performed on a patient with a double-chambered right ventricle.

The complex procedure group (**Table 3**) included two patients with transposition of the great arteries that had undergone a Senning procedure in childhood. Both patients presented with severe atrioventricular regurgitation, so tricuspid valve replacement and mitral valve repair was performed. One female patient with congenitally corrected transposition of the great vessels, whom we have already mentioned, had two separate procedures performed: implantation of a bioprosthetic valve into the pulmonary shunt and tricuspid

TABLE 2. Number of operations with detailed diagnoses and surgical procedures classified as moderate complexity category

Diagnosis	Surgical procedure	Number of operations
BAV+dysplastic valve	AVR+aortic root enlargement	9
Tetralogy of Fallot	ToF correction	1
	ToF correction+MVP	1
Pulmonary valve regurgitation	PVR	4
RVOT stenosis	RVOT enlargement	6
S/p Ross	PVR	3
	PVR+Op. sec. Bentall	3
Pulmonary valve anomaly	PVR	5
Mb. Ebstein	TVR	5
Thoracic aorta aneurysm	Aneurysm resection	1
MR+TR+ASD	ASD closure+MVR+TVP	1
PAPVR	Intracardiac repair	7
PAPVR+TI	Intracardiac repair+TVP	1
Scimitar sy.	Intracardiac repair	1
PAVC	PAVC correction	2
PAVC+dysplastic mitral valve	PAVC correction+MVR	4
VSD+RVOTO	VSD closure+RVOT enlargement	3
PI+TI after cacinoid	PVR+TVR	1
Double chambered right ventricle	Muscle bundle resection	1

BAV = bicuspid aortic valve; AVR = aortic valve replacement; ToF = Tetralogy of Fallot; MVP = mitral valve repair; PVR = pulmonary valve replacement; RVOT = right ventricle outflow tract; TVR = tricuspid valve replacement; ASD = atrial septal defect; MVR = mitral valve replacement; TVP = tricuspid valve repair; PAPVR = partial anomalous pulmonary veins return; TI = tricupid valve insufficiency; PAVC = partial atrioventicular canal; VSD = ventricular septal defect; PI = pulmonary valve insufficiency.

TABLE 3. Number of operations with detailed diagnoses and surgical procedures classified as complex category

Diagnosis	Surgical procedure	Number of operations
S/p Senning	TVR+MVP	2
ToF	HTx	1
CAVC.TAPVR.S/p B-T shunt	HTx	1
Cor triatriatum	Membrane resection	1
ccTGA	Conduit replacement	1
	TVR	1

TVR = tricuspid valve replacement; MVP = mitral valve repair; ToF = tetralogy of Fallot; Hex = heart transplantation; CAVC = complete atriventricular canal; TAPVR = total anomalous pulmonary veins return; B-T = Blalock-Taussig; ccTGA = congenitaly corrected transposition of great arteries.

organskog zatajenja. To je ujedno i jedini letalni ishod svih bolesnika s PSGO-om u KBC-u Zagreb (0,9 %). Svi su ostali s dobrim općim stanjem otpušteni na kućnu njegu.

Rasprava

Bolesnici s PSGO-om u naprednom dijelu svijeta danas postaju skupina koja vrlo brzo raste. Napredak pedijatrijske kar-

bioprosthetic valve replacement. Two female patients underwent heart transplant surgery due to different diagnoses. One was a young patient presenting with cardiomyopathy after tetralogy of Fallot correction; the heart transplant was successful. The other was a 38-year-old patient with a complex heart defect. It involved heterotaxy, completely anomalous inflow of pulmonary arteries into the right atrium, a complete atrioventricular canal defect, and pulmonary stenosis. She had received palliative treatment in childhood in the form of a Blalock-Taussig shunt and had not attended follow-up since then. She presented to the University Hospital Centre Zagreb with decompensated heart failure, and due to the complexity of the defect urgent heart transplantation was indicated. The surgical procedure was performed without complications, but there was a lethal outcome on the 33rd postoperative day, presenting with the multiorgan failure. This was the only lethal outcome in any patient with ACHD in the University Hospital Centre Zagreb (0.9%). All others were discharged to home care in a good overall state.

Discussion

In the Western world, patients with ACHD are a rapidly growing group. Advancements in pediatric cardiology and cardiac surgery have allowed these patients to reach adulthood, when they present with different problems. In Canada, the number of adult patients with congenital heart diseases has already overtaken the number of pediatric patients[2]. The situation is similar in Great Britain, where that trend started as early as the nineties[4]. In Croatia, the University Hospital Centre Zagreb is the only center systematically treating congenital heart defects, which includes surgical procedures. Several years ago, patients with ACHD started to appear in Croatia as well and if there was a need for surgical intervention, they were referred to pediatric surgeons in the University Hospital Centre Zagreb. Patients with ACHD or bicuspid aortic valve (BAV) disease that presented at the hospital in adulthood had been surgically treated in childhood at the University Hospi-

diologije i kardiokirurgije omogućio je takvim bolesnicima da dožive odraslu dob te se javljaju s drugim, različitim problemima. U Kanadi je broj odraslih bolesnika s prirođenim srčanim greškama već nadmašio pedijatrijske bolesnike[2]. Slično je i u Velikoj Britaniji, gdje je taj trend počeo još devedesetih godina prošloga stoljeća[4]. Jedini centar u Hrvatskoj koji se sustavno bavi liječenjem urođenih srčanih grešaka jest KBC Zagreb. U to je liječenje uključena i kirurška terapija. Unatrag nekoliko godina i u nas su se počeli javljati bolesnici s PSGO-om. Ako se pojavila potreba za kirurškom terapijom, bilo je razumljivo da su referirani pedijatrijskim kirurzima u KBC-u Zagreb. Bolesnici s ASD-om ili bikuspidalnim aortalnim zalistkom (BAV), koji su se tek u odrasloj dobi javili sa svojim problemima, bili su i prije kirurški liječeni, kako u KBC-u Zagreb, tako i u drugim kardiokirurškim centrima u Hrvatskoj, u kojima su ih operirali opći kardijalni kirurzi. Te su greške i inače vrlo česte, mnogo godina ne stvaraju posebne probleme, pa su se bolesnici javljali u kardiološke ambulante za odrasle[5,6]. Još uvijek postoje takvi bolesnici koji su liječeni u drugim centrima u Hrvatskoj, tamo operirani te se ne nalaze u Registru za PSGO pri KBC-u Zagreb. S obzirom na to da ASD i BAV pripadaju kategoriji jednostavnih zahvata, s niskim mortalitetom, opći ih kirurzi rado stavljaju na svoje liste. Te su dvije skupine najveće i u našoj populaciji i po tome se ne razlikujemo od inozemnih centara[7]. Od 2010. godine takvi su bolesnici u našoj ustanovi referirani pedijatrijskim kirurzima te se otada vodi i registar. Godine 2015. ustanovljena je i Radna skupina za PSGO pri Hrvatskome kardiološkom društvu, ustrojena služba za takve bolesnike u KBC-u Zagreb te se i njihov broj povećao. To se povećanje vidi i na broju operiranih bolesnika u 2015. godini.

Treću veliku skupinu operiranih bolesnika čine oni sa zahvatima na desnoj strani srca, od desne klijetke, pa sve do grana pulmonalnih arterija. Najčešće se radilo o bolesnicima koji su već imali prethodne zahvate, najčešće Fallotovu tetralogiju i u kojih se poslije u životu razvija pulmonalna insuficijencija. Prema studiji iz Toronta, zbog pulmonalne insuficijencije ili stenoze provodnika nakon totalne kirurške korekcije tetralogije u djetinjstvu, u 45,75 % bolesnika bio je potreban novi zahvat[8]. To je još jedan pokazatelj kompleksnosti urođenih srčanih grešaka te da takvi bolesnici trebaju doživotnu medicinsku skrb. Kod druge podskupine trebalo je, zbog izrazitih stenoza i degeneracija, mijenjati provodnik, ili zalistak u provodniku. Incidencija ovakvih zahvata također se ne razlikuje od podataka iz literature[1]. Može se reći da su ovo tipične komplikacije za bolesnike s PSGO-om koji su kirurški korigirani u djetinjstvu te se u kasnijoj dobi javljaju s novim ili rezidualnim lezijama, kao i s degeneriranim provodnicima ili umjetnim zaliscima. Ovu su skupinu gotovo u cijelosti činile reoperacije. Ako se dodaju bolesnici iz skupine složenih zahvata te oni s prethodno učinjenim komisurotomijama aortnog zalistka, skupina reoperiranih bolesnika iznosi 44 % ukupnog broja zahvata. U cijeloj seriji imali smo samo jedan letalni ishod, koji u podskupini od 47 bolesnika kojima je učinjen ponovni zahvat čini 2,13 %. U literaturi se mortalitet nakon reoperacija kreće od 3 do 7,6 %, uz napomenu da u kasnijim serijama znatno pada, što se objašnjava većim iskustvom u takvim zahvatima[9,10]. I ukupni se mortalitet s vremenom smanjuje, pa grupa iz Toronta pokazuje smanjenje

tal Centre Zagreb or in other cardiosurgical centers in Croatia, by general cardiac surgeons. Heart defects are very common and often do not cause any special problems for many years, so these patients would go to a cardiac clinic for adults[5,6.] Some patients are still treated at other centers in Croatia, receive surgeries there, and are not in the ACHD Registry at the University Hospital Centre Zagreb. Since ASD and BAV surgeries are comparatively simple and have a low mortality, general surgeons gladly accept them to their operation lists. Those two groups are the most common in Croatia, as in other centers abroad[7]. Since 2010, such patients have been referred to pediatric surgeons in our center, which is when our patient registry was started. The Working Group for ACHD of the Croatian Cardiac Society was established in 2015, as well as a service for such patients at the University Hospital Centre Zagreb, so the number of such patients increased as can be seen in the number of surgeries performed in 2015.

The third large group of surgical patients underwent procedures ranging from those on the pulmonary valve, although performed in the right ventricle, to those on the pulmonary artery branches. These were predominantly patients with previous procedures, most commonly tetralogy of Fallot correction, who developed pulmonary insufficiency later in life. According to a study from Toronto, 45.75% of patients who underwent total surgical correction for pulmonary insufficiency or shunt stenosis in childhood require a new procedure[8]. This is another indicator of the complexity of congenital heart defects and the fact that these patients require life-long medical care.

In the second subgroup, it was necessary to replace the shunt or the shunt valve due to severe stenosis and degeneration. The incidence of these procedures at our center matches the data from the literatue[1]. We can say that these are typical complications for patients with ACHD that underwent surgical correction in childhood and later in adulthood presented with new or residual lesions, degenerated shunts, or artificial valves. In this group, almost all procedures were reoperations. If we add up the patients from the complex procedure group and those with previous aortic valve commissurotomies, reoperations made up 44% of the total number of procedures. The whole series of patients had only one lethal outcome, i.e. 2.13% in the reoperation subgroup with 47 patients. In the literature, post-reoperation mortality is between 3 and 7.6% but drops sharply in later patient series, which can be explained by increasing experience in such procedures[9,10]. Total mortality has been dropping over time as well, and the Toronto group shows a reduction from 4.7% to 1.9% over three decades[11]. The most common cause of death was multi-organ failure and/or heart failure. The total mortality of 0.9% at the University Hospital Centre Zagreb does not differ significantly from international literature. Although surgical procedures on patients with ACHD have started only recently, the experience gained on pediatric and non-congenital cases is obviously bearing fruit.

We expect an even greater influx of patients with ACHD in the future, and thus an increasing number of surgical procedures. It is not yet known in which direction the treatment of these patients will develop. Globally, the number of patients requiring surgical procedures with low complexity is going down. More and more patients with atrial septal defects or

s 4,7 % na 1,9 % u razdoblju od tri desetljeća[11]. Najčešći je uzrok smrti bilo multiorgansko zatajenje i/ili popuštanje srca. Ukupni mortalitet u KBC-u Zagreb od 0,9 % ne razlikuje se znatno od svjetske literature. Iako se bolesnici s PSGO-om u ustanovi ne operiraju dugo vremena, iskustvo operatera stečeno na pedijatrijskim, kao i na nekongenitalnim odraslim bolesnicima, očito daje rezultate.

U budućnosti očekujemo još veći priljev bolesnika s PSGO-om, pa tako i veći broj onih koji zahtijevaju kirurško liječenje. Ostat će nepoznanica u kojem će se smjeru razvijati daljnje liječenje takvih bolesnika. Naime, u svijetu se smanjuje broj bolesnika koji trebaju kardiokirurške zahvate, koji bi se, po stupnju kompleksnosti, ubrajalii u jednostavne. Sve više bolesnika s atrijskim septalnim defektima ili greškama aortnog zalistka liječi se perkutanim interventnim zahvatima. Isto se odnosi i na bolesnike s rezidualnim ventrikularnim defektima ili pulmonalnim insuficijencijama nakon Fallotove tetralogije, a koji pripadaju kompleksnijoj grupu. Računa se da bi od 30 do 46 % bolesnika, koji se danas liječe kirurški, u budućnosti bili rješavani perkutanim intervencijama[12-14]. Ta naša budućnost u zapadnim je zemljama sadašnjost.

pulmonary insufficiency are being treated with percutaneous interventions. The same is true for patients with residual ventricular septal defects or pulmonary insufficiency after tetralogy of Fallot, belonging in the group of more complex procedures. It is estimated that 30 to 46% of patients that are treated surgically today will be treated with percutaneous interventions in the future[12-14]. This may be our future, but it has become reality in many Western countries.

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