

Organizacija tercijarnog centra za prirodene srčane bolesti u odraslih

Organizing a Tertiary Center for Adult Congenital Heart Disease

**Margarita Brida,
Maja Strozzi***

Medicinski fakultet Sveučilišta u Zagrebu, Klinički bolnički centar Zagreb, Zagreb, Hrvatska

University of Zagreb School of Medicine, University Hospital Centre Zagreb, Zagreb, Croatia

RECEIVED:
January 5, 2016

ACCEPTED:
January 31, 2016



CITATION: *Cardiol Croat.* 2016;11(1-2):5-7. | DOI: <http://dx.doi.org/10.15836/ccar2016.5>

***ADDRESS FOR CORRESPONDENCE:** Maja Strozzi, Klinički bolnički centar Zagreb, Kišpatičeva 12, HR-10000 Zagreb, Croatia. / Phone: +385-1-2367-508 / E-mail: maja.strozzi@gmail.com

ORCID: Margarita Brida, <http://orcid.org/0000-0001-8754-8156> • Maja Strozzi, <http://orcid.org/0000-0003-4596-8261>

Prirodene srčane greške čine trećinu svih velikih kongenitalnih anomalija, a točan broj djece rođene s kongenitalnom srčanom manom teško je procijeniti zbog različite dostupnosti i razine zdravstvene skrbi te mogućnosti dijagnosticiranja u različitim dijelovima svijeta. Podatci govore u prilog prevalenciji većoj od 8 na 1000 živorođene djece.¹

Preživljavanje djece rođene s kongenitalnom srčanom greškom u prošlosti je bilo izrazito loše, još u četrdesetim godinama prošloga stoljeća tek do 20 % takve djece uspjelo je odrasti. To se otada drastično promijenilo. Tijekom druge polovice XX. stoljeća, pioniri kardijalne kirurgije izumili su operativna rješenja za gotovo sve kompleksne srčane greške. Napretkom kardijalne kirurgije i pedijatrijske skrbi danas više od 90 % djece s kompleksnom greškom preživljava u odraslu dob. Procjenjuje se da je danas broj odraslih bolesnika dvostruko veći od broja djece s kongenitalnom srčanom greškom.²

Iako je riječ o iznimnom uspjehu liječenja ove skupine bolesnika, većina se ne može smatrati izliječena, već kažemo da je palijativno zbrinuta. U prilog tome govori i činjenica da se unatrag 15 godina broj hospitalizacija takvih bolesnika povećao za više od 80 %. Riječ je o kroničnoj bolesti koja zahtijeva dugotrajno praćenje i liječenje te često reintervencije zbog rezidualnih anatomskih i funkcionalnih abnormalnosti. Mnogi imaju povišeni rizik za razvoj aritmija, zatajivanja srca i endokarditisa.³ U Hrvatskoj se očekuje oko 10 000 takvih bolesnika, različite kompleksnosti.

Upravo ovaj preokret, odnosno činjenica da ova skupina bolesnika koji su nekad bili isključivo u pedijatrijskog skrbi, zahtijeva razvoj novog, specijaliziranog područja u odrasloj kardiologiji i

Congenital heart disease makes up for one third of all serious congenital anomalies; the exact number of children born with congenital heart defects is hard to estimate due to differences in the availability and quality of health care and diagnosis in different parts of the world. Data indicate a prevalence of over 8 per 1000 newborns.¹

Survival rates for children born with congenital heart defects were very poor in the past, with only up to 20% of such children reaching adulthood in the 1940s. This has changed drastically. During the second half of the 20th century, pioneers of cardiac surgery came up with surgical solutions for almost all complex heart defects. With advancements in cardiac surgery and pediatric care, today over 90% of children with complex defects survives into adulthood. It is estimated that the number of adult patients is now twice as large as the number of children with congenital heart defects.²

Although this is an outstanding success in the treatment of these patients, most cannot be considered cured, but rather palliated. This is supported by the fact that during last 15 years, the number of hospitalizations for these patients increased by over 80%. These are chronic conditions that require long-term monitoring and treatment, and commonly also re-interventions due to anatomical and functional abnormalities. Many of these patients are at high risk for the development of arrhythmia, heart failure, and endocarditis.³ In Croatia we expect to have about 10,000 such patients of different complexity.

This growing group of patients that was once exclusively under pediatric care, now requires

znači novi izazov. Potreba za adekvatnim zbrinjavanjem te, rastuće skupine bolesnika prepoznata je na svjetskoj i europskoj razini te danas postoje preporuke Europskoga kardiološkog društva o organizaciji skrbi za odrasle bolesnike s kongenitalnim srčanim greškama, kao i za edukaciju kardiologa supspecijalista za liječenje kongenitalnih srčanih grešaka u odraslih unutar Europe (engl. *Grown-up Congenital Heart Disease*; GUCH).⁴

Specijalizirani se centar treba nalaziti u okruženju u kojemu postoji adultna i pedijatrijska kardiologija, kao i kongenitalna srčana kirurgija. Nadalje, uz starenje stanovništva treba biti dostupan cjelokupni spektar medicine za odrasle. Osim kardiologa koji je specijalist za prirođene srčane bolesti u odraslih (PSBO), važni su ekspert u ehokardiografiji koji poznaje napredne tehnike i intervencijski kardiolog koji je ekspert u strukturnoj srčanoj bolesti i poznaje kompleksne intervencijske zahvate. Pristup elektrofiziologiji i elektroterapiji (elektrostimulator, kardijalna resinkronizacijska terapija, defibrilator, kompleksne ablacije) od velike je važnosti zbog aritmija, koje su u bolesnika s PSBO-om česti uzrok mortaliteta i razlog hospitalizacije. Od ostalih članova tima treba spomenuti radiologa – specijalista za slikovne metode (magnetna rezonancija i višeslojna kompjutorska tomografija) te specijalista ginekologije i porodništva za žene koje su u generativnoj dobi. Trudnoća je dodatan rizik u toj skupini bolesnika te zahtijeva savjetovanje, kao i intenzivno praćenje. Konačno, potrebna je jaka veza s transplantacijskim centrom s iskustvom u zbrinjavanju bolesnika s PSBO-om. Veliki doprinos takvom centru očekuje se i od medicinskih sestara specijaliziranih za PSBO.

Otvaranje takvih centara dokazano je dovelo do smanjenja mortaliteta, kao i morbiditeta (prije svega redukcije učestalosti hospitalizacija) u takvih bolesnika.⁵ Najbolji model skrbi jest onaj u kojemu su bolesnici barem jednom pregledani u specijaliziranom centru te gdje će supspecijalist za PSBO dati daljnje preporuke o razini skrbi i intervalima praćenja ovisno o individualnim karakteristikama bolesnika. U svakom slučaju, funkcioniranje mreže između specijaliziranog centra, lokalnoga općeg kardiološkog centra za odrasle i obiteljskog liječnika od kritične je važnosti.⁶ U Hrvatskoj je, s obzirom na očekivani broj bolesnika s PSBO-om, potreban 1 do 2 takva specijalizirana centra. Pedijatrijski kardiolozi imaju veliko znanje u kongenitalnim srčanim greškama u djece, međutim, u dugotrajnom praćenju i daljnjem liječenju odraslih uz potencijalni razvoji i stečenih srčanih bolesti njihovo je znanje ograničeno. Stoga je važan tranzicijski period u zbrinjavanju bolesnika s prirođenim srčanim greškama. Najbolji je model zajednička skrb u razdoblju od 16. do 18. godine života. U tom razdoblju pedijatri mogu specijalistu za PSBO prenijeti činjenice o dosadašnjem liječenju, a pacijent stječe povjerenje u liječnika koji će brigu o njemu voditi u sljedećem životnom razdoblju.

Unatoč činjenici da je prepoznata potreba za usavršavanjem kardiologa u području PSBO-a, većina ih ima malo iskustava. Prema europskim preporukama, edukacija u trajanju od dvije godine potrebna je za usavršavanje kardiologa supspecijalista. U tom razdoblju 18 mjeseci potrebno je provesti u specijaliziranom centru i 6 mjeseci na pedijatrijskoj kardiologiji.

the development of a new subspecialty in adult cardiology and presents new challenge in this field. The need for adequate care for this growing patient group has been recognized at the global and European levels, and guidelines have been issued by the European Society of Cardiology on the organization of care for adult patients with congenital heart disease (ACHD; or Grown-up Congenital Heart Disease, GUCH), as well as the education of cardiology subspecialists in Europe on that topic.⁴

A specialized center must be located in a place with access to adult and pediatric cardiac care, as well as congenital heart surgery. Furthermore, due to the ageing of the population the whole spectrum of adult medicine must be available. In addition to a cardiologist specializing in ACHD, having an expert in echocardiography with knowledge of advanced techniques is also important, as well as a having an interventional cardiologist with expertise in structural heart disease and knowledge of complex interventional procedures. Access to electrophysiology and electrotherapy (pacemakers, cardiac resynchronization therapy, defibrillators, complex ablation) is of great importance because of the possibility of arrhythmia, which is a common cause of mortality and hospitalization in patients with ACHD. The team must also include a radiologist – imaging method specialist (magnetic resonance imaging, multislice computed tomography) and a specialist in gynecology and obstetrics regarding the care of women in generative age. Pregnancy is an additional risk factor in this group of patients and requires advisement and careful monitoring. A large contribution to such a center can be expected from medical nurses specializing in ACHD.

Opening such centers has been shown to reduce mortality and morbidity (primarily hospitalization rates) in patients with ACHD.⁵ The best care model is that in which patients undergo at least once examinations in a specialized center where a subspecialist for ACHD can give them further recommendations on the level of care and monitoring intervals depending on individual patient characteristics. In any case, a working network between the specialized center, the local general adult cardiology center, and the family physician is of critical importance.⁶ Based on the estimated number of patients with ACHD, Croatia would need 1 to 2 such specialized centers. Pediatric cardiologists have a great knowledge and experience on congenital heart defects in children, but even their knowledge is limited when it comes to long-term monitoring and further treatment of adults, especially since that can include also the potential development of acquired heart diseases. Thus, a transitional period in the care of patients with congenital heart diseases is important. The best model is joint care in the period between 16 and 18 years of age. During that period, pediatric cardiologists can inform ACHD specialists on the course of treatment so far, and the patient will get to know and trust the physician that will be taking care of them in the next phase of their life.

Despite the fact that the need for additional education in ACHD has been recognized among cardiologists, most still have little experience. According to European recommendations, a training period of two years is needed to complete subspecialty training in GUCH. During that period, 18 months

Zaključno recimo da specijalizirani centar za PSBO mora biti supraregionalni centar koji služi kao centar izvrsnosti s najvišom razinom skrbi. Također mora omogućivati edukaciju i usavršavanje u tom području, kao i promovirati istraživanja i inovacije.⁷

must be spent in a specialized center, and 6 months at a pediatric cardiology department.

In conclusion, a specialized ACHD center must be a supra-regional center serving as a center of excellence with the highest standard of care. It must also allow education in that field and promote research and innovation.⁷

LITERATURE

1. Ávila P, Mercier LA, Dore A, Marcotte F, Mongeon FP, Ibrahim R, et al. Adult congenital heart disease: a growing epidemic. *Can J Cardiol.* 2014;30(12 Suppl):S410-9. DOI: <http://dx.doi.org/10.1016/j.cjca.2014.07.749>
2. Khairy P, Ionescu-Ittu R, Mackie AS, Abrahamowicz M, Pilote L, Marelli AJ. Changing mortality in congenital heart disease. *J Am Coll Cardiol.* 2010;56(14):1149-57. DOI: <http://dx.doi.org/10.1016/j.jacc.2010.03.085>
3. O'Leary JM, Siddiqi OK, de Ferranti S, Landzberg MJ, Opatowsky AR. The Changing Demographics of Congenital Heart Disease Hospitalizations in the United States, 1998 Through 2010. *JAMA.* 2013;309(10):984-6. DOI: <http://dx.doi.org/10.1001/jama.2013.564>
4. Webb G, Mulder BJ, Aboulhosn J, Daniels CJ, Elizari MA, Hong G, et al. The care of adults with congenital heart disease across the globe: Current assessment and future perspective: A position statement from the International Society for Adult Congenital Heart Disease (ISACHD). *Int J Cardiol.* 2015;195:326-33. DOI: <http://dx.doi.org/10.1016/j.ijcard.2015.04.230>
5. Mylotte D, Pilote L, Ionescu-Ittu R, Abrahamowicz M, Khairy P, Therrien J, et al. Specialized adult congenital heart disease care: the impact of policy on mortality. *Circulation.* 2014;129:1804-12. DOI: <http://dx.doi.org/10.1161/CIRCULATIONAHA.113.005817>
6. Orwat MI, Kempny A, Bauer U, Gatzoulis MA, Baumgartner H, Diller GP. The importance of national and international collaboration in adult congenital heart disease: A network analysis of research output. *Int J Cardiol.* 2015;195:155-62. DOI: <http://dx.doi.org/10.1016/j.ijcard.2015.05.116>
7. Baumgartner H, Budts W, Chessa M, Deanfield J, Eicken A, Holm J, et al; Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology. Recommendations for organization of care for adults with congenital heart disease and for training in the subspecialty of 'Grown-up Congenital Heart Disease' in Europe: a position paper of the Working Group on Grown-up Congenital Heart Disease of the European Society of Cardiology. *Eur Heart J.* 2014;35(11):686-90. DOI: <http://dx.doi.org/10.1093/eurheartj/ehf572>



AORTNA STENOZA

3. simpozij Radne skupine za bolesti srčanih zalistaka Hrvatskog kardiološkog društva s međunarodnim sudjelovanjem

Zagreb, hotel Westin
13.-14.05.2016.



CROVALV
RADNA SKUPINA ZA BOLESTI SRČANIH ZALISTAKA
HRVATSKOG KARDIOLOŠKOG DRUŠTVA

www.crovalv2016.org

