

■ Prirodene srčane bolesti odraslih

Adult Congenital Heart Disease



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S velikim sam zadovoljstvom prihvatila biti gost urednik ovoga broja časopisa *Cardiologia Croatica*. Broj je posvećen prirodnim srčanim bolestima odraslih. Već je nekoliko puta u ovom časopisu bilo riječi o toj temi sa stajališta epidemiologije i organizacije^{1,2}, a u ovom bih se uvodniku osvrnula na neke druge teme te upozнала hrvatsku kardiološku javnost s određenim napretkom koji smo postigli u kratkom vremenu u kojemu je problem odraslih bolesnika s prirodnim bolestima srca u Hrvatskoj osviješten.

Akronim. U svrhu lakšeg razumijevanja, u literaturi koja je uglavnom engleska, pojavljuju se dva akronima za kongenitalne srčane bolesti odraslih, a to su GUCH (*grown up congenital heart disease*), više rabljen u Sjedinjenim Državama, i ACHD (*adult congenital heart disease*), koji prevladava u Engleskoj, odnosno Europskoj uniji. Razmišljajući koji bi akronim bio prihvatljiv u hrvatskom jeziku, pri osnivanju Radne skupine za prirodene srčane bolesti Hrvatskoga kardiološkog društva, na sjednici Upravnog odbora, raspravljali smo o toj temi i suglasili se s prijedlogom PSBO (prirodene srčane bolesti odraslih). O tom prijedlogu očekujemo širu raspravu te će to biti i tema okruglog stola na našem sljedećem nacionalnom kongresu. Radi jednostavnije komunikacije u ovom ćemo broju rabiti taj akronim.

Referralni centar. S obzirom na preporuke, kako smjernica³, tako i općeg konsenzusa stručnjaka iz tog područja, ovom prilikom upoznali bismo čitatelje s činjenicom pokretanja postupka osnivanja nacionalnog centra za zbrinjavanje bolesnika s PSBO-om u Kliničkom bolničkom centru Zagreb zato što ta ustanova za sada jedina ima sve elemente potrebne za multidisciplinarni pristup takvim bolesnicima. Prema broju stanovnika, Hrvatska ih može očekivati 5 – 10 tisuća te

It was a great pleasure to accept the invitation to be the guest editor for this issue of *Cardiologia Croatica*, which is dedicated to congenital heart disease in adults. This topic has been touched upon several times already in the journal from the organizational and epidemiological perspectives^{1,2}. In this editorial, I would like to address other topics and inform the general cardiologic public in Croatia about some progress achieved over a short span of time in increasing awareness of the problem of adult patients with congenital heart disease.

Acronym. For easier comprehension, two acronyms are generally used in the predominantly English language literature: GUCH (*grown-up congenital heart disease*), more common in the USA, and ACHD (*adult congenital heart disease*), more common in the UK and European Union. While deliberating on the best acronym to use in the Croatian language during the formation of the Working Group for Congenital Heart Disease of the Croatian Cardiac Society, we discussed this topic at the meeting of the Management Board and agreed on the acronym PSBO ("prirodene srčane bolesti odraslih" – congenital heart disease in adults). We expect more discussion on this topic and that this will be one of the points addressed at the round-table meeting at our next national congress. For easier comprehension, the Croatian language version of this Journal's issue will be using PSBO as an acronym.

Referral Center. Given the recommendations in relevant guidelines³ as well as from the general consensus of experts in this field, we would like to announce the beginning of efforts to found a national care center for patients with ACHD at the University Hospital Centre Zagreb, since this institution is currently the only one with all the ele-

će imati potrebe za jednim do dva takva centra. U kratkome članku u ovom broju opisana su glavna obilježja ovakvog centra, a posebno treba naglasiti da bi se svi bolesnici s PSBO-om trebali registrirati u njemu, obaviti prvu evaluaciju, a da će se većina nastaviti liječiti u lokalnim ustanovama, osim u slučaju iznimne kompleksnosti ili iznimnih situacija.

Registar. Veoma je važno i osnivanje registra. I iz drugih područja kardiologije znamo koliko nam nedostaju nacionalni podatci, i ne jednom smo o tome raspravljali (registar akutnoga koronarnog sindroma, registar perkutanih koronarnih intervencija i sl.). S obzirom na relativno ograničen broj bolesnika s PSBO-om, pruža nam se jedinstvena prilika da prvi put u Hrvatskoj imamo pravi registar, a u ovoj kategoriji on je veoma važan, jer je riječ o tzv. cjeloživotnim pacijenatima, koji se međusobno razlikuju unatoč možda istoj dijagnozi i u kojih je i sama bolest, ali i liječenje kompleksno. Registar bi svima koji dolazimo u dodir s njima znatno olakšao međusobnu komunikaciju i liječenje. Unatoč velikome trudu registar, koji vodimo u KBC-u Zagreb nešto više od godinu dana, za sada obuhvaća tek nešto više od 350 bolesnika. Nadamo se da će ovaj, tematski broj potaknuti razmjenu podataka.

Edukacija. Opće je poznato da su kardiolozi slabije obrazovani kada je posrijedi prirođena srčana bolest jer je većina nas educirana u vrijeme kada je potreba za liječenjem ovakvih bolesnika bila mala. Iako kasnimo za razvijenim zemljama u zbrinjavanju bolesnika s PSBO-om, isti je problem prisutan i tamo te se u smjernicama⁴, ali i u raznim člancima govori o kurikulu obrazovanja kardiologa subspecialista za PSBO. Smatramo da je važno da neki naši kolege završe edukaciju koju preporučuje Europsko kardiološko društvo, ali je još važnije da svi mi postanemo svjesni problema da takve bolesnike prepoznamo, da ih pravilno usmjerimo te da se dalje brinemo za one niže ili srednje kompleksnosti, pružajući im potrebnu razinu skrbi, bilo u svojoj ustanovi bilo upućujući ih u specijalizirani centar za PSBO. Kao mali doprinos u postizanju tog cilja željeli smo izdati i tematski broj časopisa.

U ovom su broju tiskani članci koji sadržavaju teme iznesene na I. sastanku Radne skupine za PSBO Hrvatskoga kardiološkog društva, održanom 12. studenoga 2015. u Hrvatskoj kući srca u Zagrebu, uz neočekivano veliki interes te sa zadovoljstvom mogu istaknuti da je dvorana bila puna tijekom cijelog jednodnevnog sastanka. Nadamo se da će teme iz ovog broja časopisa *Cardiologia Croatica* zainteresirati čitatelje, a iznesene su kao pregledni članci, prikazi slučajeva i stručni članci te da će prenijeti informacije i poruke koje smo tijekom sastanka odaslali.

ments needed for a multidisciplinary approach to these patients. Based on demographics, Croatia can be expected to have 5-10 thousand such patients, and will need one or two such centers. A brief article in this issue describes the main characteristics of such a center. It is especially important to note that all patients with ACHD should register and receive the first evaluation at the center, after which they will continue treatment in their local institutions except in extremely complex or extraordinary cases.

Registry. Establishing a patient registry is very important as well. Other cardiologic issues have shown how badly we lack national data, a problem that has been discussed several times (acute coronary syndrome registry, percutaneous coronary intervention registry, etc.). Due to the relatively limited number of patients with ACHD, we are presented with a unique opportunity to create a real registry for the first time in Croatia, which is especially important for ACHD, since these are "lifelong" patients with a complex disease that requires complex treatment due to differences in clinical presentation even under the same diagnosis. A registry would significantly improve communication among all physicians coming into contact with these patients. Despite great effort, the registry currently contains just over 350 patients we have had at the University Hospital Centre Zagreb for over a year. It is our hope that this issue of our Journal will encourage data-sharing.

Education. It is well known that cardiologists lack education on congenital heart diseases, since most of us received our degrees at a time when the need for treatment of these diseases was small. Although we are behind other countries in the organization of care for patients with ACHD, the same problem is present in those countries as well, and guidelines⁴ as well as other articles discuss the need for the education of cardiologists, subspecialists for ACHD. We believe it is important that some of our colleagues complete courses recommended by the European Society of Cardiology, but that it is even more important that we all become aware of the problem, recognize the patients, advise them appropriately, and treat cases of lower or moderate complexity by providing them with an adequate level of care either in our own institutions or by sending them to specialized ACHD centers. As a small contribution to this goal, we decided to dedicate this Journal issue to that topic.

This Journal issue contains articles addressing topics discussed at the 1st meeting of the ACHD Working Group of the Croatian Cardiac Society, which took place in the Croatian Heart House in Zagreb on November 12, 2015 and was met with great interest. I can say with pleasure that the conference hall was full during the whole one-day meeting. We hope that readers will find the topics addressed in this issue of *Cardiologia Croatica* to be interesting reading; they are presented as review articles, case reports, and professional articles and are intended to convey the information and conclusions we arrived at during the meeting.

LITERATURE

1. Strozzi M, Brida M, Anić D. Adult Congenital Heart Disease Treatment Problems in Croatia: Organization in a Tertiary Center and Report of Three Cases. *Cardiol Croat.* 2015;10(1-2):3-10. DOI: <http://dx.doi.org/10.15836/ccar.2015.3>
2. Šeparović Hanževački J, Malčić I, Ivanac Vranešić I. Congenital heart diseases in Croatia – a review of current state and goals. *Cardiol Croat.* 2012;7(11-12):276-82.
3. 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/eh572>
4. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, et al; Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPIC); ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J.* 2010;31(23):2915-57. DOI: <http://dx.doi.org/10.1093/eurheartj/ehq249>