Ehokardiografski pokazatelji jednostavnih prirođenih srčanih bolesti odraslih

Echocardiographic Indicators of Simple Adult Congenital Heart Diseases

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SAŽETAK: Prirođene srčane bolesti odraslih (PSBO), osim složenih srčanih grešaka otkrivenih u djetinjstvu, obuhvaćaju i sasvim jednostavne jednostruke srčane greške koje se zbog maloga hemodinamskog značenja i težine otkrivaju tek u odrasloj dobi. Pokazatelji jednostavnih PSBO-a na lijevome srcu i aorti, znakovi su tlačno i volumno remodelirane lijeve klijetke (LK) te patološke morfologije zalistaka (najčešće je to dvolisni aortni zalistak – BAV i miksomatozni mitralni zalistak), kao i koarktacije aorte. Volumno opterećenje LK zbog povećanog utoka u lijevo srce dovodi do ekscentrične hipertrofije LK (normalna debljina stijenki, povećani volumeni, kuglasti oblik) razmjerno težini hemodinamske greške, pa što je volumen regurgitiranja ili šanta veći, uvećanje je veće. U BAV-u su česta i proširenja ascendentne aorte. Tlačno opterećena LK remodelirana je po tipu koncentrične hipertrofije (malog volumena u sistoli i dijastoli, zadebljanih stijenki). Najčešći prirođeni uzroci tlačnog opterećenja LK jesu: prirođena aortna stenoza, subaortna stenoza (izolirani subaortni fibrozni prsten i subaortna membrana) te koarktacija aorte (češće povezana s BAV-om) te stanja nakon prethodnih operativnih zahvata. Osim veličine šupljina i debljine stijenki, lijevog i desnog srca, važan je i dinamičan odnos razlike tlakova tijekom srčanog ciklusa. Kada postoji poremećaj tog odnosa, dolazi do pomaka ventrikulskog ili interatrijskog septuma prema šupljini s manjim tlakom. U tlačnom opterećenju desne klijetke (DK) dolazi do pomaka septuma prema lijevo u sistoli i stvaranje tzv. D-oblika LK u sistoli. U volumnom opterećenju DK dolazi do pomaka septuma prema lijevo u dijastoli tzv. „D-oblik" LK u dijastoli. U zaključku možemo istaknuti da se ehokardiografski pokazatelji jednostavnih i manje teških PSBO-a mogu svrstati u pet osnovnih kategorija: 1. veličina i morfologija LK; 2. veličina desnog atrija, položaj interatrijskog septuma; 3. veličina i mofologija DK; 4. plućna hipertenzija, 5. gradijent tlaka protoka preko aortnog i plućnog zalistka (AV/LVOT i PV/RVOT). Nalaz bilo kojeg od gore navedenih pokazatelja (s kardiološkim simptomima i znakovima ili bez njih) treba pobuditi sumnju na postojanje PSBO-a, što potom treba dokazati ili isključiti.

SUMMARY: Adult congenital heart diseases (ACHD), in addition to complex heart defect diagnosed in childhood, also include very simple singular heat defects that are diagnosed only in adulthood due to their minor hemodynamic influence and severity. The indicators of simple ACHD on the left side of the heart and the aorta are signs the left ventricle (LV) has been remodeled by volume, pathological morphology of the valve (most commonly bicuspid aortic valve (BAV) disease and myxomatous mitral valve disease), and coarctation of the aorta. Volume load of the LV due to increased inflow into the left side of the heart leads to eccentric hypertrophy of the LV (normal wall thickness, increased volume, and ball-like shape) proportionate to the severity of the hemodynamic defect – the greater the regurgitation or shunt volume, the greater the dilation of the heart. Dilatation of the ascending aorta is also common in BAV. Volume load on the LV causes concentric hypertrophy (low systolic and diastolic volume, thickened walls). The most common natural causes of volume load on the LV are: congenital aortic stenosis, subaortic stenosis (isolated subaortic fibrous ring and subaortic membrane), coarctation of the aorta (more often associated with BAV), and conditions after previous surgical procedures. Other than the size of the cavities and the thickness of the walls and the left and right heart, the dynamic relationship of pressure changes during the cardiac cycle is important as well. When this relationship is disrupted, the ventricular or inter-atrial septum is displaced towards the area of lower pressure. Pressure load of the right ventricle (RV) causes the septum to be displaced towards the left in the systole and create a so-called D-shaped left ventricle. Volume load of the RV causes the septum to be displaced to the left in the diastole, causing a so-called D-shaped left ventricle in the diastole. In conclusion, we can group the echocardiographic indicators of simple and less severe ACHD into 5 basic categories: 1) size and morphology of the LV; 2) size of the right atrium, position of the inter-atrial septum; 3) size and morphology of the RV; 4) pulmonary hypertension; 5) flow pressure gradient across the aortic and pulmonary valve (AV/LVOT and PV/RVOT). Finding any of these indicators (with or without

accompanying cardiologic signs and symptoms) should raise suspicion of the existence of ACHD, which should then be established or eliminated from the differential diagnosis.

KLJUČNE RIJEČI: ehokardiografija, prirođene srčane bolesti, volumno opterećenje, tlačno opterećenje.

KEYWORDS: echocardiography, adult congenital heart disease, volume overload, pressure overload.

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Prirođene srčane bolesti odraslih (PSBO) poseban su klinički problem u kardiologiji koji se najčešće poistovjećuje sa složenim morfološkim i višestrukim funkcijskim greškama, potpuno ili djelomično operativno riješenima u djetinjstvu.[1] Međutim, PSBO obuhvaćaju i sasvim jednostavne jednostruke srčane greške koje se zbog maloga hemodinamskog značenja i težine otkrivaju tek u odrasloj dobi. Čak i u svojoj jednostavnosti neke od tih bolesti vrlo često su kliničko-ehokardiografski zahtjevne i rijetko se navrijeme dijagnosticiraju. Osim niske učestalosti, nekoliko je razloga tomu. Morfološki jednostavne greške povezane su s malo ili nimalo simptoma i znakova bolesti pa stoga potreba za aktivnim traženjem greške izostaje.[2] Nedovoljno poznavanje ehokardiografskih pokazatelja za PSBO i nedostatni ehokardiografski pregledi rezultiraju čestim neprepoznavanjem simptomatske PSBO, a asimptomatski bolesnici i unatoč rutinskim sistematskim pregledima ostaju neotkriveni. Transtorakalna ehokardiografija često je sasvim dovoljna u otkrivanju i postavljanju dijagnoze jednostavnih PSBO-a pod uvjetom da se izvodi u skladu s preporukama za standardizirani protokolirani pregled.[3] Ovaj članak upućuje na morfološke i hemodinamske pokazatelje jednostavnih i manje teških PSBO-a koji se trebaju uočiti tijekom ehokardiografskoga pregleda ili probiranja ove skupine bolesnika. S obzirom na široku morfološku različitost PSBO-a, njihovi daljnji specifični dijagnostički pristupi nisu obuhvaćeni u ovome članku.

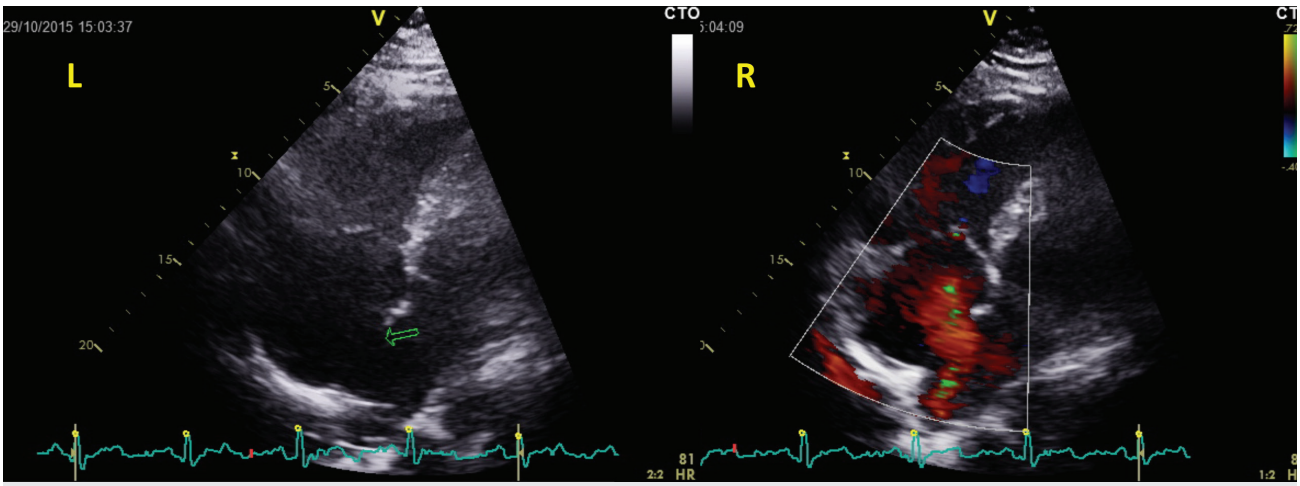
Primjer 1.

Bolesnica sa zaduhom u naporu unatrag godinu dana, klasifikacije tegoba NYHA I. U medicinskoj se dokumentaciji pronalazi u jednom navratu zabilježen sistolički šum nad bazom, koji kasnije u nalazima nije opisivan. S obzirom na progredirajuće tegobe, više puta učinjene pretrage (dinamički elektrokardiogram, test opterećenja te transtorakalna ehokardiografija u dvama laboratorijima, pulmološki pregled) nisu objasnile uzrok zaduhe. Tijekom ponovljenog ehokardiografskog pregleda utvrđeni su umjereno proširenje desne pretklijetke i blago proširenje desne klijete sa znakovima volumnoga hemodinamskog opterećenja. U aktivnom traženju uzroka volumnom opterećenju desne strane srca nađe se široka komunikacija na razini interatrijskog septuma (**slika 1**) koja nije bila primijećena u prethodnim nalazima, vjerojatno zbog vrlo slabo izražene turbulencije šanta na tako širokom otvoru (**slika 1**). Protoci preko mitralnog ušća, kao i utok iz plućnih vena bili su povećanih brzina kao odraz značajnoga lijevo-desnog šanta.

Adult congenital heart diseases (ACHD) represent a special clinical problem in cardiology that is typically seen as including complex morphological and multiple functional defects, which are fully or partially operable in childhood.[1] However, ACHD also includes very simple singular heart defects that are diagnosed only in adulthood due to their minor hemodynamic influence and severity. Even these simple diseases, however, are often clinically and echocardiographically demanding and are rarely diagnosed in a timely manner. Other than the low incidence, there are several reasons for this. Morphologically simple defects are often associated with few or no signs and symptoms, so no active searching for the defect takes place.[2] Inadequate knowledge of echocardiographic indicators of ACHD and incomplete echocardiographic examinations result in symptomatic ACHD being overlooked and asymptomatic patients remaining undiagnosed despite routine systematic check-ups. Transthoracic echocardiography is often sufficient to discover and diagnose simple ACHD provided it is performed according to the guidelines for standardized check-up protocols.[3] This article describes the morphological and hemodynamic indicators of simple and less severe ACHD that should be noted during echocardiographic examination or screening of this group of patients. Due to the morphological variety of ACHD, specific diagnostic follow-up for ACHD variants are beyond the scope of this article.

Example 1

A female patient presented with shortness of breath during exertion a year before admission, in class I of the New York Heart Association (NYHA) Functional Classification. Medical documentation showed that a systolic murmur over the base of the heart had been found at an earlier point, but not mentioned again. Since her issues kept progressing, repeated examinations were performed (dynamic electrocardiogram, exercise stress test, and transthoracic echocardiography in two laboratories, as well as a pulmonology exam) but did not identify the cause of the dyspnea. Repeating the echocardiographic imaging established moderate dilation of the right atrium and mild dilation of the right ventricle with signs of volume hemodynamic load. While actively searching for the volume load of the right side of the heart, broad communication was found at the level of the inter-atrial septum (**Figure 1**), which had not been noticed during previous echocardiographic exam likely due to weak shunt turbulence in such a large opening (**Figure 1**). Flow across the mitral and inflow



Broad communication at the level of the inter-atrial septum (arrow) – atrial septal opening in an apical 4 chamber view (L). A significant left-to-right shunt at the atrial level in color Doppler imaging, with small turbulence at the opening.

FIGURE 1.

Ovaj nas primjer upozorava nam na nekoliko bitnih načela u kliničkom prosuđivanju i dijagnosticiranju jednostavnih PSBO-a.

Tegobe bolesnika i njihovo detaljno ispitivanje, kao i klinički pregled s posebnim naglaskom na auskultaciju srca trebali bi biti neizostavni i vodeći dio pristupa bolesniku tijekom ehokardiografskog pregleda. Ako u prvom susretu s bolesnikom propustimo pronaći vodeće tegobe i srčane znakove, to nas može odvesti u krivom smjeru i u dugotrajnu nesvrsishodnu dijagnostičku obradu. Nakon anamneze i pregleda postavljamo radnu dijagnozu ili kliničko pitanje, odnosno sumnju postoji li srčana greška u odraslog bolesnika.

Ehokardiografski postupnik

Ehokardiografija je prva slikovna metoda kojom se koristimo u dijagnostičkom postupniku. Ona nam treba odgovoriti na postavljenu sumnju i kliničko pitanje.

Transtorakalna ehokardiografija razvija se već desetljećima u kompleksnu morfološku analizu i hemodinamsko ispitivanje funkcije srca koristeći se svim novim suvremenim metodama. Ova nam pretraga služi u postavljanju točne dijagnoze, tj. potvrde dijagnoze, kao i u praćenju bolesnika nakon izvedenih jednostavnih ili složenih operativnih zahvata.

Klinička ehokardiografija bit će svrsishodna ako se vodimo nekim bitnim osnovnim načelima.

1. Standardizirani protokol i arhiviranje zapisa. Standardizirani protokol podrazumijeva da se tijekom pregleda pridržavamo točnog redoslijeda i sadržaja što i kako prikazati i arhivirati da bi pregled bio potpun i sadržavao sve potrebne elemente za kasniju analizu i mjerenja koji nas vode do konačne dijagnoze.[2-4] Ehokardiografski je protokol propisan smjernicama i preporukama Europskog udruženja za kardiovaskularno oslikavanje (EACVI prema engl. *European Association of Cardiovascular Imaging*). U bolesnika s PSBO-

from the pulmonary veins were increased in velocity as a result of a significant left-to-right shunt.

This example illustrates several important principles in the clinical assessment and diagnosis of simple ACHD.

Examining patient complaints in detail and a clinical examination focusing on auscultation of the heart should be integral and primary elements of echocardiography examinations. If we fail to find the most important issues and cardiac signs during the first contact with the patient, we can be led astray into long and ineffective diagnostic processing. After patient history and physical examination, a tentative diagnosis or clinical question should be put forward, i.e. whether a heart defect is suspected in the adult patient.

Echocardiographic procedures

Echocardiography is the primary imaging method used in diagnostic procedures to answer the suspicions and clinical questions we are dealing with.

Transthoracic echocardiography has been in development for decades and has grown into a complex morphological analysis and hemodynamic testing of the function of the heart through the use of all new diagnostic methods. This test allows us to give an accurate diagnosis, i.e. establish it, as well as follow up the patient after simple or complex surgical procedures.

Clinical echocardiography will be fit for purpose if we are guided by the following basic principles.

1. Standardized protocol and data archiving. Standardized protocols mean that the physician will perform examination while carefully adhering to the correct order and content with regard to what to image and archive, so as to ensure the examination is complete and contains all necessary elements for later analysis, as well as measurements that lead to the establishment of a diagnosis.[2-4] Echocardiographic protocols

om posebno je važno držati se preporuka za mjerenje desne klijetke (DK) i desne pretklijetke (DA) i za procjenu funkcije desnog srca te provoditi strukturiranu analizu morfologije srčanih šupljina.[5,6]

2. Aktivni pristup u traženju greške. Tijekom pregleda očekujemo objašnjenje i odgovor na kliničko pitanje koje smo sebi postavili pa, ako na njega nismo našli odgovor iz ehokardiografskog preliminarnog nalaza, potrebno je protokol dopuniti novim prikazima koji bi odgovarali očekivanoj grešci (npr. neobjašnjen patološki šum – potrebno je prikazati subaortni dio membranoznog septuma i tražiti mali ventikulski septalni defekt itd.).[7]

3. Razumijevanje neočekivanih mjerenja. Kada tijekom ehokardiografskog pregleda izmjerimo neočekivano visoke ili niske brzine protoka, proširene šupljine i sl., navedeni je pokazatelj potrebno objasniti i dokazati njegovo postojanje u logičnome slijedu kliničkog pitanja i odgovora (npr. sistolički šum nad Erbom ne može se objasniti proširenom aortom i sl., turbulencija u izgonskome traktu desne klijetke nije patološki šant ako nismo dokazali povećane protoke preko pulmonalnog zalistka).

4. Praćenje s vremenskom odgodom. U donošenju odluke o daljnjem postupku s bolesnikom pokatkad je potrebno utvrditi kakvu progresiju ima određena greška. Na osnovi jednog pregleda to nije moguće pa je stoga potrebno odgoditi odluku na određeno vrijeme u kojem se očekuje promjena, u skladu sa smjernicama Europskog kardiološkog društva (ESC) i EACVI-a.[8-11]

Nasuprot kompleksnoj ehokardiografiji je ehoskopija tzv. FOCUS (engl. *Focused cardiac ultrasound*) pregled. Njime se koristimo kao površnim jednostavnim pogledom na srce u hitnoj službi. To je orijentacijski pregled kojim se isključuje srčana bolest kao uzrok hitnoga stanja. Ovaj pregled usmjereno odgovara s da/ne na nekoliko pitanja (npr. sistolička disfunkcija – da/ne, perikardni izljev – da/ne, valvularna greška – da/ne). Za to služe vrlo mali jednostavni uređaji veličine dlana i skromnih vizualnih mogućnosti. Vrlo su korisni u bolesnika s poznatim PSBO-om koji se prezentiraju u hitnoj službi s kardiološkim tegobama.[9]

LIJEVO SRCE I AORTA

Pokazatelji jednostavnih PSBO-a na lijevom srcu i aorti znakovi su tlačno i volumno remodelirane lijeve klijetke (LK) te patološka morfologija zalistaka (najčešće je to dvolisni aortni zalistak i miksomatozni mitralni zalistak) te koarktacija aorte. Izolirano uvećanje lijeve pretklijetke (LA) najčešće nije specifičan nalaz kod PSBO-a.

a) Volumno opterećenje lijeve klijetke

Povećani utok u lijevo srce dovodi do ekscentrične hipertrofije LK (normalna debljina stijenki, povećani volumeni, kuglasti oblik) razmjeran težini hemodinamske greške, što je volumen regurgitiranja ili šanta veći, uvećanje je veće. U blažim i jednostavnim PSBO-ima koji dovode do volumnog opterećenja, sistolička i dijastolička funkcija LK dugo ostaju neoštećene, a bolesnici imaju vrlo malo tegoba. Stoga se pri ehokardiografskom pregledu i u praćenju bolesnika treba pomno procijeniti hemodinamsko opterećenje i funkciju te pravodobno uputiti

are based on the guidelines and recommendations of the European Association of Cardiovascular Imaging (EACVI). In examining patients with ACHD, it is especially important to adhere to the guidelines for the measurement of the right ventricle (RV) and right atrium (RA), assessment of the function of the right side of the heart, and the structured analysis of the morphology of the cavities of the heart.[5,6]

2. Actively searching for errors. During the course of the examination, we expect to find an answer and explanation for the clinical question we have asked ourselves; if the answer is not forthcoming based on the preliminary results, the protocols must be expanded with new images that correspond to the expected error (e.g. in case of an unexplained pathological murmur – imaging of the subaortic part the membranous septum and looking for a small ventricular septal defect, etc.).[7]

3. Understanding unexpected measurements. During echocardiographic examination, if we measure unexpectedly high or low flow rates, dilated cavities, etc., it is necessary to explain those clinical signs and explain their existence logically, based on clinical questions and answers (e.g. systolic murmur over the Erb's point cannot be explained with a dilated aorta, turbulence in the outflow tract of the right ventricle is not a pathological shunt if we have not established increased flow over the pulmonary valve, etc.).

4. Follow-up with time delay. When deciding the further course of treatment, it is sometimes necessary to determine the progression on a specific defect. This cannot be done in a single examination, so decisions have to be postponed for a time sufficient for changes to be noted, in line with the guidelines of the European Society of Cardiology (ESC) and EACVI.[8-11]

Contrasting with the complexities of echocardiography is the so called FOCUS (focused cardiac ultrasound) examination. It is used as a simple, surface look at the heart for emergency services. It is an orientation examination used to eliminate heart disease as a cause of the emergency. The examination gives a clear yes/no answer to several questions (e.g. systolic dysfunction – yes/no; pericardial effusion – yes/no; valvular defect – yes/no). Simple, palm-sized devices with low imagining power are used. These examinations are very useful in patients with known ACHD that present to emergency service with cardiac issues.[9]

LEFT HEART AND THE AORTA

ACHD indicators in the left heart and the aorta are signs of volume and pressure remodeling of the left ventricle (LV), the pathologic morphology of the valve (usually a bicuspid aortic valve and myxomatous mitral valve), and coarctation of the aorta. If isolated, dilation of the left atrium (LA) is usually not specific to ACHD.

a) Volume load to the left ventricle

Increased inflow to the left heart leads to eccentric hypertrophy of the LV (normal wall thickness, increase volume, ball-shaped) in proportion to the severity of the hemodynamic defect – greater regurgitation volume or a larger shunt leads to more dilation of the ventricle. In milder and simpler ACHD that can lead to volume load, the systolic and diastolic func-

na potrebne zahvate kako se ne bi propustila mogućnost pozitivnog remodeliranja miokarda u cijelosti.[8,11] Najčešći razlozi volumnog opterećenja LK u PSBO-u:

- aortna regurgitacija (prirođena morfološka patologija aortne valvule)
- mitralna regurgitacija (miksomatozni mitralni zalistak s prolapsom, neprepoznati atrio-ventriukularni kanal)
- ductus Botalli (spoj između silazne aorte i lijeve pulmonalne arterije /pumonalnoga stabla kada manji do umjereni šant dovodi do opterećenja samo LK i LA).

Prolaps mitralnog zalistka s regurgitacijom lako se otkrije i često nije dijagnostički problem. Međutim, dvolisni aortni zalistak treba aktivno tražiti ako je uzrok volumnog opterećenja aortna regurgitacija. Osobitu pozornost treba usmjeriti prema traženju ductusa Botalli (neobjašnjenom uvećanju LK s normalnom funkcijom i sistoličko-dijastoličkim šumom). U parasternalnom poprečnom presjeku nad bazom srca treba tražiti turbulentni mlaz sistoličko-dijastoličkog šanta u stablu plućne arterije ili na njenom grananju. On se dokazuje zapisom kontinuiranog doplera s velikim vršnim brzinama u sistoli i manjim u dijastoli (što je karakteristično za manje spojeve koji ne dovode do plućne hipertenzije).[1,7]

Dvolisni aortni zalistak i proširenje uzlazne aorte

Tijekom ehokardiografskog pregleda prikazujemo morfologiju aortnog zalistka iz više prikaza (uzdužni i poprečni parasternalni) kako bismo dokazali prisutnost triju listića, ali i oblik otvaranja. Često tek iz oblika otvorenih listića potvrđujemo sumnju na dvolisni aortni zalistak (BAV, prema engl. *bicuspid aortic valve*). Naime, otvoreni dvolisni zalistak čini elipsoidni otvor usmjeren uzdužno s dvjema polaznim točkama na komisurama (**slika 2**). Dvolisni zalistak može biti suženog otvora te je potrebno utvrditi hemodinamski značajnu stenozu (napomena: uvijek mjerimo i brzine u izgonskome traktu LK i uvrštavmo u računanje gradijenta ako su brzine veće od 1 m/s). Osim suženja, BAV često ima i nedovoljnu

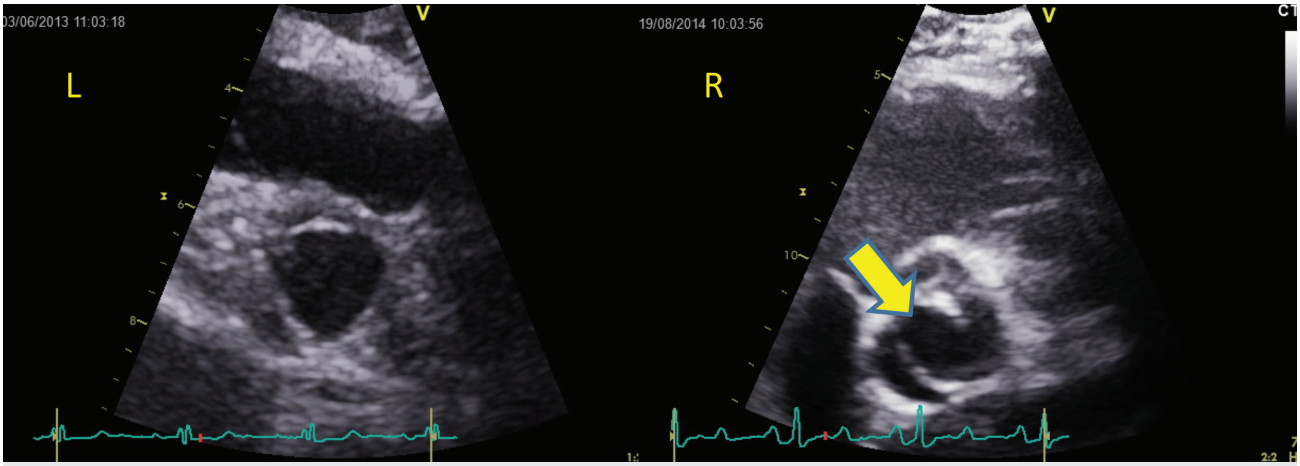
tions of the LV stay undamaged for a long time, and patients have very few issues. Thus it is important to assess the hemodynamic load and function during echocardiographic examination and monitoring, and make timely treatment decisions to avoid missing opportunities for full myocardial remodelling.[8,11] The most common causes of volume load to the LV in ACHD cases are:

- Aortic regurgitation (congenital morphologic pathology of the aortic valve)
- Mitral regurgitation (myxomatous mitral valve disease, unrecognized atrioventricular channel)
- Ductus Botalli (the connection between the descending aorta and the left pulmonary artery/pulmonary trunk when a small to medium shunt creates load to LV and LA).

Mitral valve prolapse with regurgitation is easy to diagnose and is rarely a diagnostic problem. However, a bicuspid valve must be actively searched for if it is causing volume load and aortic regurgitation. Special care should be taken to look for signs of ductus Botalli (the unexplained dilation of the LV with normal function and systolic-diastolic murmur). Parasternal transverse section over the base of the heart should be used in checking for the turbulent flow of the systolic-to-diastolic shunt in the pulmonary truncus or its branching. This is established by continuous wave Doppler imaging with ceiling velocities in the systole and lower velocities in the diastole (which is characteristic of smaller connections that do not lead to pulmonary hypertension).[1,7]

Bicuspid aortic valve and dilation of the ascending aorta

Apical views of the several planes are needed to establish the presence of three leaflets, but also the way they open. Often only the shape of the open leaflets confirms the suspected bicuspid aortic valve (BAV). Namely, an open bicuspid valve makes an ellipsoid opening directed longitudinally, with two points of origin from the commissures (**Figure 2**). A bicuspid



Tricuspid aortic valve open during systole – triangular opening (L), bicuspid aortic valve open during systole – ellipsoid opening (R). Cross section at the base of the heart using the parasternal view.

FIGURE 2.

koaptaciju s posljedičnom regurgitacijom. Težinu regurgitacije utvrđujemo objektivnim mjerenjima mlaza i određivanjem volumena regurgitacije u skladu s preporukama EACVI-a.[11] Obojeni dopler koristi nam samo u otkrivanju regurgitacije te pri utvrđivanju smjera, dok u procjeni težini greške prednost dajemo objektivnim mjerenjima.

Posebnu pozornost potrebno je posvetiti otkrivanju proširenja uzlazne aorte i njezinu praćenju na kontrolama. Uzlaznu aortu mjerimo u uzdužnome parasternalnom prikazu (LAX), pri čemu os mjerenja treba biti okomita na obje stijenke kako ne bismo dobili lažno veća mjerenja zbog dijagonalnoga presjeka. Također treba imati na umu da u nekim slučajevima smjer uzlazne aorte nije paralelan s prikazom te mjerenja mogu odstupati pa ih je stoga potrebno potvrditi iz najmanje dvaju prikaza (osim LAX-om, koristiti se i poprečnim parasternalnim ili jugularnim). Ako nema znatnog proširenja i hemodinamske greške, dovoljne su kontrole svake 2 – 3 godine.[12]

b) Tlačno opterećenje lijeve klijetke

Tlačno opterećena LK remodelirana je po tipu koncentrične hipertrofije (malog volumena u sistoli i dijastoli, zadebljanih stijenki).

Najčešći prirođeni uzroci tlačnog opterećenja LK koje treba tražiti jesu:

- prirođena aortna stenoza (najčešće BAV) (vidjeti gore)
- subaortna stenoza (izolirani subaortni fibrozni prsten, subaortna membrana)
- koarktacija aorte (češće povezana s BAV-om)
- stanja nakon prethodnih operativnih zahvata.

Na subaortnu stenozu treba posumnjati kada se turbulentni sistolički protok nalazi ispod granice aortnog zalistka. On se dokazuje velikim brzinama u sistoli mjerenim kontinuiranim doplerom u izgonskome traktu iz apikalnog prikaza pet šupljina. U procjeni težine stenoze izgonskoga trakta ne smijemo izostaviti i procjenu aortnog zalistka jer stenoze mogu biti višestruke.[11]

Koarktacija aorte koja se otkriva u odrasloj dobi manje su teške stenoze silazne aorte ili imaju znatna suženja, ali s obilnom kolateralnom cirkulacijom. Bolesnici s arterijskom hipertenzijom i znakovima lošije perfuzije donjih ekstremiteta te tlačnim opeterećenjem LK moraju pobuditi sumnju na koarktaciju. Jedna od manifestacija mogu biti i disekcija aorte, endokarditis i hemoragijski moždani udar. Suženje silazne aorte nalazi se najćešće na mjestu pripoja arterijskog voda (ispod potključne arterije) i obuhvaća različite forme suženja (hipoplastični silazni dio luka ili preduktalna, paraduktalna i postduktalna). Možemo je prikazati suprasternalnim, tj. jugularnim prikazom, u kojem vidimo morfološki mjesto i oblik suženja te turbulentni protok.[7,8,12] Suženje dokazujemo velikim brzinama protoka na tom mjestu, dok za dokaz teškog suženja, osim visoke vršne brzine, trebamo imati i tipičnu repnu turbulenciju u dijastoli (pod uvjetom sačuvane rastegljivosti stijenke distalne aorte). Jednako tako, gradijent preko suženja može biti lažno smanjen ako je kolateralna cirkulacija obilno razvijena.[10]

valve can result in a smaller passage, so it is necessary to determine whether there is a presence of hemodynamically significant stenosis (note: we also always measure LV outflow and include them in gradient calculations if the velocity is above m/s). In addition to stenotic lesion, BAV often results in insufficient coaptation and consequent regurgitation. The severity of the regurgitation is determined objectively by measuring the jet and determining the regurgitation volume, in line with the EACVI guidelines.[11] Colour Doppler flow imaging is only used to detect the regurgitation and direction; we favor objective measurements in assessing the severity of the defect.

Special attention should be given to discovering possible dilation of the ascending aorta and its monitoring in follow-up. The ascending aorta is measured with the aortic arch long axis (LAX) view, in which the axis of measurement must be perpendicular to both walls so as to avoid overestimated measurements due to the diagonal cross section. It should also be noted that the direction of the ascending aorta is in some cases not parallel to the image and the measurements can be incorrect. They should thus be verified using at least two views (using short axis and jugular views in addition to LAX). If there are no significant dilation and hemodynamic defects, follow-up every 2-3 years is sufficient.[12]

b) Pressure load to the left ventricle

A pressure-loaded LV undergoes concentric hypertrophy remodeling (low volume in the systole and diastole, thickened walls).

The most common congenital causes of pressure load to the LV that should be suspected are:

- Congenital aortic stenosis (most commonly BAV) (see above)
- Subaortic stenosis (an isolated subaortic fibrous ring, subaortic membrane)
- Coarctation of the aorta (often associated with BAV)
- The result of previous surgical procedures

Subaortic stenosis should be suspected when the turbulent systolic flow is below the aortic valve. This is demonstrated by high velocities in the systole, measured in the apical view of five chambers by continuous Doppler in the left ventricular outflow tract (LVOT). When assessing the severity of the stenosis, we must also take into consideration the assessment of the aortic valve, since multiple stenoses are possible.[11]

Coarctation of the aorta diagnosed only in adulthood is usually a less severe stenosis of the descending aorta or has significant constriction but abundant collateral circulation. Coarctation should be suspected in patients with arterial hypertension and signs of poor peripheral perfusion and volume load to the LV. Other manifestations can be the dissection of the aorta, endocarditis, and hemorrhagic stroke. Constriction of the descending aorta most commonly is seen at the joint of the arterial ductus (below the subclavian artery) and includes various forms of constriction (hypoplastic descending part of the arch or pre-, para-, and postductal constriction). It can be seen on the suprasternal, i.e. jugular view, which shows the morphological location and type of constriction as well

DESNO SRCE

Tijekom pregleda bolesnika sa sumnjom ili u probiranju na jednostavnu PSBO posebnu pozornost potrebno je posvetiti desnom srcu. Pokazatelji tlačnog ili volumnog opterećenja (npr. povećana šupljina DA ili DK, hipertrofija DK i sl.) upućuju na grešku koju treba detaljnije tražiti.

a) Tlačno opterećenje

Osim veličine šupljina i debljine stijenki, lijevog i desnog srca važan je i dinamičan odnos razlike tlakova tijekom srčanog ciklusa. Kada postoji poremećaj tog odnosa, dolazi do pomaka ventrikulskog ili interatrijskog septuma prema šupljini s manjim tlakom.[1,7]

U **tlačnom opterećenju** DK dolazi do pomaka septuma prema lijevo u sistoli i stvaranje tzv. D-oblika LK u sistoli (**slika 3**). Tijekom dugotrajnijega tlačnog opterećenja nastaje i koncentrična preobrazba DK. Ona uključuje debljinu stijenke veću od 0,5 mm u dijastoli (na razini kordi trikuspidnog zalistka u supkostalnom prikazu (**slika 4**) kao znak povećane masu miokarda desnoga srca, te smanjena šupljina klijetke u sistoli i

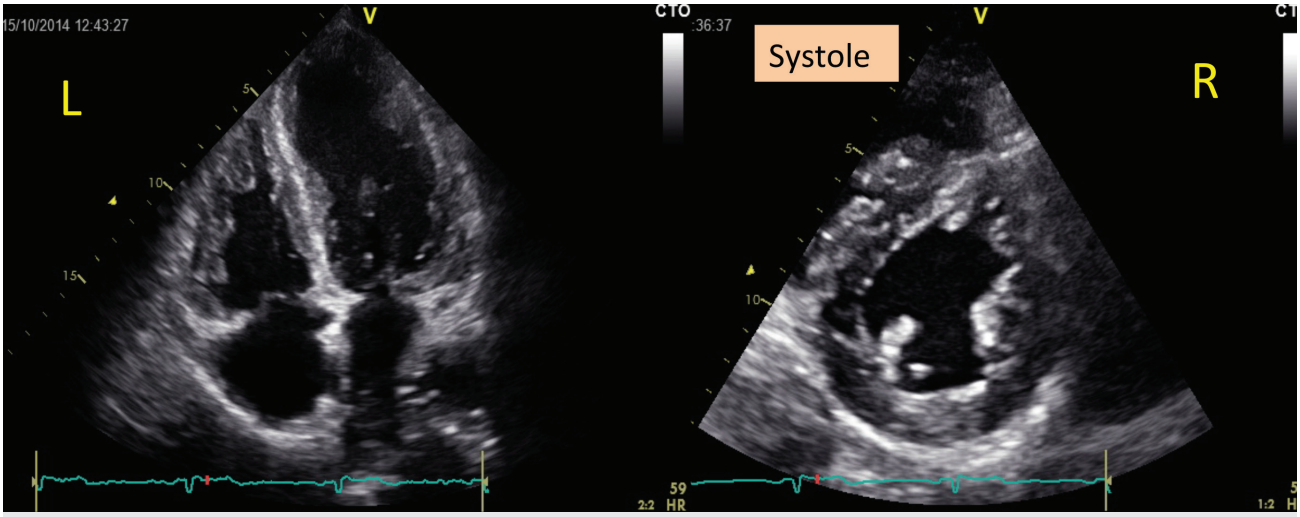
as the turbulent flow.[7,8,12] The constriction is verified by high flow velocities at the location, whereas for severe constriction we should also see the typical diastolic tail turbulence (if the elasticity of the wall of the distal aorta is intact). Additionally, the gradient over the constriction can be misleadingly low if the collateral circulation is well developed.[10]

THE RIGHT HEART

When examining a patient suspected or being screened for simple ACHD, the right heart should receive special attention. Indicators of pressure or volume load (e.g. dilated cavity of the RA or RV, RV hypertrophy, etc.) indicate a defect that should be examined further.

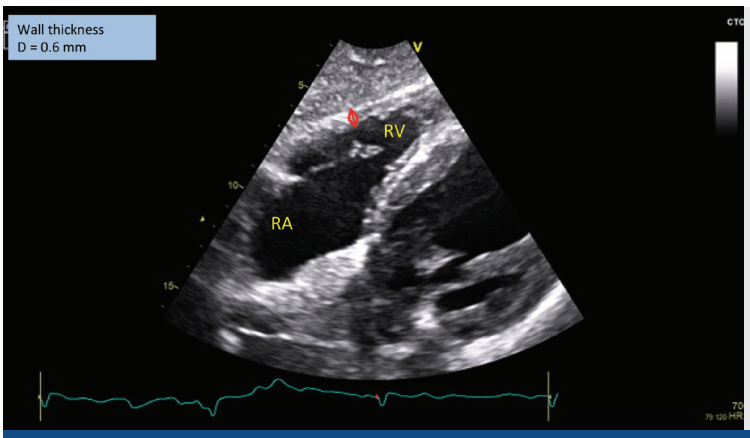
a) Pressure load

In addition to cavity size and wall thickness in the left and right heart, the dynamic relationship of the pressure differential during the cardiac cycle is important as well. When it is disturbed, the ventricular or interatrial septum is displaced towards the cavity with the lower pressure.[1,7]



Pressure load to the right heart. Concentric hypertrophy of the right ventricle (L), systolic flattening of the interventricular septum, D– systolic shape of the left ventricle (R).

FIGURE 3.



Hypertrophy of the right ventricle. Wall thickness measurement in the right ventricle at the level of the chords of the tricuspid valve, in subcostal view in the diastole. Wall thickness >0.5mm

RA = right atrium; RV = right ventricle

FIGURE 4.

dijastoli.[9] Dokaz postojanja tlačnog opterećenja u očuvanoj sistoličkoj funkciji jest plućna hipertenzija s vršnim sistoličkim tlakom u DK i plućnoj arteriji većim od 40 mmHg.[5,7,11]

Među češćim uzrocima tlačnog opterećenja desnog srca u PSBO-u jesu:

- pulmonalna stenoza (subvalvularna, valvularna i supravalvularna) (uključujući i stenoze umjetnih proteza i umetaka)
- opstrukcija izgonskoga trakta desne klijetke (RVOT) (npr. nakon operirane Fallotove tetralogije (TOF)
- „atrial switch" operacija transpozicije velikih krvnih žila (TGA)
- prirođeno korigirana transpozicija velikih krvnih žila (ccTGA)
- periferne stenoze plućne arterije.

b) Volumno opterećenje

U **volumnom opterećenju** DK dolazi do pomaka septuma prema lijevo u dijastoli – „D-oblik" LK u dijastoli (**slika 5**). Za lakše snalaženje u vremenskom određivanju pomaka septuma unutar srčanog ciklusa korisni su M-mode presjeci. Osim pomaka septuma, volumno opterećenje s vremenom dovodi do ekscentrične hipertrofije desnoga srca, povećanog volumena, izmijenjenog zaobljenog oblika (ali održane debljine stijenki). Nužno je što objektivnije izmjeriti volumene/površine DK i DA, kao i utvrditi sistoličku i dijastoličku funkciju desnoga srca. U hemodinamskoj procjeni korisni su nam protoci u RVOT-u i preko PV-a koji su veći zbog većeg volumena, ali bez gradijenta između izgonskoga trakta i zalistka, jer su brzine u RVOT-u i preko PV-a podjednake (napomena: brzine u RVOT-u mjerimo koristeći se pulsirajućim doplerom, a preko zalistka brzine mjerimo kontinuiranim doplerom).[7,11]

Kod znatno uvećane šupljine DK i DA može doći do širenja trikuspidnog prstena i, posljedično, do funkcionalne trikuspidne regurgitacije. Brzine regurgitirajućega mlaza i razlike tlakova su male, PAP ≤ 40 mmHg.

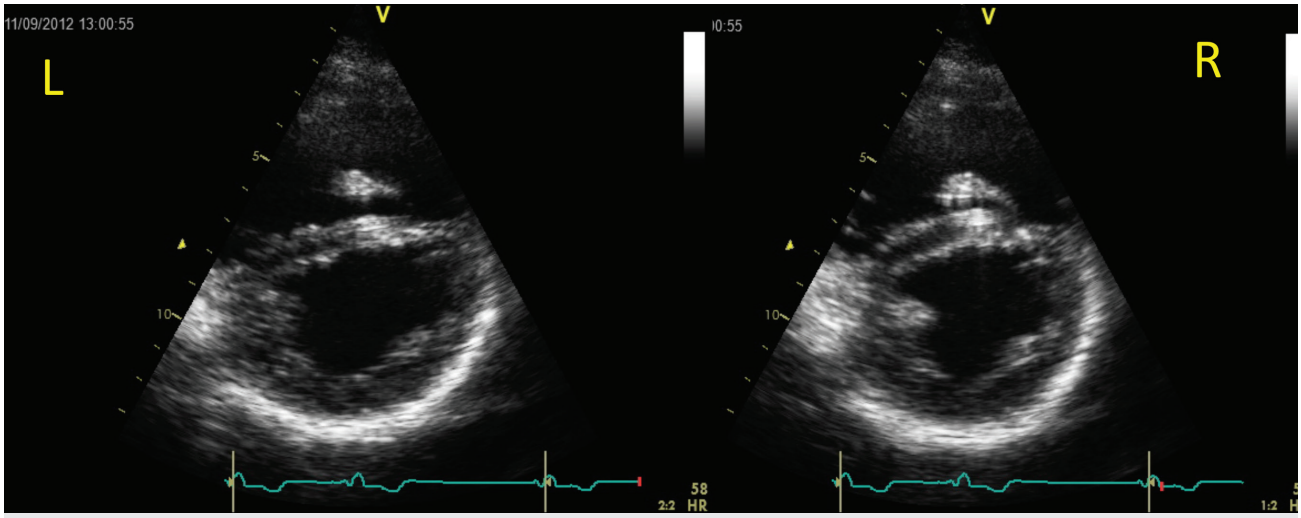
Pressure load of the RV causes displacement of the septum to the left in the systole and the so called D-shaped left ventricle (**Figure 3**). Long-term pressure load causes concentric remodeling of the RV as well. This includes diastolic wall thickness above 0.5 mm at the level of the chords of the tricuspidal valve (**Figure 4**), as a sign of increased right heart myocardial mass, and cavity reduction in the ventricle in the systole and diastole.[9] The evidence for the existence of pressure load in preserved systolic function is pulmonary hypertension with ceiling systolic pressure in the RV and 40 mmHg in the pulmonary artery.[5,7,11]

Common causes of pressure load to the right heart in ACHD cases include:

- Pulmonary stenosis (subvalvular, valvular, and supravalvular) (including stenosis of prosthetics and implants)
- Obstruction of the outflow tract of the right ventricle (RVOT) (e.g. after tetralogy of Fallot (TOF) repair)
- Atrial switch procedure for the transposition of the great arteries (TGA)
- Congenitally corrected transposition of the great arteries (ccTGA)
- Peripheral stenosis of the pulmonary artery

b) Volume load

Volume load to the RV causes diastolic displacement of the septum to the left – D-shaped left ventricle (**Figure 5**). Cross-section imaging with M-mode facilitates in determining the point in the cardiac cycle that the septum displacement occurs. In addition to septum displacement, volume load eventually leads to eccentric hypertrophy of the right heart, increased volume, and change in the shape of the curve (but wall thickness remains constant). It is crucial to measure the volume/surface of the RV and RA as objectively as possible, as well as determine the systolic and diastolic function of the right heart. Hemodynamic assessment is based on RVOT



Volume load to the right ventricle with diastolic flattening of the interventricular septum; D-shape in the diastole (L); septum is normalized in the systole (R).

FIGURE 5.

Najčešći uzroci volumnog opterećenja DK u odraslih bolesnika bez otprije poznate PSBO mogu biti:

- anomalni utok plućnih vena
- sinus venosus (otvor uz gornju ili donju šuplju venu)
- atrijski septalni otvor (ASD)
- pulmonalna regurgitacija
- primarna trikuspidna regurgitacija (svi oblici Ebsteinove anomalije).

Najčešći uzroci izoliranog proširenja desne pretklijetke nalazimo kod anomalnog utoka plućnih vena, malih atrijskih septalnih otvora te prirođene bolesti trikuspidnog zalistka.[7]

U zaključku možemo reći da se ehokardiografski pokazatelji jednostavnih i manje teških PSBO-a mogu razvrstati u pet osnovnih kategorija:

I. veličina i morfologija LK
II. velična desnog atrija, položaj interatrijskog septuma
III. veličina i morfologija DK
IV. plućna hipertenzija
V. gradijent tlaka preko AV/LVOT i PV/RVOT.

Nalaz bilo kojeg od gore navedenih pokazatelja s kardiološkim simptomima ili znakovima ili bez njih treba pobuditi sumnju na postojanje PSBO koju potom treba dokazati ili isključiti.

and PV flow, which are increased due to increased volume but with no gradient between the ejection tract and the valve since the RVOT and PV velocities are comparable (note: RVOT velocity is measured with pulsed wave Doppler imaging, and flow across the valve is measured with continuous Doppler imaging.[7,11]

In cases of significant dilation of the RV and RA, the tricuspid ring can expand with consequent functional tricuspid regurgitation. The velocity of the regurgitation flow and the pressure differentials are low, PAP≤40 mmHg.

The most common causes of volume load to the RV in adult patients with no previously diagnosed ACHD can be:

- Anomalous pulmonary venous return
- Sinus venosus (an opening at the superior or inferior vena cava)
- Atrial septal defect (ASD)
- Pulmonary regurgitation
- Primary tricuspid regurgitation (all forms of Ebstein's anomaly)

The most common causes of isolated dilation of the right atrium are found in anomalous pulmonary venous, small ASD, and congenital tricuspid valve disease.[7]

In conclusion, we can say that echocardiographic indicators for simple and less complex ACHD can be divided into five basic categories:

I. LV size and morphology
II. Size of the right atrium, position of the interatrial septum
III. RV size and morphology
IV. Pulmonary hypertension
V. AV/LVOT and PV/RVOT pressure gradients

The presence of any of the above indicators with or without cardiologic symptoms or signs should make us suspect ACHD, which should then be established or eliminated.

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