Prikaz dvaju slučajeva atrijskoga septalnog defekta otkrivenog u odrasloj dobi

Two Cases of Atrial Septal Defects Diagnosed in Adult Patients

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SAŽETAK: Atrijski septalni defekt (ASD) najčešća je srčana greška u odrasloj dobi. Često se kasno dijagnosticira. Potrebno je posumnjati na ovaj defekt kad se ehokardiografski registrira uvećana desna klijetka. Prikazana su dva slučaja pacijenata starijih od 50 godina s novodijagnosticiranim ASD-om. U obaju bolesnika greška se prezentirala s aritmijom (fibrilacija atrija, odnosno totalni atrio-ventrikulski blok) i učinjena je uspješna kirurška, odnosno intervencijska korekcija defekta. Ako nema kontraindikacija, a najčešća je razvoj značajne plućne hipertenzije, odnosno plućne vaskularne rezistencije, preporučuje se aktivni stav, tj. zatvaranje defekta, jer znatno pridonosi kliničkom poboljšanju, ali i smanjenju mortaliteta.

SUMMARY: Atrial septal defects (ASD) are the most common heart defect in adult patients and are often diagnosed late. This defect should be suspected whenever echocardiographic imaging finds an enlarged right ventricle. We report two cases of patients with newly-diagnosed ASD that were older than 50 years of age. In both cases the patients presented with arrhythmia (atrial fibrillation, i.e. total atrioventricular block), and a successful correction of the defect was performed. If there are no contraindications, that is primarily the development of significant pulmonary hypertension i.e. pulmonary vascular resistance, the active approach – defect closure – is recommended, since it significantly contributes to clinical improvement and reduction in mortality.

KLJUČNE RIJEČI: prirođene srčane greške u odraslih, atrijski septalni defekt, ehokardiografija, plućna hipertenzija, Amplazer okluder.

KEYWORDS: adult congenital heart disease, atrial septal defect, echocardiography, pulmonary hypertension, Amplatzer occluder.

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Uvod

Atrijski septalni defekt (ASD) jedna je od najučestalijih prirođenih srčanih grešaka, a prema podatcima, više od 50% upravo se otkrije u odrasloj životnoj dobi. Na desno-lijevu komunikaciju potrebno je posumnjati uvijek kada ehokardiografski utvrdimo uvećanu desnu stranu srca. Indikacija za zatvaranje ASD-a jest signifikantan defekt, tj. ≥ 10 mm sa znakovima volumnog opterećenja desne klijetke, a bez znakova značajne plućne vaskularne rezistencije (< 5 Woodovih jedinica). Isto tako bolesnici sa sumnjom na paradoksalne embolizacije uz isključenje drugih uzroka trebaju biti razmatrani za zatvaranje defekta.[1] Perkutano zatvaranje metoda je izbora kod ASD-a tipa secundum ako defekt nije preveliki (< 38 mm) te ako

Introduction

Atrial septal defects (ASD) are one of the most common congenital heart defects, and according to data more than 50% are diagnosed in adulthood. Right-to-left communication should be suspected whenever echocardiographic imaging establishes that the right side of the heart is enlarged. Closure is indicated for ASD when there is a significant defect of ≥10 mm and indication of right ventricular volume overload, but with no indication of notable pulmonary vascular resistance (<5 Wood units). Closure should also be considered in patients with suspected paradoxical embolizations, if other causes have been eliminated.[1] Percutaneous closure is the method of choice for ostium secundum ASD if

postoje adekvatni rubovi od minimalno 4 – 5 mm na koje se može adekvatno „usidriti" uređaj. Sukladno navedenom, kada je defekt izrazito veliki, bez adekvatnih rubova i kada postoji opasnost od ozljede okolnih struktura indicirano je kiruško zatvaranje.[2]

Ehokardiografija je postala dominantna metoda u dijagnosticiranju, procjeni veličine i oblika defekta, anatomije rubova, anomalnih plućnih vena i okolnih struktura. 3D ehokardiografija u današnje vrijeme dodatno nam pomaže upravo da bolje vizualiziramo navedene strukture.[3,4] Korist je od zatvaranja ASD-a višestruka. Fiziološki dolazi do redukcije u veličini desne klijetke i povećanja veličine i funkcije lijeve klijetke.[5] Bolesnici prelaze u niži funkcijski NYHA status već unutar nekoliko mjeseci od zatvaranja defekta te dolazi do povećanja fizičkog kapaciteta čak i neovisno o NYHA funkcionalnom statusu i dobi.[6] Konačno, uz fiziološko i simptomatsko poboljšanje dokazano je da zatvaranje signifikantnog ASD-a produljuje preživljavanje. Naime, neoperirani bolesnici imaju znatno skraćeno preživljenje u usporedbi s općom populacijom i povećani morbiditet uz razvoj kardiopulmonalnih komplikacija. S druge strane, preživljavanje je komparabilno s općom populacijom ukoliko je zatvaranje ASD-a učinjeno što ranije u životu, a idealno prije 24. godine života.[7]

Slučaj 1.

Pacijentica u dobi od 58 godina hospitalizirana je putem hitne službe zbog paroksizmalne fibrilacije atrija praćene zaduhom. Do tada nije teže bolovala. Vrlo brzo nakon hospitalizacije uz medikamentnu terapiju dolazi do konverzije fibrilacije atrija u stabilni sinusni ritam. U 12-kanalnom elektrokardiogramu (EKG) razabiru se desna električna os i inkompletni blok desne grane. U sklopu kliničkoga pregleda treba izdvojiti tih sistolički ejekcijski šum nad prekordijem i fiksno cijepan drugi ton. Labaratorijski su nalazi bili u granicama normale. U nastavku su učinjene transtorakalna (TTE) i transezofagusna ehokardiografija (TEE) kojom se prikaže uredna veličina i funkcija lijeve klijetke (**slika 1**). Desna klijetka (DK) bila je uvećana, očuvane funkcije (RVEDd 4Ch 4,0 cm, FAC 37%, TAPSE 1,9 cm), bez znakova plućne hipertenzije (TR V maks. 2,6 m/s, VCI diam 2,0 cm, PVAccT 120 ms, PAP 32 mmHg), međutim, uz prisutne znakove volumnog opterećenja (pomak i izravnavanje ventrikulskog septuma prema lijevoj klijetki tijekom dijastole). Osim blage trikuspidne insuficijencije, ostale valvule bile su uredne morfologije i funkcije. Na 3D TEE jasno je bio vidljiv atrijski septalni defekt ovalnog oblika (15 x 10 mm) lokaliziran u području fose ovalis sa značajnim lijevo-desnim šantom (**slika 2**). Omjer plućnog prema sistemnom optoku (Qp/Qs) iznosio je 2,9 : 1. Prikazane su sve četiri plućne vene s adekvatnim utokom u lijevi atrij, bez anomalnih utoka. Nije bilo drugih srčanih grešaka. Pretraga magnetnom rezonancijom (MR) srca prekinuta je zbog panične atake uzrokovane klaustrofobijom. Desnostrana kateterizacija srca pokazala je skok saturacije na razini DK te potvrdila nalaz ASD-a uz uredne tlakove u plućnoj cirkulaciji bez značajne plućne vaskularne rezistencije. U bolesnice je implantiran Amplazer septalni okluder bez periproceduralnih komplikacija (**slika 3** i **slika 4**) i bolesnica je već sljedeći dan otpuštena iz bolnice uz preporuku uzimanja acetilsalicilatne kiseline i male doze beta-bloka-

the defect is not too large (<38 mm) and if an adequate rim is present, minimally 4-5 mm, on which the device can be anchored. Surgical closure is indicated when the defect is large, has no adequate rim, and if there is danger of damage to the surrounding tissue.[2]

Echocardiographic imaging has become the dominant method for diagnosis and assessing size, rim anatomy, anomalous pulmonary veins, and surrounding structures. Today, 3D echocardiographic imaging is an additional tool that helps us better visualize the above elements.[3,4] The benefits of ASD closure are many. Physiologically, the right ventricle is reduced in size while the left ventricle improves in both size and function.[5] Patients move to a lower New York Heart Association (NYHA) class shortly after defect closure, and physical capacity improves even regardless of NYHA class and age.[6] Finally, in addition to physiological and symptomatic improvements, it has been shown that closure of significant ASD improves survival as well. Patients that did not undergo intervention have a significantly shorter survival rate and higher mortality in comparison with the general population as well as increased morbidity from cardiopulmonary complications. In patients who underwent ASD closure early in life, ideally before the age of 24, survival rates are comparable to the general population.[7]

Case 1

A female patient was hospitalized at the age of 58 via emergency medical service for paroxysmal atrial fibrillation and dyspnea. She had had no significant prior illnesses. Soon after hospitalization, atrial fibrillation was medically converted to stable sinus rhythm. A 12-lead electrocardiogram (ECG) showed right electrical axis deviation and an incomplete right bundle branch block. During clinical examination, a quiet systolic ejection murmur was noted over the precordium as well as a fixed split second tone. Laboratory test results were within normal ranges. Transthoracic (TTE) and transesophageal echocardiography (TEE) was performed, which showed normal size and function of the left ventricle (**Figure 1**). The right ventricle (RV) was enlarged, its function intact (RVEDd 4Ch 4.0 cm, FAC 37%, TAPSE 1.9 cm), with no signs of pulmonary hypertension (TR V max 2.6 m/s, VCI diam 2.0 cm, PVAccT 120 ms, PAP 32 mmHg). However, there were signs of volume overload (bowing and flattening of the ventricular septum toward left ventricle during diastole). Other than mild tricuspid insufficiency, the other valves had normal morphology and function. 3D TEE clearly showed an oval atrial septal defect (15x10 mm) localized in the fossa ovalis area with a significant left-to-right shunt (**Figure 2**). The ratio of pulmonary to systemic blood flow (Qp/Qs) was 2.9:1. All four pulmonary veins were seen to have adequate flow into the left atrium, with no anomalous inflow. There were no other heart defects. Magnetic resonance (MR) imaging of the heart was interrupted due to a claustrophobia-induced panic attack. Right-sided cardiac catheterization showed an elevation of saturation at the level of the RV and confirmed the diagnosis of ASD, with normal pressures in the pulmonary circulation and without significant pulmonary vascular resistance. An Amplatzer septal occluder was implanted with no complications during the procedure (**Figure 3** and **Figure 4**); the patient was

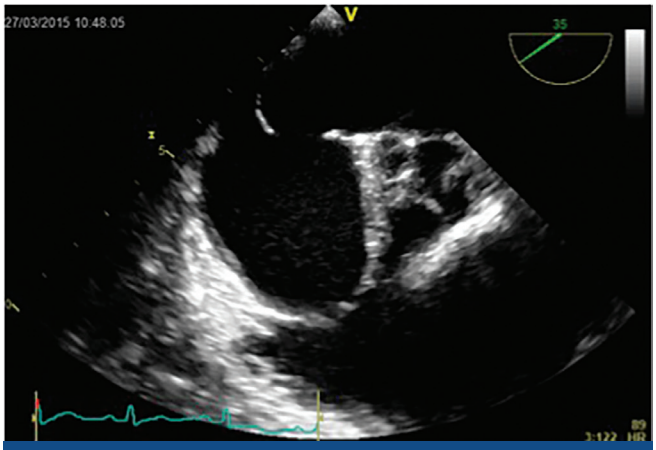

FIGURE 1. Transoesophageal short axis view of atrial septal defect.

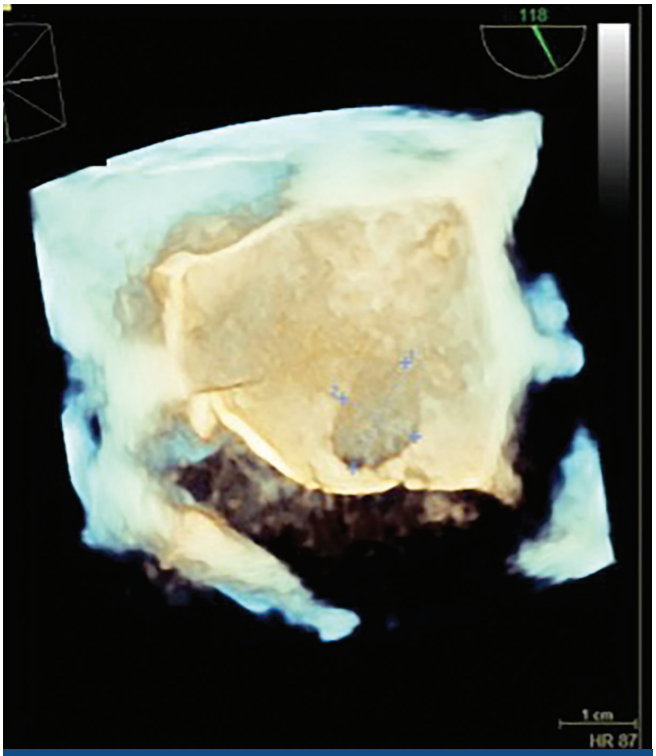

FIGURE 2. Transoesophageal 3-dimensional view of the atrial septal defect.

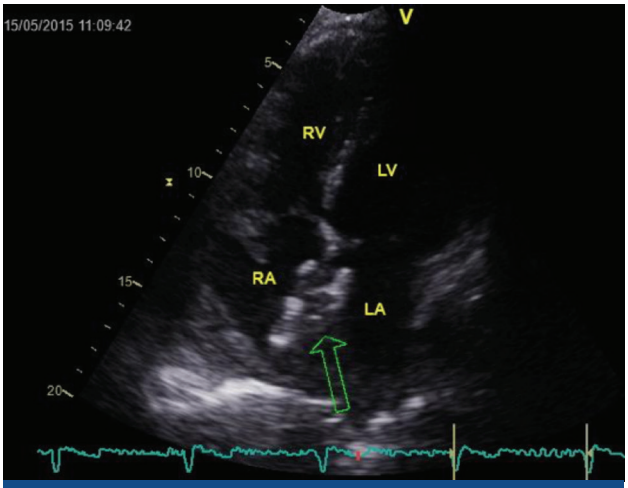

FIGURE 3. Transthoracic short axis view at the aortic level of atrial septal defect Amplatzer occluder (arrow) at the end of the procedure.

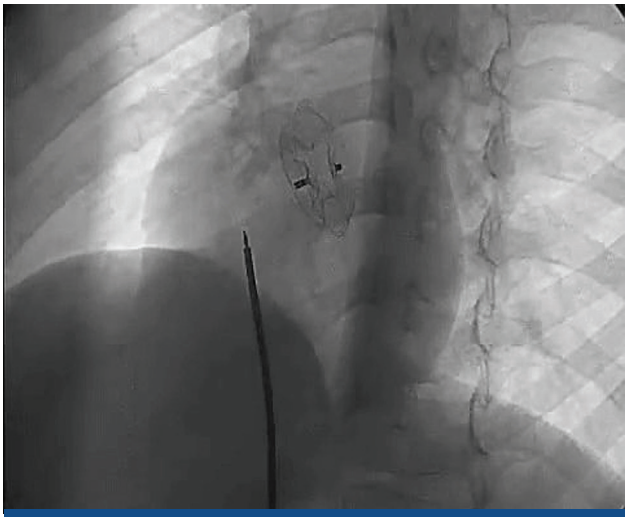

FIGURE 4. X-ray of atrial septal defect Amplatzer occluder at the end of the procedure.

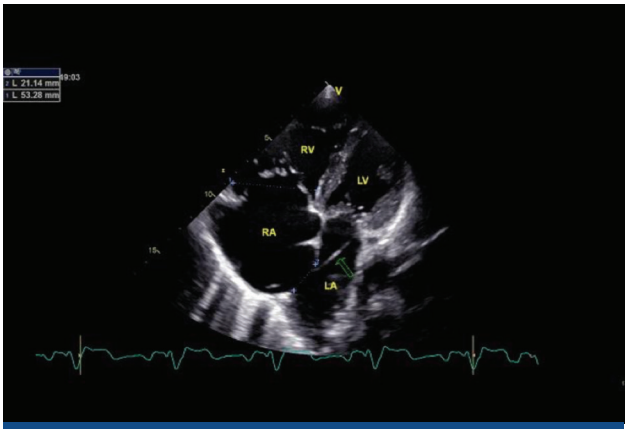

FIGURE 5. Transthoracic apical four chamber view of the atrial septal defect with a pacemaker electrode (arrow) passing through interatrial communication and stimulating the left ventricle.

tora. Preporučena je profilaksa bakterijskog endokarditisa do epitelizacije Amplazera (6 mjeseci).

Slučaj 2.

Pacijentica u dobi od 66 godina, koja dotad nije teže bolovala, hospitalizirana je putem hitne službe zbog intolerancije napora i omaglica, a verificiran je totalni atrio-ventrikulski blok frekvencije ventrikula oko 35/min. U bolesnice je implantiran elektrostimulator srca nakon kojeg se u elektrokardiogramu bilježi ritam elektrostimulatora s jasno vidljivim „šiljcima-spikovima" izgleda bloka desne grane. Pregled TTE-om razot-

krio je razmjerno veliki ASD tipa ostium secundum promjera 26 mm te se vidjela elektroda elektrostimulatora srca koja ulazi kroz venu kavu superior u desni atrij te preko ASD-a prolazi u lijevi atrij i preko mitralnog zaliska vrškom završava u području lateralne stijenke lijeve klijetke (**slika 5**). ASD je bio s dominantno lijevo-desnim šantom uz znakove volumnog opterećenja DK i umjerenu trikuspidnu insuficijenciju. Ehokardiografijom i desnom kateterizacijom srca verificira se i umjerena plućna hipertenzija (PAP do 50 mmHg) uz odnos plućne prema sistemnoj cirkulaciji (Qp/Qs) 2 : 1 te plućna vaskularna rezistencija od 3 Wooda. U bolesnice je učinjeno operativno zatvaranje ASD-a uz ekstrakciju i repozicioniranje elektrode elektrostimulatora srca. Postoperativni je tijek protekao uredno i pacijentica je bez znatnih poteškoća u šestomjesečnom praćenju.

Zaključak

S obzirom na navedeno, u svakog bolesnika s verificiranim signifikantnim ASD-om koji nema kontraindikacija za zatvaranje, potreban je proaktivni pristup. Odluka o načinu zbrinjavanja, bilo intervencijskom ili kiruškom metodom, donosi se na osnovi individualnih karakteristika svakog bolesnika.

discharged the next day and was advised to take aspirin and a small dose of beta blockers. Prophylaxis of bacterial endocarditis was advised until the epithelization of the Amplatzer occluder (6 months).

Case 2

A 66-year-old female patient with no previous history of severe illness was hospitalized via emergency medical service for exertion intolerance and dizziness. A total atrioventricular block was identified with a ventricular frequency of about 35/min. A pacemaker was implanted, after which the ECG showed the pacemaker spikes with QRS appearance of a right bundle branch block. TTE examination showed a relatively large ASD of the ostium secundum type, 26 mm in diameter. The pacemaker lead could be seen entering through the vena cava superior into the right atrium and going through the ASD into the left atrium, with the tip of the lead going over the mitral valve and stimulating the lateral wall of the left ventricle (**Figure 5**). The ASD had a dominant left-to-right shunt with signs of RV volume overload and moderate tricuspid insufficiency. Echocardiography and right-sided cardiac catheterization also showed moderate pulmonary hypertension (PAP up to 50 mmHg) with a Qp/Qs ratio of 2:1 and pulmonary vascular resistance of 3 Wood units. Surgical closure of ASD was performed, including pacemaker lead extraction and repositioning. Postoperative course was uneventful, and the patient was without significant problems after 6 months of follow-up.

Conclusion

Based on the above, a proactive approach is recommended in any patient with established significant ASD and with no contraindications for closure. Treatment decision, either interventional or surgical, is made on the basis of the individual characteristics of every patient.

LITERATURE

1. 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 (AEPC); ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J. 2010;31:2915-57. **DOI:** http://dx.doi.org/10.1093/eurheartj/ehq249
2. Bissessor N. Current perspectives in percutaneous atrial septal defect closure devices. Med Devices (Auckl). 2015;8:297-303. **DOI:** http://dx.doi.org/10.2147/MDER.S49368
3. Roberson DA, Cui VW. Three-dimensional transesophageal echocardiography of atrial septal defect device closure. Curr Cardiol Rep. 2014;16(2):453. **DOI:** http://dx.doi.org/10.1007/s11886-013-0453-4
4. Faletra FF, Pedrazzini G, Pasotti E, Muzzarelli S, Dequarti MC, Murzilli R, et al. 3D TEE during catheter-based interventions. JACC Cardiovasc Imaging. 2014;7(3):292-308. **DOI:** http://dx.doi.org/10.1016/j.jcmg.2013.10.012
5. Veldtman GR, Razack V, Siu S, El-Hajj H, Walker F, Webb GD, et al. Right ventricular form and function after percutaneous atrial septal defect device closure. J Am Coll Cardiol. 2001;37(8):2108-13. **DOI:** http://dx.doi.org/10.1016/S0735-1097(01)01305-5
6. Brochu MC, Baril JF, Dore A, Juneau M, De Guise P, Mercier LA Improvement in exercise capacity in asymptomatic and mildly symptomatic adults after atrial septal defect percutaneous closure. Circulation. 2002;106(14):1821-6. **DOI:** http://dx.doi.org/10.1161/01.CIR.0000029924.90823.E0
7. Murphy JG, Gersh BJ, McGoon MD, Mair DD, Porter CJ, Ilstrup DM, et al. Long-term outcome after surgical repair of isolated atrial septal defect. Follow-up at 27 to 32 years. N Engl J Med. 1990;323(24):1645-50. **DOI:** http://dx.doi.org/10.1056/NEJM199012133232401