Cor et Vasa

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/crvasa

Kasuistika | Case report

When is it too late for a correction of an atrial septal defect secundum type in an adult patient?

Stefan Naydenov Naydenov[a], Nikolay Margaritov Runev[a], Emil Ivanov Manov[a], Rabhat Ahmet Shabani[a], Temenuga Ivanova Donova[a], Pencho Doychev Kratunkov[b]

[a] *Department of Internal Diseases "Prof. St. Kirkovich", Medical University of Sofia, Sofia 1431, Bulgaria*
[b] *"St Ekaterina" University Hospital, Sofia 1431, Bulgaria*

ARTICLE INFO

Article history:
Received: 10. 5. 2015
Received in revised form: 9. 6. 2015
Accepted: 19. 6. 2015
Available online: 15. 7. 2015

Klíčová slova:
Defekt síňového septa
typu secundum
Dospělí
Korekce

Keywords:
Adults
Atrial septal defect secundum type
Correction

SOUHRN

Defekty síňového septa (atrial septal defect, ASD) patří mezi nejčastější vrozené srdeční anomálie zjišťované v dospělosti; jsou charakterizovány defektem v mezisíňovém septu umožňujícím průtok krve z levé síně do síně pravé. V tomto článku popisujeme případ 39leté ženy s nekorigovaným ASD typu secundum, který byl diagnostikován v dospělosti poté, co se u ní projevily morfologické a funkční změny. V době vyšetření bylo nutno vyloučit život ohrožující komplikace, jakou je infekční endokarditida v místě mezisíňového defektu. Po důkladném vyšetření bylo u naší pacientky zvoleno chirurgické řešení, kdy byl defekt uzavřen perikardiální záplatou s následným rychlým zotavením a významným zlepšením klinických parametrů nemocné.

ABSTRACT

Atrial septal defects (ASD) are among the most commonly recognized congenital cardiac anomalies presenting in adulthood, characterized by a defect in the interatrial septum allowing blood from the left atrium to pass to the right atrium. In this article we present a clinical case of a 39-year-old female patient with uncorrected ASD secundum type, diagnosed in adulthood after cardiac morphologic and functional changes, have been developed. At the time of presentation, clinical symptoms required exclusion of life-threatening complications such as infective endocarditis, located at the interatrial defect. After careful evaluation, surgical approach was chosen for our patient and the defect was closed using pericardial patch with rapid patient's recovery and significant clinical improvement.

Address: Stefan Naydenov Naydenov, MD, PhD, Department of Internal Diseases "Prof. St. Kirkovich", Medical University of Sofia, Georgi Sofyiski Str., Sofia 1431, Bulgaria, e-mail: snaydenov@gmail.com
DOI: 10.1016/j.crvasa.2015.06.004

Please cite this article as: S.N. Naydenov, et al., When is it too late for a correction of an atrial septal defect secundum type in an adult patient?, Cor et Vasa 58 (2016) e439–e443 as published in the online version of *Cor et Vasa* available at http://www.sciencedirect.com/science/article/pii/S0010865015000703

Introduction

Congenital heart defects (CHD) are the most common types of birth defects, and most babies born with these conditions are living nowadays longer and healthier lives. Data from the European Surveillance of Congenital Anomalies central database for 29 population-based congenital anomaly registries in 16 European countries show that the average total prevalence of CHD is 8.0 per 1000 births [1–3]. The prevalence of some CHD, especially mild types, is increasing, while this one among the other types has remained stable [1–6].

Congenital heart defects cause high morbidity and mortality among infants and affect the quality of life during childhood and adulthood, depending on the progression of the disease [2–4]. While some newborns with a cardiac disorder are symptomatic and identified soon after birth, many others are not diagnosed until the disease progresses into a severe stage. Data from the Northern Region Pediatric Cardiology database suggest around 1 in 4 cases of congenital heart disease in the UK are diagnosed later in childhood or adulthood [1–6].

Atrial septal defects (ASD) are one of the more commonly recognized congenital cardiac anomalies presenting in adulthood, characterized by a defect in the interatrial septum allowing pulmonary venous return from the left atrium to pass directly to the right atrium [1,3,7]. Depending on the size of the defect, size of the shunt, and associated anomalies, this can result in a spectrum of diseases from no significant cardiac sequelae to right-sided volume overload, pulmonary arterial hypertension, and even atrial arrhythmias [8–10].

Secundum type ASD represents 80% of ASDs and is usually located in the region of the fossa ovalis and its surrounding [1–4]. Patients frequently remain asymptomatic until adulthood; however, the majority develop symptoms beyond the fourth decade including reduced functional capacity, exertional shortness of breath, and palpitations (supraventricular tachyarrhythmias), and less frequently pulmonary infections and right heart failure. Life expectancy is reduced overall, but survival is much better than previously assumed [7–11].

In this article we present a case of a patient with uncorrected ASD secundum type, diagnosed in adulthood after cardiac morphologic and functional changes have been developed.

Case presentation

A 39-year-old female patient from the city of Sofia, Bulgaria was admitted to our hospital with symptoms of viral respiratory infection – fever up to 38.5 °C and productive cough (October 2013). The patient had never had complaints consistent with a cardiovascular disease so far. From the medical history we found Hashimoto thyreoiditis and diabetes mellitus type 2 to be theconcomitant diseases.

Physical examination on the day of hospital admission: satisfactory general condition; Pulmonary system: acute vesicular breathing, no crackles and crepitations; Cardiovascular system: rhythmic heart rate ~90 beats per minute (bpm), clear heart sounds, largely split and accentuated second hear sound, proto-mesosystolic 2/6 murmur at the second left intercostal space, next to the sternum, arterial blood pressure 125/80 mmHg; Abdomen: soft abdominal wall, not painful on palpation, respiratory mobile, liver and spleen – not enlarged; Extremities: no edema, preserved peripheral arterial pulsations.

From the instrumental investigations (October 2013):

Electrocardiography: sinus rhythm, heart rate 83 bmp, right axis deviation, incomplete right bundle branch block (RBB), criteria presented for possible right ventricular hypertrophy and right atrial enlargement (Fig. 1).
The X-ray showed mild to moderate pulmonary congestion, no infiltrations, enlarged cardiac silhouette, double-figured descendent aorta (suspicions for aortic coarctation), marked conus pulmonalis.
From the laboratory: fasting glucose 9.0 mmol/L; all other routine laboratory findings within normal ranges.

Transthoracic echocardiography:

- Marked dilation of the right chambers, the heart apex is formed by the right ventricle – right ventricle diameter 35 mm, right atrium – 62/46 mm, left atrium – 65/43 mm; Large aneurysm of the atrial septum with deviation from the main axis up to 19 mm; Mobile formation in the right atrium, contacting the anterior cusp of the tricuspid valve.
- Atrial septal defect secundum type 32/29 mm, significant left-to-right shunt, reversed septal motion;
- Left ventricular (LV) end-diastolic diameter 37 mm, LV end-systolic diameter 23 mm, LV end-diastolic

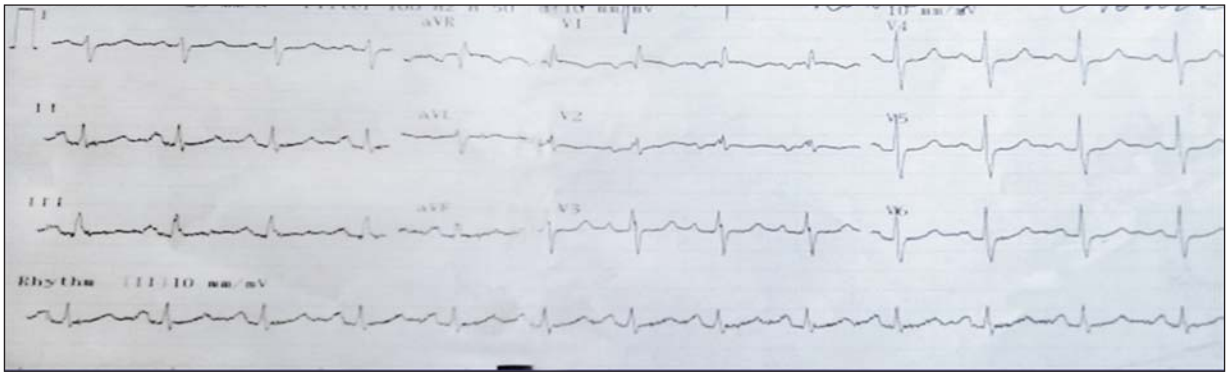

Fig. 1 – Electrocardiographic record of the patient on the day of hospitalization.

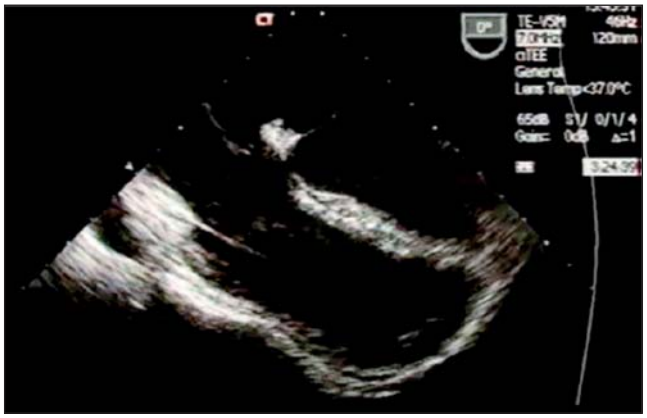

Fig. 2 – Transoesophageal echo of the patient: large atrial septal defect type ostium secundum and large aneurysm of the atrial septum with deviation.

volume 48 ml, LV end-systolic volume 16 ml, LV ejection fraction 56% (by Simpson's rule);
- Septum 10 mm, posterior LV wall thickness 10 mm
- Moderate tricuspid valve insufficiency; Tricuspid annular plane systolic excursion 23 mm
- Systolic pulmonary pressure 55 mmHg;
- Pericardial effusion ~ 250 ml.

Transoesophageal echocardiography: Marked Eustachian valve, reaching the coronary venous sinus and forming Chiari's network in the right atrium; Large atrial septal defect type ostium secundum and large aneurysm of the right atrium with deviation (32/33/20 mm) from the main axis up to 17 mm; Pulmonary hypertension (Fig. 2).

Invasive cardiac evaluation (February 2014) showed:
- Clear coronary arteries
- No segmental LV wall motion abnormalities
- LV ejection fraction 62%
- Large atrial septal defect visualized with left-toright shunt from the left to the right atrium
- Pressures: moderate pulmonary hypertension, peak pulmonary artery pressure 43 mmHg
- Oxymetry: O2 saturation step-up between vena cava superior and the pulmonary artery with large left-to-right shunt – Qp:Qs > 2.5. (Fig. 3A and 3B).

The data from the instrumental cardiac evaluation justified surgical closure of ASD. For this reason the patient was directed to a cardiac surgery clinic after the hospital discharge. The surgical intervention was performed on February 18th, 2014 and included pericardial patch closure of ASD under extracorporeal circulation. The post-procedural period was smooth, without complications and with rapid patient's recovery.

Post-surgical electrocardiography: sinus rhythm, heart rate 72 beats per minute, right axis deviations, no significant changes compared to the baseline electrocardiography.

Post-surgical transthoracic echocardiography: end-diastolic LV volume 67 ml, end-systolic LV volume 27 ml, LV ejection fraction 59% (by Simpson's rule), mild tricuspid regurgitation, systolic pulmonary arterial pressure 30 mmHg, no pericardial effusion, and pleural effusions on both sides ~350–400 ml. The other finding were similar to the baseline echocardiography.

The medical treatment prescribed after hospital discharge included: Acetyl-salicylic acid 100 mg daily, Torasemide 5 mg daily, Levothyroxine 50 µg daily, Gliclazide retard form 60 mg daily, Omeprazole 20 mg daily and Colhicine 1 mg daily (for 10 days).

Control clinical visits on 15th and 30th days were performed. The patient reported to feel significant improvement of the clinical symptoms. There were no clinical and instrumental signs for postoperative complications.

Discussion

Congenital heart disease accounts for nearly one-third of all major congenital anomalies with ASD secundum type being one of the most common among the population with CHD [1–3,7]. The majority of ASDs are detected in childhood, although a significant number of them are diagnosed in adult life [9,11]. That is the case presented in this article – our patient was first diagnosed at the age of 39 years. The signs and symptoms of CHD depend on the type and severity of the disease and an ASD could remain "silent" even until 4–5th decade before the first symptoms and signs appear [1–6]. A few ASDs present in childhood with breathlessness, or even heart failure, but most of them are detected by echocardiography after a child has been referred for other reasons [2–4,7,8]. In adult life, presentation commonly occurs because of breathlessness, atrial arrhythmias or heart failure [10,11].

In high-volume cardiovascular centers, a considerable number of ASD patients with late complications are admitted. However, our patient has been absolutely asymptomatic so far – she came to us with clinical manifestations of a viral infection. Furthermore, the detection of the CHD required exclusion of infective endocarditis (IE) [1–4,11]. Virtually any congenital cardiac defect may predispose to the development of IE although the risk of IE in an isolated uncorrected ASD is considered to be negligible compared for example to an uncorrected ventricular septal defect in which the risk for IE by the age of 30 years is about 10% [1–5].

We found no signs of intracardiac infection but the echocardiographic examination showed the typical evolution of a moderate to large ASD when the enlargement of the atria and the right ventricle, the tricuspid valve insufficiency as well as the pulmonary hypertension left uncorrected. The physiological consequences of ASD essentially are independent of their site and depend mostly on their size and the amount of the shunt which in turn depends on the compliance of the two ventricles and the size of the defect [1,2,5]. A small defect (<5 mm) is associated with a small shunt that is without hemodynamic consequences and a defect >20 mm on the other hand is associated with a large shunt that can have considerable hemodynamic effects [3,4]. A large uncorrected ASD causes dilatation of both left and right atria, the right ventricle and the pulmonary arteries in order to accommodate the increased blood volume. Ultimately either the right ventricle fails, or right ventricle compliance falls, resulting in a reduction in the magnitude of

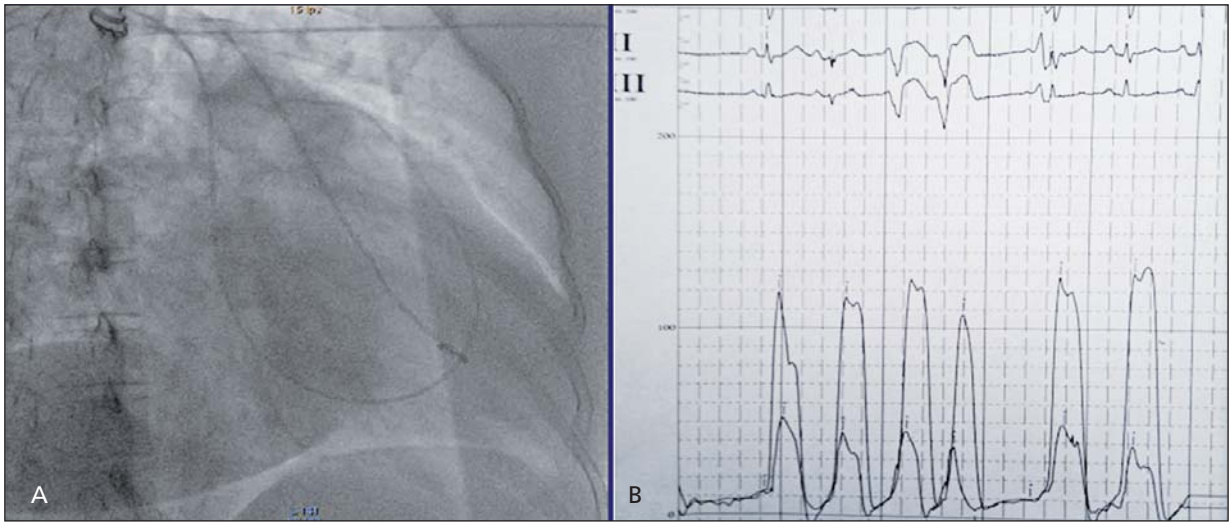

Fig. 3 – (A, B) Invasive cardiac evaluation: large atrial septal defect with left-to-right shunt from the left to the right atrium, moderate pulmonary hypertension, peak pulmonary artery pressure 43 mmHg.

shunting, or even flow reversal [3–6,8]. Although pulmonary hypertension is quite common with increasing age, it is rare for the pulmonary vascular resistance to exceed 500 dyn/s/cm^2 [4–7]. The ASD of our patient was relatively large with moderate left-to-right shunt and although the typical auscultation signs were present (fixed splitting of the second heart sound with a systolic murmur at the auscultation site of the pulmonic valve) the diagnose was delayed for decades [6–10]. Many authors explain this delay with the lack of enough experience of physicians in the general practice who meet usually "asymptomatic ASD patients with stable functional status" and a murmur that is attributed to a negligible valve dysfunction [7–11]. With this case we would like to point out the important role of the careful physical examination (auscultation, palpation, etc.) since its findings could direct our clinical cogitation toward a possible ASD, leading to timely referral of the patients for further specific investigations. Detailed medical history is also important since it could give us clues about the correct diagnosis, i.e. the reliable patient's information could help to differentiate a post--rheumatic from a congenital disorder.

The treatment approach in ASD patients depends mostly on the defect size, the clinical state of the patient, developed complications, concomitant conditions and diseases. In small defects and in the absence of signs of ventricular dilatation, with only a modest increase in pulmonary blood flow, closure has often been thought to be unnecessary [2–4,10]. However, atrial septal defects, associated with a greater than 50% increase in pulmonary blood flow should be closed in order to prevent the development of right ventricle dysfunction and atrial arrhythmias [11]. In patients who have undergone closure before 25 years of age, life expectancy and functional outcome are usually normal [11]. Unfortunately later closure remains a risk for premature late death. Traditionally, the closure of ASDs has required surgery. The era of transcatheter closure has now arrived, and today most ASDs can be closed percutaneously [11]. The long-term results of the devices used to close these defects are currently unknown, although the short-term results are excellent [3–9].

The treatment approach was discussed by an interdisciplinary team, including non-invasive and invasive cardiologists and cardiac surgeons as well. Endovascular repair was initially taken into consideration but after thorough evaluation of the patient this option was not approved for the following reasons: 1) Presence of a large atrial septal aneurysm with deviation from the main axis up to 17 mm and 2) A huge mobile formation in the right atrium of unknown origin, contacting the anterior cusp of the tricuspid valve – the differential diagnosis included: Chiari's network, a vegetation, a thrombus and a tumor in the right atrium. These were the main reasons for which surgical closure of ASD was performed.

The recovery of the patient was rapid and the clinical improvement was significant despite the morphological and functional cardio-pulmonary changes.

In conclusion, patients with hemodynamically significant ASDs could remain undiagnosed even until their 4-5th decade mostly due to their asymptomatic state. The presence of a fixed split of the second heart sound and a systolic murmur at the auscultation site of the pulmonic valve requires ASD exclusion. The clinical manifestation and complications of an uncorrected large ASD are due to the enlargement and malfunction of the atria and the right ventricle, the pulmonary hypertension and later due to the reverse of the shunt (Eisenmenger's syndrome). The treatment approach is invasive – surgical or non-surgical and is determined by careful clinical and instrumental patient's evaluation.

Conflict of interest
None declared.

Funding body
None.

Ethical statement
Authors state that the research was conducted according to ethical standards.

Informed consent

The authors of this article (Stefan Naydenov Naydenov, Nikolay Margaritov Runev, Emil Ivanov Manov, Temenuga Ivanova Donova, Rabhat Ahmet Shabani and Pencho Doychev Kratunkov) agree that the article be published in its current version.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.crvasa.2015.06.004.

References

[1] K. Bjornard, T. Riehle-Colarusso, S. Gilboa, A. Correa, Patterns in the prevalence of congenital heart defects: metropolitan Atlanta, 1978 to 2005, Birth Defects Research. Part A, Clinical and Molecular Teratology 97 (2) (2013) 87–94.

[2] L. Botto, A. Correa, D. Erickson, Racial and temporal variations in the prevalence of heart defects, Pediatrics 107 (2001) 32.

[3] S. Boulet, S. Grosse, T. Riehle-Colarusso, A. Correa-Villasenor, Health care costs of congenital heart defects, in: D.F. Wyszynski, A. Correa-Villasenor, T.P. Graham (Eds.), Congenital Heart Defects: From Origin to Treatment, Oxford University Press, Inc, New York, 2010 493–501.

[4] D. Linde, E. Konings, M. Slager, Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis, Journal of the American College of Cardiology 58 (2011) 2241–2247.

[5] R. Hartman, S. Rasmussen, L. Botto, et al., The contribution of chromosomal abnormalities to congenital heart defects: a population-based study, Pediatric Cardiology 32 (2011) 1147–1157.

[6] J. Hoffman, S. Kaplan, The incidence of congenital heart disease, Journal of the American College of Cardiology 39 (2002) 1890–1900.

[7] A. Miller, T. Riehle-Colarusso, C. Alverson, et al., Congenital heart defects and major structural noncardiac anomalies, Atlanta, Georgia, 1968–2005, Journal of Pediatrics 159 (2011) 70–78.

[8] C. Limperopoulos, A. Majnemer, M. Shevell, et al., Neurodevelopmental status of newborns and infants with congenital heart defects before and after open heart surgery, Journal of Pediatrics 137 (2000) 638–645.

[9] N. Oyen, G. Poulsen, H. Boyd, et al., Recurrence of congenital heart defects in families, Circulation 120 (2009) 295–301.

[10] M. Reller, M. Strickland, T. Riehle-Colarusso, et al., Prevalence of congenital heart defects in Atlanta, 1998–2005, Journal of Pediatrics 153 (2008) 807–813.

[11] C. Warnes, R. Liberthson, G. Danielson Jr., et al., Task Force 1: the changing profile of congenital heart disease in adult life, Journal of the American College of Cardiology 37 (2001) 1170–1175.