Folia Cardiologica 2023
vol. 18, no. 1, pages 55–58
DOI: 10.5603/FC.a2022.0049

ISSN 2353–7752
e-ISSN 2353–7760

A rare case of a double atrial septum in a 4-year-old boy with a large an ostium secundum atrial septal defect

Rzadki przypadek podwójnej przegrody międzyprzedsionkowej u 4-letniego chłopca z dużym ubytkiem przegrody międzyprzedsionkowej typu *ostium secundum*

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Abstract

The work presents a case of a 4-year-old boy diagnosed with a large atrial septal defect type ostium secundum (ASD2) and additional abnormal structures in the direction of the left atrium in the echocardiogram. The patient was qualified for interventional defect closure, however, in the transoesophageal echocardiography, in addition to a very large ASD2 (approx. 2 cm), a double interatrial septum was diagnosed. The extra wall layer was parallel to the atrial septum and did not obstruct blood flow from the pulmonary veins or through the mitral valve. No signs of clotting material were observed in the space between the two layers. The closure was withdrawn during cardiac catheterization and the boy was qualified for scheduled surgery. Currently, the boy is asymptomatic and leads a normal lifestyle.

Key words: double atrial septum, interatrial space, congenital heart disease

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Introduction

Double atrial septum (DAS) is a very rare congenital defect of the interatrial septum, where there is a free space in the atria between the native septum and the accessory septum located in the left or the right atrium (RA) [1]. As an etiology, Roberson [2] suggests that if the accessory membrane is in the left atrium (LA), it may be an abnormality of the atrial primum septum, while when the layer is on the right side, then it is formed by accessory structures.

According to van Praagh and Corsini [3] the accessory septal structure distinguishes this space from the RA, it is presumed it might be a persistent left ventricular valve attached to the sinus venosus in the fetal period.

Taking into consideration the fact that the double septum may be secondary to an intramural hematoma, that may delaminate the interatrial septum. The extra layer of DAS must be distinguished from the cor triatriatum, where the supplemental membrane runs parallel to the mitral annulus (cor triatriatum sinistrum) or parallel to the tricuspid

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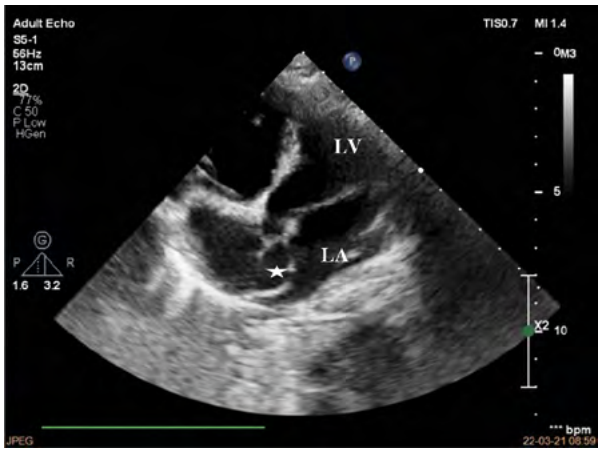

Figure 1. Echocardiography, 4-chamber view. In the LA additional septum. Free space and no thrombus between 2 layers (asterisk); LA — left atrium; LV — left ventricle

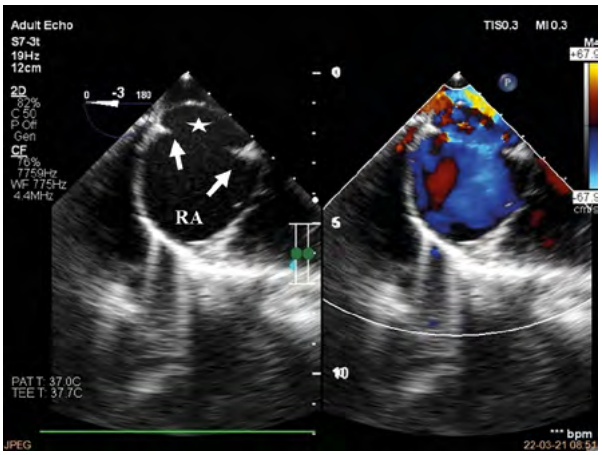

Figure 2. Transoesophageal echocardiography. Large intra-atrial defect (2 cm) of the ostium secundum type (arrows). Enlarged right atrium (RA). In the left atrium additional septum and free space. No thrombus between 2 layers (asterisk). In the color Doppler, undisturbed blood flow between the atria

annulus (cor triatriatum dextrum). What is vital, in DAS the mitral valve connects freely with the pulmonary veins, unlike the triatrial heart [4].

In children DAS is usually asymptomatic and, similarly to adults, it is diagnosed incidentally [5]. However, clots, that may later become an embolic material, can form in the space between the 2 layers, causing a transient ischemic attack or full-spectrum ischemic stroke [6, 7]. Single cases of DAS have been described in the world literature [8]. Among children, one of the youngest patients described in the literature was a case of a 6-year-old boy, whose only symptom was a murmur over the heart. Further diagnostics of the murmur revealed DSA, which was not taken into consideration in the initial differential diagnostics [9].

Case report

A 4-year-old boy was referred to the pediatrician due to recurrent infections and the next he was referred for a cardiological consult. Transthoracic echocardiography revealed a large defect in the atrial septal defect type ostium secundum of 2 cm in diameter and within the lumen of the LA, the presence of a second septum was established (Figure 1). In addition, the RA and the right ventricle were enlarged, as well as the pulmonary artery was dilated. Color Doppler examination revealed a profuse left-right shunt through the atrial septum. The electrocardiogram showed a right axis deviation and features of a partial right bundle branch block. The child was qualified for transoesophageal echocardiography (TEE), which was performed in a hemodynamic laboratory with intention of simultaneous interventional closure.

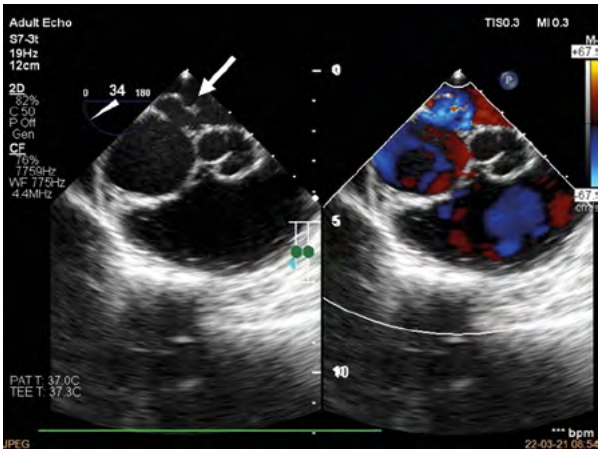

Figure 3. Transoesophageal echocardiography. In the left atrium normal and additional septum and free space between them (arrow)

TEE examination confirmed the presence of two partitions in the atrium, with a space in between the layers, excluding the existence of a thrombus (Figure 2 and 3). The decision on interventional closure of atrial septal defect has been changed and the boy was qualified for cardiac surgery.

Discussion

The interatrial defect is one of the most common congenital heart defects [10], while the detection of an additional septum inside the RA or LA is extremely rare. There are

single case reports in the world literature [8, 9]. The present patient was only 4 years old and, to the authors' knowledge, is probably one of the youngest to be diagnosed with this defect. If the accessory layer is located on the side of the LA, then the space between the septum may be a source of thrombus and cerebral embolism or other complications [6, 7]. Therefore, especially in the case of an increased risk of embolism (e.g., congenital thrombophilia), the patient should be qualified for surgical removal of the accessory septum and defect closure. Transthoracic echocardiography, supplemented with TEE, is usually a modality sufficient enough to assess the defect, although it sometimes requires an extension to computed tomography or magnetic resonance imaging.

The differential diagnostics of DAS should include the three-atrial heart (most often left-sided), in which the additional septum is located in the LA and is parallel to the mitral annulus

Conclusions

During echocardiography, the assessment of the atrial septum must be conducted with precision. Any unusual image of the atrial septum should prompt TEE and other imaging methods. The double atrial septum creates a high risk of thrombus formation and stroke.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organizations that could adversely affect the content of the publication and claim the right to this publication.

Funding

None.

Streszczenie

Praca dotyczy przypadku 4-letniego chłopca, u którego w badaniu echokardiograficznym zdiagnozowano duży ubytek przegrody międzyprzedsionkowej typu *ostium secundum* (ASD2) oraz dodatkowe nieprawidłowe struktury od strony lewego przedsionka. Pacjent został zakwalifikowany do interwencyjnego zamknięcia ubytku, jednak w echokardiografii przezprzełykowej oprócz bardzo dużego ASD2 (ok. 2 cm) rozpoznano podwójną przegrodę międzyprzedsionkową. Warstwa dodatkowej ściany była równoległa do przegrody międzyprzedsionkowej i nie utrudniała przepływu krwi z żył płucnych ani przez zastawkę mitralną. W przestrzeni pomiędzy dwiema warstwami nie stwierdzono skrzeplin. Zrezygnowano z zamknięcia ASD2 podczas cewnikowania serca i pacjenta zakwalifikowano do planowego leczenia operacyjnego. Obecnie chłopiec nie wykazuje objawów i prowadzi normalny tryb życia.

Słowa kluczowe: podwójna przegroda przedsionkowa, przestrzeń międzyprzedsionkowa, wrodzona wada serca

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