Introduction

The Amplatzer septal occluder (ASO) has become the device of choice for interventional closure of atrial septal defects (ASDs) in many institutions during the last decades. Although excellent results have been reported for the device, concerns have arisen about the long-term complications (1). Of these complications, thrombus formation was rarely seen after 1 year in patients (2–4). This is the first report of a pediatric patient of a huge thrombus developing on an ASO device detected by transthoracic echocardiography on a routine examination after 1 year of implantation without a risk factor.

Case Report

A 17-year-old boy had been diagnosed with an ASD during evaluation for cardiac murmur. Transthoracic echocardiography (TTE) showed a 14-mm ASD and moderate dilation of the right ventricle. Transesophageal echocardiography during cardiac catheterization revealed a 16-mm ASD with the balloon sizing technique. The pulmonary-to-systemic flow ratio was 2.8. An 18-mm ASO was successfully implanted, and the patient was discharged with the prescription of aspirin (300 mg/day) for 6 months. TTE 4 weeks, 3 months, and 6 months after the procedure showed the device in place.

At the 1-year follow-up, TTE revealed huge mobile thrombus with a diameter of 34×62 mm attached to the left atrial disk of the device (Fig. 1 and Video 1). He was taken to surgery for removal of the thrombus and the device. After right atriotomy, a well-endothelialized occluder device was seen and was excised with the large thrombus (Fig. 2). There was no device fracture or dislocation. The newly created ASD was closed by pericardial patch. The patient had an uneventful recovery and was discharged the third day after surgery. The pathological examination of the material was compatible with thrombus formation. The thrombus consisted of peripheral blood elements and fibrin. There was no acute inflammation and granulomatous inflammation.

Coagulation assays were performed in order to identify an inherited thrombosis disposition. The screening, which included the measurement of protein C and S, antithrombin III, homocysteine, anti-phospholipid antibodies, and lupus anticoagulance were normal. Furthermore, the patient had no factor II or factor V Leiden mutation.

Discussion

Thrombus formation on the transcatheter closure devices, which could lead to systemic embolization, is one of the major concerns with these implants. Although it has been noted up to 5 years it was usually seen in early period after device placement (5, 6). All commercially available devices had at least one reported case of thrombosis, but in a recent study involving 407 patients with ASDs, the Amplatzer occluder has been found to be less thrombogenic than the other devices (2, 7). There was only one child with ASD that presented with a very late device thrombus formation after percutaneous Amplatzer device closure (8). Furthermore, the predisposition to thrombosis was present in most of patients reported at previous studies.
Recommendations regarding specific anticoagulation therapy after device implantation remain controversial (9). It is accepted that 6 months of aspirin alone is usually effective in preventing early thrombus formation on the device. However, there was one patient with nonendothelialization of the left atrial disk 32 months after ASO device placement (10). Heparin, at a dose of 100 U/kg during implantation, was given to the patient, and aspirin alone was used to prevent thrombosis for 6 months since our patient had no history of pre-thrombotic event. Also, there was no coagulation disorders detected in our patient. Nevertheless, huge thrombus was detected at the central part of the left side of the well endothelialized Amplatzer device after 1 year of implantation. Therefore, additional long-term follow-up studies are needed to reevaluate the duration or type of anticoagulation in children with closure devices.

Although TEE was shown to be more sensitive than TTE in detecting thrombus formation in adults, follow-up by TTE as an imaging perspective might be sufficient for younger children. We also preferred to follow-up all children with closure devices by TTE because of good echocardiographic windows. For this reason, it is not clear whether the thrombus was present at the early period after device implantation with TEE imaging. Therefore, it was emphasized that more studies are required to determine the choice of imaging method after device implantation in children with ASDs even with a good quality of transthoracic imaging.

**Conclusion**

To the best of our knowledge, this is the first reported case of a child with late huge thrombus on an Amplatzer device without any known risk factor. Additional longer follow-up studies are warranted in children to determine the duration and the type of antiplatelet therapy and the preference of imaging technique after device implantation.

**References**


**Video 1.** Transthoracic echocardiography of the patient with huge thrombus on the left atrial disk of the device (parasternal long-axis view).

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**Treatment of pulmonary hypertension in three patients with β-thalassemia intermedia using pulmonary arterial hypertension-specific medications**

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**Introduction**

Pulmonary hypertension (PH) is frequent among patients with β-thalassemia intermedia (TI) and β-thalassemia major (TM) (1). Almost 60% of all TI patients develop PH (2). However, no randomized controlled trials have evaluated this condition-specific treatment options. Recent guidelines for the treatment of PH offer no specific recommendations for these patients; moreover, the classification of chronic hemolytic anemia was changed from group I PH to group V PH, in which group pulmonary arterial hypertension (PAH)-specific therapy is not recommended (3). We report three patients with β-TI who developed severe PH and were successfully treated with PAH-specific therapies.

**Case Reports**

**Case 1**

A 39-year-old man with β-TI was admitted with new-onset dyspnea and fatigue. On further examination, systolic pulmonary arterial pressure (sPAP) was measured as 115 mm Hg on trans-