Fungal Endocarditis After Hybrid Periventricular Closure of Muscular Ventricular Septal Defect by Amplatzer Occluder in a Child

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A 2-year-old boy was admitted for a 16-day history of intermittent fever (up to 40.5°C) with no other symptom. He had been followed since birth for a large midmuscular ventricular septal defect with a left-to-right shunt and signs of congestive heart failure. At 2 months of age, the patient had a pulmonary artery banding, and at 16 months of age he underwent hybrid periventricular closure of a muscular ventricular septal defect by Amplatzer occluder and removal of the pulmonary band.

A 12-mm Amplatzer muscular ventricular septal defect (VSD) occluder was implanted through a direct puncture of the right ventricular free wall. Postoperative echocardiography initially showed a 7-mm residual muscular VSD at the inferior border of the device with a moderate left-to-right shunt that progressively decreased to a 3- to 4-mm residual VSD over the next few months. During postoperative follow-up, the patient’s condition improved with decreasing heart failure signs. Treatment with diuretics and angiotensin-converting enzyme inhibitor, and aspirin as well, was continued. Over the next 8 months he received several courses of antibiotics for various infections.

On admission, the patient was subfebrile (37.7°C). His heart rate was 144 bpm, blood pressure was 121/75 mm Hg. Chest examination revealed a holosystolic murmur at the midleft sternal border with irradiation to the apex. There were mild subcostal retractions, diminished vesicular sound, and expiratory crackles on the right basal lung field. Laboratory examination showed elevated inflammatory signs: leukocytosis at 24.6 g/L, elevated C-reactive protein at 283 mg/L, and elevated sedimentation at rate 71 mm/h. Chest x-ray showed diffuse bilateral infiltrates and right lower lobe condensation.

Endocarditis was suspected by transthoracic echocardiography and confirmed by transesophageal echocardiography and blood cultures.

Transesophageal echocardiography showed a large irregular echogenic mass measuring 22×12 mm, attached to the right disc of the VSD occluder, that did not compromise the tricuspid valve nor cause tricuspid regurgitation (Movie I in the online-only Data Supplement). A 4×3 mm residual muscular VSD was seen at the lower border of the VSD occluder device.

Successive blood cultures were positive for multisensitive Candida albicans.

Therefore, the patient was started on intravenous antifungal treatment with caspofungin. His physical condition improved, but the fever persisted. Laboratory values for inflammation decreased. A repeated transesophageal echocardiogram 6 days later showed no changes and no reduction in the size of the large vegetation.

Subsequently, because of the fungal nature of the endocarditis and the infection of prosthetic material, the patient underwent surgery with cardiopulmonary bypass. The VSD occluder, and the large vegetations and infected tissues as well, were removed (Figure 1). The VSD was closed by pericardial patch.

The macroscopic examination of the device showed a partially endothelialized device and focal vegetations (Figure 2). The microbiological analysis of the removed vegetation revealed the presence of C albicans as in the previous blood cultures.

Intravenous treatment with caspofungin was continued for 12 weeks, and thereafter he was placed on oral fluconazole for 9 months. The patient has now remained asymptomatic for 15 months since surgery without any signs of recurrent infection.

To our knowledge, this is the first report of candida endocarditis on a VSD occluder device implanted by hybrid therapy in a child. Hybrid therapy rather than conventional surgery in this case was chosen for several reasons. First, the distal localization of the muscular VSD, and the extension from the inlet to the outlet part of the interventricular septum as well, made a surgical closure technically difficult. In fact, the exposure of distal muscular VSD can be problematic because of the numerous trabeculations. Second, the hybrid approach permitted the avoidance of a right or left ventriculotomy. Only a direct puncture in the right ventricular free wall was performed. A surgical approach with a right or left ventriculotomy to access the muscular VSD can lead to extensive...
myocardial damage, ventricular dysfunction, and ventricular arrhythmia. Third, this approach permitted the extraction of the pulmonary band and the closure the VSD during the same intervention, avoiding cardiopulmonary bypass.

Current guidelines for the indication for VSD closure are well defined. Hybrid therapy has several advantages over surgical treatment or catheter intervention alone, but the patient selection for this approach has not yet been well established.\(^1\) Some groups of patients have been identified to be better managed with a hybrid approach, such as infants with isolated muscular VSDs, patients with muscular VSD and other associated lesions, and patients with muscular VSDs and pulmonary artery band.\(^2\)

As previously reported, residual shunting is frequently seen after VSD device closure.\(^1\) In this case, the initial residual VSD progressively reduced in size and had no hemodynamic impact. Therefore, there was no indication for reintervention. According to current guidelines for prevention of infective endocarditis, repaired congenital heart diseases with residual defects at the site or adjacent of the prosthetic device are an indication for endocarditis prophylaxis, because it can inhibit endothelialization. Fungal endocarditis is a rare entity among immunocompetent children. However, congenital heart defects such as VSDs, corrective therapy with prosthetic material, and multiple broad-spectrum antibiotics are known risk factors associated with developing fungal endocarditis.\(^3\)

This case illustrates that persistent or prolonged fever in children with residual VSDs after device occluder placement should raise the suspicion of endocarditis. Fungal infections are infrequent; however, previous multiple antibiotic trials, as shown in our case, are found to be a risk factor. Treatment combines antifungal therapy with ablation of the device.

**Disclosures**

None.

**References**


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