

Miniivasive hybrid closure of multiple muscular ventricular septal defects in a premature infant with novel use of Amplatzer Duct Occluder II – a case report

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Abstract

Muscular ventricular septal defects (mVSD) appearing together with other septal defects are frequently regarded as “concomitant” pathologies, that nevertheless should be considered while the patient is referred for intervention. We followed a conception of mVSDs’ miniivasive treatment with a hybrid approach based on perventricular implantation of occluding devices. In this paper we report a hybrid procedure performed in a premature infant referred for surgical correction of a large perimembranous VSD with a simultaneous perventricular approach for concomitant muscular ventricular septal defect. The device of choice, because of the patient’s small size and weight, was the Amplatzer Duct Occluder II. Colour Doppler showed complete closure of all VSDs 8 months after surgery with no complications related to the procedure.

Key words: ventricular septal defect, device closure, hybrid procedures, prematurity, congenital heart defects, miniinvasive cardiac surgery.

Introduction

Muscular ventricular septal defects (mVSD) appearing together with other septal defects are frequently regarded as ‘concomitant’ pathologies, that nevertheless should be considered while the patient is referred for intervention.

Therapeutic decisions for accompanying mVSDs that do not close spontaneously in early infancy are usually undertaken with regard to fundamental defects and the risk of congestion, or the effects of haemodynamically significant pulmonary shunt in the postoperative course. Heart failure, or ongoing development of pulmonary vascular disease, is not to be neglected in the natural history of under-treated patients with the “Swiss cheese” phenomenon.

Our surgical strategies for multi-VSDs thus far have rarely led to definitive septation and certain closure of mVSDs in infancy [1]. Different surgical approaches that have been proposed gave overall mortality and residual shunt risk higher than surgery for neonatal perimembranous VSDs [2]. A commonly followed strategy, although regarded “old-fashioned”, is a two-stage approach with pulmonary artery banding, guided by expectations that multi-VSDs in older children will be a challenge for interventional cardiologists.

The promising conception of multi-VSD closure in infants seems to be a hybrid approach with intraoperative perventricular implantation of occluding devices [2, 3]. There are several technical limitations, such as diameters of the occluder and delivery sys-

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tem, that need to be considered before implantation in the ventricular septum in an infant.

In this paper we report a miniinvasive hybrid procedure performed in a premature infant referred for surgical correction of a large perimembranous VSD with a simultaneous periventricular approach for concomitant muscular ventricular septal defect. The device of choice, because of the patient's small size and weight, was the Amplatzer Duct Occluder II. We chose the device as a substitute for VSD occluders because of the delicate discs and small dimensions, not available in ventricular defect closure implants.

Case report

A congestive premature male child, of 3 kg body weight, was admitted in the 2nd month of life to our department with the diagnosis of a large perimembranous VSD, patent arterial duct (PDA), and muscular ventricular septal defects, with respiratory disorders because of bronchopulmonary dysplasia (SaO_2 85%). Precise preoperative echocardiography showed the largest (8.6 mm) perimembranous defect, with left-to-right flow and peak systolic gradient of 10 mmHg, and multiple VSDs in the muscular part of the ventricular septum ("Swiss cheese" type defect) (Figure 1). Left ventricle enlargement (130% of normal size) and poor contractility were found. The aortic valve and arch vessels were normal sized, without any symptoms of dysfunction.

Prior to admission the boy deteriorated despite maximal medication and permanent oxygen supply,

and therefore was referred for urgent surgical treatment because of pulmonary overflow and significant desaturation ($\text{SaO}_2 < 85\%$).

After median sternotomy standard Pacifico system cardiopulmonary bypass (CPB) was commenced and the 8.6 mm perimembranous VSD was closed after hypothermic cardioplegic arrest with a Dacron patch in a classic mode. Muscular VSDs were difficult to localize in flattened trabeculations of the septum. After closing the heart and the reperfusion epicardial echocardiography showed maximal diameter of the largest muscular VSD of 4 mm, the pump was stopped. The right ventricle was punctured under epicardial echo imaging and the guide wires followed by a 6 F vascular sheath were inserted through the largest muscular VSD. A 4-mm Amplatzer Duct Occluder II (ADO II) (AGA Medical, Golden Valley, Minnesota, USA) was implanted into the septal defect on the beating heart (Figure 2). Control echocardiography showed complete closure of perimembranous and concomitant muscular defects. ADO II position was stable with the right disc close to the VSD patch, and no residual left-to-right leakages between ventricles were found (Figure 3).

The early postoperative period was uneventful, and the boy was successfully extubated 12 hours after the procedure. No arrhythmias or ischaemia were found, although dyspnoea and desaturation related to bronchopulmonary dysplasia needed continuation of oxygen mask delivery. Nevertheless, the boy was transferred to the neonatology department on postoperative day 8 for further treatment. The observation

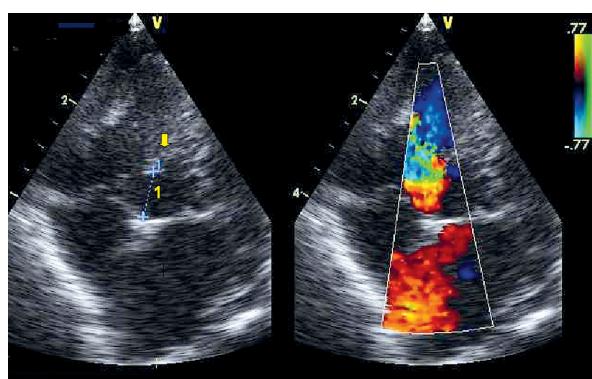


Figure 1. Preoperative echocardiography showed the largest (8.6 mm) perimembranous ventricular septal defect (1), with left-to-right flow and peak systolic gradient of 10 mmHg, and accompanying additional VSDs in muscular part of ventricular septum (arrow)

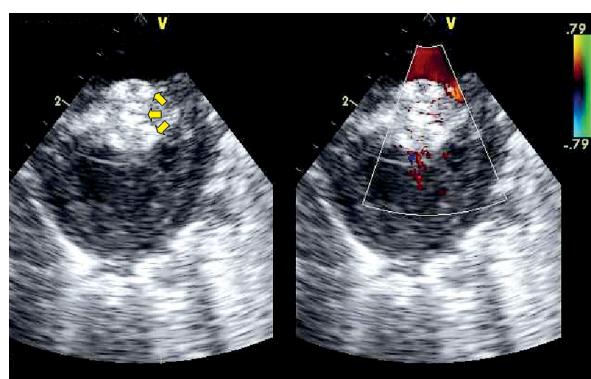


Figure 2. Four-millimetre Amplatzer Duct Occluder II (AGA Medical, Golden Valley, Minnesota, USA) was implanted into the largest muscular septal defect (arrows) – epicardial echocardiography

period after the hybrid procedure is 8 months. Control echocardiography after discharge showed improved left ventricular function, normal atrioventricular valves, and correct position of both the Dacron patch and the ADO II occluder in the muscular septum. The antiplatelet aspirin was administered for 3 months after hybrid surgery.

Discussion

Coexistence of congenital heart defects and additional muscular septal defects leading to oversized systemic-to-pulmonary blood flow increases the risk of pulmonary hypertension and heart failure [4]. In this specific group the decision of challenging surgical correction is questionable with regard to morphology of basic defects, patients' weight and age, as well as additional problems. Therapeutic strategies in small premature babies are more complicated, because it seems beneficial to postpone the time of surgical intervention until the child reaches proper weight. Nevertheless, classic surgical closure of muscular defects remains troublesome because of bad visualization of the muscular septum in the empty heart. Muscular VSDs are usually hidden in the coarse right ventricular trabeculations [1, 2]. These arguments prompt some authors to wait with definitive surgery until body weight is suitable for transcatheter VSD closure [1, 2]. In decompensated patients with a bad response to intensive medical treatment, failing to thrive, low body weight or prematurity problems, the only option is emergency surgery in early infancy [2].

In the presented child the decision to perform surgical correction was made because of coexistence of pulmonary overflow and bronchopulmonary dysplasia, that necessitated perimembranous VSD closure. PA banding remained suboptimal because of pulmonary problems and initial desaturation. The ADO II occluder, our device of choice, was the smallest and was delicate enough to be implanted in the small, thin intraventricular septum of the premature infant. An occluder of appropriate size was not offered in the catalogues of commercially available VSD occluding devices.

The key to success of every minimally invasive hybrid approach is perfect cooperation of interventional cardiologists, surgeons and anaesthesiologists. In the presented child the appropriate approach and exposure of the heart were easier because of

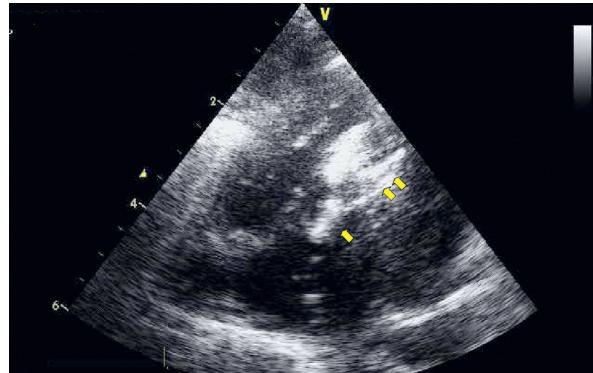


Figure 3. Control echocardiography showed complete closure of concomitant muscular defects with Amplatzer Duct Occluder II (AGA Medical, Golden Valley, Minnesota, USA), which is shown (two arrows) near perimembranous patch (arrow), with no residual left-to-right shunt between ventricles

stand-by CPB, which was stopped before the hybrid procedure. We achieved satisfactory epicardial echo imaging which precisely showed the defect and enabled a successful periventricular implantation procedure. Transoesophageal echocardiography was difficult because of the patient's low weight and probe size.

In the reported patient the device implantation supplemented complete closure of septal defects, which could not be achieved in a classic surgical approach. Implant endothelialisation, a reaction that starts early after the procedure, usually results in definitive closure of residual shunts after 6 to 10 months, as was confirmed in postoperative echo controls.

Different hybrid approaches have been reported in small groups, but there are several limitations of hybrid procedures in borderline children that are related to technological development of miniaturized implants and delivery systems [4]. After a hybrid approach with artificial elements implanted on the beating heart, heparin is recommended in the early postoperative period, with antiplatelet agents finally administered orally. The most dangerous complications related to hybrid surgeries are displacement of artificial elements, heart injuries, arrhythmias and thromboembolic complications [2, 5]. Taking these limitations into consideration, we chose the ADO II and avoided any complications in our patient, and in summary, we achieved a good final result, with 8 months of follow-up.

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