The Heart Surgery Forum #2010-1081
14 (1), 2011 [Epub February 2011]
doi: 10.1532/HSF98.20101081

Online address: http://cardenjennings.metapress.com

Delayed Closure of Multiple Muscular Ventricular Septal Defects in an Infant after Coarctation Repair and a Hybrid Procedure—A Case Report

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ABSTRACT

There are several strategies of surgical approach for the repair of multiple muscular ventricular septal defects (mVSDs), but none leads to a fully predictable, satisfactory therapeutic outcome in infants. We followed a concept of treating multiple mVSDs consisting of a hybrid approach based on intraoperative perventricular implantation of occluding devices. In this report, we describe a 2-step procedure consisting of a final hybrid approach for multiple mVSDs in the infant following initial coarctation repair with pulmonary artery banding in the newborn. At 7 months, sternotomy and debanding were performed, the right ventricle was punctured under transesophageal echocardiographic guidance, and the 8-mm device was implanted into the septal defect. Color Doppler echocardiography results showed complete closure of all VSDs by 11 months after surgery, probably via a mechanism of a localized inflammatory response reaction, ventricular septum growth, and implant endothelization.

BACKGROUND

Congenital multiple muscular ventricular septal defects (mVSDs) remain a difficult and troublesome clinical problem, especially in newborns and infants with a high-pressure systemic-to-pulmonary shunt. A residual left-to-right shunt in hemodynamically compromised babies is usually comparable to a single-ventricle pathophysiology. The natural history of such pathologies often leads to heart failure or to ongoing development of pulmonary vascular disease and fatal Eisenmenger syndrome, because of the flow-related pulmonary hypertension at a young age.

There are several strategic approaches for the surgical correction of multiple mVSDs, but none leads to a fully predictable, satisfactory therapeutic outcome in infancy, ie, with definitive septation and a 4-chamber physiology [Wollenek 1996]. The different surgical approaches that have been proposed have an overall mortality rate and a risk of residual shunts that are higher than those for the repair of isolated perimembranous VSDs [Holzer 2004]. Multiple mVSDs are still a challenge for interventional cardiologists in search of an optimal treatment and the application of minimally invasive technologies.

A promising concept for the closure of multiple mVSDs in infants is a hybrid approach that involves intraoperative perventricular implantation of occluding devices [Spevak 1993; Holzer 2004]. Good results with this technique, as well as for transcatheter occluder implantation, have been described and recommended for treating mVSDs in children older than 12 months. In this report, we describe a 2-step procedure for infant patients consisting of a hybrid approach of an initial coarctation repair with pulmonary artery (PA) banding in the newborn and subsequent repair of multiple mVSDs in the infant.

CASE REPORT

A female child, 3 kg body weight, was admitted on the fifth day of life to our department with an initial diagnosis of critical aortic coarctation, patent ductus arteriosus (PDA), and mVSDs. Continuous prostaglandin E infusion permitted a precise preoperative echocardiography evaluation, which revealed multiple mVSDs in the muscular part of the ventricular septum ("swiss cheese" type of defect). The diameter of the largest defect was 5 mm, with a left-to-right flow and a peak systolic gradient of 15 mm Hg. Left-to-right atrial flow through a 5-mm-wide foramen ovale was also observed. The aortic valve was bileaflet with no signs of dysfunction. The distal part of the aortic arch was hypoplastic (less than one third of the ascending aorta diameter) with a typically localized 1.7-mm coarctation and a 4-mm PDA with blood flow from the PA to the descending aorta.

Surgical repair of the coarctation with an "extended end-to-end" anastomosis was performed via a left thoracotomy on the sixth day of life, with PA banding and PDA closure. The postoperative course was uneventful.

A control echocardiography examination after 2 months revealed a slight hypertrophic right ventricle with active

Received June 19, 2010; received in revised form August 19, 2010; accepted August 26, 2010.

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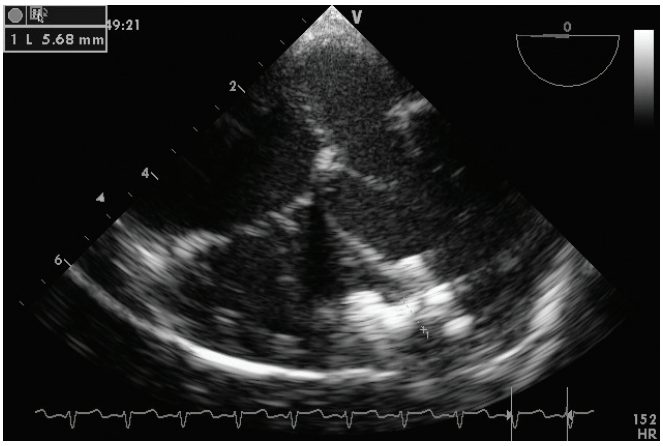

Transesophageal echocardiography image of an AMPLATZER Muscular VSD Occluder (AGA Medical) implanted into a muscular septal defect.

multiple mVSDs and balanced flow. The largest defects were decreased to 4 mm in diameter; nearby defects were 1 to 2 mm in diameter. The PA band gradient was 60 mm Hg; the peripheral oxygen saturation was 90%.

In the seventh month of life, the girl was referred for surgical treatment because of significant desaturation (oxygen saturation <85%). She had an appropriate weight (5 kg) for perventricular hybrid VSD closure and PA debanding. After a median sternotomy and standby cardiopulmonary bypass, the PA band was removed. After meticulous resection of pericardial adhesions, we performed a transesophageal echocardiography (TEE) evaluation, which showed a mild pressure gradient over the PA (<20 mm Hg). The maximum diameter of the largest VSD measured after PA debanding was 6 mm. The right ventricle was punctured under TEE guidance and the guidewires, followed by a 6F vascular sheath, were inserted through the largest muscular VSD. An 8-mm AMPLATZER Muscular VSD Occluder (AGA Medical, Golden Valley, MN, USA) was implanted into the septal defect (Figure). TEE imaging showed partial closure of concomitant smaller defects that were close to the largest one, along with residual left-to-right leakage between ventricles.

The postoperative period was prolonged. The girl was ventilated with a peak positive-pressure mode typical for patients with pulmonary overflow and was extubated successfully on the sixth postoperative day. We found no arrhythmias or ischemia, although we did detect questionable peripheral leakage in the occluder area. Nevertheless, the girl was discharged home on postoperative day 14 in good general condition. A planned scintigraphy evaluation at 1 month after the operation revealed a pulmonary-to-systemic flow ratio (Qp:Qs) of 1.8:1.

The observation period after the hybrid procedure is now 24 months. A control echocardiography examination at 10 months after discharge showed a normal left ventricular function, mild right ventricular hypertrophy, normal atrioventricular valve functions, physiological flow in the aortic arch (1.8 m/s), a residual PA gradient (10 mm Hg), and a correctly positioned muscular VSD occluder. A color Doppler evaluation 11 months after surgery showed complete closure of all VSDs. Antiplatelet aspirin therapy was stopped 3 months after the hybrid surgery.

The results of additional follow-up evaluations were uneventful, and at 2 years after the procedure, the girl remains free of any cardiac symptoms and medications, with normal growth and neurophysiological development.

DISCUSSION

Multiple mVSDs coexist with other congenital heart defects, and several reports have described the coexistence of mVSDs with coarctation of the aorta [Crossland 2008]. Concomitant congenital heart defects and excess systemic-to-pulmonary blood flow may increase the risk of pulmonary hypertension and left heart failure. In this particular group of patients, the decision to perform a challenging surgical correction is uncertain and depends on the morphology of the defects, the patient's weight and age, the time of the intervention, and the overall therapeutic strategy. In smaller babies, it is beneficial to postpone the time of surgical intervention until the child reaches the proper weight. Nevertheless, classic surgical closure remains troublesome because of the expected poor visualization of muscular VSDs in the empty heart, which are hidden by the courses of right ventricular trabeculations [Wollenek 1996; Holzer 2004]. Some authors have therefore argued that definitive surgery should wait until the patient's body weight is suitable for transcatheter VSD closure. In patients with a bad response or contraindications to intensive medical treatment, a failure to thrive, a low body weight, and a significant risk of pulmonary hypertension, the best option is to commence any surgery at the end of late infancy [Holzer 2004].

In the presented patient, the decision for a 2-step strategy was made because of the coexisting critical aortic coarctation, which necessitated surgical repair of the arch, and the left thoracotomy approach, which was more suitable for PA banding than for multiple mVSD surgery. Another important argument for our decision was the induced stimulation of right ventricular hypertrophy and the ventricular septum in response to PA banding.

Success with the hybrid approach depends on perfect cooperation between the interventional cardiologists, surgeons, and anesthesiologists. In the presented child, the appropriate choices regarding the approach, exposure of the heart, and ischemia-protection protocols (preconditioning period, optimal exposure of ventricular septum) were similar to those of the off-pump coronary bypass strategy in adult patients. We achieved satisfactory TEE imaging, which enabled the precise and successful completion of the perventricular implantation procedure.

In the early postoperative period, we observed symptoms of pulmonary edema because of the slow decrease in the excess pulmonary blood flow and unrestrictive physiology of the pulmonary bed, which was flooded because of residual left-to-right shunts, after PA debanding.

Given our observations of the delayed closure of the residual shunts, we believe it is unnecessary to close all exposed

small coexisting VSDs that are near each other [Spevak 1993]. In our case, the occluder device stimulated delayed closures of nearby defects, probably via a mechanism of a localized inflammatory response reaction, ventricular septum growth, and implant endothelization. These reactions led to complete closure of residual shunts after 6 to 10 months, a result that was confirmed in postoperative echocardiographic control examinations.

The use of a hybrid approach has been reported for small groups of patients, but there are several limitations on the use of hybrid procedures in children [Crossland 2008]. After a hybrid approach involving the implantation of artificial elements in the beating heart, heparin administration is recommended in the early postoperative period, followed finally with orally administered antiplatelet agents. The most dangerous complications related to hybrid operations are the displacement of artificial elements, heart injuries, arrhythmias, and thromboembolic complications [Holzer 2004]. With these limitations in mind, we were able to avoid any complications in our patient.

In summary, we have achieved a good final result, with 24 months of uneventful follow-up. After obtaining these promising initial results, we have recommended the use of this strategy for other patients. Thus, we have begun a hybrid surgery program in our department for the repair of multiple mVSDs.

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