Hybrid VSD closure in a 4.5 kgs Neonate Case Report and Literature Review

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Abstract

Ventricular septal defects are the most common congenital heart defects. They account for approximately 20% of all forms of congenital heart disease as an isolated lesion. Perimembranous Ventricular septal defects are the most common form (70%), followed by muscular (15–20%).Muscular VSDs are a challenging problem in neonates and infants when they present with significant congestive heart failure from interventricular shunting. However, with careful adjustments to technique, most of these can be closed.

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Abstract

Ventricular septal defects are the most common congenital heart defects. They account for approximately 20% of all forms of congenital heart disease as an isolated lesion. Perimembranous Ventricular septal defects are the most common form (70%), followed by muscular (15–20%).Muscular VSDs are a challenging problem in neonates and infants when they present with significant congestive heart failure from interventricular shunting. However, with careful adjustments to technique, most of these can be closed.

Introduction

Ventricular septal defects are the most common congenital heart defects. They account for approximately 20% of all forms of congenital heart disease as an isolated lesion. When associated with multiple congenital heart defects, the incidence rises to 40%. Perimembranous Ventricular septal defects are the most common form (70%), followed by muscular (15–20%) and sub arterial (5%) types of ventricular septal defect[1]. Depending on the size of the defect pulmonary hypertension may develop even as early as 18 months to 2 years

of age if a large VSD is left unrepaired [2]. Smaller infants with muscular VSDs presented inherent limitations of both surgical and percutaneous device closure so a hybrid approach, also called "perventricular closure", was introduced [2]. The initial descriptions included six infants and seven mini pigs[3] and showed encouraging results in both cases, without the need to utilize cardiopulmonary bypass[3,4]. Muscular VSDs are a challenging problem in neonates and infants when they present with significant congestive heart failure from interventricular shunting. However, with careful adjustments to technique, most of these can be closed[5].

Case Report

A 4-day old male neonate was transferred to our neonatal unit from the local hospital for continuation of care. He had been diagnosed at the 22+6-week scan with common atrioventricular septal defect with a small ventricular septal defect and a large ostium primum septal defect. An amniocentesis yielded normal karyotype results. The neonate was born via caesarean section due to previous caesarian at 40 weeks gestation with a birth weight of 4350 grams.

He was transferred to our hospital due to desaturations and perioral cyanosis. At 20 days of age, he started to have episodes of tachypnea and diaphoresis during feeds. On Day 26 of life, he underwent a pulmonary artery banding. The post op echocardiogram showed "Common atrioventricular canal type A, loose Pulmonary Artery banding with a PG 17.5 mmHg(Figure 1), large mid muscular Ventricular septal defect measuring 5.8-6.7 mm with bilateral flow, small inlet ventricular septal defect measuring 2 mm, large ostium primum atrial septal defect and large ostium secundum atrial septal defect". He was difficult to wean off the ventilator and required inotropic support. Therefore, on the 38th day of life, he underwent a cardiac catheterization (Figure 2) which showed

1)Large muscular VSD after PA banding, PAP 37/10 mean 20mmHG, Qp/Qs1.3 L/min, PVR 1.46 WU 2) Hypertrophied right ventricle with good contractility 3) Moderate muscular type ventricular septal defect with smaller outlets towards the right ventricle 4) Large Atrial septal defect 5) Moderate Reversible pulmonary hypertension.

He underwent a new hybrid cardiac operation with patch closure of the atrial septal defect, closure of the ventricular septal defect with a device, repair of the left and right atrioventricular valve and lung biopsy. The device was positioned after right atriotomy and under direct vision, through the tricuspid valve. The device had a diameter of 5mm.Amplatzer duct occluder ii size 9-2-6-4 was used with disc diameter 12mm, waist 6mm and waist length 4mm (figure 3). The device charging system was used but a short 6Fr sheath was used as a release system. The lung biopsy showed moderate architecture disturbance, heterogenous thickening interalveolar diaphragms, fibroblast hyperplasia without fibrosis, siderophages as well as mild to moderate pulmonary hypertension (Heath Edwards I), mild chronic inflammatory infiltration.

On the 103 day of life, he underwent a new tightening of the Pulmonary artery banding. There were numerous attempts at weaning off the ventilator, which the patient did not tolerate, and he underwent a tracheostomy insertion on day 139. The post operative course was complicated by thrombosis of right common and superficial femoral vein, with concomitant oedema of the surrounding tissues and lymph glands on day 53, managed tinzaparin. On Day 115, after an episode of sepsis, he had an encased fluid collection was observed in the minor pelvis. The venous catheter was removed and the anticoagulation was reversed with tranexamic acid. A CT pelvis was done on day 116 and revealed hematoma applying external pressure on the bladder. 5 days later, he was taken to theatre, for removal of the foreign body from the left iliac vein. On the 29th day of life, after starting enteral feeds, he developed chyloperitoneum was made and enteral feeding was stopped. He was fed enterally with MCT milk and octreotide, and TPN until day 41 when octreotide was stopped. He remained well, tolerating enteral nasogastric feeds. The final echocardiogram showed, PA banding with PG max 35 mmHg (mean 17.6mmHg), small residual flow from left to right by the VSD closure device and a 2nd apical muscular VSD with left to right flow with a PG 24mmHg, 1+/4+ regurgitation of the left atrioventricular valve with a PG of 75 mmHg, intact atrial septum. He remained well. He established feeds. The tracheostoma was closed successfully at 6 months of age.

Discussion

Even as early as 1999, perventicular as well as interventional ventricular septal defect closures proved effective even is small infants from 3.2 to 8.9 kgs [6]. Over the course of the next 8 years perventicular approach in mostly muscular Ventricular septal defects device placement was successful (88.5%) even on large defects in infants 3.5-14 kgs [7]. Similarly, larger cohorts of infants from 25 days to 8.9 years old weighing 4-12.9 kgs have demonstrated success rates upwards of 90%[8,9,10]. Similar success rates for perventicular muscular VSD closure have been reported in infants 3.4-10 kgs as well[11,12]. More, in the case of muscular VSD as residual lesions for a more complex congenital heart disease, perventricular approach has been used with shorter procedural times and good result[13]. Successful implementation of perventricular approach has been reported is isolation [14], in conjunction with other hybrid procedures [15], in other lesions such as truncus arteriosus [16], and various types from apicomuscular [17] to swiss cheese VSDs[18]. The hybrid procedure has been performed on children with a larger weight range from 3 to 30 kgs with a good safety profile [19]. As in our case, collaboration between interventional and surgical teams is required [20,21]. However, reported complications included death, esophageal perforation, complete heart block [22] and pulmonary hypertensive crisis [23].

Conclusion

In small children <15 kgs hybrid perventricular approach provides an excellent alternative access to the heart especially in low-birth-weight infants to prevent hemodynamic instability or in small children requiring large delivery sheaths [25]. The hybrid perventricular technique not only reduces the risk of significant complications compared with the conventional surgery, but also produces not inferior results compared with the transcatheter occlusion in selected perimembranous VSD patients [26]. Although transthoracic device closure of VSD induces a systemic inflammatory response, it seems to be less traumatic and involves a quicker recovery[26].

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