

Hybrid management of a large atrial septal defect and a patent ductus arteriosus in an infant with chronic lung disease

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ABSTRACT

We report a case wherein a dysmorphic four-month-old infant (weighing 4.5 kgs) with an 8 mm atrial septal defect (ASD), a 1.5 mm patent ductus arteriosus (PDA), a 2 mm mid-muscular ventricular septal defect (VSD) associated with chronic lung disease, and severe pulmonary hypertension, was successfully managed using a hybrid approach, without the use of cardiopulmonary bypass (CPB). Through a median sternotomy, the PDA was ligated and the ASD was closed with a 9 mm Amplatzer septal occluder implanted through peratrial access. The VSD was left untouched. Serial echocardiograms showed complete closure of the ASD and PDA, with progressive normalization of the pulmonary artery (PA) pressures within three months. The child rapidly gained weight and was weaned from sildenafil and oxygen administration. After 12 months, the VSD closed spontaneously and the child remained well, with normal PA pressures. A hybrid approach without the use of CPB should be considered in the management of infants with congenital heart disease, associated with chronic lung disease and pulmonary hypertension.

Keywords: Congenital heart disease, interventional catheterization, surgery

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INTRODUCTION

It is generally accepted that the presence of congenital heart defects with significant left-to-right shunts, such as, patent ductus arteriosus (PDA), ventricular septal defect (VSD), and atrial septal defect (ASD), can aggravate the clinical conditions of infants with chronic lung disease, resulting in pulmonary hypertension and increased oxygen and medication requirements.^[1-4] In this situation, surgical repair of the intracardiac defects carries a high risk due to the need of cardiopulmonary bypass (CPB) with its attendant morbidity, especially considering the possible occurrence of pulmonary hypertensive crises in the postoperative period.

In the last decade, a hybrid approach has been successfully employed to treat neonates and infants with a variety of congenital heart defects, such as, hypoplastic left heart syndrome (HLHS) and VSDs.^[5-8] In this article, we report a case in which a dysmorphic four-month-old infant with an 8 mm ASD, a 1.5 mm PDA, and a 2 mm mid-muscular VSD, associated with chronic lung disease and severe pulmonary hypertension was successfully managed without the use of CPB using a novel hybrid approach.

CASE REPORT

A 4 month-old female infant with dysmorphic features (excessive material in chromosome 8) was born premature at a community hospital, at 36 weeks of gestation, weighing 2.5 kgs. She needed mechanical ventilation for a week, due to respiratory distress and infectious complications. After extubation, she remained tachypneic with arterial saturations running in the high 80s range, requiring continuous oxygen administration. A mild regurgitant systolic heart murmur was heard in the left lower sternal border with an increased component of the second heart sound. The chest X-ray showed mild cardiomegaly, a patchy interstitial infiltrate, and increased pulmonary vascular markings. The electrocardiogram (ECG) showed biventricular hypertrophy. The transthoracic echocardiogram (TTE) demonstrated a small PDA (1-1.5 mm), an 8 mm secundum ASD, and a 2 mm mid-muscular VSD, with all defects shunting left-to-right. The systolic pulmonary artery pressure was estimated at two-thirds of the systemic levels. The infant was placed on diuretics (furosemide), vasodilators (captopril), and digoxin. She was discharged from the neonatal unit at

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52 days of age on continuous oxygen administration. At 3.5 months of age (3.9 kgs) she had an adenoviral pulmonary infection, which required mechanical ventilation for two weeks at a pediatric community hospital. Within the first days on ventilation, she had repeated episodes of severe pulmonary hypertensive crisis with desaturation and bradycardia. Suprasystemic pulmonary pressures were estimated by TTE. She was kept on nitric oxide while on ventilation, and sildenafil (2 mg/kg/day) was started at that time. After extubation, she continued to require oxygen despite the completion of a two week-course of corticosteroids treatment. The pulmonary artery systolic pressure was estimated at two-thirds of the systemic on sildenafil. Therefore, she was transferred to our cardiology center for further management. Among the several treatment options, including pharmacological, surgical, and interventional management, a decision was made to perform a hybrid approach with surgical PDA ligation and peratrial device closure of the ASD. Informed consent was obtained from the parents.

The procedure was performed in the operating room through a median sternotomy, under standard transesophageal echocardiography (TEE) and fluoroscopic guidance using a portable C-arm machine. Before surgery, the child weighed 4.3 kgs. TEE confirmed the previous TTE findings and showed an 8 mm ASD with a deficient anterosuperior rim [Figure 1]. The length of the interatrial septum was 25 mm. The PDA was ligated in the usual fashion and the right atrium was punctured at the base of the right atrial appendage. This location was chosen as the entry site after we made sure it would provide the straightest wire course toward the left atrium by pushing the right atrial wall with the index finger tip in various locations and assessing the angle by TEE. The ASD was then crossed using the standard 0,038" J tip short guide wire that came with the short percutaneous

sheath [Figure 2a]. Measurements were made prior, to determine the distance between the entry site and the posterior wall of the left atrium, so that the standard short sheath could be marked with a cotton suture to avoid perforation of the posterior LA wall. This sheath was then carefully advanced over the wire towards the left atrium until the pre-marked point reached the right atrial wall [Figure 2b]. After the wire and dilator had been taken out, a 9 mm Amplatzer ASD device was loaded in the usual fashion and transferred to the 7 Fr sheath. The TEE probe had to be pulled up for some seconds in order to avoid compression of the posterior LA wall and make room for full deployment of the left atrial disk [Figure 3]. The waist of the device was exposed and the whole system was brought back toward the septum as a unit. The left disk approached the interatrial septum at a perfect angle, coming in a parallel fashion toward it [Figure 4 a and b]. The right disk was carefully deployed in the right atrium using the "two-hand" technique, by simultaneously and very slowly retracting the sheath and pushing on the delivery cable [Figure 4c]. The perfect position of the device was confirmed by TEE [Figure 4d]. There was complete closure of the ASD immediately after release of the device [Figure 5 a and b]. The chest was closed in the usual fashion and the postoperative period was unremarkable, with the child being extubated uneventfully the following day. A significant improvement in the respiratory pattern was noted, which allowed for a progressive weaning from the oxygen. She was discharged two weeks after the procedure with a significant weight gain (+500 g), on nasogastric tube feeding. Serial TTEs showed complete closure of the ASD and PDA, and a progressive decrease in the pulmonary artery systolic pressure. After three months, the estimated systolic pressure in the pulmonary artery was 35 mmHg, which allowed for discontinuation of sildenafil, digoxin, vasodilators,

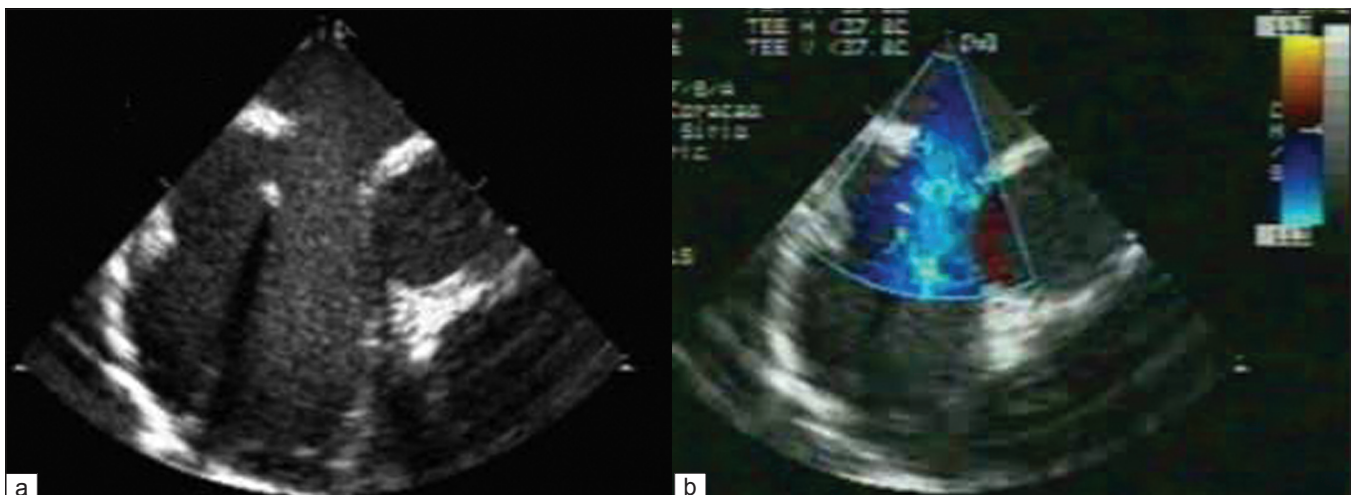


Figure 1: Transesophageal pictures of the atrial septal defect before closure a) Short axis view showing an 8 mm defect with a deficient anterosuperior rim behind the aorta b) Same view with color flow mapping showing left-to-right shunt

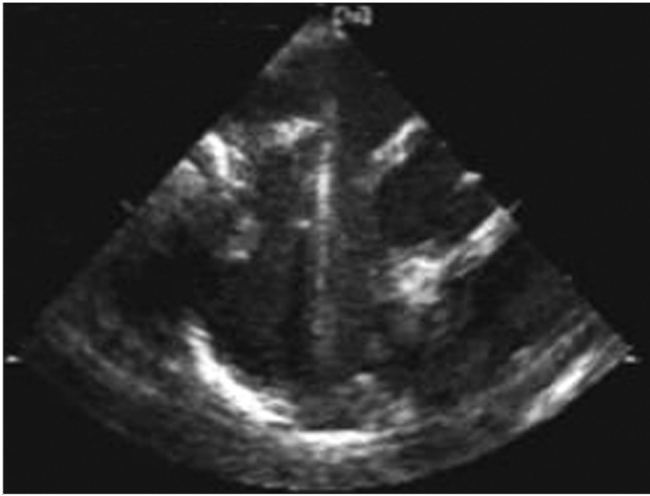


Figure 2a: Transesophageal picture of the procedure displaying the guide wire crossing the atrial septal defect. The base of the right atrial appendage had been punctured with a 17 gauge needle

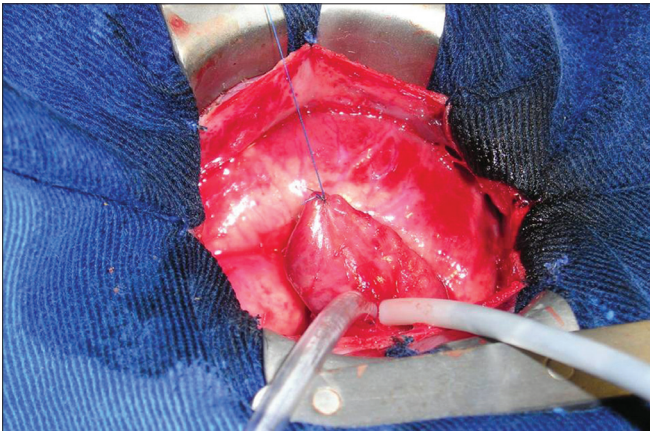


Figure 2b: Surgical view of the heart after a median sternotomy (surgeon's perspective). A 7 Fr sheath was placed in the left atrium through the base of the right atrial appendage over the guide wire. The right atrial and right ventricular dilatation is conspicuous



Figure 3: Still fluoroscopic frame performed with a portable C arm equipment showing the left atrial disk being deployed in the left atrium after the transesophageal probe had been pulled up, to avoid compression of the left atrium

and oxygen. She remained asymptomatic from the cardiovascular standpoint, despite having feeding and respiratory issues due to severe gastroesophageal reflux that required further hospitalizations and surgery. After a year, the TTE revealed spontaneous occlusion of the muscular VSD, good device position, complete closure of the ASD and PDA, normal cardiac dimensions, and normal estimated pulmonary artery pressures, with no cardiovascular medications.

DISCUSSION

Although the association between chronic lung disease and PDA has been well-documented in infants,^[1,9] other congenital heart defects with left-to-right shunts, such as an ASD and VSD, may also have similar effects on the pulmonary vasculature, further complicating the clinical course of such patients. Indeed, it has been demonstrated that atrial septal defects may deteriorate the respiratory status and prolong oxygen and / or ventilatory requirements in these infants.^[2-4] Surgical and catheter closure of such defects result in rapid respiratory improvement, with early extubation and oxygen weaning.^[2,3] Unfortunately, this patient was referred at a later age to our center and the positive effects of early PDA ligation on the underlying respiratory status could not be assessed. It is likely that early PDA ligation in the neonatal period could have averted the chronic lung disease and the need for the procedure described herein. On the other hand, at the time the patient was transferred to our unit, the three sources of increased pulmonary blood flow (ASD, VSD, and PDA) certainly contributed to the severe nature of the respiratory status, with pulmonary hypertension and oxygen dependency. Although a higher incidence of congenital heart defects has been reported in patients with chromosomal abnormalities,^[10] including those with defects in chromosome 8,^[11] whether the underlying clinical picture was intrinsically associated with the excessive material in chromosome 8 in our patient is speculative.

Although the negative impact of multiple congenital heart defects on the respiratory function was clear in the case presented herein, the decision with regard to what defect should be addressed and how it should be tackled was less straightforward. Several aspects were taken into account in the decision-making process, including the natural history of the underlying defects, the presence of severe pulmonary hypertension, and the morbidity of the available surgical and interventional strategies. Because the mid-muscular VSD was small with a high likelihood of spontaneous closure, it was left untouched. This proved to be the right decision, since the defect did close spontaneously after a year of follow-up. On the other hand, there was a consensus among our team that cardiopulmonary bypass had to be avoided due

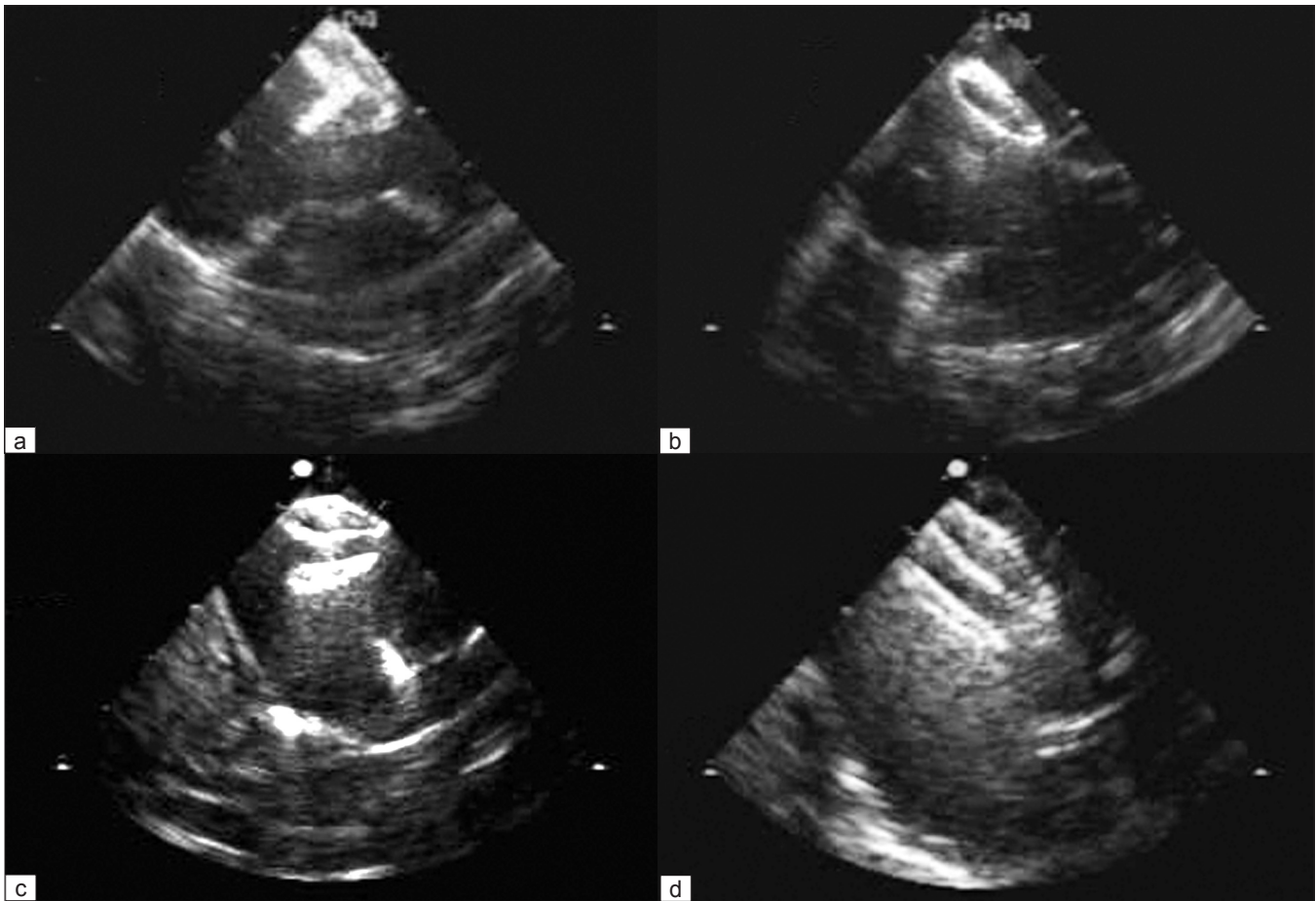


Figure 4: Transesophageal echocardiography pictures during implantation. a) The left atrial disk is seen deployed in the left atrium coming in a parallel fashion toward the septum. b) The left atrial disk is seen very close to the septum and the waist is within the defect. c) The right atrial disk is seen deployed in the right atrium still attached to the delivery cable. d) The device is seen in a perfect position after it had been released from the delivery cable. There is no prolapse of the device through the anterior rim of the defect



Figure 5a: Transesophageal echocardiography with color flow mapping showing complete closure of the defect



Figure 5b: Still frame of the fluoroscopy using the portable C arm equipment showing the device after release

to its attendant morbidity, especially considering the possible occurrence of a pulmonary hypertensive crises in the postoperative period. The option between a pure interventional approach and a hybrid approach was a difficult one.

Although percutaneous closure of the PDA has been performed in infants and even in preterms,^[12] the rate of complications in this age group, such as, arterial occlusion and protrusion of the device causing either left pulmonary artery stenosis or coarctation are higher than

at older ages. Also, the procedure is not as easy from the technical standpoint and devices that are not promptly available in our hands may be required for optimal positioning and conformation to ductal anatomy.^[13] As ASDs seldom cause significant symptomatology in small children, the experience with percutaneous closure of these defects is limited in this age group, especially in small infants.^[14] Patients who need ASD closure at a younger age^[15] and undergo this kind of procedure usually have associated comorbid conditions including genetic syndromes and noncardiac anomalies such as prematurity, chronic lung disease, diaphragmatic hernia, and tracheoesophageal fistula.^[14,15] Our patient fit well in this scenario. Although percutaneous closure of an ASD has been reported recently in a preterm infant^[2] (after the procedure described here had been performed), the issue of what would be the best access for such a small patient was a crucial one. Defects with no anterosuperior rim, such as the one seen in our patient, may offer challenges with regard to positioning the device in place, which may require more manipulation of sheaths from the femoral vein and / or application of less standard techniques.^[16] It was likely that the limited size of this child would make these maneuvers more difficult, less effective, and even risky. However, the transhepatic approach has been employed to perform a variety of different procedures, including ASD closure^[17] and some others, even in neonates,^[17,18] this access would have had the same limitations as described earlier, to deal with the PDA. Based on the recent reports of successful initial palliation of neonates with hypoplastic heart syndrome^[5] and more importantly, periventricular closure of muscular VSDs,^[6-7] the hybrid approach has emerged as an attractive alternative to deal with both ASD and PDA at the same time in the case presented here.

Although peratrial ASD closure has been performed earlier in a child with an associated muscular VSD,^[19] it is worthwhile discussing some technical aspects of the procedure. We feel that the key issue for successful implantation was the site for puncturing the right atrial wall at the base of the right atrial appendage. Using previously described techniques for periventricular closure of VSDs,^[6-8] this location was chosen so as to provide the straightest course to the left atrium, crossing the interatrial septum in a perpendicular fashion. In this manner, the left atrial disk could approach the interatrial septum at a parallel angle, avoiding prolapse through the deficient anterosuperior rim. We decided not to balloon size the defect for some reasons. First, the profile and the lengths of the available sizing balloons would have made the sizing procedure, through a peratrial approach, a cumbersome or even an impossible one, in such a small infant. Second, ASD closure has been performed without stretching the defect by using devices that are 10 – 30% larger than it.^[16] Therefore, we arbitrarily chose

a device that was about 10 – 15% larger than the defect, taking into account that the left disk would fit in the left atrium. This was achieved because the left disk of 9 mm Amplatzer ASO device had a diameter of 21 mm, while the length of the interatrial septum was 25 mm. Also, the optimal angle of attack toward the interatrial septum helped to place an ‘undersized’ device correctly. With regard to left disk manipulation, we felt that the standard TEE probe, due to its size and the close vicinity to the esophagus and the posterior left atrial wall, exerted some compression effect on the left atrium and could have impaired left disk deployment. This may not be an issue in small infants when the intracardiac probe (AcuNav) is used for TEEs.^[18] Also, because there was not enough room in the right atrium, the technique of deployment of the right atrial disk had to be slightly modified. By slowly retracting the sheath and pushing on the delivery cable, the right disk was reconfigured close to the septum and away from the entry site within the right atrial wall. Finally, TEE monitoring with optimal imaging of all the steps of the procedure was crucial for successful wire and sheath manipulations and adequate implantation.

In conclusion, the case presented herein illustrates another useful application of the so-called ‘hybrid approach’ to successfully manage a high-risk infant with chronic lung disease, severe pulmonary hypertension, and multiple congenital heart defects, with a left-to-right shunt, without the need for a cardiopulmonary bypass.

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