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Original article

Hybrid stage I palliation for hypo-plastic left heart condition without a hybrid suite: Suggestions for developing nations

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ABSTRACT

Cardiac hybrid procedures are performed in modern, spacious, and highly equipped hybrid suites in developed countries. Organizing such expensive suites in countries with an emerging economy is difficult from both a financial and logistics point of view. We share our experience of safely performing a Hybrid stage I palliation procedure for Aortic atresia with ventricular septal defect on a 2-month-old infant weighing 3.35 kg using minimal resources in a conventional catheterization laboratory.

1. Introduction

In developed countries, hybrid procedures for Hypoplastic left heart conditions were initially performed in well equipped operating rooms or catheterization laboratories (CL) and then shifted to Hybrid Suites.[1,2] There are very few reports of successful hybrid stage 1 palliation from developing nations because organizing such suites is expensive and difficult.[3]

2. Case details

A 2 months old full term born male infant weighing 3.35 kg who presented with cyanosis (saturating 80%) was diagnosed to have an uncommon variant and a milder form of Hypoplastic left heart complex[4–6] with aortic atresia and large ventricular septal defect (VSD). The aortic valve was atretic with severe hypoplasia of ascending aorta (2 mm) and arch, till the level of left subclavian artery and duct-dependent

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systemic circulation. There was a large inlet ventricular septal defect with anterior muscular extension. The mitral valve had parachute abnormality without obstruction or regurgitation (12 mm annulus; normal Z score) and the left ventricle was good-sized. There was a stretched patent foramen ovale (5 mm) shunting bidirectionally and there was severe pulmonary arterial hypertension (Fig. 1).

He underwent hybrid stage 1 palliation that included bilateral branch PA banding followed by intraoperative ductal stenting and transfemoral stenting of retrograde aortic arch orifice (RAAO) obstruction.

3. Technique

The procedure was done in the conventional CL and was performed according to previously described standard techniques for hybrid stage I palliation.[7,8] The stents used were 8 × 18 mm balloon expandable Genesis bare metal stent (Cordis J&J interventional systems Co, Miami, USA) for ductal stenting, and 4 × 12 mm coronary bare metal stent (Vision, Abbott, Santaclara, USA) for the stenosed retrograde aortic orifice.[8,9]

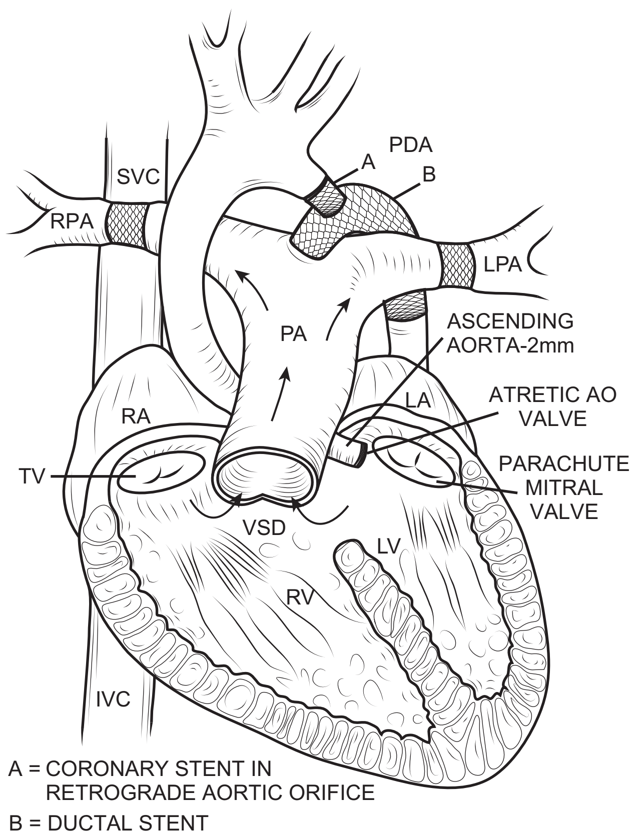

Fig. 1 – Diagram showing the uncommon and milder form of hypoplastic left heart condition with aortic atresia, ventricular septal defect, normal sized mitral valve and left ventricle. The branch pulmonary artery bands, ductal stent and the stent in the retrograde aortic orifice are shown in position. SVC – superior vena cava, IVC – inferior vena cava, RA – right atrium, LA – left atrium, PDA – patent ductus arteriosus, RPA – right pulmonary artery, LPA – left pulmonary artery, TV – tricuspid valve, MV – mitral valve, VSD – ventricular septal defect, LV – left ventricle, RV – right ventricle.

4. Discussion

In the spectrum of Hypoplastic left heart complex that may include hypoplasia of the mitral valve, Left ventricle, aortic valve and aorta, the most severe form may have severe hypoplasia of these structures which renders the left heart incapable of supporting the systemic circulation, and requires staged palliation. However there are less severe forms, in which aortic valve or aortic hypoplasia exists in the presence of adequate LV and mitral valve dimensions[4–6] as seen in our case and the left heart may be suitable for inclusion into the systemic circulation. Primary biventricular repair (BVR) in these patients require that the mitral valve and LV end diastolic dimensions be near normal.[5,6] We chose the approach of initial palliation followed by subsequent consideration of a BVR for our patient because of parachute MV. For the first stage palliation, we decided to perform Hybrid procedure instead of the Norwood operation because of risk factors like severe PAH, age beyond neonatal period and severe Tricuspid regurgitation.

Hybrid procedure is considered a safe treatment option for first stage palliation of HLHS in the following situations[7–11] (a) Infants with aortic atresia, ventricular septal defect and severe pulmonary arterial hypertension who present late and are unsuitable for Norwood operation or primary biventricular repair as seen in our case. (b) Premature/low birth weight infants. (c) Infants with severe co-morbidities. (d) As a bridge to biventricular repair in neonates with aortic atresia, VSD and two good sized ventricles. (e) As a bridge to cardiac transplant in complex cases where three stage approach is not feasible. (f) As an alternative to Norwood operation in institutions where Norwood programme is not well established. Hybrid approach would cost less as Norwood operation has a longer convalescence. In the developing world, there would be more time to arrange for financial support for a major second stage surgery.

Hybrid suites are usually very spacious and have advanced imaging facilities like biplane fluoroscopy, 3D/4D imaging and rapid post processing capabilities like image fusion. The high end facilities available in these suites prevent collision of imaging equipments with anesthesia equipment and allows access to patients from all sides. Considering the establishment and maintenance cost of hybrid suites, we present the following suggestions for institutions in developing nations who might be considering hybrid stage 1 palliation in the conventional CL.

1. Preoperative echocardiographic assessment plays an important role in planning the hybrid procedure. The essential points to be noted include thickness of interatrial septum, size of ASD, other sources of intracardiac mixing, diameter and flow acceleration in the transverse arch, PDA dimensions, presence of Isthmus shelf/gradient, tricuspid regurgitation and right ventricular function.
2. Building a Hybrid Team (Table 1) is essential for a successful Hybrid programme. The team should be a multi-disciplinary cardiac team with a limited number of members to avoid over crowding in the cathlab.

3. Besides arranging the necessary surgical and interventional equipment, educating the team members and assigning responsibilities is necessary for good team coordination during the procedure.
4. The primary reason for performing Hybrid procedures in the CL is to allow fluoroscopy. The images obtained from fixed single or biplane fluoroscopy in the CL are of higher quality than those obtained with mobile fluoroscopy used in the OR due to the higher heat storage capacity of fixed fluoroscopy.[12,13] This allows the performance of the procedure comfortably in the conventional CL and saves the cost of establishing mobile fluoroscopy facility in the operating room. The quality of angiographic image obtained with our conventional fixed single plane fluoro system with a single image display unit was good to obtain accurate measurements necessary for deciding the stent size and position.
5. Enough Space to accommodate at least 6 team members, Anesthesia equipment, CPB machine as backup, two tables for surgical equipments and hardware is necessary to safely perform the procedure in the CL (Fig. 3). Although hybrid stage 1 palliation does not require cardiopulmonary bypass, it should be available on standby. Performing the Branch PA banding in the operating room and then shifting to CL for ductal stenting is an alternative but cumbersome.
6. Special attention is paid to coordination of movement of various equipment and personnel to prevent collision and catastrophe. Proper positioning of the patient, chest retractors and the chest electrocardiographic leads should be done before the procedure to enable uninterrupted movement of the fluoroscopy and clear visualization of angiographic images. Positioning and adjustments during the procedure can lead to dislodgment of the sheath in the MPA and catastrophic bleeding. Radiolucent electrocardiographic leads and chest retractors would be very useful for angiographic analysis during hybrid congenital cardiac procedures[14] In our case, fluoroscopy in the lateral view helped us to avoid removing the retractors (Fig. 2A) and prevented any disturbance of the sheath in the MPA. The cathlab technician should inform the anesthesiologist every time he rotates the C arm from PA to lateral in order to avoid dislodgement of ventilator tubings. The Surgeon and Cardiologist will interchange their position depending up on the steps of the procedure. The surgeon maintains the MPA sheath in position while the Cardiologist performs the ductal stenting.
7. Both self expandable and balloon expandable stents can be used for ductal stenting since the type of stent has no relation to the development of RAAO stenosis.[9] Closed cell stents have higher radial strength, while it is easier to enlarge the cells of an open stent to relieve RAAO obstruction. Although we used a less expensive closed cell stent, we were able to cross and successfully stent the retrograde aortic orifice.
8. Congenital RAAO obstruction should be suspected if there are incipient features in the echocardiography such as juxta ductal shelf, mild isthumal narrowing and mild flow acceleration at the isthumus. Based on echocardiography, we anticipated the need for stenting the retrograde aortic orifice after the ductal stenting. Hence we cannulated the right femoral artery (22G cannula) and prepared the necessary hardware for aortic stenting in anticipation. RAAO stenosis was noticed in this infant following ductal stenting. We decided to stent the stenosed RAAO through transfemoral route after chest closure in order to avoid maintaining the sheath in the MPA for a longer period in a beating heart. Prior femoral artery cannulation helped us to avoid femoral puncture in a heparinised child and also saved time. Reverse BT Shunt was not considered due to the potential complications of compromised coronary flow and ventricular dysfunction reported in the literature.[1,10]
9. The limitations of performing the procedure in the CL relates mainly to maintaining sterility. To allow sternotomy in the CL, it is imperative to follow the infection control guidelines and maintain the environment in the CL like in OR.[15]
10. Postoperatively these infants are not as sick as the Norwood candidates and convalescence is more rapid, thus reducing the cost. Postoperatively this infant's oxygen saturation was 70% and haemodynamically stable.

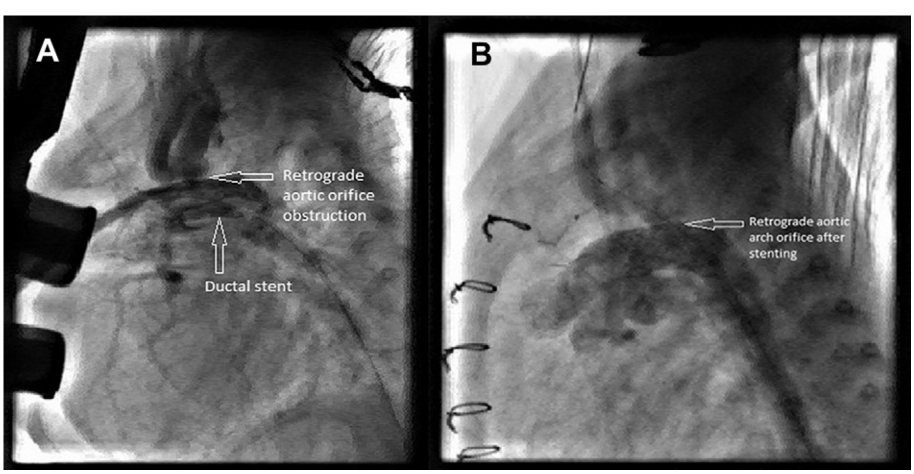

Fig. 2 – (A) – Angiography in the main pulmonary artery after ductal stenting showing stenosis of the RAAO and (B) – angiography in the descending aorta showing improved flow through RAAO after deployment of the coronary stent across it.

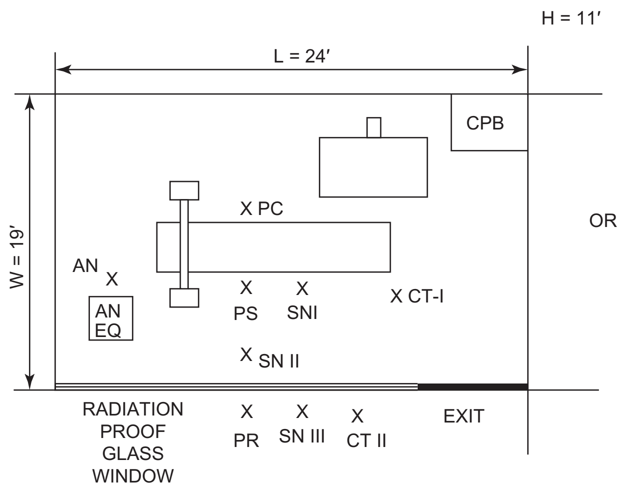

Fig. 3 – Schematic diagram showing the recommended location and dimensions of conventional cathlab and arrangement of equipments which would allow a successful hybrid procedure. H – height (floor to roof distance), L – length, W – width, PC –pediatric cardiologist, PS – pediatric surgeon, AN – anesthesiologist, SN-staff nurse, CT –cathlab technologist, PR – perfusionist, AN EQ – anesthesia equipment, CPB – cardiopulmonary bypass equipment.

He was discharged on the eighth postoperative day with antiplatelet medication (ASA) and followed up periodically. Balloon atrial septostomy is usually done before discharge in typical HLHS patients to allow time for increase in Left atrial size. It was not needed in this infant due to good intracardiac mixing at the level of the VSD and no significant stenosis at the parachute mitral valve.

11. As the small stent in the RAAO is prone to instent stenosis, it is necessary to closely monitor these infants during the inter stage. As the risk of interval mortality is similar, the followup protocol should also be the same as that of Traditional Norwood operation. Weekly review with assessment of neurological status, ventricular function and stent flow by echocardiography is essential. The same protocol was followed for this infant and he was well at 6 months followup with normal milestones, good ventricular function and unrestricted flow in the ascending aorta. Elective comprehensive second stage surgery is being planned.

Hybrid stage I palliation is a reasonable and cost effective option for developing nations. Pre procedure planning and building a “Hybrid team” should make the procedure easier and safer in the CL, without the need for hybrid suites.

Appendix A

Table 1 – Hybrid team: essential team members with defined responsibilities to perform hybrid stage 1 palliation in conventional cathlab (total – 6 members).

Personnel	No.	Responsibilities
Pediatric cardiac surgeon	1	Performing Bilateral branch PA banding. Providing the access in the MPA during ductal stenting. Sterility measures during the procedure.
Pediatric interventional cardiologist	1	Performing the ductal stenting and the stenting of the retrograde aortic orifice if necessary. Educating the entire team about the procedure including sterility measures. Assigning responsibilities to the team members.
Pediatric cardiac anesthesiologist	1	Ventilation, anesthesia and invasive vitals monitoring/care. Adequate preparation for handling bleeding complications. (Should make sure that blood products are in reserve.)
Catheterization lab technologist	1	Handling the fluoroscopy. (Single plane or biplane, floor or ceiling mounted fluoroscopy.) To make sure the correct placement of chest leads and monitor wires/cables before starting the procedure to allow uninterrupted movement of fluoroscopy. Providing adequate light source for the procedure. (Ceiling or wall mounted lights in the cathlab and the head light used by the surgeon.) Arranging the necessary hardware for stenting procedure in consultation with pediatric cardiologist. Helping the cathlab staff nurse to prepare the appropriate stents and other hardware for catheterization. Obtaining angiographic measurements during the procedure.
Operating room staff nurse	1	Responsible for fumigation of cathlab and other sterility measures. Strict precautions as followed in the operating room should be followed. To arrange surgical equipments necessary for Bilateral PA banding including polytetrafluoro ethylene graft. To assist the pediatric cardiac surgeon during the procedure and maintaining count of surgical equipments.
Cathlab staff nurse	1	To arrange catheterization hardware for stenting of the duct and the retrograde aortic orifice including the appropriate stents. To go through a check list for the entire procedure. Coordinate the activities of all team members during the procedure. To assist the pediatric interventional cardiologist during the procedure.
Backup personnel who would stay outside the procedure room	3	Perfusionist – 1, staff nurse – 1, cathlab technologist – 1.

MPA – main pulmonary Artery, CPB – cardio pulmonary bypass.

Conflicts of interest

All authors have none to declare.

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