CARDIAC INTERVENTIONS IN PEDIATRIC CARDIOLOGY: THE FUTURE

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Pediatric Cardiac interventions have come a long way from the initial intervention in the 1950's. Balloon angioplasty has been accepted as the procedure of choice in several congenital anomalies. Apart from balloon angioplasties/ valvuloplasties, Atrial Septal Defect, Ventricular Septal Defect (muscular) device closure have been FDA approved with adequate world wide clinical experience and long-term follow-up. In addition, newer procedures are under clinical trial for perimembranous VSD device closure in the catheterization lab; per operative closed heart procedure in the operation theatre or as a hybrid procedure. Palliative procedures like flow restriction to lungs with devices to equate with surgical pulmonary artery banding; stenting of the patent ductus arteriosus in duct dependent cyanotic heart disease in the newborn or a combination of these form transcatheter Norwood stage I in the cath lab. Experience and technology will also help make transcatheter Fontan operation possible and that does not seem too far. The emphasis in pediatric cardiac interventions shall always remain that the decision, procedure, and management of their complications is a joint effort of the surgeon and the interventionalist.

Key words: Pediatric, Cardiology, Stent, Interventions, PDA, ASD, VSD.

IT WAS the pioneering work of Rubio-Alverez in 1950's that initiated pediatric cardiac interventions. Interventional Pediatric Cardiology since then has come a long way and several lesions are now primarily treated by interventional catheterization. These include pulmonary valve stenosis, aortic stenosis, patent ductus arteriosus, aortic recoarctation. In other situations, interventions are used preferably over surgery including native coarctation of aorta, stenting for branch pulmonary arteries, pulmonary valve balloon dilatation in tetralogy of Fallot's, re-dilatation of the stenosed conduit placed in the right ventricular outflow tract, stenosed Blalock-Taussig shunt and coil embolization of collateral vessels. In the last decade, the spectrum of interventions have increased to an extent that surgical procedures are being replaced by interventional procedures, making pediatric cardiology one of the more advanced fields in medicine and further increasing the challenges of pediatric cardiac surgery.

We start with an established intervention i.e., ASD device closure and discuss newer interventions which may be incorporated into clinical practice in the next few years. *(Table* 1*).*

ATRIAL SEPTAL DEFECT CLOSURE

Ever since the pioneering transcatheter device closure of a secundum atrial septal defect by King [1] in 1974 and the subsequent work of Rashkind [2], cardiologists have been trying to make interventional closure the preferred mode of treatment. The first device used by King required a 23F sheath. Subsequently the Clamshell device (11F sheath), Sideris button device (8F), Cardioseal (11F), Angelwings (11F) and the ASD Occluder System device (10F) also require large delivery sheaths. The major development in this field has been the ability to deliver the repositionable, retractable Amplatzer

Table 1. ***Pediatric cardiac interventions currently in the process of being incorporated into clinical practice.***

S.No.	Interventions
1.	Perimembranous VSD Device Closure (Phase 1 Clinical trial) a. Transcatheter b. Peroperative (beating heart surgery) c. Hybrid Surgery
2.	Flow Restrictor Implantation
3.	PDA stenting in Ductal dependent cyanotic CHD's
4.	Vascular Plug implantation
5.	Norwood Stage I in Cath Lab (PDA stenting + Flow Restrictor Implantation)
6.	Muscular VSD Device Closure (Clinical Trial complete)
7.	ASD Device (FDA approved)
8.	PDA Device (FDA Approved)

VSD = ventricular septal defect; CHD = congenital heart disease; ASD = atrial septal defect; PDA = patent ducuts arteriosus.

device through a sheath as small as the 6F or 7F. This technological advancement took place due to the use of Nitinol material for cardiac devices. After initial trials, the Amplatzer device was approved for clinical use in the United States by the FDA in 2002. The Amplatzer device for ASD closure is the one most frequently used in clinical practice. The device has 3 parts connected to each other (*Fig.* 1)*:* the left atrial disc is the largest (about 14mm larger than the size of the ASD); the right atrial disc is slightly smaller though still larger than the ASD. The 2 discs are connected by the "waist" or the stent which is the size of the ASD itself. Of the devices available presently, the early closure rates at 3 months for this device (>98%) are the highest. Incidence of severe complication for Amplatzer device has been amongst the least (0.3 %). Modes of retrieval of embolized device remain difficult though not impossible.

Limitations of the Procedure

The procedure can be carried out only in select patients of secumdum ASD with good rim margins. It is carried out under general anesthesia mainly for purposes of the prolonged trans-esophageal echo (TEE) which needs to be performed during the procedure.

The limitation of device closure is that none of the present devices currently are able to close ASD's with small rims. All patients with large ASD's, ASD's with small rim and sinus venosus and primum ASD's have to undergo surgical closure. There has been a recent trend towards use of intracardiac echocardiography (ICE) for better anatomy delineation during the procedure[4]. This is done via a catheter; it requires at least a 9F sheath and this maybe the limiting factor for its use in the pediatric age group.

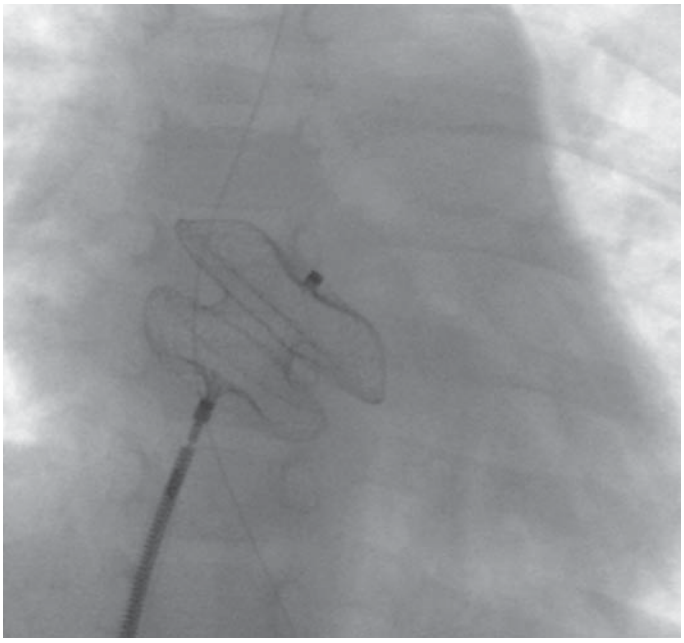

Fig. 1. The ASD device is seen held with the larger LA disc, the connecting "stent" and the RA disc. LA = left atrium; RA = right atrium

Outcomes–immediate, intermediate and late

The immediate safety of the procedure has been proven with several thousand device closures which have been performed. Closure rates were also shown to be very high with majority studies showing complete closure of the ASD by 3 months. As more and more studies have shown better outcomes, the focus is shifting to late and long-term effects of non-surgical closure.

Recent reports of neurodevelopment assessment of children who have had surgical closure of ASD compared to device closure have shown interesting results. The patients with surgical closure of ASD showed significantly lower visual-spatial/visual-motor skills. This may be related to the cardiopulmonary bypass [5].

VENTRICULAR SEPTAL DEFECT CLOSURE: MUSCULAR

Muscular ventricular septal defects can be very challenging to close surgically and several variations of the surgical technique have been discussed in literature.

These include left ventriculotomy which may be associated with an aneurysm or electrical problems in the future [6].With this in mind the interventional closure of muscular VSD has taken precedence over surgical closure and now several techniques for closure of muscular VSD by device have been developed and reported. These include per-operative closure of the VSD; the Hybrid technique for closure of the VSD and beating heart closure of the VSD. These are in addition to the now established and accepted transcatheter non-surgical closure of the VSD. The device is in the process of and nearing FDA approval in 2004.

The initial results of muscular VSD closure by Amplatzer device which is clinically the most often used device for all septal defects are encouraging. The devices have been implanted in congenital, post-operative residual VSD, in post-myocardial infarction VSD and in post-traumatic VSD.

In the largest reported single study for muscular VSD's (MVSD) thirty-two patients underwent attempted transcatheter closure of an MVSD. Of these 19 had congenital unoperated MVSD, twelve had post-MI MVSD and 1 patient had an acquired VSD post-surgical repair of hypertrophic cardiomyopathy. The median age of patients was 11.5 years. The device was implanted successfully in 30 patients. The authors consluded that the Amplatzer MVSD occluder is a safe and effective device for closure of MVSDs up to 14 mm in diameter and further clinical trials with this device are needed [7].

VSD DEVICE CLOSURE-PERIMEMBRANOUS VSD

Perimembranous VSD's are very close to and just below the aortic valve. This makes the procedure of device closure more challenging. The device has been designed in such a way that its LV disc is eccentric without any rim towards the Aortic valve. Initial results have been encouraging with the eccentric Perimembranous VSD closure device. The long-term results would decide the final outcome of whether this becomes a standard procedure or not.

Percutaneous closure of perimembranous ventricular septal defects (VSDs) has been feasible, safe, and effective with the new eccentric Amplatzer membranous septal occluder in a small number of patients [8,9]. Ten patients with volume-overloaded left ventricles underwent closure under general anesthesia and transesophageal guidance (TEE). Implantation was successful in all patients. Nine patients had complete occlusion within 1-3 months. Device-related aortic or tricuspid insufficiency, arrhythmias, and embolization were not observed. Two patients had slight gradients across the left ventricular outflow tract, normalizing after 3 months.

A new technique has been under trial for closure of the defects under echocardiographic guidance without cardiopulmonary bypass to prevent the deleterious effects of bypass. In the study carried out on pigs with naturally occurring perimembranous ventricular septal defects underwent closure of the defect in the operating room by using the perventricular technique. The device was deployed under echocardiographic guidance with the heart beating. The perventricular technique appears to be excellent for closure of perimembranous ventricular septal defects in the operating room. The technique might be feasible in smaller babies, who are high-risk candidates for closure in the catheterization laboratory. Cardiopulmonary bypass and prolonged hospital stay are avoided [10]. This has subsequently been tried in humans with good results though trial on large number of patients is awaited.

FLOW RESTRICTOR IMPLANTATION AS AN ALTERNATIVE TO PULMONARY ARTERY BANDING

Pulmonary artery banding is used as a palliation prior to muscular VSD closure or as part of other procedures like Damus-Kaye-Stansel or Norwood Stage I. It may also be used for reducing the pulmonary pressures in patients with high flow Single Ventricle physiology. The inability to predict the final PA pressure while banding the artery has resulted in several alternative techniques being looked at including externally adjustable bands. Flow restrictor implantation is also an attempt at replacing a surgical palliative procedure with a non-surgical interventional procedure. The procedure, as per personal communications, has been tried on about 14

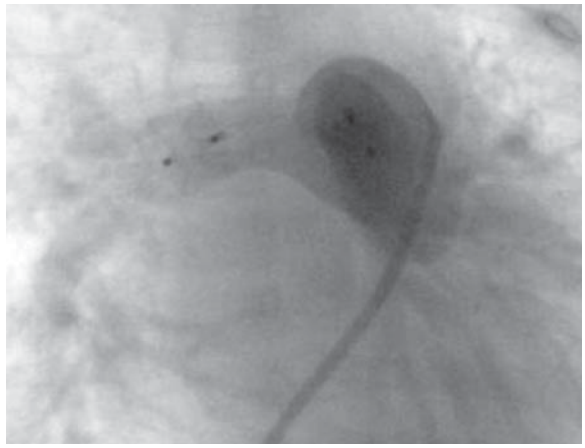

Fig. 2. The angiogram still frame shows bilateral flow restrictor implantation in each branch pulmonary artery.

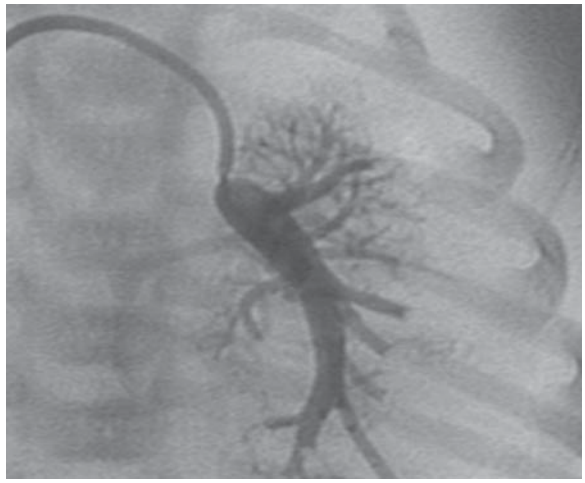

Fig. 3a. The catheter course via right aortic arch to left subclavian artery to (closed) ductus arteriosus is noted. The LPA arises from the PDA as shown here.

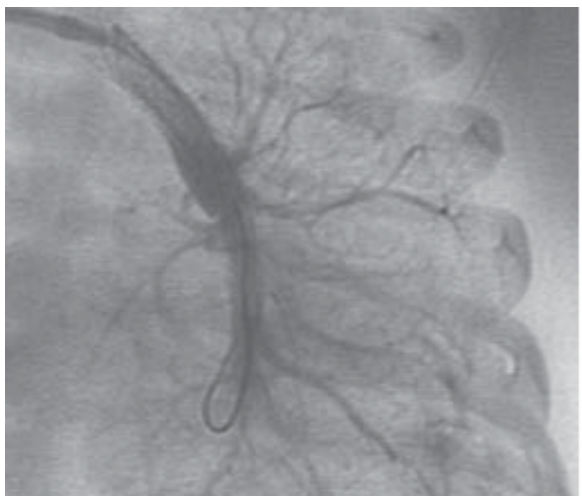

Fig. 3b. The PDA has been stented and the continuity of left subclavian artery, PDA and LPA is noted. LPA = left pulmonary artery; PDA = patent ducuts arteriosus;

Key Messages

- Pediatric Cardiac Interventions have advanced significantly in the last decade that, apart from balloon angioplasties, certain other procedures have already been FDA approved and incorporated in clinical practice (ASD Device closure, PDA Device Closure).
- Some other procedures are in the process of completion of clinical rials e.g. Muscular VSD device closure. Innovative procedures like per-operative (beating heart) device closure, Hybrid technique.
- Several patients have safely undergone Perimembranous VSD closure in the catheterization lab with the Eccentric VSD Device: long-term results will help in deciding the true safety of this procedure.
- Innovative procedures like PDA stenting in Duct dependent cyanotic congenital heart disease and Flow Restrictor implantation in high flow situations are in the stage of clinical trials.
- Norwood Stage I (Flow Restrictor Implantation with PDA stenting) and Fontan Completion (with prior surgical preparation) in catheterization lab are other interventions one can look forward to in the coming several years.

patients as part of Norwood Stage I (associated with PDA stenting) and seven of these patients have gone on to have the surgical stage II. The author has performed this procedure in 2 children (*Fig.* 2). The procedure involves inserting an ASD like device within each branch pulmonary artery. The device has 2 holes in the center which allow some flow and there by restrict majority of the flow through it. The procedure is referred to as flow restriction and the device as Flow Restrictors. The diameter (outermost) would be similar to that of the branch pulmonary artery diameter and the axis of the device has to be at 90 degrees to the long axis of the branch pulmonary artery. This is able to generate enough gradient within the branches to prepare them for single ventricle palliation [11]. *Figure* 2 shows the angiographic picture of one of the patients who has undergone this procedure by the author and at 6 months follow-up has 70 mmHg gradient across the flow restrictor.

STENTING OF THE NEWBORN PDA IN DUCTAL DEPENDENT CONDITIONS

Stenting of the newborn duct in ductal dependent conditions has long been the objective of interventional cardiologists. It has only been recently that this has become a reality in the newborn period mainly due to the advancements in the hardware available for arterial access in a newborn baby and for delivering a premounted stent. Recent results have shown this to be a safe intervention with good results [12]. Personal experience confirms the same for selected patients with favourable duct anatomy. *Figures 3a & 3 b* show the duct being stented in a patient with tetralogy of Fallot's with right aortic arch, left pulmonary artery from PDA which arose from the left subclavian artery. With the duct closing, the flow to left lung via left pulmonary artery was completely stopped. PDA stenting restored this.

In one of the larger series, balloon-expandable stents were implanted in 21 patients with ductal dependent cyanotic congenital heart disease. The PDA was dilated to a final diameter of 4-5 mm without procedural deaths. Overall survival rate after 6 years was 86%. Corrective surgery was possible in six patients.

In another large study, 69 patients with duct-dependent pulmonary circulation underwent cardiac catheterization with the intent of PDA stenting as first palliative procedure. Patent ductus arteriosus stenting was successful in 51 patients (91.1%) and failed in 5 patients (8.9%). There was no procedure-related mortality. Patent ductus arteriosus stenting proved to be an attractive alternative to surgical shunt in a majority of patients with duct-dependent circulation. An absolute contraindication to this technique is the presence of branch pulmonary stenosis [13].

PDA DEVICE CLOSURE

This device intervention technique is being discussed towards the end because it is the most evolved intervention amongst device closure. The procedure has replaced surgical ligation of PDA except in the smallest of babies (premature or newborn babies). After the limitations of the Rashkind Umbrella were realized that in the late 90's the Amplatzer duct occlude became available. The major advantages are that it can be used to close PDA of all sizes via a small size sheath. The closure rates are close to 98% instantaneously and 100% at 3 months. The complications if any reported are minor related to access for arterial catheterization.

CONCLUSION

Pediatric cardiac interventions are a rapidly advancing field with several new procedures replacing some of the simpler surgeries (PDA ligation) or replacing some of the palliative surgeries (Stenting of duct for BT shunt; Flow restrictor implantation for PA banding; Norwood stage I in catheterization lab). One can look forward to more interventions getting established in the coming years including transcatheter Fontan completion.

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