PROGRESS IN PEDIATRIC CARDIAC CATHETERIZATION WITH SPECIAL REFERENCE TO INTERVENTIONAL PROCEDURES

MOHAMED A. ALI KHAN, MD, FRCP(E), FACC

IN 1844 CLAUDE BERNARD performed the very first cardiac catheterization. 1 The subject was a horse and both right and left ventricles were entered via the jugular vein and carotid artery. In 1929 Werner Forssman2 reported an experiment on himself, inserting a catheter into a peripheral vein in the left arm and advancing it into the heart. Courand and coworkers3 in 1941 introduced this procedure for the evaluation of heart disease. Dexter, in 1947,4 utilized the technique in congenital heart disease cases. In the next 20 years, stimulus was given to improve catheterization results and to prevent complications. Rashkind's5 balloon atrial septomy in 1966 stands as one of the most important therapeutic procedures developed in the past 25 years for the treatment of certain types of congenital cardiac defects. This probably stimulated pediatric cardiologists to use the Swan Ganz balloon catheters to flow guide the catheter tip for difficult entry into certain cardiac chambers. Another result of this concept of balloon-tipped catheters was the development of Gruntzig's method to dilate renal arteries6 and coronary arteries.7,8

This method was later applied in congenital heart disease, pulmonary stenosis, coarctation of aorta, and aortic stenosis by Kan,9 Lock,10 and Lababidi.11 There are many other landmarks that could be mentioned and many individuals whose contributions should be recognized, but this is beyond the scope of this paper.

From the Division of Pediatric Cardiology, Armed Forces Cardiac Centre, Riyadh, Saudi Arabia.

Address reprint requests and correspondence to Dr. Ali Khan: Section of Pediatric Cardiology, Lorna Linda University Children's Hospital, 11234 Anderson Street, Lorna Linda, CA 92354, USA.

Objectives

The objectives of cardiac catheterization are to delineate the anatomy of the heart, assess hemodynamics, assess myocardial function, evaluate results of surgery, measure cardiac output, and conduct electrophysiological procedures. 12,13 With the improvement in noninvasive techniques, many of these functions can be achieved by M-mode, two-dimensional echocardiography and Doppler, and color-flow mapping.14 In the last decade, more emphasis was given to therapeutic interventional procedures to delay or avoid surgery for some cases of congenital cardiac defects.

Factors Contributing to Improve Morbidity of Cardiac Catheterization

The factors that have reduced morbidity and mortality in infants during cardiac catheterization are the treatment of metabolic disorders, pulmonary problems and congestive heart failure, and control of hypothermia. 15 Blood gases are routinely monitored during catheterization and appropriate measures are taken to treat acidemia and hypoxemia. Blood glucose levels are monitored in all neonates with marked hypoxemia and also in those with severe congestive heart failure. The use of prostaglandin E1 (PGE1)16-20 infusion in ductus-dependent lesions, such as pulmonary atresia, has revolutionized the morbidity and mortality of catheterization and surgery in the neonate. The availability of image intensification and biplane angiography is crucial for instant playback and avoidance of more than one injection at a site.21 The improvement in morbidity and mortality is also associated with a decrease in the total amount of non-ionic contrast
medium used and modifications of the types of catheters used. All the above measures taken have a profound effect in the outcome of cardiac catheterization and safety of sick infants and children.

**Special Procedures**

There are many special procedures performed in infants and children, some for diagnosis and others for therapy. For diagnosis, flow-directed balloon catheters, pulmonary vein wedge injections, pulmonary artery wedge angiograms, and pericardial taps are performed. From the therapeutic point of view, balloon atrial septostomy, blade septostomy, balloon dilatation of blood vessels and cardiac valves, balloon and coil embolization of vessels and shunts, foreign body retrieval from cardiovascular system, transvenous closure of patent ductus arteriosus, atrial septal defect, ventricular septal defect, and use of stents in stenotic vessels (Figure 1) are carried out.

![Figure 1. Transcatheter approach with various balloons and interventional catheters in cardiology, only the distal ends of catheters are shown: (A) blade septostomy catheter (open); (B) delivery system for umbrella closure of patent ductus arteriosus. Arrow indicates a 17-mm double-disc umbrella device; (C) cordis biotome for myocardial biopsy; (D) Rashkind balloon septostomy catheter; (E) Inoue balloon for mitral valvuloplasty; (F) balloon catheter for various valvuloplasties; and (G) bifoil balloon with pigtail for valvuloplasty.](image)

**Percutaneous Procedures**

The technique for percutaneous insertion of catheters was described by Seldinger in 1953. Lurie and associates modified the method in 1963 for use in infants and children, and later Desilet and Hoffman further improved the procedure. The procedure has distinct advantages - a vein or artery that has previously been used for a percutaneous or cut-down procedure can be used again for a percutaneous procedure. The rapidity of catheter placement and removal and the very low incidence of complications make it a suitable and accepted procedure in infants and children.

**Transseptal Procedures**

Transseptal left heart catheterization was first introduced by Cope in 1959 and modified by Brockenbrough in 1960. Pediatric application became popular after Mullins and Duff invented the introducer set in 1978. The new introducer set is a long sheath and dilator manufactured precisely to fit the Brockenbrough transseptal needle. The dilator component is 1 cm shorter than the needle. The sheath is thin-walled Teflon and is 4 cm shorter than the dilator portion of the set, available in pediatric 54-cm length and adult 68 cm in various sizes from 6F to 14F. The contraindications for transseptal puncture include left atrial thrombus and left atrial tumors such as myxoma or lack of access due to absent or thrombosed inferior vena cava. Certain conditions require special precautions in transseptal left heart catheterization; they include small or large left atrium, large dilated ascending aorta, large coronary sinus, cardiac malpositions, thoracic spine deformities, postseptum primum defect, and post-Mustard operation, and when the left femoral vein is used. We have performed over 350 transseptal procedures between October 1983 to October 1992. The results of our first 220 procedures confirm the success of this technique. The procedure was successful in over 99% of patients. Pericardial puncture occurred in three patients, though without tamponade or other sequelae. There was no morbidity or mortality.

**Balloon Atrial Septostomy**

Since its introduction in 1966, the balloon atrial septostomy (BAS) procedure has changed
the outlook for survival of children with transposition of the great arteries. The catheter is 4F or 5F (USCI, Rashkind, Miller Edwards) introduced percutaneously through a sheath from the femoral or umbilical vein and advanced to the right atrium, then through the foramen ovale to the left atrium. It is inflated with diluted contrast material between 3 to 6 mL and rapidly pulled to the right atrium, inferior vena caval junction and deflated. The procedure can also be performed under two-dimensional echocardiography. The effective palliation rate is approximately 90%, and the long-term survival rate is approximately 80%. The indications include conditions needing increased mixing at the atrial level or vent decompression of the right or left atrium. Examples include transposition of the great arteries, tricuspid, pulmonary or mitral atresia, total anomalous pulmonary venous drainage, and complex cyanotic congenital heart diseases. Reported complications are perforation of atria, tear of atria, damage to atrioventricular valves, thrombosis of inferior vena cava, and thromboembolism. Our experience at the Armed Forces Hospital since 1982 includes over 350 procedures with very low morbidity.

**Blade Atrial Septostomy**

An interatrial opening is important in transposition of great arteries, as well as for obligatory interatrial shunt lesions such as mitral atresia, tricuspid atresia, and total anomalous pulmonary venous drainage. It is also beneficial in patients with double-outlet right ventricle and a restrictive ventricular septal defect and those with an univentricular heart and an unilateral restrictive atrioventricular valve. Rashkind and Millers BAS has been useful in infants. After infancy, the atrial septum is thickened and a repeat BAS is often ineffective. In 1973 a Park catheter with a built-in surgical blade was developed to enlarge the interatrial opening in such cases. Clinical use of this technique has been successful. The catheter is a 5F or 6F, 65-cm long polyethylene tubing (William Cook Europe, Bjaeverskov, Denmark) with the tip consisting of a 3.5-cm section of stainless steel tubing, containing a small blade. The blade catheter is advanced through a long 6F or 7F sheath placed in the left atrium.

Several cuts are made in the atrial septum followed by Rashkind's balloon septostomy procedure. Adequate septostomy indicators are clinical improvement, equalization of atrial pressures, change in arterial oxygen saturation, angiogram of defect before and after septostomy, increase in systemic arterial pressure, decrease in pulmonary artery pressure, balloon calibration of defect, and echo visualization of defect. 33

Blade septostomy risks are blood loss, laceration of the heart, and embolization; all are avoidable. The collaborative study in 52 cases showed improvement in 79% of cases, and 4 years follow-up showed excellent results in 84% and reduced interatrial opening in 16%. We have performed over 80 blade septostomies and concur with the results of the collaborative study.

**Balloon Pulmonary Valvuloplasty**

Dotter and Judkins in 196435 introduced the concept of dilating an atherosclerotic obstruction using a series of progressively larger catheters for the treatment of peripheral vascular disease. In 1976 Gruntzig modified the technique by placing a polyvinyl chloride balloon on the tip of the catheter for angioplasty of renal6 and coronary vessels. 7,8 The success of percutaneous transluminal coronary angioplasty (PTCA) prompted the application of this principle to pulmonary valve stenosis by Kan,9 Pepine,36 and others by using larger size balloons.37-39 Once the clinical, electrocardiographic, and echocardiographic diagnoses are made of the moderate-to-severe pulmonary valve stenosis, the hemodynamic study and angiography are performed and the gradients measured.

For single-balloon valvuloplasty, the catheter is advanced over a guidewire and positioned across the stenotic pulmonary valve. The catheters utilized are 5F to 9F with a balloon 5 to 20 mm in diameter; the balloon selected is approximately 100% to 130% larger than the pulmonary valve annulus. For double-balloon technique, balloon catheters are introduced from both groins. The combined diameter of the two balloons is 120% to 150% of the diameter of the pulmonary valve annulus.

Balloon valvuloplasty in critical pulmonary valve stenosis has been limited in infants who are already compromised by hypoxemia or acidemia.
Potential difficulties include guide wire positioning, perforation of the right ventricular outflow tract, and inability to advance the necessary calibre balloon. In such cases a 0.014-in guidewire is introduced, and a coronary angioplasty balloon catheter as small as 2 mm in diameter is used for the initial dilatation and subsequently replaced by larger guidewires and suitable-sized balloons for definitive dilatation. 

Using these procedures, we have performed 257 procedures between August 1984 to October 1989 at the Riyadh Armed Forces Hospital in Saudi Arabia. The age range was from one day to 12 years with a mean age of 46.4 ± 44 months. Twenty-three patients were less than one month of age, a total of 84 patients under 12 months, and the remaining over 12 months of age. The weight range was between 2.4 to 46.8 kg with a mean of 13.7 ± 8.5 kg. Thirty-one patients were less than 5 kg; 80 patients weighed between 6 and 10 kg. Twelve technical failures and 14 complex lesions were excluded from the following analysis. In the remaining 232 procedures, the pre balloon valvuloplasty peak systolic gradients ranged from 30 to 310 mm Hg with a mean of 97 ± 49 mm Hg. After balloon valvuloplasty the gradients ranged from 0 to 175 mm Hg with a mean of 22 ± 17 mm Hg, indicating a significant 77% reduction. There was 75% to 100% reduction in the gradient in 125 of 232 procedures (54%), with 50% to 75% reduction in 80 patients (35%). A total of 89% had the gradient reduced by 50% or more. One hundred and three patients underwent recatheterization at an average period of 8 months after the initial procedure. The mean follow-up gradient was 32 ± 3 mm Hg. The remaining patients were followed by echo-Doppler study. Twenty-one patients were reballooned because of increase in gradients. The maximum follow-up was 8 years. Sixteen patients required surgery, including the technical failures and inadequate relief of gradients due to dysplastic valves. Two infants, aged 5 months and 9 months, died during the procedure.

So far, we have performed over 400 dilatations of pulmonary stenosis. Long-term follow-up in balloon pulmonary valvuloplasty has been reported. In dysplastic pulmonary valves the results have been poor inspite of reballoon dilatation, although in partial dysplasia the results are satisfactory. 

### Balloon Dilatation of Coarctation of Aorta

A similar technique has been utilized in cases of coarctation of aorta restenosis and, more recently, in native coarctation. Patients are heparinized for this procedure. After the coarctation angiogram, a wire is advanced through an end-hole catheter, passed retrogradely across the coarctation. The flexible end of the wire is placed in the ascending aorta. The catheter is then exchanged with an appropriate size balloon catheter and advanced over the wire to place the balloon across the coarctation and inflated once or twice until there is no waisting. The balloon chosen should be the size of the isthmus, just distal to the origin of the subclavian artery proximal to coarctation. After the procedure, the gradients are remeasured and the angiogram is repeated. Our experience is only with native coarctation. Between November 1984 and December 1989, 137 dilatations were performed in 116 patients between the ages of one day and 12 years, with a mean of 0.95 years. The weight range was 1.3 and 44 kg with a mean of 7.4 kg. The male to female ratio was 2:1. Sixty percent of the patients were under 6 months of age. A significant drop in the coarctation gradient was noted in all age groups, with a better response noted in neonates. Recatheterization was performed 3 to 12 months after the procedure in 62% of patients. An increase in the gradient was noted mostly in younger patients, especially in neonates of whom 80% showed recoarctation. The following complications were observed: blood loss requiring transfusion in 21 patients, decreased femoral pulses in 23, requiring heparin in 19, streptokinase in 5, and surgery in 3 patients. Serious arrhythmias were noted in 3 patients. Intimal irregularities at the dilatation site were seen in 15 patients and aneurysm formation in 9 patients. No mortality occurred in our cases. Recoarctation appears to be related to age, femoral injury is preventable, and reballoon dilatation can be performed for recoarctation. Aortic aneurysm is a major complication and needs further follow-up. Until now, there have been over 250 dilatations of
native coarctation with one month to nine-year follow-up.

Mitral Valve Balloon Dilatation

Balloon dilatation of rheumatic mitral stenosis in adults is quite common, using one or two balloons by transseptal techniques. 55-57 The commonest technique involves a transseptal entry into the left atrium, advancing single or double-balloon catheters over the wires across the stenotic mitral valve and inflating the balloons to split the commissures. Patients are heparinized as soon as the left atrium is entered. Balloon selection depends on the size of the child and the mitral valve diameter measured by the echocardiogram. The balloons are fully inflated until the "waist" disappears, then deflated. The procedure is repeated one to three times. The balloon combination must not be oversized to cause any significant mitral regurgitation. Inoue balloon technique is rather precise and easier to use but most centers still utilize the conventional balloon catheters. These are rare reports in the literature of success in dilating congenital mitral stenosis.58,59 In congenital mitral stenosis because of the complexity of the lesion, balloon dilatation has not gained major success.

Aortic Valve Balloon Dilatation

Balloon dilatation of congenital aortic valve stenosis can be performed with a single or double balloon.11,60,61 The balloon size chosen for single balloon is 90% of the aortic valve annulus diameter, and for double balloon the size is 110% to 120% of the annulus. The risk of the procedure in neonates is high, especially when they have a small left ventricular cavity. At the present time, surgery can be postponed for many years until the child is older. Twenty-five percent to 30% of these patients may develop aortic regurgitation after the procedure. Restenosis is not uncommon.62 A similar technique has been utilized for thin membranous subaortic stenosis. In all these cases, indications for balloon dilatation are the same as surgical indications.

Catheter Closure of Patent Ductus Arteriosus

Porstmann63 in 1967 pioneered nonsurgical catheter closure of the patent ductus arteriosus (PDA), although Gross had performed the first surgical closure of PDA in 1938. The Porstmann procedure consisted of insertion of an Ivalon plug into the ductus using a transfemoral arteriovenous guide catheter loop. This procedure was quite cumbersome for children. The initial studies of Rashkind on ductus closure involved the fabrication of a miniature grappling hook device that could be inserted through the femoral artery. The system was redesigned because of incomplete plugging and embolizations.

In 1979 Rashkind64 developed a new collapsible, double disc, hookless umbrella device that could be passed through the venous system. Mullins further modified the procedure by introduction of a long sheath through the pulmonary artery via the PDA into the descending aorta.65-70 A right heart catheter study is completed, including a descending aortogram through the ductus arteriosus in anteroposterior and lateral projections. The narrowest diameter of the ductus is measured; this should be less than 4 mm to obtain adequate fixation of a 12-mm occluder device, and the 17-mm device is required for a ductus greater than 4 mm in diameter. 68 An appropriate umbrella device is advanced through a long Mullins sheath into the ductus and released. A repeat aortogram is performed 15 minutes later. Intravenous antibiotics are given immediately after the release of the device. Echocardiopple studies are performed at 12 to 24 hours, 6 weeks, and 6 months after the procedure.71

We have performed 368 such procedures between December 1987 and November 1993. The age range was from 8 months to 45 years with a median age of 5 years. The male to female ratio was 1:2.4. Fifty-seven patients were less than 2 years of age. Implantation was successful in 358 attempts and unsuccessful in 10 patients. A second device was implanted in 49 patients because of residual shunt present at 6 months after the first implantation. Two patients required a third stent to achieve closure. Four patients required surgical closure of the ductus and retrieval of the device after it embolized to the
lung; they had an uneventful recovery. At 6 months follow-up, only 23% of 319 patients examined had patent. Although it is more difficult to implant in a long ductus, the ductus shape did not change the outcome of shunt patenty. The concerns in catheter closure of PD A are embolization to pulmonary or systemic circulation, endocarditis, and hemolysis. The limiting factors in the use of the occluder device are the size and weight of the patient. In young patients because of narrow anteroposterior diameter of the chest, the sheath may kink, making device insertion difficult. The longest follow-up has been 72 months and 20% of patients have residual small shunts either on clinical examination or echo-Doppler with color flow mapping only. These patients are still being followed for an indefinite time with a hope of spontaneous closure.

Transcatheter Atrial Septal Defect Closure

Transvenous atrial septal defect (ASD) closure was first reported by Mills and King in 1972.72 Rashkind in the late 1970s utilized a single-disc hooked prosthesis.73 In the early 1980s he utilized a double-disc device. The defect is sealed on both sides. Because of residual shunts and non-seated prosthesis, it was further modified by Lock 74 to a double-hinge clamshell device with arms folded against each other providing stability for the thin atrial septum. The procedure is still experimental. The delivery system pod size is UF. The device sizes are between 17 to 40 mm. The device chosen is between one and a half to twice the size of stretched ASD diameter. Only a few medical centers have experience with this device since it is presently under an investigational protocol. The person should be familiar with the anatomical landmarks of pulmonary veins, coronary sinus, superior vena cava, mitral valve, and location and size of the atrial defect. Residual shunts and embolization are the two main problems. This procedure should not be done without the help of transesophageal echocardiography in the catheter laboratory. The clamshell device has also been used in selected cases of muscular and apical ventricular septal defects.74

Coil Embolization

Various materials are utilized to occlude an unwanted vessel, including clots of the patient's own blood,75 small bits of Gelfoam,76 polymer glues,77 small coils of spring guidewire embedded with strands of fabric, 78 and small detachable mini balloons.79 Coil embolization was first described by Gianturco in 1975.80 It has only recently become widespread in congenital heart disease. The coils come in various diameters, lengths, and thicknesses and can be delivered through small catheters such as 4F for a mini coil, SF for a standard coil, and 6F or 7F catheter for large ones. The coils are made of preformed steel, have Dacron strands attached to them, and are loaded into cylinders with an internal diameter that is either 0.038 in or 0.025 in. Once the coil is extruded out by advancing a guidewire, the coil reforms to its stated diameter (2- to 12-mm sizes). The extended coil is thrombogenic and in a low flow vessel will create occlusion in 5 to 15 minutes. These are presently used to block an unwanted vessel or arteriovenous fistulas, such as pulmonary arteriovenous malformations to improve arterial saturation and also in cases of systemic bronchial vessel collaterals. Care must be taken to identify the diameter of the vessel being embolized. The coil diameter should be 20% to 30% larger than the stretched diameter of the vessel to be occluded.78 In general, the first coil should be the largest, preventing distal embolization with subsequent smaller coils delivered to "pack in" the occlusion site. The most likely complication appears to be inaccurate coil positioning and embolization of the coil to a normal vessel. A second complication has been arterial occlusion at the entry site.

Myocardial Biopsy

Intravascular endomyocardial biopsy method was first introduced in Japan in 1962 and subsequently modified by several laboratories, improving considerably in recent years. The availability of small, very soft SF, 104-cm long biopettes (modified by Lurie; Cordis, Miami, Florida) and preformed long sheaths (6F) have
made this method a rather safe procedure, even in infants.81-83 The three clear-cut indications in children for endomyocardial biopsy are the diagnosis of myocarditis, detection of cardiac transplant rejection, and detection and quantification of doxorubicin-induced myocardial disease. In other myocardial diseases which can be diagnosed with clinical examination and other investigations, the indication for myocardial biopsy is doubtful. In general, four samples are obtained from both ventricles, mostly from the septum with changing sheath position. The complications with either left or right ventricular biopsy have been few. The most common complication is cardiac perforation and tamponade. Other complications have included pneumothorax, heart block, atrial or ventricular arrhythmias, air embolism, recurrent laryngeal nerve paresis, and right phrenic nerve paresis.

Removal of Thrombus and Foreign Bodies

The technique of arterial embolectomy was first described by Fogarty in 1963.85 In this technique a Fogarty balloon catheter is used which in its original design was 80 cm long and 3F in diameter, with a latex balloon at its distal end. The catheter is threaded into the obstructed vessel by way of an arteriotomy and gently advanced past the thrombus. The balloon is then expanded with saline to a predetermined amount and slowly withdrawn, extracting the clot. The process is repeated as necessary.

Catheter techniques have been devised for the retrieval and removal of foreign bodies such as fragments of polyethylene central venous catheters, guidewire fragments, and fragments of pacemaker catheters.86-88 Three basic tools are used: the snare, the wire basket, and the endoscopy forceps.

Stents

In recent years, intravascular stents are being utilized in adult89,90 and pediatric cardiology.91,92 The procedure is still at an experimental stage. Few centers have used stents in humans in the pediatric age group to keep the stenotic pulmonary vessels expanded. Palmaz stainless steel or titanium (Johnson and Johnson Co.) stents have a 3-mm diameter and are 3 cm long which can be expanded with a balloon catheter placed in the stenotic lesion up to 12 mm and maximally to 18 mm. It is advanced through an IIF long sheath placed in the region of the stenotic vessel. A 7F balloon catheter is used and advanced through the stent and long sheath to the stenotic region and slowly inflated and deflated. If sustained expansion is needed, a high pressure Meditech Blue Max balloon catheter is used. These are thin balloons and can take up to 17 atmospheric pressure. The patients are heparinized during the first 12 hours after the procedure and then started on aspirin for 6 months. One of the complications is embolization of the stent by stent extrusion from the inflated balloon. Rupture of the vessel maybe a potential danger. These stents can be further expanded after initial installation.

Conclusion

The concept of cardiac catheterization has changed quite rapidly in the last decade, from diagnostic to aggressive interventional procedures in all age groups. Today, many therapeutic options exist and these include balloon atrial septostomy, blade septostomy, balloon dilatation of blood vessels and cardiac valves, balloon and coil embolization of vessels and shunts, foreign body retrieval from the cardiovascular system, transvenous closure of patent ductus arteriosus, closure of atrial and ventricular septal defects, and the use of stents in stenotic vessels.

Acknowledgment

The author is indebted to Mr. Joseph Franke for his technical expertise in the preparation of this manuscript.

References

PEDIATRIC CARDIAC CATHETERIZATION


