Pediatric and Congenital Therapeutic Cardiac Catheterization

Charles E. Mullins, MD

The concept that a catheter could be used for intracardiac therapy was first put into practice in 1964 with the dilation of acquired peripheral vascular stenosis by graduated rigid dilators. Isolated cases of intracardiac therapy with a catheter had appeared as early as 1953; however, the balloon atrial septostomy developed by Dr. Rashkind and reported in 1966 was the first pediatric and the first intracardiac therapeutic procedure with a catheter to be used as a routine form of therapy. This extremely innovative yet relatively crude procedure rapidly became the standard emergency therapy for the newborn with transposition of the great arteries. The balloon atrial septostomy not only proved lifesaving for hundreds of critically ill infants, but the procedure established that successful therapy could and should be accomplished with a catheter. In doing so, the Rashkind procedure also stimulated the imagination of other investigators in the field of catheter therapeutics.

Septostomies

Although there have been some improvements in the balloon septostomy catheter and in the technique of introduction of the balloon into the vascular system, the actual atrial septostomy technique and the indications for septostomy have remained unchanged for more than 2 decades. Because of its lifesaving capabilities, the balloon atrial septostomy still is a procedure that every pediatric cardiologist caring for infants with congenital heart disease must be able to perform skillfully and safely. In the era of echocardiographic diagnosis and prostaglandin therapy, the balloon atrial septostomy for infants with transposition of the great arteries remains one of the few indications for emergency cardiac catheterization of newborns with congenital heart disease.

The use of the balloon atrial septostomy was extended to patients with other complex congenital heart lesions, where inadequate mixing or venting of blood at the atrial level further interfered with a compromised circulation. For example, infants with tricuspid or pulmonary valve atresia, where systemic venous return could not reenter the effective circulation, were treated by septostomy, as were infants with mitral valve atresia, where the pulmonary venous blood could not join the systemic output. A third group of patients, where the restrictive atrial communication prevents both the systemic and the pulmonary venous blood from reentering the effective systemic circulation, is that with total anomalous pulmonary venous connection. Many patients from each of these groups have been successfully treated by a balloon atrial septostomy. However, in many of the slightly older infants, as a result of their tougher and more resistant septum, the balloon atrial septostomy has not been effective, or the effect has been very transient. This inadequacy of the procedure was a particular problem in the patients without transposition who often presented significantly later in life compared with the infant with simple transposition. This limitation of the balloon septostomy procedure led to the next significant development in therapeutic catheterizations.

In 1975, the blade septostomy procedure was introduced as an extension of the balloon septostomy. The blade septostomy procedure is used in conjunction with a balloon atrial septostomy and is indicated for any patient who is over 1 month of age and who requires an atrial septal defect for intracardiac mixing or atrial level venting. The blade septostomy is a safe and established procedure that should eliminate the need for a surgical septostomy. Blade and balloon atrial septostomy procedure has been performed successfully as late as the 3rd decade of life for palliation of right heart failure in patients with obstructive pulmonary vascular disease. The development of a larger 2-cm long blade and the use of valvuloplasty dilation balloons to enlarge the septostomy after the blade septostomy facilitated the procedure in these larger patients.

Valve Dilations

In the early 1980s, the next significant development in pediatric therapeutic catheterizations appeared with the dilation of valvar pulmonic stenosis. The use of small cylindrical fixed-diameter balloons, inflated at high pressures to

From Baylor College of Medicine and the Texas Children's Hospital, Houston, Texas.

Address for correspondence: Charles E. Mullins, MD, Cardiac Catheterization Laboratory, Texas Children's Hospital, 6621 Fannin, Houston, TX 77030.

Received November 2, 1988; revision accepted February 14, 1989.
generate rigid wall tension, had been introduced to dilate coronary arteries of adults in 1978. The same balloon concept was applied to larger balloons in 1982 for the dilation of valvar pulmonic stenosis. This technique gained rapid acceptance and wide investigational clinical use in pulmonic valve stenosis in pediatric patients. In a unique voluntary registry of 27 pediatric cardiac centers, data on more than 800 pulmonary valve dilations, compiled by the Valvuloplasty and Angioplasty of Congenital Anomalies Registry (VACA), showed that the balloon technique was safe and effective.

From early experience, it became clear that an effort had to be made to ablate the pulmonary valve. To accomplish that, a single balloon 20–40% larger than the valve anulus had to be used for an effective and lasting pulmonary valve dilation. In the process, pulmonary valve insufficiency is created by the pulmonary valve dilation. The insufficiency is considered to be hemodynamically inconsequential because of the normally low pulmonary diastolic pressures and small regurgitant fraction. Double- and even triple-balloon catheters were introduced for the larger patients with valve anuli greater than the available balloon diameters. Now, combined balloon diameters as much as 160% of the valve anulus are used. The double-balloon technique also was shown to have advantages in the smaller patients for ease of introduction and less damage to vascular structures by the use of two smaller profile balloons.

There does not appear to be any restenosis of adequately dilated pulmonary valves. The only patients who do not seem to respond to balloon dilation of the pulmonary valve are those with definitely dysplastic valves. Balloon dilation of pulmonary valvar stenosis has been approved by the United States Food and Drug Administration (FDA), for routine use, and now is the standard accepted therapy for isolated valvular pulmonic stenosis. Balloon dilation is recommended for any patient having valvular pulmonic stenosis with a gradient of 50 mm Hg or greater. It may be used in some patients with slightly smaller gradients in the presence of right ventricular hypertrophy.

After the success of the pulmonary valve dilation procedure, there was a tremendous increase in therapeutic procedures in the pediatric catheterization laboratory. Dilation of valvar aortic stenosis was reported in 1984. Initially, many pediatric cardiologists had concerns about the possibility of creating significant aortic valve insufficiency by the balloon technique. As a consequence, the aortic valve dilations were cautiously pursued. Gradually, reports appeared with recommendations for some modifications and limitations for the technique. The results have been satisfactory; however, there has been an incidence of significant aortic regurgitation between 10% and 15% but without high incidence of other significant systemic complications.

The technique for dilation of the aortic valve is surgically more conservative than that for the pulmonic valve because no attempt is made at full commissure to commissure splitting of the valve. The technique requires an accurate measurement of the aortic valve anulus and use of a balloon in diameter equal to or slightly less than the anulus diameter. A double-balloon technique allows the use of smaller profile balloons and reduces the trauma of the single larger balloon on the artery. When two balloons are used, a combined diameter of the two balloons should be between 10% and 20% larger than the valve anulus. Balloons 5.5 or even 8.0 cm in length are recommended for aortic valve dilation to prevent the balloons from being squeezed back from the valve into the aorta or forward into the ventricle during inflation.

There is a large variability and lack of predictability in the results of aortic valve dilation. This probably is related to the different structure of the valvular stenosis and to the necessarily conservative approach to the actual dilation. Because of these mixed results, the lack of long-term follow-up information and the demonstrated incidence of aortic regurgitation, only patients who otherwise unequivocally would require surgical intervention should undergo aortic valve dilation. The procedure itself remains investigational and should be performed only by those centers performing it regularly and gathering valid ongoing data to answer the unknowns about the technique.

There has been limited experience with the balloon dilation of the mitral or tricuspid valves in pediatric patients. Rheumatic valve disease has become relatively rare in developed countries. The bizarre structure of the congenital atrioventricular valve stenosis, along with the poor surgical experience in dilating these valves, has discouraged the use of balloons for these lesions. Several approaches and different techniques have evolved, and there now has been extensive and favorable experience in dilation of adult rheumatic mitral stenosis. These techniques are applicable to pediatric patients with rheumatic mitral stenosis. Successful dilation of congenital mitral stenosis has been reported and has been moderately successful in the Texas Children’s Hospital experience without significant complications. The double-transseptal, double-balloon approach, with a combined diameter of the two balloons equal to the calculated anulus diameter, appears most satisfactory for the mitral valve dilations in the pediatric patient. Because of the complexity of the basic technique and of the lesions themselves, dilation of atrioventricular valve stenosis in pediatric patients remains investigational. Mitral valve dilation should be performed only in centers having physicians with extensive experience in both the transseptal technique and with other balloon valvuloplasty procedures. The dilation of the congenital mitral stenosis should only be performed as a near-lifesaving procedure when the patient’s only other alternative would be valve replacement.
Vessel Dilation

Simultaneous with the use of fixed-diameter balloons for the dilation of valves, studies and reports on the use of the same balloons and similar techniques for vessel dilation have appeared.20–22 Recoarctation of the aorta was the first major vessel dilation attempted successfully in the pediatric population.23 This lesion appeared ideal for a transcatheter technique. The restenosis usually is discrete. The previous surgery results in scar and adhesions surrounding the lesion, making it less appealing for reoperation and, presumably, somewhat protectively “encased” by those adhesions. These theoretical considerations proved essentially correct, and an extensive body of data soon was accumulated in the VACA registry showing a large experience with the dilation of recoarctation of the aorta. The technique involves the retrograde arterial introduction of the dilation balloon. The balloon diameter should be equal to, or only slightly larger than, the nearest normal (usually proximal) aortic diameter. This procedure appears safe, and the results are satisfactory although not perfect. Frequently, there is a small residual gradient or slight residual arterial narrowing shown by angiography. However, considering the alternatives, balloon dilation now is considered a reasonable alternative approach for recoarctation of the aorta. With the exact anatomic consequence of the dilation of this lesion and the long-term results yet to be determined, this procedure still must be considered investigational.

With success of the recoarctation dilation, the natural extension of the same technique was to dilate the native or unoperated coarctation of the aorta. With that same basic technique, the immediate results of native coarctation dilation were even more satisfactory and with no more immediate complications than the recoarctation dilation.23,24 With follow-up studies of these lesions, however, there appeared reports of unusual dilatations or “aneurysms” occurring at the site of the previous coarctation.25,26 The consequence of these “aneurysms” is only speculative. However, because of these reports, many centers have abandoned the dilation of native coarctation, whereas those centers continuing to use this procedure are doing so under very strict investigational protocols with very close follow-up of each individual patient.

Similar to the situation with recoarctation of the aorta, stenoses of branch pulmonary arteries, whether congenital or surgically acquired, are difficult to approach surgically. Furthermore, surgical results have not been very satisfactory. This combination of factors, plus the relative ease at approaching these areas with a catheter, makes the stenoses of branch pulmonary arteries seem optimal for catheter dilation. Experimental evidence has shown that the normal pulmonary arteries can be safely overdilated severalfold.20 Clinically, the recommended technique is to use a balloon three to four times the diameter of the narrowing or up to two times the diameter of the adjacent normal vessel. Although most of the pulmonary branch stenoses can be acutely dilated, as many as 50% of the stenoses returned to the same degree of stenosis immediately after dilation.27,28 In addition, the early experience showed that there was a small but definite risk of vessel rupture from the procedure.28 Despite these drawbacks, the alternatives of surgery or of doing nothing are so unsatisfactory that an attempt at dilation of severe branch pulmonary artery stenosis is definitely indicated. This procedure should be attempted only in those centers performing such dilations regularly and with immediately available surgical back-up.

Even before the various arterial dilations, attempts had been made to dilate pulmonary vein stenosis29 and systemic venous, or “baffle,” stenosis after a “Mustard” repair of transposition of the great arteries.30 These early attempts showed that these venous channels could be acutely dilated. Although initially they appeared very successful, the stenoses recurred over a short period of time. With the newer balloon technology, there has been a renewed interest in these areas.31 The data in the VACA registry suggest more prolonged success at least in the area of the post-Mustard systemic venous baffle dilations. The use of balloons with combined diameters of at least two times the diameter of the nearest normal vessel seems to be effective. Again, because of the poor therapeutic alternatives, dilation of significant postoperative systemic venous obstruction is recommended.

Dilation of several other miscellaneous lesions has been reported with various success. The dilation of discrete membranous subaortic stenosis is reportedly acutely successful in approximately 50% of patients attempted.32 Even with significant relief of the stenosis, the lesion probably is never eliminated. If similar to the residual surgical lesions, recurrence can be expected. The mixed success may be related to subtle variations in the structure with some or many of the lesions actually being fibromuscular.

Stenotic anastomotic sites, such as post-Blalock-Taussig shunts, have been dilated, again, with mixed success probably related to the exact type of narrowing. Stenotic conduit tissue valves and anastomoses also have been subjected to attempts at balloon dilation with totally unpredictable but usually unsatisfactory results.33 Under very controlled investigational situations, attempts at these miscellaneous lesions should be attempted when surgery otherwise would be required. Even partial success at dilation may postpone the eventual and inevitable reoperation in these patients.

Vascular Stents

The high incidence of recurrence of vessel stenoses after dilation in both adult and pediatric patients has generated much interest and investiga-
tion into various devices for supporting or stenting these lesions. There are now some promising data on stents from animal studies and some early clinical data from studies in adults with atherosclerotic lesions. For the past 2½ years, the Palmaz stent has been used experimentally in pulmonary arteries and systemic veins in animals at Texas Children’s Hospital. The delivery technique has been perfected, and there are some very encouraging short-term and relatively long-term results from animal studies. On the basis of these studies, FDA approval is being requested to begin a limited and compassionate clinical investigational trial of the stents in selected severely affected children. Assuming the results from animal studies can be duplicated in children, these stents offer new hope, particularly, for those vascular lesions that up to now have been dilated successfully but have had a high immediate or long-term recurrence rate.

Occlusion Devices

The therapeutic catheter techniques discussed so far have been related to opening or dilating narrowed or occluded structures. In addition to the many narrowed or stenosed lesions among the congenital heart lesions, there are a large number of abnormal openings or communications that require closing. The closure of an opening by a catheter technique introduces a separate problem. With the exception of the vascular stents, which are yet to have clinical use in pediatric patients, the dilating procedures do not require the deposition of any foreign material in the body. All catheter techniques for the closure of discrete openings or vessels require depositing a device or material intended to remain in the patient for a lifetime with all of the potential unknowns of the effect of those materials on the tissues over a long period of time.

The earliest attempt at the transcatheter closure of defects occurred more than 2 decades ago in 1967 when the first report appeared of a catheter technique with a sponge plug for the closure of a patent ductus arteriosus without surgery. This particular technique was successful; however, it required very large introducing catheters in both the artery and vein. As a consequence, although it still is in limited clinical use for large children and adults, it was never applicable for infants or even small children.

In 1979, Rashkind used a small hooked umbrella and reported the successful closure of a patent ductus arteriosus in a 3.5-kg infant. A modification of this umbrella device and its delivery system into the present Rashkind double-umbrella ductal occlusion device has been the most successful of the catheter techniques for closing defects. This device has been used in investigational clinical trials since 1981, and since then, there have been improvements in the device and in the delivery technique with a continually improving success rate. No major morbidity or any mortality has occurred because of the device or the delivery technique. The present Rashkind devices are available in 12-mm and 17-mm diameters and are delivered through 8 French and 11 French delivery sheaths, respectively. The smaller device is usable for ductus up to 3–4 mm in the narrowest diameter, whereas the larger device is used for all ductus of 4 mm or larger in the narrowest diameter. Although the Rashkind device can be delivered from either the arterial or the venous route, most of the current procedures are performed through the venous route, thus avoiding any potential arterial complications. The small device can be used in infants weighing as little as 4 kg, although for elective closure, patients weighing over 10 kg are preferable should the use of the larger device be necessary.

The device is still investigational, however, and is approved for use in 12 centers in the United States and in four centers outside the United States. The most active eight centers in the collaborative study have used the double-umbrella device in over 380 patients. With the current device and delivery technique, there have been successful implants in over 95% of the cases, and complete closure of the ductus has been possible in 80–85% of these cases. The residual leaks are, for the most part, only detectable as tiny trickles noted only by high-quality contrast injection or by very detailed Doppler studies but are not detectable by auscultation. Several very large ductus that did have audible residual leaks after attempted closure have been completely occluded 6–12 months later with a second device.

The current device and delivery technique require very high-quality radiographic imaging and some special catheterization skills and training. The device is currently being reviewed by the FDA with the hope that it will become available for use on a noninvestigational basis by specifically trained individuals in selected institutions. With further experience and potential changes in the device, surgical therapy for the patent ductus soon should be relegated to only the premature and small infants who require urgent correction because of refractory heart failure.

The Rashkind device has been successfully used to occlude other structures on an individualized and compassionate basis in patients who otherwise were not good candidates for surgery. These other structures include large systemic to pulmonary collaterals, persistent abnormal venous channels and atrial septal defects after the Fontan procedure, a Potts shunt, and ventricular septal defects. The ductus device was not designed for, nor is it ideal for, any of these lesions. Not any of these uses can be considered routine nor are they presently used in any type of protocol study. However, these exceptional uses do highlight the potential for this type of catheter therapy. On the individual and compassionate basis, the ductus device should be available for use in designated centers already expert with the
standard implant of the device for these unusual situations. Catheter therapy for a variety of lesions should increase with the development of different types of occluding devices.

An umbrella device was used for the occlusion of atrial septal defects before the use of the umbrella device to occlude the ductus. The first successful implant of an atrial septal occluding device was reported in 1974. This device, a large rigid umbrella, had a drawback similar to the original Portman ductal device. It required a very large delivery catheter that made it unusable in most pediatric patients and totally unacceptable in the younger preschool-aged children when most atrial septal defects were being closed electively. Despite several successful atrial defect occlusions with the device, but presumably because of its size, this device never gained acceptance or any further use outside of the developing institution. A modification of the original Rashkind hooked ductal occluding umbrella was used successfully to occlude an atrial septal defect in 1977 and placed in very limited clinical trials in 1981. Although several successful implants were accomplished, the hooks of the device tended to attach to various unintended structures within the left atrium (appendage and mitral valve!) before attaching to the atrial septum. Consequently, emergency open-heart surgical removal of the device along with closure of the atrial septal defect was required. Because of this problem, the clinical trials for the hooked device were discontinued in 1987. The few successes with this device and the basic concept did stimulate the development of a new prototype atrial septal defect occluding umbrella that has proved successful in animal studies. This device, if approved for clinical trials, will remain investigational for some time before it will be generally available for closure of atrial septal defects. Eventually, this should be the preferred treatment for the small-to-moderate secundum atrial septal defects. This same prototype device, or a slight modification of it, may also prove satisfactory for the occlusion of some muscular and other intraventricular septal defects that are located away from the aortic valve.

Many techniques have been used for the occlusion of smaller vessels by the embolization of various types of materials to accomplish the occlusions. These techniques are best suited for small “end vessels” or tubular vessels with an area of discrete or localized narrowing. The various materials used include clots of the patient’s own blood, small bits of Gelfoam, polymer glues, small coils of spring guide wire imbedded with strands of fabric, and very small detachable miniballoons.

The preclotted blood and Gelfoam are used primarily for multiple small and diffuse end-vessel arteriovenous malformations and tumors structures. By themselves, they have little use in discrete congenital heart lesions. The glue materials are not approved for use in children. The detachable miniballoons are very effective in reaching more distal lesions with a tortuous afferent vessel. They also have the advantage of allowing a “test occlusion” before the actual release of the balloon. On the other hand, they are available in only two sizes and require tension against the vessel wall to become fixed securely in place. Unless placed in an area of significant discrete stenosis, the miniballoons tend not to fix securely, and in a vessel with high flow, they are drawn further into the vessel or even through the fistulous communication.

The most commonly used material or device for small vessel occlusions in congenital lesions are the coils. The straightened coils are delivered by pushing them through a thin-walled, end-hole catheter into the vessel to be occluded. As they are extruded from the end of the catheter, they coil into a predetermined diameter. The irregular spring configuration of the implanted coils fixes them securely against the vessel wall. The filament of fabric embedded in the coil initiates thrombosis in the vessel. The coils are available in sizes from 2 to 8 mm in diameter and thus are applicable to a wide range of vessel diameters. This type of device is used most frequently for the occlusion of persistent aortic to pulmonary collateral vessels in patients after surgery for severe right ventricular outflow tract obstruction and even for the occlusion of previously created “tubular” aortic to pulmonary shunts. The coils also have been used in conjunction with the PDA occlusion device for completion of the occlusion of larger tubular intravascular communications.

**Foreign Body Retrieval**

Another area of therapeutics in the cardiac catheterization laboratory that is gaining momentum is the catheter retrieval of intravascular foreign bodies. For the most part, the foreign bodies have been pieces of indwelling catheters that have become severed during attempted removal of the line. The severed piece then migrates to the right heart or pulmonary arteries. More recently, errantly embolized Rashkind occluding devices have been added to the materials retrieved from the circulation by a catheter technique. There are a variety of devices available for snaring and removing foreign bodies, which include wire snares, wire retrieval baskets, “grabber” forceps, and even bioptome forceps, all now usually used in conjunction with a long sheath. The device that is used depends upon the type of foreign body and the location of the foreign body at the time of the removal. Since 1988, most intracardiac or intravascular foreign bodies can and should be removed by a catheter technique.

Clearly, many therapeutic procedures can be performed successfully and safely in the catheterization laboratory—some more predictably than others. There is less mortality, less morbidity, and less cost for the procedure performed in the catheterization laboratory than for the comparable surgi-

References


Pediatric and congenital therapeutic cardiac catheterization.

C E Mullins

Circulation. 1989;79:1153-1159
doi: 10.1161/01.CIR.79.6.1153

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1989 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on
the World Wide Web at:
http://circ.ahajournals.org/content/79/6/1153.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/