Pediatric and Congenital Therapeutic Cardiac Catheterization

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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
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 valvular pulmonic stenosis.
This technique gained rapid acceptance and wide
investigational clinical use in pulmonic valvuloplasty
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 bal-
loon 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 pro-
cess, 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-7
and even triple-balloon8 catheters were introduced
for the larger patients with valve anulus 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.7

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 pul-
monary 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 pres-
ence of right ventricular hypertrophy.

After the success of the pulmonary valve dilation
procedure, there was a tremendous increase in
therapeutic procedures in the pediatric catheteriza-
tion 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.11,12

The technique for dilation of the aortic valve is
surgically more conservative than that for the pul-
monary 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.7
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 predictabil-
ity in the results of aortic valve dilation. This
probably is related to the different structure of the
valvular stenosis and to the necessarily conserva-
tive approach to the actual dilation. Because of
these mixed results, the lack of long-term follow-up
information and the demonstrated incidence of aor-
tic regurgitation, only patients who otherwise
unequivocally would require surgical intervention
should undergo aortic valve dilation. The procedure
itself remains investigational and should be per-
formed only by those centers performing it regu-
larly and gathering valid ongoing data to answer the
unknowns about the technique.

There has been limited experience with the bal-
loon 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 experi-
ence 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.13–16 These
techniques are applicable to pediatric patients with
rheumatic mitral stenosis. Successful dilation of con-
genital mitral stenosis has been reported, and has
been moderately successful in the Texas Chil-
dren’s Hospital experience without significant com-
lications. 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 dila-
tions in the pediatric patient. Because of the com-
plexity of the basic technique and of the lesions
themselves, dilation of atrioventricular valve steno-
sis 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.\textsuperscript{20–22} Reoperation of the aorta was the first major vessel dilation attempted successfully in the pediatric population.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{27,28} In addition, the early experience showed that there was a small but definite risk of vessel rupture from the procedure.\textsuperscript{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 stenosis\textsuperscript{29} and systemic venous, or “baffle,” stenosis after a “Mustard” repair of transposition of the great arteries.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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\textsuperscript{34-36} and some early clinical data from studies in adults with atherosclerotic lesions.\textsuperscript{37} 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.\textsuperscript{38} 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\textsuperscript{39} 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,\textsuperscript{39} 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.\textsuperscript{40} 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,\textsuperscript{41} and since then, there have been improvements in the device and in the delivery technique\textsuperscript{41} with a continually improving success rate. No major morbidity nor 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.\textsuperscript{42,43} 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 Portsman 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 interventricular 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 tumorous 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 embo-

ized 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” forcesps, and even bioptome forcesps, 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-
cal procedures. However, the exact patient selection and in turn the immediate results, the duration of the improvement, and finally, the net risk to benefit ratio for these therapeutic catheterization procedures are still to be determined. This information must be obtained by very careful and well-documented prospective studies carried out at least over the next several decades. With the exception of the balloon atrial septostomy, the blade atrial septostomy, the dilation of the valvular pulmonary stenosis, and the retrieval of foreign bodies, all of the other procedures should still be on carefully controlled investigational protocols. Centers providing these other therapeutic catheterization procedures should have both specially trained physicians and technicians to perform them. They must have the necessary high-quality imaging equipment as well as the large inventory of special catheters, devices, and support equipment to complete any procedure attempted. To fulfill these requirements and to maintain proficiency with these techniques, the laboratories engaged in all types of therapeutic catheterizations should perform them regularly and in adequate numbers.

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