TRANSCATHETER treatment of congenital heart disease has captured the imagination of cardiologists for some time. In 1968, Dr. Helen Taussig wrote these words of encouragement to Dr. William Rashkind "It would be wonderful if we can do some of the simpler operations without opening the chest. I think that is a real advance and a real look in to the future."1 Real advances have been made and the era for nonsurgical treatment of congenital heart lesions has begun. The closure of defects such as the patent ductus arteriosus (PDA),1-10 atrial septal defect (ASD),1,12,13 and, more recently, ventricular septal defect (VSD)14 can now be accomplished with catheters during cardiac catheterization.

Historical Perspective

Although the PDA was the first congenital heart lesion to be treated successfully by surgery,15 this defect was also the first to undergo closure by transcatheter technique.2 Beginning in 1966, Postmann et al2 used Ivalon foam plastic plugs, delivered by a transfemoral catheter technique, to occlude the PDA in 62 patients. Complete closure was achieved in 56 patients (90%) with no mortality and minimal morbidity. However, this technique was limited because it could only be performed in older children and adults with a conical-shaped patent ductus. Sato et alP subsequently modified the transfemoral approach so that it could be used in patients as young as three years of age, regardless of the shape of the ductus. The plug needed to be inserted into a surgically exposed femoral artery with a minimum of a 3 mm diameter and passed retrogradely via the arterio-ductal-venous loop as described by Porstmann.2

In 1979, Rashkind and Cuas04 reported a technique for the closure of the PDA in infants and young children using a single foam disk, with a miniature grappling hook made of stainless steel. The prosthesis was passed retrogradely from the femoral artery inside a delivery system and extruded into the ductus. With later developments, the PDA occluder was altered to improve the ease of implantation and rate of complete occlusion. I,3 The occluder now consists of a hookless device of two polyurethane foam disks mounted on two opposing, four-arm spring assemblies resembling a double-umbrella. The delivery system has also been modified so that the occluder can be inserted by the transvenous femoral approach using a long, transeptal sheath.5,6,9,10

Nonoperative closure of an ASD was first reported by King et al after some initial experimental work with dogS.11,12 They used a 35 mm diameter double-umbrella device of Dacroncovered stainless steel to close a secundum-type ASD of 25 mm diameter in a 17-year-old-girl. Adapting devices that were originally designed for PDA closures, Rashkind and his colleagues used initially the single-disk hooked device and, later, the double-disk hookless prosthesis to close ASDs in children and adults.1 More recently, the Rashkind double-umbrella device has been used to close a number of congenital and postoperative defects,13 including VSDS.14

Closure of Patent Ductus Arteriosus

Since most of the initial experimental work and clinical trials in transcatheter closure of congenital lesions were performed on PDA, this type of
lesions will be discussed first. The technology and experience with transcatheter device closure have advanced and developed, from the days when Porstmann et al.2 performed transfemoral plug closure of the PDA in hospitalized adults and older children to today, when infants and small children can undergo outpatient closure of the PDA using the Rashkind double-umbrella PDA occluder.9

The indications for closure of the PDA are discussed elsewhere.16 Since the smallest diameter device (12 mm), currently available, requires an 8-French delivery system, the age and the weight of the patient remain a restriction. The decision for surgical or nonsurgical closure of the PDA will also depend on the presence of associated cardiac anomalies that may necessitate future treatment. Lesions, such as common atroventricular canal defects, large ventricular septal defects, and most cyanotic heart lesions, associated with a PDA will require surgery and it, therefore, may be more appropriate to ligate the ductus at the time of repair. However, there may be circumstances when early closure of the PDA is desirable, even in the presence of other significant cardiac lesions such as infants with respiratory syncytial viral bronchiolitis or bronchopulmonary dysplasia who have severe respiratory insufficiency and are high risk candidates for surgery; lessening the amount of left-to-right shunting of blood by closure of the PDA may be beneficial in these circumstances.

Generally, however, transcatheter closure of the PDA is offered as an alternative to surgery in infants and children who have an isolated, restrictive PDA (i.e., without pulmonary hypertension) of less than 6 to 8 mm maximum diameter in medical centers that specialize in this technique. The advantages of transcatheter closure are that it can be performed on an outpatient basis without prolonged convalescence or extended hospital stay and a thoracotomy is avoided. There have been no reported deaths due to the procedure, and it is associated with minimal morbidity. The long-term concerns of late recanalization and infective endocarditis using this technique remain unanswered.

The technique for double-umbrella closure of the PDA has been well-described6-10 and recently has been modified to achieve a reliable and safe closure as an outpatient procedure.9 Vascular access is obtained percutaneously using the femoral vessels; a 7-French sheath is placed in the right femoral vein and the femoral artery is cannulated with a 4-French to 6-French ultra-thin-walled Teflon pigtail catheter. The catheters are flushed with heparinized saline, but systemic heparinization is avoided. A retrograde aortogram is performed to determine the precise size, configuration, and location of the PDA; the narrowest part of the PDA is measured using the lateral projection, usually between the anterior and posterior tracheal borders on the lateral projection. Although the Rashkind PDA occluder comes in 2 sizes (12 mm and 17 mm), the 12 mm occluder with an 8-French delivery system is used for the small PDA (4 mm or less), and the 17 mm umbrella with an 11-French delivery system is used for the larger PDA, up to 6 to 8 mm in diameter.

The Rashkind PDA occluder has a number of components: a double umbrella prosthesis that has a prosthetic ring for attachment, two foam pads attached to each set of umbrella arms, a machined clear plastic prosthesis loader, and the delivery system catheter. Included in the catheter is a central pin used to secure the prosthesis, a small sleeve attached to a second larger wire, the delivery pod inside which the prosthesis folds, a flushing seal and port, a black plastic locking collar, and the pin control-clamp and spring at the very proximal end of the catheter delivery system. Both the delivery device and prosthesis should be carefully inspected before use to ensure that they are fully operational and that there are no defects in the umbrella foam pad.

Using a 7-French balloon end-hole catheter and a 0.038 inch straight guide wire, the PDA is crossed from the main pulmonary artery to the descending aorta. With the guide wire placed down in the descending aorta, the venous catheter. and sheath are exchanged for the appropriate sized (Mullins) sheath and dilator (8-French or 11French). The curve on the long sheath may need to be partially straightened to about 900 with the use of steam or boiling water. The sheath and dilator are advanced together across the PDA over the guide wire, and an injection of contrast, by hand, is again performed through the sheath or the pigtail arterial catheter to confirm the precise PDA position; the transvenous placement of the long sheath through the PD A can distort the ductal position (usually posteriorly) by as much as 5 to 8 mm. The sheath is then flushed, prior to loading.
the PDA occluder.

Before loading the umbrella, the umbrella foam disks are soaked in topical thrombin for 5 to 10 min to promote thrombosis after placement. The double-umbrella is loaded into the pod by one operator holding the prosthesis ring gently against the fat part of the pin and partially engaging it, while a second person slowly pulls the pin control-clamp back, bringing both the pin and prosthesis ring back into the sleeve; the prosthesis is then securely locked into place, and by pulling hard on the loader-handle while the knuckle control clamp and rod are held firmly, the distal arms of the umbrella should collapse forward and slide into the plastic loader. The prosthesis is then inspected through the plastic to ensure that the 6 or 8 arms of the umbrella are properly collapsed with the distal ones forward and the proximal ones backward, and that there are no tears in the foam. Finally, the pod is directed into the loader to fit snugly into the large bore cylinder, and the pin control-clamp and rod are pulled smoothly back in order to load the umbrella device into the pod. The prolene sutures, which are tied to the prosthesis and the loader-handle, are cut and removed, leaving the device in the pod ready for placement.

The patient should now be adequately sedated (we use intravenous Ketamine) and extra local anesthesia should be given, if needed, prior to inserting the delivery system with the loaded device into the sheath. The pod is then advanced into the sheath up to the level of the tricuspid valve, taking care not to buckle the sheath. Since the right ventricle curves sharply from the tricuspid valve to the main pulmonary artery, the pod will dig into the sides of the sheath and causes it to buckle if attempts are made to advance it beyond this level. Therefore, the prosthesis is released into the sheath at this point by slowly pushing the control-clamp with the attached rod beyond the pod. The arms of the umbrella device will remain collapsed inside the barrel ofthe sheath, and they can now be advanced to the end ofthe sheath, still attached to the pin and wire. The tip ofthe sheath is held constant in the aorta just beyond the ductal opening, and the prosthesis is positioned directly over the center of the ductus. The pin control-locking spring is rotated so that the pin and wire curves are pointing posteriorly.

With the umbrella device at the end of the sheath and centered just past the ductus, the long sheath is slowly withdrawn until the distal arms open widely. As the entire system is moved back gently, the arms of the distal umbrella will partially collapse against the aortic margin of the POA (200 inward for a short, large POA and 450 for a small, long one). The prosthesis position is maintained absolutely constant as the sheath is then further withdrawn into the main pulmonary artery, now releasing the proximal umbrella. Both umbrellas, with all 6 or 8 arms, are now open in, presumably, the correct position, but the device is still attached to the pin and wire in the sleeve and can be retrieved, if necessary.

The position of the device can now be checked by moving the wire, gently back and forth, and watching the arms fold in and out to make sure that the prosthesis is not free in the aorta or pulmonary artery. If the umbrella is freed, in either the aorta or pulmonary artery, the wire is withdrawn back fully, pulling the prosthesis into the pod so that the device and the entire system can be removed, and another attempt can be made to occlude the POA. If the prosthesis is in the correct position, the device can be released by lifting the copper locking spring and sliding the pin control-clamp forward, thus dislodging the pin from the prosthesis ring. At this point, the pin may become entangled in the proximal foam disk and snare the device; if this occurs, the long sheath can be advanced over the wire and used to pin the proximal umbrella against the pulmonary end of the POA, while the wire and pin are retracted without dislodging the prosthesis.

After the device has been released, a repeat aortogram is performed with the pigtail catheter. If there is more than trivial residual ductal flow, then the delivery system can be removed and replaced with a 7-French balloon end-hole catheter which is used to "tamponade" the ductal flow, by pressing the inflated balloon against the pulmonary side of the umbrella for 10 to 15 min. This may help promote thrombosis and eventual occlusion of the PDA. When the procedure is completed, a chest X-ray is obtained 4 to 6 hours later to confirm the position of the device, and the patient can be discharged that same day after a brief period of observation.
The potential complications associated with this technique are embolization of the device, residual ductal leak, late recanalization, and infection with endarteritis. Embolization can occur because of incorrect placement of the device or using a device that is relatively too small for the ductus. The 17 mm diameter device should be used to occlude the PDA, with the greatest diameter at either the aortic or pulmonary end (4 mm or larger and up to a maximum of 6 to 8 mm). If incorrect placement is suspected at any stage prior to the release of the pin from the prosthesis ring, then the device should be withdrawn into the pod and another attempt made with a new device. If the device becomes dislodged after attempted placement and embolizes into the distal pulmonary arteries (most commonly, the left), then the options are to retrieve the device, either during catheterization with a snare or surgically at the time of PDA ligation if needed; or if the device is very peripheral in the distal arteries and not occluding any major vessel, it may be left to endothelize.

Retrieval of the umbrella (especially, a 17 mm device) through the right heart by snare or other devices can be hazardous if the device becomes entangled in valvular structures, since the valve tissue and apparatus can be damaged. In this circumstance, removal is best achieved at surgery. If the device embolizes to the systemic arterial circulation, then retrieval is mandatory.

Residual ductal leak after proper placement of the device can usually be dealt with at the time of cardiac catheterization. If "tamponading" the trans-PDA flow by temporarily occluding the ductus with a balloon-tipped catheter fails, then an additional device can be used at a later time to occlude the defect. Care must be taken not to dislodge the first device. Small leaks may resolve with time and, therefore, a period of observation for several months is warranted with follow-up Doppler echocardiography. Residual, large transPDA flow will likely necessitate surgical ligation of the ductus. Recanalization of a successfully occluded PDA has, as yet, not been reported, despite follow-up from 1981 with double-umbrella devices and from 1976 with single, hooked-disk umbrellas.

Because of the potential risks for infection with the implantation of a foreign device, we have routinely given prophylactic antibiotics at the time of the procedure, along with the use of strict asepsis.

No case of early or late infection with endocarditis has been reported to date. Since the incidence of infective endocarditis with PD A is low, the long-term follow-up of large numbers of patients with PDA, occluded by transcatheter devices, is needed to assess this potential complication, along with late recanalization.

Using the technique described, our recent experience at the Children's Hospital in Boston with over 75 cases would indicate a clinical success rate of over 90% in restrictive PDA. For nonrestrictive, hypertensive PDA, the closure rate has been less than 50%. We have encountered no significant morbidity.

Closure of Atrial Septal Defects

Using modifications of the Rashkind PDA occluder, transcatheter umbrella closures of congenital defects, other than the PDA, have been performed. Initially, the single-disk, hooked prosthesis was used to close ASDs and, subsequently, the double-disk umbrella with recent modifications has been used. The disadvantages of the single-umbrella technique with "fishhooks" are that it requires a larger delivery system (IS-French), it can only be used to close small defects since the valve tissue and apparatus can be damaged. In this circumstance, removal is best achieved at surgery. If the device embolizes to the systemic arterial circulation, then retrieval is mandatory.

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right-to-left shunting have been closed by umbrella devices. A coronary sinus septal defect was closed by Lock et al,13 but attempts have not been made to close other ASDs, i.e., sinus venosus or primum-type septal defects, because of the proximity of important intracardiac structures such as the pulmonary veins and the atrioventricular valves.

Similar to device closures of the PDA, preliminary assessment with echocardiography is important to measure the size and location of the defect, in particular, the supero-inferior margins and their respective distances from the right pulmonary veins, the coronary sinus, and the atrioventricular junction. It is also important to exclude other cardiac anomalies that will later require surgical repair, thus, mitigating against device closure of the atrial defect. In some cases, though, device closure may be used as an adjunct to surgery; one such case is the postoperative patient, with tetralogy of Fallot repair or pulmonary atresia with intact ventricular septum and right ventricular outflow tract repair, who had residual right ventricular dysfunction with right-to-left shunting at the atrial level via a patent foramen ovale. In these circumstances, after testing the suitability of the closure of the atrial defect by prior balloon occlusion in the cardiac catheterization laboratory, the transcatheter device closure may be beneficial.

The technique for implanting a double umbrella prosthesis across the atrial septum is similar to the closure of the PDA. Firstly, the defect needs to be “sized” and “tested” together with a balloon catheter prior to device closure. A 7-French balloon-tipped catheter (Critikon, Inc.) is advanced through the defect and the balloon is inflated with 1.5 cc of carbon dioxide, giving an external balloon diameter of 11 to 12 mm. If the defect cannot be occluded with this balloon size, then the large 17 mm diameter PDA occlusion device will be unable to effectively close this defect; closure may be better achieved by the newer, larger devices that are currently undergoing clinical trials. If occlusion is successful, then gentle traction should be maintained on the balloon catheter so that the defect can be occluded for 10 to 15 min and that the hemodynamics can be reassessed to determine if there is a favorable change with the closure of the defect. The precise position of the defect should be noted during balloon occlusion, since this may cause displacement of the defect which should be duplicated at the time of actual umbrella closure. Next, the balloon is slowly deflated until it just "pops" through the defect, hence, the size of the defect can be estimated from this critical balloon diameter. The 12 mm double-umbrella device can be used for defects 4 mm or smaller, and the 17 mm device can be used for larger diameter septal defects, up to 10 to 11 mm diameter; these recommendations may change pending the results of the clinical trials with the larger devices.

Secondly, the precise size and location of the defect are confirmed by angiography, preferably with multiple views to profile the defect, its shape and size, and its relationship to other structures. The sheath and dilator are placed across the defect, as in the case for PDA closure, having been "pre-shaped" in steam or boiling water to achieve an angle of 90° with the atrial septal wall. Unfortunately, the delivery system tends to straighten the angle of the sheath while retraction is applied during the release of the device, thus, presenting a relatively acute angle with respect to the plane of the septum. When the device is being released, the distal arms of the umbrella should open correctly, but the proximal arms may straddle the defect instead of being completely opened on the right atrial surface. The straddling arms of the proximal umbrella may be sufficient to securely close small defects (less than 6 mm in diameter) but would not achieve satisfactory closure of large defects. The newer devices may prove to be more effective in closing these defects because of their larger size and their better ability to self-center across a defect.

The double-umbrella device is loaded, delivered, and released in a similar manner as described for the PDA closure. One hundred units/kg of sodium heparin is routinely given intravenously for systemic heparinization at the commencement of the procedure because of the potential risks of clot formation and paradoxical embolization across the ASD; this is not done for PDA closures.

The potential complications of embolization of the device, infection with endocarditis, and residual shunt also exist for umbrella device closures of ASDs. These complications can be dealt with in a similar manner as those complications for device closures of the PDA. Again, prophylactic antibiotics are given routinely during the procedure.
dure to avoid potential infection which is introduced at the time of the device placement. There have been no reported cases of infective endocarditis or late recanalization of a device-occluded ASD in patients.

Closure of Ventricular Septal Defects

Transcatheter closure of VSDs using the Rashkind double-umbrella device is feasible but only in very selected cases, and the experience with this technique is limited. Attempts to close VSDs with this device have been confined to patients in whom closure of the defect is medically indicated but are poor or unsuitable candidates for surgery. These include patients with postinfarction VSDs who are hemodynamically unstable, those who have residual defects after repeated surgical attempts for VSD closure, and those in whom closure of the defect is warranted but refuse surgery. In addition to these limitations, the size and location of the VSD are factors to be considered in patient selection. Defects with maximum stretched diameters of less than 10 mm can be safely occluded by the large, 17 mm Rashkind double-umbrella, although current trials are in progress using larger devices to close septal defects. The VSD should be situated sufficiently far from the semilunar and atrioventricular valves to allow the umbrellas to open without impinging on these structures. Ventricular septal defects of the atrioventricular canal type and of the posterior muscular septum are unsuitable for transcatheter closures.

Similar to PDA and ASD closures, the size and site of the VSD need to be assessed by two-dimensional echocardiography and angled angiography, prior to attempted closure. The distance from the VSD to the aortic valve is measured, and the relationship to the tricuspid valve is assessed. The size of the defect and the potential hemodynamic effects of closure are firstly assessed by balloon-occlusion with a balloon end-hole catheter. Because of the heavily trabeculated right ventricle, it is easier to cross the VSD from the left side. Also the trabeculae may cross the defect, making it more difficult for the central placement of the guide wire which is needed for delivery of the device. Therefore, a 7-French balloon end-hole catheter is passed retrogradely from a femoral artery sheath into the left ventricle which is manipulated by looping the catheter and using a curved guide wire so that the balloon inflated tip is "seated" in the VSD. Either the partially deflated balloon or a soft J-tipped wire is passed through the VSD and advanced into the right atrium or pulmonary artery, where the catheter can then be exchanged for a 400 cm double-exchange 0.035 inch guide wire. This wire is then snared by a transvenous catheter and extruded from the body, creating a transfemoral arterial-VSD-transvenous loop. The transvenous entry site for snaring the guide wire will be determined by the location of the VSD. The right internal jugular approach is best suited for mid-muscular or apical muscular VSDs, whereas perimembranous or anterior muscular VSDs are best approached from the femoral vein; these entry sites are chosen because they allow for a more suitable approach for the long venous sheath and dilator that will be used to deliver the device.

Following the extrusion of the 0.035 inch 400 cm guide wire from the venous sheath, a second 7French balloon end-hole catheter is passed from the venous end of the wire through the right ventricle, across the VSD, into the left ventricle. The inflated balloon is used to assess the closure and the size of the VSD, as in the case for ASD device closure. It is important to assess the "stretched" diameter for closure. As mentioned above, the maximum diameter of the VSD should be 10 mm for a 17 mm device, although in 2 reported cases, the size of the VSD was 11 mm.14

Once the size of the VSD has been assessed and the location is confirmed to be suitable, the device is delivered to the defect in a similar manner as described in PDA and ASD closures, using a long venous sheath. The device, which has been pre-soaked in topical thrombin for 5 min, is released into the sheath at the level of the tricuspid valve and then advanced, still attached to the pin and wire across the VSD, to the end of the sheath. When the distal arms of the device are extruded into the left ventricular cavity, then the entire sheath-umbrella system is withdrawn until the proximal arms are opened with the prosthesis, now straddling and closing the VSD. After confirming that the position is correct and that the device is not free in the ventricular cavities, the prosthesis is released. Repeat hemodynamic and angiographic assessments of the closure are made prior to completing the procedure. Again, as in
the case of ASD closures, systemic anticoagulation is required at the start of the procedure in order to avoid clot formation and the complication of systemic embolization; a prophylactic antibiotic is given to avoid infective endocarditis with the introduction of a foreign body.

The potential complications for this technique are similar to those described for PDA and ASD closures; the management of these complications is also similar. These include embolization of the device, residual shunt flow, and infection. The long-term risk of late recanalization and infective endocarditis is unknown. From experimental animal data, these devices are known to be completely sealed and covered with endothelium within several months of implantation, 1,7 and in a postmortem examination of one patient who had a 17 mm double-umbrella inserted in an aortopulmonary connection 12 months previously, the communication was closed and completely covered by endothelium13; therefore, the risk of endocarditis with complete closure of a defect, without residual shunt flow, should be negligible.

An additional potential complication with implanting the current umbrella devices across the ventricular septum is that the arms of the pros thesis may not fully open and the device may partially collapse in the defect with arms protruding into the left ventricular outflow tract. Although this complication has not caused problems, such as outflow obstruction or a nidus for clot formation and infection, the experience and follow-up are limited.14 This emphasizes the need for devices that will fully expand when released and remain flush with the septal surface to allow complete endothelization later.

Conclusion

Transcatheter closures of the PDA, ASD, and VSD with the Rashkind double-umbrella device or the clamshell occluder are now feasible and are an alternative to surgery for small PDA, secundum-type ASDs, and apical muscular VSDs. It is important to assess the size and location of the defect by echocardiography and angiography and to determine the maximum distensible diameter of a septal lesion prior to umbrella device closure. The procedure should be performed by personnel who are experienced in interventional cardiac catheterization, and the technique should be adapted as a therapeutic intervention rather than a diagnostic procedure. Short-term and intermediate-term follow-up suggest that this technique is associated with minimal morbidity and mortality, but long-term follow-up is needed to determine the incidence of the late complications of recanalization and infective endocarditis.

References