Transcatheter umbrella closure of congenital heart defects

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ABSTRACT Between October 1984 and September 1986, we attempted transcatheter umbrella closure, using the Rashkind PDA occluder, of 12 congenital or postoperative cardiovascular defects (other than patent ductus arteriosus [PDA]) in 11 patients. In each, we used the umbrella for closure because the defect was too short and/or too large to close with conventional transcatheter methods. The defects included three post-Glenn venous communications (superior vena cava–right atrium, n = 2; azygos vein to inferior vena cava), four congenital “interatrial defects” producing cyanosis (“coronary sinus” septal defect, left superior vena cava to left atrium, patent foramen ovale, left inferior vena cava to left atrium), and five non-PDA systemic–to–pulmonary arterial communications (two congenital and three postoperative). Ten of 12 defects were embolized successfully; nine had complete or subtotal closure, and one was partially closed. The first attempted closure resulted in embolization of a 12 mm device to a lower-lobe pulmonary artery, without clinical sequelae. No other complications occurred. Clinical improvement was most dramatic in those patients whose cyanosis was relieved and less obvious when pulmonary blood flow was reduced. The Rashkind umbrella device, originally designed for closure of PDA, considerably expands the list of congenital or operative defects that can be closed nonsurgically.


TRANSCATHETER CLOSURE of congenital or postoperative cardiovascular defects, until recently a medical curiosity,1,2 has in the last 5 years become standard practice in some centers.3,4 Without exception, the vessels closed to date have been channels with either distal stenoses, unwanted capillary beds, or tortuous arteriovenous malformations. In these cases, the vascular anatomy has been such that distal migration of large embolic devices would be quite unlikely, allowing the safe use of either Gianturco coils or detachable balloons.

There are many cardiovascular defects whose anatomy has not been so favorable for transcatheter closure: defects that are quite short cannot be safely straddled by a coil or a balloon, and defects larger than 8 or 9 mm in diameter are too large to safely trap the currently available coils or balloons.

The patent ductus arteriosus (PDA) is such a defect: generally short and frequently large, the PDA cannot be closed with coil or detachable balloon. Postmann et al.5 and others6,7 took advantage of the “intracardiac” location of the PDA to develop a long wire and plug technique to effect transcatheter PDA closure. More recently, Rashkind and Cuaso8 developed a double-umbrella prosthesis that could be delivered to the duct via No. 8F or 11F catheters. This device, recently modified, has been used to close PDAs in experimental animals9 and children10 with increasing success.11

These umbrella devices appear well suited to close other “unfavorable” congenital or postoperative cardiovascular defects of various shapes and sizes. However, the methods for patient selection, preliminary evaluation, and sizing of defects, the techniques for delivery and release of the umbrella device, and the determination of the success and complication rates can be determined only from experimental clinical trials.

Materials and methods

Patient selection. Patients were considered candidates for transcatheter umbrella closure only if three criteria were met: (1) the defect could not be safely closed by standard transcatheter techniques (e.g., coils, detachable balloons); (2) the defect produced significant cardiovascular symptoms or disability; and (3) if cardiac surgery for other abnormalities was envisioned,
the defect could not be easily closed from the same operative approach.

Patients who met the above conditions were presented to a cardiology/cardiac surgery joint conference. Transcatheter closure was attempted only if recommended in that conference. When such a defect was encountered unexpectedly during a diagnostic cardiac catheterization and umbrella closure appeared warranted, prior consultation with a staff cardiac surgeon was obtained before proceeding with closure.

The FDA protocol for transcatheter closure of PDAs was approved by the Committee on Clinical Investigation at the Children’s Hospital. This modified use of the Rashkind double umbrella for non-PDA structures was approved in advance, whenever feasible, on a case-by-case basis by the Committee on Clinical Investigation. In each case, informed consent was obtained from the patient and/or the patient’s parents.

Procedure
Preparation for closure. Vascular access for the delivery system was obtained percutaneously in each case: via the femoral or subclavian vein in seven cases and via the femoral artery in five cases. In the first four patients, we used simple variations of the long-sheath PDA occlusion technique of Bash and Mullins9 as subsequently modified. After the difficulties encountered in the first few patients (see below), a protocol designed specifically for closure of non-PDA structures was employed.

1. The precise size and location of the defect to be closed was determined angiographically. Preferably, multiple views were obtained to optimally profile the defect, its shape, and its diameter.

2. A No. 7F balloon-tipped catheter (Critikon, Inc.) was advanced to and through the defect. The balloon, inflated with 1.2 to 1.5 cc of CO₂, was used to “test” occlude the defect. During test occlusion, we determined several things. (a) Was the defect occluded? If a 1.5 ml balloon (diameter 11 to 12 mm) did not occlude a defect, that defect was deemed too large for any transcatheter device, regardless of the angiographically determined diameter. (b) Did stretching markedly change the size of the defect? Most vessels are compliant to varying degrees. A vessel may be 6 mm in diameter on the initial angiogram but may stretch to 9 mm when test occluded (figures 1 and 2). It was the occluded diameter that we used to determine which device to employ. (c) Were the hemodynamics favorably altered by test occlusion? In most cases, the physiologic success of a procedure could be determined in advance. (d) Does test occlusion alter the precise position of the defect? To occlude something, the balloon was passed distal to the defect, inflated, and withdrawn until snug. This withdrawal occasionally displaced some defects 1 cm or more. Since this displacement would be duplicated at the time of actual umbrella closure, it was ascertained in advance. (e) What was the distance from the vascular entry site to the defect? This should be noted, using the 10 cm markers on the catheter side. Since any sheath more than 75 cm long cannot be used to successfully deliver the device and defects more than 75 cm from the groin cannot be closed via the femoral vein (using the sheath technique), a closer vascular entry site should be sought.

Closure. Once evaluation of the defect was complete and closure was thought to be warranted and possible, the defect was crossed with an end-hole catheter (0.035 to 0.038 inch lumen). We use either a 12 mm double-umbrella prosthesis (defects 4 mm or smaller) or a 17 mm umbrella (defects 5 mm or larger). After the first case, we have used only 17 mm umbrellas.

For the 17 mm device, a No. 11F long sheath (U.S.C.I., or Cook Inc.) was selected. The shape of the sheath was altered by steam, the dilator, and stiff wires, to fit the catheter course. For example, the curve in a long sheath held in boiling sterile water for 60 sec while straightened over the curved end of the long dilator will change from 180 to 90 degrees; if the sheath is then held in boiling water for 3 min, straightened by the straight end of the dilator, it will further change to a 40 degree curve.

Once the sheath was prepared and the device selected and flushed, the patient was anesthetized (generally ketamine) to ensure immobility at the time of implantation. The device was loaded as previously described.9, 10 Before loading, the devices and loading system were carefully inspected to ensure that there were no metal burrs on the release mechanism and that the

FIGURE 1. Angiographic demonstration of a moderately long, large aortopulmonary collateral in a patient (No. 6) with repaired tetralogy of Fallot. The diameter of the collateral is clearly smaller than an 8 mm coil laid on the patient’s chest for purposes of comparison (open arrow).
polyurethane foam was intact and attached to the metal arms.

After the No. 11F sheath and dilator were passed over the exchange guidewire through the defect, the dilator was removed so that the long sheath was positioned at least 1 to 2 cm past the defect. As during a PDA closure, the delivery catheter was advanced to about 5 to 10 cm short of the sheath tip, stopping short of any acute or subacute bends in the long sheath. By means of the delivery wire, the umbrella was advanced out the end of the delivery catheter and into the lumen of the long sheath, thus using the long sheath as an extension of the delivery catheter (figure 3). With the center of the double-umbrella device held carefully in (or just distal to) the center of the defect to be closed, the sheath was withdrawn until only the distal arms were open. The entire system (sheath, device, and catheter) was withdrawn as a unit until the distal arms were seen to flex inward. With the umbrella fixed in this flexed position, the long sheath was withdrawn 2 to 3 cm until both distal and proximal sets of arms were open, straddling the defect. The fully sprung (but unreleased) device was nudged back and forth to be certain it was correctly positioned, and the device was then released as previously described.9,10

Hand injections of contrast through the long sheath 1 to 2 min after implantation usually indicated nearly complete closure; if not, a balloon-tipped catheter was advanced through the long sheath and inflated near to the umbrella for 5 to 10 minutes to occlude flow and promote clot formation. If the defect remained patent for more than 20 min and if the anatomy was favorable, a second umbrella or 8 mm coils were delivered upstream to the first umbrella to further reduce flow.

Miscellaneous. Other than making a unit of blood available, no special precautions were undertaken. All patients received the usual dose of heparin (100 U/kg) at the beginning of the procedure. Although our surgical colleagues were notified of each case, we did not make a cardiac operating room available. Most patients received one dose of prophylactic antibiotics just before implantation of the device. All but one patient recovered on the general cardiac ward overnight, with discharge the day after implantation. Each patient has been followed clinically and with Doppler echocardiography.

Data analysis. All combined data are expressed as the mean ± 1 SD. No tests of statistical significance were used.

Results

We attempted to close 12 defects in 11 patients. Patients’ characteristics and diagnoses are listed in table 1. Closure was required because of persistent cyanosis (n = 7) or persistent pulmonary hypertension (n = 5).

A wide range of rather uncommon diagnoses was encountered, including a left inferior vena cava connecting to the left atrium (via hepatic veins) in a patient with heterotaxy, a dilated septal coronary vein (vein of Kugle) connecting the coronary sinus to the left atrium, and a short, squat systemic-to-pulmonary collateral (a “Potts-like” collateral). All 10 patients had had at least one prior cardiac operation, and three patients (Nos. 1, 4, and 11) had had at least one prior operation that unsuccessfully attempted to close the residual systemic-to-pulmonary connection.

Procedure

Preparation for closure. In each of 12 cases, we were able to define the defect angiographically and pass a catheter across it. The defects were relatively short and PDA-like in eight cases and relatively long but wide in the other four. All 12 defects were entered via the femoral artery or vein, but in two cases (patients 5 and 10b) the defect was more than 65 cm from the femoral vein, making a second percutaneous entry into the subclavian vein necessary.

During test occlusion, one defect (the patent foramen) was found to be highly mobile; the others moved less than 0.5 cm when occluded with a balloon catheter. In two patients (Nos. 6 and 7) the defects enlarged from 6 to 9 mm and from 7 to 11 mm, respectively, indicating that occlusion with an 8 mm coil would have been hazardous.

One patient, not included in this report, had a double-inlet left ventricle and a regurgitant right-sided atrioventricular valve. A Fontan repair included sewing a patch just above the right atrioventricular valve ring, with the coronary sinus between the valve and the patch. At postoperative catheterization, regurgitation across the valve produced high pressures in the coronary sinus, which now communicated with the innominate vein via a restrictive left superior vena cava (LSVC). The tricuspid regurgitation caused, in effect, a left to right shunt. Although transcatheter closure of the LSVC appeared technically straightforward, test occlusion of the LSVC produced coronary sinus pressures in excess of 30 mm Hg, and the patient was
TABLE 1
Patient characteristics

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Weight (kg)</th>
<th>Diagnoses, surgery</th>
<th>Hemodynamic disorder</th>
<th>Device</th>
<th>Positioned correctly?</th>
<th>Complete, subtotal or partial closure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8</td>
<td></td>
<td>Tetralogy of Fallot, pulmonary atresia; midaorta and collateral vessels used to fashion neopulmonary artery; residual aortopulmonary window</td>
<td>Pulmonary hypertension</td>
<td>12 mm</td>
<td>Yes</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>12</td>
<td></td>
<td>Tricuspid atresia; S/P Glenn shunt; residual SVC-to-right atrial hole</td>
<td>Cyanosis</td>
<td>17 mm</td>
<td>Yes</td>
<td>Complete</td>
</tr>
<tr>
<td>3</td>
<td>10</td>
<td></td>
<td>Pulmonary atresia; intact septum; patent foramen ovale</td>
<td>Cyanosis</td>
<td>17 mm</td>
<td>No*</td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>31</td>
<td></td>
<td>Tetralogy of Fallot S/P Potts shunt S/P repair; persistently patent Potts</td>
<td>Pulmonary hypertension</td>
<td>17 mm</td>
<td>No*</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>14</td>
<td></td>
<td>Single ventricle; S/P Fontan; left SVC to left atrium</td>
<td>Cyanosis</td>
<td>17 mm</td>
<td>Yes</td>
<td>Subtotal</td>
</tr>
<tr>
<td>6</td>
<td>18</td>
<td></td>
<td>Tetralogy of Fallot; pulmonary atresia; S/P repair; large aortopulmonary collateral</td>
<td>Pulmonary hypertension</td>
<td>17 mm</td>
<td>Yes</td>
<td>Complete</td>
</tr>
<tr>
<td>7</td>
<td>4</td>
<td></td>
<td>Single ventricle; abdominal heterotaxy; S/P modified Fontan; persistent left inferior vena cava to left atrium</td>
<td>Cyanosis</td>
<td>17 mm</td>
<td>Yes</td>
<td>Subtotal</td>
</tr>
<tr>
<td>8</td>
<td>10</td>
<td></td>
<td>Tricuspid atresia S/P Fontan; persistent communication from enlarged coronary sinus to left atrium via vein of Kugle</td>
<td>Cyanosis</td>
<td>17 mm</td>
<td>Yes</td>
<td>Complete</td>
</tr>
<tr>
<td>9</td>
<td>10</td>
<td></td>
<td>Tetralogy of Fallot; multiple large collaterals with one very large short aortopulmonary collateral</td>
<td>Pulmonary hypertension</td>
<td>17 mm</td>
<td>Yes</td>
<td>Partial</td>
</tr>
<tr>
<td>10a</td>
<td>26</td>
<td></td>
<td>Single ventricle S/P Glenn; persistent opening between SVC-right atrium</td>
<td>Cyanosis</td>
<td>17 mm</td>
<td>Yes</td>
<td>Subtotal</td>
</tr>
<tr>
<td>10b</td>
<td>—</td>
<td></td>
<td>Largeazygos vein draining to inferior vena cava</td>
<td>Cyanosis</td>
<td>17 mm × 2</td>
<td>Yes</td>
<td>Complete</td>
</tr>
<tr>
<td>11</td>
<td>19</td>
<td></td>
<td>Tetralogy of Fallot; residual Potts shunt</td>
<td>Pulmonary hypertension</td>
<td>17 mm</td>
<td>Yes</td>
<td>Subtotal</td>
</tr>
</tbody>
</table>

S/P = status post; SVC = superior vena cava.
*Device embolized to left lung. See text.
*Devices retrieved before release.

referred for operative closure of the coronary sinus ostium.

Closure. In 10 of 12 attempts, the umbrella was successfully delivered to the defect and released correctly. In two cases (patients 3 and 4), one with a postoperative Potts shunt and one with a patent foramen ovale, the distal arms were opened correctly distal to the defect and the device was withdrawn to the orifice until two of the arms flexed. In both cases, the axis of the catheter withdrawal was not at all perpendicular to the “wall” of the defect to be closed (figure 4). With distal arm flexion, the proximal arms were opened but in both cases the device did not appear stable when nudged. In both, it appeared as if six of the eight arms were opened on the distal side of the defect. The devices were not released, but rather were pulled back inside of the sheath and delivery catheter and removed. These two procedures were terminated, and both patients underwent subsequent operative closure.

In retrospect, it seems likely that because of the angle of approach, the umbrellas (with the distal arms opened) were not pulled firmly enough against the defect to engage all four arms against the sides of the defect (figure 4). Thus, when the proximal arms were opened, only two of them straddled the defect. It may still be possible to close such defects, although the defect size would need to be smaller (i.e., no more than 6 to 8 mm); in small defects approached from an acute angle, only two of the arms would be needed to restrain the umbrella during retraction. Changing the

FIGURE 4. The angle of catheter “attack,” if acute, will render umbrella delivery difficult. The distal arms may be opened correctly (left) and withdrawn until the leftward arms are seen to flex (center). If the sheath is then drawn back, the proximal arms may straddle the defect (right) so that six of eight arms are on the same side.
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The other 10 umbrellas were correctly and successfully positioned on the first attempt. However, in patient 1 a 12 mm device was positioned across a 6 mm surgically created aortopulmonary window. After release, it remained in position for 6 heart beats but was then swept into a branch lower-lobe pulmonary artery.

Given the complex nature of the right ventricular-to-pulmonary “artery” conduit, we were unable to retrieve the umbrella. Thirty minutes later, the pulmonary artery branch was still unobstructed angiographically. Since two previous attempts at surgical closure of this aortopulmonary window had been unsuccessful and since angiography indicated that the embolized lower-lobe artery was still patent, we proceeded to close the aortopulmonary connection with a 17 mm device without incident.

In seven of 10 vessels embolized successfully, a single umbrella caused complete or near-complete cessation of flow (figures 5 and 6). Of the three with persistent flow, two were in patients with very large (more than 10 mm) venous channels. In one (No. 10b), a second 17 mm umbrella (figure 7) completely occluded flow. In the other venous channel (No. 7) several 8 mm coils were packed in behind the umbrella to occlude flow. Only in patient 9 did significant (albeit markedly reduced) flow persist. This patient subsequently underwent successful operative repair of his tetralogy of Fallot, without requiring further operative control of the collateral.

Complications. Other than the embolized device noted above, complications were few and insignificant. No patient developed apparent vascular compromise despite the large (No. 11F) sheaths used. During retrieval of one of the unreleased umbrellas (patient 4), the arms would not fold completely back into the catheter or sheath: the umbrella dislodged partly through the cutaneous fascia at the groin but was retrieved via a 5 mm local incision.

FIGURE 5. Injection into a dilated coronary sinus (top) causes cephalad filling of an atrial septal vein (closed arrow) and hence the left atrium (patient 8). After positioning a double umbrella (center) with arms straddling the distal (narrowest) end of the defect, right to left shunting is eliminated.

FIGURE 6. Persistent shunting from superior vena cava (large arrow) to left atrium follows a Fontan procedure, producing cyanosis. A double umbrella (small arrow) occludes flow.
No other complications occurred. Specifically, there have been no signs of emboli (pulmonary or systemic), endocarditis, or device dislodgment.

Clinical follow-up. The patients with systemic-to-pulmonary communications remained largely unchanged after the procedure, at least for the first 6 months of follow-up. Those with cyanosis caused by right-to-left cardiac shunts had, in all cases, improved exercise tolerance and/or oxygenation after embolization as well as diminished cyanosis.

One child (No. 1) began to develop signs of progressive right heart failure 6 months after embolization. These findings were attributed to progressive pulmonary vascular disease and perhaps pulmonary arterial obstruction. She died 12 months after umbrella occlusion. At postmortem examination, the 17 mm (correctly positioned) device was completely covered with endothelium, except for two metal arm tips that were visible from the aortic end (figure 8). The aortopulmonary connection was closed. The 12 mm device had lodged in a lower-lobe branch pulmonary artery with the two polyurethane discs parallel to the axis of blood flow. Despite being covered with fibrin, the umbrella neither occluded nor significantly obstructed flow to the distal branch pulmonary artery.

Discussion

Congenital cardiovascular defects that have been previously closed with transcatheter devices include arteriovenous malformations, systemic-to-pulmonary arterial collaterals, the PDA, and atrial septal defects. In this report, we have described the adaptation of the Rashkind double umbrella to close a variety of congenital and postoperative cardiac defects; the expanded list of defects that can be closed with transcatheter therapy now includes superior vena cava to right atrial openings, persistent left inferior and superior vena cava to left atrial channels, a “coronary sinus septal” defect, postoperative aortopulmonary windows, Potts shunts, and a large azygos vein. In this process, several important observations were made on the adaptation of double-umbrella closure of unconventional cardiac defects.

Defect sizing. Angiographic determination of defect size is not always adequate before transcatheter closure. Angiography alone will not assess defect compliance; some defects (especially large veins) may stretch considerably when an occlusion device such as a balloon is inserted (figures 1 and 2). It appears that defect sizing with a balloon-tipped catheter is an important first step before umbrella closure of unconventional cardiovascular defects is attempted.

Catheter angle. Ideally, when using a double umbrella the delivery catheter or sheath approaches the defect...
at a right angle, allowing the distal arms to self-center the device. A very steep angle may allow poor device centering, so that the arms will not correctly straddle the defect (figure 4). It would seem that such defects will be successfully closed only if they are less than half the size of the umbrella, making proper centering less important, since only two of the arms would be needed to restrain the device.

Device delivery. The shortest, most direct, and most perpendicular approach to a defect, coupled with the use of the long sheath and dilator to cross the defect over an exchange guidewire, allowed us to successfully cross each defect we considered closing. Determination of the degree of defect displacement that occurs with test occlusion allowed proper positioning, even in very mobile defects.

Using these technical modifications, we occluded or substantially reduced flow through 10 of 12 congenital or postoperative cardiac defects, including the last eight in a row. This technique is new, and undoubtedly further modifications will be needed. Furthermore, the late effects of these devices remain unknown. Nonetheless, given that operation for some of these defects may be either difficult or hazardous and that complications of transcatheter closure were few and clinically insignificant, it seems likely that transcatheter Rashkind umbrella closure of various congenital cardiovascular defects will frequently replace operative management for many lesions.

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