

EDITORIAL

The Use of Catheter Intervention Procedures for Congenital Heart Disease*

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The pioneering proposals to use catheter procedures to treat congenital heart disease were published over 20 years ago (1-3). Only in the last half decade, however, have most of these procedures achieved sufficient clinical maturity to affect the delivery of cardiac care to children. In some centers, such procedures now constitute a large percentage of all cardiac catheterizations performed (Fig. 1).

During the last 5 years, various workers have laid the experimental foundations for most of these transcatheter techniques. Others have described the initial clinical experiences for balloon angioplasty of pulmonary veins (4), pulmonary arteries (5), pulmonary valves (6), native (7) and recurrent (7,8) aortic coarctations, aortic valves (9) and obstructed intracardiac baffles (10). In addition to these balloon angioplasties and valvuloplasties, other transcatheter techniques have been successfully applied to congenital heart disease, including knife atrial septostomy (11), closure of the patent ductus arteriosus (12,13), closure of large systemic pulmonary artery vessels (14,15) and arteriovenous fistulas (16). As a group, these procedures are still too new to identify their indications and contraindications with any precision. Nonetheless, enough clinical experience has accrued to allow a discussion of how these techniques appear to fit into the current practice of pediatric cardiology at our own institution.

Procedures Currently Indicated*Balloon Catheter Procedures*

Balloon atrial septostomy. This procedure, the first catheter intervention to prove useful in cardiac medicine

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(2), remains the standard initial palliation in transposition of the great arteries. It is safe and very effective.

Pulmonary valve stenosis. Balloon valvuloplasty for typical pulmonary stenosis outside the newborn period is a very safe procedure; several hundred have been performed in the past 2 years without a reported fatality or major morbid event. Initial reports (17,18) on the use of balloons less than the size of the anulus described gradient reductions of 50 to 60%. Subsequent use of balloons 20 to 40% larger than the anulus have produced gradient reduction (70 to 75%) equivalent to that of surgery (19). Short-term (1 to 3 years) follow-up has indicated no significant incidence of restenosis or important regurgitation. The initial notion that the severely dysplastic pulmonary valve generally cannot be dilated has, in our experience, held up; although it is reasonable to attempt dilation in patients with such valves, most will require operative resection of valvular tissue, together with anular augmentation. Thus, we now consider balloon valvuloplasty the treatment of choice for typical valvular pulmonary stenosis.

Recurrent postoperative aortic obstructions. Despite the ongoing advances in surgical management of aortic arch obstruction, the incidence of recurrent narrowing after repair in infancy of coarctation, interruption of the aortic arch and hypoplastic left heart syndrome remains 5 to 20%. Several centers have reported good results from reoperations with low mortality rates and gradient reductions approaching 70%. It is now clear that equivalent results can be achieved with balloon angioplasty (20).

The major unresolved issue in these patients is that of late aneurysm. Neither postoperative nor postangioplasty patients have been studied carefully over a long period of time to ascertain the incidence of this complication. Despite the limited information now available, we consider angioplasty the treatment of choice in all such patients.

Hypoplastic or stenotic pulmonary arteries. Balloon angioplasty of such lesions remains technically difficult, and there is a small (1 to 2%) but significant mortality rate associated with the procedure. Because operative management of such lesions outside the mediastinum is unsuccessful, angioplasty remains the only therapeutic option for

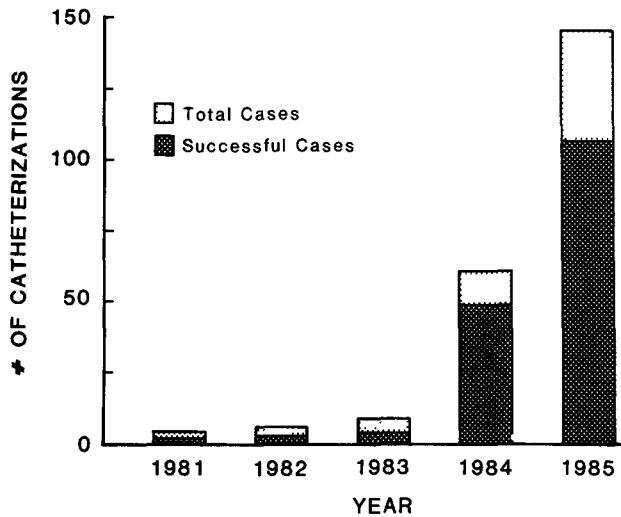


Figure 1. Therapeutic cardiac catheterizations at Children's Hospital, Boston. This increase in therapeutic catheterizations occurred with total case volumes ranging from 650 (in 1983) to 825 (in 1985), representing an increased incidence of procedures from 1.5 to 18%.

most patients. Fifty percent of such lesions can be successfully dilated (increasing the diameter of the narrowed artery by 50% or more) with higher success rates in patients less than 3 to 4 years of age.

Any child with symptomatic right heart failure, cyanosis or suprasystemic right ventricular pressures due to pulmonary artery stenosis should undergo balloon angioplasty, preferably in the first 3 to 4 years of life. Children with a near systemic right ventricular pressure or marked flow abnormalities (that is, <15% of total cardiac output going to one lung) should certainly be considered for balloon angioplasty. It is clear that even minor degrees of pulmonary artery narrowing are poorly tolerated after the Fontan operation; lesions that produce mean gradients as small as 3 to 4 mm Hg after a Fontan procedure should probably be dilated in symptomatic children.

Finally, balloon angioplasty may be very useful as part of the pulmonary vascular rehabilitation of patients with tetralogy of Fallot, pulmonary atresia and small pulmonary arteries. In such patients, we are attempting early establishment of central pulmonary artery flow from the right ventricle, followed by angioplasty of the residual narrowings.

Obstructed intracardiac baffles. Increasing use of the arterial switch operation and improved atrial techniques have made this complication of repaired transposition of the great arteries increasingly rare. When obstruction occurs, it appears to be at the level of the old interatrial septum. The use of very large balloons will enlarge the narrowed channel and decrease the gradient in almost every patient. Although late follow-up is not yet available, such patients can remain

symptom-free for 2 to 3 years. Thus, angioplasty should probably be considered as the initial treatment in such patients.

Other Catheter-Directed Procedures

In addition to balloon angioplasty catheter techniques, several other forms of catheter-directed therapy have become standard practice in the past few years.

Drainage of pericardial effusions. Accurate two-dimensional echocardiography has revolutionized the management of pericardial fluid in children (21). The localization and quantification of the fluid around the heart has made percutaneous drainage an extremely safe procedure even in patients with poor hemodynamic status, localized fluid or a bleeding diathesis. Once the correct needle course has been identified and a guide wire positioned in the pericardial space, large bore (8F) catheters can be introduced over the wire for long-term effective drainage. Even in diagnostic pericardiocentesis, the most prudent course might be to introduce a catheter and, once the nature of the fluid has been established, decide whether to remove the catheter or leave it in for long-term drainage. The only patients whose pericardial fluid cannot be drained percutaneously are those whose fluid is inaccessible from a precordial, subxiphoid or subcostal approach or those whose pericardium needs to be removed.

Obstruction of the interatrial septum. In patients with mitral atresia or stenosis and either an anatomic or a functional single ventricle, wide patency of the interatrial septum is needed to allow unobstructed pulmonary venous drainage. Although surgical atrial septostomy or septectomy has been standard in these children, the use of the Park blade catheter followed by balloon septostomy has considerable advantages; it has been proven to be safe, it can be repeated at low risk and its short-term efficacy has been good. Because the ultimate palliation for these children will be a Fontan type of operation, one needs to lower left atrial pressure to rather low levels (that is, <8 mm Hg). Once again, late follow-up is lacking for both surgical and catheter techniques to open the atrial septum but, at present, the transcatheter technique as the initial procedure appears to offer considerable advantages.

Occlusion of redundant systemic pulmonary collateral arteries. In patients with tetralogy of Fallot and pulmonary atresia, or similar lesions, blood supply to the lungs frequently occurs through systemic pulmonary collateral arteries (so-called bronchial arteries). These vessels frequently anastomose with true pulmonary arteries; after shunts into the pulmonary arteries, the lung may have a dual supply to the same capillary bed. At the time of complete repair, these (now redundant) systemic pulmonary collateral arteries need to be occluded: they will complicate intraoperative management by allowing pumped blood to reach the lungs during

cardiopulmonary bypass, and leave an unwanted left to right shunt in the postoperative period. Operative ligation of such vessels can be difficult, especially from an anterior approach at the time of complete repair.

The use of either detachable balloons or preformed steel coils to occlude such vessels allows precise definition of the vascular anatomy at the time of occlusion and can be performed with considerable safety and accuracy. Perhaps 75 or 85% of collateral arteries have a long and straight enough course to allow transcatheter closure.

Procedures Currently Considered Experimental

Aortic valve stenosis. Surgical aortic valvotomy, outside of the newborn period, is a relatively safe procedure in pediatric patients, mortality rates are 1 to 2%, and fewer than 5% of such patients require valve replacement in the perioperative period because of iatrogenic aortic regurgitation. However, the procedure is clearly not curative, pressure gradient reductions are only 50 to 70%, restenosis is not uncommon and there is a distressingly high incidence of deaths in the decades after surgery (22). In that context, balloon angioplasty, albeit imperfect, offers several advantages: it appears to provide roughly the same degree of pressure gradient reduction (at least over the short term) and can be repeated a number of times should valve restenosis occur. Because most of the patients develop at least some degree of valvular regurgitation (usually mild) in the post-dilation period, and because the impact of balloon valvuloplasty on late gradients and late mortality remains unclear, it must still be viewed at this time as an unproved form of therapy.

Patent ductus arteriosus. The surgical closure of the patent ductus arteriosus offers superb results at a very small mortality or morbidity rate. Using the double umbrella device developed by Rashkind (12), there has been a promising start in several centers for transcatheter closure (success rates over 70% with no mortality and minimal morbidity). In patients who are at increased risk for thoracotomy for any of a number of reasons, transcatheter patent ductus arteriosus closure may well be indicated. However, because many patent ductus are closed to prevent bacterial arteritis, and because the impact of transcatheter closure on the late incidence of endocarditis is unknown, it will be some years before the ultimate role of this procedure becomes clear.

In our opinion, this device may already be very useful to close large nonpatent ductus arteriosus openings elsewhere in the vascular tree. In many such cases (very large systemic pulmonary collateral vessels, persistent left superior vena cava to the left atrium and others) the surgical risks may be considerably higher than those for a simple patent ductus arteriosus ligation, thus making transcatheter closure a more attractive option.

Unoperated (native) aortic coarctations. Studies have now shown (23) that, after infancy, the native aortic coarctation can be dilated with very good short-term gradient reduction and minimal morbidity. However, both animal and now some clinical trials (24) have demonstrated a significant incidence of "aneurysm" formation, 6 to 12 months after the procedure, in the area of the old coarctation. Although none of these lesions have produced symptoms, their angiographic appearance has prompted prophylactic excision of the affected area. At present, not all groups engaged in angioplasty of native coarctations have observed late aneurysms, and it is possible that some (as yet unidentified) variations in catheter technique or aortic arch anatomy may predispose to aneurysm formation. If so, such aneurysms may be avoidable. Given the good results of surgery for coarctation, this procedure must be viewed as experimental until more late follow-up data become available.

Even with the potential risks of late aneurysm formation, there are occasional clinical circumstances in which standard operative management becomes a high risk procedure because of severe associated cardiac or noncardiac defects. In such cases, angioplasty can prove invaluable as an initial therapy to allow normalization of blood pressure and reduction of ventricular afterload.

Conclusions

Transcatheter therapy has substantively altered cardiac care for children in the past 5 years. Although no firm conclusions can be reached until more extensive follow-up is available, it appears that a number of procedures have already replaced operative management. Even the procedures we currently consider experimental will undoubtedly play some role in the management of congenital cardiac disease.

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