cations may help to demonstrate the presence of a unilateral intrinsic carotid bruit in these two situations. One other potential source of error is that as the arterial stenosis increases toward total occlusion, the intensity of the bruit decreases. It may become of lower intensity than the radiated murmur, or so faint that its signal-to-noise ratio is too low for spectral analysis. In our experience, this occurs very rarely.

In summary, recording of murmurs and bruits over the base of the heart and along the course of the carotid arteries, combined with spectral analysis, usually permits differentiation of intrinsic carotid artery bruits from those due to radiated basal heart murmurs and allows quantitation of the extent of carotid stenosis even in the presence of radiated murmurs.

References


Long-term Mitral Valve Replacement in Young Children
Influence of Somatic Growth on Prosthetic Valve Adequacy

Sidney Friedman, M.D., L. Henry Edmunds, Jr., M.D., and Charles C. Cuaso, M.D.

SUMMARY Long-term clinical and laboratory findings in three children who required mitral valve replacement below age four years are reported. In each instance a second valve replacement was necessary approximately 8 1/2 years after the initial one, following a two and one-half fold increase in body weight. Inadequate mitral valve orifice size was found in each instance, producing a hemodynamic picture equivalent to mitral stenosis: congestive heart failure, pulmonary hypertension and atrial fibrillation. A second valve was placed without mortality in each instance and relieved the mitral valve obstruction. Pulmonary resistance increased postoperatively in two patients and failed to decrease in the third. Pulmonary arterial hypertension and left ventricular hypertrophy persisted as long as 13 to 37 months after the second valve placement in all patients. The consequences of increasing body size and the long-term interposition of a rigid prosthesis in a growing heart introduce additional complications to mitral valve replacement in childhood. Frequent hemodynamic observations and the use of a prosthesis other than the ball-cage variety is recommended for improved management.

SEVERE MITRAL VALVE DEFORMITY producing intractable heart failure is an indication for prosthetic valve replacement even in very young patients. The clinical and hemodynamic improvements which follow valve replacement are often dramatic and the long-term hazards of this procedure have been steadily decreasing as improved valve forms and surfaces have been developed.1-3 Well recognized complications of prosthetic valve use are thrombembolism, bleeding related to anticoagulation, bacterial endocarditis, cardiac arrhythmias and prosthesis breakdown; these occur with approximately equal frequency in adults and children.4 In the pediatric age group, an additional consideration is the effect of body growth on the long-term functional adequacy of a prosthetic device, and the in situ effect of the valve prosthesis on growing myocardium.

The purpose of this report is to describe the clinical, hemodynamic and surgical observations in three children who were treated by mitral valve (Starr-Edwards) replacement under the age of four years and who were subsequently followed for periods of 7 1/2, 9 and 9 1/2 years prior to reoperation. During these intervals, each child showed a near normal somatic growth rate and was relatively free of cardiac symptoms. Eventually, however, all three developed serious cardiac problems which necessitated a second, larger mitral valve prosthesis.

From the Divisions of Cardiology and Cardiothoracic Surgery, The Children's Hospital of Philadelphia; and the Departments of Pediatrics and Surgery, School of Medicine, University of Pennsylvania, Philadelphia, Pennsylvania.


Received November 16, 1977; revision accepted December 28, 1977.
Case 1

R. S., a white male, born November 10, 1960, was recognized to have a serious variety of acyanotic congenital heart disease in early infancy. In July 1963, because of intractable heart failure, resection of coarctation of the aorta was performed. This surgery failed to produce adequate relief of heart failure and in July 1964, at a body weight of 13.4 kg, a parachute mitral valve was replaced by a #0 Starr-Edwards mitral valve prosthesis. Cardiac catheterization performed prior to valve replacement revealed a pulmonary artery pressure of 50/30 mm Hg (mean 41). Following open heart surgery, heart failure disappeared. In January 1970 recurrent coarctation was resected without event. An aortic insufficiency murmur was noted at this time. Three months later, a transient, mild cerebrovascular accident occurred. Thereafter the patient was maintained on Warfarin sodium and has shown no further thromboembolic phenomena.

In February 1974, at the age of 13½ years, cardiac status deteriorated abruptly over a period of 10 days with symptoms suggesting severe mitral stenosis. Cardiac catheterization was performed and revealed a severe elevation of pulmonary artery pressure to systemic levels, 85/46 mm Hg (mean 60). Pulmonary vascular resistance was calculated to be 8.9 units. Mean diastolic gradient at the mitral valve was 27 mm Hg. The patient developed acute pulmonary edema during catheterization. On the following day, the #0 Starr-Edwards prosthesis was excised and replaced with a #29 Bjork-Shiley prosthesis. Fibrous tissue had overgrown each of the four struts and attached the cage to the endocardium of the left ventricle (fig. 1). The valve anulus was enlarged by excising a rim of fibrous tissue anteriorly beneath the aortic valve and along the ventricular septum. The new valve was sutured to the fibrous anulus of the excised valve; it was not recognized that the posterior rim of this anulus was not at the arioventricular junction.

In September 1974, signs and symptoms of heart failure gradually reappeared. Restudy demonstrated the presence of a small paravalvular leak, mild aortic insufficiency and that the Bjork-Shiley valve was not located at the true mitral valve anulus. The mitral diastolic pressure gradient was only 5 mm Hg but left ventricular end-diastolic pressure gradient had increased markedly to 23 mm Hg. A portion of the left atrium was situated beneath the level of the prosthetic valve and showed paradoxical wall movement.

In January 1975 restudy revealed severe mitral insufficiency and increased evidence of aortic insufficiency. Pulmonary artery pressure had increased (90/57 mm Hg, mean 69). Left ventricular ejection fraction was 36%. End-diastolic pressure in the left ventricle was again 23 mm Hg.

In March 1975, the Bjork-Shiley mitral valve was replaced with a new, slightly smaller Bjork-Shiley (#27) valve. The posterior rim of the anulus at the true arioventricular junction was located and sewn to the new prosthesis. The aortic valve was also replaced because of a 4 mm perforation in the noncoronary cusp; a #21 Bjork-Shiley prosthesis was implanted. The tricuspid valve was inspected; mattress sutures were placed in each commissure of the dilated tricuspid anulus to correct tricuspid insufficiency.

Four days following operation, the patient was free of any evidence of valvular dysfunction and was making a satisfactory recovery. On the fifth postoperative day, the murmur of tricuspid insufficiency appeared together with evidence of right heart failure. This has persisted as has pulmonary hypertension, and a severe degree of biventricular myocardial dysfunction.

Repeat cardiac catheterization performed two years following the double valve replacement again showed moderate pulmonary hypertension. The pulmonary artery pressure was 65/30 as compared to an aortic pressure of 101/69. Mean pulmonary artery wedge pressure was 24 mm Hg and end-diastolic pressure in the left ventricle averaged 24 mm Hg. No diastolic pressure gradient was found at the mitral prosthesis. The calculated pulmonary vascular resistance was 11.3 units; right ventricular end-diastolic pressure was 17 mm Hg.

Case 2

B. L., a white female born September 5, 1962, was seen initially at the Children’s Hospital of Philadelphia in May 1966 at the age of three years with severe heart failure, refractory to medical management. Cardiac catheterization demonstrated the presence of severe mitral insufficiency and a pulmonary artery pressure of 56/32 mm Hg (mean 40). On July 1, 1966, at age three years and ten months and at a body weight of 12.2 kg, the mitral valve was explored and found to have a parachute mitral valve deformity which was treated by replacement with a #0 Starr-Edwards ball-valve prosthesis. The immediate postoperative course was stormy.
and mechanical ventilation was required for a period of three weeks. Gradual improvement followed and was attended by a decrease in heart size and a disappearance of the signs and symptoms of heart failure.

At five years of age, while not receiving anticoagulant medication, the patient developed a left hemiparesis. Subsequently, she has made an excellent recovery of neuromuscular function and has been taking Warfarin sodium without further thromboembolic complications. Bronchiectasis involving the left lower lung field was recognized in this period.

At age twelve years, a slight decrease in exercise tolerance was reported. Cardiac catheterization revealed the presence of moderate pulmonary hypertension (65/30 mm Hg, mean 40) and a significant diastolic gradient at the level of the mitral valve (mean 22 mm Hg). Left ventricular end-diastolic pressure was 5.5 mm Hg. On November 12, 1975, at age thirteen, the original Starr-Edwards valve was replaced by a larger (#2M) of the same type. Left ventricular muscle was hypertrophied and abutted against the cage struts; no fibrous tissue had grown onto the cage. Postoperatively she required catecholamine support for 36 hours and discharge was delayed to the fourteenth postoperative day by fever due to bronchiectasis. Cardiac catheterization performed six months postoperatively demonstrated partial relief of the mitral diastolic gradient (7 mm Hg), a marked reduction in left atrial size, but no change in pulmonary artery pressure (57/27 mm Hg, mean 40). Pulmonary vascular resistance measured 7.4 units. Left ventricular end-diastolic pressure had increased slightly to 5–11 mm Hg. Exercise tolerance returned to normal; bronchiectasis has been controlled by conservative measures.

Nineteen months postoperatively the patient was recatheterized. The pulmonary artery pressure remained elevated at 60/25 mm Hg (mean 40) as compared to an aortic pressure of 120/80 mm Hg (mean 95). Mean diastolic gradient across the mitral valve was 14 mm Hg and pulmonary vascular resistance was 4.3 units. Left atrial mean pressure and mean pulmonary artery wedge pressures were identical (22 mm Hg). Cineangiography demonstrated no insufficiency of the mitral valve prosthesis; some left atrial enlargement persisted. The left ventricle contracted normally, but left ventricular free-wall thickness was increased. Simultaneous echocardiographic study showed a left ventricular free wall thickness of 0.95 cm, a value slightly above the top normal value of 0.8 cm.

Case 3

S. B., a black male born December 4, 1966, was recognized to have mitral insufficiency at age five months. At eighteen months he presented with severe heart failure, refractory to medical management. Cardiac catheterization confirmed the presence of severe mitral insufficiency with a pulmonary artery pressure of 45–50/22 mm Hg. Surgical exploration of the mitral valve was performed at nineteen months at a body weight of 10.9 kg; a grossly deformed mitral valve was found. Intraoperatively, an attempt was made to carry out a mitral valvuloplasty but this was functionally unsuccessful and a #00 Starr-Edwards mitral valve prosthesis was inserted. The histopathologic diagnosis of the excised valve tissue was endocardial sclerosis. Relief of heart failure was prompt, but little reduction in the heart size occurred and several severe episodes of upper and lower respiratory tract infection followed. At age twenty-six months, repeat cardiac catheterization revealed a large left atrium which emptied slowly; there was angiographic evidence of residual mitral regurgitation. Over the next fifteen months, the patient gradually improved.

At age seven years, exercise intolerance was reported. Cardiac catheterization was repeated at age eight years and revealed moderate pulmonary hypertension (55–65/22 mm Hg, mean 37), a mean diastolic gradient at the prosthetic mitral valve of 17 mm Hg, and a moderate paravalvular leak. Left ventricular end-diastolic pressure was 7 mm Hg. At nine years of age the patient suddenly developed atrial fibrillation which was accompanied by signs and symptoms of heart failure. The arrhythmia was controlled by digoxin and quinidine sulphate, but the appearance of the cardiac arrhythmia and heart failure prompted replacement of the original Starr-Edwards prosthesis on February 17, 1976. No fibrous tissue was attached to the excised cage. Anular scar tissue was aggressively excised and the incision was carried down to the ventricular muscle along the posterior rim of the anulus near the posterior commissure. A #25 Hancock porcine heterograft was inserted into the enlarged anulus. Convalescence was uneventful and the child was discharged home on the tenth postoperative day without anticoagulant medication.

Approximately four months postoperatively cardiac catheterization demonstrated moderate residual pulmonary hypertension (50/20 mm Hg, mean 35) and a mean diastolic gradient of 5 mm across the porcine heterograft. There was no evidence of mitral insufficiency. The clinical state of the patient improved remarkably with a dramatic reduction in heart size (fig. 2). The cardiac arrhythmia has not recurred despite the discontinuation of antiarrhythmic medications.

Thirteen months after insertion of the porcine heterograft, cardiac catheterization showed no change in pulmonary artery pressure (55/25 mm Hg, mean 35). Pulmonary vascular resistance was 5.2 units. Mean diastolic gradient across the Hancock valve was 10 mm Hg. Left ventricular end-diastolic pressure was 10 mm Hg. Cineangiography showed no evidence of prosthetic mitral valve insufficiency; the left ventricular free wall appeared to be thickened. Echocardiographic measurements made at the same time showed a left ventricular free-wall thickness of 1.1 cm as compared to the normal of 0.7 cm. The left ventricular chamber was also enlarged, measuring 5 cm (upper limit of normal, 4.6 cm).

Discussion

The three children varied in age from 1½ to 3½ years at the time of first valve replacement; in each instance the indication was intractable heart failure due to severe mitral insufficiency, with or without mitral stenosis. In two children the pathology was a parachute mitral valve, and in one of these the additional lesions of the Shone Complex (R.S.) were present. In the third patient, the histopathologic findings of endocardial sclerosis were found, offering no opportunity for plastic repair. In the preoperative catheterization studies, the pulmonary artery pressures were 50/30, 56/32 and 50/22 mm Hg, representing approximately 50% of
systemic pressure levels. The prosthetic valves used for the initial replacement were Starr-Edwards valves of #00 and #0 size (1.27 and 1.54 cm²). The clinical improvement in each child was prompt, dramatic and maintained over a period of 7½ to 9½ years.

The anthropometric weight curves of the three children are depicted in figure 3; two follow the lower 3rd and 10th percentile curves while the third follows the 90th percentile. In no instance was there a deviation from the weight percentile pathway despite the presence of a prosthetic mitral valve. Mean body weight of the three children increased from 12.2 kg prior to the initial valve replacement to 29.5 kg at the time of the second replacement. Body weight had increased nearly ½ times during the interval that the first valve was in place, thereby providing a crude measure of the change in body weight which may be associated with symptomatic or hemodynamic evidence of inadequate valve size.

In earlier considerations of the effect of somatic growth on the adequacy of prosthetic valves inserted in small children, most predictions have suggested that the small Starr-Edwards valve (#0, #00) would accommodate the normal cardiac output of older children or adults. This optimistic attitude was supported by calculations of the valve diameters and by the fact that most children requiring prosthetic valves have a marked degree of cardiac enlargement and anular dilatation. The observations in the three children refute these projections since somatic growth did result in a valve orifice of inadequate size. Two pathologic mechanisms were operative: first, failure of the fixed diameter of the originally inserted valve prosthesis to accommodate the augmented cardiac output associated with increased body size, and second, the development of paravalvular fibrous tissue overgrowth which occurred in the patient with Shone Complex (R.S.). In each instance, the cardiac difficulties which resulted from inadequacy of the mitral valve orifice were the equivalent of severe mitral stenosis; congestive heart failure, pulmonary hypertension, and a cardiac arrhythmia were the major manifestations.

Introduction of a second prosthesis with a larger orifice area did relieve the mean mitral valve diastolic gradient in each instance (fig. 4). However, an unanticipated sequel to
mitral valve replacement in the growing child was the failure of the pulmonary artery pressure to return to normal after reoperation. This experience is contrary to extensive observations in adults in whom a prompt fall in pulmonary artery pressure and pulmonary vascular resistance follows relief of mitral stenosis and insufficiency by prosthetic valve replacement. Moderate pulmonary hypertension was present before and after the second valve replacement. The persistent pulmonary hypertension was associated with an increased pulmonary vascular resistance; the latter did not decrease after the second valve but actually increased in two of the three children (fig. 5). This persistent elevation of pulmonary vascular resistance is unexplained. Perhaps the elevated pulmonary vascular resistance represents a persistence of the fetal pattern of pulmonary vasculature resulting from the presence of chronically elevated pulmonary venous pressure due to the congenital mitral valve deformity. Another theoretical possibility is that in growing children as compared to adults, pulmonary venous hypertension may more readily cause irreversible pulmonary vascular changes.

A third unfavorable consequence of long-term mitral valve replacement was the persistence of left ventricular wall hypertrophy long after mitral insufficiency had been corrected by valve replacement. By direct inspection at the time of surgical exploration, cineangiographic observations and by echocardiographic measurements, all three patients retained evidence of increased left ventricular wall thickness. The electrocardiograms, however, did not show the criteria of left ventricular hypertrophy. In each instance multiple potential causes for left ventricular hypertrophy were present, including long standing valvular insufficiency. In addition, it is possible that the long-term physical presence of the bulky caged-ball prosthesis contributed to the left ventricular myocardial alterations in these small subjects, perhaps by the mechanism of obstructing the left ventricular outflow tract. The change from the original Starr-Edwards valve to a disc-type prosthesis or a porcine heterograft did not result in a reduction of the left ventricular wall thickness over an 18-24 month interval.

The experiences with the three children can be used to make several recommendations concerning postoperative management of children who require mitral valve replacement. Two of the three patients developed thromboembolic complications while not receiving anticoagulant prophylaxis; this hazard is a real one and is not age-related. Despite the technical difficulties involved, small children with caged-ball or discoid prosthetic valves should be maintained on anticoagulant prophylaxis, using the same criteria of management as in adults. In the single patient who suffered no thromboembolic complications a Hancock porcine prosthesis was inserted as a second valve replacement and no anticoagulant medication has been used to date.

Frequent hemodynamic studies are needed to evaluate the size and adequacy of the mitral valve orifice as well as the pulmonary artery pressure and pulmonary vascular resistance. The timing of such studies should be influenced by the size and age of the patient, particularly the percentage of adult size achieved. Hemodynamic studies should be carried out after the patient’s weight has doubled from the time of the first valve replacement. Thereafter, it would seem wise, especially with the availability of the percutaneous technique, to perform catheterization studies at two or three year intervals in small children for the purpose of making serial measurements of the pulmonary artery and right ventricular pressures, mitral valve gradients, pulmonary artery wedge pressure and left atrial pressure.

The need for accurate hemodynamic information was emphasized by the lack of usefulness of the physical examination in predicting the pulmonary pressure. On auscultation in the second left interspace, two widely spaced heart sounds were regularly audible at the end of systole. In retrospect, the second component, which was never grossly accentuated, was probably not a pulmonary valve closure sound but rather a sound generated by the prosthesis itself. Similarly, a review of the serial X-ray films failed to demonstrate any clear-cut evidence of pulmonary venous obstruction, despite the subsequent measurement of high left atrial and pulmonary venous pressures. The electrocardiograms did indicate the development of left atrial enlargement and progressively increasing right ventricular hypertrophy, as manifested mainly by increasing depth of the S waves in the left chest leads.

A second valve replacement can be carried out with low mortality risk in childhood and should be performed prior to
the development of the cardiac and pulmonary vascular complications which were observed in these three children. Nine to thirteen years ago when the original mitral valve prostheses were introduced, only the Starr-Edwards ball-cage valve was available. Since that time a number of other artificial valves have been offered for clinical use. In view of the development of left ventricular wall thickening in all three cases and the extension of fibrous tissue onto the valve struts in one, it would seem wise to avoid the use of prosthetic valves which may partially occlude the left ventricular outflow tract in small children. The glutaraldehyde-preserved porcine heterograft currently possesses the greatest number of advantages for use in this age group because of its central flow characteristics, favorable effective orifice to anular diameter ratio, low thromboembolic rate without anticoagulants, and proven durability of five or more years.

References


The Murmur of Pulmonic Regurgitation in Tetralogy of Fallot with Absent Pulmonic Valve

MARY E. FONTANA, M.D. AND CHARLES F. WOOLEY, M.D.

SUMMARY Absent pulmonic valve (APV) in tetralogy of Fallot produces a pulmonic regurgitation murmur (PRM) which is usually late in onset after A2, low pitched, and of crescendo-decrescendo character. We have seen three adult patients with tetralogy of Fallot with APV and have done intracardiac sound and pressure studies in two. The PRM was loudest in the RV outflow tract (RVOT), where the onset was earlier than the murmur recorded on the chest wall. The crescendo portion of the PRM occurred during an abnormally slow decline in the RVOT pressure pulse after the crossover of PA and RVOT pressures. The RVOT pressure reached its minimum 30 msec after the RV body pressure, resulting in a pressure gradient between the two. The PRM peaked 30 msec later in the RV body than in the RVOT. The delayed precordial onset of the PRM after A2 is likely due to failure of transmission of early vibrations through the chest wall. The morphology of the PRM in tetralogy of Fallot with APV may be related to delayed relaxation with altered diastolic compliance of the RVOT which is subjected to a large regurgitant volume from the massively dilated pulmonary arteries.

TETRALOGY OF FALLOT with absent pulmonic valve is an entity which can be differentiated from the classical tetralogy of Fallot. Distinctive features are those of right ventricular dilatation as well as hypertrophy, obstruction usually at a small pulmonic valve ring with absent or mild infundibular obstruction, absent pulmonic valve cusps, and aneurysmal proximal pulmonary arteries. On clinical examination the harsh systolic ejection murmur along the left sternal border and absent pulmonic second sound generally occur in classic tetralogy, but the low pitched, crescendo-decrescendo murmur of pulmonic regurgitation heard along the left sternal border is not heard in unoperated tetralogy of Fallot.

The determinants of the characteristics of the pulmonic regurgitation murmur in tetralogy of Fallot with absent pulmonic valve have not been documented.

Three adults in whom the diagnosis was established by operation and/or autopsy had complete clinical, phonocardiographic, and catheterization studies. In addition, two had intracardiac sound and pressure studies performed to define the mechanism of production of the murmur.

Materials and Methods

Diagnostic cardiac catheterization was performed with standard fluid-filled catheter systems, using hydrogen
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prosthetic valve adequacy.
S Friedman, L H Edmunds, Jr and C C Cuaso

Circulation. 1978;57:981-986
doi: 10.1161/01.CIR.57.5.981
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1978 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

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