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# Congenital Aortic Regurgitation: Natural History and Management

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***Objectives and Background.*** **Congenital aortic regurgitation is rare as an isolated lesion. We describe seven children with no physical features of the Marfan syndrome in the patients or their families and no other cardiac lesions who had congenital valvular aortic regurgitation.**

***Methods.*** **From 1954 to the present, seven children with auscultatory and physiologic characteristics of aortic regurgitation were evaluated for a total of 108 patient-years. We report on their natural history, clinical and laboratory findings, management and outcome.**

***Results.*** **In five of the seven children congenital aortic regurgitation was diagnosed in infancy. In four, progressive severity of the regurgitation led to valve replacement at age 3, 10, 15 and 20 years, respectively, and to resection of an aneurysm of the ascending aorta in the 10-year old patient. Two patients had cystic medial necrosis on aortic biopsy. One of these patients died after reoperation for dissecting aneurysm of the thoracic aorta at 22 years of age; the other died after dissection and rupture of the ascending aorta at age 25 years. After obstructing pannus developed, the 3-year old patient underwent replacement of the St. Jude valve at age 10 years. The other three patients were asymptomatic at last follow-up at age 8, 10 and 20 years, respectively.**

***Conclusions.*** **Supportive management is recommended until it becomes necessary to intervene surgically when regurgitation becomes severe. The need for surgical treatment is indicated by the appearance of a diastolic thrill, left ventricular strain on the electrocardiogram or other evidence of left ventricular dysfunction on the echocardiogram or exercise stress testing by treadmill or radionuclide cineangiocardiography. Close follow-up of these patients is important to detect progression of aortic regurgitation, especially in the presence of cystic medial necrosis.**

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Congenital insufficiency of the aortic valve is rare as an isolated lesion. It is more often associated with the stenotic bicuspid aortic valve, with discrete subaortic stenosis, as a result of prolapse of the aortic valve with a ventricular septal defect or as a component of the Marfan syndrome (1,2). Aortic-left ventricular tunnel is a cause of congenital aortic regurgitation not due to a valvular lesion but to an abnormal connection between the aorta and left ventricle. We report on seven children with no physical features of the Marfan syndrome in the patients or their families and no other intracardiac lesions who had congenital valvular aortic regurgitation.

## Methods

**Study patients.** From 1954 to the present we evaluated and kept under cardiac surveillance seven children (four male, three female) with the auscultatory and physiologic characteristics of aortic regurgitation. The condition was detected at birth in two, by age 8 months in three and by age 3 and 5 years, respectively, in two (Table 1). They were under observation for a total of 108 patient-years. We report on their natural history, clinical and laboratory findings, management and outcome. During these years of observation, diagnostic and surgical techniques and the concepts of patient management were evolving.

**Natural history and clinical aspects.** Detection of a murmur on routine examination led to the diagnosis of congenital aortic regurgitation. Signs of left ventricular burden from volume overload and increasing loudness of the diastolic murmur and accompanying systolic murmur preceded symptoms in the four patients with progressively severe aortic regurgitation. The other three patients were asymptomatic at last examination at age 8, 10 and 20 years, respectively. The clinical highlights and the age when major events occurred are shown in Table 1. Brief summaries of each case follow.

## Case Reports

### *Case 1*

Moderately severe aortic regurgitation with dilation of the ascending aorta on the 1st day of life had become severe by

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**Table 1.** Clinical Data in Seven Patients With Congenital Aortic Regurgitation

| | Relation of Event to Patient Age (yr) | | | | | | |
|---|---|---|---|---|---|---|---|
| | Pt 1 (M) | Pt 2 (M) | Pt 3 (F) | Pt 4 (M) | Pt 5 (F) | Pt 6 (M) | Pt 7 (F) |
| Diagnosis | 1 day | 5 mo | 3 | 5 | 8 mo | 1 day | 2 mo |
| Symptoms | 2.5,10 | 13 | 8,22 | 20 | 0 | 0 | 0 |
| Cardiac failure | 2.5 | 0 | 8 | 0 | 0 | 0 | 0 |
| Aortography/cardiac cath | 2.5 | 0 | 3,8,11 | 20 | 4,9 | 4 | 4 |
| Valve replaced | 3 | 15 | 10 | 20 | 0 | 0 | 0 |
| Reoperation | 10 | 0 | 22 | 0 | 0 | 0 | 0 |
| Death | 0 | 0 | 22 | 25 | 0 | 0 | 0 |
| Age at last follow-up | 11 | 17 | 22 | 25 | 20 | 8 | 10 |
| Current NYHA class | I | I | — | — | I | I–II | I |

Cath = catheterization; F = female; M = male; NYHA class = New York Heart Association functional class; Pt = patient.

age 2 years, leading to cardiac failure and replacement of the aortic valve with a St. Jude mechanical valve at age 3 years. Pannus formation beneath the valve caused obstruction and led to replacement with a larger valve at age 10 years. Although the aorta had become enormously dilated, sections did not show cystic medial necrosis.

**Clinical features.** At birth this male infant had a soft systolic and prolonged diastolic murmur in the second right interspace and enlargement of the left ventricle and ascending aorta on the chest X-ray film. The electrocardiogram (ECG) showed left axis deviation and left ventricular hypertrophy. At age 3 months the two-dimensional echocardiogram showed left ventricular hypertrophy, thickened aortic valve leaflets and a dilated ascending aorta. By age 7 months the diastolic murmur was accompanied by a thrill. At age 2.5 years the ECG showed T wave changes of left ventricular "strain." The echocardiogram revealed more marked dilation of the ascending aorta as well as diastolic flutter of the anterior leaflet of the mitral valve and septum. The cardiothoracic ratio increased to 59%. Aortography confirmed severe aortic regurgitation and dilation of the ascending aorta and left ventricle (Fig. 1). Nine months later, the child began to tire on exertion and developed cardiac failure that responded to digoxin and diuretic drugs. The cardiothoracic ratio was 71%.

**Surgical findings.** Operation was performed through the dilated aorta. The aortic valve had four leaflets because of an accessory leaflet between the left and right coronary cusps. The commissural attachments and the orifice of the right coronary artery were superiorly displaced. The thickened and rolled leaflets did not coapt. An oval hole was present in the right coronary cusp. The valve was replaced with a 19-mm St. Jude's mechanical valve. On microscopic examination, focal calcification was seen in the valve. There was no evidence of cystic medial necrosis on biopsy of the ascending aorta.

**Follow-up.** Medication with warfarin was begun and continued and the patient became symptom-free. The cardiothoracic ratio decreased to 54% but two-dimensional echocardiography showed that the size of the aorta was increasing to

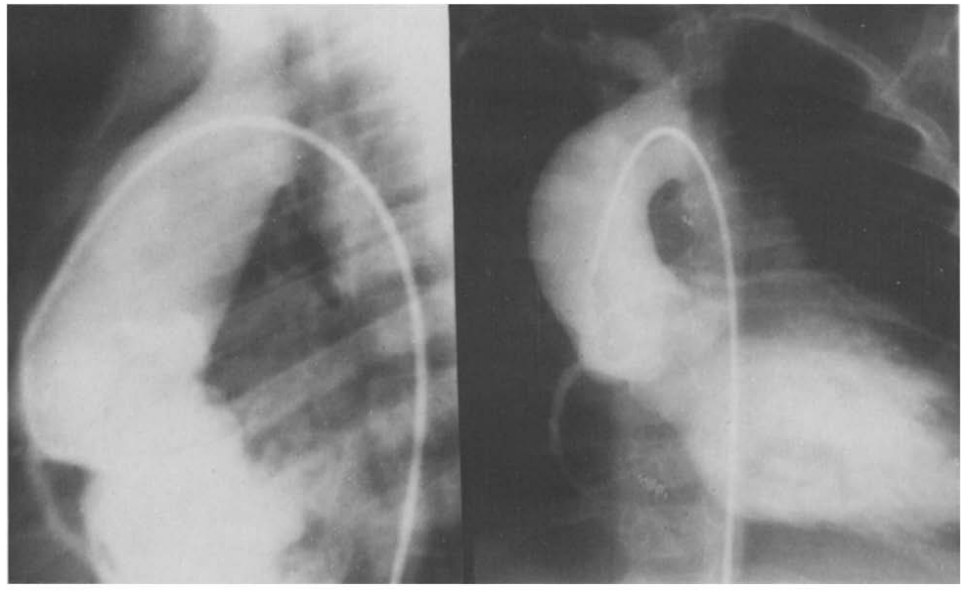

**Figure 1.** Case 1. Selective injection of contrast medium into the root of the aorta in lateral (**left**) and simultaneous frontal (**right**) views shows dilation of the ascending aorta and prompt reflux of contrast medium that fills the dilated left ventricle. Coronary arteries arise and course normally.

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aneurysmal proportions. Doppler studies revealed a steady increase in peak instantaneous systolic pressure gradient from 20 to 49 mm Hg across the valve. Cardiac catheterization confirmed the obstruction of or around the aortic valve. Left ventricular pressure was 173/20 mm Hg and simultaneous aortic pressure was 120/90 mm Hg. The ascending aorta was aneurysmally dilated.

*Reoperation* disclosed a ring of fibrous tissue beneath the valve that obstructed the subvalvular area but did not involve the valve itself. This tissue and the prosthetic valve were removed and a 21-mm St. Jude valve was placed. On microscopic examination, the pannus consisted of dense fibroconnective tissue with scattered lymphocytes and a small aggregate of acute inflammatory cells. Stains for bacteria and fungi and cultures were negative. Biopsy of the ascending aorta again failed to show cystic medial necrosis. He is once more symptom free and taking warfarin.

## *Case 2*

Mild aortic regurgitation at age 5 months progressed slowly for 15 years. This male patient is now aged 17 years, asymptomatic and on warfarin therapy after undergoing valve replacement at age 15 years. The resected aortic valve and biopsy of the ascending aorta showed myxoid change.

**Clinical features.** At age 4.5 years the ECG showed left ventricular hypertrophy. The chest X-ray film showed left ventricular enlargement; the diastolic murmur had increased to grade 4/6 in intensity. A two-dimensional echocardiogram showed a trileaflet aortic valve with eccentric closure and no evidence of a subaortic shelf. At age 7 years the study showed flutter of the anterior mitral leaflet and at 10 years, extension of the regurgitant jet midway into the left ventricle. At age 13 years the aortic valve prolapsed into the left ventricle. At that time, leads $V_5$ and $V_6$ on the ECG showed ST segment depression and inverted T waves of left ventricular "strain." Nonetheless, the patient reported no symptoms.

*A treadmill exercise test* revealed a normal endurance time but persistent ST segment depression and ventricular premature beats that increased during early recovery. The pulse pressure was 100 mm Hg at peak exercise when the pulse rate was 174/min. Radionuclide cineangiography during supine bicycle exercise showed that the left ventricular ejection fraction increased from 59% at rest to 71% during exercise. We interpreted this as evidence of preservation of left ventricular function.

**Surgical findings.** Operation showed a moderately dilated ascending aorta and a redundant thickened trileaflet aortic valve that prolapsed into the left ventricle. A 23-mm St. Jude mechanical valve replaced the aortic valve. Biopsy of the aorta and the aortic valve showed slight to moderate myxoid change but no cystic medial necrosis.

**Follow-up.** Postoperatively, the patient has been maintained on warfarin therapy. Electrocardiography showed disappearance of left ventricular hypertrophy and "strain" and echocardiography showed normalization of left ventricular chamber size by 18 months.

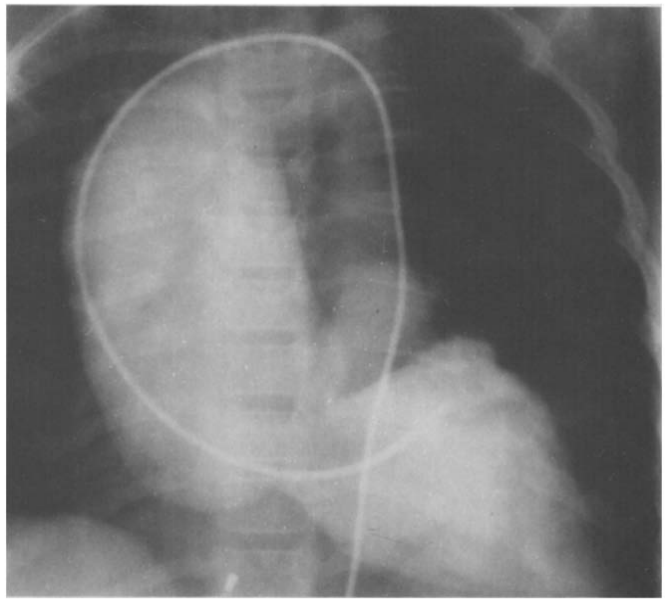

**Figure 2.** Case 3. On aortography, the ascending aorta is greatly dilated. Contrast medium and the catheter tip reflux into the dilated left ventricle.

## *Case 3*

In this female patient, mild aortic regurgitation became progressively more severe, causing cardiac failure at age 9 years and replacement of the aortic valve and ascending aorta at age 10 years. Dilation of the main pulmonary artery appeared 1 year later. At age 20 years aneurysmal dilation of the entire thoracic aorta with dissection led to death after surgery. The patient had cystic medial necrosis of the aorta; she may have had a mild form of the Marfan syndrome without skeletal or ocular abnormalities or a nonMarfan condition.

**Clinical features.** At 3 years of age, cardiac catheterization confirmed the diagnosis of mild aortic regurgitation and showed a markedly dilated ascending aorta. Gradually, the soft diastolic murmur increased to grade 4/6 intensity. At 8 years of age, she complained of frequent pounding occipital headaches and a tight feeling in her chest. During the next year, she developed increasing fatigue and signs of heart failure that improved after digitalis therapy. Cardiac catheterization showed normal right heart chambers, a dilated left ventricle and marked aortic regurgitation (Fig. 2). Three aortic cusps were identified; the right and posterior cusps were remarkably enlarged.

**Surgical findings.** The patient underwent operation in 1968 at age 10 years. The ascending aortic aneurysm measured 9 cm in diameter and 10 cm in length from the aortic anulus to the innominate artery. The anulus was dilated and the trileaflet valve was grossly incompetent. The cusps were

excised and a Starr-Edwards valve was placed. The aneurysm was removed, leaving a rim around the coronary ostia. The ascending aorta was reconstituted with a 2.5-cm segment of Dacron prosthesis 30 mm in diameter. The pathologic report on the aorta revealed cystic medial necrosis on both elastic and mucin stains. The aortic valve showed focal thickening.

**Follow-up.** One year postoperatively, the size of the left ventricle on the chest roentgenogram had decreased to normal, but the pulmonary artery was dilated, a new finding. Cardiac catheterization showed that the Starr-Edwards valve functioned well. Left ventricular systolic pressure was 138/9 mm Hg. On pullback of the catheter across the valve and proximal to the aortic anastomotic site, there was a 34-mm Hg peak systolic pressure gradient. The patient continued to do well with normal activity.

While in college at age 22 years she became hoarse and was found to have left vocal cord paralysis secondary to stretching of the recurrent laryngeal nerve from massive dilation of the aorta, now involving the entire thoracic aortic arch and left subclavian artery as well as the sinuses of Valsalva. She underwent operation to replace the aortic valve and aortic arch with a valve and composite graft. She died 2 h postoperatively. Autopsy revealed cystic medial necrosis and an acute dissecting aneurysm of the descending thoracic aorta with occlusion.

## *Case 4*

In this male infant, mild aortic valve disease with regurgitation was not noted until age 5 years. Slow progression of regurgitation led to valve replacement with a Starr-Edwards valve and trimming of aortic tissue at aortotomy closure at age 20 years. Biopsy revealed cystic medial necrosis. The patient died of ruptured dissecting aneurysm of the aorta 5 years later.

**Clinical features.** From the outset, the ECG showed left ventricular hypertrophy and the X-ray film showed an average heart size but a moderately dilated ascending aorta. At age 15 years a thrill accompanied the increasingly loud diastolic murmur and at age 18 he had elevated systolic and decreased diastolic (145/60 mm Hg) pressure. At age 20 years cardiac catheterization disclosed an end-diastolic pressure of 25 mm Hg in the dilated left ventricle. Ventricular function was not impaired. Aortography confirmed massive regurgitation and extreme dilation of the aorta.

**Surgical findings.** Operation in 1973 showed a trileaflet aortic valve with a small and partly calcified right coronary cusp and thickened left and noncoronary cusps. The valve was replaced with a Starr-Edwards valve and the dilated aorta made smaller by excision of 1 cm from each side of the aortotomy. Aortic biopsy disclosed cystic medial necrosis.

**Follow-up.** Postoperatively, the patient felt well and entered medical school but died suddenly in his 4th year at age 25 from rupture and dissection of the ascending aorta. Autopsy showed cystic medial necrosis of the aorta, coronary arteries and pulmonary artery. The fabric on the Starr-Edwards valve had completely disappeared.

## *Case 5*

The diagnosis of congenital aortic regurgitation was made at age 8 months in this female patient. In follow-up to age 20 years, she has remained symptom free and the regurgitation is stable.

**Clinical features and follow-up.** Cardiac catheterization at age 4 years established the diagnosis of isolated aortic regurgitation; repeat catheterization at age 9 years showed no change. The ECG studies have shown normal findings. Serial two-dimensional echocardiograms showed moderate aortic dilation and grade 2/3+ aortic regurgitation with multiple echoes of eccentric aortic valve closure. At age 14 years, the valve appeared thickened and at age 20 years, valve prolapse during diastole was evident. There was no evidence of left ventricular compromise. Results of treadmill exercise stress test (Bruce protocol) were normal. The patient continues to be asymptomatic, attends college and undergoes regular cardiac follow-up.

## *Case 6*

Congenital aortic regurgitation due to apparent absence of the noncoronary cusp was noted on the 1st day of this male infant's life. It caused no symptoms through the last follow-up examination at age 8 years.

**Clinical features.** At age 4 years, in 1960, at the time of aortography, the systolic and diastolic murmurs had increased in intensity at the right upper sternal border and a diastolic thrill was now present. The ECG fulfilled voltage criteria for left ventricular hypertrophy. The chest X-ray film showed moderate enlargement of the left ventricle and marked enlargement of the ascending aorta. Aortography confirmed the aortic dilation and showed immediate reflux into the enlarged left ventricle. The posterior noncoronary cusp of the aortic valve appeared to be absent. The patient was subsequently lost to follow-up.

## *Case 7*

In 1954 this 2-month-old female infant was the first of these patients to present with the murmur of aortic regurgitation. Her brother had a ventricular septal defect. She was still asymptomatic when lost to follow-up at age 10 years.

**Clinical features.** The ECG and chest X-ray film were unremarkable. In 1960, at age 5 years, aortography showed dilation of the sinuses of Valsalva but not of the ascending aorta and demonstrated moderate regurgitation through the posterior aspect of the aortic valve region, perhaps through an abnormal or rudimentary cusp (Fig. 3) (3).

## *Physical Findings*

A long diastolic murmur in the aortic area became louder and was accompanied by a thrill and an increasingly loud

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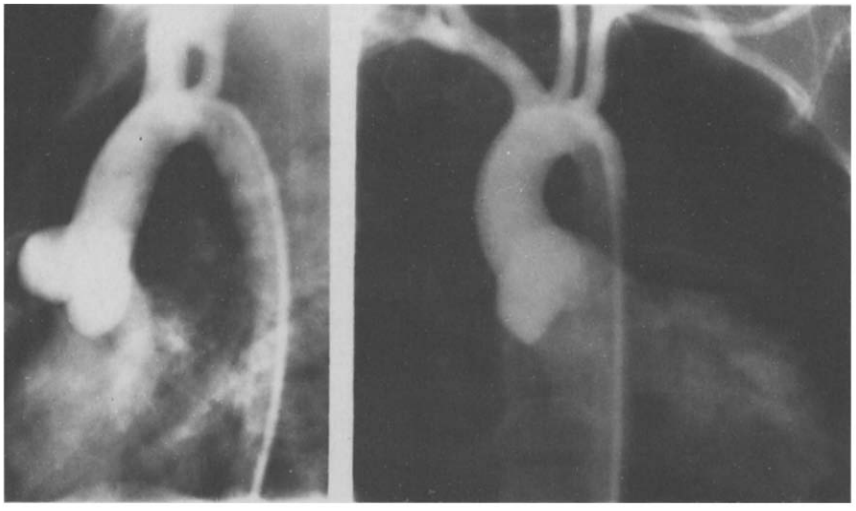

**Figure 3.** Case 7. Aortogram in the lateral projection (**left**) and simultaneous frontal view (**right**) shows dilation of the aortic sinuses of Valsalva but not of the ascending aorta. Contrast medium regurgitates through the posterior aspect of the aortic valve.

aortic systolic murmur as aortic regurgitation worsened in Cases 1 to 4. Pulse pressure widened and arterial pulses were bounding. Height and weight during the growing years were in the normal range.

## *Laboratory Findings*

**Radiologic abnormalities.** At diagnosis, these consisted of left ventricular enlargement with mild dilation of the ascending aorta in all but Patient 7. The dilation became aneurysmal in Patients 1, 3 and 4. Ventricular enlargement disappeared within months after valve replacement in Patients 1 to 3.

**Electrocardiograms.** These fulfilled voltage criteria for left ventricular hypertrophy in all but Patients 4 and 6. Left ventricular "strain" (inversion of T waves in leads $V_5$ and $V_6$) preceded symptoms in Patients 1 and 2 and abnormalities regressed after valve replacement.

**Cardiac catheterization and aortography.** Except in Patient 2, these procedures were undertaken to confirm the diagnosis, judge physiologic severity and exclude associated anomalies, especially discrete subaortic stenosis. Dilation of the aortic sinuses and ascending aorta (Fig. 1 and 2) was noted in all but Patient 7, in whom the dilation was limited to the aortic sinuses (Fig. 3). Because echocardiography provided sufficient information in Patient 2, the most recently operated on, catheterization was not undertaken.

**Two-dimensional echocardiography with Doppler studies.** These studies were performed in Patients 1, 2 and 5. The other patients were studied before the advent of this technique. Dilation of the aortic root progressed to aneurysmal proportions in Patient 1, >95th percentile for age for our laboratory (4). Flutter of the mitral valve and ventricular septum occurred in Patients 1 and 2, as well as prolapse of the aortic leaflets (Patients 1, 2 and 5). Although left ventricular chamber size and wall thickness were increased, left ventricular function was preserved at the time of operation. Postoperatively, left ventricular chamber size normalized. Doppler measurement of obstruction to left ventricular outflow by the prosthesis was helpful, especially in Patient 1, who developed pannus beneath the valve. Flow velocity (measured in m/s) was translated into peak instantaneous gradient for comparison with gradients obtained at catheterization. In Patient 2, the Doppler gradient increased steadily from 20 to 49 mm Hg, values that corresponded well with the gradient of 53 mm Hg determined at cardiac catheterization 1 week later. In this patient the peak instantaneous gradient was 25 mm Hg 1 year after valve replacement.

**Radionuclide cineangiocardiography.** This study was performed in Patient 2 to assess left ventricular function at rest and during supine bicycle exercise. Just before valve replacement, he had a near normal endurance time and a normal ejection fraction that increased with exercise.

## *Management and Outcome*

Management consisted of regularly scheduled cardiac follow-up to observe signs of increasing physiologic burden and early appearance of symptoms. In the four patients who underwent valve replacement, signs preceded symptoms of fatigability, pounding headache or cardiac failure.

*The surgical aspects are presented in Table 2.* The timing of operation was influenced by the risk of anticoagulant therapy in healthy active children versus the risk of irreversible damage to the left ventricle from long-standing severe aortic regurgitation. Fortunately, in each patient radiologic and ECG evidence of left ventricular dysfunction disappeared. We were also concerned that the size of the valve that could be placed in a young child might be too small for an adult. It came as a surprise that in Patient 1, operated on at age 3 years, the obstruction to left ventricular outflow that developed over the next 7 years occurred not because the patient "outgrew" the valve but because pannus developed below the valve.

*We considered a mechanical valve preferable to a tissue valve*, which might calcify and deteriorate in these young

**Table 2.** Surgical Data in Four Patients With Congenital Aortic Regurgitation

| | Patient 1 | Patient 2 | Patient 3 | Patient 4 |
|---|---|---|---|---|
| Ascending aorta | Aneurysm | Moderate dilation | Aneurysm | Aneurysm |
| Anatomy of valve | Quadricuspid | Trileaflet, redundant | Trileaflet, redundant | Trileaflet, with small right coronary cusp |
| Valve calcification | + | – | – | + |
| Cystic medial necrosis | – | – | + | + |
| Mechanical valve | St. Jude | St. Jude | Starr-Edwards | Starr-Edwards |
| Aortic surgery | – | – | Aneurysm removed; Dacron prosthesis | Aortotomy edges of aneurysm trimmed |

– = absent; + = present.

patients (5). The choice of mechanical valve changed as newer valves became available. In 1968 and 1973, the surgeon selected the Starr-Edwards valve. In Patient 3, he left intact the rim of aorta that gave rise to the coronary arteries, replacing the resected distal aneurysm with a Dacron prosthesis. Today, a composite graft of a mechanical valve with a Dacron prosthesis for replacement of the ascending aorta after implantation of the coronary arteries or a cryopreserved aortic homograft would probably be chosen (6).

*Anticoagulant therapy with warfarin* was continued only briefly for 2 months after placement of the Starr-Edwards valves, with no evidence of embolization at follow-up. The two boys with a St. Jude valve have been maintained on warfarin therapy.

## Discussion

Congenital aortic regurgitation as an isolated finding is so rare that a discussion of it cannot be found in most pediatric cardiology textbooks. In a review (7) of congenital heart disease from 1950 to 1973, investigators from The Hospital for Sick Children reported a 0.3% incidence rate (49 in 15,104 cases). Anderson (8) stated that the most common cause of congenital aortic regurgitation is the aortic-left ventricular tunnel, a defect that does not involve the aortic valve itself in most instances. Isolated valve defects are common after rheumatic fever and most reports relating to the diagnosis and treatment of aortic regurgitation come from patients in this group.

**Cystic medial necrosis and Marfan syndrome.** These seven children were normal in outward appearance, without signs of the Marfan syndrome (1,2) or evidence on study of other congenital cardiovascular anomalies. All had dilation of the aortic sinuses and all but one had dilation of the ascending aorta. In two of the four children with valve replacement, cystic medial necrosis was identified; both later developed dissecting aneurysm of the thoracic aorta. The microscopic lesion of focal separation of the elastic and fibromuscular elements of the tunica media by cystic-appearing spaces would not have been found without biopsy of the aorta. The lesion was not identified in the other two boys, even though Patient 2 had the most marked dilation of the aorta among all the children.

Although aortic regurgitation occurs in the Marfan syndrome (1,2), it can also be found in patients with non-Marfan disease. A report (9) on 93 such patients came from the Cleveland Clinic. The youngest patient was 26 years old. Ninety patients underwent operation and 34 died during follow-up, chiefly from cardiovascular causes. In another report (10), a family without clinical features of the Marfan syndrome had nine members over two generations with aortic dissecting aneurysm.

**Quadricuspid aortic valve.** This was present in Patient 1 and is indeed unusual (11–16). In 1923, Simonds (11) reviewed existing published studies and found only five cases of a quadricuspid aortic valve. In 1968, Robicsek et al. (12) identified nine published cases and reported a 10th case. A 1990 survey (13) reported that of 700 operations for pure aortic regurgitation, only 2 were for quadricuspid valve. The embryologic basis of this defect is unknown, but Simonds (11) theorized that an additional pad formed within the trunk of the aorta. Another possibility, suggested in a case described initially by Abbott (14), is that the supernumerary cusp, which is not well formed, is due to an inflammatory reaction.

**Pannus formation in valve prostheses.** In Patient 1, the late pannus formation under the first St. Jude valve is curious. The chronic and acute inflammatory cells in the fibrous tissue suggest an ongoing process in this child, who had experienced no recognized febrile illness. Pannus in itself is rare after left-sided prosthetic valve replacement. It is theorized (17) that the high flow system on the left side guards against fibrous tissue deposition and prevents pannus formation in most cases. In this patient, the pannus formed independent of the valve or suture line on the left side. Although the 19-mm valve should have provided adequate flow without resistance, perhaps because the patient needed valve replacement at the early age of 3 years, the increasing turbulence across the valve caused the pannus to form below and adhere to the left ventricular outflow tract wall despite adequate anticoagulant therapy.

**Indications for operation.** Indications for operation and the timing of valve replacement require individualized assessment based on regular examination. Echocardiography, exercise testing by treadmill or radionuclide cineangiocardiography and ECG studies should help in the regular noninvasive evaluation of left ventricular dysfunction and the severity of aortic regurgitation. In our patients, abnormalities on these tests preceded the onset of symptoms. We

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believe that laboratory evidence of left ventricular dysfunction should be the indication for operation.

Pediatric cardiologists and surgeons may have an understandable reluctance to replace an aortic valve in young growing children. Outgrowth of the valve with need for later replacement as well as problems inherent in maintaining optimal anticoagulation without hemorrhage in active children are considerations. Fortunately, the outcome after operation was favorable in that all patients had regression of radiographic left ventricular enlargement and two of these had echocardiographic confirmation as well. However, they acquired mild aortic stenosis secondary to the artificial valve. This progressed in the youngest child because of subvalvular pannus formation.

**Postoperative follow-up.** At operation, when the aorta is noted to be markedly dilated and the frozen section at biopsy indicates cystic medial necrosis, our experience suggests that use of a composite graft to replace the regurgitant valve and dilated ascending aorta may be indicated. Furthermore, because of the risk of progressive dilation, dissection and rupture of the aorta, these patients require close postoperative follow-up with current imaging techniques. These might include nuclear magnetic resonance imaging and other tests of left ventricular dysfunction, such as echocardiography, treadmill exercise testing and radionuclide cineangiocardiography. In the four surgically treated patients, left ventricular volume overload and dysfunction were reversed.

Close follow-up of patients with congenital aortic regurgitation is important, especially in those with cystic medial necrosis and the prospect of progressive aortic disease.

**Conclusions.** Congenital valvular aortic regurgitation in the absence of overt signs of the Marfan syndrome or other cardiac anomalies is a rare defect, recognized by the aortic diastolic murmur heard early in life. We recommend supportive management until it becomes necessary to intervene surgically when the regurgitation becomes severe. The need for surgical treatment is indicated by findings such as the appearance of a diastolic thrill, left ventricular "strain" on the ECG and evidence of left ventricular dysfunction on echocardiography or exercise testing.

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