The Surgical Treatment of Dissecting Aortic Aneurysm Due to Idiopathic Cystic Medial Necrosis in a Six-Year-Old Child*

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The clinical, hemodynamic and surgical findings of a six-year-old boy with an incomplete dissection of an aortic aneurysm and aortic valve regurgitation due to idiopathic cystic medial necrosis are reported. The aortic aneurysm was successfully excised while the aortic valve incompetence, despite annuloplasty, remained unchanged.

Aneurysm of the thoracic aorta usually results from syphilis, arteriosclerosis, trauma or from inherent structural defects of the aortic wall itself. In childhood, aneurysm of the aorta is very rare; in a few recently reported cases, the aneurysm was the cardiovascular manifestation of Marfan's syndrome. Microscopically, the process in the aortic wall begins usually at the annulus of the aortic valve and terminates proximally at the origin of the innominate artery.

Dissecting aneurysm of the aorta is most unusual in childhood. A review of the literature revealed only six children with dissecting aneurysm with Marfan's syndrome being the cause in only two.

The purpose of this communication is to report the clinical and surgical findings of a six-year-old boy with an incompletely dissected ascending aorta aneurysm and aortic valve regurgitation due to idiopathic cystic medial necrosis. The other stigmata of Marfan's syndrome were absent.

CASE REPORT

A six-year-old boy was hospitalized in December, 1966 for an upper respiratory tract infection. His previous history revealed that a heart murmur suggesting aortic valve disease was discovered on routine examination at the age of two years. His parents reported obscure diffuse chest pain, palpitations and occasional dyspnea on exertion accompanied by fatigue since the age of three. Syncope or nocturnal dyspnea were denied. He was the product of a normal pregnancy and delivery.

The physical examination revealed a well built, well developed, stocky boy; cyanosis and congestive heart failure were absent. The peripheral arterial pulses were bounding with a blood pressure of 100/60 mm Hg. Prominent pulsations of the carotid arteries were seen. The neck veins were not distended. The heart size was mildly enlarged with the cardiac impulse palpable at the 5th left intercostal space at the mid clavicular line. A systolic thrill was palpable at the second right intercostal space as well at the jugular notch. The heart sounds were clear; the second sound at the second right intercostal space was normal. The pulmonic closure sound was also normal. There was a grade 4/6 loud rough ejection systolic murmur maximal at the second right intercostal space, radiating toward the neck as well as along the left sternal border accompanied by

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SURGERY FOR DISSECTING AORTIC ANEURYSM

FIGURE 2. Moderate left ventricular enlargement.

Figure 2. Moderate left ventricular enlargement.

a median sternotomy. About one inch above the aortic valve annulus, there was an aneurysm of the aorta about the size of a large egg (Fig 4). The aneurysm bulged from the anterior aortic wall and projected to the left. A systolic thrill accompanied by strong pulsations were felt. Over the aortic root and the left ventricle a diastolic thrill was also palpable.

The patient was connected to the heart-lung machine in the usual fashion. After total bypass institution, the patient was cooled to 30° C. The ascending aorta above the aneurysm was cross-clamped. A longitudinal aortotomy was performed through the aneurysm. There was an incomplete dissection of the aorta beginning about the aortic annulus and terminating at the upper border of the aneurysm, approximately 6 to 8 cm in length. The aortic valve leaflets appeared normal with moderately enlarged and distorted aortic annulus. Two mattress sutures were applied: one at the anterior commissure between the right and the non coronary cusp, and a second between the non coronary and the left cusp, in order to reduce the annulus diameter. Following that, the aneurysm was excised and the ascending aorta restored by approximating both edges of the dissection utilizing two rows of silk material. During this stage of the procedure, the left coronary artery was continuously perfused. After the patient was disconnected from the heart-lung machine, the ascending aorta was wrapped with a Dacron graft from the base of the aorta up to the innominate artery and tightly sutured (Fig 4).

The patient made an uneventful recovery. The specimen obtained at surgery showed typical changes of cystic medial necrosis and also a part of the aortic wall where the dissection process began (Fig 5).

Four months after surgery, he underwent a second left heart hemodynamic study (Table 1). The postoperative

Table 1—Hemodynamic Data

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure</th>
<th>Site</th>
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<tr>
<td>Left Ventricle</td>
<td>110/10 mm Hg</td>
<td>Left Ventricle</td>
<td>115/10 mm Hg</td>
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<td>Ascending Aorta (mean: 55)</td>
<td>mm Hg</td>
<td>Ascending Aorta (mean: 77)</td>
<td>mm Hg</td>
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<td>Femoral Artery (mean: 57)</td>
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Figures 3 and 4. Figure 3. The retrograde aortogram shows a large irregular aneurysm located anteriorly and to the left of the ascending aorta. There is also marked regurgitation of contrast material into the left ventricle.

Figure 4. Anterior aspect of the heart and the ascending aorta. An aneurysmatic formation arising from the ascending aorta and projecting anteriorly is clearly seen.
to be congenital in two \(^1\) and the etiology remained unspecified in the other four \(^1\) (Table 2). Although the manifestations of Marfan’s syndrome are widely known, the syndrome is usually diagnosed in its full form. Occasionally, however, an isolated aortic aneurysm with secondary aortic regurgitation were observed without the other stigmata of Marfan’s. The lack of skeletal or eye abnormalities in these patients should not deter from the correct diagnosis, since the characteristic cystic medial necrosis is universally present. A few authors have recently reported dilated aortic root with isolated aneurysmatic dilation of the ascending aorta and aortic regurgitation. The structural defect of this syndrome labeled “annulo aortic ectasia” is cystic medial necrosis which may or may not be accompanied by the other stigmata of Marfan’s. This syndrome so far was observed only in adults and the youngest reported patient was 21 years old.\(^1\)

The aortic valve regurgitation brought our patient to attention. The aneurysm was not clinically suspected. The clue to the correct diagnosis was provided by the aortography. At this stage several etiologic possibilities were considered: mycotic vascular aneurysm, aortico-left ventricular canal\(^1\) and form fruste of Marfan’s syndrome. The aortico-left ventricular canal was readily excluded in the absence of the typical channel that characterizes this.

DISCUSSION

Aneurysm of the aorta is uncommonly observed in childhood. Javet and Kahn in 1952\(^1\) reviewed 53 cases of aneurysm of the aorta in patients under 18 years of age and pointed out the difficulties in establishing the correct etiology in the majority of the cases. Subacute bacterial endocarditis or sepsis were thought to be, in their review, the most common cause of aneurysm of the aorta in childhood.

A review of the literature since 1952 revealed 14 additional children with thoracic aortic aneurysm;\(^1\),\(^4\),\(^9\),\(^1\),\(^1\),\(^1\) eight showed the full stigmata of Marfan’s syndrome;\(^4\),\(^7\),\(^9\) the aneurysm was thought.
entity on aortography. The microscopic changes helped to classify our patient as a form fruste of Marfan's or within the group of patients with annulo aortic ectasia due to idiopathic cystic medial necrosis.14

The surgical treatment of aortic aneurysm in Marfan's syndrome or in patients with annulo aortic ectasia seems to be the most desirable and effective method of improving the otherwise poor outlook of this malformation.13-19 These patients are commonly confronted with a double threat: death from progressive left ventricular failure or dissection and rupture of the aorta.

Dissecting aneurysm in childhood is very uncommon and carries a bad prognosis. Dissection was observed in six of the above mentioned 14 patients with aortic aneurysm reported since 1952.14,16,19 (Table 2). Two of the six showed the full stigmata of Marfan's. Aortic valve regurgitation was present in only one prior to death.6 Chest trauma sustained during a ball game was the cause of dissection in a 12-year-old boy. None of the six patients was surgically treated and none survived. The incomplete dissection observed in our patient was apparently of recent origin, indicating the stage to which the disease had advanced.

The surgical procedure performed in our patient is not usually advised for patients with aortic aneurysm and aortic valve regurgitation, because of the patient's age. The aneurysm was excised and the aorta was wrapped to minimize the possibility of recurrence; however, it was clear that a prosthetic valve would not fit at the present time to correct the aortic regurgitation. Therefore, an attempt was made to diminish the incompetence by aortic annuloplasty. It is evident that in the future, if anatomic conditions permit, valve replacement may be considered. To the best of our knowledge our patient is the youngest surgically treated for aortic aneurysm with incomplete dissection due to idiopathic cystic medial necrosis.

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REFERENCES


Reprint requests: Dr. Gueron, Negev Central Hospital, Beersheva, Israel

Table 2—Thoracic Aorta Aneurysm in Childhood

<table>
<thead>
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<th>Author</th>
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