Combined Supravalvular Aortic and Pulmonic Stenosis

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SIGNIFICANT outflow obstruction of both ventricles on a congenital basis is rare. Only 11 cases have been reported1-7 and, in most instances, the stenosis was either valvular or subvalvular on both sides. Supravalvular stenosis occurring in the pulmonary arteries or in the root of the aorta, although uncommon, has been reported frequently in recent years. The association of these two lesions in the same patient, however, has not, to our knowledge, been previously described. Two unusual cases recently reported by Soulié and co-workers8 involved the coexistence of stenosis or hypoplasia of the primary pulmonary artery branches and hypoplasia of the thoracic and abdominal aorta, in the absence of supravalvular aortic stenosis. These authors also mention a similar case reported by Bulgarelli9 in which narrowing of the main pulmonary artery and its branches was associated with hypoplasia of the entire aorta.

The purpose of this report is to describe the combination of supravalvular aortic stenosis and postvalvular pulmonic stenosis seen in three patients during the past 3 years and to stress the common features of the two conditions.

Case Reports

Case 1

S.M., a 16-year-old boy, was admitted to the hospital in February 1960, complaining of marked diminution of his exercise tolerance because of dyspnea, fatigued cough, and hemoptysis. A heart murmur had been discovered at the age of 18 months, following an attack of cyanosis. At the age of 10 years, he began to experience dyspnea upon moderate exertion. After the age of 13, he was unable to participate in competitive athletics. Five months prior to admission, after running 200 yards, he experienced intense dyspnea and gross hemoptysis. From then on, he had bouts of hemoptysis almost daily following slight exertion. This was accompanied by frequent coughing and marked dyspnea on exertion. Moderate orthopnea was also present. There was no history of dizziness, syncope, or angina pectoris. The patient was a tall, muscular adolescent. The blood pressure was 95/65 mm. Hg in the right arm, 100/70 mm. in the left arm, and 125/70 mm. in the lower extremities. The apex of the heart was palpated in the left fifth intercostal space, slightly outside the midclavicular line. A rough ejection-type systolic murmur, grade III/IV, was maximal at the right second intercostal space and radiated over all the precordium and to the interscapular area. The aortic and pulmonic components of the second sound were both decreased in intensity, the aortic component to a greater degree. The lung fields had inconstant moist rales, but there was no other evidence of heart failure.

The electrocardiogram showed an axis at +90°, a vertical electrical position, atrial enlargement, and predominant left ventricular hypertrophy pattern of the systolic overload type (fig. 1). The vectocardiogram showed left ventricular hypertrophy predominantly, but signs of right ventricular hypertrophy were also present. Chest X-ray and fluoroscopy revealed generalized cardiomegaly with evidence of enlargement of all chambers (fig. 2). The aortic arch was small, and pulmonary markings were normal.

Right heart catheterization demonstrated systolic pressure gradients between the main pulmonary artery and its right and left branches. Retrograde left heart catheterization showed a 125 mm. Hg systolic pressure gradient between a supravalvular chamber and the ascending arch of the aorta (table 1). Angiocardiographic studies were not performed.

On March 23, 1960, cardiac surgery for repair of the aortic lesion was undertaken under extracorporeal circulation. The heart appeared grossly enlarged and both coronary arteries were huge, especially the right, which was the size of the surgeon’s little finger. The root of the aorta was small, tough, and thickened. A longitudinal incision was made in this area. The aortic wall was hypertrophic, and the lumen was markedly reduced. This was, in essence, a supravalvular coarctation that seemed to originate from the heart itself and extend distally over a distance of 2 to 3 cm. It was impossible to visualize the...
aortic valves and coronary ostia. The coarctation was deemed impossible to repair adequately. However, longitudinal streaks were made in the aorta in an attempt to enlarge its lumen. Closure of the aortic incision was complicated by severe hemorrhage, and the patient died from heart failure approximately 12 hours after the onset of the operation.

At autopsy, the heart weighed 550 Gm. There was generalized myocardial hypertrophy. All the cardiac valves were normal, except for slightly thickened aortic leaflets. The coronary ostia had a diameter of 4 to 5 mm. The coronary arteries were markedly dilated and contained numerous arteriosclerotic plaques. At a distance of approximately 1½ cm. above the insertion of the aortic valves, there was a fibrous ring with a superior opening of only 2 to 3 mm. This was followed by a narrowed ascending aorta with an external diameter of 1.2 cm. at the area of maximal stenosis. The wall of the ascending aorta averaged 4 mm. in thickness and did not have the elastic consistency of a normal aorta. The thickness of the wall decreased progressively after the origin of the innominate trunk. This vessel as well as the left carotid and subclavian arteries also had thick walls and a narrowed lumen at their origin. The pulmonary artery trunk had an external diameter of 1.8 cm. Its wall was thick, devoid of flexibility and elasticity, and in all respects identical to that of the ascending aorta. After its division into right and left pulmonary arteries, its wall decreased in thickness. At each subsequent division of the pulmonary arteries, the wall thickened again resulting in a decreased size of the lumen over a short distance and followed by a poststenotic dilatation until the next division. The venae cavae at their origin also had thick walls. Microscopically, the heart showed marked interstitial fibrosis and signs of coronary arteriosclerosis.

Case 2

J.D., a 21-year old married woman, the mother of two children, was asymptomatic until 4 months prior to her admission to the hospital in December

*Performed by Dr. Nicolas Aérichidé.
1959. She had a known heart murmur since early infancy. Her two previous pregnancies had been without incidents. Chief complaints were dyspnea and angina on moderate exertion, easy fatigability, palpitations, and frequent ankle edema. On occasions, she had noted some muscular weakness of the left hand.

The pulses of the right arm were stronger than those of the left arm. The blood pressure was 125/55 mm. Hg in the right arm, 90/60 mm. in the left arm, and 130/90 mm. in the lower extremities. The cardiac impulse was felt in the fifth intercostal space, 8 cm. to the left of the midsternal line. There was a well-defined systolic thrill, maximal in the suprasternal notch, of lesser intensity over the two carotid arteries, and still less intense over the aortic and pulmonic areas, where it was equally felt. A very loud coarse systolic murmur was heard all over the precordium and also over both sides of the back. It was coarsest in the suprasternal notch and over the carotid arteries. It was heard with about equal loudness in the aortic and pulmonic areas. The pulmonic second sound was present but the aortic second sound was very faint.

The electrocardiogram showed an axis at +90° and a vertical electrical position. It showed a left ventricular hypertrophy pattern of the systolic overload type (fig. 1). The vectorcardiogram showed bilateral ventricular hypertrophy with left ventricular predominance. On chest x-ray and fluoroscopy, the heart was slightly enlarged, with predominant left ventricular enlargement (fig. 2). Right ventricular enlargement was also evident. The aorta was small and poorly visualized, and the pulmonary vascularity was normal.

Right heart catheterization showed a systolic pressure gradient of 60 mm. Hg between the main pulmonary artery and the right pulmonary artery. Another small systolic gradient of 8 mm. Hg was present within the right pulmonary artery. The left pulmonary artery was not catheterized. Retrograde left heart catheterization showed a 140 mm. Hg systolic pressure gradient across a stenotic area in the ascending aorta (table 1). An angiogram done in the outflow tract of the right ventricle demonstrated the presence of multiple bilateral stenosis of the pulmonary artery (fig. 3).

The patient was referred to the Mayo Clinic, where surgical repair of the supravalvular stenosis was performed by Dr. John W. Kirklin on September 8, 1960. Prior to repair the systolic pressure of the left ventricle was about 200 mm. Hg, whereas that of the ascending aorta was 90 mm. There was a rather tough, thickened area in the aorta, just downstream of the sinuses of Valsalva. The sinuses themselves were markedly enlarged. The right coronary artery was quite large. The orifices of both coronary arteries were normal in size. At the attachment of the commissures of the aortic leaflets, there was a thick ridge producing a narrowing in that area. The orifice measured 6 mm. in diameter. The orifice of the left coronary artery was deep in the sinus of Valsalva. Between this ridge-like aortic obstruction and the left coronary cusp, there appeared to be virtually no opportunity for blood to get into the left coronary artery. The orifice of the right coronary artery seemed slightly downstream and appeared to receive blood adequately. The ridge-like thickening was carefully resected. In order to assure a more adequate orifice, the

Figure 2
Posteranterior roentgenograms of the chest showing, in all cases, mild to moderate generalized cardiomegaly.
incision was extended deep into the noneorornary sinus of Valsalva. A diamond-shaped piece of woven Teflon was used to enlarge the area as a plastic revision. After repair, the systolic pressure in the cavity of the left ventricle was 125 mm. Hg and 85 to 90 mm. in the ascending aorta. At the time of her dismissal, the patient was improved and did not require any cardiac medication.

Case 3

C.D., a 4-year-old girl, was born at term following a normal gestation and delivery. A heart murmur was discovered at the age of 6 months. She had frequent upper respiratory infections and was unable to keep up with other children because of easy fatigability. She was admitted to the hospital in April 1961 for investigation.

She was normally developed. Her blood pressure was 120/80 mm. Hg in the left arm, 130/110 mm. in the right arm, and 120/90 mm. in the lower extremities. The apex beat was felt in the fifth interscostal space at the left midclavicular line. A palpable systolic thrill was maximal in the suprasternal notch and also felt over both carotid arteries and aortic and pulmonic areas. A grade III/IV ejection-type systolic murmur was heard at the base and along the left sternal border, with maximal intensity at the left second interscostal space. It was transmitted to the neck and over both sides of the back. The aortic second sound

### Table 1

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<th>Case I</th>
<th>Case II</th>
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<td>24/14</td>
<td>20/10</td>
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<td>..</td>
<td>12/8</td>
<td>16/6</td>
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<tr>
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<td>..</td>
<td>32/8</td>
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<tr>
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<td>Wedge (mean pressure)</td>
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<tr>
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was markedly decreased and the pulmonic second sound was present.

The electrocardiogram showed an axis at +105° with a vertical electrical position, left ventricular hypertrophy, and incomplete right bundle-branch block (fig. 1). The vectocardiogram showed left ventricular hypertrophy and incomplete right bundle-branch block. Chest x-ray and fluoroscopy showed slight cardiomegaly, with enlargement of both ventricles and the left atrium (fig. 2). The aorta was small, and the pulmonary vascularity was normal.

Right heart catheterization revealed bilateral systolic pressure gradients at the origin of the right and left pulmonary arteries and at their point of division into secondary branches. At retrograde left heart catheterization, a systolic pressure gradient of 65 mm Hg was present across a stenotic area located approximately 2 to 3 cm. above the aortic valve in the ascending aorta (table 1). A selective angiocardiogram was done in the outflow tract of the right ventricle (fig. 4). The right ventricle was moderately enlarged. The main pulmonary artery measured 2 cm. in width, whereas the right pulmonary artery measured 10 mm. in diameter. The inferior lobar branches of the right and left pulmonary arteries showed visible small narrowings with poststenotic dilatations. The left ventricle was large, and its wall measured 10 mm. in thickness. In the proximal portion of the ascending aorta, immediately above the sinuses of Valsalva, there was a transversal annular narrowing, forming a clear band and measuring 8 mm. in transverse diameter as compared to a poststenotic ascending aorta measuring 12 to 13 mm. in diameter.

In view of her young age and absence of important symptoms, cardiac surgery was deferred.

Discussion

In true supravalvular aortic stenosis, marked thickening of the plica at the upper margin of the sinuses of Valsalva forms an encircling ridge of fibrous tissue that narrows the aortic lumen. In some cases, coarctation of the ascending aorta follows and may extend beyond the origin of the left subclavian artery. Stenosis of a subclavian artery, especially the left, at or near its origin is also frequent. Fibrous bands often extend from the free margins of the aortic leaflets to the plica, causing valvular insufficiency and impairment of coronary flow. A less common and nonobstructing form of supravalvular lesion is a fibrous band, cord, or membrane crossing or subtending the lumen of the aorta, just above the cusps. Except in one case in which the membrane extended over three fourths of the internal circumference of the lumen, this entity does not usually cause aortic stenosis, and represents an incidental autopsy finding. Before 1958, only four pathologic reports of true supravalvular aortic stenosis were published. The condition was first diagnosed clinically by Denie and Verheught and Morrow and associates. Since 1959, many reports have stressed the
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diagnostic and surgical aspects of this entity.\textsuperscript{16–22}

Postvalvular pulmonic stenosis most commonly consists of a constriction at or in the vicinity of the bifurcation of the main pulmonary artery. When proximal to the bifurcation, it usually extends over a short segment and realizes a coarctation of the pulmonary artery. These lesions are frequently accompanied by multiple stenoses at the site of division of the smaller branches of the pulmonary arteries. They usually give rise to significant right ventricular and pulmonary artery hypertension. A common variant, which is not hemodynamically significant, is the unilateral stenosis of a main branch of the pulmonary artery at its origin, involving especially the right pulmonary artery. The pathologic entity of postvalvular pulmonic stenosis was first described by Oppenheimer in 1938.\textsuperscript{23} Clinical, hemodynamic, and angiocardiographic aspects of this condition have been well documented since 1953.\textsuperscript{24–33}

In spite of these significant pathologic findings, there are few symptoms or clinical signs distinctive of either entity or of their association. In contrast to the other types of aortic obstruction, angina pectoris is usually absent, since the coronary arteries originate below the stenosis and are subjected to a high perfusion pressure. Dizziness and syncope are also rare and sudden death was seen only once.\textsuperscript{35}

A basal systolic thrill and ejection-type murmur are common to both entities, and maximal intensity on either side of the sternum may suggest especially one or the other condition. Their distribution, however, is different and transmission of the murmur to the suprasternal notch and carotid arteries is typical of the aortic stenosis, whereas transmission to both axillae and the back is due to the pulmonary stenoses. A difference in the pulses and blood pressure of the upper extremities is a frequent and valuable sign in supravalvular aortic stenosis. It is presumably due to stenosis of a subclavian artery,\textsuperscript{16} it involved the left side in two of our patients.

A small ascending aorta and absence of post-stenotic dilatation or valvular calcifications practically rule out valvular aortic stenosis of a significant degree. Signs of right ventricular hypertrophy on the electrocardiogram, vectorcardiogram, or chest roentgenogram, in addition to left ventricular hypertrophy, indicate the presence of an associated lesion and the need for further investigation.

Combined right and left heart catheterization clearly establishes the diagnosis of both conditions (table 1). A large systolic pressure gradient at the bifurcation of the main pulmonary artery was demonstrated in all our patients. Smaller pressure gradients were also present in the peripheral pulmonary arteries. In addition, characteristic and very similar withdrawal pressure tracings can be obtained from both ventricular outflow tracts.\textsuperscript{14, 34} (figs. 5 and 6). The ejection period of both ventricles is prolonged because the semilunar valves remain open as long as the ventricular pressure exceeds that distal to the stenosis. During this period, the arterial pressure beneath the stenosis is identical in height, contour, and timing to the corresponding ventricular pressure. On the other hand, the diastolic arterial pressure proximal and distal to the stenosis has the same level and morphology. This results, in the supravalvular chamber, in an abnormally wide pulse pressure, with a descending limb of the pressure tracing which is steeper than in the area distal to the stenosis. The morphology of the diastolic pressure, in all segments of the aorta and in the pulmonary artery, is characterized by a deep dicrotic notch followed by a low diastolic plateau (figs. 5 and 6). These changes are probably due to the fact that, owing to the supravalvular stenosis, the retrograde surge of blood is greatly restricted in early diastole.\textsuperscript{34}

According to Agustsson and associates,\textsuperscript{34} a normal progressive fall in diastolic pressure, such as that registered in our second case, is only seen with unilateral stenosis of a main branch of the pulmonary artery (fig. 6). Since the left pulmonary artery was not cath-
Figure 5

Continuous recordings of aortic and left ventricular pressures in cases 1, 2, and 3. The diastolic aortic pressures, proximal and distal to the stenosis, show a deep dicrotic notch and a low diastolic plateau in all three cases.

eterized in this patient, it is probable that there was no constriction in the left proximal branch and that the hypertension present in the main pulmonary artery was produced by the multiple peripheral pulmonary stenoses.

In two of our patients, the pressure tracings registered in the right subclavian or brachial artery showed the usual anaerotic notch and slow upstroke time of aortic stenosis. However, the dicrotic notch remained very low on the descending limb of the pressure tracing and the dicrotic wave was replaced by the characteristic diastolic plateau (fig. 7). This is rare in other forms of aortic stenosis and may suggest the diagnosis of supravalvular aortic stenosis prior to left heart catheterization.

In our patient who came to autopsy, the striking lesion, also shown in previous pathologic reports of supravalvular aortic steno-
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sis,12-15, 22 was segmental fibrosis of the wall of the blood vessels involved. This produced thickening of all the areas of division of the pulmonary arteries and even at the origin of both venae cavae in addition to the supra-valvular aortic narrowing and stenosis of the vessels arising from the aortic arch. Diffuse fibrosis of the myocardium, previously noted frequently in supravalvular aortic stenosis,2-5, 19, 20 was also present. Arteriosclerotic plaques in the coronary arteries and other manifestations of coronary arteriosclerosis have also been found in other cases15, 22 and are presumably due to the marked hypertension present at the root of the aorta.

Both conditions are generally regarded as congenital in origin. The discovery of a heart murmur in infancy in all our patients gives support to this opinion. Marfan’s features and a true Marfan syndrome,12, 14 a typical facies, and mental retardation,20 have all been described with supravalvular aortic stenosis.

In postvalvular pulmonic stenosis, similar characteristics have also been noted occasionally.30, 32 Such associated anomalies were not present in any of our cases. On the other hand, the association and similarities of these two unusual entities in our patients is probably more than coincidental. As indicated by the pathologic findings in our first case, the underlying lesion could be a fibrotic process, of developmental origin, involving the large vessels at the base of the heart and their branches.

The surgical technic used by Dr. John W. Kirklin to repair the supravalvular aortic stenosis in our second case is the one now commonly used. At least seven others have been operated on successfully by this method.16, 18, 20, 21 A long supraaortic coarctation was not found in any of these patients. On the other hand, at least four patients with lesions very similar to our first case have died following surgery.14, 15, 19, 22 It is then impera-

Figure 6
Continuous recordings of pulmonary artery and right ventricular pressures in cases 2 and 3. Note the resemblance of these tracings with the left heart pressures. The absence of a diastolic plateau of the pulmonary artery pressure in case 2 is probably due to unilateral stenosis of the right pulmonary artery in this case.
Peripheral arterial pressure in case 2. Note the persistence of a characteristic diastolic pressure with a low dicrotic notch and diastolic plateau, instead of the usual high dicrotic notch and prominent dicrotic wave.

tive that a thorough investigation be made prior to surgical repair, especially since the diagnosis can be established with certainty in practically all cases.

In postvalvular pulmonic stenosis, the pulmonary artery and its branches have been dilated successfully in a few reported cases.\textsuperscript{24, 30}

Summary

The association of supravalvular aortic stenosis and postvalvular pulmonic stenosis is described in three patients. In addition to the clinical findings resulting from obstruction to both ventricular outflow tracts, other unusual features were also present, such as a difference in the pulses and blood pressure of the upper extremities and absence of poststenotic dilatation of the ascending aorta. Diagnosis was established at cardiac catheterization by the demonstration of important pressure gradients in the aorta and in the pulmonary arteries with characteristic and almost identical pressure tracings on both sides. The pathologic lesions demonstrated in our patients by selective angiocardigraphy, surgery, and autopsy are discussed. The possibility of a common etiology for both entities is suggested.

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