CLINICAL CONFERENCE

Editor: EDGAR V. ALLEN, M.D.
Associate Editor: RAYMOND D. PRUITT, M.D.

Congenital Aortic Stenosis

By NORMAN J. SISSMAN, M.D., CATHERINE A. NELL, M.D.,
FRANK C. SPENCER, M.D., and HELEN B. TAUSSEG, M.D.

DR. NORMAN J. SISSMAN, Moderator: Interest in aortic stenosis has recently been greatly enhanced because of the surgical advances in its alleviation. The following 4 cases are presented to illustrate some of the problems in diagnosis and treatment of this lesion and to record 2 unusual pathologic variants.

Case 1. J.S. (HLH Number A-96356), a white girl, was observed in the Cardiac Clinic of the Harriet Lane Home from the age of 15 months until her death at the age of 5 1/2 years.

The family history was not pertinent. There were no siblings. The patient was born at full term; birth weight was 6 pounds, 14 ounces. A heart murmur as well as unusual facies and webbed neck were noted at birth. Her psychomotor development was retarded; she was not able to sit up even with support until the age of 12 months; she began to walk only at the age of 2 years and she did not use words until the age of 3 1/2 years. Her past medical history was negative. The only cardiac symptoms noted by the parents were slight dyspnea, fatigue, and dizziness around the lips and nose after exertion.

Physical examination at 15 months revealed an average-sized, mentally retarded child. The face was unattractive: the nose was broad and upturned, there were bilateral slight ptosis of the eyelids, a suggestion of micrognathia, and minimal bilateral webbing of the neck. There was bilateral cubitus valgus. The left anterior chest wall was prominent. The peripheral pulses were of good volume in all 4 extremities. The systolic blood pressure was 90 mm. Hg in the arm and 120 in the leg. The heart was moderately enlarged to the left. There was an intense systolic thrill over the entire precordium, felt best along the upper left sternal border. There was a grade-IV, long, harsh systolic murmur, loudest between the second and fourth intercostal spaces along the left sternal border, heard well over the back and in both axillae, but poorly transmitted to the great vessels of the neck. No second sound could be heard at the base of the heart on either side of the sternum. A spectrophonocardiogram taken by Dr. V. A. McKusick confirmed the auscultatory findings. It showed that the systolic murmur had the "Christmas tree" configuration usually observed in the sounds produced by the ejection of blood through a stenotic area. No clear second sound was recorded anywhere except at the localized area in the left midprecordial region; even there it was faint and had a low frequency. Fluoroscopic examination revealed the cardiothoracic ratio to be increased to about 60 per cent. The right atrial shadow was prominent. The main pulmonary artery segment was concave. The pulmonary vascularity appeared normal in the hilar areas but was slightly decreased in the periphery of the lung fields. In the left anterior oblique view, the right ventricular shadow was enlarged anteriorly and the left ventricular shadow did not clear the spinal column until the patient was rotated to an angle of 75 degrees anterior to the upright fluoroscopic table. The electrocardiogram (fig. 1) was interpreted as showing right ventricular hypertrophy and possibly also some left ventricular hypertrophy.

Examinations over the next 4 years showed little change in the cardiac findings. The blood pressure showed a narrow pulse pressure; it was 110/80 mm. Hg in the arm and 100/60 mm. Hg in the leg.

From the Departments of Pediatrics and Surgery, Johns Hopkins University School of Medicine, Cardiac Clinic of the Harriet Lane Home, Johns Hopkins Hospital, Baltimore, Md.

Aided by grants from the Suffolk County (N.Y.) Heart Chapter of the American Heart Association, and the Department of Health, State of Maryland (U.S. Children’s Bureau).
CONGENITAL AORTIC STENOSIS

at the age of 4½, and 6 months later it was 118/68 mm. Hg in the arm and 130/100 mm. Hg in the leg. There was a slight increase in the cardio-thoracic ratio over the 4-year period, from 59 per cent to 62 per cent (fig. 2). At the age of 3 years, the liver was palpable 7 to 8 cm. below the right costal margin; it was smooth but not tender and did not pulsate. An endocrinologic consultant reported a slightly advanced bone age, a female chromosomal pattern on skin biopsy, and no specific endocrinologic abnormalities. Repeated blood counts, urinalyses, and liver function tests were normal. At the age of 5 years, right heart catheterization was attempted but was unsuccessful.

The clinical diagnosis was pulmonary stenosis, either valvular, infundibular, or both. Because of the gradually but progressively increasing cardiac size, operation was recommended. This was performed by Dr. Frank C. Spence who employed hypothermia. Systolic pressures were recorded, by direct measurement as follows: right atrium, 15 mm. Hg; left atrium, 25 mm. Hg; pulmonary artery, 30 mm. Hg; right ventricle, 175 mm. Hg. The pulmonary artery was opened and a valvular stenosis corrected. Infundibular stenosis was also noted, and this was excised from below through a right ventriculotomy. Following these procedures the right ventricular pressure varied from 25 to 75 mm. Hg and the pulmonary artery pressure was 10 mm. Hg. Although a loud systolic murmur persisted, the postoperative course appeared satisfactory until 8 hours after surgery, when she developed sudden respiratory distress and died within 10 minutes.

Autopsy (no. 27006), performed by Dr. E. Hurst, revealed combined pulmonary and aortic stenosis and pulmonary congestion. The heart was greatly enlarged. The right atrium was normal. The foramen ovale was sealed and the tricuspid valve was normal. There was extreme hypertrophy of the right ventricle; its wall measured 12 mm. in thickness. There was marked stenosis in the infundibular area; the opening in this region in the fixed state admitted only the tip of a pair of scissors. A portion of the localized constricting muscle on the medial aspect of the infundibulum had been removed surgically. The pulmonary valve revealed a ring of diminished size (10 mm. in diameter) and the cusps were thickened and distorted, especially at their free edges. Recent cuts had been made along the commissures; at autopsy the valve admitted the tip of an index finger. The pulmonary artery above the valve was slightly dilated. The pulmonary veins entered normally into the left atrium, which was normal. Examination of the mitral valve disclosed some shortening and thickening of the chordae tendineae and the valve itself seemed smaller than normal with thickening of the leaflets; however, there was no stenosis. The left ventricle was slightly hypertrophied; its wall was 16 mm. in thickness. The cusps of the aortic valve were thickened and showed slight fusion of the posterior and right commissures; the valve ring was also 10 mm. in diameter. There were thickening and scarring of the endocardium beneath the aortic valve which, together with a fibrous band that ran from the base of the aorta.

Fig. 1. Case 1. Electrocardiogram at the age of 5½ years. Note the deep S wave in V1 and the tall peaked T wave in V3R.
to the mitral valve (fig. 3), caused a subvalvular obstruction in the left ventricular outflow tract. The coronary ostia were wide and the coronary arteries had delicate walls. Blood-stained mucus was found in the bronchial tree, and the parenchyma of the lungs showed congestion and diminished aeration.

Dr. SISSMAN: This is the first case of combined aortic and pulmonic stenosis with an intact septum that we have encountered in this clinic. Dr. Taussig, would you comment on some of the clinical aspects of this case that could have given us clues to the correct diagnosis?

Dr. HELEN B. TAUSSIG: I think there were 3 main clinical clues. The first was the presence of left ventricular hypertrophy on fluoroscopy. Several observers thought that the left ventricle looked enlarged; others thought it was being pushed posteriorly by the enlarged right ventricle. It would have been wise to hunt for the interventricular groove in the left anterior oblique position. Usually, with extreme enlargement of the right ventricle, one can easily see the interventricular groove because it is displaced posteriorly. Secondly, the electrocardiogram differed from the pattern usually seen in cases of severe "pure" pulmonic stenosis in that lead V₃ did not show the usual tall R wave with an inverted T wave; the deep S wave in this lead should have made us suspicious of additional left ventricular hypertrophy. The tall T waves in V₃ are thought by some to be associated with left ventricular hypertrophy, although they are supposed to reflect "diastolic overload" of the left ventricle, which was not present in this case. Also, the patient did not have the high pyramidal P waves commonly seen in severe pulmonic stenosis. Thirdly, the repeated auscultatory observation that the second sound was not present on either side of the sternum or at the apex should have caused us to wonder if the aortic as well as the pulmonic valve was involved.

Dr. SISSMAN: How would more detailed physiologic investigations have helped us?

Dr. TAUSSIG: It is unfortunate that the right heart catheterization was unsuccessful; but had it been completed, it would have merely confirmed our clinical impression of a rather severe pulmonic stenosis. Left heart catheterization would have established the presence of obstruction to left ventricular outflow but here as with other similar cases, one must balance carefully the possible complications of such a procedure with the value of the information obtainable. I do think that, knowing this was not a typical pulmonic stenosis, we should have suggested to the surgeons that at the time of operation they
CONGENITAL AORTIC STENOSIS

461

take pressures from the left-sided chambers of
the heart as well as from the right; this would
have enabled them to proceed differently. Had
the presence of the additional aortic stenosis
been recognized at the time of surgery, proba-
bly it would have been wiser to back out
immediately and re-operate with use of open
heart technics.

**Dr. Sissman**: Dr. Spencer, Dr. Taussig has
suggested a course for the surgeons! Would
you tell us what you would have done had
the correct diagnosis been established before
the operation?

**Dr. Frank C. Spencer**: There is no obvious
reason why combined aortic and pulmonic
stenosis, either valvular or infundibular, could
not be relieved during the same operation.
The best approach would include using the
heart-lung machine. The actual procedure in
such an instance would be to relieve the pul-
monic stenosis first while on the pump with-
out inducing cardiac standstill. After relief
of this stenosis, the heart could be stopped, the
aorta opened, and then the aortic stenosis
relieved and the heart re-started. This method
would minimize the period of cardiac isch-
emia.

**Dr. Sissman**: In this case, in which there
was a fibrous band extending from the sub-
aortic region to the mitral valve, could the
aortic subvalvular stenosis have been relieved
without creating mitral regurgitation?

**Dr. Spencer**: Yes, by avoiding extensive
resection in this area.

**Dr. Taussig**: In general, I think that the
less resection there is of actual muscle, the
less chance there will be of subsequent scar-
rine. In this connection, Dr. Mary Allen
Engle recently described 3 cases in which
there appeared to be combined valvular and
infundibular stenosis of the pulmonary out-
flow region and in which the latter ‘resolved’
after the relief of the valvular stenosis alone.
It was postulated by her that the infundibular
‘stenosis’ was the result of hypertrophy of the
crista supraventricularis and she thinks that
usually there is no necessity for surgical
resection of the muscular stenosis.

**Dr. Sissman**: This subaortic stenosis was
fibrotic rather than muscular. Dr. Neill,
would you say something about this lesion
from an embryologic point of view?

**Dr. Catherine A. Neill**: There are fibrotic
lesions on both sides of the heart and they
extend a considerable distance down from
the valves. It appears as if the chambers
themselves were formed normally and the
fibrosis took place at a considerably later
stage in embryonic development than we
are accustomed to associate with the forma-
tion of cardiac anomalies.

**Dr. Sissman**: Dr. Eugene Braunwald of the
National Heart Institute tells us that his
group has seen 3 cases of combined aortic
and pulmonic stenosis. The patients appeared
with symptoms of aortic stenosis and were
diagnosed by left and right heart catheteri-
izations. A full report on these cases is
being prepared. Another case of this type
has already been reported in the literature.2
This combination of lesions is certainly one
for which we should be on the alert.

**Case 2. S.A.C.** (HLH Number B-2306), a
white girl, was first seen in the Cardiac Clinic of
the Harriet Lane Home in September, 1955, at
the age of 19 months because of a heart murmur
which had been heard at birth, failure to thrive, and
episodes of ‘congestive failure.’

The family history revealed no significant dis-
seases. One previous pregnancy had terminated
in a miscarriage at 4 months. The mother’s health
during this pregnancy was good. The patient was
born without complication at term. The present
illness dates from birth. A heart murmur had
been heard at birth and persisted. She became ill
on the third day of life and remained in the hos-
pital for 2 months. During the first month she
was periodically febrile and suffered from dyspnea
and cyanosis, which required continuous admin-
istration of oxygen. She took her feedings poorly,
and even at the age of 2 months had failed to
regain her birth weight. After discharge from
the hospital, she continued to do poorly. She had
frequent respiratory infections, appeared pale and
weak, and usually breathed heavily. Between the
ages of 5 and 19 months, she had 3 episodes of
what was called ‘congestive heart failure.’ These
attacks were characterized by the sudden onset of
irritability, prolonged continuous crying, refusal
to eat, rapid respirations, pallor, and excessive
sweating. Hepatomegaly and tachycardia were
also noted at these times. It was because of the
third attack of this nature that she was referred to the Cardiac Clinic.

Physical examination at the age of 19 months showed an underdeveloped, undernourished child, in acute distress with rapid respirations and suprasternal retraction. The heart rate varied from 130 to 180 per minute; the rhythm was irregular. The blood pressure was 90/60 mm. Hg in the arm and 110/70 in the leg. The left chest was prominent. The point of maximal apical impulse was 2 cm. to the left of the midclavicular line. A systolic thrill was felt along the right sternal border. There was a harsh systolic murmur which was loudest over the upper and middle sternal areas, but could be heard over the entire precordium and back. The second sound at the base to the right of the sternum was diminished in intensity. Fluoroscopy showed the heart to be enlarged with a cardiothoracic ratio of approximately 70 per cent. In the oblique views both ventricles appeared to be enlarged. The main pulmonary artery segment was prominent and pulsed vigorously, but the peripheral pulmonary vascularity appeared normal. A barium swallow showed a left aortic arch and moderate left atrial enlargement. The electrocardiogram showed atrioventricular disso-

![Fig. 4. Case 2. A. Typical preoperative resting electrocardiogram, at the age of 2⅔ years. B. Electrocardiogram during an attack of angina pectoris, age 3 years. C. Electrocardiogram, taken 3⅔ weeks after operation.](image-url)
CONGENITAL AORTIC STENOSIS

ones. Examination at that time showed her to be in acute distress with pallor, continuous crying, and marked diaphoresis. The pulse was 160 per minute and the respirations 40 per minute. The physical findings remained unchanged. An electrocardiogram taken at this time (fig. 4B) showed slight S-T depressions and marked inversion of the T waves in leads I, aV_L, and V_3L through V_6, consistent with myocardial ischemia. She was re-admitted to the hospital and responded within a few hours to treatment with morphine and oxygen. An electrocardiogram taken the following day showed reversion to its previous pattern.

Because of the character and location of the murmur and the repeated episodes of what appeared to be angina pectoris, the diagnosis of aortic stenosis was entertained. Right heart catheterization was performed, however, to determine the presence or absence of other cardiac lesions. This study showed an increase in oxygen saturation from 67 per cent to 86 per cent between the right atrium and the right ventricular outflow region. The pressure in the pulmonary artery was 36/13 mm. Hg; in the right ventricle it was 60/4 mm. Hg. The femoral artery pressure was 132/97. The pulmonary flow was approximately 4 times the systemic flow. Therefore, the diagnosis was changed to a ventricular septal defect.

In June 1957 operation was performed by Drs. H. Bahnsen and F. Spencer for direct-vision closure of the lesion with the aid of a Gaertner-Kay heart-lung pump. The defect, which was small and lay in the membranous septum behind the crista supraventricularis, was closed completely. There was no evidence of aortic stenosis. Postoperatively, her course was satisfactory. When examined 3 months postoperatively, she was asymptomatic. Physical examination demonstrated only a short, soft systolic murmur along the upper left sternal border and no thrill. The heart size had decreased considerably. The electrocardiogram (fig. 4C) showed complete right bundle-branch block and decrease in the depth of the Q waves over the left-sided precordial leads.

**DR. SISMAN:** Although this case is not one of aortic stenosis, it is included because the clinical picture was more suggestive of aortic stenosis than of a ventricular septal defect. Dr. Taussig, would you comment on the differential diagnosis of these 2 lesions?

**DR. TAUSSIG:** The diagnostic difficulties in this patient were the reverse of those usually encountered in childhood. More commonly, early in life the murmurs of aortic stenosis mimic those of a ventricular septal defect. Thus, in infancy and early childhood the stenotic murmur may be located quite low down along the left sternal border and does not radiate into the neck. Later, the murmur becomes loudest in the second right parasternal area and changes in character. In this case, the murmur was maximal over the base of the heart and to the right of the sternum and this, together with the absence of the usual evidence of increased pulmonary flow and the presence of attacks of angina, misled us. This is the first time I have ever seen angina in a ventricular septal defect, but I see no reason why a sudden decrease in systemic flow at times might not reduce coronary flow to such a degree as to cause temporary myocardial insufficiency, such as occurred in this patient.

**DR. SISMAN:** Systemic flow at the time of catheterization was not reduced, but of course this was not determined during an anginal attack. I know of no specific studies of coronary flow in ventricular septal defects. Dr. Neill, would you comment on the electrocardiographic changes?

**DR. NEILL:** The S-T and T-wave changes in the electrocardiogram during the anginal attack strongly suggest myocardial ischemia. The return to the usual pattern by the following day excluded the possibility of infarction. In all the records, the Q waves in lead I and over the left side of the precordium were unusually prominent for a young child with a ventricular septal defect. The combination of these Q waves and the inverted T waves over the left precordium made us favor aortic stenosis.

**DR. SISMAN:** Dr. Spencer, will you answer 2 questions for us? Were the findings at operation out of the ordinary, and, do you think the location of the defect would help explain the physical findings of aortic stenosis?

**DR. SPENCER:** The ventricular defect was located just proximal to the crista supraventricularis, which has been a common location for the ventricular defects in our series. The crista seemed unusually hypertrophied;
this may have given rise to the marked murmur and thrill which were present. I have no other explanation for the signs suggesting aortic stenosis.

Dr. Taussig: The finding of a pressure differential of 30 mm. Hg across the pulmonary valve at the time of catheterization also might be explained by the large crista.

Dr. Sissman: Recently, there has been a report by Hancock et al.\(^9\) from Boston concerning 7 persons over 50 years of age who were diagnosed as having aortic stenosis from their histories and physical findings, but were found subsequently to have either severe myocardial disease or unsuspected disease of the mitral valve.

Dr. Taussig: We have run into similar situations on several other occasions. In atypical cases, aortic stenosis remains one of our most tricky diagnostic problems.

Dr. Sissman: This case indicates that ventricular septal defects must be included in the differential diagnosis of aortic stenosis.

Case 3. P.W., Jr. (HLH Number A-71350) was first seen in the Cardiac Clinic in July 1949 at the age of 6 years, when he was admitted to the Harriet Lane Home because of subacute bacterial endocarditis superimposed upon congenital heart disease. Thereafter, his case was followed until his death at the age of 11½ years.

The family history revealed that his maternal grandmother (see case 4 below) had congenital heart disease. Following his birth, his mother had 2 unsuccessful pregnancies, both complicated by pre-eclampsia. The mother had pre-eclampsia during this patient's gestation, and he was born prematurely. A heart murmur was heard shortly after delivery and persisted. When he was 4 months old, the mother noted minimal cyanosis during a severe upper respiratory infection. Throughout his life, his activity had been moderately restricted. He remained relatively asymptomatic until 2 months before admission, when he developed an intermittent fever, anemia, and had a severe epistaxis. He was admitted to another hospital where a diagnosis of septicemia was made and where he was treated with blood transfusions, penicillin, sulfonamide, and aureomycin. After his discharge from this hospital, his fever reappeared and petechiae were noted. Two weeks after the recurrence of these symptoms, he was admitted to the Harriet Lane Home.

On examination the heart was moderately enlarged to the left; the rhythm was regular, and the heart sounds were of good quality. There was a pathologic third sound at the apex. There was a systolic thrill along the left sternal border and a harsh systolic murmur which was audible over the entire chest but was loudest in the second and third interspace at the left sternal border. The blood pressure was 105/75 in the arm. Fluoroscopy showed a moderately enlarged globular heart. The pulmonary vessels were enlarged and pulsed more vigorously than normal. In the left anterior oblique position, the left ventricle did not clear the shadow of the spine until the patient was rotated to an angle of 85 degrees anterior to the upright fluoroscopic table. The electrocardiogram showed right axis deviation, a moderately deep Q wave and inverted T wave in lead III, and a decreased ratio of the height of the R wave to the depth of the S wave in V1. Blood cultures established the diagnosis of subacute bacterial endocarditis due to an alpha hemolytic streptococcus. He was treated with a 6-week course of penicillin and made an uneventful recovery.

Over the next 4½ years, his heart showed striking progressive enlargement. In 1949, the cardiothoracic ratio was 53 per cent; by 1954 it had increased to 67 per cent (fig. 5). Periodic fluoroscopy indicated that the enlargement was mainly left ventricular. The electrocardiograms also showed increasing left ventricular hypertrophy: the electric axis became balanced, the voltage in the left-sided precordial leads increased, the QRS complexes showed widening and lengthening of the ventricular activation times over the left ventricle, and the ST-T segments showed changes of so-called left ventricular strain (fig. 6). Repeated physical examinations showed that the systolic murmur radiated well into the neck and ob-
seured the second sound in the second interspace to the right of the sternum. The third heart sound persisted and was interpreted as a gallop. His blood pressures averaged 100/75. The boy showed increasing dyspnea on exertion and ease of fatigability. In early 1954, he began to have attacks of dyspnea, wheezing, and coughing which resembled asthma. In late 1954, he developed frank congestive failure, which progressed despite vigorous therapeutic measures. The diagnosis of congenital aortic stenosis had been made, but, to exclude the presence of other lesions, venous catheterization was performed; this showed no shunt of blood. The pulmonary artery pressure was 70/41 mm. Hg, and the right ventricular pressure was 85/0.5 mm. Hg. The femoral artery pressure tracing showed a flat curve consistent with aortic stenosis.

Because of his intractable heart failure, an operative attempt to relieve the stenosis was recommended. A transventricular valvulotomy was performed under general anesthesia by Dr. H. Bahnson. There was great resistance to the opening of the dilator; the amount of relief obtained was questionable. After closure of the ventriculotomy, the patient developed ventricular fibrillation and died on the operating table.

Autopsy (no. 25546), performed by Dr. S. Wood, revealed that the heart weighed 650 Gm. The right side of the heart was normal. The left ventricle was greatly dilated and hypertrophied. Its wall measured 1½ cm. in thickness. The mitral valve was normal. The aortic valve measured 4 cm. in circumference at the valve ring. One centimeter above the aortic valve, the inner surface of the aorta was markedly roughened due to a heaping up of the endothelium. This roughening caused the aorta to be constricted to a circumference of 3 cm. The constricted area extended a distance of 2 cm. up the aorta from its proximal level. The changes in the aortic wall were attributed to healed subacute bacterial endocarditis. Beyond this abnormal area the aorta was normal. The aortic valve itself showed marked thickening of the valve leaflets, which must have produced a significant degree of stenosis. The coronary arteries were normal but there was diffuse scarring of the left ventricular myocardium.

Dr. Taussig: It should be remembered that when this patient was first seen by us in 1949, the protean manifestations of aortic stenosis were not familiar to us. It is of interest, for example, that this patient had right axis deviation in his electrocardiogram even at the age of 6 years and that the axis did not shift into the normal quadrant until after he had subacute bacterial endocarditis.

Dr. Sissman: Was it not significant that his clinical course rapidly worsened after his endocarditis?

Dr. Taussig: Yes. In our present age of antibiotics, I think it is important to emphasize that many patients still do poorly despite bacteriologic cures.
Dr. Sissman: Perhaps this is due to myocardial involvement. Perry, Edwards, et al. analyzed 52 autopsied cases of subacute bacterial endocarditis and found myocardial lesions of varying degrees of severity in all but 1 case; in that one the vegetation were limited to the tricuspid valve and the mural endothelium of the right ventricle. The lesions consisted of miliary infarcts, nonspecific inflammatory changes (both interstitial and perivascular), perivascular fibrosis, actual emboli and thrombi, and petechiae. There was no direct correlation between the severity of the lesions and the presence or absence of myocardial failure but in some instances, although the changes were microscopic, they were so numerous it seemed that they must have had some deleterious effect on myocardial function. In 9 cases treated with penicillin, there was no difference in the incidence or type of lesions except that there were some additional foreign-body granulomata found. Saphir et al. found a high incidence of electrocardiographic changes in patients with bacterial endocarditis who came to autopsy. These changes consisted of generalized low voltage and S-T and T-wave abnormalities which were thought to reflect myocardial disease. In our case, the electrocardiographic pattern was one of "strain" but, nevertheless, there was diffuse myocardial scarring, which probably contributed to the progressive cardiomegaly and eventual failure.

Dr. Taussig: Recently we have seen a 15-year-old girl who had electrocardiographic changes of left ventricular strain following a proved subacute bacterial infection. Despite pronounced physical signs of aortic stenosis, her aortic valve at operation was found to be normal.

Dr. Sissman: Dr. Spencer, would you tell us how the surgical approach to aortic stenosis has changed since this patient was operated upon in 1955?

Dr. Spencer: The operation for congenital aortic stenosis by the introduction of a dilator through the left ventricular wall is probably quite hazardous. The stenosis is of a type that does not readily open with a dilator. Thus, with this approach, the valve may stretch and not be significantly widened; furthermore, the leaflet rather than the commissural line may tear. Also, an incision in the left ventricular wall is probably in itself hazardous. For these reasons, the open approach with extracorporeal circulation in which the stenosis is relieved under direct vision from above through an opening in the aorta is strongly preferred. In this hospital, open operation in 12 cases of congenital aortic stenosis by this method has demonstrated rather uniformly a valve with an opening of 3 to 4 mm. in diameter. The commissures have been present and there has been no calcification in any of the valves. A rather complete opening along the commissures has been made in each instance. All the patients survived operation and showed a marked decrease in the pressure gradient across the valve, although the gradient was not completely relieved. Two patients had regurgitation following the operation. Four patients in the group had sub-aortic stenosis; this too was resected through the aortic opening. The advantage of extracorporeal circulation is that an unhurried approach can be used to open the valve very carefully along the commissures and thus to avoid the production of later regurgitation; the advantage of the supravalvular approach is that the left ventricular wall is intact. The time actually required for the corrective procedure has varied from 10 to 15 minutes. The most encouraging reported series of open operations for congenital aortic stenosis is that of Dr. Henry Swan who operated upon 11 patients, using hypothermia, and had 3 operative deaths. Of the 8 survivors had some regurgitation.

Dr. Sissman: This patient had an unusual lesion of the intima of the aorta above the valve, which has been attributed to his subacute bacterial endocarditis. Recently, we received the specimen of the heart of this patient's grandmother and it showed a congenital lesion of the aorta which was entirely supravalvular. She is the subject of our final presentation. We are greatly indebted
CONGENITAL AORTIC STENOSIS

to Drs. J. Hirschfeld and V. H. Norwood, of Baltimore, for permission to include this case in this conference and for assistance in supplying the details of the clinical and pathologic picture.

Case 4. Mrs. A. H. (Church Home and Hospital, Baltimore, no. 93788), white, the maternal grandmother of case 3 (P.W., Jr.), died in 1956 at the age of 70 years of an acute cerebrovascular accident.

The family history was interesting. The patient was an only living child. Her mother was said to have had congenital heart disease and died during childbirth. One sister died at the age of 16 years because of heart disease which she was supposed to have had since birth. The patient had known "valvular disease" since childhood. Some degree of cardiac decompensation had been present since the age of 60 years but the symptoms had been mild and responded well to treatment with digitalis and mild diuretics such as amphetamine and ammonium chloride. Hypertension had been present since 1946, with systolic levels between 158 and 180 mm. Hg and diastolic levels between 88 and 105 mm. Hg. There was no history of rheumatic fever.

The salient cardiac findings as recorded in 1951 showed a full-sized heart which was enlarged to the left. There was a forceful thrust at the apex and an occasional extrasystole. There were "M^1 and A^1 blows"; the aortic murmur was the louder of the 2, and neither radiated to the axilla. One observer described a marked systolic murmur heard well at both the apical region and the aortic area. A chest x-ray taken in 1954 showed slight cardiac enlargement with a cardiothoracic ratio of 56 per cent. There was lengthening of the left ventricular border; the shadow of the great vessels was narrow. No electrocardiogram was taken and fluoroscopy was not done. Laboratory data showed a negative blood serology, a normal hemoglobin, and normal "routine" chemistries. She died shortly after being admitted to the Church Home and Hospital with symptoms of a cerebrovascular accident.

Autopsy (no. 1872, Church Home and Hospital), performed by Dr. V. H. Norwood, showed the heart to weigh 440 Gm. Both atria were dilated. The foramen ovale was closed and the right atrium and ventricle and the tricuspid valve were normal. The right ventricle was 4 mm. thick at a level 1 cm. below the tricuspid valve. The ventricular septum was intact. At the level of 2 cm. below the mitral valve, the left ventricular wall measured 14 mm. in thickness. The mitral valve was somewhat thickened, but otherwise it was not remarkable. The cusps of the aortic valve appeared to be shortened and thickened but movement was still possible. One to 2 cm. above the aortic valve, an annular ring, which was completely circumferential and extended into the lumen of the aorta, was found. The ring was calcified and of sufficient size to cut down the lumen of the aorta to a diameter of approximately 1 cm. (fig. 7). No sign of the ring could be seen from the exterior. The aorta distal to the ring, in the region of the origin of the innominate, the left internal carotid, and the subclavian arteries, was somewhat dilated but beyond this point it was slightly hypotrophic. The right and left coronary arteries appeared patent throughout their larger ramifications. There were mild atheromatous changes in the aorta in the thoracic and abdominal regions. Microscopically, the heart muscle fibers showed definite hypertrophy but were uniform in size. Sections from the aorta through the congenital ring showed that the ring projected as a shelf-like structure of compact hyaline collagogenous tissue covered by a single layer of epithelial cells and that it made a sharp protrusion into the lumen of the vessel.

DR. TAUSSIG: This case is presented primarily because of its unusual morphology. Although the lesion undoubtedly contributed to the cardiac physical findings, it was prob-
ably not of physiologic significance, for the patient lived a normal life span and died of unrelated causes.

REFERENCES


Caloric intakes of 20 schizophrenic men, who were otherwise healthy, were increased without changing physical activity. Diet was constant and adequate in vitamins and protein. Carbohydrate furnished two thirds and fat one third of added calories. As a result, the proportion of calories due to fat decreased although total fat intake was higher. Calories were thus increased 8 to 39 per cent for 20 weeks. On this regimen weight gain varied from 2.5 to 22.2 Kg. (average 0.5 Kg. per week). Total serum cholesterol increased 20 mg. per cent during the first 5 weeks, but then leveled off even though gain in weight continued. On the other hand, $S_{12-20}$ lipoprotein increased from the tenth to twentieth week even though cholesterol was not changing at this time. There was some increase in circulating plasma and blood volume during the early part of the overeating period. The authors consider that these data support the thesis that serum cholesterol concentration is determined by the fat transport load per unit of circulation imposed on the blood.
Congenital Aortic Stenosis

NORMAN J. SISSMAN, CATHERINE A. NEILL, FRANK C. SPENCER and HELEN B. TAUSSIG

Circulation. 1959;19:458-468
doi: 10.1161/01.CIR.19.3.458

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1959 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/19/3/458.citation

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/