SUBAORTIC STENOSIS*

DAVID G. MASON, M.D., San Francisco, Calif., and
WARREN C. HUNTER, M.D., Portland, Ore.
(From the Department of Pathology, University of Oregon Medical School, Portland, Ore.)

The less frequent cardiac malformations are always of pathologic interest. The example of subaortic stenosis encountered by us is especially interesting because a correct diagnosis was made during life.

REPORT OF CASE

History. R. H. K., a white male, 26 years old, entered the Multnomah County Hospital on March 23, 1935, complaining of cough, weakness and a swollen jaw. At the age of 10 years the patient had scarlet fever. Following this he learned that he had a "blow" in the heart and found he could not run as fast as his playmates. For the past 2 years he had not felt well, had tired very easily and had noted an increased pallor. Three months before admission sweats and chills developed, and for the last 3 to 4 months he had been coughing up about an ounce of sticky sputum a day. Following extraction of several carious teeth 1 month earlier, several small, painful, hemorrhagic spots developed at the tips of the fingers. There was no history of chest pain, nocturia or dependent edema. At the age of 16, after hard physical exertion, he was cyanotic for a few hours.

Physical Examination. Pulse, 110 per minute; respirations, 24; temperature, 101.0° F.; blood pressure 102/80. The important physical findings were as follows: The gums were swollen and bled very easily. There was no cyanosis. The heart was enlarged chiefly to the left. There was no lower sternal propulsion noted. A definite systolic thrill, detected over the second right interspace, was transmitted to the great vessels of the neck and down the left side of the sternum. The pulmonary second sound was not felt. A purely systolic murmur, most evident in the second right interspace, was transmitted to the vessels of the neck and down across the sternum to the left. The heart rhythm was regular. The aortic second sound had a clicking character. It was short, sharp and sounded close to the ear. The peripheral pulse was small and poorly sustained. The spleen was enlarged to palpation.

Laboratory Data. On entrance the following determinations were made: hemoglobin, 38.2 per cent; red blood cells, 2,490,000; color index, 0.72; white blood cells, 7,050; polymorphonuclear leukocytes, 61 per cent; lymphocytes, 21 per cent; staff cells, 12 per cent; sedimentation rate, 110 and 156 mm. (15 and 45 minutes). Evidence of a moderate degree of erythropoiesis was observed in the blood film. The Kolmer and Kahn reactions were negative. Urine: specific gravity, 1,010; yellow; acid, and cloudy with occasional hyaline and granular casts. Pus cells ranged from ++ to +++ and erythrocytes were listed as + in one of two examinations. Streptococci and gram-negative bacilli were found in the blood cultures but were not identified further. Interpretation of the electrocardiographic tracings was "questionable coronary disease."

Clinical Diagnosis. (Dr. Maurice F. Gourley.) Rheumatic aortic endocarditis, probably with considerable calcification; superimposed subacute bacterial endocarditis; subaortic stenosis.

* Received for publication, July 5, 1941.

After a progressive downward course, the patient died 42 days after hospitalization. The temperature varied from 100° to 103° F., occasionally rising to 104°, while the pulse ranged from 90 to 110 per min.

POSTMORTEM EXAMINATION

The postmortem examination was performed 15 hours after death. The body appeared slightly undernourished and pallor was evident. Aside from edema of the ankles there were no external changes. About 150 cc. of transudate were present in the peritoneal cavity. Approximately a liter of similar fluid filled the right pleural cavity while the left contained about 200 cc. The spleen weighed 540 gm. and contained a large, yellow, anemic infarct. A branch of the splenic artery supplying this portion of the spleen was occluded by a septic thrombotic embolus in which there were plates of lime salt like those in the aortic valve. The kidneys were swollen, soft and varied from yellow to bright red. Microscopically, they showed typical subacute proliferative glomerulonephritis with occasional partially infarcted glomeruli attributable to embolic occlusion. The other salient findings were encountered only in the heart.

Heart

Gross Examination. There were approximately 100 cc. of clear fluid in the pericardial sac. The heart had a diameter of 14.5 cm. and weighed 450 gm. The left ventricle, from the base to the apex, measured 10 cm. The thickness of the myocardium on the left and at the base was 2.2 cm., and 0.7 cm. at the apex. The chamber seemed relatively small in comparison to the dilated right cavity. The wall of the right ventricle was 0.7 cm. in thickness. One cm. inferior to the base of the aortic valve ring there was an annular, shelflike stenotic ring consisting of pale, grayish white, smooth, firm tissue covered everywhere by intact endocardium. This band was located 0.6 cm. caudal to the right posterior aortic valve cusp, but approximated and fused with the bases of the anterior and left posterior leaflets (Fig. 1). The ridgelike structure extended 0.6 cm. into the ventricular cavity and was not more than 2 mm. thick at any point. The endocardium between the posterior aortic valve cusps and the subaortic ring was smooth, glistening, opaque and slightly irregular. Here was found an endocardial pocket, or pseudo-valve, with an orifice measuring 0.6 by 0.2 cm. Its ostium lay close to, and opened toward, the base of the left posterior aortic valve cusp. Through it a probe could be passed towards the infra-aortic ring for a distance of 0.5 cm. The ring had a circumference of 2.5 cm. It extended well onto the ventricular surface of the aortic leaflet of the mitral valve but lacked 0.8 cm. of reaching the junction with its chordae tendineae. The anterior leaflet of the aortic valve was normal

save for slight fenestration along its free edge. The right posterior leaflet, however, was quite large, measuring 2.5 cm. in breadth at its commissural attachments and 1.5 cm. in height. The left posterior leaflet was 1.7 cm. in breadth. The large right posterior cusp was thickened and indurated. Adherent to the ventricular surface of the half nearest the left posterior leaflet was a firm, grayish yellow to pink nodular vegetation measuring 0.6 by 0.9 cm. Adherent to the aortic surface of this same cusp (Fig. 1) was a moderate amount of granular, pinkish yellow material. Similar excrescences were present on the ventricular aspect of the left posterior cusp, mainly involving the half of the valve near the commissure separating the two posterior leaflets. The aortic valve ring measured 6 cm. in circumference. Over the intima of the ascending aorta about the orifice of the left coronary artery there was a yellowish to grayish pink sessile elevation measuring 0.6 to 0.7 cm.; otherwise the inner surface of the aorta was smooth and glistening. Except for the extension of the subaortic annular ring onto the mitral, this structure was negative. The mitral valve ring measured 8.0 cm. in circumference. A few small focal areas of fibrosis were seen in the papillary muscles of the mitral valve. There were no associated anomalies other than a slight anatomical patency of the foramen ovale.

Microscopic Examination. The free border of the subaortic band consisted of hyalinized fibrous connective tissue which was largely covered by intact, flattened endothelium. At the junction of the ring with the myocardium there was a single focus of lymphocytes. The endothelium along the inner edge of the aortic valve and over the ventricular surface was replaced by a thin zone of polymorphonuclear leukocytes. Near the base of the valve the tissue was thicker, hyalinized, contained more polymorphonuclear leukocytes caught in fibrin, and showed areas of calcification. Bacterial stains revealed moderate numbers of gram-positive cocci in the central portion of the vegetation and along the free ventricular border of the valve. Sections of the myocardium proper showed a few minute foci of recent necrosis, unaccompanied by inflammation, and slight hypertrophy of the muscle cells. No structures that could be interpreted as active or healed Aschoff bodies were observed.

Comment

Judging from the slight degree of cardiac hypertrophy (450 gm.), maximum thickness of the left ventricular wall (2.2 cm.) and the slight dilatation of the left chambers, the subaortic annular stenotic ring and thickened aortic cusps failed to produce much obstruction or incompetence, yet formed a sufficient barrier to the passage of blood to evoke

the systolic thrill and murmur detected clinically. Another fact indicative of diminution in output was the small and poorly sustained peripheral pulse. Clinically, the clear-cut aortic second sound was interpreted as evidence that this valve was functioning properly. The diagnosis of subaortic stenosis was based upon the evidence of aortic obstruction with retention of the second aortic tone. The presence of a small endocardial "bird's nest" pocket between the aortic valve and the subaortic ring is an additional point against full competency of the valve. Such endocardial pockets are usually found in association with valvular insufficiency, and, as in this instance, open in the direction of the regurgitation. The fact that the nidus lay superior to the subaortic ring and was somewhat recessed makes it difficult to believe that it could have been responsible for the clinical observations or that it contributed appreciably to the changes observed at necropsy. Before the annular ring was sectioned at necropsy the right posterior aortic valve leaflet was found to be directly in line with the probable flow of blood coming from the ventricle. Inasmuch as the largest vegetation was present here, it further strengthens the view that trauma plays a decided etiologic rôle in acquired valvular affections. Clinically it was thought that the bacterial endocarditis was secondary to carious teeth. Both may have been responsible for the associated glomerulonephritis. This latter condition, together with the blood-stream infection, adequately explains the intense hypochromic anemia. The pulmonary edema was thought to be secondary to cardiac failure.

Discussion

The subject of subaortic stenosis has been thoroughly studied by Abbott [1] who, in 1927, was able to catalogue twenty-six instances. Enzer,[2] in 1927, described an anomalous subaortic bicuspid valve, located 3 cm. below the aortic valve. This anomalous valve had chordae tendineae attached to a papillary muscle. This is not a clear-cut instance of subaortic stenosis and is not included in the total list of cases. Sternberg,[3] in 1930, described two cases of this condition. One occurred in a woman, 27 years old, in whom heart disease had been clinically recognized since the age of 5. At the age of 19 years a diagnosis of aortic stenosis was made. The illness that led to her death was a septic sore throat. At necropsy a subaortic annular ring was found 6 to 7 mm. below the aortic valve ring. This fibrous band was only 2 mm. in height and appeared smooth and white. The ventricles and auricles were slightly dilated; no other congenital or acquired lesions were present. The second case was that of a man, 77 years old, who had arteriosclerosis, pulmonary emphysema and, later, obstructive icterus. In this heart a small, irregular, fibrosed band extended from the lower part of

the membranous interventricular septum onto the aortic leaflet of the mitral valve. The aortic valve showed extensive calcification. Sergent, Launay and Imbert,[4] in 1932, reported an example of congenital subaortic stenosis occurring in a male, 21 years old, which was complicated by an acute aortic bacterial endocarditis. In this case the subaortic ridge rose 1 cm. above the level of the interventricular wall.

A recent report,[5] of a boy, aged 14 years, with congenital subaortic stenosis and narrowing of the brachiocephalic venous trunks, is a clinical study and makes no factual mention of subaortic stenosis. This instance consequently is not included in the present review.

Dormanns,[6] in 1939, reported an instance of subaortic stenosis in a male adolescent, 16 years and 9 months of age. This person had had measles as a child. On entering school a systolic heart murmur was discovered accidentally. Subsequent examinations revealed this murmur to have become more intense and audible over the entire cardiac area. Nevertheless he was active in athletics and had no apparent difficulty in various competitive sports. One morning, without significant previous exertion, he suddenly collapsed and died within 2 hours. Autopsy, performed 72 hours after death, revealed a heart weighing 375 gm., which had an anomalous subaortic ring and thickened aortic valve leaflets. The collar-shaped subaortic ring lay 7 mm. below the aortic valve ring and was wedge-shaped in cross section, measuring 3 mm. in width at its base and 3 mm. in height. It was located 21 mm. below the upper border of the aortic valve and 8 cm. from the apex of the left ventricle. The aortic valve ring was 5.5 cm. in circumference and all three of its leaflets were thick, whitish and opaque. The left ventricle was enlarged and dilated. The anomalous ring was formed of fibrous connective tissue lying beneath an intact endocardium. There was no significant cellular infiltration. In the aortic leaflets an increase of fibrous connective tissue was apparent as well as considerable myxomatous tissue beneath the endocardium over the free edge of the valve. Here, as in the stenotic ring, significant cellular infiltration was absent. The myocardium showed no changes. Stating that the examination revealed no evidence of fetal or post-fetal endocarditis, Dormanns explained the subaortic stenosis and the changes in the aortic valve as due to a complicated arrest of development. No other cardiac abnormalities were found.

We feel that our case is a true example of subaortic stenosis, bringing the apparent total to thirty-one.

As to the pathogenesis of subaortic stenosis we can do no better than to quote Abbott:[1] "Two groups are to be differentiated, (*a*) those in which the lesion is associated with other anomalies, in which case a true arrest of development with survival of the bulbus cordis in the aortic

vestibule has occurred, and (*b*) a subaortic stenosis of inflammatory origin due to an infective process setting in [in] early postnatal or late prenatal life." While not susceptible of proof, the appearance of cardiac symptoms following scarlet fever at the age of 10 years, together with the lack of associated cardiac anomalies or evidences of arrested development, are points much in favor of an acquired lesion due to inflammation. Abbott [7] further emphasized that the lesion "does not in itself interfere with cardiac function but is nevertheless of serious significance on account of the fact that it is liable to become the seat of a subacute infective process." Cyanosis, clubbing of the fingers and evidence of aortic valvular stenosis are usually absent. In our case the subaortic ridge escaped but the aortic valve and the aortic intima were the seats of bacterial infection.

SUMMARY

The example of subaortic stenosis forming the basis of this report brings the total of reported cases to thirty-one. There is evidence suggesting, but not proving, that in this instance the original process was acquired. Bacterial infection was present on the aortic valve but not on the subaortic ridge when the patient died at the age of 26 years. The clinical and pathologic aspects of this case are presented and the incidence and characteristics of subaortic stenosis are discussed.

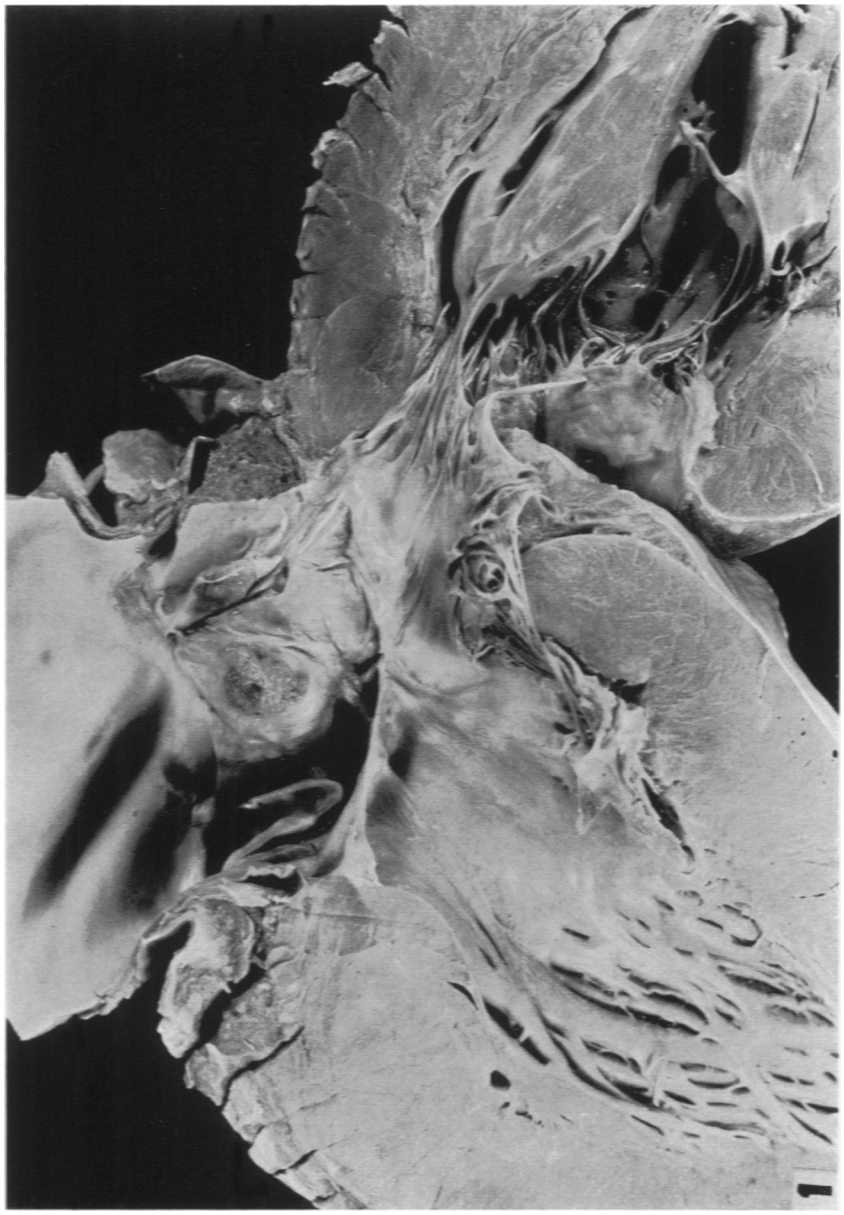
REFERENCES

1. Abbott, M. E. Congenital Cardiac Disease. In: McCrae, Thomas. Osler's Modern Medicine. Lea & Febiger, Philadelphia, 1927, ed. 3, **4**, pp. 743–746.
2. Enzer, Norbert. Anomalous congenital bicuspid subaortic valve of the heart. *Arch. Path.*, 1927, **4**, 966–973.
3. Sternberg, Carl. Ueber infravalvuläre Konusstenosen. Zur Pathologie der Grenze zwischen Herzmuskulatur und Herzskelett. *Verhandl. d. deutsch. path. Gesellsch.*, 1930, **25**, 238–251.
4. Sergent, Émile; Launay, Clément, and Imbert, Fr. Endocardite maligne aiguë greffée sur un rétrécissement sous-aortique congénital. *Paris méd.*, 1932, **83**, 520–522.
5. Faz Tabio, H. Un caso de estenosis sub-aórtica congénita, con estrechez de los troncos venosos braquio-cefálicos. *Bol. Soc. cubana de pediat.*, 1938, **10**, 675–688.
6. Dormanns, E. Zur sogenannten linksseitigen Conusstenose. *Beitr. z. path. Anat. u. z. allg. Path.*, 1939, **103**, 235–244.
7. Abbott, M. E. Atlas of Congenital Cardiac Disease. The American Heart Association, New York, 1936, p. 24.

DESCRIPTION OF PLATE

PLATE 59

FIG. 1. Photograph of left ventricle and aortic valve showing clearly the shelflike, annular stenotic ring below the aortic valve. The dark, oval, ragged areas on the right posterior aortic valve cusp and aortic intima are ulcerative vegetations.



Mason and Hunter Subaortic Stenosis