# DIAGNOSIS OF AORTIC STENOSIS

## BASED ON A STUDY OF 25 PROVED CASES

BY

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Aortic stenosis is an important clinical entity which often escapes recognition in life. Reliance upon gross physical signs of the valvular lesion will allow diagnosis of only one-quarter of the cases to be found at necropsy (Parkinson, 1949). Routine necropsies also show that in one-half of the cases in which stenosis is present there is no other valvular lesion, and that most patients with *lone* aortic stenosis eventually die from heart failure or infective endocarditis as a result of their valvular disease.

This paper, based on a study of 25 proved cases, shows that aortic stenosis is common, and that a correct clinical diagnosis should be possible in a high proportion of cases. It is also submitted that lack of familiarity with the natural history of aortic stenosis as well as the demand for too rigid diagnostic criteria is chiefly responsible for the common failure to diagnose the condition.

### Material

Amongst 2,245 routine necropsies performed at the Postgraduate Medical School of London during the seven-year period ending in February, 1950, aortic stenosis was found in 45 cases (1.8%). Of these 45 cases calcareous aortic stenosis was the only significant valvular lesion in 25; in the remaining 20 all degrees of aortic stenosis were found—16 were complicated by mitral stenosis, three by gross aortic incompetence, and one by tricuspid stenosis and mitral stenosis.

As it was thought that the clinical problem of aortic stenosis could be studied satisfactorily only when uncomplicated by other valvular lesions, detailed analysis has been confined to the first group of 25 cases (Table IV, overleaf). The degree of stenosis at necropsy was graded as mild, moderate, or severe, depending upon whether the aortic orifice would admit the index finger, a finger-tip, or less than a finger-tip respectively (Kumpe and Bean, 1948).

Of these 25 cases significant aortic stenosis was a coincidental necropsy finding in three in which death was due to non-cardiac causes. Twenty-two patients first came under medical observation on account of cardiac or cerebral symptoms; 15 died from heart failure consequent upon the valvular lesion and four from infective endocarditis (Table I).

TABLE I.—*Cause of Death in 25 Cases of Aortic Stenosis*

| | No. of Cases |
|---|---|
| Heart failure from aortic stenosis | 15 |
| Heart failure from aortic stenosis and myocardial infarction | 2 |
| Bacterial endocarditis | 4 |
| Heart failure | 3 |
| Cerebral embolism | 1 |
| Rupture of dissecting aortic aneurysm | 1 |
| Bronchopneumonia | 2 |
| Cerebral metastasis from bronchial carcinoma | 1 |

### Clinical History

The majority of patients had spent a long life of activity and usefulness untroubled by heart complaints until the sixth or seventh decade. The onset of symptoms was commonly sudden (Tables II and III), and was attended by a serious prognosis.

The total duration of cardiac symptoms was less than four years in four-fifths of the cases. Valvular disease had been diagnosed 16 to 30 years before symptoms appeared in four patients; all lived active lives in the interim, and two were professional footballers. The extent of calcification of the aortic valves at necropsy indicated that significant stenosis had been of long standing in every case in the series. There was thus good reason to believe that most of them had enjoyed long periods of good health in the presence of unsuspected valvular disease.

TABLE II.—*Presenting Symptoms in 22 Cases of Aortic Stenosis*

| Symptom | No. of Cases |
|---|---|
| Exertional dyspnoea | 11 |
| Nocturnal dyspnoea | 3 |
| Cardiac pain | 3 |
| Syncope | 2 |
| Giddiness | 2 |
| Stroke | 1 |

TABLE III.—*Frequency of Symptoms in 22 Cases of Aortic Stenosis*

| | No. of Cases |
|---|---|
| Exertional dyspnoea | 19 |
| Nocturnal dyspnoea | 10 |
| Cardiac pain | 10 |
| Palpitation | 4 |
| Attacks of pallor and sweating | 4 |
| Cerebral symptoms: | |
| Giddiness | 8 |
| Syncope | 4 |
| Mental confusion | 4 |
| Fits | 2 |
| Maniacal violence | 2 |
| Stroke | 1 |

Characteristic clinical histories demonstrate how the patients first came under medical observation, as well as the course of the terminal illness.

### Case 1: Lassitude and Giddiness

A 70-year-old baker had had valvular disease diagnosed 16 years previously. In spite of this he had enjoyed good health until three months before admission, when he complained of gradual failure of vision and increasing lassitude. Two months later postural giddiness set in. Ten days before admission he experienced a sensation of "water streaming down in the air" past his head. This symptom never left him and kept him awake at night. On admission there were classical signs of aortic stenosis (Table IV). Heart failure was absent and the electrocardiogram showed sinus rhythm and left bundle-branch block. His symptoms improved in hospital, but persisted until the final admission a year later. Ten days previously, while taking a meal, a small piece of bread stuck in his throat, and he gasped for breath. Severe breathlessness persisted from this moment. Unable to lie flat, he remained in a chair until admission.

On examination there was severe congestive heart failure. The pulse rate was 32 a minute, and the electrocardiogram confirmed complete heart-block. The aortic systolic thrill previously felt could not be detected terminally. Heart failure showed little response to treatment, and he became mentally confused, with lucid intervals during the day. Death took place suddenly while drinking a cup of tea, two weeks after admission and 15 months after the first symptom.

Post-mortem examination revealed acute pulmonary oedema and severe aortic stenosis with gross calcification of the cusps. Calcium deposits encroached upon the interventricular septum, and microscopically there was myocardial fibrosis underlying the calcification in the neighbourhood of the bundle of His.

### Case 4: Cardiac Pain and Giddiness

A 63-year-old foreman had been well until six months previously, when he noticed pain in the throat and upper chest, brought on by exercise and relieved by rest. Giddiness on exertion was also troublesome. On the day of admission, while walking in the street, he bent down to do up a shoelace, and suddenly became extremely short of breath. Struggling to his doctor, he began coughing up blood-stained sputum. On admission a diagnosis of hyper-

TABLE IV.—*Clinical and Post-mortem Findings in 25 Cases of Calcareous Aortic Stenosis*

| Case | Sex | Age | No. of Admissions | History of Rheumatism | Total Length of History | Pulse: Rate/min. | Pulse: Rhythm | Pulse: Volume | Apex Beat Displaced | Thrill | Systolic Murmur: Aortic | Systolic Murmur: ∧ ≡ ∨ | Systolic Murmur: Mitral | Diastolic Murmur | Aortic 2nd Sound | Triple Rhythm | Blood Pressure | Degree of P.M. Stenosis | Weight of Heart (g.) | Clinical Diagnosis |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 1 | M | 70 | 2 | − | 15 mths | 32 | Reg. | Small | ++ | Aortic | ++ | ∨∧ | ++ | ++ | Absent | − | 170/60 | Severe | 690 | Aortic stenosis. Complete heart-block |
| 2 | M | 63 | 2 | + | 5 ,, | 100 | ,, | Collapsing | − | ,, | − | − | − | − | − | − | 120/60 | Mod. | 540 | Infective endocarditis Aortic valve |
| 3 | M | 65 | 1 | − | 5 yrs | 100 | ,, | Normal | + | − | + | − | + | − | (I & II) (distant) | − | 150/110 | Mild | 490 | Chronic bronchitis. Emphysema. Heart failure |
| 4 | M | 63 | 2 | − | 9 mths | 88 | ,, | ,, | − | − | + | − | + | + | Normal | − | 105/75 | Severe | 750 | Calcareous aortic stenosis |
| 5 | F | 72 | 1 | − | 1 yr | 130 | Aur. fib. | Small | + | Aortic | + | ∧ ≡ | + | + | − | − | 130/85 | Mod. | 780 | Thyrotoxic heart failure |
| 6 | M | 57 | 1 | − | 3 mths | 138 | ,, | Bounding | + | − | + | ∧ ∨ | + | + | (I & II) (distant) | − | 134/105 | Severe | 450 | Aneurysm of aorta. Aortic stenosis |
| 7 | M | 65 | 1 | − | 2 wks | 60 | Reg. | Small | − | − | + | − | + | + | − | − | 105/75 | Mild | − | Coronary thrombosis |
| 8 | M | 53 | | − | − | 80 | ,, | Normal | No | − | + | − | + | − | − | − | 120/80 | Mild | 300 | Carcinoma of stomach |
| 9 | M | 78 | | − | − | 88 | ,, | Full | No | − | + | − | + | − | (I & II) (distant) | − | 115/75 | Mod. | 300 | Aortic incompetence. Bronchopneumonia |
| 10 | M | 49 | 2 | − | 4 yrs | 82 | ,, | Anacrotic | + | Aortic and mitral | + | ∨∧ | + | + | Dim. | + | 120/80 | Mod. | 730 | Aortic stenosis. Heart failure |
| 11 | M | 77 | 1 | − | 10 days | 160 | Aur. fib. | − | − | − | Nil | − | Nil | Nil | − | − | 108/85 | Mild | 350 | Hypertensive heart disease |
| 12 | M | 71 | 1 | − | 7 wks | 80 | Reg. | Normal | + | Aortic | + | ∧ ≡ | + | + | − | − | 130/85 | Mod. | 450 | Aortic stenosis. Anaemia |
| 13 | M | 43 | 1 | + | 5 ,, | 102 | ,, | Collapsing | + | Aortic and mitral | + | − | + | + | − | − | 140/70 | ,, | 660 | Aortic stenosis and incompetence. Bacterial endocarditis |
| 14 | M | 61 | 2 | − | 6 mths | 132 | ,, | Dicrotic | + | Aortic | + | ∧∧∧ | + | + | Absent | − | 110/90 | Severe | 765 | Aortic stenosis. Heart failure |
| 15 | F | 75 | 1 | + | 3 ,, | 100 | Aur. fib. | − | + | − | + | − | + | + | − | − | 120/65 | Mod. | 740 | Rheumatic heart disease. Heart failure |
| 16 | F | 78 | 2 | − | 1 mth | 84 | ,, | Dicrotic | + | Mitral | + | − | + | + | Absent | − | 120/70 | ,, | 560 | Aortic stenosis. Bacterial endocarditis |
| 17 | M | 72 | 2 | − | 14 wks | 80 | Reg. | ,, | + | − | Nil | Nil | Nil | Nil | (I & II) (distant) | − | 170/100 | Mild | 535 | Hypertensive heart failure |
| 18 | M | 52 | 3 | + | 12 mths | 84 | ,, | − | + | Aortic | + | − | + | + | Loud | + | 110/60 | Mod. | 1,020 | Aortic stenosis and incompetence. Mitral stenosis |
| 19 | M | 64 | 1 | − | 6 wks | 80 | ,, | Small | + | ,, | + | ∧∨∨∧ | + | + | (I & II) (distant) | + | 120/60 | ,, | 565 | Aortic stenosis. Syphilitic aortitis |
| 20 | F | 74 | 2 | − | 7 ,, | 84 | ,, | Anacrotic | + | − | + | − | + | + | Dim. | + | 170/140 | ,, | 575 | Hypertensive heart failure |
| 21 | F | 56 | 2 | − | 2 yrs | 112 | ,, | − | + | Mitral only | + | − | + | + | Absent | − | 100/85 | Severe | − | Calcareous aortic stenosis. Heart failure |
| 22 | F | 78 | 2 | − | 9 mths | 118 | ,, | Full | No | − | + | − | + | + | Normal | − | 115/75 | Mild | 360 | Aortic stenosis. Heart failure |
| 23 | M | 68 | − | − | − | 80 | ,, | − | + | − | + | − | − | + | − | − | 180/80 | ,, | − | Carcinoma of bronchus. Aortic stenosis |
| 24 | M | 76 | 1 | − | 4 yrs | 75 | ,, | Normal | No | − | + | ∨∧ | + | + | (I & II) (distant) | − | 140/85 | Mod. | 574 | Coronary artery disease. Heart failure |
| 25 | M | 62 | 3 | − | 4 ,, | 85 | ,, | Small | − | − | + | − | + | + | Normal | − | 130/80 | ,, | 470 | Carcinoma of bronchus |
| Totals or averages | M 19<br>F 6 | 63·6<br>72·2 | 1·8 | 4 | 14 mths | 94 | Aur. fib.<br>5 | Small 9<br>Full 5 | 16 | Aortic 9<br>Mitral 4 | 21 | | 21 | 12 | Absent 4<br>Dim. 2<br>Normal 3 | 4 | 130/80 | Mild 7<br>Mod. 13<br>Severe 5 | 389<br>597<br>664 | Aortic stenosis 13 |

+ = Present. − = Not recorded. P.M. = Post-mortem examination. >, =, or < = Greater than, equal to, or less than, respectively. Aur. fib. = Auricular fibrillation.

tensive left ventricular failure was made. Response to treatment was satisfactory, and he was discharged within a month. On readmission two months later, little history could be obtained on account of the patient's confused mental state. He told of four weeks' nocturnal breathlessness and severe exertional chest pain, radiating down the left arm.

On examination there was little evidence of heart failure, but respiration was of the Cheyne–Stokes type. Aortic stenosis was diagnosed on the basis of a rough aortic systolic murmur (Table IV). The patient subsequently became violent, abusive, and mentally confused, though he had remarkably lucid intervals, and died six days after admission, nine months after his initial symptoms. The mode of death was curious: there was no complaint of breathlessness or any true orthopnoea. There was no peripheral oedema, and the jugular venous pressure was not raised. Mental confusion was the sole manifestation of failure; yet the patient was able to recognize his own Cheyne–Stokes respiration by remarking that he breathed "too much in fits and starts."

Post-mortem examination showed severe aortic stenosis and acute pulmonary oedema.

## Case 12: Syncopal Attacks

A healthy old-age pensioner of 71 years, while sitting in a public-house, fell unconscious without warning. There was no fit; recovery of consciousness was rapid, and he soon felt well enough to go home. One week later, while sitting in front of the fire, he had another similar fainting attack. He recovered consciousness after two minutes and passed the incident off as "a joke." Subsequently he felt well and was admitted for investigation of the syncopal attacks, which were his only complaint. A clinical diagnosis of aortic stenosis and anaemia without heart failure was made (Table IV). Nine days after admission and seven weeks after the first syncopal attack he became mentally confused, with temporary cyanosis followed by pallor, urgent dyspnoea, and moist sounds throughout both lung fields. Heart failure, which proved to be terminal, showed no response to treatment, and death took place from left ventricular heart failure 30 hours later.

Post-mortem examination confirmed the clinical findings, showing a moderate degree of aortic stenosis.

## Case 13: Infective Endocarditis

A 43-year-old accountant was admitted to hospital on account of weakness and chest pain. Valvular heart disease had been diagnosed at the age of 10 years, following rheumatic fever. Subsequently he had enjoyed good health apart from recurrent winter bronchitis until one month before admission. After removal of septic teeth at this time he noticed increasing lassitude and breathlessness on exertion. Two weeks later sharp back pain, which lasted a week, was followed by persistent "gnawing" pain in the chest, radiating down the inner and outer aspects of the left arm.

On admission continuous chest pain was his chief complaint. A clinical diagnosis of bacterial endocarditis developing upon aortic valve disease was made (Table IV). Heart

failure was absent at first, but the electrocardiogram showed changes consistent with posterior cardiac infarction. The infection was treated with sulphonamides without response, and rapidly progressive and unresponsive heart failure supervened. Ten days after admission and five weeks after the first symptom there was sudden collapse, with pallor, sweating, and intense dyspnoea, and death took place in six hours.

Post-mortem examination confirmed pulmonary oedema as the cause of death. Friable vegetations were confined to a moderately stenosed and incompetent aortic valve, but a mycotic aneurysm was found arising from the anterior sinus of Valsalva. This had compressed the left coronary artery, with consequent recent anterior myocardial infarction.

### Case 19: Left Ventricular Heart Failure

A previously healthy 64-year-old bricklayer developed a nocturnal cough which was relieved by sitting up in bed. He also noticed exertional dyspnoea during the day: "If I walked 100 yards it felt as though I had run the same distance." His nights became progressively more disturbed by cough, breathlessness, and, latterly, by upper-abdominal pain. He was admitted to hospital after an attack of severe abdominal pain and breathlessness during the early hours of the morning. Congestive heart failure was not evident at this time, and the signs in the chest were regarded as due to pneumonia, although the chest film suggested pulmonary oedema. Aortic stenosis was recognized clinically (Table IV); but treatment was unsuccessfully directed towards bronchopneumonia, which was strongly suggested on clinical grounds. Five days after admission and six weeks after the first symptom of illness the patient died suddenly from acute pulmonary oedema while using a bedpan.

Post-mortem examination showed moderate aortic stenosis, acute pulmonary oedema, and large bilateral pleural effusions, but no evidence of pulmonary infection.

## Physical Signs

*Pulse.*—The rhythm was regular except in five cases with auricular fibrillation. Of the 20 cases in which the volume was recorded a small pulse was noted in less than half; an anacrotic pulse in five cases was always good evidence of moderate or severe stenosis, but in an equal number, including two cases with advanced stenosis, the pulse was full or collapsing. In one-third of the cases correctly diagnosed in life the pulse was recorded as normal or full.

*Apex Beat.*—The cardiac impulse was displaced to the left in two-thirds of the cases; in five it was heaving, and was twice noted to be of a slowly rising character. In one-fifth of the series the apex beat was in a normal position.

*Aortic Systolic Thrill.*—A systolic thrill felt in the second right intercostal space in one-third of the cases invariably indicated moderate or severe stenosis. A systolic thrill was confined to the mitral area in one case with a slit-like valvular opening, 4 by 1 mm. In a similar case, in which the aortic orifice was described as of "pinhole" size, a basal systolic thrill was hardly palpable. Two cases with moderate stenosis had thrills equally intense at apex and base, and in a third case it was felt only in the third left intercostal space. A correct clinical diagnosis was made in five cases without a basal systolic thrill.

*Aortic Systolic Murmurs.*—A loud harsh aortic systolic murmur conducted to the neck was heard in four-fifths of the cases. In an equal proportion a harsh mitral systolic murmur was recorded. In six cases, of which five had advanced stenosis, the mitral murmur was loud and rough, exceeding in intensity the signs at the base. Widespread systolic murmurs were noted in one-fifth of the series; in two cases no murmurs were heard at any time throughout the illness although they were repeatedly sought. A correct clinical diagnosis was made in three cases in which a harsh aortic systolic murmur was the only physical sign of the valvular lesion.

*Aortic Diastolic Murmurs.*—These were recorded in just under half of the series, and were most obvious when bacterial endocarditis involved the aortic cusps. Diastolic murmurs, sometimes noted as harsh, but usually short and soft, were heard in both the cases of extreme stenosis.

*Aortic Second Sound.*—The aortic second sound was absent in four patients, in three of whom stenosis was severe; in two with moderate stenosis it was diminished; and in three, including one with gross stenosis, it was normal. Frequently both heart sounds were equally diminished, but in one case of moderate stenosis the aortic second sound was accentuated.

*Blood Pressure.*—Blood-pressure readings did not conform to the expectation that both systolic and pulse pressures should be low (Christian, 1931). The average reading for the series was 130/80 mm. Hg. Four cases with a pulse pressure less than 30 mm. Hg had severe stenosis. Otherwise blood-pressure readings were of little value in diagnosis. Wide pulse pressures in the absence of significant aortic incompetence were noted, the highest figure of 110 mm. Hg (Case 1) being recorded in the presence of extreme stenosis. Diastolic blood pressures of 100 mm. Hg or more were found in four cases. It was important to distinguish, retrospectively, between benign hypertension and hypertension contingent upon left ventricular failure in this series. This may not always be possible when a case of aortic stenosis is seen for the first time in acute heart failure. Normal systolic and diastolic pressures greatly increased when terminal heart failure supervened in two patients, and in another the sudden onset of left ventricular failure raised the blood pressure from 170/140 to 250/160 mm. Hg (confirmed by repeated observations).

*Triple Heart Rhythm.*—This was heard in six cases; in four it persisted throughout the final illness and in two it was transient.

## Electrocardiography

Limb lead tracings, and usually chest leads, were recorded in 20 cases. In no case was a normal tracing seen (Table V). Left ventricular preponderance was common, but its detection was not always possible from the standard leads alone, especially in the presence of bundle-branch block. Myocardial infarction with necropsy control was diagnosed once. A cardiographic diagnosis of pericarditis was later confirmed at necropsy in one case to be due to intrapericardial haemorrhage from a ruptured aorta. Only once was aortic stenosis diagnosed on the basis of characteristic S–T changes, but the clinical signs of the lesion were obvious.

TABLE V.—*Electrocardiographic Findings in 20 Cases of Aortic Stenosis*

| | No. of Cases |
|---|---|
| Left ventricular preponderance: | |
| Slight | 4 |
| Moderate | 6 |
| Severe | 3 |
| Auricular fibrillation | 4 |
| Prolonged P–R interval | 4 |
| Bundle-branch block (left 5, right 1) | 6 |
| Complete heart-block | 1 |

### Radiography

Every patient was submitted to radiological study. This was often confined to portable chest films, as many were too ill for more detailed investigation. Cardiac enlargement was reported in 21 cases and isolated or disproportionate left ventricular enlargement in 18 of these. Pulmonary congestion or pleural effusion was a usual accompaniment of cardiac enlargement. Aortic unfolding was the rule and frequently this was advanced. Associated heart failure and unfolding of the aorta obscured the classical radiological appearances of aortic stenosis in every case.

Gross aneurysmal dilatation of the ascending aorta was observed in two patients. In Case 6, at post-mortem examination, a syphilitic aneurysm was found to be associated with extreme aortic stenosis. Histological examination failed to demonstrate either a syphilitic or rheumatic basis for the stenosis. In Case 7 a dissecting aneurysm of the aorta, which finally ruptured into the pericardial sac, was associated with moderate aortic stenosis. Microscopical examination revealed medial necrosis of the aorta.

Calcification of the aortic valves, demonstrated in three of the five cases in which it was specifically sought and observed only by fluoroscopy, permitted a certain clinical diagnosis of aortic stenosis in one case in which the only other physical sign of the lesion was an aortic systolic murmur.

### Clinical Course

A rapidly downhill course with death from heart failure was the rule, and was seen in 22 cases. In this group the average survival was 14 months from the first symptom of illness. In the final phase heart failure was peculiarly resistant to treatment, and death took place within three weeks of hospital admission in more than two-thirds of the cases. Nearly half the patients died on the first admission, and only two survived a second stay in hospital.

Sweating attacks associated with striking pallor were seen in four cases during the terminal stages of heart failure. Mental confusion, epileptiform convulsions, or maniacal violence was a terminal event in seven cases. Seven patients, all of whom were in heart failure, died suddenly or unexpectedly within seconds or minutes. Three of these gave a clear history of anginal pain; acute pulmonary oedema was confirmed at necropsy in five, and in one there was recent myocardial infarction.

*Frequency of Correct Clinical Diagnosis.*—Aortic stenosis was recognized in life in 13 of the 25 cases. The frequency of correct diagnosis was related to the degree of stenosis found at necropsy (Table IV). Clinical diagnosis was always correct when stenosis was severe; 6 of the 13 moderately severe cases were diagnosed in life, while five of the seven mild cases escaped clinical detection.

## Discussion

### Incidence of Aortic Stenosis

This study confirms the frequent observation that the maximum incidence of symptoms due to an isolated aortic stenosis is in the sixth and seventh decades and that males are affected about three times as often as females (Table IV). The incidence of all cases of aortic stenosis (with and without other valvular lesions) in the present series agrees precisely with the findings of McGinn and White (1934) in 6,800 routine necropsies (1.8%). Amongst 630 patients dying from rheumatic valvular disease, Clawson *et al.* (1938) found that calcified nodular deformity of the aortic valve constituted 41% of valvular deformities and 31.5% of all rheumatic heart disease. Finally, Kumpe and Bean (1948) identified *isolated* calcareous aortic stenosis in 0.72% of 15,016 necropsies, comparable to the present findings of nearly 1%.

Compared with congenital heart disease aortic stenosis is common. Gelfman and Levine (1942), analysing 34,023 necropsies, discovered significant congenital cardiac defects after the age of 2 years in 0.53% of cases. These figures suggest that after infancy aortic stenosis, both isolated and combined with other valve lesions, is more than three times as common as congenital heart disease.

### Clinical Features

Karsner and Koletsky (1947) have put forward convincing pathological evidence that the aetiology of aortic stenosis is nearly always rheumatic. A past history of rheumatism, however, contributes even less to the diagnosis of aortic stenosis than it does in mitral valvular disease. This is indicated by a rheumatic history in less than one-sixth of the present series, and by the extreme variations in the incidence of past rheumatism in other published figures: 66% (Kumpe and Bean, 1948); *nil* (Kiloh, 1950).

The degree of necropsy calcification which has developed upon a still older scarring process in the aortic valve suggests a very prolonged and slow development. This is borne out by the not infrequent recognition of aortic stenosis in early middle life—a symptomless course, lasting for decades, with frequent termination after the age of 60. When symptoms finally develop the earliest complaints are commonly exertional or nocturnal dyspnoea, cardiac pain, giddiness, or syncope. Mental disorder may appear early (Kumpe and Bean, 1948). Breathlessness, whether exertional or paroxysmal, is often precipitated by unaccustomed exertion or emotion, and tends to progress rapidly. Cardiac pain, which is common, may be independent of any associated hypertension, aortic incompetence, or coronary vascular disease (Contratto and Levine, 1937). The present study confirms the observation of Kumpe and Bean (1948) that cardiac pain is an index of advanced stenosis and suggests that patients afflicted by it are liable to sudden death.

Attacks of giddiness or syncope assume special diagnostic importance when they follow some unaccustomed exertion. Syncope, however, may occur at rest and, rarely, may be precipitated by the development of complete heart-block complicating the valvular lesion. Stokes–Adams attacks may occur (Contratto and Levine, 1937). Sudden death, so common during the final illness in this series, may be the only clinical manifestation of aortic stenosis (Marvin and Sullivan, 1935; Dry and Willius, 1939).

The high incidence of infective endocarditis in this series (4 cases out of 25) agrees closely with the findings of Contratto and Levine (1937). Early diagnosis of symptomless aortic stenosis is therefore of the utmost importance in that steps may be taken to reduce the very real risk of infective endocarditis to which these patients are liable.

On the basis of the classical signs of (1) a slowly rising pulse, (2) an aortic systolic murmur and thrill, and (3) an absent aortic second sound, only one-sixth of the present cases would have been recognized clinically. This fits in with the statement by Wiggers (1944), that so long

as compensation persists the aortic orifice must be reduced to less than one-quarter of its natural size before systolic discharge, blood pressure, or pulse form is affected.

Lewis (1940) taught that a slowly rising pulse was an essential accompaniment of any significant degree of aortic stenosis. The findings in this series and the observations of McGinn and White (1934) are opposed to the general truth of this teaching. Even allowing for the fact that the art of detecting minor changes in pulse volume at the bedside is fast being lost, the present study has shown that advanced aortic stenosis may be accompanied by a full or even a collapsing pulse.

A systolic thrill in the second right intercostal space was the most reliable sign of aortic stenosis in this series. It is well known that in the presence of heart failure the systolic thrill of aortic stenosis may become less intense or even disappear. In lesser degrees of aortic stenosis, and especially in the presence of heart failure, a thrill may be detected only by placing the outstretched hand firmly against the praecordium during expiration with the patient sitting well forward.

The finding of a thrill at or near the mitral area in nearly one-sixth of the reported cases emphasizes a variant in aortic stenosis which is often misinterpreted at the bedside and is rarely mentioned in clinical teaching.

A thrill felt in the neck should be accepted as evidence of aortic stenosis only when it is long and is accompanied by a rough systolic murmur in the aortic area. A short thrill in the root of the neck may occur in health, especially in young people. In the presence of significant incompetence the systolic thrill of aortic stenosis may reveal itself by a carotid shudder visible in the neck. When this sign is present it is by itself reliable evidence of the dual lesion (Evans and Lewes, 1945).

A loud harsh systolic murmur in the second right intercostal space, conducted into the neck, was the commonest clinical sign of aortic stenosis. It was a careful appraisal of this murmur that led to a correct clinical diagnosis in four cases without a systolic thrill. It is important to emphasize how often the systolic murmur of aortic stenosis is audible over a wide area and how commonly the murmur is loudest in the mitral area, to which it may occasionally be confined. The recognition of this fact should help to guard against the unwarranted diagnosis of mitral incompetence in such cases.

Aortic diastolic murmurs may be anticipated in about half the cases of aortic stenosis. The discovery of a harsh aortic diastolic murmur in cases of valvular disease should suggest the possibility of aortic stenosis as well as of incompetence.

The appraisal of a harsh basal systolic murmur in the absence of any of the expected signs of aortic stenosis may offer a difficult diagnostic problem. The teaching of Sir Thomas Lewis (1940), however, that it is unpardonable to diagnose aortic stenosis on the basis of an aortic systolic murmur is not generally accepted (McGinn and White, 1934; Contratto and Levine, 1937; Willius, 1939; White, 1944; Evans, 1948). The statement of White (1944) that the diagnosis can be made on the aortic systolic murmur alone in a patient without aortic dilatation or hypertension, provided the murmur is loud and harsh, is supported by this study. There is abundant evidence that aortic stenosis may declare itself by an aortic systolic murmur as the only physical sign; for both Willius (1939) and Baker *et al.* (1943) emphasize that patients seen one or two decades before death from aortic stenosis had basal systolic murmurs, often without symptoms and sometimes dismissed as innocent or functional. All cases of suspected heart disease, therefore, with rough systolic murmurs, especially at the aortic area, should be carefully examined with aortic stenosis in mind, and submitted to fluoroscopy for the detection of calcification of the aortic valve.

An absent second sound in the aortic area is good evidence of advanced stenosis but it was not a common finding in this study. Evaluation of a diminished second sound at this site is often difficult, especially in the presence of heart failure, when the pulmonary second sound may be accentuated. Since a normal and even accentuated second sound may accompany established stenosis, failure to detect any audible abnormality other than a systolic murmur should not weigh too heavily against the diagnosis.

The unreliability of blood-pressure readings as a diagnostic guide in the study is also emphasized by the finding by McGinn and White (1934) of "an unexpectedly high figure" of 60 mm. Hg for the mean pulse pressure in their series and by the observation of Kumpe and Bean (1948) that in their cases blood-pressure readings were so different from the classical description that sometimes they actually biased the observer against the correct diagnosis.

Triple rhythm, rarely mentioned in the literature, is reputed to be exceedingly uncommon. Evans (1948) states that he has not met with it in aortic stenosis, and Kumpe and Bean (1948) record "gallop rhythm" in only 5 of their 107 cases. The finding of unquestionable triple rhythm in four cases in the present series is therefore of importance in that this sign should not influence the observer against a diagnosis of aortic stenosis. Possibly this increased incidence reflects a greater interest in triple rhythm in recent years, stimulated by the work of Evans (1943).

### Special Investigations

Electrocardiography contributed little to the diagnosis of the valvular lesion, although several tracings suggested a diagnosis of aortic stenosis retrospectively and showed the distinctive graph described by Evans (1948). Electrocardiography was of most value in elucidating rhythm disorders and in revealing conduction defects, which were frequent.

Auricular fibrillation is often regarded as rather rare in aortic stenosis (Contratto and Levine, 1937; Dry and Willius, 1939; Scherf and Boyd, 1948); yet it was present in five of the present cases. A comparable incidence is noted by Kumpe and Bean (1948). It is considered, therefore, that the presence of auricular fibrillation should not weigh too heavily against the diagnosis of isolated aortic stenosis on the one hand, or demand too strongly the coexistence of mitral stenosis as an explanation of the rhythm disorder on the other. The finding of left bundle-branch block in one-quarter of the cases supports the view of Berk and Dinnerstein (1938) that left bundle-branch block is found more often in aortic stenosis than in any other condition, with the possible exception of hypertensive disease.

In this series radiology was of limited diagnostic value. There is no doubt, however, that routine screening of the heart and a careful search for calcification of the aortic valve, using a proper technique, is a most valuable diagnostic procedure (Sosman and Wosika, 1933; Parkinson, 1949). Sosman (1939) has demonstrated the remarkable precision that may be achieved

in the diagnosis of intracardiac calcification, and Parkinson (1949) states that calcification of the aortic valve is so rarely absent in aortic stenosis that its demonstration radiologically is comparable in diagnostic worth with enlargement of the left auricle as evidence of mitral stenosis. Calcification of the aortic valve was observed in 26 of 32 cases of aortic stenosis reported by Contratto and Levine (1937), who state that this fluoroscopic sign was always indicative of significant aortic stenosis. The value of tomography has been shown by Davies and Steiner (1949), who report 14 cases of aortic stenosis all diagnosed by tomographic record of the calcified valve.

If every case of suspected heart disease with a rough aortic systolic murmur were screened by the proper technique, or submitted to tomography, few cases of established aortic stenosis would be missed clinically. Routine investigation in this manner wou'd also serve to prevent unwarranted cardiac invalidism through the making of a diagnosis of aortic stenosis in such conditions as hypertension and aortic sclerosis, which are often accompanied by an aortic systolic murmur.

### The Terminal Illness

The onset of symptoms, whether cardiac or cerebral, carries a grave prognosis. The course of the illness is then relentless; the response to treatment is poor, and death follows from heart failure within two years of the first symptom. The average survival following the first symptom in this series (14 months) is comparable with the findings of Contratto and Levine (1937) that the duration of life after the onset of dyspnoea was 23 months, of syncope 9.1 months, and of oedema 9.3 months. Rapidly progressive left ventricular failure soon follows symptoms of giddiness, syncope, cardiac dyspnoea, or anginal pain in most cases, although syncopal attacks sometimes occur years before the terminal illness. Patients rarely survive a second attack of acute heart failure. This march of events, so clearly demonstrated by the present series, confirms the experience of McGinn and White (1934) and contrasts sharply with the usual progress of heart failure from mitral stenosis or hypertension.

Fami'iarity with the natural history of aortic stenosis, once symptoms develop, may assist diagnosis, especially when the expected signs of the valvular lesion are lacking. In the absence of gross coronary artery disease, hypertension, and aortic incompetence, the rapid development, in an elderly subject, of heart failure which is peculiarly resistant to treatment should suggest the diagnosis, especially if the rhythm be normal. If in addition there is a history of syncopal attacks and nocturnal dyspnoea, and a harsh basal systolic murmur is present, aortic stenosis is likely. During the terminal phase the occurrence of sweating attacks and pallor, which may be associated with mental confusion and maniacal symptoms, or even the manner of death, may give a clue to the diagnosis when this has remained in doubt for want of better evidence.

Sudden death in hospital practice is perhaps too often attributed to pulmonary embolus or cardiac infarction. The recognition that one in every five cases of aortic stenosis ultimately dies suddenly or unexpectedly may permit a tardy but correct clinical diagnosis in some cases of obscure heart disease.

### Accuracy of Clinical Diagnosis

A correct clinical diagnosis in 52% of this series compares favourably with corresponding figures of 52.9% (Thompson and Levine, 1936), 33% (McGinn and White, 1934), 25% (Karsner and Koletsky, 1947), and 24% (Kumpe and Bean, 1948). The present figure, although encouraging, is by no means satisfactory.

In six of the cases incorrectly diagnosed in life but with grossly calcified valves, fluoroscopy could have settled the diagnosis, although it is admitted that some of the patients were too ill for this to be carried out. Closer attention to a long history of repeated attacks of fainting and giddiness in one patient with a harsh basal systolic murmur but a normal blood pressure should have led to recognition of the valvular lesion. A short history of heart failure in three patients with normal blood pressures and basal systolic murmurs, but without electrocardiographic evidence of cardiac infarction, who developed acute mental symptoms during the final illness which terminated suddenly, offered enough clinical evidence for a confident diagnosis of aortic stenosis.

## Summary

Significant aortic stenosis, almost always calcareous, is a common clinical and pathological entity. In this study, it occurred in 1.8% of routine necropsies.

In 25 proved cases of isolated calcareous aortic stenosis, heart failure and infective endocarditis were the commonest causes of death. In only three cases was aortic stenosis an incidental post-mortem finding.

A correct clinical diagnosis was made in just over half of the cases. On the basis of the classical signs of a slowly rising pulse, an aortic systolic murmur and thrill, and an absent second sound, less than one-sixth of the cases would have been recognized clinically.

Reasons are given to show that considerably more than half the reported cases might have been correctly diagnosed in life. A more careful evaluation of harsh aortic systolic murmurs, and demonstration by fluoroscopy of grossly calcified aortic valves, could have settled the diagnosis in some cases; a closer study of the natural history of the disease after the onset of symptoms should have directed attention to the valvular lesion in others.

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