TRICUSPID STENOSIS ; WITH PARTICULAR REFERENCE TO DIAGNOSIS AND PROGNOSIS

BY

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Recent experience with several cases of tricuspid valve disease, attending out-patient clinics without arousing any thought of this valve lesion, has stimulated our interest in the subject and has emphasized the inadequacy of most discussions on the subject.

It has become almost traditional to state that tricuspid stenosis is rare and that the diagnosis is difficult. Herrick (1897) wrote that the disease is so rare that the full details of every case should be reported. Mackenzie (1908) wrote that he had heard a tricuspid murmur only three times in his life. Osler and Gibson (1915) advocated caution in the diagnosis—"As a rule the physician is in a safer position if he limits his diagnosis to two valves: clinically, when lesions of three or four valves are determined with accuracy, mortifying post-mortem disclosures are not unlikely to follow." Lewis (1933) rather implies that the diagnosis is not worth making when he writes "I have not known the diagnosis, when made, affect the management of any case." Strümpell and Seyfarth (1928) say that tricuspid valve disease is so rare as to have no practical importance. The stress that has been laid on the rarity and difficulty of diagnosis of the affection combined with statements such as those above, have tended to make the average practitioner forget that the tricuspid valve is not infrequently affected. Of the 250 reported cases collected by Zeisler (1932) the diagnosis had been made before death in only 31: of those, 14 were made by Dressler and Fischer (1929-1930) who reported 33 autopsied cases, but in their series it was only after their interest had been aroused that the diagnosis began to be made, resulting in the correct interpretation in 11 out of the last 14 cases.

We have, therefore, thought it worth while to consider if the diagnosis of tricuspid stenosis is after all entirely unimportant and to determine as clearly as possible the diagnostic criteria.

According to the pathologist, the tricuspid valve becomes stenosed when the ostium is reduced to between 10 and 11 cm., the normal being between 12 and 14 cm. (Cabot, 1926). White (1937) writes that stenosis of clinical importance

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and of such degree as to be diagnosable clinically in some cases is not reached until the ostium is reduced to 8 cm. or less.

The valve becomes incompetent for three reasons: (1) extensive scarring in which regurgitation accompanies stenosis; (2) slight shortening of the chordæ tendineæ or fibrosis of the valve edge resulting in regurgitation, without necessarily causing stenosis; and (3) dilatation of the valve ring due to myocardial infection, long standing congestive failure, or constitutional disease such as anæmia. The valve is diseased in the first and second instances, and defective though not diseased in the third; the functional disorder of the third instance may actually be more serious than organic valvular disease, at least as a manifestation of important trouble, though often it is only a transient condition.

INCIDENCE AND PATHOLOGY

A false idea of the incidence of tricuspid valve stenosis has been given by the numerous compilations of isolated case reports of the disease. There exists, moreover, a good deal of discrepancy between several of the analyses that have been published, doubtless dependent largely on the degree of stenosis. In Baltimore only 7 cases were found in 24,000 autopsies (Hirschfelder, 1910). Coombs (1924) gives the following figures of the incidence of valve injury in 97 cases of rheumatic heart disease, including the lesser as well as the greater degrees of involvement.

Total	Mitral	Aortic	Tricuspid	Pulmonary
97	97	57	35	2

These were combined in the following ways:

Mitral alone	..	..	..	..	..	..	27 cases
Mitral and aortic	..	..	..	..	..	..	35 "
Mitral and tricuspid	..	..	..	..	..	..	12 "
Mitral, aortic, and tricuspid	..	..	..	..	..	..	21 "
Mitral, aortic, tricuspid, and pulmonary	..	..	..	..	..	..	2 "

He does not give the relationship of these cases to the clinical signs, but later states that the incidence at Bristol of clinical tricuspid stenosis in rheumatic heart disease was 14 per cent. Cabot (1926) gives the incidence at 15 per cent while Dressler and Fischer put the incidence at 24 per cent.

Bland, Jones, and White (1935) analyzed the pathological findings in 100 cases of fatal rheumatic disease below the age of 21 in whom the diagnosis of tricuspid valve disease had not been made during life. Out of 100 cases, the mitral valve was affected in 98 instances, the aortic in 71, the tricuspid in 30, and the pulmonary valve in 5, and there was one case with no valves at all affected. The valve lesions, many of them acute with little or no deformity, occurred alone or combined as follows:

Mitral alone	..	..	..	..	..	..	23 cases
Aortic alone	..	..	..	..	..	..	1 case
Mitral and aortic	..	..	..	..	..	..	45 cases
Mitral and tricuspid	..	..	..	..	..	..	5 "
Mitral, aortic, and tricuspid	..	..	..	..	..	..	20 "
Mitral, aortic, tricuspid, and pulmonary	..	..	..	..	..	..	5 "

This group is selected, in that it is composed of cases dying of rheumatic fever: therefore one might suggest that they had more extensive heart involvement than a similar group having rheumatism over the same period of time, but surviving; and that therefore the incidence may naturally be expected to be higher. On the other hand, valve deformity sufficient to produce stenosis is less common in these younger cases, even though fatal; the myocardium is more involved than the valves. Yet it is fairly certain that cases surviving show a rather high incidence of tricuspid involvement, even though rarely progressing to a degree of much stenosis. Von Glahn (1927) examined 109 rheumatic heart cases at autopsy, grouping them into acute or chronic lesions, and found 19 acute and 26 chronic tricuspid valve lesions making a total of 45 or 41 per cent. Libman (1923) has always emphasized the high incidence of tricuspid involvement (not stenosis as such) in rheumatic heart disease; in one small series, he found the valve involved in 12 out of 18 cases of endocarditis. Thayer (1925) also found a high involvement (44 per cent). Cabot (1926) records 33 cases of tricuspid stenosis among 4000 autopsies between 1896 and 1919 at the Massachusetts General Hospital.

OUR SERIES OF CASES

We have analyzed the data on 30 cases of tricuspid stenosis proved post-mortem, and on 12 cases that we ourselves have studied during life—3 confirmed by autopsy, the others still alive but almost certainly correctly diagnosed. At the Massachusetts General Hospital between the years 1920 and 1937 inclusive, there have been 4300 autopsies; these included 217 cases of rheumatic heart disease, and in that series there was involvement of the tricuspid valve in 47 cases. Of these 47 there were 17 in whom the affection was probably of no clinical significance, being either an acute terminal rheumatic process or a bacterial endocarditis or one of the group that Libman states is common, slight scarring and fibrosis of a limited segment of the valve ring.

The valves affected in this whole group from 1920 to 1937 at the Massachusetts General Hospital consisting of 217 cases of rheumatic heart disease were as follows:

Mitral alone	..	..	..	..	..	..	..	59 cases
Aortic alone	..	..	..	..	..	..	..	11 "
Mitral and aortic	..	..	..	..	..	..	..	100 "
Mitral and tricuspid	..	..	..	..	..	..	..	7 "
Mitral, aortic, and tricuspid	..	..	..	..	..	..	..	35 "
Mitral, aortic, tricuspid, and pulmonary	..	..	..	..	..	..	..	5 "
Total	..	..	..	..	..	..	..	217 "

Thus there were 47 cases of tricuspid disease (22 per cent) among the 217, but only 30 (14 per cent) that were important in themselves (as definite tricuspid stenosis).

There is unfortunately no satisfactory way in the series of cases noted above of correlating the post-mortem data with the clinical signs of tricuspid disease that may have been present. The diagnosis was made before death in only 1 of the 30 cases and suspected in only 2 other cases. These 30 cases have

therefore been divided up by us quite arbitrarily into three clinical diagnostic groups according to whether or not the cases might have shown signs during life. Accordingly the clinical diagnosis justified by the data of the 30 cases of tricuspid stenosis was as follows:

1. Tricuspid stenosis and regurgitation of varying degree .. 19 cases
2. Possible functional impairment of tricuspid valve .. 9 „
3. Tricuspid ring dilatation alone 2 „

If these assumptions are correct, there would be roughly 9 per cent (that is, 19 out of 217 cases of rheumatic heart disease) of all cases of rheumatic valvular disease showing clinical signs of tricuspid stenosis and regurgitation.

There is one further fact to be considered statistically. In adults, tricuspid stenosis produces in its later stages a life of chronic invalidism, which lasts for very much longer than the invalidism produced by mitral valve disease alone. For this reason it is probable that these cases find their way to homes for the chronic sick rather than to the general hospitals, and so lower the post-mortem incidence of tricuspid disease in the hospitals.

As yet there is no large series of rheumatic heart cases that has been followed through from beginning to end, to give us a better idea of the true incidence. From the facts at present available, it is probable that one case in every ten of rheumatic heart disease of more than a few years' duration has organic disease of the tricuspid valve, at least moderate in degree.

AGE AND SEX

Herrick (1908) gives a summary of the ages at death of 187 reported cases of tricuspid stenosis: in his series and in our own series of 30 cases studied post-mortem they were as follows:

		Herrick's Series	Our Series
0-10 years	..	0	1
10-20 „	..	16	8
20-30 „	..	59	7
30-40 „	..	38	6
40-50 „	..	28	5
50-60 „	..	10	3
60-70 „	..	6	—
Age not given	..	30	—

Usually females have been more frequently affected than males (Duroziez, 1868; Herrick, J. B., 1897; Dressler and Fischer, 1929). In our 30 cases, 16 were males and 14 were females. In an additional later group of 12 cases that we have observed clinically, 7 were females and 5 were males.

CLINICAL DIAGNOSIS

Duroziez (1868) reported 10 cases of tricuspid stenosis with ages varying from 22 to 64, and asserted that tricuspid disease was more common than was usually accepted. He was impressed by their marked cyanosis and by the fact that many could lie flat in bed in spite of gross cardiac deficiencies: in spite of marked valvular damage some were able to live to old age, and so he concluded

that the tricuspid lesion did not aggravate the mitral lesion. He drew attention to the systolic or diastolic murmur at the lower end of the sternum that pointed to the diagnosis, and wrote: "The disease should be diagnosed when the patient is a female, has a history of rheumatism, and of dyspnoea, palpitation, oedema, often with remissions and exacerbations, is cyanosed, has mitral stenosis with an enlarged right heart, particularly if the auricular enlargement can be made out. Especial value attaches to the persistent cyanosis. If in addition to this the patient has a separate murmur best heard over the ensiform or the fifth or sixth right costal cartilage, then the diagnosis becomes reasonably certain." This admirably summarizes the chief points. There is no one infallible diagnostic sign but there are numerous signs, any one of which should initiate a search for other clues.

The diagnosis may be suspected from the *history* alone as Levine and Thompson (1937) have pointed out. Any adult patient who has repeated attacks of oedema and ascites and yet is able to lead a sedentary life with the aid of diuretics and carry on with symptoms that ordinarily would cause the early demise of the patient, probably has tricuspid stenosis.

Several observers, most recently Wearn (1936), have commented on the peculiar colour, a mixture of jaundice and cyanosis, that is presented by these patients, pointing out that this should always suggest the possible diagnosis of tricuspid stenosis. Much reliance cannot, however, be placed on this sign inasmuch as its commonest pathogenesis lies in the inability of an engorged or diseased liver to excrete rapidly enough the blood pigment from a pulmonary infarct (so common in mitral stenosis with congestive failure), whether mitral stenosis alone or mitral stenosis with tricuspid valve disease is responsible for the trouble with the liver.

A patient with a rheumatic heart and ascites who is *able to sleep without extra pillows* probably has tricuspid stenosis. This is due to the lack of pulmonary vascular engorgement, as confirmed by X-ray study, resulting from the obstruction to the free flow of blood through the right heart.

Pulsation in the neck veins has for over one hundred years excited comment, with much argument as to whether the jugular polygram is diagnostic. Mackenzie (1894) pointed out that the deep (internal) jugular pulse in the neck is frequently thought to be arterial in origin because it is systolic in time. As a matter of fact it can very easily be shown to be venous in origin by light compression over the base of the neck (jugular bulb) with obliteration of the pulse (Fig. 1). The significance of this venous pulsation has recently been studied by us (White and Cooke, 1939). We found that although this pulse in the neck signifies only tricuspid regurgitation, whether due to organic disease or to functional regurgitation, *there is, nevertheless, a very strong probability, in fact almost a certainty, of the presence of tricuspid stenosis in the case of a patient in the fourth or fifth decade with rheumatic heart disease who has for years marked systolic deep jugular pulsation with little or no congestive failure* (see Cases 1 and 2, and Fig. 2).

The same conclusion holds good about *liver pulsation*. It is interesting that the observation is frequent in the follow-up records of a large number of the

histories we have studied, "that the patient still shows some signs of failure as evidenced by his large liver." On the other hand tricuspid stenosis may be

FIG. 1.—(A) Simultaneous tracings of electrocardiogram (lead I), above, and of deep jugular pulsation, below, in a case of rheumatic, aortic, mitral, and tricuspid stenosis, with auricular fibrillation. Note the slow upward stasis curve with superimposed systolic pulse.

(B) Simultaneous tracings of electrocardiogram (lead I), above, and of deep neck pulsation, below, after compression of the jugular bulb. The receiving cup has not been moved in its position. The jugular pulse has been obliterated and only the underlying carotid pulse remains (it is anacrotic).

present without venous pulsation either in the neck or in the liver (Clements, 1927; and others).

Tschilikin (1930) states that the only pathognomonic sign of tricuspid stenosis is a *localized diastolic murmur* at the lower end or to the right of the sternum. However, this sign is often, probably usually, absent, as in the

FIG. 2.—Case 3. (A), (B), (C) Sequential exposures taken from a moving picture film of the systolic deep jugular pulse of a young man with tricuspid stenosis: recorded in the sitting position. Early diastole in A, full systole in C, and midway in B.

majority of Dressler and Fischer's cases. It was present in 7 of the 12 cases that we have ourselves examined clinically.*

Auricular fibrillation and polycythaemia occur just as frequently in cases of pure mitral valve disease, especially the so-called "tight type," as in tricuspid stenosis. There is no evidence in the group of cases here reported that pulmonary infarcts are more frequent with tricuspid stenosis than in mitral stenosis alone.

No diagnostic significance can be attached to the electrocardiogram.

We have estimated the *venous blood pressures* and *circulation rates* in seven of these patients. All the cases, although ambulatory and comparatively well, gave readings between 19 and 27 cm. of water (method of Burwell *et al.*). This has been noted in the absence of œdema, constrictive pericarditis, and mediastinal obstruction on several occasions (e.g. Friedlander and Kerr, 1936; Altschule and Blumgart, 1937) and should be taken as a strong diagnostic point in favour of tricuspid stenosis. The circulation times were all markedly prolonged, the right heart times averaging 20 seconds (ether method) against a normal average of 6 seconds.

Dressler and Fischer emphasized as an important diagnostic pointer the presence of *marked enlargement of the heart to the right by X-ray* especially when associated with absence of pulmonary congestion at the hilus of the lung.

Lian and Marchel (1936) drew attention to the *deviation of the œsophagus to the left* in the presence of marked enlargement of the heart to the right. They suggested that this finding, while not pathognomonic of tricuspid stenosis, indicates enlargement of the right auricle and should lead to the suspicion of tricuspid stenosis. We have found in 4 of the cases here reported that the œsophagus is deviated to the left and in other instances appears to travel down the front of the spinal column in the midline. Certainly the incidence of this left deviation of the œsophagus in our small group of cases warrants some consideration being paid to this finding (Fig. 3).

Castex, Battro, and Quirno (1939) reported two cases of tricuspid disease diagnosed during life. Kymograms of both patients suggested the diagnosis of tricuspid regurgitation by the presence of systolic pulsation in the right auricle and in the dilated superior vena cava.

We are herewith presenting the histories and essential findings in 17 of our cases, the first 3 in some detail as classical examples of the condition and the last 14 in brief. Details are summarized in the table on p. 157. In the 217 autopsies of patients with rheumatic hearts already referred to, there were 5

* An interesting possible but doubtless very rare explanation for a middiastolic murmur at the lower end of the sternum, not transmitted from the apex and not due to organic tricuspid stenosis, is a mechanism in the right ventricle similar to that in the left ventricle which causes the Austin Flint murmur, namely, pulmonary regurgitation with dilated right ventricular cavity. It happens that recently (December 1939) one of us (P. D. W.) has encountered such a case, a patient in failure with marked mitral stenosis, functional pulmonary regurgitation giving rise to a Graham Steell murmur, and a well marked localized middiastolic rumble in the 4th intercostal space at the left border of the sternum just below and distinctly different and separate from the pulmonary systolic murmur. Tricuspid stenosis was diagnosed antemortem on this basis but not found at autopsy, which disclosed marked mitral stenosis, dilated partially thrombosed pulmonary artery and left auricle, and dilated right ventricle.

A

B

FIG. 3.—(A) Antero-posterior X-ray view of the heart shadow of case of mitral and tricuspid stenosis showing barium-filled oesophagus deviated to the left.

(B) Left (II) oblique view of same case.

who had marked or clinically “pure” tricuspid stenosis, and 25 others with slight to moderate tricuspid stenosis, making a total of 30 cases.

CASE REPORTS

Case 1, female, aged 54 years. An attack of rheumatism at the age of 32 kept her in bed a short time, but she was back at work within three weeks. She first attended Massachusetts General Hospital in December 1935, for abdominal swelling of 18 months' duration. Mitral, tricuspid, and aortic stenosis and regurgitation were diagnosed. Since then she had carried on her housework and had entered the hospital every two or three weeks for abdominal paracentesis and occasional doses of salyrgan intravenously. Her condition remained fairly constant for the three years that she was under observation. She never used more than one pillow at night although the veins of the neck, forehead, and arms were remarkably distended when she lay down. She was slightly cyanosed and slightly icteric. The neck veins showed a systolic deep jugular pulsation extending to the angle of the jaw and easily obliterated by light pressure over the jugular bulb at the base of the neck. There was no engorgement of the superficial veins. Venous pressure was 26 cm. water. Circulation time: ether, 20 seconds (normal 5–10); and saccharin, 50 seconds (normal 15–25).

The heart was enlarged both to right and left and the oesophagus was deviated in its lower half to the left of the vertebral column. B.P., 115/85 mm. On auscultation there were systolic and mid-diastolic murmurs at the apex, and a loud aortic systolic murmur with a palpable thrill at the base, but no tricuspid murmur could be defined. E.C., auricular fibrillation with moderate right axis deviation, slight slurring of the QRS waves, and “digitalis” T waves.

She was admitted to the hospital for the last time after a cerebral vascular accident and died a few days later in April 1939.

Autopsy.—The superior and inferior vena cavæ were much dilated, measuring 2.8 and 4 cm. respectively in diameter. The heart weighed 410 g. The right auricle

was much dilated and its walls hypertrophied. The cardiac apex was formed by the large right ventricle although the walls were not thicker than those of a normal right ventricle. The tricuspid valve admitted two fingers only and measured 10.5 cm. in circumference (Fig. 4A); all the leaflets were thickened and fused to form a slightly

FIG. 4.—Case 1. (A) Markedly stenosed (button-hole) tricuspid valve seen from below, in a case of chronic rheumatic heart disease.

(B) Stenosed mitral valve seen from above, in the same case.

nodular band 12–13 mm. in width; the chordæ were not appreciably thickened. The left auricle was also much dilated, approximating in size that of the right. The mitral valve was of the fish mouth type, 6 cm. in circumference but with remarkably narrowed and rigid fused cusps and an opening approximately 2.5 by 0.5 cm. (Fig. 4B). The aortic valve was approximately 7 cm. in circumference, with fusion of the valve cusps and poor approximation. The liver was enlarged and showed cirrhosis which was thought to be cardiac in origin.

Case 2, female, aged 49. No history of joint pains, rheumatic fever, chorea, or scarlet fever during childhood. At the age of 14, she had a chest cold which confined her to bed for a fortnight and left her very weak for some months. She was, however, able to play games actively as a child. At the age of 22 she began to notice some undue shortness of breath while doing heavy housework, and began to complain of attacks of praecordial pain which have been present more or less ever since. At the age of 27, she was rejected for life insurance owing to a murmur in her heart, of which she had been unaware. At 43 years she had some signs of congestive failure which cleared up but have recurred from time to time ever since. The following year she was admitted to the hospital with slight congestive signs and menorrhagia due to fibroids. She was digitalized and was able to continue her work in the household and at a sewing centre. A diagnosis of tricuspid stenosis was queried. At the age of 47, she was again admitted with abdominal swelling of two years' duration and examination at that time showed cyanotic lips, malar flush, distension of the neck veins, markedly engorged

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TABLE SHOWING IMPORTANT FINDINGS IN 17 CASES OF TRICUSPID STENOSIS

	*Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7	Case 8	Case 9	*Case 10	Case 11	Case 12	*Case 13	*Case 14	*Case 15	*Case 16	*Case 17
Age	54	49	40	54	60	36	57	50	39	34	58	29	21	41	42	30	41
Sex	F.	F.	M.	M.	F.	F.	M.	M.	F.	F.	M.	F.	M.	F.	F.	F.	F.
Rheumatic fever	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Chorea	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Duration of symptoms (in years)	5½	6	8	?	10	6	3	2	5	1½	4	?	6	7	?	2½	5
Cyanosis	..	sl.	..	sl.	sl.	sl.	..	..	sl.	sl.	..	sl.	..	..	..	sl.	..
Jaundice	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Systolic deep jugular pulsation	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Enlargement of liver	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Pulsation of liver	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Ascites	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Diastolic murmur localized at lower end of sternum	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Enlarged right heart	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Deviation of oesophagus to left of midsternum (X-ray)	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Arterial pressure (mm. mercury)	115	150	100	150	235	120	130	120	110	135	120	120	140	115	150	120	125
Systolic	85	80	70	80	100	80	80	90	70	80	90	70	55	30	80	75	85
Diastolic	26	22	24	22	21	—	—	19	24	—	—	—	—	—	—	—	—
Venous pressure (cm. water)	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..
Circulation time (sec.): saccharin (arm to tongue)	50	30	19	45	44	—	—	60	44	—	—	—	—	—	—	—	—
Circulation time (sec.): ether (arm to lungs)	20	15	15	20	15	—	—	0	20	—	—	—	—	—	—	—	—
Auricular fibrillation	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..	..

The last 5 cases (Nos. 13-17 inclusive) presented marked tricuspid stenosis at autopsy, as did also Case 10. The first 12 cases were carefully studied during life by ourselves.

The cases were comprised of 6 males and 11 females, whose ages ranged from 21 to 60 with an average of 43 years : 4 had no history of rheumatism, and 2 had had chorea only.

liver, palpable spleen, heart enlarged to the left, irregular rhythm, and systolic and diastolic murmurs at the apex. Over the next two years the enlargement of the liver was found constantly.

On examination at present her condition is good. She is able to do her housework and carries on with her sewing work. She has deep and superficial systolic pulsation in the neck veins and is a little cyanosed in lips and finger tips.

The heart is enlarged by X-ray to both right and left, but the lung hilar shadows are not strikingly abnormal. The heart is "mitral" in shape. The œsophagus runs down the front of the vertebral column. On auscultation there are systolic and mid-diastolic murmurs at the apex, and localized systolic and faint diastolic murmurs, separate and distinct, at the lower end of the sternum. The sounds at the aortic area are quite clear. The liver is enlarged two fingers' breadth below the costal margin, and pulsates with systole. B.P., 140/80 mm. E.C., auricular fibrillation, no abnormal axis deviation, and digitalis T waves. Venous pressure 21–24 cm. water. Circulation time: ether 15 seconds (normal 5–10); lobeline over 30 seconds (normal 15–20).*

Case 3, male, aged 40 years. First seen by us at the age of 21, following rejection for life insurance. He had run as a member of his high school team and had been apparently quite healthy until one year previously when he had some mild rheumatic pains and some paroxysms of tachycardia. At that time he had a normal sized heart, a blood pressure of 90/65, an accentuated and reduplicated pulmonary second sound, a blowing watery systolic murmur nearer the sternum than the apex, and a short early diastolic murmur in the second and third left interspaces, while the E.C. showed normal rhythm with probable early intraventricular block. For the next ten years he was fairly well, working hard, until in 1931 at the age of 32 he developed auricular fibrillation and congestive failure. He made a good recovery, and in 1934 visible venous pulsation in the neck, an enlarged and pulsating liver, and a very large heart were found; in spite of this he was able to walk five miles a day without undue discomfort. The diagnosis of tricuspid stenosis was then queried. In 1935 a note was made that the mid-diastolic murmur was louder at the lower end of the sternum than at the apex. At the end of 1936 ascites began to be a prominent symptom and necessitated paracentesis every ten to fourteen days, 10–12 litres being withdrawn each time. In 1937 his activities were extremely limited by fatigue and dyspnoea, and a prognosis of only a few months of life was given. However, over the past two years his condition has not grown any worse and at present he spends his time resting, walking about the house, and going out for car rides.

On examination his facial appearance is evident in Fig. 2. He shows slight cyanosis, emaciated musculature, a prominent abdomen, and marked systolic, deep jugular pulsation in the neck veins (Fig. 2 A and B); on auscultation there are systolic and diastolic murmurs at the apex, and at the lower end of the sternum there are two other distinct murmurs; the liver is enlarged and pulsating, and there is marked ascites. B.P. 100/70. X-ray examinations, over the years from 1931 to the present, have revealed a steadily increasing heart size, so that now the shadow of the right heart border touches that of the right chest wall. The œsophagus is deviated slightly to the left in the lower half of its course through the thorax. E.C., auricular fibrillation and intraventricular block of the right branch type. Vital capacity in 1931, 3550 c.c.; in 1939, only 1900 c.c. The venous pressure is 23.5–24.5 cm. water. Circulation rate: ether time 19 seconds (normal 5–10).

Case 4, male, aged 54 years. Rheumatic fever at the age of 34, in bed 14 weeks. Worked steadily thereafter as a mechanic. Examined after a transient attack of blurring of vision and weakness, there was slight cyanosis, systolic deep jugular pulsation, a slightly enlarged liver with definite expansile pulsation, localized systolic and diastolic murmurs at the lower end of the sternum, and in addition the murmurs diagnostic of mitral and aortic stenosis and regurgitation, and auricular fibrillation. X-ray showed enlargement of the heart to right and left. He is still working as a mechanic.

* For later notes and autopsy see Addendum on p. 165.

Stenosis and regurgitation of the mitral, aortic, and tricuspid valves was diagnosed. Venous pressure was 22 cm. water. Circulation rates: ether time, 20 seconds; saccharin time, 45 seconds. A point of special interest in this case, besides that of his relatively good condition, is that his systolic jugular pulsation was mistaken for a carotid pulse in the medical clinic.

Case 5, female, aged 60 years. Rheumatic fever at the age of 40. She has had twelve children and two miscarriages. She was first seen for her heart trouble because of attacks of tachycardia and fibrillation at the age of 45. First admitted to hospital for congestive failure at the onset of permanent fibrillation ten years ago, at the age of 50 years. At that time she had an enlarged and pulsating liver and ascites and one of us questioned the diagnosis of tricuspid stenosis. For the past ten years she has been much the same, becoming dyspnoëic on slight exertion and presenting on X-ray examination a heart enlarged both to right and left, and on physical examination an enlarged and pulsating liver. She lives alone on a hill and is able to do her housework and some shopping and comes to the cardiac clinic once a month.

At present she is slightly cyanosed with systolic pulsation of the neck veins, which has been present for at least one year. Although the liver is large and pulsating, there is no ascites or œdema of the ankles. There is no localized tricuspid murmur. E.C., auricular fibrillation and intraventricular block. B.P., 235/100. A diagnosis of mitral and aortic stenosis and regurgitation, tricuspid regurgitation and probable tricuspid stenosis, and hypertension has been made. Venous pressure 21 cm. water. Circulation time: ether, 15 seconds; saccharin, 44 seconds.

Case 6, female, aged 36. First attack of rheumatic fever at the age of 24 and a second attack two years later. She was admitted to the hospital in congestive failure in 1933 and for this a total thyroidectomy was performed. Since then she has been troubled with symptoms of myxœdema and hypo-parathyroidism, and was always on the edge of congestive failure with repeated attacks of ascites and always with an enlarged pulsating liver. A diagnosis of mitral stenosis and regurgitation, tricuspid regurgitation and probable tricuspid stenosis, and aortic stenosis has been made.

Autopsy in January 1941 confirmed the clinical diagnosis of rheumatic heart disease with pancarditis, panvalvulitis, and pericarditis, chronic; cardiac hypertrophy and dilatation; mitral, aortic, and tricuspid stenosis and regurgitation, slight œdema, slight ascites, slight bilateral hydrothorax, pulmonary congestion, and confluent bronchopneumonia of the right upper lobe and right lower lobe.

Case 7, male, aged 57. At the age of 13 years his mother told him that he had valvular heart disease, but he had no definite history of rheumatic fever. His presenting symptoms on admission to the hospital were œdema of the ankles and dyspnoëa of three years' duration and abdominal swelling of one year's duration. On the basis of his liver enlargement and ascites, systolic jugular pulsation, enlargement of the heart to right and left, and a localized diastolic murmur, a diagnosis of tricuspid stenosis, in addition to mitral stenosis, was made. He responded well to digitalis, and is now ambulatory and comparatively well though somewhat limited in his activities, twelve months after discharge from hospital.

Case 8, male, aged 50. Rheumatic fever at the age of 17. Otherwise well until two years ago when he had an attack of swelling of the joints, following which he got increasingly short of breath, and his abdomen swelled. A diagnosis of tricuspid stenosis and regurgitation was made on the basis of the history, the marked systolic jugular pulsation, a large pulsating liver that was not tender, ascites, some enlargement of the heart to the right, and a localized systolic murmur at the lower end of the sternum, in addition to the signs of mitral and aortic valve lesions. At present, six months later, he is able to help in the house, do light gardening, and keep his chickens, but he is not able to perform any strenuous work. Venous pressure, 19 cm. water. Circulation rate, using decholin, 60 seconds.

Case 9, female, aged 39. Repeated attacks of polyarthritis from the age of 16 to 34, although she never took any prolonged rest in bed and was doing fairly heavy work

as a housemaid for most of this time. Four years ago admitted to the hospital in congestive failure from which she made a good recovery; systolic jugular pulsation was noted on this occasion. Her last admission was precipitated by excessive exertion on coming to the city every day to go to a sewing centre.

A diagnosis of tricuspid stenosis and regurgitation was made on the basis of the history, the jugular pulsation, a pulsating enlarged liver, a localized systolic murmur at the lower end of the sternum, and X-ray enlargement to the right and left with the œsophagus passing down the front of the vertebral column. The venous pressure was 24 cm. water, and circulation rates were prolonged: with ether, 20 seconds, and with saccharin, 44 seconds. She is ambulatory, but restricted in her activities.

Case 10, female, aged 34 years. Rheumatic fever at the age of 7 and again at 13 with pericarditis. In bed for two years from the age of 20 to 22, and then able to work for eleven years as a stenographer. She was admitted to the hospital with recurrent rheumatism, auricular fibrillation, and congestive failure, twenty months before death. Ascites and œdema and the enlarged pulsating liver showed little variation except for temporarily yielding to diuretics. On account of the persistence of the failure and the pulsation of the liver and in the neck, and a localized diastolic murmur at the lower end and to the left of the sternum, a diagnosis of tricuspid stenosis in addition to mitral and aortic valve disease was made. She died suddenly.

Autopsy showed marked mitral, tricuspid, and aortic stenosis.

Case 11, male, aged 58. Well and active as a youth. Rheumatic fever, at the age of 37, kept him out of work for 4 years, and affected his heart. He felt well, however, thereafter for 13 years. He has had dyspnoea for 4 years and swelling of his abdomen, for the past 2 years, for which he has had to have abdominal paracentesis every few weeks.

A diagnosis of tricuspid stenosis in addition to mitral stenosis was made on the basis of the history of systolic jugular pulsation, an enlarged liver with ascites, a large heart, and localized systolic and diastolic murmurs at the lower end of the sternum.

Case 12, female, aged 29. Rheumatic fever with joint pains; at the age of 7, chorea for six weeks, recurring each winter until the age of 13 when a heart lesion was first diagnosed. Married at the age of 18, there was a normal pregnancy and delivery at 22. The auricles began to fibrillate at 23 and she developed congestive failure. Since then marked limitation of activities, with three attacks of congestive failure and probable rheumatic fever. For the past ten months she has had ascites and œdema of the legs and relatively little dyspnoea, and has been living a bed-and-chair existence.

A diagnosis of tricuspid stenosis has been made upon the history suggestive of long-standing right heart failure, a large pulsating liver, a localized diastolic murmur at the lower end of the sternum, marked enlargement of the heart to the right by X-ray, and slight systolic deep jugular pulsation.

Case 13, male, aged 21, negro. Rheumatic fever at the age of 6; no recurrences. His activity was not restricted. He played games, graduated from high school, and worked as a day labourer until he developed acute rheumatic fever two months previous to his admission to hospital. He was admitted with tremendous œdema of the legs and abdomen. Fibrillation of the auricles was present.

Autopsy showed marked stenosis of mitral, aortic, and tricuspid valves.

Case 14, female, aged 41. Chorea at 12 and 14. She married and had five children, the last at the age of 24 years without any difficulties. At 34, she was admitted to the hospital owing to increasing shortness of breath and attacks of pulmonary œdema and a three months' pregnancy which was terminated. X-ray showed enlargement of the heart to the right. The cardiac rhythm was regular. She was able to do heavy housework with the aid of full digitalization, until two weeks before her death from cerebral embolism.

Autopsy showed chronic rheumatic endocarditis, with marked stenosis of both mitral and tricuspid valves, cardiac hypertrophy, embolism and thrombosis to the bifurcation of the abdominal aorta, both external and internal iliac, and both renal arteries, embolus at bifurcation of the basilar artery with occlusion of postero-median

ganglionic branches, infarction of the spleen and both kidneys, pulmonary congestion, and cholelithiasis.

Case 15, female, aged 42. Whooping cough at 5, pneumonia at 9 and 12, and also "rheumatism" in childhood. She had suffered from ill health most of her life, with cough, sputum, and hemoptysis, thought to be due entirely to bronchiectasis. Her third hospital admission found her with marked congestion (anasarca); she died at this time.

Autopsy showed tightly stenosed mitral and tricuspid valves.

Case 16, female, aged 30. At 16, because a rapid regular cardiac impulse was noticeable on the chest wall, she was put to bed for one month, although she had no symptoms at all. After this she resumed her usual active life. At the age of 22 she had paroxysmal tachycardia but was able to play tennis and dance energetically. Two and a half years before death she developed generalized œdema of the extremities, chest, back, and face, with ascites, for which she was bedridden. She was not relieved by diuretin or digitalis.

Autopsy revealed mitral and tricuspid stenosis and active rheumatism.

Case 17, female, aged 41. Chorea at 12 and 14, and a syncopal attack at 17, following which valvular disease was discovered. At 27 she collapsed while nursing and from then onwards she was much limited by dyspnœa. Ten years ago (at 31) œdema of her ankles set in and marked cyanosis appeared two years ago. Following her mother's death she became psychopathic and was admitted to the hospital. At this time her lungs were clear, but she showed a slightly enlarged liver, ascites, and slight œdema of the legs. She grew worse and died.

Autopsy showed marked stenosis of aortic, mitral, and tricuspid valves, the tricuspid valve barely admitting the tip of the finger.

DISCUSSION

Tricuspid regurgitation as evidenced by pulsating neck veins, a distended and pulsating liver, tricuspid murmurs, and in some cases by the absence of any respiratory distress in spite of marked ascites and œdema, is not uncommon in children suffering from rheumatic fever. The signs may persist for some months and only disappear with the disappearance of the rheumatic fever and the recovery of normal function of the heart muscle. It is therefore unwise, save in exceptional cases, to make a diagnosis of tricuspid valve disease in the first two decades of life, or in a patient within five or six years of the onset of his rheumatic fever.

Cases presenting the full tricuspid syndrome of ascites, œdema, pulsating neck veins, and surprising functional ability have been reported, in whom autopsy has revealed only an irreversible stretching of the valve and no endocarditis (Fischer, 1928, Holzman, 1932, and White and Cooke, 1939). There appears to be no method of avoiding this diagnostic pitfall. The stretching of the tricuspid ring, which occurs during failure but may persist even after the right ventricle has recovered in large part or wholly, presents the chief diagnostic difficulty. This stretching of the valve ring may be present in a case of isolated mitral stenosis, in which instance the full tricuspid syndrome will be simulated.

Some cases of long-standing systemic arterial hypertension develop this "tricuspid syndrome" over the last three to four years of their life. At autopsy the only abnormality is some dilatation of the tricuspid valve ring with the production of incompetence.

Finally, of course, cases of constrictive pericarditis must be distinguished. The absence of heart murmurs and of any more than the slightest enlargement of the heart should be sufficient to distinguish these cases from rheumatic heart disease.

For interest we determined the average age at death of 160 consecutive autopsied cases of rheumatic heart disease, excluding those with tricuspid involvement. This proved to be 43.5 years. The same determination was carried out in the series of 30 autopsied cases of tricuspid valve disease referred to earlier in this paper with the finding of 23.4 years; however, the average in our series of 17 cases whose histories we have presented was 43 years, with several patients still alive.

Gross, Kugel, and Rothschild (1934) in careful investigations showed that in the first five decades of life, lesions indicative of active rheumatism were present in over 80 per cent of cases with rheumatic heart disease dying of congestive failure. As patients grow older the frequency of attacks of rheumatic fever tends to grow less. In the young age group the attacks are frequent. In the first three decades, therefore, it is probably true to say that it is the state of the myocardium that plays the dominant part in the survival of the patient, while afterwards, the mechanical factors produced by the valve lesions play an increasingly important role in determining whether the heart can continue to function adequately. The whole group can consequently be divided into two, a young group in the first three decades of life and an old group. In the young group the greatest number of autopsies will occur in a general hospital as the patients are often acutely ill with rheumatic fever. The more severe the infection, the more likelihood there is that the tricuspid valve is affected. If, therefore, one can make a diagnosis of tricuspid stenosis in this first group, it indicates an extremely grave prognosis as to future life.

In the older group, the mechanical factors produced by the valve deformity begin to play an important role. The safety valve function has been recognized for many years, acting either by regurgitation or stenosis in preventing the dangerous distention of the right ventricle or of the lung capillaries that attends marked mitral stenosis. This function is very well illustrated by *Case 1*, a patient who was able to carry on with her household tasks in spite of such marked mitral and tricuspid stenosis as is shown in the illustration (Fig. 4 A and B). Her venous pressure was 21 cm. of water, and she always insisted on sleeping with one pillow only, an observation that is not in keeping with Altschule and Blumgart's contention that orthopnœa is closely related to a raised venous pressure.

The importance of recognizing tricuspid valve disease in this older group is, again, but for a different reason, in establishing the correct prognosis. As has already been stressed, these patients, although they will probably die at an earlier age than similar patients without the tricuspid valve affected, are able for some years to live a sheltered and useful life, although constantly presenting signs and symptoms that would lead one to expect a much earlier demise.

Thus, although well-marked tricuspid valve disease (stenosis) is found in

cases of rheumatic heart disease who live somewhat shorter lives than do those without tricuspid valve disease, this lesion is found in those cases who survive the longest (a matter of several years at least, as a rule) after the systemic veins (including the jugulars) and the liver become permanently engorged, with the appearance of ascites.

The realization of the true state of affairs in these older cases indicates the correct line of treatment. The chief complaint is usually recurrent or persistent ascites and œdema. Fluids must therefore be restricted and carefully balanced with the urinary output, a procedure these patients perform for themselves. Mercurial diuretics at regular intervals, or from time to time when required, give good results. Digitalis may or may not be indicated, depending chiefly on the heart rate when auricular fibrillation is present; as a rule it is necessary. Occasionally these measures are not sufficient and then abdominal paracentesis may be carried out when needed. It is unfortunately still true in many places that the auscultation of murmurs indicative of rheumatic heart disease in these patients leads automatically to the administration of large doses of sodium salicylate and sodium bicarbonate. Such drugs in these patients can only increase the patients' discomfort by causing increased water retention. For this reason, also, diets should be kept as low as possible in salt, and frequent saline diuretics such as those frequently advertised in the press for the production of a feeling of "well being" should be avoided.

It will be noted that the cases we are reporting are, with two exceptions, in the fourth, fifth, and sixth decades. The antecedent histories in all these cases are similar—the usual story being: (1) a single attack of rheumatic fever, leaving no demonstrable lesions until some years later when heart disease is discovered on routine examination; (2) attacks of chorea without any apparent heart involvement; or (3) isolated attacks of rheumatic fever in the fourth or fifth decade. In none of these cases was there a period of chronic ill health which is the commonest course for rheumatic fever to follow. In the past histories this group is very similar to a series with so-called "pure" mitral stenosis reported by Walsh, Bland, and Jones (1940). They found that patients who had had a single attack of rheumatic fever in childhood without any residual heart damage or those who had no definite previous rheumatic symptoms frequently appeared later with well marked mitral stenosis. A suggested explanation of this marked stenosis is that there never has been any great dilatation of the heart and that therefore gradual and progressive fibrosis of the valve has been allowed to proceed undisturbed during a long continued active rheumatic infection that has largely passed unnoticed.

SUMMARY AND CONCLUSIONS

Between 1920 and 1937 there were 217 cases of rheumatic heart disease occurring in 4300 autopsies at the Massachusetts General Hospital. In 47 of these the tricuspid valve was affected, but in only 30 was tricuspid stenosis thought to be of sufficient degree to be of clinical significance.

In addition 12 cases of tricuspid valve stenosis have been examined

clinically by the authors during the past three years including 3 cases that came to autopsy.

There were 21 males and 21 females in the combined groups of 33 autopsied and 9 clinical cases.

The ages at death varied between 10 and 59 years, and the average age for the 30 cases with autopsies was 23 years. The average age at death of 160 cases of rheumatic heart disease in the same hospital was 42 years.

The cases of tricuspid valve disease may be divided into two groups: a young group in the first three decades dying of rheumatic fever, and an older group in whom the mechanical factors induced by the lesions played an increasingly important part.

The symptoms in the younger group were almost indistinguishable from those of rheumatic fever: the older group was characterized by the relatively long survival after the appearance of congestive symptoms and signs indicative in most other circulatory disorders of death in the near future.

The diagnosis of tricuspid disease in the young group indicates serious involvement of the myocardium and a poor prognosis; in the older group owing to the "safety valve" function of the tricuspid valve, the patients may live many years providing there is no recurrence of severe rheumatic fever.

The diagnosis of tricuspid disease is difficult, but when due attention is paid to the history, clinical examination of the patient, and X-ray of the heart, the diagnosis should be made more frequently. No one sign is pathognomonic, but in the order of importance the clues may be listed as follows (their chief value lies in combination): a mid-diastolic murmur localized over the tricuspid area, chronic and well-marked systolic pulsation of the deep jugular veins, ascites in the absence of lung congestion, enlargement of the heart shadow to the right, deviation of the œsophagus to the left, cyanosis and sometimes jaundice, enlarged liver with or without pulsation, persistently raised venous pressure, and prolonged right heart circulation time. The chief reason that the diagnosis is not made more often is that these clues and signs are not looked for.

The diagnosis of tricuspid disease is important as an aid in the more accurate determination of prognosis and treatment.

ADDENDUM

Later Notes of Case 2.

A steady increase of dyspnœa, œdema, and ascites over a period of four months brought her into the hospital again on June 20, 1941, with marked anasarca, and she died two days later. Post-mortem examination showed a moderately hypertrophied heart with marked enlargement of the right ventricle which formed a large portion of the apex of the heart. Weight 450 grams. Right ventricle wall thickness was 5 mm. Mitral valve stenosed, buttonhole type; the circumference was 8 cm.; small thrombus in the left auricular appendage. Aortic valve very slightly involved by rheumatic adherence of the cusps. Aorta smooth. Pulmonary valve normal. Right auricle very large. Tricuspid valve moderately stenosed with shortened chordæ tendineæ. No thrombi in right auricle. Small recent pulmonary embolus and infarct (probably the immediate cause of death). Liver cirrhotic and congested. Kidney congested. Spleen normal.

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