Primary Abnormalities of the Mitral Valve in Marfan's Syndrome

Electrocardiographic Findings

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Bowers (1961a) described ST segment depression and T wave inversion in electrocardiographic leads II, III, and aVF in patients with Marfan's syndrome and abnormalities of the mitral valve. At that time, no attempt was made to distinguish those patients with primary abnormalities of the mitral valve from those patients whose mitral abnormalities were secondary to aortic disease. This electrocardiographic pattern was confirmed in three reports (Bowers, 1962; Perrin *et al.*, 1966; Dietzman *et al.*, 1967) of patients with Marfan's syndrome and mitral regurgitation, but could not be confirmed in a study (Read, Thal, and Wendt, 1965) of patients with the "floppy valve syndrome" (considered to be a *forme fruste* of Marfan's syndrome), nor in a study (Barrett *et al.*, 1964) of Marfan patients with mitral regurgitation secondary to aortic dilatation. Further review of the electrocardiogram in Marfan patients with mitral valve abnormalities appeared necessary.

SUBJECTS AND METHODS

Bowers (1969) reviewed the pathogenesis of primary mitral valve abnormalities in patients with Marfan's syndrome. Cases were included in that review only if they satisfied stringent criteria for the diagnosis of Marfan's syndrome, and if they met the requirement of necropsy confirmation of mitral valve deformity and exclusion of aortic valve deformity.

Fifteen cases already reported in various medical journals were considered to satisfy these specifications. Electrocardiograms from 7 of these 15 patients were illustrated with the original case reports; electrocardiograms from an additional 6 of these 15 patients were obtained by the author through personal communications with their attending physicians. The present paper reviews the electrocardiographic findings in these 13 patients (Table I). Electrocardiograms from 2 of the

15 patients, reported by Olcott (1940) and by Lutman and Neel (1949), were not available for study.

The electrocardiograms were interpreted by the author using currently accepted diagnostic criteria.

RESULTS

Sinus rhythm was present in tracings from 9 of the 13 patients, and atrial fibrillation in the remaining 4. In the 9 tracings showing sinus rhythm, first degree atrioventricular block was present in 2; the remaining tracings showed normal atrioventricular conduction. In one tracing, the QRS interval was prolonged to 0.12 sec., with a late R wave in lead V6; in the remaining tracings, the intraventricular conduction time was normal.

The abnormalities of repolarization observed in leads II, III, and aVF, and the left praecordial leads are presented in Table II.

DISCUSSION

In a previous study of 55 unselected patients with Marfan's syndrome (Bowers, 1961b), 29 per cent of the patients had abnormal electrocardiograms.

The electrocardiograms reviewed in this paper have been obtained from a selected group of patients; the criteria for the diagnosis of Marfan's syndrome have been strict; pathological confirmation of mitral valve deformity and exclusion of aortic disease have been mandatory. It is impressive that the electrocardiograms from all these patients were abnormal.

In the previous study of electrocardiograms from unselected patients with Marfan's syndrome, the most common abnormal electrocardiographic pattern was the pattern indicative of left ventricular hypertrophy. In the electrocardiograms from these 13 Marfan patients with primary mitral valve abnormalities, the most common abnormal pattern was

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TABLE I
IDENTIFICATION OF ELECTROCARDIOGRAMS FROM PATIENTS WITH MARFAN'S SYNDROME AND
NECROPSY-PROVEN PRIMARY ABNORMALITIES OF MITRAL VALVE

Previous publication(s) of patient's case history	Previous publication of electrocardiogram	Comment
McKusick (1966) Fig. 3-18C Keith, Rowe, and Vlad (1958); Bowers (1961a) Case 1; Wagenvoort, Neufeld, and Edwards (1962) Case 3; McKusick (1966) Fig. 3-6A Bowers (1961a) Case 5; McKusick (1966) Fig. 3-36 Miller and Pearson (1959); Bowers (1961a) Case 3 McKusick (1966) Fig. 3-35 Vivas-Salas and Sansón (1948) Wunsch, Steinmetz, and Fisch (1965) Gordon (1959); Bowers (1961a) Case 4 Bolanle (1963) Case C.R.; Bolande and Tucker (1964) Case C.R. Favara, Nomura, and Bowden (1963); Bowden, Favara, and Donahoe (1965) Van Buchem (1958) Case 1; Van Buchem (1959) Case 7 Raghib <i>et al.</i> (1965) Shankar <i>et al.</i> (1967)	No Bowers (1961a) Fig. 1 No Miller and Pearson (1959) Fig. 4 No Vivas-Salas and Sansón (1948) Fig. 4 Wunsch <i>et al.</i> (1965) Fig. 3 Bowers (1961a) Fig. 2 No Bowden <i>et al.</i> (1965) Fig. 1A Van Buchem (1958) Fig. 4; Van Buchem (1959) Fig. 8 Raghib <i>et al.</i> (1965) Fig. 1 Shankar <i>et al.</i> (1967) Fig. 2A	* Not suitable for reproduction * Two tracings available; one with sinus rhythm; other, taken 5 years later, shows atrial fibrillation; latter tracing is considered here * Not suitable for reproduction — * Not suitable for reproduction — * Leads I, II, III, and IV only * Leads I, VI, and V6 only Possibly made 5 years before patient's death — —

* Obtained by personal communication.

TABLE II

ABNORMALITIES OF REPOLARIZATION IN ELECTROCARDIOGRAMS FROM PATIENTS WITH MARFAN'S SYNDROME AND PRIMARY ABNORMALITIES OF MITRAL VALVE

	ST segment depression	T wave inversion
Lead II	9/12	8/12
Lead III	9/12	8/12
Lead aVF	6/8	5/8
Left precordial leads	7/12	7/12

Note: In each instance, the first figure indicates the number of electrocardiograms showing the abnormality, the second figure the number of electrocardiograms in which the lead was recorded.

the pattern of ST segment depression and T wave inversion in leads II, III, and aVF.

Because the electrocardiograms from some of these 13 patients consisted of fewer than the now standard 12 leads, and because it could not be determined whether or not these patients were taking digitalis at the times these electrocardiograms were recorded, one can do no more than speculate on the cause of the ST segment and T wave abnormalities in leads II, III, and aVF in these electrocardiograms. It is thought probable that the abnormalities of repolarization in these leads were due to right ventricular hypertrophy, digitalis, or both.

Patients with abnormalities of both the aortic and mitral valves have been excluded from this study. One might expect to see in such patients with bivalvular disease the electrocardiographic patterns indicative of left ventricular hypertrophy or combined right and left ventricular hypertrophy. The potential value of the electrocardiogram in distinguishing between isolated mitral valve abnormalities and combined mitral and aortic valve abnormalities

in patients with Marfan's syndrome remains for further study.

SUMMARY

Electrocardiograms from 13 patients meeting stringent criteria for the diagnoses of (a) Marfan's syndrome and (b) primary abnormalities of the mitral valve (that is, not secondary to aortic disease) have been reviewed.

All 13 electrocardiograms were abnormal. The most common single abnormal pattern was ST segment depression and T wave inversion in leads II, III, and aVF.

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