An Electrocardiographic Pattern Associated with Mitral Valve Deformity in Marfan’s Syndrome

By Dorrance Bowers, M.D.

In a recent review of the electrocardiograms of 55 patients with Marfan’s syndrome, a pattern of S-T depression or T-wave inversion in leads II, III, and aVF* was observed in 7 patients. Subsequently, it was found that in 2 of these 7 patients (previously coded as M.Mc.G. no. 45 and as W.H. no. 50), deformity of the mitral valve had been demonstrated at the time of necropsy examination. This observation prompted a review of the electrocardiographic findings in other patients with Marfan’s syndrome and proved deformity of the mitral valve. It has been readily possible to collect from recent medical literature 3 additional examples of mitral valve deformity in patients with Marfan’s syndrome. In these 3 patients also, there were abnormalities of repolarization in electrocardiographic leads II, III, and aVF.* It is the purpose of this collective review to document briefly this association between electrocardiographic and anatomic abnormalities.

Report of Cases

Case 1

(Previously coded as M.Mc.G. no. 45, this patient was illustrated by McKusick in figure 1c in the first edition2a of his monograph and in figure 4e in the second edition;2b his code number A92675.)

This white girl was 10 years old at the time of her death at the Hospital for Sick Children* in Toronto. She presented the following stigmata of Marfan’s syndrome: bilateral ectopia lentium, scoliosis, and pectus excavatum. Electrocardiograms made on July 18, 1951, when the patient was 5 years of age, and on November 9, 1956, shortly before her death, are presented in figure 1. At necropsy examination the mitral valve was found to have 3 cusps and abnormally short chordae tendineae.

Case 2

(Previously coded as W. H. no. 50.) This white man with Marfan’s syndrome was studied at the Toronto General Hospital.4 An electrocardiogram made on August 30, 1954, when the patient was 19 years old showed atrial fibrillation and S-T depression and T-wave inversion in leads II, III, and aVF. At the time of necropsy examination it was observed that the endocardium of the left atrium and over the anterior leaflet of the mitral valve was abnormally thick and yellow; the mitral valve orifice measured 17 cm. in circumference (the normal value for this measurement is 7.5 cm.)

Case 3

(This patient was reported5 from the United States Naval Hospital, Bethesda, Maryland, by Drs. Russell Miller, Jr. and R. J. Pearson, Jr.)

This Negro patient was 27 years old at the time of her examination. She had bilateral ectopia lentium, pectus carinatum, kyphoscoliosis, and arachnodactyly. Her electrocardiogram (presented in this figure 4 by Miller and Pearson5) in addition to other abnormalities, shows S-T depression and T-wave inversion in leads II, III, and aVF. In this patient, at the time of necropsy examination, it was observed that the anterior mitral cusp was thickened, calcified, and studded with vegetations; the chordae tendineae were thickened, and one of the chordae had ruptured close to its attachment to the valve leaflet.

Case 4

(This patient was reported6 from the Jewish Hospital, Louisville, Kentucky, by Dr. Abraham Gordon and coded as no. 32436.)

This patient was a 24-year-old white man at the time of his death. The lens had been surgically removed from his right eye; the left lens was dislocated upwards. In addition, he had a moderate pigeon-breast deformity. There was an extensive family history of Marfan’s syndrome. An electrocardiogram (kindly supplied7 to the author by Dr. Gordon) made on November 29, 1946, is reproduced in figure 2. At the time of necropsy examination the superior half of the mitral valve proved to be densely adherent to the endocardial surface of the left ventricle anteriorly, and at the upper margin of the valve bony spicules protruded into the ventricular cavity. On microscopic examination the mitral valve proved to be hyalinized and acellular.

*When lead aVF was not recorded, the author has assumed that the pattern of repolarization in lead aVF would have been similar to that in leads II and III.

From the Knox Clinic and Kelowna General Hospital, Kelowna, British Columbia, Canada.
Figure 1
Electrocardiograms from case 1. The tracing on the left was made on July 18, 1951. Note the sinus rhythm, the S-T depression in lead II, and the inversion of the T waves in leads III and aVF. The tracing on the right was made on November 9, 1956. Note the atrial fibrillation and the S-T and T-wave abnormalities in leads II, III, and aVF.
Case 5

(This patient was reported from Johns Hopkins Hospital, Baltimore, Maryland, by Dr. Victor McKusick. She was coded as M.E.C. (J.H.H. A98174) and described on page 60 in the first edition of his monograph, and on page 91 in the second edition. She has also been mentioned in 2 other reports.

This patient was a 15-year-old white girl at the time of her death. She was tall and slender, and had bilateral ectopia lentium and scoliosis. Her electrocardiogram showed changes in the ST-T complexes in leads II and III interpreted as "right ventricular strain pattern." At the time of necropsy examination the mitral valve was found to have 5 cusps with nodular thickening along the line of closure. On microscopic examination this area showed basophilic degeneration.

Discussion

Deformity of the mitral valve in a patient with Marfan's syndrome was first recorded by Salle in 1912. Subsequently, Traisman and Johnson and McKusick have emphasized that mitral valve abnormalities occur not infrequently in patients with Marfan's syndrome. In some of these patients the mitral valve deformity is probably an intrinsic part of the genetically determined abnormality of connective tissues; in others, the mitral valve abnormalities appear to have resulted from complicating rheumatic fever or subacute bacterial endocarditis. Since both rheumatic fever and subacute bacterial endocarditis may pass unrecognized during life, one can only speculate, at the present time, on the relative importance of genetic and environmental factors in the causation of the mitral valve deformities in these patients.

The pathogenesis of the S-T and T-wave abnormalities in the electrocardiograms of these 5 patients is also not clear. Barnes (prior to the introduction of unipolar electrocardiographic leads) noted that S-T depression or T-wave inversion in standard leads II and III was often associated with right ventricular strain or with the administration of digitalis. Presumably these same factors were operating in the patients described in this report; but again, one can only speculate on the relative importance of right ventricular hypertrophy and administration of digitalis in the genesis of the electrocardiographic abnormalities in these 5 patients.

It is of interest that, in this series of patients, the significant clinical cardiovascular abnormality was mitral valve insufficiency. In a review of the electrocardiograms of 23 patients with "pure mitral insufficiency," S-T and T-wave abnormalities in leads II, III, and

*It is the author's presumption that this pattern consisted of S-T depression or T-wave inversion.

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aVr were not mentioned. Thus, the electrocardiogram may serve to distinguish between the mitral valve insufficiency of rheumatic heart disease and of Marfan's syndrome.

In recent years aortic valvar insufficiency in Marfan's syndrome has attracted wide interest.16 The electrocardiographic pattern of left ventricular hypertrophy is to be expected in such patients.1 In a patient with Marfan's syndrome, S-T depression or T-wave inversion in electrocardiographic leads II, III, and aVr should arouse suspicion of mitral valve deformity.

Summary

A collected series of 5 patients with Marfan's syndrome is reported in whom S-T depression and T-wave inversion in electrocardiographic leads II, III, and aVr were associated with necropsy-proved deformity of the mitral valve.

References


In so complicated a subject as the animal body, all things are wisely adjusted in number, weight and measure, yet with such complex circumstances as require many data from experiments, whereon to found just calculations: but though many of the following calculations are founded only on such inaccurate mensurations as the nature of the subject would allow of; yet may we thence fairly draw many rational deductions in relation to the animal oeconomy.—STEPHEN HALE, B.D., F.R.S. Haemastatics. Preface, Ed. 3.
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