Chronic Atrial Flutter in Brothers with the Marfan Syndrome

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Structural defects of the heart or great vessels are common in patients with the Marfan syndrome. The precise incidence is unknown, partly because of difficulty in defining forms frustes, but it is probably between 30 and 60 per cent. The electrocardiogram is also frequently abnormal, and several varieties of arrhythmia or conduction defect have been described. However, we have found no reference to the occurrence of atrial flutter.

In this paper we record the presence in each of two teenage brothers with the Marfan syndrome of chronic atrial flutter with no evident associated structural cardiovascular defect.

Case Reports

Case 1

A young man (P.W.) aged 19 years had severe bilateral foot-drop caused by weakness of the anterior tibial muscles. He had previously undergone uneventful operations for triple arthrodesis of the right foot and for transplantation of tendons. In June 1961 he was readmitted to the hospital for the first of similar operations on the left foot, but was found to have a highly irregular pulse rate. He was digitalized and discharged home. The arrhythmia persisted and he was referred 2 months later for further management to the West Virginia University Hospital.

He was a tall thin youth with a pinched adenoidal facies, a receding mandible, an extremely high and narrow palate, an elongated head and mild dysarthria. His eyes were normal. The subcutaneous tissue was sparse and muscle tone was poor. The feet were long and thin; there was pronounced foot-drop on the left side and scars of previous operations were noted on the right side. The chest was flat in the posteroanterior diameter. The fingers were long and slender. The pulse was highly irregular. There was an early systolic ejection murmur along the upper left sternal border. This was quite variable in intensity from time to time, almost disappeared during a full inspiration, and was felt to be related to the flatness of his thorax. The electrocardiogram (fig. 1, bottom) showed atrial flutter with an irregular ventricular response and occasional ventricular ectopic beats. X-ray films of the chest showed a slender cardiac silhouette with no enlargement of any chamber and with normal vascular markings in the lung fields. The metacarpal index2 was 10.0.

He was given full doses of digoxin and then quinidine; after receiving 1.4 Gm. of quinidine he suddenly reverted to sinus rhythm. He was discharged from the hospital, maintained on digoxin, and subsequently had an operation for triple arthrodesis of the left foot and transfer of the peroneal and posterior tibial tendons to the dorsum of the left foot.

In July 1963 his rhythm reverted to atrial flutter, a few days after he had stopped taking digoxin as a protest against his parents' unwillingness to purchase an automobile for him. He was readmitted to the hospital but the flutter persisted, despite full doses of digoxin and quinidine. At his next visit to the out-patient clinic, however, normal sinus rhythm was present and has persisted since then.

Case 2

A younger brother (J.W.) aged 17 years, was seen while visiting P.W. during his first admission to this hospital. His appearance was almost identical. He was tall and slender, with a long narrow face, a high arched palate, crowded teeth, nasal speech, weakness of the back muscles and of the dorsiflexors of the feet, and high pedal arches. The eyes were normal. His pulse was highly irregular, and an electrocardiogram (fig. 1, top) showed coarse atrial flutter with a variable ventricular response. There were no cardiac murmurs. X-rays confirmed the presence of a long, flat thorax, with normal cardiovascular silhouette and pulmonary vascular markings. The metacarpal index was 9.5. He too had been seen by
orthopedic surgeons, who thought that the deformities of the feet were insufficient to require braces or corrective surgery. He was admitted to the hospital and conversion to normal sinus rhythm was achieved with the use of digitalis and quinidine. He has cooperated in his continued treatment and has remained in sinus rhythm up to the present time.

Discussion

Bowers emphasized the relative frequency of abnormalities of the electrocardiogram in patients with the Marfan syndrome. In most cases they can be explained by coexistent major structural defects of the heart or aorta. Thus, the most common pattern is that of left ventricular hypertrophy, which results from aortic valve regurgitation. The second most common pattern, consisting of depression of the S-T segment or inversion of the T wave in leads II, III, and aVF, probably reflects right ventricular or biventricular hypertrophy or digitalis effects.

Several instances of atrial fibrillation, paroxysmal supraventricular tachycardia, bundle-branch block, and complete heart block have been recorded. McKusick suggested that these arrhythmias also are probably the functional consequence of underlying lesions such as aortic valve regurgitation and atrial septal defect, and in most instances this appears to be a reasonable explanation.

Both brothers reported here had classical features of the Marfan syndrome, including dolichocephaly, arachnodactyly, the metacarpal indices being 10.0 and 9.5 as compared with the normal range of 5.5 to 8.0, (3) high narrow arched palate, (4) pectus planum, (5) weakness of muscle groups, and (6) deformities of the feet. Neither had ectopic lenses. However, this finding is not a sine qua non for the diagnosis. The presence of atrial flutter with an irregular ventricular response was detected in the elder brother during a physical examination performed as a preliminary to an orthopedic operation. The identical arrhythmia was detected in the younger brother during a subsequent survey of close relatives.

Atrial flutter is rare in perfectly normal hearts. Its occurrence in two teenage brothers with an heredofamilial disorder therefore is presumptive evidence of underlying cardiovascular disease. Since neither brother has evidence of any gross structural defect of the
heart or major vessels, the basis for the arrhythmias may be an alteration in the fine structure of the atrial myocardium. This hypothesis is supported by the recent demonstration at necropsy, in two patients with the Marfan syndrome, of pathologic changes in the small arteries supplying the sinoatrial and atrioventricular nodes.

Summary
Chronic atrial flutter was found in two teenage brothers with classical features of the Marfan syndrome. Since neither patient has a gross cardiovascular defect, it is postulated that the arrhythmias may be secondary to an alteration in the fine structure of the atrial myocardium.

References

Enigmas
Consider such common and serious diseases as exophthalmic goitre, diabetes, and disseminate sclerosis. They have been under investigation a long time and with an increasing refinement of method, yet, however much we may have learnt of their mechanism, we still know nothing about their ultimate causation. Are they likely to be brought under causal mechanisms we already know, or are they and other diseases in a similar situation, like the residuary ‘bubble of air’ in Henry Cavendish’s eudiometer, awaiting for their explanation some radical fresh development? In answer to these questions we cannot make even a plausible guess.—The Collected Papers of Wilfred Trotter, F.R.S. London, Oxford University Press, 1946, p. 139.
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