

# INTERESTING ELECTROCARDIOGRAM

## EBSTEIN'S ANOMALY AND PRE-EXCITATION OF WPW TYPE

By M. Irene Ferrer, M.D.\*

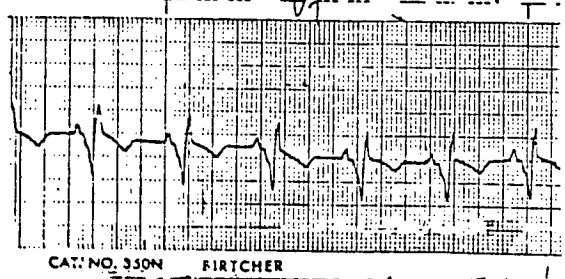
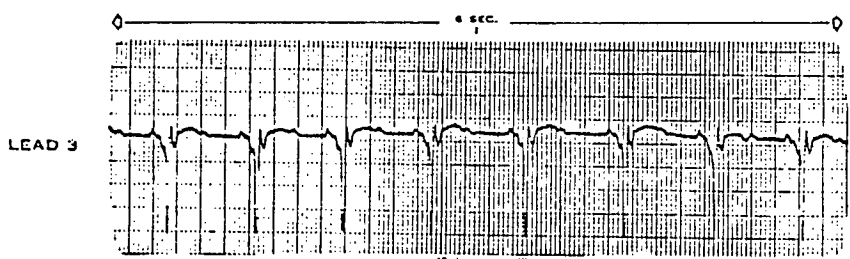
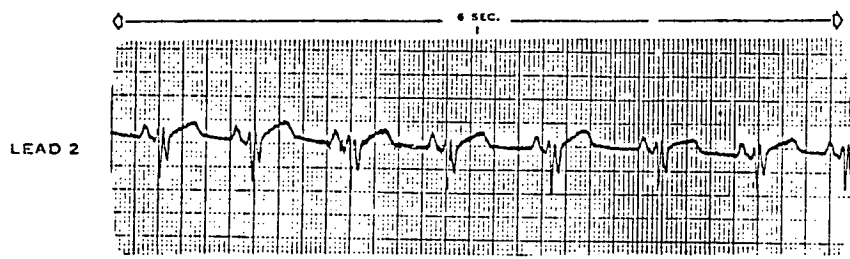
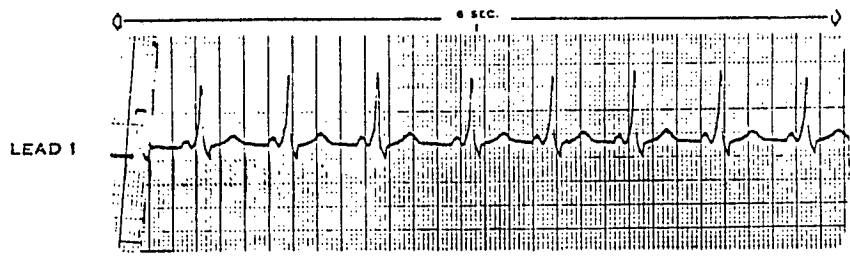
The two electrocardiograms shown here were obtained on an 18-year old male insurance applicant. He had a history of palpitations and in February 1981 was investigated by an internist who obtained a resting ECG (Figure 1) and a tracing during one of his spells of palpitations (Figure 2). This material was made available to the Journal by Dr. D. M. Fleming, Medical Director, North American Life Assurance Company, Toronto, Canada.

Figure 1 shows sinus rhythm and the short PR and prolonged QRS of pre-excitation of the Wolff-Parkinson-White-type. The QRS configuration, especially as seen in V1-V6, identifies this as a bypass tract located in the posterior septal part of the heart (in the old terminology, a type A). Noteworthy also in this record are the peaked tall and widened P waves (see leads II and aVF) and a QR in V1. Both these features strongly suggest Ebstein's Anomaly is present. During the supraventricular tachycardia (Figure 2) there is, of course, a different pathway of excitation to the ventricles, - i.e., no pre-excitation, and therefore the QRS is different in form. The arrhythmia is atrial flutter with 2:1 AV response, an atrial rate of 272 and a ventricular rate of 136 per minute. At first glance the QRS during flutter might appear wide if one overlooked lead I where clearly the QRS is normal in duration. Leads II and III display QRS complexes which contain a flutter wave and hence appear, but are not, wide. This is clarified by V1 where the first upward wave of the QRS (marked with an f) is a flutter wave as its form is the same as the second flutter wave seen between the QRS complexes. Of special importance during flutter is the finding of a marked left axis deviation of -107 degrees and no evidence of any IV conduction defect such as RBBB. This marked left axis deviation is seen in Ebstein's Anomaly because there is a vestigial, or very small and underdeveloped, right ventricle in these patients, allowing all the anterior forces of the left ventricle to be virtually unopposed.

Hence review of these two electrocardiograms suggests the combined diagnosis of pre-excitation and Ebstein's Anomaly. The association of pre-excitation and Ebstein's Anomaly is not uncommon, with an incidence ranging from 6 to 26 percent (1). The bypass tracts in Ebstein's Anomaly are always right-sided, as one would expect with a congenital defect involving the tricuspid valve and right AV ring. Pre-excitation is a congenital defect due to faulty formation of the AV ring.

*Ebstein's Anomaly* is an uncommon congenital heart defect characterized by malformation and downward displacement into an underdeveloped right ventricle of the septal and posterior leaflets of the tricuspid valve. This valve may also have a balloon-like and enlarged anterior leaflet as well as be fenestrated and deformed. Palpitations may occur even when pre-excitation is not an associated finding. The diagnosis can be suspected from the electrocardiogram but is confirmed by echocardiogram or angiography of the right side of the heart.

The prognosis of Ebstein's Anomaly has recently been greatly clarified allowing a new view of these subjects in relation to life insurance. There is a rather wide spectrum of function in this Anomaly. When the Anomaly is seen in infancy and childhood, one-half of the cases have an interatrial communication (patent foramen ovale or ostium secundum interatrial septal defect). They rarely have an associated ostium primum, pulmonic stenosis or atresia or IV septal defect. In childhood the symptoms come early in life and consist of the appearance of cyanosis and congestive heart failure. The ECG usually shows a prolonged PR, giant P waves and RBBB. If pre-excitation is also present, its ECG features are seen. These young patients will probably not be involved in seeking insurance unless surgically treated.



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Figure 1

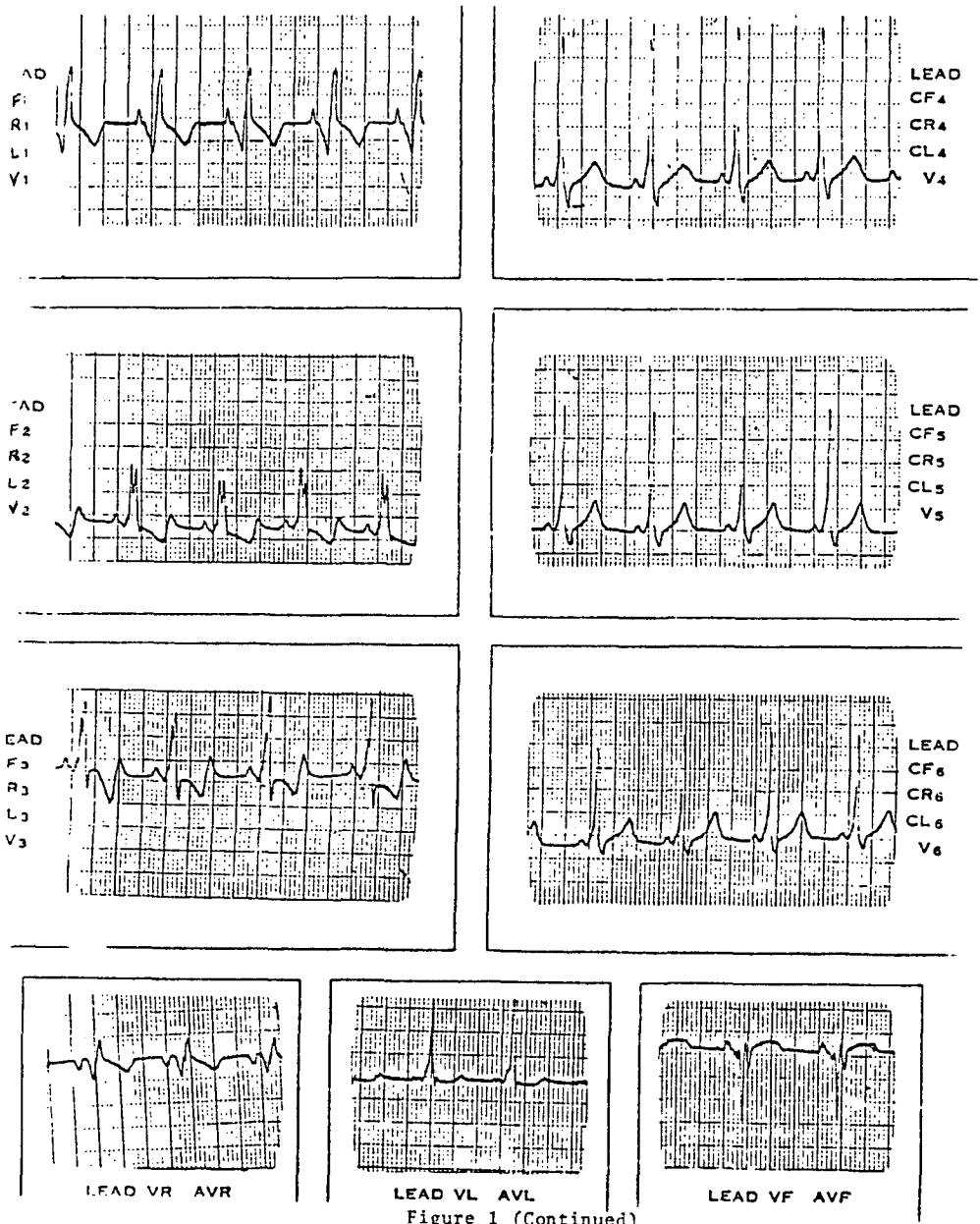


Figure 1 (Continued)

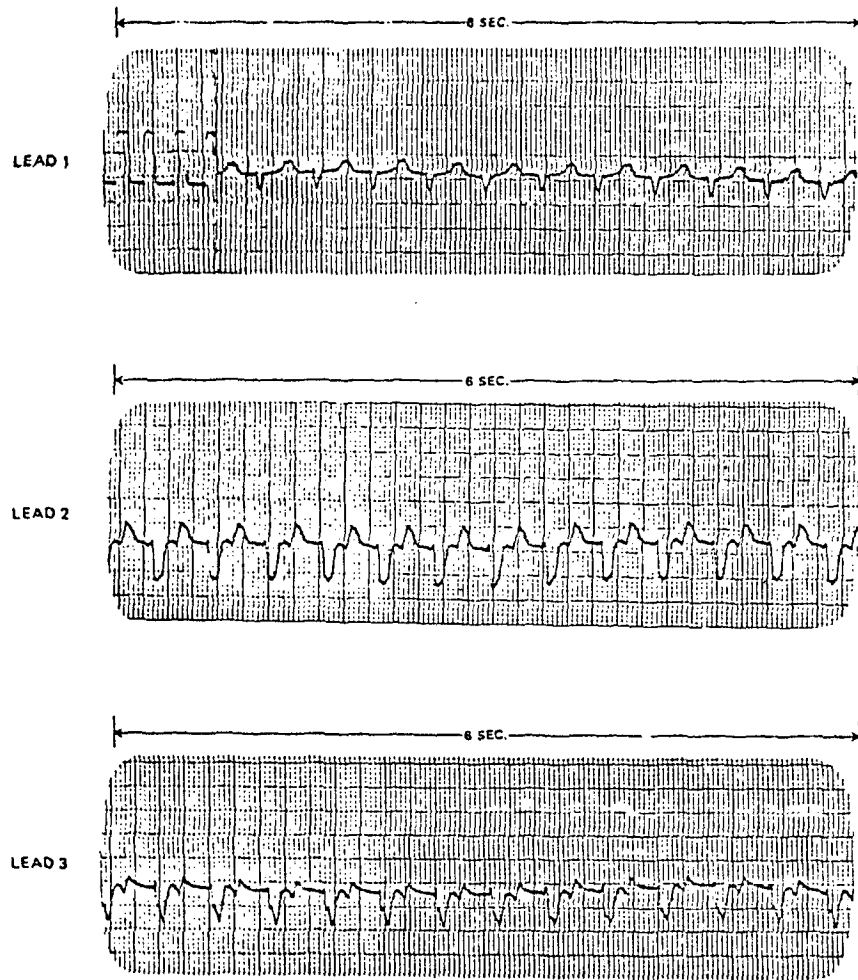


Figure 2

Beyond childhood, the diagnosis seldom depends on symptoms unless palpitations occur. The appearance of dyspnea, fatigue and cyanosis may be insidious and occur late. The syndrome in the adult is compatible with long life however even though 75 percent of adults have some cyanosis, either only on exertion or mildly at rest. The functional state of the hypoplastic right ventricle is the key to the prognosis. If relatively good, a normal life expectancy is possible with some patients reaching 79 years. If arrhythmias occur the outlook is less favorable if

ventricular rates are very rapid. If bypass tracts co-exist, these can be treated and ablated in subjects with serious arrhythmias (1) using modern surgical or laser beam techniques. Thus it is possible today to make a reasonable insurance offer to such patients when a full description of their cardiac status is available. The anatomy and function of the right ventricle can be assessed by ehocardiogram, angiogram and, as functional evaluation, the right ventricular ejection fraction can be measured using the newer radionuclide (technitium-99m) myocar-

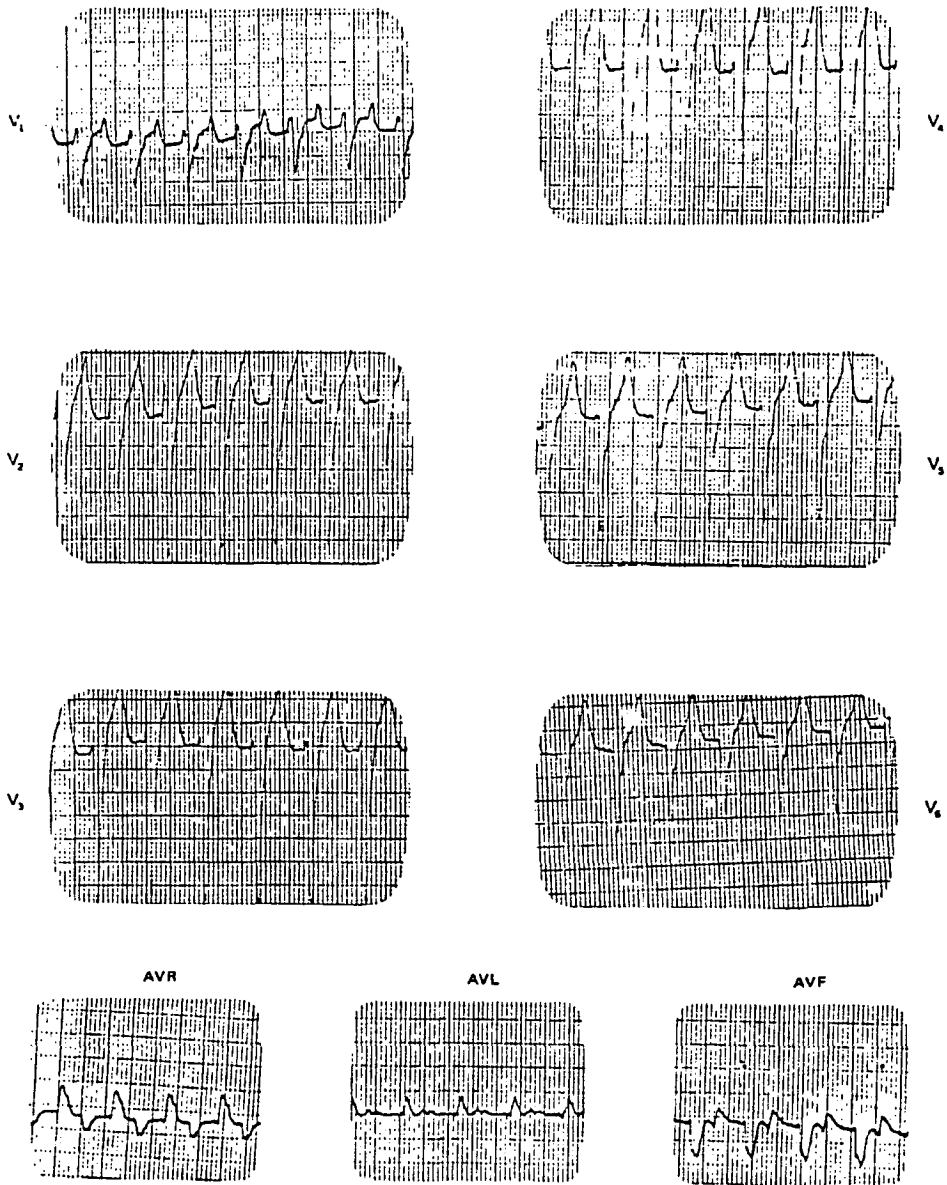


Figure 2 (Continued)

dial imaging techniques. A full appraisal of right ventricular function and the role of arrhythmias will then allow a good insurance evaluation.

REFERENCE(1) Smith, W.M., Gallagher, J.J., Kerr, C.R., Sealy, W.C., Kasell, J. H., Benson, Jr., D.W., Reiter, M.J., Sterba, R., Grant, A.O. *The Electrophysiologic Basis and Management of Symptomatic Recurrent Tachycardia in Patients With Ebstein's Anomaly of the Tricuspid Valve.* *Am. J. Cardio.* 49: 1223, April 1982.

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