Marfan's Syndrome Associated with Wolff-Parkinson-White Syndrome Type B

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SUMMARY

A case of Marfan's syndrome associated with Wolff-Parkinson-White (WPW) syndrome is described and a discussion follows on the possible relationship of the two conditions. Isolated cases of such an association have been previously reported.1,2) It is possible that the association is more common than was previously recognized and the two conditions may share a common pathological basis. Moreover, WPW syndrome may contribute to the cardiac morbidity of Marfan's syndrome.

Additional Indexing Words:
Marfan's syndrome    Wolff-Parkinson-White syndrome

MARFAN'S syndrome is a generalized disorder of connective tissue involving mainly the skeletal, ocular and cardiovascular systems. Structural cardiovascular changes are well recognized. Cardiac conduction defects are less widely seen and Marfan's syndrome associated with Wolff-Parkinson-White syndrome has been previously reported twice.1,2) Such a case is presented and a discussion follows.

CASE REPORT

A 26 year old Caucasian female was routinely referred for a cardiology consultation since she was 18 weeks pregnant and known to have Marfan's syndrome with aortic regurgitation. Her past medical history was uneventful; she had one normal pregnancy and delivery 2 years earlier. She had no symptoms that could not be accounted for by a normal second trimester pregnancy. She had never had syncopal episodes or palpitations. Her father and grandfather had both had Marfan's syndrome; no further details were known.

Physical examination revealed a Marfanoid habitus; she was 179 cm in
height which was over the upper third centile for her weight. She had
arachnodactyly and a high arched palate. Cardiovascular examination re-
vealed a grade 2/4 early diastolic decrescendo murmur maximal at the lower
left sternal border that was loudest on expiration, and a grade 2/6 late systolic
murmur maximal at the apex and radiating into the axilla, again loudest on
expiration. Pulse and blood pressure were normal. Abdominal examina-
tion showed an 18 week gravid uterus. There were no other abnormalities.

ECG showed a PR interval of 0.10 sec, delta waves in the QRS complex
and a predominantly negative deflection in lead VI—consistent with Wolff-
Parkinson-White syndrome type B. Echocardiography confirmed mitral valve
prolapse and aortic regurgitation due to outflow tract dilation. The patient
was followed by the cardiologists.

DISCUSSION

Cardiovascular complications of Marfan's syndrome occur in 30–60% of
cases depending on the series studied. Several complications have been re-
ognized. Mitral valve disease with regurgitation is found in nearly 50% of
cases. Most commonly this is mitral valve prolapse, however, severe mitral
regurgitation may develop requiring valve replacement.

In adults dilation of the aortic ring with aortic regurgitation and slowly
progressive left ventricular failure can occur. Chest pain with anginal fea-
tures may be secondary to aortic insufficiency, dilation of the aorta or coronary
ostial involvement. Dissecting aortic aneurysm is a common finding. Infec-
tive endocarditis of the mitral valve in not uncommon and has a significant
mortality.

Rarer complications have been described and include:
1. Aneurysmal dilation of the sinuses of Valsalva and pulmonary ar-
teries.
2. Cystic medial necrosis of the extrathoracic arteries.
3. Aneurysmal dilation and rupture of the intraventricular septum.
4. Endocardial fibroelastosis—like changes in the endocardium.
5. Coarctation of the aorta.

Cardiac conduction defects are also recognized in Marfan's syndrome.
James et al (1964), described 2 cases of non-specific cardiac conduction ab-
normalities, both of which included prolonged PR intervals—the opposite of
the change seen in Wolff-Parkinson-White syndrome. In both cases arrhy-
thmias resulted in sudden death and post mortem examination showed prolif-
erative changes in the tunica media of the nutrient arteries of the sinus node
and the atrioventricular node.
In Wolff-Parkinson-White syndrome most patients have normal hearts on gross examination. However, cardiomyopathies are occasionally seen. Possibly, an accessory pathway due to some muscular ischemia may have been present in the case discussed here, although there is no direct evidence for such a mechanism.

Wolff-Parkinson-White syndrome is also associated with mitral valve prolapse. The mechanism of this is not clear, but since diastolic depolarization of muscle fibers in the anterior mitral leaflet in response to stretch has been demonstrated experimentally, the abnormal stretch of the prolapsed leaflet may be of pathogenic significance. Clearly, the same process may be occurring in Marfan's syndrome.

Thus, to conclude, Wolff-Parkinson-White syndrome in this case of Marfan's syndrome may have three explanations:
1. Presence of mitral valve prolapse causing an abnormal conduction pathway.
2. Coincidental presence of the two conditions.
3. Intrinsic cardiac muscle ischemia due to vasculitis producing aberrant cardiac conduction—unlikely.

Further studies may produce a clearer understanding of this complex subject.

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References

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