Adams-Stokes attack in the patient with Congenital Complete A-V Block - 2009

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Because of initial Adams-Stokes attack in the patient with Congenital Complete A-V Block may (CCAVB) sometimes prove fatal, there is a need to be able to identify the patient at great risk. 24 children with CCAVB were followed up for 1 to 19 years to determine the efficacy of current methods of predicting risk for Adams-Stokes syncope and the usefulness of pacemaker therapy in relieving symptoms.

The HR at rest, configuration of surface ECG complexes, data obtained during intracardiac Electrophisiologic Study (EPS) and response to graded treadmill exercise testing were compared in children with and without syncope.

One or more Adams-Stokes episodes were experienced by 8 children, 1 of whom died. Only a persistent HR at rest ≤ 50 beats/ min demonstrated any significance. Intracardiac EPS was of little benefit because of site of block did not correlate with syncope.

Although the increase in HR during treadmill exercise testing showed no correlation with prevalence of syncope or location of block, exercise-induced PVCs may have predictive value in older children and young adults.

Ventricular pacemakers were implanted in 10 children. Each child was asymptomatic over a 1 to 10 year follow-up period.

Because extreme bradycardia may contribute to the prevalence of Adams- Stokes attacks in children with **CCAVB**, careful evaluation of HR at rest may be an effective means of differentiating patients at risk of syncope.

Pacemaker therapy is a feasible and effective method of treatment in young children and relieves symptoms (1). Risk for HF, syncope, and SCD is present at any age in **CCAVB** including fetal life.

Unfavorable prognostic signs in uterus are low and decreasing HR, hydrops, AV valve regurgitation, and low aortic flow velocity. Indications for pacing in infancy are CHF, HR < 55 bpm in isolated block and < 65 bpm with associated disease, prolonged OTc interval, syncope attacks, frequent PVCs, and alternating ventricular pacemakers.

Indications for immediate pacing in childhood and adult life are syncope, presyncope, HR lower than median for age, periods of junctional exit block, prolongation of QTc and mitral regurgitation, and change of ventricular pacemaker.

Pacing is recommended to all patients older than 15 years (2).

The prognosis of **CCAVB** is usually considered favorable in adults. This belief is based on studies comprising a limited number of patients and with rather short observation times.

The natural history of the disease was investigated by a prospective follow-up through decades of adult life of patients with a large group having well-defined permanent **CCAVB** (your case is intermittent).

Patients all without symptoms during their first 15 years of life, were selected. Because of the high incidence of unpredictable Stokes-Adams attacks with considerable mortality from first attacks, a gradually decreasing HR, significant morbidity, and a high incidence of "acquired" mitral insufficiency.

Prophylactic PM Treatment is recommended even for symptomfree adults with **CCAVB** because of the high incidence of unpredictable Stokes-Adams attacks with considerable mortality from first attacks, a gradually decreasing HR, significant morbidity, and a high incidence of "acquired" mitral insufficiency (3).

References

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