Pacemaker Implantation in Familial Congenital A-V Block Complicated by Adams-Stokes Attacks

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Congenital complete heart block is rarely accompanied by Adams-Stokes attacks (Campbell and Thorne, 1956; Wood, 1956), and because of this an extrinsic cardiac pacemaker is seldom necessary. A case is reported of a boy with congenital complete heart block causing Adams-Stokes attacks; other members of his family also had heart block.

Case Report

A 2½ year-old boy was seen by one of us (O.V.) 6 days after he had suffered from a convulsive attack with loss of consciousness. He had previously been treated with anticonvulsive drugs for similar episodes without improvement. A striking clinical finding was an irregular bradycardia, with a heart rate of 20 a minute. The blood pressure was 110/60 mm. Hg. The venous pulse showed independent "a" waves and occasional "cannon waves". The apex beat was displaced 1 cm. to the left of the mid-clavicular line in the fifth intercostal space. On auscultation the first sound varied in intensity and was split; there was an ejection systolic murmur grade 1/6 at the left sternal edge; the second sound was widely split but moved normally with respiration. Occasional atrial sounds could be heard.

The chest x-ray film revealed moderate cardiac enlargement. The electrocardiogram demonstrated complete A-V block. Treatment was initiated with intravenous isoprenaline which produced an increase in heart rate to 40 a minute (Fig. 1). Nevertheless, further Adams-Stokes attacks occurred, and on two occasions cardiac arrest required external cardiac massage. It was therefore decided to implant an Electrodyn cardiac pacemaker utilizing intramyocardial electrodes. Pacing was satisfactory and 16 days later he was discharged free of symptoms with a heart rate of 88 a minute. Three months later he fell and fractured an electrode wire. Adams-Stokes attacks recurred within a week, and a

second thoracotomy was required to replace the wire. At the present time two years later the patient is symptom free, and pacing satisfactorily.

Family History. The boy's mother, aged 25, later presented with episodes of dizziness without loss of consciousness. Her electrocardiogram (Fig. 2) also showed complete heart block. There was no other cardiac abnormality. She is being treated with long-acting isoprenaline.

Three sibs are living. Two sisters, aged 5 and 1½, are well and have normal electrocardiograms. A brother, aged 7, has second-degree (2:1) A-V block (Fig. 3). He has had one Adams-Stokes attack and has received no treatment.

The mother had given birth to three other children, all boys. One was stillborn; another was born prematurely and died 5 days after birth; the third died at the age of 18 months in a convulsive attack, having had several convulsive episodes during the last 12 months of his life.

No member of the family, had a positive complement-fixation reaction of Machado Guerreiro modified by Freitas (1952) and Maekelt (1957) for Chagas' disease, and there is no clinical indication of an underlying cause.

Discussion

Adams-Stokes attacks in congenital complete heart block are uncommon. Keith, Rowe, and Vlad (1958), during 17 years at the Sick Children's Hospital of Toronto, found only 17 cases and none presented with Adams-Stokes attacks. In Wood's series of 15 cases (1956) none had syncopal attacks, and in Nadas' (1963) series of 27 cases only one had Adams-Stokes attacks.

The treatment of Adams-Stokes attacks in congenital heart block should be no different from the treatment for attacks occurring with acquired heart block (Zoll, Frank, and Linenthal, 1964; Glenn *et al.*,

FIG. 1.—Electrocardiogram of 2½-year-old boy showing complete A-V block. The atrial rate is 150 a minute, and the ventricular rate is 40 a minute. There is left axis deviation.

FIG. 2.—Electrocardiogram of boy's mother shows complete A-V block and left axis deviation.

FIG. 3.—Electrocardiogram strip of boy's brother lead II, showing 2:1 A-V block.

1964). The young age of the patient should not be a deterrent to implantation of a pacemaker if the need exists (Michaelsson and Petersson, 1965; Martin *et al.*, 1966). This patient satisfied the criteria for pacemaker implantation, namely heart block leading to Adams-Stokes attacks uncontrolled by drug treatment.

Of further interest is the fact that other members of the patient's family have complete heart block. The mother and one brother might well require pacemaker implantation in time.

Other families with this malady in various members have been documented in the past (Kay, 1948; Wallgren and Agorio, 1960; Nakamura and Nadas, 1964; Gazes *et al.*, 1965), but apparently in only one family (Kariv *et al.*, 1966) has a pacemaker been implanted.

Summary

A family is reported in which several members have complete heart block. Numerous Adams-Stokes attacks necessitated pacemaker implantation in one of them aged 2½ years.

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