ARTIFICIAL PACING FOR CONGENITAL COMPLETE HEART BLOCK*

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Most cases of congenital complete heart block do not require artificial pacing. In these cases the heart rate is normally faster than 60 beats/minute.[1,2] The rate and stroke volume can also be increased more during exercise than is the case with acquired complete heart block. These patients are asymptomatic. Stokes-Adams attacks are rare, and formerly it was stated that artificial pacing was never required.

More recently several instances[3,5] of severe symptoms due to heart failure, Stokes-Adams attacks, and even death in infants with extremely slow ventricular rates and otherwise apparently normal hearts, have been noted. For such children artificial pacing is a life-saving procedure. Disappearance of a congenital complete heart block was observed in one case only.[1]

The purpose of this paper is to review the clinical findings and operative procedure in an infant who required artificial pacing.

CASE HISTORY

On 19 June 1967 the patient's general practitioner noticed a slow foetal pulse of ± 50/min. Two days later the baby was delivered by a caesarean section as the gynaecologist thought the slow pulse rate was due to foetal distress. Immediately after birth the heart rate was 47/min. The infant was sent to another hospital for observation and was discharged home after 8 days. He developed normally until May 1968, when he was readmitted because of a cough and a history of syncopal attacks. His pulse rate this time was noted to be 15/min. at times during sleep, and 24/min. when awake. The syncopal attacks were attributed to the patient's chest infection. He was put on Ephedrine gr. ½ 12-hourly and later on gr. ¼ 8-hourly. He was discharged home after 3 weeks, the pulse rate being 30 - 40/min. (Fig. 1).

At home he again started having syncopal attacks, which increased in frequency. He also had a few convulsions, not associated with cyanosis or incontinence.

He was admitted to our intensive care unit on 11 July 1968.

He was a chubby, healthy-looking baby, with a pulse rate of 38/min. His blood pressure was 110/50 mm.Hg. There was a grade II systolic murmur at the apex. On X-ray there was a slightly enlarged cardiac shadow and

*Date received: 14 November 1968.

the ECG showed complete heart block, an atrial rate of 108/min. and a ventricular rate of 38/min. with a right bundle-branch pattern.

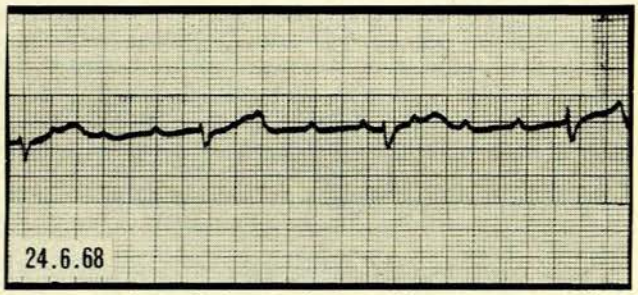

Fig. 1. Before discharge of the patient on conservative treatment with Ephedrine. The ECG shows complete heart block with atrial rate of 150/minute and ventricular rate of 43/minute.

Treatment

He was put on Saventrine ½ tab. 2-hourly. There was no increase in the pulse rate and it was decided later that same day to insert a temporary endocardial pacemaker. This was done transvenously, using the right external jugular vein. Many endocardial areas were tried but the threshold remained high, ranging from 0·9 to 3·0 volts. A stable site with a threshold of 1·2 volts was eventually selected.

On 15 July 1968 the threshold went up to 3·85 volts. The catheter was repositioned and the lowest threshold obtained was 2·4 volts.

On 18 July 1968 the threshold had gone up to 4·1 volts, although according to electrocardiographic analysis the pacemaker spike vector had remained the same, suggesting that the catheter had not changed its position. It was then decided to use epicardial electrodes for permanent pacing.

Operation—22 July 1968

The chest was opened through a left anterolateral thoracotomy below the sixth rib. The pericardium was split transversely to expose the heart. Two Cordis epicardial leads were attached to the left ventricle—one at the apex and the other in the mid-portion of the left ventricle. The distal ends of the electrodes were led through a tunnel passing from the pericardium into the subcutaneous tissue of the abdominal wall below the left costal margin.

The thresholds of both leads were tested separately and combined before they were connected to the pacemaker. The apical lead registered 0·8 volts, the mid-anterolateral lead 0·45 volts and the combined leads 0·8 volts.

A pocket was now created for the pacemaker in the subcutaneous tissue overlying the lateral border of the rectus sheath, and the pacemaker unit connected. Loops were created in both leads within the pericardium and the subcutaneous pocket to allow for future growth of the child. The wounds were closed separately.

The maximum output of the Devices St George's axilla pacemaker that was used was 3·8/3·6 volts.

The patient did well until the morning of 26 July 1968, when it was found that he was being paced intermittently every 3rd - 4th beat (Fig. 2).

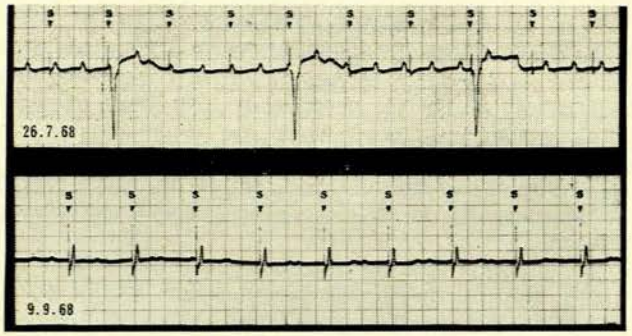

Fig. 2. Intermittent pacing showing the pacemaker spikes without subsequent ventricular depolarization. Ventricular rate 25/minute; atrial rate 160/minute.
Fig. 3. Successful pacing showing the pacemaker spikes and subsequent ventricular depolarization at a rate of 72/minute.

A temporary endocardial electrode was introduced, under local anaesthesia, via the right long saphenous vein, and the patient was paced on an external pacemaker. He was reoperated on that same afternoon as endocardial pacing had previously been found unsatisfactory.

The thresholds of the epicardial leads were tested before opening the chest. The apical lead registered 3·4 volts, the mid-anterolateral lead 2·0 volts and the bipolar (combined) lead 3·8 volts. It was decided to sacrifice the apical lead and to institute pacing, using the anterolateral lead and an indifferent electrode. The apical electrode was cut, insulated and tied off. The patient was then paced using an epicardial unipolar system with the indifferent electrode buried in the subcutaneous tissue. The threshold was now 1·5 volts, using this system. The patient returned to the ward and was pacing well (Fig. 3). Further tests in the ward showed no deterioration in the spike potential.

A month after discharge from hospital the patient was seen at the outpatient follow-up clinic and was doing well. He had had no more Stokes-Adams attacks, no apparent residual cerebral damage, and no deterioration in the spike potential.

The underlying aetiology of the heart block is obscure, but the presence of the exceptionally high threshold at all points within the right ventricular cavity might be taken to suggest a thickened endocardial surface due to a lesion such as fibro-elastosis.

DISCUSSION

The aetiology of congenital complete heart block is still obscure and causative factors vary considerably. Genetic influence has been reported; it is believed to be transmitted by an autosomal dominant trait with an incomplete but high degree of penetrance.[4] A study by Combrink *et al.*[5] showed all 4 siblings of a family to have varying degrees of right bundle-branch block.

Evidence of *in utero* infection as a causative factor has been suggested by necropsy studies which demonstrate scarring and calcification in the bundle of His.[6]

Congenital absence of the AV node[6,7] or a portion of the bundle of His[8] has been found by several investigators

after detailed histological examination of the conducting system in congenital complete heart block.

Underlying cardiomyopathy cannot be totally excluded.[9] In the case here presented, with an apparently normal heart but high endocardial threshold, the question of endocardial fibro-elastosis arises. Other congenital cardiac abnormalities are commonly found with congenital complete heart block, but that the latter is commonly associated with ventricular septal defect is untrue; although it is not uncommon with corrected transposition of the great vessels.

Complete heart block is very often missed in the infant because of a faster pulse rate.[1,2]

The first diagnosis of congenital heart block was reported by White and Eustis[10] in 1921. A slow foetal pulse was detected 6 hours before birth, and complete heart block was confirmed electrocardiographically immediately after birth. Nahamura and Nadas reported 5 cases diagnosed *in utero*.[3]

It is generally accepted that children with congenital complete heart block usually do quite well without symptoms or major difficulty. On the other hand, children with acquired complete heart block have a poor prognosis with a high incidence of fatal Stokes-Adams attacks.[2]

Review of the literature suggests that the prognosis in congenital heart block is related primarily to cardiac adaptation to stress and to the occurrence of Stokes-Adams attacks. Reports of mortality occurring in the newborn period suggest that infants with heart rates of less than 50 beats/minute are especially likely to succumb.

Therefore, implantation of a cardiac pacemaker in any child who has had two or more Stokes-Adams attacks should be strongly considered.[3,11] After persistent surgically induced heart block, if continuous oral treatment with Saventrine is not satisfactory, a transistorized pacemaker should be implanted before the patient is discharged from hospital.[12]

According to Nahamura and Nadas,[3] there are only 2 principal indications for therapy in congenital complete heart block. These are Stokes-Adams attacks and congestive cardiac failure. A cardiac pacemaker should be implanted if ordinary medical treatment fails.

The selection of electrode placement depends mainly on the endocardial threshold, as was seen in this case. Endocardial pacing is indicated with a normal endocardial threshold. Furthermore, in children with endocardial pacing there is always the possibility of movement of the electrode because of the activity of the child. Endocardial pacing can be done under local anaesthesia; epicardial pacing needs general anaesthesia and a thoracotomy. Because of the precision with which the electrodes can be placed on the epicardium, this is thought to be the site of choice. The problem of lengthening of the electrodes with future growth of the child has not yet been solved. Loops were created in the electrodes, as was mentioned earlier, in the hope that these will allow for future growth.

Another definite problem with permanent pacing in an infant is the possibility of wire fracture due to overactivity, as opposed to adults. It is therefore wise not to implant the pacemaker unit low down in the abdominal wall, in order to try to minimize flexion and extension of the electrodes.

SUMMARY

A case of congenital complete heart block which required artificial pacing is presented. The slow pulse rate was noted before birth, but was thought to be due to foetal distress. The diagnosis was made electrocardiographically only 2 days after birth.

Possible causes of congenital complete heart block are discussed and also the indications for artificial pacing in these patients.

The problems of future growth of the child and fracture of electrodes are pointed out and may be prevented through the way the electrodes and pacemaker unit are inserted.

REFERENCES

1. Campbell, M. and Emanuel, R. (1967): Brit. Heart J., **29**, 577.
2. Ayres, C. R., Boineau, J. P., Spach, M. S. and Durham, S. C. (1966): Amer. Heart J., **72**, 381.
3. Nahamura, F. F. and Nadas, A. S. (1964): New Engl. J. Med., **270**, 1261.
4. Gazes, P. C., Culler, R. M., Taber, E. and Kelly, T. E. (1965): Circulation, **32**, 32.
5. Combrink, J. M., Davis, W. H. and Snyman, H. W. (1962): Amer. Heart J., **64**, 397.
6. Latta, H. and Crittenden, I. H. (1964): Lab. Invest., **13**, 214.
7. Lev, M. (1964): Ann. N.Y. Acad. Sci., **111**, 187.
8. Yater, W. M., Lyon, J. A. and McNabb, P. E. (1933): J. Amer. Med. Assoc., **100**, 1031.
9. Wagner, C. W. jnr and Hall, R. J. (1967): Amer. J. Cardiol., **19**, 593.
10. White, P. and Eustis, R. (1921): Amer. J. Dis. Child., **22**, 299.
11. Veracochea, O., Zerpa, F., Morales, J., Hernandez, O. and Waich, S. (1967): Brit. Heart J., **29**, 810.
12. Cooley, D. A. and Hallman, G. L. (1966): *Surgical Treatment of Congenital Heart Disease*, p. 97. Philadelphia: Lea & Febiger.