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Postoperative complete heart block in children[1]

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Experience with 40 children who developed complete heart block after open intracardiac repair of congenital heart disease is described. The incidence of this complication over a twelve-year period fell from 25 to 10 per cent. The incidence of permanent complete heart block has fallen to less than 2 per cent.

The mortality rate in the immediate postoperative period (27·5%) was twice that of patients without complete heart block. Twenty-nine patients survived the immediate postoperative period. Eighteen (62%) reverted to sinus rhythm, 11 (38%) developed permanent complete heart block, and this group had a high mortality without pacemaker implantation. Of 9 medically treated patients, 6 succumbed (67%). The mode of death in all instances was a syncopal episode. Three patients have permanent pacemakers and lead active, normal lives.

The occurrence of an Adams-Stokes attack as an indication for pacemaker implantation was unsatisfactory because the first attack was fatal in 4 patients. Criteria such as the presence of residual lesions, response to sympathomimetic drugs, and the width of the QRS complex were also of limited usefulness.

The uncertainty of the prognosis has led to a policy of pacemaker implantation in all patients with surgically induced permanent complete heart block regardless of symptomatology.

Improved surgical techniques and better understanding of the anatomical pathways of the cardiac conduction system have led to a decreased incidence of surgically induced complete heart block (Gadboys and Litwak, 1964; McGoon, Ongley, and Kirklin, 1964). There is general agreement on the management of this condition in the immediate postoperative period, but the long-term use of pacemakers in children remains controversial (Hurwitz, Riemenschneider, and Moss, 1968; Murphy *et al.*, 1970). This paper describes our experience with 40 children who developed complete heart block after open intracardiac repair of congenital cardiac lesions. The usefulness of specific prognostic criteria are evaluated, and methods of treatment are compared.

Patients and methods

A study was made of 263 children with congenital heart disease who underwent open intracardiac repair from January 1957 to August 1969. The study was limited to those patients at risk for the development of complete heart block and included the following lesions: ventricular septal defect – 114, tetrad of Fallot – 117, endocardial cushion defect – 28, subaortic stenosis – 4. Forty of these patients developed complete heart block after operation. Criteria for inclusion into this group were the development of heart block during the surgical procedure and its persistence into at least the immediate postoperative period. Several patients developed transient complete heart block during operation, which reverted to sinus rhythm before they returned to the recovery ward, and are not included in this study.

Diagnosis of complete heart block was based on standard criteria (Massie and Walsh, 1960). Electrocardiographic tracings were obtained in all but two patients with a twelve lead recorder. In these two patients the diagnosis was made from an oscilloscopic tracing.

All patients still living have been evaluated recently by the authors. Patients who succumbed were all seen shortly before death, and in most cases at the time of death by one of the authors.

Cardiopulmonary bypass was accomplished with a Clark bubble oxygenator and pump (Clark *et al.*, 1958). In the first two years cardiac arrest was achieved with potassium citrate. From 1959 hypothermia was used routinely. The aorta was clamped for as long as 10 minutes at a time to achieve ischaemic cardioplegia.

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Before 1962 patients with complete heart block were managed in the immediate postoperative period with a continuous infusion of isoprenaline. From 1962 onwards when complete heart block occurred during operation, a conduction suture was sewn into the body of the right ventricle and the indifferent electrode placed subcutaneously in the anterior chest wall. These wires were attached to an external pacemaker unit[1] which was activated when cardiac pacing was necessary to achieve a satisfactory heart rate in order to maintain cardiac output and systemic blood pressure.

In this study permanent complete heart block is defined as that persisting beyond the third postoperative week. For permanent pacing a pacemaker[2] was implanted in the subcutaneous tissue of the axilla or the epigastrium. In small thin children we have on occasion placed the unit in the extrapleural space. The impulse was delivered either by leads sewn into the right ventricular myocardium or by a bipolar pacing catheter[3] inserted transvenously into the right ventricular apex.

Results

Incidence of complete heart block In the twelve-year period of 1957 to 1969 surgically induced complete heart block was encountered in 40 patients after 263 open intracardiac operations for a variety of congenital cardiac lesions (Table 1). This complication occurred most frequently with repair of endocardial cushion defects, especially if the ventricular septum was involved in the defect. Repair of isolated ventricular septal defects was associated with the lowest incidence of complete heart block. For all lesions there has been a progressive decline in the occurrence of complete heart block from 25 per cent in the early period of the study to the more recent figure of 10 per cent. The incidence of permanent complete heart block after operation has fallen from 12 per cent to less than 2 per cent (Fig. 1).

Prognosis of complete heart block in the early postoperative period The outlook for patients with complete heart block in the immediate postoperative period has altered significantly in the period under review (Fig. 2). There has been a fourfold increase in the number reverting to sinus rhythm and a corresponding fall in the incidence of permanent complete heart block. Less dramatic has been the decline in the mortality rate.

Of the 40 children with complete heart block, 11 died in the immediate postoperative

[1] External pacemaker, Model 5800, Medtronics Inc., Minneapolis, Minnesota, U.S.A.
[2] Model 5860, Medtronics, Inc., Minneapolis, Minnesota, U.S.A.
[3] United States Catheter Inc. Co., Glens Falls, New York, U.S.A.

TABLE 1 *Incidence of surgically induced complete heart block*

Lesion	*Number*	*Complete heart block*	*Per cent*
Atrioventricular canal (incomplete)	5	3	60
Ostium primum atrial septal defect	23	5	22
Tetralogy of Fallot	117	22	18
Ventricular septal defect	114	9	8
Subvalvular aortic stenosis	4	1	25
Total	263	40	

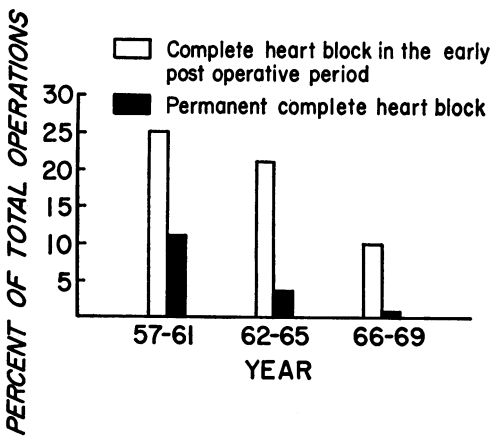

FIG. 1 *The fall in incidence of acute and permanent surgically induced complete heart block from 1957 to 1969.*

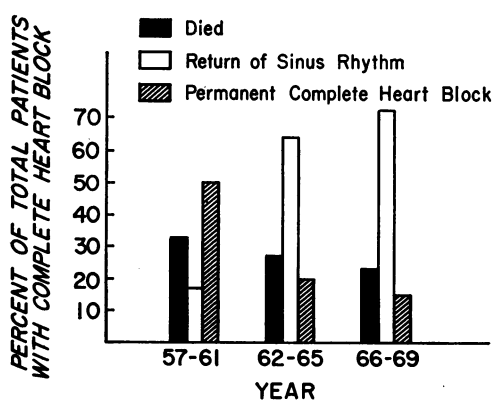

FIG. 2 *Prognosis of surgically induced complete heart block in the early post-operative period.*

TABLE 2 *Patients with permanent complete heart block*

Case No.	*Age (yr) and sex*	*Lesion*	*Year of operation*	*Residual defect*	*Adams-Stokes attacks*	*Cardiac failure*	*Long-term management*	*Outcome*	*Follow-up period*
1	9 F	Tetrad of Fallot	1957	Yes	One	Yes	Medical	Died	4 wk
2	2½ F	Ventricular septal defect	1957	No	None	No	Medical	Excellent	12 yr
3	5 F	Ostium primum atrial septal defect	1958	No	Several	Yes	Medical, then pacemaker	Fair	12 yr
4	9 F	Atrioventricular canal (incomplete)	1959	No	Three	No	Medical	Died	10 yr
5	9 F	Subvalvular aortic stenosis	1960	No	Two	No	Medical	Died	4 yr
6	4 M	Ostium primum atrial septal defect	1960	Yes	One	Yes	Medical	Died	7 yr
7	13 F	Ventricular septal defect	1962	No	None	No	Medical	Excellent	8 yr
8	13 M	Atrioventricular canal (incomplete)	1964	No	One	No	Medical	Died	4 wk
9	3 M	Ventricular septal defect	1967	Yes	One	Yes	Pacemaker, then medical	Died	7 mth
10	6 M	Ventricular septal defect	1968	No	None	No	Pacemaker	Excellent	1 yr
11	10 F	Tetrad of Fallot	1969	Yes	None	Yes	Pacemaker	Fair	8 mth

period. The primary cause of death was related to low cardiac output in 9, while the other 2 succumbed with Adams-Stokes attacks. Eighteen patients reverted to sinus rhythm, and all did so within the first three weeks (Fig. 3). Complete heart block persisted in the remaining 11 children.

Patients with permanent complete heart block The clinical data of the 11 children with permanent block are presented in Table 2. Nine children were managed with isoprenaline for a period of months to years. Seven were treated solely with isoprenaline while the other two (Cases 3 and 9, described below) were managed at different times with either isoprenaline or a pacemaker. One child (Case 9) had a pacemaker implanted four weeks after his surgical repair, but unfortunately the myocardial leads broke and he was subsequently treated with isoprenaline.

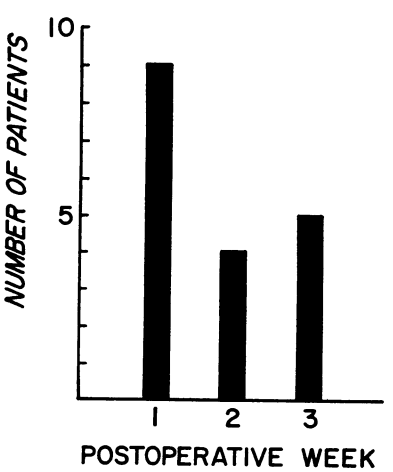

FIG. 3 *Time of reversion to sinus rhythm.*

The other child (Case 3) was managed with isoprenaline for five years during which time she had several Adams-Stokes attacks. A permanent pacemaker was subsequently implanted. She has been relatively well since though initially she required repeated surgical procedures to repair the myocardial wires. Of the 9 medically treated patients, 6 died (67%). One patient (Case 3) had a pacemaker implanted as described above, while the remaining 2 patients (Cases 2 and 7) continue to lead active, normal lives 8 and 12 years after operation. One (Case 7) is now 21 years old and recently had a healthy baby delivered by caesarean section. A temporary pacing electrode had been positioned transvenously before delivery, but pacing was not required.

Three patients (Cases 9, 10, and 11) had pacemaker implantation before leaving the hospital when it became obvious that their conduction disturbances were permanent. One of these children (Case 9) was described above and is included in the medically treated group because of his subsequent long-term management with isoprenaline. The other two at the present time lead active, normal lives.

Significance of Adams-Stokes attacks The two children (Cases 10 and 11) with permanent implanted pacemakers have not had Adams-Stokes attacks and are excluded. Seven patients with permanent complete heart block had one or more Adams-Stokes attacks (Fig. 4). These episodes generally occurred within one year of operation, but in two instances the first episode was delayed for 7 and 10 years, respectively. *The first Adams-Stokes attack was fatal in 4 patients* (57%). One of the three who survived the first episode was described above (Case 3). The other two are presented here.

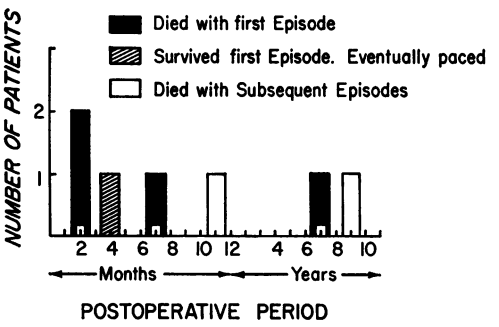

FIG. 4 *The time of occurrence and outcome of the first Adams-Stokes attack in 7 children with permanent surgically induced complete heart block.*

Case 4 had repair of an incomplete atrioventricular canal in 1959. Though left with permanent complete heart block, her subsequent course was benign until she had her *first syncopal episode 10 years after operation.* She refused a pacemaker, and 12 months later in the space of a few hours she had two Adams-Stokes attacks and died.

Case 5 had successful relief of discrete subvalvular aortic stenosis, but permanent complete heart block developed postoperatively. Fifteen months later she reported a transient feeling of faintness without loss of consciousness. We were reluctant to recommend permanent pacemaker implantation at that time especially in the light of a vague nonspecific complaint. She was asymptomatic for the next three years but then died suddenly and unexpectedly at home.

Significance of residual lesions Four patients with permanent complete heart block were considered to have haemodynamically significant lesions as judged clinically, by catheterization, or at necropsy: 3 had ventricular septal defects, and the fourth had moderate mitral incompetence. Congestive cardiac failure was present in all 4 patients with residual lesions but in only 1 of the 7 without a significant lesion. There was little correlation between the presence of residual defects and the occurrence of Adams-Stokes attacks. (Cases 10 and 11 are excluded because their natural history was altered by early pacemaker implantation.)

Analysis of electrocardiogram The patients with permanent complete heart block had ventricular rates in the immediate postoperative period in the range of 60 to 80 a minute. After a period of weeks to months the rate decreased to 44 to 65 a minute. There was no correlation between the ventricular rate and the occurrence of Adams-Stokes attacks.

Table 3 presents the duration of the pre- and postoperative QRS interval of 9 patients, excluding the 2 who had early pacemaker implantation. Of the 3 patients with little change in the QRS interval, one (Case 4) had Adams-Stokes attacks; the other 2 (Cases 2 and 7) remained asymptomatic. Six patients had significant widening of the QRS complex after operation, and all suffered from Adams-Stokes attacks.

Discussion

Surgically induced complete heart block is generally accepted as being due to trauma of the bundle of His as it courses along the caudal edge of the membranous septum. Studies by Lev (1960) and Hudson (1965) have shown close correlation between the antemortem presence of heart block and lesions of the conduction system in those cases coming to necropsy. The usual finding has been that of haemorrhage or oedema or both produced by a suture passing directly through or closely adjacent to the conduction system. It has been suggested that some cases may be due to anoxia (Thung *et al.*, 1962).

Complete heart block has been reported after almost all types of open heart operation (Gadboys and Litwak, 1964). The frequency of this complication in any individual cardiac lesion is related to the complexity of the lesion and the proximity of the vulnerable bundle to the site of the repair. The highest incidence is found in those lesions requiring the greatest surgical manipulation adjacent to the vulnerable conduction system. This is well exemplified by comparing the incidence of complete heart block after repair of ostium primum atrial septal defects with that of the

TABLE 3 *Pre- and postoperative QRS width and occurrence of Adams-Stokes attacks*

Case No.	*QRS duration (msec)*		*Adams-Stokes attacks*
	preoperative	*postoperative*	
1	60	100	Yes
2	80	80	Asymptomatic
3	80	120	Yes
4	70	80	Yes
5	70	120	Yes
6	80	120	Yes
7	80	80	Asymptomatic
8	80	120	Yes
9	70	100	Yes

secundum variety. In a series of 130 secundum atrial septal defect repairs we have not encountered a single case of surgically induced block (unpublished data). Complete heart block is more frequently associated with correction of tetrad of Fallot than with repair of isolated ventricular septal defect, because in the former lesion the ventricular defect almost invariably lies below the crista supraventricularis involving the anterior portion of the membranous septum. Such an anatomical situation is less common in isolated interventricular communications. The decreased incidence of surgically induced complete heart block may be attributed to a large extent to the care taken by the surgeon when suturing the posterior, inferior margin of membranous defects (Kirklin, McGoon, and DuShane, 1960; Gerbode and Keen, 1962; McGoon *et al.*, 1964).

In the early days of intracardiac surgery patients with complete heart block either reverted spontaneously or died in the immediate postoperative period (Lillehei *et al.*, 1963). The advent of isoprenaline and later the introduction of temporary cardiac pacing (Weirich, Gott, and Lillehei, 1957) has resulted in a fall in the mortality rate in the immediate postoperative period.

In these critically ill children it is frequently difficult to determine what part complete heart block played in their demise. In our own series the early postoperative mortality of patients with complete heart block was twice that of patients with similar lesions but without block (Table 4). Generally the mode of death was related to low cardiac output. It has been suggested that even when the ventricular rate is deemed adequate the cardiac output is limited at this critical time by the lack of synchronous atrial contraction (Lillehei *et al.*, 1963). It is not known whether the mortality rate could be reduced further by means of an atrial triggered pacemaker. Two patients died during a Adams-Stokes attack. In both instances pacing had been discontinued on the third postoperative day because of resumption of sinus rhythm. The next day both patients died after return of complete heart block.

TABLE 4 *Early postoperative mortality rate of patients with and without surgically induced complete heart block*

	No.	*Died*	*Per cent*
With complete heart block	40	11	27·5
Without complete heart block	223	28	13

Intermittent atrioventricular conduction disturbances of variable degree alternating with periods of sinus rhythm are common in the early postoperative period, and generally sinus rhythm returns in such cases. However, during this transitory period the patient is particularly liable to Adams-Stokes attacks, and we now continue pacing for several days after reversion to sinus rhythm. Competitive rhythm has not been a problem. The conduction wires are left in place for 10 to 14 days because resumption of pacing may become necessary.

The management of the child with persistent surgically induced complete heart block remains controversial (Hurwitz *et al.*, 1968; Murphy *et al.*, 1970). Knowledge of the natural history of this disorder is hindered by the small experience of any paediatric centre and the lack of long-term observation. Our own experience of 6 deaths in 9 medically treated patients (67%) confirms the high mortality rate reported by Lillehei *et al.* (1963) and Sayed (1965).

Though pacemaker implantation is technically feasible in very small children (Harris, Bowman, and Griffiths, 1966; Martin *et al.*, 1966), there is an understandable reluctance to recommend such a procedure because of the need for repeated surgical revisions to replace batteries or repair defective wires. In order to circumvent such awesome decisions, efforts have been made to determine prognostic criteria which have largely been extrapolated from what is known of the natural history of congenital complete heart block and degenerative block in the elderly. Such criteria have included the occurrence of Adams-Stokes attacks, the presence of residual intracardiac defects, the site of the pacemaker impulse as indicated by the ventricular rate and width of the QRS complex, and the response to sympathomimetic drugs. The most prognostic weight has been placed on the presence or absence of Adams-Stokes attacks. However, it has been our unfortunate experience that more than half the patients with Adams-Stokes attacks died during the first episode. Because the occurrence of the first Adams-Stokes attack cannot be predicted, patients with surgically induced complete heart block are at great risk. In addition, the length of the symptom-free period does not exempt a patient from the possibility of sudden death as evidenced by the 3 children who died 4, 7, and 11 years after operation.

There have been several reports commenting on the poor prognosis of surgically induced complete heart block complicated by residual anatomical lesions (Sayed, 1965;

Hurwitz *et al.*, 1968). Scarpelli and Rudolph (1964) described the excessive haemodynamic load in patients with congenital complete heart block with associated left-to-right shunt, and hypothesized from the work of Paul and Rudolph (1958) the enormous ventricular effort required to maintain adequate cardiac output in complete heart block complicated by outflow obstruction. In our experience congestive cardiac failure was an invariable occurrence in the presence of a significant residual defect and was unusual in the absence of such a complication. Adams-Stokes attacks were common in both groups, and the absence of a residual lesion did not exempt a patient from an unfavourable outcome.

Since prolongation of the QRS interval has been associated with a poor prognosis in *congenital* complete heart block (Nakamura and Nadas, 1964; Trusler, Mustard, and Keith, 1968), we analysed the postoperative QRS complex in our patients with permanent block. It was appreciated that intraventricular conduction disturbances which produce a wide QRS interval are common after ventriculotomy or repair of ventricular septal defects (Bristow *et al.*, 1960; Titus *et al.*, 1963), and that this might invalidate any significance ascribed to a wide QRS complex in surgically induced complete heart block. It was of interest that all patients with a prolonged QRS interval suffered Adams-Stokes attacks but that these occurred in only 1 of 3 patients with a normal QRS interval. It would be wrong to base any conclusions on a small number of patients, but our observations do suggest that patients with surgically induced complete heart block who have a wide QRS complex from whatever cause lead a more precarious existence.

Our experience with permanent pacemaker implantation in children is limited to the 4 patients described in this paper and an additional 3 patients with congenital complete heart block. Effective pacing allows these children to lead normal lives free from the risk of Adams-Stokes attacks and sudden death. Malfunction of the pulse generator or lead systems (transvenous and myocardial) has become less frequent with technical advances in their manufacture. The longest interval between successive operations in our experience has been 23 months. Permanent pacemakers are usually implanted in the axilla or epigastrium. In small thin children we have on occasion placed the unit in the extrapleural space. The pacing impulse is delivered either by leads sutured directly into the myocardium or by insertion of a transvenous electrode into the apex of the right ventricle. The anticipated duration of pacing in children may be measured in decades so that repeated replacement of pulse generators and lead systems will almost certainly be necessary. The disadvantage of the transvenous method is that it may be impossible to remove the pacing electrode should it become defective. Furthermore, repeated replacement of transvenous electrodes will result in the gradual sacrifice of readily accessible veins. The morbidity of thoracotomy in children is low and at the present time we prefer to suture the pacing lead into the myocardium under direct vision and to preserve the transvenous route for emergency pacing should this become necessary.

Conclusion

Surgically induced complete heart block continues to complicate up to 10 per cent of intracardiac operations, and despite modern advances is still associated with considerable early mortality. Two-thirds of the patients who survived the immediate postoperative period reverted to sinus rhythm. Complete heart block persisted in the remaining third.

The application of such criteria as the occurrence of Adams-Stokes attacks, the presence of significant residual lesions, response to sympathomimetic drugs, and prolongation of the QRS interval postoperatively are of limited usefulness in managing patients with permanent complete heart block. The mortality of these patients when treated medically is high (67%). Adams-Stokes attacks are invariably the primary cause of death, and the first attack is, more often than not, fatal.

Implantation of a permanent pacemaker in patients with surgically induced permanent complete heart block appears, at the present time, the most satisfactory management to prevent sudden death.

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