PERMANENT COMPLETE HEART BLOCK FOLLOWING SURGICAL CORRECTION OF CONGENITAL HEART DISEASE

F. EDWIN, E. ANITEYE, M. TETTEY, L. SEREBOE, D. KOTEI, M. TAMATEY, K. ENTSUA-MENSAH and K. FRIMPONG-BOATENG

National Cardiothoracic Centre, Korle Bu Teaching Hospital, P. O. Box KB 846, Korle Bu, Accra

Corresponding Author: Dr. Frank Edwin ***Email:*** *fedwin68@yahoo.com*

Conflict of Interest: *None declared*

SUMMARY

Background: The risk of complete heart block (CHB) from congenital heart repairs in Ghana is unknown. This information is important for referring physicians and in pre-operative counselling of patients and facilitates the process of obtaining informed consent for such repairs.

Objectives: This study was undertaken to determine the incidence of permanent post-operative CHB requiring pacemaker implantation; and the post-operative problems related to the pacemaker.

Design: Retrospective study design.

Setting: The National Cardiothoracic Centre (NCTC), Korle-Bu Teaching Hospital, Accra, Ghana.

Method: Review of all patients who had intra-cardiac repair of congenital heart disease known to predispose to post-operative complete heart block from January 1993 to December 2008 was carried out with computation of the frequency of complete heart block according to the intra-operative diagnoses.

Results: Six out of 242 patients (2.5%) developed permanent post-operative CHB. All underwent closure of a large perimembranous ventricular septal defect (VSD) either as an isolated defect (2 of 151 or 1.3%) or in the setting of conotruncal anomalies (4 of 73 or 5.5%). The dominant parental concern relating to the implanted device was the financial implications of future multiple surgeries to replace a depleted pulse generator.

Conclusion: Permanent post-operative complete heart block occurred in 1.3% of patients undergoing VSD repair and 5.5% of those undergoing repair of conotruncal anomalies (Fallot's tetralogy). The dominant anatomic risk factor was a large perimembranous VSD as an isolated defect or as part of a conotruncal anomaly. Permanent pacemaker implantation in this setting is attended by a low morbidity.

Keywords: Congenital heart defect, Complete Heart Block, Pacemaker, Ventricular Septal Defect, Morbidity.

INTRODUCTION

Permanent complete heart block (CHB) is a significant complication of intracardiac repair for congenital heart disease. It refers to post-operative heart block that does not spontaneously revert to the pre-operative rhythm (usually within 10 days of the operation). Treatment entails lifelong dependence on an artificial pacing system requiring replacement at least once every decade. The relevant part of the cardiac conduction system in this setting is the atrioventricular (AV) node and the bundle of His.

The AV node is located in the triangle of Koch within the floor of the right atrium and continues as the His bundle which penetrates the right fibrous trigone to emerge at the base of the non-coronary aortic cusp in the upper interventricular septum. The bundle of His (and its divisions) is located within the ventricular septum and is thus vulnerable to injury during surgical procedures on the ventricular septum. CHB has been reported after repair of lesions such as isolated ventricular septal defect (VSD), atrioventricular septal defects (AVSD), VSD in the setting of conotruncal anomalies {tetralogy of Fallot (TOF), double outlet right ventricle (DORV), and others} and subaortic stenosis (SAS).

The risk of post-operative permanent CHB has decreased since Lev and others[1, 2, 3] delineated the course of the conduction tissue in various types of congenital cardiac malformations of the ventricular and adjacent atrial septa. In earlier reports[4] of 1971, the risk of surgical CHB was as high as 25%. Improved surgical techniques and better understanding of the anatomy of the conduction tissue in various congenital cardiac anomalies have reduced this risk to 1-4% in the current era[5]. Anderson et al reported a risk of 0.7% for closure of isolated VSD[6]. It is however a real risk that both the surgeon and his potential patient must confront. Open-heart repairs for congenital cardiac anomalies have been performed on a consistent basis since 1992 at the National Cardiothoracic Centre (NCTC) in Accra, Ghana.

Unlike other centres worldwide, the incidence of post-operative heart block for congenital heart disease in Ghana has not been reported. This information is important for referring physicians, in pre-operative counselling of patients and parents; it also facilitates the process of obtaining informed consent for patients undergoing such repairs. This study was undertaken to determine the incidence of permanent post-operative CHB and the post-operative problems related to the pacemaker.

PATIENTS AND METHODS

A retrospective analysis of all patients who underwent intra-cardiac repair at the NCTC from January 1993 to December 2008 with a diagnosis of congenital heart disease known to predispose to post-operative complete heart block was carried out. These lesions[6] generally include a ventricular or atrioventricular septal defect either in isolation or as part of a more complex anomaly.

Subaortic stenosis (SAS) resulting from a subaortic shelf was included because the shelf directly overlies the conduction tissue in the upper part of the ventricular septum. Resection of the shelf may induce CHB from direct sectioning of the conduction tissue. Patients with a ventricular septal defect (VSD), atrioventricular septal defect (AVSD), conotruncal anomalies (Fallot's tetralogy and double outlet right ventricle) or SAS were enlisted in the study. A review of the operative records covering the study period was performed and patients who met the study criteria were selected.

Patients who underwent permanent pacemaker implantation following congenital heart repairs were selected and their case notes and operative records reviewed. The morbidity relating to the implanted device was assessed by direct questioning of parents and review of the follow up records regarding history of specific complaints, physical examination findings and investigations (electrocardiography, chest radiography and transthoracic echocardiography).

RESULTS

In the period under review, a total of 242 patients underwent intra-cardiac repair of congenital heart disease predisposing to postoperative CHB. There were 127 males and 115 females (ratio of 1.1:1). Their ages ranged from 10 months to 70 years (mean 11.9 years, median 8.0 years). Follow up ranged from 0.1 – 15.8 years (mean 6.0 years, median 4.8 years). Six patients (2.5%) developed postoperative CHB of broad QRS complex type and required permanent pacemaker implantation after a mean duration of block of 11.8days (range 5-18days).

Table 1 Diagnosis-specific risk of post-operative heart block

Diagnosis	Number (n)	Per-centage (%)	Num-ber of CHB (x)	Abso-lute Risk (x/n)
VSD (Isolated) (+ASD, PDA or RVOTO)	151 112 39	62.4	2	1.3%
CTA (TOF) (DORV[TOF type])	73 68 5	30.2	4 4 0	5.5% 5.9% 0
AVSD Partial Complete	15 9 6	6.2	0	0
SAS	3	1.2	0	0

CHB-complete heart block; **VSD**-ventricular septal defect; **ASD**-atrial septal defect; **PDA**-patent ductus arteriosus; **RVOTO**-right ventricular outflow tract obstruction; **CTA**-conotruncal anomalies; **TOF**-tetralogy of Fallot; **DORV**-double outlet right ventricle of Fallot type; **SAS**-subaortic stenosis; **AVSD**-atrioventricular septal defect.

In these patients, the pre-operative clinical evaluation was in keeping with their primary cardiac conditions requiring operation. None of them showed clinical or electrocardiographic (ECG) evidence of complete heart block before surgery. All were in sinus rhythm pre-operatively.

Intra-operative rhythm was sinus as shown by continuous ECG monitoring until the institution of cardiopulmonary bypass and cold cardioplegic arrest. The intracardiac anatomy of their defects confirmed at operation is shown in Table 2.

All the VSDs of patients developing post-operative permanent CHB were perimembranous in location and large (>50% of the patient's aortic root; mean of 57.8%, range 52%-67%). In 3 of 4 patients with TOF developing CHB, the perimembranous VSD extended into the inlet of the right ventricle beneath the septal leaflet of the tricuspid valve.

Temporary epicardial pacing was routinely employed in all patients after separation from cardiopulmonary bypass before the heart fully regained normal rhythm and chronotropy from the effects of cardioplegia and hypothermia. For most patients this was necessary for less than 24 hours. For patients with post-operative CHB, attempted cessation of this temporary pacing revealed a bradycardia and hypotension.

Continuous ECG monitoring in the Intensive Care Unit post-operatively showed CHB. Temporary pacing was

then continued with the expectation of spontaneous resolution within 7-10 days.

Table 2 Complete Heart Block – patient characteristics

Patient	Age (Years)	Intracardiac Anat-omy	POD	Pace-maker Implant Route	Pacing Thresh old Voltage
SS	5	TOF	13, 60, 68	E, E, T	2.8V, ND, 0.4V
DFR	10	TOF	18	T	Miss-ing record
AT	11	TOF	16	T	0.77V
QCE	1	VSD	11	E	2.64V
PA	5	VSD	8	T	0.64V
LY	8	TOF	5	T	ND

TOF-tetralogy of Fallot; **VSD**-ventricular septal defect; **POD**-post-operative day on which pacemaker was implanted; **E**-epicardial; **T**-transvenous; **ND**-not documented.

Permanent postoperative CHB was treated with a ventricular demand pacemaker with rate responsiveness. The route of implantation and pacing thresholds are shown in Table 2. The case notes of patient DFR could not be traced and therefore detailed information on intra-operative findings could not be obtained. As of the last follow-up visit to the out-patient clinic 3 years before this report, he was doing well and had no complications. Patient SS underwent replacement of the exhausted pulse generator in August 2007 (6 years post-implantation).

The pulse generator of patient QCE was exhausted in October 2008 (5 years post-implantation). She has an intrinsic rate of 50/minute and moderate exercise limitation; a pulse generator replacement is scheduled. The remainder of the patients are in good health with functioning pacing systems. In patient SS, the pacing system (epicardial implantation in both) failed twice within a 3 month period and required two revisions. In both instances, the patient presented with undue fatigue on exertion and bradycardia with the ECG showing failure of artificial pacing. No specific complaints relating to device malfunction was recorded in the other patients.

Four parents expressed concern regarding:
1. The future multiple surgeries to replace the pulse generator when exhausted
2. The financial implications of future replacements of the pulse generator and
3. The risk of trauma to the device during play or sporting activities.

No complaints of upper limb swelling or discomfort indicating possible venous thrombotic obstruction were lodged among the children who underwent trans venous pacemaker implantation. There was no history suggestive of a thromboembolic event in any of the patients followed up to date. No instances of pulse generator migration were recorded. The electrocardiography (ECG) findings have been in keeping with a functioning pacemaker in all the patients except in patient QCE (exhausted pulse generator) and patient DFR (lost to follow up). No evidence of intra-cardiac or intravascular thrombosis or tricuspid incompetence associated with the intracardiac electrode was found on echocardiography.

DISCUSSION

CHB following surgical repair may be transient or permanent. Transient CHB in this setting generally reverts to sinus rhythm within the first 7-10 days after surgery though recovery several months later is not unknown. By definition then, permanent CHB is not expected to exhibit spontaneous recovery. Weindling et al. have shown that 63% of patients with CHB after surgery regained atrioventricular conduction in the first post-operative month; in the majority (97%) this occurred in the first 9 days after surgery.[7]

Belated recoveries after permanent pacemaker implantation occur in a small but significant number of patients. Batra et al noted that after insertion of a pacemaker, recovery of atrioventricular conduction occurred in 7 of 72 patients (9.6%) at a median of 41 days (range 18-113 days) after the initial cardiac operation.[8] The true incidence of transient CHB for this report was not studied because of inconsistencies with its documentation.

This report therefore focused on permanent post-operative CHB for which accurate documentation existed and also has a more significant post-operative impact in terms of morbidity and cost. This study shows the commonest indication for surgery on the ventricular septum relates to the presence of a VSD which was present in 230 (95.0%) patients; the only exceptions were 3 patients with SAS and 9 with AVSD. In our experience, permanent complete heart block occurred in 6 of 242 (2.5%) patients which compares favourably with others.[5, 9, 10]

All the patients who developed complete heart block had large perimembranous VSDs. In a perimembranous VSD, the His bundle is intimately related to the posterior and inferior margins of the defect making it

vulnerable to operative trauma; the margin of error can be very small indeed[10]. It is generally held that surgical heart block is due to direct operative trauma to the conduction tissue. In all 6 instances, the QRS morphology was of broad-complex type, indicating possible injury to the distal His bundle or the bundle branches.

Studies which identify the electro physiologic sites of CHB following open-heart surgery remain scanty. Available reports are limited to small case series and lack data on prevalence. Recent reports in the surgical literature on this subject are lacking. Generally, wide complex surgical CHB indicates an infra-Hisian level of conduction injury[11] and would be in keeping with our findings. However, some workers[12] have suggested that the QRS morphology on routine ECG may not accurately predict the electro physiologic site of conduction delay.

In the report of Anderson and colleagues[12], a patient developing wide QRS-complex block after tetralogy repair was found on His Bundle recordings to have developed a supra-Hisian type of block. This would not have been predicted from the routine ECG. Again, contrary to what would have been expected, their second patient demonstrated a narrow QRS-complex block even though the site of block was found to be distal to the His bundle following tetralogy repair. No proven explanations have been put forward to account for such variability in electro cardiographic manifestations.

It may be speculated that anomalous conduction pathways and/ or pacemaker sites may be involved. We did not perform His bundle ECGs on any of our patients and therefore the site of block in our patients is difficult to ascertain with certainty. It is also noteworthy that all the patients who suffered surgical heart block in our study had ventricular septal defects that were larger than 50% of the aortic orifice. The designation of a VSD as large or small is arbitrary but useful. In this report, a large VSD is arbitrarily taken to be greater than 50% of the size of the aortic root.

It may be inferred that the larger the defect, the more intimate and extensive is the relation between the conduction tissue and the VSD margin. Titus et al[13] noted that when the defect is small relative to the size of the heart, no part of the conduction system was intimately related to the defect, apparently because the defect is too small to impinge on the tissues normally carrying conduction fibres.

It is striking also that 4 out of 68 (5.9%) patients with tetralogy of Fallot developed CHB after repair as opposed to 2 out of 151 (1.3%) patients with isolated VSD. The intracardiac anatomy in all these patients however conformed to a large perimembranous VSD with inlet extension in 3 of 4 patients. Our experience is at variance with other workers who have reported a relatively low risk of CHB following repair of TOF. Andersen and colleagues[6] reported a risk as low as 0.1% in this setting. In Hussain et al's report[14], the risk was quoted as 1.7%. Anderson's group[6] attributed their findings to the fact that the bundle of His is better protected in the setting of Tetralogy of Fallot.

They however pointed out that this may not be so in the situation of combined perimembranous and muscular defects, and straddling and overriding of the tricuspid valve. The anatomic studies of Dickinson et al suggests that the disposition of the VSD in TOF and it's relationship to the conduction tissue may have a significant bearing on the propensity towards CHB following repair[15]. When the VSD in tetralogy is of the outlet type, it is relatively remote from the conduction tissue and less vulnerable to operative trauma.

Perimembranous defects on the other hand, especially those involving the inlet are more vulnerable to CHB after repair[16]. Variations in the morphology of the VSD seen in our cases that developed CHB may explain the relatively high risk of CHB seen among TOF patients in this study. Because conclusive differentiation between perimembranous and muscular outlet defects is not always possible on gross morphological examination, it is recommended that the crest of the muscular septum and fibrous tissue in the posterior rim of the VSD should be avoided during closure of the defect.[15]

Our technique of closing these defects follow established surgical principles[17] based on the anatomy of these defects – using relatively oversized patches, suturing to the right ventricular septal surface at least 2 mm away from the rim of the VSD, especially around the postero-inferior border of the defect with placement of superficial sutures in this area. The fact that a relatively high incidence of CHB occurred nevertheless among TOF patients in this study calls for further study in this patient subset.

Post-operative CHB persisting beyond 7-10days generally requires pacemaker implantation but optimism for spontaneous recovery of sinus rhythm sometimes pushes the time limit further out. In this report, the mean waiting period was 11.8days.

For patient LY, an unstable temporary epicardial electrode implanted at the time of total correction was the reason for a much shorter waiting period of 5 days. The pacing leads that may be used for children are either trans venous or epicardial (myocardial screw-in).

For the trans venous approach there is concern about vascular obstruction, tricuspid valve integrity, and limitations of lead accommodation in relation to somatic growth. Standard epicardial leads tend to have a relatively high failure rate[10, 18] necessitating repeat procedures to reposition the lead.

Their high failure rate has been attributed to high chronic thresholds and a vulnerability to lead fracture and dislodgement. We encountered some of these problems in patient SS who had failure of two epicardial leads resulting ultimately in conversion to a transvenous lead. Higher energy demands in children (higher heart rate at rest and with exercise) shortens pulse generator longevity in children relative to adults.

The pacing features of patients requiring pacemaker implantation in this study indicate a 300% higher acute pacing threshold of epicardial relative to transvenous systems (2.6v against 0.6v). Rising chronic thresholds ultimately lead to the phenomenon of exit block in such epicardial systems. In less severe cases, the battery longevity is compromised[19]. In small children, small upper chest vein size may preclude a transvenous implantation technique.

The development of steroid-eluting epicardial electrodes is proving to be an effective method of overcoming such problems. Their use has been shown to give success rates and generator longevity comparable to transvenous electrodes.[19] Parental concern regarding the need for repeated pulse generator replacements is one of the key morbidity issues found in this study. The primary factors here are both economic and procedural. The risk of trauma to the device is another understandable concern of parents. Unfortunately, this is not an easy issue to resolve but a special protector over the pulse generator may help prevent trauma to the device.[20]

CONCLUSION

Permanent post-operative complete heart block occurred in 1.3% of patients undergoing VSD repair and 5.5% of those undergoing repair of conotruncal malformations of the Fallot type. The dominant anatomic risk factor was a large perimembranous VSD. Permanent pacemaker implantation in this setting is attended by a low morbidity.

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