Cardiology in the Young

cambridge.org/cty

Permanent pacing post-Fontan is not associated with reduced long-term survival

Sally J. Hall, Damien Cullington, John D. R. Thomson and James R. Bentham

Department of Paediatric Cardiology, Yorkshire Heart Centre, Leeds General Infirmary, Leeds, UK

Original Article

Cite this article: Hall SJ, Cullington D, Thomson JDR, and Bentham JR (2019) Permanent pacing post-Fontan is not associated with reduced long-term survival. *Cardiology in the Young* **29**: 1432–1434. doi: 10.1017/S104795111900194X

Received: 17 January 2019
Revised: 10 June 2019
Accepted: 19 July 2019
First published online: 11 October 2019

Keywords:
Congenital heart disease; Fontan procedure; pacemaker

Author for correspondence:
Dr J. Bentham, Department of Paediatric Cardiology, Yorkshire Heart Centre, Leeds General Infirmary, Great George Street, Leeds, LS1 3EX, UK. Tel: +44 (0) 1133928239; E-mail: Jamie.bentham@nhs.net

Abstract

Background: The Fontan procedure is the final stage of surgical palliation for a single-ventricle circulation. Significant complications are common including rhythm disturbance necessitating implantation of a permanent pacemaker. This has been widely considered a negative prognostic indicator. *Methods:* This single-centre, retrospective case control study involved all patients who underwent the Fontan procedure at the Leeds Congenital Heart Unit between 1990 and 2015 and have had regular follow-up in Yorkshire and Humber, United Kingdom. 167 Fontan patients were identified of which 2 were excluded for having a pre-procedure pacemaker. Of the remainder, 23 patients required a pacemaker. Outcomes were survival, early and late complications, need for further intervention and oxygen saturation in long-term follow-up. *Results:* There was no difference in survival (30-day survival pacemaker 92.6%, sinus rhythm 90.5%, $p = 0.66$, 1-year pacemaker 11.1%, sinus rhythm 10.1%, $p = 1$). The pacemaker group was more likely to have cerebral or renal complications in the first-year post-procedure (acute kidney injury: sinus rhythm 0.8%, pacemaker 19.1%, $p = 0.002$). No difference was observed in longer term complications including protein losing enteropathy (sinus rhythm 3.5%, pacemaker 0% $p = 1$). There was no difference in saturations between the two groups at follow-up. Paced patients were more likely to have required further intervention, with a higher incidence of cardiopulmonary bypass procedures (sinus rhythm 6.3%, pacemaker 35%, $p < 0.001$). *Conclusions:* Despite an increase in early complications and the need for further interventions, pacemaker requirement does not appear to affect long-term survival following the Fontan procedure.

Patients with complex CHD in whom a biventricular repair cannot be achieved undergo staged palliation finally culminating in all systemic venous blood being routed to the pulmonary circulation directly.[1] This is normally a three-staged process beginning with either a high-pressure systemic to pulmonary shunt or a band to secure pulmonary blood flow in early infancy followed by a superior caval vein to pulmonary artery anastomosis (Glenn) and finally inferior caval vein to pulmonary artery connection.[2] The last of these stages is generically called a Fontan operation, a term used to encompass multiple different operations to connect the lower body systemic venous blood to the pulmonary artery.[1,3,4] It was first performed in 1968 as an atrio-pulmonary connection but more recently as a lateral tunnel or extracardiac conduit connection with different advantages and disadvantages to each approach.[4–6] Over the decades, timing of the operation has changed but in general it is performed in mid-childhood (3–8 years).[3,6] Complications related to this form of circulation are common and medium and long-term survival remains a significant concern (5-, 10-, 20-, and 30-year survival 85, 74, 61, and 43% respectively).[3] A common complication of the Fontan procedure is arrhythmia and this may occasionally require a permanent pacemaker.[6] This is largely considered to be a poor prognostic indicator but there is very little evidence to support this conclusion.[6] This study sought to evaluate the longer term implications of pacing in this complex patient group.

Methods

This was a single-centre retrospective cohort study involving a review of the electronic records of all patients who have undergone a Fontan operation at the Leeds Congenital Heart Unit and received regular follow-up in the Yorkshire, Humber, and Lincolnshire regions of the United Kingdom between 1990 and 2015. Patients with a pacemaker prior to surgery (2) were excluded. Groups were assigned according to whether the patient required a pacemaker either at the time of the Fontan operation or at a later point and those that did not.

The primary outcomes were 7 day, 30 day, 1 year, and 5-year survival. Secondary outcomes included incidence of acute complications: cerebral, renal, or thrombotic events; incidence of chronic complications: protein losing enteropathy, pulmonary hypertension, and plastic

CAMBRIDGE UNIVERSITY PRESS

Table 1. Comparison of demographics of patients undergoing the Fontan Procedure at Leeds General Infirmary 1987–2015.

	Pacemaker		No pacemaker		
	Number	%	Number	%	*P*
Gender					
Male	15	65.2	83	57.6	0.69
Diagnosis					
Right heart lesion	15	65.22	98	69.01	0.81
Left heart lesion	1	4.35	15	10.56	0.7
Atrioventricular septal defect	1	4.35	20	14.08	0.48
Ventricular septal defect	8	34.78	48	33.80	1
Double inlet left ventricle	*7*	30.43	*15*	10.56	*0.017*
Transposition of the great arteries	13	56.52	53	37.32	0.11
Operation					
Extracardiac conduit Fontan	13	59.09	85	75.89	0.12
Atriopulmonary Fontan	6	27.27	14	12.50	0.09
Lateral tunnel Fontan	3	13.64	12	10.71	0.71

Note: The values in italics indicate statistical significance.

Table 2. Mode of pacing used.

Pacemaker		
Mode	Number	%
DDD	15	65.2
AAI	4	17.4
VVI	3	13
Unknown	1	4.3

Table 3. Time spent paced for patients with dual-chamber pacemakers.

Pacemaker mode: dual chamber		
Percentage of time paced	Number	%
<10%	1	9.09
10–25%	1	9.09
25–50%	1	9.09
50–75%	1	9.09
75–99%	1	9.09
>99%	6	54.55

bronchitis; oxygen saturation level at long-term follow-up and the requirement for further intervention (both surgery and cardiac catheterisation).

Baseline patient demographics including diagnosis and type of Fontan procedure were analysed to identify any differences between the two cohorts. Information about the patient's survival, complications, further procedures, and saturations as documented in clinic were collected from the electronic record.

Table 4. Incidence of death following the Fontan procedure at Leeds General Infirmary.

	Pacemaker			No pacemaker			
	Total	Deaths	%	Total	Deaths	%	p
7 days	22	0	0	141	9	6.38	0.366
30 days	22	2	9.5	141	9	6.38	0.61
1 y	18	2	11.1	119	12	10.08	1
5 y	15	1	6.67	74	10	13.51	0.68

y = years post-Fontan procedure

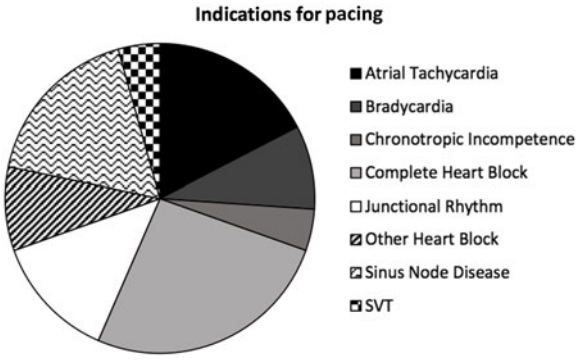

Figure 1. Implications for pacemaker implantation. SVT = supraventricular tachycardia.

For statistical analysis, median values are given with interquartile ranges. Fisher exact test for small numbers and Chi square tests were used to compare categorical outcomes between the two groups of patients. A paired t-test was used to analyse patients' saturation levels. Kaplan–Meier plots were constructed for survival analysis.

Results

A total of 167 patients underwent a Fontan procedure. Two of these patients were excluded from the study for having pacemakers prior to their initial Fontan procedure.

Of the remaining 165 patients, 23 patients required a pacemaker either during or following their Fontan operation (13.9%). There was a wide range of pacemaker indications (Fig 1). There were no differences in age or gender of the paced and non-paced patient groups, and no difference in the type of Fontan procedure performed (atrio-pulmonary, lateral tunnel, or extra-cardiac conduit). In terms of underlying diagnosis, double inlet left ventricle was more frequent in the paced Fontan group (7 of 23, 30.4% permanent pacemaker, 15 of 142, 10.6% sinus rhythm; $p = 0.017$, Table 1). In the paced group, the most commonly used setting was DDD (65%) (Table 2). Patients with dual-chamber pacemakers spent 0.4–99.6% of time paced (Table 3).

There was no significant difference between the groups in terms of survival at any stage post-Fontan procedure with up to 23 years of patient follow-up (Table 4, Figs 2 and 3).

Acute complications were significantly more likely to affect patients who required a pacemaker (8 of 23, 34.8% permanent pacemaker versus 4 of 142, 2.8% sinus rhythm, $p < 0.001$),

https://doi.org/10.1017/S104795111900194X Published online by Cambridge University Press

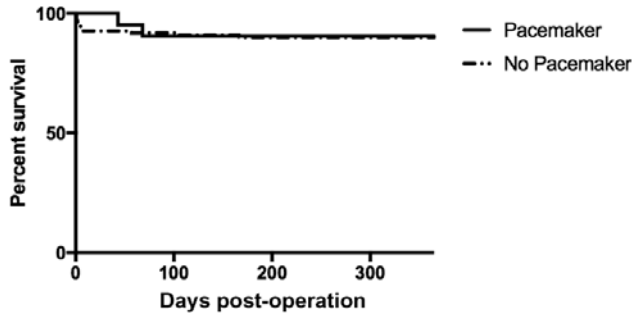

Figure 2. A Kaplan–Meier plot demonstrating survival rates throughout the first-year post-Fontan procedure comparing patients with a pacemaker to those without.

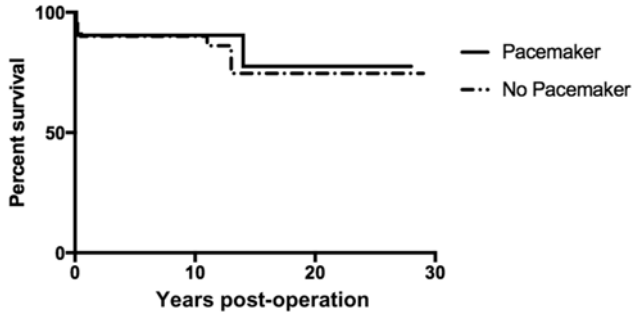

Figure 3. A Kaplan–Meier plot demonstrating survival rates post-Fontan procedure comparing patients with a pacemaker to those without.

including greater likelihood of acute kidney injury (4 of 23, 17.4% permanent pacemaker versus 1 of 142, 0.7% sinus rhythm, $p = 0.0013$). There was no difference in the incidence of chronic complications, including symptoms indicative of Fontan failure (1 of 23, 4.7% permanent pacemaker versus 11 of 142, 7.4% sinus rhythm, $p = 1$). Patients in the pacemaker group were far more likely to require further bypass procedures (8 of 23, 35% permanent pacemaker versus 9 of 142, 6.3% sinus rhythm, $p < 0.001$), although there was no difference in the incidence of interventional catheter procedures (4 of 23, 17.4% permanent pacemaker versus 37 of 142, 26% sinus rhythm, $p = 0.45$). Interventions related directly to the pacemaker were excluded (Table 3).

There were no differences between groups in oxygen saturation level as recorded in clinic at any stage post-Fontan procedure (Table 4).

Discussion

This study analyses the requirement for a post-operative pacemaker and its effects on the mortality and morbidity of patients who have undergone the Fontan operation for palliation of a single-ventricle circulation. The data provides some reassurance that pacemaker requirement does not, in the long term, appear to impact negatively on survival. However, this study does demonstrate that this group is at risk of early complications that are likely to relate to a propensity for lower cardiac output in this group (acute kidney and cerebral injury).

Pacemaker requirement had no effect on the incidence of protein losing enteropathy or other chronic complications. This suggests that pacemaker requirement in the long term is not associated strongly with symptoms suggestive of Fontan failure.[7]

A significantly higher proportion of patients in the pacemaker group had double inlet left ventricle amongst their initial diagnoses, and further analysis reveals a cohort of patients with double inlet left ventricle and transposition of the great arteries. Ventricular septal defect enlargement will have been necessary for these patients; this is often performed at the time of Fontan operation, with potential for disruption of the septal conduction pathway, increasing the likelihood of pacemaker requirement. In current practice, this patient group would have a Damus–Kaye Stansel anastomosis performed at first palliation, obviating the need for subsequent ventricular septal defect enlargement.[8]

The main limitations of this study are the retrospective design and the small number of patients requiring a pacemaker, which in turn limits the power to detect any association of pacemaker requirement with symptoms suggestive of Fontan failure in this series. A national multi-centre study would provide more evidence on the importance of pacemaker requirement and outcome post-Fontan.

Conclusions

Pacemaker requirement post-Fontan is associated with more complex surgery performed during the same procedure. This is associated with an increase in short-term morbidity. However, despite this, there appears to be no long-term propensity to Fontan failure or adverse effect on longer term survival.

Acknowledgements. None.

Financial Support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of Interest. None.

References

1. McGuirk SP, Griselli M, Stumper OF, et al. Staged surgical management of hypoplastic left heart sundrome: a single institution 12 year experience. Heart 2006; 92: 364–370.
2. Bando Ko, Turrentine MW, Sun K, et al. Surgical management of hypoplastic left heart syndrome. Ann Thorac Surg 1996; 62: 70–76.
3. Pundi KN, Johnson JN, Dearani JA, et al. 40-year follow-up after the Fontan operation. Am J Cardiol 2015; 66: 1700–1710.
4. Pigott JD, Norwood WI. The Fontan procedure. Curr Opin Cardiol 1989; 4: 286–290.
5. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971; 26: 240–248.
6. Anderson PAW, Sleeper LA, Mahony L, et al. Pediatric heart network investigators. Contemporary outcomes after the Fontan procedure. Am J Cardiol 2008; 52: 85–98.
7. John AS, Johnson JA, Khan M, et al. Clinical outcomes and improved survival in patients with protein-losing enteropathy after the Fontan operation. Am J Cardiol 2014; 64: 54–62.
8. Franken LC, Admiraal M, Verrall CE, et al. Improved long-term outcomes in double-inlet left ventricle and tricuspid atresia with transposed great arteries: systemic outflow tract obstruction present at birth defines long-term outcome. Eur J Cardiothorac Surg 2017; 51: 1051–1057.

https://doi.org/10.1017/S104795111900194X Published online by Cambridge University Press