

Longitudinal Echocardiographic Parameters Before and After Pacemaker Placement in Congenital Complete Heart Block

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Background

Congenital Complete Heart Block (CCHB) with a structurally normal heart is a rare diagnosis, seen in approximately 1:15,000 – 1:20,000 live births.

Pacemaker placement is performed in 69% of patients by age 1 year and in 74%-96% by 20 years.

Risk of LV dysfunction has been reported in the literature in 7-23% of subjects.

There is inconsistent consensus on the most important risk factors for LV dysfunction, including the effect of chronic cardiac pacing.

Study Objective

In this study, we investigate serial changes in LV chamber size as estimated by **LV end diastolic dimension (LVEDD) z-score** and LV systolic function as estimated by **LV ejection fraction (EF)** before and after pacemaker placement in patients with CCHB by:

1. Comparing changes in outcome measures over time (slopes) before and after pacemaker placement.
2. Comparing post-pacemaker implantation outcome measures with those assessed at time of pacemaker placement.

Study Methods

Retrospective cohort study of all CCHB subjects seen at CHOP between 1976 – 2018.

Subjects excluded from cohort if AV block identified subsequent to or in association with a known acquired cause, if they had structural congenital heart disease, if age at diagnosis was > 21 years.

Subjects included in the analysis if echo data available from before and after pacemaker implantation.

Statistical Methods

Linear mixed effect models of LVEDD z-score and LVEF on time with subject-specific random slope and intercept.

Model covariates: sex, race/ethnicity, birth era, age at diagnosis of CCHB, fetal diagnosis, maternal antibody status, baseline echo parameter defined as normal or abnormal, age at pacing, and initial pacemaker mode.

Estimated population slopes in LVEDD z-score and LVEF measures over time before and after pacemaker implant were compared to evaluate the effect of pacemaker implantation on these parameters during the study period.

All data management and statistical analyses were performed using SAS, version 9.4 (SAS Institute Inc., Cary, NC, USA) and R (version 4.0.4).

Table 1: Demographic and Clinical Characteristics of the Cohort

Characteristics	Overall (N = 52)
Age at last visit (years)	17.1 (11.4-20.3)
Observation period (years)*	12.7 (8.6-15.4)
Birth Era	
1970-1990	10 (19.2)
1990-2000	15 (28.8)
2000-2010	20 (38.5)
2010-2018	7 (13.5)
Sex	
Male	21 (40.4)
Female	31 (59.6)
Race/Ethnicity	
Non-Hispanic White	38 (73.1)
Non-Hispanic Black	9 (17.3)
Hispanic	4 (7.7)
Asian	0 (0.0)
Unknown	1 (1.9)
Age at CCHB diagnosis (years)	0.6 (0.0-3.5)
Fetal diagnosis	
Yes	12 (23.1)
No	33 (63.5)
Unknown	7 (13.5)
Maternal antibody status	
Positive	19 (36.5)
SSA	2 (10.5)
SSB	6 (31.6)
Specific Antibody Unknown	11 (57.9)
Negative	19 (36.5)
Unknown if any test performed	14 (26.9)

Categorical variables displayed as number (%), continuous variables as median (interquartile range)
*Defined as duration of time from first visit to most recent visit

Table 2: Pacemaker Characteristics of the Cohort

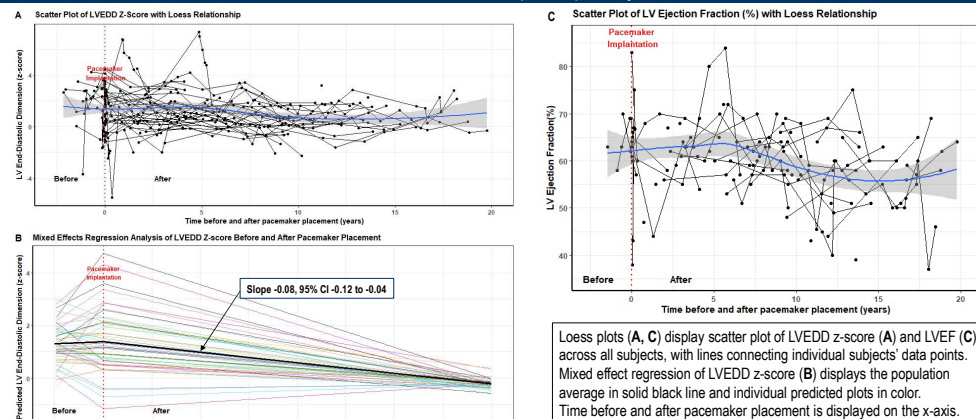
Characteristics	Overall (N = 52)
Pacemaker placement on first day of life	8 (15)
Permanent pacemaker	52 (100.0)
Age at placement of first pacemaker (years)	3.4 (0.5-9.0)
Initial pacemaker mode - VVI	30 (58)
Initial pacemaker mode - DDD	13 (25)
Initial pacemaker mode - VDD	9 (17)
Total pacemaker generators placed	2 (1.3)
Duration of pacing (years)	10.8 (5.2-13.7)
Duration of epicardial pacing (years)	4.5 (0.0-11.1)
Duration of transvenous pacing (years)	1.9 (0.0-7.0)

Categorical variables displayed as number (%), continuous variables as median (interquartile range)

Echo Data Points

- Total: 337
- LVEDD (z-score) measures: 328
- LVEF measures: 138
- Echo visits per subject: 5 (IQR, 3-9)
- Years between first and last echo: 8.4 (IQR, 3.1-13.2)

Results: LV End-Diastolic Dimension (Z-Score) & LV Ejection Fraction



- 6 months pre-pacemaker: Estimated LVEDD z-score 1.37 (95% CI, 0.78-1.97)
- There was no significant change in LVEDD z-score between 6 months pre-pacemaker and time of pacemaker placement, with an estimated z-score of 1.39 (95% CI, 0.83-1.95) at time of placement.
- Following pacemaker implantation, significant decrease in LVEDD z-score relative to measures at time of implant, with slope -0.08 (-0.12 to -0.04, p=0.002).
- Mean estimated LVEDD z-score:
 - 5 years post-placement: 0.99 (95% CI, 0.58-1.40)
 - 10 years post-placement: 0.60 (95% CI, 0.25-0.95)
 - 20 years post-placement: -0.21 (95% CI, -0.82 to 0.4)
- Observed changes in LVEDD z-score following pacemaker placement did not vary by baseline demographic or clinical characteristics of interest

Conclusions

- In a cohort of 52 paced subjects with CCHB:
- LVEDD z-score decreases significantly following pacemaker placement.
 - With increased chronotropy following pacemaker implantation, it is physiologically appropriate and expected that the LVEDD z-score would decline.
 - Observed changes in LVEDD z-score following pacemaker placement did not vary by baseline demographic or clinical characteristics of interest.
 - LV EF appears to remain preserved for the first 5.5 years following pacemaker placement, after which it appears to decline
 - Chronic electrical and mechanical dyssynchrony from right ventricular pacing and abnormal histologic myocardial changes that have been demonstrated in the setting of chronic pacing may potentially account for this estimated decline.
 - This data may inform appropriate clinical timelines for echocardiographic screening and aid counseling of families of children and young adults with CCHB.

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