

**Paediatric Heart Failure from Heart Muscle Disease in the UK and Ireland: 5+ Year Follow Up.**

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**Introduction**

Our original study, the first national prospective study of new-onset heart failure from heart muscle disease in children, showed an incidence of 0.87/100,000 with an overall 1-year survival of 82%, and an event (death or transplantation)-free survival of 66%.<sup>(1)</sup> This study aimed to evaluate 5+ year outcomes of this important cohort of patients.

**Methods**

All centres in the UK and Ireland with event-free survivors from the original study participated (n=14). Consent for inclusion was sought from patients & families. Anonymised data based on the last hospital attendance and echocardiograms (where available) were forwarded for analysis. The investigator (RA) was blinded to outcome at the time of echo review.

**Results**

Of 69 event-free survivors at 1 year, data was obtained on 64, with 3 lost to follow-up & 2 moved abroad. Results of investigations or further clinical developments resulted in a changed diagnosis in 16%. There were 3 deaths at 2.2, 3.3 & 9.0 years after presentation and 1 transplant, at 5.2 years. Overall 5-year survival was 79% and event-free survival 63%; or 78% and 61% respectively at last follow-up. For the 60 event-free survivors, median(range) follow-up duration was 9.04(5.0-10.33) years for those still under review (n=46), or time to discharge 5.13(0.67-10.0) years (n=14). Fifty-eight were in NYHA Class 1, and two in Class 2. Twenty-seven patients (45%) required one or more readmissions to hospital. On echocardiogram, 41/60 had normal studies at last follow-up. Fourteen cases had mild functional impairment, and one moderate. Mitral regurgitation was mild in 8 and moderate in 1. No intracardiac thrombi were seen. Twenty-three patients remained on medication including ACE inhibitors(n=22),  $\beta$ -blockers(n=13 of which 9 on carvedilol), diuretics(n=2), digoxin(n=3), aspirin(n=3), losartan(n=1), & flecainide(n=1).

**Conclusions**

Children who survive the first year following a diagnosis of heart failure from heart muscle disease have a good longer-term outcome and quality of life although there remains a small attrition rate. Good heart failure management remains important in those with persisting echocardiographic abnormalities or symptoms.

**Reference:**

1. New Onset Heart Failure from Heart Muscle Disease in Childhood: A Prospective Study in the United Kingdom and Ireland. *Circulation* 2008;117:79-84.