

## Idiopathic Restrictive Cardiomyopathy in Children and Young Adults: A Clinical Update

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**Background:** Idiopathic restrictive cardiomyopathy (IRCM) is a rare condition characterized by reduced ventricular compliance. Children with IRCM have poor outcomes with most patients proceeding to cardiac transplantation. We sought to analyze our institutional experience and assess contemporary outcomes for children with IRCM.

**Methods:** We reviewed the medical record for all patients (< 21 years old) evaluated for a primary diagnosis of IRCM between 1975 and 2013 at our institution. Demographic, clinical, echocardiographic, and catheterization data were abstracted. Patients with cardiomyopathies other than IRCM were excluded. The patients were divided into two groups comprising a historical cohort (HC) (diagnosis: 1975-1993, n=8) and a contemporary cohort (CC) (diagnosis: 1994-2013, n=12).

**Results:** Twenty children were identified with IRCM (mean age at presentation  $9.7 \pm 6.5$  yrs, 55% female). Mean length of follow up was  $6.5 \pm 8.4$  years (range 0.1-35.6 years). In the CC, 6 of 12 patients (50%) progressed to cardiac transplantation (mean age of  $9 \pm 4$  yrs at transplant, mean interval from diagnosis of IRCM:  $1.5 \pm 0.9$  yrs). Overall survival was improved significantly in the CC compared to the HC (80% vs. 37%,  $p = 0.02$ ), but transplantation free survival was no different between the CC and HC (50% vs. 37%,  $p = 0.16$ ). In the CC, degree of elevation of mitral valve Doppler E/e' ratio on echocardiography was associated with increased mortality ( $p = 0.01$ ).

**Conclusions:** IRCM continues has a poor prognosis. Early referral for transplantation was associated with improved overall survival in the modern era. Patients with markedly elevated E/e' ratio have increased risk of death.

Figure Headings:

Figure 1: (a) Kaplan-Meier curve of overall survival for IRCM patients in historical and contemporary cohorts ( $P = 0.02$ ). (b) Kaplan-Meier curve of transplant free survival for IRCM patients in historical and contemporary cohorts ( $P = 0.36$ ).

Figure 1a

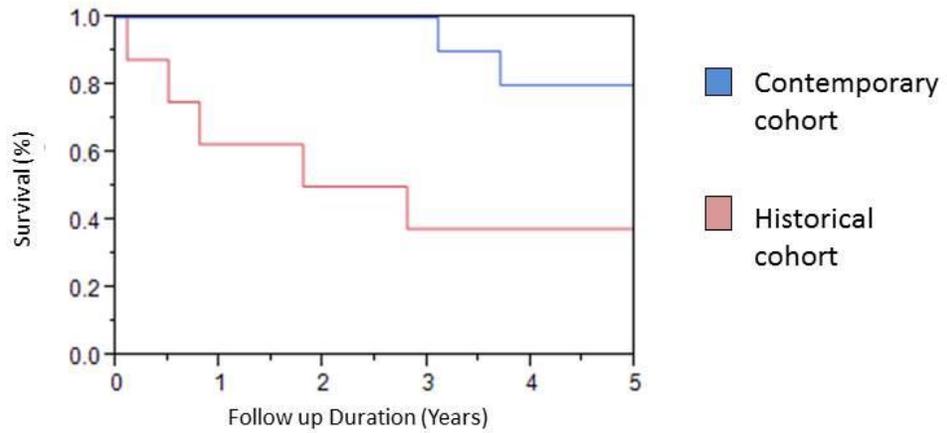


Figure 1b

