

**Clinical Outcomes of Restrictive Cardiomyopathy in the paediatric population**

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**Introduction**

Restrictive cardiomyopathy (RCM) is rare in childhood. It is characterised by diastolic dysfunction with normal or near normal systolic function, atrial enlargement and normal or near normal left ventricular wall thickness. Diastolic failure leads to congestive heart failure and pulmonary hypertension (PHT). The aim of this study was to review our institutional experience in patients with RCM.

**Methods**

Retrospective review of all patients diagnosed of RMC between September 2002 and October 2018. Demographic data, mode of presentation, echocardiographic and haemodynamic findings and clinical outcome were evaluated.

**Results**

8 children with RCM were identified (4 males, median age 3 years). At the time of diagnosis, 5/8 were considered to have pure RCM and 3/8 RCM/non-compacted cardiomyopathy (LVNC). All the cases were primary cardiomyopathies except one patient with endomyocardial fibrosis due to schistosomiasis.

Mean follow-up was 37.9 months (0.5 - 91). Overall, 6/8 patients developed congestive heart failure and 6/8 PHT. 3/8 patients had arrhythmic events during follow-up: 1 patient had a thromboembolic complication after atrial fibrillation, another developed complete heart block and cardiogenic shock and 1 patient had a sudden cardiac death on the transplant list. All patients with arrhythmias were dead or transplanted at the end of the study.

Out of the all the CT patients with cardiomyopathies in our institution, 5 (14%) were due to RCM. 4 patients received CT and 1 cardiopulmonary transplant. 2 patients required pre-transplant mechanical circulatory assistance (1 Berlin Heart; 1 ECMO and Berlin Heart). 1 patient underwent ECMO post-CT because of RV failure.

**Conclusions**

Due to the poor outcomes of RCM, early consideration for heart transplant is recommended. Arrhythmias are an important clinical finding in patients with RCM and prophylactic pacing or implantable cardioverter-defibrillator system has to be considered as paediatric patients with RCM are at risk of sudden cardiac death.