**COMPARISON OF ECHOCARDIOGRAPHIC CRITERIA FOR DIAGNOSING LEFT VENTRICULAR NONCOMPACTION IN PATIENTS WITH CLINICAL CARDIOMYOPATHY**

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**Background:** Isolated noncompaction of the ventricular myocardium (INVM) is a cardiomyopathy characterized by excessive prominent trabeculations with deep intertrabecular recesses in the left ventricular apex. Associated findings include systolic dysfunction, ventricular arrhythmias, and systemic emboli. Management is challenging not only due to variability in developing clinical myopathy, but also because multiple diagnostic criteria are in use. INVM remains most commonly diagnosed by echocardiography with 3 criteria in use; the criteria proposed by Lai based on the study by Chin, and subsequent criteria proposed by Jenni and Stollberger.

**Objective:** We evaluated INVM criteria to compare the sensitivity for detecting patients ultimately developing clinical cardiomyopathy (increased LV trabeculations, LV systolic dysfunction > 2 SD below normal).

**Methods:** 10 echocardiograms from 4 patients with clinical INVM were reviewed independently by 2 pediatric cardiologists and the inter-observer agreement was assessed.

**Results:** The average fractional shortening measured 21.9% ± 6.2%. The Chin criteria X:Y ratio was 0.31 ± 0.1 (normal > 0.5) with 100% interobserver agreement. Fractional shortening and X:Y ratio showed modest correlation with a R2 =0.43. Using the Stollberger criteria, INVM was likewise diagnosed in all patients, but interobserver agreement was 56%. The Jenni criteria diagnosed INVM in 1 of 4 patients by one cardiologist, and 4 of 4 patients by the second cardiologist with an interobserver agreement of 29%.

**Conclusion:** The Chin criteria most consistently diagnosed INVM in patients with clinical cardiomyopathy with the best inter-observer agreement. Criteria for diagnosis were met in some patients prior to developing clinical cardiomyopathy indicating there may be a predictive value in using these criteria. Further, there is modest correlation between function and X:Y ratio suggesting a correlation between degree of hypertrabeculation and severity of ventricular dysfunction in INVM. However, all patients were ultimately diagnosed with INVM using any of 3 criteria on at least one echocardiogram by at least one cardiologist.