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ORIGINAL ARTICLE

Left Ventricular Non-Compaction: A Cardiomyopathy With Acceptable Prognosis in Children

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Background Data on children with left ventricular non-compaction (LVNC) is sparse. The purpose of this study was to evaluate its clinical profiles in a population of Chinese children.

Methods From January 2010 to March 2016, consecutive Chinese children (aged <18 years) with LVNC diagnosed by cardiovascular magnetic resonance (CMR) were prospectively recruited at Fuwai Hospital.

Results A total of 41 Chinese children (male: 28%; mean age: 14 ± 4 years) were included in this study. Left ventricular non-compaction was not detected in 13 (32%) patients at initial echocardiographic evaluation. Congenital heart disease (CHD) was found in 11 (27%) patients. Four (10%) patients had Wolff-Parkinson-White (WPW) syndrome. Mean left ventricular ejection fraction (LVEF) was 41 ± 15%. Late gadolinium enhancement (LGE) was detected in 8 (20%) subjects. During a mean follow-up of 2.9 years, 4 (9%) patients died or received heart transplantation. These patients had lower systolic blood pressure (91 ± 10 vs. 108 ± 14 mmHg; p = 0.02), diastolic blood pressure (57 ± 7 vs. 68 ± 8 mmHg; p = 0.007) and LVEF (19 ± 7 vs. 44 ± 12%; p = 0.002) than the survivors. In addition, advanced heart failure (100% vs. 16%; p = 0.002) and LGE (50% vs. 5%; p = 0.04) were detected more in these subjects.

Conclusions Left ventricular non-compaction is easily overlooked at echocardiographic assessment. Congenital heart disease and WPW syndrome were relatively common in LVNC children. The prognosis of children with LVNC seemed to be better than previous studies reported, and its long-term prognosis needs to be further investigated.

Keywords Left ventricular non-compaction • Chinese children • Children • Clinical presentation • Prognosis

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Introduction

Left ventricular non-compaction (LVNC) is morphologically characterised by a two-layered structure of the myocardium, which consists of a thick non-compacted inner layer and a thin compacted outer layer. Some LVNC cases are congenital and the others seem to be acquired [1–4]. It may occur in isolation or in association with congenital heart disease (CHD). Different phenotypes can be found in LVNC, including benign, dilated, hypertrophic, restrictive and biventricular subtype [5]. Therefore, LVNC is still classified as an unclassified cardiomyopathy by the European Society of Cardiology although it is considered as a primary cardiomyopathy by the American Heart Association [6,7].

Echocardiography is the first-line imaging technology to assess this cardiomyopathy. However, LVNC is likely to be overlooked due to its limited imaging resolution [8]. Cardiovascular magnetic resonance (CMR) is superior to echocardiography in recognising non-compacted myocardium because of its higher spatial and temporal resolution, and it is also free of limitations in body habitus or imaging planes [9]. In addition, late gadolinium enhancement (LGE) technique can assess myocardial fibrosis or infiltration [9].

Left ventricular non-compaction shows different clinical presentations, ranging from asymptomatic condition, congestive heart failure, arrhythmia to systemic embolism or even sudden cardiac death (SCD). Initially, LVNC was considered a rare disease [10]. However, more and more LVNC cases have been detected during the past two decades [11,12]. Considering the clinical heterogeneity and the large population, there is an urgent need to screen the patients at high risk for unfavourable outcomes. Most previous LVNC studies focussed on the characteristics of adult patients [11,13–15]. But data from young patients has been sparse. Therefore, we performed this study to investigate the clinical profile and prognosis of LVNC in a cohort of Chinese children, who were diagnosed on the basis of CMR.

Methods

Patients

From January 2010 to March 2016, consecutive Chinese children with LVNC (aged <18 years) were prospectively recruited at Fuwai Hospital. Patients and available family members received systemical clinical assessment. Left ventricular non-compaction was diagnosed in reference to the CMR criteria suggested by Petersen [16]. These criteria comprised: (1) Two-layered structure of the myocardium; (2) prominent myocardial trabeculations and deep intertrabecular recesses in communication with the left ventricular (LV) cavity; (c) end-diastolic ratio between noncompacted and compacted myocardium >2.3. All enrolled patients were willing to take part in this research and the parents or guardians together with the children provided their informed consent documents. This study followed the Declaration of Helsinki and was approved by the Ethics Committee of Fuwai Hospital, Peking Union Medical College, Beijing, China.

Follow-Up

The primary endpoints were cardiovascular death (including SCD, heart failure-related death and unclassified death) and heart transplantation. Sudden cardiac death was defined as unexpected natural death from a cardiac cause occurring within one hour after the onset of symptoms. Heart failure-related death was defined as death occurring in the context of long-time cardiac decompensation with progression of the disease over the previous year. Unclassified death was defined as death related to LVNC but could not be classified as SCD or heart failure-related death. Other major adverse cardiovascular events included hospitalisation for heart failure, sustained ventricular tachycardia/ventricular fibrillation (SVT/VF), appropriate defibrillation shock from an implantable cardioverter defibrillator (ICD) and systemic embolism.

Statistical Analysis

Data were expressed as mean ± standard deviation for continuous variables or as percentages for categorical variables. Differences in continuous variables were evaluated with the Student t test. The Wilcoxon rank-sum test was used when the distributions were unsymmetrical. The χ^2 test was used for categorical analysis of nominal data. The Fisher's exact test was used if the patient number in any group was less than five. Statistical analyses were conducted with SPSS statistical software (version 20.0; IBM Corp., Armonk, NY, USA). All statistical tests were two-sided and p values <0.05 were considered to be statistically significant.

Results

Baseline Characteristics

There were 41 Chinese children enrolled in this study (Table 1). The mean age of these patients was 14 ± 4 years (range 6.9–17.9 years) and 28 (68%) were male. Thirteen (32%) patients were not found with LVNC in echocardiographic evaluation. Among these patients, four were diagnosed with DCM, two HCM and one, RCM. The remaining six patients were diagnosed with unexplainable left LV dilation. A CHD was detected in 11 (27%) patients. Atrial septal defect was the most common type and found in three (7%) patients. Two (5%) patients had neuromuscular disorders, including one Becker muscular dystrophy and one myasthenia gravis. A history of systemic embolism was found in one (2%) patient. Four (10%) children had a family history of cardiomyopathy. Of all the children, 31 (76%) patients were symptomatic. Exertional dyspnoea was the most common symptom and found in 22 (54%) subjects. Ten (24%) patients had advance heart failure (New York Heart Association functional class III/IV). Electrocardiographic abnormality was observed in 37 (90%) patients. High voltage QRS complex was the most common finding and documented in 10 (24%) subjects.

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