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Left Ventricular Noncompaction in Childhood: Four Years Single Center Experience from Turkey

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Introduction: Left ventricular noncompaction (LVNC) is a specific cardiomyopathy that occurs following a disruption of endomyocardial morphogenesis. This study presents clinical findings, diagnostic features, treatment and follow-up of pediatric LVNC patients.
Methods: Enrolled in the study were patients that applied to our clinic between January 2010 and December 2013 with isolated LVNC or LVNC with congenital heart disease (CHD). We evaluated patients for rhythm disorders, and in cases with CHD, for surgery and complications.
Results: Study population was comprised of 50 patients, 24 male (48%). LVNC was isolated in 29 (58%) and with CHD in 23 (46%). Clinical features are summarized in Table 1. Mean age at diagnosis was 4.35 years ± 88 months (4 days-19 years). Findings at admission were heart murmur in 28 (56%), congestive heart failure in 10 (20%), dyspnea in 5 (10%), cyanosis in 4 (8%), growth and developmental retardation and pneumonia in 3 (6%). Echocardiography revealed a shortening fraction of 29±11% (9–50%), ejection fraction of 52.7±19.3 (14-80), and LVDD of 34±13 mm (17-64 mm). Noncompaction was apical in 9 patients (18%), free wall in 14 (28%), apical and free wall in 20 (40%), biventricular in 2 (4%) and less typically located in 5 (10%). Electrocardiographic abnormalities were present in 35 patients (70%). Typical findings were ST-T changes 22%(n=11), LV hypertrophy 10%(n=5) and total AV block 6%(n=3). The more rare findings included left axis deviation 8%(n=4), first-degree AV block 6%(n=3), low voltage 4%(n=2), VES 4%(n=2), nonspecific intraventricular conduction delay 2%(n=1), VT/VF 2%(n=1), biventricular hypertrophy (1), RBBB 2%(n=1) and LBBB 2%(n=1). Mean follow-up period was 13 months ± 220 days (range, 1-46 months). During follow-up, 10 patients developed cardiac insufficiency, 7 rhythm disoders and 3 intracardiac thrombosis. One of these underwent transvenous ICD implantation due to VF. No patients died; 3 are currently on transplant waiting lists.
Conclusions: LVNC is a primary myocardial disease that presents with varied clinical, electrocardiographic and echocardiographic findings in childhood. Concomitant LVNC must especially be considered in CHD accompanied by LV dysfunction. Early diagnosis can prevent possible life-threatening complications.

Table 1. Classification of LVNC Patients

LVNC Category			**n (%)**
• **Total**			**50 (100)**
• **Isolated**			29 (58)
• **With Congenital Heart Disease**			21 (42)
***Acyanotic**	**13**	***Cyanotic**	**8**
– VSD	4	– TA, VA concordance, VSD	2
– MVP, myxomatous MV, Cleft, MI	2	– c-TGA, VSD, PS	1
– VSD, ASD	2	– Critical PS, PDA	1
– PDA	2	– SSD, PA, ARV/PA, VSD	1
– BAV, PDA	1	– TA, RVH, VSD, ASD, PDA	1
– BAV, AS, AI	1	– PA/IVS	1
– ASD, Ebstenaoid TV, TI	1	– Unguarded TV, severe PI,	1

BAV: bicuspid aortic valve, **AI**: aortic insufficiency; **AS**: aortic stenosis; **VSD**: ventricular septal defect; **MI**: mitral insufficiency; **PA**: pulmonary atresia; **PS**: pulmonary stenosis; **PH**: pulmonary hypertension; **PI**: pulmonary insufficiency; **TV**: tricuspid valve; **TI**: tricuspid insufficiency; **TA**: tricuspid atresia; **c-TGA**: corrected transposition of great artery; **RVH**: right ventricular hypoplasia; **VA**: ventriculoarterial; **SSD**: situs solitus dextrocardia; **ARV/PA**: aorta arising from right ventricle with pulmonary atresia, **PA/IVS**: pulmonary atresia with intact ventricular septum