Isolated left ventricular noncompaction

Sri Ramachandra University, Forur, Chennai, India

Background: Left ventricular noncompaction is a very rare entity with prevalence rate of 0.014% in western population. There is no report from our country.

We report consecutive cases of isolated left ventricular noncompaction of SIX. We discuss historical, clinical, surface electrocardiogram, and echo-cardiographic features of this condition.

Result: One case has been linked with positive family history, wherein father and son diagnose to have left ventricular noncompaction. In this series of cases, common localization of noncompacted segments were in the apex and lateral wall of left ventricular myocardium. Severe LV systolic dysfunction (EF < 35%) seen in four cases. Grade II mitral regurgitation in four cases and two cases had pulmonary artery hypertension.

Conclusion: High degree of suspicion in all cases of unexplained severe left ventricular dysfunction is necessary to identify left ventricular noncompaction.

Heart rate manipulation in dilated cardiomyopathy: Assessing the role of ivabradine

Deep Chand Raja, Aditya Kapoor*, Roopali Khanna, Sudeep Kumar, Naveen Garg, Satyendra Tewari, P.K. Goel
Department of Cardiology, SGPGI, Lucknow 226014, India

Background: Heart rate reduction has been proven to be of benefit in patients of chronic heart failure (HF). Despite advances in management of HF, the morbidity and mortality remains high, necessitating the development of novel therapeutic strategies. Ivabradine is a selective sinus rate slowing agent which inhibits the cardiac If current that is responsible for the spontaneous diastolic depolarization in the sinus node. The role of ivabradine in HF secondary to dilated cardiomyopathy (DCM) merits further study.

Methods: The study included 103 patients with DCM (mean age 47.9 ± 14.8 years, NYHA class 3.2 ± 0.4, BNP 742 ± 489 pg/ml, LVEF 26.3 ± 3.6%). Patients were randomized to receive ivabradine (initiated at 2.5 mg BD titrated up to 7.5 mg BD at intervals of 2 weeks, titrated up to 7.5 mg BD, n = 52, Group 1) added to standard therapy (beta blockers, ACE inhibitors, diuretics, n = 51, Group 2). Functional class, BNP levels, and echocardiographic parameters including LV volumes, LV end-systolic stress (LVESS), LV mass, LV work, and global LV strain were assessed at baseline, 3 months and 6 months follow-up.

Results: Baseline demographics including age, NYHA Class, BNP levels, 6 min WT and MLWHF (Minnesota Living with HF questionnaire) scores were comparable between the two groups. There was no difference in echocardiographic systolic and diastolic parameters amongst Groups 1 and 2 at baseline, including LVEF 26.06 ± 3.5% vs 26.7 ± 3.1% (p = 0.34), LVESS 205 ± 44.3 vs 193 ± 5.8 (p = 0.2), LV mass 121.7 ± 32 g vs 126 ± 39 g (p = 0.5), LV work 531.2 ± 146 vs 469 ± 126 mmHg/lm (p = 0.06), and LV global strain −10.4 ± 1.3% vs −10.1 ± 1.6% (p = 0.2). The mean dose of ivabradine used was 12.1 mg ± 2.4 mg (range 5–15 mg); all patients tolerated ivabradine well, except for rise in serum creatinine requiring withdrawal of the drug in one patient.

3-months follow-up: Both groups had significant improvement in mean NYHA class (Gp 1 from 3.3 ± 0.4 to 2 ± 0.0, p = 0.001, Gp 2 from 3.2 ± 0.4 to 2 ± 0.3, p = 0.03), 6 min walk test (Gp 1: 326 ± 42 to 370 ± 38 ml, p = 0.001, Gp 2: 336 ± 76 to 364 ± 77 ml, p = 0.03), MLWHF questionnaire (Gp 1: 74 ± 14 to 58 ± 11, p = 0.001, Gp 2: 78 ± 8 to 65 ± 10, p = 0.001) and fall in BNP (Gp 1: 760 ± 490 to 382 ± 193 pg/ml, p = 0.001, Gp 2: 724 ± 492 to 420 ± 289 pg/ml, p = 0.01). However, the % change in 6 min WT, MLWHF and BNP was much higher in Ivabradine group (14%, −21% and −35% vs +9.4%, −16% and −30% respectively with corresponding p values of 0.06, 0.02, 0.05). While both groups demonstrated reduction in HR (95.6 ± 12 to 80.3 ± 7 bpm, p = 0.001 in ivabradine group, 94.6 ± 9 to 87.3 ± 7 bpm, p = 0.001 in controls), the % change was significantly higher in the ivabradine group (−15% vs −10.2%, p = 0.03). No significant reduction in systolic or diastolic BP was seen in the ivabradine group.

At 3 months, amongst those receiving ivabradine, there was significant improvement in indexed LV end-systolic (LVESSVI, 86 ± 19 to 73 ± 15 ml/m², p = 0.001) and LV end-diastolic volumes (LVESDVI 117 ± 34 to 103 ± 19 ml/m², p = 0.001), MPI (0.82 ± 1.2 to 0.7 ± 0.1, p = 0.001) and LVEF (26 ± 3.5 to 29 ± 3.6%, p = 0.001). The % change in LVESVI, LVEDVI, MPI and LVEF was −15%, −15%, −11% and +12.2% respectively. In contrast, amongst controls there was no significant change in LVESVI (83 ± 20 to 81 ± 21 ml/m², p = 0.2), LVEDVI (113 ± 27 to 113 ± 24 ml/m², p = 0.9), MPI (0.79 ± 1 to 0.75 ± 0.1, p = 0.06) and LVEF (26.7 ± 3.7 to 26.7 ± 4.5%, p = 0.9).

6 months follow-up: At 6 months, while both group of patients had further improvement in NYHA class, 6 min WT, MLWHF score and fall in BNP, the % change in all parameters was significantly higher with ivabradine group (6 min WT: (19% vs 11.6%, p = 0.01, MLWHF score 22% vs 16%, p = 0.01, fall in BNP 64% vs 31%, p = 0.001). At 6 months, the % reduction in HR was also significantly greater in those on ivabradine (−19.5% vs −7.5%, p = 0.001). Importantly, 96.1% achieved HR < 70/min at 6 months on therapy with ivabradine as compared to only 25.6% of those on standard treatment.
Only patients in ivabradine demonstrated further improvement in LVEFVi (73 ± 15 ml/m² to 62 ± 15 ml/m², p = 0.001), LVEDVi (103 ± 19 ml/m² to 95 ± 18 ml/m², p = 0.001), MPI (23% vs 27%, p = 0.001), LVEF (29 ± 3.6 to 34.5 ± 5.4%, p = 0.001) and LV Global Strain (−13.3 ± 1.6 to −20.2 ± 4.4%, p = 0.001) while those in the control group did not show any further change. Parameters like E/A, E/A VTI, MPI, E'/E' septal and E'/E' lateral showed sustained improvement at 6 months only in the ivabradine group – E/A (1.5 ± 0.5 to 1.1 ± 0.3, p = 0.001), A/E VTI (1.6 ± 0.5 to 1.1 ± 0.3, p = 0.001), E'/E' septal (11.6 ± 3.3 to 9.5 ± 2.2, p = 0.001), E'/E' lateral (11.6 ± 3.2 to 9.2 ± 2.5, p = 0.001).

On multivariate analysis, heart rate reduction was a stronger predictor of improvement in LVEF (p = 0.03) than ivabradine treatment (p = 0.5). Change in HR significantly correlated with improvement in NYHA class (r = +0.4, p = 0.01), fall in BNP levels (r = −0.4, p = 0.001), better EF (r = −0.38, p = 0.001) and reduction in global LV strain (r = +0.28, p = 0.03) at 6 months.

Conclusion: Ivabradine added to standard therapy in patients of DCM with chronic HF produces greater % change in all assessed parameters including functional class and echocardiographic parameters especially global LV strain. Ivabradine therapy was four times more likely to result in HR reduction to <70 bpm at 6 months. Heart rate manipulation with ivabradine may play an important role in the management of such patients.

**Cardiac beriberi presenting as severe PAH with impending congestive heart failure in infants of poor socioeconomic background**

P.A. Kasar*, A.N. Patnaik
STAR Hospitals, 8-2-594/B, Road No. 10, Banjara Hills, Hyderabad 500034, India

**Introduction:** Severe pulmonary artery hypertension (PAH) in infants is rare presentation without congenital heart disease (CHD). Cardiac beriberi is often a missed diagnosis and is still common. Thiamine supplements in unexplained congestive heart failure (CHF) with severe PAH in a patient from poor socioeconomic (SE) status may be life saving.

**Materials and methods:** A total of 25 infants (male 18, female 7) were found to have unexplained severe PAH during period of June 2012 to June 2015 were included. All infants were exclusively breast fed belonging to low SE strata with their mothers from predominant rice eating population. Infants with CHD were excluded. Presentation was poor feeding, excessive cry and/or respiratory distress. 2D Echo done showed severe PAH and RV dysfunction in all; 2 patients had associated pericardial and pleural effusion. Diagnosis of cardiac beriberi was made and thiamine supplement were started immediately. No other PAH medications were given.

**Results:** The average age of presentation was 3.5 months (range 1–6 months); weight 4.3 kg (2.6–5.6 kg). Arterial blood gas (ABG) analysis showed high lactate levels (mean 6.4, range 3.4–17 mmol/l) at presentation. Mean PA pressures were 74 mmHg (range 50–110 mmHg) at admission. The lactate levels were higher in infants less than three-month age (n = 10) when compared with older infants (n = 15) between 3-month and 6-month age group (lactate 9.6 mmol/l vs 4.2 mmol/l respectively, p < 0.05). The infants presenting before three months had severe CHF and two infants presented with acute gasping state with severe lactic acidosis requiring mechanical ventilation. The response of thiamine supplement was rapid, ABG