Left ventricular non-compaction (LVNC) is a congenital dysfunction of ventricular morphogenesis. The World Health Organization has categorised it as an unclassified cardiomyopathy. It is thought to be the result of an arrest in the normal endomyocardial embryogenesis, which leads to the persistence of intertrabecular recesses and the development of a spongy myocardium. LVNC is now being diagnosed with increasing frequency, either in isolation or combined with congenital heart disease or neuromuscular disorders. Here we report the case of a 2-month-old male infant who presented with congestive heart failure (HF) where isolated LVNC was found on echocardiography.

Case report
A 2-month-old male infant was admitted to the paediatric intensive care unit (ICU) of Krishna Hospital, Karad, with shortness of breath and respiratory distress which commenced one day prior. The baby was born preterm at 30 weeks of gestation to non-consanguineous parents and was admitted for preterm care in the neonatal ICU. The hospital stay was unremarkable and the infant was discharged at a weight of 1.6 kg after 21 days. The baby was asymptomatic thereafter.

On admission the patient had features of congestive HF. Cardiovascular examination revealed a decreased volume peripheral pulse and no obvious murmur was appreciated. There was hepatomegaly 4 cm below the right costal margin. No dysmorphic features were observed. A chest X-ray revealed substantial cardiomegaly. Electrocardiography showed left ventricular overload, but no signs of ventricular arrhythmias. No neuromuscular abnormality was found.

An echocardiogram showed situs solitus, left aortic arch and atrioventricular-ventriculoarterial concordance; pulmonic veins drained normally to the left atrium and inferior vena cava to the right atrium. The inter-atrial and interventricular septums were intact. The left ventricle (LV) was markedly dilated with a severely decreased global systolic function (ejection fraction 30%). Hypertrabeculation was evident with multiple crypts at the apical posterolateral aspects of the LV (Fig. 1). Colour Doppler examination showed prominent LV trabeculations with deep intertrabecular recesses in continuity with the LV cavity. The ratio of non-compacted to compacted layer of the myocardium was >2 (Fig. 2).

The patient was treated with inotropes, decongestive therapy and captopril. His condition stabilised within 1 week; he was discharged and prescribed captopril and frusemide, and advised to follow-up regularly.

Discussion
Non-compaction of the myocardium was first described by Bellet et al., from an autopsy carried out on a newborn in whom aortic
Isolated LVNC is a rare, genetic, congenital disorder. Due to easy availability of diagnostic modalities and awareness of this new type of cardiomyopathy, early recognition and prompt treatment are possible. Patients should also be screened for thromboembolism and arrhythmias. Due to the genetic transmission of LVNC, screening of close relatives is recommended, and the screening of the patient for neuromuscular diseases is also advised.

References