Sir,

Noncompaction of the ventricular myocardium (NCVM) is a primary genetic cardiomyopathy.\(^1\) We observed an association of NCVM with mitral valve deformity leading to mitral regurgitation (MR) before, where strict criteria were used, including the measurement of compacted to noncompacted layer thickness ratio of \(>2:1\) and demonstration of intertrabecular recesses by color flow mapping.\(^2,3\)

We recently found three patients (all females) with this association presenting with heart failure due to MR. Symptoms started at the age of 3-7 years. All patients had echocardiographic thickening and retraction of both mitral valve leaflets and a zig-zag appearance of anterior leaflet which coapted superior to the posterior leaflet [Figures 1 and 2]. Severe MR was seen in all patients. The left ventricle ejection fractions were 60-65%.

The usual cause of MR in patients with NCVM is mitral valve annular dilatation due to myocardial dysfunction, however, ejection fraction was preserved in our patients. In addition, changes in the leaflets indicated a distinct pathology. Fibrotic and calcific changes were documented on histopathological examination in one of our previous patients as well as in a series of 14 patients reported by Burke.\(^2,4\) These reports support our observations.

Distinction of this new association from rheumatic MR is particularly important especially in older patients. In rheumatic MR, the posterior mitral leaflet is usually retracted and tethered to the left ventricle posterior wall while the anterior leaflet is hypermobile (personal observations), findings not present in our patients.

NCVM can have significant implications on the surgical treatment of patients with MR. In our experience with seven patients with NCVM and congenital heart disease who underwent surgical repair or palliation five out of seven (71%) developed post operative complications.\(^5\) In conclusion, we describe distinct echocardiographic features in three more cases with this new association: NCVM-MR that to the best of our knowledge have not been reported before except in our reports.\(^2\)

Sulafa Khalid M Ali
Department of Paediatrics and Child Health, Sudan Heart Centre and University of Khartoum, Faculty of Medicine, Khartoum, Sudan.
E-mail: sulafakhalid2000@yahoo.com

REFERENCES

1. Hare JM. The dilated, restrictive, and infiltrative cardiomyopathies. In: Libby P, Bonow RO, Mann DL, Zipes DP, editors. Braunwald’s Heart Disease A textbook of Cardiovascular Medicine. 8th ed, Vol. 2. Philadelphia: Saunders Elsevier; 2008. p. 1739-62.

2. Sulafa KM, Omran AS, Najm H, Godman MJ. Noncompaction of ventricular myocardium with mitral regurgitation and preserved ventricular function. J Am So Echocardiogr 2004;1:87-90.

3. Sulafa KM Ali. Noncompaction of the ventricular myocardium and mitral regurgitation: A unique association. Congenital Cardiology Today
Sir,

Transcatheter atrial septal defect (ASD) closure is now a widely recognized alternative to surgical closure for suitable secundum ASDs. We report closure of a large ostium secundum ASD (OS-ASD) which measured 40 mm on trans-esophageal echo (TEE) and was closed with a 46 mm device (Lifetech Scientific Inc., China). To the best of our knowledge, this is the largest size of the device used for closure of ASD till date.

A 36-year-old man from Bangladesh had complaints of shortness of breath on exertion and palpitation. On examination, he had wide and fixed split of the second heart sound with a soft ejection systolic murmur over the left second and third intercostal space. Trans-thoracic echo revealed a big OS-ASD measuring 38 mm with left to right flow and good rims of septal tissue all around with evidence of mild pulmonary arterial hypertension (PAH) and good biventricular function.

The patient preferred a nonsurgical mode of treatment and hence an ASD device closure trial was decided upon. His precatheterization investigation profile was unremarkable. TEE in the cath-lab measured 40 mm OS-ASD with adequate septal rim all around [Figure 1]. The patient was intubated and 7 French (Fr) venous and 6 Fr arterial access was obtained. Initial right heart catheterization was done. Calculated Qp/Qs was 2.1:1. His pulmonary artery pressure was 37/19 (mean 23) mm Hg. A 46 mm (Searcare Lot No 10012711) device was loaded using the standard technique in a 14 Fr delivery sheath. The 7 Fr venous sheath was exchanged with the delivery sheath. The device was placed in the right upper pulmonary vein (RUPV) with part of left atrial disk in RUPV. First the right atrial disk was delivered following which the left atrial disk fell into its place completely occluding the shunt. Device placement was checked with TEE [Figure 2], which showed no residual shunt and unobstructed flow across both the atrioventricular valves and systemic and pulmonary veins. Left ventricular end diastolic pressure remained same at 8 mm Hg both before and after the device placement. The device was deployed under cine guidance. The patient tolerated the procedure well and was extubated on table. He was given unfractionated Heparin infusion for 24 hours postprocedure along with an oral combination of aspirin and clopidogrel which he was advised to continue for 6 month. He remained asymptomatic after the procedure; his device position was confirmed before discharge. On follow-up he became asymptomatic and TTE and TEE after the first and sixth months showed the device in situ without evidence of any complication.

With the improvement in technology even very large ASDs can now be closed with the device. Devices of larger size are now available. Large size, full occluder devices, and large delivery sheaths are needed to maintain stability and also to deliver the device into the right atrium.