# Significant biventricular obstruction in non-syndromic hypertrophic cardiomyopathy

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## DESCRIPTION

Although hypertrophic cardiomyopathy (HCM) is classically considered a disease of the left ventricle (LV), we present a rare case of biventricular HCM presenting in middle age.

A 57-year-old man without a family history of HCM and sudden death presented with presyncope and New York Heart Association (NYHA) class II breathlessness. Clinical examination was normal except for ejection systolic murmur. ECG showed biventricular hypertrophy with deep T-wave inversions. Echocardiography demonstrated pronounced asymmetric LV hypertrophy and thickened right ventricle (RV) muscular components (figure 1A–C; videos 1–3). Doppler revealed a peak LV mid-cavity gradient of 109 mm Hg and subvalvar obstruction of the RV outflow tract with a peak gradient of 138 mm Hg (figure 2A–C). The patient underwent successful surgical myectomy and has been stable during the course of follow-up. The histopathological



**Video 1** Parasternal long axis view showing biventricular hypertrophy.

specimen revealed myofibril disarray as is expected in a case of HCM (figure 3). Genetic testing for Noonan's syndrome and three of the most common mutations were performed which were negative.
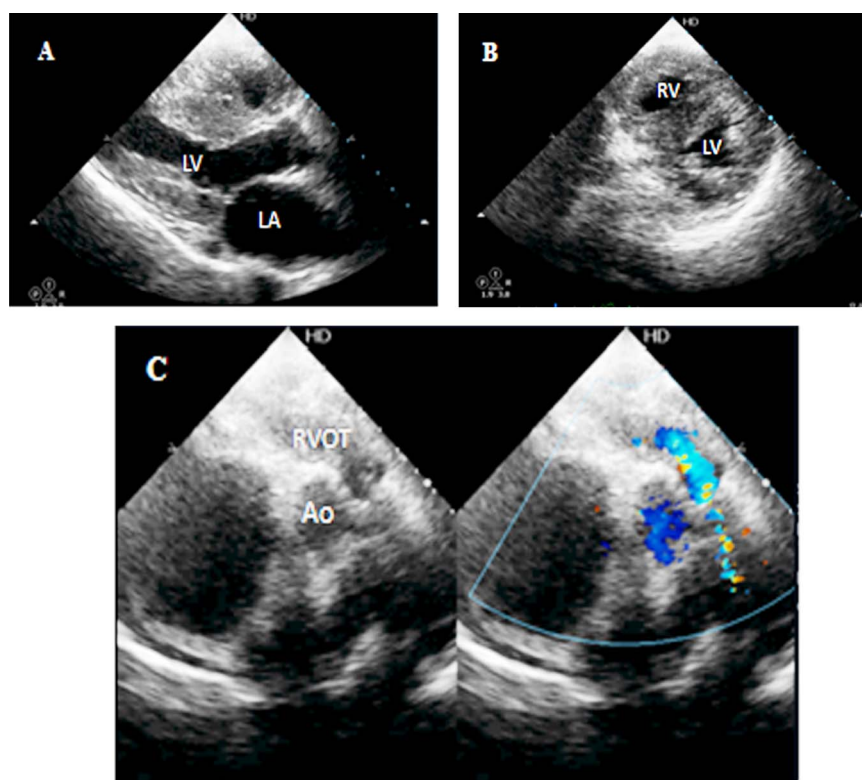


**Figure 1** (A) Parasternal long-axis view showing asymmetric septal hypertrophy. (B) Parasternal short-axis view showing biventricular hypertrophy. (C) Parasternal long-axis view showing hypertrophied muscle bundles in right ventricular (RV) outflow tract with colour flow turbulence (LV, left ventricle; RVOT, right ventricular outflow tract).

**Video 2** Parasternal short axis view at mid cavity level showing significant bi-ventriulcar hypertrophy.
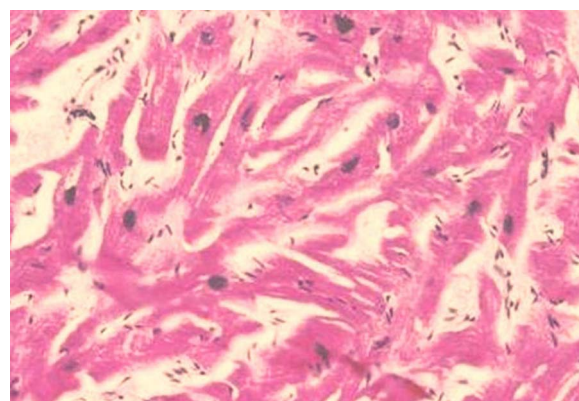


**Figure 3** Photograph of the histopathological specimen showing myofibril disarray.

**Video 3** Parasternal short axis view at the level of right ventricular outflow showing turbulence in the color Doppler signal across the RVOT suggesting RV outflow obstruction.
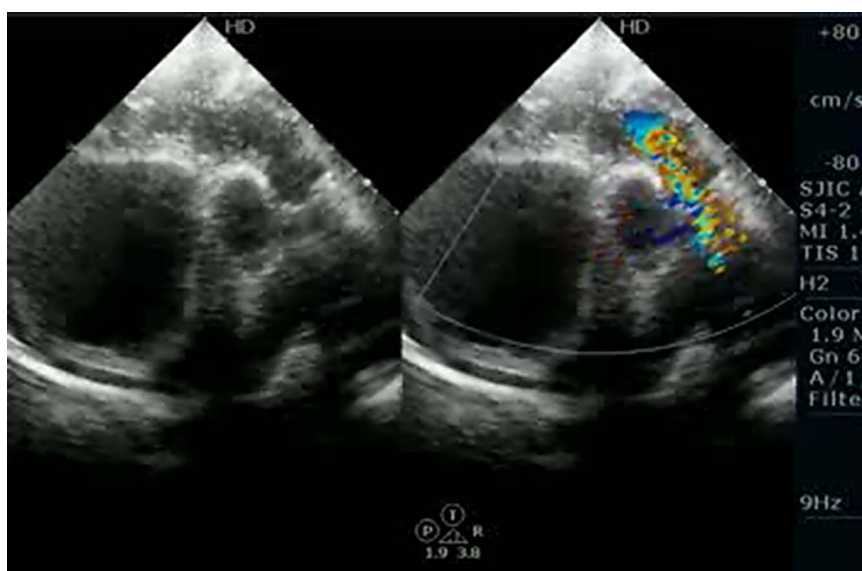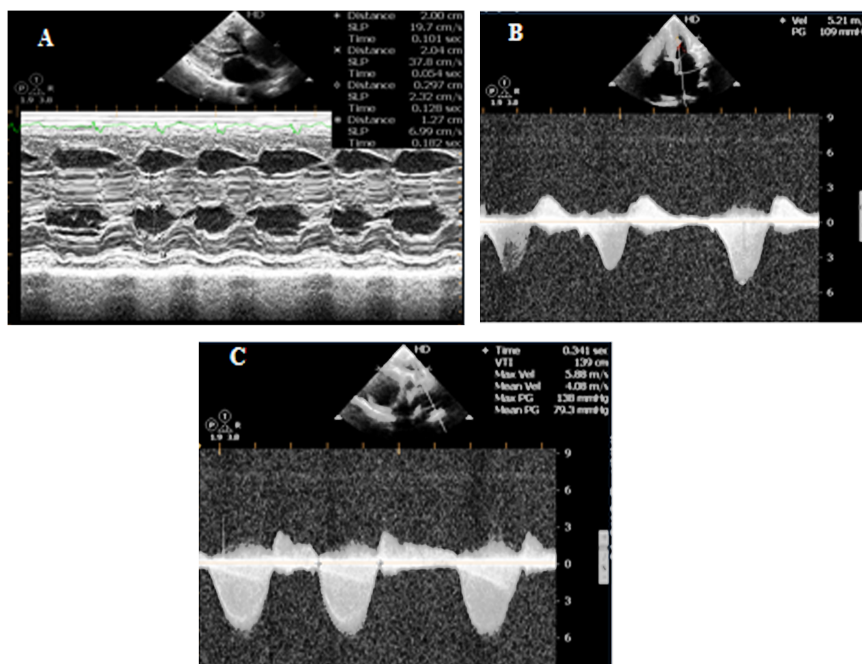


**Figure 2** (A) M-mode across left ventricle (LV) showing significant biventricular hypertrophy. (B) Continuous wave Doppler showing an LV mid-cavity gradient. (C) Continuous wave Doppler across right ventricular outflow tract showing significant gradient.

## Learning points

▶ Biventricular obstruction can occur in cases of non-syndromic hypertrophic cardiomyopathy, which should be kept in mind while evaluating the patient. Rare cases may present very late in the course of the disease, as in our case.

▶ The occurrence of significant right ventricular outflow obstruction can alter the management of the patient and can make percutaneous management extremely challenging and the results sometimes unsatisfactory.

▶ Surgical correction can provide a lasting outcome where myectomy is performed in both the chambers.

▶ The effect of the surgery on conduction blocks in cases with biventricular hypertrophy needs to be evaluated and can complicate an otherwise haemodynamically successful procedure. An extra bit of caution needs to be taken.

Others were not performed due to financial constraint on the part of the patient.

Biventricular HCM and valvular pulmonary stenosis is relatively common in Noonan's syndrome. However, most cases present early and this condition is rare in adults without a dysplastic pulmonary valve.[1] [2] It appears that biventricular HCM in young patients can initially be localised to only the RV.[3] In contrast to LV outflow obstruction, which is characteristically dynamic and due to mitral valve systolic anterior motion, RV obstruction is the result of a static and fixed impediment by excessive hypertrophy of muscular components. RV cardiomyopathy may or may not correlate with LV. While the genetics of RV involvement have not been well characterised, histological findings appear similar to those in the LV, suggesting similar pathogenesis. Surgical correction often gives successful and lasting outcome.[4]

**Competing interests** None.

**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

## REFERENCES

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