Aberrant atrial insertion of the mitral valve chord

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A 71-year-old female, with a clinical history of hypertension and hypothyroidism, presented with exertional breathlessness (New York Heart Classification II symptoms). Clinical examination revealed a pan systolic murmur of mitral regurgitation. Serum N-terminal pro b-type natriuretic peptide was elevated (238 ng/l).

Transthoracic echocardiogram demonstrated normal left ventricular chamber size and systolic function (ejection fraction >55%) with moderate mitral regurgitation. An echo-bright structure was attached to the lateral wall of the left atrium (LA; Figure 1a).

Cardiovascular MRI was performed that showed normal biventricular size and systolic function (left ventricular end-diastolic volume index of 62 ml/m^2) with appearance of localised prolapse of the middle posterior scallop of the mitral valve (P2). There was associated moderate mitral regurgitation (regurgitant fraction 38%) extending to the free wall of the LA with the suspicion of an aberrant mitral valve chord (Figure 2, Supplementary Videos 4 & 5).

The patient underwent transoesophageal echocardiogram that confirmed a single aberrant primary mitral valve chord. This arose from the ventricular surface of the posterior mitral valve leaflet, herniating through the mitral valve and attaching to the intra-atrial septum. There was moderate mitral regurgitation (vena contracta 0.4 cm, proximal isovelocity surface area radius 0.6 cm and regurgitant orifice area 0.32 cm^2) due to interference of valve closure by the papillary muscle head (Figure 1b–d, Supplementary Videos 1–3).

Figure 1 (a) Transthoracic echocardiogram showing echo-bright structure (arrow) attached to the atrial side of the basal interatrial septum. (b) Transoesophageal echocardiogram showing mitral regurgitation extending towards the free wall of the left atrium. (c) Long-axis view showing single aberrant mitral valve chord (arrow) arising from left atrium and attached to the posterior mitral valve leaflet. (d) Short-axis view showing aberrant mitral valve chord (arrow) arising from the lower portion of interatrial septum

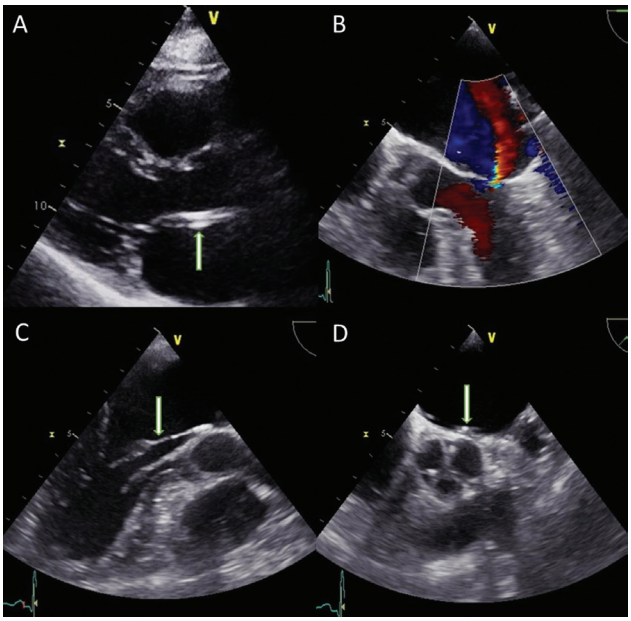

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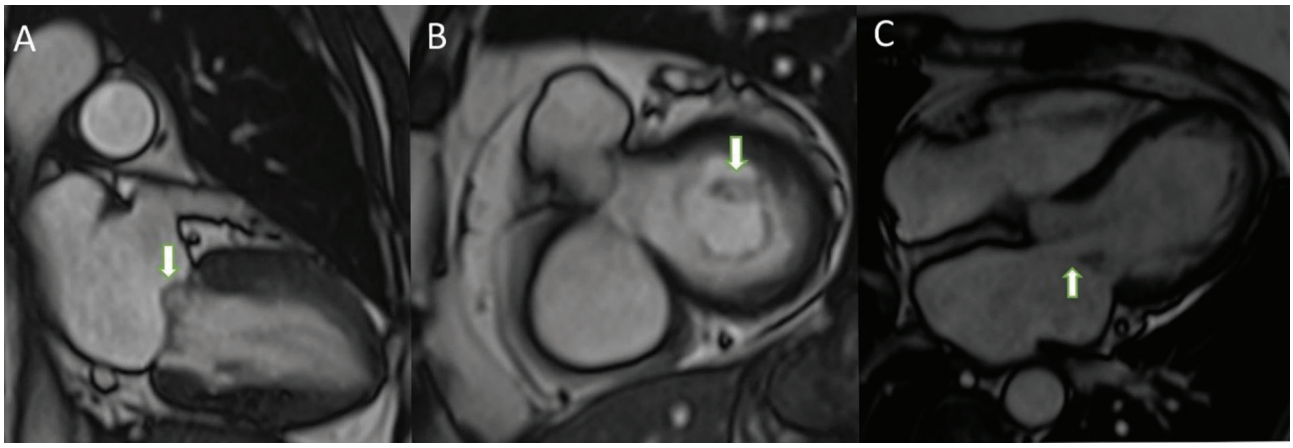
Figure 2 Cardiovascular steady-state free precession MR images. (a) Vertical long-axis view and (b) short-axis view showing localised prolapse (arrows) of the middle posterior scallop of the mitral valve (P2). (c) Four-chamber view showing suspected aberrant mitral valve chord (arrow) attached to interatrial septum



Aberrant mitral valve chord with resulting mitral regurgitation has been described as early as 1958.[1] Since then there has been a handful of cases describing the condition with resulting valve degeneration and mitral regurgitation.[2–4]

Aberrant mitral valve chord with anomalous insertion into the atrial wall should be regarded as a rare but important mechanism of congenital mitral regurgitation. When identified, aggressive management of hypertension in an attempt to prevent atrial dilatation and increasing severity of the mitral regurgitation is warranted.

Online Supplementary Material

Supplementary Videos are available with the online version of this paper, which can be accessed at https://www.rcpe.ac.uk/journal.

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