Isolated cleft posterior mitral valve leaflet: an uncommon cause of mitral regurgitation

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A 53-year-old woman with a history of hypertension was referred for an echocardiogram by her primary care physician after an unspecified abnormal ECG. The echocardiogram showed normal left ventricular size and function; however, an isolated cleft posterior mitral valve leaflet was identified with concomitant bileaflet prolapse and mild mitral regurgitation. She was subsequently referred to a cardiologist for clinical evaluation. Cleft mitral valve leaflet (CMVL) is an uncommon congenital cause of mitral regurgitation. Clefts, defined as slit-like holes or defects, are hypothesized to be a result of incomplete expression of an endocardial cushion defect which most commonly involves the anterior mitral valve leaflet with a paediatric incidence of 1:1340. Clefts affecting only the posterior mitral valve leaflet are extremely rare with only four cases being reported in the medical literature. Important co-existing anomalies with either posterior and/or anterior CMVL include counterclockwise rotation of the papillary muscles, the presence of an accessory papillary muscle or mitral valve leaflet, atrial septal defects, and mitral valve prolapse. Regurgitation from CMVL can lead to important physiological and anatomical changes within the cardiac system. Regurgitation results from blood flow directly through the cleft itself or from malcoaptation from accessory chordae with or without papillary muscle distortion. Significant chronic mitral regurgitation elevates left atrial filling pressures and leads to chamber enlargement and eccentric left ventricular hypertrophy. Early detection through two-dimensional echocardiography can provide accurate anatomical images of the various mitral valve structures and identify associated congenital anomalies. Early surgical correction is preferred before mitral regurgitation causes unfavourable remodelling. Most mitral valve cleft defects can easily be repaired by suturing the edges of the cleft. If a cleft resection leads to limited residual valve tissue, the leaflet of the mitral valve can be reconstructed using an autologous pericardial patch pre-treated with buffered glutaraldehyde. Posterior CMVL is an uncommon but clinically important cause of mitral insufficiency. Early recognition of this rare clinical entity and possible co-existent anomalies can identify the patients who would benefit from surgical intervention before compensatory left ventricular remodelling and contractile dysfunction develop.

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A 53-year-old asymptomatic woman with a history of hypertension was referred for transthoracic echocardiography by her primary care physician after an unspecified abnormal ECG. Significant physical examination findings included a II/VI holosystolic murmur at fourth intercostal space with radiation to the axilla. The echocardiogram showed normal left atrial and ventricular size and function with an isolated cleft posterior mitral valve leaflet that bisected into two separate leaflets of identical morphology. Concomitant bileaflet prolapse was also present with an eccentric, posteriorly directed regurgitant jet visualized with colour flow Doppler. The normal left atrial dimensions, low density of the spectral Doppler regurgitation envelope, small proportion of the area of the left atrium occupied by regurgitant colour pattern and the normal E-wave velocity of mitral inflow categorized this as mild regurgitation1 (see Supplementary data, Images 1–4). The patient was subsequently referred to a cardiologist for clinical evaluation.

Cleft mitral valve leaflet (CMVL) is an uncommon congenital cause of mitral regurgitation. Clefts are slit-like holes or defects hypothesized to be a result of incomplete expression of an endocardial cushion defect and most commonly involve the anterior mitral valve leaflet with a paediatric incidence of 1:1340. Clefts affecting only the posterior mitral valve leaflet are extremely rare with only four cases being reported in the medical literature. Important co-existing anomalies with either posterior and/or anterior CMVL include counterclockwise rotation of the papillary muscles, the presence of an accessory papillary muscle or mitral valve leaflet, atrial septal defects, and mitral valve prolapse. Regurgitation from CMVL can lead to important physiological and anatomical changes within the cardiac system. Regurgitation results from blood flow directly through the cleft itself or from malcoaptation from accessory chordae with or without papillary muscle distortion. Significant chronic mitral regurgitation elevates left atrial filling pressures and leads to chamber enlargement and eccentric left ventricular hypertrophy. Early detection through two-dimensional echocardiography can provide accurate anatomical images of the various mitral valve structures and identify associated congenital anomalies. Early surgical correction is preferred before mitral regurgitation causes unfavourable remodelling. Most mitral valve cleft defects can easily be repaired by suturing the edges of the cleft. If a cleft resection leads to limited residual valve tissue, the leaflet of the mitral valve can be reconstructed using an autologous pericardial patch pre-treated with buffered glutaraldehyde. Posterior CMVL is an uncommon but clinically important cause of mitral insufficiency. Early recognition of this rare clinical entity and possible co-existent anomalies can identify the patients who would benefit from surgical intervention before compensatory left ventricular remodelling and contractile dysfunction develop.

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CMVL include counterclockwise rotation of the papillary muscles, the presence of an accessory papillary muscle or mitral valve leaflet, atrial septal defects, and mitral valve prolapse.7–10 Acquired causes of clefts include infective endocarditis or trauma from surgical exploration.

Regurgitation in CMVL results from blood flow directly through the cleft itself or from malcoaptation from accessory chordae with or without papillary muscle distortion. Early detection through two-dimensional echocardiography can provide accurate anatomical images of the mitral valve structure and identify associated congenital anomalies.11 Surgical correction is a class I recommendation in symptomatic patients with or without left ventricular dysfunction; and for asymptomatic patients demonstrating a left ventricular ejection fraction <50% or signs of left ventricular dilation.12 Mitral valve clefts may be repaired by suturing the edges of the cleft. If this is not possible due to fibrous tissue on the edges of the cleft, this tissue is resected and mitral valve repair using a pericardial patch can be performed.13

Posterior CMVL is an extremely rare cause of mitral insufficiency. Early recognition of this rare clinical entity and co-existent anomalies can identify afflicted patients who can be closely monitored for the progression of symptoms as well as ventricular dysfunction. Given the asymptomatic status of the patient and a lack of ventricular dysfunction or remodelling, she was treated with conservative management of her hypertension coupled with close clinical follow-up and periodic serial echocardiograms.

**Supplementary data**

Supplementary data are available at *European Journal of Echocardiography* online.

**References**