

was unpredictable on the basis of periprocedural echocardiography but might be most probably related to the mechanical characteristics of this old generation of PFO Star device and the fact that one of the arm of the implant was directly protruding into the aorta.

To our knowledge, only three similar cases of aorto-atrial fistulas following transcatheter closure of PFO or atrial septal defect have been reported so far in the literature.^{1–3} Two case reports refer to fistula between aorta and right or left atrium, respectively, following percutaneous implantation of Amplatzer septal occluder for atrial septal defects. One case of fistula between aorta and right atrium after PFO closure with the PFO Star device has been briefly mentioned. Diagnosis was made 1 month after closure during routine echocardiography and the authors report a spontaneous closure after discontinuation of oral anticoagulation.

Our report of late spontaneous fistula closure after discontinuation of antiagregant therapy in an asymptomatic patient is of paramount interest in

the management of such rare potential complication of percutaneous PFO closure as this procedure is emerging as the treatment of choice for selected patients who suffered cryptogenic stroke due to paradoxical embolism.

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10.1016/j.euje.2005.11.001

Isolated cleft of the anterior mitral valve leaflet

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Received 17 November 2005; received in revised form 25 November 2005; accepted 4 December 2005
Available online 23 January 2006

KEYWORDS

Isolated cleft of the
mitral valve;
Congenital mitral
regurgitation

Abstract Isolated anterior mitral leaflet cleft (not associated with atrio-ventricular septal defect) is a rare cause of congenital mitral regurgitation, and the treatment consists of direct suturing of the cleft. We present a clinical case with this entity.
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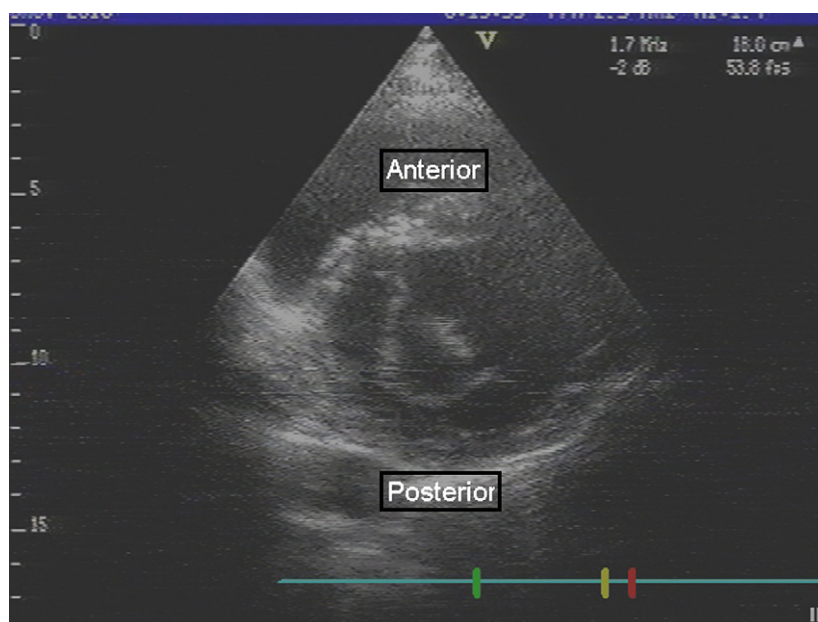


Figure 1 Mitral valve with dysplastic leaflets. The valve has a triangular opening, with the anterior leaflet divided into two (transthoracic echocardiogram, parasternal view, short axis).

Introduction

Isolated mitral cleft is a rare congenital cause of mitral insufficiency.¹ Its association with other cardiac malformations has been previously described.² The clinical manifestations are those of mitral insufficiency. Pre-operative diagnosis is sometimes difficult. Echocardiography shows the cleft and any associated anatomic anomalies.¹

Clinical case

A 32-year old male, with a previous history of surgery to close a patent arterial duct at the age of 9 years was considered in subsequent echocardiograms, to have some degree of mild mitral valve insufficiency due to a probable mitral valve prolapse. He remained stable over the years, and was completely asymptomatic. In 2002, became symptomatic with development of slight dyspnoea and fatigue. The patient improved after starting diuretics. In 2004, there was a new worsening of his clinical condition (NYHA class III). The echocardiogram showed the presence of severe mitral regurgitation and a cleft of the anterior mitral valve leaflet was evident (Fig. 1). There was also some dilatation of the left ventricle (Table 1). Transesophageal echocardiography confirmed those findings (Figs. 2 and 3). The degree of regurgitation was quantified as severe, with systolic inversion of the pulmonary venous flow. There were no other anomalies.

The patient underwent surgery in January 2005, with confirmation of the cleft in the anterior leaflet (Fig. 4). A suture of the entire length of the cleft was performed with a simple, interrupted suture. The intra-operative transesophageal echocardiogram showed very mild mitral insufficiency. At the end of surgery, the patient had a very severe allergic reaction to protamin, with cardiac arrest. The remaining post-operative course was uneventful and he was discharged 15 days after surgery. The echocardiogram performed on discharge showed a left ventricle yet dilated, with moderate systolic dysfunction. There was moderate mitral insufficiency (one central jet and another eccentric, originating in the middle portion of the anterior mitral leaflet). We considered an eventual rupture of the suture

Table 1 Left ventricular dimensions over time

	2002	2004	Discharge	2 months	6 months
LVIDD (mm)	58	65	62	59	57
LVIDD (mm/m ²)	32	36	35	33	32
LVIDS (mm)	30	44	48	43	44
SF (%)	48	32	22	28	24

LVIDD – left ventricular internal diameter in diastole; LVIDS – left ventricular internal diameter in systole; SF – shortening fraction.

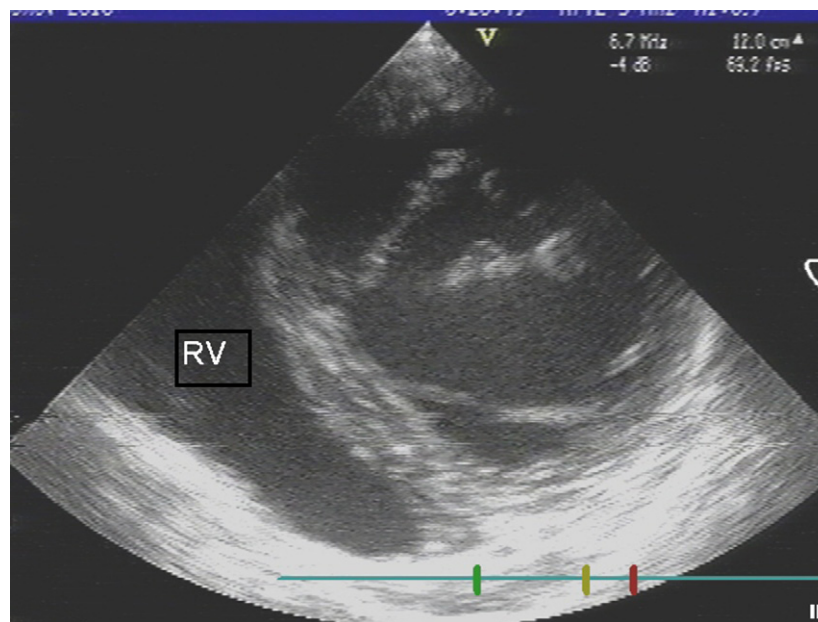


Figure 2 Dysplastic mitral valve, with a triangular opening, with the anterior leaflet divided into two by a cleft (transesophageal echocardiogram, transgastric view, 0 degree).

after surgery, related with the resuscitation performed. Nevertheless, the patient is presently in NYHA class I (12 months after surgery), and is professionally active with no limitations. The 6-month echocardiogram showed recovery of left ventricular dimensions and function, but maintenance of the two regurgitant jets described at discharge.

Discussion

Mitral valve clefts not associated with a septal defect of the endocardial cushion defect type (atrio-ventricular septal defect), also called isolated mitral cleft, is a rare cause of congenital mitral insufficiency,¹ described for the first time in 1954.³ It is defined by the Congenital Heart

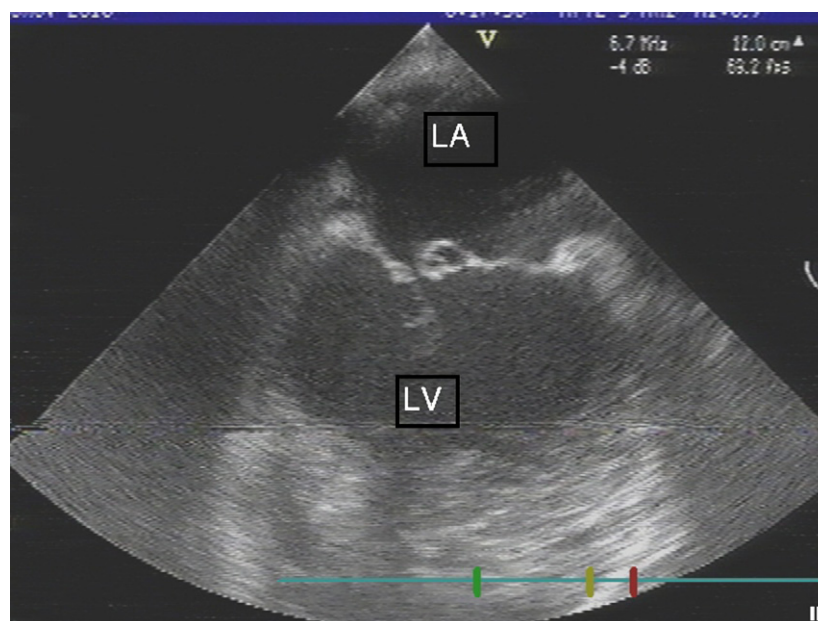


Figure 3 Prolapse of the anterior mitral leaflet that caused severe mitral regurgitation (transesophageal echocardiogram, mid-oesophagus, 90 degrees).

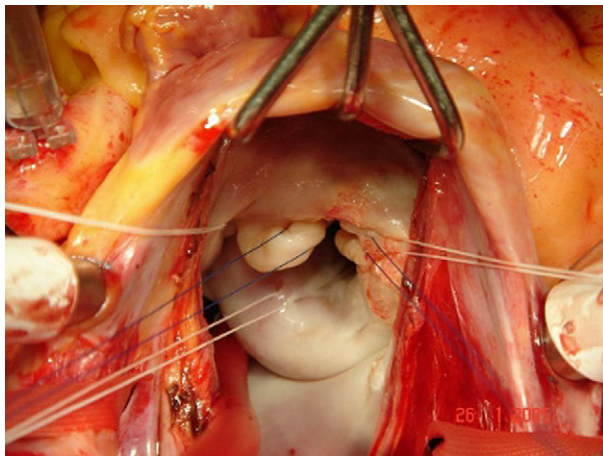


Figure 4 Surgery, with visualization of the anterior leaflet cleft, without significant dilatation of the annulus.

Surgery Nomenclature and Database Project as a "cleft of the anterior mitral valve leaflet not associated with a primum atrial septal defect or other features of atrio-ventricular septal defect (with or without other associated defects)".⁴ An association with other cardiac anomalies such as secundum type atrial septal defect, transposition of the great arteries, ventricular septal defect, tricuspid atresia, patent arterial duct, coarctation of the aorta, double outlet right ventricular and anomalous pulmonary venous connection have been previously described.² In the present patient, there was an association with patent arterial duct.

The usual manifestations are the ones that are found in patients with mitral insufficiency. Pre-operative diagnosis is sometimes difficult, due to the position, dimensions and shape of the cleft. Echocardiography can demonstrate the cleft and the anatomical malformations eventually

associated, such as accessory papillary muscles, papillary muscle displacement, laxity/rupture of chordae and thickening of the edges of the cleft with increasing age. At younger ages, clefts cause mild mitral insufficiency. Perier and Clausnizer³ studied older patients and detected more severe degrees of mitral insufficiency, suggesting some worsening with age, as happened in our patient. In our case, the cleft was not identified in early childhood, and the mitral insufficiency worsened in subsequent years, with need for late cardiac surgery. The technique used was the one described for these cases. Unfortunately, an anaphylactic reaction occurred, with need for cardiac resuscitation measures, which could have compromised in part the success of the intervention, eventually by suture rupture. Nevertheless, 6 months after surgery, the patient resumed his professional activity, is in NYHA class I, and has normalized the left ventricular dimensions, in spite of persistence of residual moderate mitral insufficiency (less significant than at the time of surgery).

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