Idiopathic isolated annular dilatation causing congenital mitral regurgitation

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ABSTRACT

Isolated annular dilatation is an extremely uncommon cause of congenital mitral regurgitation. We report a case of a 5-year-old child with idiopathic isolated annular dilatation causing severe congenital mitral regurgitation.

Keywords: Annular dilatation, congenital mitral regurgitation, isolated annular dilatation

INTRODUCTION

Isolated congenital mitral valve disease is present in less than 0.4% of patients with congenital heart defects.[1] Annular dilatation is a common morphological finding in congenital mitral regurgitation; it is usually secondary to either left ventricular (LV) dilatation caused by some other lesion in the mitral valve or other anomalies like ventricular septal defect causing LV volume overload.[2]

CASE REPORT

A 5-year-old girl presented to our institute with dyspnoea (NYHA class IV), hepatomegaly, and ascites. She did not have any marfanoid features. The chest radiography showed a cardiothoracic ratio of 0.8, left atrial and LV enlargement with dilated pulmonary trunk. Electrocardiogram demonstrated biventricular ventricular hypertrophy and biatrial dilatation. Transthoracic echocardiography revealed severe mitral regurgitation [Figures 1 and 2] due to noncoapting leaflets and severe tricuspid regurgitation with severe LV dysfunction (ejection fraction-30%). The valve leaflets were not myxomatous and there was no leaflet prolapse. She was first diagnosed as having severe

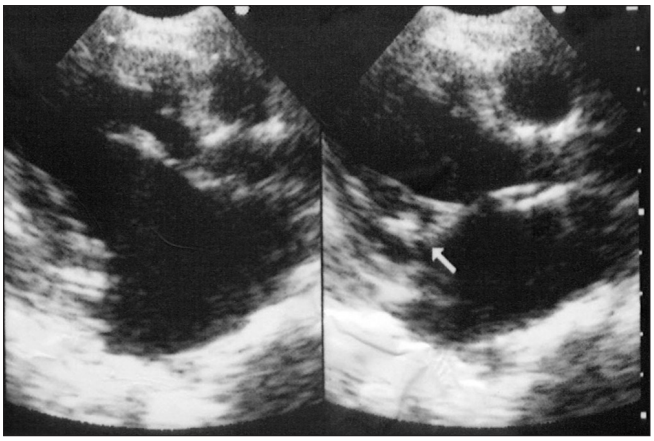

Figure 1: Preoperative parasternal long axis transthoracic echocardiography showing noncoapting mitral leaflets (arrow)

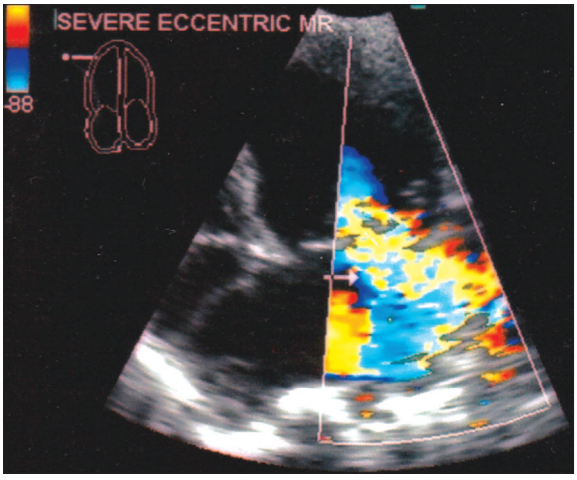

Figure 2: Preoperative apical four chamber transthoracic echocardiography showing severe mitral regurgitation (arrow)

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mitral regurgitation at nine months of age; however, her LV function was preserved at that time. Her functional class and LV function gradually deteriorated since then. During surgery the mitral valve annulus was found to be grossly dilated with normal leaflets and subvalvular apparatus. Mitral valve repair was performed using 26 mm Carpentier–Edward classic mitral annuloplasty ring (Edwards Lifesciences, Irvine, CA, USA) based on the size of her anterior mitral leaflet. Tricuspid repair was performed using modified De Vega's annuloplasty. Postrepair transesophageal echocardiography revealed trivial mitral regurgitation and trivial tricuspid regurgitation. She was transferred out of the intensive care unit on eighth postoperative day and discharged after 16 days of surgery. She is doing well 1 year postrepair, with echocardigraphy [Figure 3] showing mild mitral regurgitation and mild tricuspid regurgitation with a sustained LV function of 30% (NYHA class I) with improved symptomatic status.

DISCUSSION

Congenital malformations of the mitral valve are relatively rare and present with a wide spectrum of morphologic abnormalities and a high rate of concomitant cardiac anomalies. Isolated congenital mitral valve disease is rare.[2,3]

Most of the large series on congenital mitral regurgitation have reported annular dilation as a lesion contributing to mitral regurgitation; however, it is still unclear whether an entity like isolated annular dilation exists [Table 1].[4-6]

Carpentier and colleagues[2] reported isolated annular dilatation in 8 (17%) of 47 cases with congenital mitral valve disease; however, some deficiencies of commissural tissue were implicated by their description. Four of these patients had asymmetrical dilatation of the annulus involving only the posterior leaflet and commissural area. This form of annular dilatation is commonly seen with ventricular dilatation. Four patients had symmetrical dilatation hinting toward its primary nature.

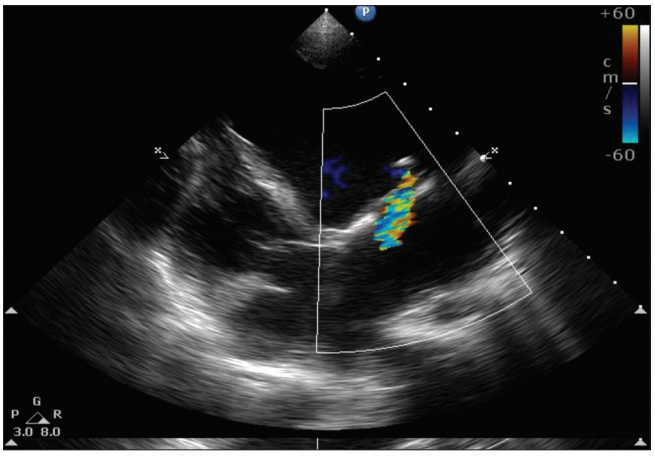

Figure 3: Postoperative apical four chamber transthoracic echocardiography after 1 years howing mild mitral regurgitation

Chauvaud *et al.*[4] reported congenital mitral regurgitation in 145 patients. They described annular dilatation in seven (4.8%) patients with normal leaflet motion. The dilatation mainly affected the posterior annulus and was associated with minor anomalies such as imperforate interchordal spaces.

Uva *et al.*[7] described ten patients with congenital mitral regurgitation. Annular dilatation was a constant feature in all patients but isolated annular dilatation was seen in only three patients.

Coles *et al.*[8] reported their experience with surgical management of congenital mitral valve disease. They described 22 patients with congenital mitral regurgitation. Sixteen had annular dilatation of which seven were isolated annular dilatations.

McCarty *et al.*[9] reported their 10-year experience of surgical management in congenital mitral valve disease. Thirteen patients had congenital mitral regurgitation, all of them due to annular dilatation but none of them was isolated. There was either a primary leaflet pathology or LV dilatation.

Oppido *et al.*[6] described 60 patients with predominant congenital mitral regurgitation, of which 24 patients had nondysplastic and normal leaflets with annular dilatation. However, annular dilatation in these patients was secondary to LV dilatation due volume overloading by a large left to right shunt. Thus, there were no cases of isolated annular dilatation in their series.

The embryological origin of idiopathic isolated annular dilatation is still unknown. The surgical management involves annuloplasty and a durable repair is usually expected.[4-6]

Our case underscores the importance of recognizing isolated idiopathic annular dilatation as a cause of congenital mitral regurgitation.

Table 1: Studies on congenital mitral regurgitation with annular dilatation as a cause

Authors	Study period	No. of cases of congenital MR	No. of cases of annular dilatation	Isolated annular dilatation
Carpentier *et al.*	1970–1976	47	14	8
Chauvaud *et al.*	1970–1995	145	7	0
Uva *et al.*	1980–1993	10	10	3
Coles *et al.*	1962–1986	22	16	7
McCarthy *et al.*	1983–1994	13	13	0
Oppido *et al.*	1996–2006	60	25	0

MR: Mitral regurgitation

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