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Successful Percutaneous Balloon Dilatation of Membranous Sub-aortic Stenosis in a 22 Year Old Pregnant Patient

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Authors' contributions

This work was carried out in collaboration among all authors. Author GS designed the manuscript, provided the clinical finding, the laboratory findings and images for the study and wrote the first draft of the study. Authors MB, AJ and AG managed the literature searches. Authors AB and NOB revised the manuscript. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Congenital obstruction of the left ventricular outflow tract comprises a heterogeneous group of disorders, with obstruction potentially occurring below, above, or at the level of the aortic valve. Subvalvular stenosis is the second most common type of left ventricular outflow tract obstruction, of which discrete membranous type is the most common. Although surgical resection of the subaortic membrane is the treatment of choice in discrete membranous subaortic stenosis, in selected patients with isolated membranous subaortic stenosis, without significant aortic insufficiency, percutaneous balloon tearing of the membrane results in reduction in the degree of left ventricular outflow tract obstruction and symptomatic relief. We report a case of 22 year old pregnant patient admitted with NYHA class III breathlessness, found to have discrete membranous subaortic stenosis. Balloon aortic valvuloplasty was performed in the patient with good result. Patient underwent normal vaginal delivery at 38 weeks. Both mother and newborn were asymptomatic. Patient is asymptomatic on subsequent follow-ups.

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Keywords: Left ventricular outflow tract obstruction; subaortic stenosis; balloon aortic valvuloplasty.

ABBREVIATIONS

LVOT : Left ventricular Outflow Tract
BAV : Balloon Aortic Valvuloplasty
NYHA : New York heart Association

1. INTRODUCTION

Congenital obstruction of the left ventricular outflow tract (LVOT) comprises a heterogeneous group of disorders, with obstruction potentially occurring below, above, or at the level of the aortic valve. Each of these scenarios represents a distinct disease process with unique ontogeny and natural history. Valvar aortic stenosis constitutes the most common type of congenital LVOT obstruction, accounting for approximately 70 to 85% of cases [1,2]. The second most common type is fixed subvalvar stenosis, which accounts for approximately 15% of total cases and the supravalvar stenosis, is the least common type [3]. Balloon aortic valvuloplasty (BAV) is typically reserved for patients with valvar aortic stenosis, although there is evidence that thin, discrete subaortic membranes may be effectively treated with balloon valvuloplasty as well. In most patients, BAV leads to tearing of the membrane which results in a drastic reduction in the degree of LVOT obstruction with little or no increase in the degree of aortic insufficiency.

2. CASE REPORT

22 year old female, primigravida, with 26 weeks pregnancy was admitted in our hospital with NYHA class III breathlessness since 20 days. There was no significant past history of any illness. On examination, pulse was 92/ minute, blood pressure was 100/70 mm Hg. On Cardiovascular examination, apex beat was forceful and sustained, systolic thrill was noted in left 3rd /4th intercostal space. A harsh, grade IV/VI, ejection systolic murmur was heard in left 3rd/4th Intercostal space near sternum. Electro-cardiogram showed left ventricular hypertrophy. Echocardiography showed turbulence in Left ventricular outflow tract, with a fibrous membrane clearly seen below the tricuspid aortic valve (Fig. 1A, 1B, 1C). The peak and mean gradient was 88/56 mmHg, with mild aortic regurgitation. Membrane was confirmed on transesophageal echocardiography (Fig. 2A, 2B). Routine blood investigations were normal. The treatment options were explained to the husband and parents.

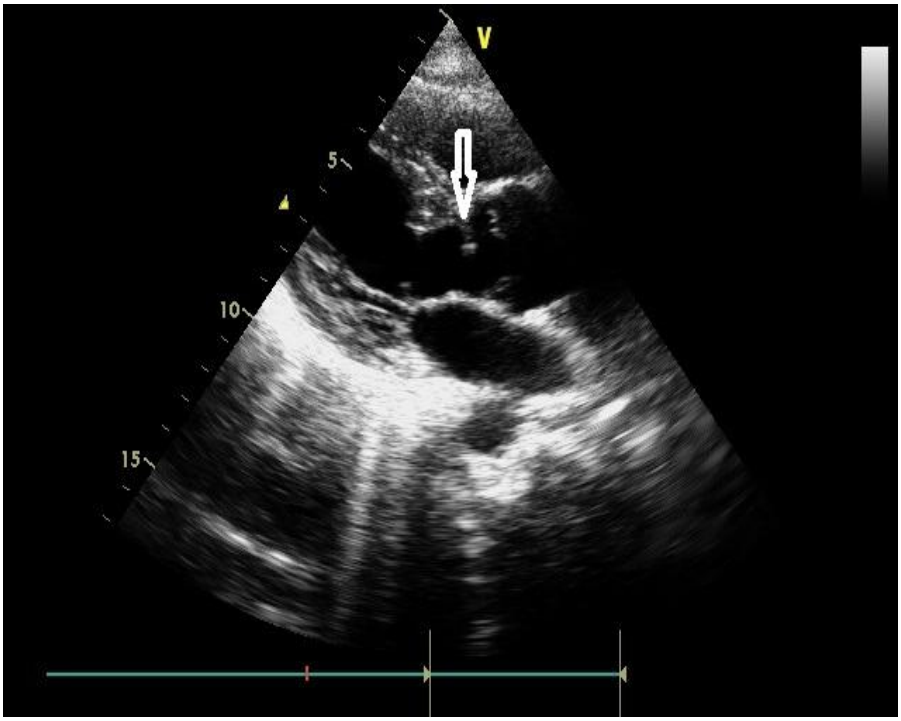

Fig. 1A. Echo – parasternal long axis view showing membrane just below the aortic valve

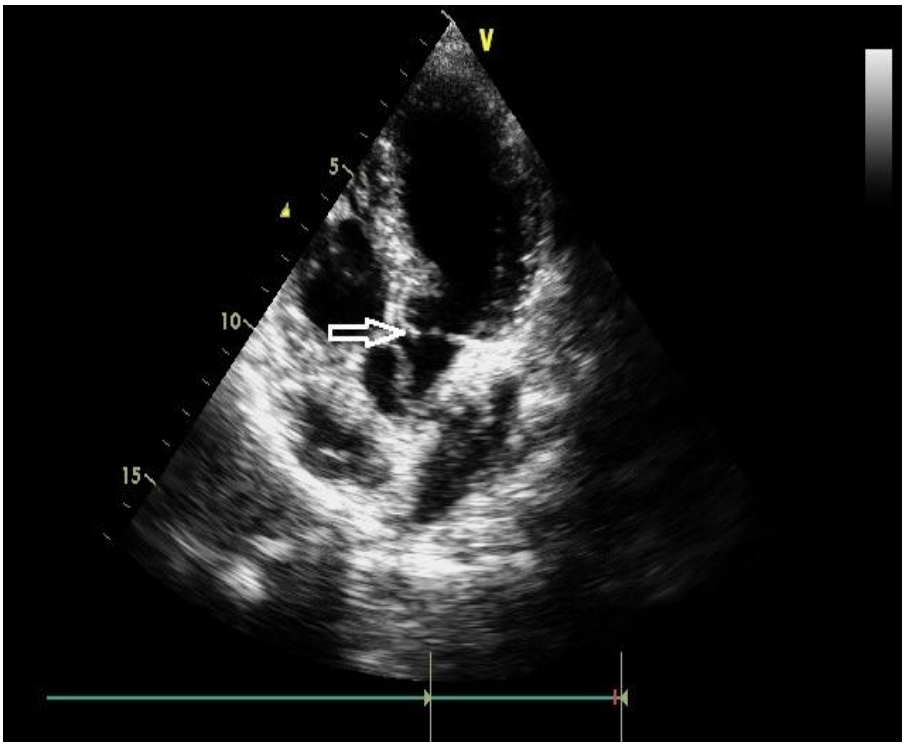

Fig. 1B. Echo- apical five chamber view showing membrane just below the aortic valve

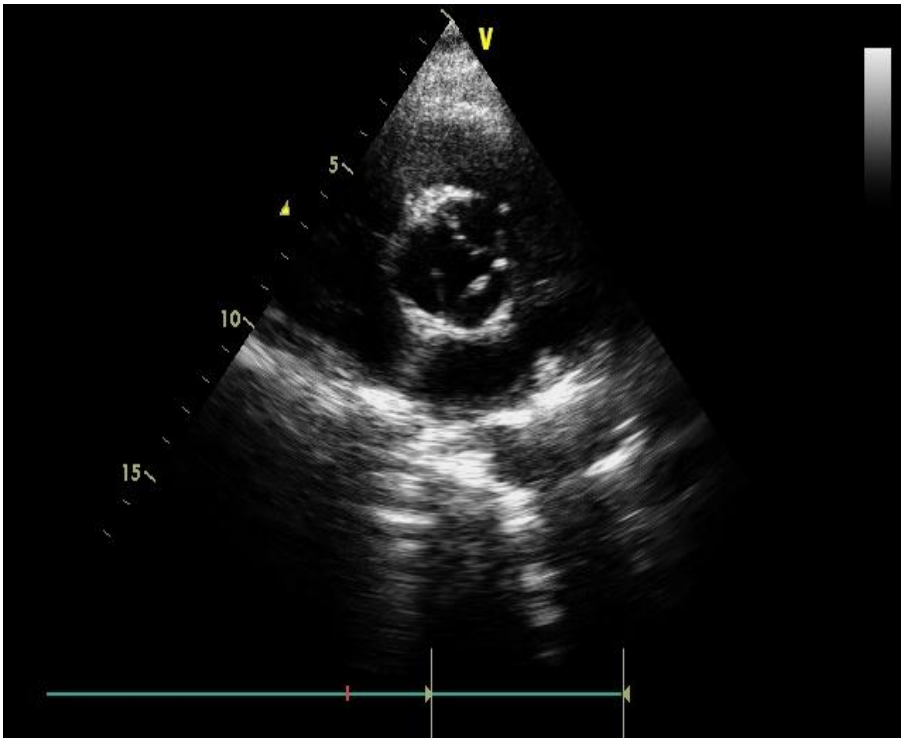

Fig. 1C. Parasternal short axis view showing tricuspid normal aortic valve

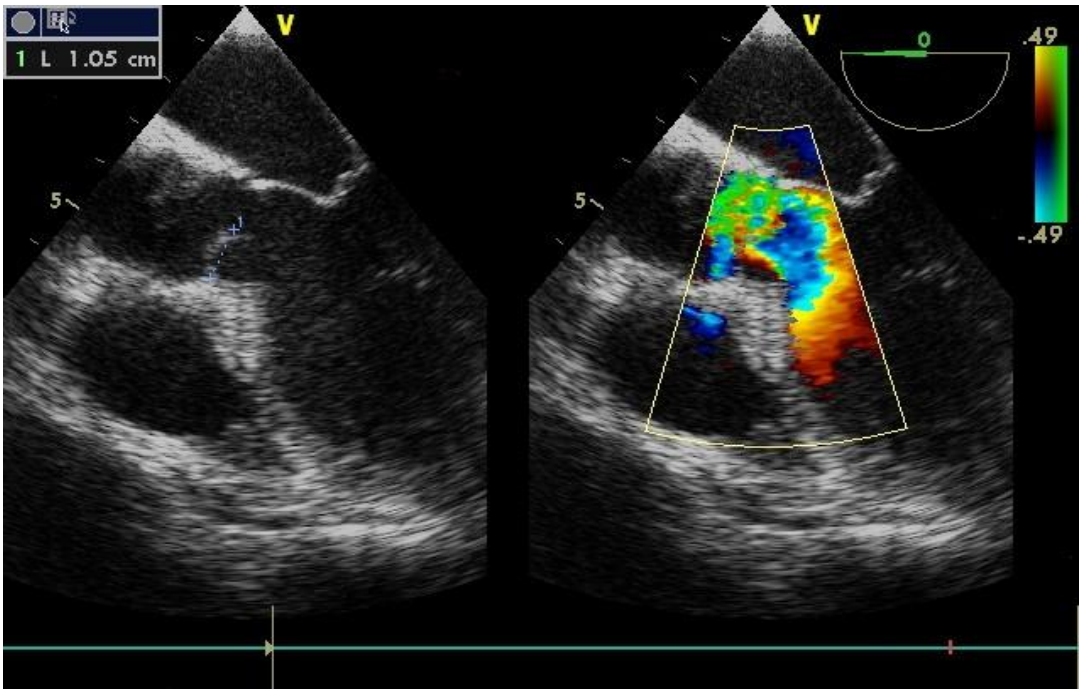

Fig. 2A. TEE showing fibrous membrane in LVOT (0° view)

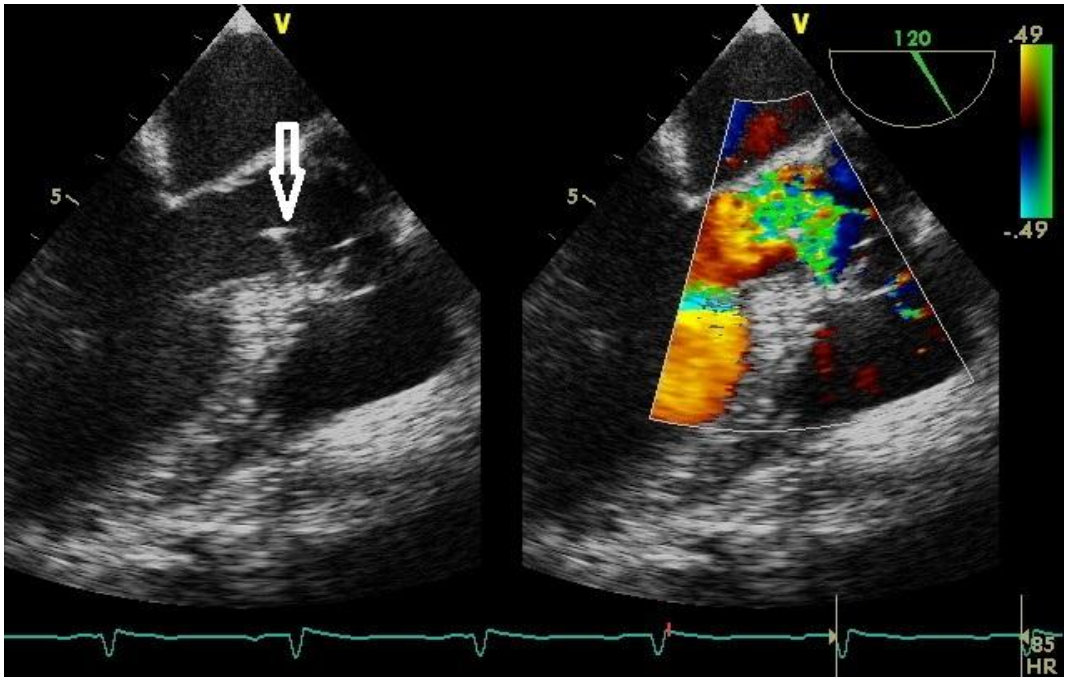

Fig. 2B. TEE showing fibrous membrane in LVOT just below the aortic valve and turbulence in LVOT across the membrane on colour Doppler (120° view)

Percutaneous balloon dilatation of the membrane was planned. Right femoral venous and arterial access was obtained. Cardiac catheterization showed PCWP- 22 bmmHg, LV systolic pressure- 316 mmHg, Aortic pressure- 92/72 mmHg, with peak gradient of – 124 mmHg (Fig. 3). The position of the membrane was confirmed on left ventriculogram (Fig. 4). BALTON 23mm X

50 mm balloon was used based on the measurement of aortic annulus (Fig. 5A, 5B). After inflation, LV systolic pressure reduced to 125 mm Hg (Fig 6A) and systolic aortic pressure was 106 mmHg (Fig 6B) with gradient reduced to 19 mmHg from 124 mmHg. The procedure was uneventful. The patient was discharged after 2 days. There was a normal vaginal delivery at 38 weeks. Both mother and newborn were asymptomatic. Patient is asymptomatic on subsequent follow-ups.

Fig. 3. LV pressure (Systolic- 316 mmhg, end diastolic pressure- 18 mmHg) aortic pressure (92/72 mmHg) peak gradient of 124 mmHg

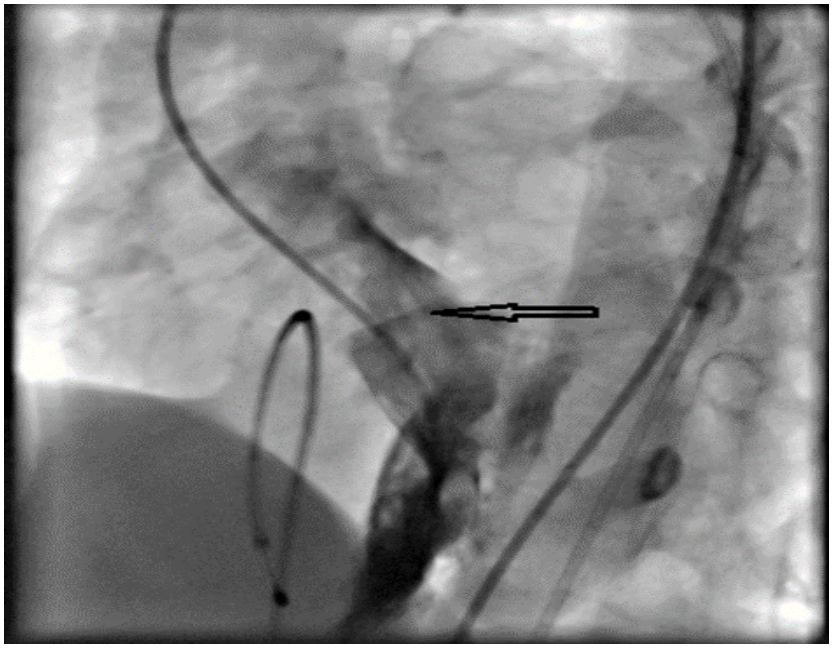

Fig. 4. LV angiogram in LAO cranial view showing membrane just below aortic valve (marked by an arrow)

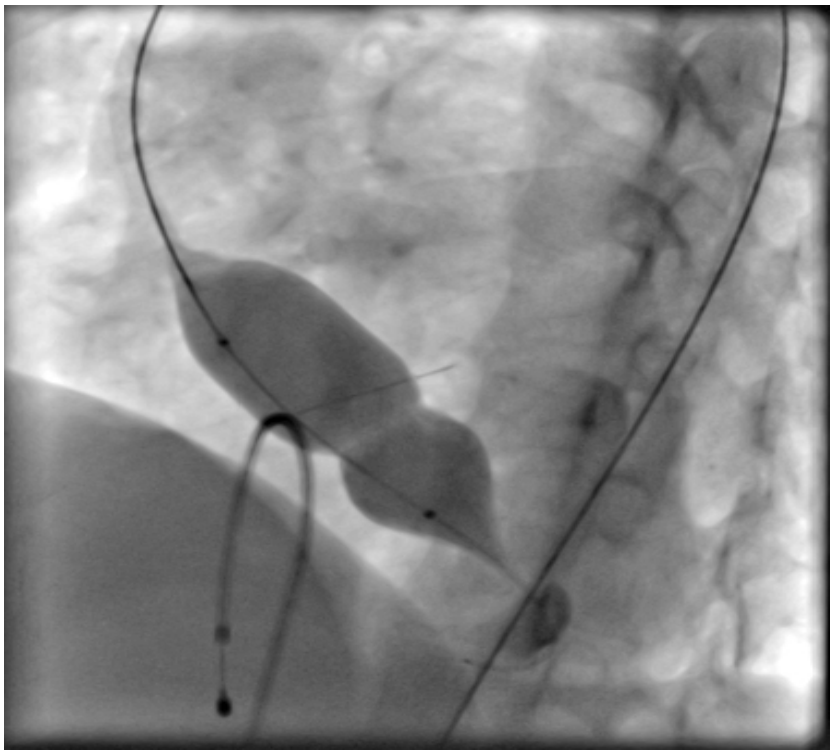

Fig. 5A. Balloon dilatation across the membrane showing the waist during initial part of inflation

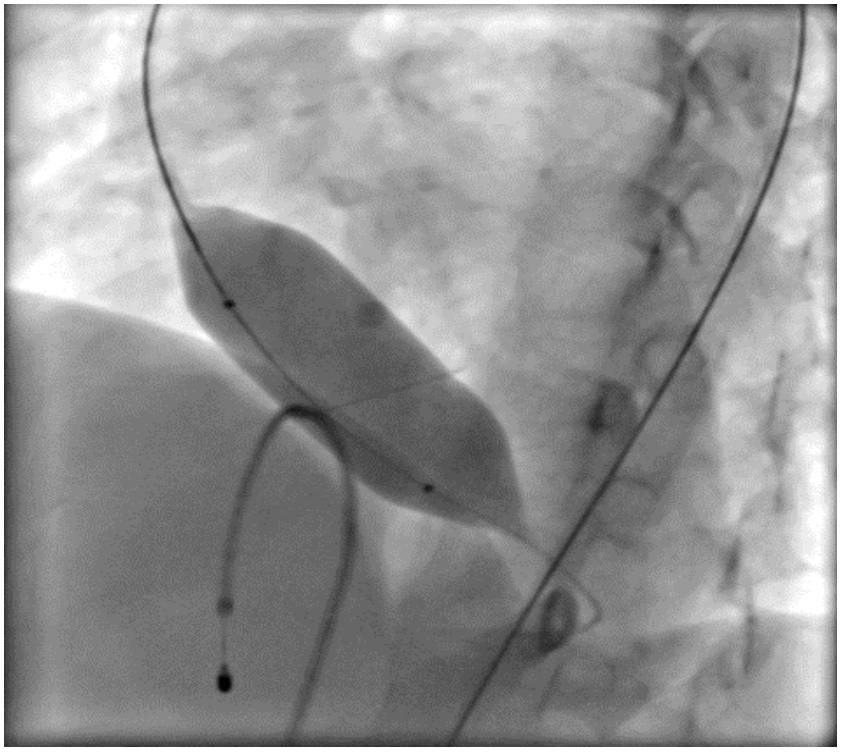

Fig. 5B. Full dilatation of the balloon

Fig. 6A. LV pressure after balloon dilatation (systolic- 125 mmHg, end diastolic- 20 mmHg)

Fig. 6B. Aortic pressure after balloon dilatation (106/74 mmHg) with final gradient of 19 mmHg

3. DISCUSSION

The most common form of subvalvular aortic stenosis is the membranous subaortic stenosis, characterized by a thin, fibrous membrane just proximal to the aortic valve, accounting for 70% to 80% of cases [4]. The membrane is generally very thin (1 to 2 mm), located within several

millimeters of the aortic valve, and is often circumferential, including attachments to the anterior leaflet of the mitral valve. The second most common type of subaortic stenosis involves a fibromuscular ridge, thicker than the membrane and frequently located slightly more inferior to the aortic valve [4,5]. Both of these types of discrete subaortic stenosis are rarely present in infancy and are often considered acquired conditions. On the most severe end of the subaortic stenosis disease spectrum is “tunnel-type” obstruction, characterized by muscular hypertrophy and narrowing of the left ventricular outflow tract that extends for several centimeters below the valve [7,8]. While aortic valve morphology is most often normal, a bicuspid aortic valve is identified in 10% to 25% of patients [4,9]. Aortic insufficiency develops in up to 70% of patients over time [3]. Most common additional cardiac malformations is a ventricular septal defect, found in 10% to 48% of patients [9,10] followed by coarctation of the aorta, which is present in 6% to 20% of patients [5,6].

Subvalvar stenosis is generally a progressive disease, although the rate of progression can be highly variable. Echocardiography is most important in the diagnostic evaluation of suspected aortic stenosis. Cardiac catheterization has largely been replaced by echocardiography for the diagnosis of aortic stenosis, but there is still a prominent role for catheterization and balloon aortic valvuloplasty in the treatment of patients with aortic stenosis. For patients with severe stenosis who do become pregnant, intervention on the aortic valve during pregnancy is recommended if there is hemodynamic deterioration or the development of NYHA class III or IV heart failure symptoms [11].

Management of patients with membranous subaortic stenosis is surgical which involves relatively straight forward membrane resection, a low-risk procedure with early mortality between 1% and 2% [12,13]. Balloon aortic valvuloplasty is typically reserved for patients with valvar aortic stenosis, although there is evidence that thin, discrete subaortic membranes may be effectively treated with balloon angioplasty as well [14]. In most patients, balloon tearing of the membrane results in a drastic reduction in the degree of left ventricular outflow tract obstruction with little or no increase in the degree of aortic insufficiency [14]. Most patients (77%) had sustained relief at subsequent follow-ups without restenosis, the need for surgery, progression to muscular obstructive disease, or an increase in the degree of aortic regurgitation over the period of 25 years.

4. CONCLUSION

Although surgical resection of the subaortic membrane is the treatment of choice in discrete membranous subaortic stenosis, in selected patients with isolated membranous subaortic stenosis, without significant aortic insufficiency, percutaneous balloon tearing of the membrane results in reduction in the degree of LVOT obstruction and symptomatic relief.

CONSENT

Written informed consent was obtained from the patient for publication of this report and any accompanying images.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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