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Long-Term Outcome of Patients With Isolated Thin Discrete Subaortic Stenosis Treated by Balloon Dilation

A 25-Year Study

José Suárez de Lezo, MD, PhD; Miguel Romero, MD, PhD; José Segura, MD, PhD; Manuel Pan, MD, PhD; Javier Suárez de Lezo, MD, PhD; Djordje Pavlovic, MD, PhD; Francisco Mazuelos, MD, PhD; Mónica Delgado, MD, PhD; Dolores Mesa, MD, PhD

Background—Transluminal balloon tearing of the membrane in a thin discrete subaortic stenosis is an alternative to membrane surgical resection. However, the long-term outcome of patients with isolated thin discrete subaortic stenosis treated by transluminal balloon tearing remains unknown.

Methods and Results—This 25-year study describes findings from 76 patients with isolated thin discrete subaortic stenosis who underwent percutaneous transluminal balloon tearing of the membrane and were followed up for a mean period of 16 ± 6 years. The age at presentation had a wide range (2–67 years). The mean age at treatment was 19 ± 16 years. Immediately after treatment, the subvalvular gradient decreased from 70 ± 27 to 18 ± 12 mm Hg ($P<0.001$). No significant postprocedural aortic regurgitation was observed. After a mean follow-up time of 16 ± 6 years, 11 patients (15%) developed restenosis, 3 patients (4%) progressed to muscular obstructive disease, and 1 patient (1.3%) developed a new distant obstructive membrane. Twelve patients (16%) were redilated at a mean of 5 ± 3 years after their first treatment, and 4 patients (5%) underwent surgery at a mean of 3 ± 2 years after their first treatment. Fifty-eight patients (77%) remained alive and free of redilation or surgery at follow-up. Larger annulus diameter and thinner membranes were independent factors associated with better long-term results.

Conclusions—Most patients (77%) with isolated thin discrete subaortic stenosis treated with transluminal balloon tearing of the membrane 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. (*Circulation*. 2011;124:1461-1468.)

Key Words: balloon ■ discrete subaortic stenosis ■ follow-up studies

Balloon tearing of a fixed subaortic thin membrane is an effective and safe method for reducing subaortic obstruction in patients with discrete subaortic stenosis (DSS). Since 1985,^{1,2} several studies have confirmed that persistent pressure relief is achieved after balloon dilation.^{3–13} Tearing of the membrane by the balloon provided a mean gradient reduction of 70% in 11 series of patients without resultant aortic or mitral valve dysfunction. However, given the progressive nature of the disease, the long-term efficacy of this treatment remains unknown. At present, only a few follow-up studies after membrane tearing are available.^{6,7,9}

Clinical Perspective on p 1468

The purpose of this retrospective 25-year study was to describe the findings obtained from 76 patients with isolated thin DSS who underwent percutaneous transluminal balloon tearing (TBT) of the membrane and were followed up for a mean period of 16 ± 6 years.

Methods

Study Patients

We retrospectively studied 76 consecutive patients with isolated thin DSS who were treated by balloon dilation and followed up for a mean period of 16 ± 6 years (range, 0.5–25 years) at our institution from January 1985 to June 2010. Table 1 shows the main baseline data from this group of patients. The age at presentation ranged from 2 to 67 years (mean, 19 ± 16 years); 39 patients were children, 17 patients were adolescents, and 20 patients were adults. Noninvasive and invasive studies were carried out in all candidates. The inclusion criteria for the study restricted the study to only patients with a diagnosis of isolated DSS who had a thin (<3 mm) subaortic membrane, crescent or circumferential, from the base to the orifice in the outflow tract and a significant subaortic gradient or symptoms (dyspnea or effort angina) or ECG signs of left ventricular strain. Fifty-six patients had a subvalvular gradient >50 mm Hg (mean, 78 ± 23 mm Hg). Twenty patients with baseline gradient <50 mm Hg (mean, 44 ± 4 mm Hg) had symptoms (dyspnea or effort angina) or signs of left ventricular strain. Patients having any type of fibromuscular component, including those patients with a thin

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From the Instituto Maimónides de Investigación Biomédica de Córdoba (José S.d.L.) and Department of Cardiology, University Hospital “Reina Sofía,” (M.R., J.S., M.P., Javier S.d.L., D.P., F.M., M.D., D.M.), Córdoba, Spain.

Correspondence to José Suárez de Lezo, MD, PhD, Department of Cardiology, University Hospital “Reina Sofía,” Avda Menéndez Pidal s/n, 14004-Córdoba, Spain. E-mail grupo_corpal@arrakis.es

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Table 1. Baseline Data (n=76)

Clinical	
Age, y	19±16
Median (25th–75th percentiles)	14 (9–19)
Body surface area, m ²	1.38±0.44
Median (25th–75th percentiles)	1.43 (0.96–1.75)
Sex, n (%)	
Male	50 (66)
Female	26 (34)
Echocardiographic Doppler gradient, mm Hg	74±22
Angiographic and procedural	
Peak systolic ventricular pressure, mm Hg	169±33
Degree of aortic insufficiency, n (%)	
0	25 (33)
I	48 (63)
II	3 (4)
Mitral regurgitation	0
Left ventricle-to-aorta gradient, mm Hg	70±27
Valve-to-membrane distance, mm	8.6±5.4
Annulus size, mm	19±5
Membrane thickness, mm	1.97±0.8
Largest balloon diameter, mm	21±6
Annulus/balloon ratio	0.93±0.11

Data are expressed as mean±SD when appropriate.

membrane but a fibromuscular base in the outflow tract, were excluded. These forms of the disease, which are almost always collar type, have shown poor results^{3,8} and were not attempted for catheter intervention. We also excluded patients with any degree of aortic valve gradient and those with a thin membrane on the outflow tract that becomes markedly reduced in systole, contributing in some way to the stenosis. This type of fibromuscular stenosis may potentially cause dynamic obstruction, and patients with these characteristics are not candidates for an interventional procedure.

Echocardiographic Studies

Echocardiographic techniques have improved significantly during the last 25 years. Consequently, the image quality and assessment of patients for selection, evaluation of results, and follow-up have also favorably evolved through years. The left ventricular outflow tract was always carefully inspected, and the relationship of the membrane to other structures such as the aortic or mitral apparatus was established. Proximity of the membrane to the aortic annulus could favor the extension of fibrous tissue onto the aortic valve, but this extension was quite difficult to assess. In terms of the mitral valve, most circumferential membranes may have attachments to the fibrous tissue continuity between the aortic and mitral valves. Possible attachments of the membrane to the free border of the anterior mitral leaflet were also inspected. Echocardiographic measurements included the peak instantaneous Doppler gradient. Continuous-wave Doppler was performed by multiple acoustic windows (apical, suprasternal, right parasternal, or supraclavicular) to determine the highest velocity. This assessment was available in our center starting in 1989. Thus, 21 patients treated between 1985 and 1989 had only M-mode and 2-dimensional echocardiographic assessment without Doppler gradients before and after treatment. However, all treated patients had serial Doppler gradient assessment at follow-up. The aortic annulus diameter, membrane thickness, and distance from the aortic annulus to the membrane were also measured by transthoracic or transesophageal echocardiogram. Measurements were made in diastole on the long-axis aortic view. The

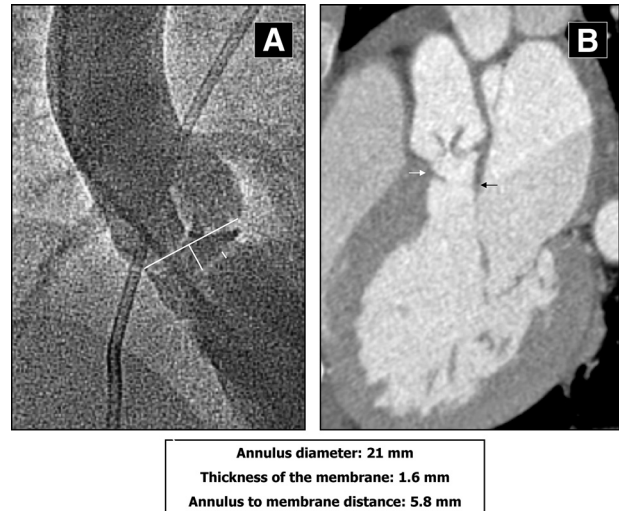


Figure 1. Angiographic (A) and 64-slice computed tomographic (B) diastolic images of the left ventricular outflow tract in a patient with a circumferential membrane. Measurements were obtained by angiography as depicted.

aortic annulus diameter was the distance between the insertion of the noncoronary leaflet on the aortic wall to the junction of the right coronary leaflet with the anterior aortic wall. The degree of the aortic valve incompetence before and after treatment was evaluated in 55 patients. The same assessment was performed serially in all successfully treated patients at follow-up. The degree of aortic regurgitation was determined by the depth and width of the jet with color Doppler in multiple acoustic windows. It was graded as 0 (no) to I to IV degrees of aortic regurgitation. All parameters were obtained by consensus of 2 expert echocardiographers according to standard published guidelines.

Hemodynamic and Angiographic Studies

These studies were always the last evaluation before proceeding with an intervention and were repeated immediately after treatment. The studies included simultaneous right and left ventricular pressure measurements, subvalvular gradient recordings, and cardiac output determination. Angiographic measurements of the aortic annulus, valve-to-membrane distance, thickness of the membrane, and left ventricular volumes and function were obtained from 2 left ventricular angiograms (30° right anterior oblique and 45° left anterior oblique with 25° cranial angulations). All measurements were made in diastole, as shown in Figure 1. The distance between the midpoint of the aortic annulus and the center of the membrane was considered the valve-to-membrane distance. The angiographic thickness of the membrane was also obtained in diastole. However, in 5 patients with a very thin membrane (<1 mm) that was not well visualized in a fixed angiogram, we obtained this quantitative parameter by echocardiography. A 60° left anterior oblique or lateral aortogram was always performed to evaluate the aortic valve function. The degree of aortic regurgitation was evaluated by the classic method of Sellers et al.¹⁴ All quantitative parameters were obtained by consensus of 2 expert angiographers.

Therapeutic Phase

The treatment technique has evolved significantly in this 25-year period. The therapeutic target was always the rupture of the fibrous membrane without any subsequent damage to any valve apparatus. A significant advancement has been new balloon sizes, which optimize the achievement of clinical objectives. The balloon must be quickly and fully inflated for a diameter size that is 1 to 2 mm larger than the aortic annulus in the left ventricular outflow tract. If the balloon remains stable during inflation, despite the continuously beating heart, and a notch in the balloon disappears at full inflation, then the

membrane has been torn. The procedure is most likely completed, and no further inflations are needed. However, maintaining a stably inflated balloon during potent left ventricular contractions can be challenging. Therefore, 2 methods were developed to facilitate balloon stability during inflation. One method was to simultaneously administer 6 to 12 mg adenosine through the pulmonary catheter with the balloon catheter in place to obtain a transient cardiac arrest or block during fast balloon inflation in the left ventricular outflow tract. The second method was to pace the right ventricle at a high rate (180 to 200 bpm) during balloon inflation. This condition always generates a transient low cardiac output during balloon inflation and allows stable and full balloon expansion. After treatment, a new hemodynamic and angiographic evaluation was performed under the same conditions as the baseline study.

Complications of the puncture site may create problems. However, its management has also favorably evolved with time, significantly decreasing puncture-site complications. This has resulted from using sheaths and developing percutaneous methods to solve problems when they appear. Now, we always introduce and retrieve a large balloon through a long valve cannula. Using the Perclose Prostar system for the final suture of the artery hole has also significantly decreased the frequency of groin complications.

Follow-Up Studies

Predischarge echocardiographic evaluation was always performed. In addition to repeating echocardiographic measurements under conditions similar to pretreatment measurements, we focused on the remaining membrane in the left ventricular outflow tract. The residual Doppler gradient was obtained, and aortic valve competence was evaluated. Both parameters were valuable during subsequent follow-up studies.

Patients were followed up closely by telephone calls and outpatient clinical evaluations, which were performed at 6 months, 1 year, and every subsequent year. Serial transthoracic echocardiographic studies were performed during follow-up visits. In 31 patients, a new hemodynamic and angiographic evaluation was performed at a mean of 4 ± 3 years after initial intervention. Recurrence was defined either as a late residual gradient >40 mm Hg or a loss of $\geq 50\%$ of the initial gain. When these findings were obtained by echocardiography, they were always confirmed by cardiac catheterization. The evolution of aortic valve function was also evaluated at long-term follow-ups by echocardiography. Factors associated with long-term results were investigated. Clinical, echocardiographic, and angiographic parameters were analyzed to determine the cumulative 25-year recurrence-free probability after TBT.

Statistical Analysis

Quantitative data are expressed as mean \pm SD. The paired *t* test was used to compare 2 mean values. Event-free probability curves were constructed by the Kaplan-Meier method. To identify the factors that influence long-term results, we calculated the 25-year recurrence-free probability in the different groups (Table 2). The log-rank test was used to determine the statistical significance of the differences in cumulative recurrence rate. All variables identified as significant ($P < 0.05$) or marginally significant in the univariate analysis were considered in a Cox proportional hazards regression analysis (SPSS 17.0). Independent predictors of late recurrence were identified.

Results

Immediate Results

After balloon dilatation, the left ventricular-to-aortic gradient significantly decreased, from 70 ± 27 to 18 ± 12 mm Hg ($P < 0.001$). In 6 patients (8%), the gradient disappeared completely, and in 21 patients (28%), the immediate residual gradient was <10 mm Hg. The mean percent gradient reduction was $73 \pm 16\%$. In 54 patients with echocardiographic Doppler assessment, we observed significant correlations between peak hemodynamic and peak instantaneous Doppler

Table 2. Factors That Influence Recurrence After Treatment

	20-Year Recurrence-Free Probability, %*	<i>P</i>
Age at treatment, y		
>13 (n=39)	69 \pm 20	0.002
≤ 13 (n=34)	42 \pm 11	
Body surface area, m ²		
≥ 1.3 (n=42)	67 \pm 20	0.009
< 1.3 (n=31)	43 \pm 11	
Aortic annulus, mm		
≥ 20 (n=38)	93 \pm 1	0.001
< 20 (n=35)	35 \pm 11	
Valve-to-membrane distance, mm		
≥ 9 (n=34)	78 \pm 15	0.001
< 9 (n=39)	37 \pm 12	
Membrane thickness, mm		
< 2.5 (n=55)	66 \pm 12	0.007
≥ 2.5 (n=18)	19 \pm 16	

P values are from the log-rank test. No events were observed beyond the chosen time point.

*Kaplan-Meier method, presented as mean \pm SE.

gradient both before ($r=0.73$, $P < 0.001$) and after ($r=0.72$, $P < 0.001$) treatment. Angiographically, no significant changes were observed in the degree of aortic regurgitation; in 6 patients (8%), regurgitation disappeared immediately after TBT of the membrane. Mild mitral regurgitation induced by balloon dilatation of the membrane was observed in 1 patient (1.3%). In this particular patient, there was echocardiographic evidence of attachments between the free border of the anterior mitral leaflet and the fibrous tissue of the membrane. The echocardiogram after balloon dilatation showed integrity of the chords but with mild regurgitation through a cleft-like small tearing of the anterior mitral leaflet. After TBT of the fibrous diaphragm of the membrane, a fluttering and widely mobile remaining structure was clearly visualized on angiography in the outflow tract, which was confirmed by echocardiography (Figure 2). At the base of the membrane, a small remnant was sometimes observed as a fixed structure. However, when the membrane was very thin (<1 mm), no significant residual structure was seen in the outflow tract. Improvements in aortic valve opening were also frequently noted, and a higher degree of pressure relief was associated with a larger annulus diameter and a longer valve membrane distance.⁹

One patient died after emergency surgery for wall perforation during dilatation maneuvers (1.3%), and another patient needed surgical repair at the access site. However, no other major complications occurred. Some degree of myocardial contusion was observed in 17 patients, as represented by transient A-V nodal rhythm in 1 patient and transient left bundle-branch block in 16 patients. Five patients had a decreased femoral pulse at the puncture site without ischemic compromise; 8 patients had blood loss during the procedure that required blood transfusion. All patients without complications during the 88 therapeutic procedures (first treatment

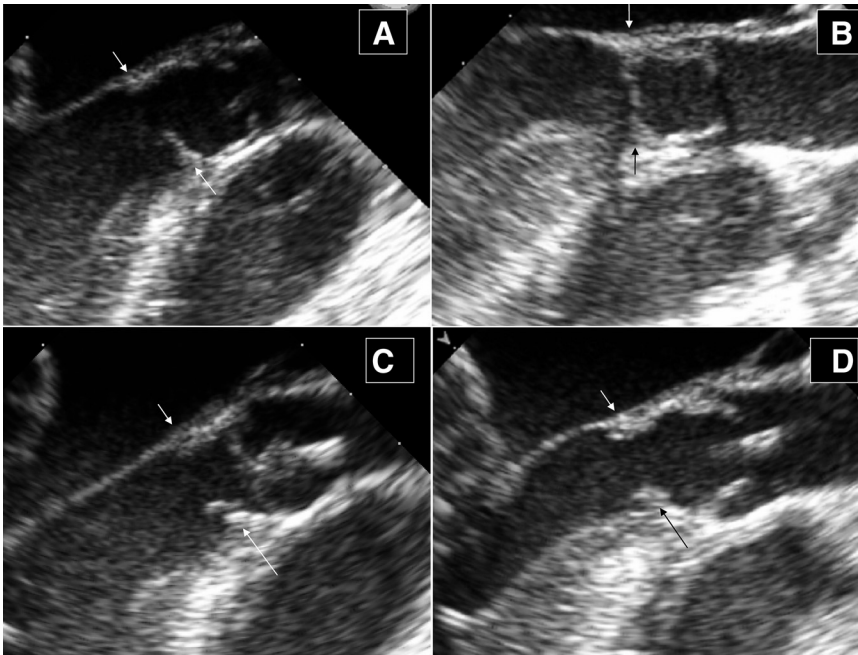


Figure 2. Diastolic (A and C) and systolic (B and D) transesophageal echocardiographic images of a patient with discrete subaortic stenosis before (A and B) and immediately after (C and D) transluminal balloon tearing of the membrane. With a catheter still in place, a torn membrane can be seen after treatment. Arrows show the level of the aortic annulus and the membrane.

and redilation) were discharged 2 to 6 days after the procedure (mean, 2.7 days).

Follow-Up

Five patients (7%) were lost to follow-up 3 ± 2 years after treatment; none of them had recurrence before being lost. The clinical follow-up ranged from 0.5 to 25 years (mean, 16 years). One 55-year old patient died of noncardiac causes 1 year after treatment. Seventy-two patients remained asymptomatic, and 2 patients still had mild dyspnea. No syncope or palpitations were reported. No patients developed endocarditis or other complications. During this time period, late events occurred in 17 patients (23%). Recurrence of left ventricular outflow obstruction appeared in 15 patients (20%). Eleven patients (15%) had restenosis of the membrane 4 ± 2 years after treatment. The echocardiographic and angiographic features of the membrane after restenosis were similar to those observed at the first attempted dilation, indicating a regrowth of fibrous tissue at the same site as the cause of restenosis. In fact, mobility of the membrane was never observed when restenosis was detected. Redilation was always attempted in these patients, which resulted in pressure relief similar to that observed after the first dilation.⁹ The gradient decreased again from 58 ± 11 to 24 ± 8 mm Hg ($P < 0.01$). One patient with a recurrence of a significant subvalvular gradient 12 years after treatment showed good evolution of the torn membrane at first dilation, and a new distant membrane was the cause of late recurrent obstruction. The patient was successfully redilated 12 years later (Figure 3). Figure 4 is a graphic representation of the gradient evolution in nonrecurrent and redilated groups of patients. Three patients developed progression to subaortic muscular obstructive disease at follow-up (1, 4, and 6 years after balloon dilation) and were referred for surgery. The last peak residual Doppler gradient in 70 nonoperated patients was 31 ± 14 mm Hg at 16 ± 6 years after treatment. In terms of the

evolution of aortic valve regurgitation after treatment, only 1 patient had significant progression of aortic regurgitation at follow-up. This patient was sent to surgery for valve replacement. Figure 5 shows a graphic representation of the evolution of aortic regurgitation after balloon dilation; only minor changes were observed. The only patient who developed mild mitral insufficiency after balloon dilation of the membrane remained stable and symptom free under close follow-up monitoring. Figure 6 shows the Kaplan-Meier curves of our series.

Factors associated with better long-term results were studied. Table 2 shows a summary of the univariate analysis. As shown, the 20-year probability of being recurrence free was significantly higher in older patients (>13 years of age) who had a larger body surface area, bigger aortic annulus size, and a more distant membrane from the aortic annulus. In addition, recurrence-free survival was more frequent in patients with thinner membranes. The multivariate analysis (Table 3) identified annulus size and membrane thickness as the only independent factors that influence recurrence.

Discussion

Natural History

Discrete subaortic stenosis has been considered a mysterious disease because there are many uncertainties concerning its origin, pathogenesis, natural history, treatment options, and recurrence after treatment. The entity is surprisingly complex, and many mysteries remain unresolved. Histological and ultrastructural analysis of fibrous rings excised at operation have shown a layered arrangement of these rings that resembles that of normal endocardium.^{15,16} The ring may be localized in the left ventricular outflow tract, or it may extend fibrous connections to the aortic annulus or the mitral apparatus. Although DSS has classically been considered a congenital malformation, there is evidence for an acquired subaortic membrane during life. Certain observations on the

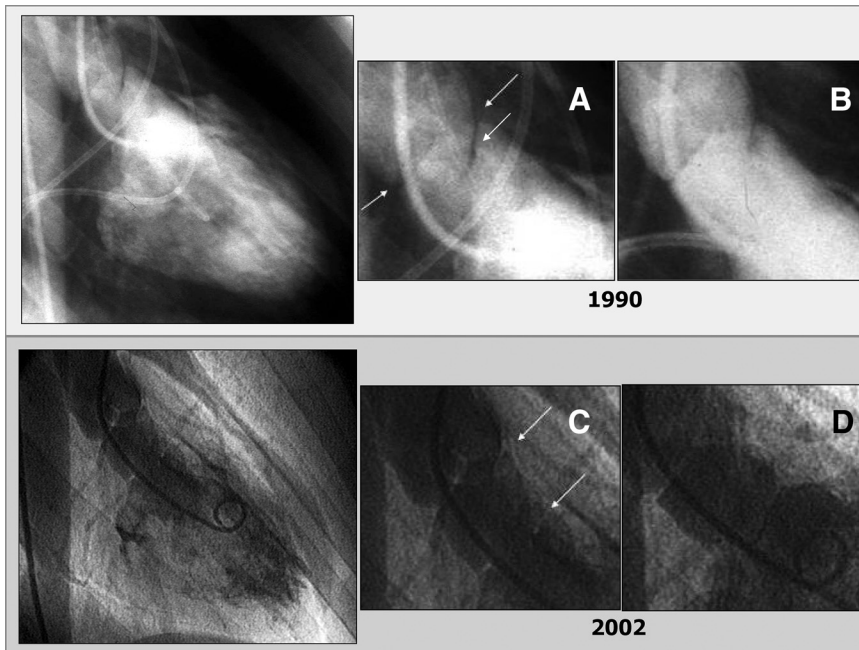


Figure 3. A series of 30° right anterior oblique left ventricular angiograms of a patient showing progression by the appearance of a new distant membrane 12 years after successful transluminal balloon tearing of a first membrane. **A**, Before first treatment; **B**, after first treatment; **C**, before second treatment; **D**, after second treatment. Arrows show the aortic annulus and the levels of both membranes.

follow-up of patients have shown how the disease may evolve from a thin membrane to a more advanced muscular obstruction. As postulated by Somerville et al¹⁷ 30 years ago, the pathological basis seems to reside in the myocardium, which could generate, by unknown mechanisms, a wide spectrum of abnormal hypertrophic responses in the left ventricular outflow tract. The “mildest” and most frequent form of the spectrum is the development of a thin (1 to 3 mm) crescent or circumferential fibrous diaphragm traversing the anterior portion of the left ventricular outflow tract, causing a fixed subaortic obstruction. The continuous high-pressure subaortic jet may damage the aortic cusps, and some degree of aortic regurgitation is frequently incurred. The degree of hemodynamic severity may also vary between a mild peak gradient (<30 mm Hg) and more severe forms (>70 mm Hg). In addition, progression of hemodynamic severity after diagnosis may develop, and it remains unknown why, when, and how it may happen. The degree of hemodynamic severity of the disease may influence the rate of possible evolving complications such as onset or worsening of aortic regurgitation, infectious endocarditis, or progression to tunnel lesion or extended muscular obstruction. For all these reasons, a thin DSS is considered a potentially progressive disease, and all diagnosed patients should be followed up indefinitely, no matter what treatment decision is made.

Surgery

Surgical intervention is an effective and safe treatment option for reducing the left ventricular-to-aortic gradient. During the last 27 years, there have been 20 published series on a total of 1098 operated patients.^{18–37} Therefore, a significant amount of information on surgical experience is now available. Different surgical techniques have been proposed. Sharp resection of the membrane or blunt enucleation, with or without myectomy or myotomy, has been applied successfully for better relief of outflow tract obstruction. However,

there is currently no consensus on which is the best option. Aggressive septal resection has been proposed³⁰ for improved relief of the outflow tract; however, it may also result in procedure comorbidities such as heart block. Aggressive resection may be needed in a more advanced muscular obstruction but may be unnecessary in patients with isolated thin membranes. Finally, surgery-related mortality ranges between 0% and 8% (mean, 1.8%).

Criteria for operation have included a pressure gradient >50 mm Hg and ECG signs of strain or symptoms such as dyspnea, angina, or syncope. However, the progressive nature of the disease has led others^{38–41} to recommend surgical treatment in patients with mild subaortic obstruction. Precise recommendations for operation based on gradient level are difficult. The postoperative gradient in some reported series³⁸ is higher than the initial gradient in treated groups with mild subaortic obstruction.^{39–40} Thus, discussion persists on when and how to operate on patients with thin DSS^{22–42} despite a general agreement that surgery is needed if progression is detected. The pressure relief after surgical treatment persists in most patients. The great majority of surgical series have shown a recurrence or progression at later follow-up, ranging from 0% to 35% (mean, 16.7%). In a recent report,³⁷ this recurrence rate was not different in patients treated with enucleation of the membrane alone (28%) compared with those who had enucleation plus septal myectomy (27%).

Aortic regurgitation can be arrested but not reversed by surgical resection.^{17,39} Some authors^{19,43} have noted that aortic regurgitation may develop even after surgical treatment. This late complication is more frequent in patients with a high preoperative pressure gradient, and it is related to the length of postoperation time. Thus, over time, a large proportion of surgically treated patients may become candidates for additional surgical procedures because of residual or recurrent stenosis or progressive deterioration of aortic valve function.

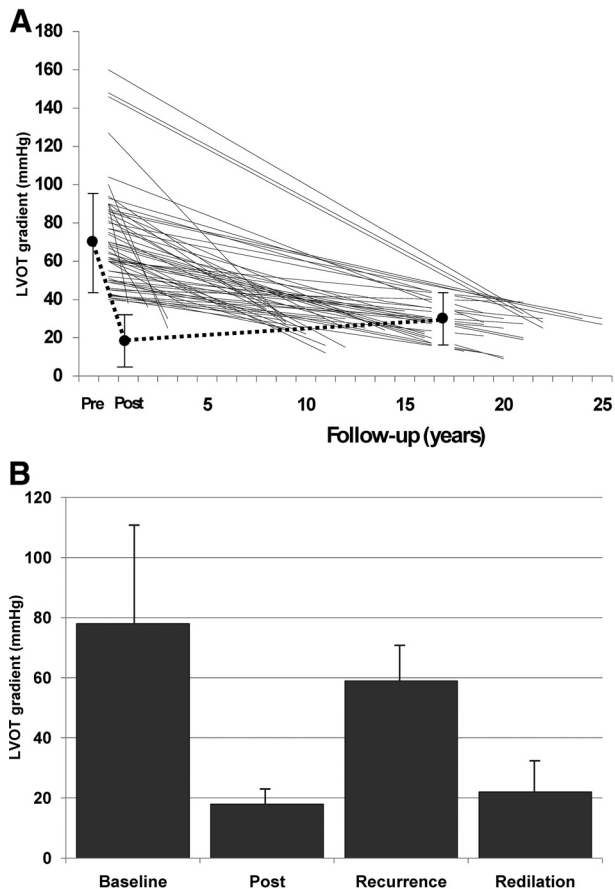


Figure 4. Subvalvular gradient evolution in nonrecurrent (**A**; n=58) and redilated (**B**; n=12) groups of patients. In **A**, the lines relate the baseline gradient of each patient to each latest gradient at follow-up; mean values before, immediately after treatment, and at mean follow-up time are also represented and connected by a dashed line. LVOT indicates left ventricular out-flow tract.

Balloon Dilation

Balloon tearing of the subaortic thin membrane is also an effective and safe method for reducing subaortic obstruction. Since its first description in 1985,^{1,2} several studies have confirmed that persistent pressure relief is achieved after balloon dilation.³⁻¹³ This treatment has not been widely used, and the experience with percutaneous treatment is very different from surgery. However, the mean gradient reduction

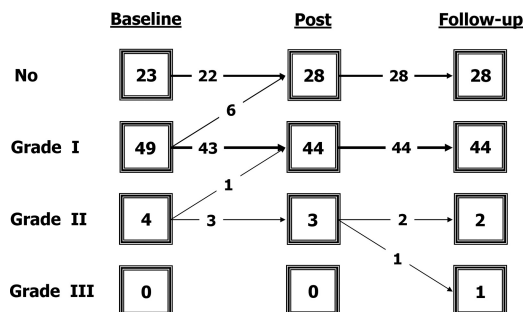


Figure 5. Evolution of the degree of aortic regurgitation after transluminal balloon tearing. The numbers in each box represent the number of patients having no or any degree of aortic regurgitation at each condition.



Figure 6. Kaplan-Meier probability curves of survival, surgery-free survival, and redilation-free survival at long-term follow-up.

in 11 series including 170 patients was 70%.³⁻¹³ Selection of patients is an important issue, and only patients with a thin membrane without any fibromuscular component should be attempted. In our series, there was a wide age range; children, adolescents, and adults were included. After successful TBT of the membrane, the remaining subaortic structure becomes widely mobile and flutters in response to flow, which suggests membrane rupture as the mechanism of relief. Hospital mortality is rare (1.3%) but always possible. Comorbidities are also rare, but puncture-site problems and the possibility of mitral dysfunction after balloon dilation should be considered. However, less is known about long-term follow-up because only a few midterm studies are available.⁶⁻⁹

Patients with a larger annulus size and a membrane more distant from the aortic valve have a better initial outcome.⁹ However, recurrence may appear over time. Our results show that such recurrence may be secondary to restenosis (15%) or disease progression (5%). This progression may lead to extended muscular obstructive disease or the development of a new distant membrane (Figure 3). In the univariate analysis, factors identified that decreased the likelihood of long-term recurrence included age at treatment >13 years, larger body surface area, larger annulus size, and thinner membranes. The annulus size has also been observed to similarly influence long-term surgical results.^{38,44} The fibrous tissue can involve the aortic valve annulus, which most likely contributes to limiting the annulus size. In addition, the distance from the membrane to the aortic annulus seems to influence long-term results after balloon dilation. A more distant and thinner membrane may result in less fibrous connections between the membrane and the annulus, which may favor better long-term results. However, the aortic annulus diameter and membrane thickness were the only independent factors that influenced late recurrence.

Table 3. Multivariate Recurrence-Free Survival Analysis (Cox Regression Model)

	β -Coefficient	Relative Risk (95% CI)	Wald	P
Annulus diameter	-0.107±0.044	0.89 (0.82-0.98)	5.98	0.014
Membrane thickness	1.131±0.501	3.09 (1.16-8.26)	5.1	0.024

CI indicates confidence interval. β -Coefficient is given as mean±SE.

The persistent pressure relief obtained in most patients after balloon dilatation may account for the favorable evolution observed in the degree of aortic regurgitation. Immediately after treatment and at follow-up, mild aortic regurgitation may not change or may even disappear; infrequently, it may also progress at follow-up. Thus, balloon dilatation does not seem to affect aortic valve competence unfavorably.⁶⁻⁹ In terms of the mitral valve, concern has been expressed about the possibility of the balloon dilatation creating new mitral insufficiency in the presence of valvular membrane attachments.²² The crescent membrane may not necessarily involve the mitral valve. Circumferential diaphragms may have attachments to the fibrous tissue continuity between aortic and mitral valves. This type of attachment does not seem to be dangerous for interventional procedures. In fact, none of the published series on balloon dilatation spanning the last 25 years had reported this complication. Nevertheless, in our experience, 1 patient (1.3%) developed mild mitral insufficiency after balloon dilatation. One possibility for mitral complication could be improper technique, including passing the guidewire and the balloon within the subvalvular chords. It is also possible that patients with attachments to the free border of the anterior mitral leaflet may experience tearing as a result of the balloon overstretching the fibrous connection. In our experience, this type of attachment is rare, but it should be ruled out before balloon dilatation is attempted.

Strengths and Limitations

This study presents the longest follow-up analysis of a series of patients with isolated thin DSS treated by TBT of the membrane. Factors influencing recurrence after treatment were identified, although the age- and body size-related factors could be interrelated. On the other hand, the study is a descriptive one. Selection of appropriate candidates for percutaneous treatment was an important issue, and only patients with a thin membrane at the outflow tract without a fibromuscular component were included. In addition, hemodynamic and angiographic assessments before and immediately after treatment were compared with similar echocardiographic parameters at follow-up. Although it would be ideal to use the same technique for assessments in all studied conditions, serial follow-up studies must be noninvasive. Thus, a combination of techniques was used for different purposes in the study, and angiographic parameters were used to study the factors that influence recurrence. Both techniques have greatly evolved over the past 25 years, and today, other imaging techniques like 64-slice computed tomography and nuclear magnetic resonance should better visualize the outflow tract anatomy. This capability could help in the selection of patients for interventional procedures.

Conclusions

We are still searching for the ideal treatment for this mysterious disease. Surgery and balloon dilatation are only palliative procedures, and they alone cannot cure the disease. Both of these procedures reduce the pressure gradient to a similar degree but do not eliminate the underlying mechanism for potential disease progression. If the gradient reduction somehow prevents further complications, then the percutaneous

technique appears to be an attractive alternative to surgery as a first-option treatment. The potential savings in patient discomfort are significant, although caution should always be taken to avoid perforation. Once a pressure gradient relief is obtained, continued follow-up is mandatory. Aortic insufficiency seems to be unaffected after balloon dilatation of the membrane. Recurrence seems to be favored by the presence of thicker membranes and smaller aortic annulus diameter. The recurrence rate may be similar to that observed in surgical patients, and both are probably influenced by the progressive nature of the disease. If restenosis of the membrane develops, balloon dilatation can be repeated successfully in most patients. With this strategy, surgery for a thin membrane may be delayed or even avoided.

Disclosures

None.

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CLINICAL PERSPECTIVE

This study presents the longest follow-up analysis of a series of patients with isolated thin discrete subaortic stenosis treated by transluminal balloon tearing of the membrane. Selection of appropriate candidates for percutaneous treatment was an important issue, and only patients with a thin membrane at the outflow tract without a fibromuscular component were included. After a mean follow-up time of 16 ± 6 years, 11 patients (15%) developed restenosis, 3 patients (4%) progressed to muscular obstructive disease, 1 patient (1.3%) developed a new distant obstructive membrane, and 4 patients (5%) underwent surgery at a mean of 3 ± 2 years after their first treatment. Fifty-eight patients (77%) remained alive and free of redilation or surgery at follow-up. Larger annulus diameter and thinner membranes were independent factors associated with better long-term results. These findings, together with current improvements in image techniques for better delineation of the outflow tract, could help in the selection of patients who can benefit from an interventional procedure as a first-choice treatment. The recurrence rate in this selected group of patients may be similar to that observed in surgical patients, and both are probably influenced by the progressive nature of the disease. If restenosis of the membrane develops, balloon dilation can be repeated successfully in most patients. With this strategy, surgery for a thin membrane may be delayed or even avoided.