# Feature Article

# Valvuloplasty for Rheumatic Mitral Stenosis: The Philippine Experience

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While acute rheumatic fever and rheumatic heart disease (RHD) have disappeared from developed countries, the global burden of the disease remains a concern with around 300,000 new cases worldwide.[1] It is estimated to affect 15.6 million people globally. There are at least 233,000 deaths per year secondary to RHD.[1]

Rheumatic Heart Disease remains to be a major cardiovascular problem in the Asia-Pacific region, with prevalence among school-age children at 0.3 to as high as 21 per 1000 children.[2] According to Dr. Santiago Guzman, the prevalence rate of RHD in the Philippines is 0.8 – 1.3/1000 after screening almost 170,000 school children in between 1987 to 1997.[2] It is very prevalent in the Philippines, as poverty malnutrition, overcrowding, poor housing, and shortage of health-care resources are determinants of this disease.[3]

Rheumatic Heart Disease (RHD) is the most common etiology of mitral stenosis. In one pathologic report where 99% of the 452 cases of mitral stenosis requiring mitral valve replace ment showed gross features attributable to rheumatic post-inflammatory disease.[4]

The dominant mechanisms causing clinically important mitral stenosis (MS) of rheumatic origin are commissural fusion, leaflet thickening and alteration of the subvalvular apparatus, with calcification and decrease in leaflet mobility as subsequent features.[5] The four forms of fusion of the mitral valve apparatus in rheumatic mitral stenosis are commissural fusion in 30%, cuspal fusion in 15%, chordal in 10% and combination of involvement in 45%.[6] It is clear that commissural fusion is a major component of MS in RHD making 75% of cases manifesting with commissural involvement.

Percutaneous transvenous mitral commissurotomy (PTMC) was first performed in 1984 by Inoue, and since then, has emerged as the treatment of choice for severe pliable rheumatic mitral stenosis (MS).[7] It is indicated in symptomatic patients with clinically significant MS (valve area $\leq 1.5$ cm$^2$), asymptomatic patients with high thromboembolic risk and/or high risk for hemodynamic decompensation (pulmonary hypertension at rest and exercise, need for major non-cardiac surgery, desire for pregnancy), for as long as the valve anatomy is favorable and there are no contraindications like left atrial thrombosis and moderate to severe mitral regurgitation, which would warrant surgical intervention instead. PTMC may also be performed as a bridge to surgery among high-risk, critically ill patients. The clinical characteristics, valve anatomy and local expertise should all be considered when deciding on the type of treatment.[8]

Echocardiography remains to be the main diagnostic modality for assessment of the severity and extent of involvement. A number of echocardiographic scoring systems have been developed to predict procedural success of PTMC; namely the Wilkin's scoring system, the scoring systems by Chen et al, Reid, Nobuyoshi and Cormier.[9] The Wilkin's scoring system, a semi-quantitative echocardiographic scoring

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system formulated by Wilkins and his associates in 1988, is the most popularly used scoring system all over the world. It is composed of four variables: (1) leaflet mobility, (2) leaflet thickness, (3) subvalvular thickness and (4) valvular calcification, each graded on a scale of 1 to 4 depending on the severity of involvement. (See Table 1). A Total score of 8 or less is usually associated with an excellent immediate and long term results, while scores of more than 8 have suboptimal results.[10] In a study by Cannan et al, there was a trend towards improved survival at 36 months, free of death, repeat percutaneous mitral balloon valvotomy or mitral valve replacement among patients with Wilkins score of 8 or less (78 ± 6% vs. 67 ± 8%, p = 0.07) The presence of commissural calcium was also assessed to be strongly predictive of outcome after PTMC, with a statistically significant higher incidence of mitral valve replacement and all end points combined among patients with commissural calcification. Thus, the simple presence or absence of commissural calcification assessed by two-dimensional echocardiography can also be used to predict outcome.[11]

A number of local studies have tried to validate the Wilkins scoring system, but came up with inconsistent results. A Wilkin's score of 11 or less had optimal outcomes in a study by Acosta et al,[12] while a score of 6 or less was predictive of success in the study by Pineda et al.[13]

One of the possible explanations for the inconsistency of these results is the failure of the Wilkin's scoring system to account for commissural involvement, despite this finding being considered as the pathognomonic sign of rheumatic MS. Previous studies have proven the presence of commissural calcium to be strongly predictive of outcomes after PTMC, with higher incidence of mitral valve replacement among patients with commissural calcification.[11] The grading system is also confusing and prone to errors, as there is significant overlap in the descriptions of the degree of valvular involvement. This poses a significant problem especially to a qualitative and descriptive scoring as this one, which has the inherent concern of interobserver variability to start with. In one local study, thirty experienced echocardiographers were made to evaluate and assess the mitral valve apparatus according to the Wilkin's echocardiographic scoring system. There was weak agreement among the raters in all the parameters assessed, although the very small sample size limited the validity of the results.[14]

The Philippine General Hospital (PGH) performed a total of 299 PTMC procedures from 1998 – 2012.

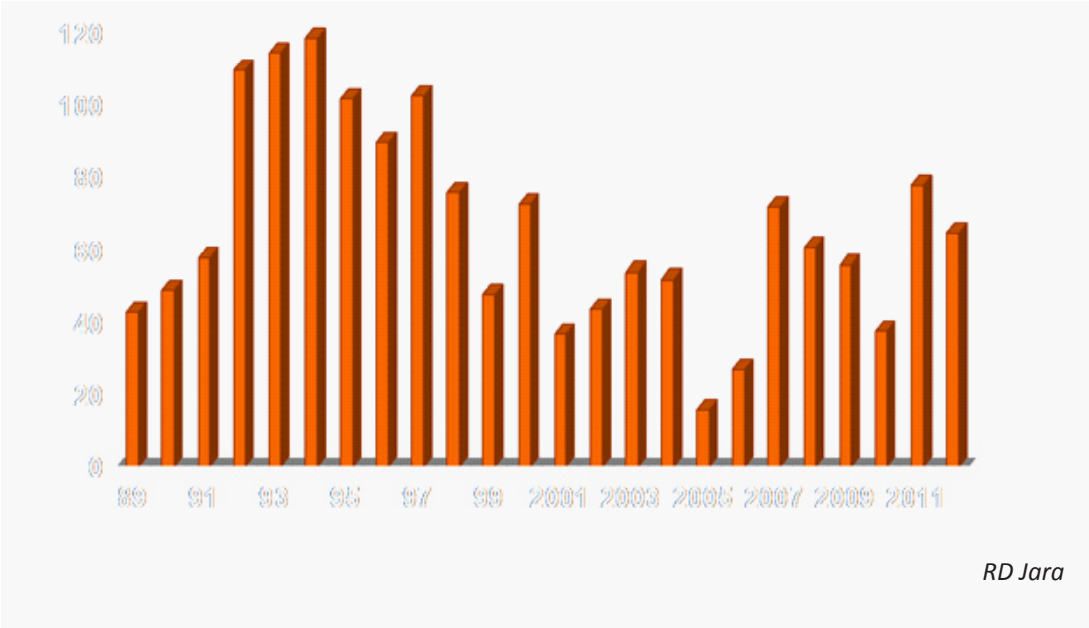


**Figure 1.** PTMC Procedure from 1989-2012 at the Philippine Heart Center

While the Philippine Heart Center (PHC) is the largest cardiac specialty hospital in the Philippines with a total of 1,586 PTMC procedures performed during the 23-year period from 1989-2012.

To address lack of consistency of the Wilkin's scoring system as a predictor of procedural outcomes, the Non-Invasive Laboratory of the Philippine Heart Center (PHC) developed its own echocardiographic scoring index for mitral stenosis. The four parameters used in the Wilkins score are still used in the scoring but with a revised grading system, and with the addition of two or more variables: (1) distribution of the commissural calcification, and (2) eccentricity of the mitral orifice. (See Table 2) This new echocardiographic scoring system was validated by Pascual AC, Jara RD, Santos RJ et al in a review of 425 PTMC procedures. The sensitivity and specificity of the PHC scoring system as predictor for procedural success were higher at 74% and 55% respectively, compared to the 57% and 52% respectively for the Wilkins score. The positive and negative predictive values of 74% and 55% respectively with the PHC score was also higher compared to the Wilkins score (67% and 41%). Commissural calcification, subvalvar involvement and leaflet mobility were found to be the most important variables predictive of outcomes for PTMC.[15]

Three-dimensional echocardiography (3DE) can also provide a more detailed morphologic display and analysis of the mitral valve apparatus and offer rapid, accurate quantification of mitral valve structure and valvular orifice area in 3D. Leaflet mobility and subvalvular apparatus involvement are predictors of optimal

**Table 1.** Wilkin's Scoring System (1988)[10]

| | |
|---|---|
| **Leaflet Mobility** | |
| Grade I | Highly mobile valve with leaflet tips only restricted |
| Grade II | Mid portion and base of leaflets have reduced mobility |
| Grade III | Valve continues to move forward in diastole, mainly from the base |
| Grade IV | No or minimal forward movement of the leaflets in diastole |
| **Leaflet Thickening** | |
| Grade I | Leaflets near normal in thickness (4 to 5 mm) |
| Grade II | Mid-leaflets normal, marked thickening of margins (5 to 8 mm) |
| Grade III | Thickening extending through the entire leaflet (5 to 8 mm) |
| Grade IV | Marked thickening of all leaflet tissue (>8 mm) |
| **Subvalvar Thickening** | |
| Grade I | Minimal thickening just below the mitral leaflets |
| Grade II | Thickening of chordal structures extending up to one-third of the chordal length |
| Grade III | Thickening extending to the distal third of the chords |
| Grade IV | Extensive thickening and shortening of all chordal structures extending down to the papillary muscle |
| **Leaflet Calcification** | |
| Grade I | A single area of increased echocardiographic brightness |
| Grade II | Scattered areas of brightness confined to the leaflet margins |
| Grade III | Brightness extending into the mid-portion of the leaflets |
| Grade IV | Extensive brightness throughout much of the leaflet tissue |

optimal PTMC success, and the incidence and severity of mitral regurgitation are associated with high-calcification score.[9,16] This technology however is not readily available in many of the institutions in our country, thus limiting its usefulness.

Rheumatic Heart Disease (RHD) is a very prevalent condition in the Philippines, and this therefore puts us in the best position to study, learn and offer the best solution to this problem. More local researches to further our knowledge on this disease should be undertaken.

**Table 2.** Philippine Heart Center (PHC) Echocardiographic Score Index for Rheumatic MS (1998) [15]

| | |
|---|---|
| **Leaflet Mobility** | |
| Grade I | Mildly reduced mobility (only the leaflet tips are restricted |
| Grade II | Moderately reduced mobility (Leaflet tip and mid portions are restricted |
| Grade III | Severely limited mobility (minimal forward movement of leaflets in diastole) |
| **Leaflet Thickening** | |
| Grade I | Mild leaflet thickening with minimal echo brightness. ( 4-5 mm) |
| Grade II | Moderate echo brightness (6-8 mm) |
| Grade III | Severe echo brightness (>8 mm) |
| **Subvalvar Thickening** | |
| Grade I | Mild thickening not extending to more than one third of the chordal length |
| Grade II | Moderate thickening extending to the distal third of the chordal length |
| Grade III | Severe calcification (areas of brightness involving most of the leaflet tissue) |
| **Leaflet Calcification** | |
| Grade I | Mild calcification (single area of increased echo brightness) |
| Grade II | Moderate calcification (scattered areas of brightness confined to the leaflet margins) |
| Grade III | Brightness extending into the mid-portion of the leaflets |
| **Eccentricity of the Mitral orifice** | |
| Grade I | Central or mild eccentricity (orifice to wall distance of the anterolateral and posteromedial commissures are equal or almost equal) |
| Grade II | Moderate eccentricity (orifice to wall distance of one commissure is 2x the orifice to wall distance of the other commissure |
| Grade III | Severe eccentricity (orifice to wall distance of one commissure is 3x the orifice to wall distance of the other commissures |
| **Distribution of commissural calcification** | |
| Grade I | Mildly disproportional (small commissural calcification) |
| Grade II | Moderately disproportional (moderate to severe calcification of one commissure) |
| Grade III | Severely disproportional or eccentric (moderate to severe calcification of both commissures) |

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