

Surgery for Rheumatic Heart Disease in Nepal

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Abstract

Rheumatic heart disease is still very common in the developing world. Nepal is no exception. Lack of health insurance for most people and monitoring of the PT/INR in a long run pose a significant financial challenge to an ordinary Nepalese family and is associated with significant lifestyle issues of the patients. This article presents the surgical issue in the management of rheumatic heart disease in Nepal.

Introduction

Rheumatic heart disease (RHD) is still very common in the developing world. Nepal is no exception. Surgical therapy for RHD has become a routine and fairly safe procedure in Nepal. At the same time, lack of health insurance for most people and monitoring of the PT/INR in a long run pose a significant financial challenge to an ordinary Nepalese family and is associated with significant lifestyle issues of the patients.

The patho-physiology of rheumatic fever (RF) and RHD is well described in the literature. It affects mostly children and young adults. The antibodies formed against the streptococcal antigen also attack the endocardial tissue, a phenomenon called molecular mimicry, thereby causing inflammation of the target tissue. It involves the endocardium and the valves of the heart, joint and serosal surfaces of the pericardium, pleura and brain.¹ Most commonly involved valves in order of frequency are mitral, mitral and aortic valve combined, and aortic valve alone followed by tricuspid valve. The pulmonary valve is almost never involved by the rheumatic process, and so, any involvement of this valve is considered to be of congenital origin unless proven otherwise.

Prevalence of Rheumatic Heart Disease (RHD) among school children in Kathmandu Valley of Nepal is about 1.2 per thousand.² Surgery for RHD is one of the commonest procedures performed in Nepal. Among the two major cardiothoracic centers of Kathmandu, ShahidGangalal National Heart Center (SGNHC) and Manmohan Cardiothoracic Vascular and Transplant Center (MCVTC), surgery for valve lesions comprises between 30-40% of all cardiac surgeries. In both the centers, vast majority of the valve lesions are of rheumatic origin. The annual number of valve surgeries at SGNHC and MCVTC is approximately 450 and 250 respectively. The cumulative number of patients who have had heart valves replaced in Nepal exceeds 5000. There are significant social and economic issues related to RHD and surgical treatment of it. RHD is more commonly seen in young girls from rural areas. The patients may have significant issues with pregnancy and marriage. While the disease affects people from lower socio-economic class, the treatment of valve surgery is costly and patients often need repeated admissions and lifelong treatment.

Historical perspectives

Surgery for isolated mitral stenosis started as early as 1965s at the Bir Hospital³; Dr. D.N. Gongal performed first few closed mitral commissurotomies with limited success. His successor, Dr. L.B.Thapa, set up a cardiothoracic surgical program, but could perform only closed heart surgeries. Later, Dr. G. Sharma and B. Koirala started closed and Open Heart surgery program at Tribhuvan University Teaching Hospital.⁴ It was not until the establishment of Shahid Gangalal National Heart Center, with B. Koirala as the chief, and R. Koirala and J. Sharma in the surgical team, that a comprehensive cardiac surgical program was established in Nepal.

Closed and Open Mitral Commissurotomy

Isolated mitral stenosis is fairly common in our daily practice. In the earlier days of history of cardiac surgery in Nepal, the only treatment for this condition was surgical closed mitral commissurotomy. The severity of stenosis and the symptoms of the patient constitute the main basis for intervention. However, other factors such as the quality of valve leaflets, involvement of the sub valvar apparatus, and presence of thrombus are important considerations in selection of a surgical procedure.

Closed mitral commissurotomy (CMC) is one of the early surgical procedures performed in the history of cardiac surgery and has given quality life to many of the patients.

Indications for surgery

CMC used to be a very common procedure in many countries, including Nepal, in the past but now has been almost entirely replaced by percutaneous transcatheter balloon dilatation (PTBD).

The typical indications for CMC or PTBD are:

- Symptomatic severe mitral stenosis with MVA less than 1.5 cm²
- Asymptomatic very severe mitral stenosis with MVA of less than 1 cm²

- Asymptomatic severe mitral stenosis with MVA less than 1.5 cm² with new onset of atrial fibrillation
- Mitral stenosis with history of PND or pulmonary edema.

Echocardiographic mean diastolic gradient across mitral valve more than 10 mm Hg is indicative of significant mitral stenosis but it is not considered as an indication independently because of its variability.

The only indication for CMC in the current era could be the need for commissurotomy when there are no PTBD facilities. It may also be performed when balloondilatation is not feasible in small children. Mitral stenosis with failed PTBD is also an indication for CMC but usually requires an open procedure. In general terms, AHA/ACC guidelines for intervention in mitral and aortic valve pathology are followed.⁵ Detailed preoperative study of valve anatomy with 2D and color flow echocardiography are mandatory. Echocardiography will demonstrate the quality of the valve tissue, degree of stenosis, presence of subvalvar involvement, presence of left atrial thrombus, size of the left atrium, left ventricular function, and degree of mitral regurgitation. Wilkins score is a useful guide to identify patients suitable for a closed or open procedure. This is calculated based on the severity of changes in each component of the mitral valve apparatus, namely leaflet thickening, calcification, leaflet mobility, and involvement of the subvalvar apparatus. Wilkins score of less than 8 out of total 16 offers high chance of having a successful closed procedure.⁶

Contraindications

The contraindications for a closed procedure (CMC or PTBD) are:

- Presence of left atrial thrombus
- Presence of more than mild mitral regurgitation
- Presence of severe subvalvar involvement
- Heavy calcification of the valve

Slight calcification of the tips of the leaflets is not a contraindication. Presence of a giant left atrium is a relative contraindication for CMC. Presence of atrial fibrillation per se is not a contraindication but needs detailed and careful evaluation for the possible presence of a left atrial thrombus. Open mitral commissurotomy (OMC) used to be a popular procedure for isolated mitral stenosis when techniques of open heart surgery were first established. OMC is seldom used these days for isolated pure mitral stenosis because most of them would have been dealt with by PTBD. The technique of open commissurotomy, however, is still a useful procedure today in various situations. The general indications for open commissurotomy are similar to those for closed intervention. However, here are certain specific conditions where OMC is particularly suitable:

- Symptomatic severe mitral stenosis with MVA <1.5 cm² and left atrial thrombus that does not disappear with anticoagulation.
- Complicated PTBD with leaflet tear causing severe MR or a failed PTBD.
- Calcific symptomatic severe mitral stenosis where PTBD is not feasible or contraindicated.
- Symptomatic moderate mitral stenosis (MVA >1.5 cm²) when a cardiac surgical procedure is being undertaken for another condition

OMC is frequently performed as part of combined procedures when severe mitral stenosis is present along with a condition

that independently requires surgical intervention. They include aortic valve procedures, closure of atrial septal defect, or repair of severe tricuspid regurgitation among others. OMC alone is generally not sufficient when the leaflets are calcified, or the subvalvar apparatus severely affected. The surgical technique of OMC is generally straight forward. The most important point in OMC is the identification of line of commissural fusion. The sharp commissure is added with resection of secondary chords and placement of a ring(s).⁷ In such conditions, either a more complicated repair procedure should be performed by a surgeon experienced in rheumatic valve repair or a more straight forward valve replacement should be carried out.

Results

The early results of CMC and an isolated OMC procedure were excellent. Operative mortality for a CMC varies according to the preoperative functional class. Stanley John, et al., in one of the largest series of CMC, reported 1.5% hospital mortality.⁶ CMC using a dilator gives an excellent postoperative MVA. Although restenosis of the valve is a common phenomenon, most commonly seen 10 to 15 years after CMC, long-term survival is excellent. A total of 5% to 10% of the patients after CMC may develop significant MR and may need early reintervention using an open procedure. Our own experience at the University Teaching Hospital and the National Heart Center in the early days of cardiac surgery in Nepal are fairly similar. OMC for isolated mitral stenosis carries a low mortality, nearing zero. The procedure achieves good postoperative valve orifice, reaching up to 3.4 cm² and 87% freedom from significant MR or MS at 10 years. Since the intraoperative transesophageal echocardiography (TEE) is available in most centers, significant residual MR should not be accepted in the current era. The comparative benefits of CMC or OMC are even more striking for patients who have difficulty in managing warfarin after mechanical valve implantation. The risk factors for restenosis after open or closed procedure are the quality of valve leaflets and degree of subvalvar involvement at the time of intervention.

Mitral valve replacement

Mitral valve replacement is probably the commonest cardiac surgical procedure in Nepal. The indication for surgery on mitral valve are well outlined in AHA guidelines.⁵ The operative mortality for this procedure is quite low, ranging from 3-7% depending upon the time era. Long term follow up has been limited due to poor direct access to patients and lack of national valve registry.

Since the first mechanical valve replacement with Starr-Edwards Ball and Cage valve in 1961, there have been significant changes in the valve design in the world. But no prosthetic valve is ideal. Most commonly used valve is mechanical bileaflet valve and only about 10-15% of all valve replacements get a tissue valve, in our set up. The technical procedure of mitral valve replacement is fairly straightforward. The surgical approach is from left atrium or by trans-septal approach, if the tricuspid valve needs to be addressed. Generally, our preferences have been placement of interrupted, pledgetted, horizontal mattress polyester sutures and place the valve in intra-annular position. Because of the ease of the procedure and predictable outcome most surgeons choose to replace a rheumatic valve. However, use of a mechanical valve comes with significant issues of anticoagulation for life time. Thrombosis of the valve with acute heart failure is not very common but a definite possibility. Warfarin induced bleeding complications are much more common in our daily practice and constitute a serious cause of morbidity after mitral (and aortic) valve replacement.

Mitral Valve Repair

The initial stage of mitral valve involvement always starts from at least some degree of mitral regurgitation. That is because of chordal lengthening and some degree of associated myocarditis. As the process progresses the leaflets start to adhere to each other, thereby creating stenosis. In general, mixed stenotic and regurgitant lesions are more common than pure mitral stenosis. The best option of treatment for a regurgitant mitral valve is the repair, because of the difficulties in monitoring of the PT/MR and follow up of the patients in our setting.

The pathological anatomy of rheumatic mitral regurgitation is complex. However, rheumatic process generally involves all components of the mitral valve apparatus, thus may have multiple mechanisms for regurgitation. Leaflet thickening, commissural fusion, chordal shortening or fusion, chord elongation, leaflet fibrosis and calcification all can be present in one diseased valve. Late presentation or referral to a surgeon makes the valve more fibrotic and or calcified making a successful repair of the valve even more challenging.

Reparative techniques to correct mitral valve disease were first introduced in 1957 by Walton Lillehei at University of Minnesota, but it did not get great popularity as mechanical and bioprosthetic valves were introduced in 1960s which became popular because of their simplicity of insertion, reliability of function. However, issues related to the replacement of valves like thromboembolism, hemorrhage and durability again forced the surgeons to look for an alternative. In the late 70s, Alain Carpentier and Carlos Duran pioneered reliable techniques for MV repair, slowly repair became popular again. Carpentier Classification is widely used to describe the regurgitation and serves as a guide for selection of repair technique.⁷ Many surgeons have reported high success rates in repair of rheumatic mitral valve disease. However, in our own experience, the reparability of all the valves presenting for surgery has reached only up to 20-25%. The bottom line of a successful repair is exact delineation of the dominant pathology using TEE and intraoperative assessment by surgeon. Correction of the dominant pathology (chord transfer, chord shortening, chordolysis, commissurotomy, leaflet augmentation, resection of the secondary chords of the posterior leaflet, use of a rigid or a semirigid ring) are always necessary. Repair cannot be accepted if there is more than mild regurgitation in post bypass TEE and prompt replacement of the valve with a suitable prosthesis is advisable in such cases. Mitral repair with intraoperative ablation can also be undertaken in patients with atrial fibrillation, if the size of the LA is less than 6 cm.

There have a huge number of publications of the techniques and the results of rheumatic mitral repair in the literature. The first landmark paper on this topic was published by Alain Carpentier and his colleagues in 1980. They reported 10 years follow up of 551 cases with 4.2% early and 7% late mortality and 11% reoperations in isolated mitral repair.⁸ Similarly, Yankah et al have shown that pure mitral incompetence may be reparable and long-lasting, whilst mixed lesions may be reparable but the results are suboptimal.⁹ Surgeon related factors, like, residual prolapse, non use of a ring, ring dehiscence, undersizing or oversizing the ring are all very important reasons for failed repair. Progression of the inflammatory process leading to either leaflet retraction or recurrent prolapse are the most important causes of late failure.^{10,11} Our experience in rheumatic mitral valve repair is limited, but encouraging in recent times.¹² Because of the complexity of the pathology, the learning curve for rheumatic mitral valve repair is long one.

Aortic valve replacement and repair

Surgery for rheumatic aortic valve disease is fairly common in Nepal. In adult population, however, degenerative and rheumatic processes compete in terms of their prevalence. Repair of an aortic valve is generally fraught with failure, especially in rheumatic and degenerative processes. Its role is more established in congenital valve lesions. Trusler's repair for an isolated cusp prolapse, leaflet extension in a retracted leaflet or just a commissurotomy for stenosis are the commonly employed techniques. Our experience with aortic valve repair is limited. More often, a more straightforward aortic valve replacement (AVR) is carried out. We prefer mechanical, bileaflet valve in any one who is less than 55 years of age. The INR can be maintained at a lower level than for mitral valve replacement and thus has less bleeding complications. A bovine pericardial or stent mounted porcine valve is preferred if the patient is in sinus rhythm and is over 55 years of age, or when the patient is a lady of reproductive age and desires to conceive. Anticoagulation with warfarin is indicated for the first three months after AVR with biological valve after which the patient can be switched to aspirin.

Organic and functional Tricuspid valve lesion in RHD

Tricuspid valve disease had been ignored for in the earlier days of heart valve surgery. Consequently, a lot of patients returned back with severe tricuspid regurgitation with serious functional disability. Currently, there has been a growing pool of knowledge on the patho-physiology of progression of tricuspid valve disease after left sided valve surgery. There are extensive publications on the functional tricuspid regurgitation in left sided valve surgery in the literature. Even in the setting like ours, where the vast majority of valve surgery is for RHD, most tricuspid regurgitations (TR) are functional in nature. As has happened with most centers in the world, we largely ignored moderate TRs in the past. The results of redo surgery for progressive TR after left sided valve surgery are poor. This is why, we need to be more meticulous and aggressive in repair of TRs during the first left sided valve surgery.^{13,14} The techniques of tricuspid repair in TR are well described in the literature. In the current era, all tricuspid valves that have moderate or more regurgitation or if the annulus is more than 40 mm in diameter are repaired using an appropriately sized tricuspid ring. In cases of organic TS or TR, where the tissue is contracted by organic involvement of the tricuspid valve, we prefer patch augmentation of TV using fresh autologous pericardium.

Minimal Access Surgery

Minimal access surgery for AVR utilizing upper mini sternotomy is in our practice for more than 15 years. Recently we have started using right thoracotomy for tricuspid or mitral valve surgery. Sutureless valve and transcatheter valve placement has become a routine undertaking in most developed countries, but have not entered our practice because of its cost and lack of health insurance.

Thrombosis and anticoagulation issues after valve surgery

Anticoagulation management is of paramount importance after a mechanical valve replacement. A mechanical valve in aortic position requires an INR between 2 and 3 and in mitral position requires INR between 2.5 and 3.5. Precise control of INR in our setting is very difficult to achieve because of poor access to good healthcare facilities and lack of education required for self management of INR. The valve thrombosis

is a dreaded complication of under-warfarinization. On the other hand, minor to major bleeding issues are much more common in our country.¹⁵ Again, careful counseling and regular monitoring of the INR through a reasonably accessible health facility is the only key to reducing anticoagulation related issues. Pregnancy after mechanical valve replacement is well tolerated by most patients provided the anticoagulation management is done carefully. Recently, a report was published on outcome of pregnancies in women with prosthetic mechanical heart valves in Nepalese population.¹⁶ It suggested that pregnancies in women with prosthetic mechanical heart valves have a favorable outcome even in the low-resource country like Nepal. Most of the patients can successfully carry pregnancy and very few may have major bleeding issues around the time of delivery, if the anticoagulation protocols are followed strictly.

Conclusions

RHD is still very prevalent in the developing world. Historically a very common and a life-saving procedure, CMC, is now largely replaced by PTBD. OMC is still a useful procedure and is applicable in a variety of valvular heart conditions. The early results of CMC or OMC are excellent with good long-term results. The knowledge and skills of these two procedures can still be useful for a cardiac surgeon in the contemporary world. Mitral valve repair is an extremely useful in our setting to avoid the use of warfarin and the consequent management of INR and bleeding issues. This demands dedication and development of special skills. Aortic valve repair, although popular in academic fora, has not taken off well in our part of the world, where the rheumatic process is the dominant one. Access to reliable INR assessment for the patients from all corners of our country is still a challenge, which we will have to address. Starting a national valve registry and systematic collection of follow up data should be our next agenda.

References

1. Cunningham MW. Streptococcus and rheumatic fever. *Curr Opin Rheumatol*. 2012;24(4):408-416.
2. KC Man Bahadur, D Sharma, MP Shrestha, et al. Prevalence of Rheumatic and Congenital Heart Disease in School Children of Kathmandu Valley in Nepal. *Indian Heart J* 2003;55:615-18.
3. Amatya R. Evolution of Anesthesia in Nepal: A historical perspective. *Journal of Society of Anesthesiologists of Nepal* 2015 Oct 3;1(1):3-6.
4. Koirala B. Beginning of Open Heart Surgery in Nepal. *J Nep Med Assoc*. 1998;37(125): 447-8.

5. Nishimura RA, Otto CM, Bonow RO, et al. 2014 AHA/ACC guidelines for the management of patients with valvular heart disease: A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *J Thorac Cardiovasc Surg*. 2014;148(1):e1-e132.
6. Abascal VM, Wilkins GT, O'Shea JP, et al. Prediction of successful outcome in 130 patients undergoing percutaneous balloon mitral valvotomy. *Circulation*. 1990;82:448-456.
7. Koirala B. open and close mitral commissurotomy for rheumatic mitral stenosis in master technique in surgery. *Cardiac surgery edited by Grover FL and Mach MJ. Wolter Kluwer Philadelphia* . 2016; PP 219-218.
8. John S, Bashi VV, Jairaj PS, et al. Closed mitral valvotomy: Early results and long-term follow-up of 3724 consecutive patients. *Circulation*. 1983;68:891-896.
9. Carpentier A. Cardiac valve surgery--the "French correction" *J Thorac Cardiovasc Surg*. 1983 Sep;86(3):323-337.
10. Yankah CA, Siniawski H, Detschades C, et al. Rheumatic mitral valve repair: 22-year clinical results. *J Heart Valve Dis*. 2011 May;20(3):257-264.
11. Chauvad S, Fuzellier JF, Berebi A et al. Long term (29 years) results of reconstructive surgery in Rheumatic Mitral Insufficiency. *Circulation*. 2001;104(suppl1):I-12-I-15.
12. Deloche A, Jebara VA, Relland JY, et al. Valve repair with Carpentier techniques. The second decade. *J Thorac Cardiovasc Surg*. 1990 Jun;99(6):990-1001.
13. Pradhan S, Koirala B, Koirala R, Sharma J, Rajbanshi B, Timala R, et al. Outcome of Repair of Rheumatic Mitral Valve (Asian Pacific Society of Cardiology Poster Session 1, The 71st Annual Scientific Meeting of the Japanese Circulation Society). *Circulation journal: official journal of the Japanese Circulation Society*. 2007 Mar 1;71:322.
14. Dreyfus GD, Martin RP, Chan J, et al. Functional Tricuspid Regurgitation: A Need to Revise Our Understanding. *J Am Coll Cardiol*. 2015;65(21):2331-2336.
15. Jorge Rodriguez-Capita, Juan J. Gomez-Doblas, Leticia Fernandez-Lopez, et al. Short and Long-term Outcomes of Surgery for Severe Tricuspid Regurgitation. *Rev Esp Cardiol*. 2013;66(8):629-635.
15. Shrestha P, Koirala B, Shrestha N, et al. Study on Complication and Monitoring of Warfarin in Post Valve Surgery. *J Nep Med Assoc*. 2009;48(174):111-115.
16. Sharma P. Outcome of pregnancies in women with prosthetic mechanical heart valves. *Nepalese Heart Journal*. 2016 12;13(1):5-7.

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