Rheumatic heart disease in pregnancy

Anand Sagar*, MBBS (3rd year resident); Col. G. Vyas†, MD; R. G. Dhwale‡*, MD; S. B. Gawani‡, MD; V. Garg‡, MD; V. Porwal‡*, MD; A. Sharma‡, MD; A. Neekhra*, MBBS (2nd year resident)

* Resident, ‡ Professor and Director, § Associate Professor, Department of Medicine
R. D. Gard Medical College, Ujjain, Madhya Pradesh, India

Abstract
Rheumatic heart disease is a common heart condition in India and poses a great burden which becomes more significant if a pregnancy is being planned or if the patient is pregnant. A good knowledge about the disease, precautions and appropriate management at the right hour changes the outcome of pregnancy, thus having a deep insight into this subject is very important.

Key Words
• Rheumatic heart disease (RHD)
• Percutaneous mitral valvuloplasty (PMV)
• Mitral stenosis (MS)
• Tricuspid stenosis (TS)
• Mitral regurgitation (MR)
• Tricuspid regurgitation (TR)

Introduction
Rheumatic heart disease remains a major health problem in the developing countries. According to World Health Organization (WHO) approximately 300,000 individuals acquire acute rheumatic fever each year.

Rheumatic heart disease follows as a non-suppurative manifestation of group A beta hemolytic streptococcal pharyngitis; which causes permanent damage to the cardiac valves, viz., mitral, aortic, tricuspid, and pulmonary valves, are involved in order of frequency of which the mitral valve is the commonest one while pulmonary valve involvement being very-very rare. Prevalence of RF/RHD has been attributed to overcrowding and unhygienic living related to low socioeconomic status. Early diagnosis and treatment are the main cornerstones for preventing the disease progression.

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Pregnancy and rheumatic heart disease

Worldwide, rheumatic fever is the predominant cause of valve disease encountered during pregnancy. It is fairly common in developing countries and as far as an Indian scenario is concerned its incidence is very high. The hemodynamic changes that occur during pregnancy pose a challenge for a physician when it is accompanied with rheumatic valvular heart disease. Management of valvular disease through pregnancy is largely the same regardless of the cause. An exception is that those with rheumatic fever as the cause should be advised of antibiotic prophylaxis against a recurrence, even during pregnancy. Before going into the disease process we must be well versed with the normal cardiovascular changes during pregnancy.

Pregnancy

Normal cardiovascular changes during pregnancy

The periods most at risk for complications are labor, delivery, and the early puerperium since acute heart failure may be precipitated by an increased venous return to the heart as a result of decompensation of the inferior vena cava and physiological return of extravascular fluid into the systemic circulation (auto transfusion).

Delivery being a state of anxiety, increased physical effort may lead to tachycardia and acute heart failure to

and left-sided heart obstruction (mitral valve area <2 cm², aortic valve area <1.5 cm²) are predictors of adverse cardiac events. When valve morphology is favorable, percutaneous mitral balloon commissurotomy is the preferred intervention.
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Abstract
Rheumatic heart disease is a common heart condition in India and it poses a great burden which becomes more significant if a pregnancy is being planned or if the patient is pregnant. A good knowledge about the disease, precautions and appropriate management at the right hour changes the outcome of pregnancy, thus having a deep insight into this subject is very important.

Key Words
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Introduction
Rheumatic heart disease remains a major health problem in the developing countries. According to World Health Organization (WHO) approximately 500,000 individuals acquire acute rheumatic fever each year.

Rheumatic heart disease follows as a non-suppurative manifestation of group A beta hemolytic streptococcal pharyngitis; which causes permanent damage to the cardiac valves, viz., mitral, aortic, tricuspid, and pulmonary valves, are involved in order of frequency of which the mitral valve is the commonest one while pulmonary valve involvement being very very rare. Prevalence of RF/RHD has been attributed to overcrowding and unhygienic living related to low socioeconomic status. Early diagnosis and treatment are the main cornerstones for preventing the disease progression.

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Rheumatic heart disease follows as a non-suppurative manifestation of group A beta hemolytic streptococcal pharyngitis, which causes permanent damage to the cardiac valves, viz., mitral, aortic, tricuspid, and pulmonary valves, are involved in order of frequency of which the mitral valve is the commonest one while pulmonary valve involvement being very very rare. Prevalence of RF/RHD has been attributed to overcrowding and unhygienic living related to low socioeconomic status. Early diagnosis and treatment are the main cornerstones for preventing the disease progression. An exception is that those with rheumatic fever as the cause should be advised of antibiotic prophylaxis against a recurrence, even during pregnancy. Before going into the disease process we must be well versed with the normal cardiovascular changes during pregnancy.

Normal cardiovascular changes during pregnancy

Pregnancy and rheumatic heart disease

Worldwide, rheumatic fever is the predominant cause of valve disease encountered during pregnancy. It is fairly common in developing countries and as far as an Indian scenario is concerned its incidence is very high. The hemodynamic changes that occur during pregnancy pose a challenge for a physician when it is accompanied with rheumatic valvular heart disease. Management of valve disease through pregnancy is largely the same regardless of the disease. An exception is that those with rheumatic fever as the cause should be advised of antibiotic prophylaxis against a recurrence, even during pregnancy.

Pregnancy

Pregnancy is associated with changes in cardiac output and aortic cross-sectional area that are most marked in the third trimester. Atrial and ventricular systolic function (ejection fraction <40%) and left-sided heart obstruction (mitral valve area <2 cm², aortic valve area <1.5 cm²) are predictors of adverse cardiac events. When valve morphology is favorable, percutaneous mitral balloon commissurotomy is the preferred intervention. In patients with calcified immobile valves and subvalvular fusion, the choice between therapeutic interventions using percutaneous mitral balloon commissurotomy, surgical commissurotomy, or mitral valve replacement (MVR) should be made according to the institutional expertise and should be performed in patients with severe MS before conception, even in the absence of symptoms. If first recognized during pregnancy and symptoms develop, standard medical therapy is appropriate. The primary aim is to reduce heart rate in order to allow improved ventricular filling. For this purpose patients should be advised to take bed rest, Beta-blockers are indicated to optimize diastolic filling and in general are relatively safe during pregnancy. Diuretics are given to relieve pulmonary and systemic congestion. Furosemide has been proven safe during pregnancy; however, it should be used with caution.

In case atrial fibrillation occurs, therapeutic-dose anticoagulation should be given and an attempt should be made to restore sinus rhythm. Treatment with low molecular weight heparins is safe during pregnancy. Despite optimal therapy, if symptoms persist then invasive treatment should be considered, when anatomically suitable valves are present percutaneous mitral valvuloplasty (PMV) has been shown to be a safe and effective treatment during pregnancy. Balloon valvuloplasty can be performed (with appropriate radiation shielding of the fetus). Mitral valve surgical commissurotomy or valve replacement has been performed, but fetal loss exceeds 30%. Percutaneous mitral balloon commissurotomy should preferably be performed after 20 weeks of gestation, the period safest for the fetus. During pregnancy atrial fibrillation is a great concern. Immediate treatment of a rapid ventricular response should include IV verapamil or cardioversion.

The periods most at risk for complications are labor, delivery, and the early puerperium since acute heart failure may be precipitated by an increased venous return to the heart as a result of decompression of the inferior cava and physiological return of extravascular fluid into the systemic circulation (auto transfusion).

Delivery being a state of anxiety, increased physical effort may lead to tachycardia and acute heart failure to
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avoid this heart rate control is indicated. Considering the mode of delivery, vaginal delivery under epidural analgesia is safe provided the patient is in stable condition. Measures to shorten the second stage of labor in parturients may be helpful. Cesarean section seems to be an upper hand as it avoids the physical stress of labor, but the hemodynamic consequences associated with anesthesia and assisted ventilation and increased risk of venous thromboembolism are the major pit falls and there has been no significant change in the outcome when compared with the vaginal delivery in various studies; but it is indicated in patients with moderate or severe MS who are in NYHA class II/IV or have pulmonary hypertension despite medical therapy, in whom percutaneous mitral commissurotomy cannot be performed or has failed. It is advised that in all cases the decision about the mode of delivery should be finalized after a discussion with the cardiologist, anesthetist, and obstetrician of the patient.

During puerperium also patients are at risk of cardiac complications because of volume overload due to autologous transfusion. Patient should be examined for signs of heart failure and clinical observation for at least three days.

Mitral regurgitation

This entity doesn’t impose a great problem as mitral regurgitation is well tolerated during pregnancy. Pregnancy can alter examination findings, but rare associated arrhythmias, endocarditis, cerebral emboli, and hemodynamically significant regurgitation are no more likely to occur during pregnancy than at other times.1

Aortic stenosis

Rheumatic disease of the aortic leaflets produces commissural fusion, sometimes resulting in a bicuspid-appearing valve. This condition, in turn, makes the leaflets more susceptible to trauma and ultimately leads to fibrosis, calcification, and further narrowing. The diagnostic criteria for aortic valve stenosis remain same during pregnancy as at other times. Pregnancy in the presence of aortic valve stenosis can be successful, but if stenosis is severe maternal deaths have occurred (1–2%), and congestive heart failure is common.3 Even if tolerated during pregnancy, aortic valve stenosis may affect subsequent maternal functional capabilities and survival.1

The offloading may have an incidence of congenital heart disease as high as 20%, which may be reduced by correcting the outflow tract obstruction before pregnancy.4

If severe stenosis is recognized before pregnancy, balloon valvuloplasty or a surgical commissurotomy is recommended prior to conception. If pregnancy does occur in the presence of severe aortic stenosis, measures to avoid hypovolemia are particularly important. If severe symptoms persist despite adequate treatment, a balloon valvuloplasty or aortic valve surgery can be performed during pregnancy.6

Aortic regurgitation

Rheumatic disease results in thickening, deformity, and shortening of the individual aortic valve cusps, changes that prevent their proper opening during systole and closure during diastole. Generally, it’s well-tolerated during pregnancy. If it is severe/symptomatic, or associated with LV dysfunction, valve surgery should be considered before pregnancy.7 If congestive heart failure occurs with pregnancy, treatment should include afterload reduction. ACE inhibitors and ARBs should be avoided. If endocarditis occurs and the infection is not rapidly controlled, mortality with medical therapy is high, and surgical therapy is indicated. If this occurs late in pregnancy, consideration of associated cesarean section is appropriate.

Tricuspid valve disease

Rheumatic fever may produce organic (primary) tricuspid regurgitation (TR), often associated with tricuspid stenosis (TS). Tricuspid regurgitation usually requires no specific therapy during pregnancy. Tricuspid stenosis is rare. If it is encountered, avoidance of intravascular volume depletion seems to be important.8

General recommendations that apply to all the pregnant RHD subjects:

1. Restricted physical activity
2. Restricted salt intake
3. Appropriate secondary prophylaxis
4. Avoiding inter-current infections
5. Assessment and selection for appropriate intervention

The following are recommendations for pregnant patients with severe valve disease.

### During pregnancy

**Medical therapy**

1. Anticoagulation should be given to pregnant patients with MS and AF unless contraindicated.
2. Use of beta-blockers as required for rate control is reasonable for pregnant patients with MS and HF symptoms.
3. Use of diuretics may be reasonable for pregnant patients with MS and HF symptoms.
4. ACE inhibitors and ARBs should not be given to pregnant patients with valve stenosis.

**Valve intervention**

1. Percutaneous mitral balloon commissurotomy is reasonable for pregnant patients with severe MS with valve morphology favorable for percutaneous mitral balloon commissurotomy who remain symptomatic with NYHA class III or IV HF symptoms despite medical therapy.
2. Severe MS and valve morphology not favorable for percutaneous mitral balloon commissurotomy only if there are refractory NYHA class IV HF symptoms.9
3. Severe AS only if there is hemodynamic deterioration or NYHA class III to IV HF symptoms.
4. Severe valve regurgitation only if there are refractory NYHA class IV HF symptoms.
5. Valve operation should not be performed in pregnant patients with valve stenosis in the absence of severe HF symptoms.
6. Valve operations should not be performed in pregnant patients with valve regurgitation in the absence of severe intraraftral HF symptoms.

### Before pregnancy

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<td>Exercise testing is reasonable in asymptomatic patients with severe AS and severe valve regurgitation before pregnancy.</td>
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<td>All patients with severe valve stenosis or regurgitation should undergo prepregnancy counseling by a cardiologist with expertise in managing patients with valvular heart disease during pregnancy.</td>
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### Valve intervention to be done in pregnancy

1. Severe AS
2. Symptomatic patients with severe MS
3. Percutaneous mitral balloon commissurotomy is recommended before pregnancy for asymptomatic patients with severe MS who have valve morphology favorable for percutaneous mitral balloon commissurotomy.
4. May be considered in the asymptomatic patient with severe MR (stage C) and a valve suitable for valve repair, but only after a detailed discussion with the patient about the risks and benefits of the operation and its outcome on future pregnancies.
5. Recommended before pregnancy for symptomatic women with severe valve regurgitation.

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1. Anecdotal evidence suggests that maternal outcomes are generally favorable for percutaneous mitral balloon commissurotomy. However, the long-term impact on fetal outcomes remains uncertain. A dedicated team of cardiologists, anesthetists, and obstetricians should be involved before pregnancy to ensure a comprehensive approach to patient care.

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It is advised that in all cases the decision about the mode of delivery should be finalized after a discussion with the cardiologist, anesthetist, and obstetrician of the patient.

During puerperium also patients are at risk of cardiac complications because of volume overload due to autologous transfusion. Patient should be examined for signs of heart failure and clinical observation for at least three days.

Mitril regurgitation

This entity doesn’t impose a great problem as mitral regurgitation is well tolerated during pregnancy. Pregnancy can alter examination findings, but rare mitral stenosis or regurgitation should undergo a clinical evaluation and TTE before pregnancy.

When pregnancy occurs with mitral regurgitation, treatment should include afterload reduction. ACE inhibitors and ARBs should be avoided. If endocarditis occurs and the infection is not rapidly controlled, mortality with medical therapy is high, and surgical therapy is indicated. If this occurs late in pregnancy, consideration of associated cesarean section is appropriate.

Tricuspid valve disease

Rheumatic fever may produce organic (primary) tricuspid regurgitation (TR), often associated with tricuspid stenosis (TS). Tricuspid regurgitation usually requires no specific therapy during pregnancy. Tricuspid stenosis is rare. If it is encountered, avoidance of intravascular volume depletion seems to be important.

General recommendations that apply to all the pregnant RHD subjects:

1. Restricted physical activity
2. Restricted salt intake
3. Appropriate secondary prophylaxis
4. Avoiding inter-current infections
5. Assessment and selection for appropriate intervention

The following may have an incidence of congenital heart disease as high as 20%, which may be reduced by correcting the outflow tract obstruction before pregnancy. If severe stenosis is recognized before pregnancy, balloon valvuloplasty or a surgical commissurotomy is recommended prior to conception. If pregnancy does occur in the presence of severe aortic stenosis, measures to avoid hypovolemia are particularly important. If severe symptoms persist despite adequate treatment, a balloon valvuloplasty or aortic valve surgery can be performed during pregnancy.

Aortic regurgitation

Rheumatic disease results in thickening, deformity, and shortening of the individual aortic valve cusps, changes that prevent their proper opening during systole and closure during diastole. Generally, it’s well-tolerated during pregnancy. If it is severe/symptomatic, or associated with LV dysfunction, valve surgery should be considered before pregnancy.5 If congestive heart failure occurs with pregnancy, treatment should include afterload reduction. ACE inhibitors and ARBs should be avoided. If endocarditis occurs and the infection is not rapidly controlled, mortality with medical therapy is high, and surgical therapy is indicated. If this occurs late in pregnancy, consideration of associated cesarean section is appropriate.

Recommendations

**Before pregnancy**

All patients with suspected valve stenosis or regurgitation should undergo a clinical evaluation and TTE before pregnancy.

All patients with severe valve stenosis or regurgitation should undergo pre-pregnancy counseling by a cardiologist with expertise in managing patients with valve heart disease during pregnancy.

Patients referred for a valve operation before pregnancy should receive pre-pregnancy counseling by a cardiologist with expertise in managing patients with valve heart disease during pregnancy about the risks and benefits of all options for operative interventions, including mechanical prosthesis, bioprosthesis, and valve repair.

**Follow-up**

Exercise testing is reasonable in asymptomatic patients with severe AS and severe valve regurgitation before pregnancy.

Valve intervention to be done in

1. Severe AS
2. Symptomatic patients with severe MS
3. Percutaneous mitral balloon commissurotomy before pregnancy for asymptomatic patients with severe MS, who have valve morphology favorable for percutaneous mitral balloon commissurotomy.
4. May be considered in the asymptomatic patient with severe MR (stage C) and a valve suitable for valve repair, but only after a detailed discussion with the patient about the risks and benefits of the operation and its outcome on future pregnancies.
5. Recommended before pregnancy for symptomatic women with severe valve regurgitation.

**During pregnancy**

Pregnant patients with severe valve stenosis or regurgitation should be monitored in a tertiary care center with a dedicated team of cardiologists, surgeons, anesthesiologists, and obstetricians with expertise in the management of high-risk cardiac patients during pregnancy.

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2. Use of beta-blockers as required for rate control is reasonable for pregnant patients with MS and HF symptoms.
3. Use of diuretics may be reasonable for pregnant patients with MS and HF symptoms.
4. ACE inhibitors and ARBs should not be given to pregnant patients with valve stenosis.

Valve intervention

1. Percutaneous mitral balloon commissurotomy is reasonable for pregnant patients with severe MS with valve morphology favorable for percutaneous mitral balloon commissurotomy who remain symptomatic with NYHA class III to IV HF symptoms despite medical therapy.
2. Severe MS and valve morphology not favorable for percutaneous mitral balloon commissurotomy only if there are refractory NYHA class IV HF symptoms.
3. Severe AS only if there is hemodynamic deterioration or NYHA class III to IV HF symptoms.
4. Severe valve regurgitation only if there are refractory NYHA class IV HF symptoms.
5. Valve operation should not be performed in pregnant patients with valve stenosis in the absence of severe HF symptoms.
6. Valve operations should not be performed in pregnant patients with valve regurgitation in the absence of severe intractable HF symptoms.
Severe AS:
- Aortic velocity >4.0 m/s or mean pressure gradient >40 mm Hg
- Valve area <0.8 cm² or valve area index <0.5 cm²/m²

Severe MS:
- Mitral valve area <1.5 cm² or valve area index <0.6 cm²/m²

Secondary prevention
In RHD it is defined as the continuous administration of specific antibiotics to patients with previous attack of rheumatic fever or a well-documented RHD.

Prophylaxis in pregnant patients with rheumatic heart disease is same as with the other cases except that oral sulfadiazine or sulfasoxazole are contraindicated in pregnancy due to teratogenic effects.

Abbreviations

References

Address for correspondence
Dr. Anand Sagar: Email: dranandsagar@hotmail.com

Abstract
Cardiovascular diseases (CVDs) are showing an escalation among the Indian population with a trend of reaching the younger age groups. It is now known to have a major share in the burden of diseases. A number of studies have been conducted time and again to find out the prevalence of CVDs and it has been found that they have a widespread prevalence in India, with regional variations. It is now affecting almost all sections of the society from young to old and most affluent to least affluent. Statistics also show an increased prevalence of CVDs in India as compared to other developing countries. Large scale and widespread incidences show downgrading of the cardiovascular health status of Indians and emergence of CVDs as a chronic manifestation across the population. Future trends predict that in the coming decade CVDs will inflict every section of the population irrespective of age, gender, economic status, and locale. Owing to such a large rate of prevalence, India is set to lose on it is productive population on accounts of CVD mortality and morbidity hampering the advantage of a positive demographic transition. This affects the country’s productivity owing to the disease-related economic burden in an otherwise beneficial phase of demographic transition. The need of the hour is to track down and closely monitor the prevalence of disease and encounter it with better intervention policies aimed at prevention, control, and treatment of CVD.

Key Words
- Prevalence
- Cardiovascular disease
- India
- Economic impact

Introduction
With the coming of the age of massive development, India has gone through dramatic lifestyle changes—moving from agrarian diets and active lifestyles to fast foods and sedentary lifestyles in a much shorter span of time than other nations. Consequently, mortality rates related to chronic non-communicable diseases like cardiovascular diseases (CVDs), coronary heart diseases (CHDs), diabetes and stroke have increased rapidly in the last decade with CVDs having a major share. Recent figures suggest that CVDs have outgrown the barriers of gender, locale, and economic status. Moreover, it is affecting the productive population imposing a huge socioeconomic burden. Future forecasts predict a continuing trend in the coming decades, where CVDs will lead to major loss of the