

stenosis in children and young adults is frequently encountered in developing countries, and up to 25% of patients are younger than 20 years of age. This disease has been called juvenile mitral stenosis.

Juvenile MS has been reported at very young age in the Indian subcontinent.

We report a case of juvenile MS at 3 year age, the youngest ever case reported

## Clinical and hemodynamic profiles of different etiologies of severe aortic stenosis



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**Aims and objectives:** To study the correlation between clinical presentation of severe aortic stenosis with echocardiographic measurements of left ventricle function and aortic valvular hemodynamics including LV strain amongst its etiologies.

**Materials and methods:** This study has been performed from June 2013 to December 2014 in cardiology department at Sri Satya Sai Institute of Higher Medical Sciences in Puttaparthi, Anantapur (Andhra Pradesh). Study included patients of severe AS coming to cardiology OPD and who were willing for the study and were eligible as per inclusion criteria. (Severe AS is defined as aortic valve area (by continuity equation)  $<1 \text{ cm}^2$ .)

**Results:** Total number of patients in study is 102. Male constitute 71 (70%) and females 31 (30%).

Mean age of presentation of rheumatic AS group 44 years, for bicuspid group is 25 years and for degenerative group is 59 years. Main symptoms of patient in all etiologies are dyspnoea, bicuspid group 96% rheumatic 100%, degenerative 72%. Angina as a predominant symptom was seen 21% of patients with degenerative group. Syncope was seen in 3 patients only. One in bicuspid group and 2 in degenerative group.

All three group of patients had average AVA (Bicuspid  $0.73 \pm 0.01 \text{ cm}^2$  Rheumatic  $0.76 \pm 0.12 \text{ cm}^2$  Degenerative  $0.74 \pm 0.08 \text{ cm}^2$ ) and AVAI (Bicuspid  $0.58 \pm 0.3 \text{ cm}^2/\text{m}^2$  Rheumatic  $0.48 \pm 0.09 \text{ cm}^2/\text{m}^2$  Degenerative  $0.46 \pm 0.03 \text{ cm}^2/\text{m}^2$ ) in the range of severe AS.

The average PG for (Bicuspid  $96 \pm 30 \text{ mm Hg}$  Rheumatic  $93 \pm 29 \text{ mm Hg}$  Degenerative  $93 \pm 31$ ) and MG (B  $62 \text{ R } 56 \pm 19 \text{ D } 55 \pm 16$ ) in the range of severe AS.

LV dimensions and LVH comparisons in etiologies.

	Bicuspid	Rheumatic	Degenerative	P value
LVIDd, mm	$43 \pm 7.17$	$59 \pm 6.71$	$47 \pm 6.7$	0.006
LVIDs, mm	$26 \pm 6.19$	$31 \pm 4.8$	$32 \pm 6.98$	0.001
LVSD, mm	$13 \pm 2$	$14 \pm 1.9$	$14 \pm 2.3$	0.005
LVPWd, mm	$12 \pm 2$	$13 \pm 1.6$	$14 \pm 1.7$	0.001
LVMI $\text{g}/\text{m}^2$	$137 \pm 13$	$148 \pm 12$	$155 \pm 9$	0.001

Comparison between SVI, LVEF and GLS between etiologies.

	Bicuspid	Rheumatic	Degenerative	P value
SVI $\text{ml}/\text{m}^2$	$60 \pm 4$	$59 \pm 3$	$56 \pm 5.2$	0.001
LVEF %	$67 \pm 8$	$64 \pm 9$	$59 \pm 9$	0.008
GLS %	$-18 \pm 6.4$	$-16 \pm 5$	$-16 \pm 4$	0.421

There was statistically significant difference between SVI and LVEF among the three groups. SVI and LVEF were higher in patients with bicuspid group compared to rheumatic and degenerative group

which suggest relatively better preserved LV systolic function in bicuspid group

LV GLS assessment results showed that GLS was decreased in all 3 group. Degenerative and rheumatic group have more decrease in GLS compared to bicuspid group but statistically not significant.

**Conclusions:**

1. The clinical spectrum of Aortic stenosis is broad. And patients with the same AVA can have different clinical profile, intracardiac hemodynamics and symptoms.
2. Patients with bicuspid aortic valve presents early, mean age 24 years, have less degree of LVH, less systolic and diastolic dysfunction.
3. While patients with rheumatic AS mean age of presentation was 44 years with predominant symptom as dyspnoea. Have significant degree of left ventricular hypertrophy and diastolic dysfunction.
4. Degenerative AS group mean age of presentation was 59 years and most common symptom dyspnoea 72%. Have significantly more left ventricular hypertrophy and diastolic dysfunction compared to other etiologies. They also have more elevated LV filling pressures ( $E/e'$ ) possibly because of higher age of patients and may have preexisting myocardial disease. Ischemic heart disease, diabetes mellitus type 2, systemic hypertension are common in this age group.
5. GLS assessment of patients with preserved LV systolic function LVEF  $>50\%$ . Showed significant abnormality in GLS suggestive of subclinical myocardial systolic dysfunction. Emphases the recommendation for routine use of strain assessment in patients of valvular AS.

## Association of VKORC1 gene polymorphism (1639G>A and 1173C>T) and acenocoumarol maintenance dosage in patients with mechanical heart valve



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**Background:** Acenocoumarol, an oral vitamin K antagonist, is a commonly prescribed anticoagulant following valve replacement surgery in India and has a narrow therapeutic index. This study aimed at investigating the prevalence of vitamin K 2,3 epoxide reductase C1 (VKORC1) gene polymorphism in south Indian patients and its influence on the inter-individual variability in response to acenocoumarol therapy.

**Methods:** The study cohort included 205 rheumatic heart disease patients with mechanical heart valves, on acenocoumarol therapy with a stable therapeutic INR between 2 and 3.5. VKORC1 1639G>A and 1173C>T genotypes were determined by PCR-RFLP method. Correlation between genotypes and the acenocoumarol dosage was evaluated.

**Results:** The most prevalent genotypes of VKORC1 in our study was wild homozygous GG for-1639G>A and CC for 1173C>T (57.6%). This was followed by heterozygous GA & CT at 36.1% and AA & TT at 6.3%. The Allele frequency for both G and C was 0.76 and for A

and T 0.24. There was complete linkage disequilibrium between -1639G>A and 1173C>T ( $r^2 = 0.98$ ,  $D' = 1.0$ ,  $LOD = 74.02$ ).

The wild type (GG and CC) of VKORC1 required a mean acenocoumarol dose of 2.78 mg, a moderately higher dose as compared to 2.39 mg for heterozygous GA and CT and 2.29 mg for homozygous AA and TT ( $p$  value 0.023). Only in male patients, bearing A allele of VKORC1 gene independently increased the odds of requiring a low dosage of acenocoumarol (adjusted OR 1.65 at 95% CI, class interval 0.99-2.76,  $p$  value-0.05). Both the wild type and alleles had a lower mean dose requirement when the patients were concomitantly prescribed Furosemide and Digoxin.

**Conclusions:** Heterozygous patients were more common in our study cohort compared to previous studies on Indian population and require a lower mean acenocoumarol dosage. Male patients bearing 'A' allele and patients concomitantly using Furosemide and Digoxin require a lower dosage. However co-prescribing low dose aspirin has no influence on the mean dose requirement.

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### C-reactive protein at admission as an independent predictor of major complications, need for urgent surgery and mortality in infective endocarditis patients: A protocol-based prospective analysis



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**Background:** Major complications in infective endocarditis (IE) remain high even in the current era of critical cardiac care. A reliable parameter that predicts complications and identifies patients requiring urgent surgery is lacking.

**Purpose:** Our aim was to study the value of baseline clinical, laboratory and microbiological parameters in IE patients. We focussed on the independent utility of inflammatory markers like leukocyte count (TLC), erythrocyte sedimentation ratio (ESR) and C-reactive protein (CRP) regardless of blood culture positivity or vegetation size.

**Methods:** This was a prospective study on consecutive IE patients (by modified Duke criteria) who were admitted to our tertiary care centre between 2012 and 2015. Predefined laboratory-microbiological sampling protocols and antibiotic-initiation protocols were followed. Peak levels of CRP and ESR in the first 3 days of admission were documented.

**Results:** Out of 101 patients treated, 71 patients with definite IE by Duke criteria were analysed. Mean age was  $43 \pm 16$  years. Blood cultures were positive in 55% ( $n = 39$ ) of which *Staphylococcus* was the most common. Major complications occurred in 72% ( $n = 53$ ) and in-hospital mortality was 31% ( $n = 22$ ). Mean ESR and CRP levels were  $102 \pm 31$  mm/hr and  $51 \pm 20$  mg/l respectively. In multivariate analysis, high CRP levels were independently predictive of poor outcomes ( $p < 0.001$ ) including major complications, embolic events and need for urgent surgery. A CRP  $>30$  mg/l predicted complications with a sensitivity of 91% and specificity of 82%. CRP level  $>40$  mg/l predicted mortality (relative risk = 8.12,  $p = 0.003$ ).

**Conclusion:** Major complications and mortality continue to remain high for IE. The interim results identify the independent value of CRP levels as a marker for early risk-stratification of IE patients. A biomarker based algorithm at admission may favourably impact treatment outcomes in IE. This study has set the stage for further randomised trials in this regard.

### A rare case of severe juvenile rheumatic triple valve disease



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**Case presentation:** A 13 years old girl presented to us with history of dyspnea on exertion since 15days. On examination she had a pansystolic murmur in mitral area and an ejection systolic murmur in aortic area. ECG done showed RBBB with features suggestive of biatrial enlargement. Patient did not give any previous history of rheumatic fever. Patient was diagnosed to have rheumatic heart disease and was advised to get 2D echocardiography. 2D echo showed dilated LA, RA and RV. The anterior mitral leaflet was thick and domed with restricted movement of PML and Doppler interrogation revealed a peak gradient of 31 mm Hg and mean gradient of 22 mm of Hg. Colour Doppler study showed severe mitral regurgitation. Aortic valve was thickened and was showing restricted movement. Doppler study showed mild AR with severe AS (peak gradient 82 mm of Hg and mean gradient of 47 mm of Hg). There was organic tricuspid valve disease with severe TR and severe TS (peak gradient 14 mm of Hg and mean gradient of 7 mm of Hg). Pulmonary valve was normal except for mild PR probably due to associated severe PAH. Patient also had LV concentric hypertrophy with normal LVEF. Final diagnosis of rheumatic heart disease with severe triple valve involvement with severe PAH was made and patient was sent for surgery.

**Discussion:** Rheumatic heart disease in India is characterized by rapid progression and death at younger age. Nearly 32% of patients die before the age of 20 years. In an autopsy study conducted on 144 children below the age of 18 years mitral valve was affected in 100% of cases. Involvement of aortic, tricuspid and pulmonary valves was seen in 63.89%, 54.86% and 12.5% respectively. Multivalvular disease was noted in 75.69% cases; but double valve and triple valve disease which possibly would have required surgery was present in only 8.33% and 3.5% cases respectively. This case showed severe rheumatic involvement of aortic, mitral, and tricuspid valve in a juvenile patient with is very rarely seen.

**Conclusion:** In India, rheumatic fever is endemic and remains one of the major causes of cardiovascular disease, accounting for nearly 25-45% of the acquired heart disease. This case demonstrates smoldering rheumatic activity that can be seen in Indian children at a very young age leading to triple valve involvement. Not only the cost of treatment in such patients are phenomenal, the morbidity and mortality is also enormous. Hence "Prevention is better than cure" is very apt for rheumatic fever.

### Percutaneous balloon mitral valvuloplasty during pregnancy: Retrospective analysis of pregnancy and neonatal outcomes



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**Introduction:** Rheumatic mitral valve stenosis is most common lesion and important contributor of mortality in pregnancy. Percutaneous balloon mitral valvuloplasty (BMV) is the intervention of choice during pregnancy. Given the procedural complexities and