

Clinical cases and conundrums in ARF & RHD

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South African giants of RHD

Disclosure: Nil

South African
cardiologists

John Barlow

Bongani Mayosi



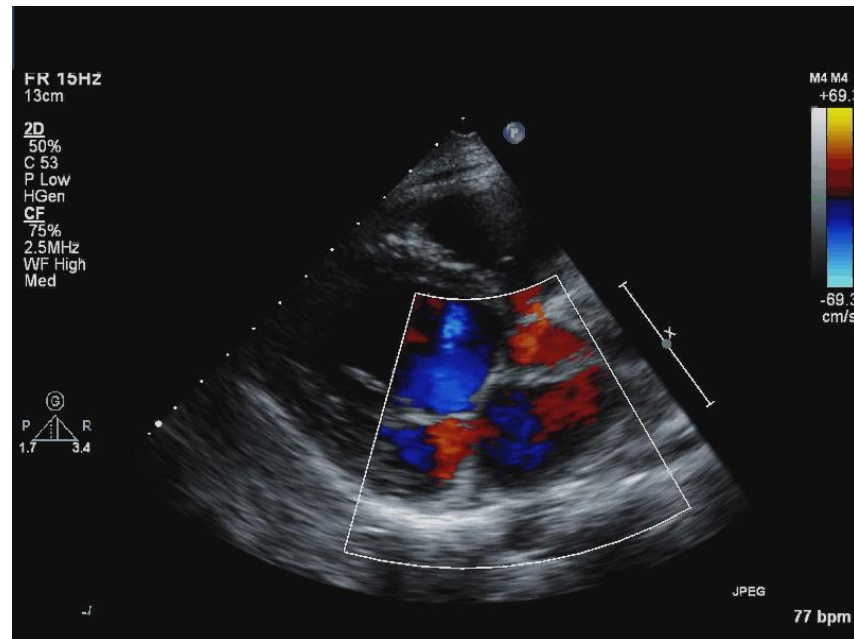
RHD difficult scenarios

1. Medical management of ARF
2. Fulminant mitral regurgitation
3. Chronic combined Mitral and aortic regurgitation
4. Endocarditis
5. Tricuspid valve regurgitation
6. Mitral stenosis in pregnancy

Case 1 mitral and regurgitation

11 yr old female

- Presented with ARF – fever, migratory polyarthrititis, mitral regurgitation, ESR 114 and recent evidence of streptococcal infection
- Management ?





ARF: medical treatment

- **Penicillin**: eradicate GAS, begin secondary prophylaxis
- Role bed rest – no good data since penicillin 1940s
- Salicylates – no influence on carditis
- Steroids – no influence on carditis: **RCTs 1950s & 60s**
- **IVIG - no influence on carditis**
- Cardiac medications:
 - proven symptoms /when ventricular dysfunction

NZ IVIG RCT Results: Presence of carditis

1 Year Follow Up n=59/61

	IVIG	Placebo	P value
Diagnosis	59%	63%	NS
			
1 year	26%	28%	NS

Voss L, Wilson N, Neutze J et al. Circulation
2001; 103:401-6

The natural history of carditis of ARF

In the absence of a recurrence of ARF

50% improvement within a year

- **clinically** *clinical improvement at 1 year*
May Wilson, Illingworth, UK US RFWP 1950s-60s
- **by echo** Voss et al Circulation

Proposed study for international multi-centre RCT medical treatment of ARF

Establish a platform group of centres

- Study 1: Corticosteroids vs placebo echo endpoints
- Study 2: steroids + hydrochloroquine vs corticosteroids
- Study 3: anti-inflammatory vs other immunomodulator
Interleukin

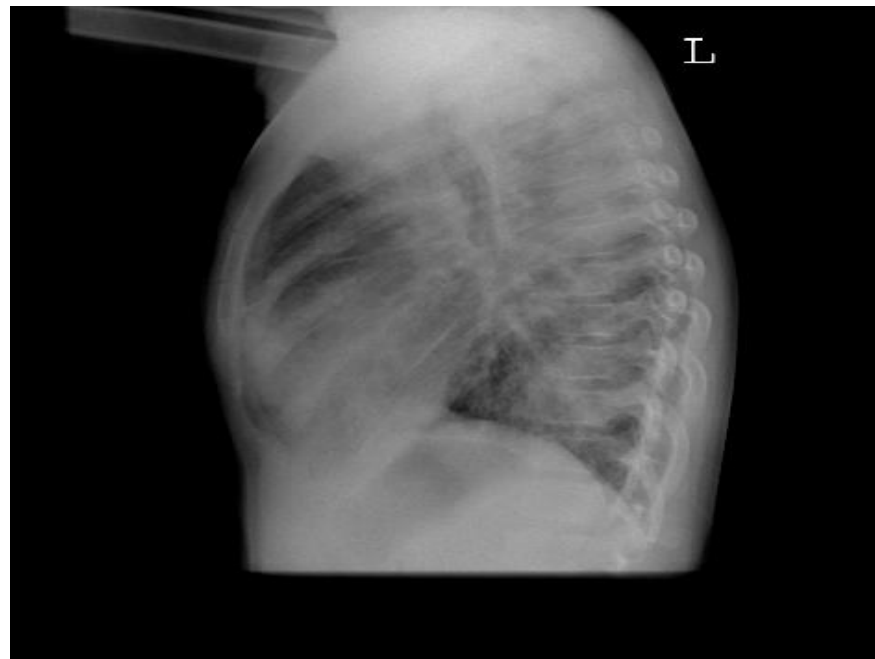
New treatments

New generation cardiology investigators

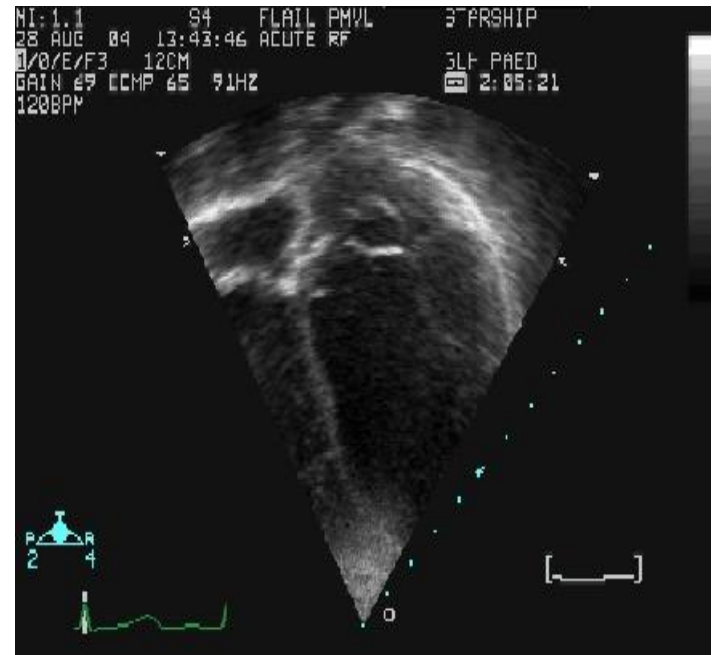
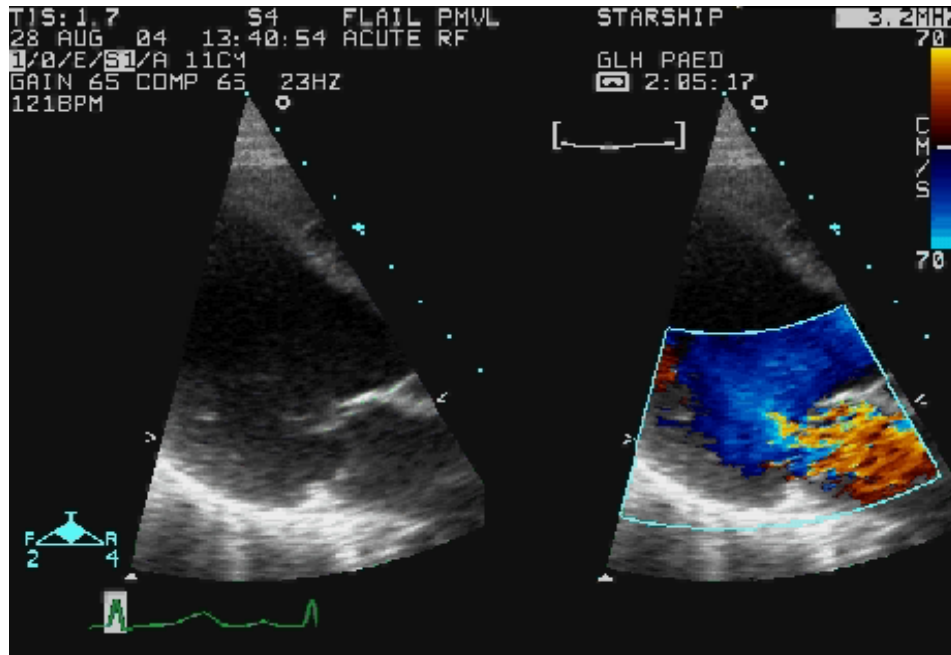
Case 2: 10 year old boy

- ❑ Played competitive rugby
- ❑ 2/7 cough
- ❑ 1/7 breathlessness/asthma
- ❑ Extremis – put on a ventilator
- ❑ Diagnosis: pneumonia

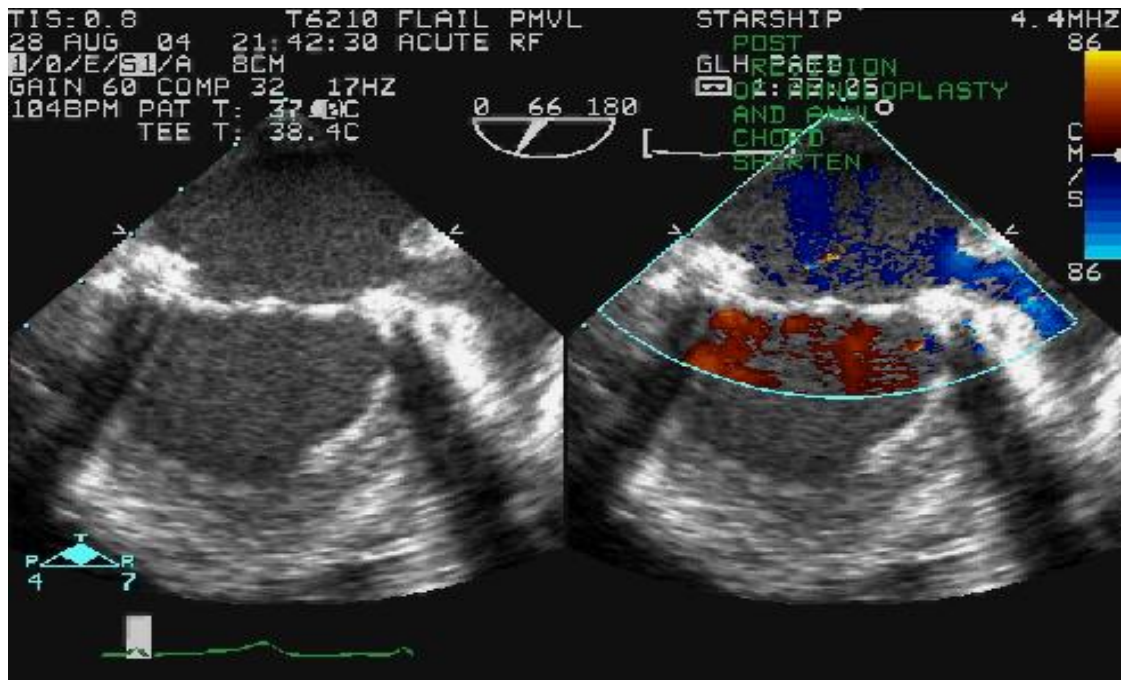
Pulmonary oedema



Echocardiogram



Transoesophageal post repair



Fulminant ARF

Acute rupture of chordae tendinae – young children

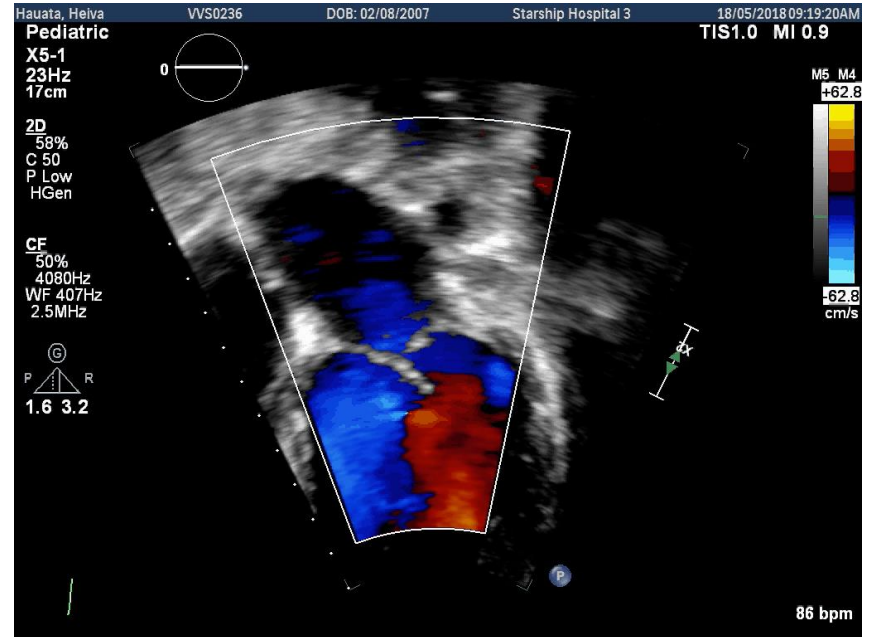
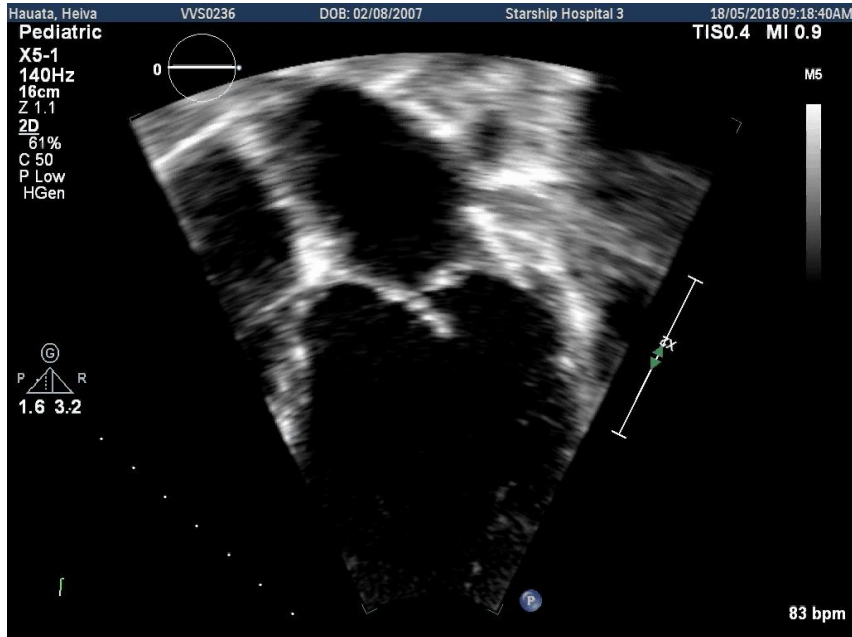
- ❑ Acute mitral regurgitation
- ❑ Rapid rise in left atrial pressure
- ❑ Pulmonary oedema = fulminant ARF (misdiagnosed as pneumonia)
- ❑ Cardiac surgery life saving

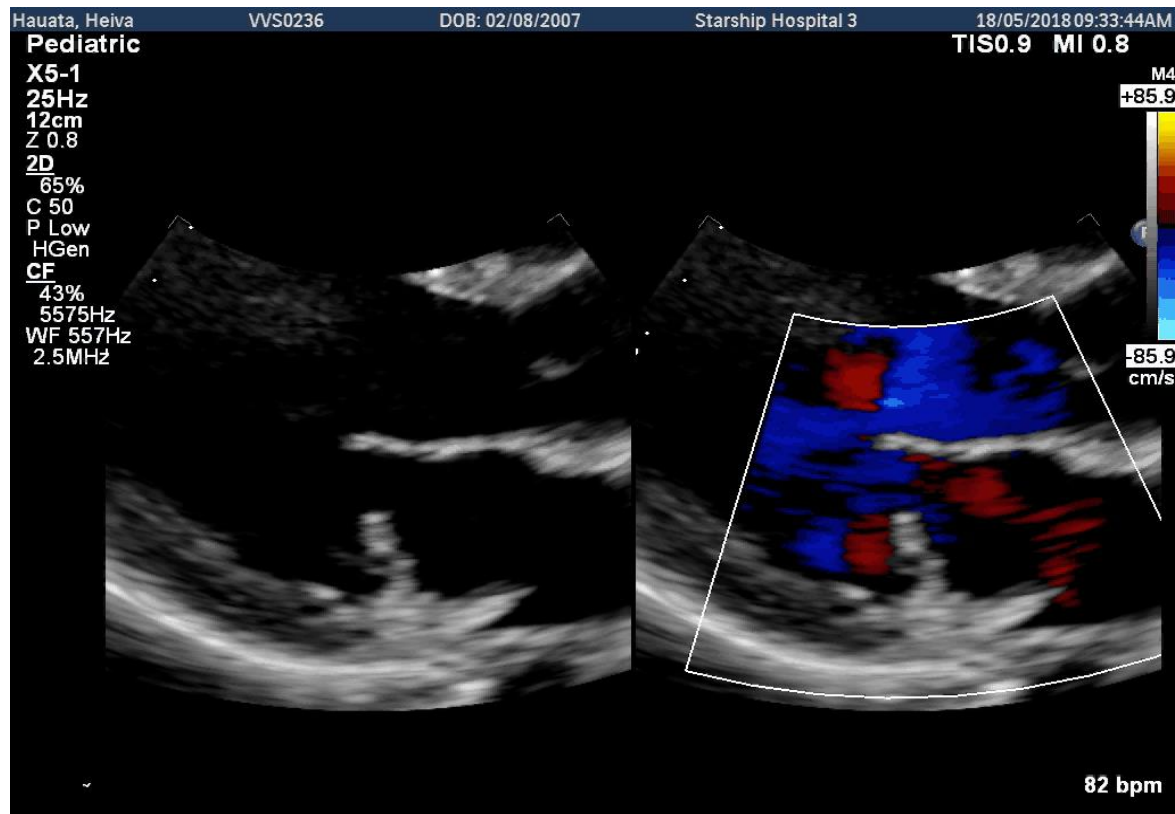
South Africa: De Moor M, et al. Rupture of tendinous chords during acute rheumatic carditis in young children. Int J Cardiol. 1986;12(3):353-7

Anderson Y, et al. Fulminant mitral regurgitation due to ruptured chordae tendinae in acute rheumatic fever. J Paediatr Child Health. 2008; 44:134-7

- ❑ Occurs in 1% of 1st episodes of ARF in New Zealand
- ❑ Not a risk for Chronic RHD

Flail PMVL





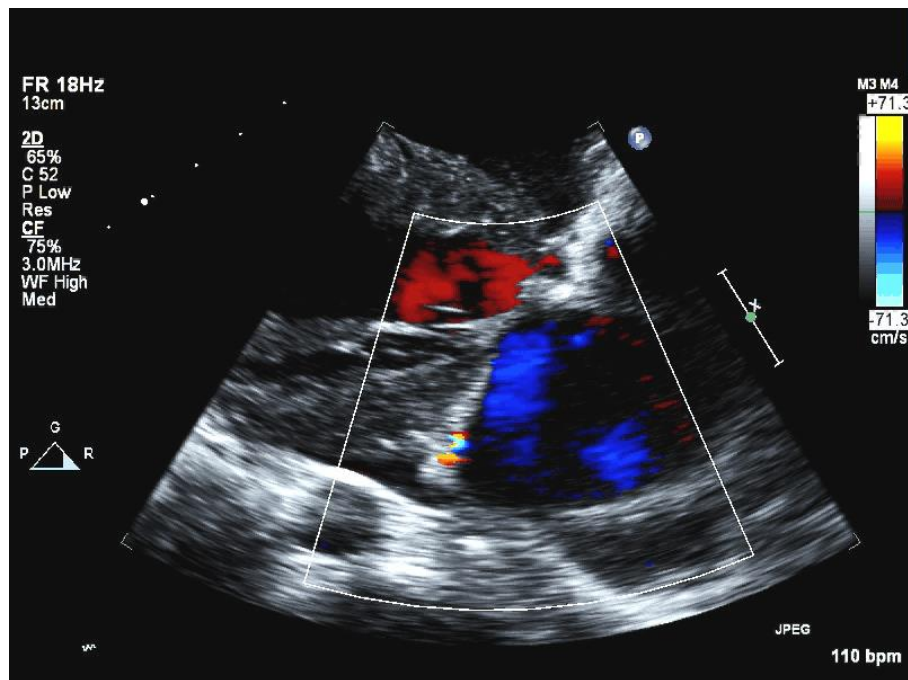
Case 3

Combined mitral and aortic regurgitation

14 yr old female

- ❑ Presented with ARF - arthritis of the small joints, mitral regurgitation, ESR 114 and recent evidence of streptococcal infection
- ❑ 86 kg, Blood pressure 102/45. MR and AR murmurs
- ❑ Observed acute phase, asymptomatic but 6 months later CRHD assessment

Case 3



Chronic RHD in adults

Indications for operation

**2014 AHA/ACC Guideline 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

*Developed in Collaboration With the American Association for Thoracic Surgery,
American Society of Echocardiography, Society for Cardiovascular Angiography and Interventions,
Society of Cardiovascular Anesthesiologists, and Society of Thoracic Surgeons*

JACC. 2014;63:e57-e185

>939 references

Isolated MR

isolated AR

2017 ESC/EACTS Guidelines for
the management of valvular heart
disease. *Eur Heart J.*
2017;38:2739-91.s 2017

Combined mitral and aortic regurgitation

□ LV function

- LVEDD 7.3 cm (Z-score = 6.24)
- LVESD 5.14 cm (Z-score = 5.57) FS 30%
- EDV-AL 279. ml (Z-score = 7.39) ESV-AL 134.ml (Z-score = 7.18), EF-AL 52.%

Meets indication for operation for severe MR **LVESD** > 4cm (AHA/ACC 2017,
> 4.5cm ESC)

Meets indication for operation for severe AR **LVEDD** > 6.5 cm (AHA/ACC)

Meets indication for operation by impaired LV function

Combined mitral and aortic regurgitation

What are the **indications** for operation for combined MR and AR?

What are the **outcomes** for combined MR and AR?

Combined (multiple) AR and MR in adults

AHA/ACC guidelines ESC guidelines 2017

Gaps in evidence

'More data on the natural history and the impact of intervention on outcome are required to better define the indications for intervention'.

MIXED Valve disease eg mitral stenosis/mitral regurgitation

For patients with mixed valve disease, there is a paucity of data on the natural history of such coexistent conditions.

Ventricular Function Before and After Surgery for Isolated and Combined Regurgitation in the Young

Thomas L. Gentles, MBChB, FRACP, A. Kirsten Finucane, MBChB, FRACS, Bo Remenyi, MBBS, FRACP, Alan R. Kerr, MBChB, FRACS, and Nigel J. Wilson, MBChB, FRACP

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- Group 1 severe AR n = 13
- Group 2 severe MR 21
- Group 3 combined AR and MR 13
all with LV dilatation. Evidence
afterload elevation in groups 1
and 3
- Post op to 18/12

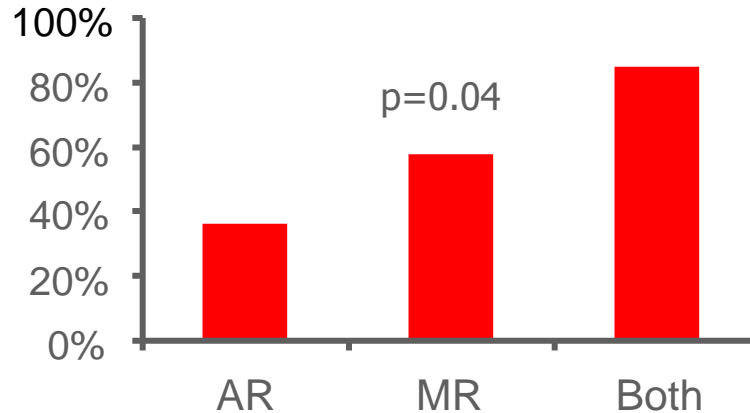
Table 2. Preoperative Left Ventricular Geometry and Mechanics

Variable	Group I (AR)	Group II (MR)	Group III (Both)
EDD z	5.2 ± 1.4 ^a	5.0 ± 1.9 ^a	5.4 ± 1.9 ^a
ESD z	4.5 ± 1.2 ^a	4.1 ± 2.1 ^a	5.2 ± 1.9 ^a
EDth/D z	-1.2 ± 1.2 ^a	-1.0 ± 1.4 ^a	-1.3 ± 0.9 ^a
Mass z	4.4 ± 1.5 ^a	4.3 ± 2.2 ^a	5.4 ± 1.8 ^a
Sphericity z	2.2 ± 1.6 ^a	3.7 ± 2.0 ^a	2.8 ± 1.6 ^a
ESFSc z	2.0 ± 2.1 ^{a,b}	0.2 ± 1.9	1.7 ± 2.3 ^a
FS z	-0.9 ± 1.7	-0.3 ± 1.9	-1.4 ± 1.6 ^a
VCFc z	-2.6 ± 1.3 ^{a,b}	0.1 ± 1.9	-2.2 ± 1.6 ^{a,b}
SVI	-1.9 ± 1.0 ^{a,b}	0.2 ± 1.4	-1.6 ± 1.3 ^{a,b}

Aortic and mitral valve surgery - postoperative LV dysfunction

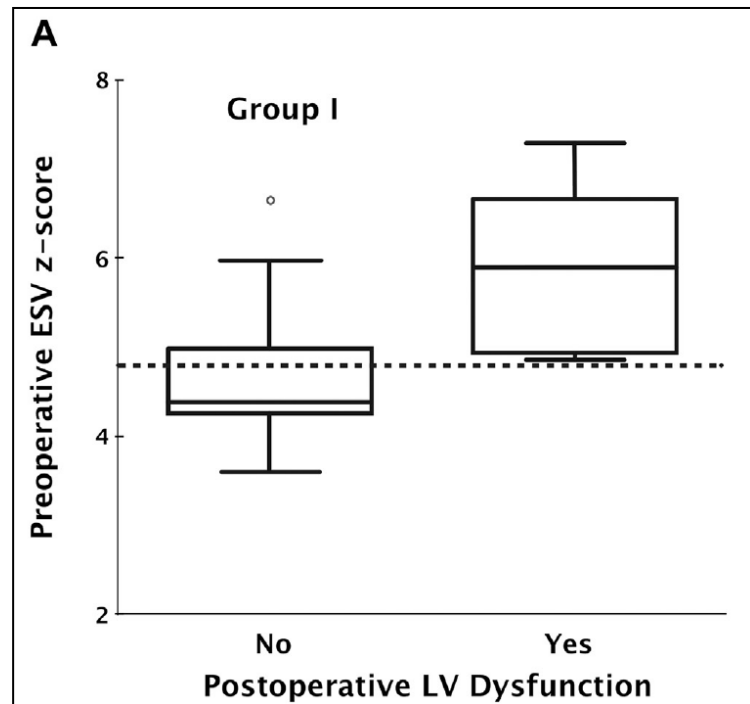
- Echocardiogram median 7 months after operation

LV dysfunction
(EF z-score < -2)

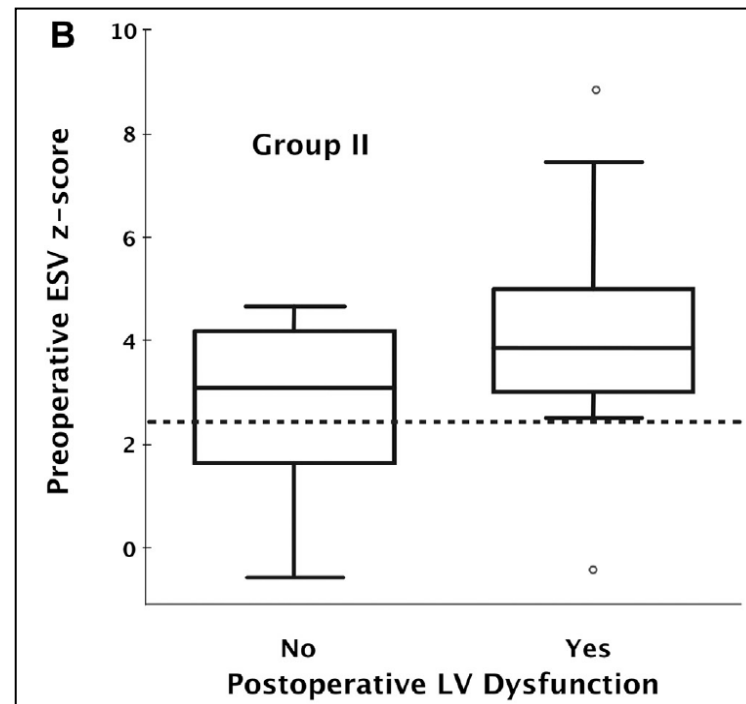


LV dysfunction more common after AR & MR surgery

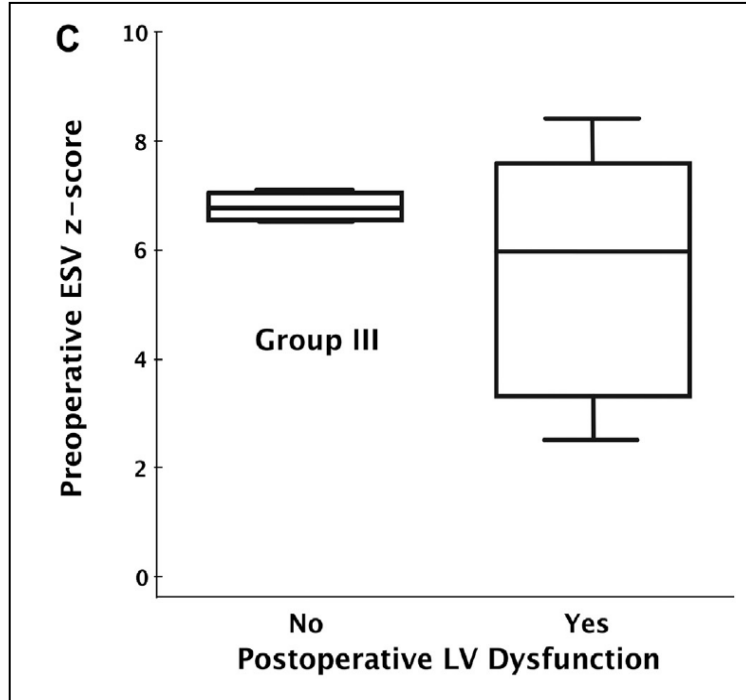
Isolated aortic regurgitation



Mitral regurgitation



Combined AR and MR

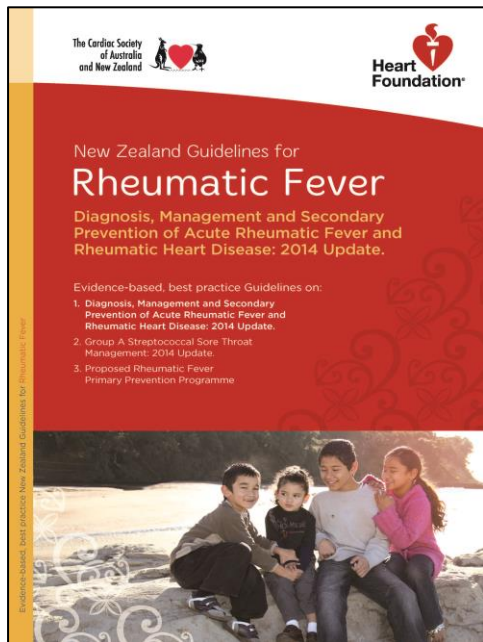


Conclusions

- Combined MR and AR were at the highest risk of postoperative LV dysfunction
- Indications for operation based on MR not AR

Indications for cardiac surgery in the young: MR

www.heartfoundation.org.nz



New Zealand RF/RHD guidelines
2014 update

A. Severe MR with symptoms of breathlessness; or

B. Asymptomatic MR and one of the following:

- Impaired LV function $<60\%$
- LVESV z-score >2.5
- Pulmonary hypertension $>50\text{mmHg}$

Case 4: Endocarditis

- ❑ 25 year old female, Pacific Island ethnicity
- ❑ CVA resulting in right sided weakness and aphasia
- ❑ CT angiogram: Left middle cerebral artery territory emboli

Continued fevers and +ve blood cultures *Streptococcus sanguis*

- ❑ ? Endocarditis
- ❑ 95kg poor dentition, pansystolic murmur

Echo: 2D moderate posteriorly directed MR

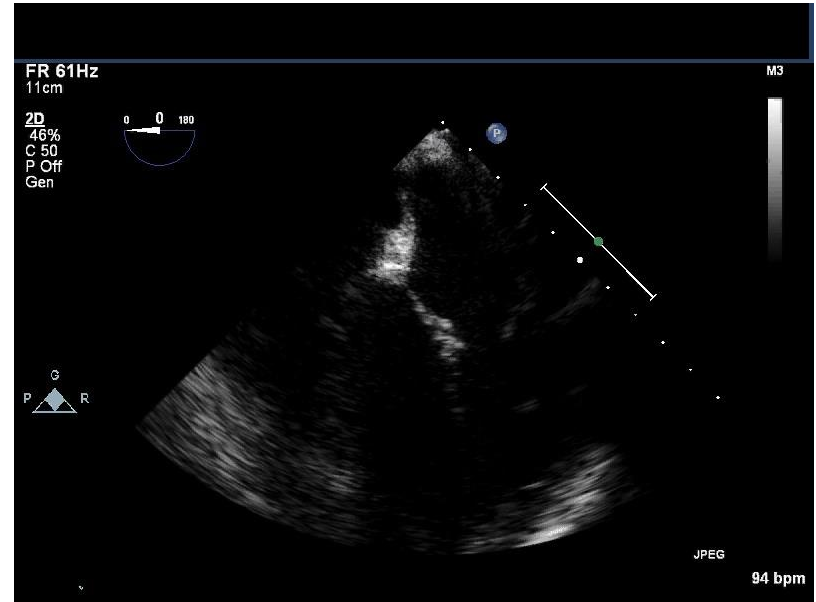
thickening AMVL? Mass?

Fibroelastoma ?

no vegetations seen

Transoesophageal Echo

- ❑ Thickened AMVL
- ❑ Eccentric posteriorly directed MR
- ❑ Vegetations diffusely involving the AMVL
- ❑ No AR, no TR



Differential diagnosis MR with endocarditis

Child

- ❑ RHD
- ❑ Mitral valve prolapse
- ❑ Congenital MV pathology
 - mitral valve clefts
 - accessory mitral valve leaflets/scallops
 - double orifice mitral valve

Young adult

- ❑ Barlow's disease 1963
- ❑ Myxomatous MV disease
- ❑ Fibroelastoma
- ❑ Connective tissue disease
- ❑ RHD
- ❑ Congenital MV pathology

Remenyi B, Gentles T Congenital Mitral valve lesions
Ann Ped Cardiol 2012

Differential diagnosis of mitral pathology

Child

- ☒ RHD
- ☐ Mitral valve prolapse
- ☐ Congenital MV pathology

Young adult

- ☐ Barlow's disease
- ☐ Fibroelastoma
- ☐ Connective tissue disease
- ☐ Barlow's disease
- ☐ Congenital MV pathology
- ☒ RHD

Implications

- 1) Secondary prophylaxis and duration
- 2) Dentition
- 3) Serial mitral and LV function

2012 WHF Echo criteria for RHD: evidence-based guidelines

Echocardiographic criteria for individuals aged >20 years

Definite RHD (either A, B, C, or D):

- A) Pathological MR and at least two morphological features of RHD of the MV
- B) MS mean gradient ≥ 4 mmHg*
- C) Pathological AR and at least two morphological features of RHD of the AV, only in individuals aged < 35 years†
- D) Pathological AR and at least two morphological features of RHD of the MV

Nature Reviews Cardiology 9, 297-309 (May 2012)

The 2012 WHF Guidelines define the **minimum** echo criteria for diagnosis of RHD in individuals without a clear history of ARF.

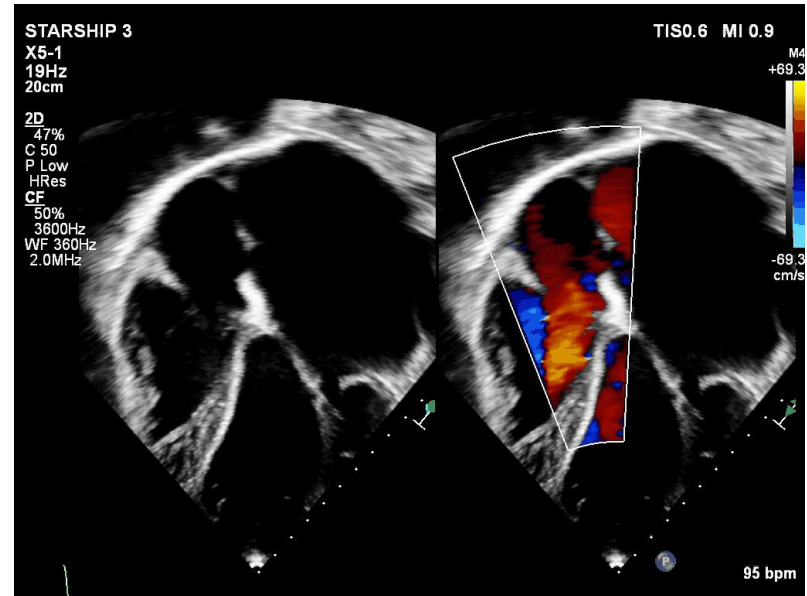
1. In the setting of RHD screening programs
2. As a clinical tool when a diagnosis of RHD is being considered or

Nature Reviews Cardiology **9**, 297-309 (May 2012)

Scenario 5: Tricuspid valve disease in RHD

“We’ll fix it”

Cardio-surgical
meeting



Tricuspid valve disease in RHD

pathology

Secondary to left heart lesions

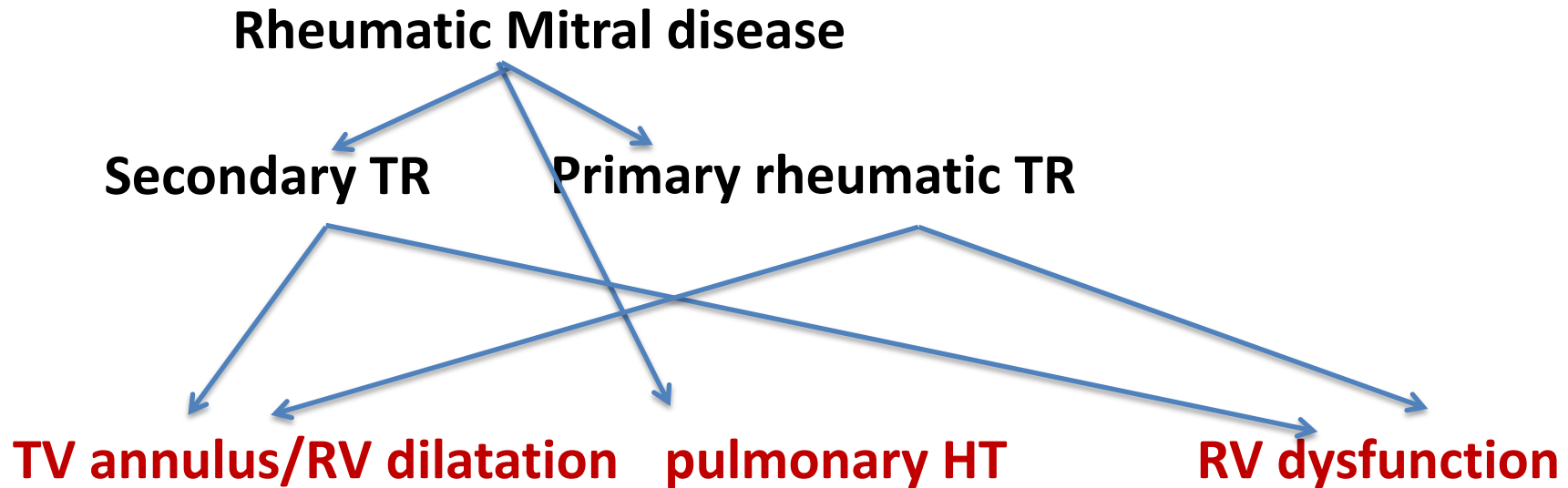
The pathological process of **annular dilatation** occurs primarily at the anterior and posterior portions of the annulus

Primary rheumatic involvement

- ❑ Anterior and posterior leaflet chordae elongated
- ❑ Leaflets can shrink, thick rolled edges
- ❑ Septal leaflet restricted, tethered against ventricular septum

Tricuspid regurgitation in RHD

pathophysiology



Tricuspid valve disease in RHD

Preoperative TTE evaluation

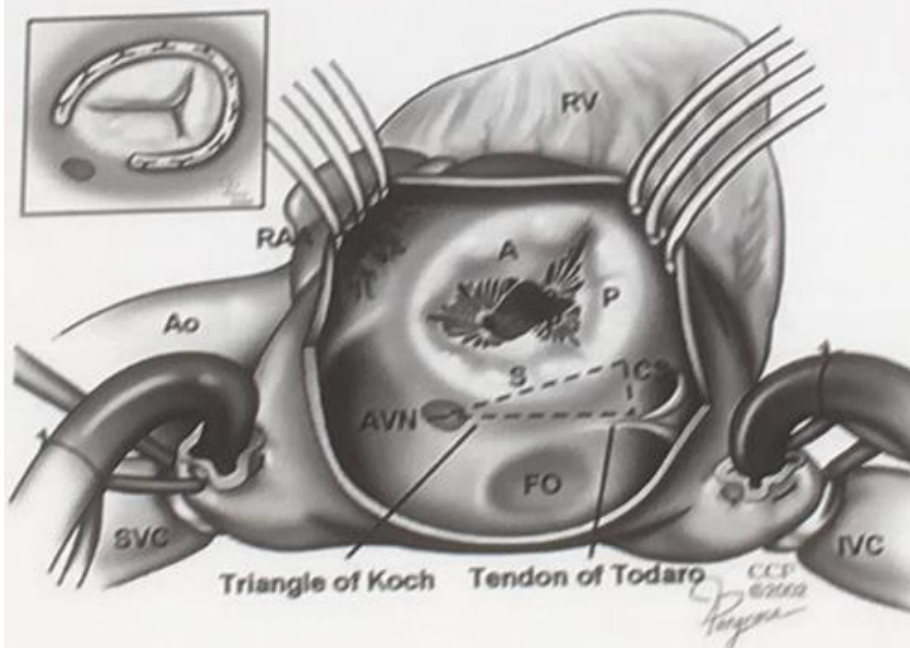
- ❑ objectively document TV dilatation
- ❑ Severity of TR regurgitation
- ❑ tethering effect on the chordae tendinae

'There is no reliable method to judge how much of the TR is reversible when the left heart valve lesions are corrected'

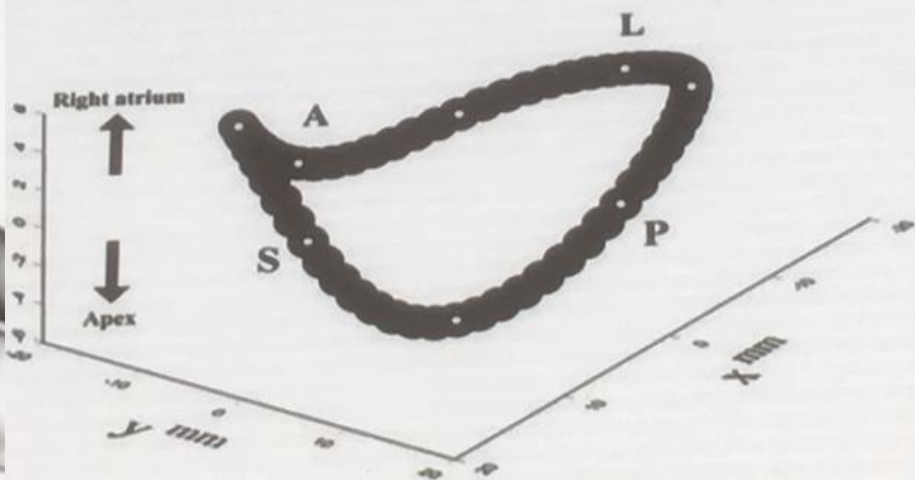
Antunes MJ, Barlow JB. Management of tricuspid valve regurgitation.

Heart. 2007;93(2):271-6.

Anatomy



3D Shape of TV Annulus



Vahanian A, et al. Guidelines on the management of valvular heart disease. **Eur Heart J. 2012;33:2451-96**

“should be considered in patients with mild or moderate secondary TR with dilated annulus (≥ 40 mm or > 21 mm/m²) undergoing left-sided valve surgery” Class IIa.

Antunes MJ, et al. Management of tricuspid valve regurgitation: Position ESC/CVS Surg on Valvular Heart Disease. **Eur J Cardiothorac Surg. 2017;52; 1022-1030.**

“address severe TR” Class I

“Trend for addressing moderate TR” Class IIa.

Case 6: Mitral stenosis in Pregnancy

- ❑ 21 year old Pacific Islands nulliparous
- ❑ Known RHD IM BPG prophylaxis
- ❑ Echo:
 - moderate-severe mitral stenosis
 - moderate MR
 - mild TR
- ❑ no significant symptoms
 - played basketball
- ❑ “advised against pregnancy”
- ❑ lost to follow-up



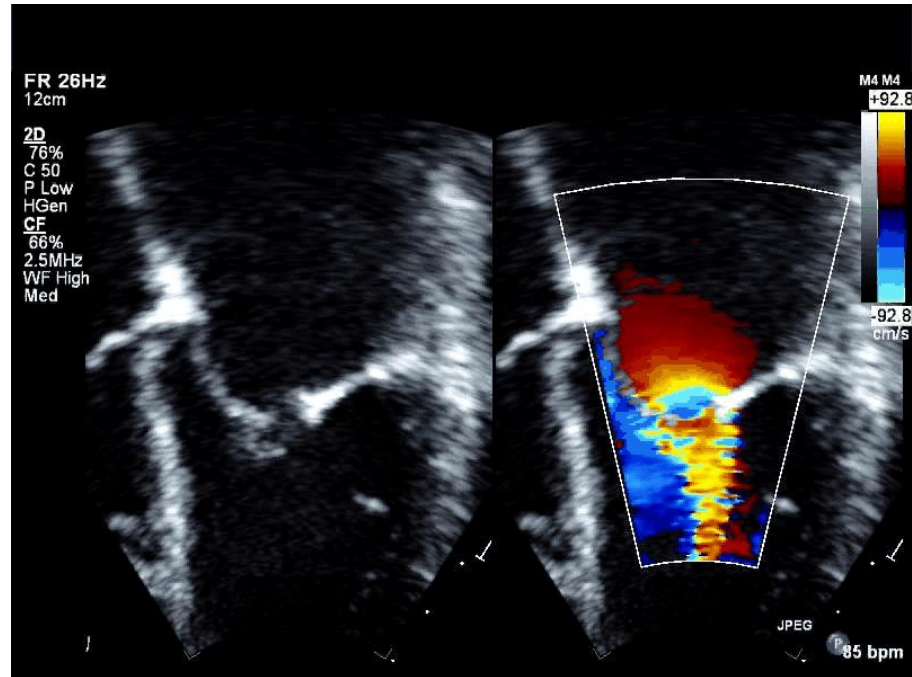
- ❑ 28/40 weeks pregnant
 - nocturnal cough, not productive, no fever
- ❑ short of breath walking upstairs
- ❑ 3 pillow orthopnoea
- ❑ pulse 105, regular
- ❑ BP 108/74
- ❑ murmurs MS, MR



New Zealand

- ❑ 33 weeks - Auckland
- ❑ pulse 86, sinus rhythm
- ❑ BP 113/69
- ❑ Echo:
 - MVA 0.8 cm²
 - MV gradient 22 mmHg
 - mild MR
 - EF 68%
- ❑ Rx metoprolol
- ❑ Rx Frusemide

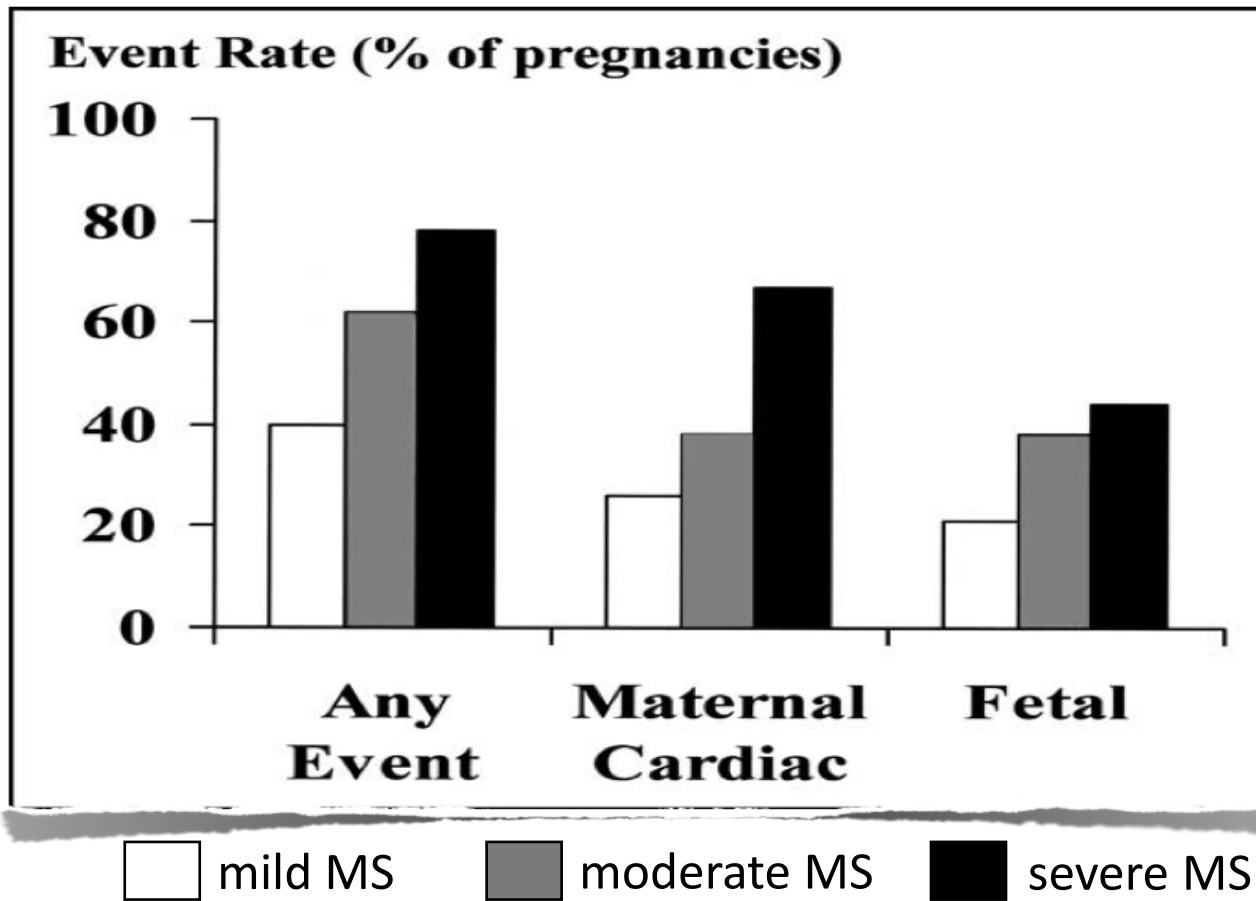




Maternal complications in women with Mitral stenosis

□ Canadian study 80 pregnancies in 73 women with MS

- mild (n=42) moderate (n=29) severe (n=9)
- NYHA class I (89%) and II (11%)
 - 40% get worse pregnancy
- Cardiac medications
 - pre-pregnancy 13% during pregnancy 68%

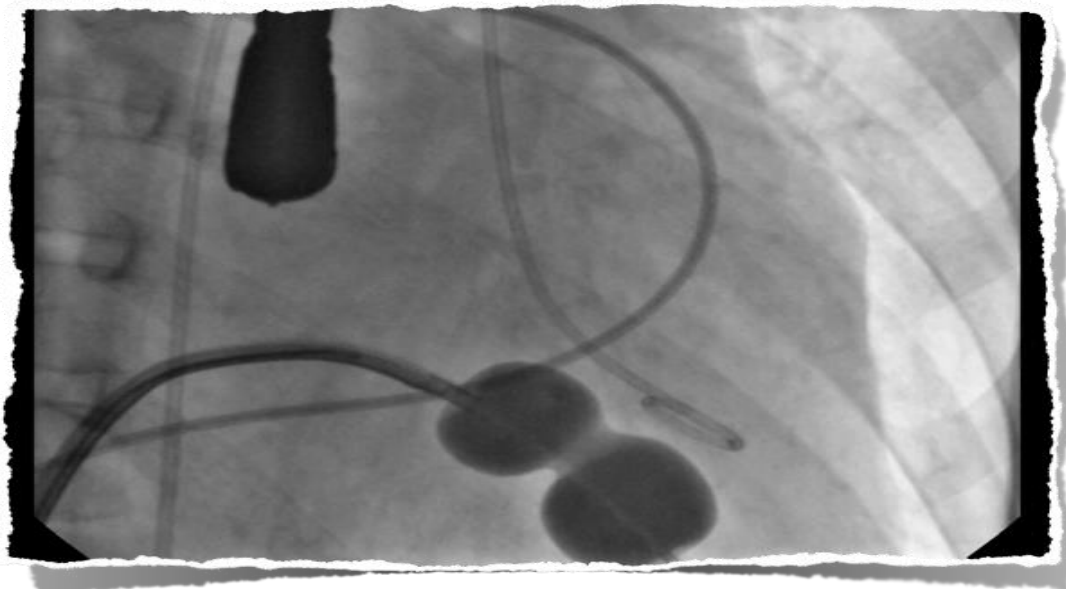


CASE: BMV and beyond

- ❑ Post-BMV
 - MVA >2cm², MV gradient 9
- ❑ Discharged - family in Auckland
- ❑ SROM @ 38+2 weeks
- ❑ 6 hours later - baby!
- ❑ Back to Pacific Islands 6 weeks postpartum

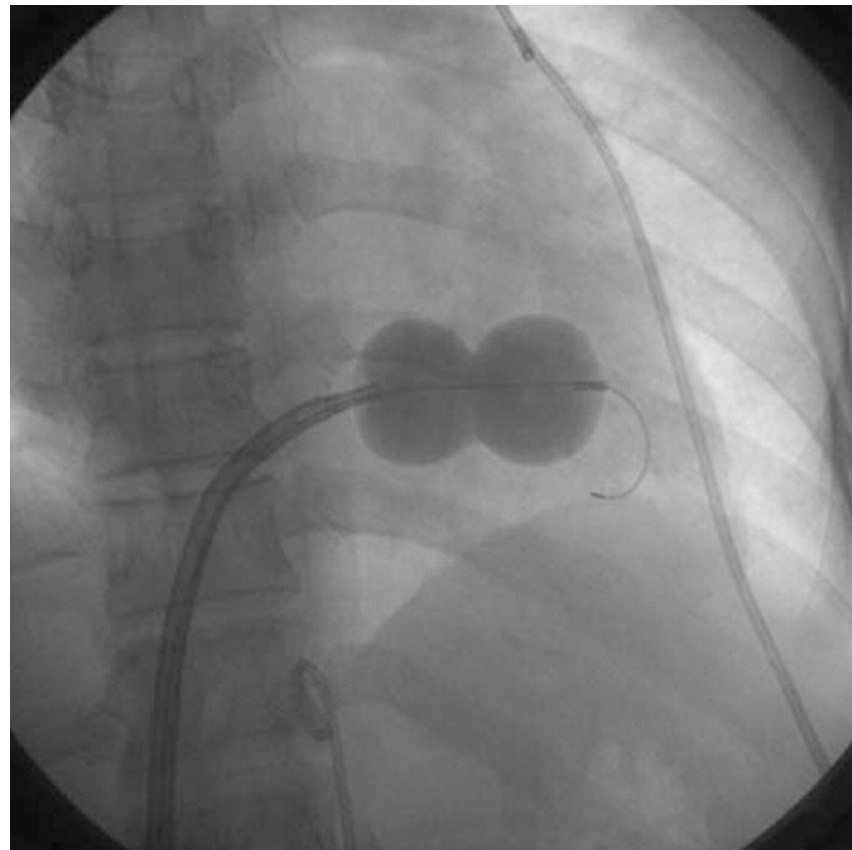
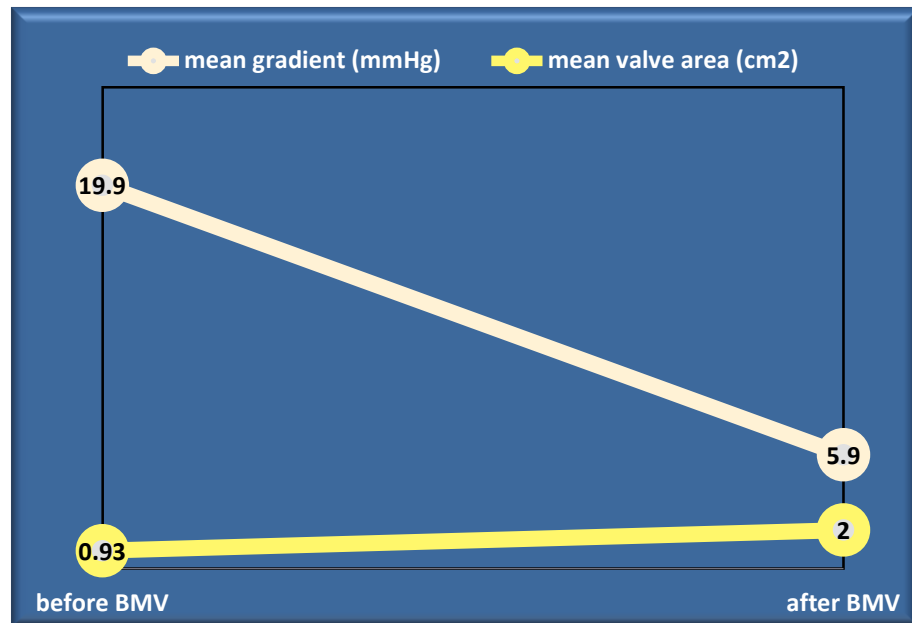
Plan

- ❑ Family planning
- ❑ Continue BPG
- ❑ Dental care



BMV in pregnancy

15 studies* of BMV in 455 pregnancies



BMV in pregnancy

15 studies* of BMV in pregnancy 455 pregnancies

Complications	n (%)
Maternal	
Mitral regurgitation	4 (0.9)
Cardiac tamponade	2 (0.4)
Suboptimal improvement MS	6 (1.3)
Fetal	
Stillbirth or fetal death	10 (2.2)
neonatal death	2 (0.4)
preterm delivery	10 (2.2)

Difficult Cases & scenarios

- ARF: need new medical managements
- ARF: Fulminant mitral regurgitation: need urgent cardiac surgery
- Combined MR and AR: Risk for LV dysfunction
- Endocarditis in the young: is this RHD ?
- Severe TR in RHD: fix it
- Mitral stenosis in pregnancy: Good outcomes with PMBV - need cardiac catheterization