

# Rheumatic Heart Disease

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# Pathology

## Overview:

- Definitions
- Epidemiology
- Pathogenesis & Anatomical Pathology
- Prognosis

# Definition

- Rheumatic fever is an acute immune-mediated multisystem disease that follows an episode of streptococcal (group A beta hemolytic) pharyngitis.
- It often involves the heart – **acute rheumatic carditis**.
- Rheumatic heart disease is cardiac inflammation and scarring triggered by an autoimmune reaction to infection by group A streptococci.
- Acute – pancarditis (myocardium, endocardium, epicardium)
- Chronic – valvular fibrosis manifesting as stenosis and /or insufficiency.

# Epidemiology

- Highly prevalent in PNG.
- Occurs in 3% of patients with group A beta haemolytic streptococcal pharyngitis
- In the last 50 years incidence in more developed countries is low due to:
  - Improved socio-economic status, rapid diagnosis and treatment of acute streptococcal pharyngitis and improvement in general health & hygiene.
- Remains a significant public health problem in other less developed countries.

# PNG Statistics

PNG Med J 1997 Sep-Dec;40(3-4)157-167

## Operation Open Heart 1995: lessons learned and thoughts for the future

TERI MILLANE<sup>1</sup>, JOHN VINCE<sup>2</sup>, ALAN GALE<sup>3</sup>, GRAHAM NUNN<sup>1</sup>, RUSSELL LEE<sup>4</sup> AND RICHARD HAWKER<sup>5</sup>

Royal Alexandra Hospital for Children, Sydney, Australia, University of Papua New Guinea, Port Moresby, Sydney Adventist Hospital, Australia, 393 Medical Centre, Mosman, Australia and IDP Education Australia, Canberra

TABLE 2

MAIN DIAGNOSTIC CATEGORIES BY CLINIC CENTRE

Centre	RHD Adult	CHD Adult	RHD Child	PDA	VSD	Tetralogy of Fallot	ASD	Complex CHD Child	Other cardiac
Goroka	18	3	2	9	15	4	2	2	9
Madang	1	0	3	1	24	4	1	8	8
Mount Hagen	6	1	1	7	9	2	1	1	7
Port Moresby	2	3	9	3	16	4	4	8	11
Rabaul	0	1	0	7	16	6	1	3	4
<b>Total</b>	<b>27</b>	<b>8</b>	<b>15</b>	<b>27</b>	<b>80</b>	<b>20</b>	<b>9</b>	<b>22</b>	<b>39</b>

RHD = rheumatic heart disease  
 CHD = congenital heart disease  
 PDA = patent ductus arteriosus  
 VSD = ventricular septal defect  
 ASD = atrial septal defect

# Clinical Presentation

Clinical diagnosis based on Jones Criteria.

Documentation of an increased anti-streptolysin O (ASO) titre in the presence of 2 major or 1 major and 2 minor criteria

Major Criteria	Minor Criteria
Polyarthrititis	Fever
Carditis	Increased ESR
Chorea (Sydenham chorea)	Leucocytosis
Erythema induration (subcutaneous nodules)	Prolong PR interval in ECG
Erythema marginatum	History of rheumatic fever or rheumatic heart disease

# Pathogenesis

- Rheumatic fever results from humoral and cellular-mediated immune responses occurring 1-3 weeks after the onset of streptococcal pharyngitis.
- Streptococcal proteins share similarities with host proteins recognised by immune system
  - especially bacterial M-proteins and human cardiac antigens such as myosin and valvular endothelium.
- **Anti-myosin antibody recognizes laminin**, an extracellular matrix protein, which is part of the valve basement membrane structure. Ag-Ab Rxn induces inflammatory response.
- Inflammatory response results in destruction of valves

# Pathogenesis

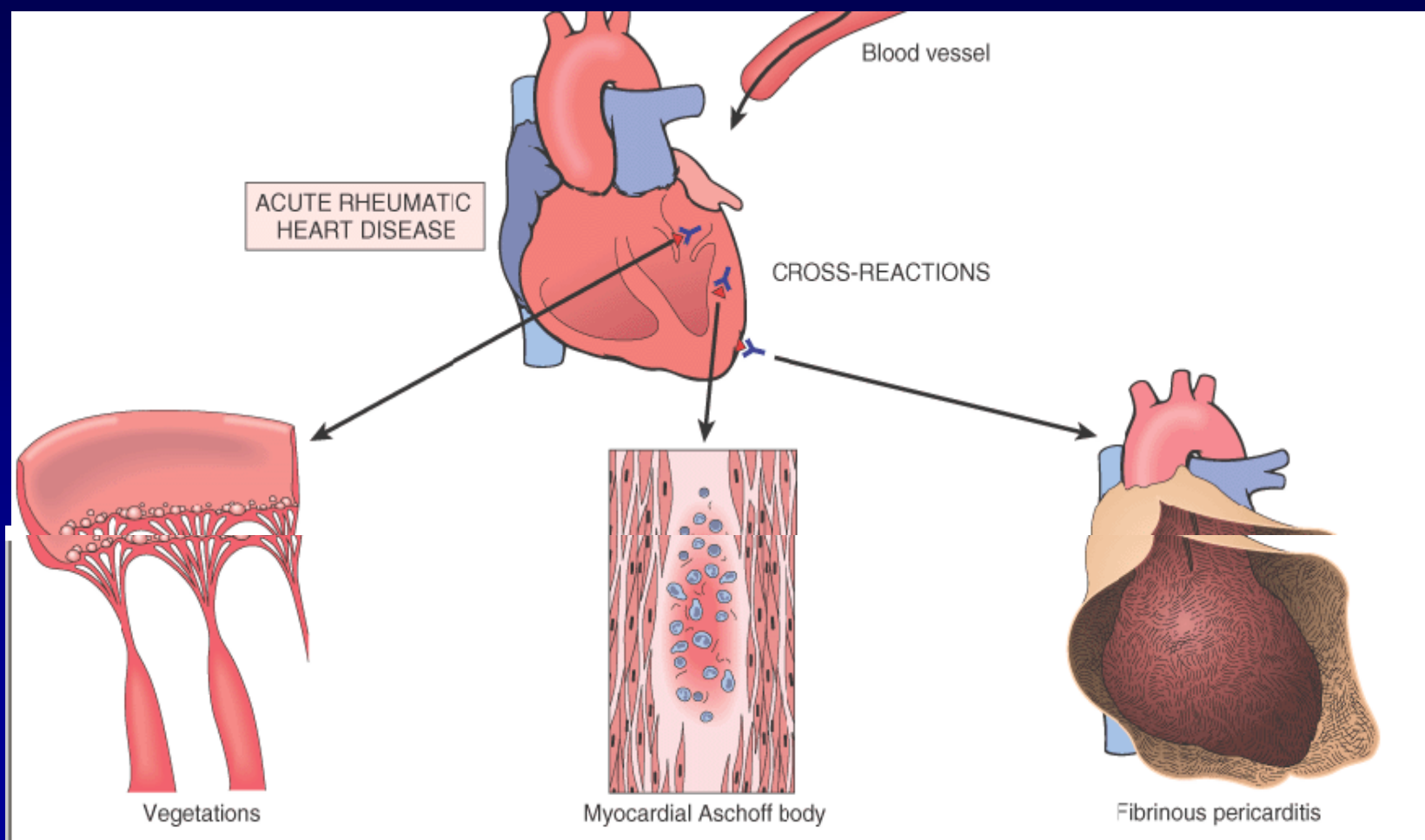
- T-cells that are responsive to the streptococcal M-protein infiltrate the valve through the valvular endothelium, activated by the binding of anti-streptococcal carbohydrates with release of tumor necrosis factor (TNF) and interleukins.
- **Rheumatic heart disease** - pancarditis, with inflammation of the myocardium, pericardium, and endocardium.
  - Carditis occurs in 40-50% of patients on the first attack
  - Pericarditis occurs in 5-10% of patients with rheumatic fever; isolated myocarditis is rare.



# Pathogenesis

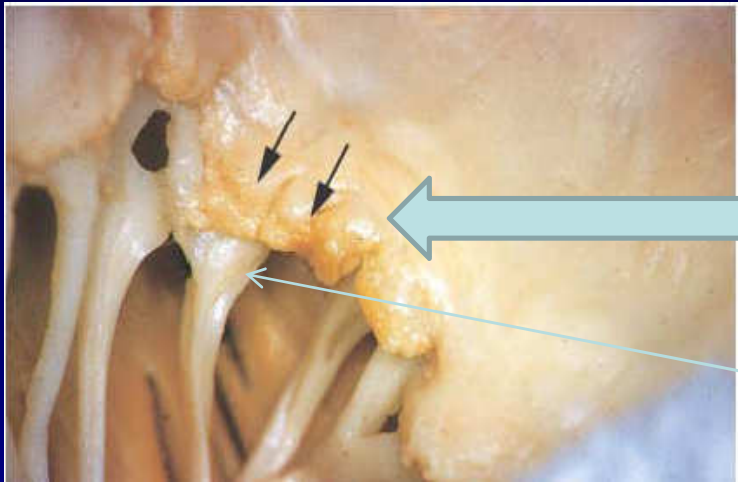
- Affected valves -mitral, aortic, tricuspid, and pulmonary valves.
  - In most cases, the mitral valve is involved with 1 or more of the other 3.
  - i.e mitral valve only or with other valves involvement
  - In acute disease, small thrombi form along the lines of valve closure.
  - In chronic disease, there is thickening and fibrosis of the valve resulting in stenosis, or less commonly, regurgitation.

# Pathology Summary

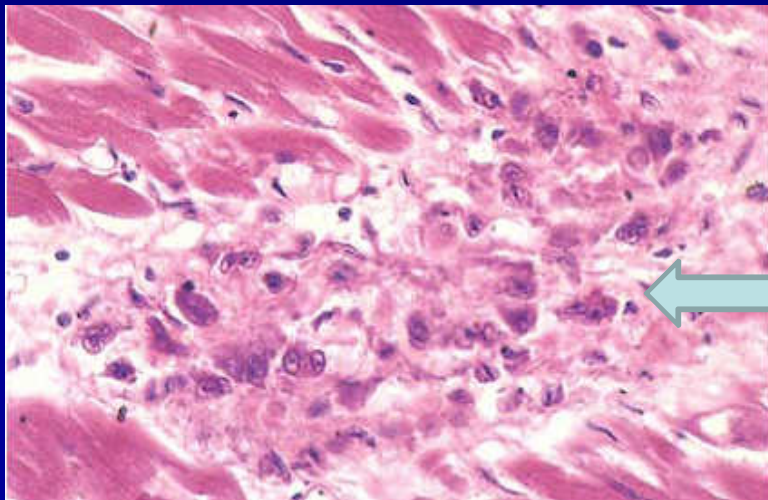


Ref: Robins Pathological Basis of Diseases.

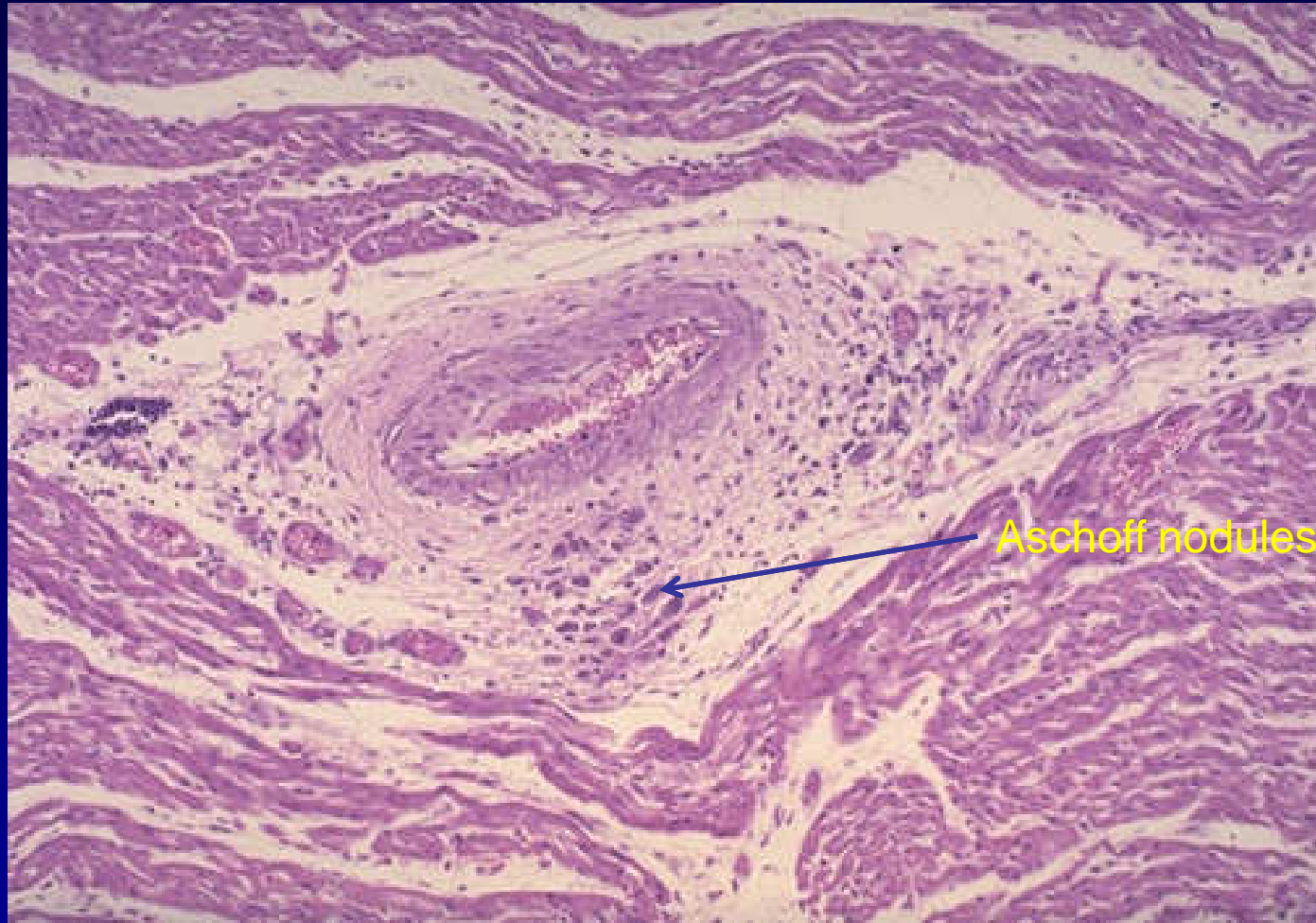
# Pathology



- **Vegetations** (verrucae – wart-like lesions) along line of closure of the mitral valve leaflet.
- Fibrous thickening and fusion of tendinous cords.

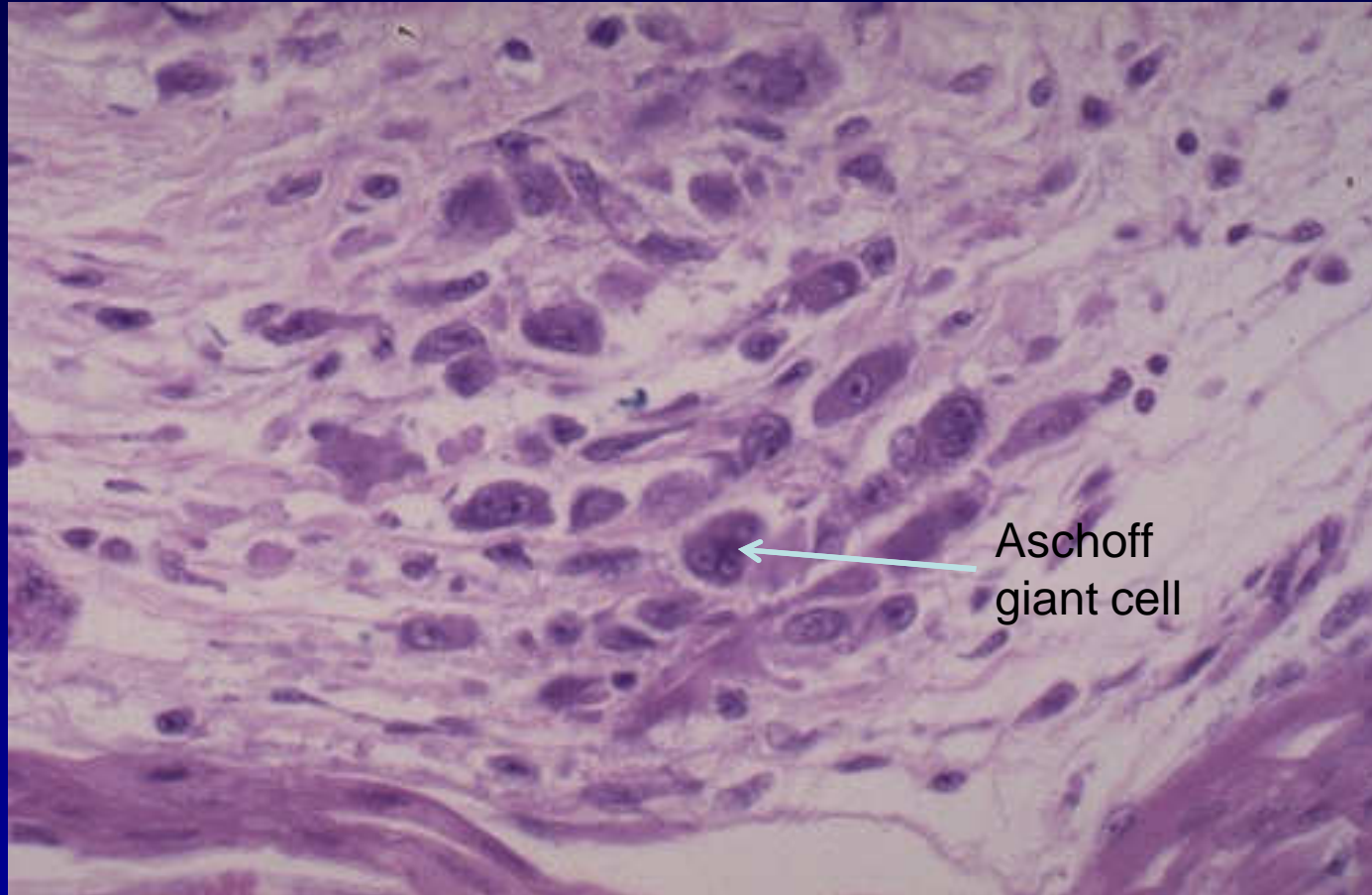


- **Aschoff bodies** within interstitial connective tissue—area of fibrinoid necrosis surrounded by macrophages.



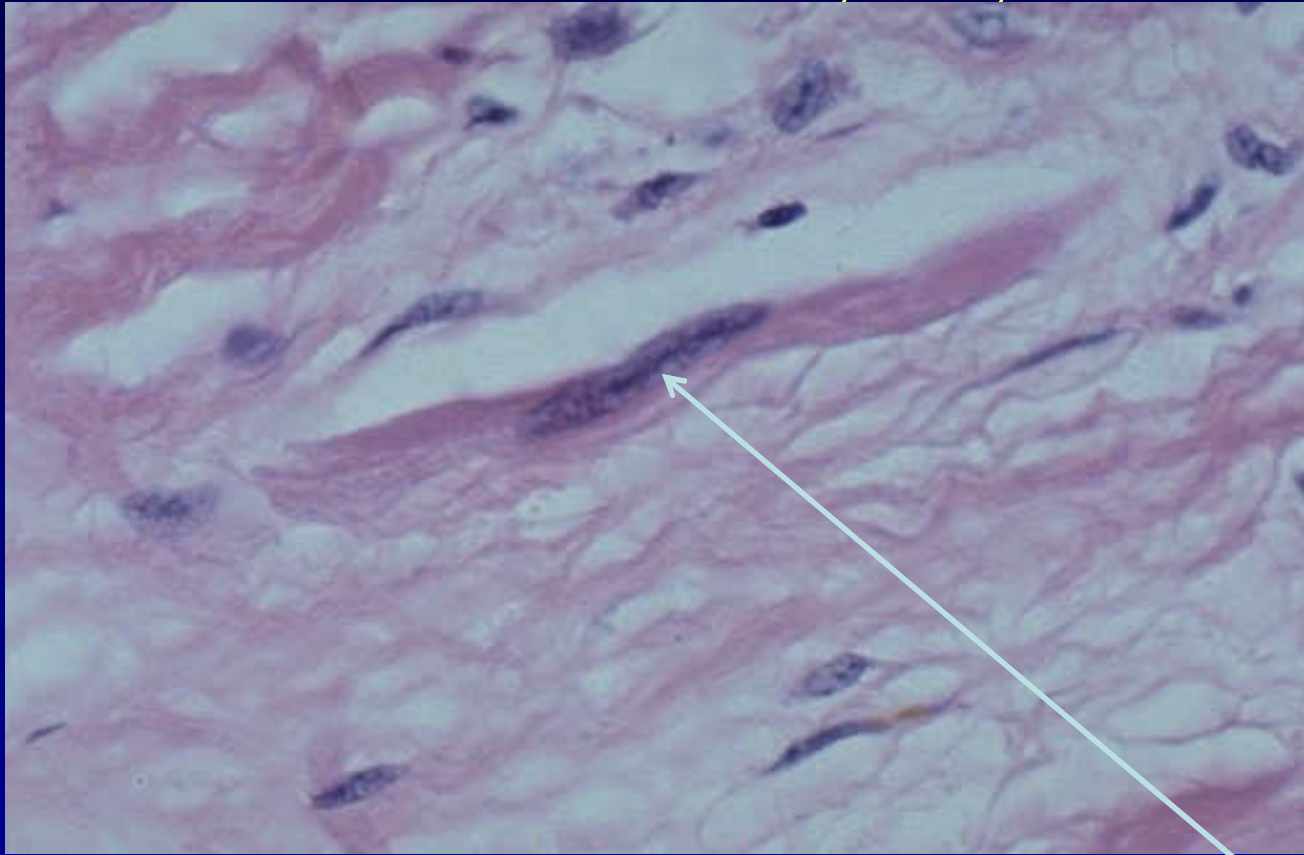
Microscopically, acute rheumatic carditis is marked by a peculiar form of granulomatous inflammation with so-called "**Aschoff nodules**" seen best in myocardium. These are centered in interstitium around vessels as shown here. The myocarditis may be severe enough to cause congestive heart failure

# Aschoff nodules



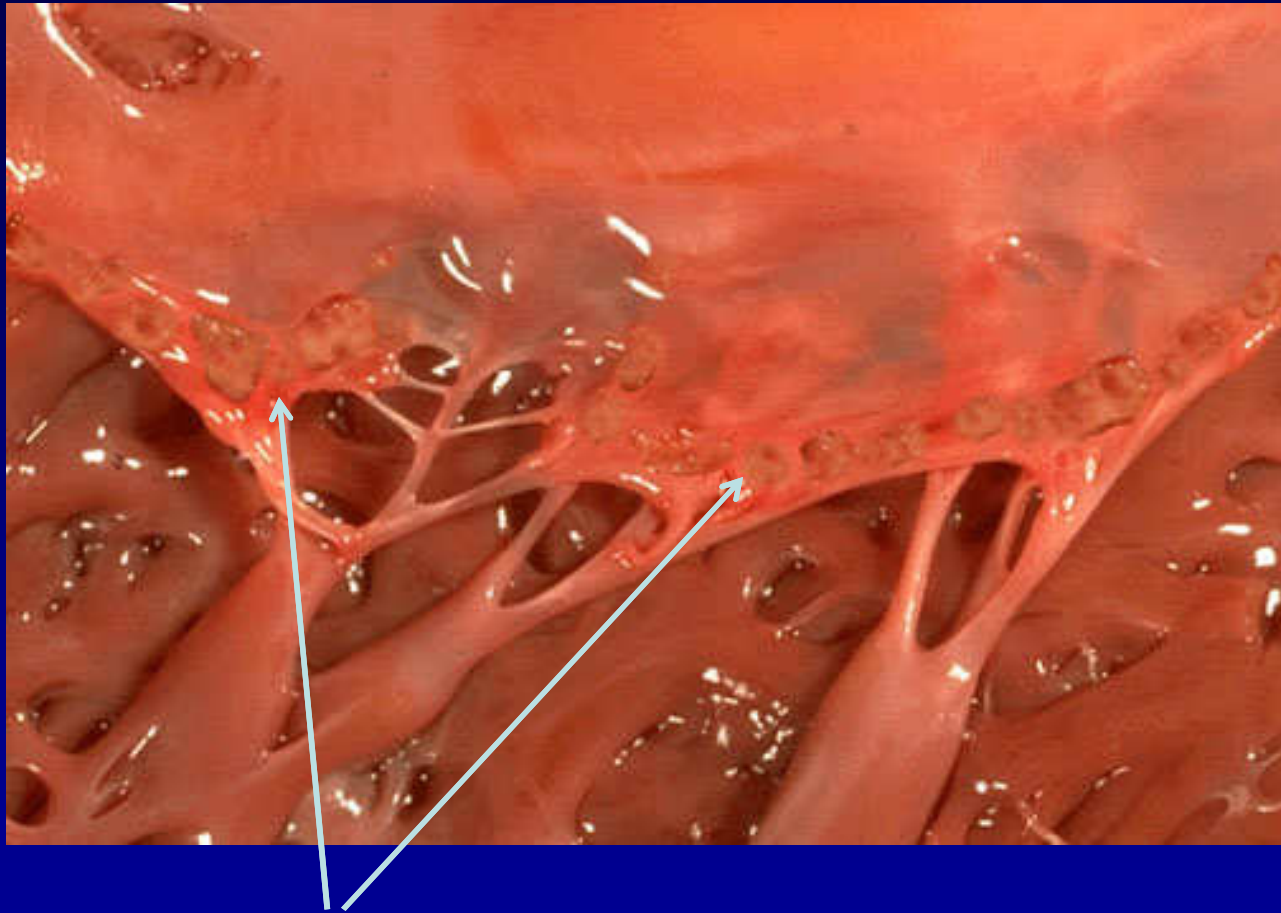
Here is an **Aschoff nodule** at high magnification. The most characteristic component is the **Aschoff giant cell**. Several appear here as large cells with two or more nuclei that have prominent nucleoli. Scattered inflammatory cells accompany them and can be mononuclears or occasionally neutrophils.

# Anitshkow myocyte



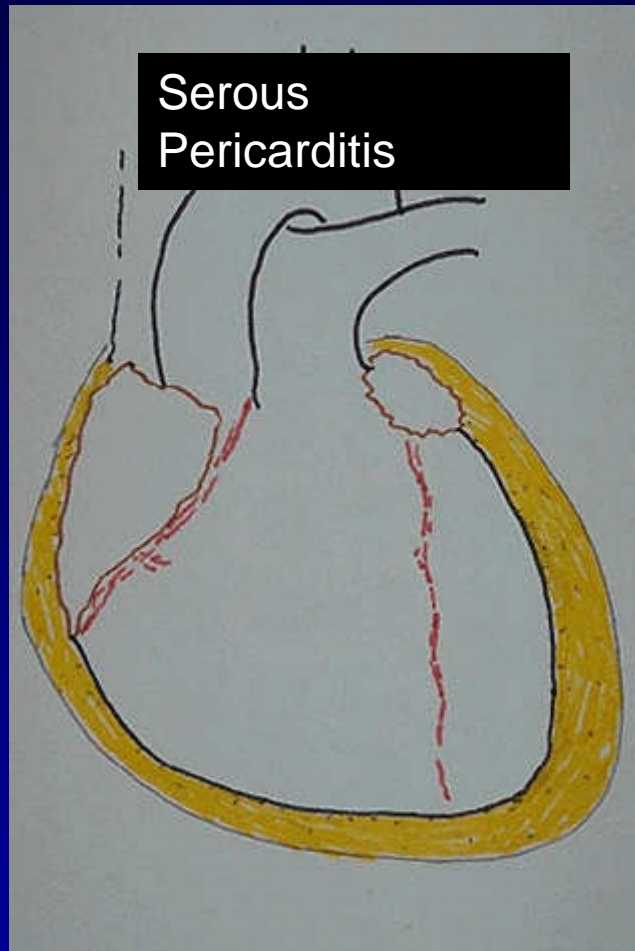
Another peculiar cell seen with acute rheumatic carditis is the **Anitschkow myocyte**. This is a long, thin cell with an elongated nucleus.

# Vegetations

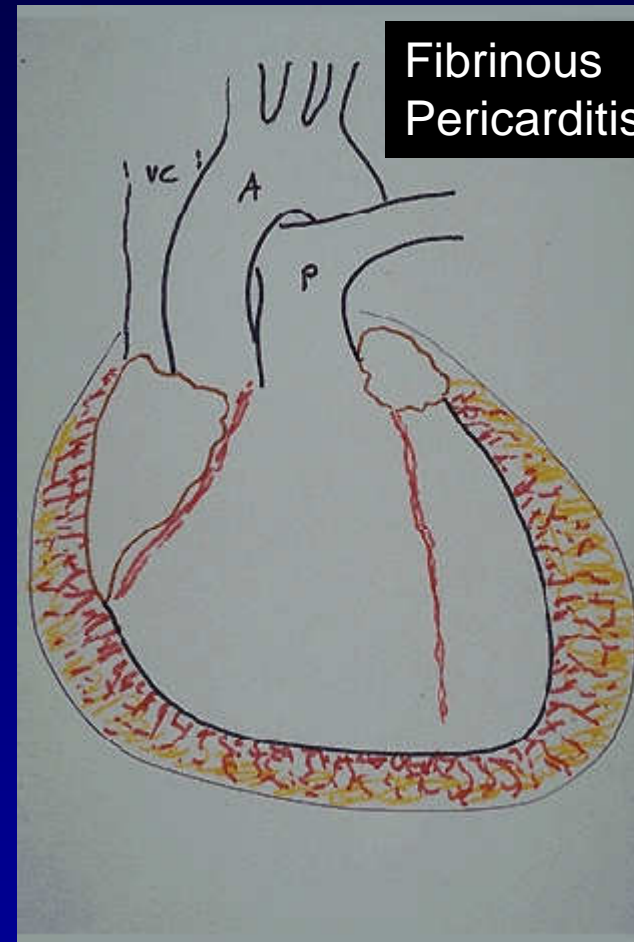


The small **verrucous vegetations** seen along the closure line of this mitral valve are associated with acute rheumatic fever. These warty vegetations average only a few millimeters and form along the line of valve closure over areas of endocardial inflammation.

# Serous Vs Fibrinous Pericarditis



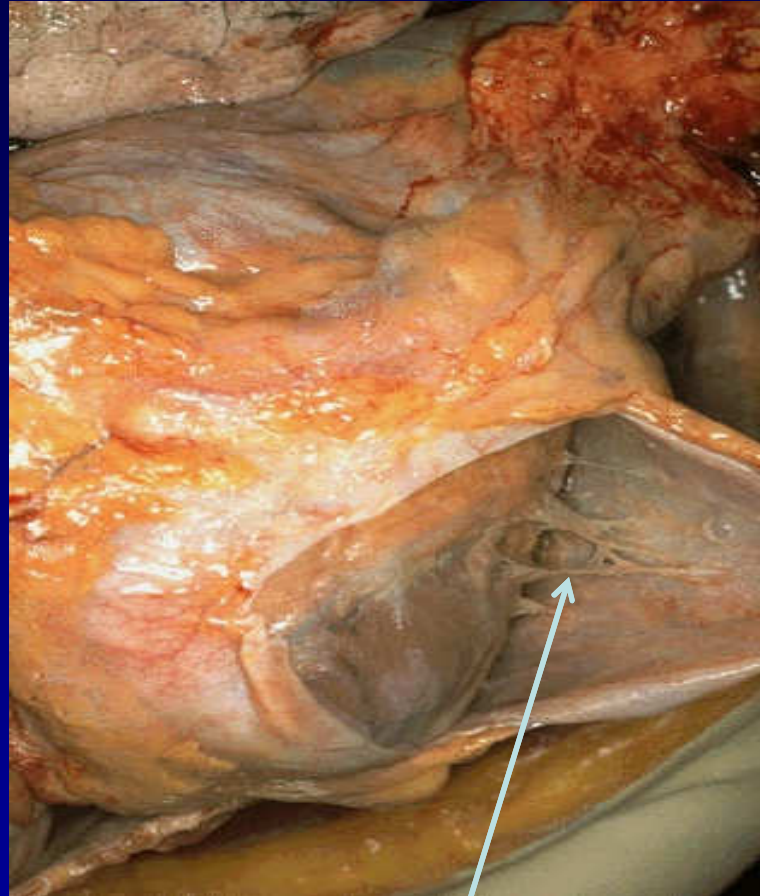
Serous fluid in pericardial space  
No fibrin  
Inflammatory cells in yellow serous fluid



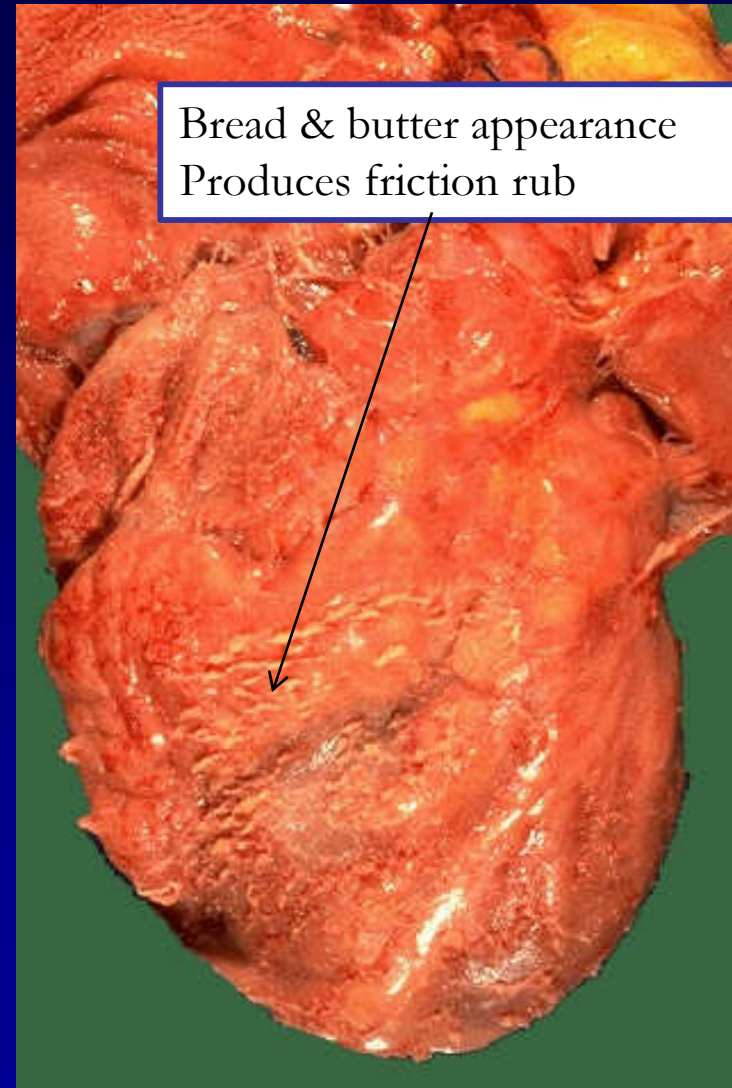
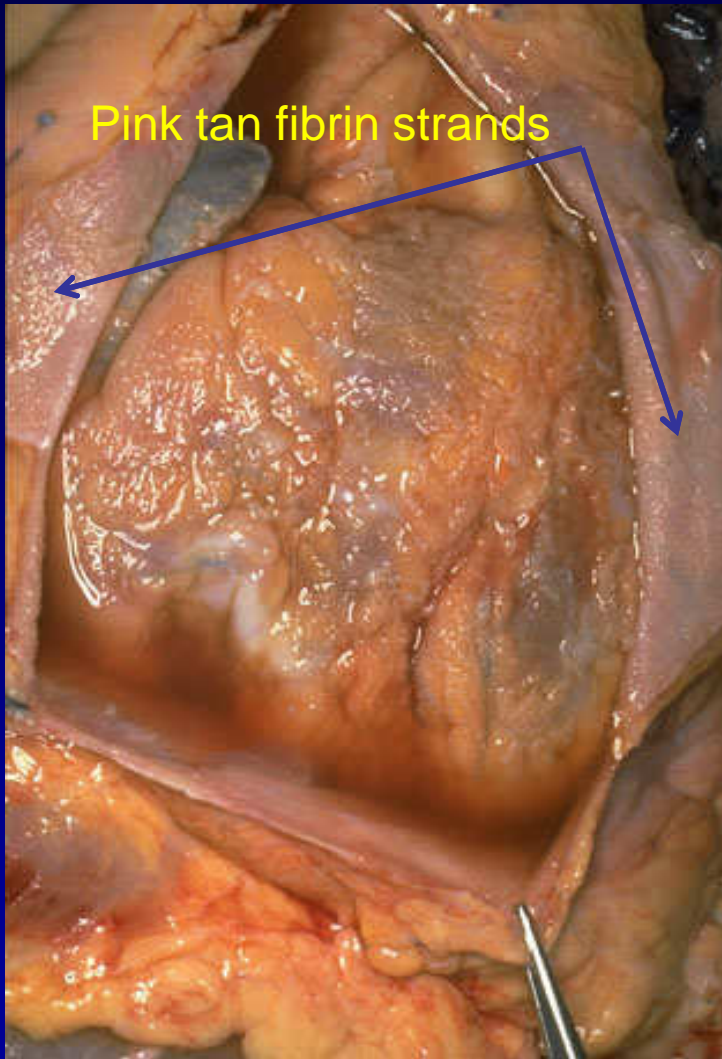
Fibrin strands extending from epicardium into yellow fluid



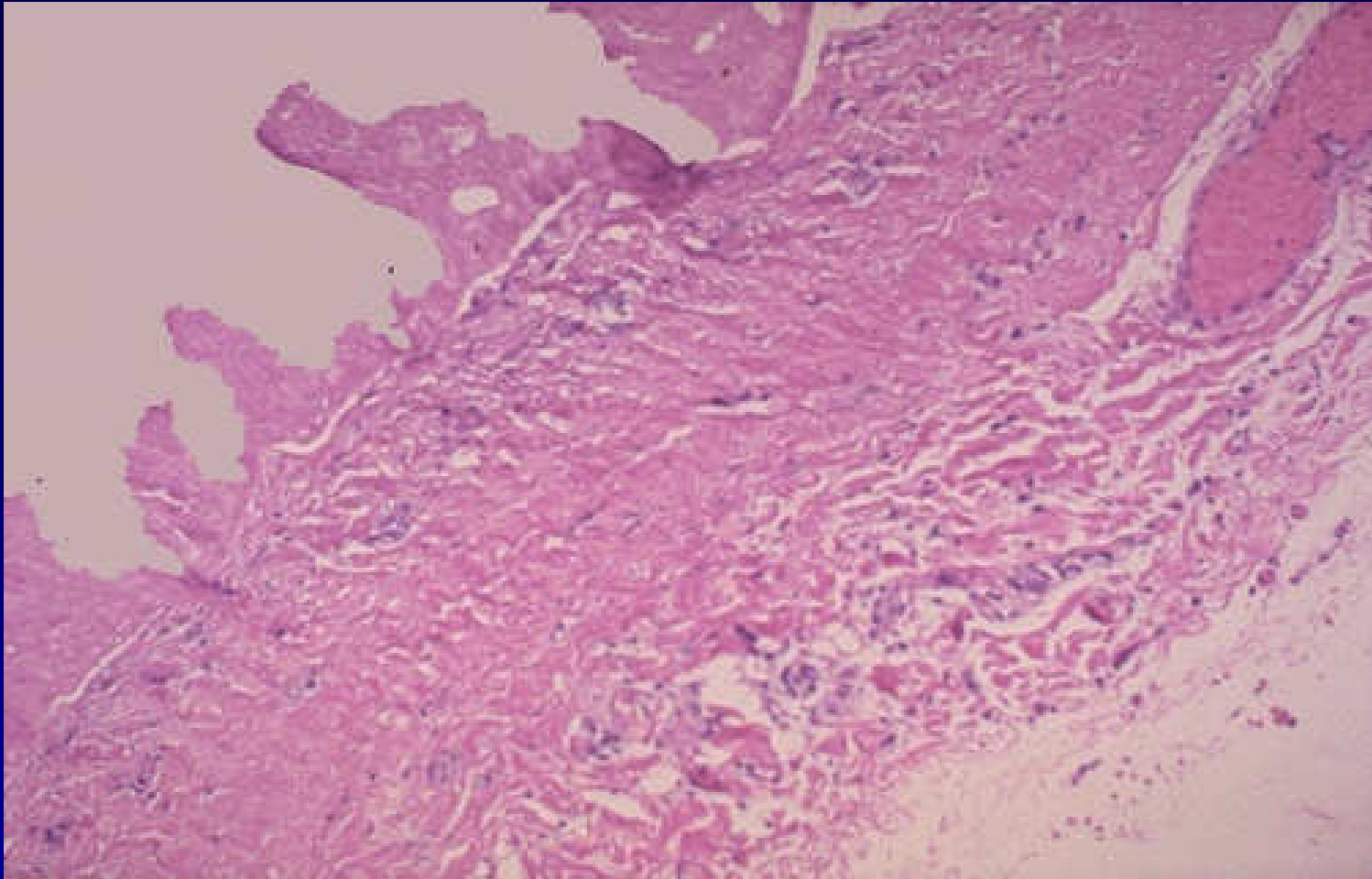
# Fibrinous Pericarditis



A window of adherent pericardium has been opened to reveal the surface of the heart. There are **thin strands of fibrinous exudate** that extend from the epicardial surface to the pericardial sac.

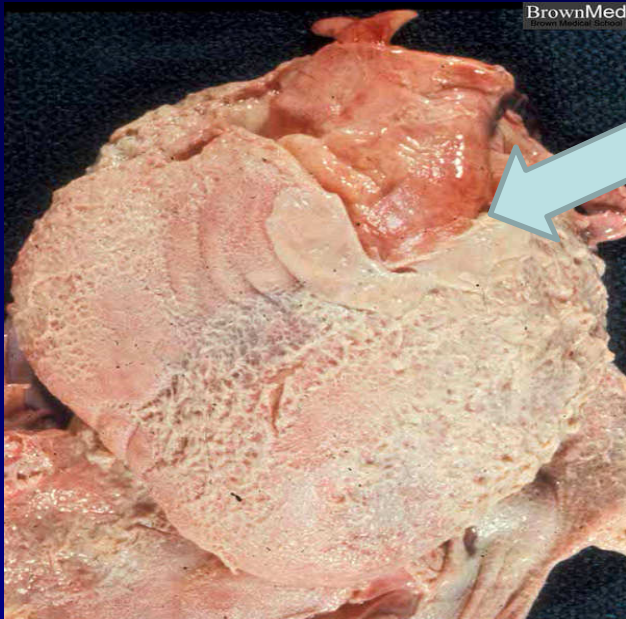


- Fibrinous Pericarditis



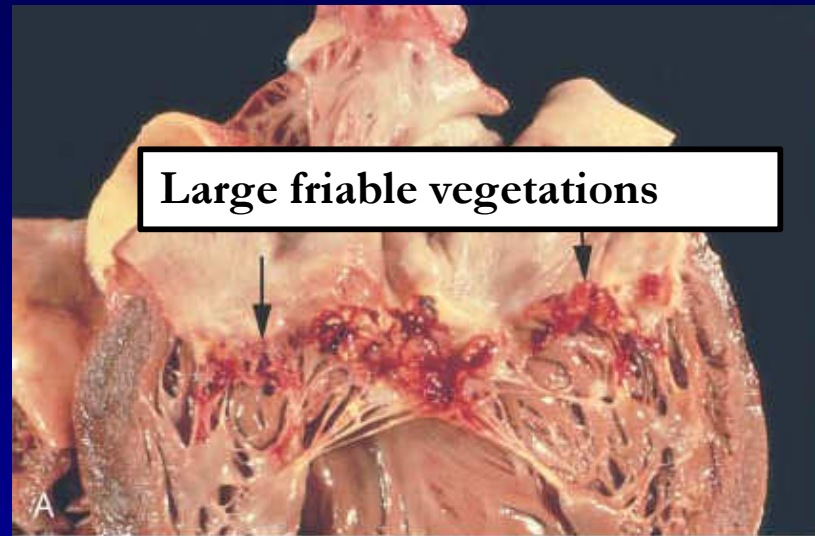
Microscopically, the pericardial surface here **shows strands of pink fibrin extending outward**. There is underlying inflammation. Eventually, the fibrin can be organized and cleared, though sometimes adhesions may remain

# Pathology



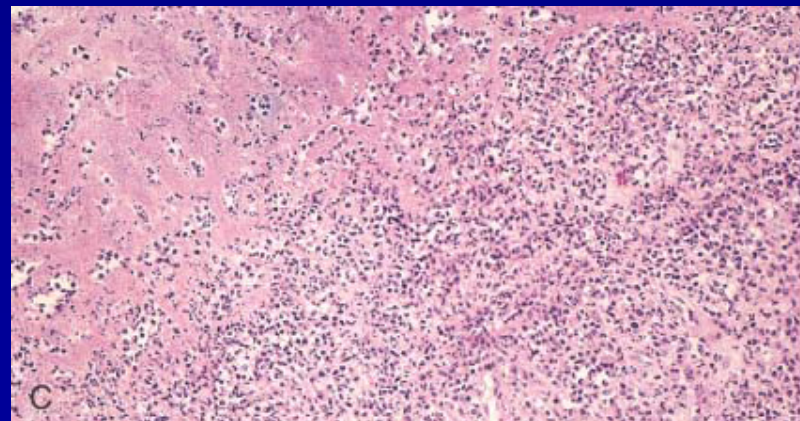
**Sterile fibrinous pericarditis** "Bread & butter" appearance of pericarditis.

Ref: [www.brown.edu](http://www.brown.edu)

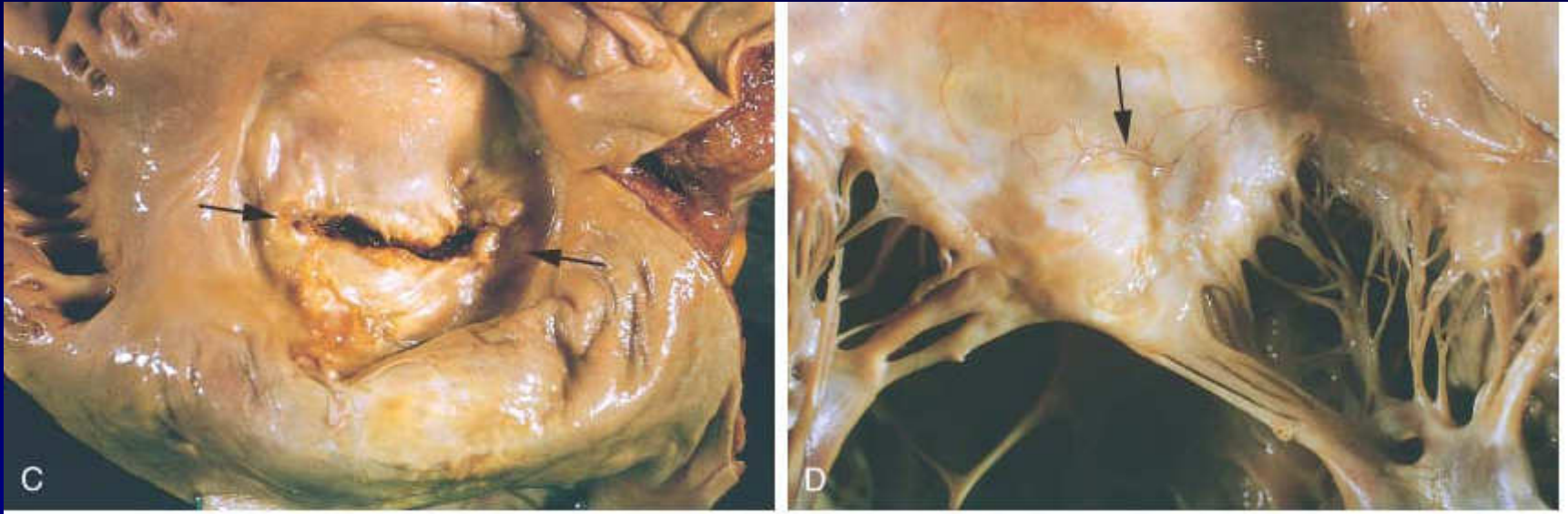


**Infective Endocarditis**

Acute inflammatory cells & fibrin



# Chronic Rheumatic Heart Disease



Characterised by: organisation of acute inflammation and subsequent deforming fibrosis. Micro: fibrosis & neovascularization. Aschoff bodies replaced by fibrotic scar.

Valvular leaflets become thickened & retracted causing permanent deformity.

Mitral valve most commonly affected. Aortic valve next common.  
Changes: leaflet thickening, commissural fusion; shortening, thickening and fusion of tendinous cords.

# Chronic rheumatic valvulitis



Shortened thickened  
chordae tendinea

Chronic rheumatic valvulitis may develop by organization of the acute endocardial inflammation along with fibrosis, as shown here affecting the mitral valve. Note the **shortened and thickened** chordae tendinea

# Rheumatic Valvular Heart Disease

- Mitral stenosis – 99%
- Mitral valve alone – 65-70% of patients.
- Mitral valve & aortic – 25% of patients.
- Tricuspid & pulmonary valve very rare involvement in rheumatic heart disease.
- Fibrous bridging across valvular commissures and calcification create “fish mouth” or “buttonhole” stenosis.
- Left atrial dilation from long standing MS.
- Long standing congestion changes in the lung induce vascular & parenchymal changes (pulmonary hypertension) causing RVH

# Mitral Stenosis: “fish mouth”



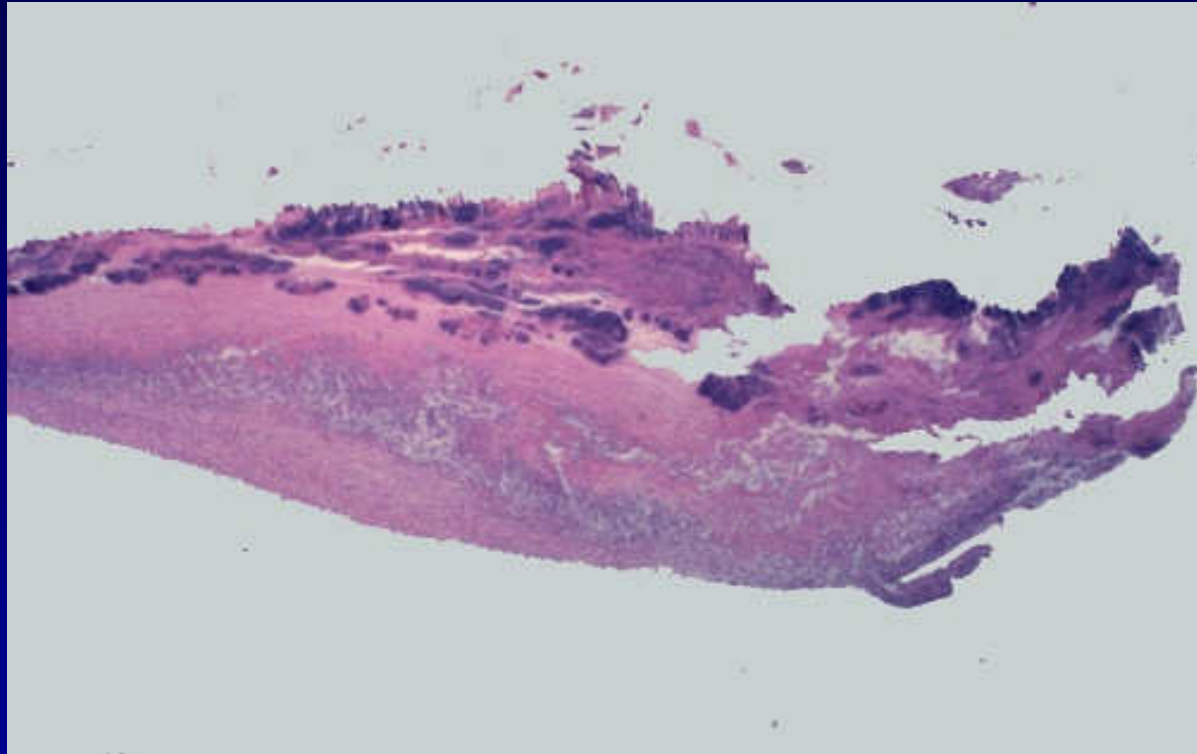
The mitral valve demonstrates the typical **"fish mouth"** shape with chronic rheumatic scarring. Mitral valve is most often affected with rheumatic heart disease, followed by mitral and aortic together, then aortic alone, then mitral, aortic, and tricuspid together.



# Complications of Rheumatic Heart Disease

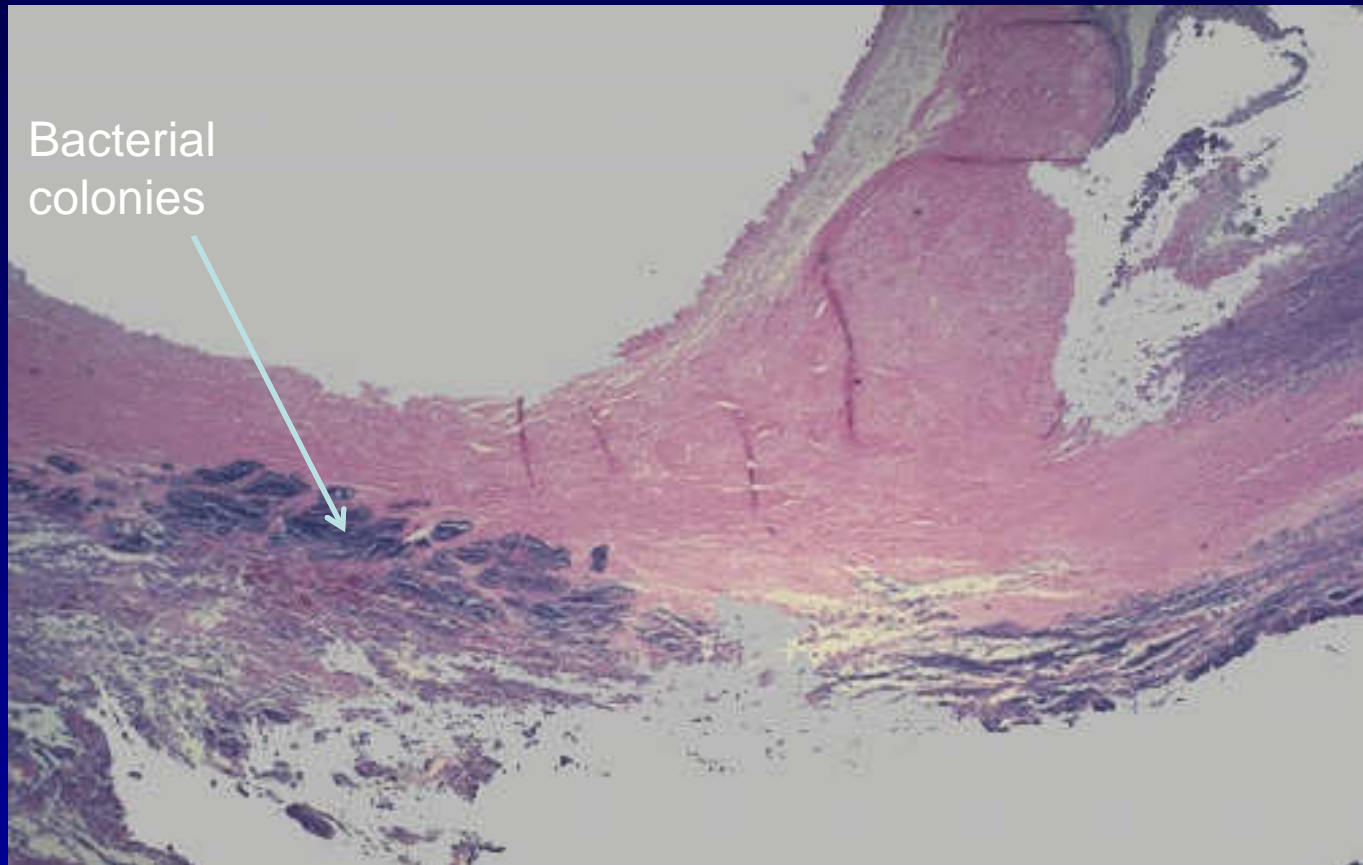
- **Infective endocarditis** – the vegetations become infected with bacteria, often streptococcus spp.
- **Embolization** – vegetation detach and embolize to the brain or other organs.
- **Valvular stenosis** – narrow of valves from fibrotic scarring and calcification of damaged valve.
- **Valvular insufficiency** – fibrosis and scarring prevent valves from closing properly leading to regurgitation of blood during systole.
- **Constrictive pericarditis** – resolution of pericarditis causing obliteration of pericardial sac.

# Infective Endocarditis



Microscopically, the valve in infective endocarditis demonstrates **friable vegetations of fibrin and platelets (pink) mixed with inflammatory cells and bacterial colonies (blue)**. The friability explains how portions of the vegetation can break off and embolize.

# Infective Endocarditis



Here is a valve with infective endocarditis. The blue bacterial colonies on the lower left are extending into the pink connective tissue of the valve. Valves are relatively avascular, so high dose antibiotic therapy is needed to eradicate the infection.

# Laboratory Diagnosis

- Demonstrating rise in ASO titre with Jones criteria.
- Echocardiography – demonstrate vegetations, valvular heart disease.
- Blood culture only if you suspect infective endocarditis. Obtain blood sample before antibiotics.

# Prognosis

- Good prognosis (depending on where you live)
- 1% die from fulminate rheumatic fever.
- Increased risk of rheumatic heart disease after 1<sup>st</sup> episode.
- Carditis likely with each subsequent episode and damage is cumulative.
- Increased risk of infective endocarditis.
- Chronic rheumatic heart disease from damaged valves.

# End

- Robins Pathological Basis of Diseases – what ever edition you have.
- Download PDF copies of seminar notes at:

[www.pathologyatsmhs.com](http://www.pathologyatsmhs.com)