Problems in the heart

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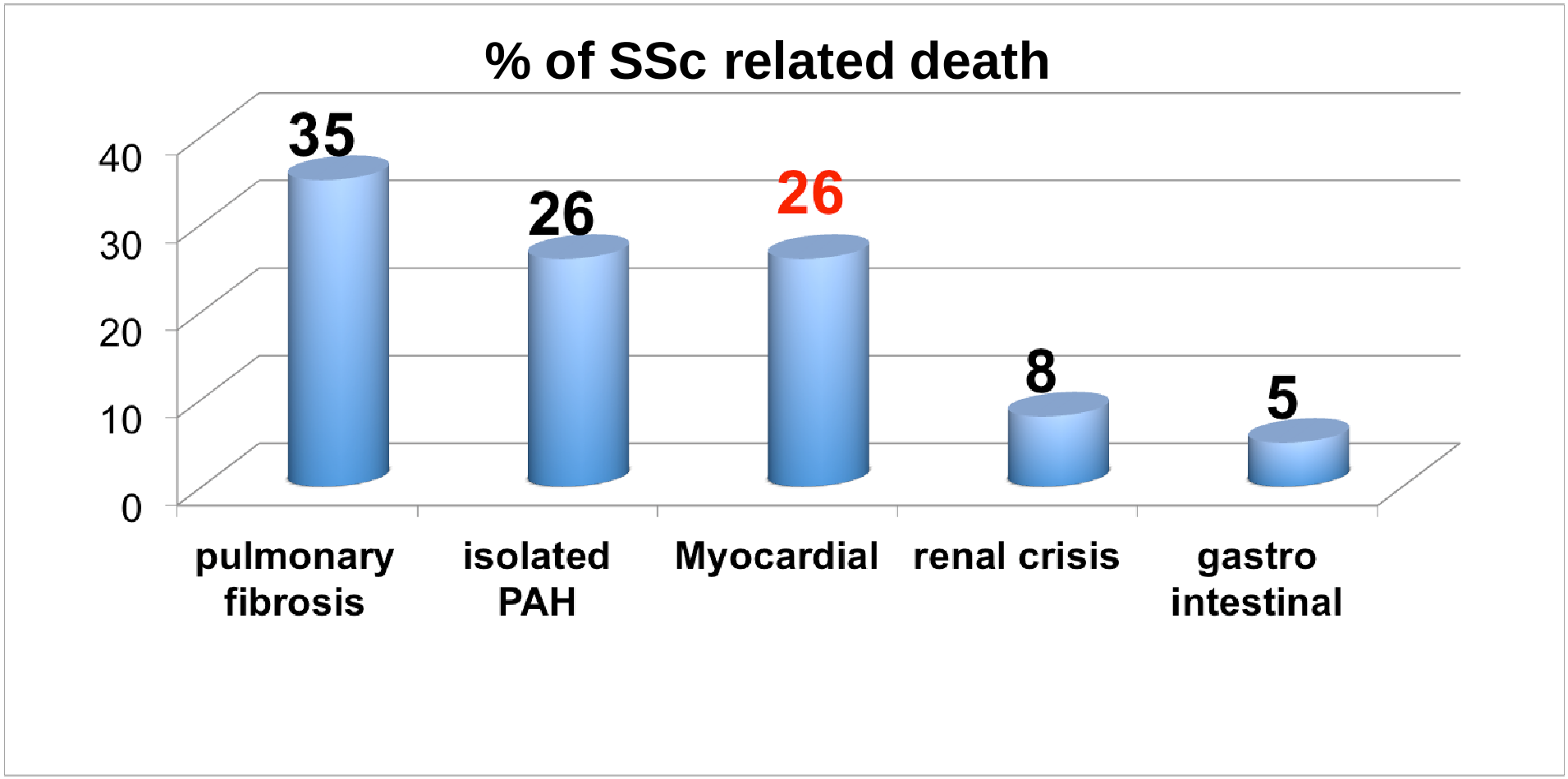
Symptoms

- **Dyspnea**
- **Chest pain**
- **Fluid retention….**

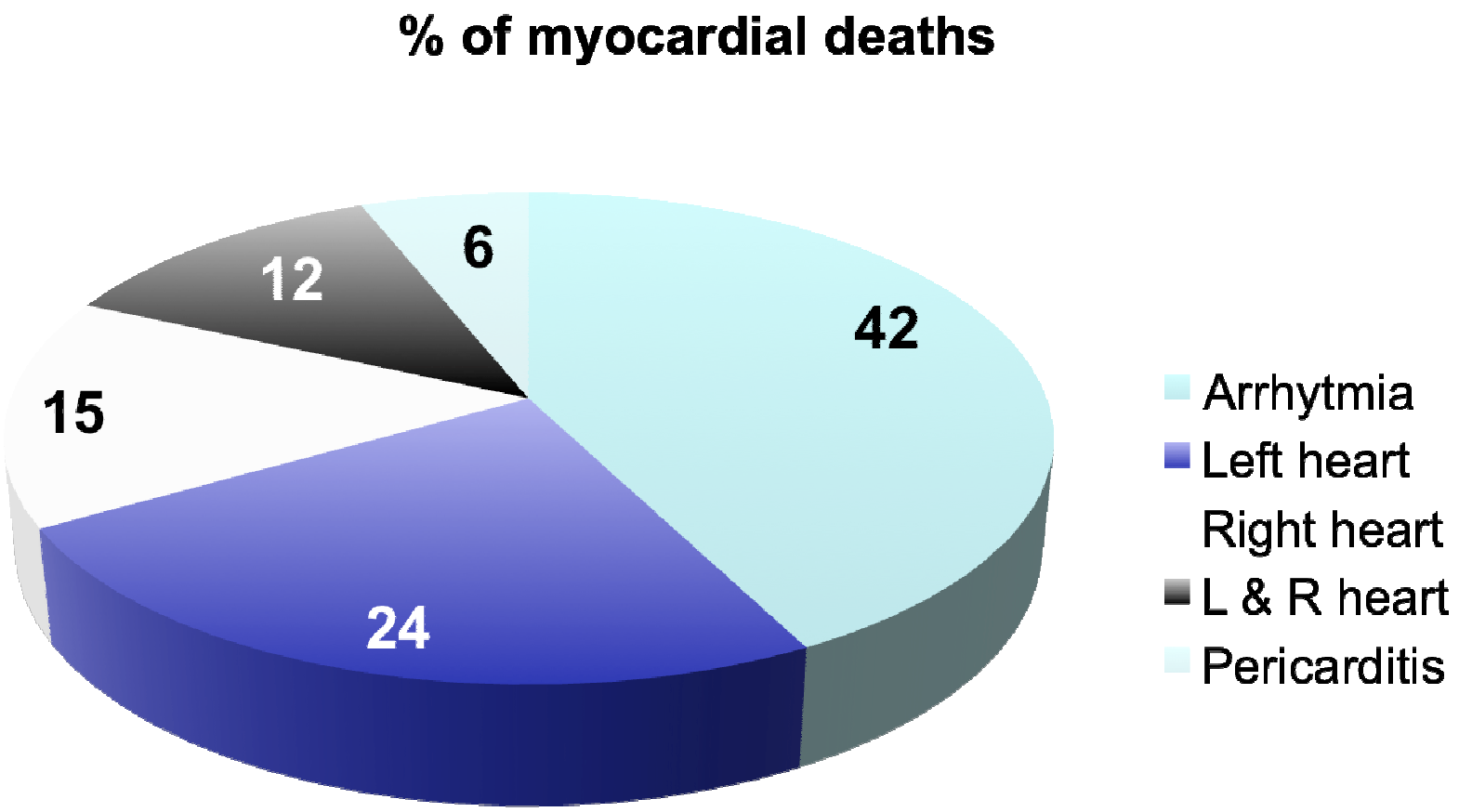
- **More specific of cardiac involvement:**
  - **Palpitations**

EUSTAR database, > 7000 patients: 234 deaths including 128 SSc-related



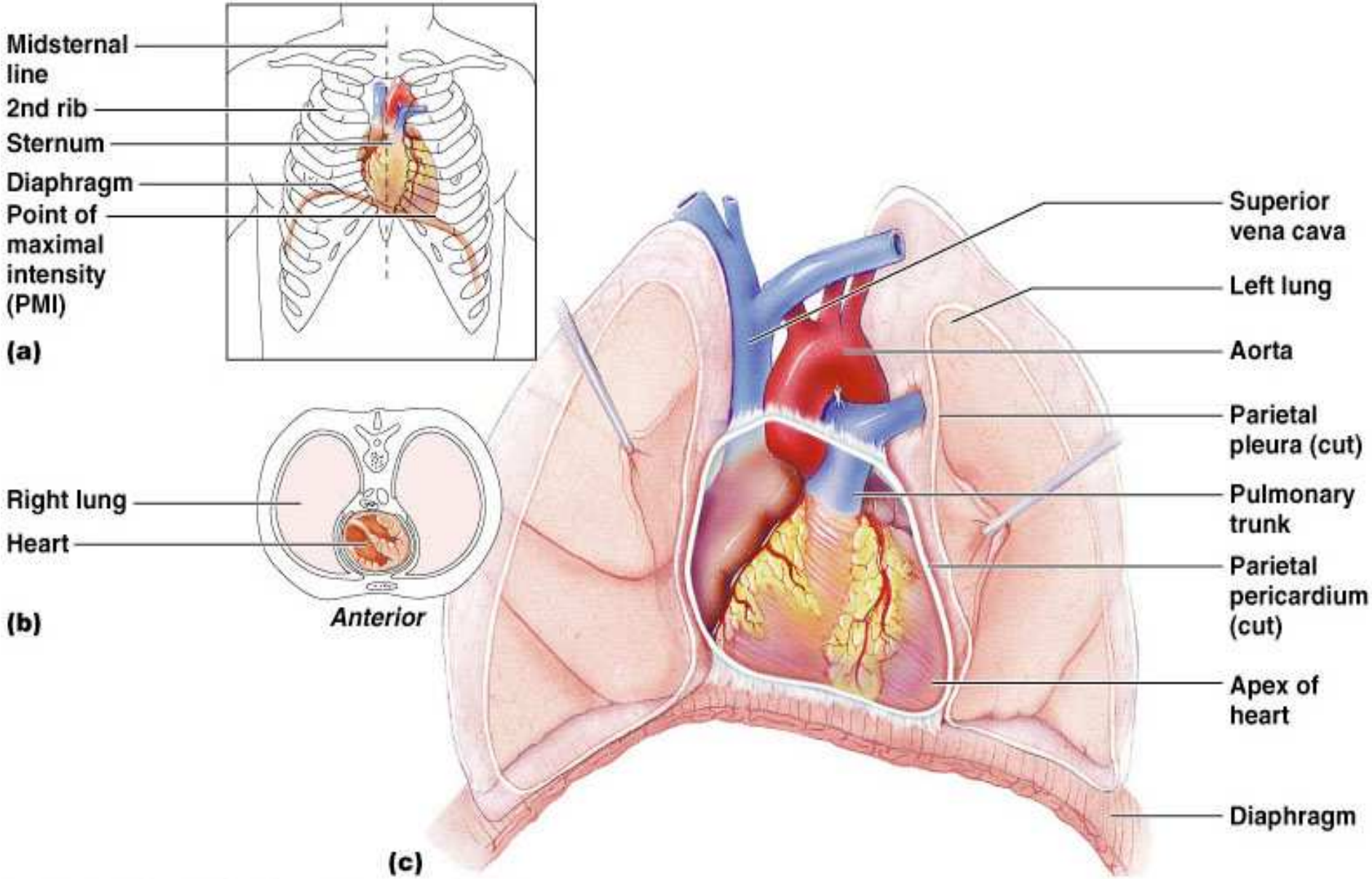
Tyndall et al, Ann Rheum Dis 2010;69:1809-1815

Cardiac deaths: n=33

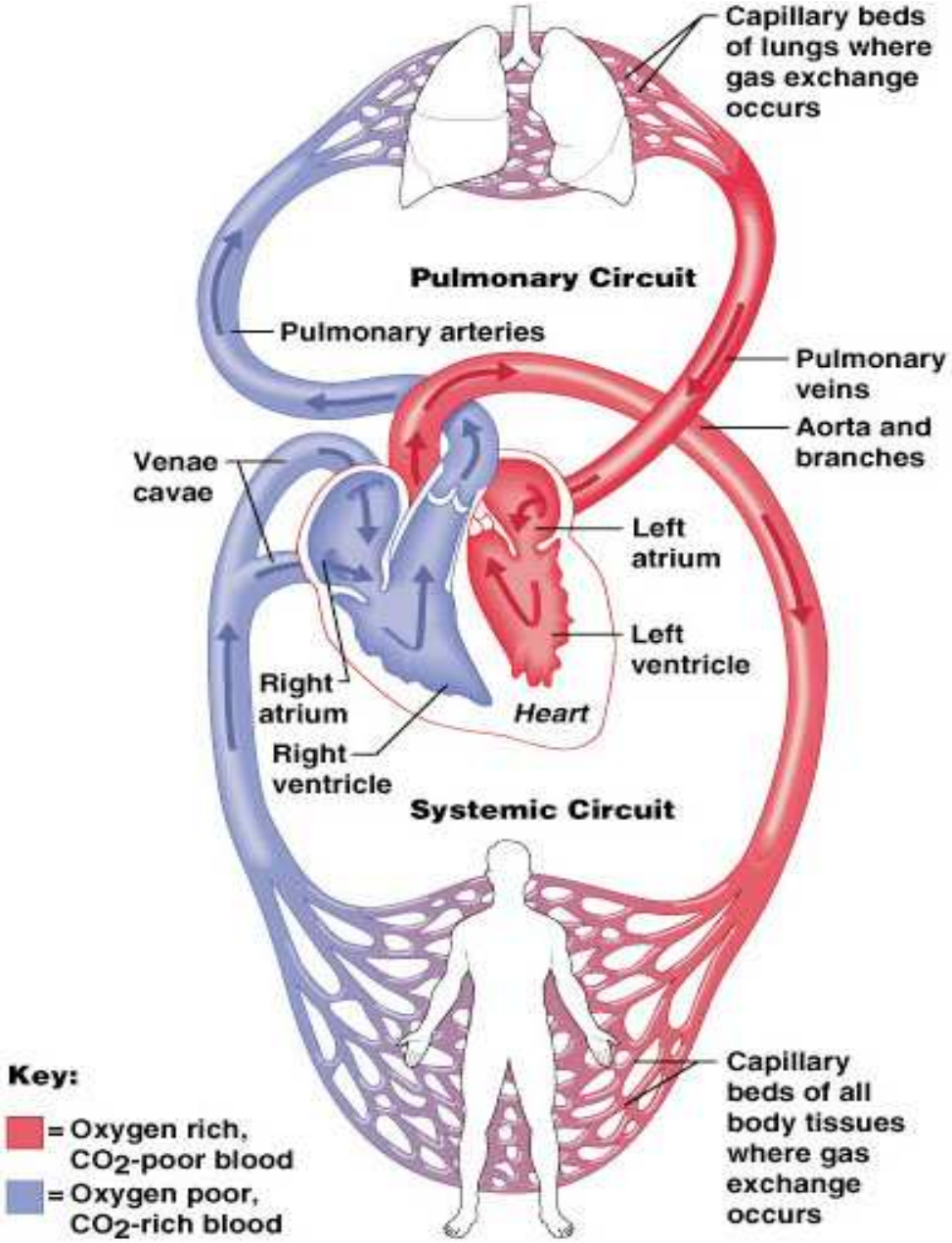


Tyndall et al, Ann Rheum Dis 2010;69:1809-1815

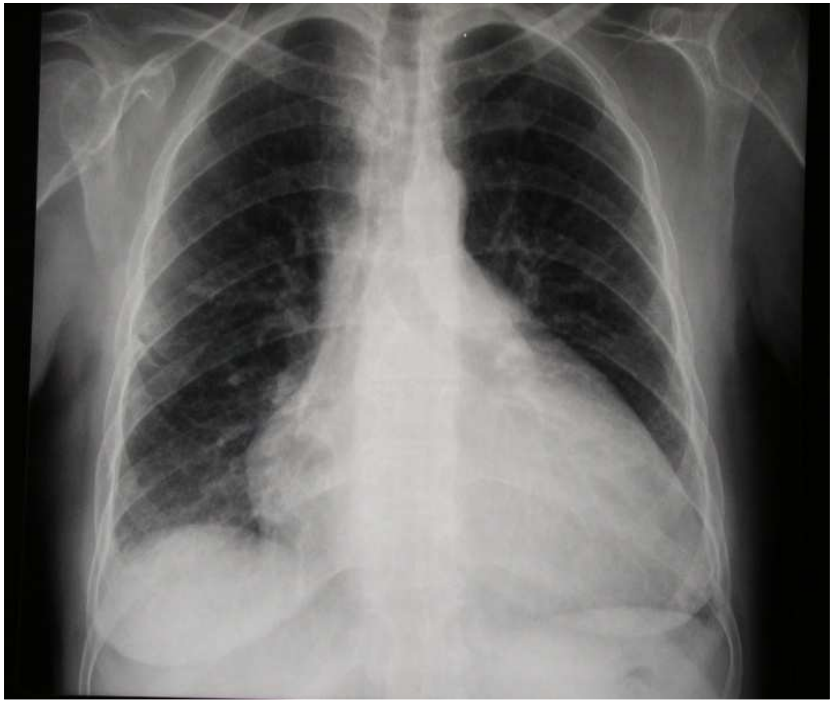
Heart Anatomy

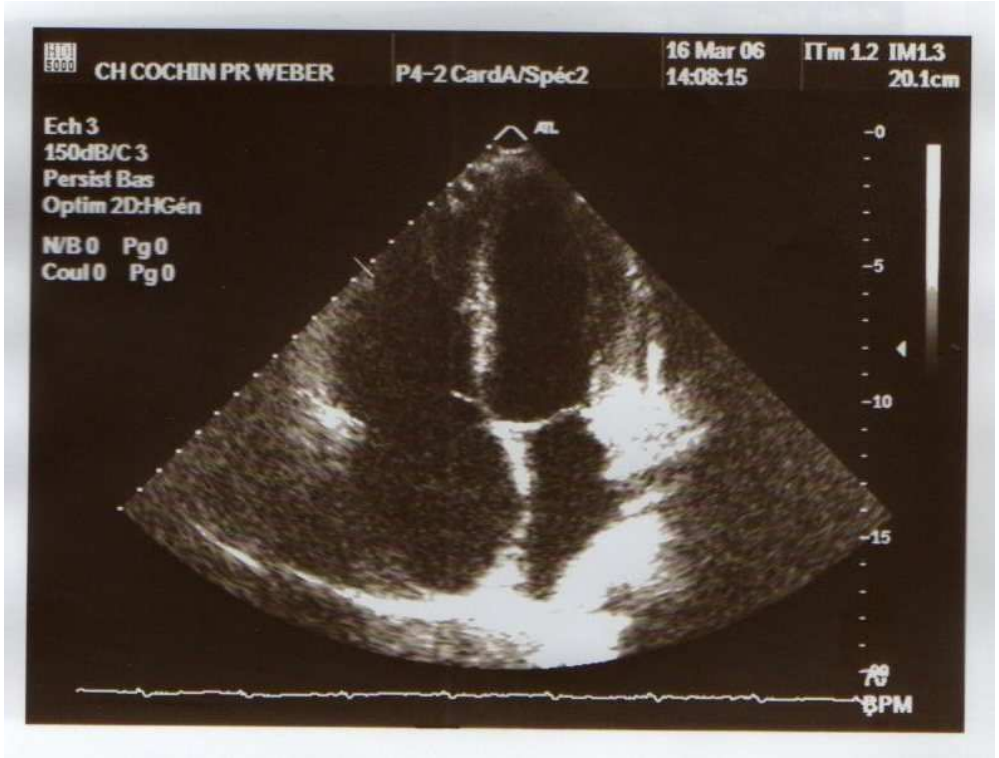


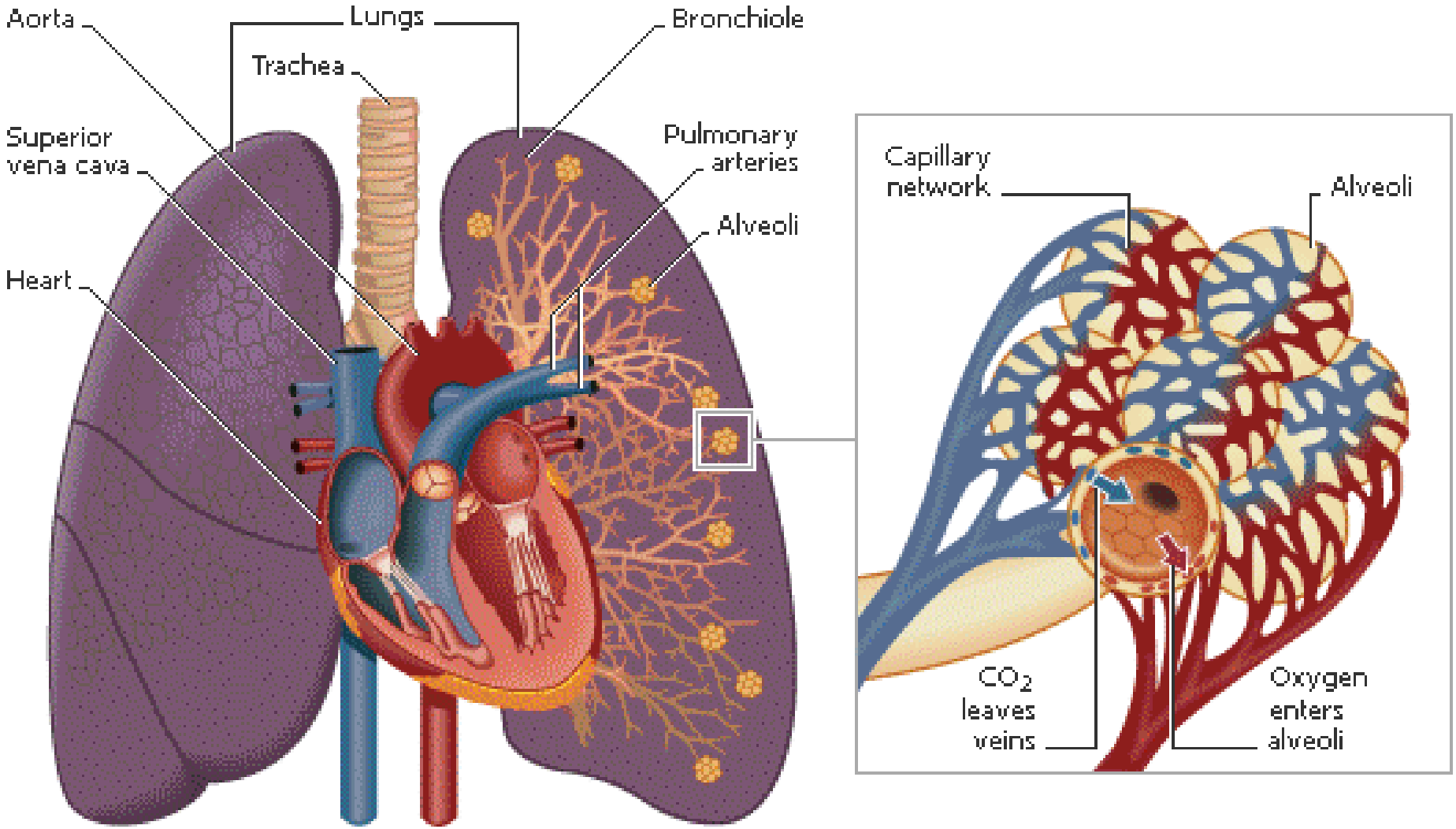
Pathway of Blood Through the Heart and Lungs

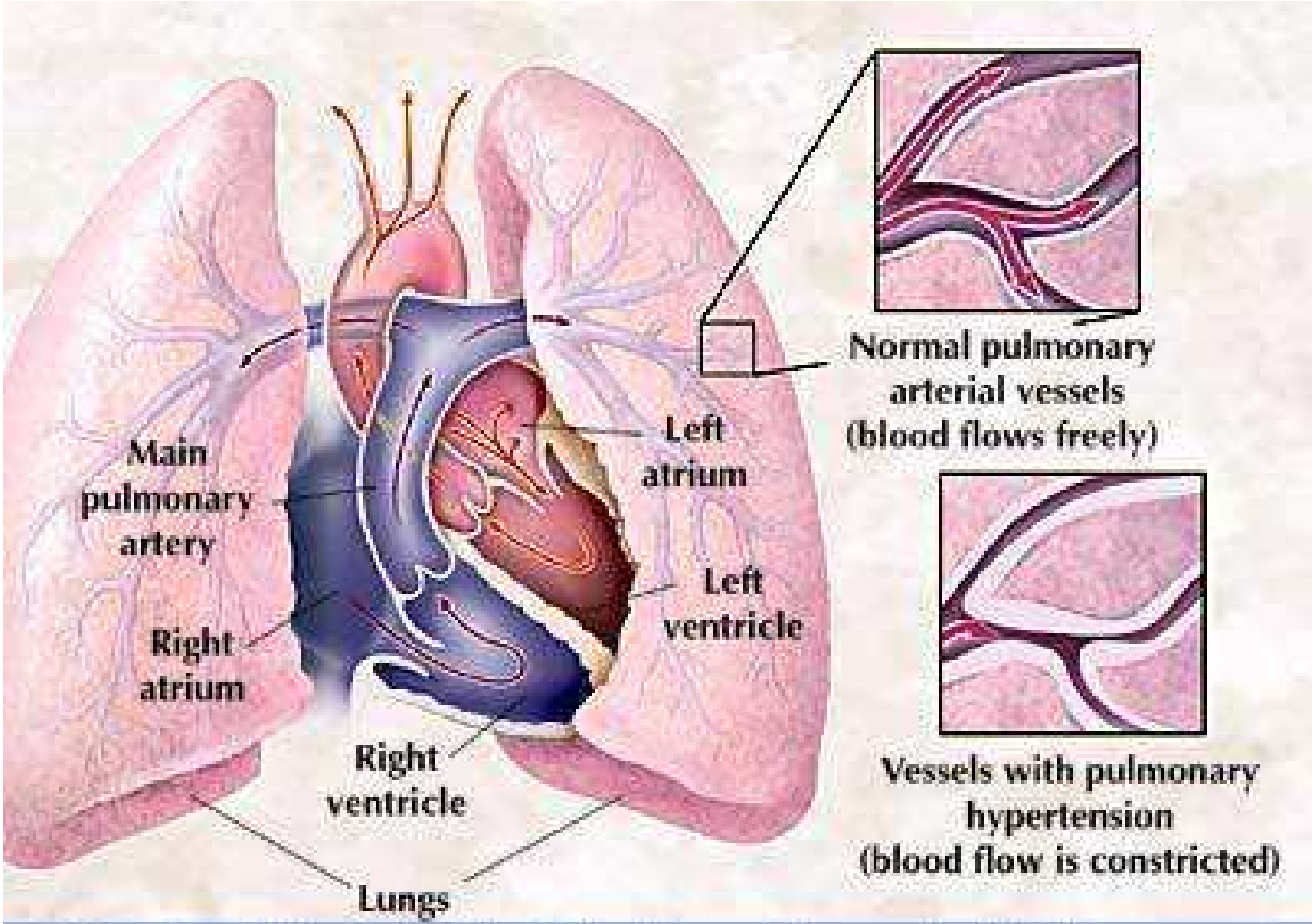


Secondary heart disease: pulmonary hypertension






Aorta
Lungs
Trachea
Bronchiole
Superior vena cava
Pulmonary arteries
Alveoli
Heart
Capillary network
Alveoli
CO_2 leaves veins
Oxygen enters alveoli



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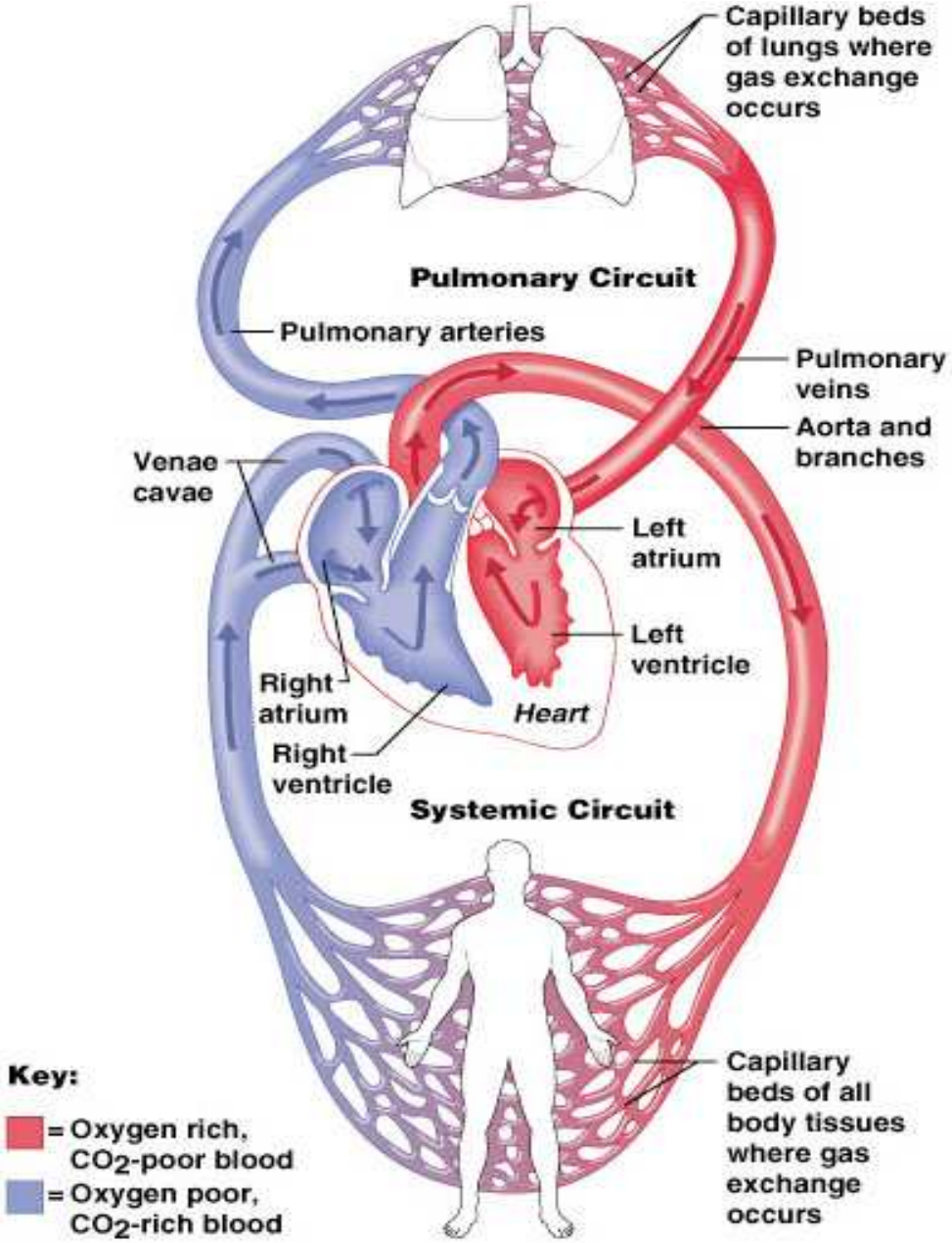
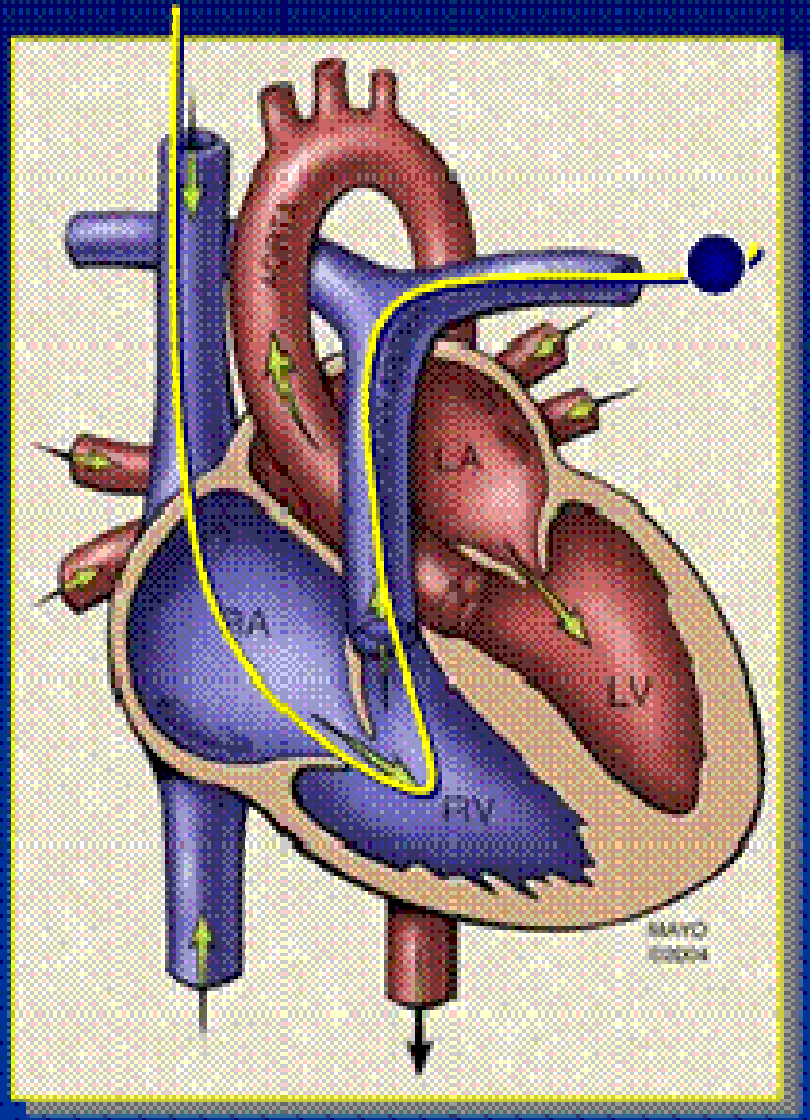


Cardiac Catheterization

A more invasive test in which the physician inserts a small tube called a "catheter" into a patient's blood vessel and passes the tube toward the heart

The cardiac catheterization measures important pressures in the heart and lungs

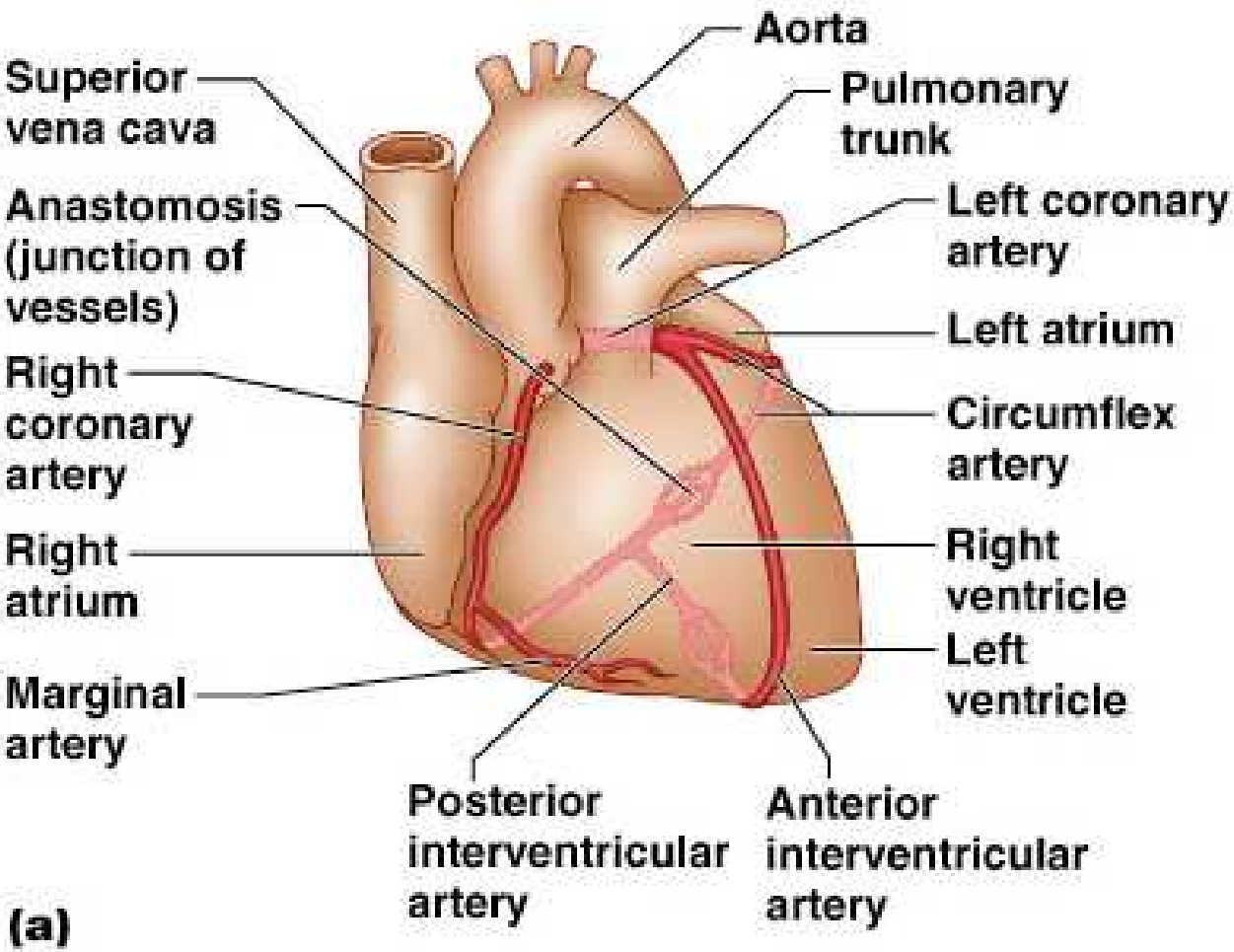


Primary heart disease: reduced systolic ejection fraction

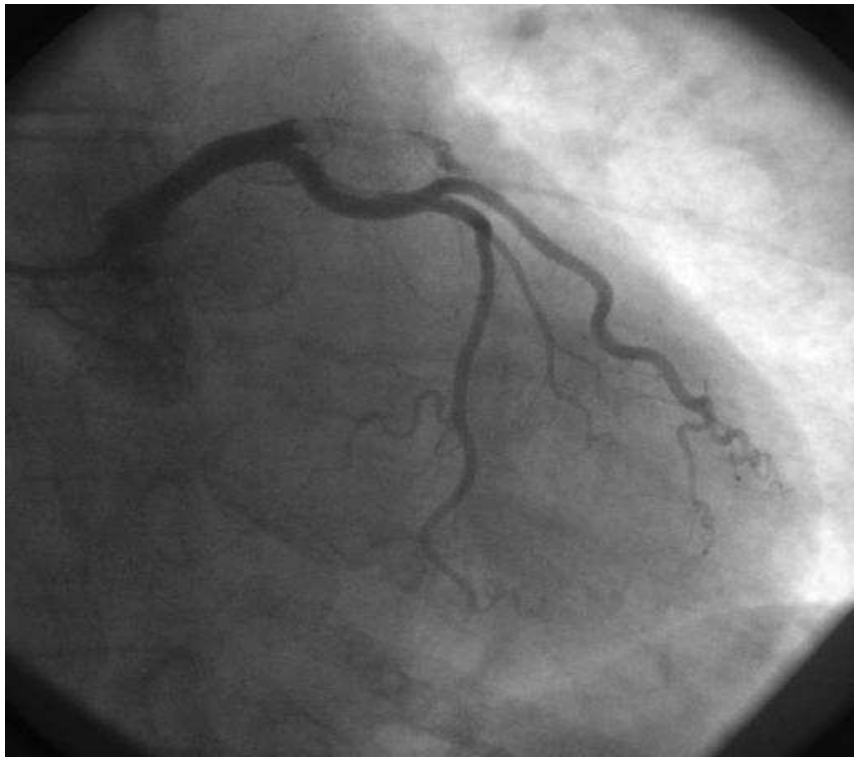
- **EUSTAR database**
- **7,077 patients included in April 2008 with available ECHO**

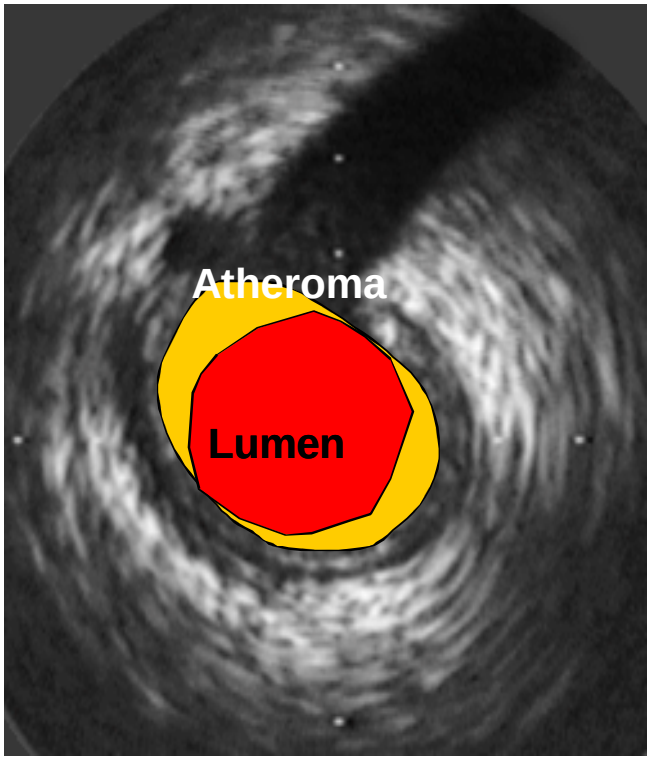
  → 383 (5.4%) had a reduced LV ejection fraction < 50-55%

Coronary Circulation: Arterial Supply

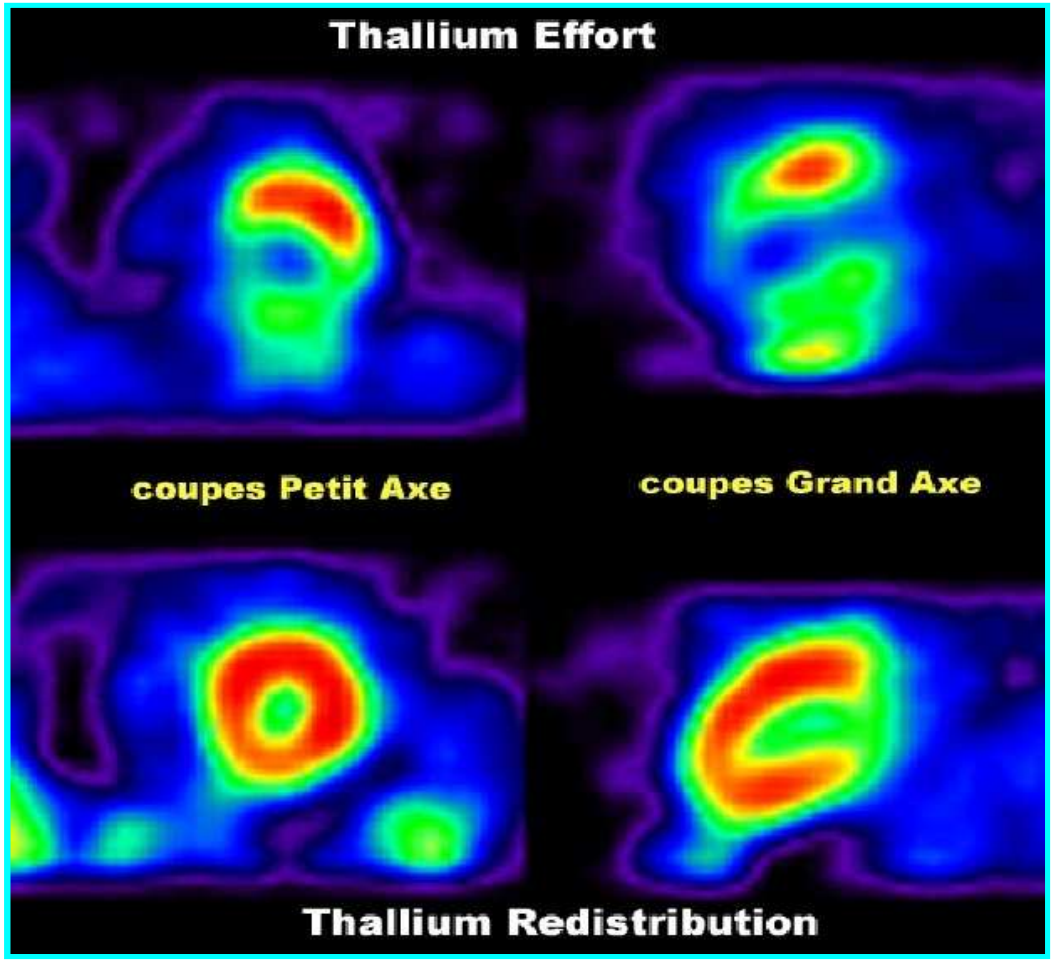


Atherosclerosis

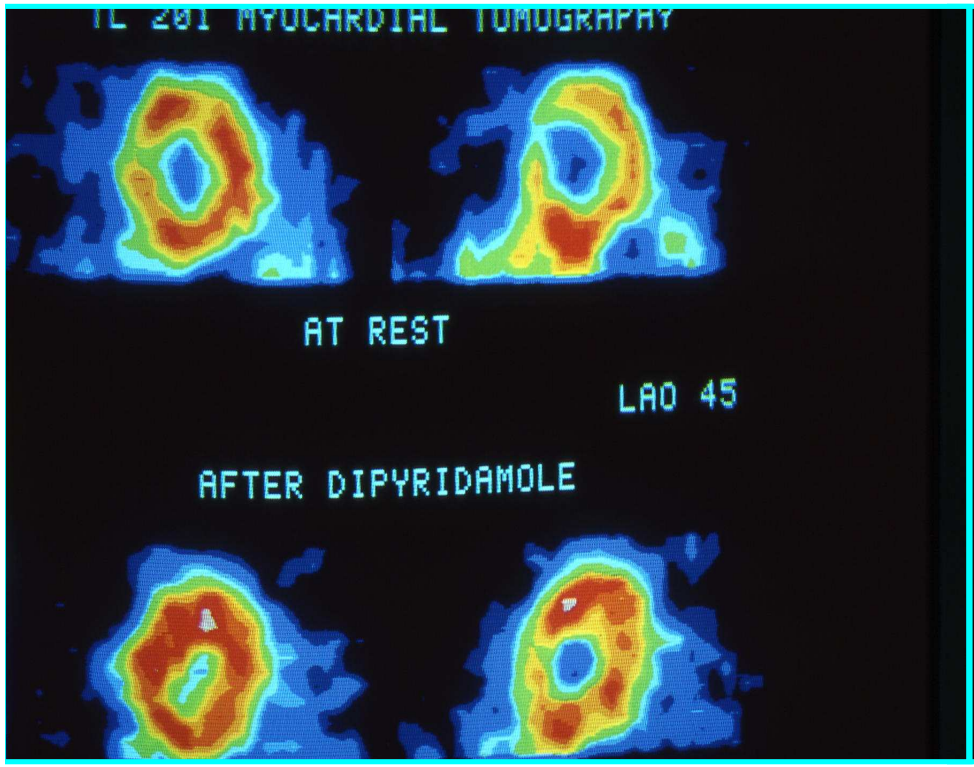


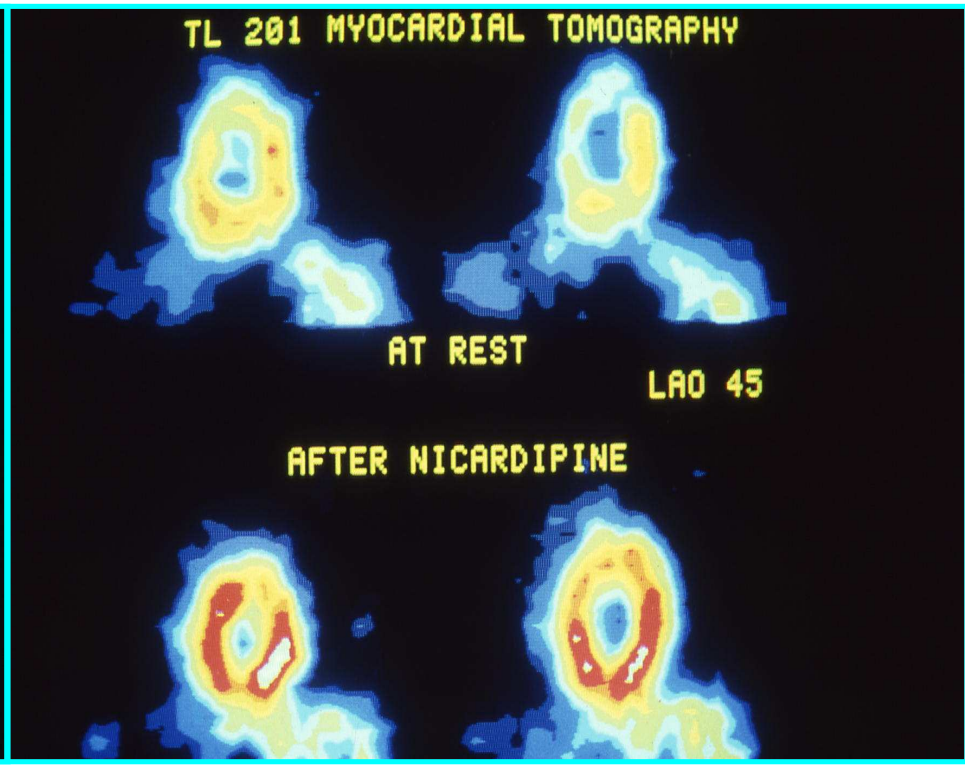


Thallium scintigraphy
Proximal coronary lesion : atherosclerosis



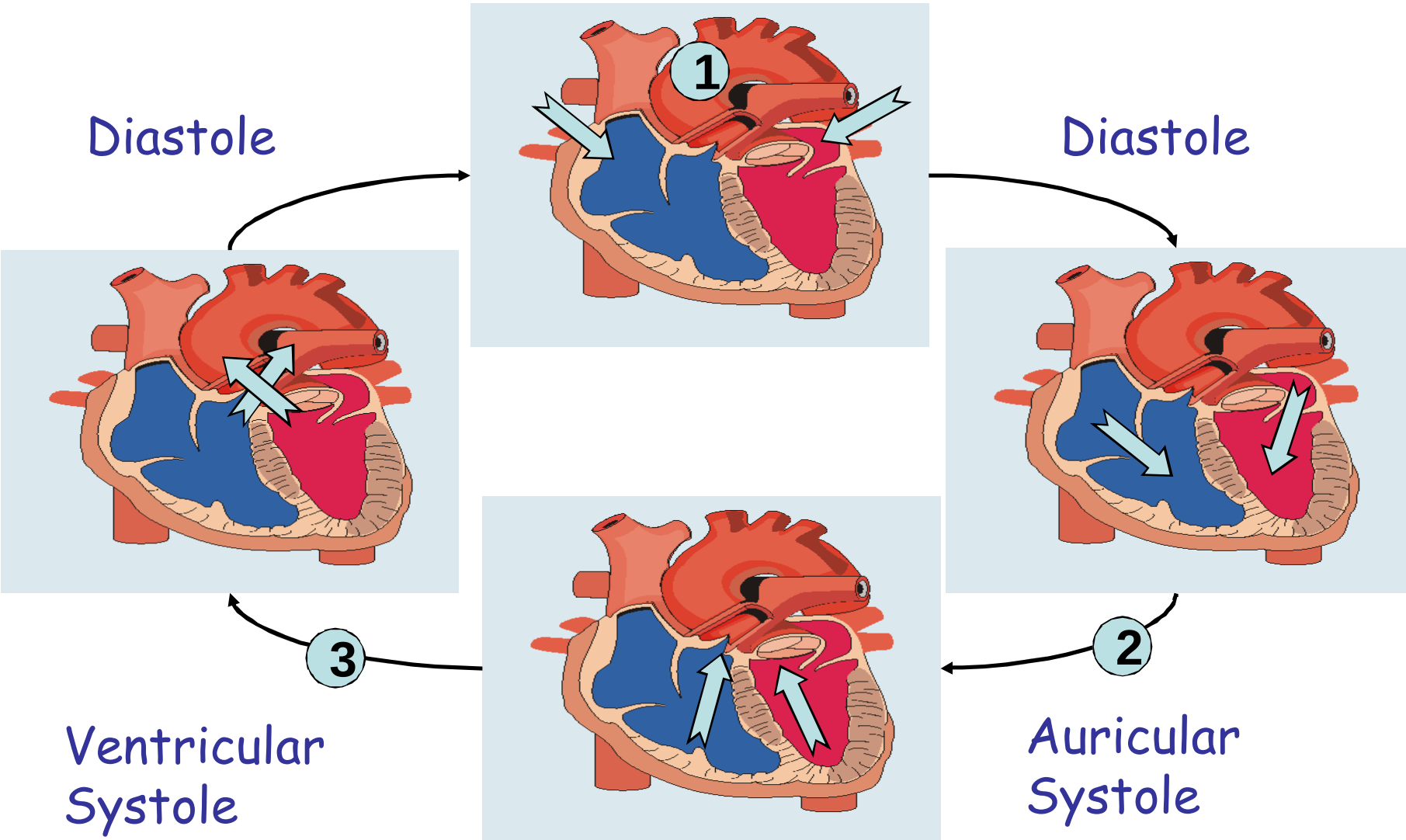
Distal coronary lesion: SSc



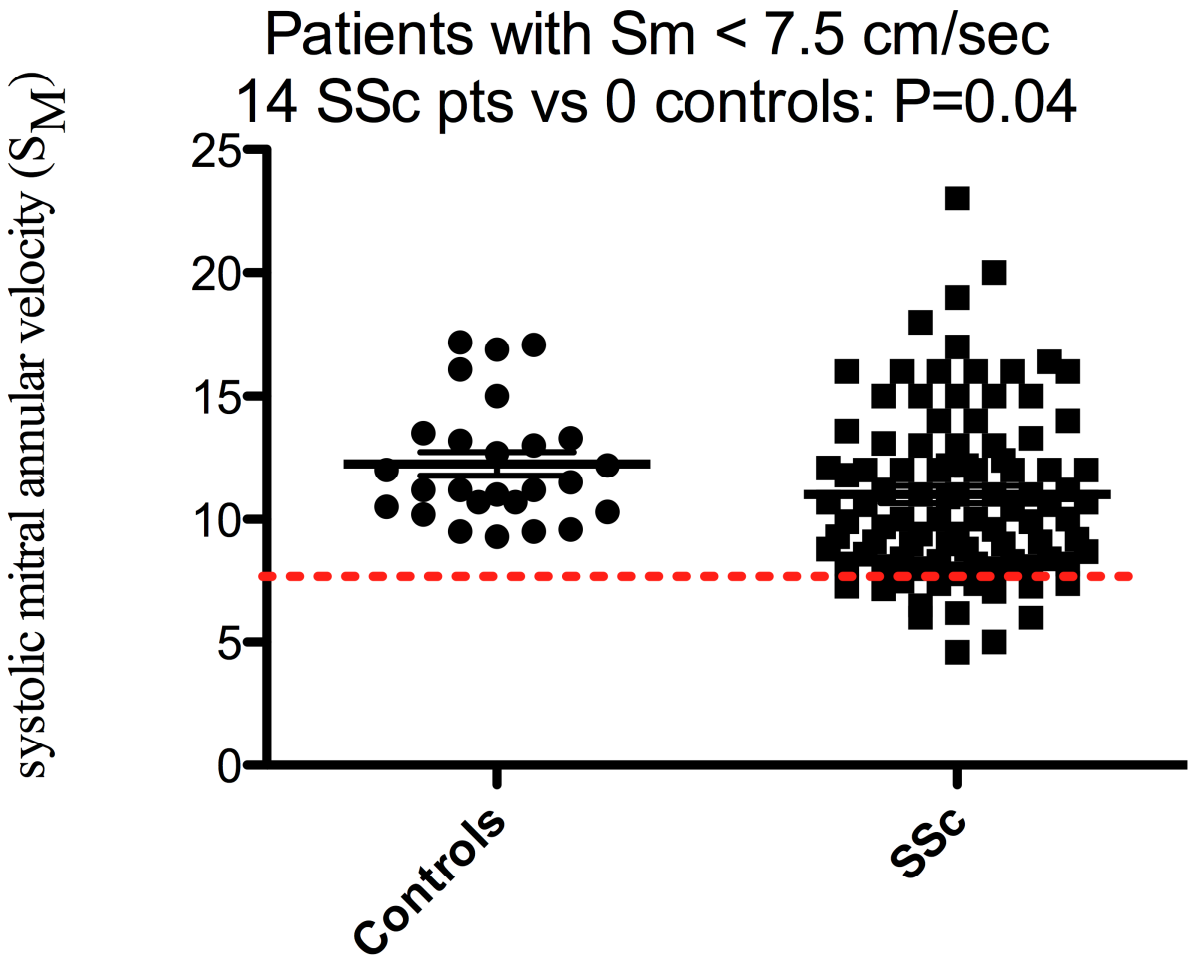


Raynaud’s phenomenon and arteriole vasospasm

Heart physiology

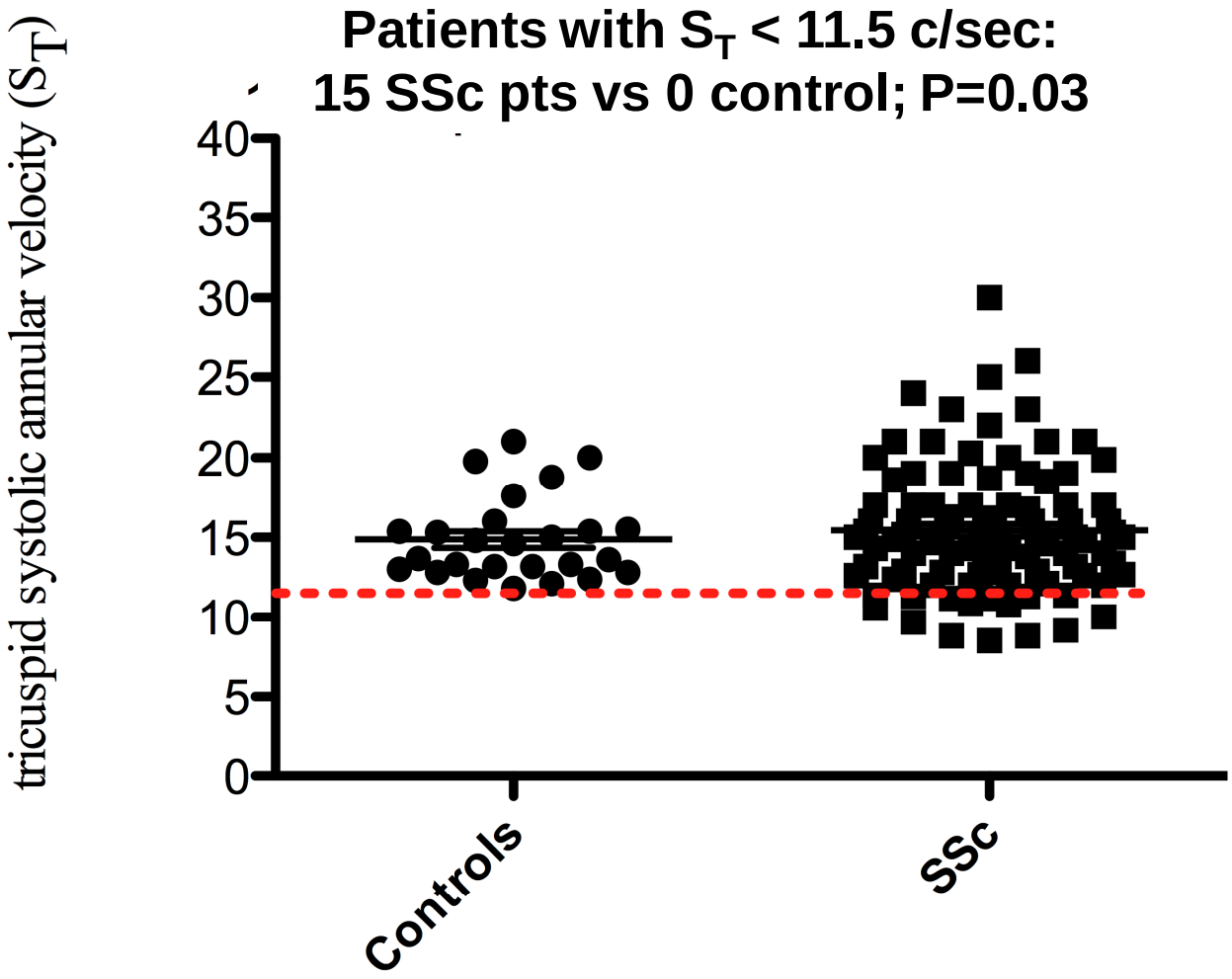


Doppler tissue echocardiography
LV contractility= LVEF



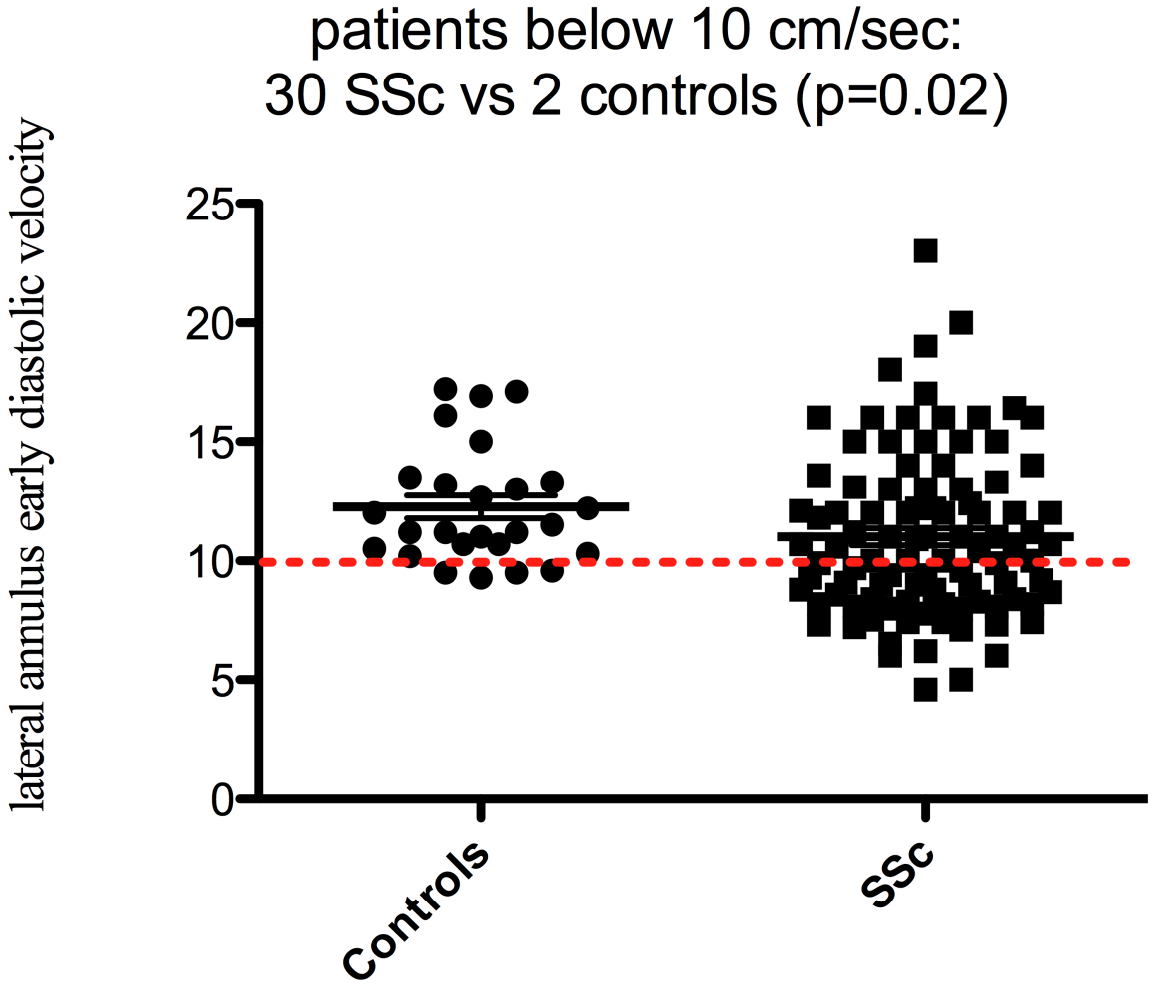
Meune et al, Arthritis Rheum 2008;58:1803-9

RV contractility: RVEF



Meune et al, Arthritis Rheum 2008;58:1803-9

LV diastolic function: LV filling

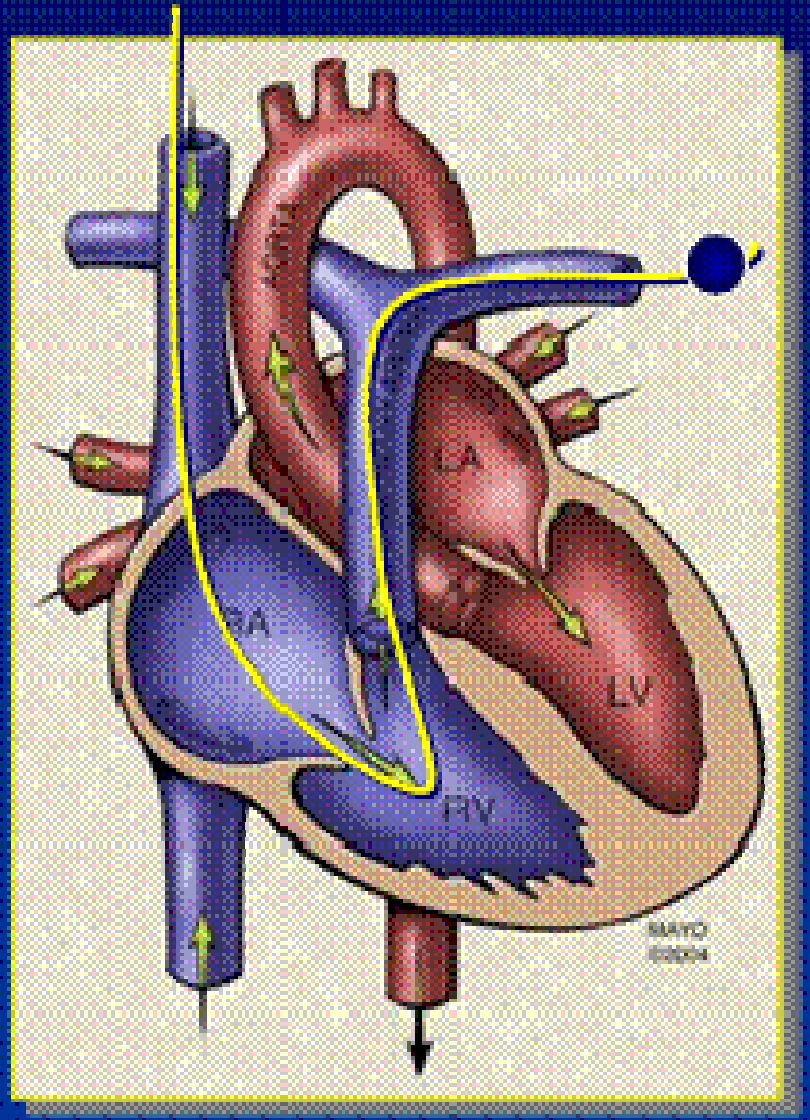


<u>Standard Echo:</u> 101 of 570 patients (18%) ***(De Groote et al, ARD 2008)*** **and 23 of 77 (29%)** ***(Maione et al, Seminars in Arthritis 2005)***

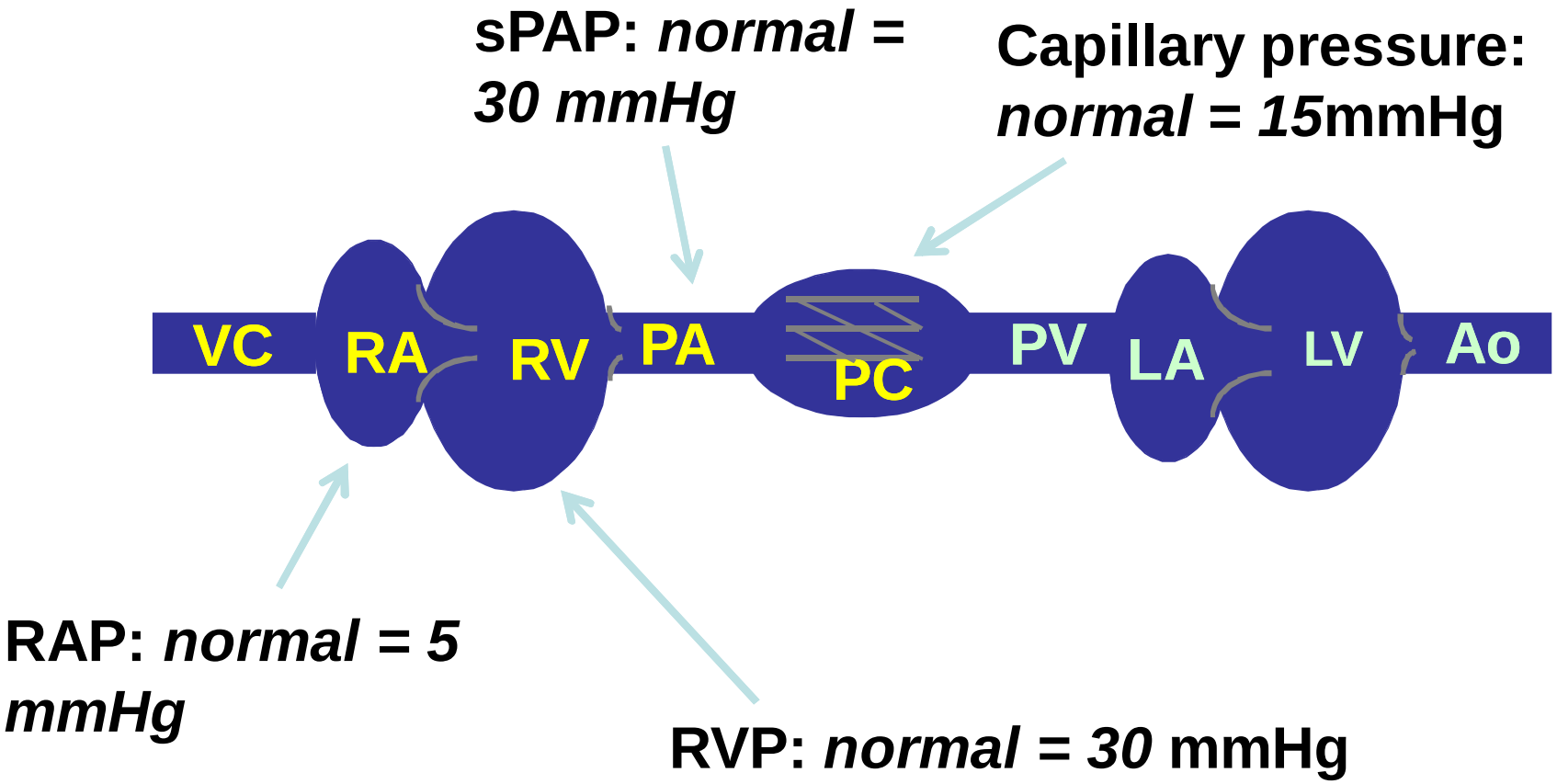
Cardiac Catheterization

A more invasive test in which the physician inserts a small tube called a "catheter" into a patient's blood vessel and passes the tube toward the heart

The cardiac catheterization measures important pressures in the heart and lungs

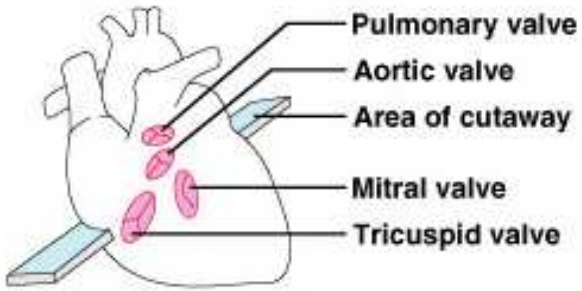


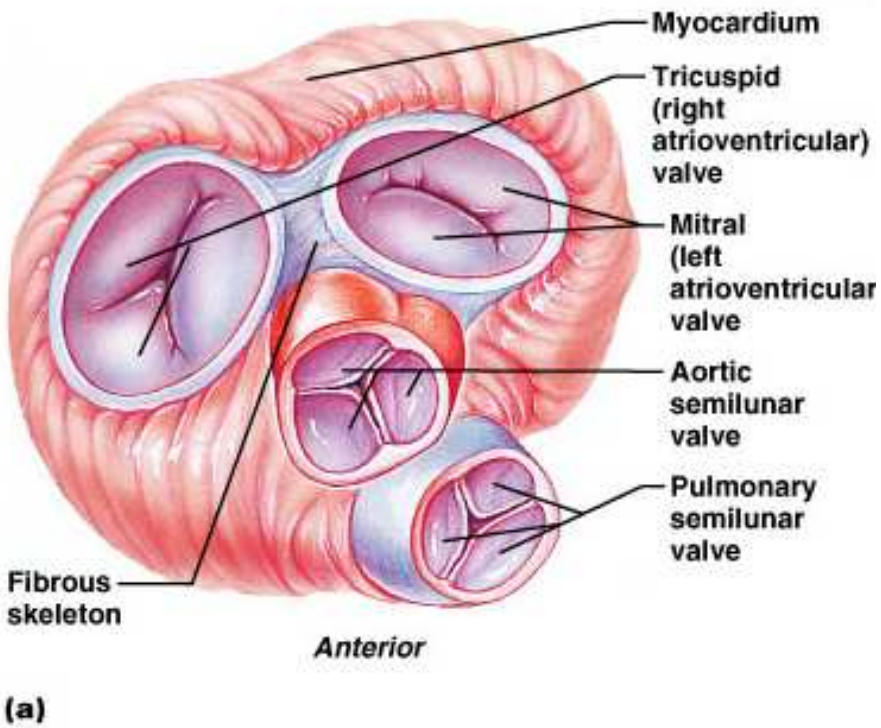
CATHETERISATION



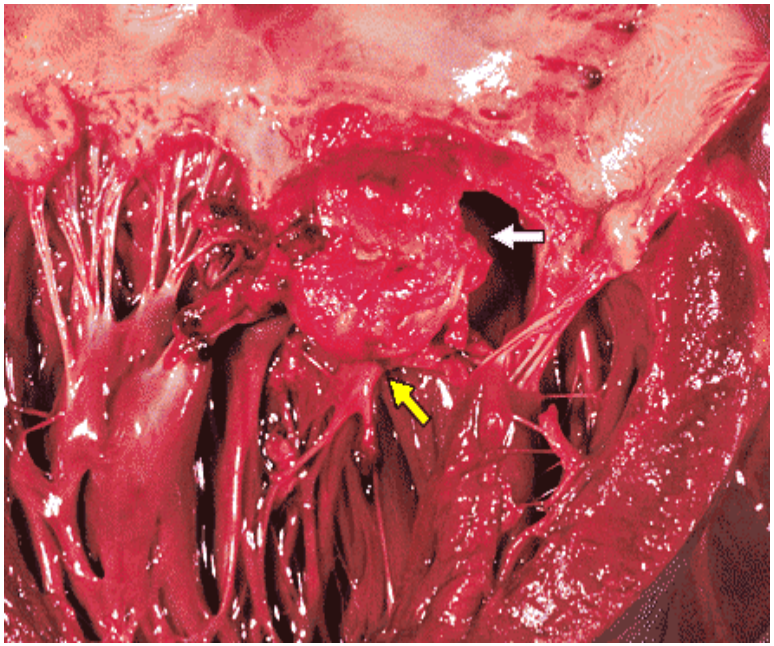
Heart Valves

allow blood flow in only one direction through the heart



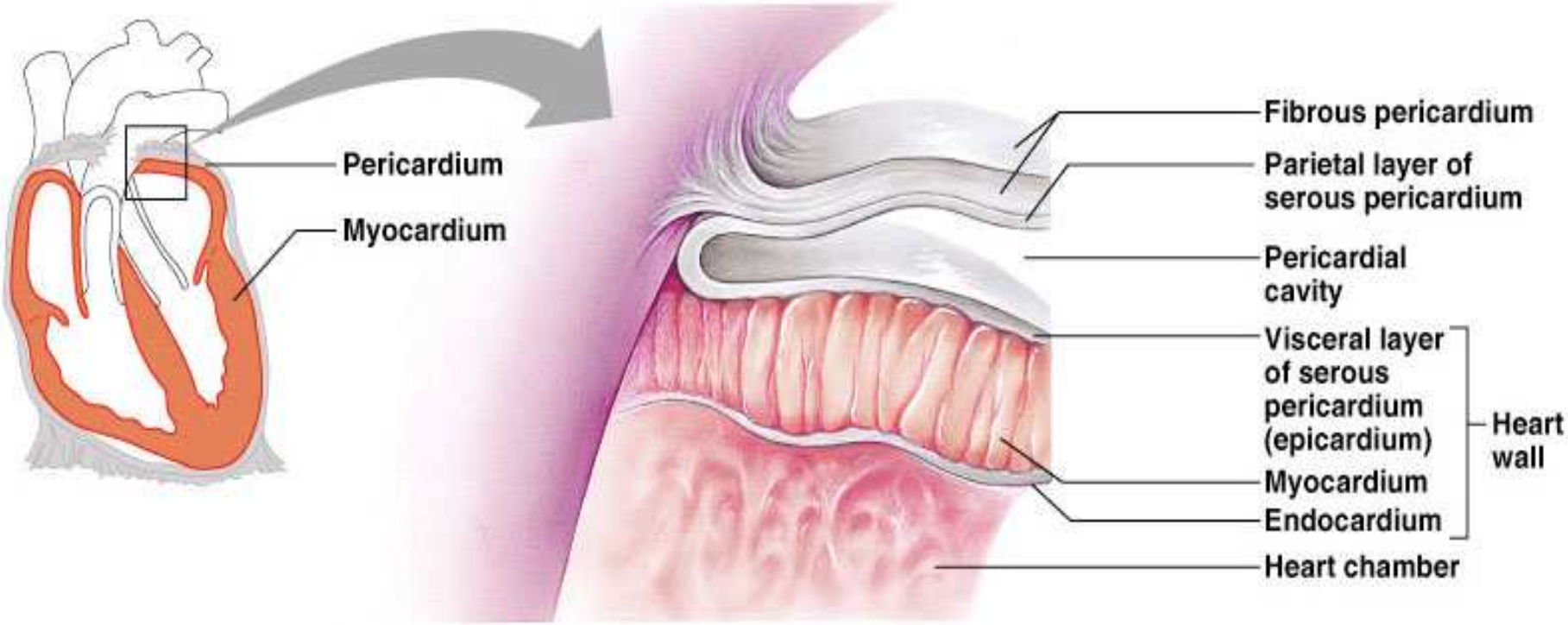


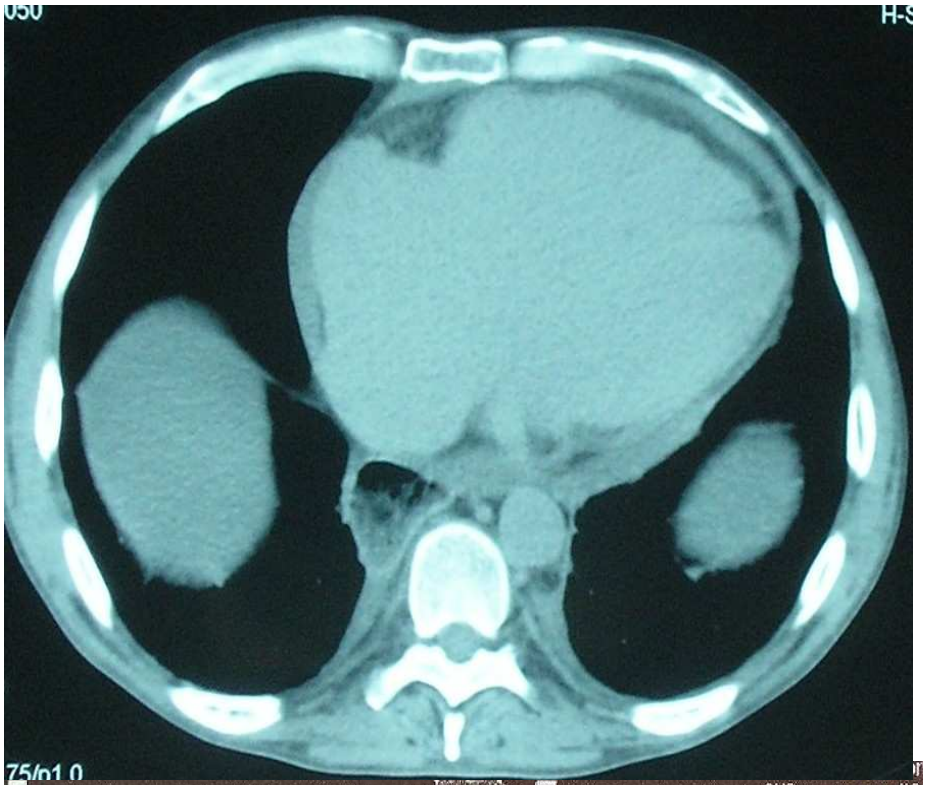
VALVULAR INVOLVEMENT

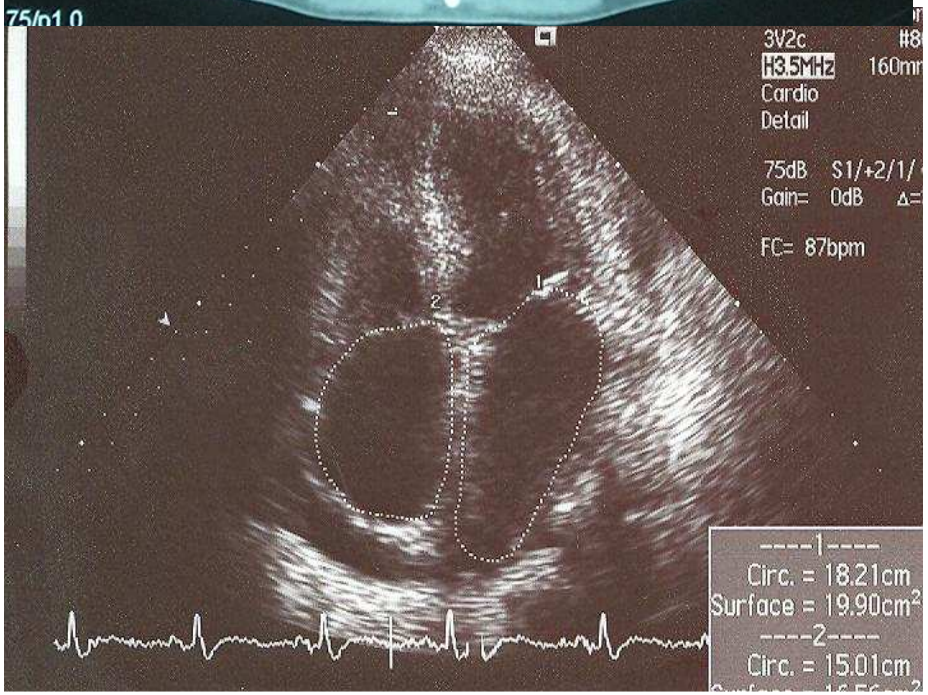


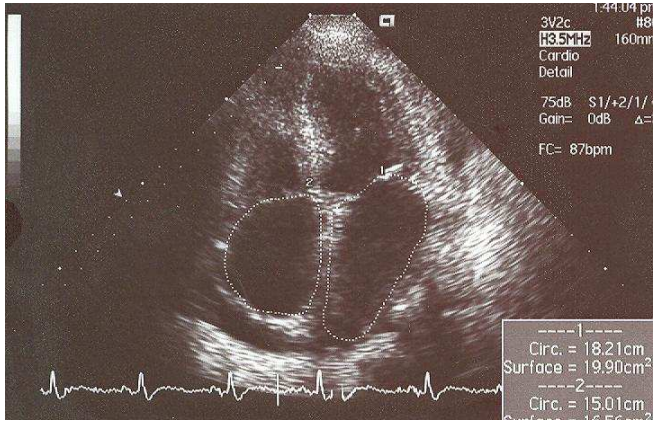
	SSc pts (n=100)	Controls (n=26)	p
Mitral valve			
Regurgitation	48	8	ns
Grade I	45	7	ns
Grade II	3	1	ns
Stenosis	2	0	
Prolapse	2	0	
Aortic valve			
Insufficiency	18	0	0.023
Grade I	15	0	
Grade II	3	0	
Stenosis	3	0	ns

Pericardial Layers of the Heart






3V2c
H3.5MHz
Cardio
Detail
75dB
Gain= 0dB
FC= 87bpm
----1----
Circ. = 18.21cm
Surface = 19.90cm²
----2----
Circ. = 15.01cm

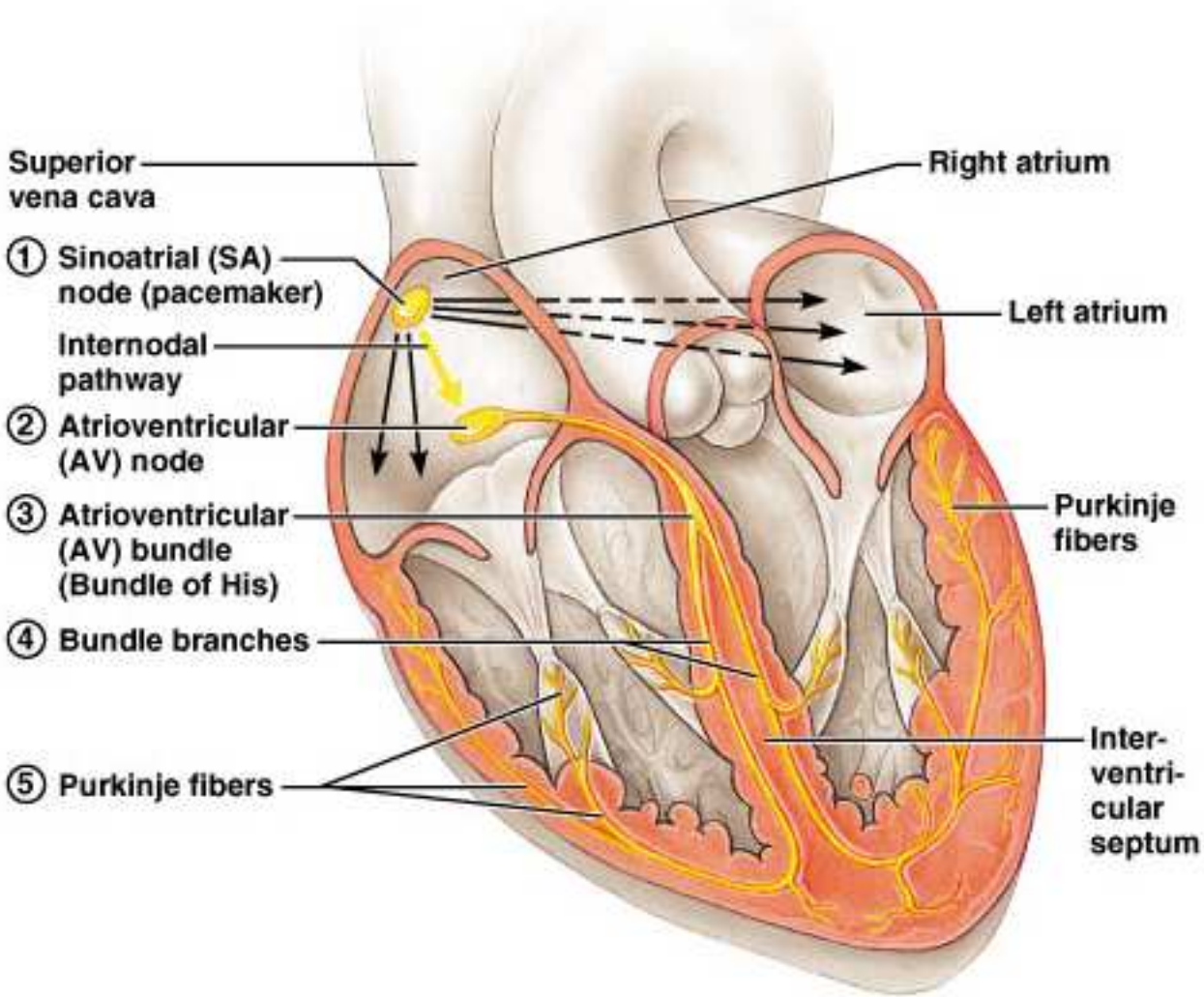


PERICARDIAL DISEASE

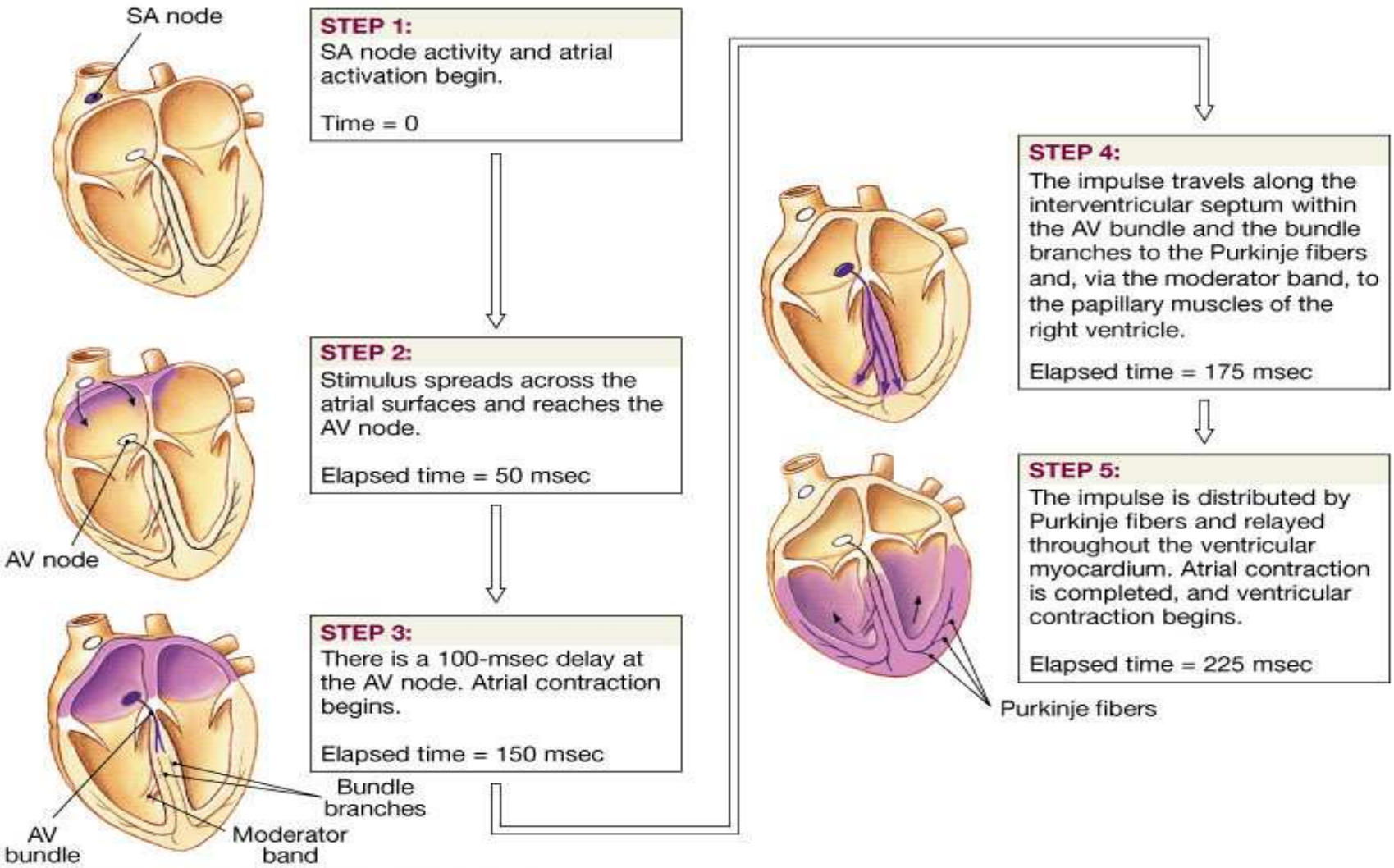
- **Autopsies: 33 to 72%**
- **Clinical controled studies:**
  - **33/77 (43%) SSc pts versus 2/45 (4%) controls, 11/77 (14%) with a significant effusion[1]**
  - **15/100 (15%) pts compared with 1/26 (4%) (NS) controls[2]**
- **Large or recurrent effusion: marker of disease activity, linked with renal crisis or PAH**

[1]Maione et al, Semin Arthritis Rheum 2005;34:721-7
[2]Meune et al, Arthritis Rheum 2008;58:1803-9

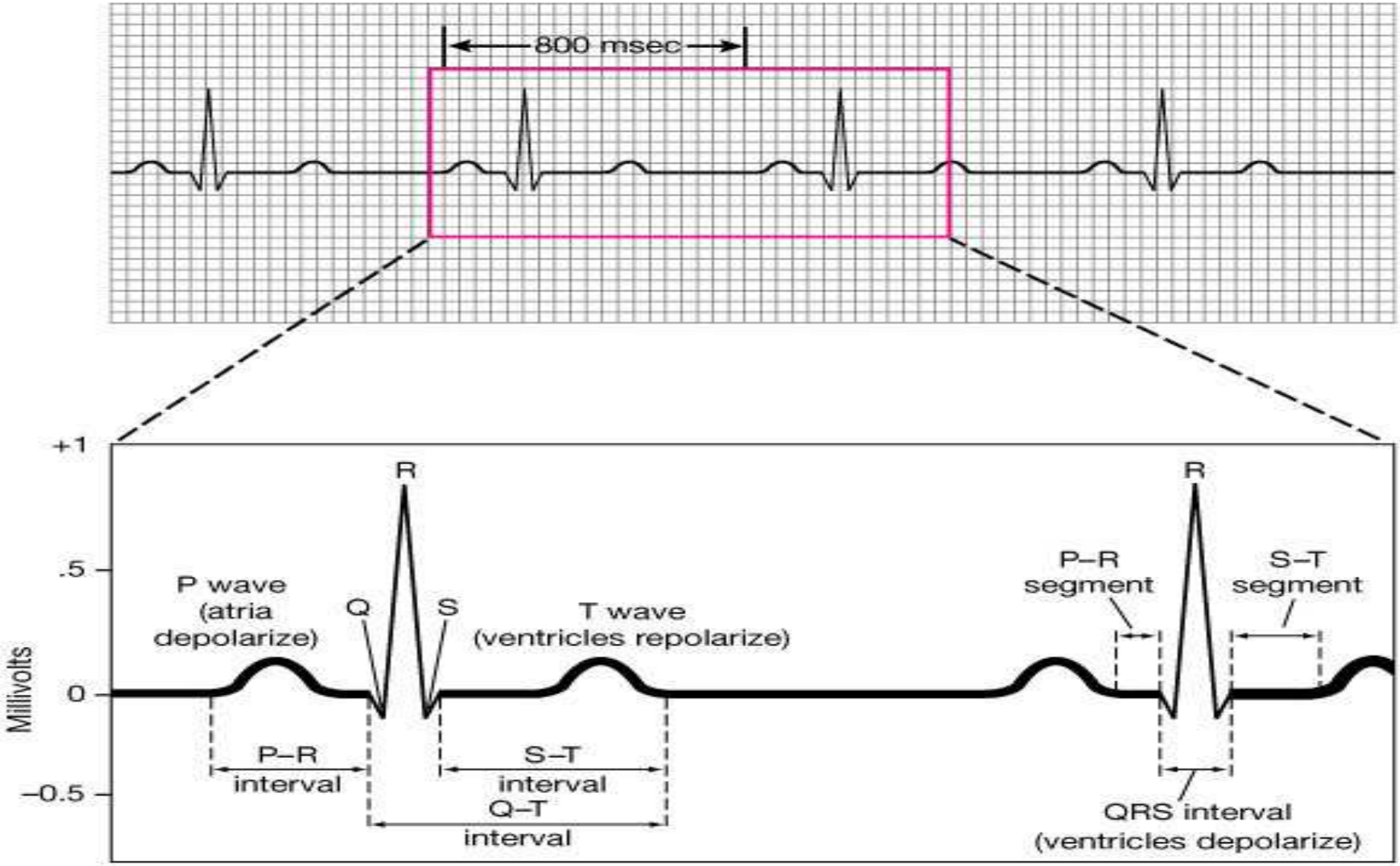
Conducting System



Impulse Conduction through the Heart



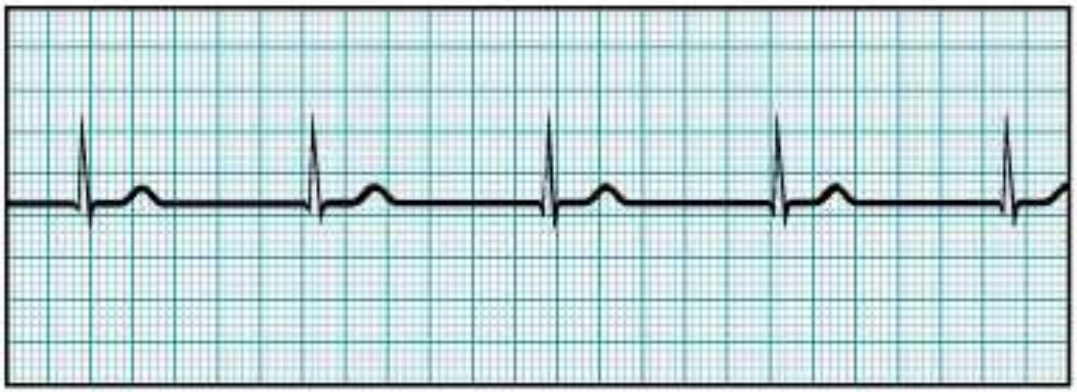
An Electrocardiogram



(b)

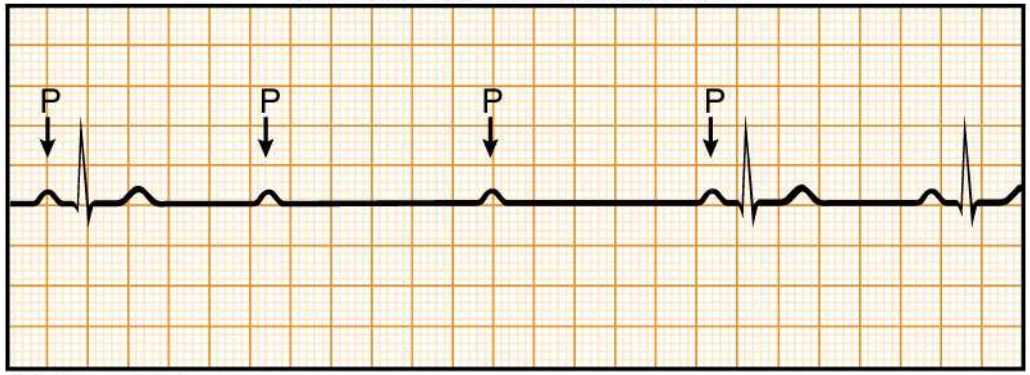
ECGs, Normal and Abnormal

(a) Sinus rhythm (normal)



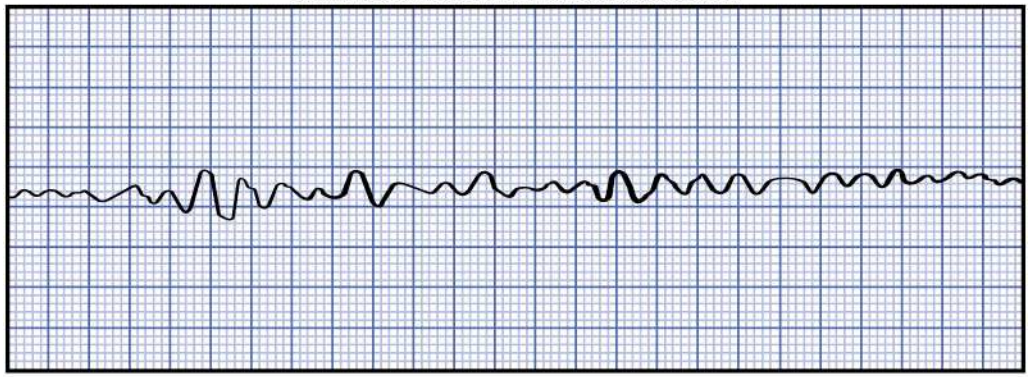
(b) Nodal rhythm – no SA node activity

ECGs, Abnormal



(c) Heart block

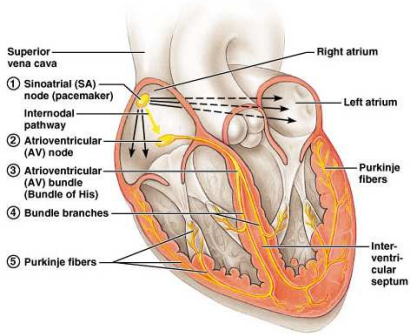
Arrhythmia: conduction failure at AV node



(e) Ventricular fibrillation

No pumping action occurs

CONDUCTION SYSTEM AND ARRHYTMIAS in SSc



- **Normal resting EKG in 50% of SSc patients**
  - **mild or aspecific abnormalities** (***Follansbee et al, Am J Med 1985;79:183)***
- **Holter EKG (n=50)** ***(Ferri et al, A&R 1985;28:1259)***
  - **20% coupled ventricular extrasystoles**
  - **10% non sustained ventricular tachycardia**
- **Higher risk in case of muscle and cardiac disease** ***(Follansbee et al, Am Heart J 1993;125:194)***

Treatments

- ✓ **Heart perfusion and function: calcium channel blockers, ACE inhibitors, diuretics**
- ✓ **Pericarditis: colchicine, NSAIDs, prednisone**
- ✓ **Conduction: pace-maker**
- ✓ **Arrhythmias:**
  - **Drugs: amiodarone**
  - **Implantable cardioverter defibrillator**

CONCLUSION
Heart involvement

- **Very common infra-clinical involvement: myocardium/pericardium/conduction system**
- **Bad prognosis when symptomatic**
- **Ischemia-reperfusion and microvascular involvement**
- **Long term benefit of vasodilators**

CONCLUSION: assessment

- **In routine:**
  - **Clinical examination: dyspnea, palpitations, chest pain**
  - **BNP or NT-proBNP**
  - **Doppler Echocardiography (and pulsed TDE)**
  - **ECG and 24h-Holter**
- **For research or difficult cases:**
  - **Cardiac MRI**
  - **Left or Right heart catheterisation**
  - **Tissular Doppler**