

Tricuspid and Pulmonic Valve Disease

The Forgotten Valves

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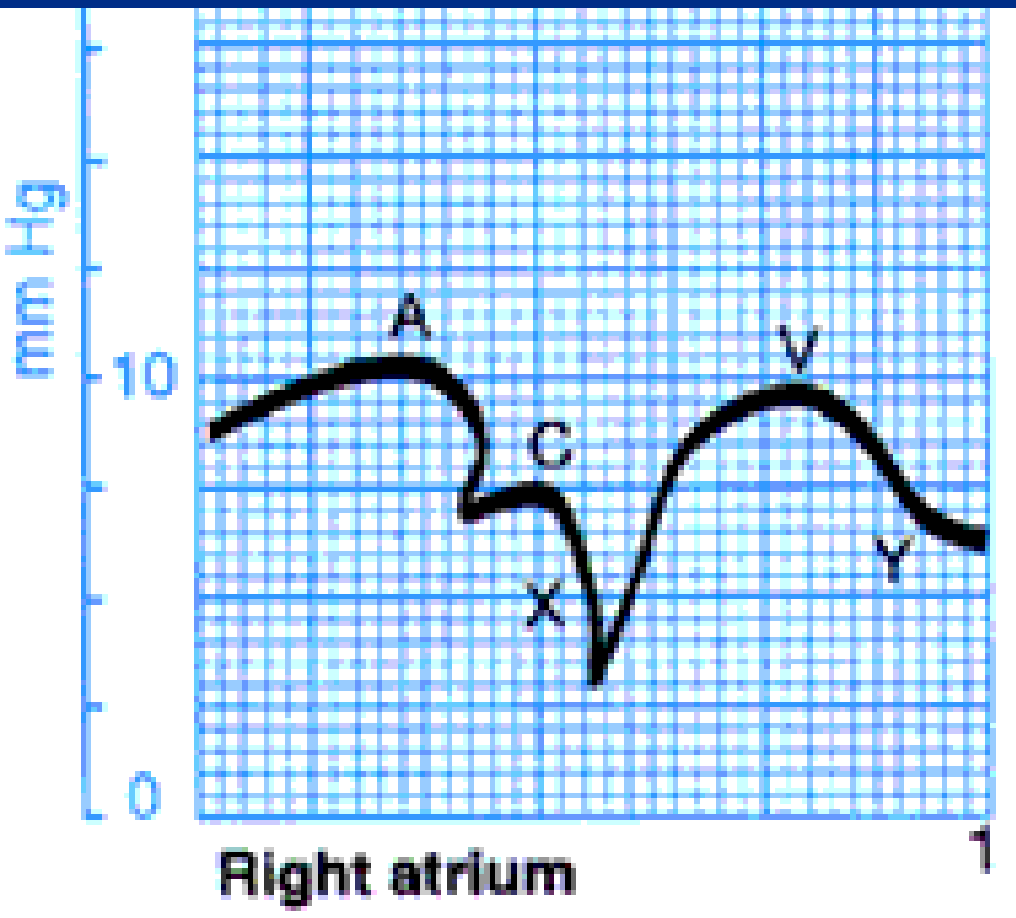
Goals of Conference

- Understand Tricuspid and Pulmonic valve stenosis and regurgitation
 - Diagnosis
 - Treatment options
- Specific conditions
 - Ebstein anomaly
 - Carcinoid Syndrome

A Few Basics

- Venous Waveform
- Understanding Echo terminology with respect to pressure gradients and stenosis

Right Atrial Waveform

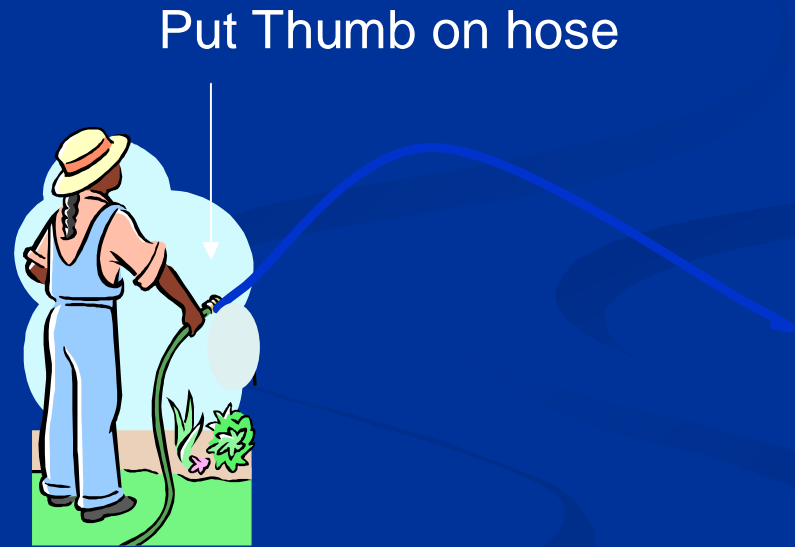


- a wave** - RA contraction
elevated in RV failure
- c wave** - tricuspid closure
- v wave** - passive filling of RA during ventricular systole = T wave on ECG
elevated in tricuspid regurgitation
- x descent** - atrial diastole
- y descent** - atrial emptying

Understanding Stenosis and Pressure Gradients

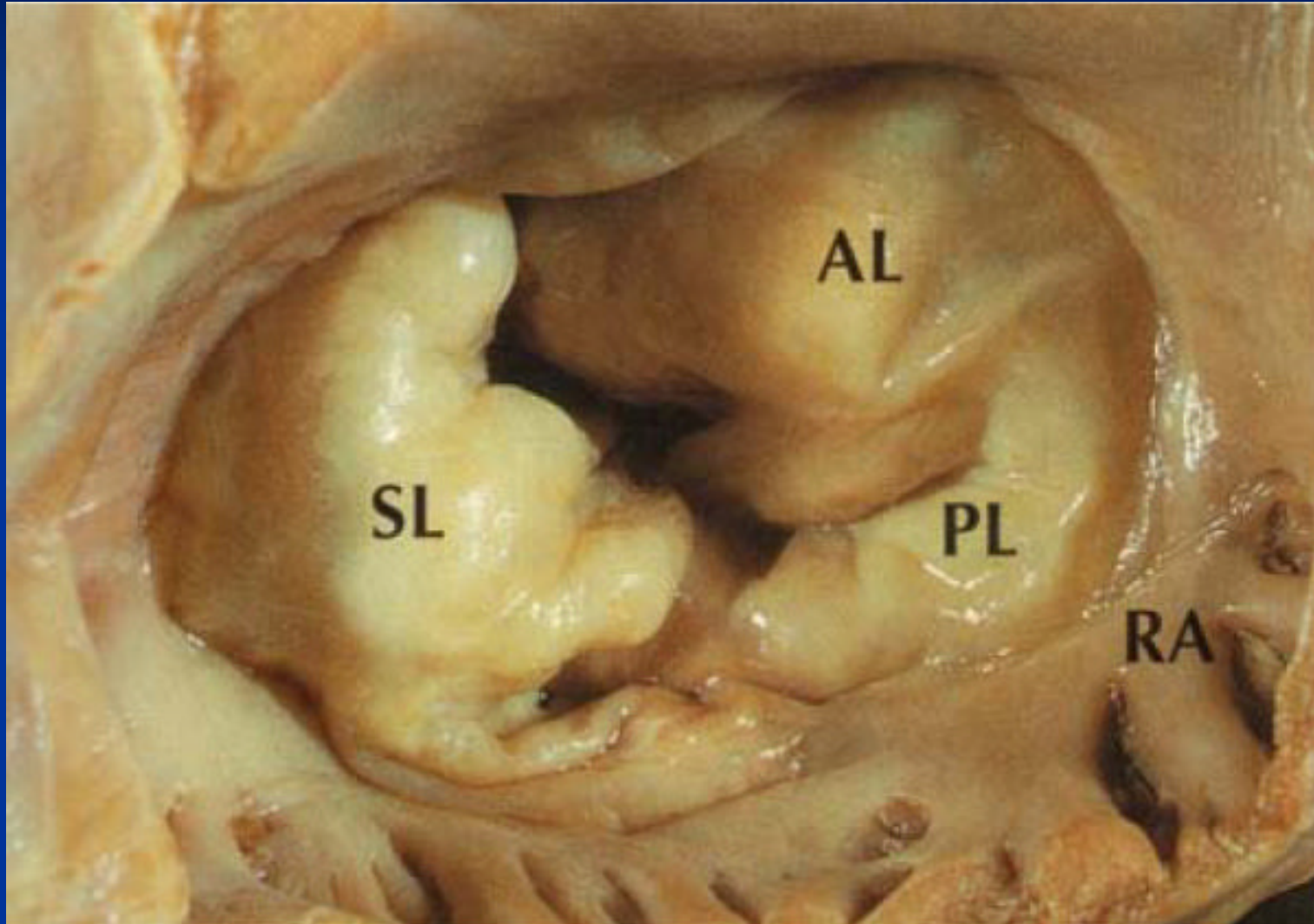


No “Stenosis” at the end of the hose
No SIGNIFICANT Pressure gradient
No impedance to water flow



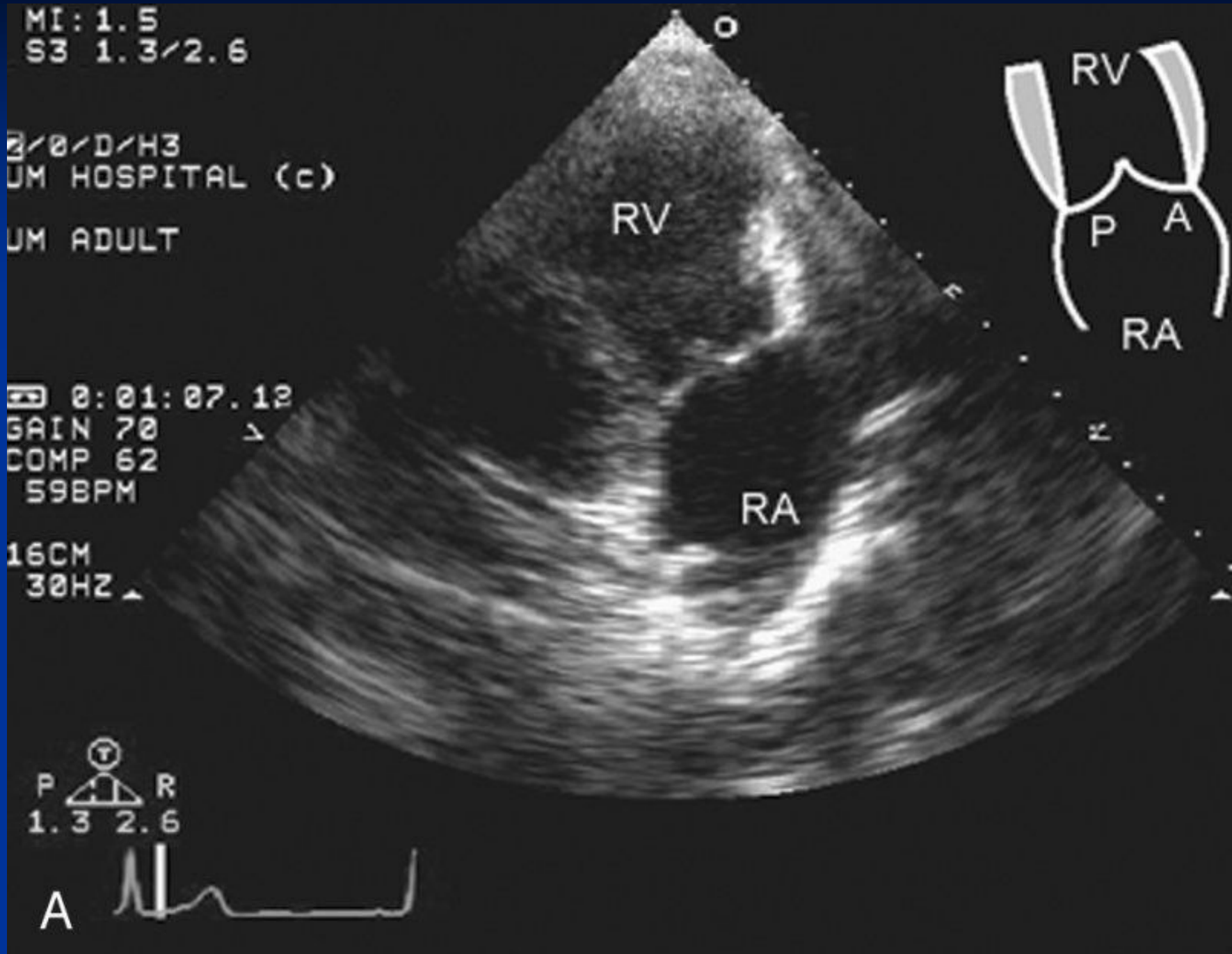
Now there is a “Stenosis” at the end of the hose
SIGNIFICANT Pressure gradient across the thumb
Water flow impeded, creating increased velocity

Tricuspid Valve Anatomy

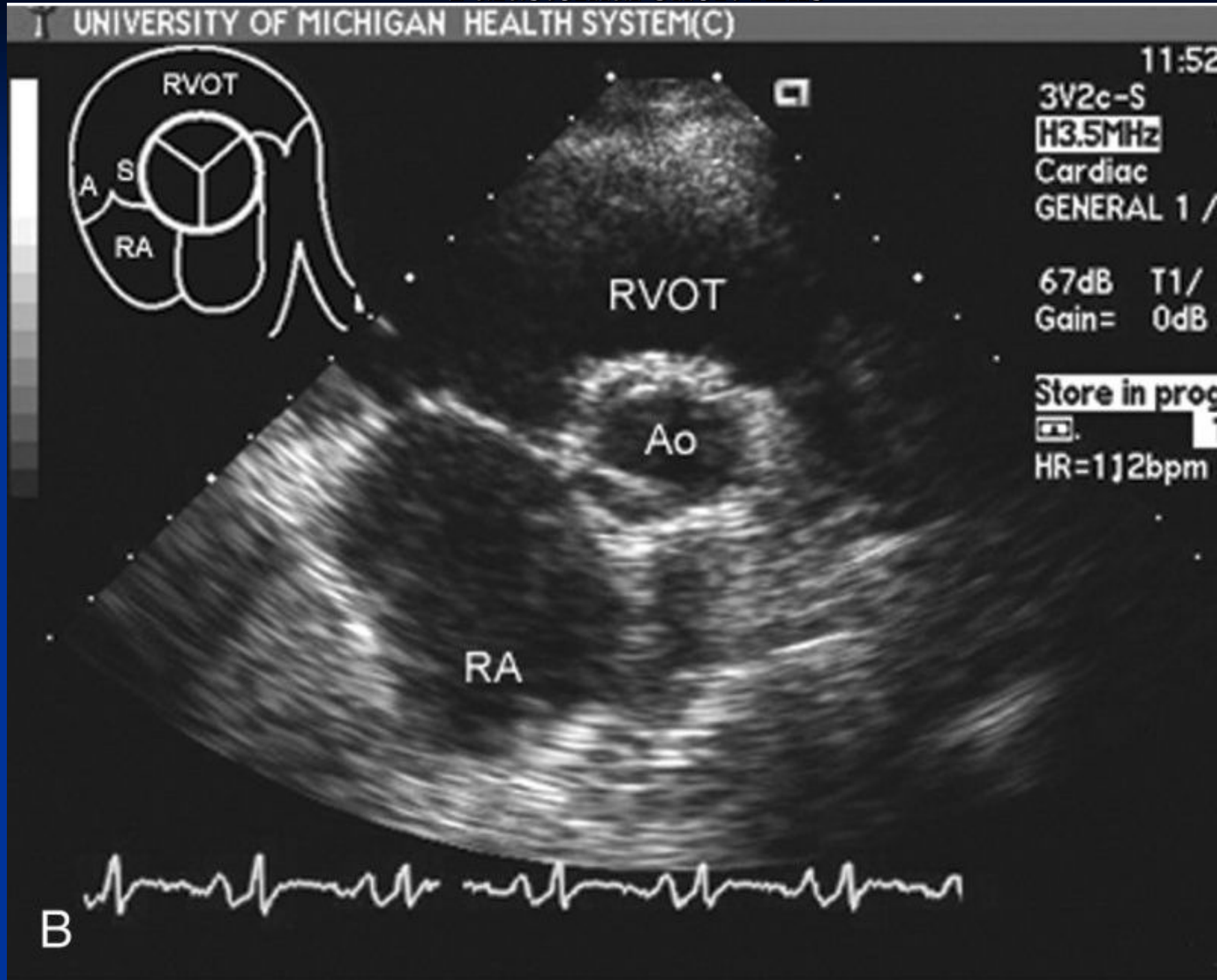


SL – Septal leaflet, AL – Anterior leaflet, PL – Posterior leaflet

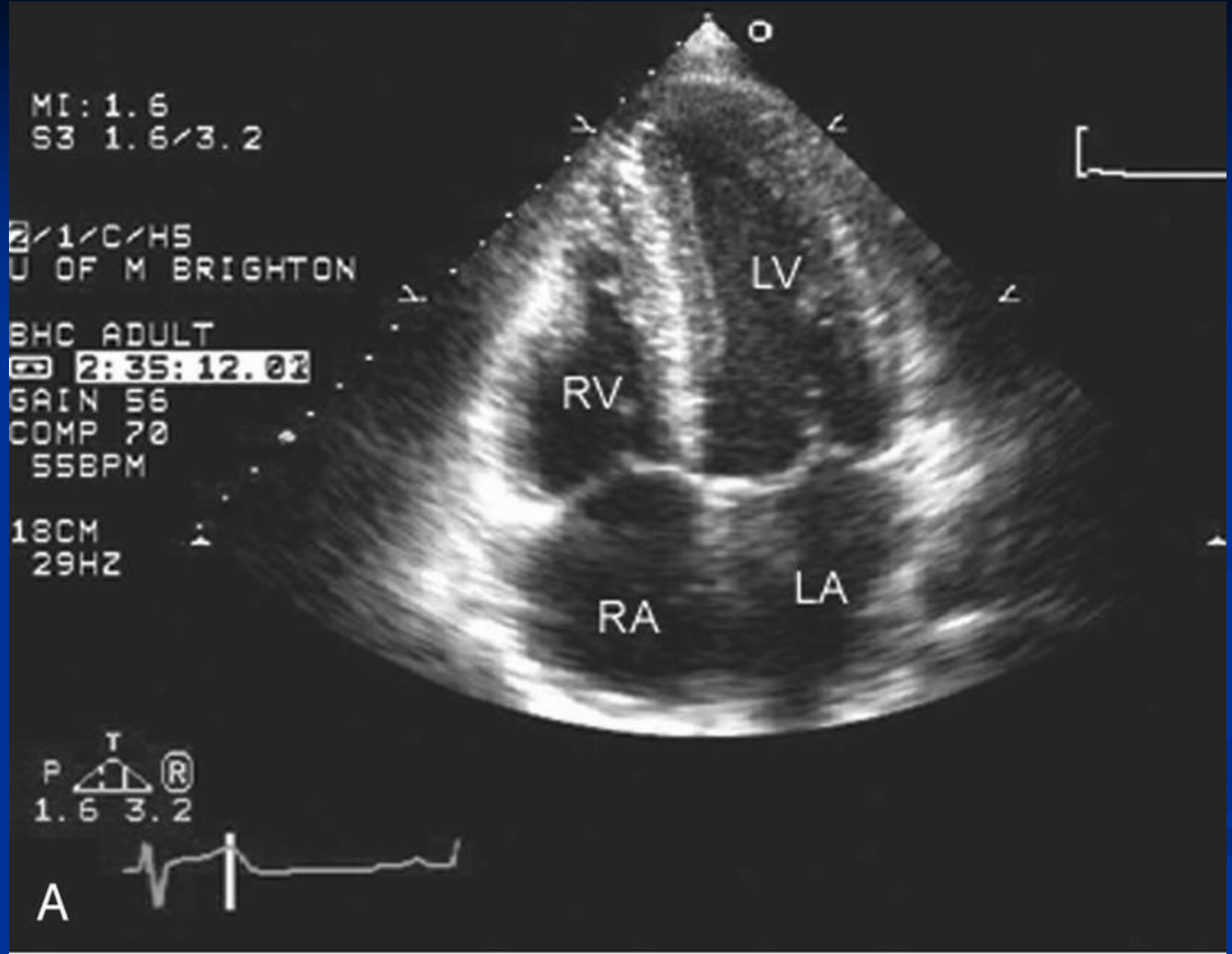
Right Ventricle Inflow



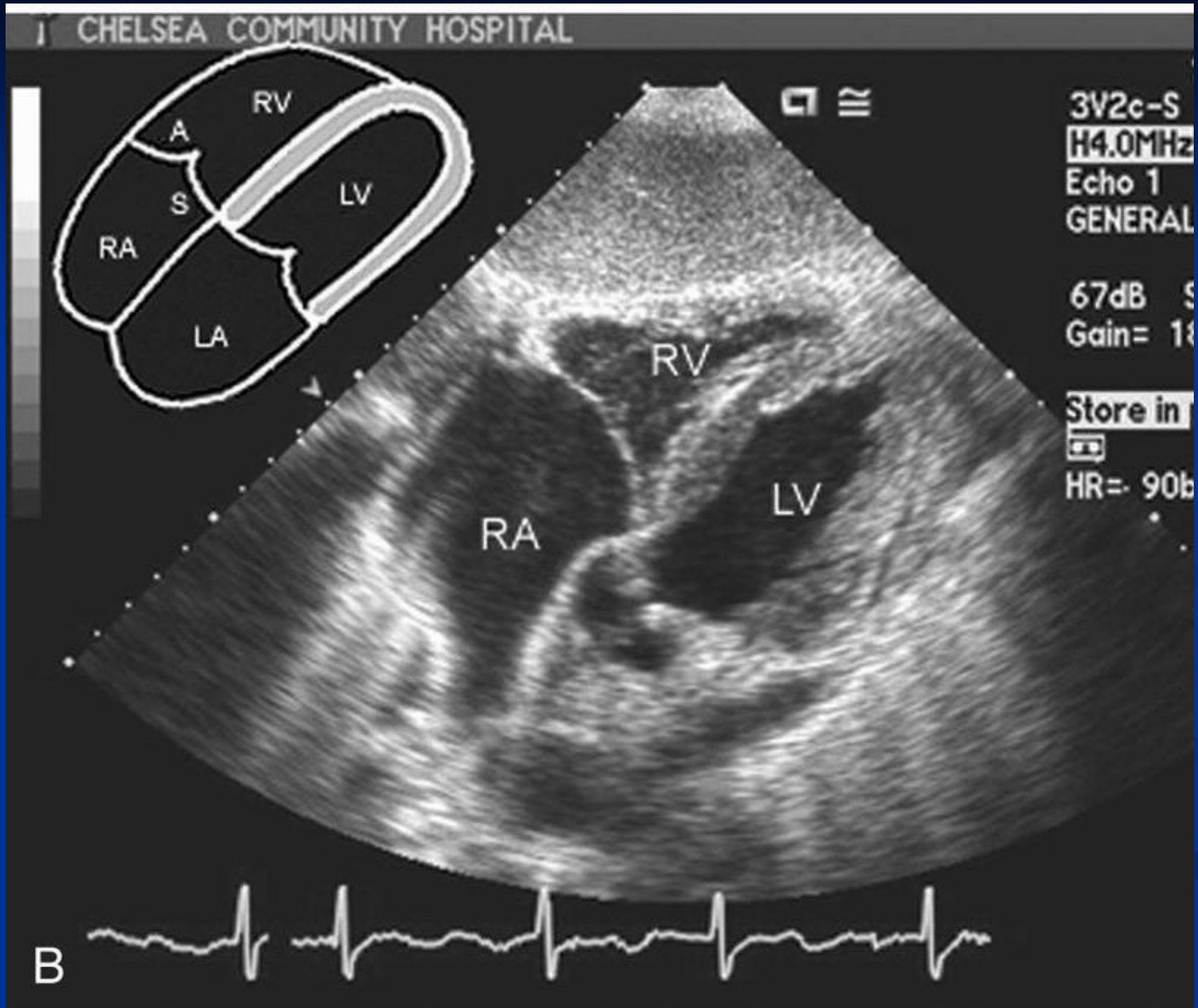
Parasternal Short Axis



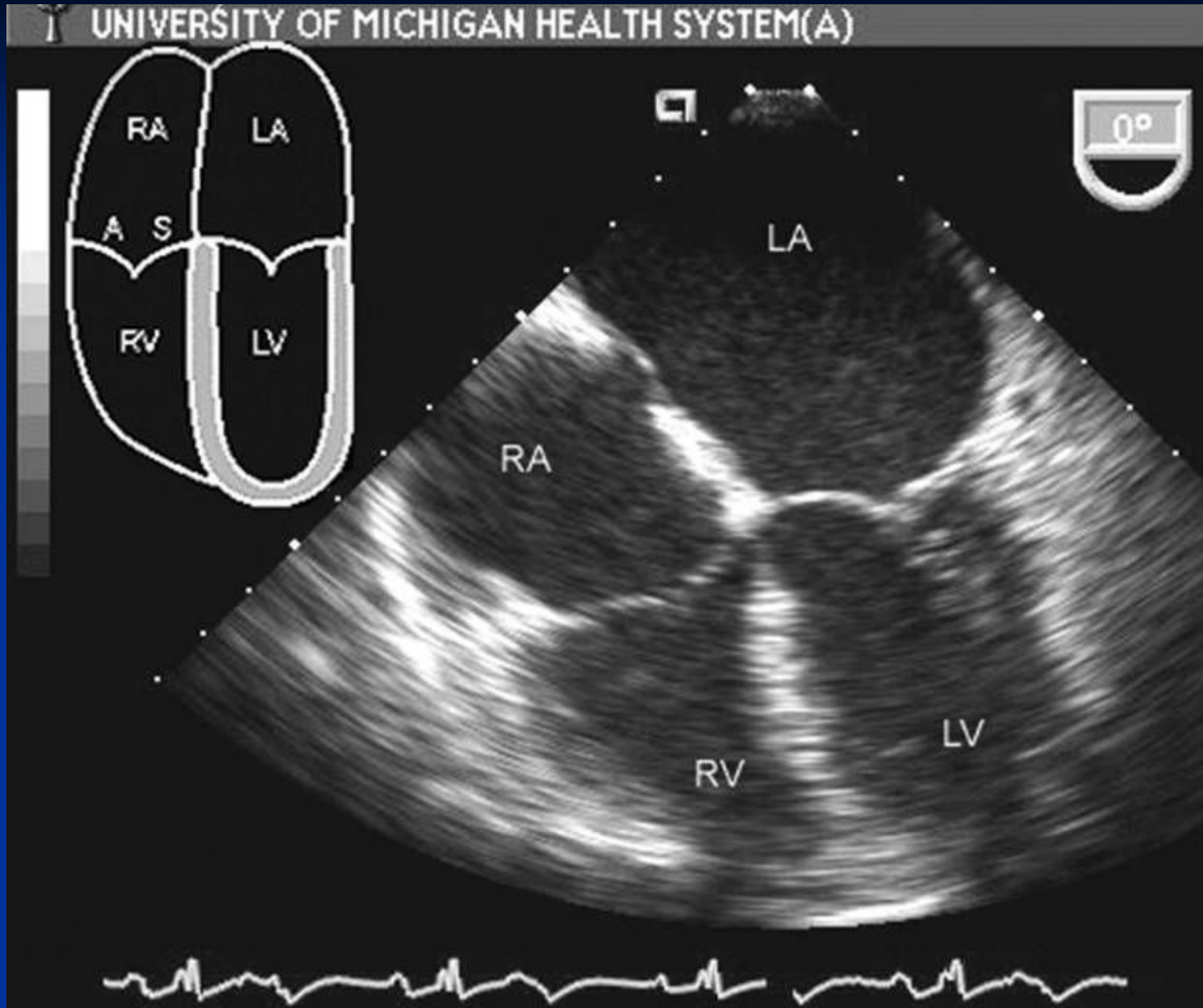
Apical 4 Chamber



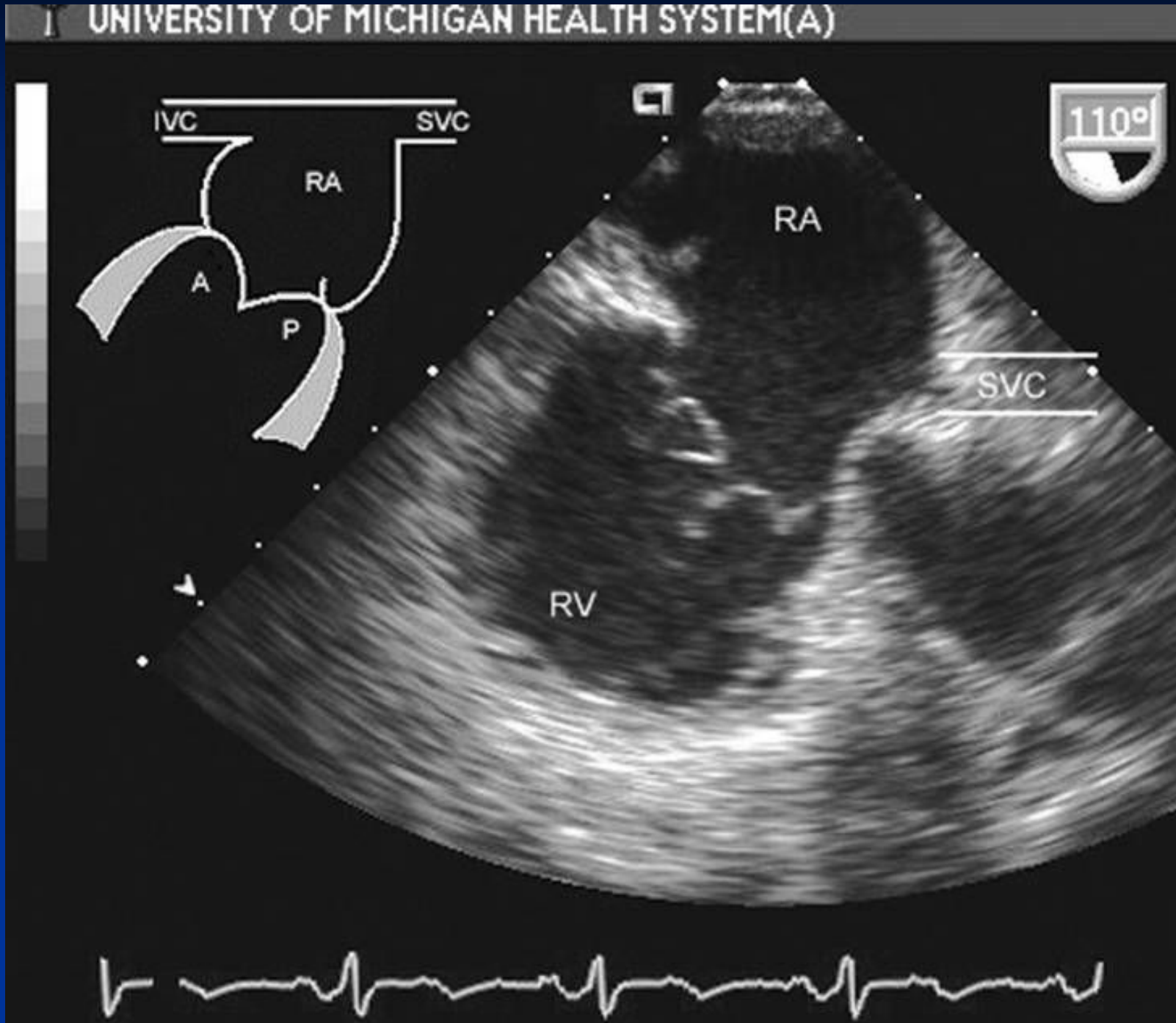
Subcostal



TEE 4 chamber



TEE RV inflow



Tricuspid Stenosis

- Etiology
 - Almost always rheumatic
 - Other causes are rare
 - Congenital tricuspid atresia
 - Right atrial tumors
 - Carcinoid syndrome
 - More often tricuspid regurgitation
 - Endomyocardial fibrosis
 - Vegetations
 - Pacemaker lead
 - Extracardiac tumors

Rheumatic Tricuspid Stenosis

- At autopsy, seen in 15% of patients with rheumatic heart disease
 - But clinically significant in only 5%
- Isolated TS is rare
 - Almost always mitral valve involvement
 - Also aortic valve involvement common

Pathologic Findings

Rheumatic Tricuspid Stenosis

- Similar to mitral stenosis
- Fusion and shortening of chordae tendineae
- Fusion of leaflet edges
- Calcification is rare
- Right atrial dilatation, wall thickening

Pathophysiology

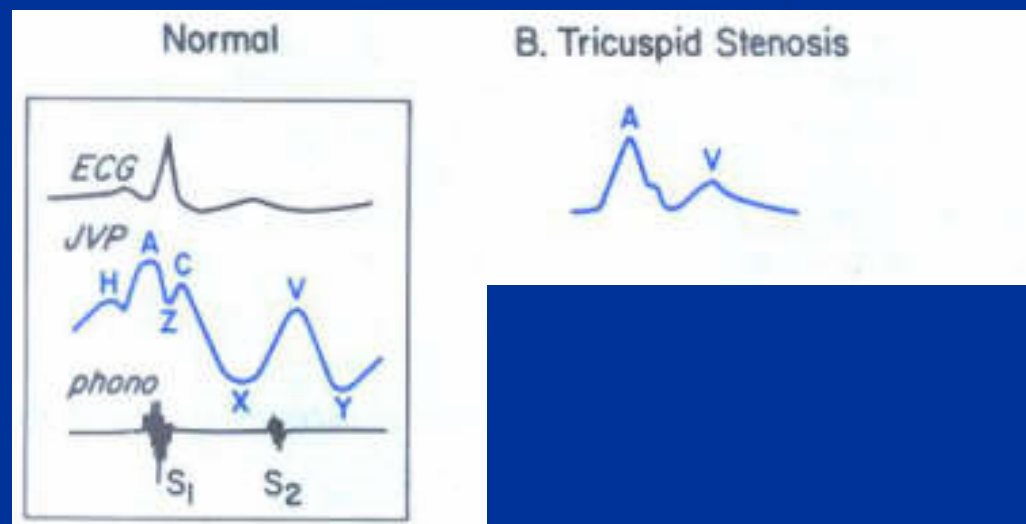
- Mean pressure gradient across tricuspid valve of 5mmHg is enough to cause symptoms
 - Jugular venous distension
 - Ascites
 - Edema
- Mean pressure gradient of 2mmHg is sufficient for diagnosis
 - Augmented by inspiration, fluid status

Symptoms

- Fatigue
- Abdominal discomfort
 - Hepatomegaly
 - Ascites
 - Anasarca
- Neck discomfort
- Dyspnea, orthopnea rare with isolated TS
 - More often these symptoms due to mitral valve disease

Physical Examination

- Diastolic rumble at left lower sternal border
 - Increased with inspiration
 - Often confused with mitral stenosis
- Tricuspid opening snap
- Neck vein distention
 - Prominent a wave
 - Slow y descent
- Hepatic pulsation
- Ascites
- Aortic and Mitral murmurs



Maneuvers to Change intensity of Murmur in Tricuspid Stenosis

- Increase Murmur
 - Inspiration
 - Mueller maneuver
 - Right lateral decubitus position
 - Leg raises
 - Squatting
 - Isotonic exercise (hand grip)
 - Amyl nitrite inhalation
- Decrease Murmur
 - Expiration
 - Strain phase of Valsalva

Diagnostic Evaluation

- Echocardiography
 - Diastolic doming of leaflets
 - Thickening of leaflets, restricted motion
 - Doppler echo shows prolonged antegrade slope
- EKG
 - Right atrial enlargement
- Chest Xray
 - Right atrial enlargement
- Catheterization
 - Largely replaced by Echo
 - Hemodynamics
 - Angiography of RA shows a small diastolic jet

Management

- Sodium restriction
- Diuretics
- Surgical management
 - Mean pressure gradient of 5mmHg
 - Valve orifice area of $\leq 2.0\text{cm}^2$
- Often coexistent Mitral disease
 - TS + MS – fix both at same time

Surgical Techniques

- Open valvotomy (Create a functional bicuspid valve)
 - Open commissures between anterior-septal and septal-posterior leaflets
 - Not between anterior and posterior leaflets
 - Leads to severe regurgitation
- Bioprosthetic valve replacement
 - Preferred over mechanical due to risk of thromboembolism
- Balloon valvuloplasty?

Tricuspid Regurgitation

Etiology

- Secondary (functional) TR
- Most Common
 - Structurally Normal Valve
 - Due to dilatation of right ventricle
 - Commonly due to mitral valve disease
 - RV systolic pressure more than 55mmHg will cause TR
 - Other etiologies
 - Pulmonary hypertension
 - Eisenmenger's syndrome/Congenital
 - RV infarction

Tricuspid Regurgitation

Etiology

- Primary Causes (abnormal valve)
- Less Common
 - Infective endocarditis
 - Ebstein anomaly
 - Prolapse (Floppy Valve)
 - Carcinoid
 - Rheumatoid Arthritis
 - Radiation Injury
 - Connective Tissue Disease
 - Cardiac Tumors (RA Myxoma)
 - Pacemaker leads
 - Repeated Endomyocardial biopsy
 - Endomyocardial fibrosis
 - Methysergide valvular disease
 - Fenfluramine-phentermine
 - Lupus

Carcinoid Syndrome

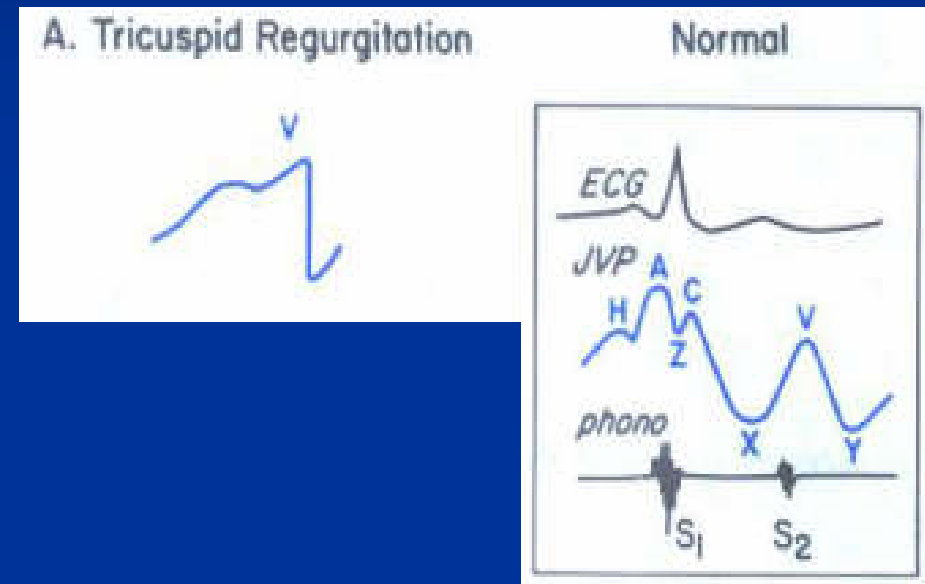
- Tricuspid regurgitation or combined stenosis/regurgitation
- Deposits of fibrous tissue on endocardial surface of valvular cusps
- Usually ventricular surface of tricuspid valve
- Causes adhesion to right ventricular wall, creating regurgitation

Symptoms

- Generally well tolerated
- Pulmonary hypertension + TR = decreased cardiac output
- Fatigue
- Abdominal discomfort
 - Hepatomegaly
 - Ascites
 - Anasarca
- Neck discomfort

Physical Examination

- Jugular venous distension
 - Prominent c-v wave
 - Sharp y descent
- Murmur
 - Associated pulmonary HTN
 - Pansystolic murmur
 - Left lower sternal border
 - Loud P2
 - Without pulmonary HTN
 - Low intensity
 - Murmur in 1st half of systole



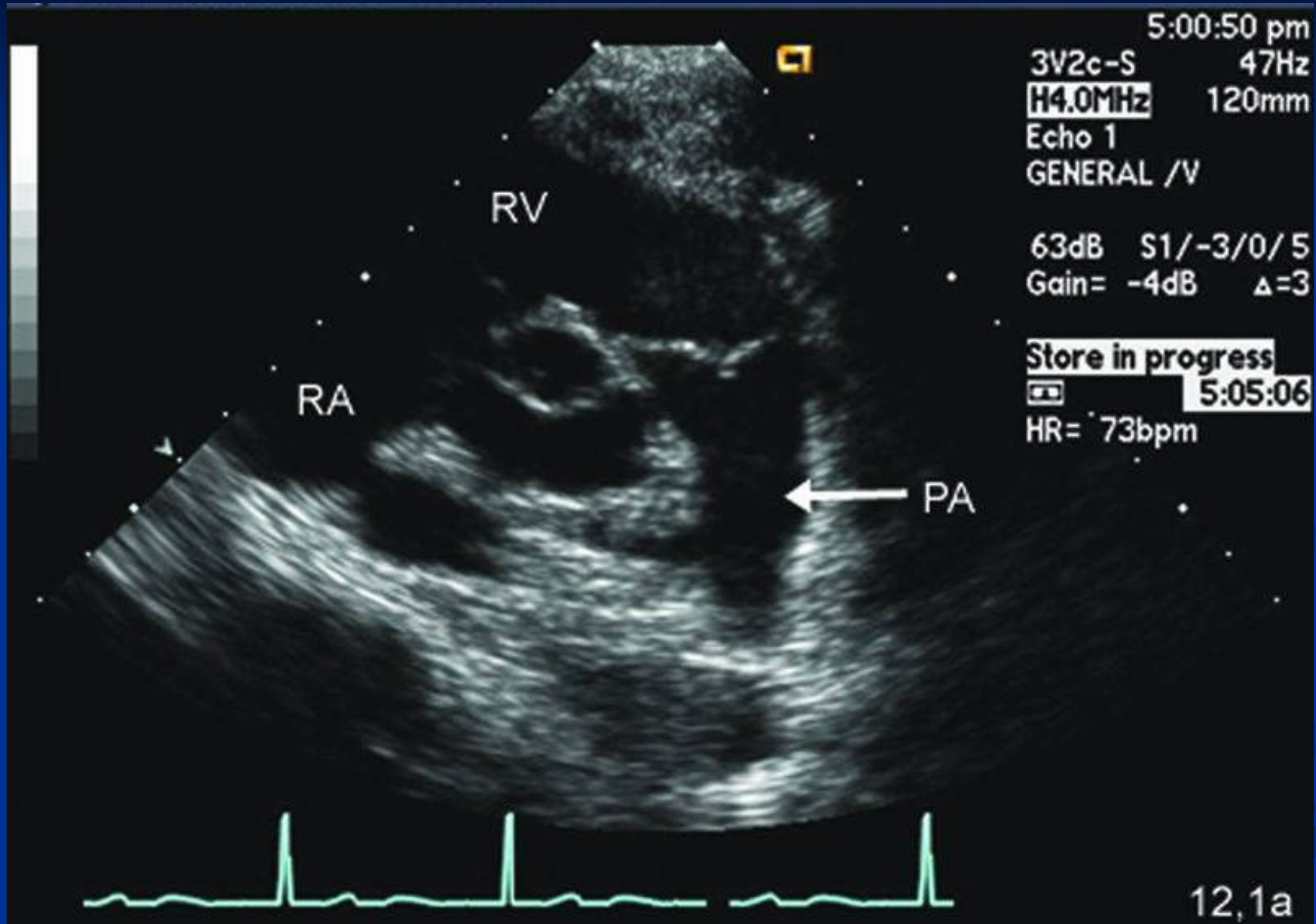
Diagnostic Evaluation

- Echocardiography
 - Tricuspid regurgitation severity
 - Right ventricular function
 - Pulmonary artery pressure
- EKG - nonspecific
 - Incomplete RBBB
 - Atrial fibrillation
- Chest Xray
 - Cardiomegaly, right atrial enlargement
- Catheterization/Hemodynamics
 - RV systolic pressure <40 mmHg favors Primary cause
 - RV systolic pressure >55 mmHg favors Secondary cause

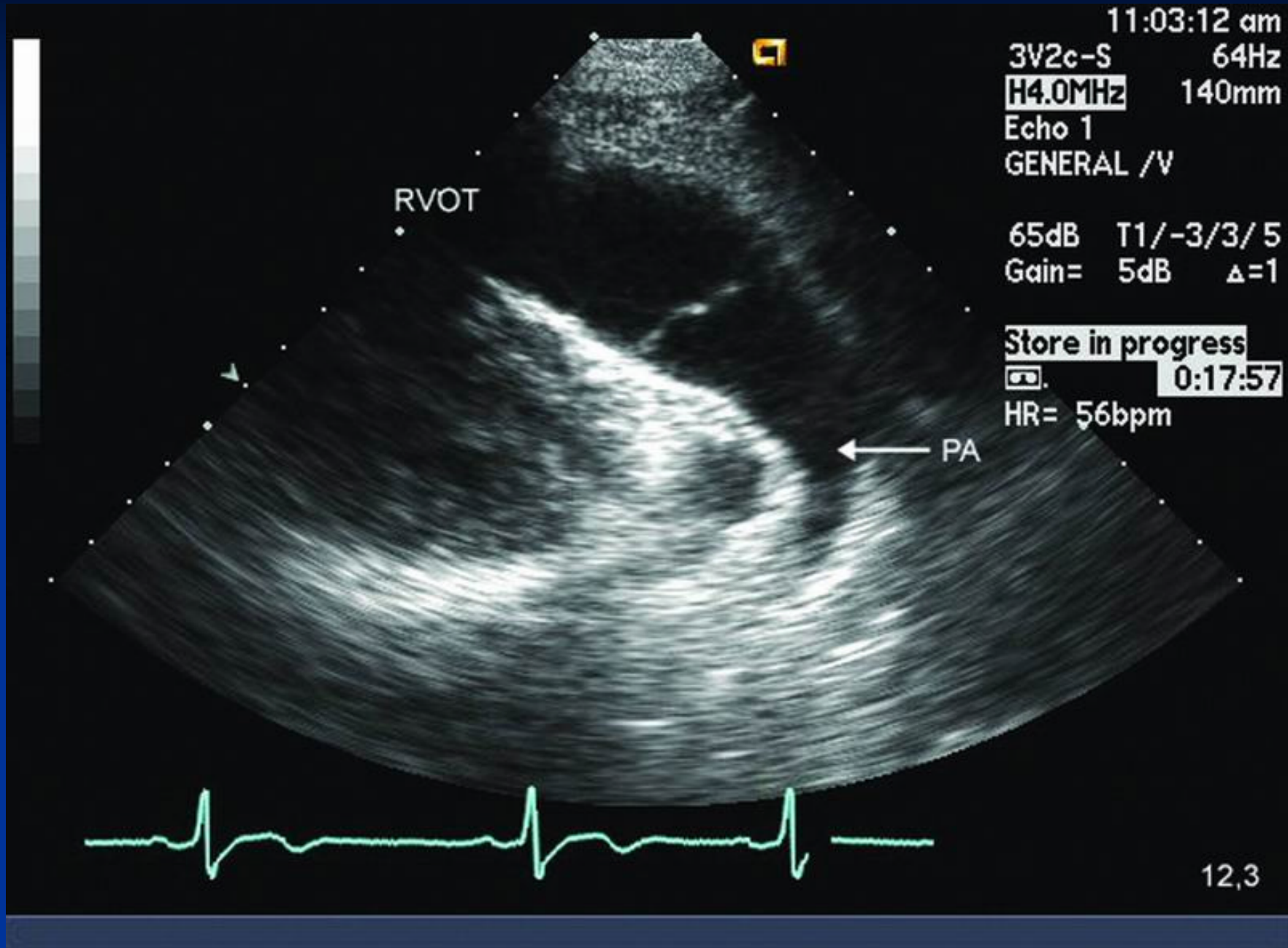
Surgical Management

- Well tolerated without pulmonary HTN
 - Valvectomy tolerated well, with right heart dilatation developing months to years after surgery
- Primary cause
 - Usually require bioprosthesis
- Secondary cause
 - Usually annuloplasty ring (in conjunction with mitral valve surgery)
 - May not require TV surgery if mild TR and normal Triscupid annulus size
- Endocarditis in IV drug use
 - Controversial
 - Valvectomy followed later by bioprosthesis?

Pulmonary Valve (TTE Short Axis)



Pulmonary Valve (TTE Long Axis)



Pulmonic Valve Stenosis

- Etiology
 - Congenital pulmonic valve stenosis
 - Variable atresia, dysplastic valve
 - Associated with Noonan Syndrome
 - May be supra-auricular
 - Rheumatic disease (uncommon)
 - Carcinoid
 - Cardiac Tumors

Physical Exam

- Decreased intensity of P2
- Possible thrill in 2nd left intercostal space

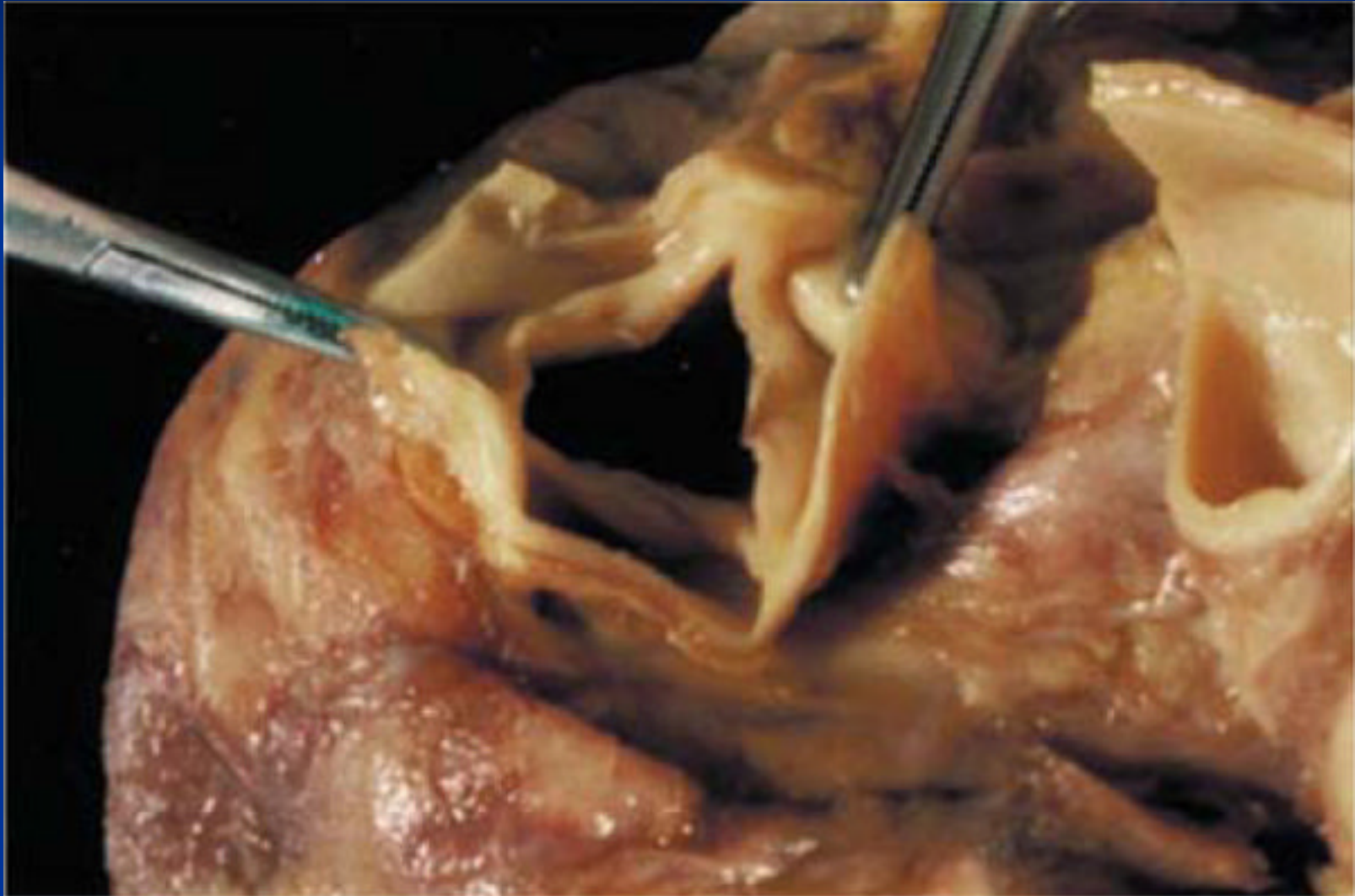
Treatment of Pulmonic Valve Stenosis

- Percutaneous Balloon Valvotomy
 - Usually when mean pressure gradient across valve is 50mmHg
 - May require additional valvotomies in future

Pulmonic Valve Regurgitation

- Etiology
 - Dilatation of pulmonic valve ring (Most Common)
 - Pulmonary hypertension (Any cause)
 - Pulmonary artery dilatation
 - Connective tissue disease
 - Infective endocarditis
- Less Common Causes
 - Surgical treatment for tetralogy of Fallot
 - Other Congenital causes
 - Carcinoid syndrome
 - Syphilis
 - Chest trauma
 - Pulmonary artery catheterization

Carcinoid of Pulmonic Valve



Symptoms

- Similar to tricuspid regurgitation
- Tolerated well as long as pulmonary artery pressure is normal

Physical Examination

- Low pitch diastolic murmur at left 3rd or 4th intercostal space
- S3 or S4 at left lower sternal border, augmented by inspiration
- Graham-Steell Murmur
 - Pulmonary pressure >55mmHg
 - High pitched, blowing decrescendo murmur
 - Left parasternal area, starts after prominent P2
 - Increases with inspiration
 - Confused with aortic regurgitation murmur
 - However AR is more common

Diagnostic Evaluation

- Echocardiography
 - Severity of regurgitation
 - Pulmonic valve annulus size
 - Pulmonary arterial pressure
- EKG
 - Without pulmonary hypertension
 - Incomplete RBBB
 - With pulmonary hypertension
 - RV hypertrophy
- XRay and Angiography – nonspecific
- Cardiac MRI – Assess PA dilatation, PR severity

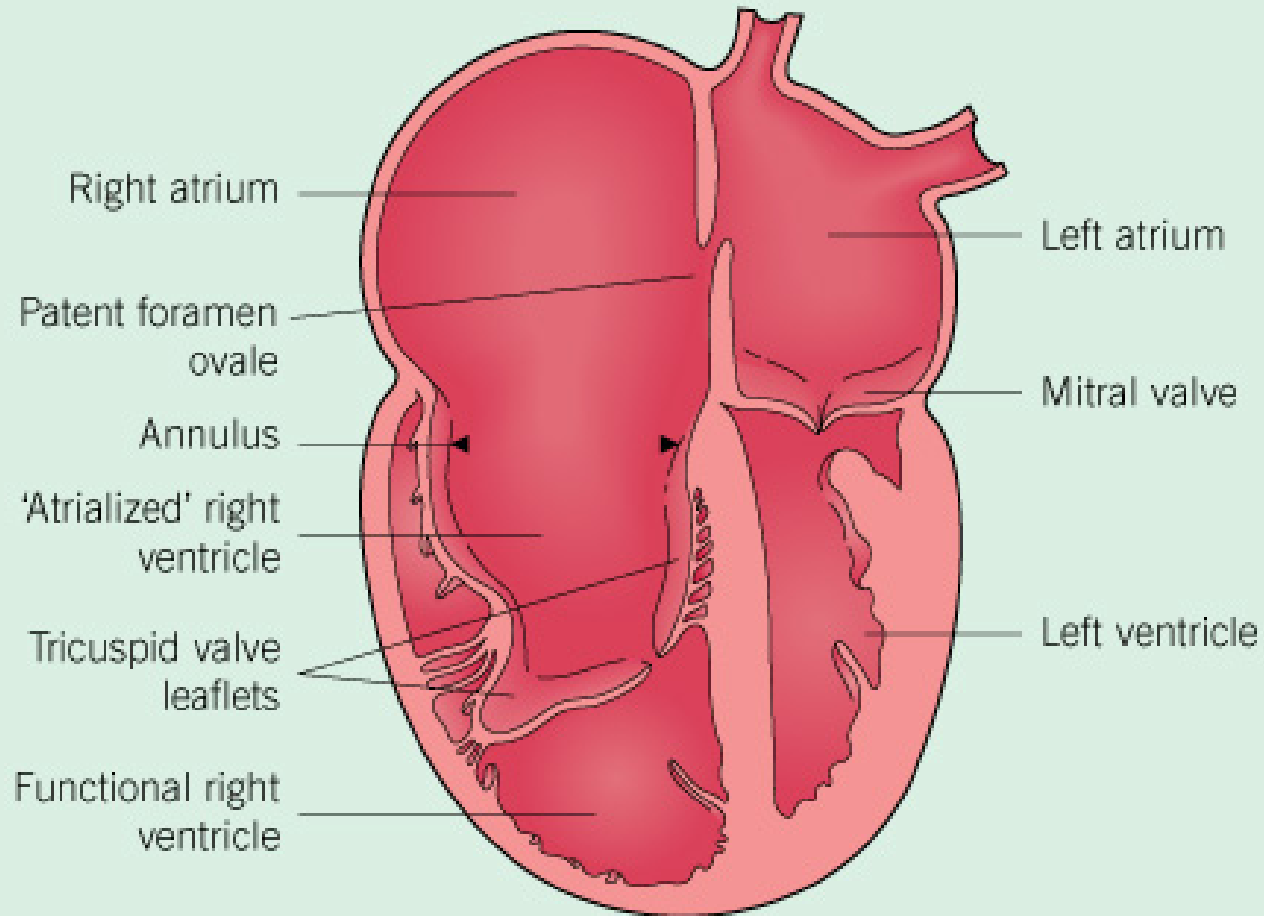
Management

- Usually not severe enough alone to require specific treatment
- Treatment of pulmonary hypertension usually sufficient
 - Mitral valve surgery
- Endocarditis may require valve replacement
- Bioprosthesis or allograft preferred for replacement

Specific Diseases

- Ebstein's Anomaly
- Carcinoid Syndrome

EBSTEIN'S ANOMALY OF THE TRICUSPID VALVE

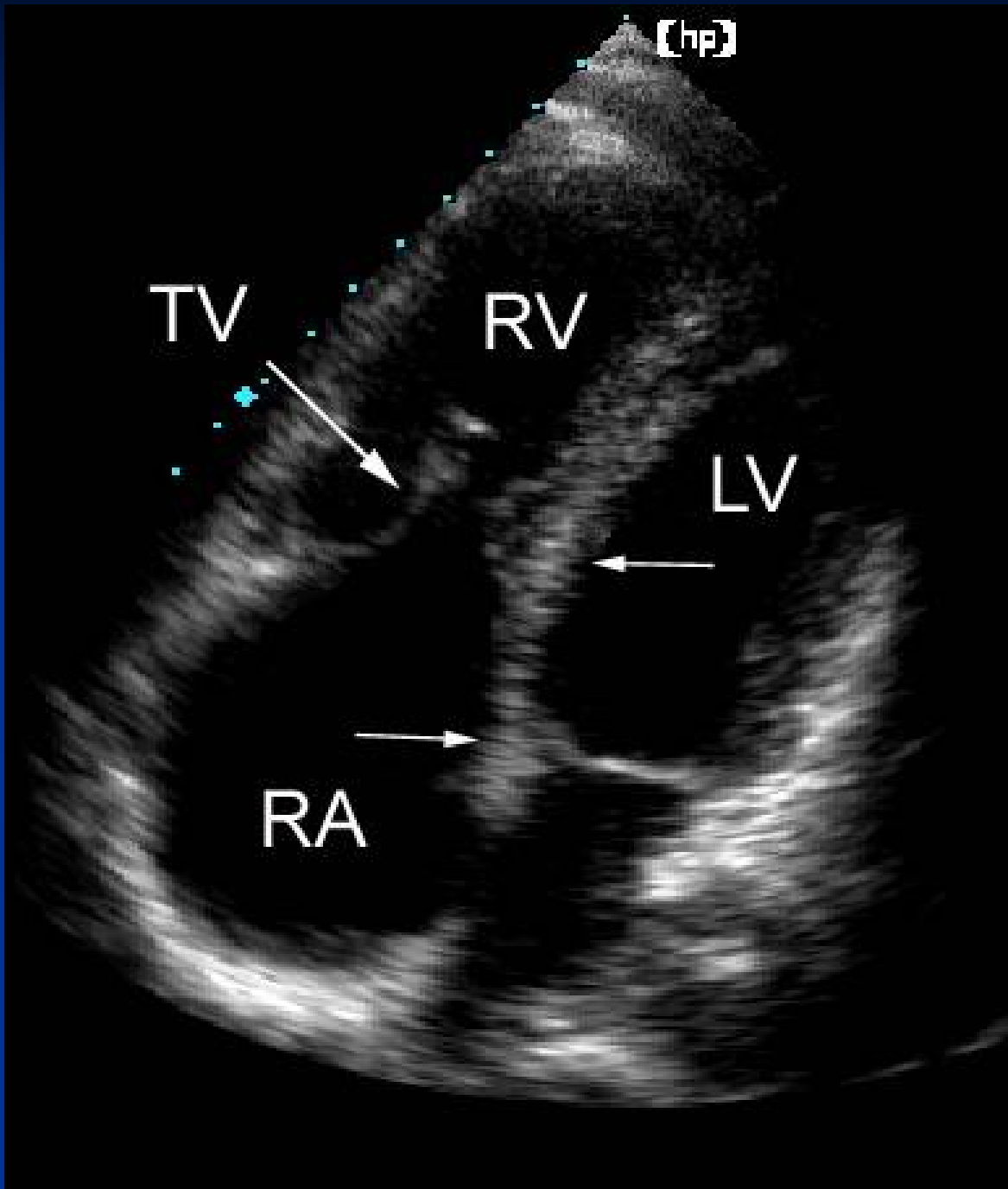


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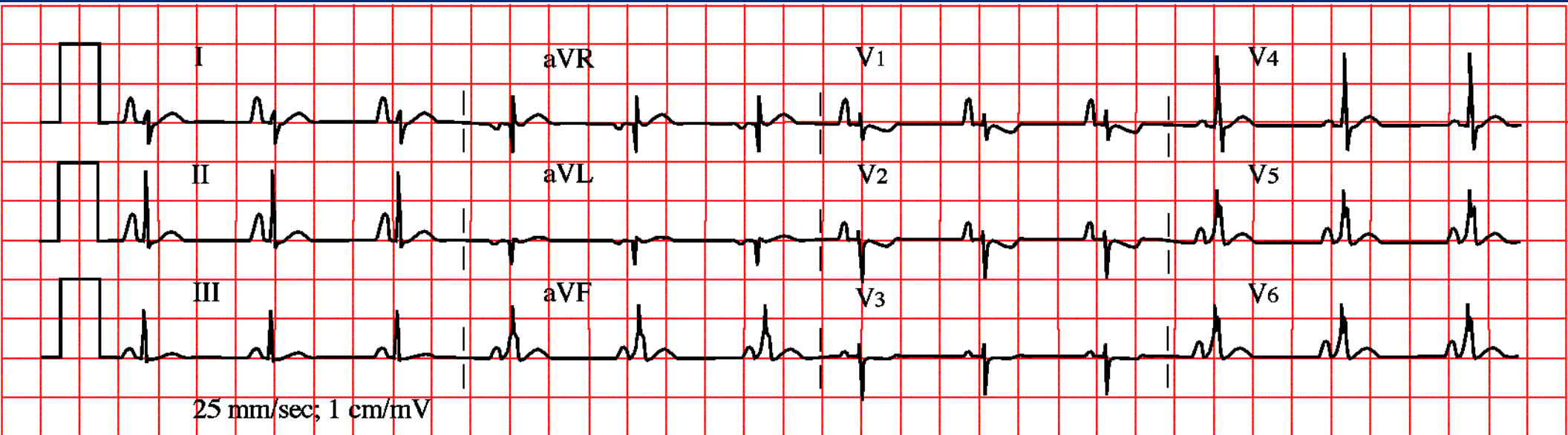
Ebstein's Anomaly

- 1-2 in 10,000 approximate incidence in live births
- Apical displacement of tricuspid valve leaflets
 - Anterior leaflet never displaced
 - Variable leaflet deformity
- “Atrialization” of right ventricular tissue
- Small right ventricle, large right atrium
- Associated lesions
 - 50% have atrial septal defect or patent foramen ovale
 - 25% have accessory bypass tract

Ebstein's Anomaly (TTE Apical 4 chamber)



EKG



Invasive catheterization of RV

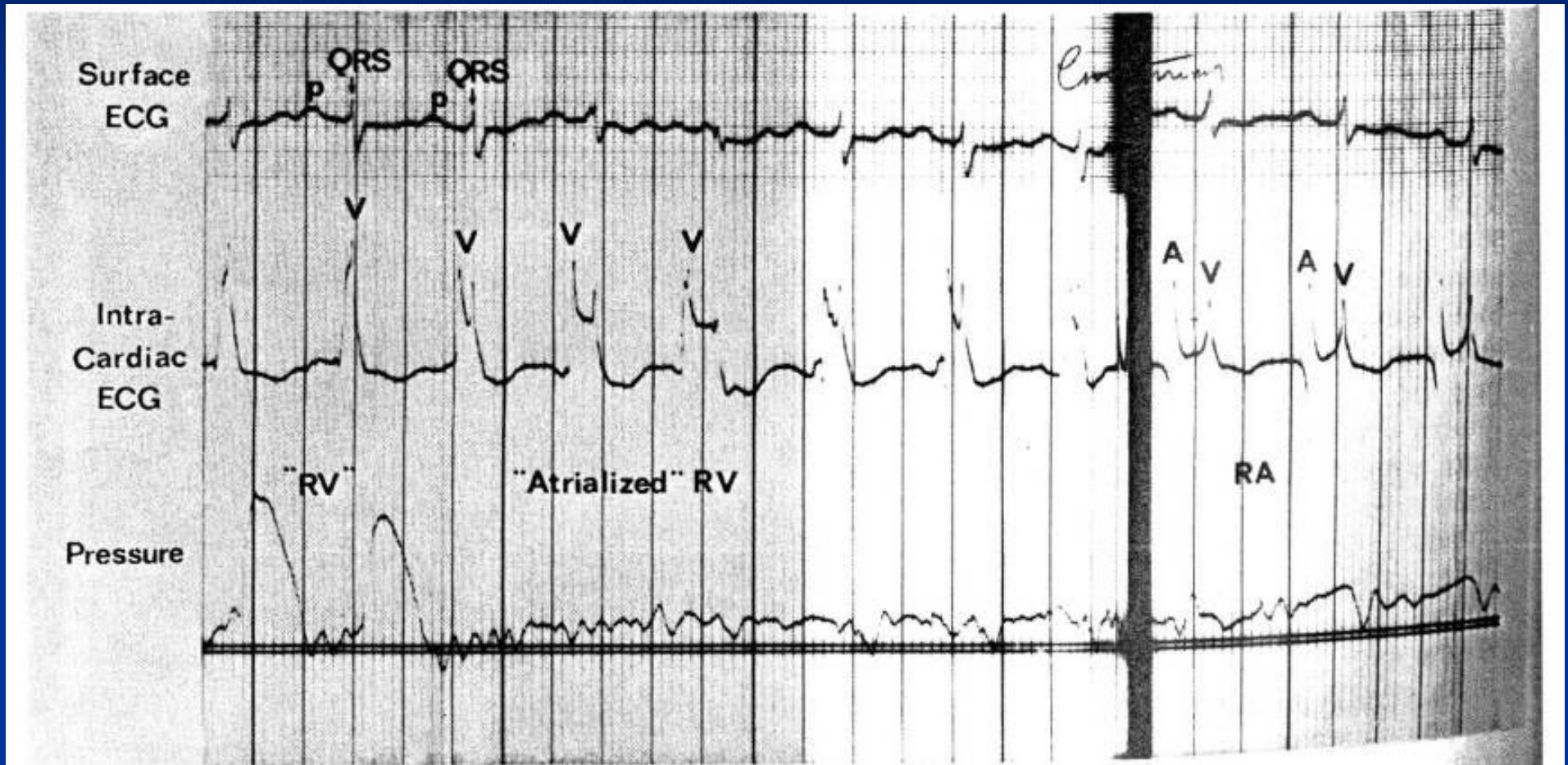
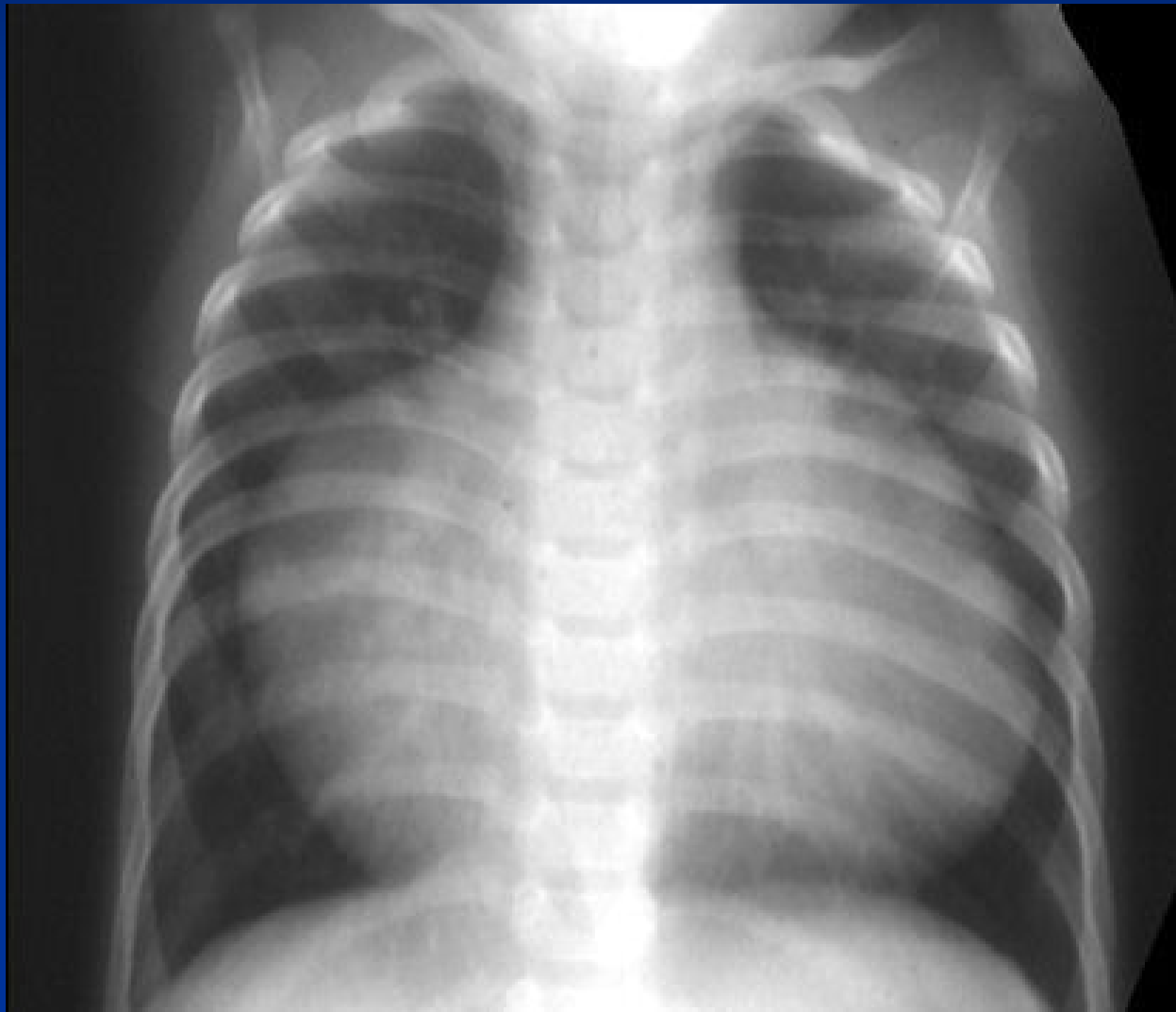


Fig. 28-16. Surface ECG, intracardiac ECG, and pressure recording in a 15-year-old girl with Ebstein's disease. As catheter is withdrawn from right ventricle (RV) (left) to right atrium (right), one sees an RV electrogram with RV pressure in the body of the RV, and RV electrogram with atrial pressures in the "atrialized" portion of the RV, and an atrial electrogram with atrial pressures in the true right atrium. V and A refer to ventricular and atrial depolarization, respectively.

Chest Xray (Infant)



Natural History

- Variable!
- If extreme, in utero death
- If mild, may live into 9th decade
- Severe valve deformity – symptoms develop in infancy
- Moderate valve deformity – symptoms in adolescence

Indications for intervention

- Significant cyanosis
- Right-sided heart failure
- Declining functional capacity (\geq Class III NYHA)
- Relative indications
 - Paradoxical emboli
 - Recurrent supraventricular arrhythmias
 - Asymptomatic substantial cardiomegaly

Surgical Techniques

- Tricuspid Valve Repair
 - Anterior leaflet cannot be tethered down
 - Repair results in “monocuspid” valve
- Valve replacement – bioprosthetic
- Concomitant
 - Accessory path ablation
 - MAZE procedure if atrial fibrillation present
 - Closure of PFO/ASD
- Unusual procedures
 - Bidirectional cavopulmonary shunt
 - Fontan
- 10-20% will develop supraventricular arrhythmias after Ebstein repair

Carcinoid Syndrome

- Caused by carcinoid producing tumor
- Symptoms of diarrhea and flushing, usually occurring together
- Asthma less common
- 10-40% incidence of cardiac involvement
- About 10% of carcinoid tumors cause the carcinoid syndrome
 - Mostly midgut tumors cause syndrome
- Caused by overproduction of 5HT
 - (5-Hydroxytryptophan)

Carcinoid Tumor Location, Frequency of Metastases, and Association with the Carcinoid Syndrome

| | Location (% of Total) | Incidence of Metastases | Incidence of Carcinoid Syndrome |
|-------------------------|--------------------------|----------------------------|------------------------------------|
| Foregut | | | |
| Esophagus | <0.1 | — | — |
| Stomach | 4.6 | 10 | 9.5 |
| Duodenum | 2.0 | — | 3.4 |
| Pancreas | 0.7 | 71.9 | 20 |
| Gallbladder | 0.3 | 17.8 | 5 |
| Bronchus, lung, trachea | 27.9 | 5.7 | 13 |
| Midgut | | | |
| Jejunum | 1.8 | 58.4 | 9 |
| Ileum | 14.9 | — | 9 |
| Meckel's diverticulum | 0.5 | — | 13 |
| Appendix | 4.8 | 38.8 | <1 |
| Colon | 8.6 | 51 | 5 |
| Liver | 0.4 | 32.2 | — |
| Ovary | 1.0 | 32 | 50 |
| Testis | <0.1 | — | 50 |
| Hindgut | | | |
| Rectum | 13.6 | 3.9 | — |

CHARACTERISTICS OF VARIOUS TYPES OF CARCINOID TUMOR

| Location | Tumor products | Symptoms |
|---|--|---|
| Foregut: (Bronchus, stomach, gynecomastia, proximal duodenum) | 5-HTP, histamine, HCG, ACTH, GH, gastrin, tachykinins, bradykinin Low serotonin | Bronchospasm, generalized flushing, Cushing's syndrome, acromegaly, dilutional hyponatremia, heart disease |
| Midgut: (Distal duodenum, jejunum/ileum, appendix, right colon) | Serotonin, tachykinins | Diarrhea, heart disease, hypotension, flushing, vasodilation, diarrhea, telangiectasias, wheezing |
| Hindgut: (Transverse/left colon, rectum) | Tachykinins | Diarrhea, facial flushing |

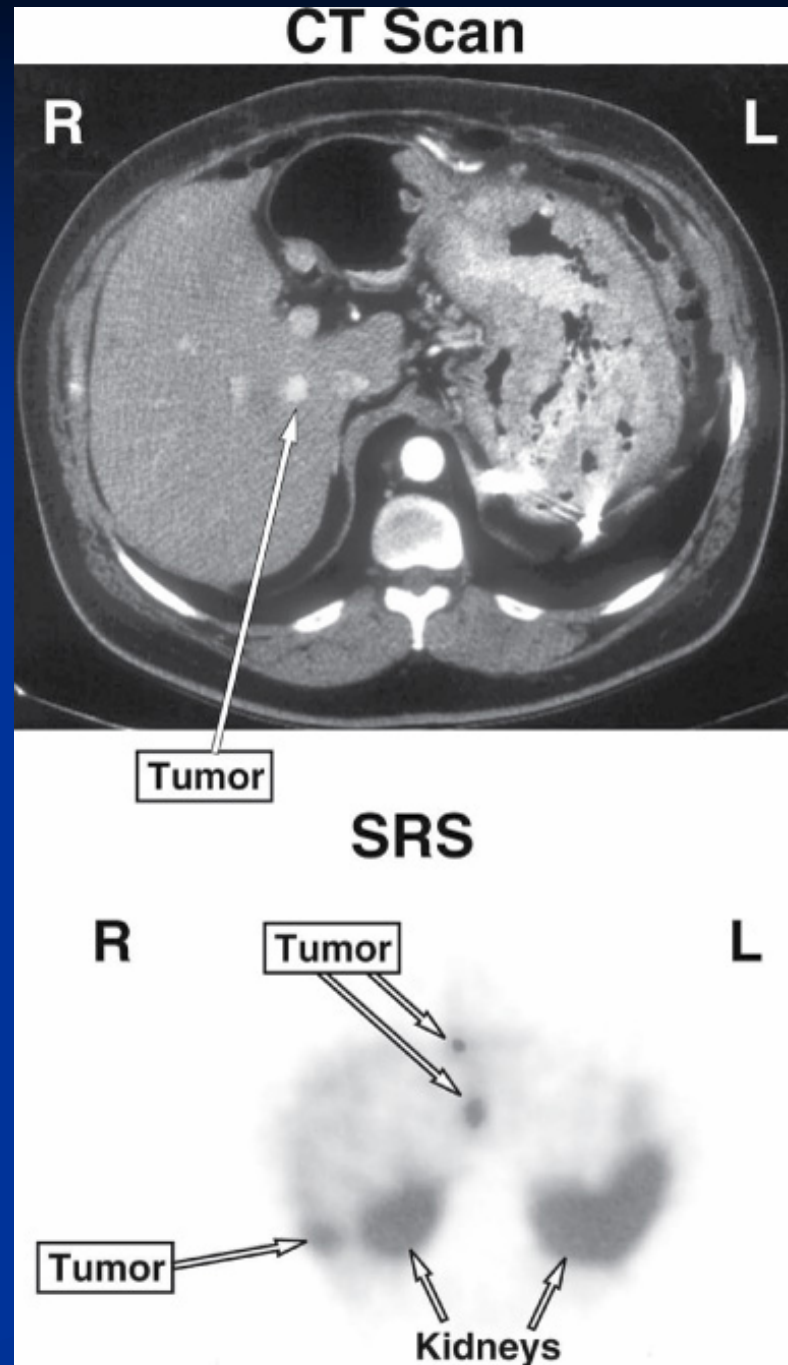
Diagnosis of Carcinoid

- Urinary 5HIAA (Hydroxyindolacetic acid)
 - Normal excretion is 2-8mg/day
 - 78% sensitivity, 100% specificity
- Platelet serotonin – more sensitive
- Plasma serotonin
- Serum chromogranin A

Anatomic Localization

- CT scan combined with nuclear imaging
- Carcinoid tumors often express high-affinity somatostatin receptors
- Indium-111-pentetreotide is a radiolabeled somatostatin analogue
- False positive uptake can occur with granulomas, wounds, thyroid disease

CT Localization combined with Somatostatin Receptor Scintigraphy



Treatment

- Carcinoid Syndrome:
 - Octreotide
 - Somatostatin analogue
 - 100mcg SQ q8h; up to 3 mg/day
 - Controls symptoms of diarrhea and flushing
 - Interferon
 - Hepatic artery embolization
- Surgical resection

Carcinoid effects on the Heart

- Seen with Carcinoid Syndrome
- Tricuspid and Pulmonic Valve
- Stenosis and Regurgitation

Summary

- Tricuspid Stenosis
 - Rheumatic
- Tricuspid Regurgitation
 - Secondary Causes
- Pulmonic Valve Stenosis
 - Congenital
- Pulmonic Valve Regurgitation
 - Secondary Causes

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