

Longevity In Unoperated Congenital Heart Disease

**The Ahmanson/UCLA
Adult Congenital Heart
Disease Center**

*Unoperated and Untreated
The First Account*

ATLAS
OF
CONGENITAL CARDIAC
DISEASE

MAUDE E. ABBOTT

Natural and Unnatural

Natural history is not
synonymous with *unoperated*.
Surgeons are not perpetrators of
the unnatural.

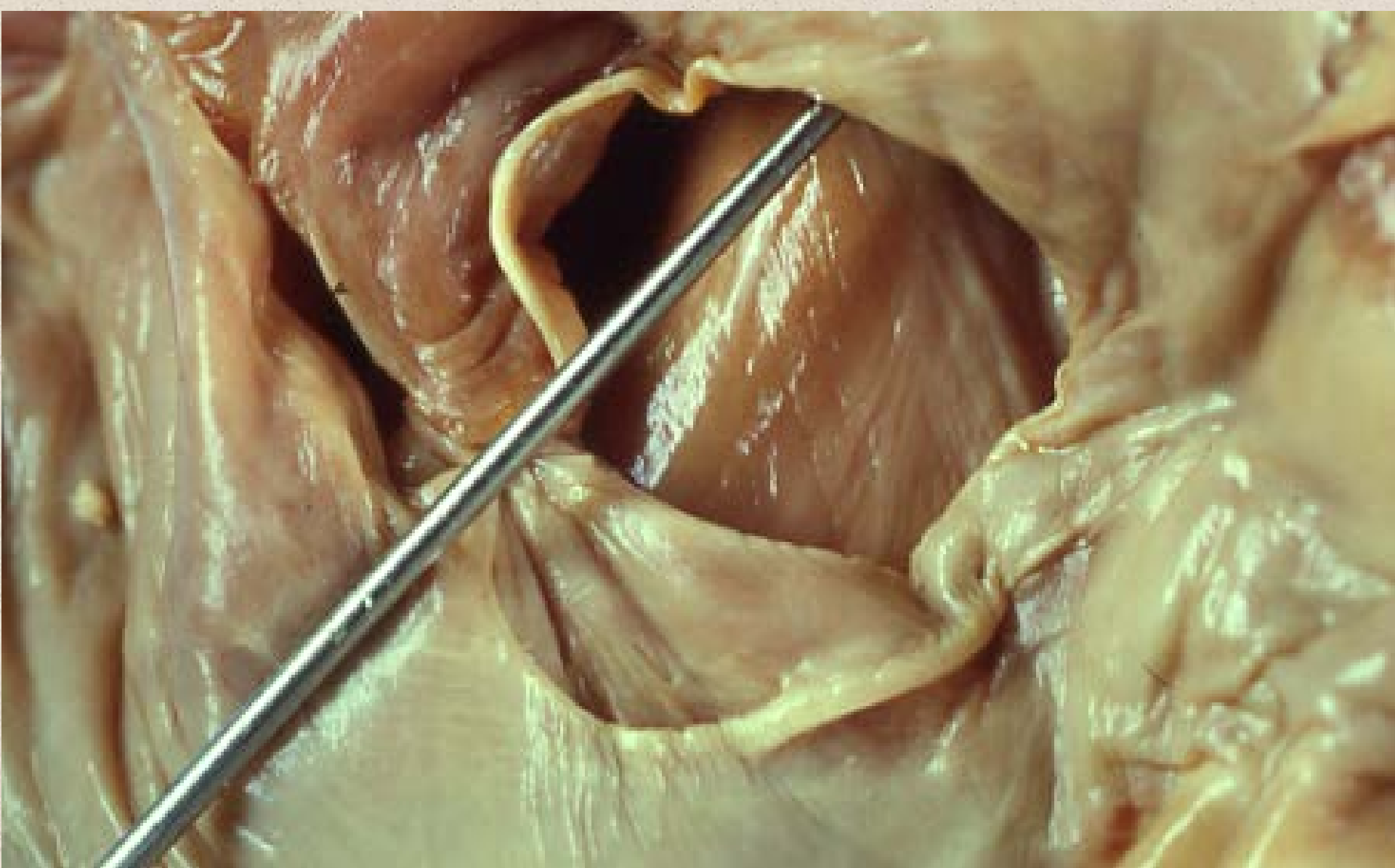
Gaussian Distribution
is a statistical article of faith.
However, it is the
rarity at the far end of the
distribution curve that
provides insight
into unoperated survival.

Longevity Without Cardiac Surgery

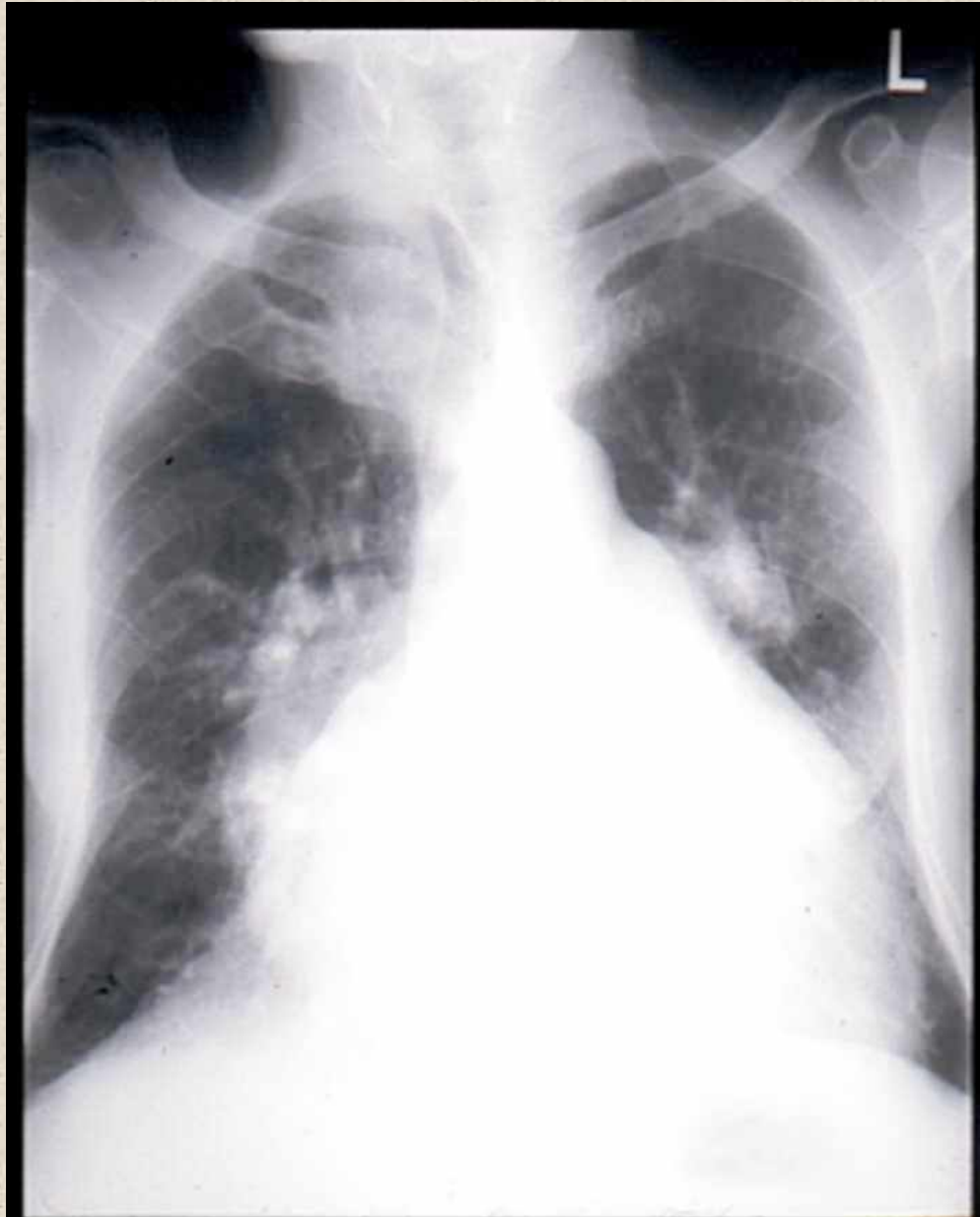
- 1. Common defects with expected adult survival but exceptional longevity.*
- 2. Common defects with unexpected adult survival and exceptional longevity.*
- 3. Uncommon defects with expected adult survival and exceptional longevity.*
- 4. Uncommon defects with unexpected adult survival and exceptional longevity. The rarest of the rare.*

*Common Defects With Expected
Adult Survival but Exceptional
Longevity*

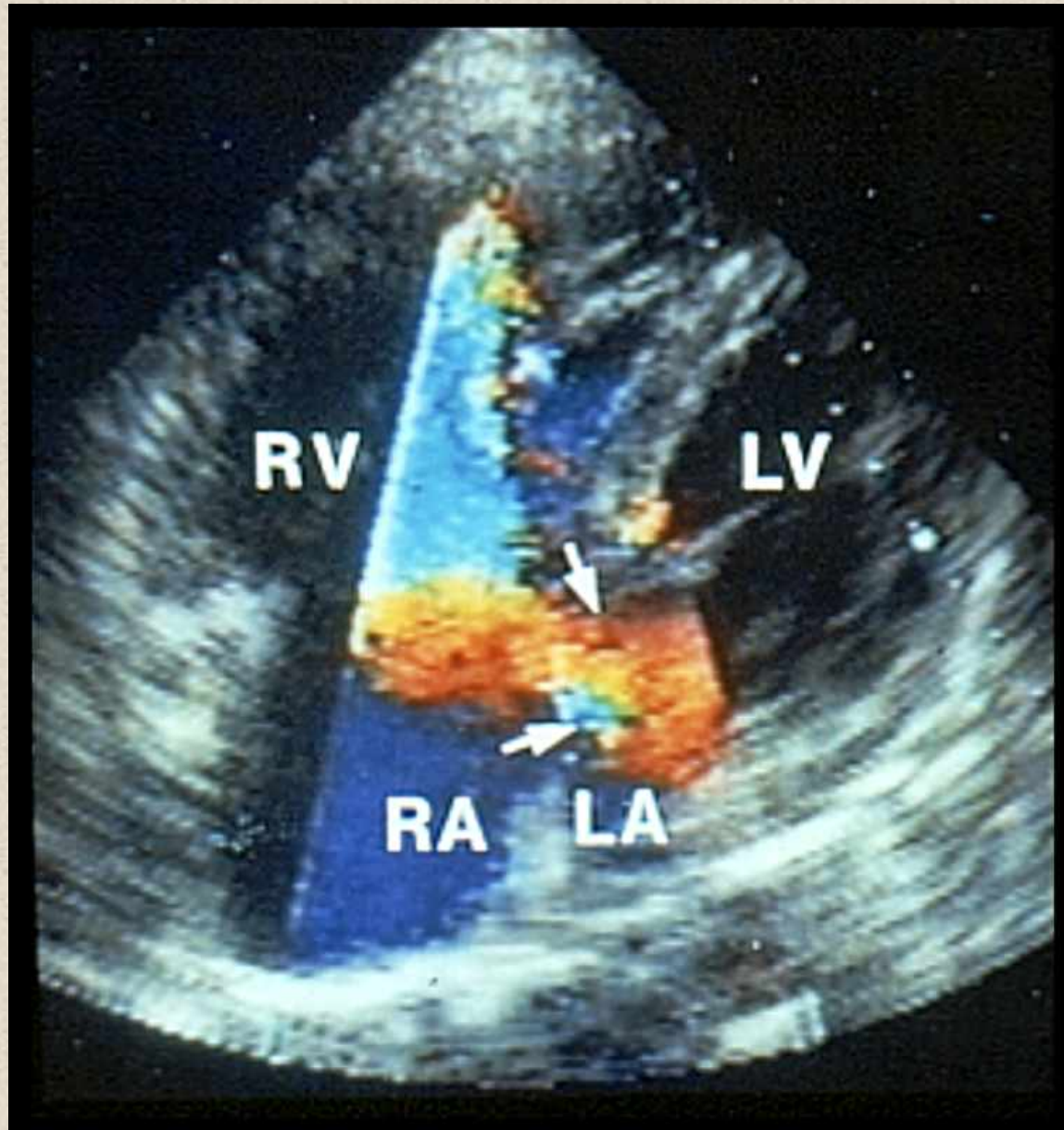
Ostium Secundum ASD Age 85



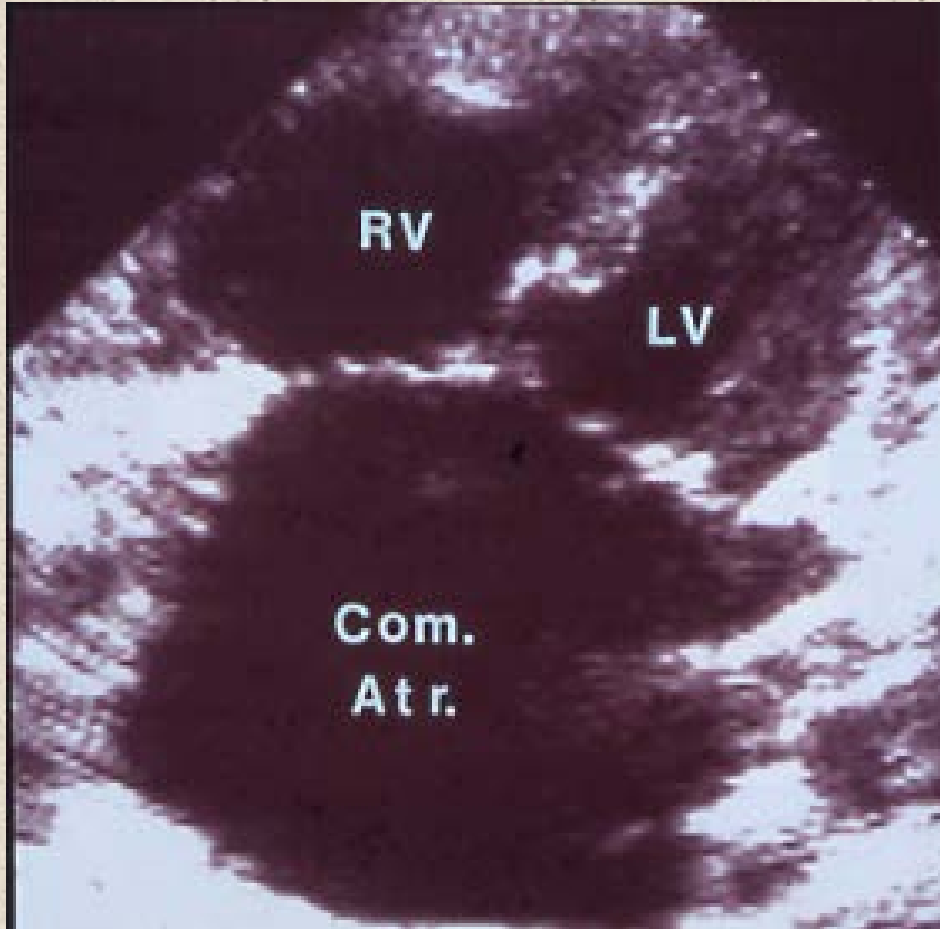
Ostium Secundum ASD Age 95



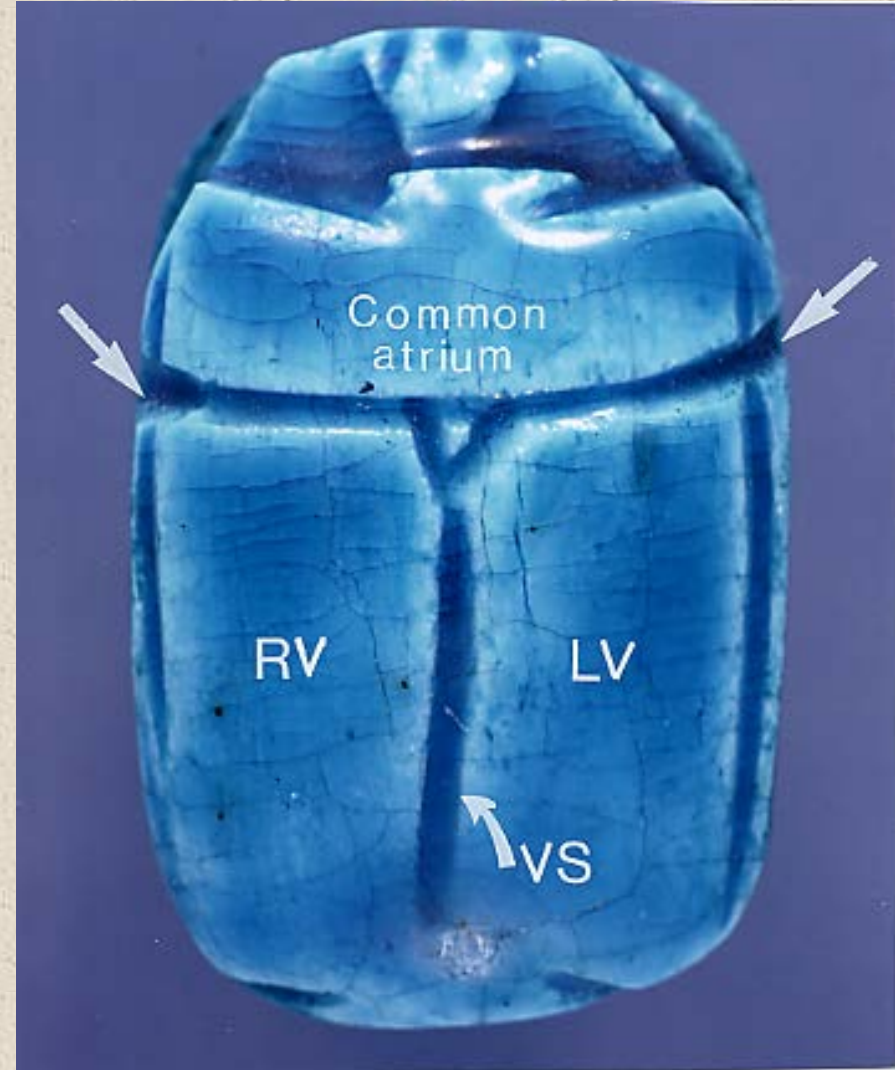
Ostium Primum ASD Age 76



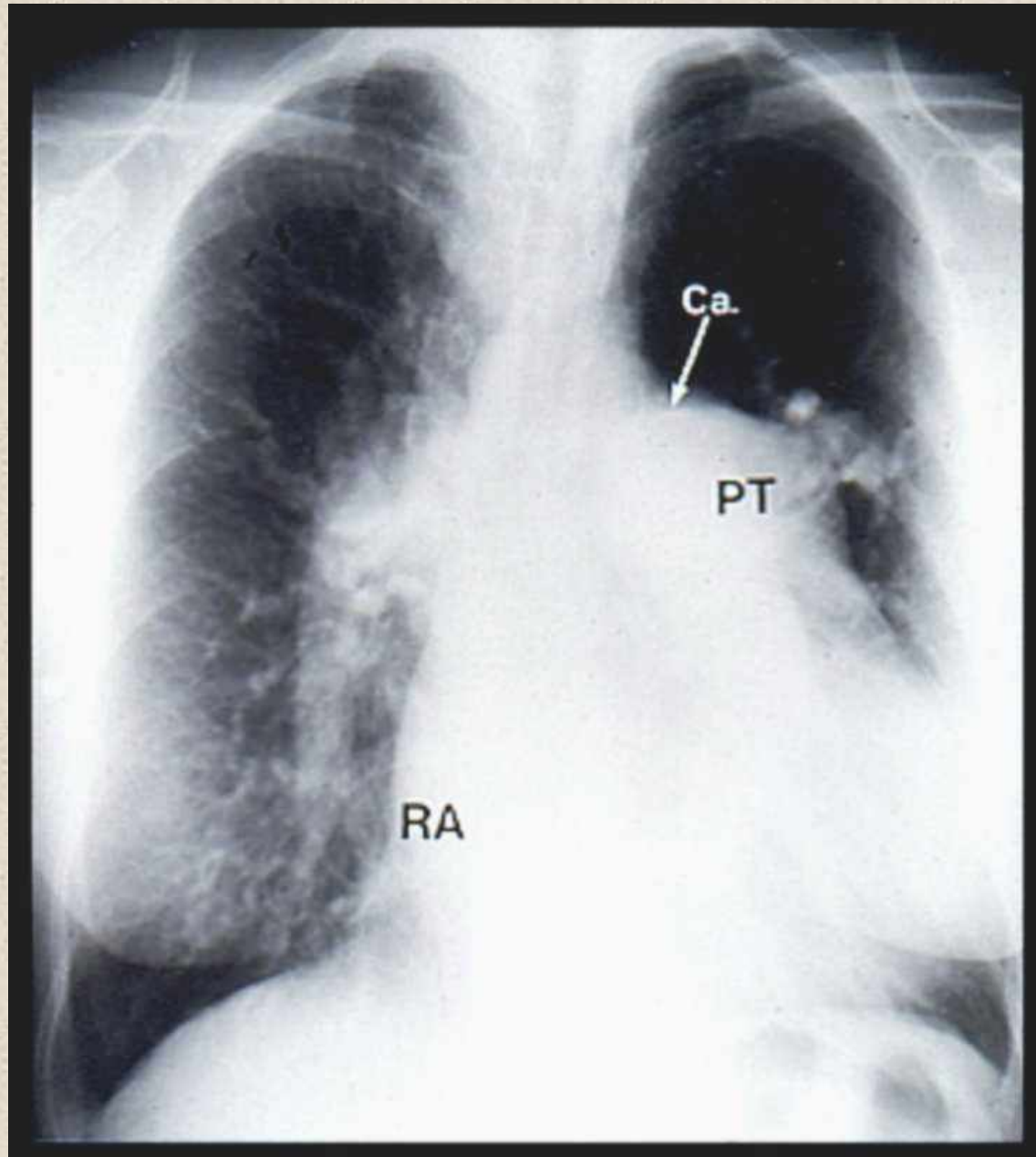
Common Atrium
Age 57



Scarab
Egyptian circa 1450 BC

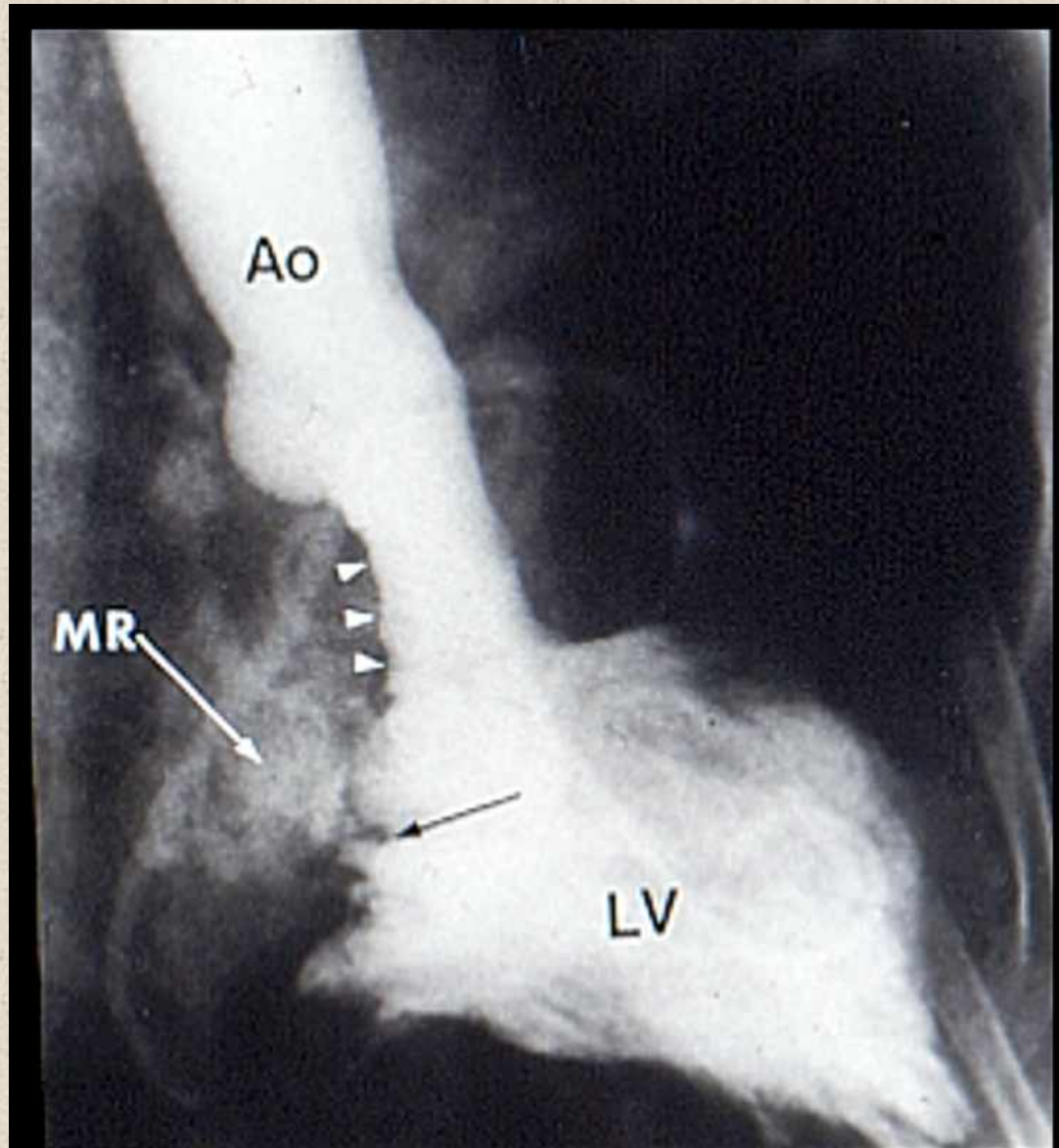


Common Atrium Age 57



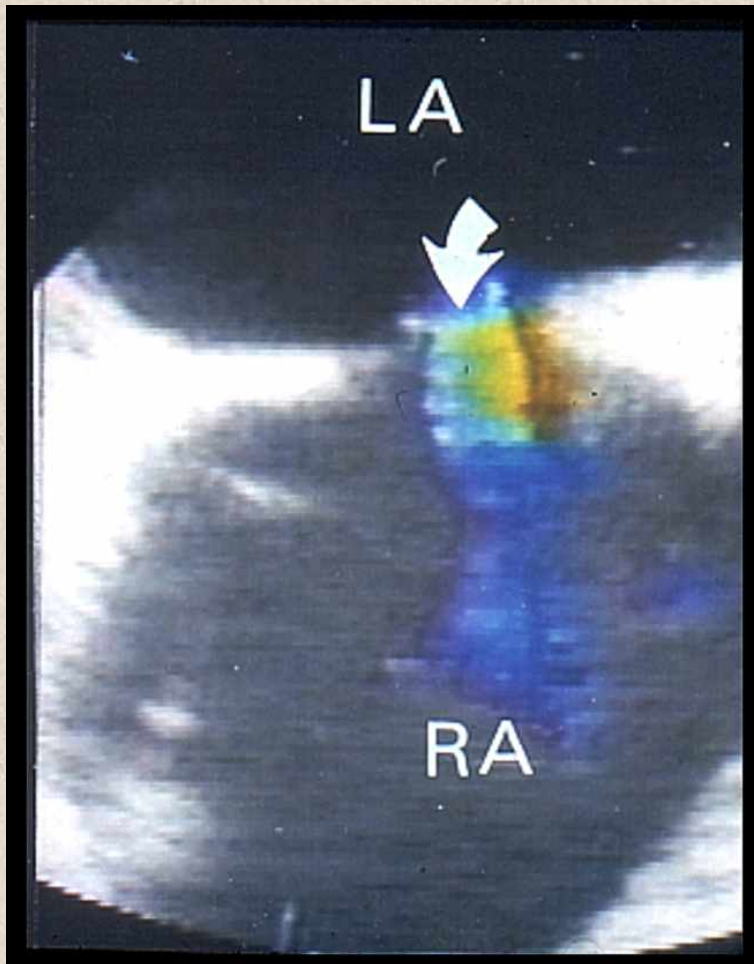
Angiogram

Common Atrium

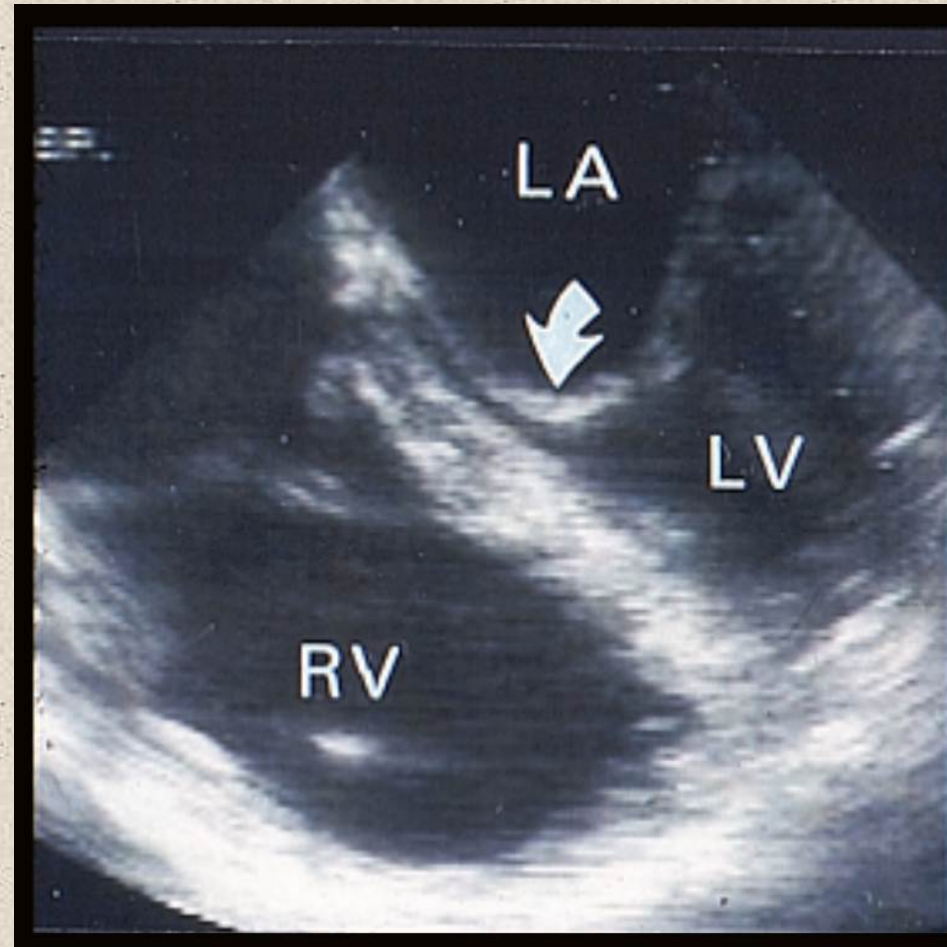


Lutembacher Syndrome

Secundum ASD



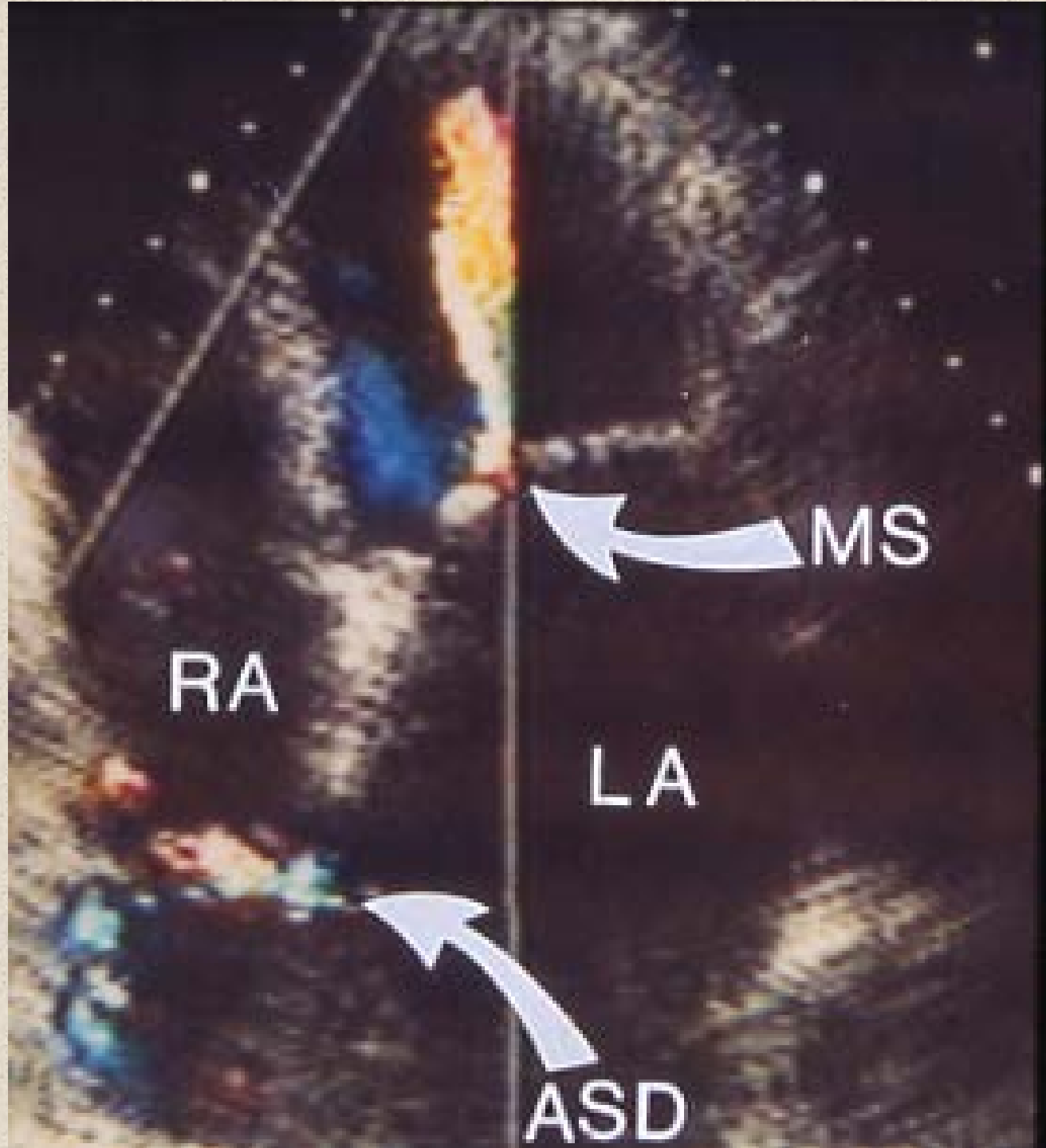
Rheumatic Mitral Stenosis



Lutembacher is a German name,
but Dr. Lutembacher used the
French pronunciation (Loo-tem-bah-
share) because he was Alsatian when
Alsace-Lorraine was part of France.
The pronunciation is now anglicized
to *Loo-tem-bah-ker*.

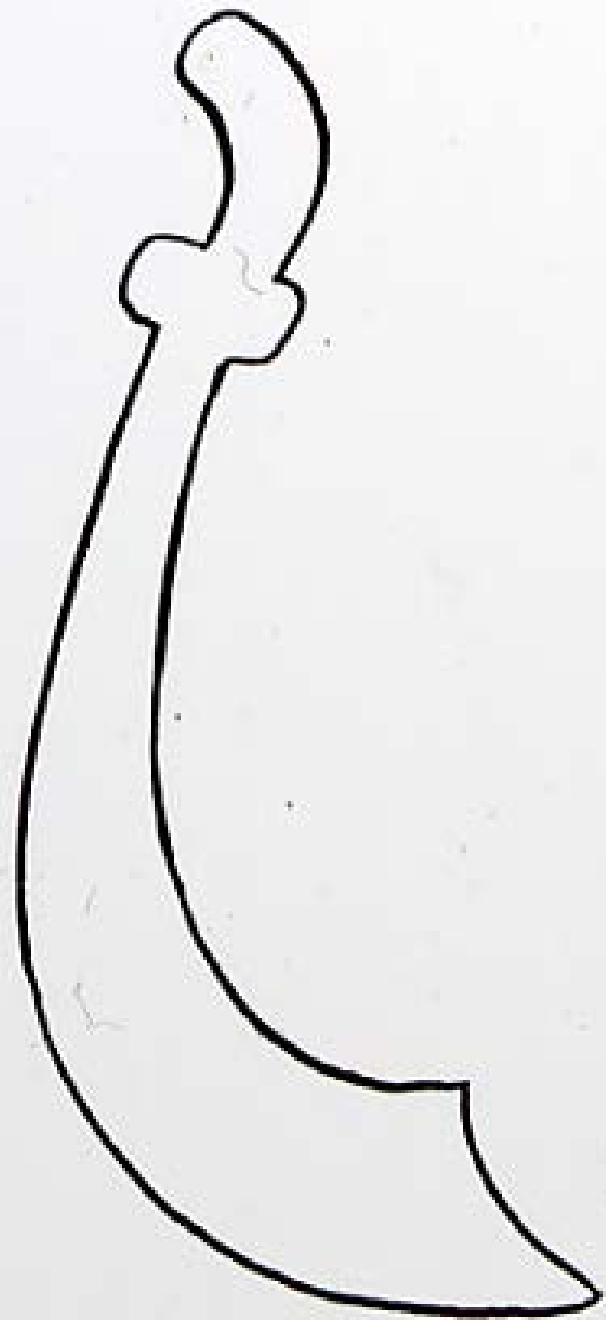
Lutembacher's original patient was a *61 year old* woman who had been pregnant seven times. Firkett's patient was a *74 year old* woman who had endured 11 pregnancies. In one instance, an *81 year old* woman with Lutembacher syndrome experienced no cardiac symptoms until her 75th year.

*Iatrogenic
Lutembacher
Syndrome*



Scimitar

***Middle Eastern or South
Asian sword with a
curved blade***



Scimitar Syndrome Age 63

Anomalous connection of the
pulmonary veins in one lung
to the inferior vena cava.



Pulmonary Valve Stenosis



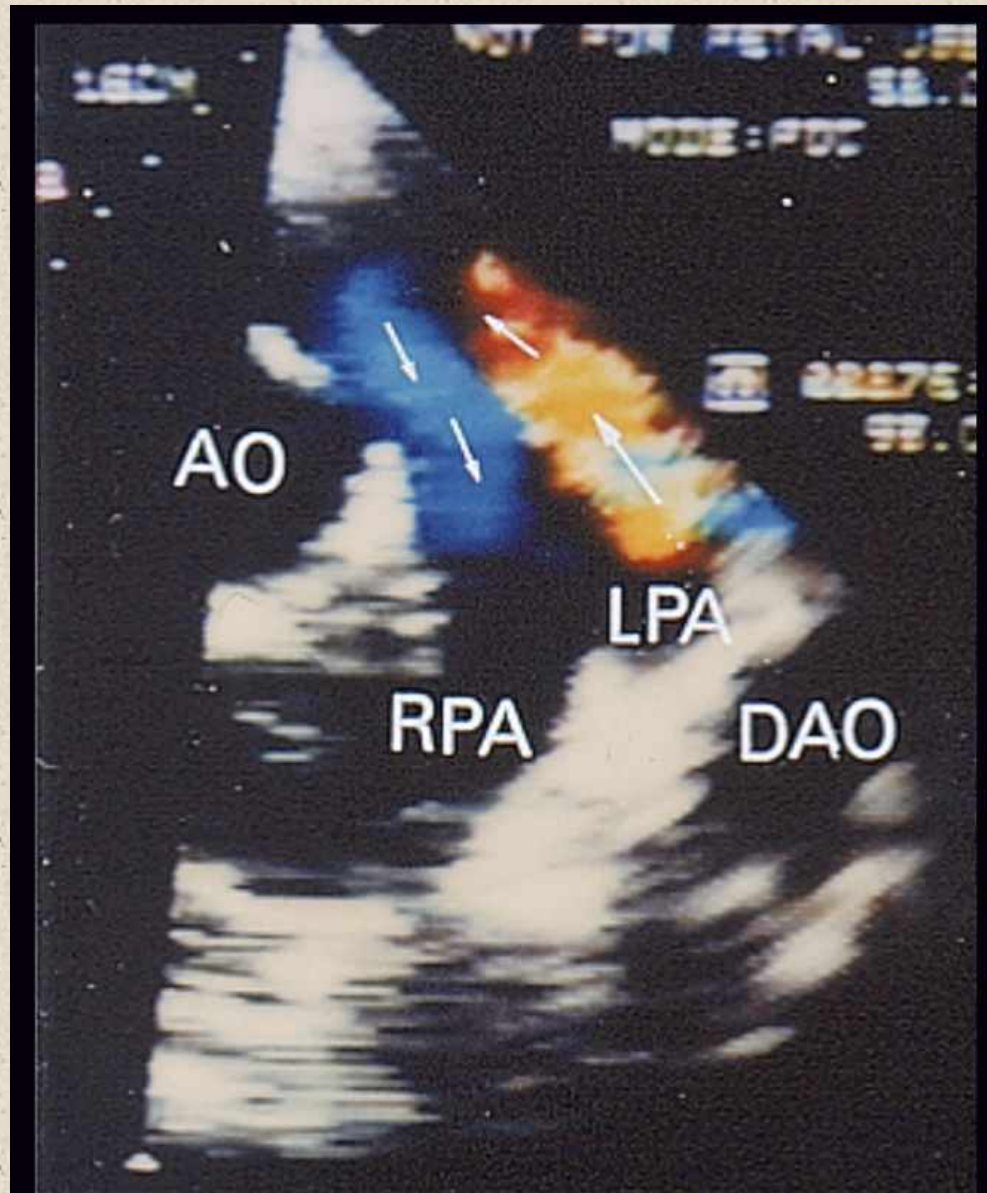
Pulmonary Valve Stenosis

An appreciable number of patients with moderate to severe congenital pulmonary stenosis claim to be virtually symptom free. A group of patients with right ventricular systolic pressures of 75 to 100mm Hg included a New Zealand long-distance swimmer, a female athlete, an English hockey captain and a long-distance runner.

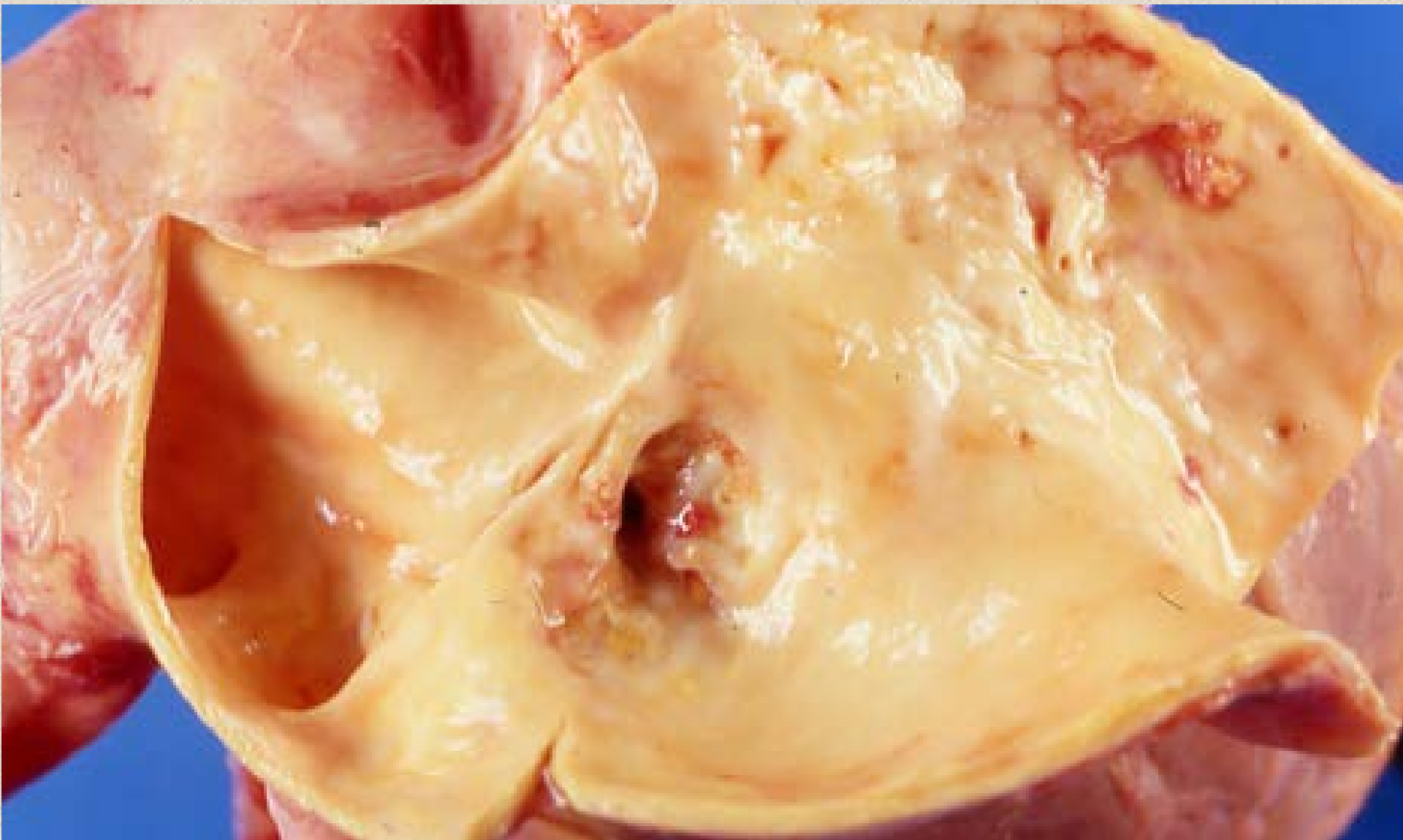
Paul Wood, OBE, MD, FRCP

The UCLA Registry includes a 17 year old boy with *pulmonary valve stenosis* who played baseball despite a right ventricular systolic pressure of 200 mm Hg, and a 32 year old man who had run the quarter mile in high school despite a resting right ventricular systolic pressure of 75 mm Hg.

Patent Ductus Arteriosus



Calcified Ductus Age 74



Calcified Patent Ductus Age 84



Coarctation Age 76

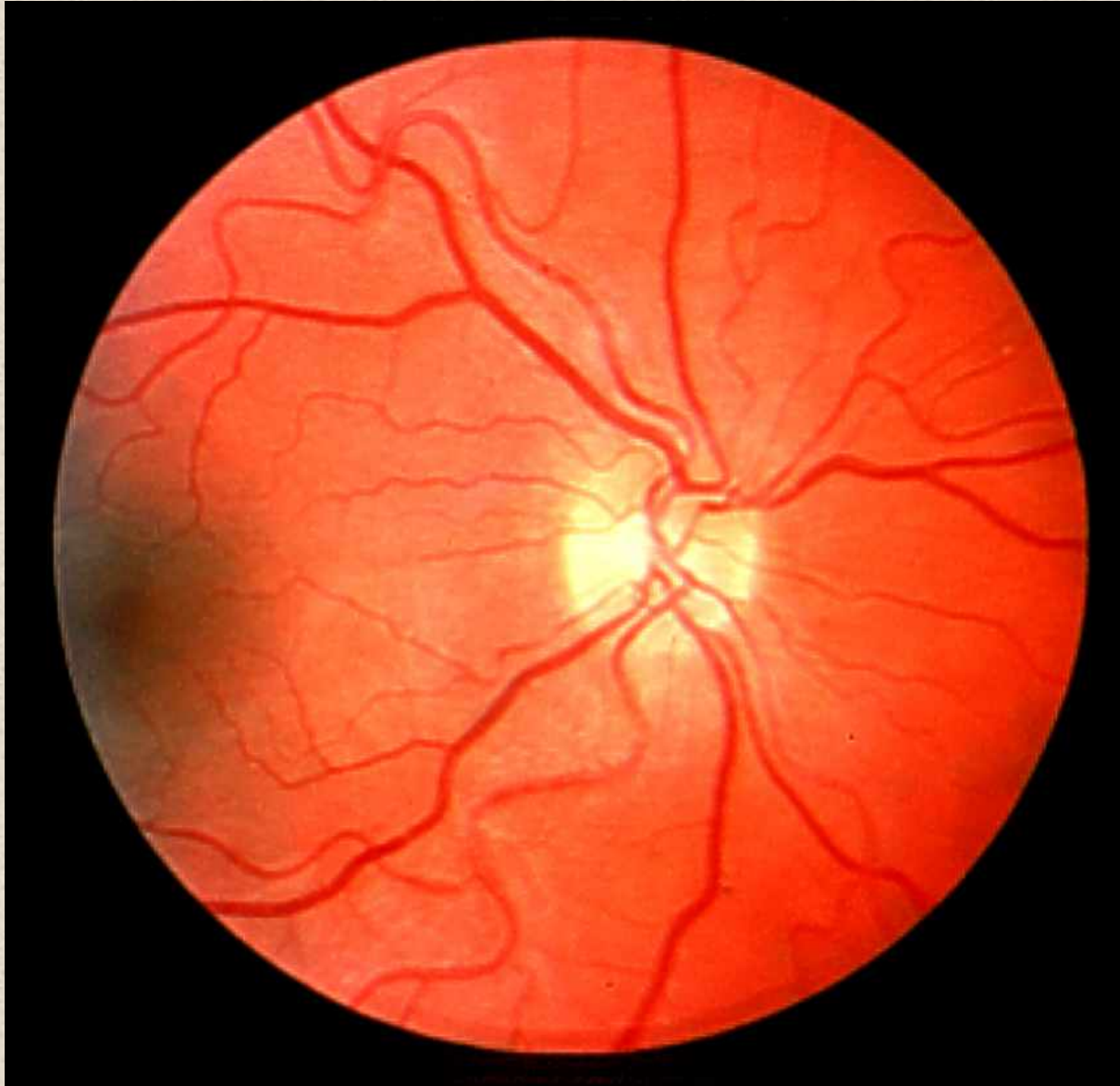


Complete Aortic Obstruction



Coarctation of the Aorta

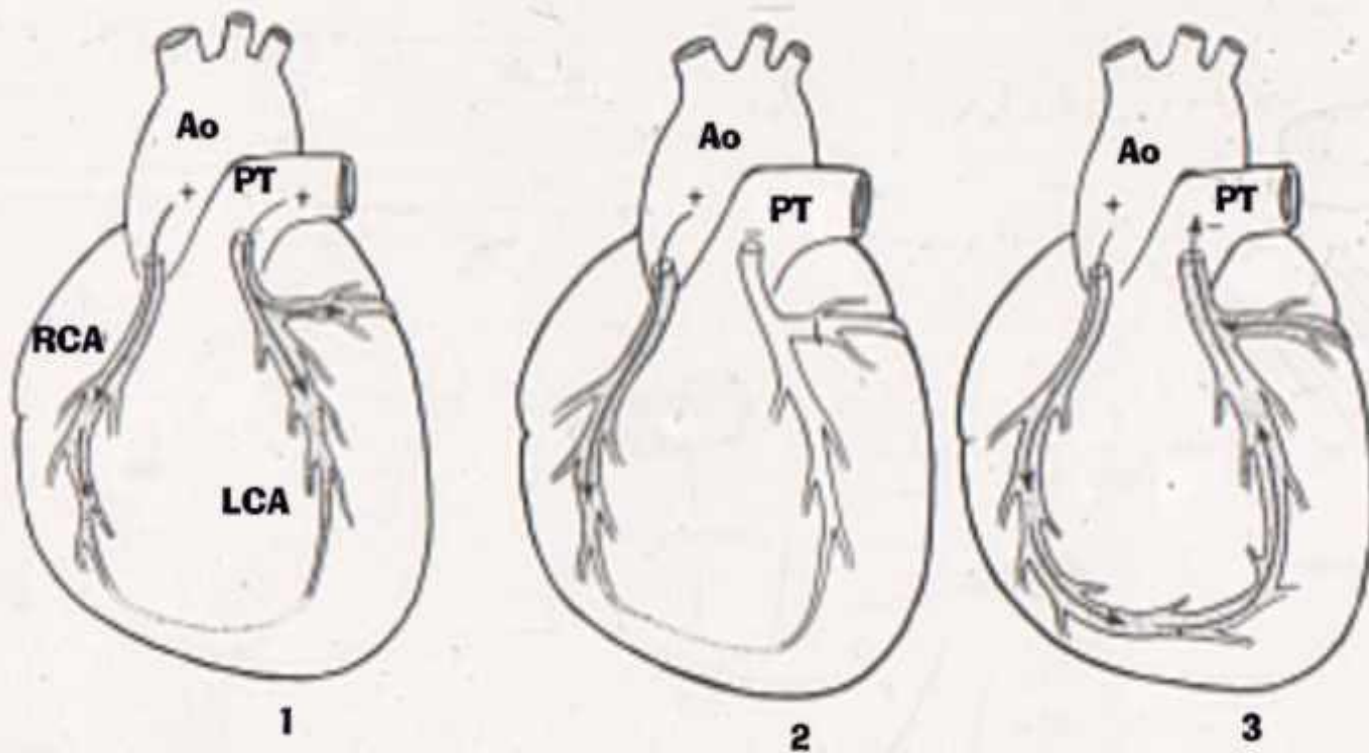
U-Shaped Retinal Arterioles



*Uncommon Defects with
Expected Adult Survival But
Exceptional Longevity*

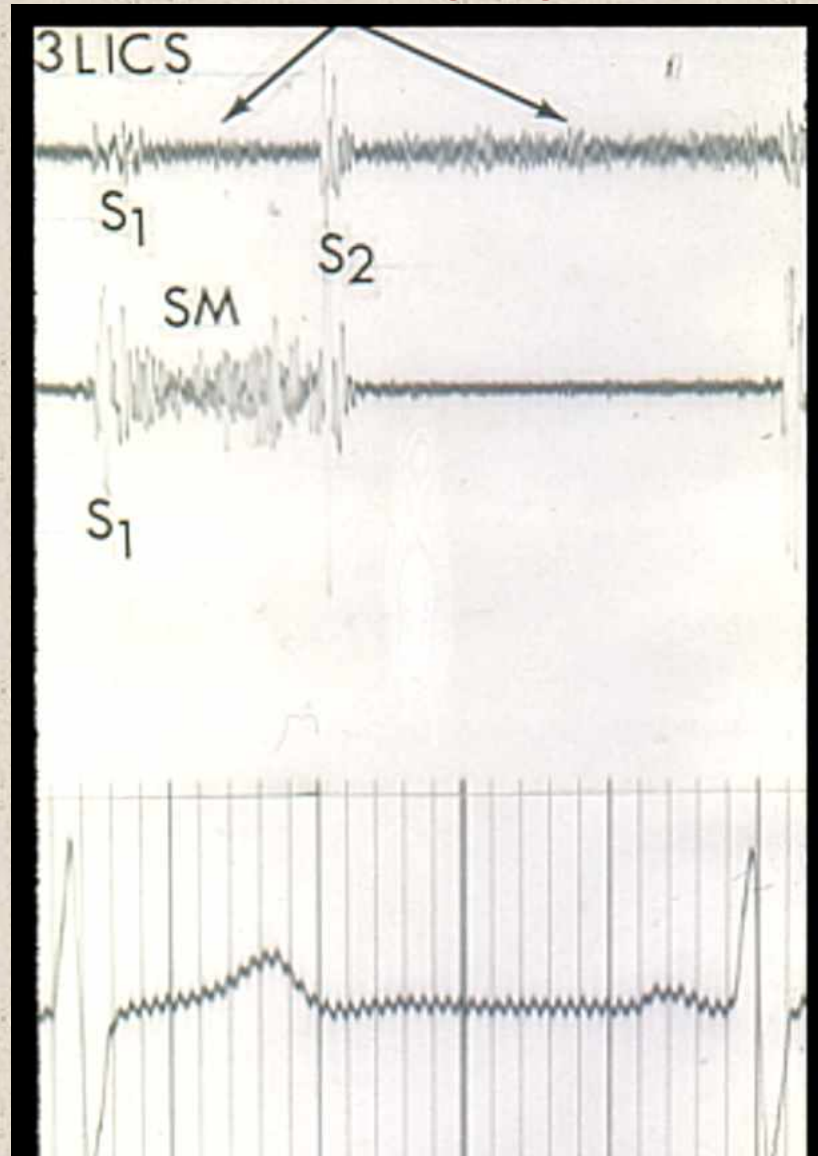
Anomalous Origin of LCA from PT

The Bland/White Garland Syndrome



Two Murmurs:

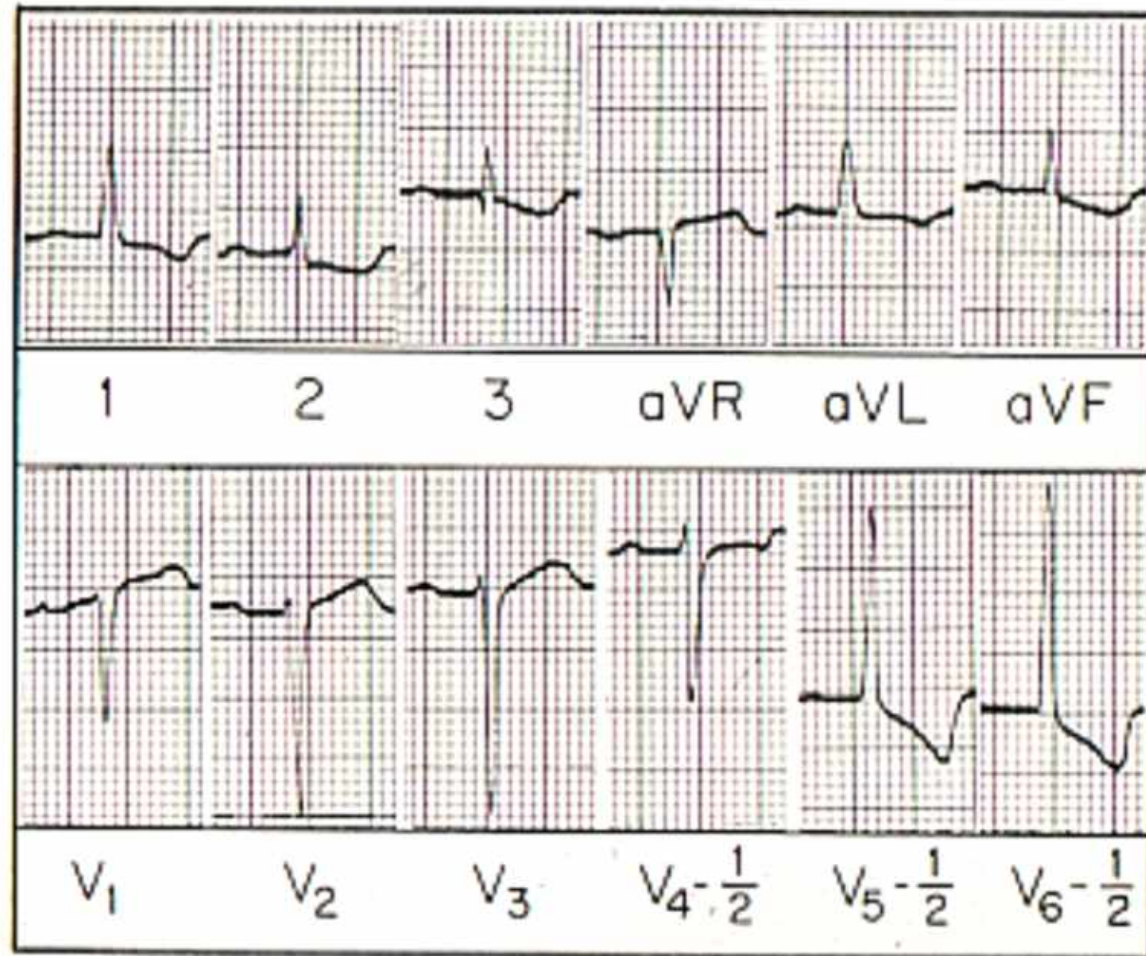
- 1) The Anomalous LCA***
- 2) Mitral Regurgitation***



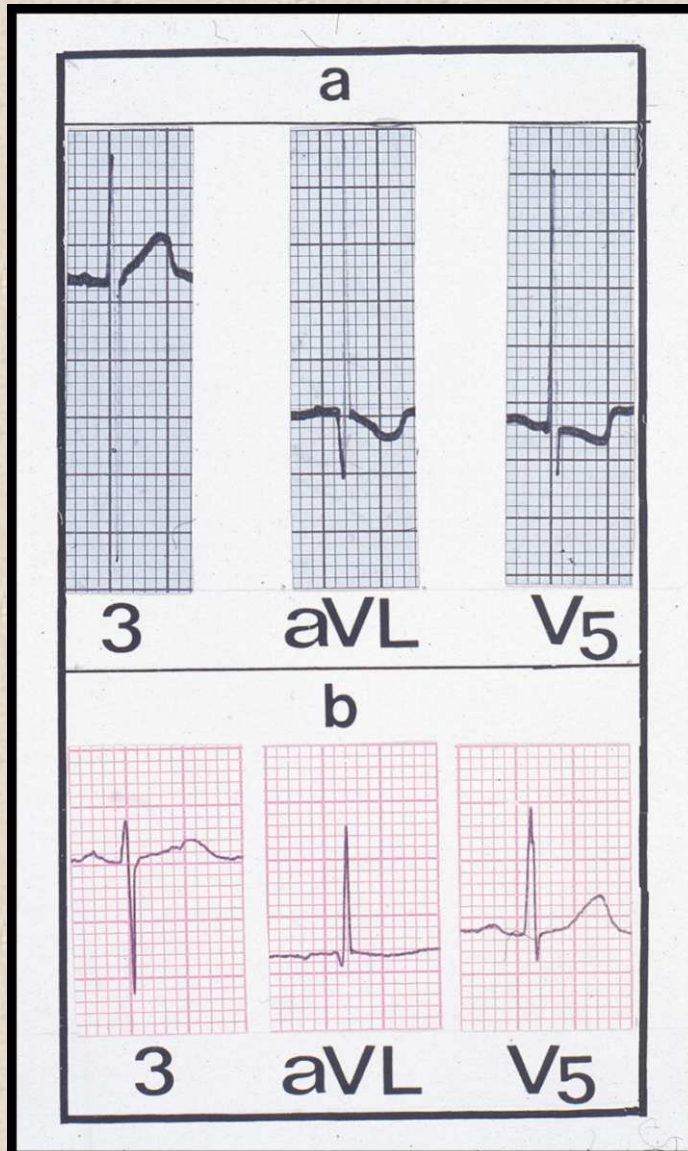
A Distinctive Electrocardiogram

Left Axis Deviation

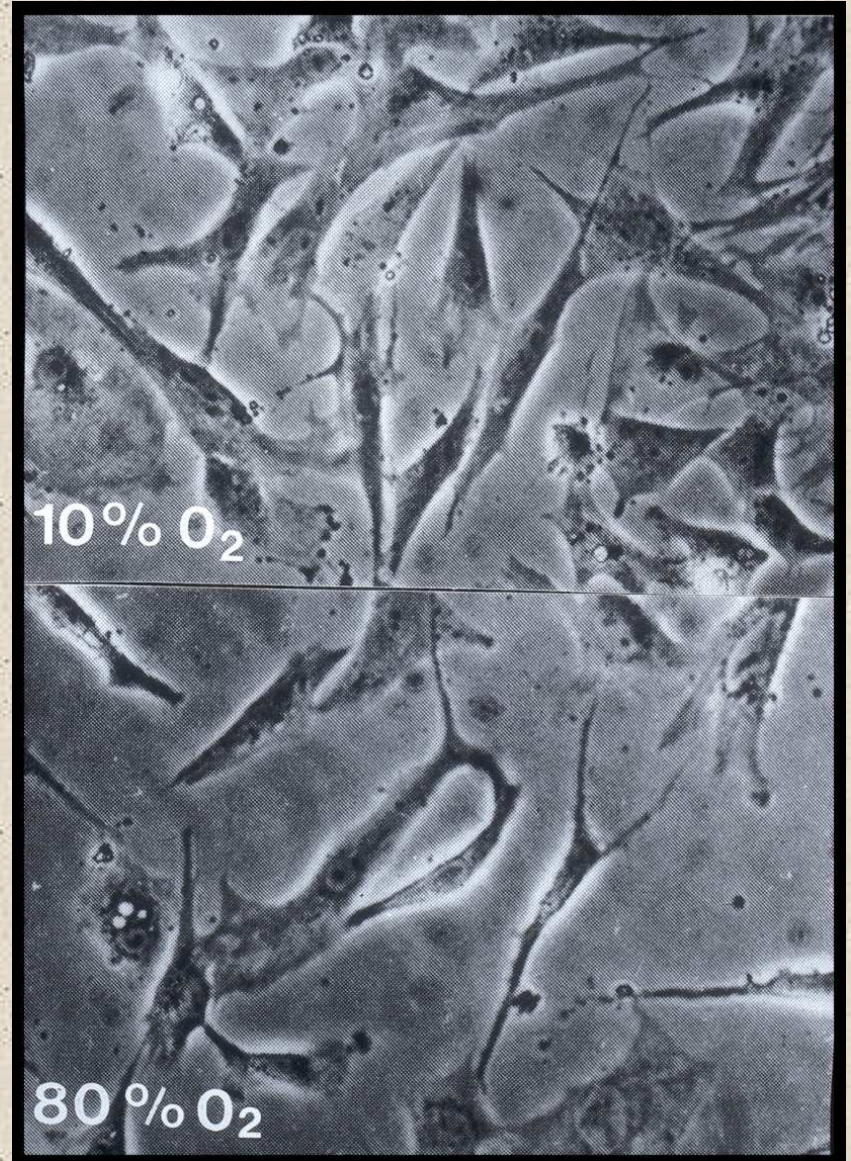
Left Ventricular Hypertrophy



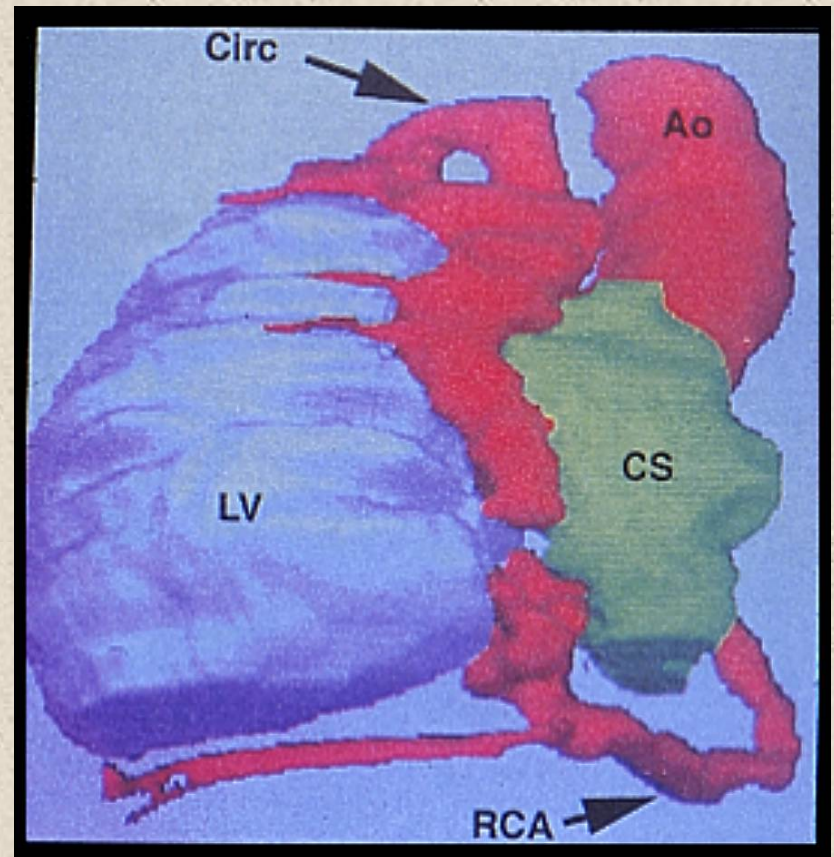
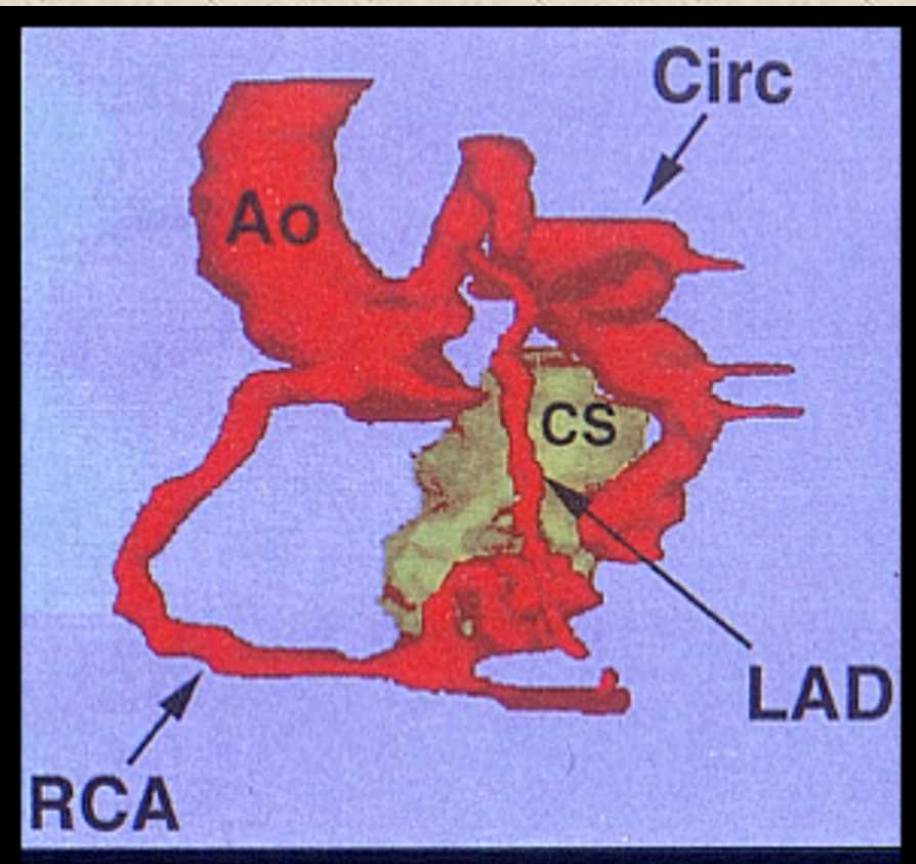
LCA from PT



Cultured Chick Cardiomyocytes



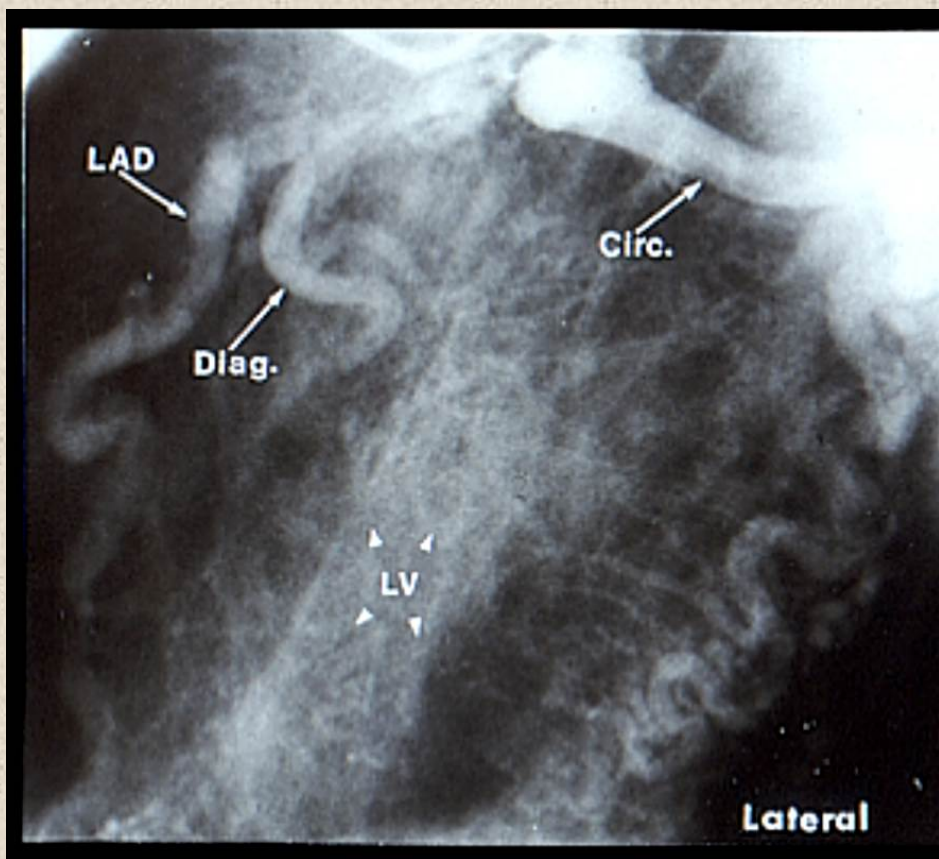
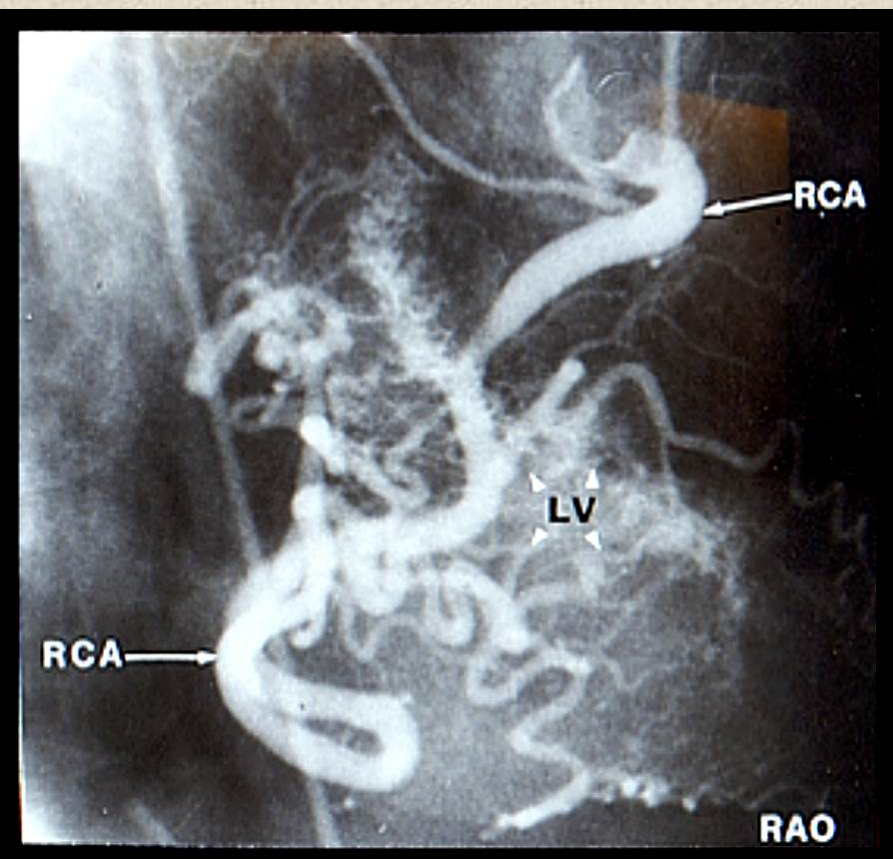
Right Coronary Artery to Coronary Sinus Fistula



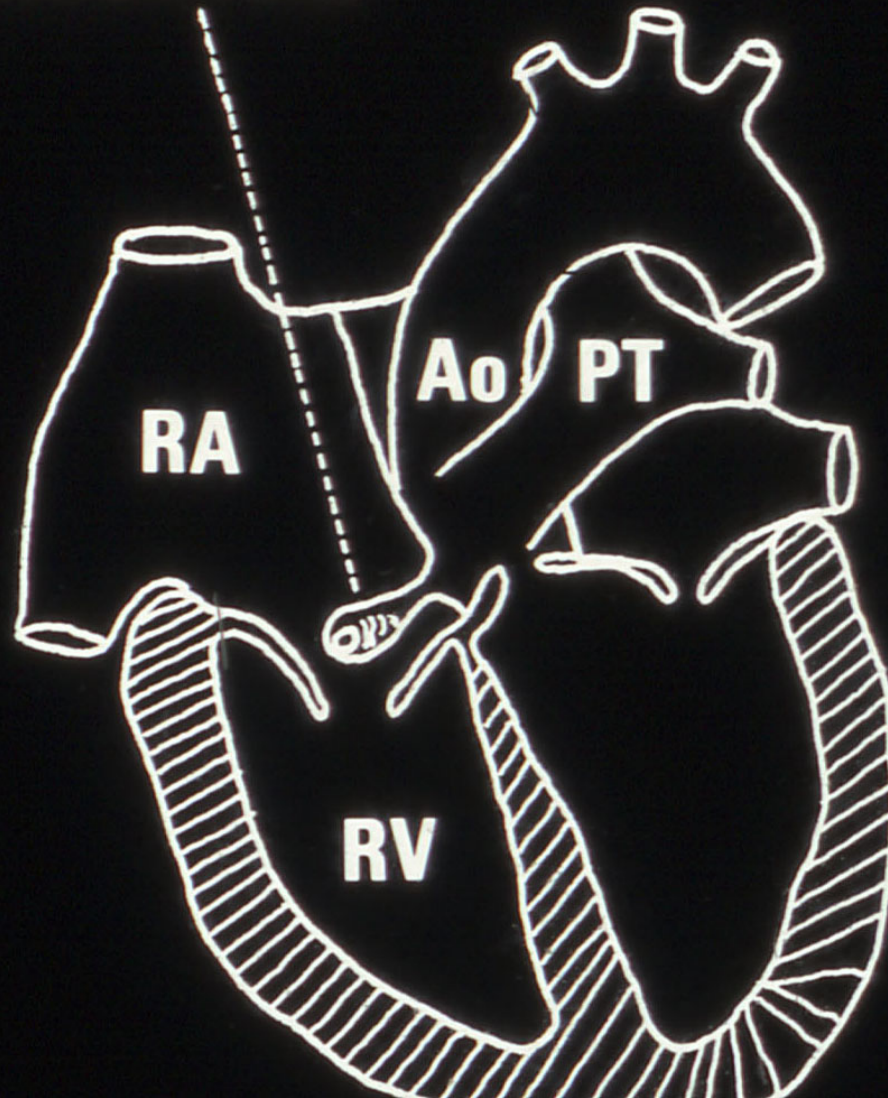
RCA to RA Fistula Age 74



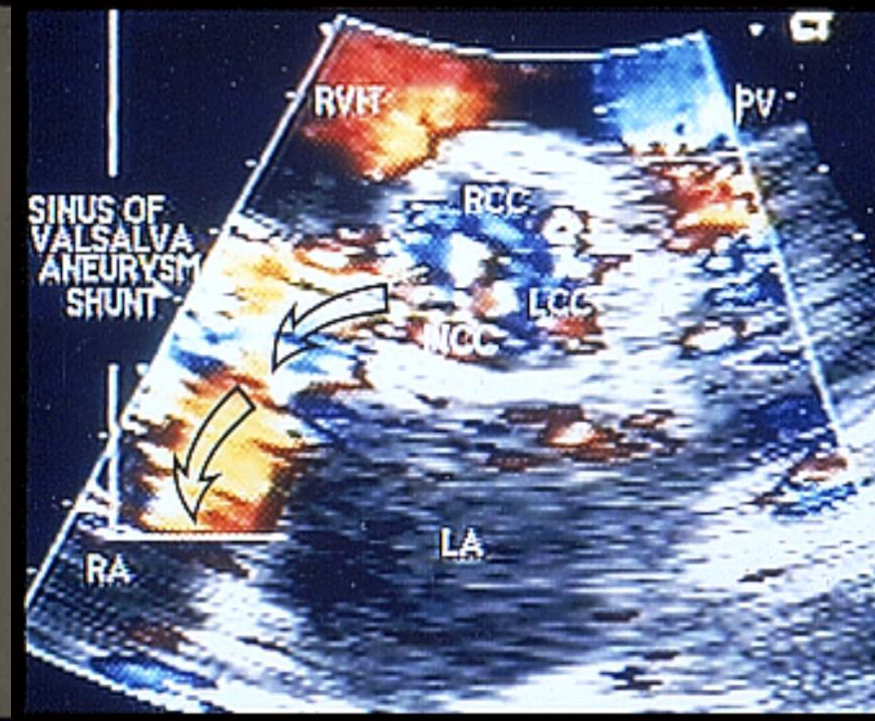
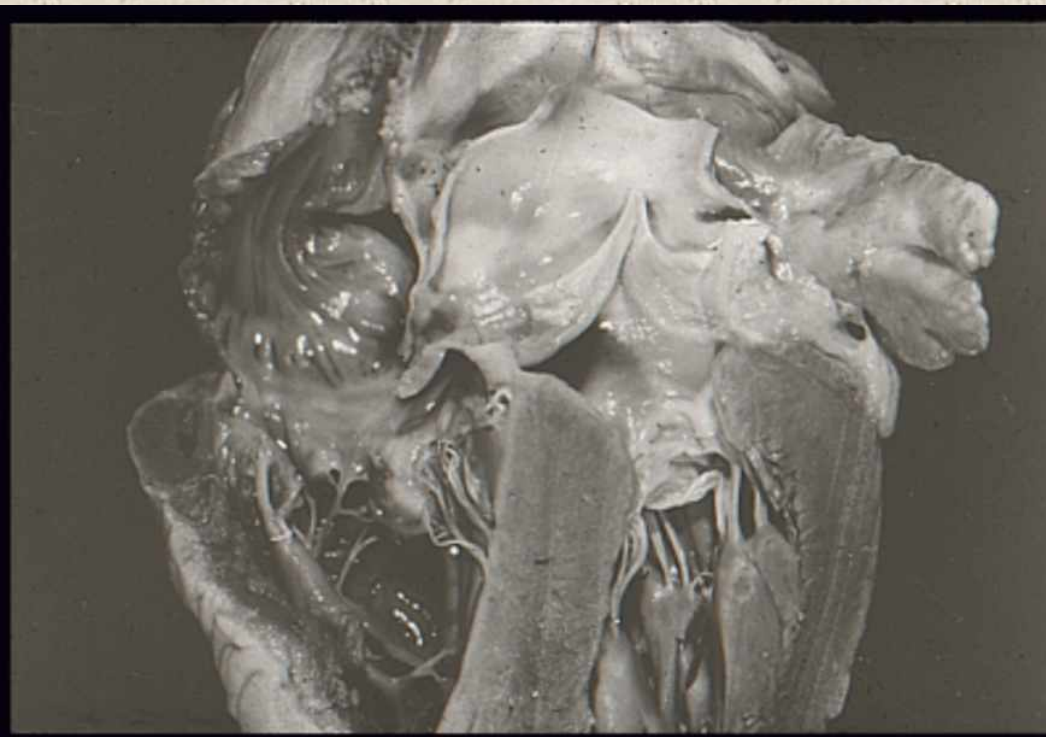
RCA to LV Fistula Age 62



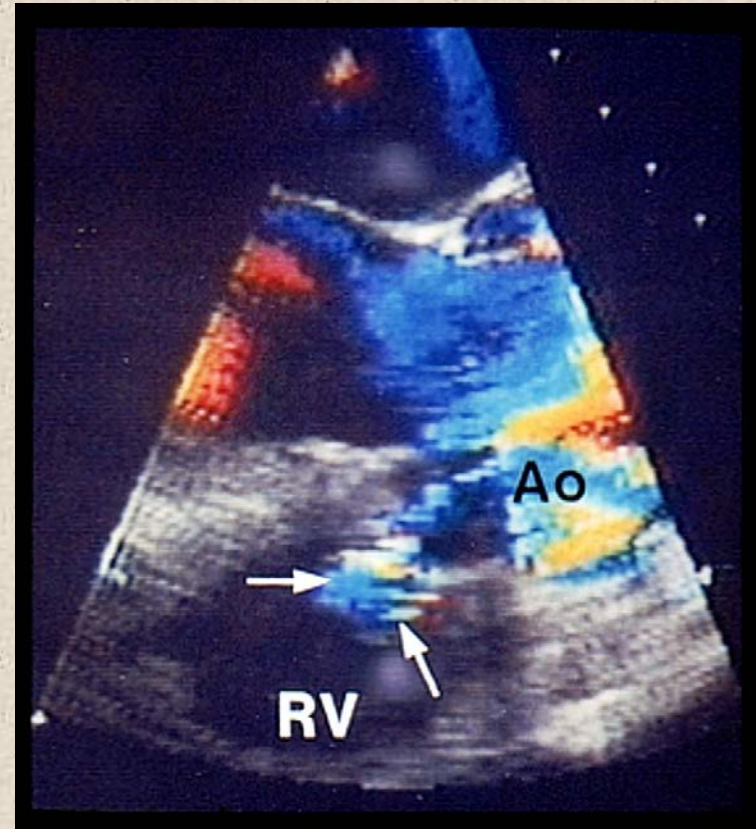
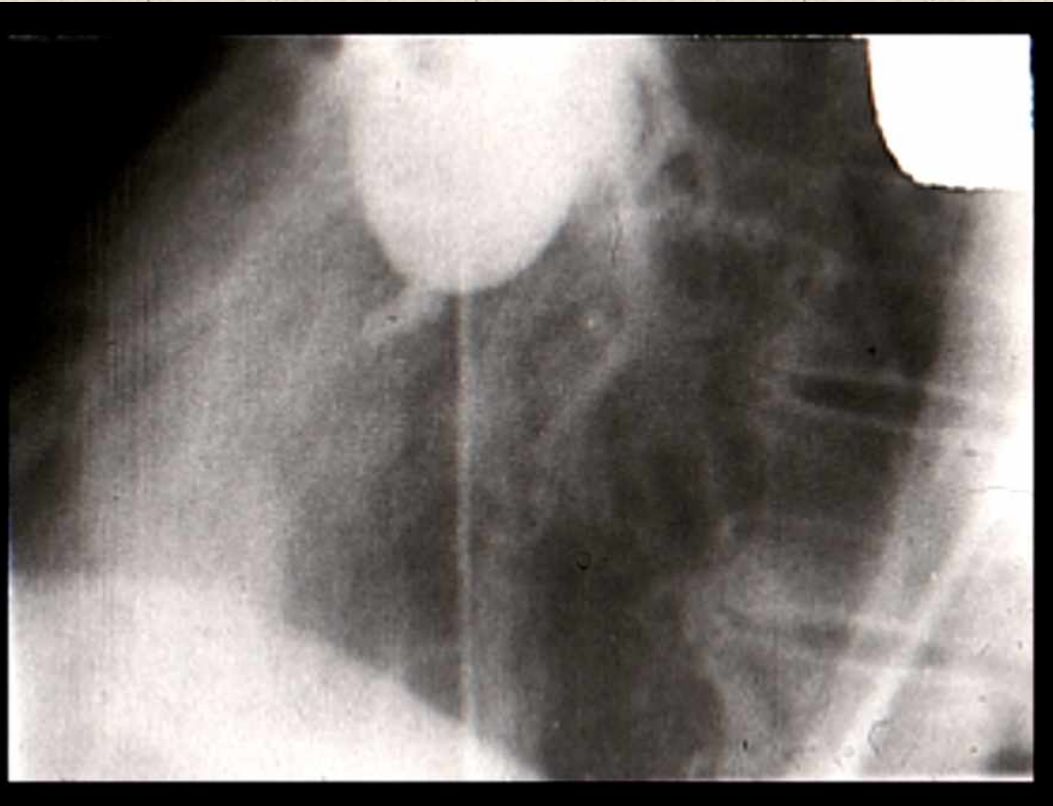
SINUS of VALSALVA ANEURYSM



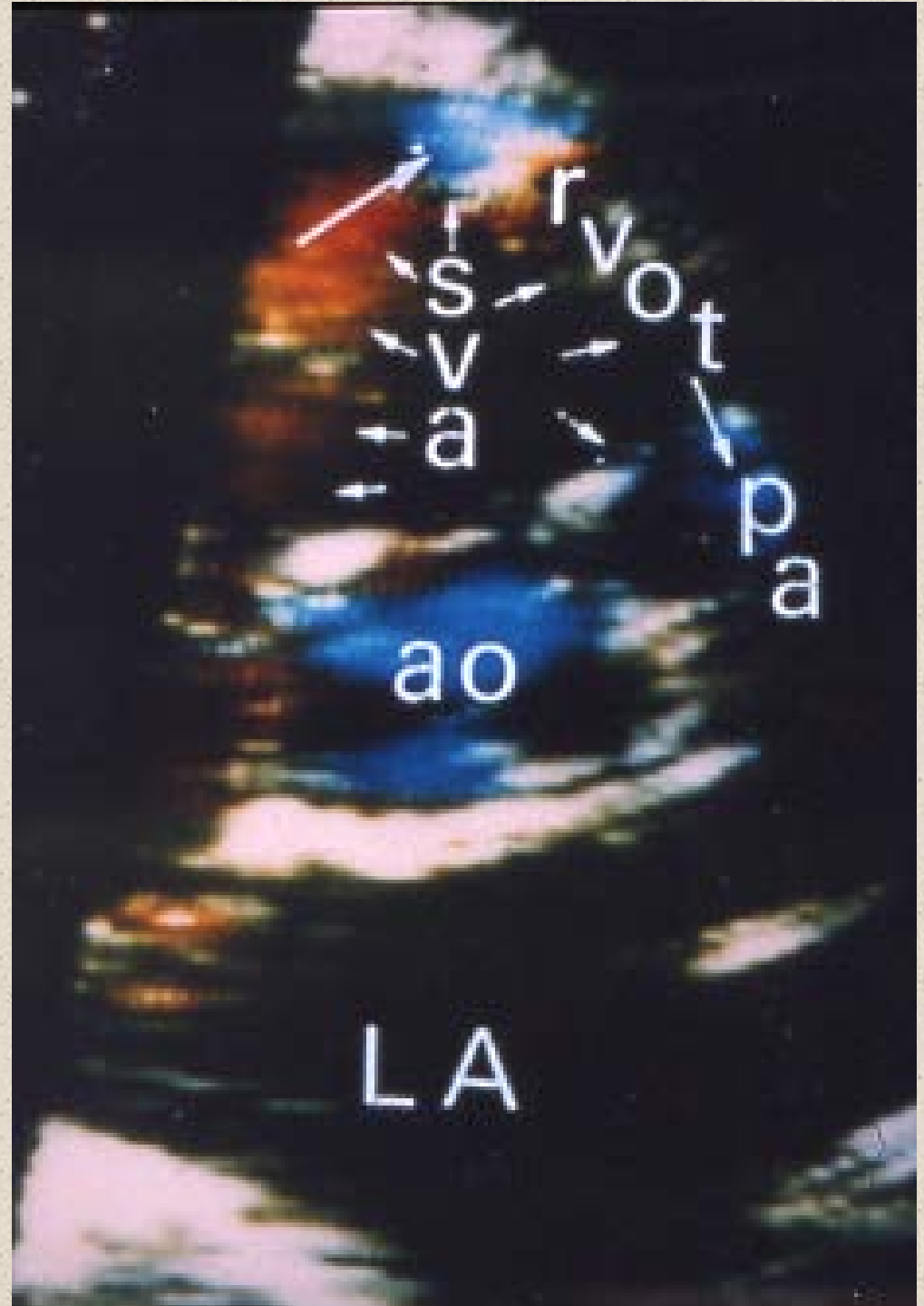
*Typical Sinus of Valsalva Aneurysm
into Right Atrium
Age 32*



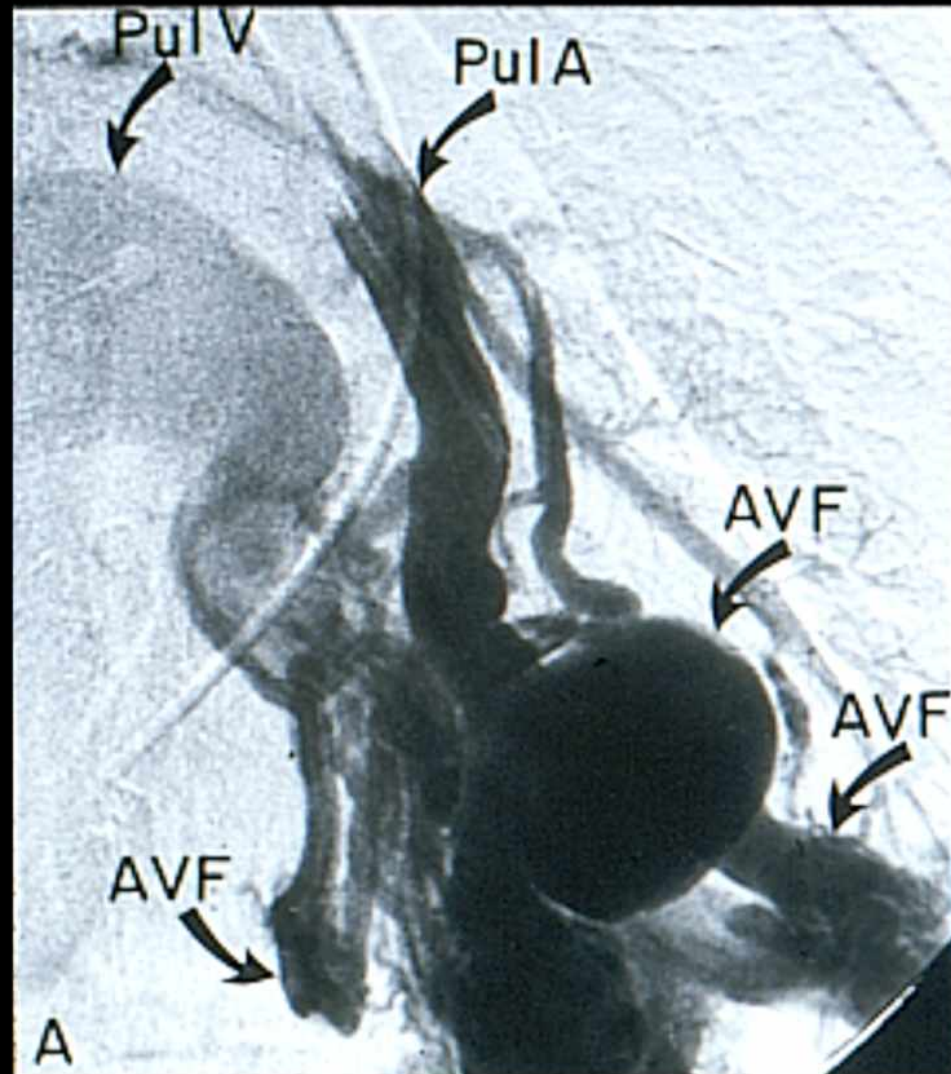
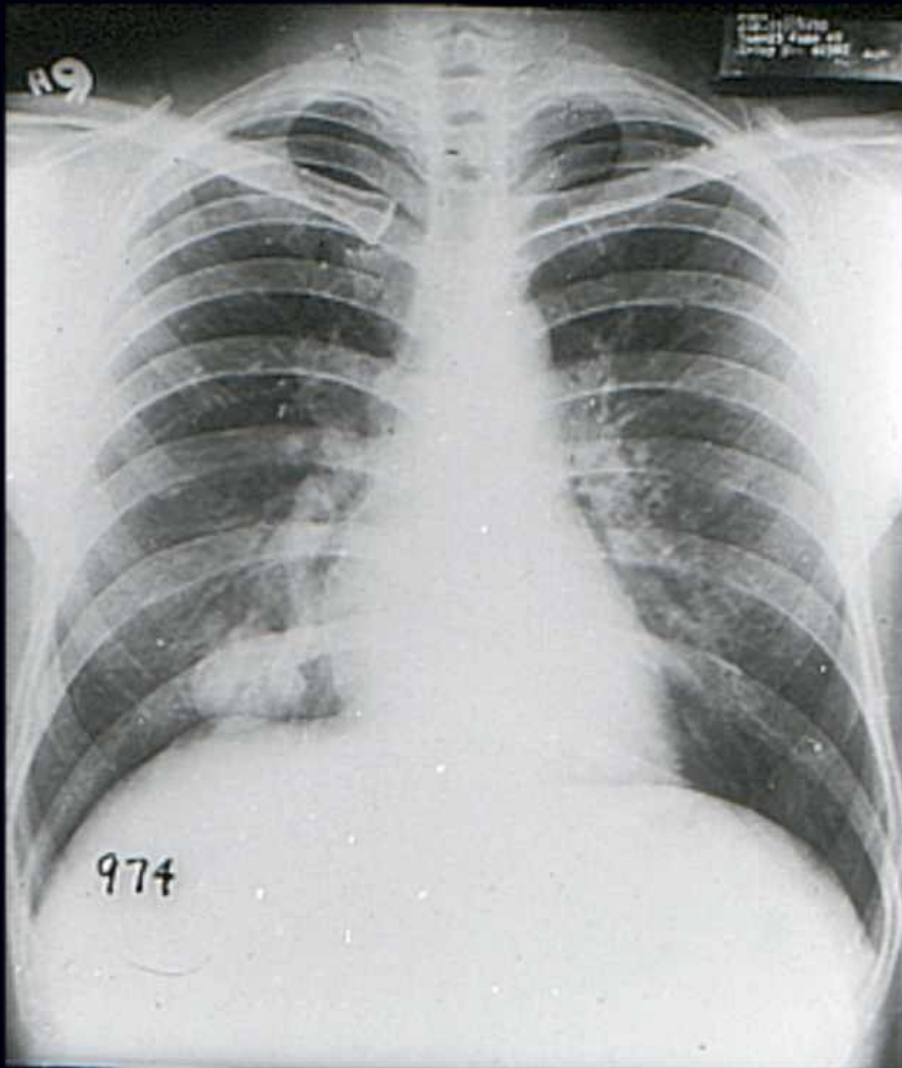
*Unusual
Sinus of Valsalva Rupture into RV
Age 35
Asymptomatic Continuous Murmur*



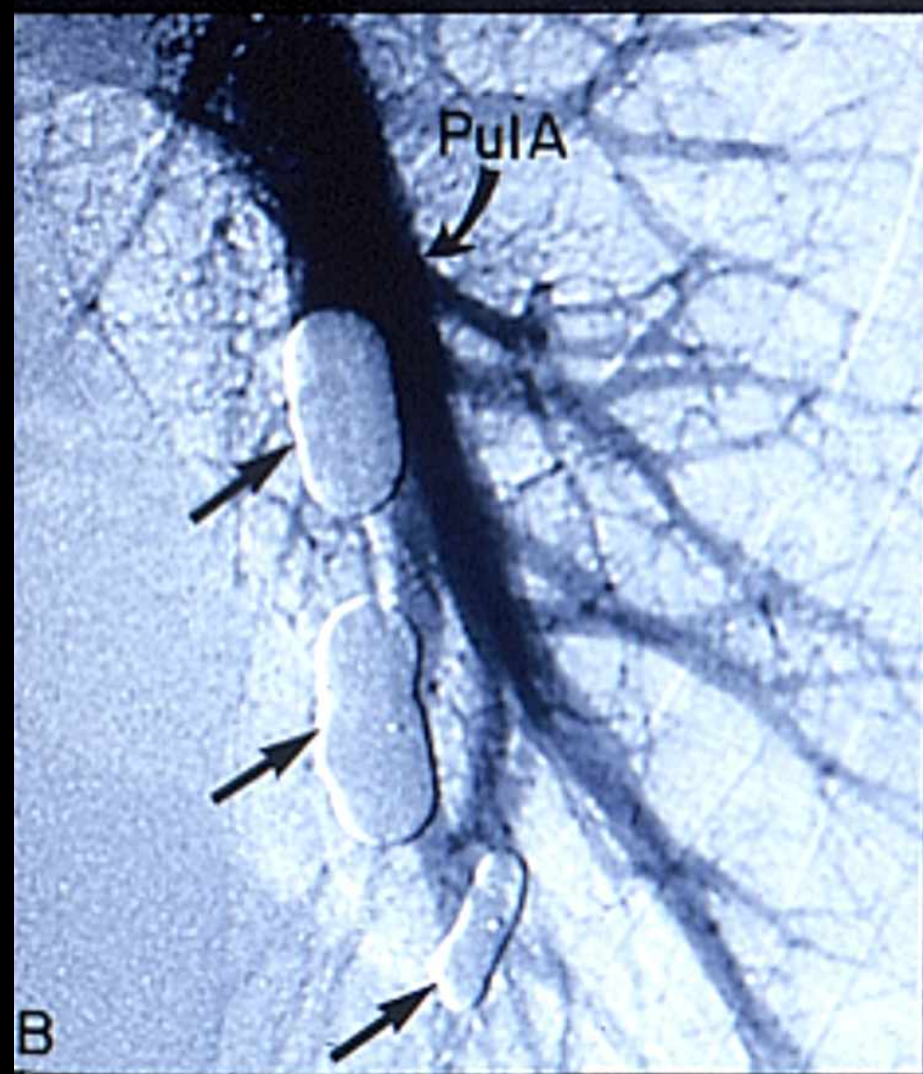
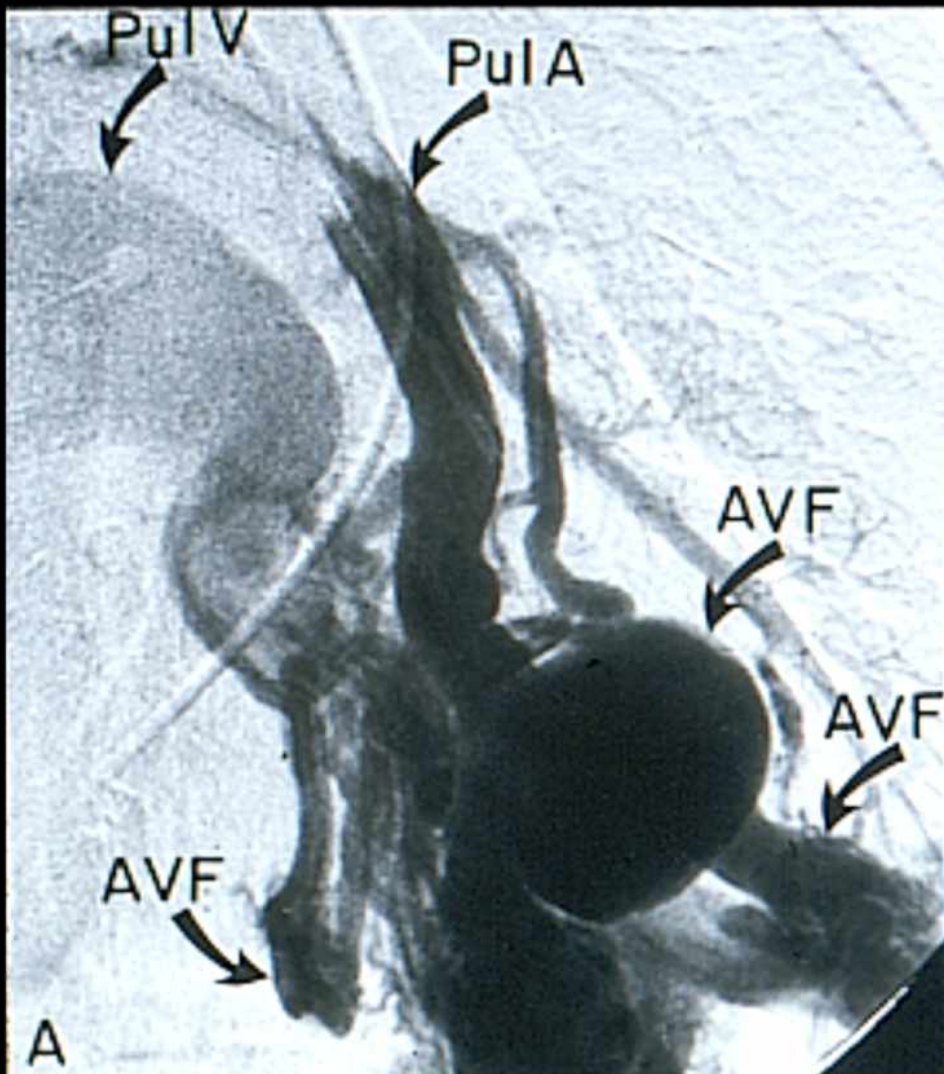
*Rare
Unruptured Sinus
of Valsalva Aneurysm
Age 85*



Pulmonary AV Fistula Age 73



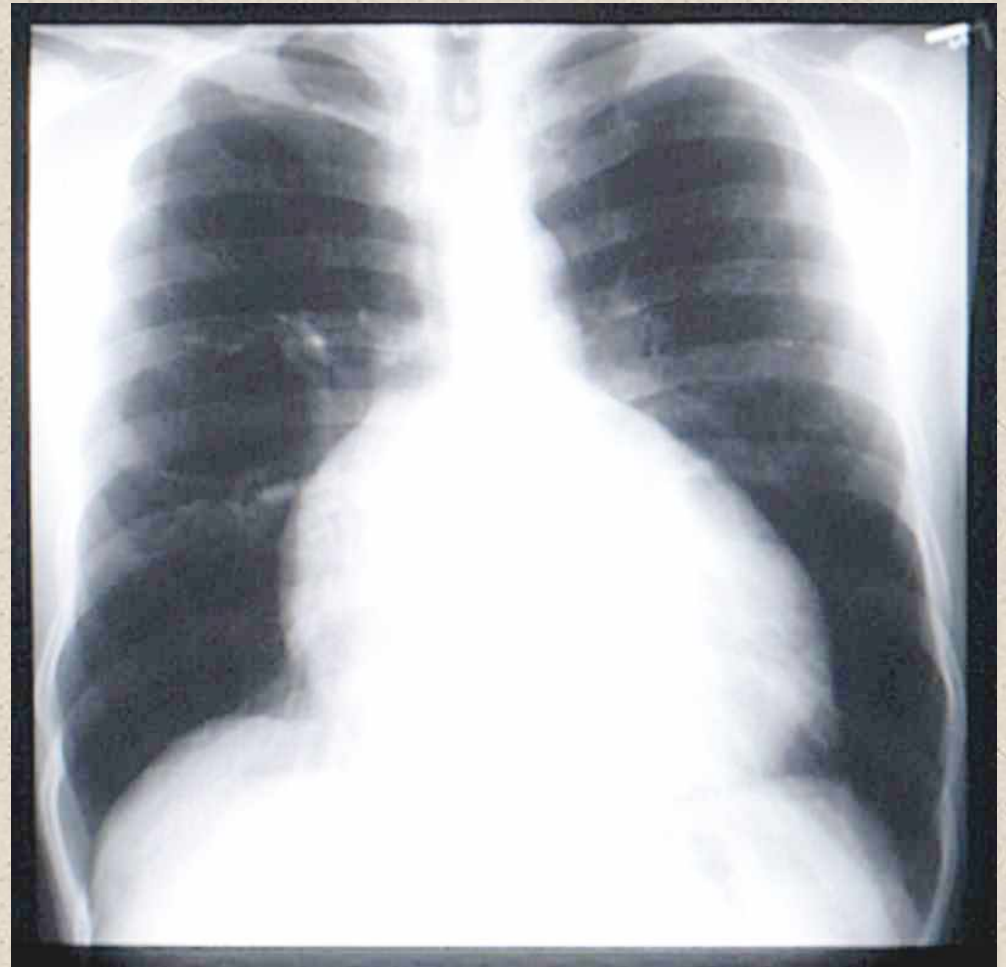
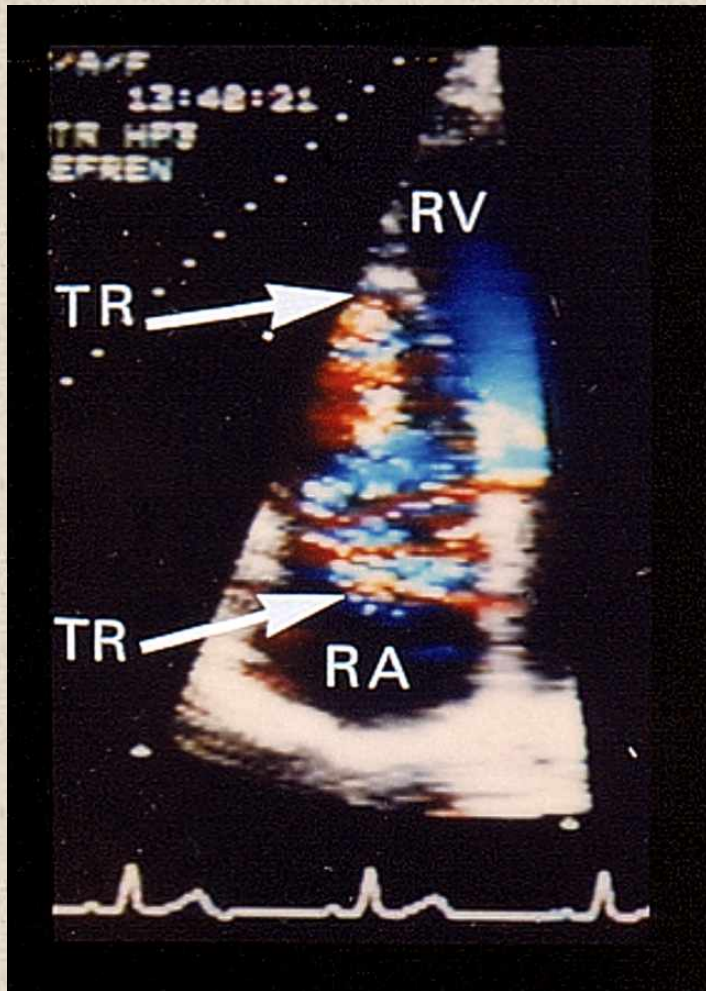
Balloon Occlusion





Wilhelm Ebstein

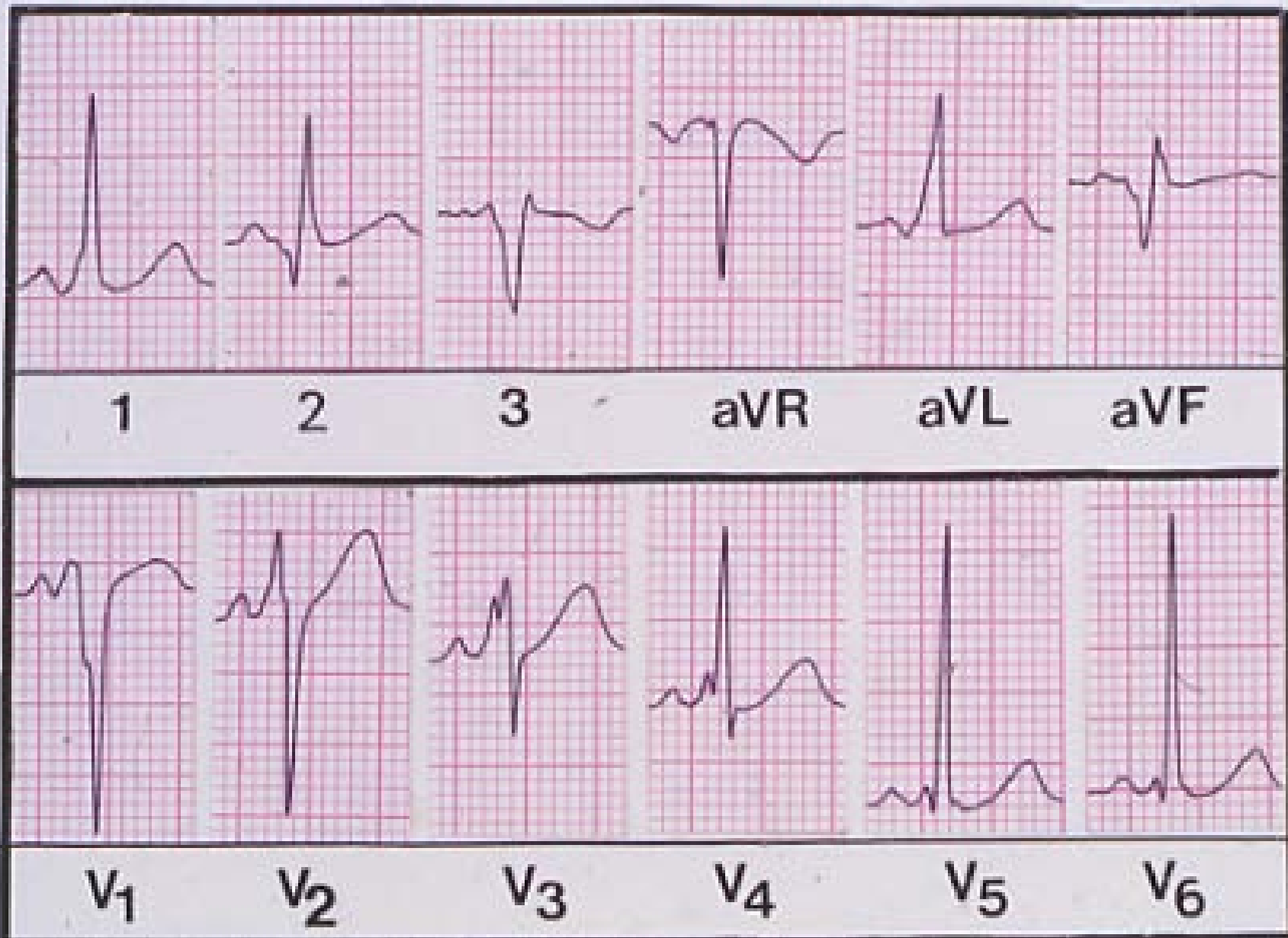
Ebstein's Anomaly

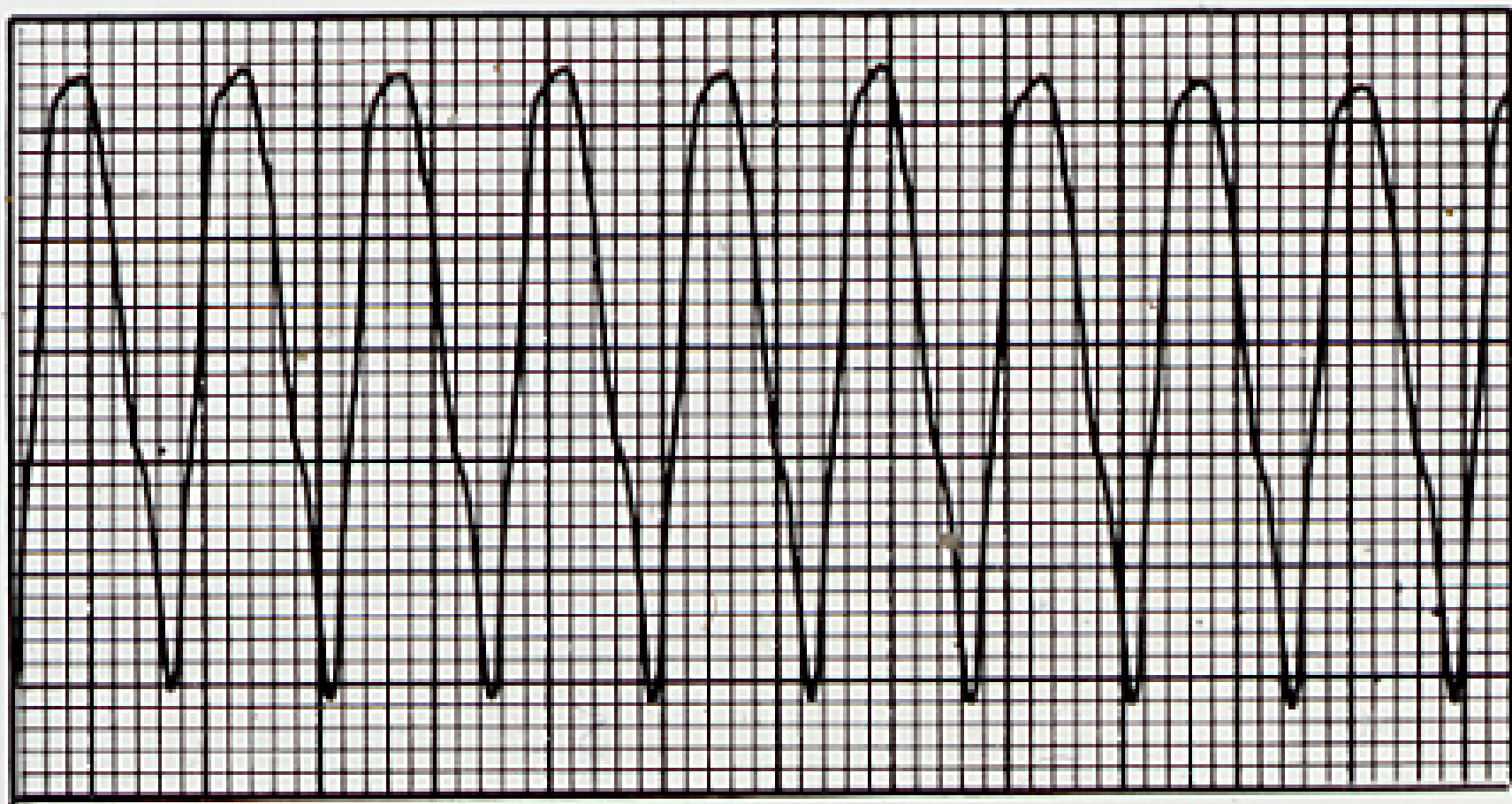


Ebstein's Anomaly of the Tricuspid Valve

There are legendary accounts of astonishing longevity with survival into the eighth and ninth decades. Ebstein's anomaly was discovered at necropsy in a 75 year old man who, in his youth, had been a lumberjack working on log booms. He was asymptomatic until his fifties, when he was obliged to outrun an irate female bear. At necropsy 25 years later, the tricuspid valve was malformed, and his right atrium was thin-walled and greatly dilated. The oldest recorded patient with Ebstein's anomaly lived to age 85 years with no cardiac symptoms until age 79 when he developed right ventricular failure.

Ebstein's Anomaly WPW Age 62





LEAD V₁

Ventricular Inversion

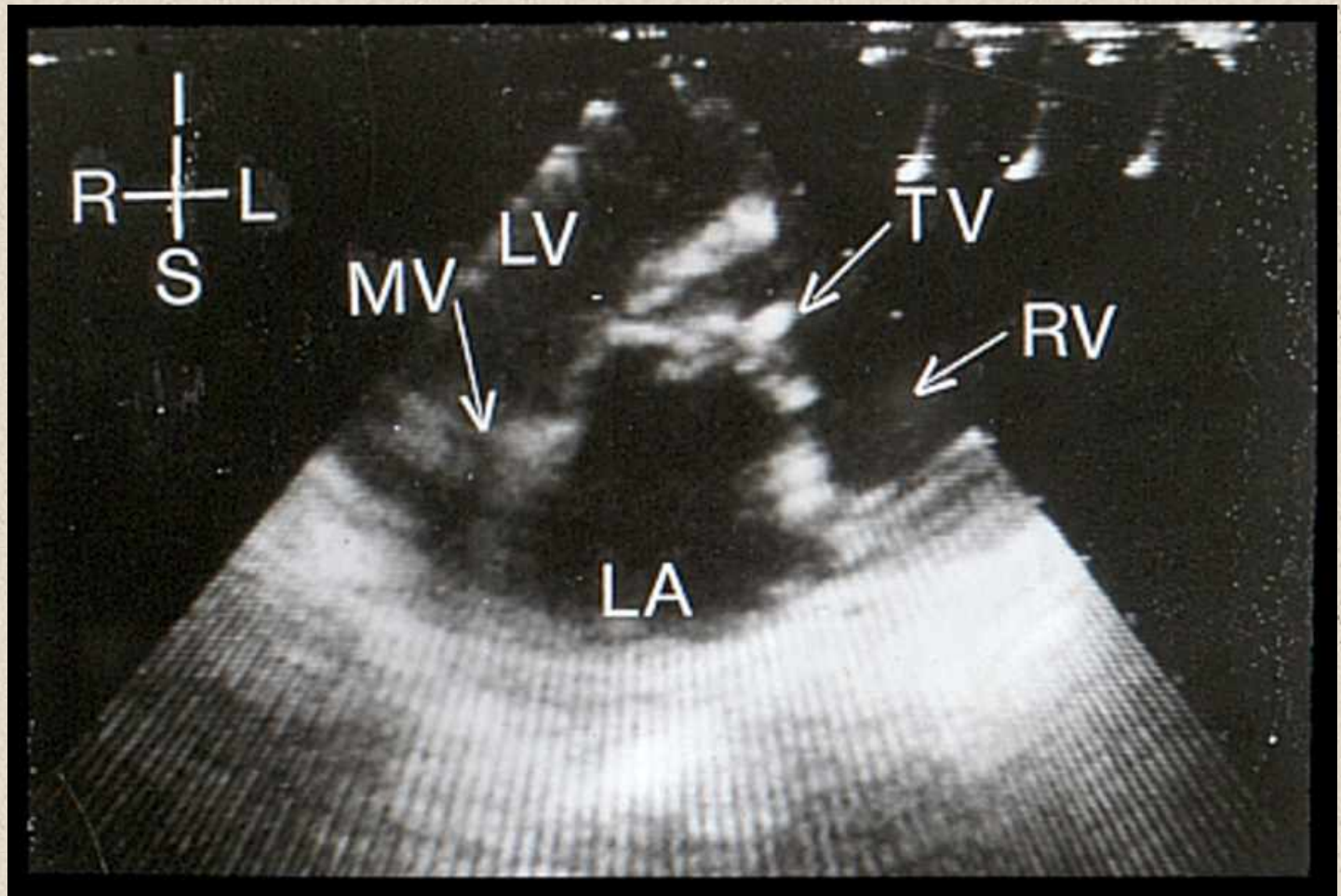
Ist der rechte Ventrikel der richtige Ventrikel?

Is the right ventricle the right ventricle?

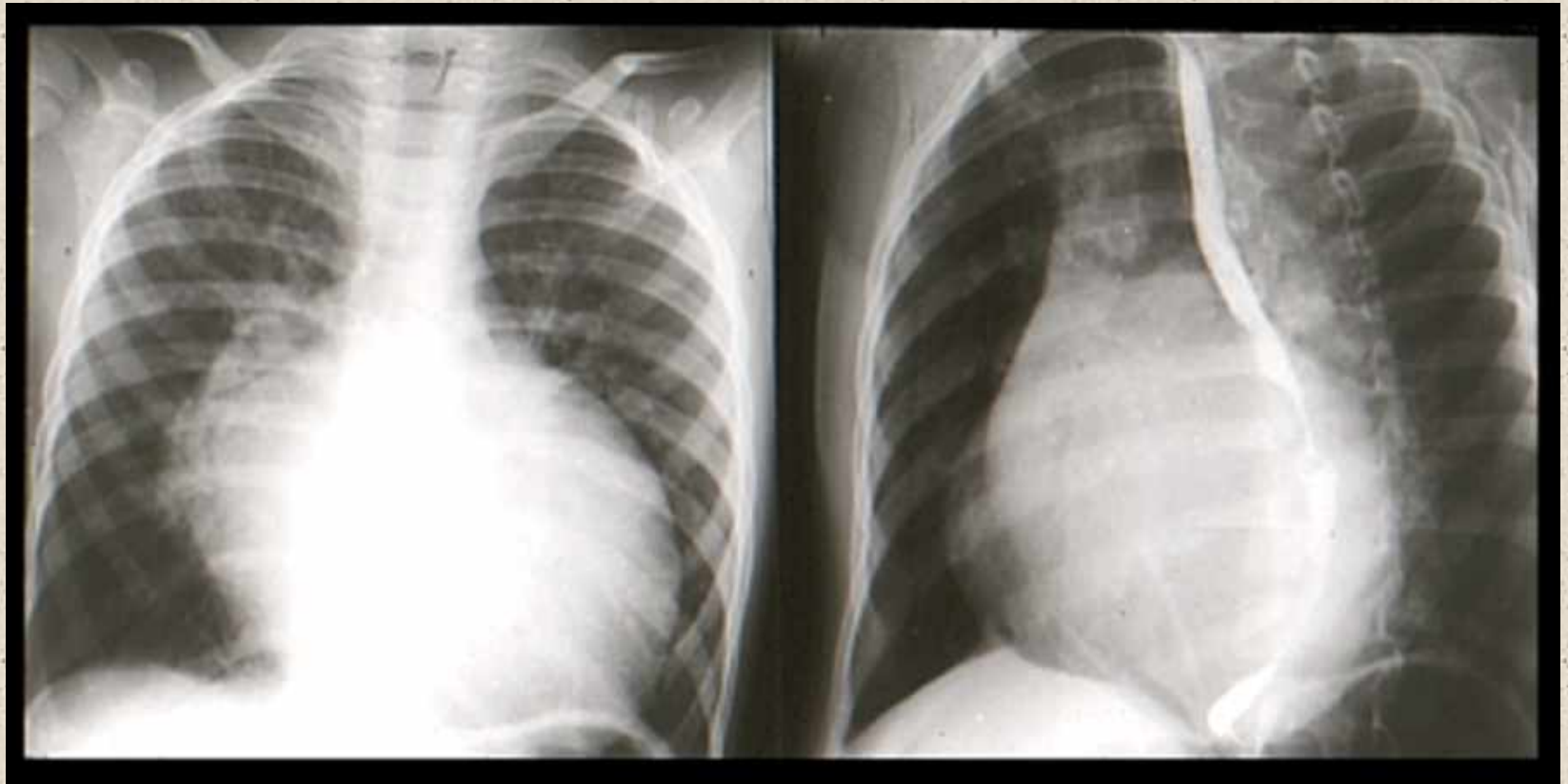
Prof. Dr H. C. Kallfelz Hannover

The vulnerable chamber is the subaortic morphologic right ventricle which is perfused by a morphologic right coronary artery and has an inherently lower ejection fraction than a morphologic left ventricle.

Left-sided Ebstein's Anomaly Congenitally Corrected TGA

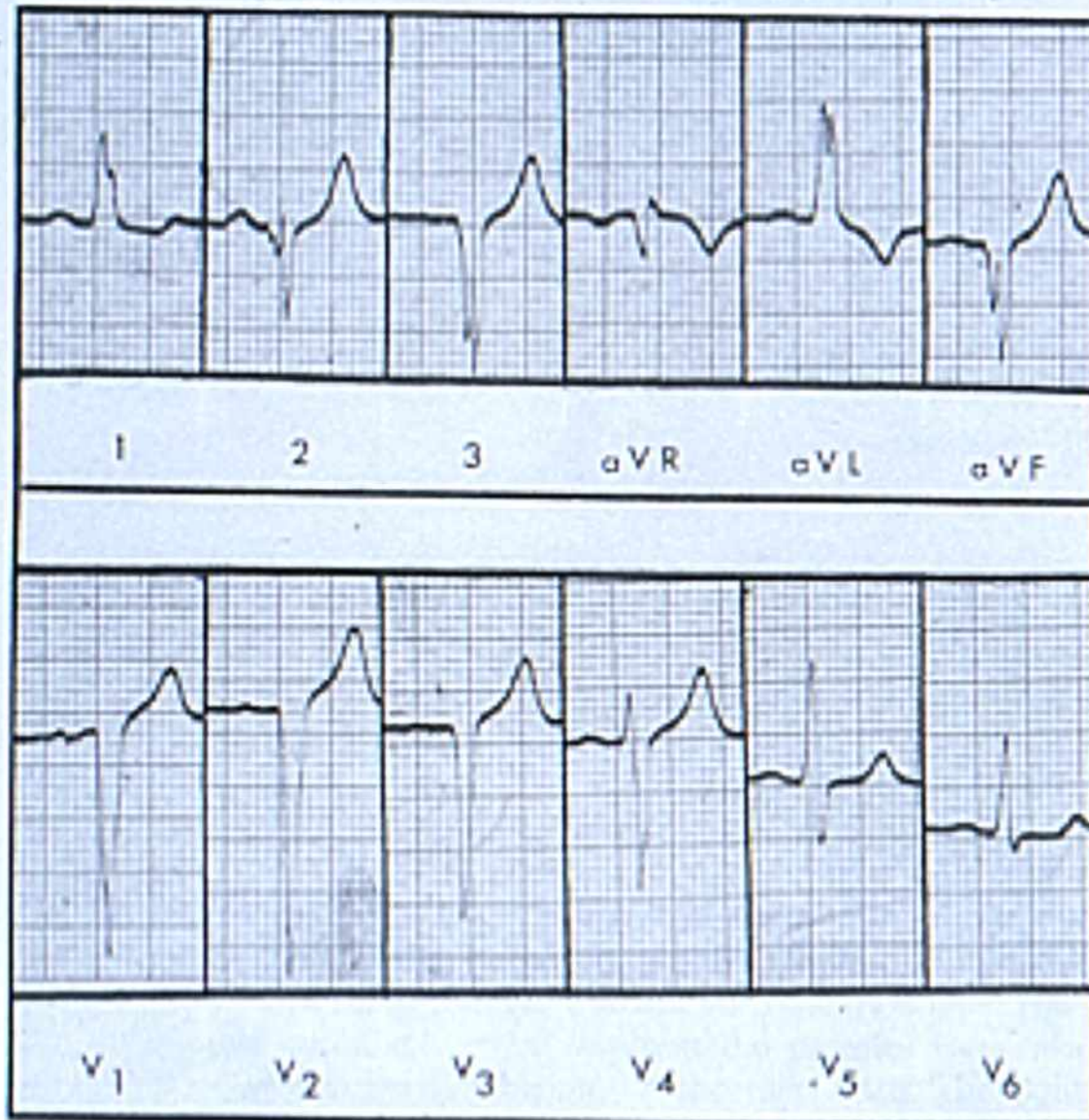


Left-Sided Ebstein's Anomaly



Ventricular Inversion

Right to Left Septal Depolarization



*Common Defects with
Unexpected Adult Survival and
Exceptional Longevity*

Complete Transposition of the Great Arteries

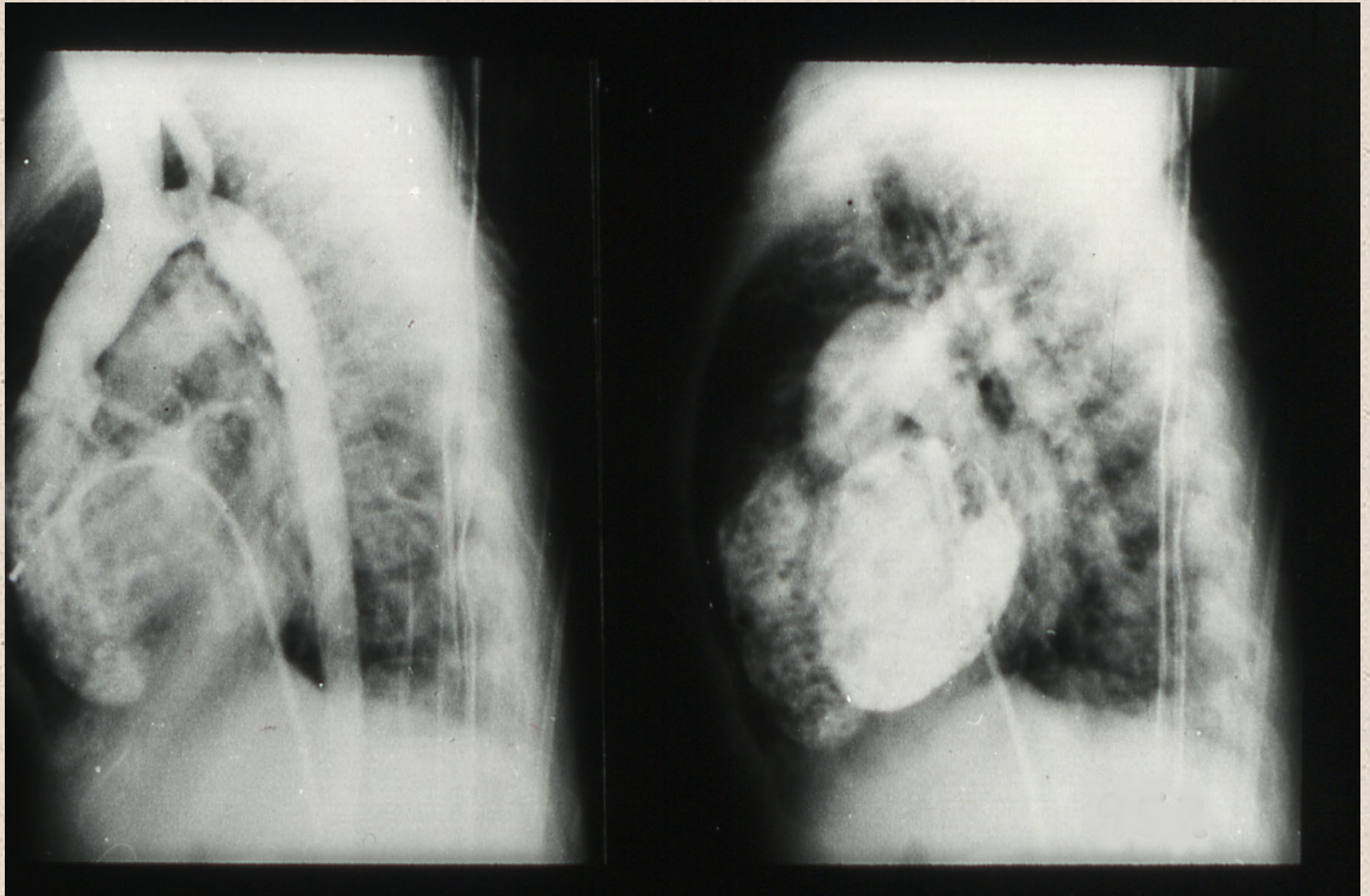
Death Rates Without Intervention:

First week 30%

First month 50%

First year 90%

Complete Transposition of the Great Arteries Age 18 Months





Merry Christmas

Against All Odds
Age 36 Years



*Common Defects with
Unexpected Adult Survival and
Exceptional Longevity*



Arthur Fallot 1888

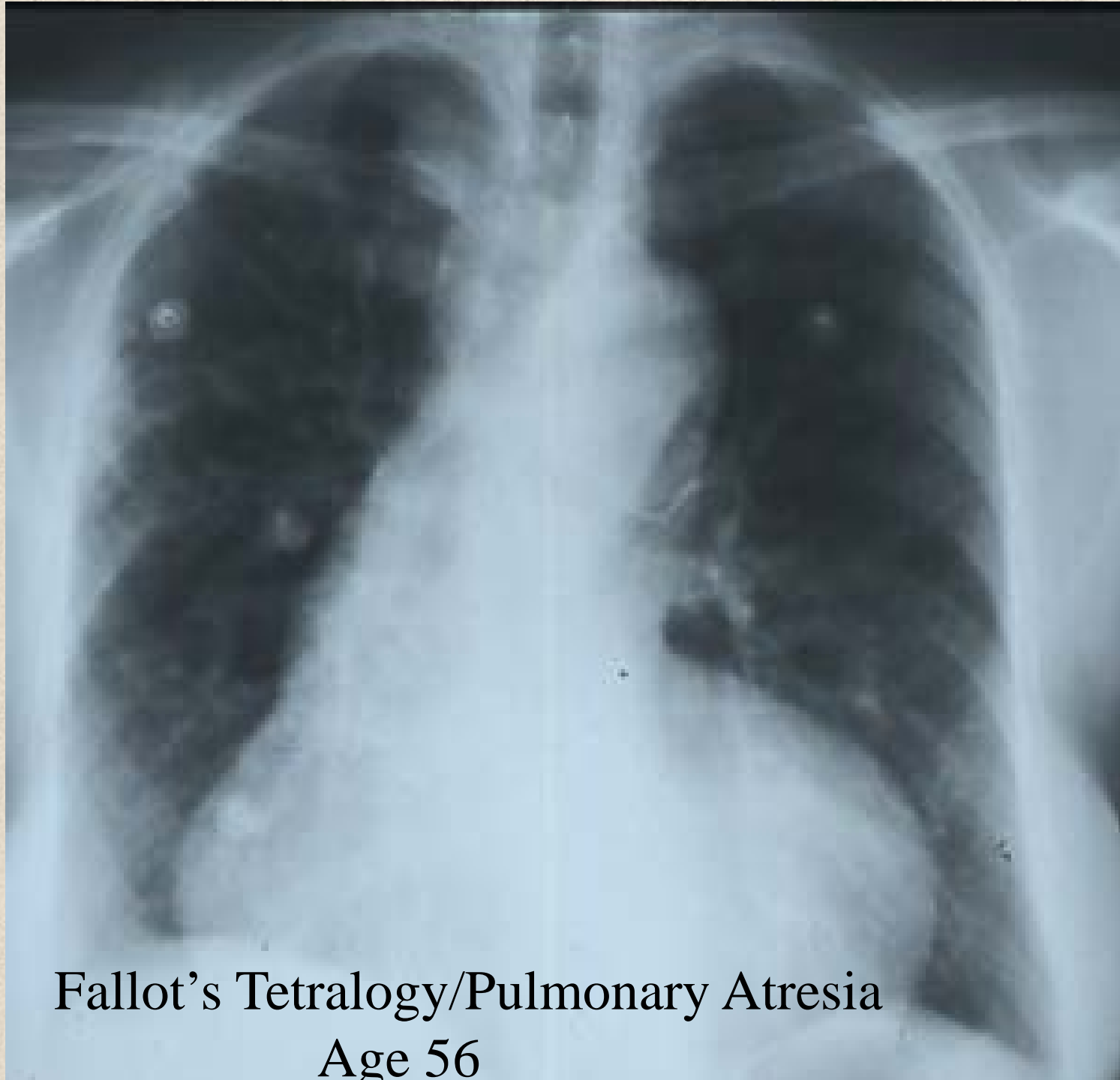
We have seen from our observations that cyanosis, especially in the adult, is the result of a small number of cardiac malformations well determined. One of these cardiac malformations is much more frequent than others.

Fallot's Tetralogy

Survival Without

Intervention

<i>1st birthday</i>	<i>---</i>	<i>66%</i>
<i>Age 3 years</i>	<i>---</i>	<i>50%</i>
<i>Age 10</i>	<i>---</i>	<i>25%</i>
<i>Age 20</i>	<i>---</i>	<i>11%</i>
<i>Age 30</i>	<i>---</i>	<i>6%</i>
<i>Age 40</i>	<i>---</i>	<i>3%</i>



Fallot's Tetralogy/Pulmonary Atresia
Age 56

Oldest Unoperated Tetralogy

Surgical Repair of Tetralogy of Fallot in a Seventy-five Year Old Patient,” the oldest known unoperated survivor and the oldest to undergo intracardiac repair.

International Journal of Cardiology 2008.

“Increasing geriatric clinical and pathological experience is beginning to shed light on a host of problems relating to the life history of cardiovascular disease. In any large series of geriatric necropsies, for example, atrial septal defect is always well represented, but where’s the Maladie de Roger? Assuming it does provide immortality, it must either close spontaneously in middle life or have long since run its mortal course.”

Paul Wood, O.B.E., M.D., F.R.C.P.

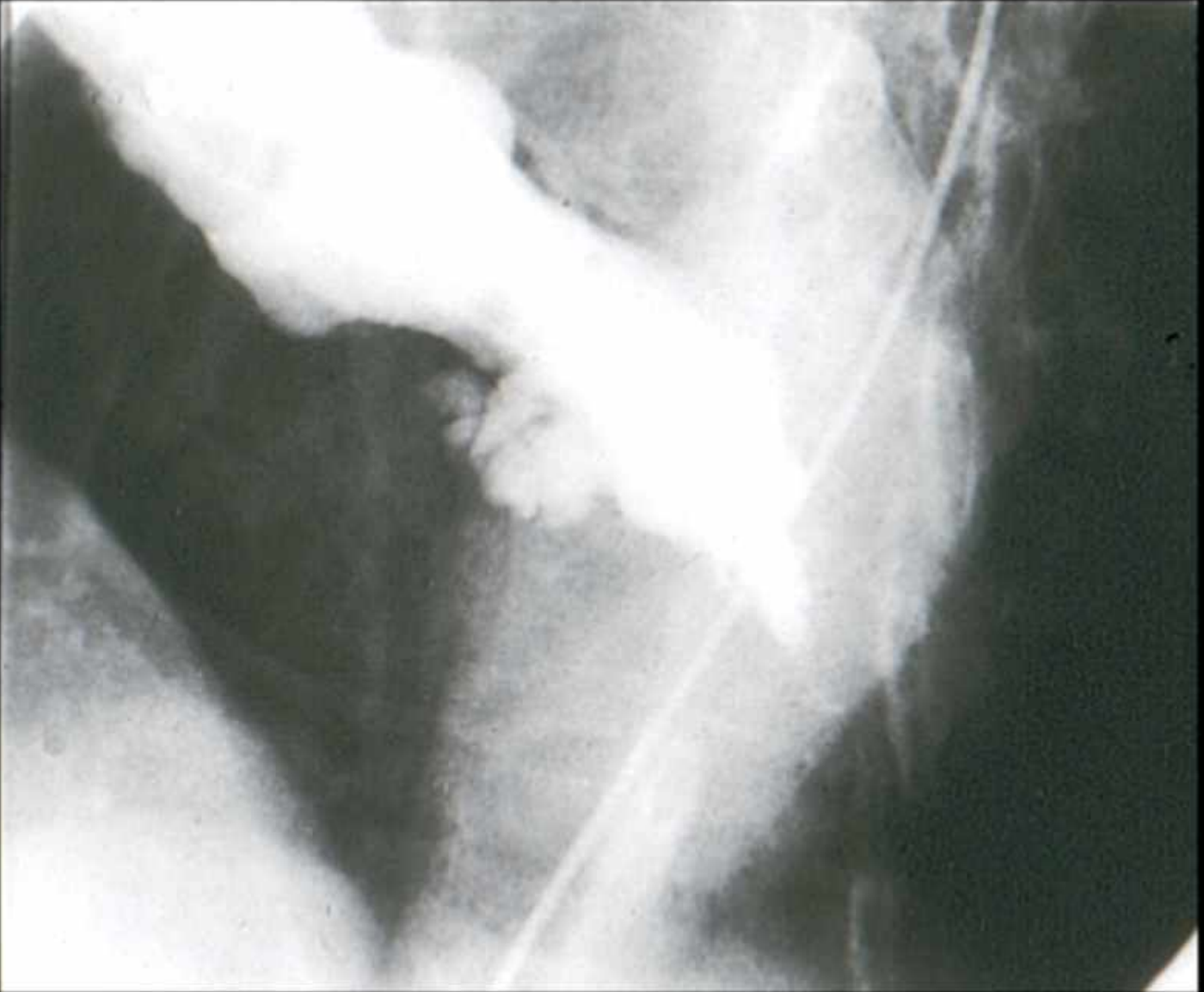
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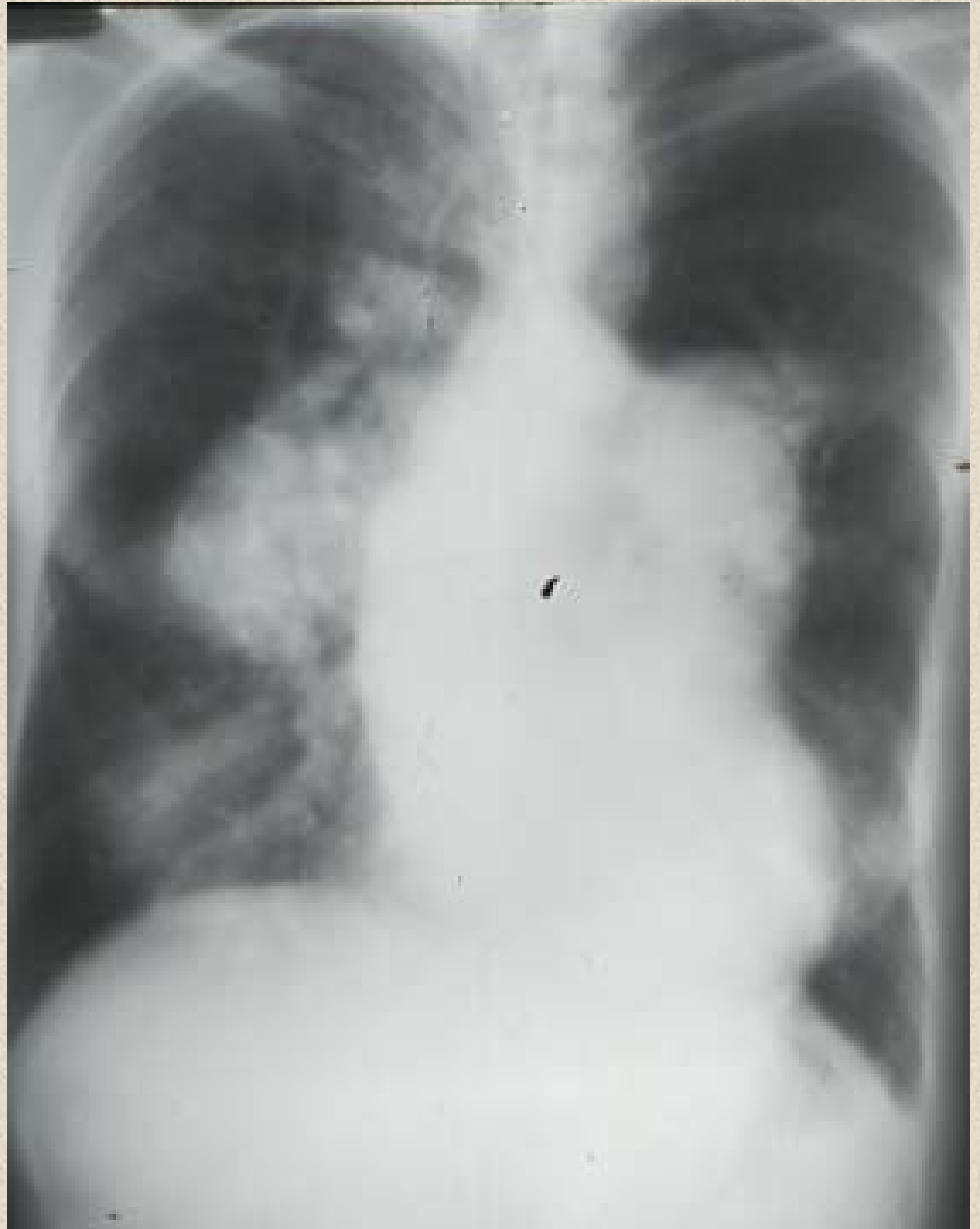
Necropsy VSD Age 68



Closed Inlet VSD
A Rarity



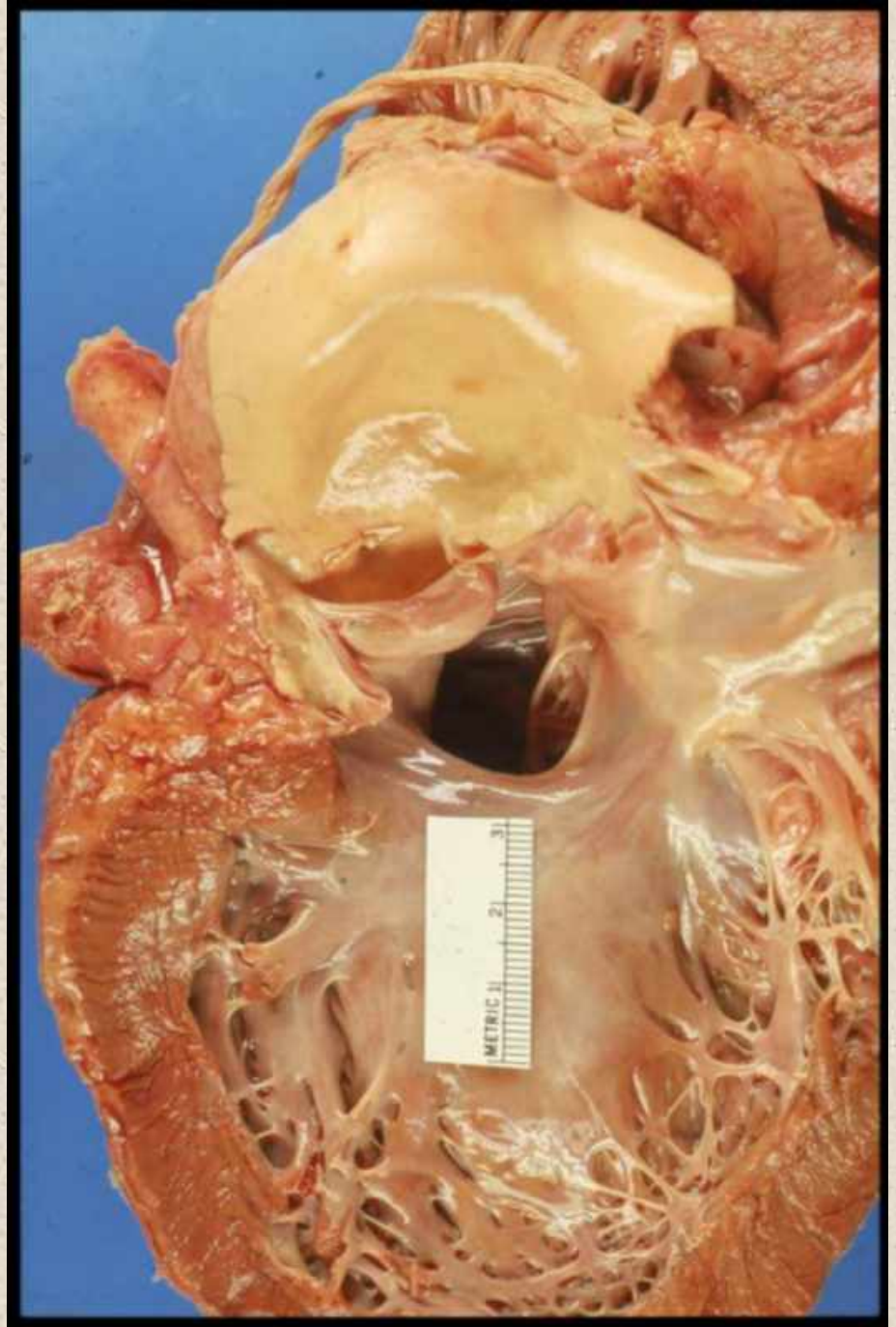
***Moderately
Restrictive
VSD
Age 46***



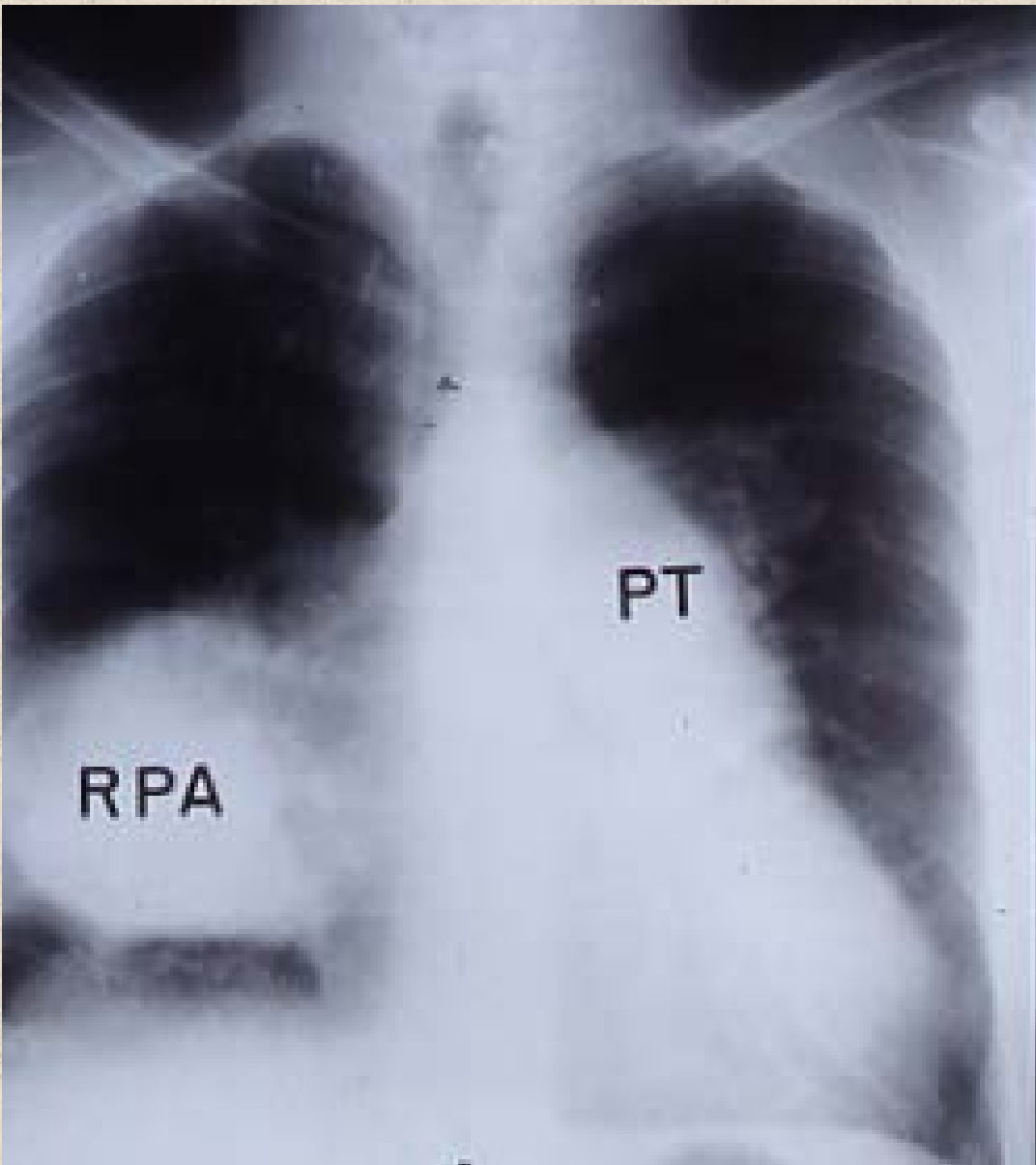
Eisenmenger VSD Age 43



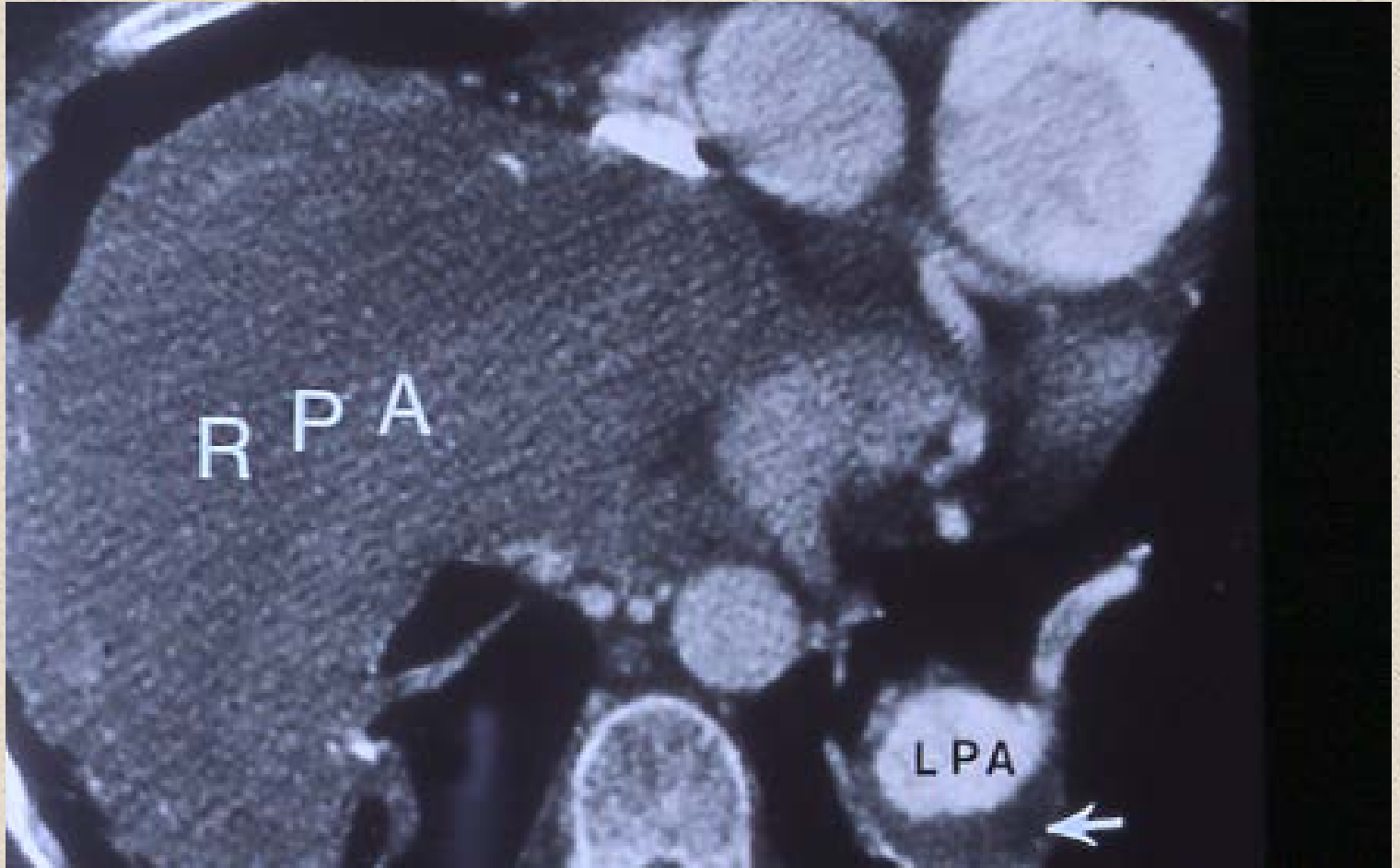
Eisenmenger
VSD
Age 54



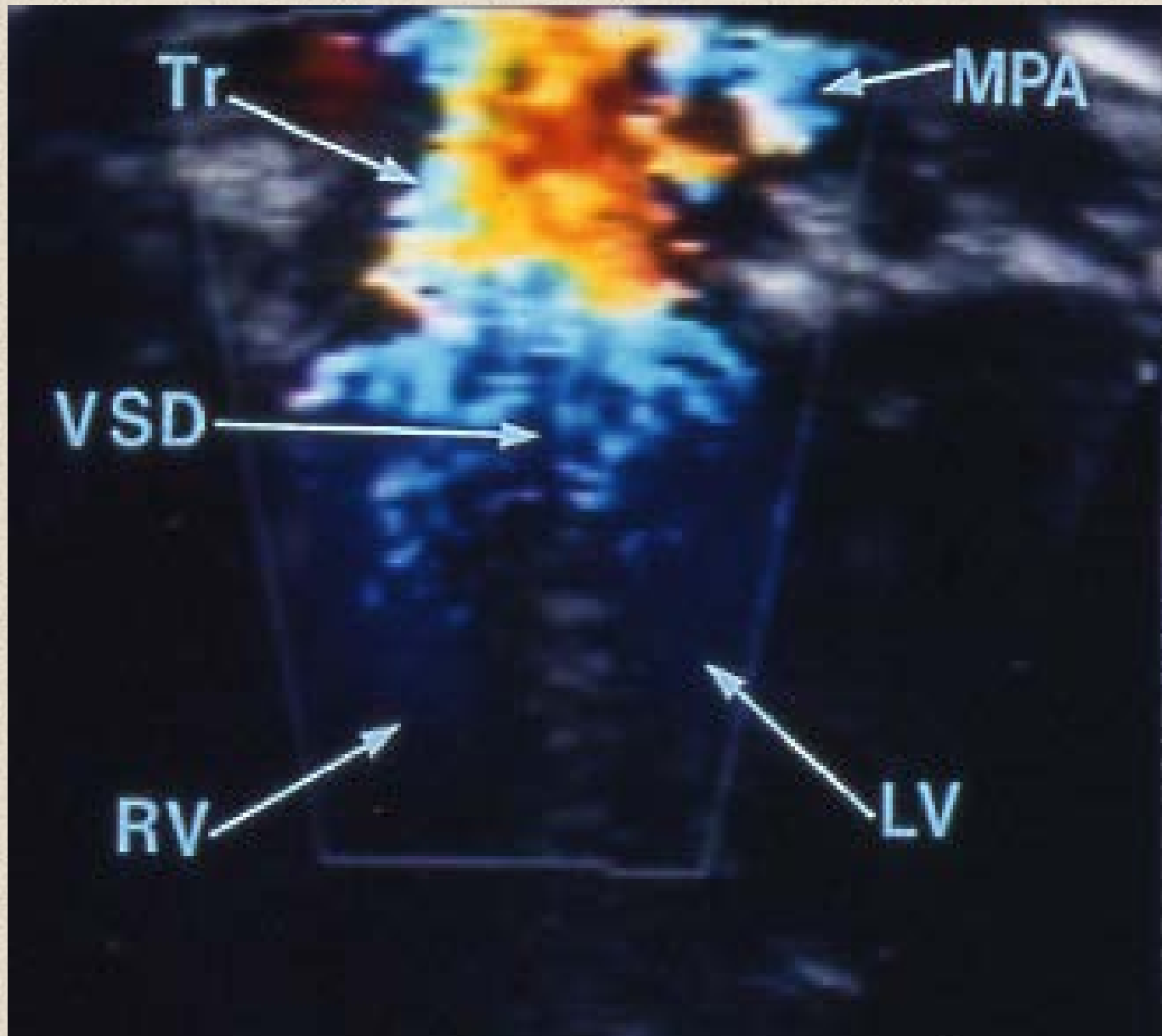
Eisenmenger VSD Pulmonary Aneurysms
Age 54



*Thrombosis in Dilated Hypertensive
Proximal Pulmonary Arteries.
A Therapeutic Dilemma*



Aortopulmonary Window Age 57



Aortopulmonary Window

Age 61

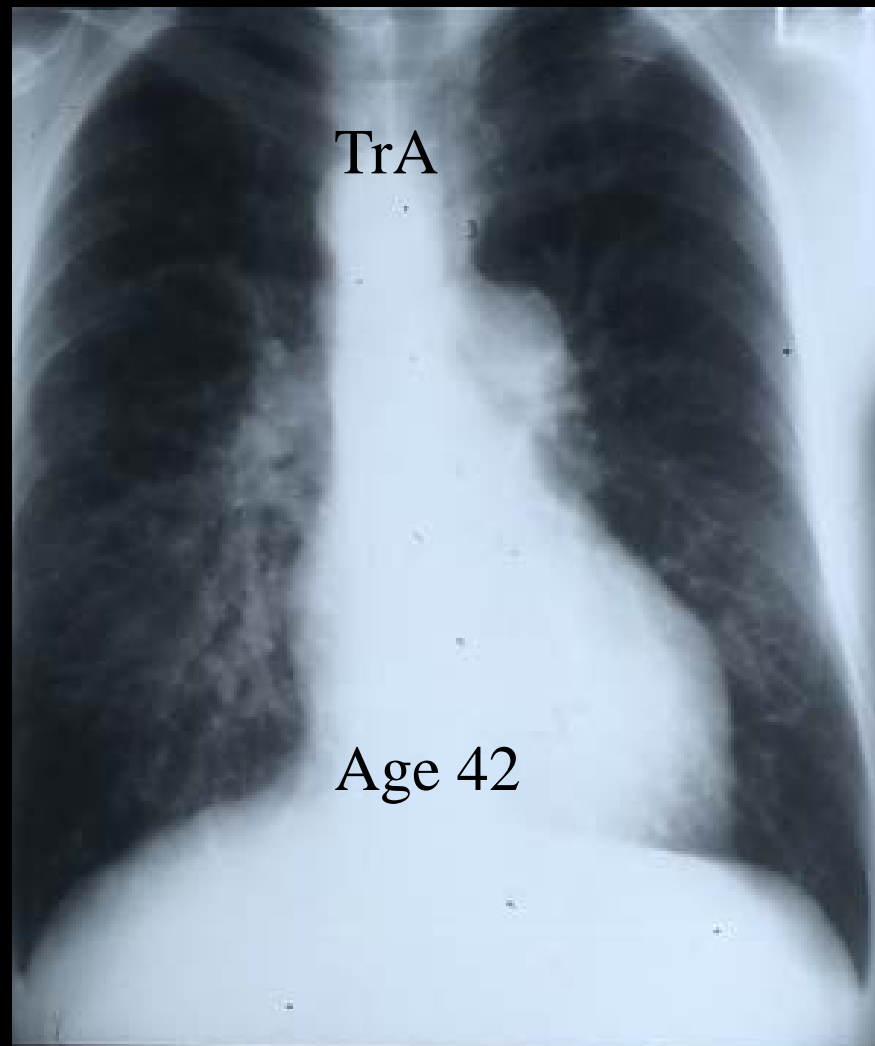
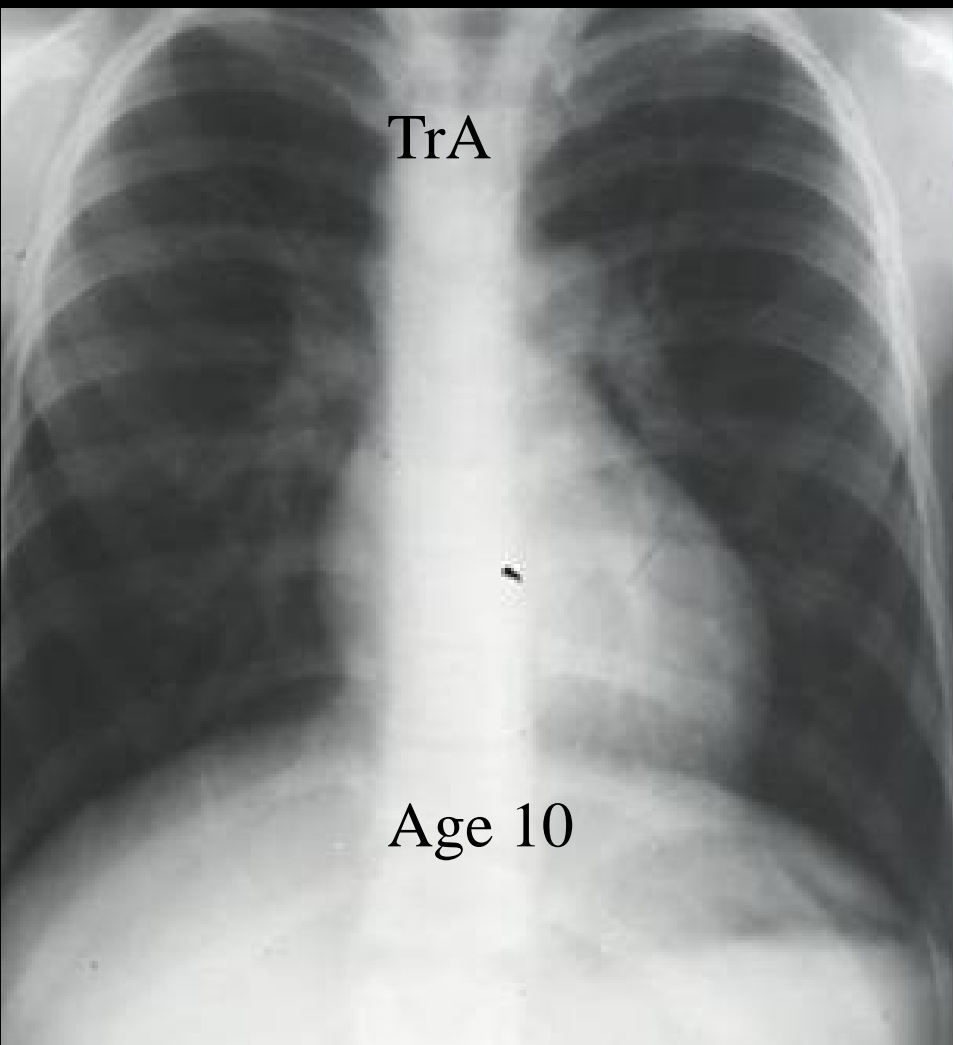


A-P Window

The Rarest of the Rare

**Uncommon Defects with
Unexpected Survival and
Exceptional Longevity**

Truncus Arteriosus



Isolated Quadricuspid Aortic Valve



Age 56



Age 62

Intrapulmonary Hemorrhage Age 55

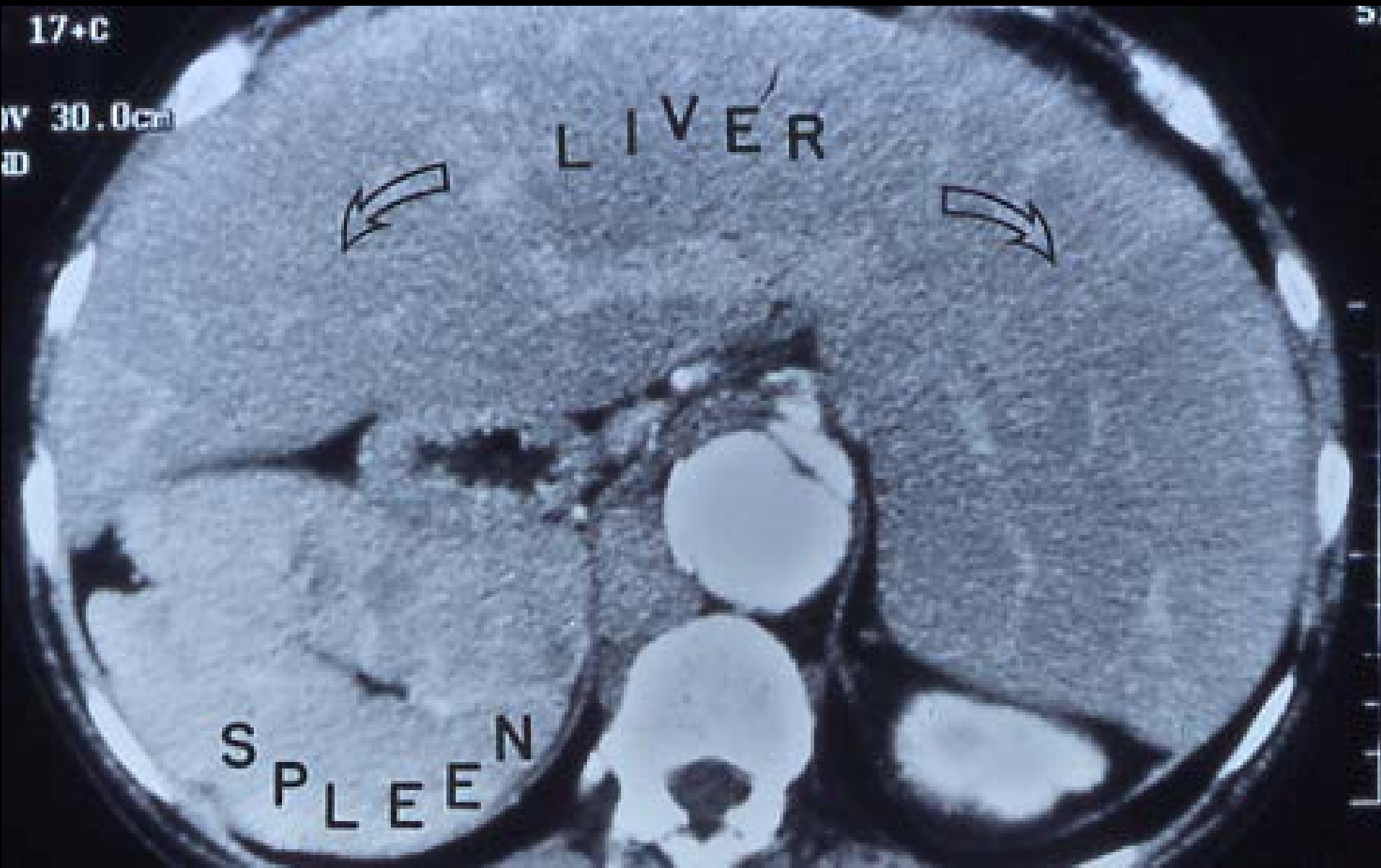
Intrapulmonary Hemorrhage Age 55



Truncus
Arteriosus
Age 54



Heterotaxy Left Isomerism



Left Isomerism

18 Months



Left Isomerism

Age 52



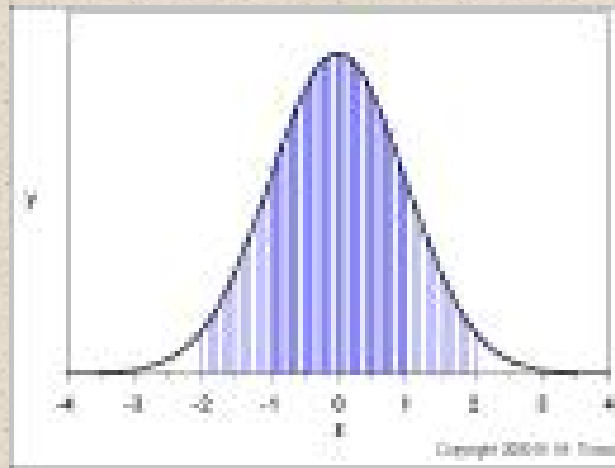
An Important Question

If as many as 90% of infants born with anomalous origin of the left coronary artery from the pulmonary trunk die in their first year, why did a patient of Maude Abbott's live to age 60 years? Why does the same disease express itself so differently?

Proteomics

Unlocking genomic information has been considered the key to understanding cellular and molecular mechanisms, but it is now clear that knowledge of the DNA sequence is essential but insufficient. A more meaningful understanding of gene expression can be achieved through characterization of the protein products of that expression---the ultimate biological determinants of disease phenotype. The term *proteome* was coined to describe the proteins encoded from a specific genome. The new discipline is called ***proteomics***.

Longevity In Unoperated Congenital Heart Disease



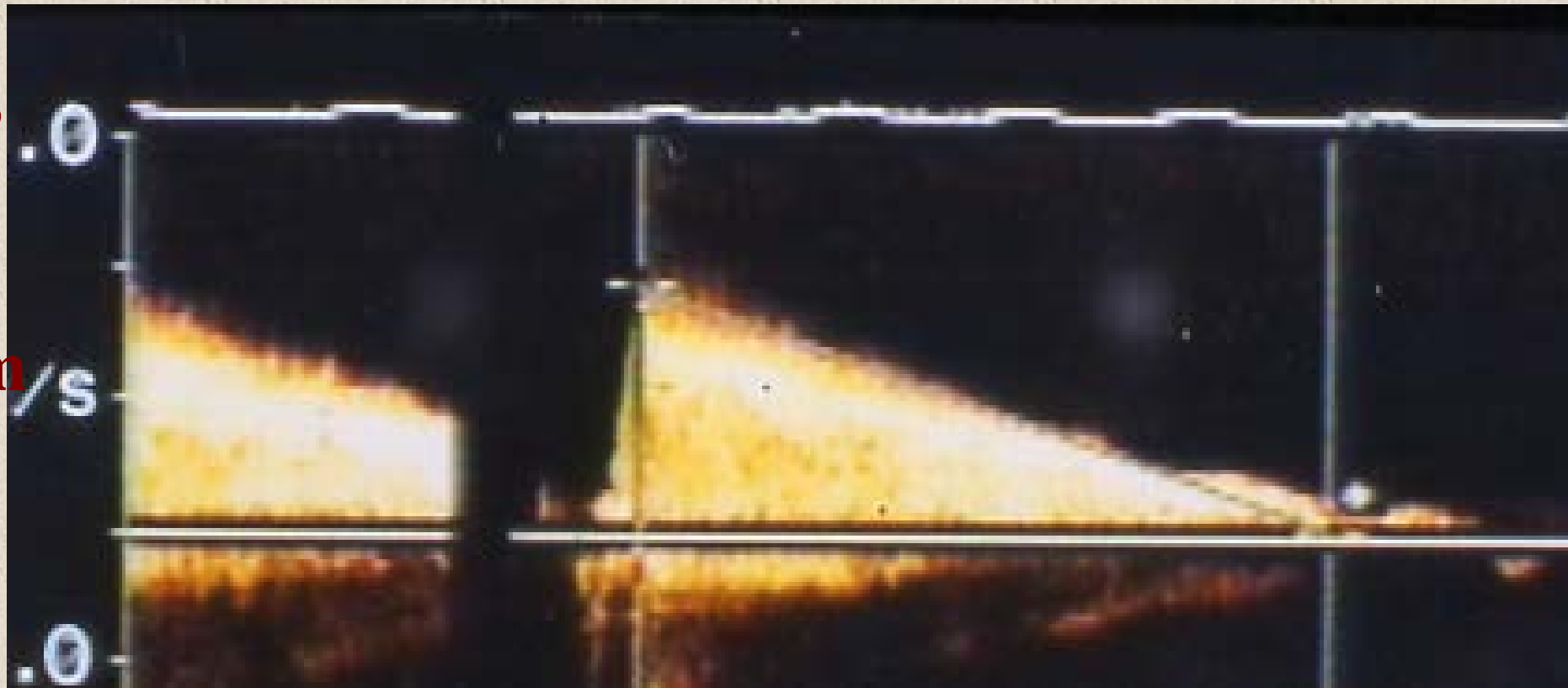
*Never Make Predictions,
Especially About the Future*

Mitral Stenosis

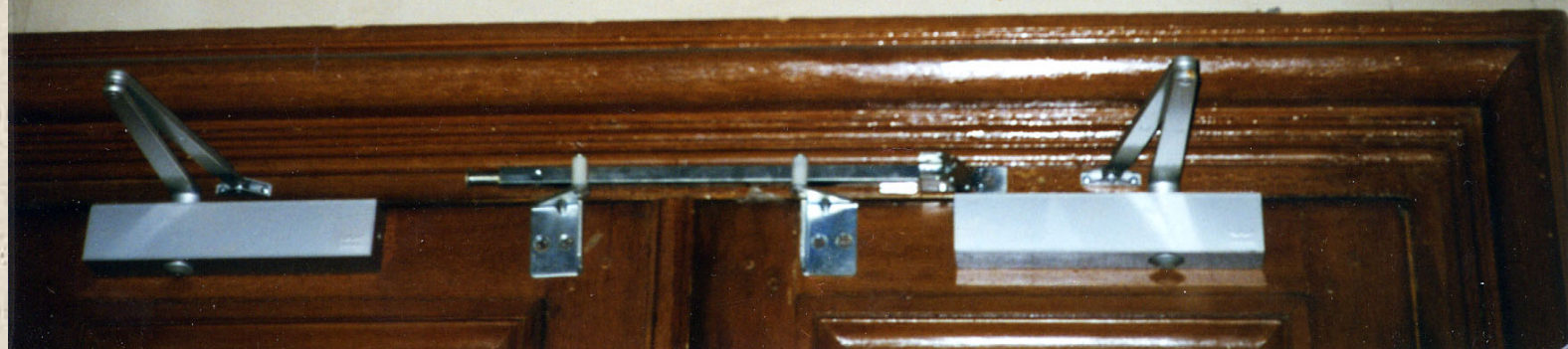
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SALLE HENRI ROGER



NATURAL HISTORY



Uncommon Defects with Expected Adult Survival But Exceptional Longevity

The natural history of any disease is a description of what happens to people with that disease who do not receive treatment for it.

Julien I. E. Hoffman

“Natural” History

- **Pharmacologic therapeutics are not natural.**
- **Anticoagulants are not natural.**
- **Pacemakers are not natural.**
- **Electrical cardioversion is not natural.**
- **Electrical defibrillation is not natural.**
- **Radiofrequency ablation is not natural.**
- **So – *naturally, natural history* is inappropriate in contemporary medicine.**

Categories

1. *Unoperated* – Unrecognized or inoperable. Longevity improved by medical management.
2. *Postoperative* – Improved longevity after operation is not due to operation alone.