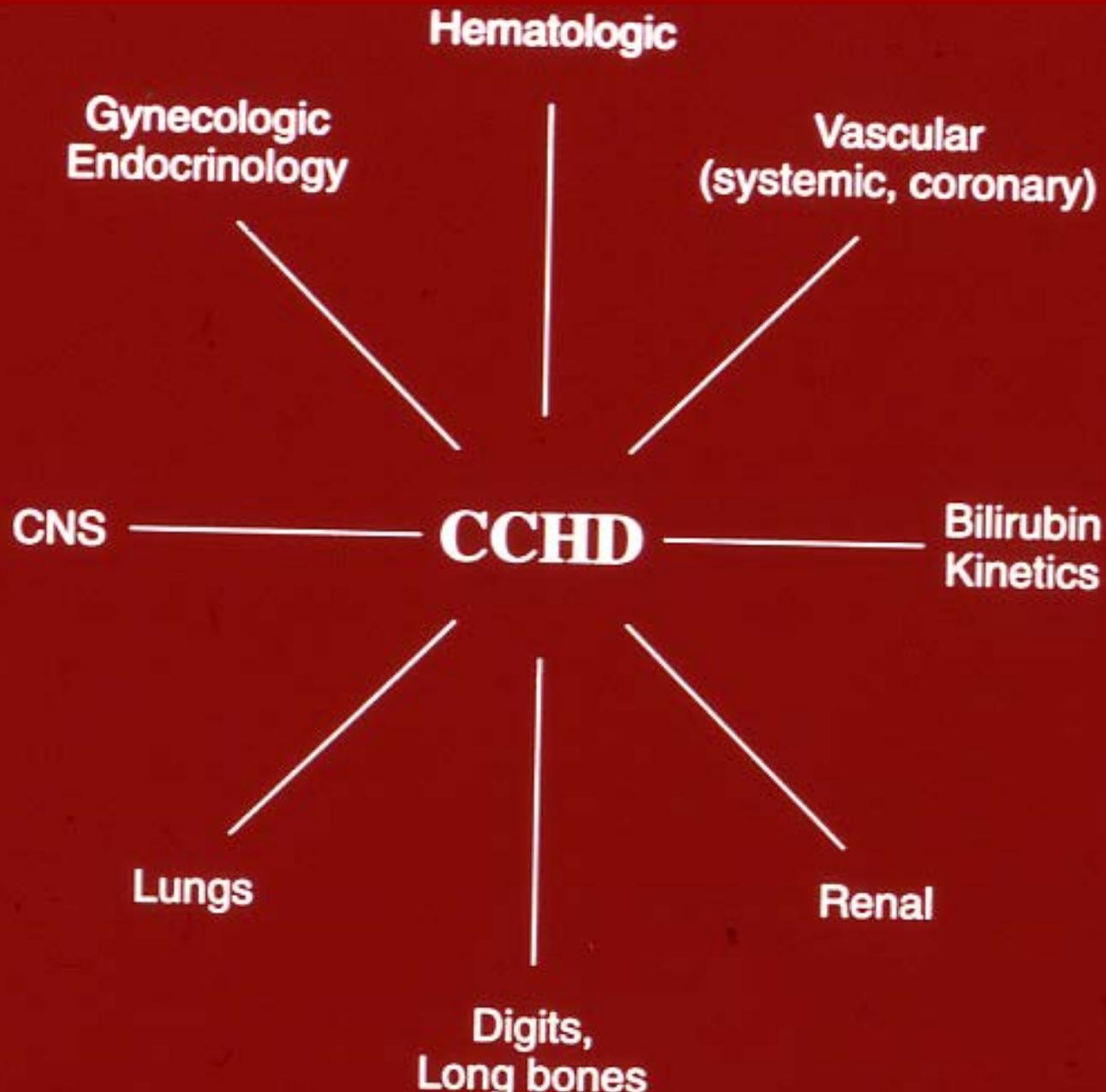


# **Medical Management of Adult Congenital Heart Disease**

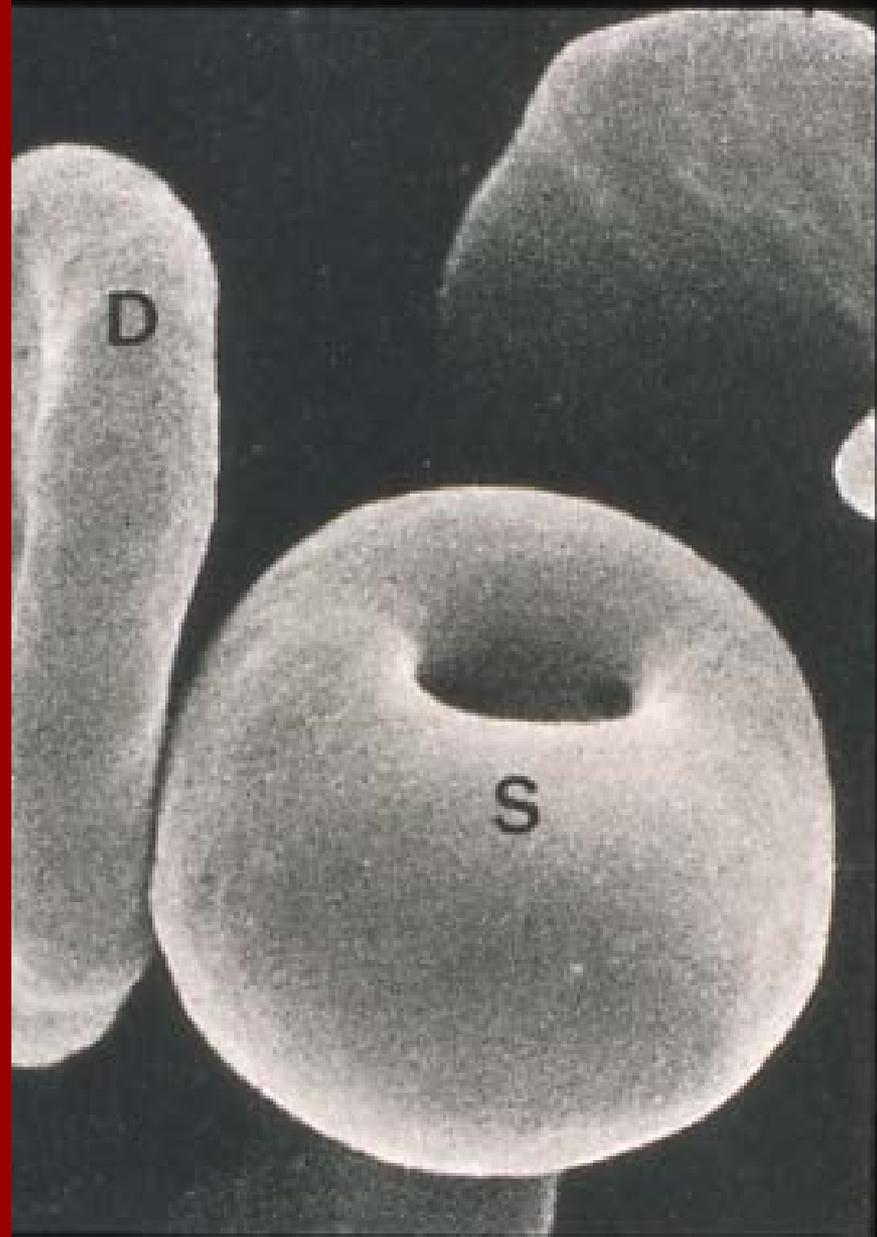
- 1. Erythrocytosis**
- 2. Pulmonary Hemorrhage**
- 3. The Thrombosis Dilemma**
- 4. Non-Cardiac Surgery:**
  - a) The anesthesiologist**
  - b) Bilirubin kinetics**
  - c) Post operative acute gouty arthritis**
- 5. Gynecologic Endocrinology**
- 6. Abnormalities of great arterial walls**

# A Multi-System Systemic Disorder



# Blood Letting





# **Cyanotic Congenital Heart Disease**

**Erythrocytosis is a physiologically appropriate response to the decrease in tissue oxygenation caused by arterial hypoxemia. Decreased tissue oxygenation stimulates renal release of erythropoietin.**

**Phlebotomy reduces red blood cell mass, reduces delivery of oxygen to metabolizing tissues, and stimulates further release of erythropoietin.**

**Deformable biconcave discs become nondeformable iron deficient microspherocytes, thus increasing whole blood viscosity.**

# The Virtues of Erythrocytosis

**The viscous erythrocytotic perfusate increases endothelial shear stress**

**The increase in shear stress causes elaboration of endothelial vasodilator substances**

**Vasodilatation augments flow to metabolizing tissues**

**NO in red blood cells enhances transfer of O<sub>2</sub> from hemoglobin to tissues**

**The transfer is enhanced by the increased number of red blood cells**

**Erythrocytosis in an iron replete state is not a risk factor for stroke due to cerebral arterial thrombotic occlusion.**

**Iron deficient erythrocytosis is a risk factor for *venous* thrombotic stroke in infants but not in older children and adults.**



# **Recommendations for Phlebotomy in an Iron Replete State**

- 1. Not based on hematocrit (automated electronic particle counter) irrespective of level.**
- 2. Recommended for temporary relief of intrusive hyperviscosity symptoms.**
- 3. Minimum phlebotomy that achieves symptomatic relief, generally one unit with isovolumetric saline replacement.**
- 4. Hydroxyurea blunts the erythropoietin-induced rebound.**

# Preoperative Phlebotomy

Whole blood is removed isovolumetrically in daily amounts of 500ml to reduce the hematocrit to just below 65%. Within hours after phlebotomy, platelet counts increase, and platelet aggregation and hemostasis improve.

**“The temptation to use the anticoagulant drugs may be great. On the basis of the present studies, their use would appear to be fraught with danger.”**

Robert C. Hartmann  
Bull. Johns Hopkins  
Hospital 1952

## **Abnormal Hemostasis in CCHD**

Intrinsic hemostatic defect(s)

Increased tissue vascularity

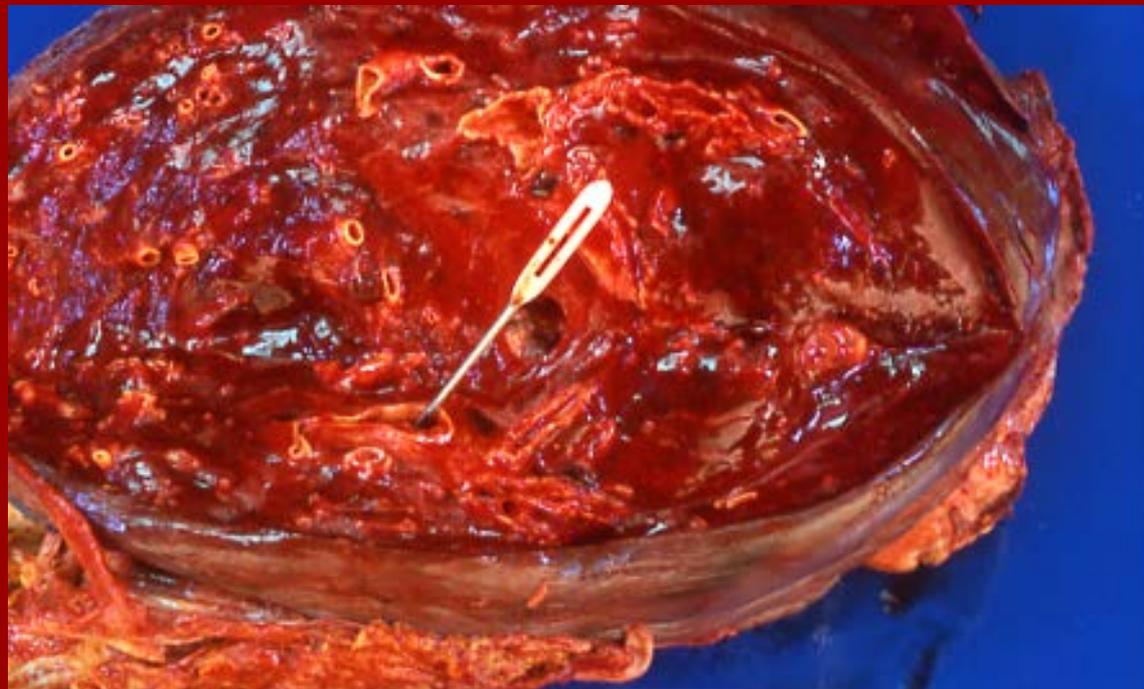
# Pulmonary Hemorrhage in Eisenmenger Syndrome

External Hemorrhage:  
Hemoptysis



**Internal  
Hemorrhage:  
Intrapulmonary**

**A common cause  
of sudden death**



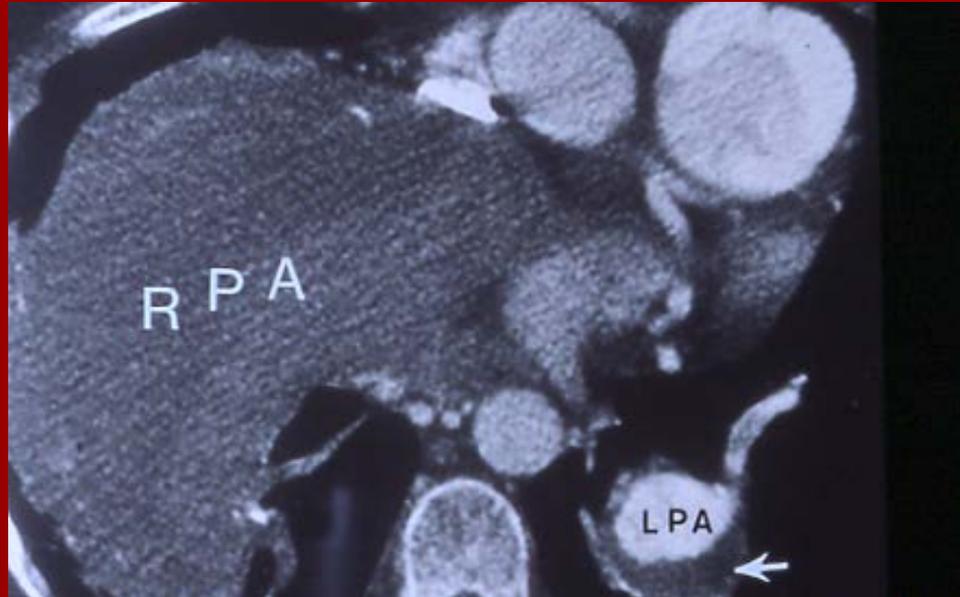
# Hemoptysis in Eisenmenger Syndrome

- 1. Do not bronchoscope.*
- 2. Determine if there is a history of antiplatelet or anti-inflammatory agents**
- 3. Chest x-ray for infiltrates of intrapulmonary hemorrhage.**
- 4. CT scan if infiltrates are present.**
- 5. Hospitalize for all but mild or moderate intrapulmonary hemorrhage.**

# Treatment of Pulmonary Hemorrhage in Eisenmenger Syndrome

1. Thrombocytopenia – platelet transfusion vs phlebotomy
2. Platelet Counts in Normal Range
  - a) Fresh frozen plasma
  - b) Cryoprecipitate
  - c) Human factor VIII – safe but efficacy unproven
  - d) Desmopressin (DDAVP), synthetic analog of vasopressin – neither safety nor efficacy has been proven
3. Excessively low HCT -- transfuse

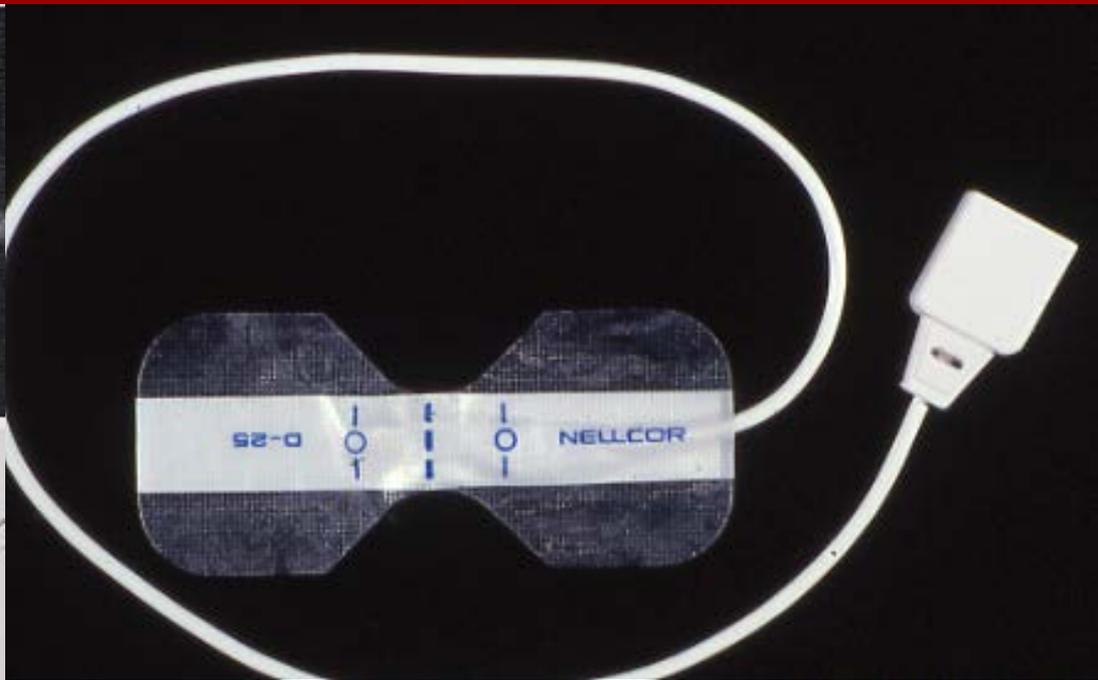
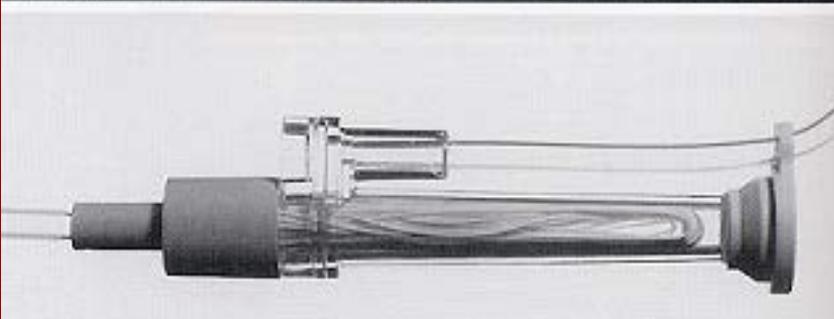
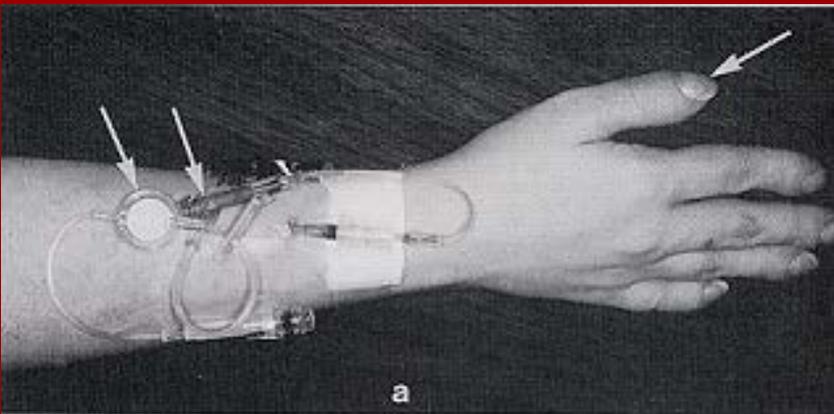
# **Thrombosis in Dilated Hypertensive Proximal Pulmonary Arteries. A Therapeutic Dilemma**



- 1. Anticoagulants – Efficacy is nil. The risk of reinforcing intrinsic hemostatic defect(s) and provoking hemorrhage is high.**
- 2. Thrombolytic Agents – The efficacy of even intrapulmonary administration is nil.**

# ***NONCARDIAC SURGERY IN ADULTS WITH CONGENITAL HEART DISEASE***

**The pivotal role of the  
cardiac anesthesiologist**



# Bilirubin Kinetics

- Bilirubin is formed from the breakdown of heme, a process that is excessive in the presence of the erythrocytosis of cyanotic congenital heart disease and that coincides with a substantial increase in the amount of unconjugated bilirubin.



935-10329



# Acute Gouty Arthritis



## Prophylaxis After Resolution

**Low dose oral colchicine is recommended because non-steroidal anti-inflammatory agents, even low doses, reinforce the hemostatic defects in CCHD.**

# CCHD

## Gynecologic Endocrinology



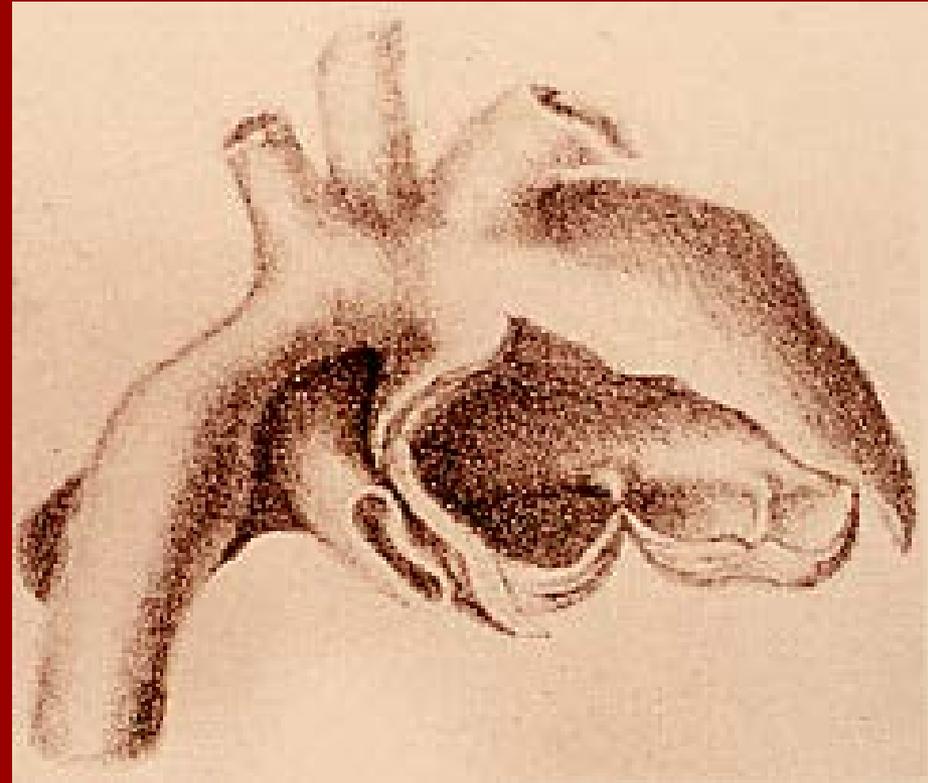
# Females with Cyanotic Heart Disease

Dysfunctional bleeding may reflect an anovulatory state. Chronic unopposed estrogen production due to anovulation leads to continuous uterine stimulation and increases the risk of endometrial hyperplasia and adenocarcinoma.

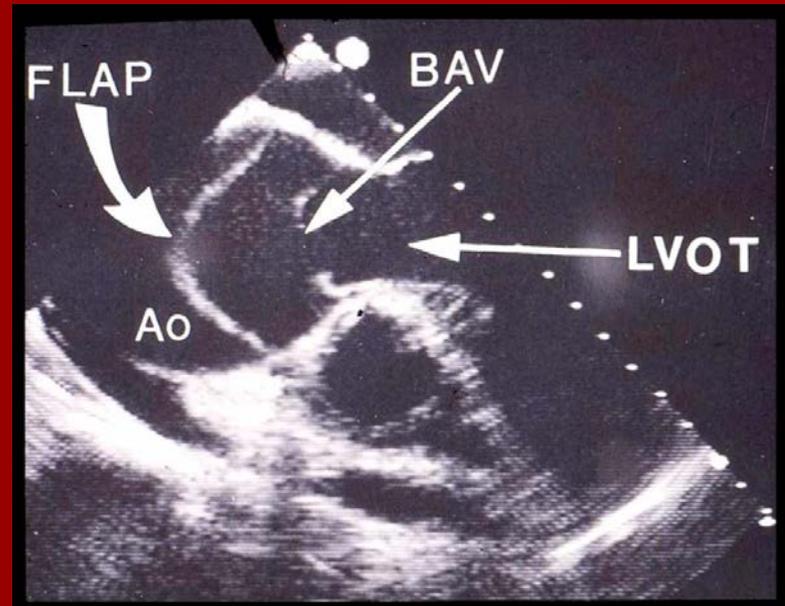
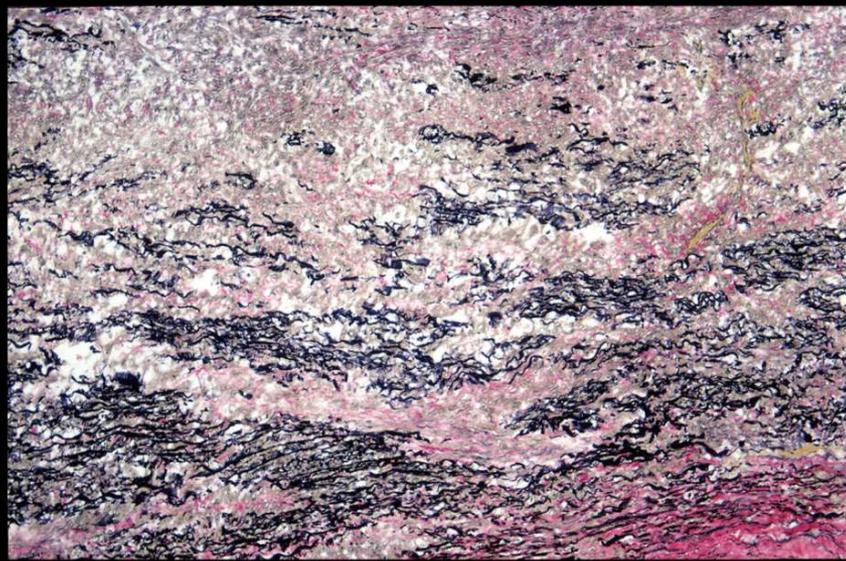
# **Abnormalities of Great Arterial Walls in Congenital Heart Disease**

**“The presence of a bicuspid aortic valve appears to indicate, at least in a portion of the cases in which it occurs, a tendency for spontaneous rupture.”**

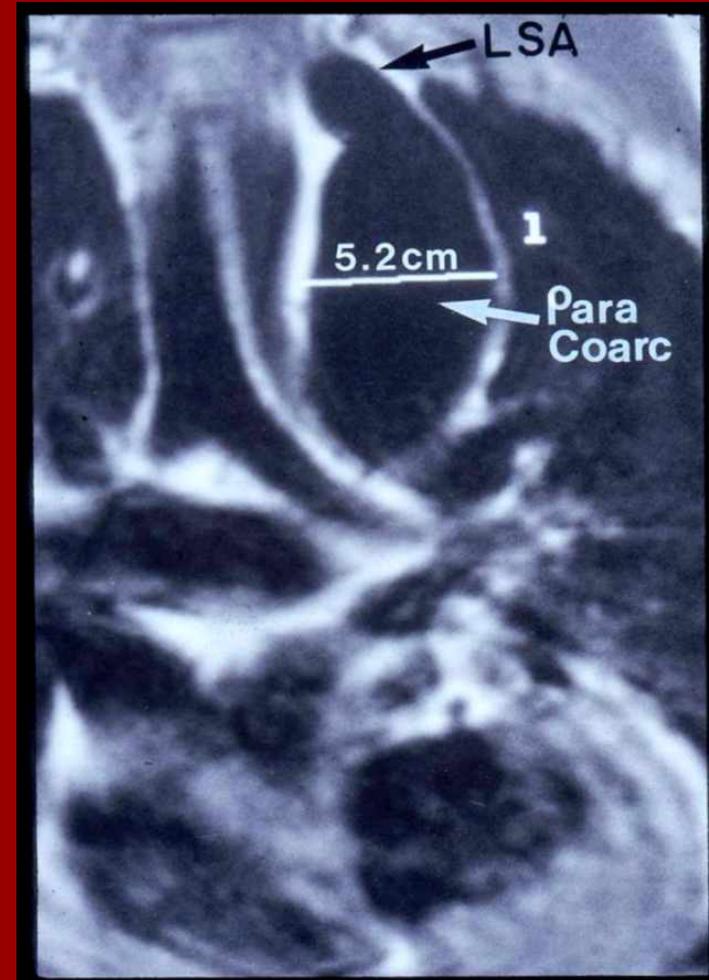
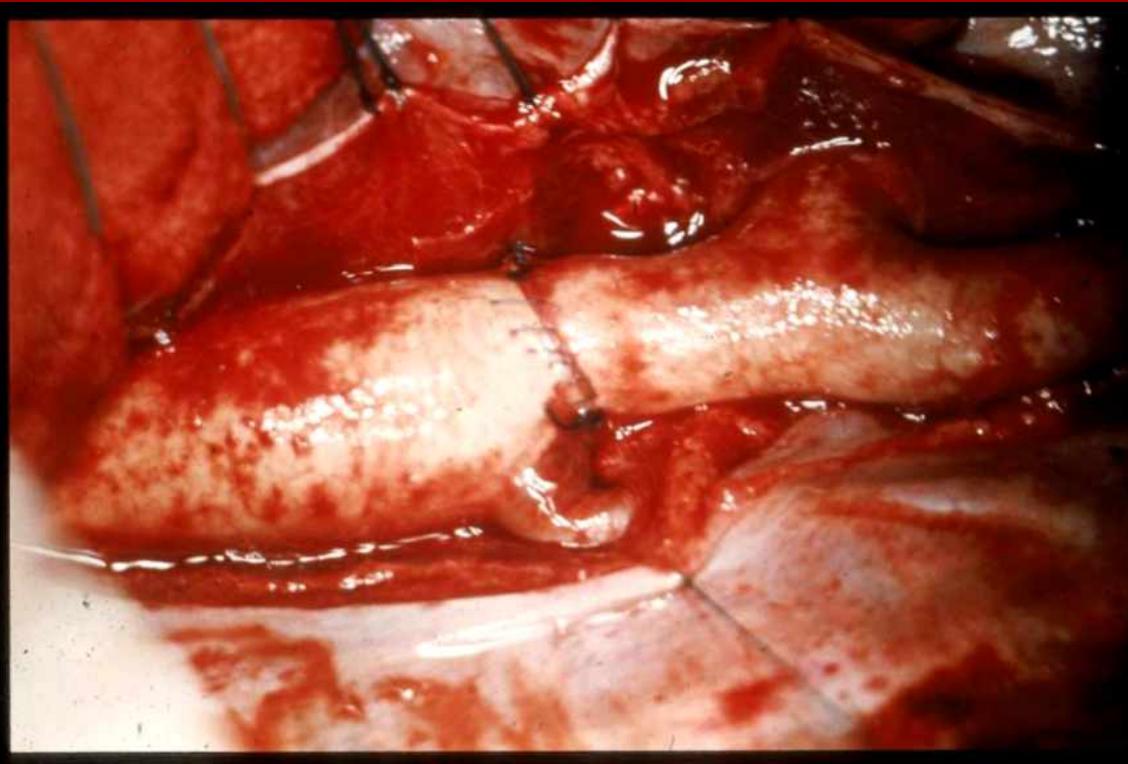
**Maude Abbott 1928**



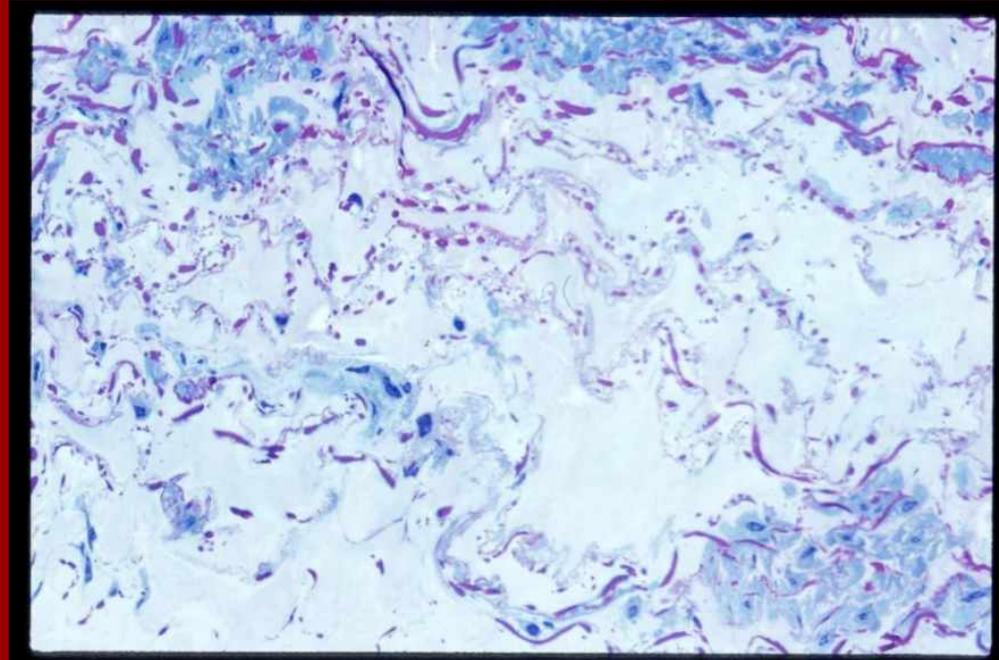
# Bicuspid Aortic Valve



# Coarctation of the Aorta



# Fallot's Tetralogy



# Fallop's Tetralogy

## Aortic Root & AR

