

Surgical Management of Adult Congenital Heart Disease

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Disclosures

No financial or
professional
disclosures



STAFF PHOTO BY TED FITZGERALD

SOUNDS MUCH BETTER: Andrea Ulloa Hallo, left, is checked over by cardiothoracic surgeon

Scope of Congenital Heart Defects

- Most common type of birth defect: global: 34 *million* people affected
- Most lethal form of birth defect: global: 323,000 deaths/year
- Incidence: 8/1000 births; approx. 50 % will require intervention
- Most cases: cause unknown
- Maternal factors: Obesity, rubella infection, alcohol, tobacco, radiation exposure
- Genetics: Ten % of offspring will have a congenital heart defect

CONGENITAL HEART DISEASE (CHD): KEY FACTS

1. PEOPLE IN THE UNITED STATES WITH CHD



≈ **2.4 MILLION**
people (children + adults)

- ~1.4 million adults
- ~1.0 million children

Source: CDC (2023)

2. ADULTS IN THE US WITH CHD



≈ **1.4–1.5 MILLION**
adults

*Adults now outnumber
children with CHD*

Source: CDC (2023)



3. SURVIVAL RATE IN THE US



>90%
of children with CHD
survive into adulthood

Source: AAP (2021)

4. SURVIVAL RATE GLOBALLY



- High-income countries:
~85–90% survival to adulthood
- Low-/middle-income countries:
substantially lower
(often <50–70% for complex CHD)

*Survival is highly dependent on access to
surgery, early diagnosis, and lifelong care.*

Sources: GBD 2017; peer-reviewed literature

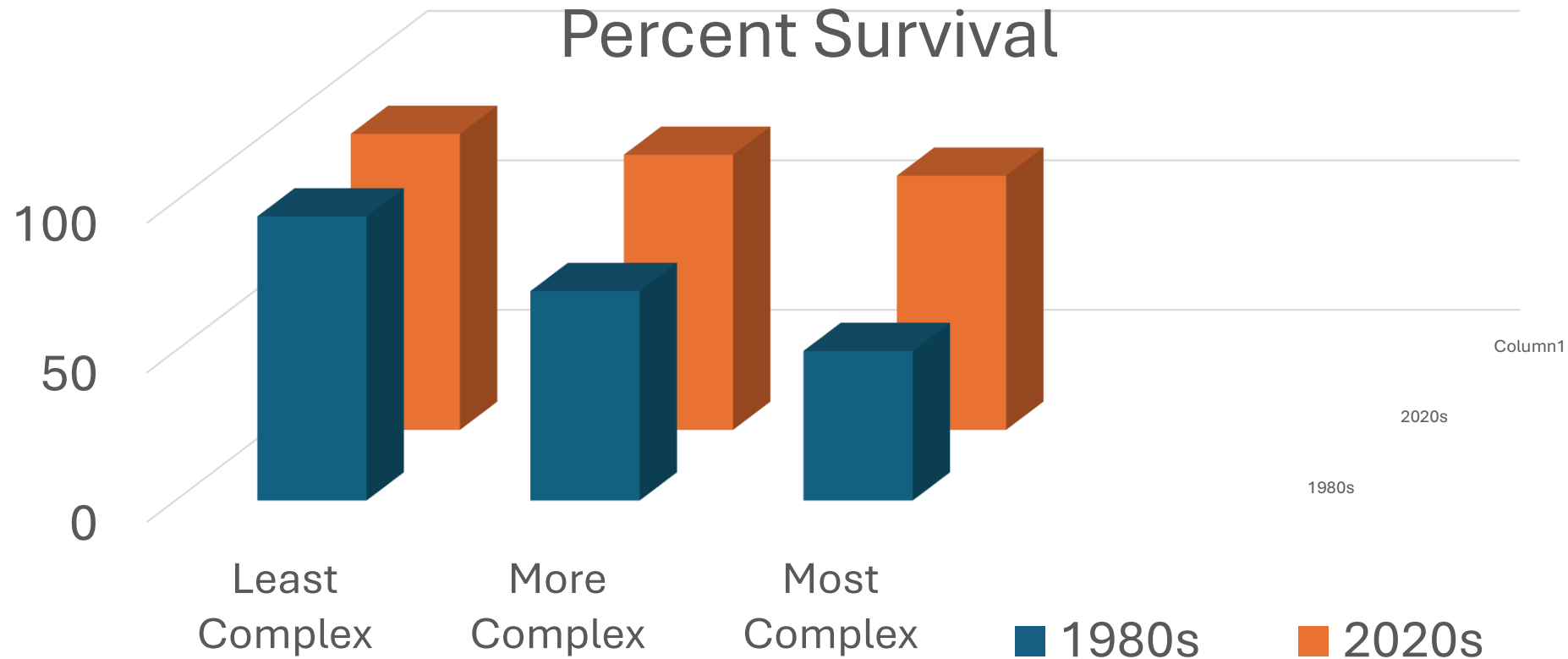


KEY TAKEAWAY:

More than 2 million people in the US live with CHD. Thanks to advances in care, >90% of children with CHD now survive to adulthood. CHD is now a lifelong, chronic condition requiring specialized care across the lifespan.

Note: Numbers are estimates and vary by data source and definition.

Childhood Survival of Congenital Heart Defects



Clinical Dilemmas in ACHD

Complication	Structural and Physiologic Etiology
Congestive Heart Failure	Systemic ventricle: Right vs Left GDMT trials excluded CHD patients
Dysrhythmias	WPW: Ebstein Anomaly; SVT: atrial suture lines; VT: Tetralogy of Fallot
Cyanosis	Right to Left shunt; Eisenmenger Syn; Pulmonary AVM; Parallel circuits: TGA; Complete Mixing: TAPVC
Thromboemboli, CVA	Hyper viscosity, Paradoxical Embolus
Endocarditis/Endarteritis	Turbulent Flow: Restrictive VSD, PDA
Aortopathy	Bicuspid Aortic Valve; Coarctation of the Aorta; Tetralogy of Fallot; Hypoplastic Left Heart Syndrome; Aortic root s/p Ross procedure
Hepatopathy, Liver cirrhosis; Protein Losing Enteropathy	Fontan pathway with elevated hepatic venous pressure

Genetic Syndromes

Diagnosis	Chromosomal Anomaly	Cardiac Defect	ExtraCardiac Anomalies
Down Syndrome	<i>Trisomy 21</i>	AtrioVentricular Septal Defects, Tetralogy of Fallot, VSD	Developmental Delay, Leukemia
Turner Syndrome	<i>Deletion X Chromosome</i>	Coarctation, Bicuspid Aortic Valve	Sterility, short stature, diabetes
Noonan Syndrome	<i>Autosomal Dominant</i>	Pulmonary Valve Stenosis, VSD	Webbed Neck, Short stature
William Syndrome	<i>Chromosome 7 Deletion</i>	Supravalvar Aortic Stenosis, Pulmonary artery stenoses	Elfin Facies, Developmental delay, hypercalcemia
DiGeorge Syndrome	<i>Chromosome 21 Deletion</i>	Interrupted aortic arch, Tetralogy of Fallot, Truncus arteriosus	Thymic aplasia, hypocalcemia, T Cell deficiency
Holt-Oram Syndrome	<i>Autosomal Dominant</i>	Atrial Septal Defect	Radial/Wrist Anomalies

Congenital Cardiac Defects

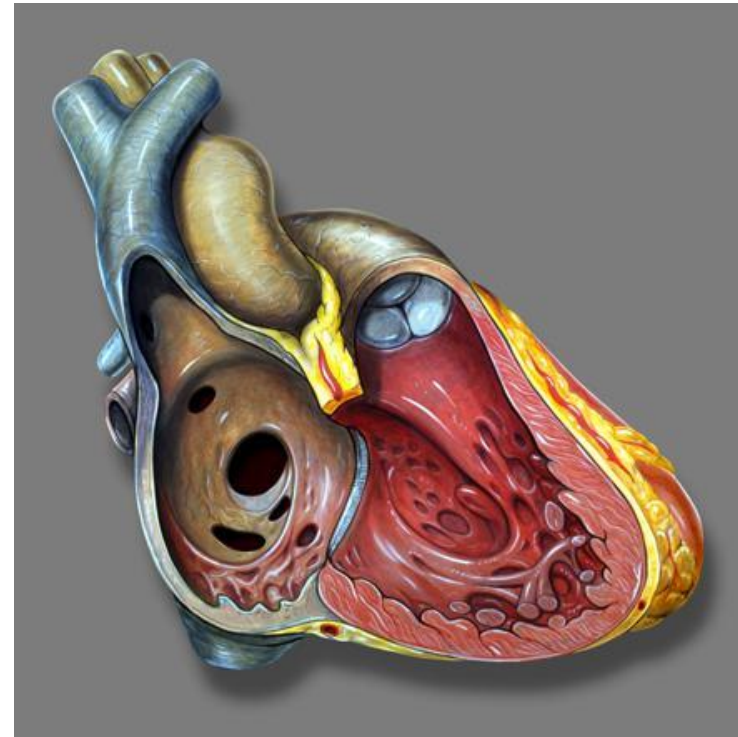
- **Left** to **Right** Shunts
- **Left** Sided Obstructive Lesions
- **Right** Sided Obstructive Lesions
- Single Ventricle Anatomy
- Complex Lesions

Left to Right Shunts

- Definition: Oxygenated blood from pulmonary venous return that is preferentially directed to the *Pulmonary Circuit (Lungs)*
- Anatomic Location: Atrial (ASD), Ventricular (VSD), and Great Vessel (PDA, Truncus Arteriosus, Aortopulmonary Window)
- Symptoms: Congestive heart Failure, Failure to thrive, Poor weight gain, Resp infections
- Magnitude of shunt dependent on
 - Size of communication
 - Relative vascular resistances (SVR vs PVR)
 - Relative diastolic compliances (LV vs RV)
 - Associated lesions

Atrial Septal Defects

- PFO
- Secundum
- Primum
 - AtrioVentricular Canal Defects
 - Trisomy 21
- Sinus Venosus
 - Partial Anom Pulmonary Venous Connection
- Common Atrium
- Coronary Sinus Septal



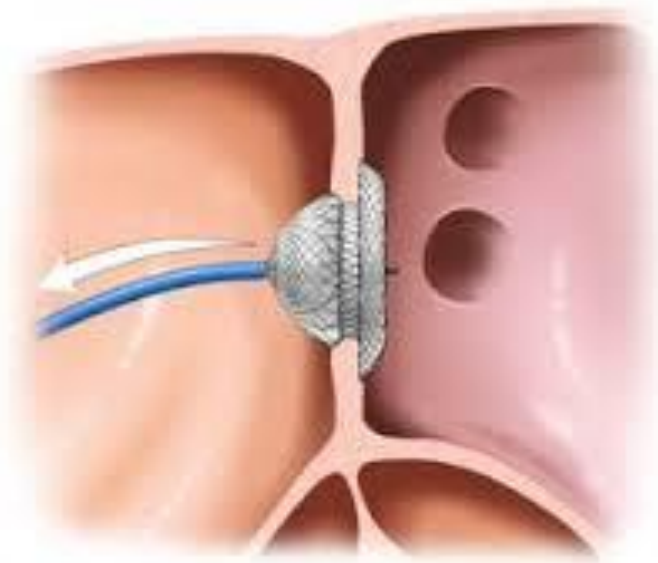
Physiology

- Excessive Pulmonary Artery Blood Flow
- Importance of Pulmonary Artery Pressures
- Development of Pulmonary Vascular Obstructive Disease (PVOD)
 - *Lesion Dependent*
 - *Age Dependent*
 - *Size of Shunt*
 - *Genetic Factors*

Symptoms of Atrial Septal Defect

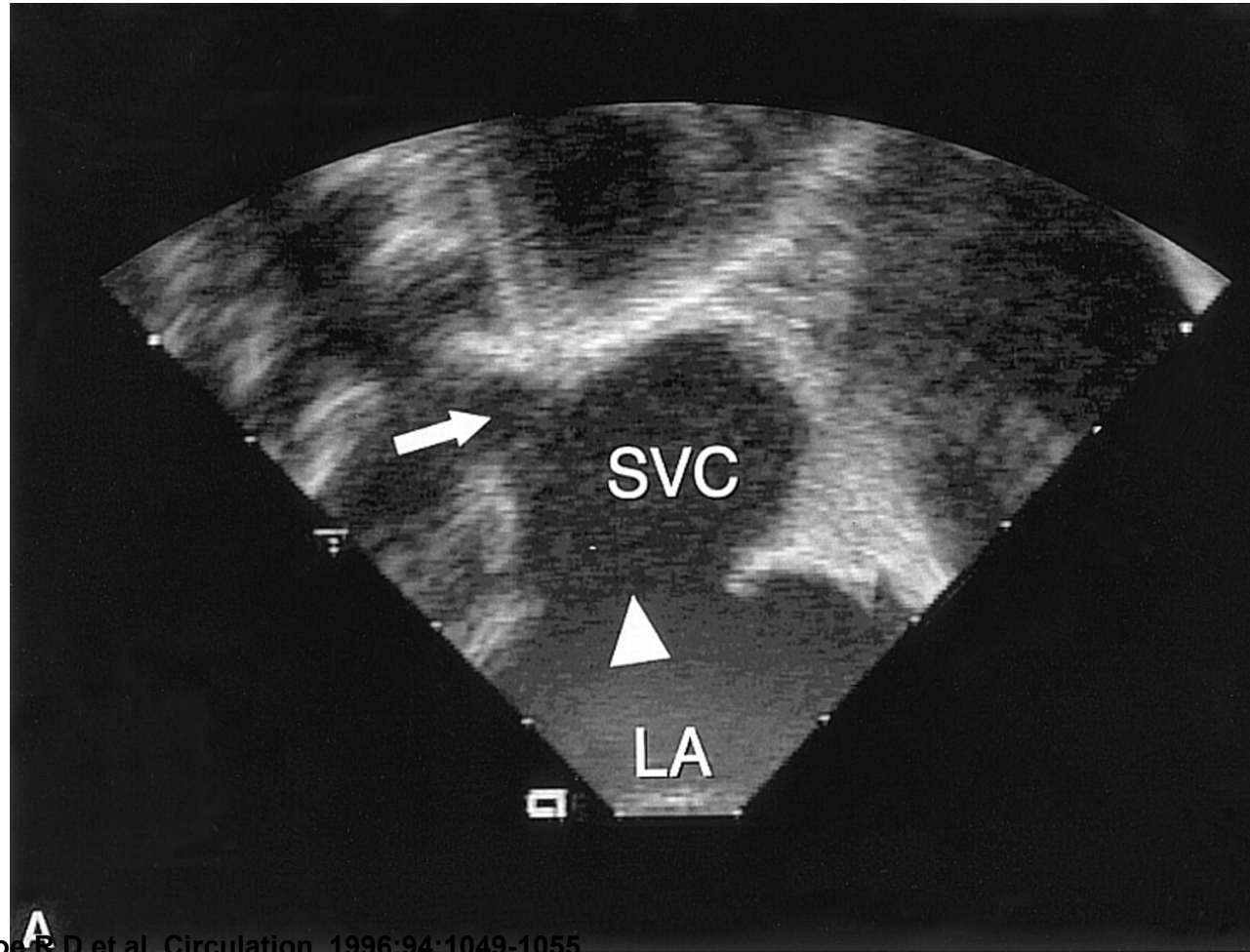
- *Exercise Intolerance*
 - Most common
 - Onset in Toddlers
 - Dyspnea and Fatigue with Exertion
- *Palpitations*
 - Atrial and Ventricular Arrhythmias
- *Paradoxical Emboli*
 - Right to Left Shunt
 - Stroke

Atrial Septal Defect Secundum



- Deficiency in Septum Primum
- Patent Foramen Ovale
- More common in females
- EKG: RBBB
- Systolic Murmur
- Repair in Childhood
- Percutaneous approach versus surgery

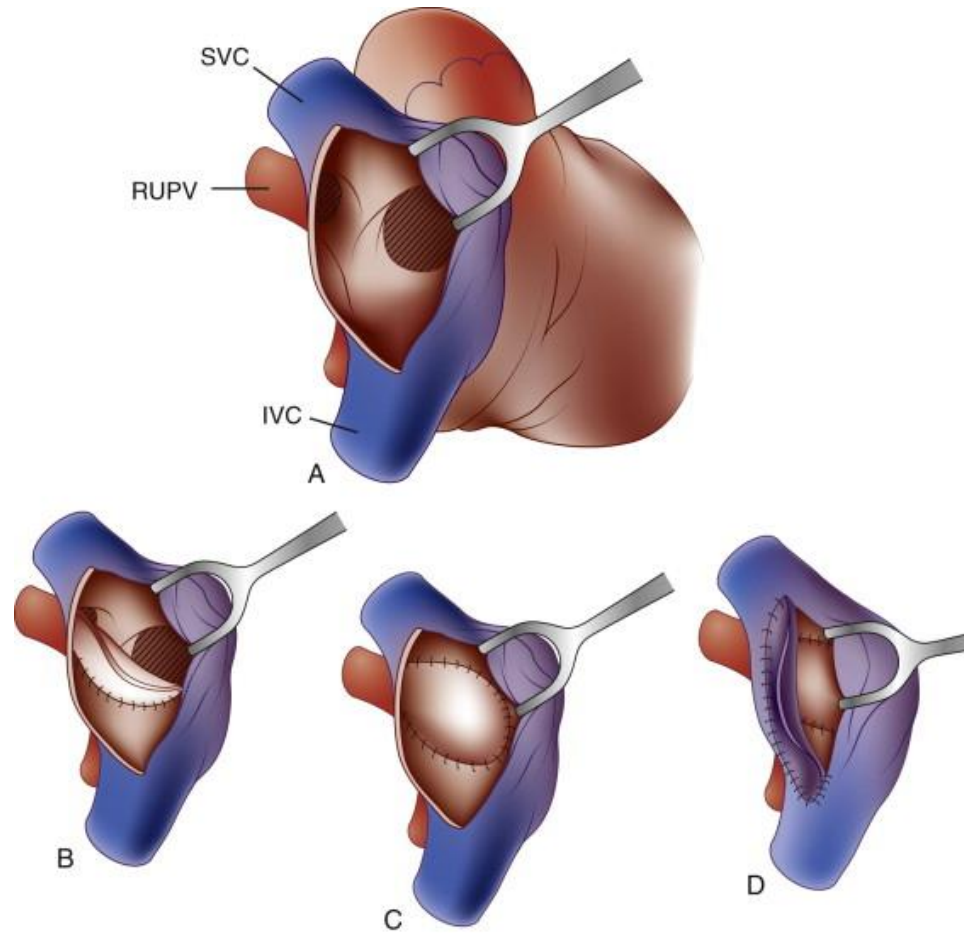
Sinus Venosus ASD



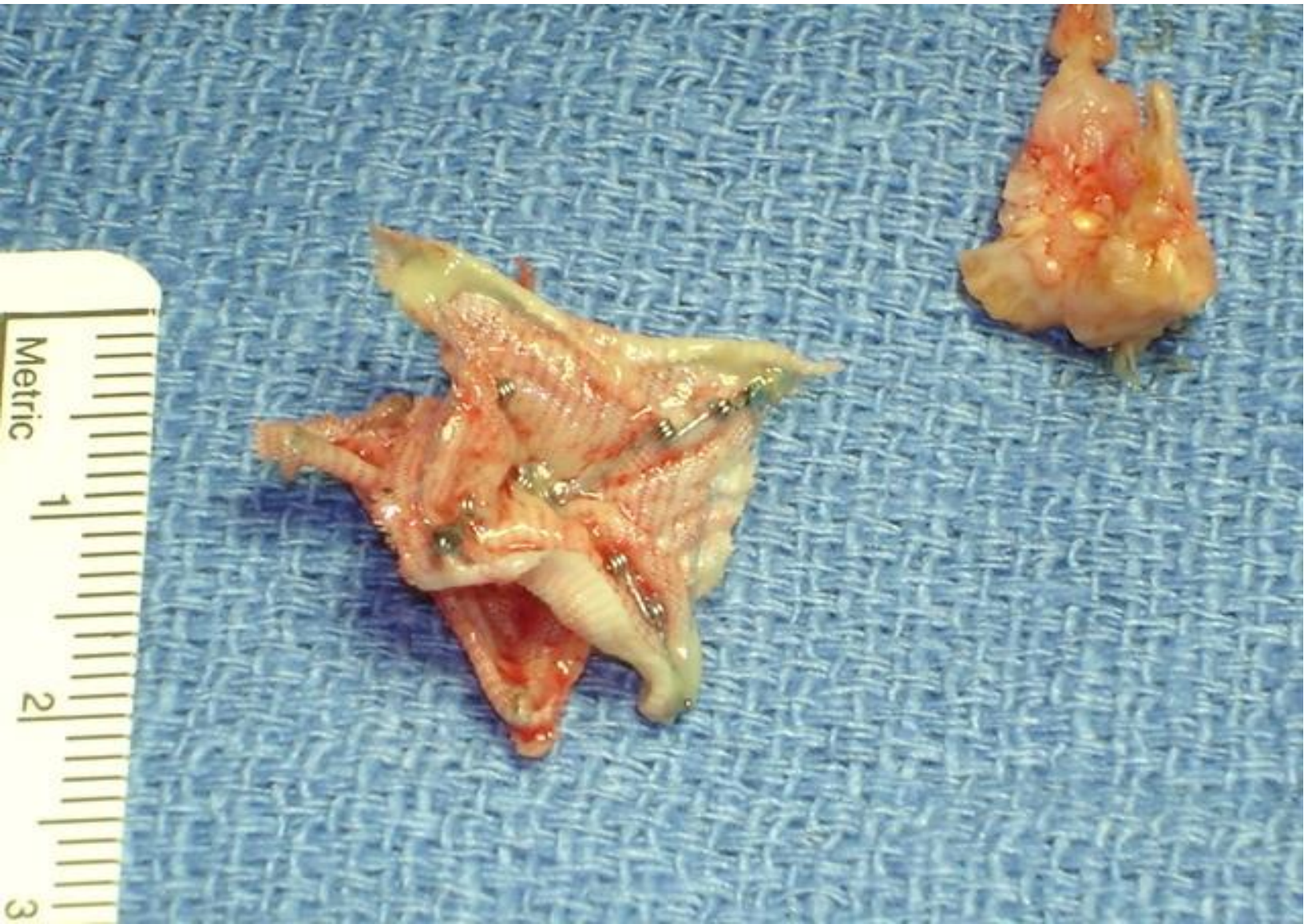
Pascoe R D et al. *Circulation*. 1996;94:1049-1055

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Sinus Venosus Defect

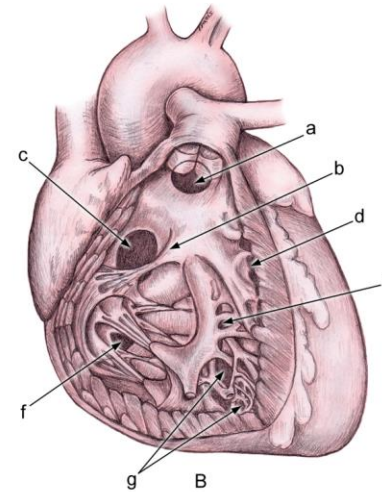
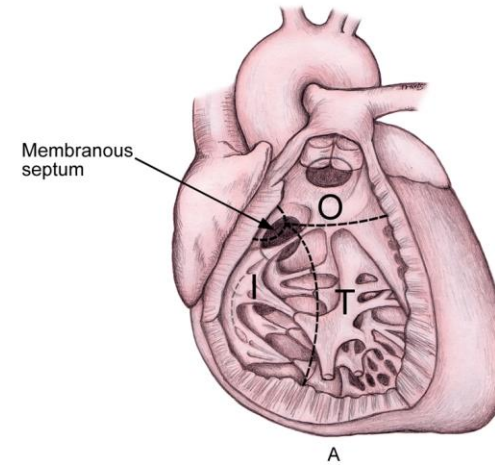


- Baffle Right UpperLobe PV to Left Atrium
- Same patch closes ASD
- Second patch to augment SVC



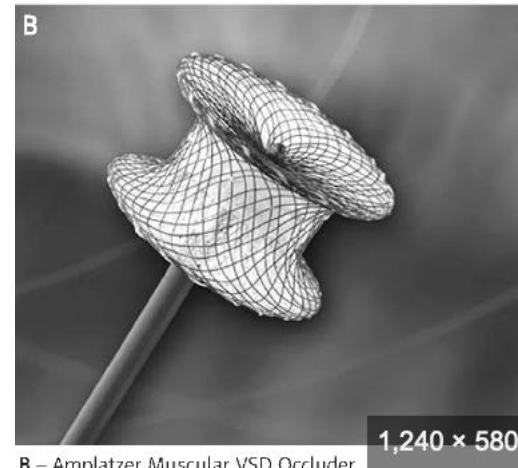
Ventricular Septal Defects

- Perimembranous
 - 80 %
 - Conventricular
- Inlet
 - AV Canal Defects
 - Trisomy 21
- SupraCristal
 - Asian Population
 - Aortic Valve Prolapse/Insuff
- Muscular
 - Single
 - Multiple: Swiss Cheese Septum
 - Types: Anterior, Mid, Posterior, Apical
 - Many will close spontaneousl



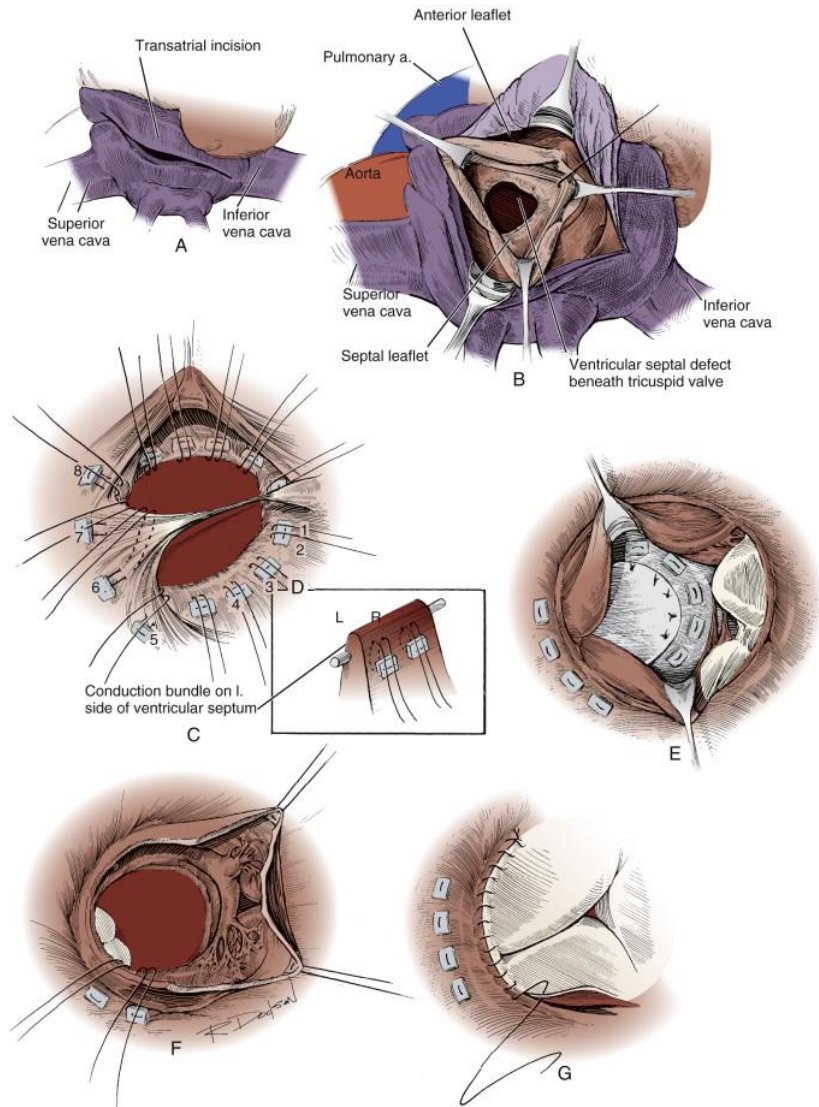
Percutaneous Closure of Muscular VSD

- Frequently performed in cardiac catheterization lab with Fluoroscopy and TEE guidance
- Occasionally performed in the operating room via direct puncture of right ventricle
- Primarily performed for muscular VSDs
- Made of Nitinol wire mesh



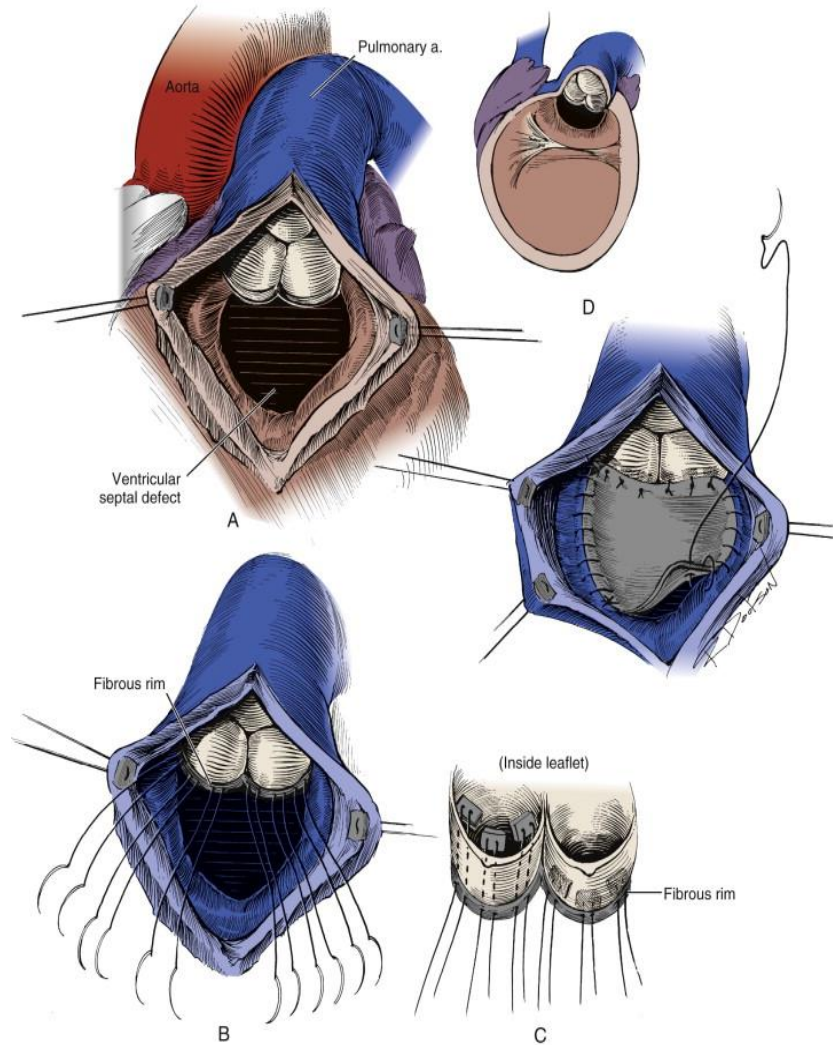
R – Amplatzer Muscular VSD Occluder

Surgical Closure of Perimembranous VSD



- Approach through right atrium
- Usually requires patch
- Either Running or Interrupted sutures
- Avoid Injury to:
 - Atrioventricular Node
 - Aortic Valve
 - Tricuspid Valve

Surgical Closure of SupraCristal VSD

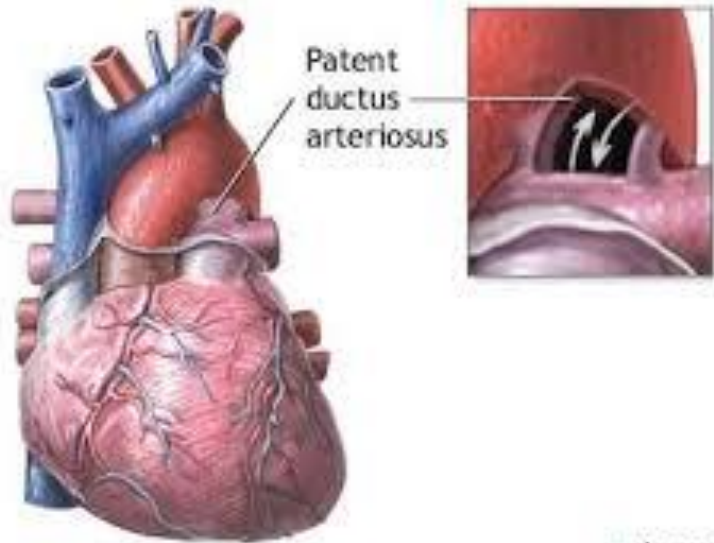


- Asian Predilection
- Aortic Valve Cusp will partially close the defect
- Approach through RV or MPA
- Always requires a patch
- Any degree of Aortic Insuff: operate
- May require aortic valve repair

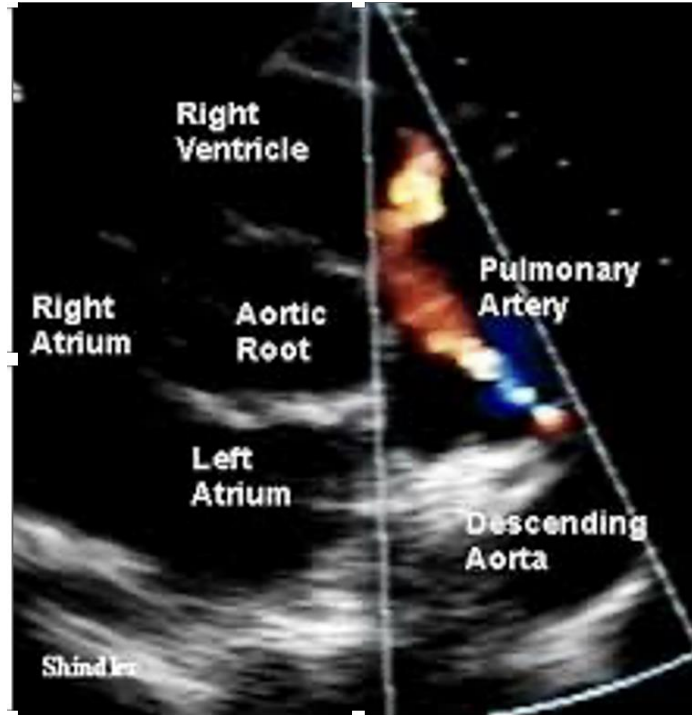
Indications for Surgery

- Onset of Symptoms
- Evidence of Elevation of Pulmonary Artery Pressures (Exam, echo, cath)
- Development of Endocarditis
- Echo evidence of Chamber Enlargement
- CXR: Cardiomegaly, Pulmonary Congestion
- $Qp/Qs = \frac{Ao\ Sat - RA\ Sat}{LA\ Sat - PA\ Sat}$
- - Large: > 2.0
 - Moderate: 1.5 to 2.0
 - Small: < 1.5

Patent Ductus Arteriosus (Great Vessel)

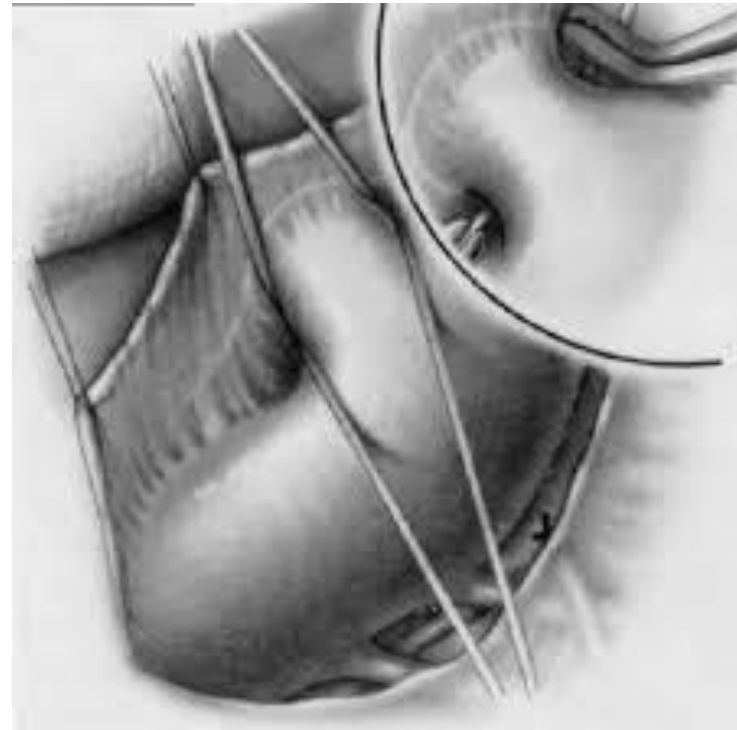


ADAM

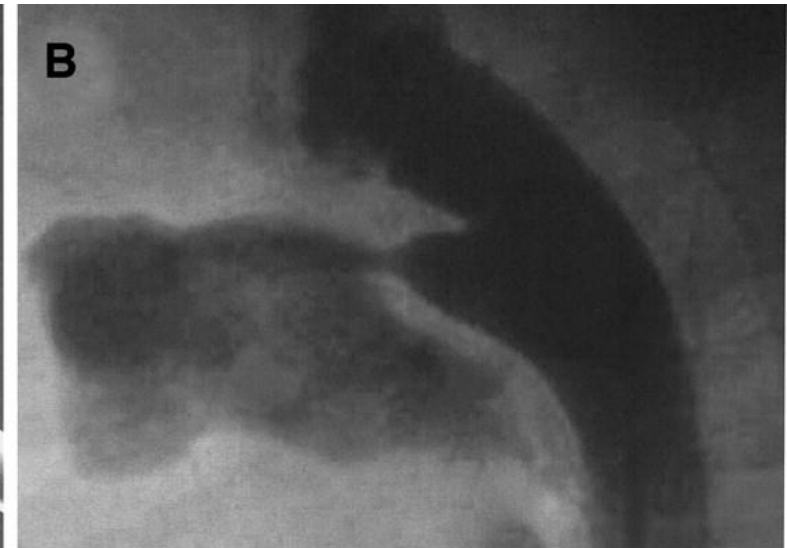
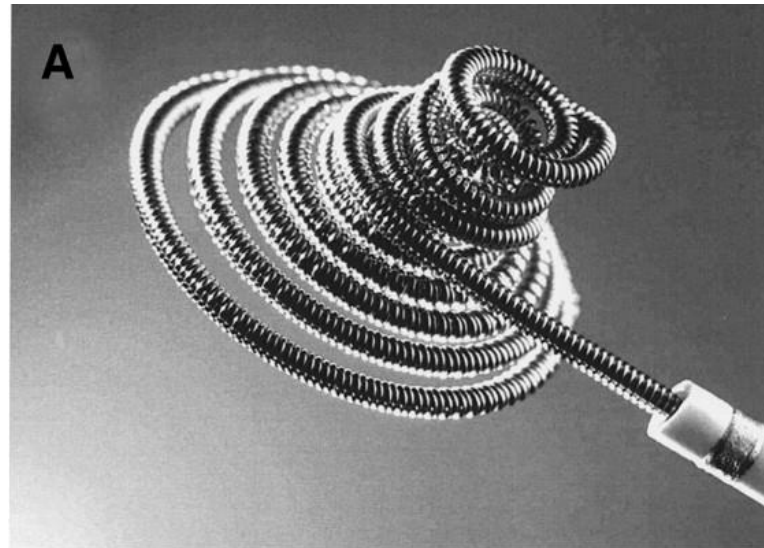


Surgical Closure of PDA in Premature Infant

- Left Thoracotomy (Open)
- Performed in NICU



Percutaneous Closure of PDA in Cardiac Catheterization Lab



Eisenmenger Syndrome

- Irreversible Histologic Changes
- $PVR > SVR$ leads to right to left shunting
- Cyanosis, Clubbing, Polycythemia, Paradoxical Embolus
- Early Death



Left Sided Obstructive Disease

- Coarctation of the Aorta
- Interrupted Aortic Arch
- SupraValvar Aortic Stenosis
- Aortic Stenosis
- Subvalvar Aortic Stenosis
- Multiple Levels of Obstruction: *Shone's Syndrome*

Coarctation of the Aorta



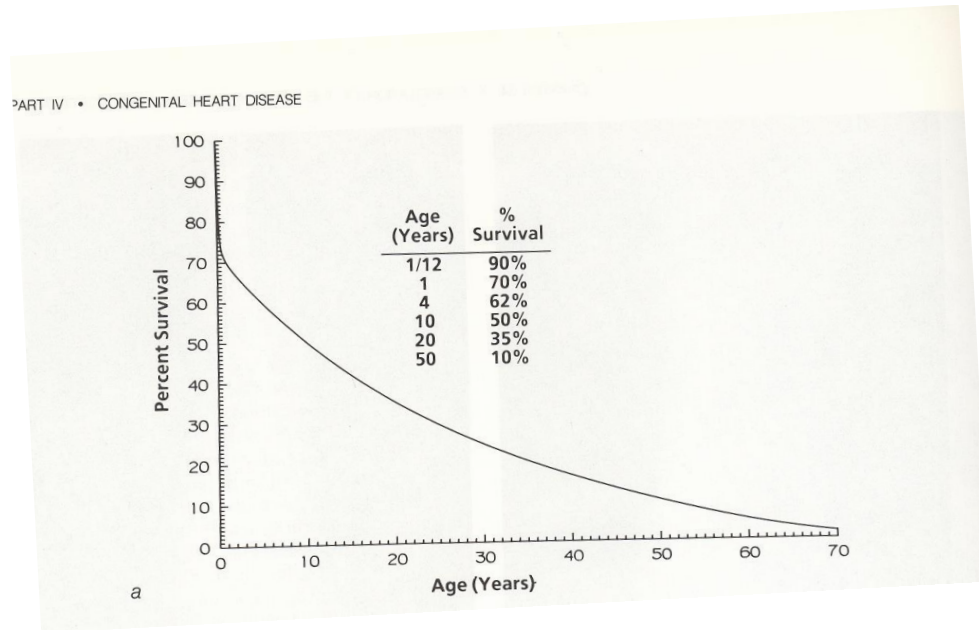
Coarctation

- Latin: coarctere: “to contract”
- First reported: Morgagni 1760
- Incidence: 0.2 to 0.6 per 1000 live births
- Male/Female ratio: 1.3 – 1.7/1.0
- 5 to 8 % of congenital heart disease
- Seasonal: late fall and winter occurrence
- Abdominal Aorta 0.5 % to 2 %

Associated Cardiac Anomalies

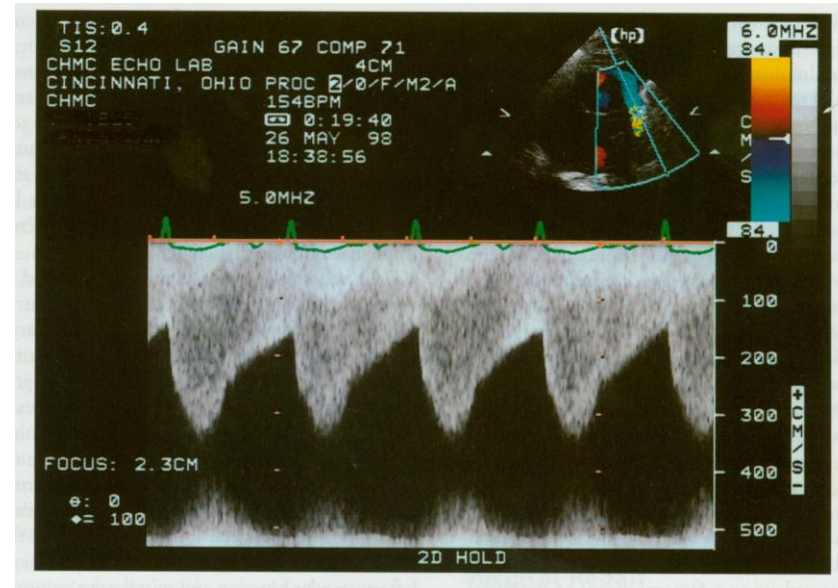
- Bicuspid aortic valve
- Ventricular Septal Defect
- Mitral Valve Stenosis
- Shone's Syndrome
 - Parachute Mitral Valve
 - Supravalvar Mitral Ring
 - Subaortic Stenosis
 - Coarctation

Premature Death

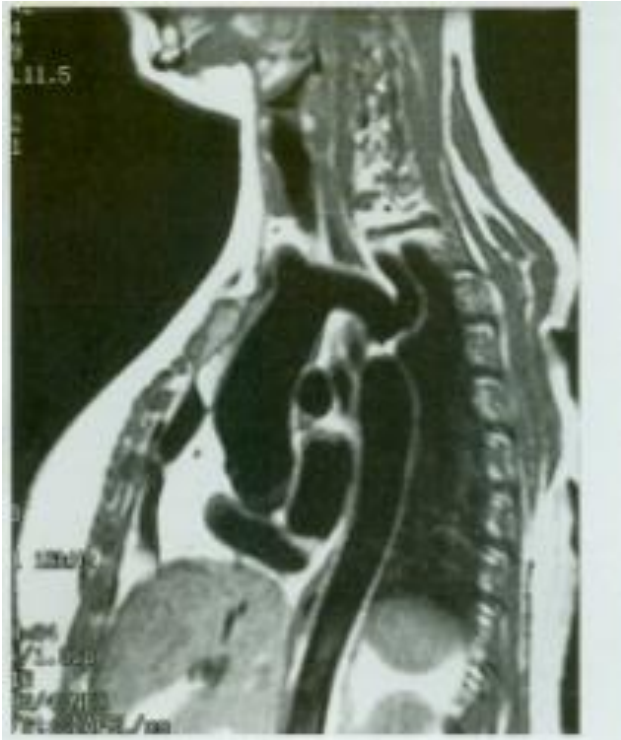


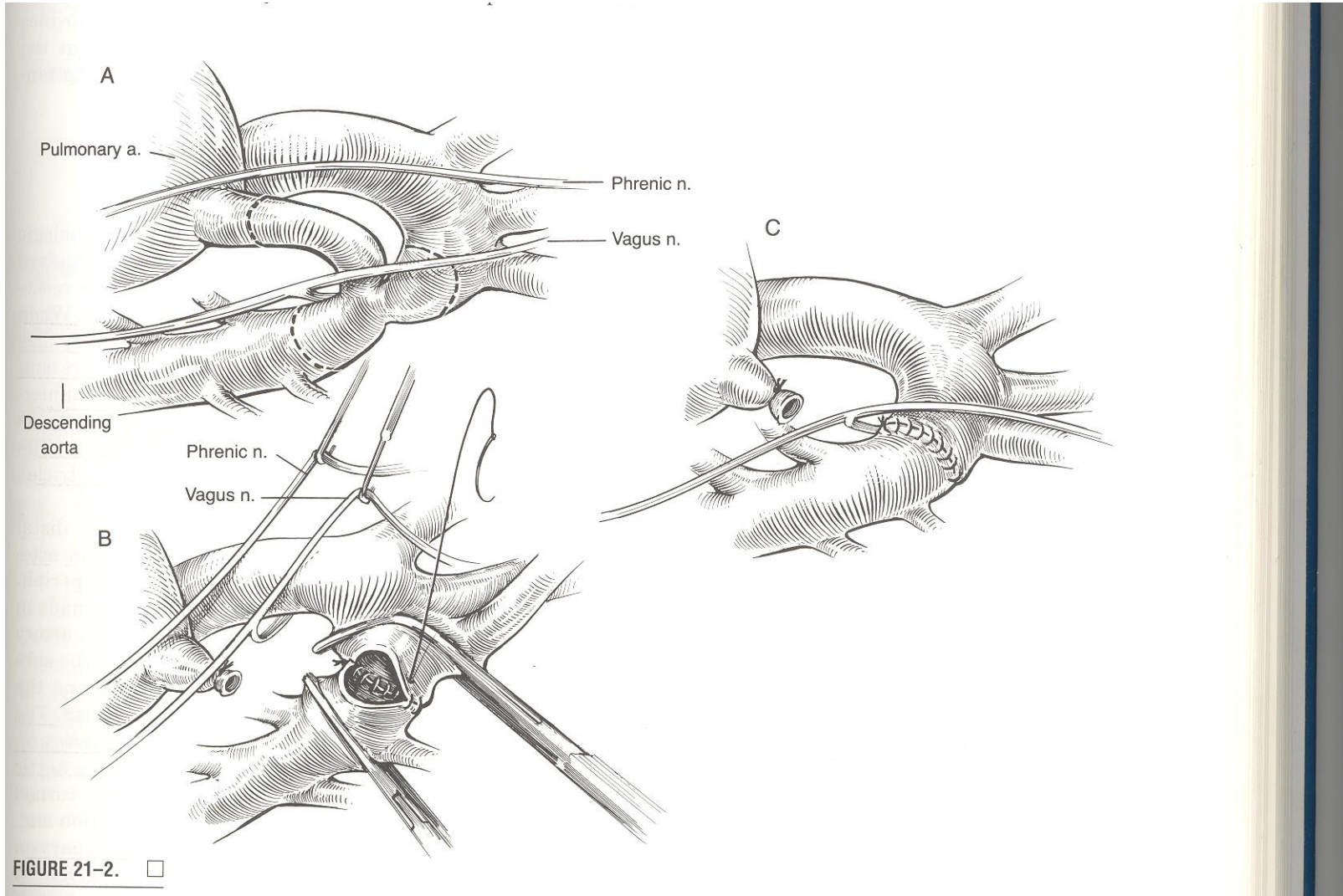
- 26 % Congestive Heart Failure
- 21 % Aortic Rupture
- 18 % Bacterial Endarteritis
- 12 % Intracranial Hemorrhage

Echocardiography

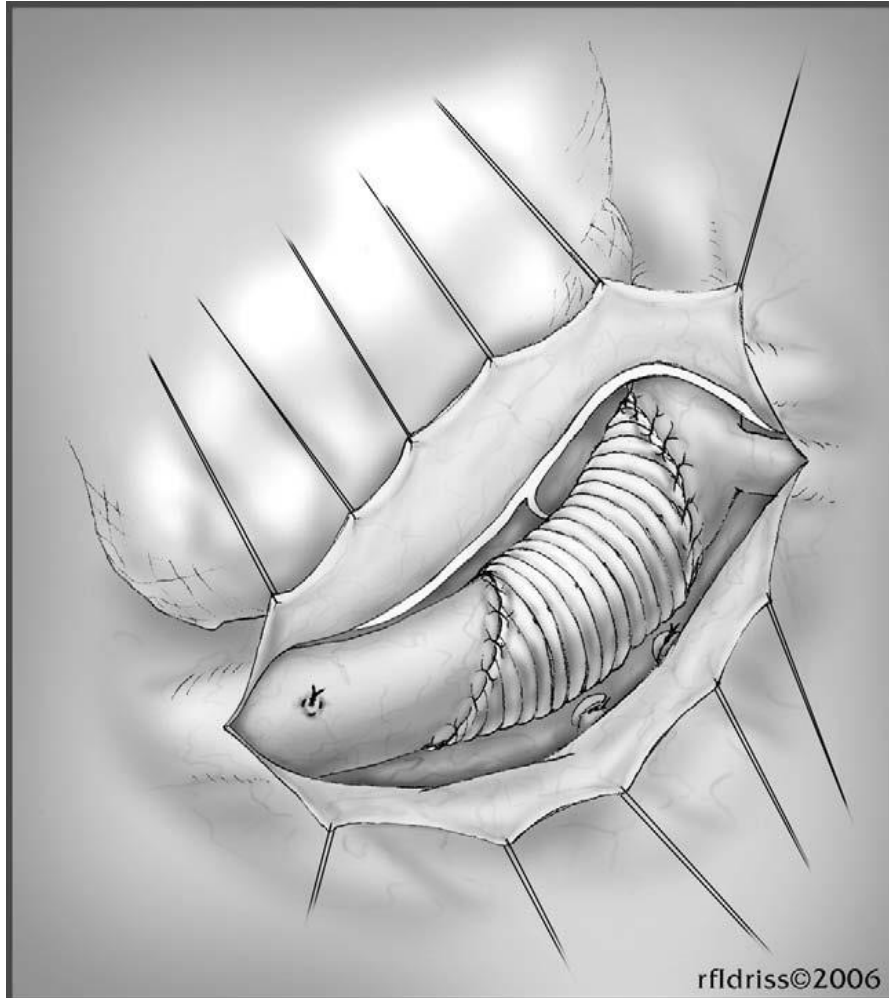


Magnetic Resonance Imaging



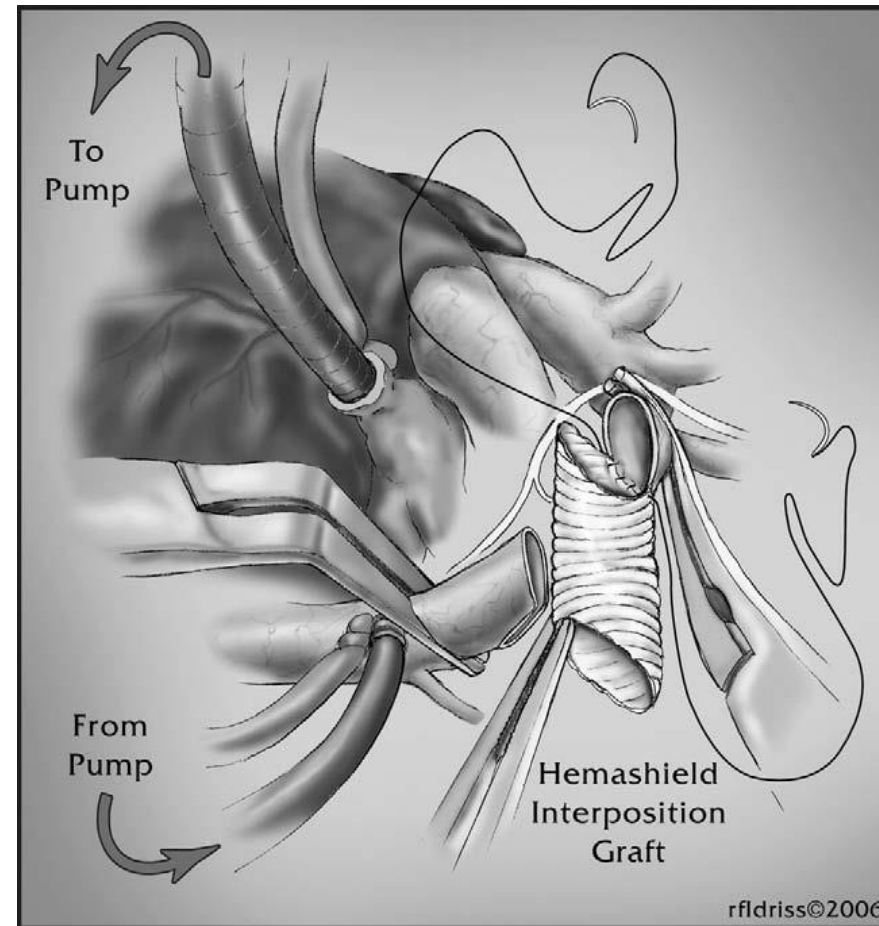
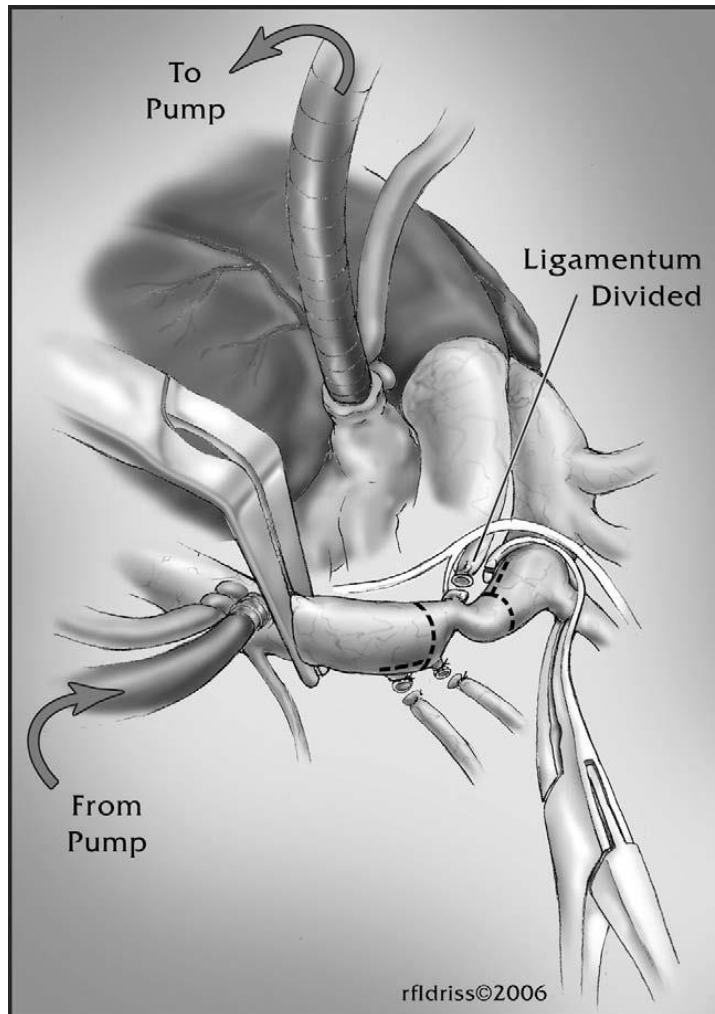


Graft Replacement

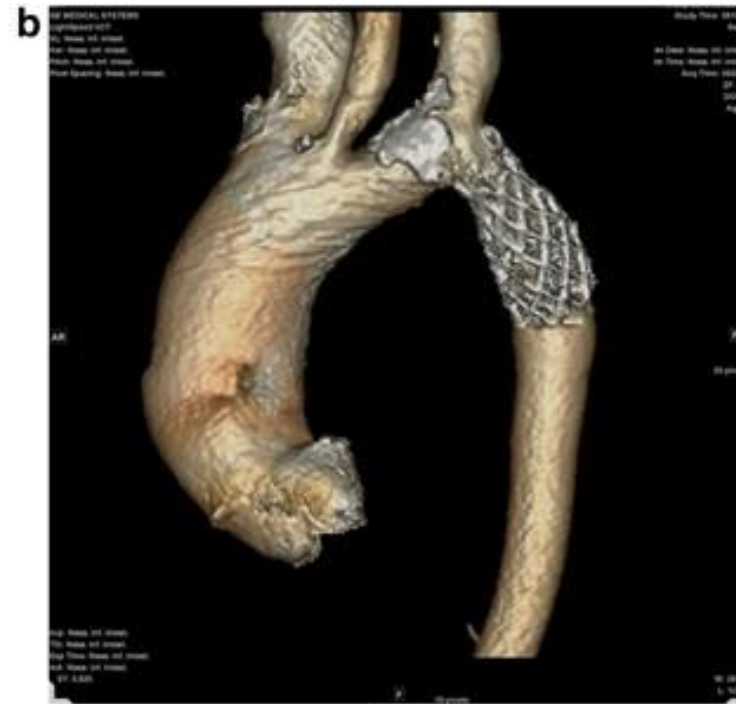
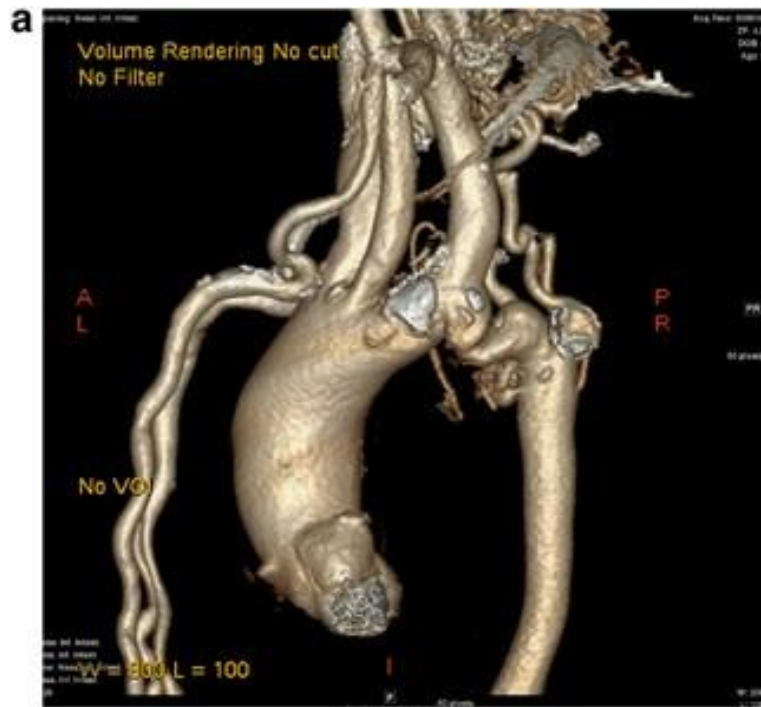


- Left Thoracotomy
- Assess collateral flow to distal aorta
- Left heart bypass if collaterals insuff
- Teenagers and Adults

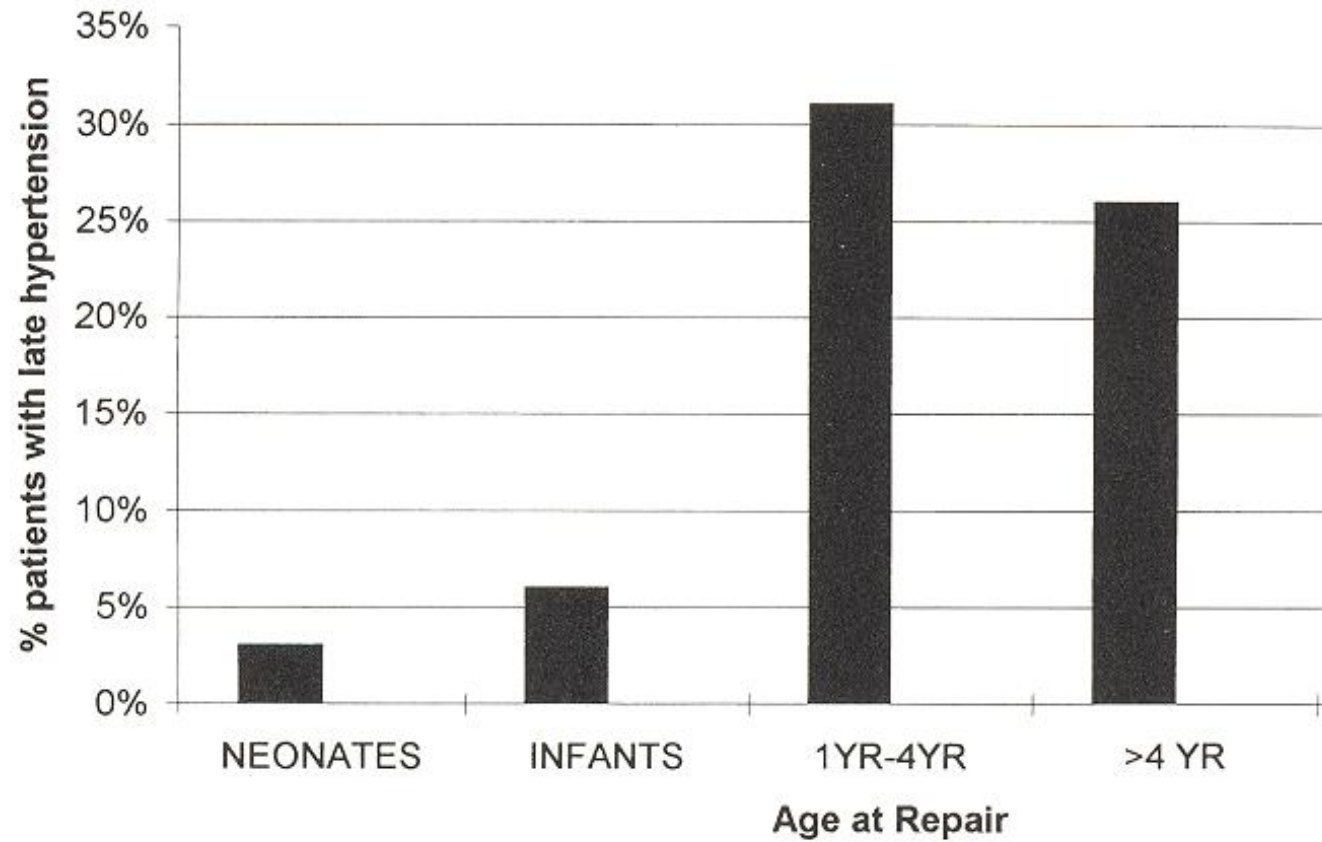
Repair Using Left Heart Bypass



Stent graft Repair of Discrete Coarctation



Ann Thorac Surg
1998;66:1378-82



Etiology of Persistent Hypertension

- Abnormal baroreceptor settings
- Reduced arterial compliance
- Endothelial dysfunction
- Subclinical residual obstruction
- Increase in collagen and decrease in smooth muscle proximal to coarctation

Right Sided Obstructive Disease

- Pulmonary Stenosis
- *Tetralogy of Fallot*
- Pulmonary Atresia
 - Intact Vent Septum
 - VSD: TOF
- Tricuspid Atresia
 - Importance of Nonrestrictive ASD
 - Usually Associated with Hypoplastic RV
 - Presents with Cyanosis

Who was Fallot ?



- Anatomic Pathologist
- *La Maladie Bleue* not simply caused by a patent foramen of Botallo (Ovale)
- Pathophysiology of cyanotic heart disease: anatomic malformations
- Marseille-Medical 1888

Tetralogy of Fallot

- Most common cyanotic heart disorder

- Incidence: 32.6 per 100,000 births

- Characterized by:

- Pulmonic Stenosis:

- Subvalvular
 - Valvular
 - Supravalvular

- RV Hypertrophy

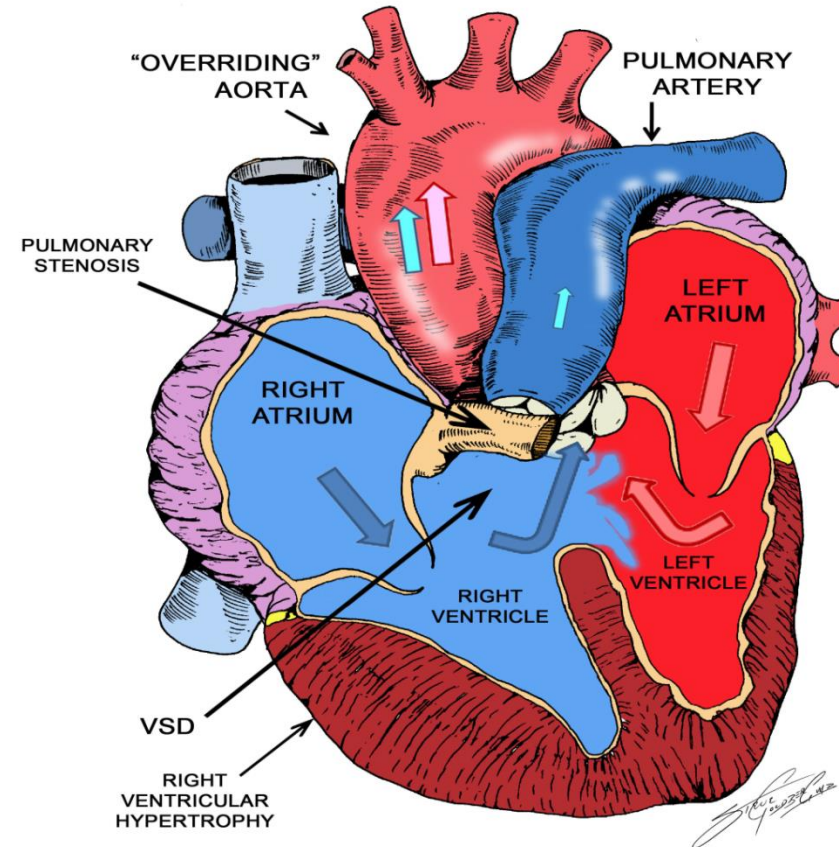
- Overriding Aorta

- Ventricular Septal Defect

- Variants:

- 25% of patients w/ R-sided aortic arch

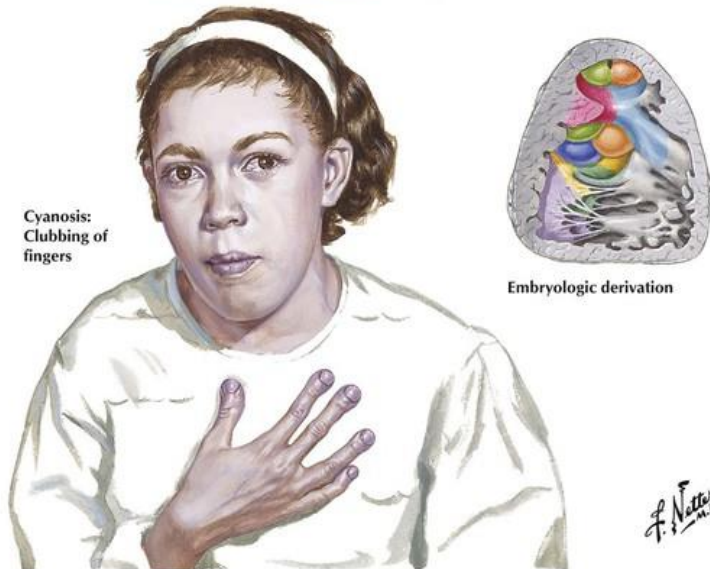
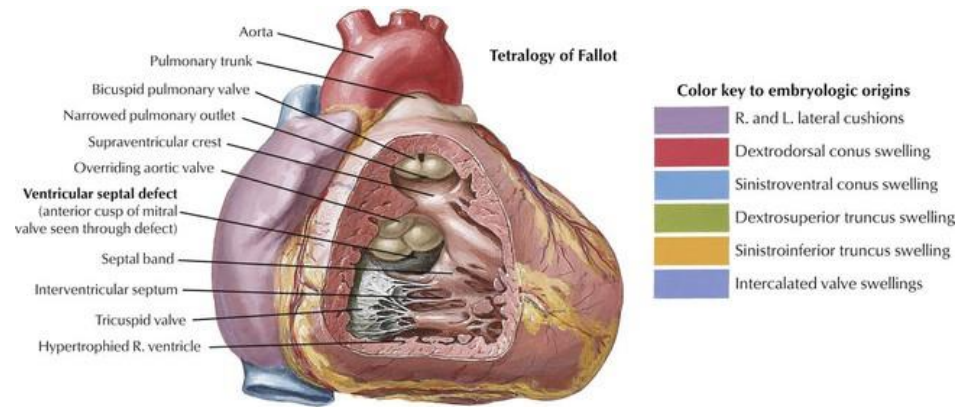
- “Pentalogy of Fallot” – ASD or PDA along with other lesions



Incidence of Tetralogy

- 3/10,000 live births
- 10 % of congenital heart disease
- Males > Females
- Down' s Syndrome 8 %
- 5 to 15 % of offspring will have congenital heart disease

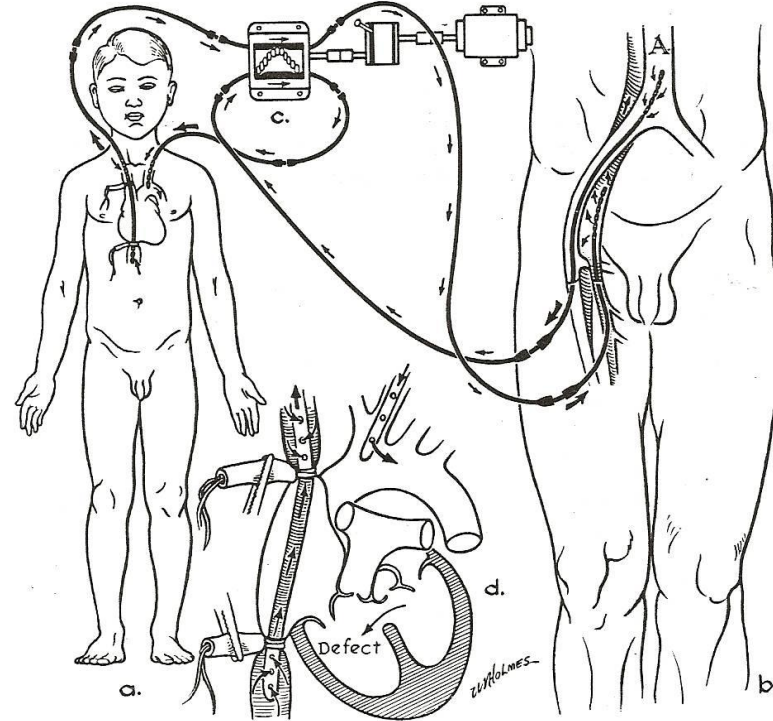
Clinical Presentation: Children and Adults



- Polycythemia
- Thromboembolic events
- Paradoxical Emboli
- Strokes, Cerebral Abscesses
- Atrial and Ventricular arrhythmias

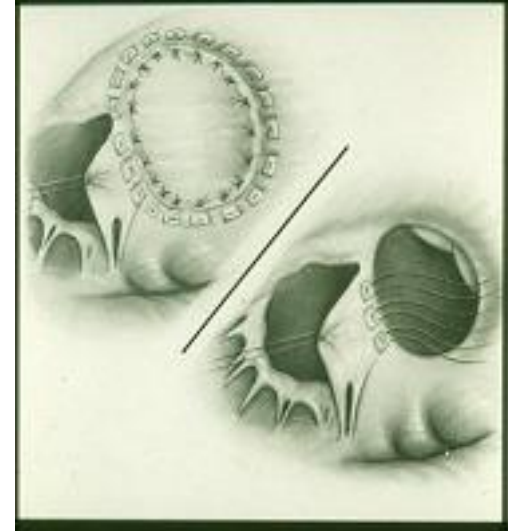
Cross Circulation

- No Oxgenator
- Low Flows
- Normothermia
- Heart Beating
- Short bypass intervals



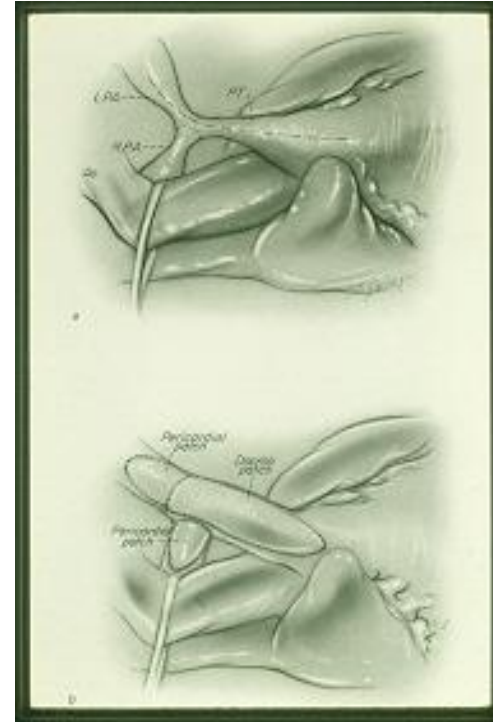
Closure of Ventricular Septal Defect

- Patch
- Interrupted versus running suture
- Right ventricular vs right atrial approach
- Septal leaflet of TV used to anchor sutures
- Avoid: aortic valve, His bundle



Relief of Outflow Tract Obstruction

- Infundibular Obstruction
- Valvar Stenosis
- Pulmonary Artery Stenoses
- Limit RV Incision
- ? Divide Pulmonary Artery Annulus



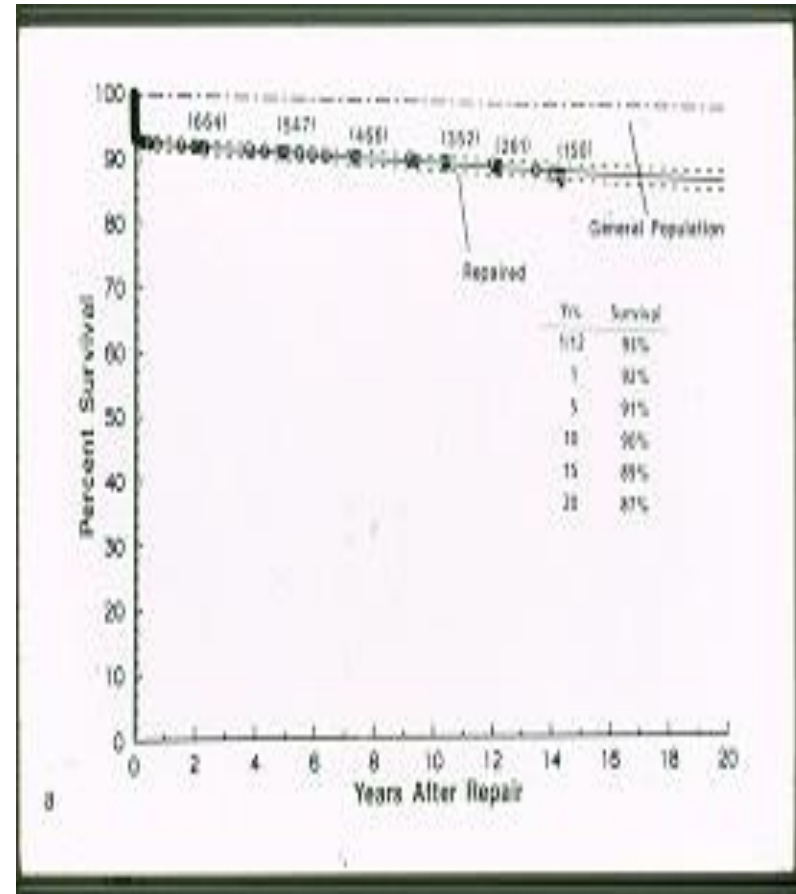
Transannular Patch Augmentation

- Approx 50 % Incidence
- Obligatory Pulmonary Insufficiency
- Monocusp Patch
- Size Annulus Before and During Surgery



Longterm Survival Following Repair of TOF

- Initial operative mortality in current era ($< 2\%$)
- Low, but detectable mid and late term mortality curve



Clinical Determinants of Ventricular Ectopy

- *Hemodynamic Burden*

- Pressure load
- Volume load
- Systolic dysfunction

- *Electrical Abnormality*

- QRS > 180 msec
- Abnormal signal EKG

- *Surgical Factors*

- Older age at repair
- Presence and size of ventriculotomy
- Length and Complexity of Repair

Treatment of High Grade Ventricular Ectopy

- *Evaluation*

- History
- Resting EKG
- Holter
- Formal EPS

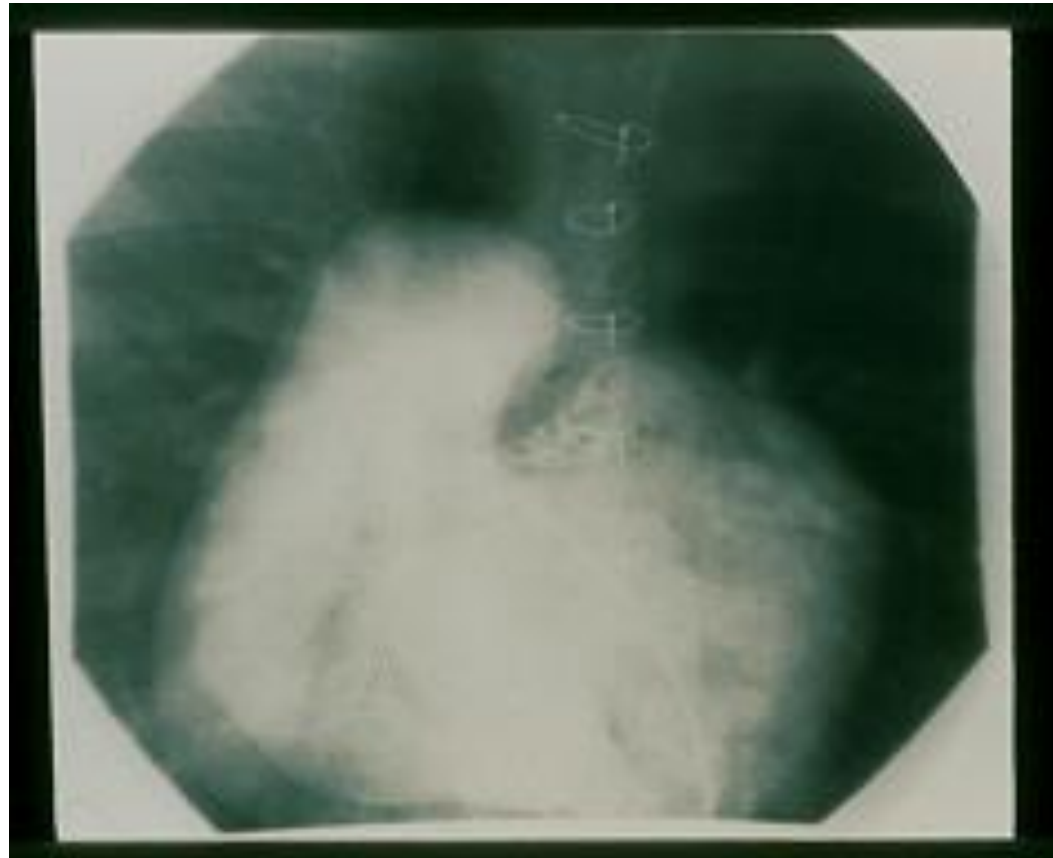
- *Indication for Treatment*

- Hemodynamic
- Electrical

Therapeutic Options

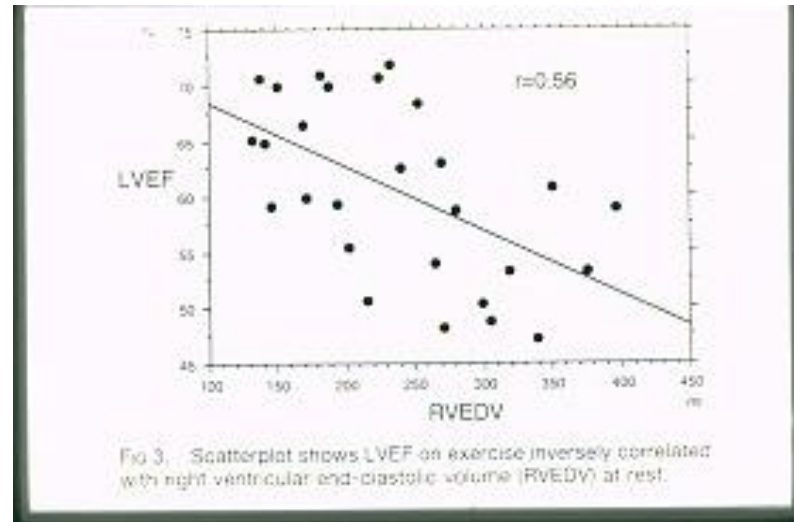
- Medical
- Surgical
 - Correct Structural Problems
 - Cryoablation
 - Endocardial Resection
- EP Intervention
 - Ablation of Focus
 - AICD

Right Ventricular Volume Overload



Effect of RV Volume Overload on LV Systolic Function

- Relationship Between Left Ventricular Function and Right Ventricular Volume Overload

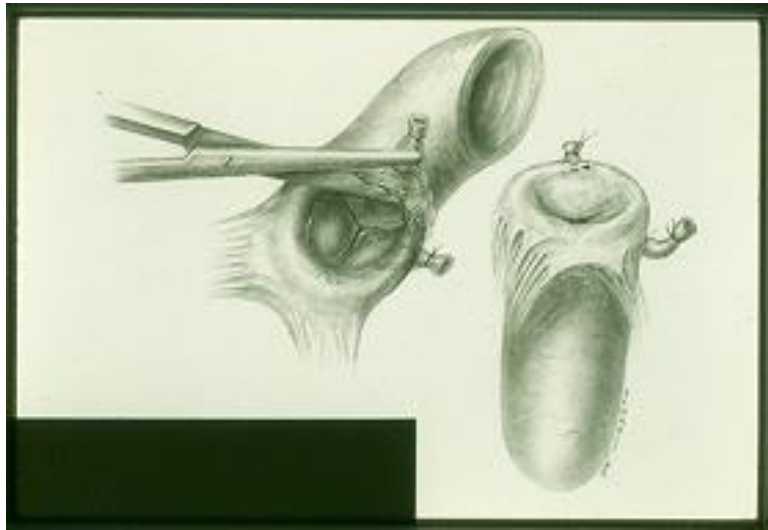


Long-Term Consequences of Pulmonary Regurgitation

- *Clinical Manifestations*
 - Diminished Exercise Tolerance
 - Chronic Fatigue
- *Right Ventricular Volume Overload*
 - Dilation of the Right Ventricle
 - Tricuspid Valve Insufficiency
 - Impaired Right Ventricular Function
- *Left Ventricular Impairment*
 - At Rest
 - During Exertion
- *Impaired Respiratory Function*
 - Resting State
 - With Physical Activity
- *Arrhythmogenic Ectopy*
 - Atrial Origin
 - Ventricular Origin

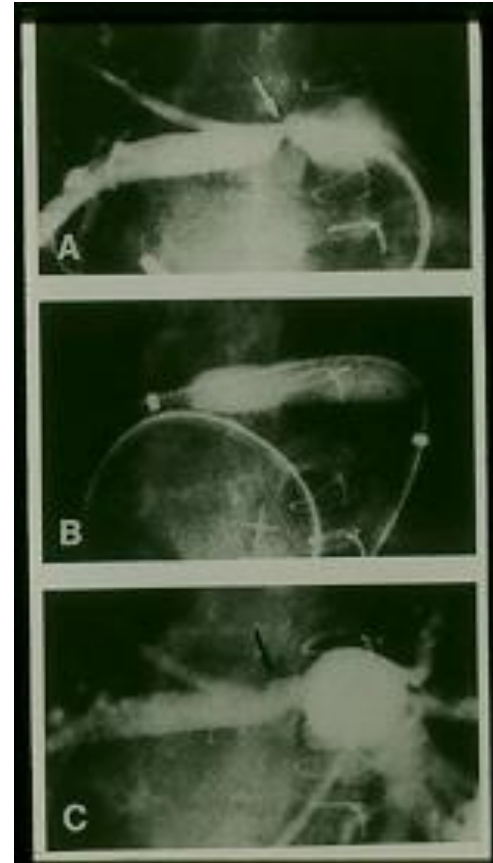
Surgical Technique

- Aortic Valve Homograft
- Orthotopic Position



Balloon Angioplasty

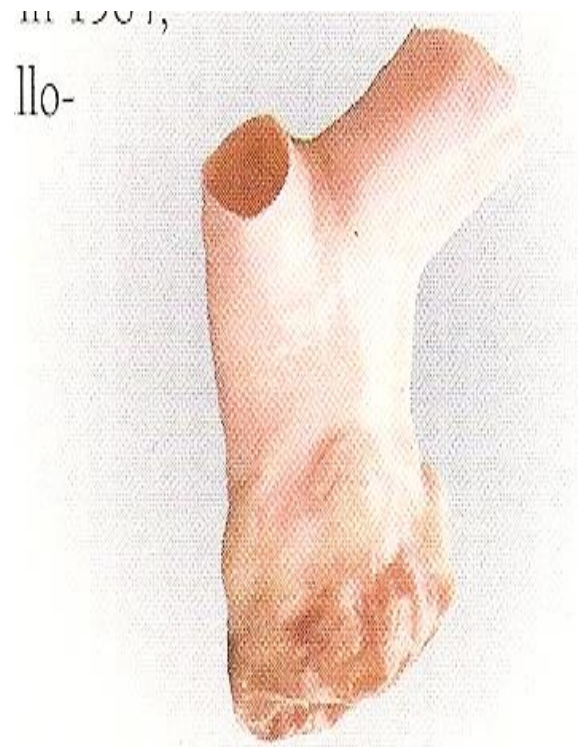
- Preoperative balloon dilation (n = 3)
- Postoperative balloon dilation (n = 4)
- Importance of relieving stenoses percutaneously or surgically



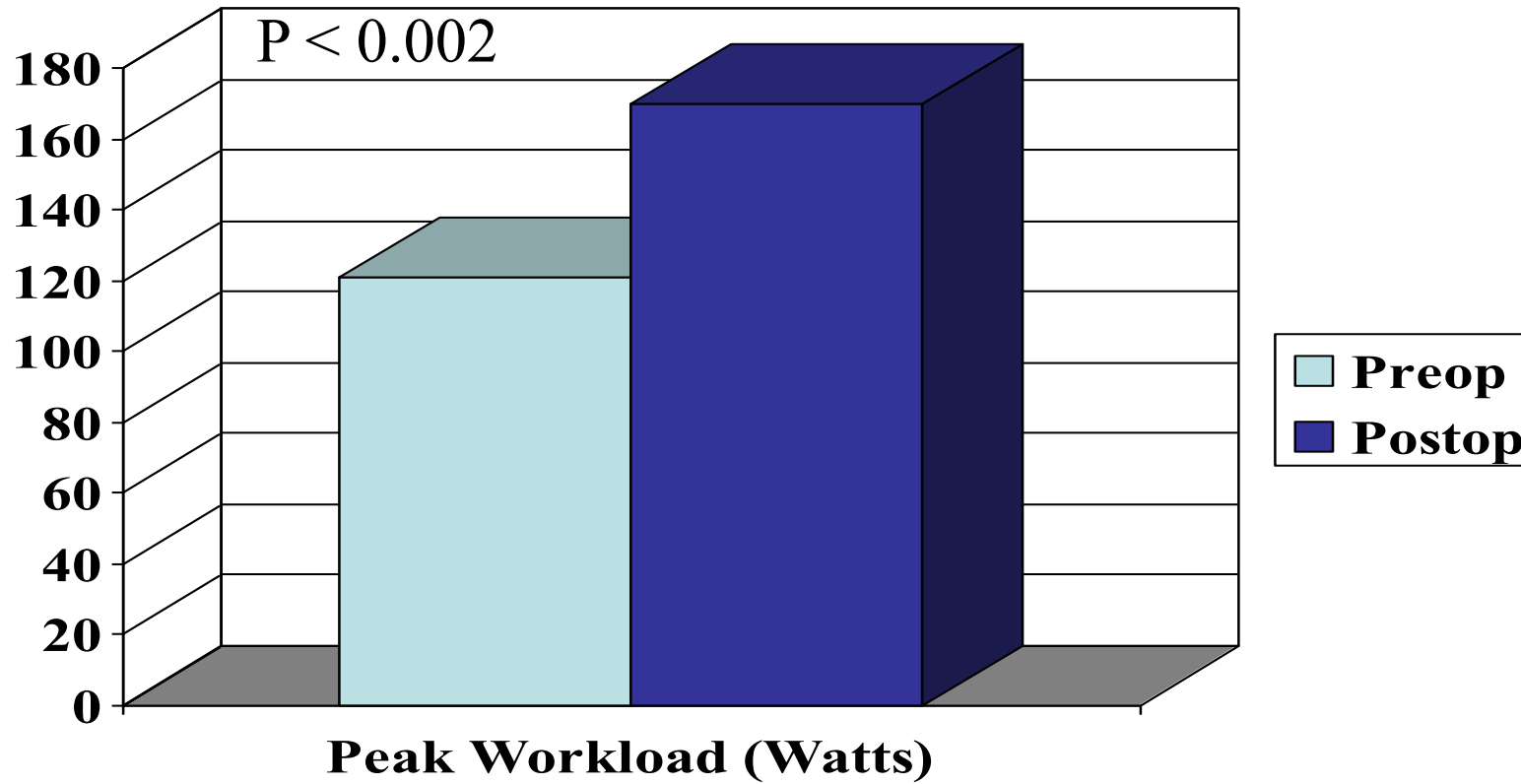
Bovine Pericardial Valve



Aortic vs Pulmonary Homografts



Exercise Performance



Clinical Symptoms

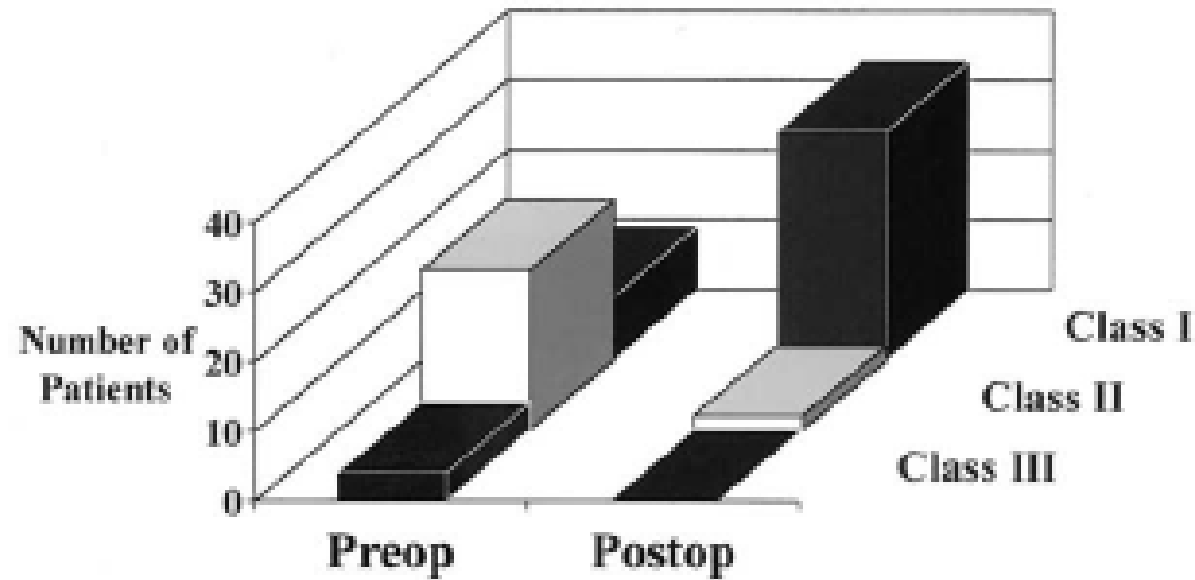
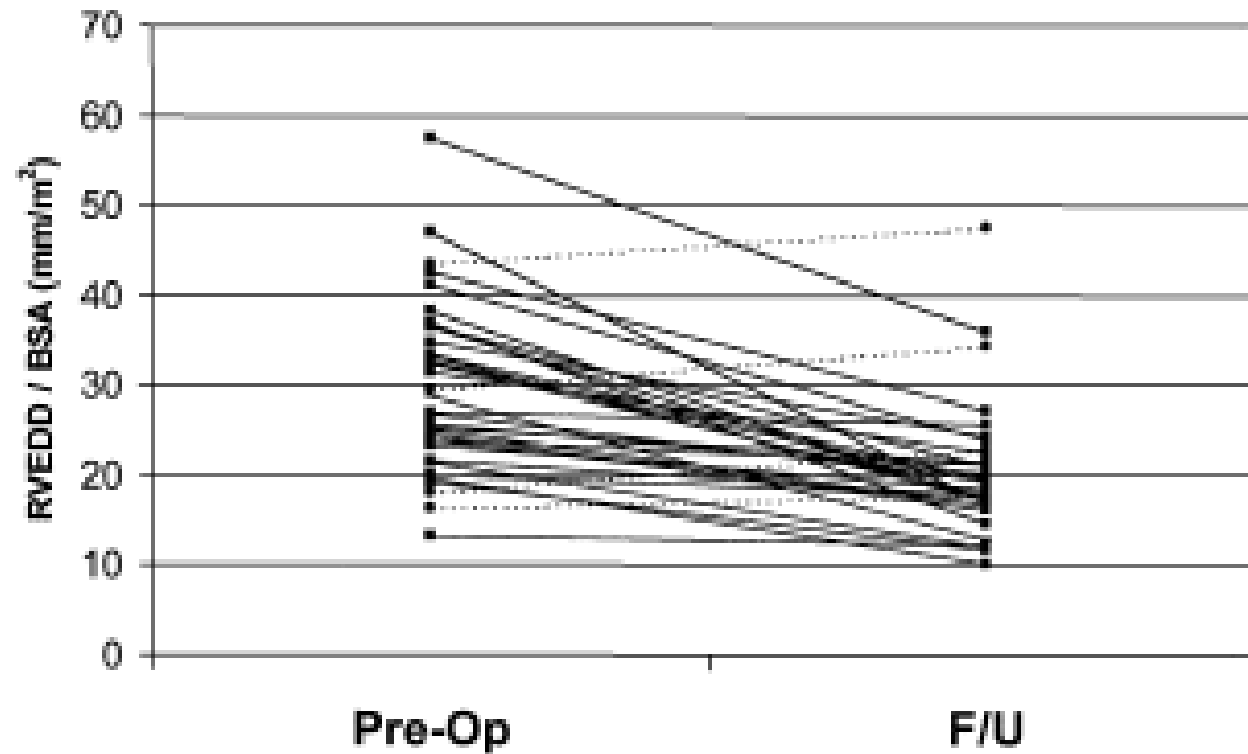
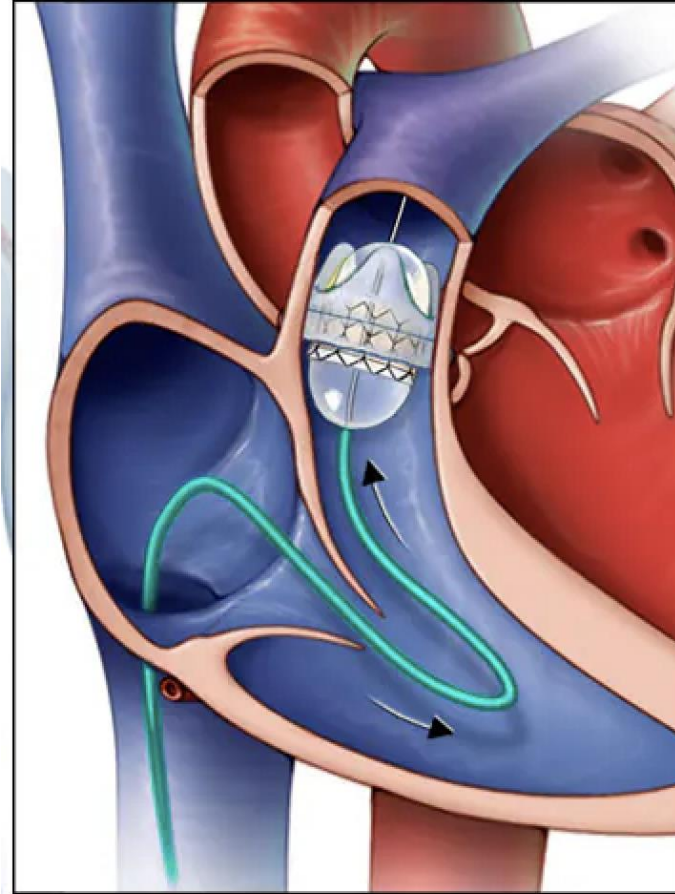


Fig 1. The patients are grouped according to their New York Heart Association classification status before ($n = 36$) and after ($n = 35$) surgery. The majority of patients were in class II before surgery. Virtually all patients were in class I after surgery at a mean follow-up of 81 months. (Preop = preoperative; Postop = postoperative.)

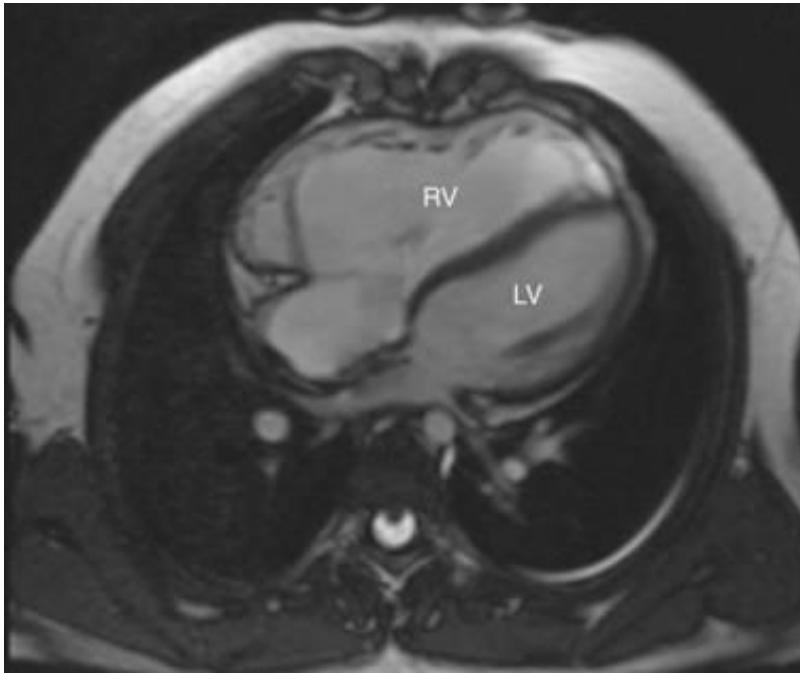
Changes in RV End Diastolic Volume



Percutaneous Pulmonary Valve Replacement (TPVR)



Cardiac MRI to Assess RV size and function



- RV Volumes: RVEDV and RVESV
- RV Ejection Fraction
- RV Regurgitant Fraction
- LVEF
- Additional lesions: pulm artery stenoses

Restoration of a competent pulmonary valve:

1. Relieves symptoms of exercise induced fatigue and dyspnea
2. Objectively improves exercise performance
3. Reduces right ventricular volume overload
4. Decreases the risk of fatal ventricular arrhythmias (?)

Pregnancy and ACHD

- Increased Risk for Mother and Fetus
 - If surgery indicated: 3rd trimester C-section followed by surgical repair
 - High rate of fetal demise if open heart procedure during 1st or 2nd trimester
 - Maternal mortality 12 % for open heart procedure
- Risk Factors:
 - Severe pulm HTN
 - Advanced cardiomyopathy
 - Obstructive lesions: aortic stenosis and mitral stenosis
 - Left to right shunts: ASD, VSD better tolerated

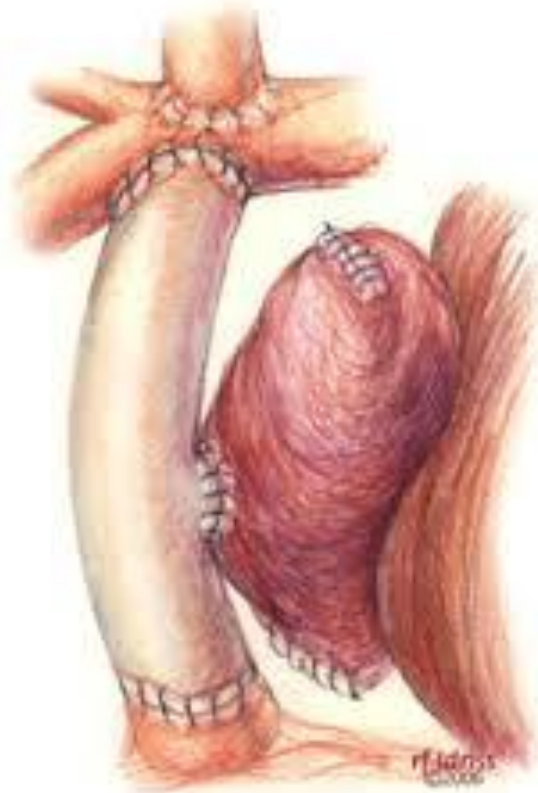
Single Ventricle Anatomy

	<i>Dominant Ventricle</i>
Tricuspid Atresia	Left Ventricle
Pulmonary Atresia/Intact Ventricular Septum	Left Ventricle
Double Inlet Left Ventricle	Left Ventricle
Complex Double Outlet RV	Right Ventricle
Hypoplastic Left Heart Syndrome	Right Ventricle
Severe Ebstein's Anomaly	Left Ventricle
Heterotaxy Syndrome	Right Ventricle

Structural Differences Right vs Left Ventricle

	Right Ventricle	Left Ventricle
<i>Shape</i>	Funnel	Conical
<i>Coronary Artery Supply</i>	RCA, LAD	LAD, Lt Circumflex, RCA
<i>Pattern of Contraction</i>	Synchronized	Simultaneous
<i>Conduction System</i>	1 fascicle	2 fascicles
<i>Papillary Muscles</i>	Numerous	Two
<i>AtrioVentricular Valves</i>	3 cusps	2 cusps
<i>Presence of Conus</i>	Yes	No
<i>AtrioVentricular Semilunar Valves Contiguous ?</i>	No	Yes

Completion Fontan Procedure



- IVC connected via Goretex tube to RPA
- Systemic and Pulm Circuits separated
- Pure laminar flow to pulmonary bed
- Importance of low PVR
- Role of Fenestration
- O2 Sats 90 % to 95 %

Lifetime Management of Congenital Heart Disease

- Congenital Heart Program
- Collaboration:
 - Cardiology
 - Cardiac Surgery
 - Electrophysiology
 - Interventionalist
 - Vascular Surgery
- Correction of Residual Lesions
- Antibiotic Prophylaxis
- Exercise programs



Citations

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