Transposition

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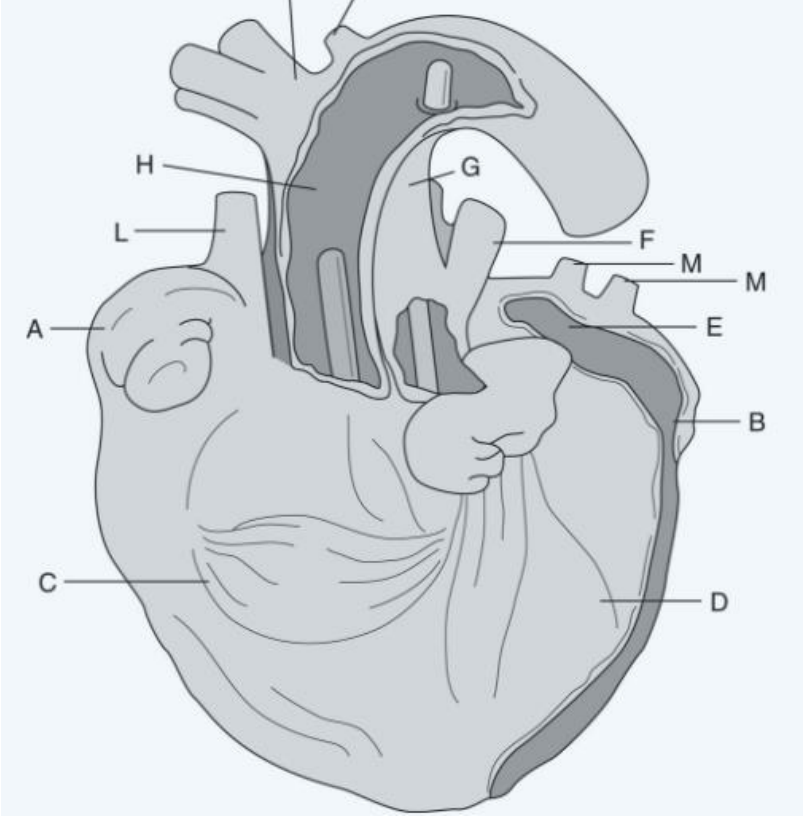
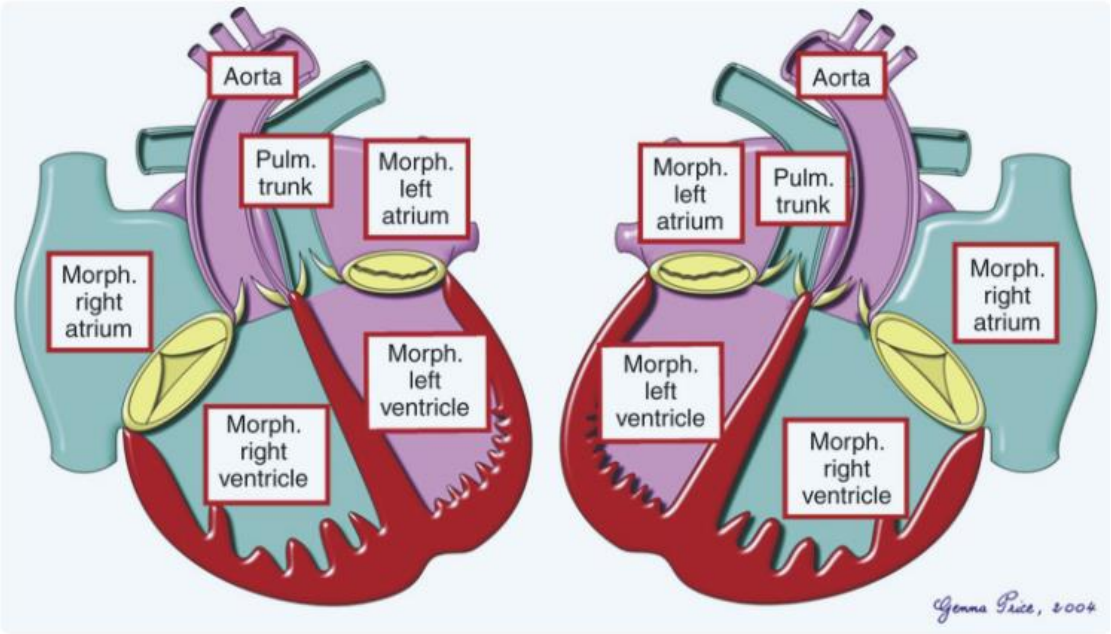
Introduction



- Matthew Baillie, first describe
- 1797
- Mr Langstaff, second describe
- 1811
- According to their ventricular origin, 1971, R. Van Praagh
- An anterior location of the aorta relative to the pulmonary trunk, 1971, L.H.S. Van Mierop

Introduction

- Concordant atrioventricular and discordant ventriculo-arterial connections

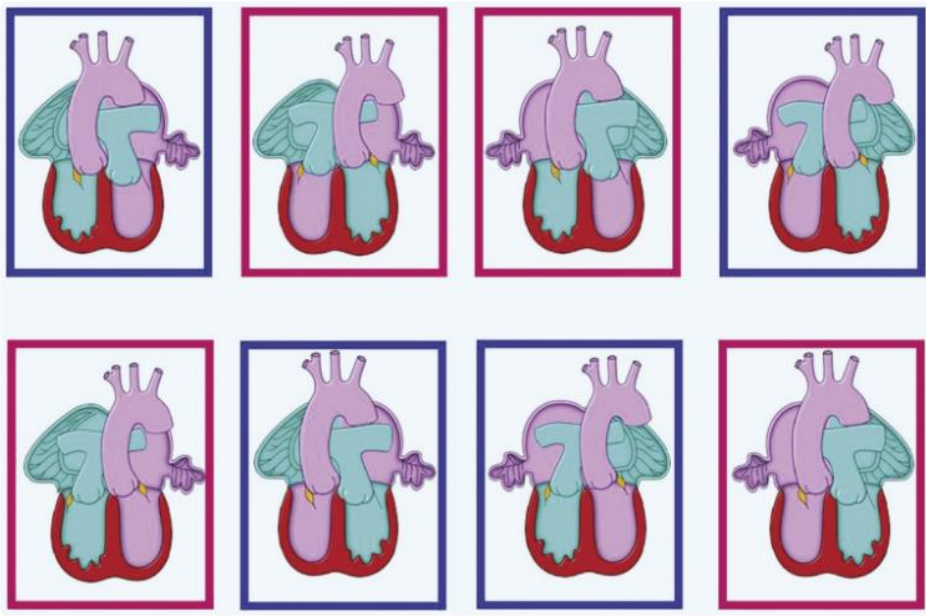


Introduction

- Discordant ventriculo-arterial connections associated also with discordant connections at the atrioventricular junction -> congenitally corrected transposition (ccTGA)
- Discordant ventriculo-arterial connections in combincation with isomeric atrial appendages -> functionally univentricular atrioventricular connections (FSV)

Historical background

- J.R. Farre, 1814, first used the term transposition
- A period becoming the norm to define the entity in terms of the anterior location of the aorta
- The aortic valve should be combined with support from a muscular infundibulum
- Produced the potential for confusion



Historical background

- Definition transposition on the basis of the origin of the great arterial trunks from morphologically inappropriate ventricles
- -> all potentials for confusion are avoided

Prevalence and aetiology

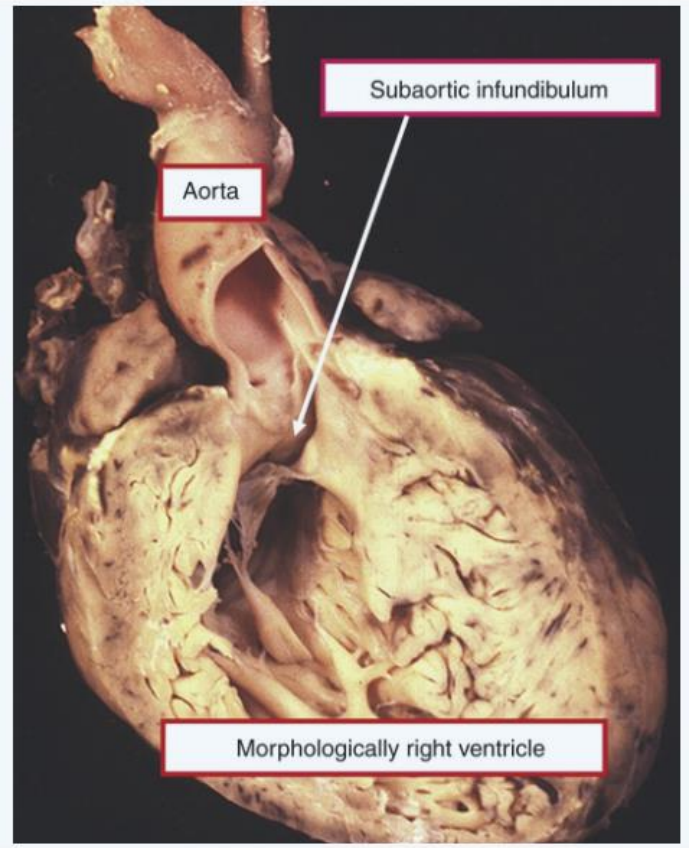
- 1/20 of all congenital cardiac malformation
- 1/5 of cardiac deaths in infancy prior to the era of surgical correction
- Boys, 2-3 times as frequently as girls
- More frequent
  - Infants of diabetic mothers, maternal intake of alcohol, poor nutrition or a stressful event during pregnancy
- Modest reduction
  - Addition of folic acid to the maternal diet

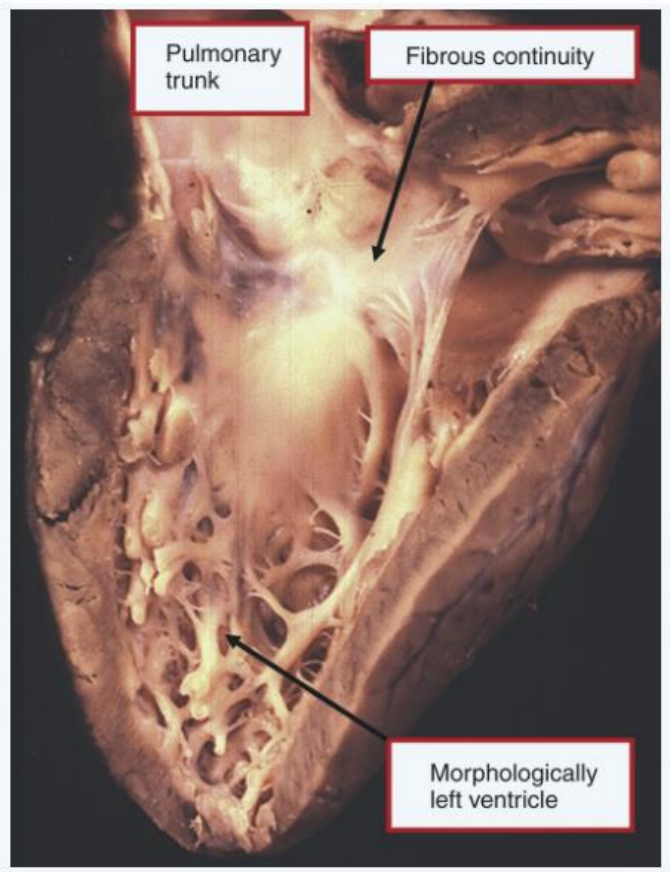
Anatomy and morphogenesis

- Anatomy
- Concordant atrioventricular and discordant ventriculoarterial connections
- Associated malformations
  - VSD
  - LVOTO

Anatomy and morphogenesis

- Basic segmental combinations



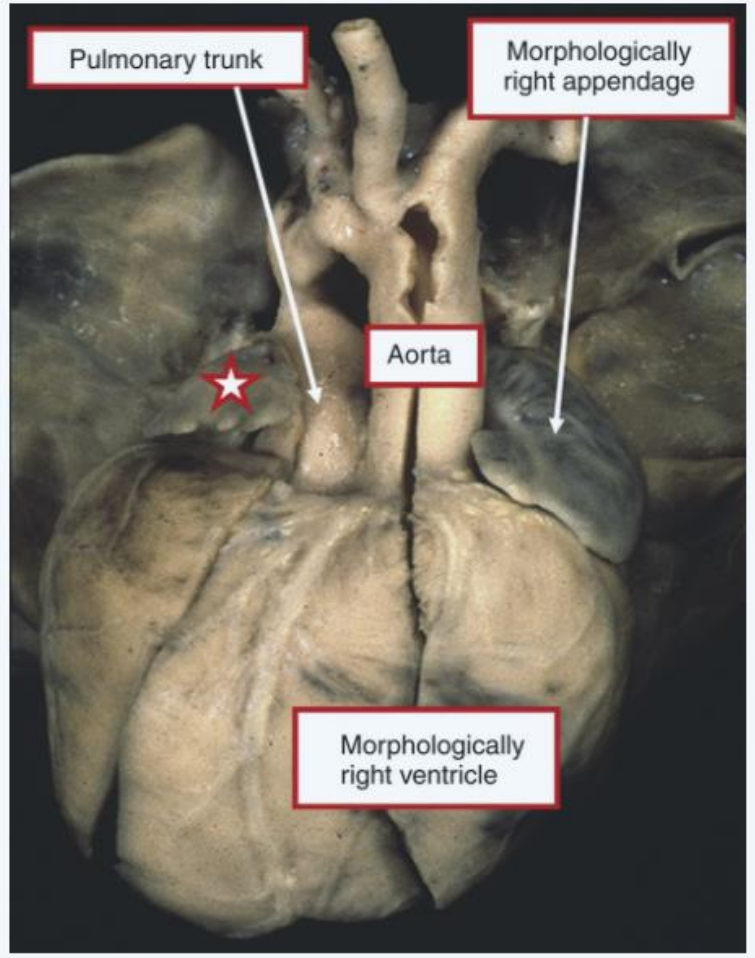


Anatomy and morphogenesis

- Basic segmental combinations
- The internal anatomy of the atrium is basically normal
- Unlike the atrial chambers, ventricular morphology is subtly different from normal
- At birth, the walls of the morphologically left ventricular wall are marginally thicker than those of the right ventricle
- The right ventricular mural thickness then rapidly increases in the first 2 years of life, becoming much thicker than that of the left ventricle

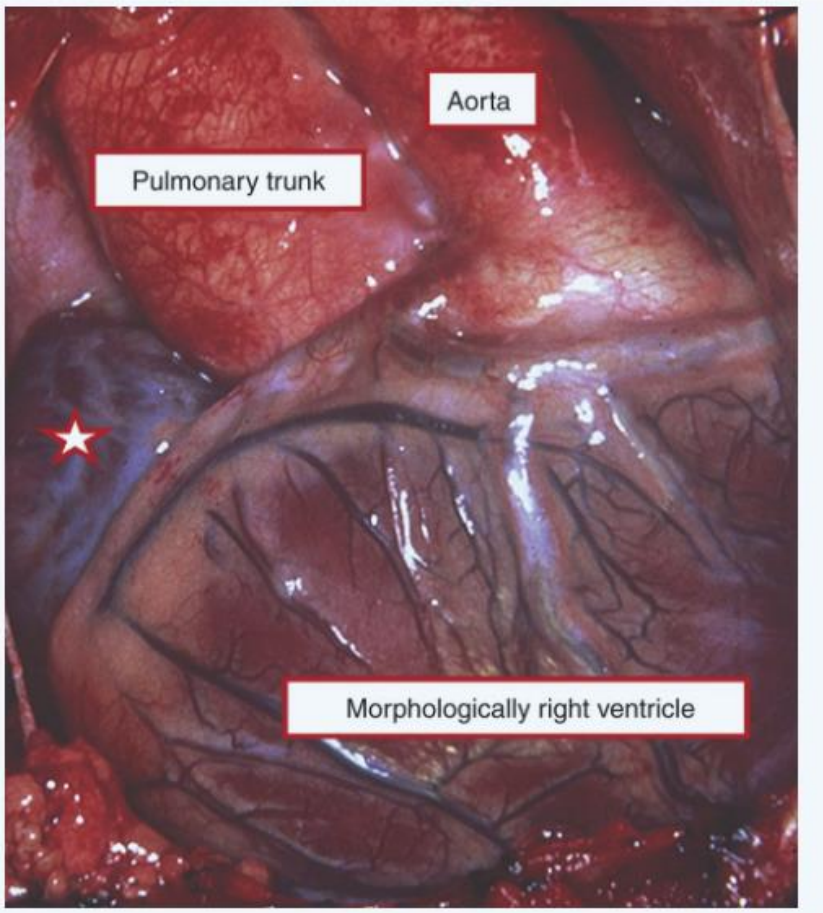
Anatomy and morphogenesis

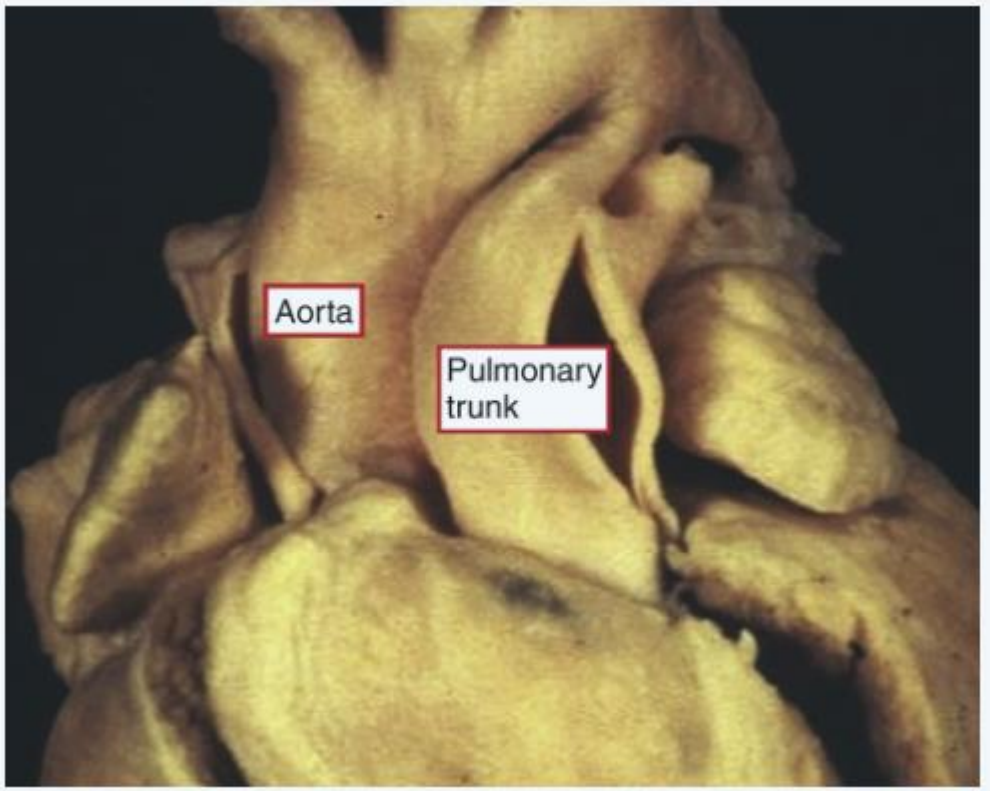
- Basic segmental combinations
- The aortic root, the right, anterior
- The left, anterior, in the mirror-imaged variant



Anatomy and morphogenesis

- Basic segmental combinations



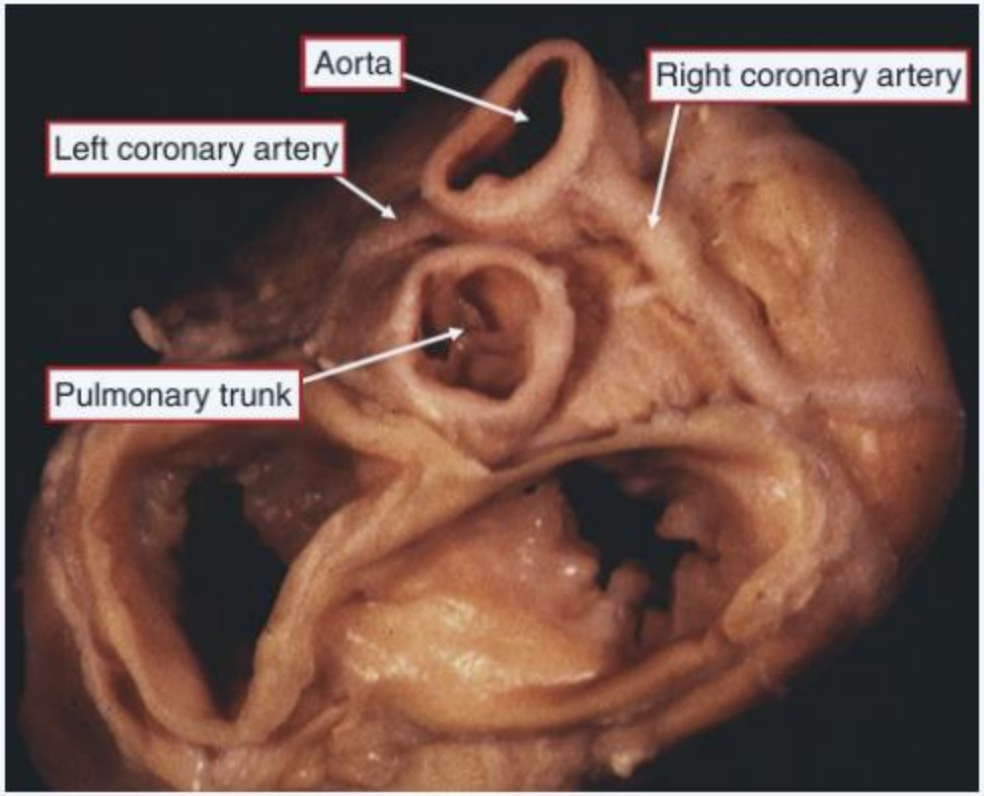


Anatomy and morphogenesis

- Infundibular morphology
- The aorta almost always has a complete muscular infundibulum
- The leaflets of the pulmonary valve are in fibrous continuity with the mitral valve

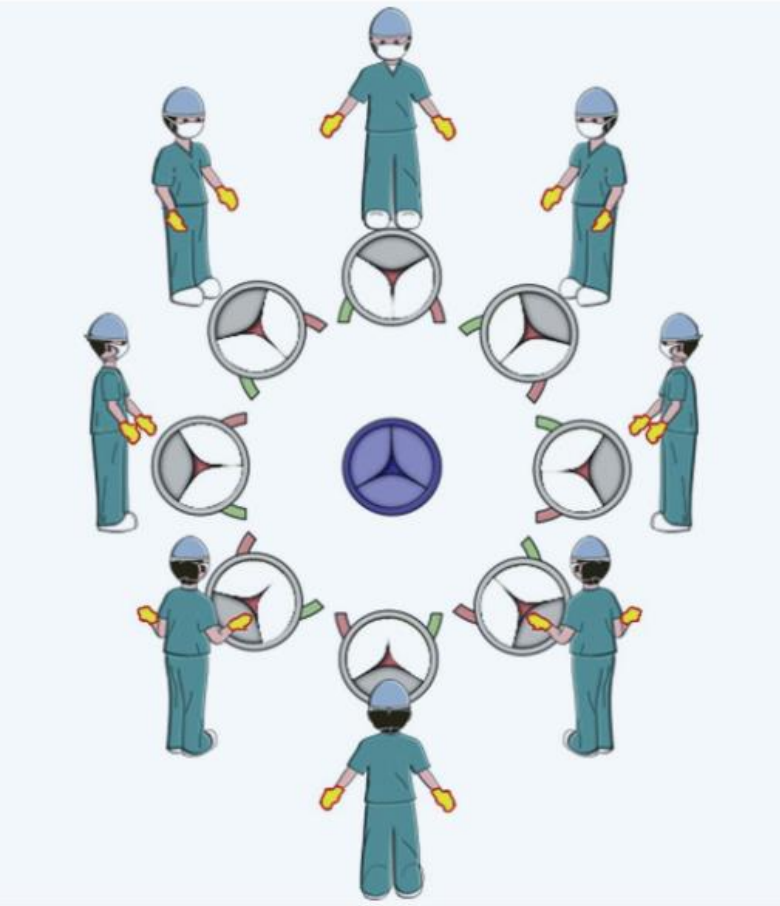
Anatomy and morphogenesis

- Coronary arteries
- The origins of the three major coronary arteries from the aortic sinuses
- Describing their course relative to the vascular pedicle, noting any intramural course
- With attention paid to the location of the artery supplying the sinus node



Anatomy and morphogenesis

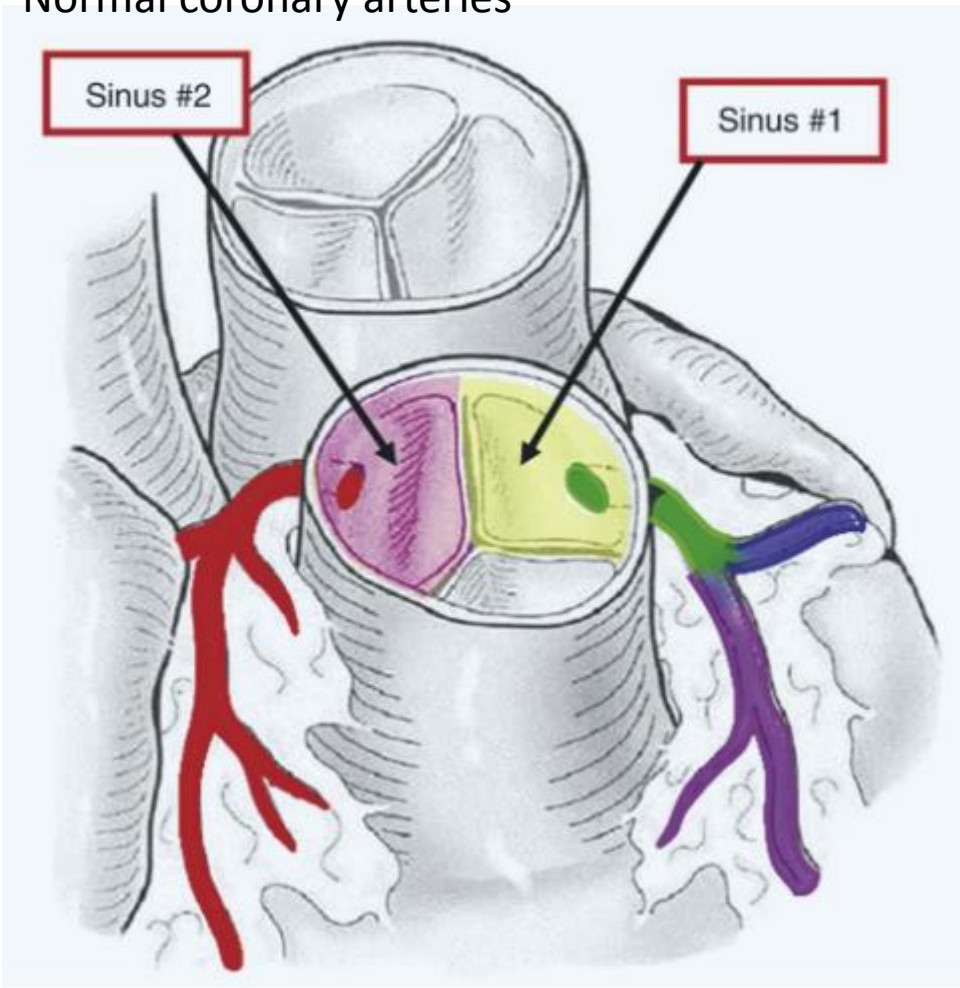
- Coronary arteries
- An observer standing in the non-adjacent aortic sinus and looking towards the pulmonary trunk
- The sinus to the right hand, sinus 1
- The left hand, sinus 2



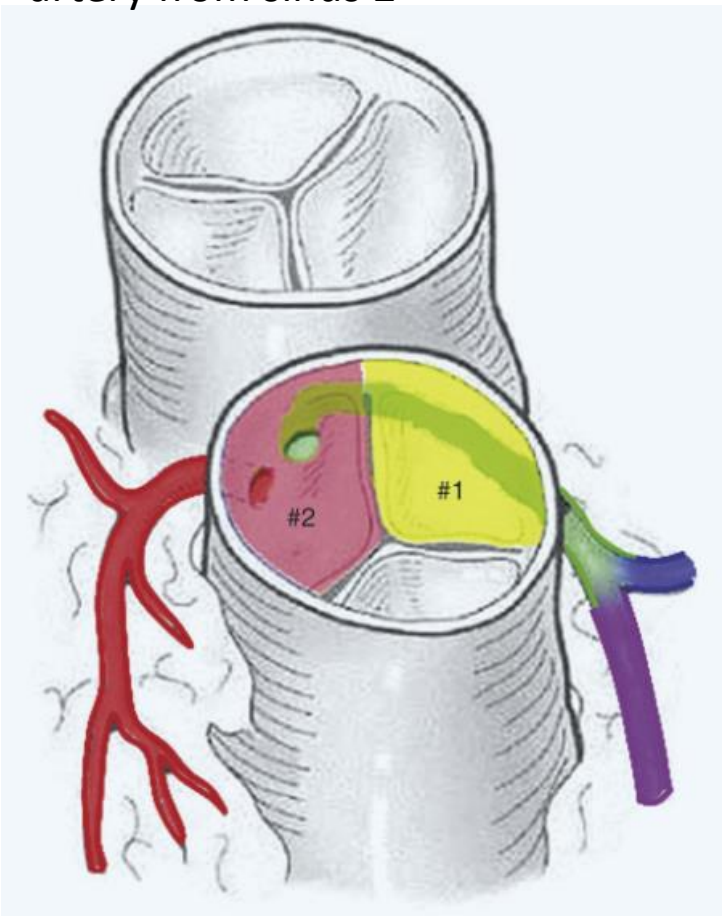
Anatomy and morphogenesis

- Coronary arteries

Normal coronary arteries

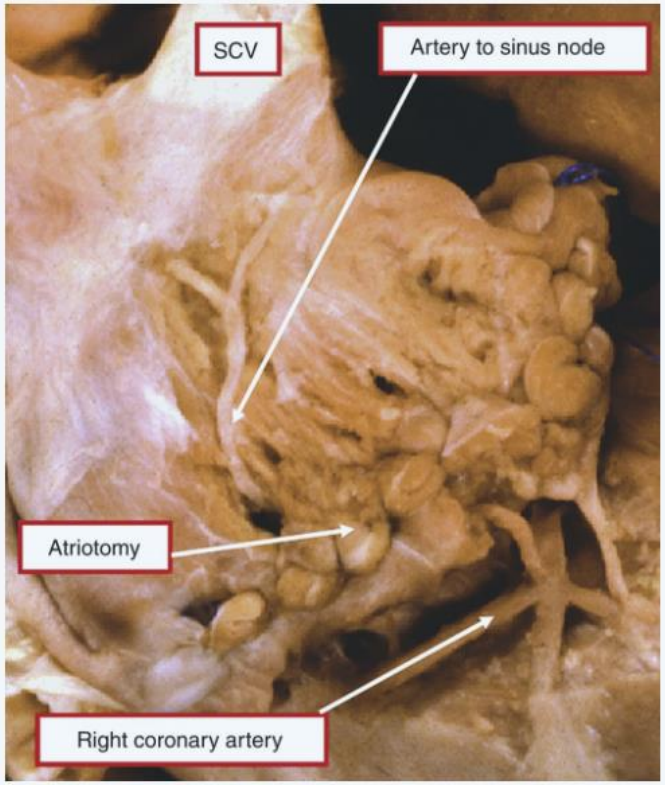


Intramural course of left coronary artery from sinus 2



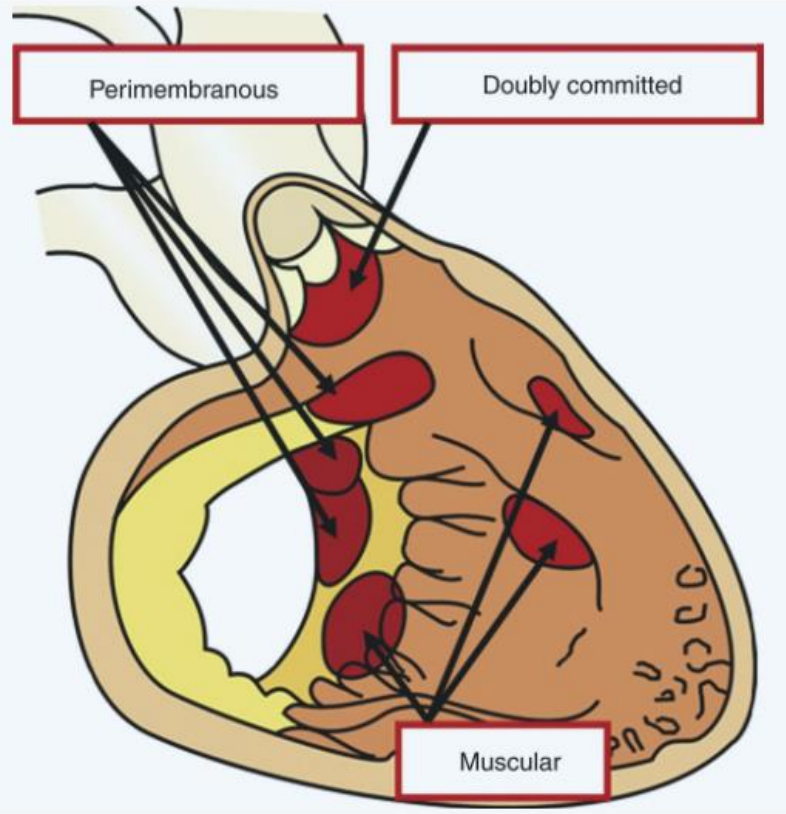
Anatomy and morphogenesis

- Coronary arteries
- Epicardial course
- Retropulmonary
- Antero-aortic position
- The artery to the sinus node
- Crosses the lateral margin of the right atrial appendage -> at surgical risk during a standard atriotomy



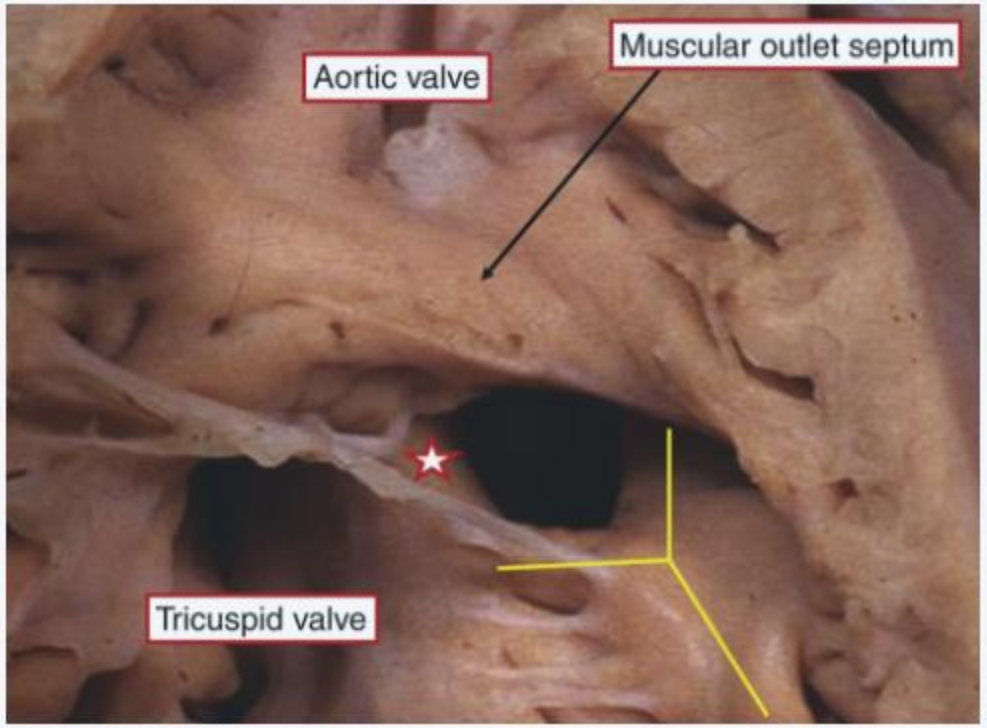
Anatomy and morphogenesis

- Ventricular septal defect
- Can be located within any part of the ventricular septum



Anatomy and morphogenesis

- Ventricular septal defect
- Outlet septum, positioned within the right ventricle
- Pulmonary valve, overrides the septum
- -> greater degrees of overrding
- -> DORV with subpulmonary VSD (Taussig-Bing anomaly)

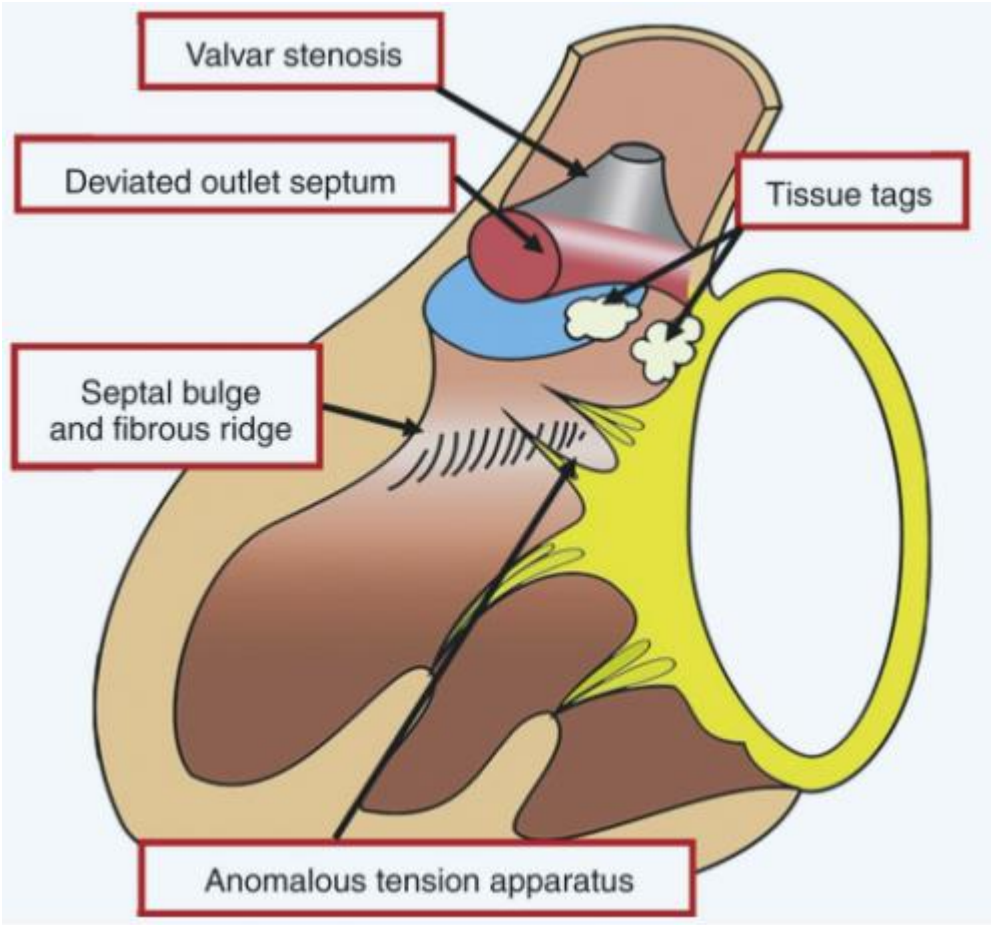


Anatomy and morphogenesis

- Ventricular septal defect
- The defect extends to open into the right ventricular inlet, potential for straddling and overriding of the tricuspid valve
- Other types of defect
  - Multiple muscular defects
  - Solitary apical muscular defects
  - Doubly committed defects

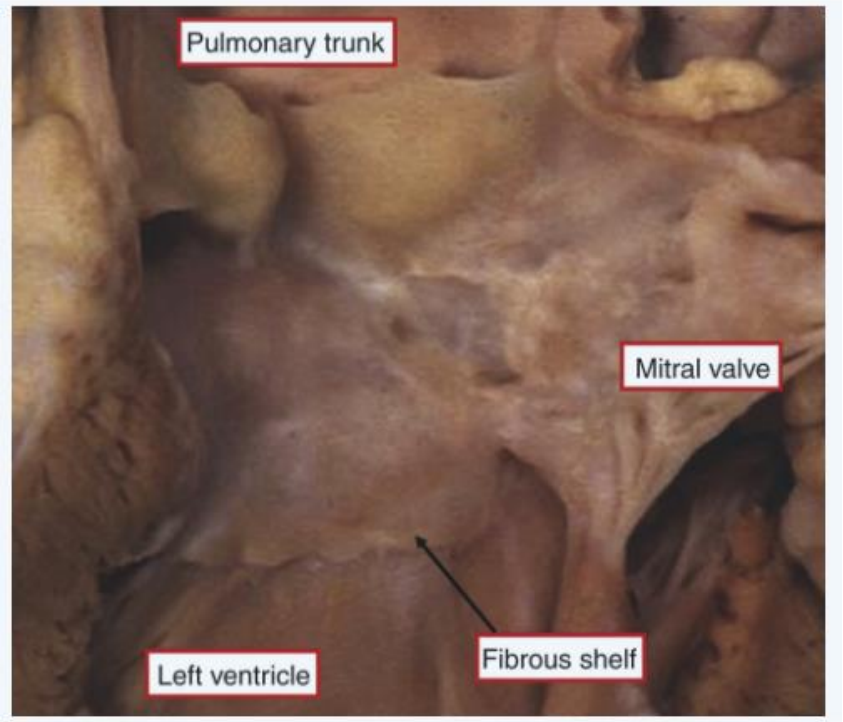
Anatomy and morphogenesis

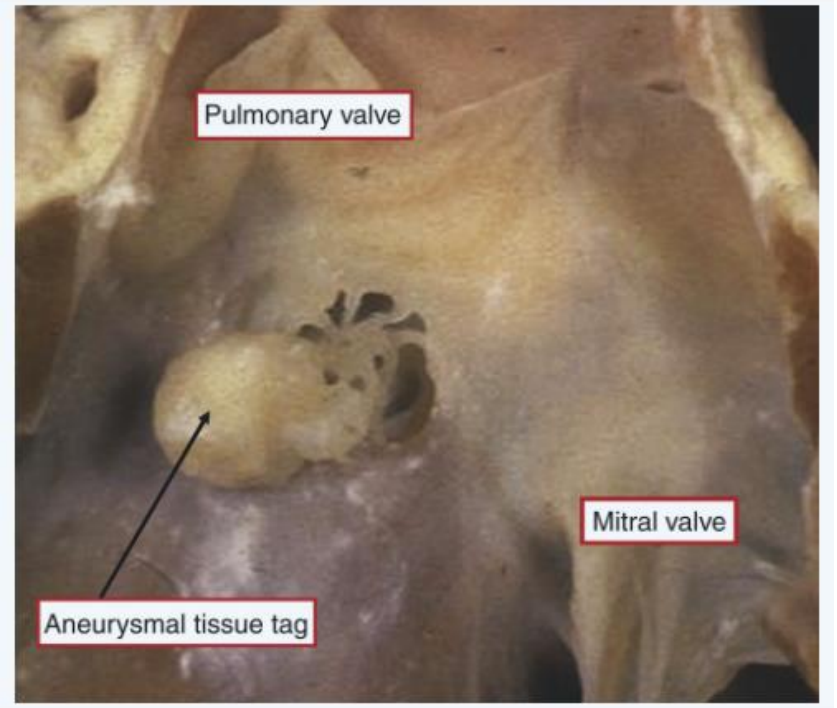
- Obstruction of the left ventricular outflow tract



Anatomy and morphogenesis

- Obstruction of the left ventricular outflow tract
- All can exist with an intact ventricular septum, or in association with a ventricular septal defect





Anatomy and morphogenesis

- Associated malformations
  - PDA
  - Stenosis of the subaortic outflow tract
  - CoA
  - Anomalous pulmonary venous connections

Anatomy and morphogenesis

- Morphogenesis
- The first suggests that the anomaly is the consequence of inappropriate separation of the arterial pole of the heart
- The second theory puts the seat of maldevelopment not in the arterial trunks, but in the ventricular outflow tracts

Pathophysiology

- Circulatory physiology
- In transposition, as two separate and parallel circuits

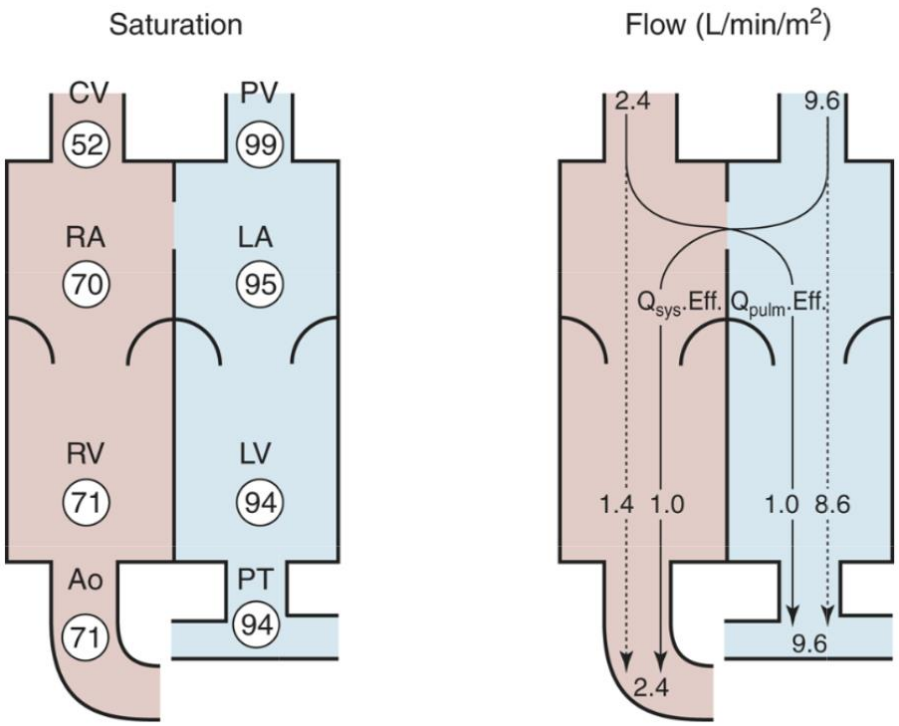


Figure 38-21

Saturations (left hand panel) and flows (right hand panel) within the heart of a patient with transposition of the great arteries and atrial mixing. Effective pulmonary (Qpulm.Eff.) and systemic (Qsys.Eff.) flows are equal (1 L/min/m2), resulting from the bidirectional shunt at atrial level. However, total systemic (2.4 L/min/m2) and total pulmonary (9.6 L/min/m2) differ considerably such that the pulmonary-to-systemic ratio is 4 to 1. Ao, aorta; CV, caval veins; LA, left atrium; LV, left ventricle; PT, pulmonary trunk; PV, pulmonary veins; RA, right atrium; RV, right ventricle.

Pathophysiology

- Determinants of systemic arterial oxygenation and mixing
- The level of systemic arterial saturation is determined by the effective systemic flow, which is proportionate to the degree of circulatory mixing
- At the level of the arterial duct, the direction of shunting largely depends upon the pulmonary vascular resistance
- High pulmonary resistance, commonly associated with a restrictive interatrial communication -> urgent decompression with an atrial septostomy

Pathophysiology

- Ventricular septal defect
- Predispose to early pulmonary vascular disease -> generally evident by 6-12 months of age
- Subpulmonary obstruction, modifies and limits the flow of blood to the lungs -> asymptomatic, protected pulmonary circulation, adequate mixing, and a well-trained left ventricle

Pathophysiology

- Growth and development during fetal life and infancy
- Smaller occipito-frontal circumferences at birth than control
- Majority of children, caught up with their peers by 2 years of age

Clinical diagnosis

- Presentation
- Antenatal diagnosis -> assess the full anatomy -> immediate postnatal initiation of PGE1 -> further intervention with a septostomy
- Cyanosis and circulatory collapse shortly after birth -> urgent balloon atrial septostomy
- Infants with a widely patent arterial duct, and a large unrestrictive VSD -> minimal cyanosis, symptomatic pulmonary over-circulation

Clinical diagnosis

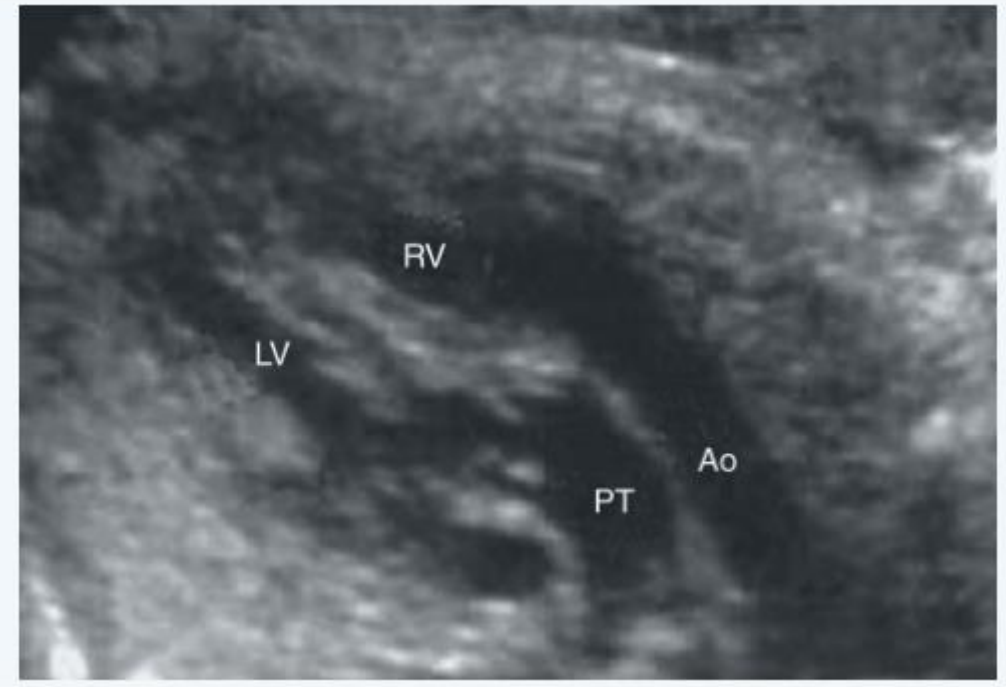
- Chest radiography
- In around 1/3 of neonates, narrow mediastinum, AP relationship of great arteries

Clinical diagnosis

- Electrocardiography
- Normal
- May be RV hypertrophy, right-axis deviation

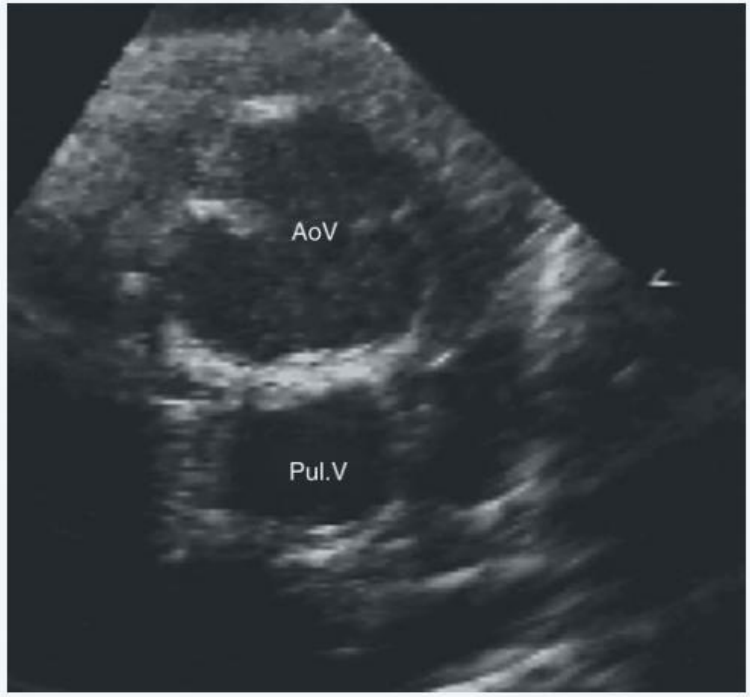
Clinical diagnosis

- Fetal echocardiography
- Optimal neonatal care and pre-operative management



Clinical diagnosis

- Postnatal echocardiography
- Typically, the aortic valve is positioned anterior and to the right of the pulmonary valve



Clinical diagnosis

- Postnatal echocardiography
- The left ventricular outflow tract, always be carefully examined to exclude any obstruction
- The atrioventricular valves, should be assessed carefully to identify any abnormal attachments of the tendinous cords
- The origin and course of the coronary arteries
- The aortic arch, CoA or interruption
- Difficult to assess, after prostaglandin is discontinued

Clinical diagnosis

- Postnatal echocardiography
- ASD
- PDA

Clinical diagnosis

- Cardiac catheterization
- No longer routinely performed

Clinical diagnosis

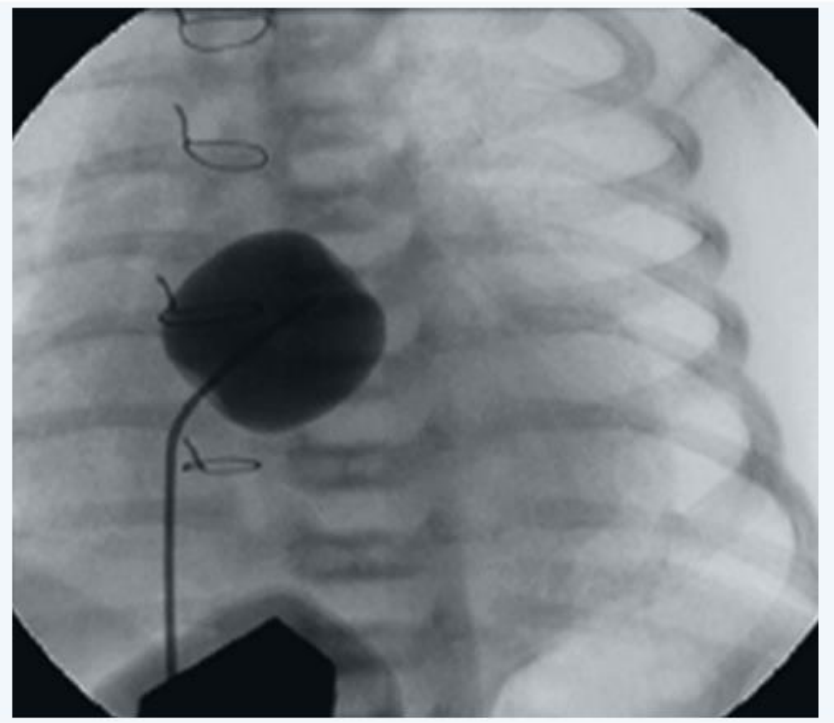
- Magnetic resonance imaging
- CT scaning

Medical management of neonates

- Postnatal stabilization
- Neonates diagnosed antenatally should ideally be delivered in a high-risk obstetric unit, with rapid access to cardiac care
- IV PGE1
- Patients with severe acidosis or hypoxemia -> immediate balloon atrial septostomy
- Inadequate intracardiac mixing and significant cyanosis -> septostomy

Medical management of neonates

- Atrial septostomy
- Introduction by Rashkind and Miller in 1966



Surgical management

- Historical perspective
- The improved survival in infancy after surgical intervention can largely be ascribed to the advent of the balloon atrial septostomy

Surgical management

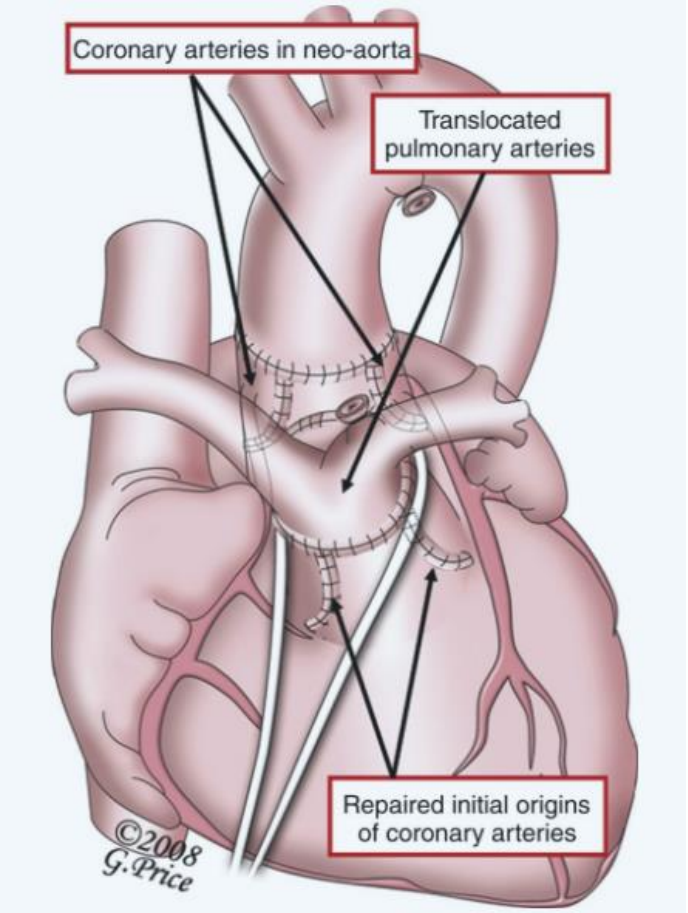
- Atrial redirection procedures
- Physiological correction
- Late complications
  - Baffle obstruction
  - Arrhythmias
  - Ventricular dysfunction
  - End-stage heart failure

Surgical management

- Mustard and Senning procedures
- Senning operation, rerouting of systemic venous blood -> achieved by means of infolding of the atrial walls
- Mustard operation, achieved using synthetic or pericardial tissue

Surgical management

- Arterial switch operation
- The first successful arterial switch operation, by Jatene and colleagues in 1975, in an older infant with an associated VSD
- De-conditioned LV, two-stage repair -> PA banding (+ systemic-to-pulmonary arterial shunting)
- Lecompte maneuver, in 1981
- The first report of successful correction of a neonate with an intact ventricular septum, in 1984
- Early experience, VSD, coronary arterial pattern, aortic arch obstruction -> impact on mortality -> these effects have decreased over time



Surgical management

- Timing of the arterial switch operation
- TGA with IVS, undertaken towards the end of the first week of life
- To avoid the deconditioning of the morphological left ventricle
- Deconditioning, arrested or slowed by
  - VSD
  - Large PDA
  - LVOTO

Surgical management

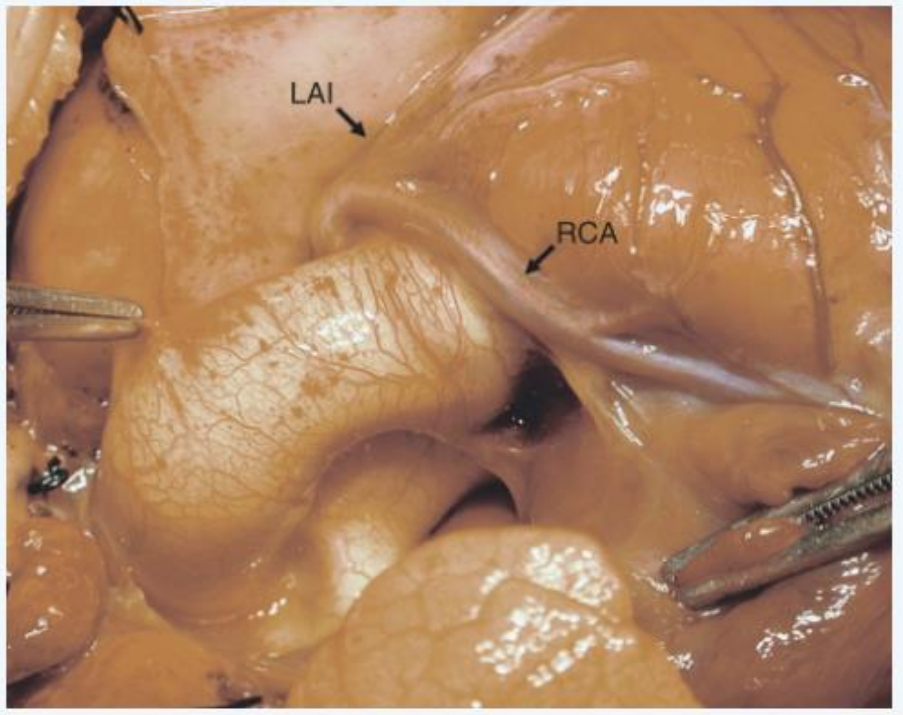
- Timing of the arterial switch operation
- By the Congenital Heart Surgeons Society in 1988, only 14 days, in TGA with IVS
- -> primary repair can be undertaken at up to 2 months of age
- -> in infants at even up to 6 months, but need for temporary mechanical support

Surgical management

- Arterial switch operation
- Ventricular training
- The potential for LV to support the systemic circulation
- PA banding (+ S to P shunt)
- At high risk patients, ASO as the primary procedure -> elective use of a left ventricular assist device immediately after CBP

Surgical management

- Arterial switch operation
- Coronary arterial anatomy
- LAD or LCx arising from sinus 2
- Intramural coronary artery
- Single origin of the coronary arteries



Surgical management

- Surgery for complex transposition
- Transposition with a ventricular septal defect
- Did not affect the outcome for the arterial switch operation in current era

Surgical management

- Surgery for complex transposition
- Surgery in infants with significant pulmonary hypertension
- Pulmonary vascular disease, develops rapidly in TGA with VSD
- PVR > 8 Wood units, insuitable
- Palliative atrial redirection
- Palliative arterial switch

Surgical management

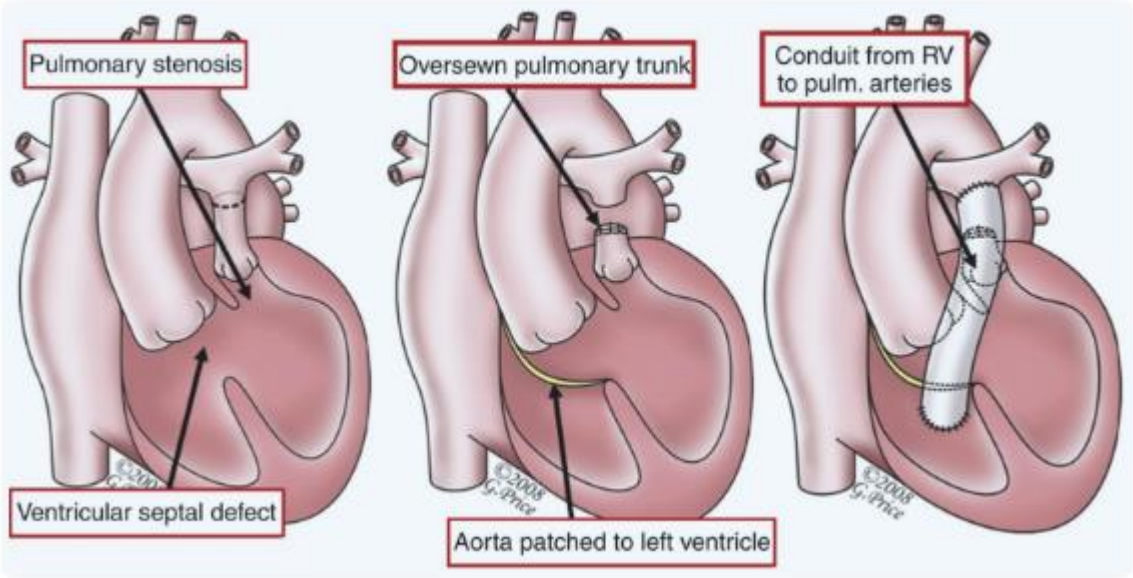
- Surgery for complex transposition
- Transposition with coarctation of the aorta of interruption of the aortic arch
- More common in those with VSD, when it is often associated with anterior deviation of the outlet septum
- Generally accepted that a single-stage operation should be performed
- Augmentation with a patch may occasionally be required

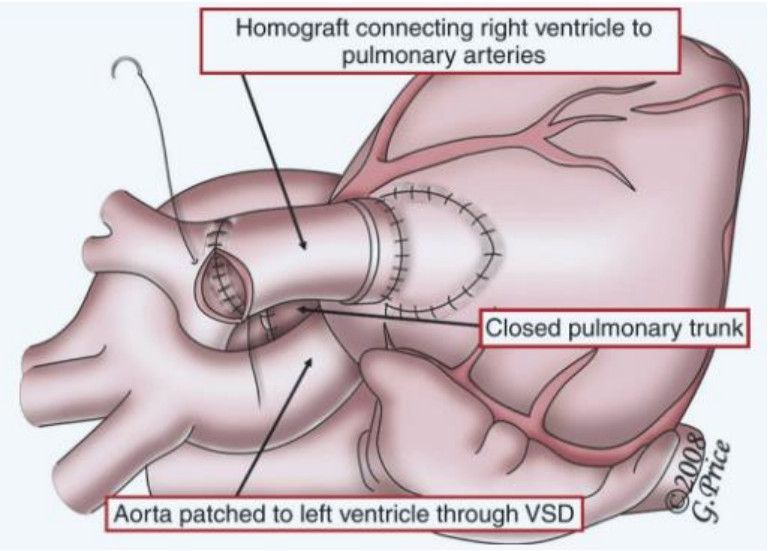
Surgical management

- Surgery for complex transposition
- Surgical options in the presence of obstruction to the left ventricular outflow tract
- LVOTO, most commonly associated with VSD
- Minor degrees, resected at the time of ASO
- Severe or complex forms, to consider an alternative procedure

Surgical management

- Rastelli procedure

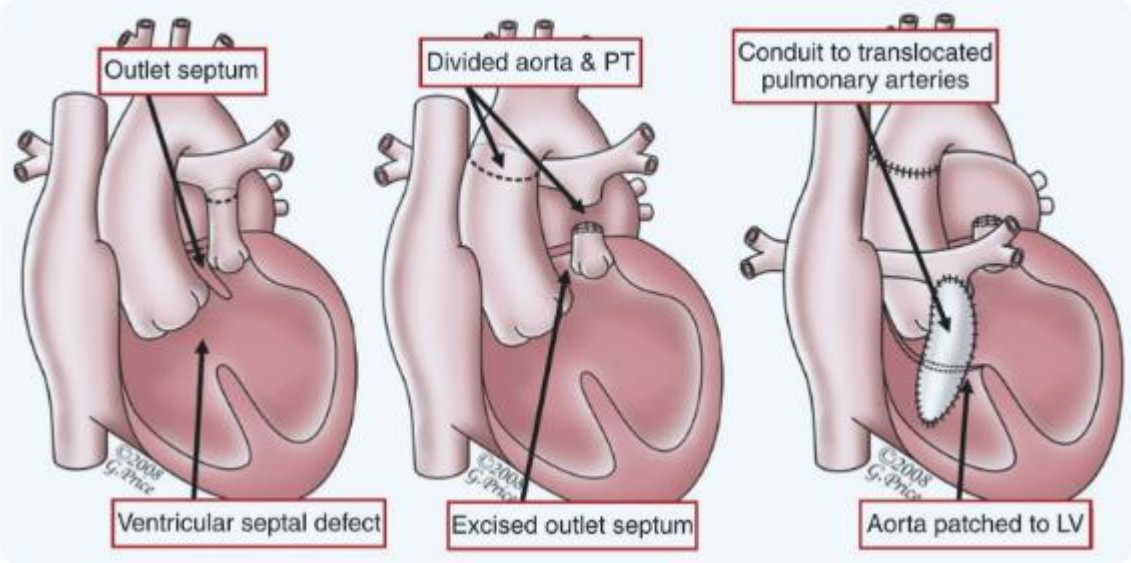




Postop. LVOTO
RV-PA conduit replacement

Surgical management

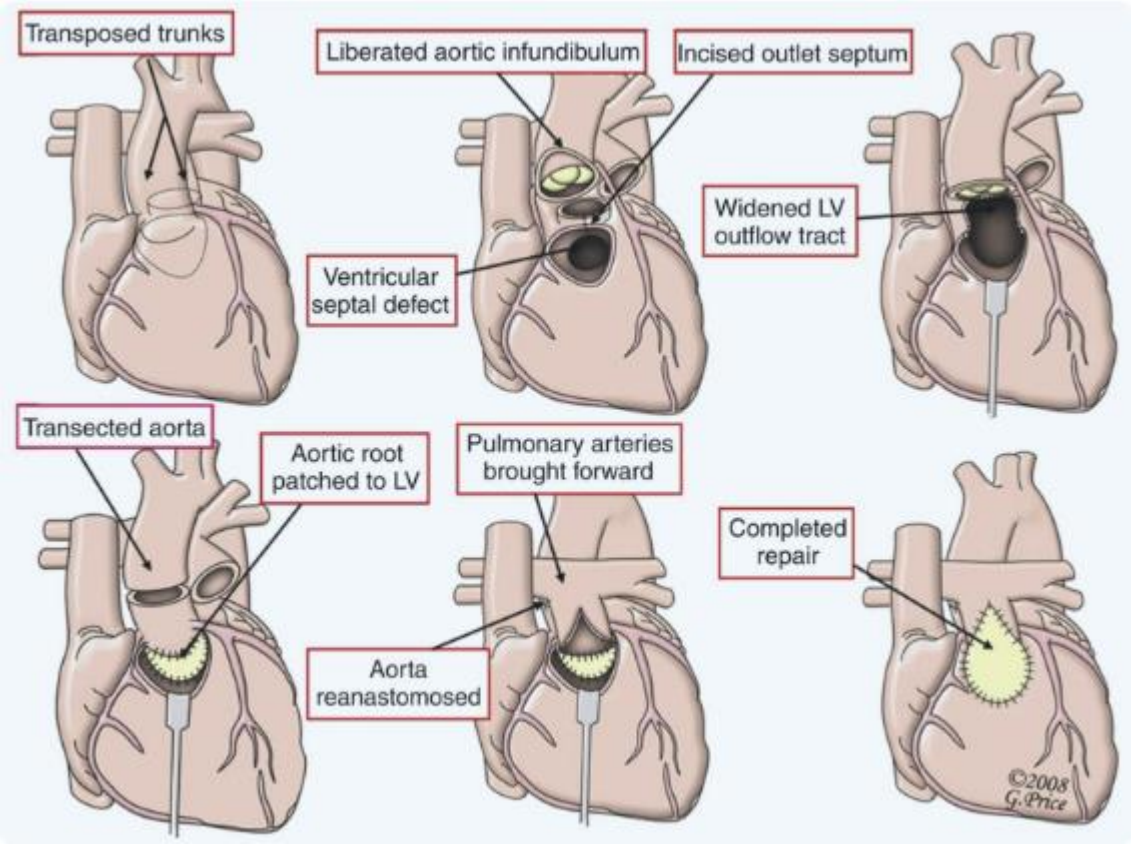
- Reparation a l'Etage Ventriculaire, or REV procedure



Post-operative RVOTO

Surgical management

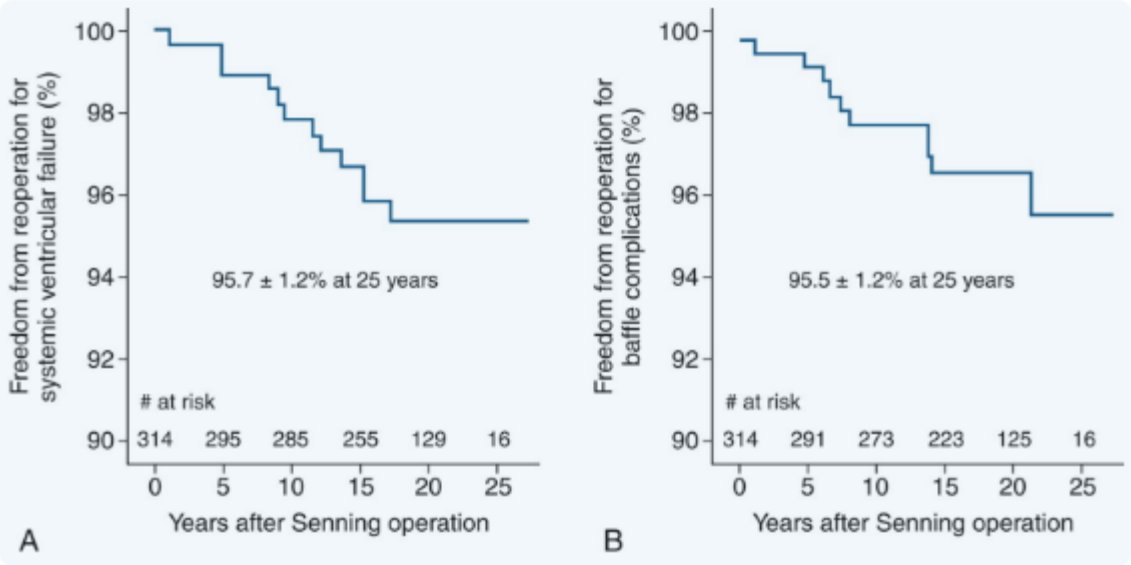
- Nikaidoh procedure, or aortic translocation



Advantage, systemic outflow is not directed through an intraventricular baffle -> lower risk of obstruction developing in the LVOT

Surgical outcomes

- Atrial redirection procedures



Main indicaions for reop.
1) Obstruction of the venous baffles
2) RV failure
3) TR
4) LVOTO

Atrial bradyarrhythmias and tachyarrhythmias

Surgical outcomes

- Late outcome after the arterial switch procedure
- General health, quality of life, and neurodevelopmental outcome
- Good overall health and quality of life, not significantly different from their peers
- Frequently perform below expectation in
  - Academic achievement
  - Fine motor function
  - Visual-spatial skills
  - Sustained attention

Surgical outcomes

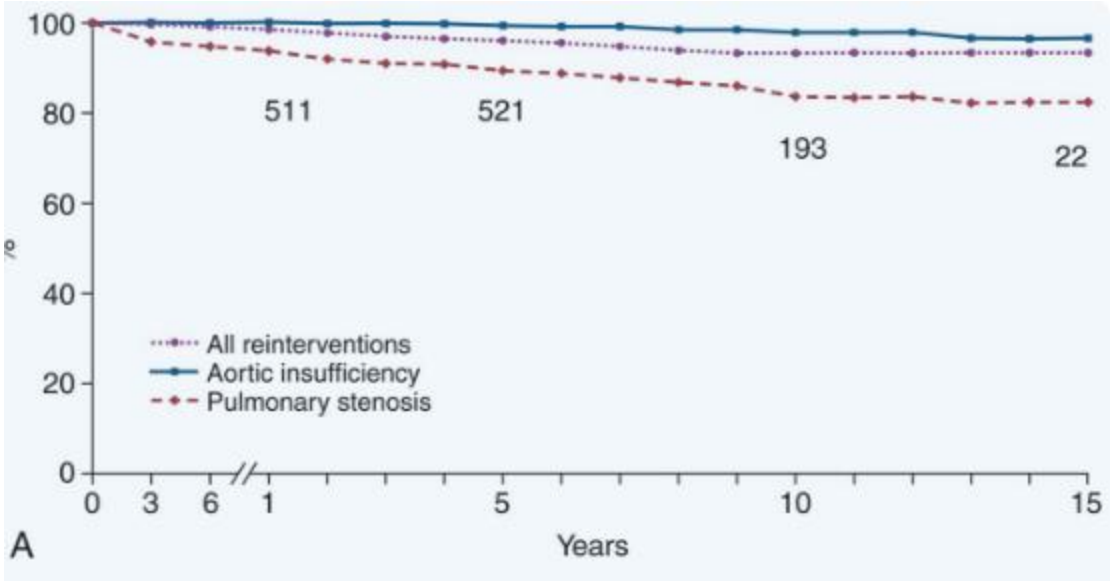
- Late outcome after the arterial switch procedure
- Fate of the coronary arteries
- Coronary arterial problems, 1/10
- 1/3 of late deaths
- Potential mechanisms
  - Anatomical distortion
  - Extrinsic compression
  - Stretching
  - Intimal proliferation

Surgical outcomes

- Late outcome after the arterial switch procedure
- Fate of the neo-aortic root and valve
- 2/5, AR, 20 months f/u
- ½, AR, 10 years f/u
- Ascending aorta dilation
  - AR
  - Abnormal angulation of the aortic arch

Surgical outcomes

- Late outcome after the arterial switch procedure
- Pulmonary arteries
- Obstruction within the pulmonary outflow tract, the most frequent residual anatomical problem
- The most frequent indication for reoperation



Recommendations for long-term follow-up

TABLE 38-1 Protocols Developed by the Canadian Adult Congenital Heart Network for the Assessment of Adults with Transposed Arterial Trunks

Modified from Canadian Adult Congenital Heart Network: Complete transposition. Available at: www.cachnet.org.

All patients should have at a minimum • A thorough clinical assessment • An electrocardiogram • A chest radiograph • Oximetry at rest and possibly with exercise
Patients who have had an atrial redirection procedure also require • An echocardiogram to detect baffle obstruction or leak, to detect regurgitation of the atrioventricular valve, to assess the function of the systemic ventricle, and to detect subpulmonary obstruction • A Holter monitor because of the high prevalence of sick sinus syndrome and atrial arrhythmias and possible ventricular arrhythmias in older patients and may require • A transoesophageal echocardiogram if there is inadequate visualisation of the intra-atrial baffle on the transthoracic study • Radionuclide assessment of myocardial perfusion (if ischaemia is suspected), or of ventricular function • Magnetic resonance imaging to evaluate the baffle for obstruction or leak and ventricular volumes, shapes, and function • Cardiac catheterisation including coronary angiography if there are doubts about additional lesions, and if surgical reintervention is planned, or if adequate assessment of the haemodynamics is not obtained by non-invasive means • Exercise testing to evaluate functional capacity, including heart rate and blood pressure measurement, and to assess whether arrhythmias may be provoked
Patients who have had an arterial switch operation also require • An echocardiogram to assess obstruction to the right ventricular outflow tract, ventricular function, neo-aortic root dilation, and regurgitation of the aortic valve and the coronary arterial orifices • Exercise testing for possible coronary ischaemia and may require • Holter monitoring if arrhythmia is suspected • Nuclear assessment of myocardial perfusion periodically • Coronary arteriography if ischaemia is documented on non-invasive testing • Cardiac catheterisation if adequate assessment of the haemodynamics is not obtained by non-invasive means or additional lesions are suspected • Magnetic resonance imaging to exclude obstruction of the right ventricular outflow tract

References

- Paediatric cardiology, third edition, by Anderson