



Wilhelmina Kinderziekenhuis



Aangeboren hartafwijkingen

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Verantwoording

- Dank aan:
 - Sprekers, Mayo Clinic
 - Cursus: Echocardiography in Pediatric and Adult Congenital Heart Disease 2015, Phoenix, Arizona
- Illustrated Field Guide to Congenital Heart Disease and Repair, 3d edition



Kindercardiologie in NL

- ♥ geboortecijfer 2014: 175.181 (CBS)
- ♥ 0.8% aangeboren hartafwijkingen
 - ongeveer 1400 kinderen per jaar
- ♥ 1/3: geringe afwijking
- ♥ 1/3: matige afwijking
 - behoeft of eenvoudige ingreep of medicamenteuze ondersteuning
- ♥ 1/3: ernstige afwijking
 - alleen palliatieve of risicovolle chirurgie, letaal



Relatieve frequentie aangeboren hartafwijkingen

♥ VSD	<i>ventrikelseptumdefect</i>	20%
♥ ASD	<i>atriumseptumdefect</i>	10%
♥ PDA	<i>persisterende ductus arteriosus</i>	10%
♥ CoA	<i>coarctatio aortae</i>	10%
♥ PS	<i>pulmonaalklepstenose</i>	10%
♥ AS	<i>aortaklepstenose</i>	10%
♥ AVSD	<i>atrioventriculair septumdefect</i>	2-5%



Relatieve frequentie aangeboren hartafwijkingen

♥TOF	<i>tetralogie van Fallot</i>	10%
♥TGA	<i>transpositie van de grote arteriën</i>	5-8%
♥PA	<i>pulmonaalklepatresie</i>	5%
♥TA	<i>tricuspidaalklepatresie</i>	3%
♥PTA	<i>(persisterende) truncus arteriosus</i>	3%
♥TAPVC	<i>tot. abn. pulmonaalveneuze connectie</i>	2%
♥HLHS	<i>hypoplastisch linker hartsyndroom</i>	2%



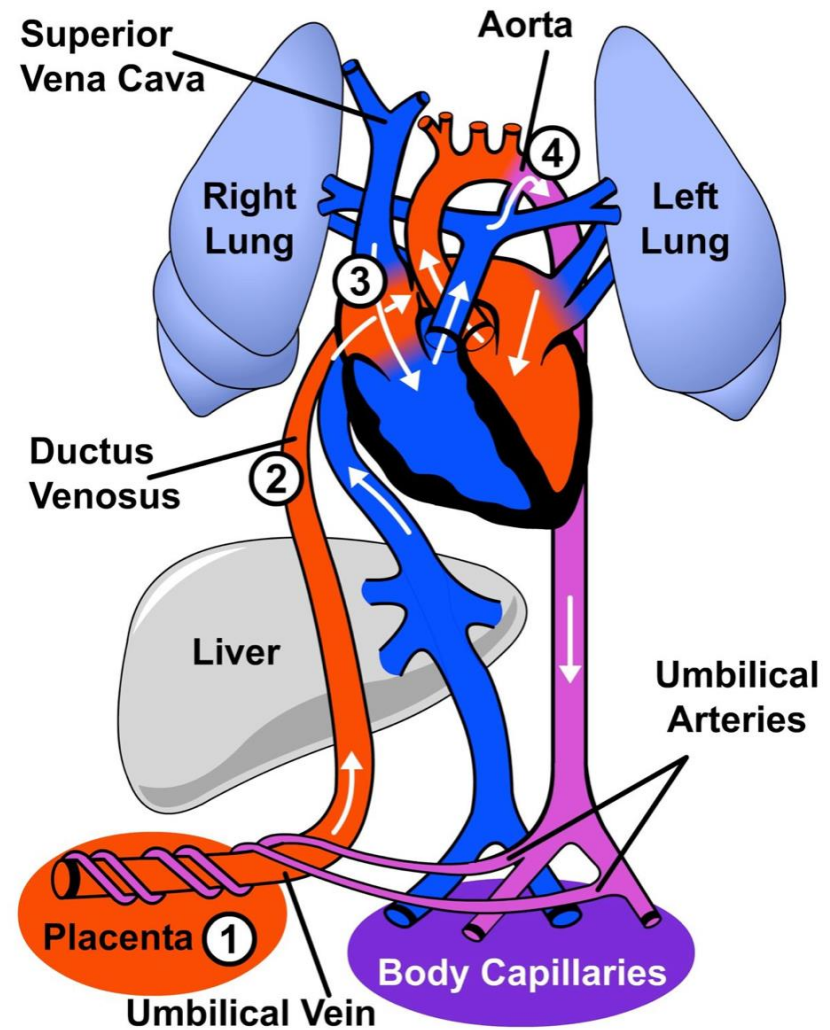
Presentatie ernstige hartafwijkingen

- binnen enkele dagen
 - ductus afhankelijk
- na 4-6 weken
 - longvaatweerstand afhankelijk



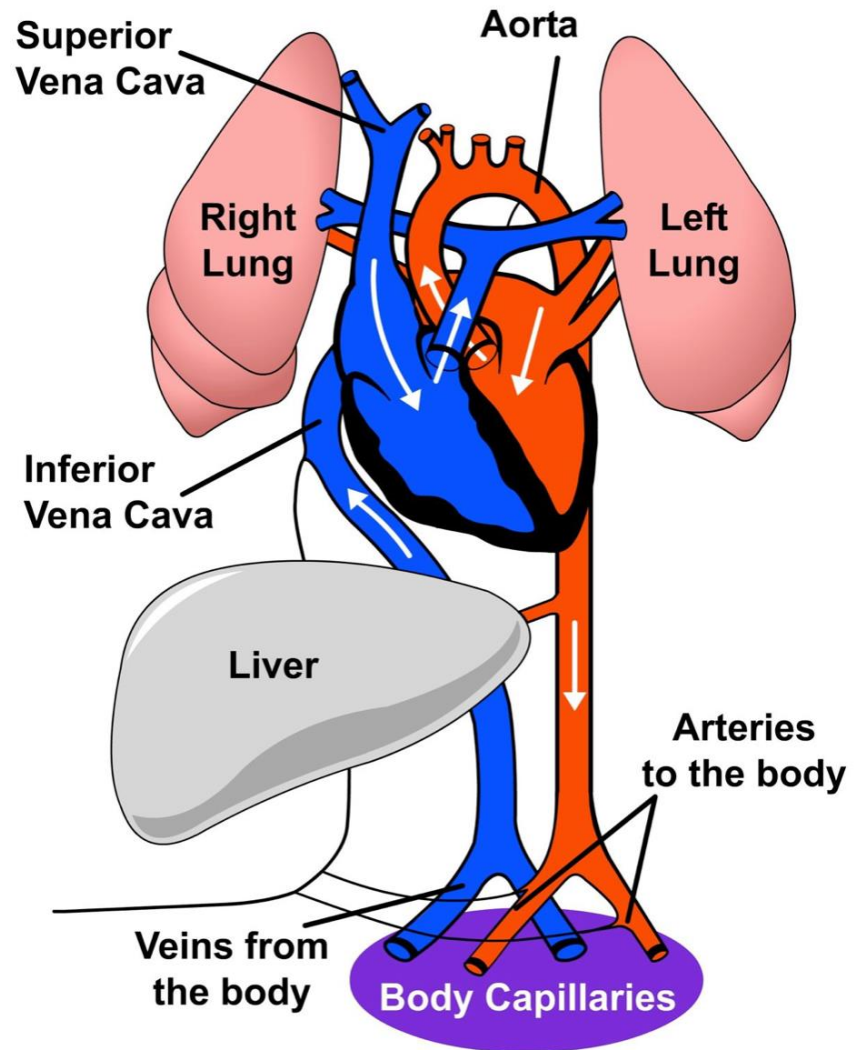
Fetale circulatie

1. Placenta/ vena umbilicalis
2. Ductus venosus
3. Foramen ovale
4. Ductus arteriosus



Postnatale circulatie

- Daling longvaatweerstand
- Stijging systeemvaatweerstand
- Functionele sluiting ductus arteriosus <24-72 uur (anatomische sluiting binnen enkele weken)



klinisch beeld en vitale parameters

♥ klinisch beeld	<i>cyanose, dyspneu, shock</i>
♥ hartactie	<i>tachycardie (... bradycardie)</i>
♥ bloedgas	<i>acidose, lactaat ++</i>
♥ thorax foto	<i>abnormaal silhouet, longvaattekening</i>
♥ ECG	<i>geleidingsstoornissen, hypertrofie, tachy-/bradyaritmie</i>
♥ SaO ₂	<i>4 extremiteiten</i>
♥ RR	<i>4 extremiteiten</i>

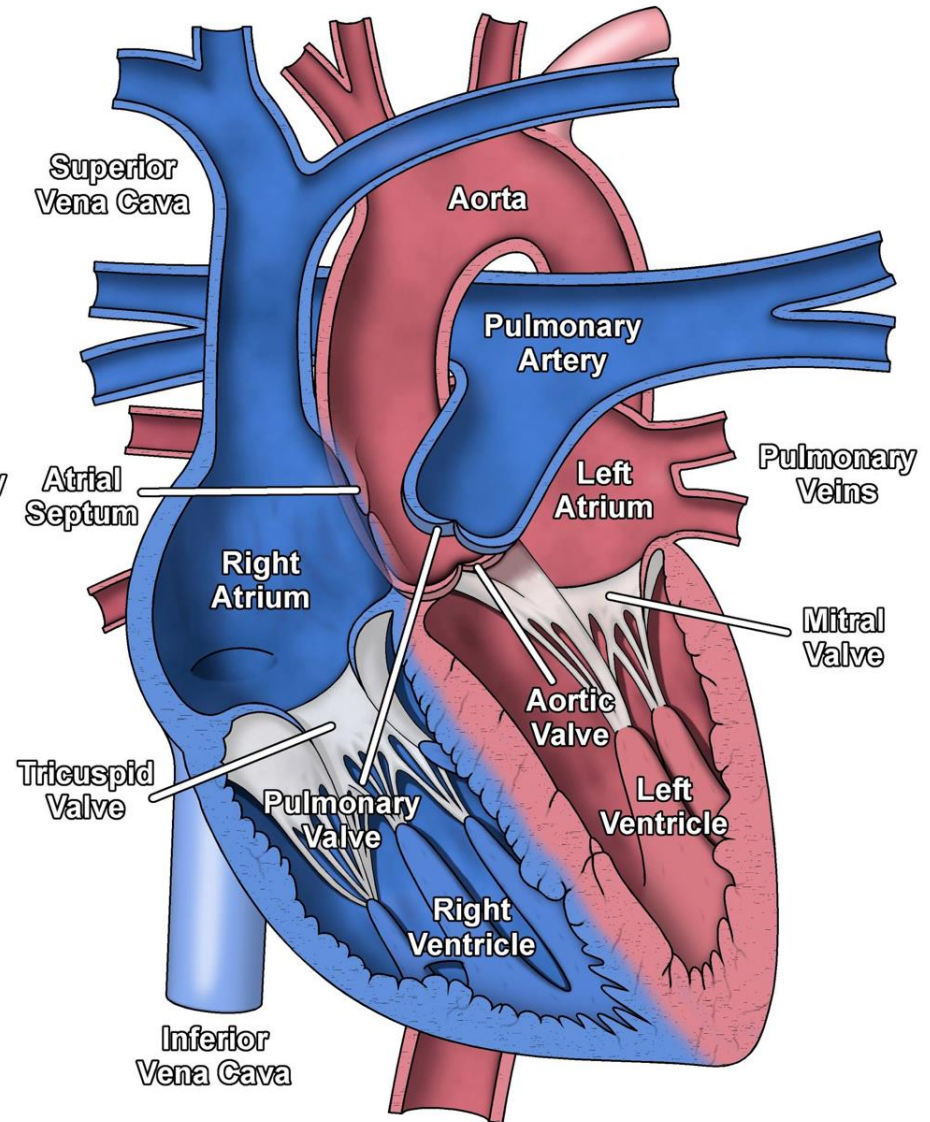
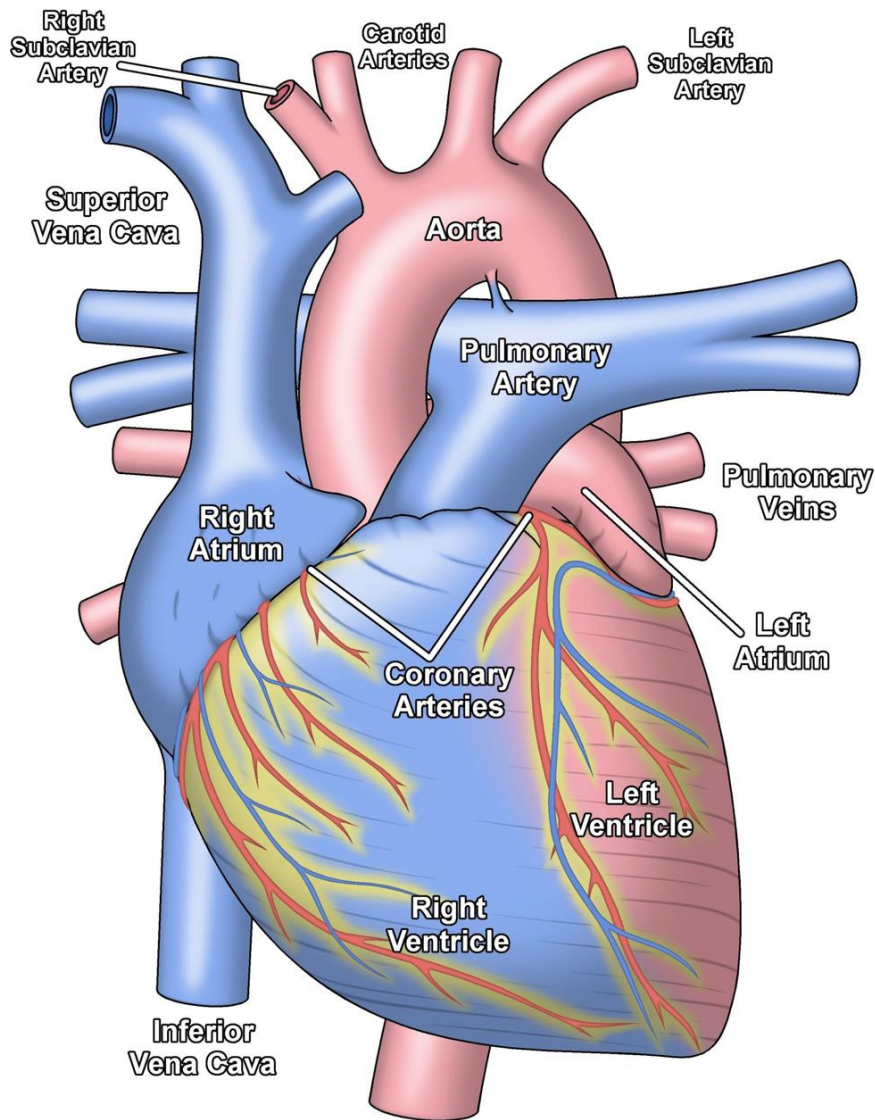


“blinde” strategie

- ♥ **Ductus openhouden !!!**
- ♥ 2 perifere veneuze lijnen
- ♥ Prostín (PGE1)(50 nanogram/kg/min)
- ♥ O₂ masker alleen als SaO₂ < 70%
- ♥ Support ademhaling (intubatie/ ventilatie)



Anatomie van het normale hart



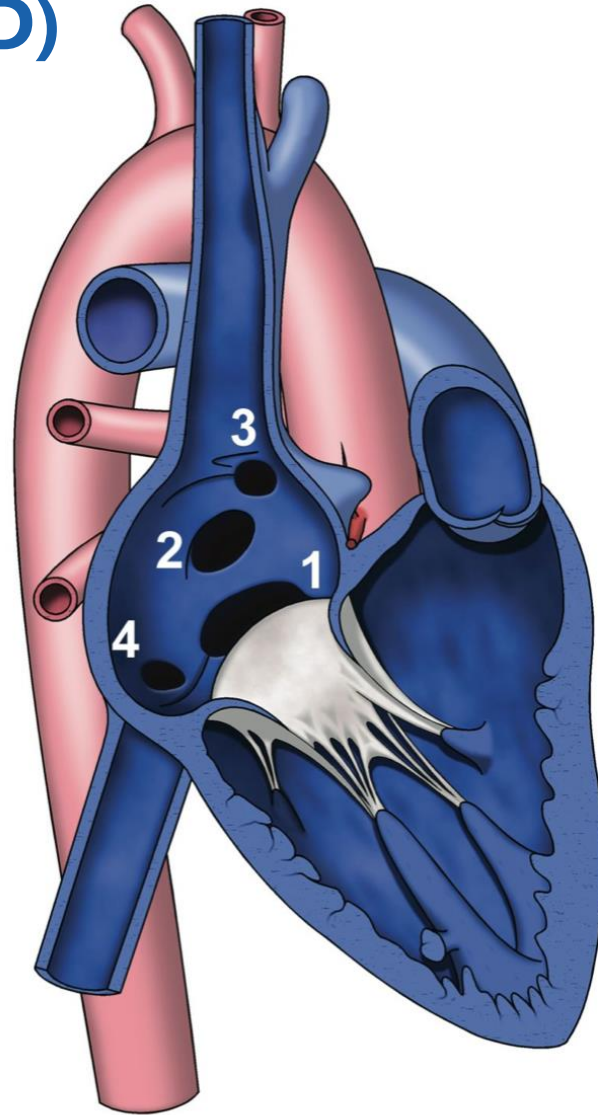
Type hartafwijking- '*shunt*' afwijkingen

- Atriumseptumdefect (ASD)
- Ventrikelseptumdefect (VSD)
- Persisterende ductus arteriosus (PDA)
- Atrioventriculair septumdefect



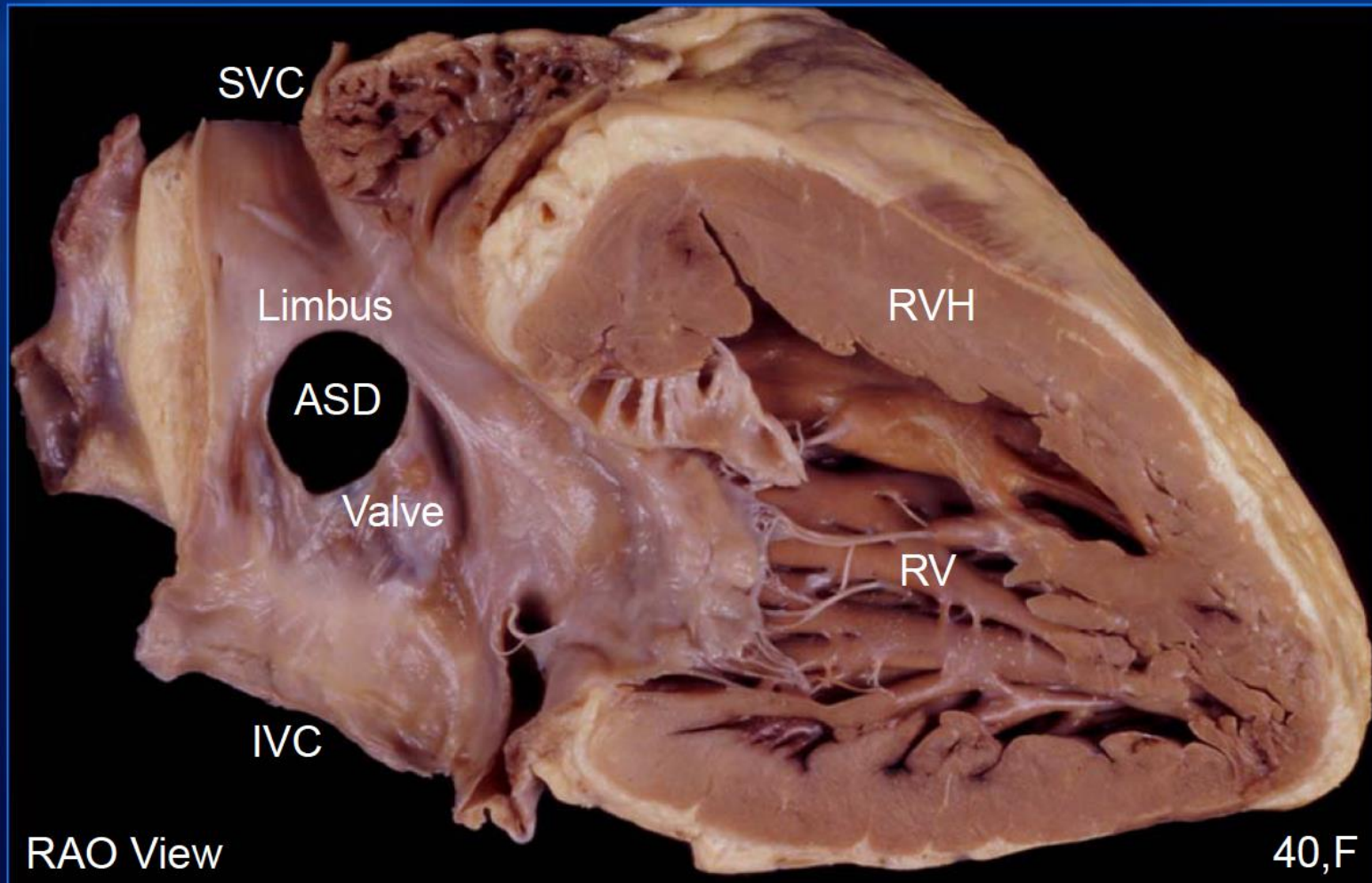
Atriumseptumdefect (ASD)

1. Primum ASD (ASD I, 20%)
2. Secundum ASD (ASD II, 70%)
3. Sinus venosus defect (10%)
 - Vaak met abnormale drainage rechter longvene naar RA
4. Sinus coronarius defect (1%)
 - Vaak met abnormale drainage van linker VCS
 - Zeer zeldzaam



Congenital Left-to-Right Shunts

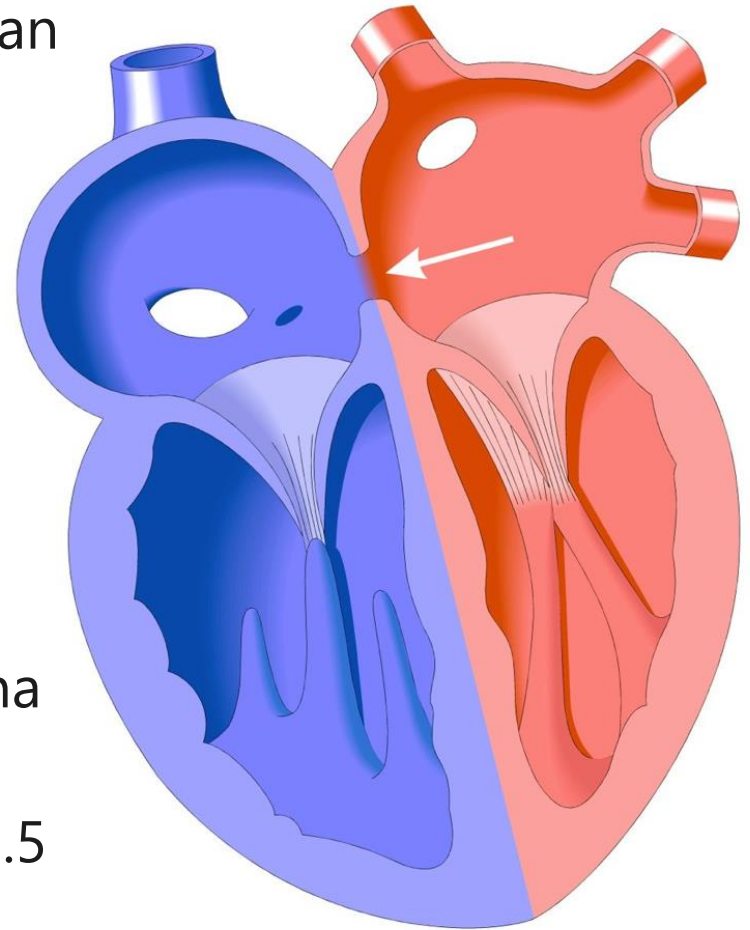
Secundum Atrial Septal Defect



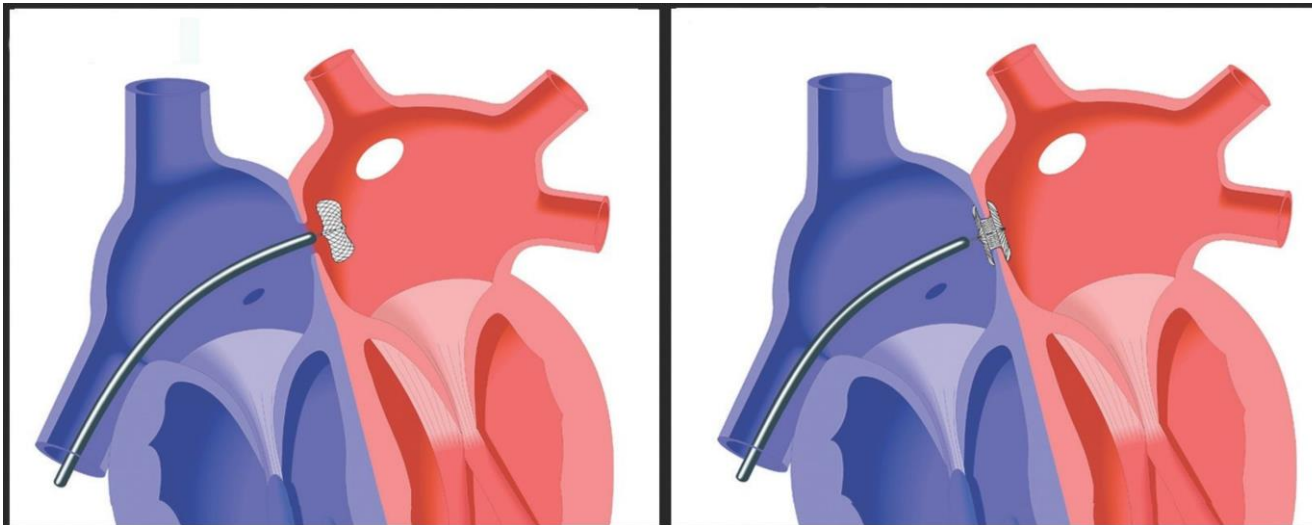
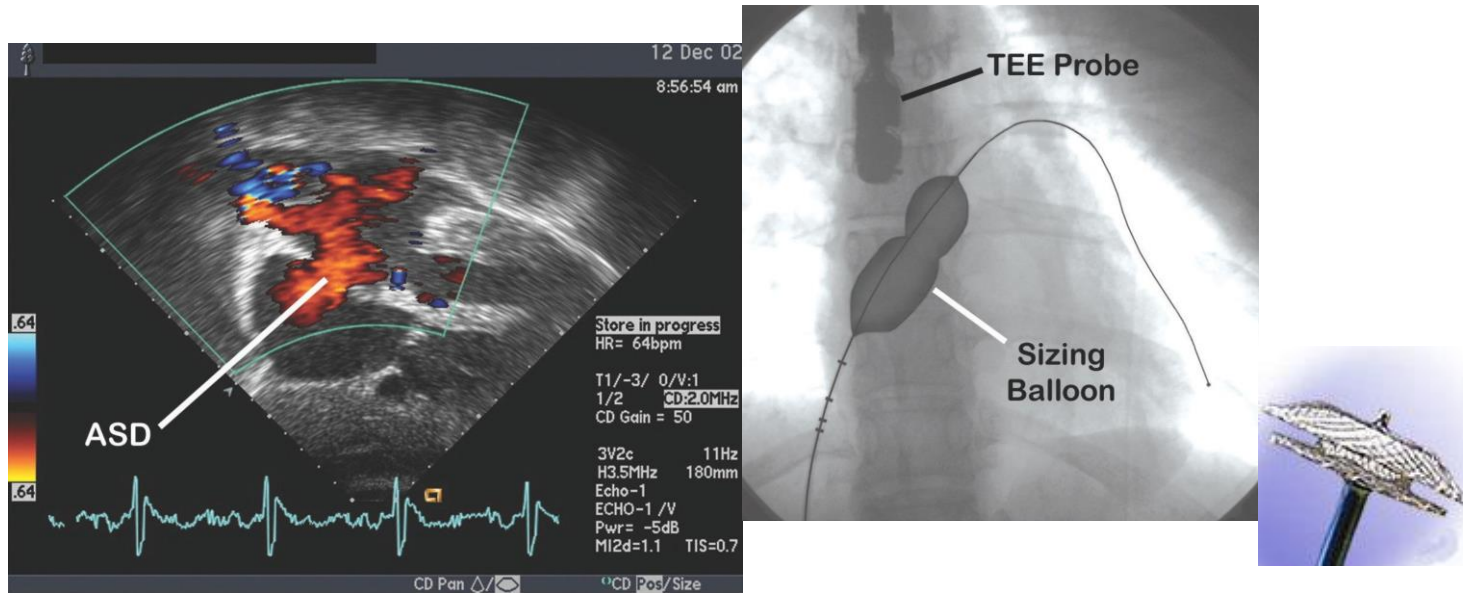
Secundum ASD (RA & RV Free Walls Removed)

Gevolgen van een ASD

- Links-rechtsshunt, afhankelijk van
 - grootte defect
 - compliance ventrikels
 - relatieve weerstand long- en lichaamscirculatie
- 'Belangrijk' vanaf 8 mm
- Shunt veroorzaakt volumebelasting RA/RV
- Meestal asymptomatisch
- Hartfalen en ritmestoornissen na 40e levensjaar
- Sluiting bij LR shunt: $Q_p:Q_s > 1.5$
 - 1e-5e levensjaar
 - interventioneel
 - chirurgisch

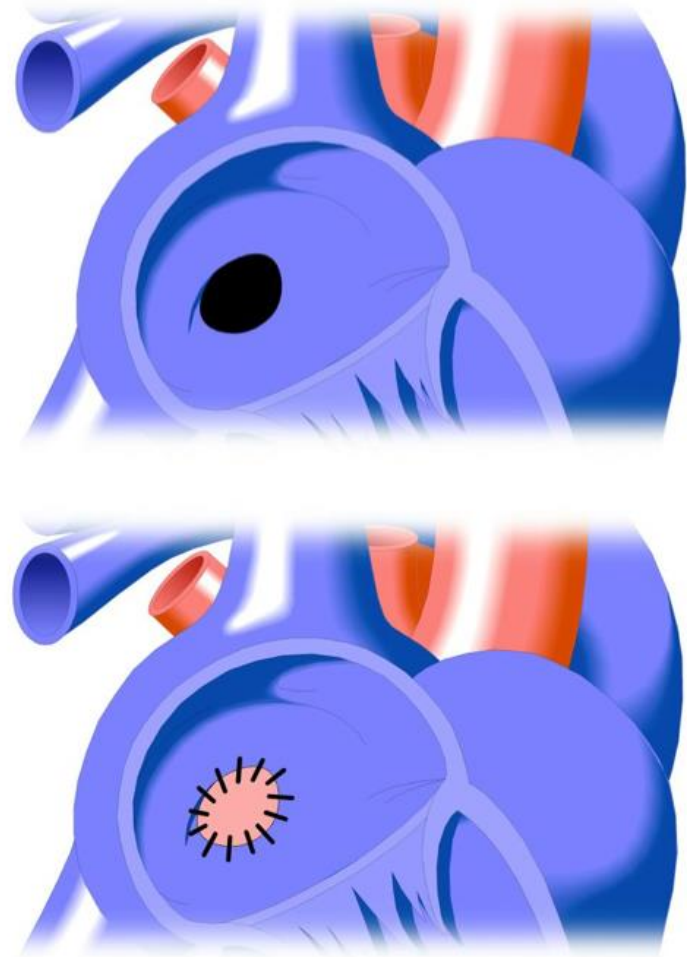


Sluiten van een ASD - *interventioneel*



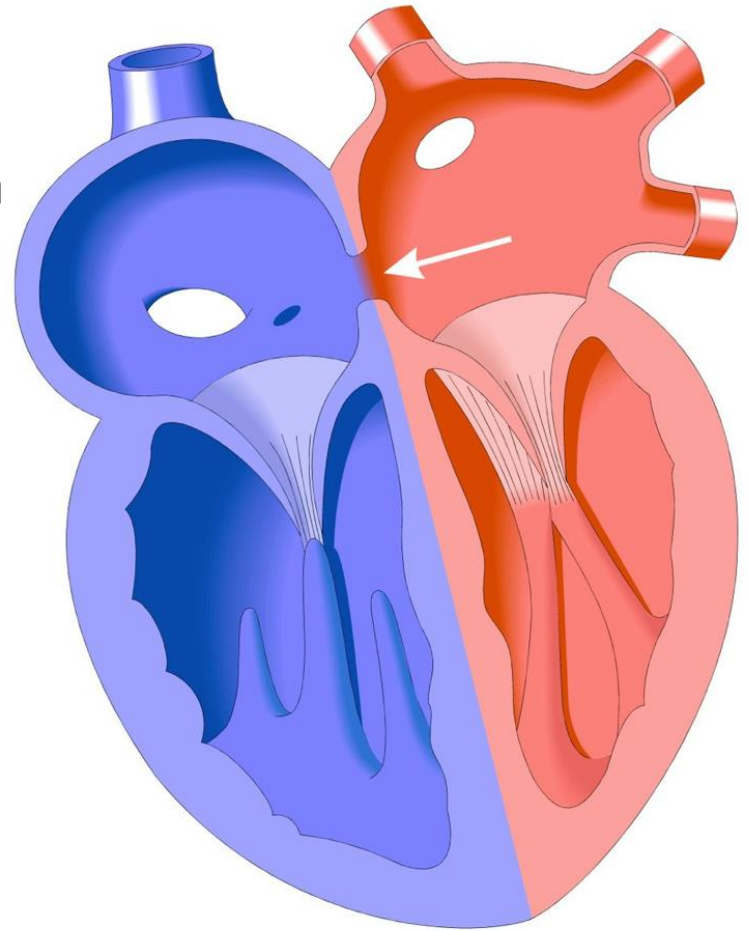
Sluiten van een ASD - *chirurgisch*

- CPB
- Patch van autoloog pericard
- Postoperatieve complicaties zeldzaam
- Mortaliteit < 1%
- Postpericardiotomie syndroom
 - malaise, koorts, thoracale pijn
 - NSAID's of prednison
 - soms: pericardpunctie/ - drainage



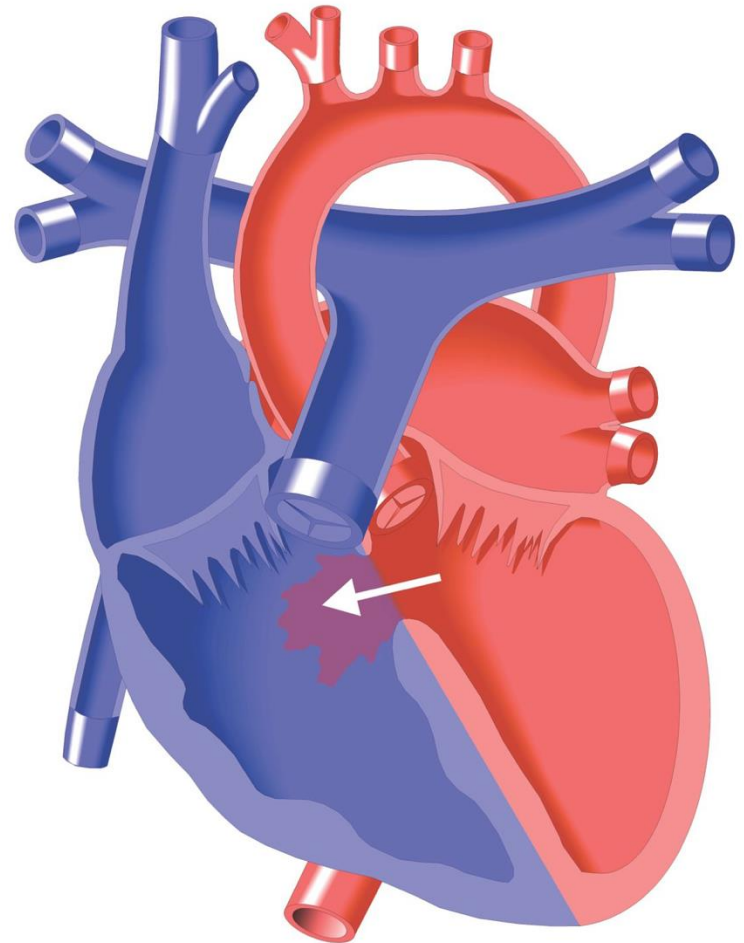
Follow-up/ prognose van een ASD

- Endocarditis profylaxe eerste 6 maanden
- Normaal inspanningsvermogen
- Normale levensverwachting



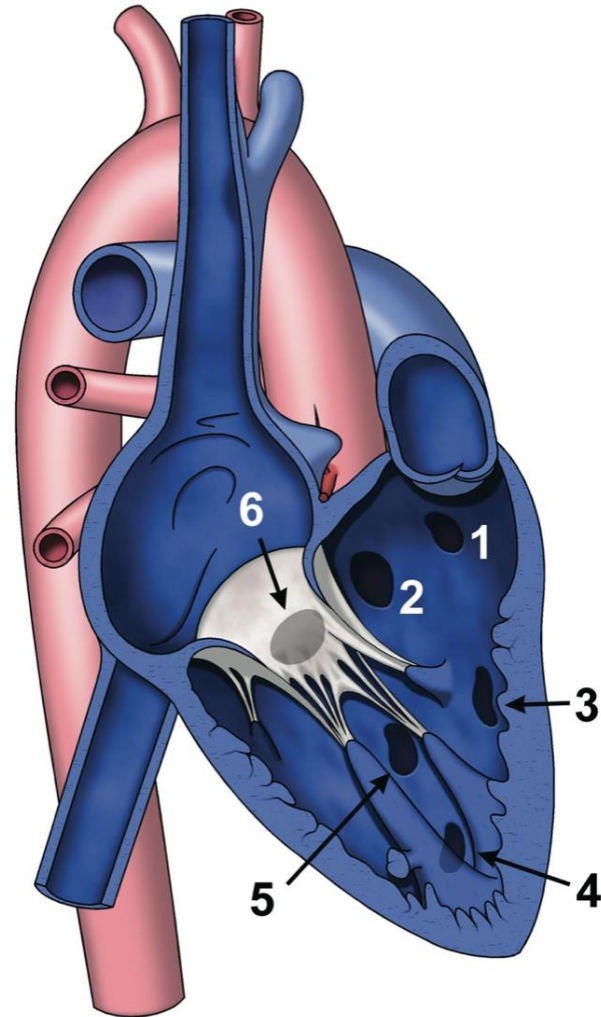
Ventrikelseptumdefect (VSD)

- Meest voorkomende hartafwijking (20-40%)
- 50% andere hartafwijking
- Tri 13/18/21, syndromen
- Kleine defecten (<5 mm) geen klachten
 - hartgeruis
- Matig grote en grote defecten
 - dyspnoe
 - afgenomen inspanningstolerantie
 - frequente luchtweginfecties



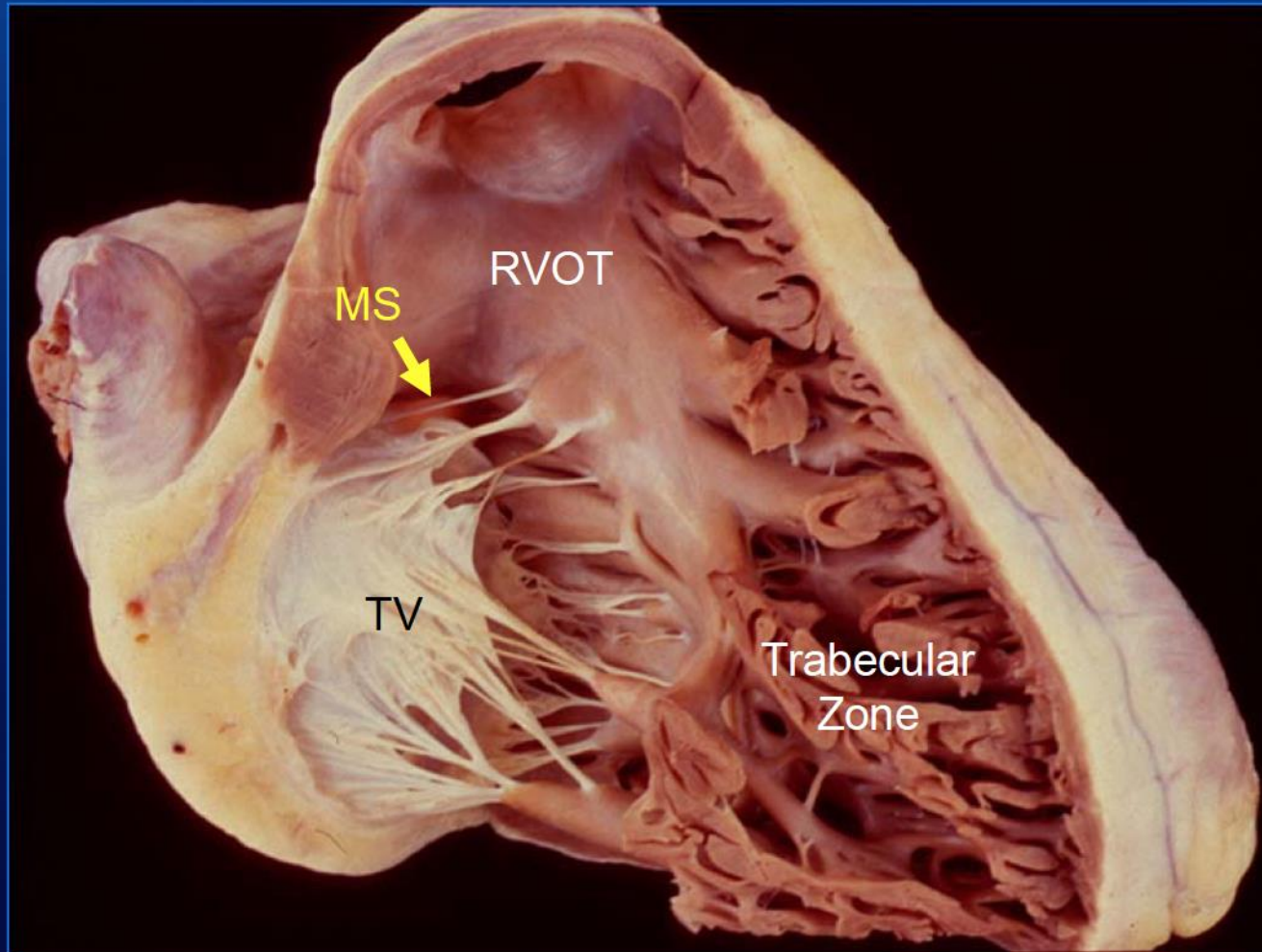
Ventrikelseptumdefect (VSD)

1. Outlet (5-8%, 30% Azië)
2. Perimembraneus (70%)
3. Musculeus, voorkant
4. Musculeus, apicaal
5. Mid-musculeus
6. Inlet (5-8%)



Ventricular Septal Defect

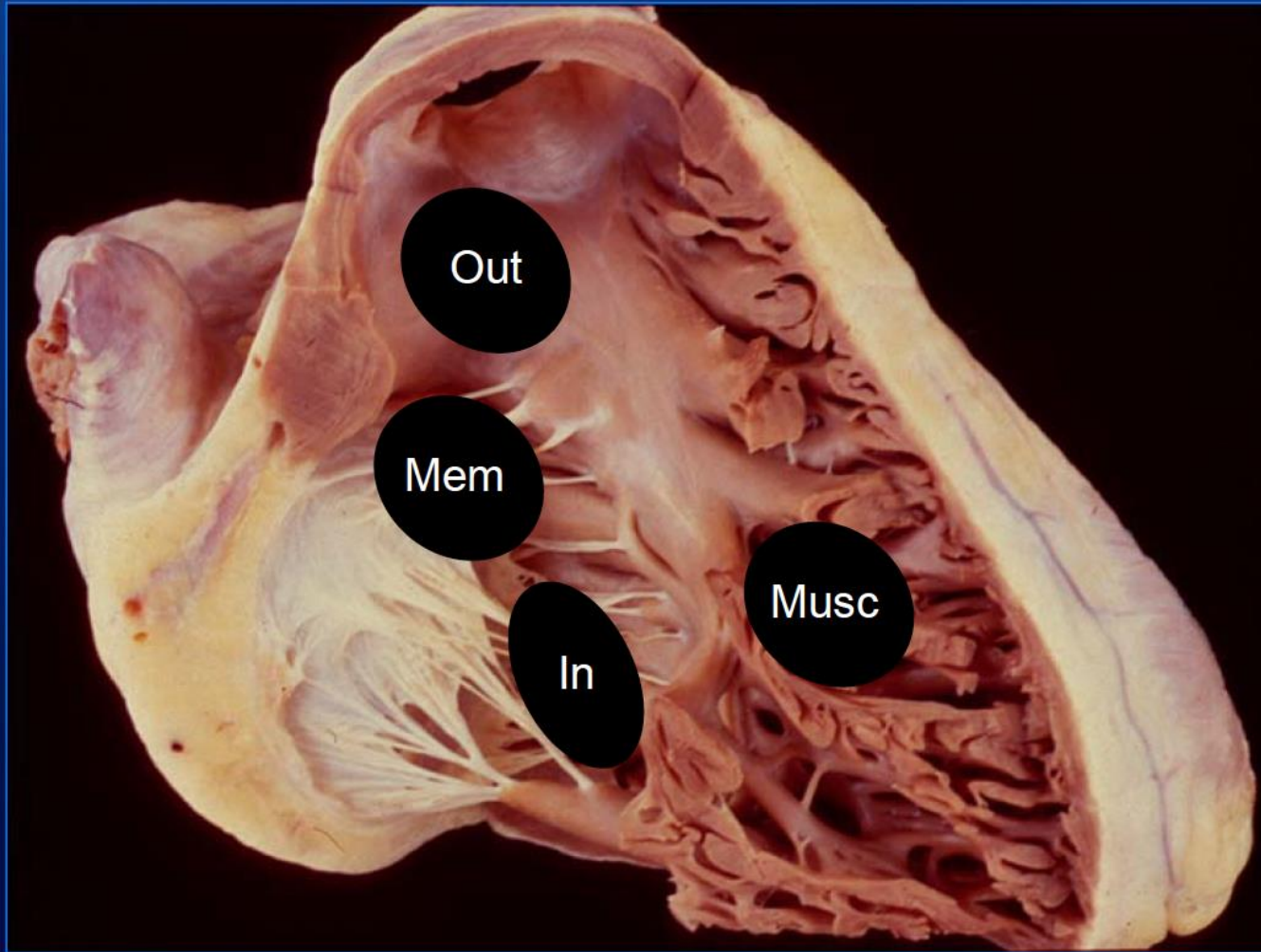
General Features



Right Ventricle (Free Wall Removed)

Ventricular Septal Defect

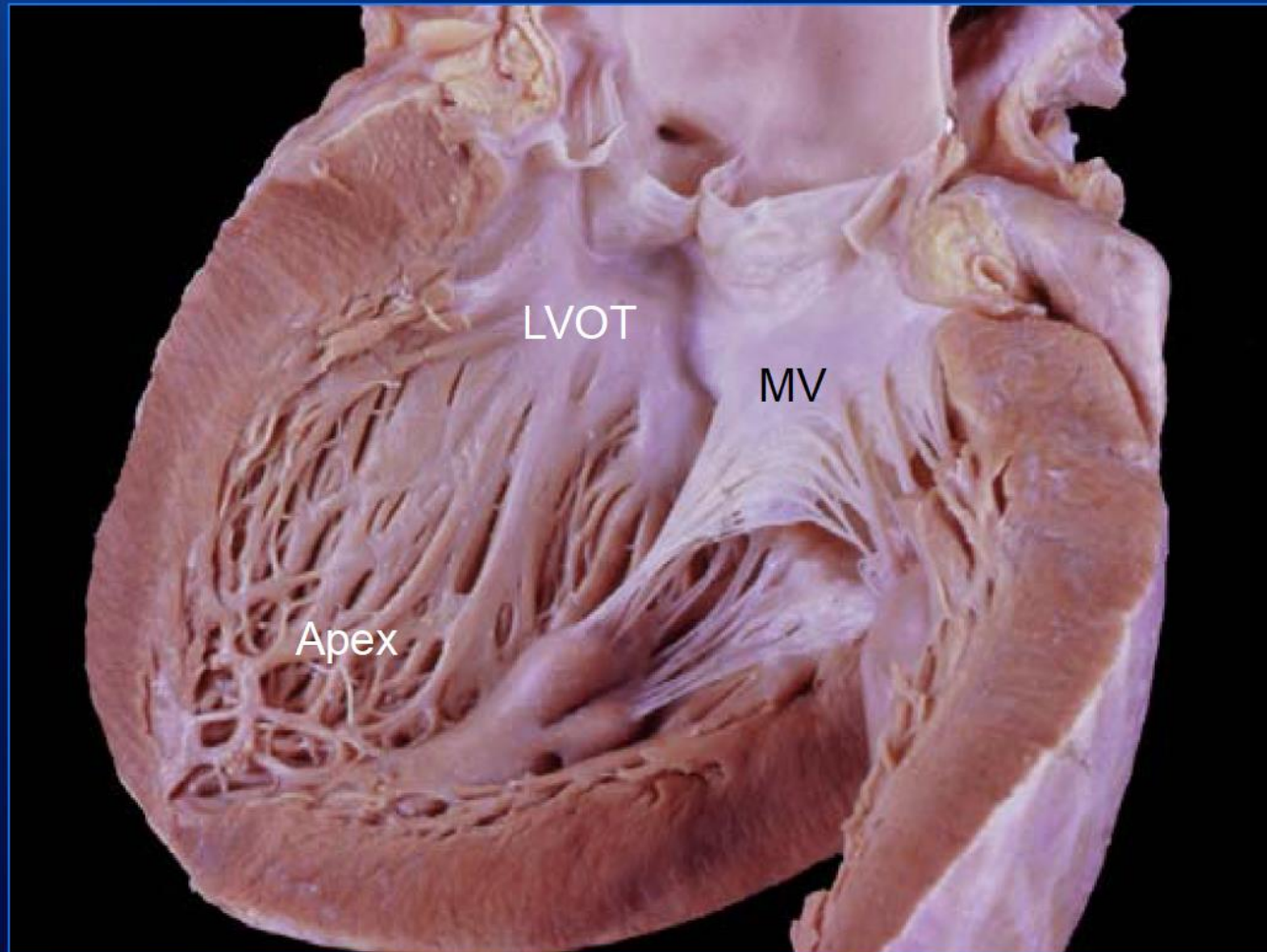
General Features



Location of Four Types of VSD

Ventricular Septal Defect

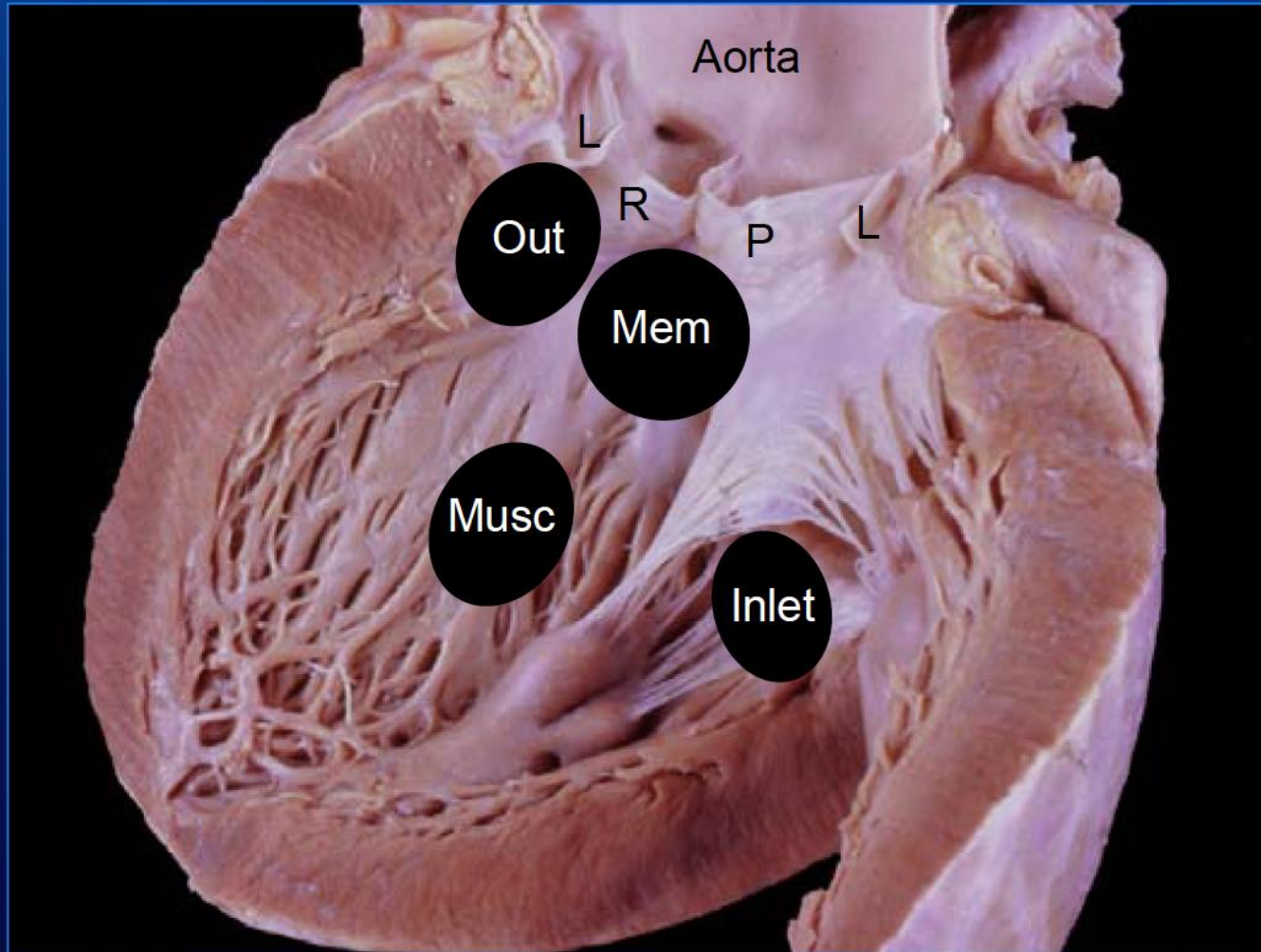
General Features



Left Ventricle (Opened Outflow View)

Ventricular Septal Defect

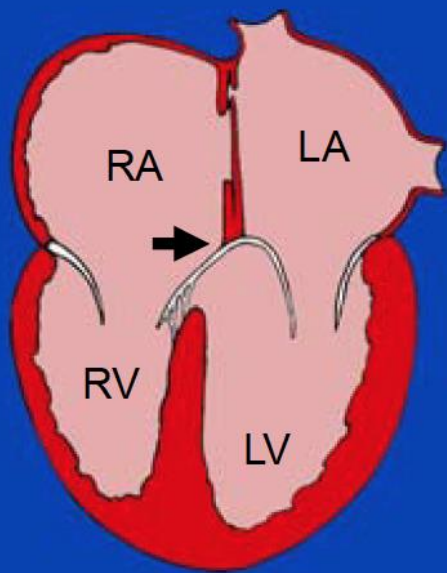
General Features



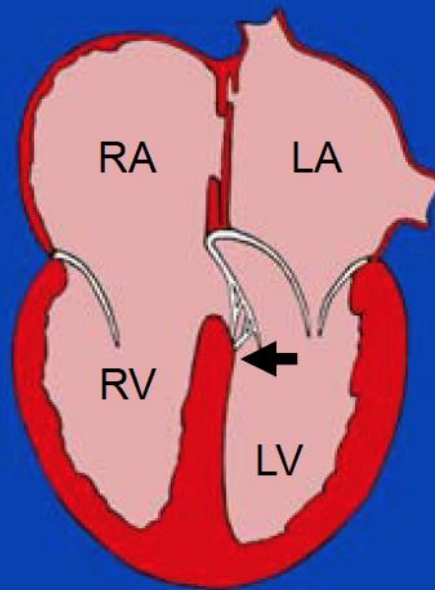
Location of Four Types of VSD

Septal Malalignment

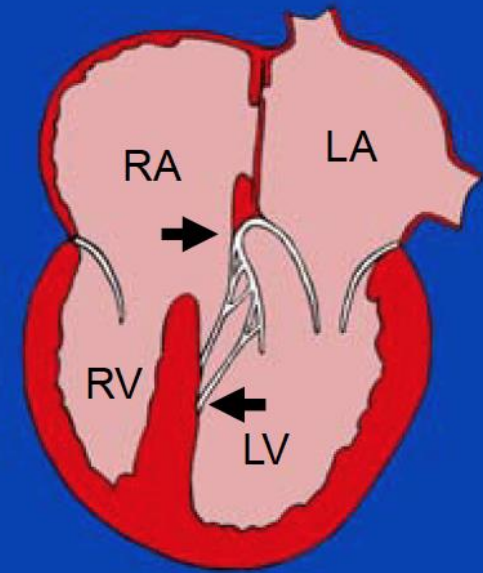
Overriding & Straddling Valves



Overriding
Only



Straddling
Only

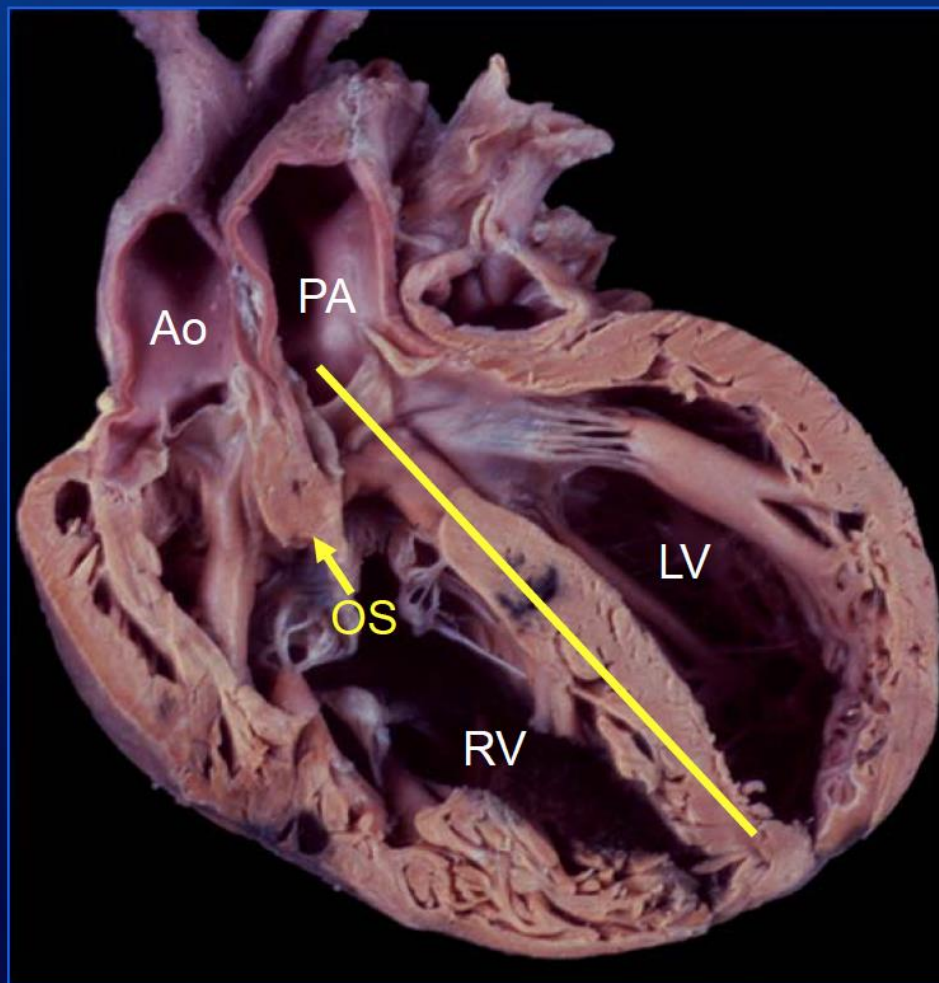


Overriding
& Straddling

Two Features (Diagram of Four-Chamber View)

Septal Malalignment

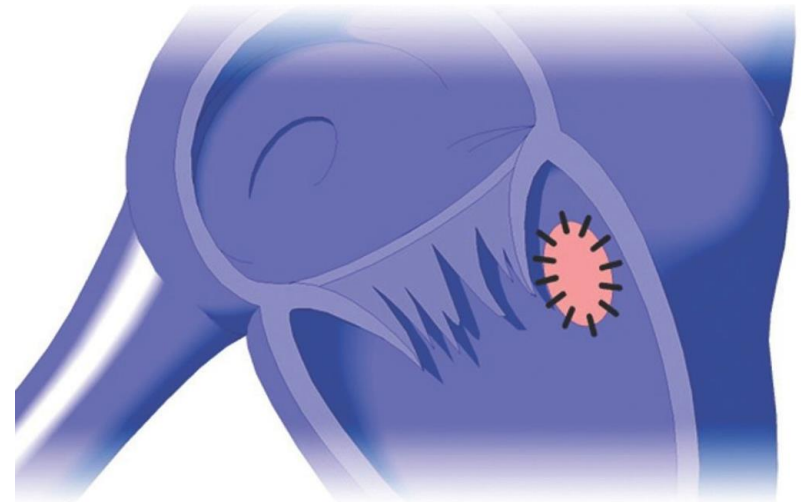
Overriding Semilunar Valve



Right Lateral Malalignment of Outlet Septum

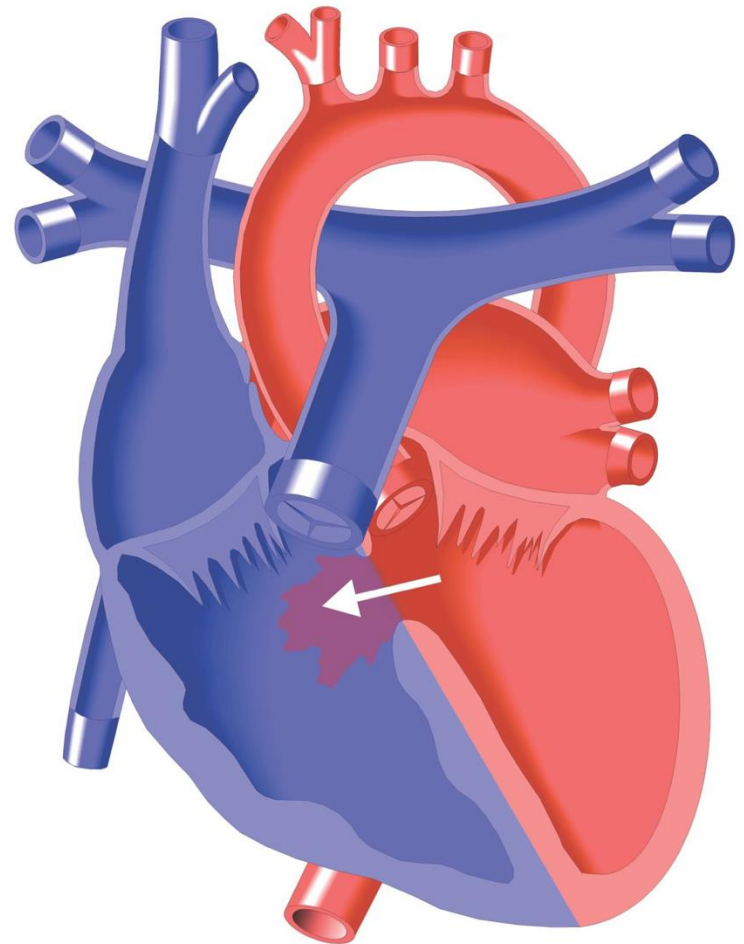
Sluiten van een VSD - *chirurgisch*

- Preoperatief
 - vochtbeperking
 - extra calorieën
 - diuretica
- Sluiten
 - hartfalen/ FTT
 - LR shunt: $Q_p:Q_s > 1.5-2$
 - aortaklep lekkage
- Via RA of PV
- Lage mortaliteit $< 2\%$
- Postoperatieve 'complicaties'
 - LCOS, PHT crise
 - ritmestoornissen (CAVB, JET)



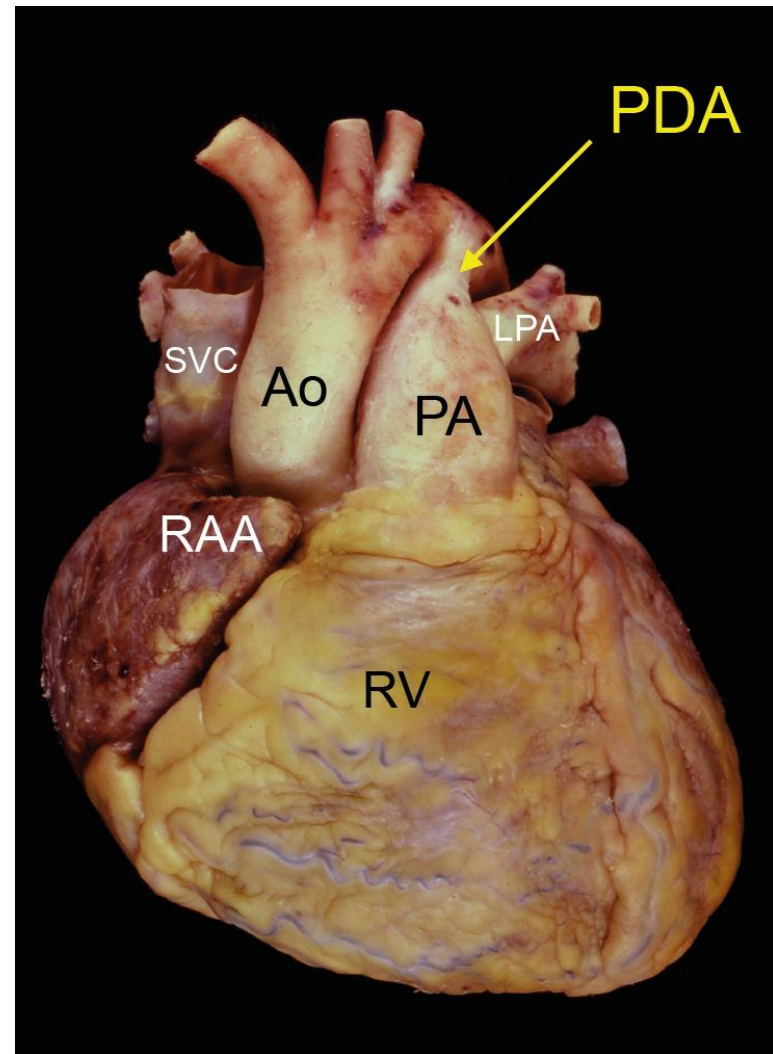
Follow-up/ prognose van een VSD

- Endocarditis profylaxe eerste 6 maanden
- Nagenoeg normaal inspanningsvermogen
- Geen sportbeperkingen mits
 - normale hartfunctie
 - geen belangrijke rest-defecten
 - geen pulmonale hypertensie
 - geen ritmestoornissen



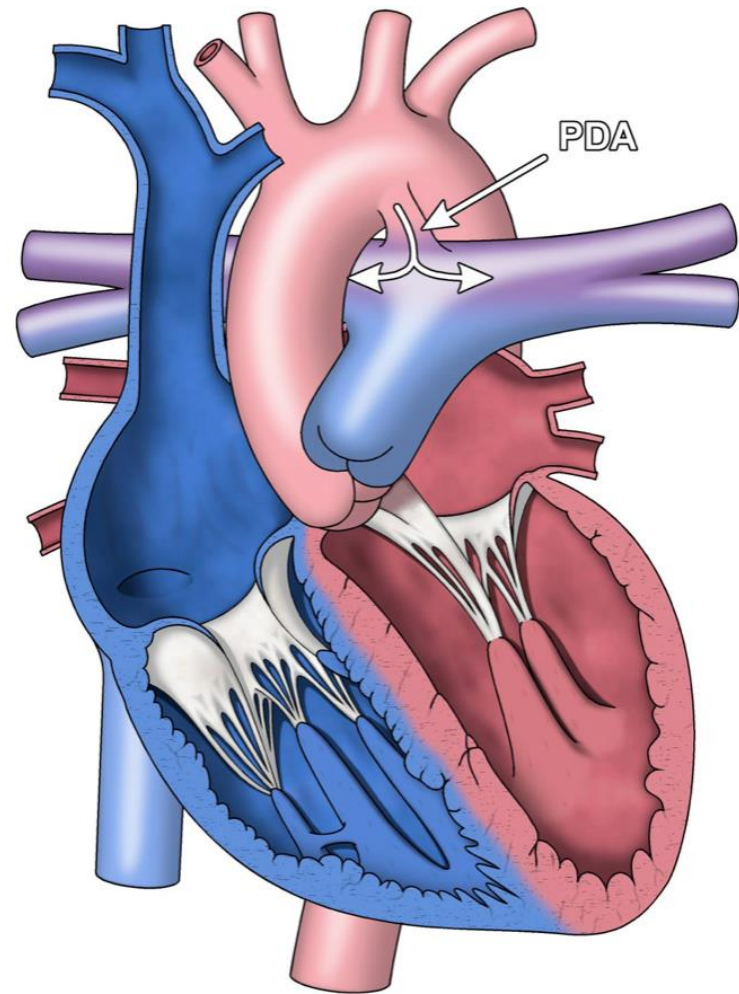
Persisterende ductus arteriosus (PDA)

- Open blijven in utero gereguleerd door prostaglandine E2 (PGE2)
- Sluit functioneel binnen 24-72 uren na geboorte
- Sluiten
 - zuurstof
 - verlaging PGE2
- Vertraagd sluiten/ open blijven vaak voorkomend bij prematuren
 - onrijpheid
 - longziekte



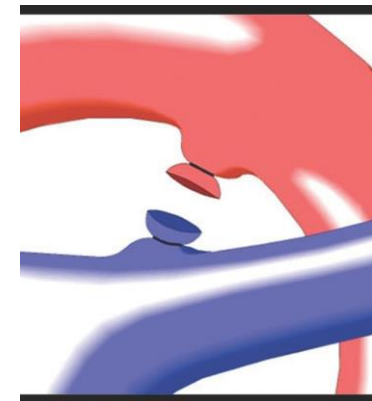
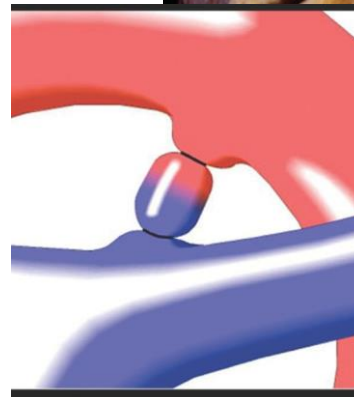
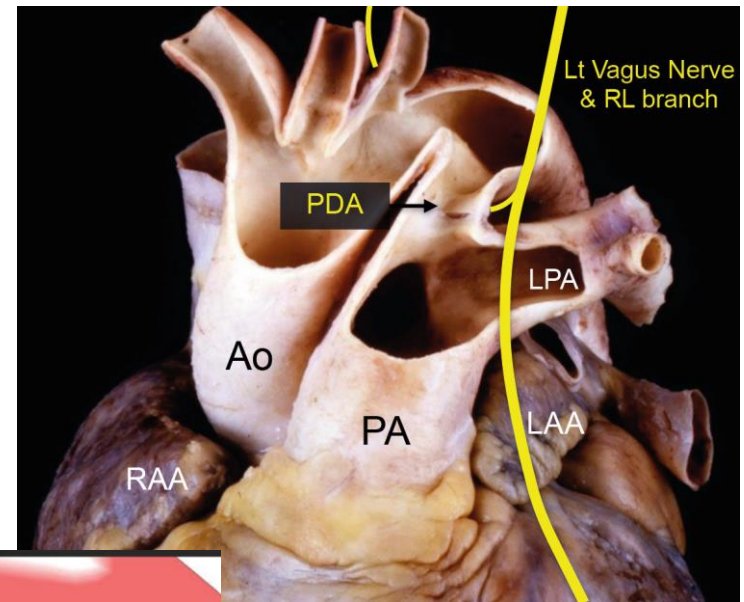
Persisterende ductus arteriosus (PDA)

- LR shunt
- Bij prematuren
 - hypotensie, NEC, longbloeding, PV bloeding
- Therapie
 - indometacine, ibuprofen
 - vochtbeperking
 - diuretica
 - chirurgisch
 - interventioneel
- Bij sommige hartafwijkingen nodig voor
 - waarborgen lichaamscirculatie
 - waarborgen longcirculatie

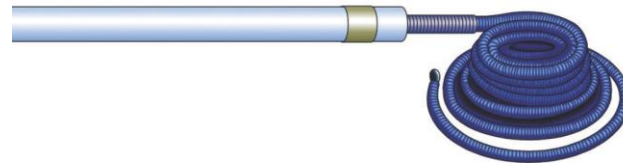
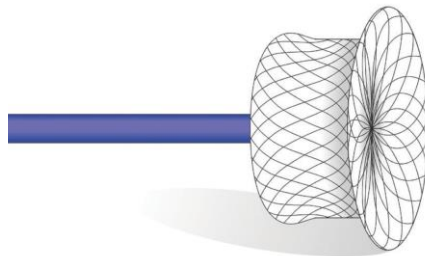
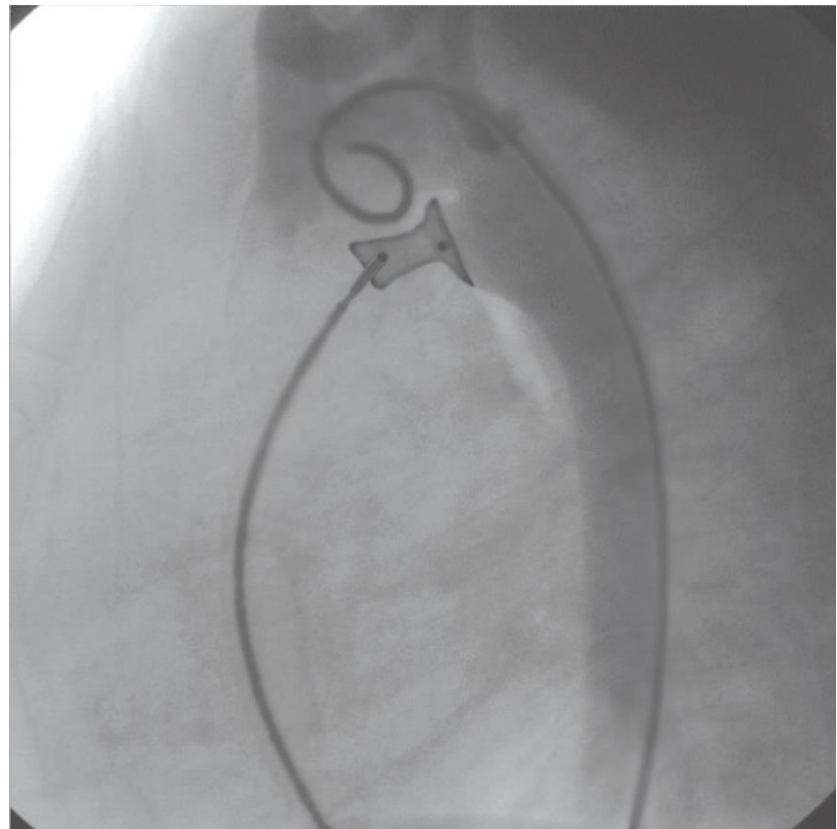
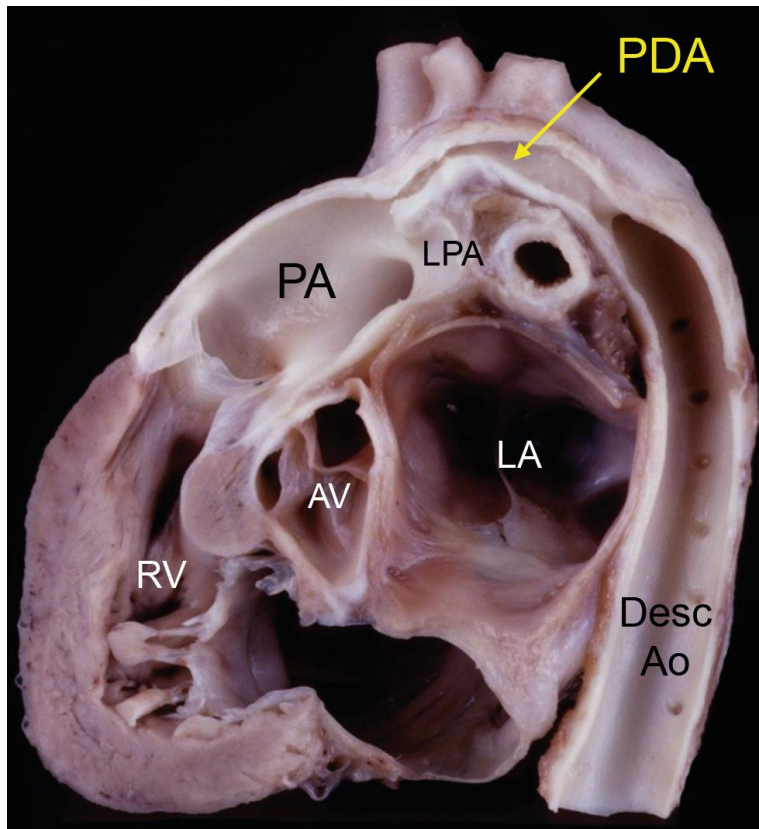


Sluiten van een PDA- chirurgisch

- Bij neonaten/ prematuren
 - bij falen indometacine therapie
- Via laterale (mini-) thoracotomie 'op zaal'
- Postoperatieve complicaties
 - stembandparese (door n. laryngeus recurrens lesie)

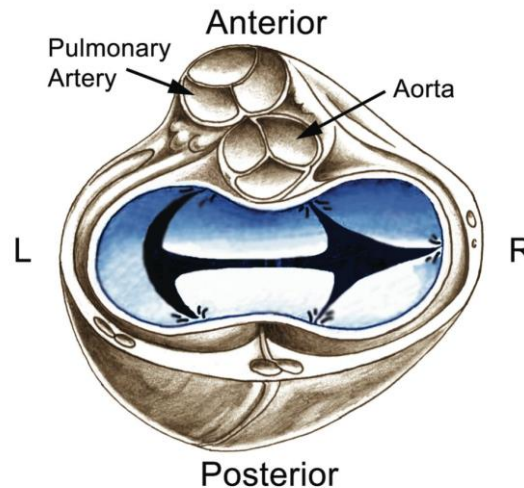
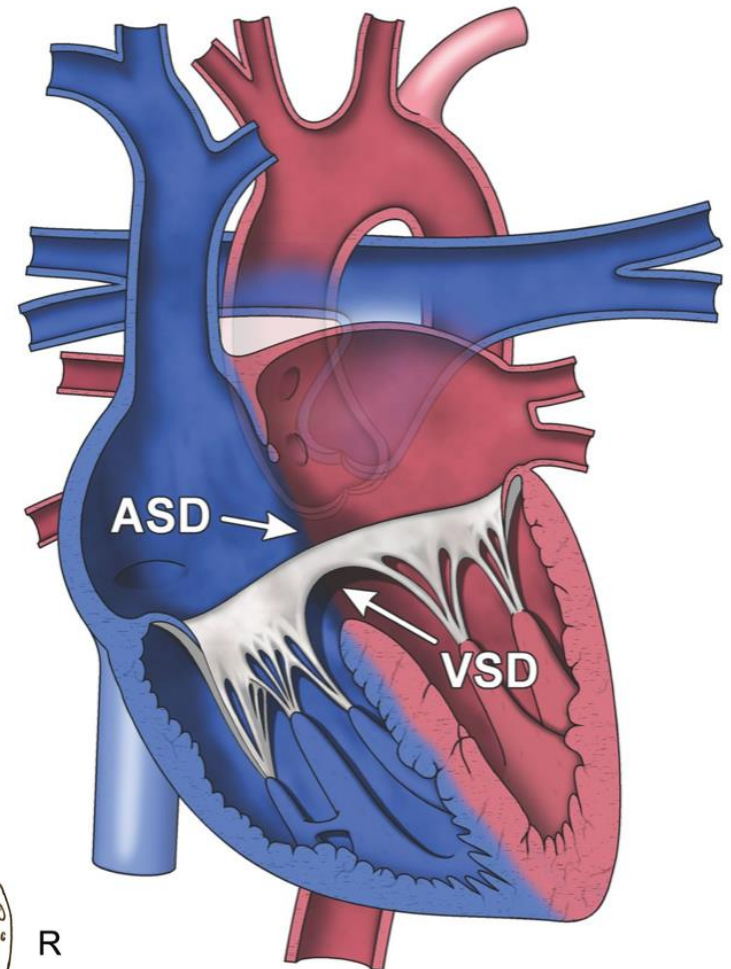


Sluiten van een PDA- interventioneel



Atrioventricular septumdefect (AVSD)

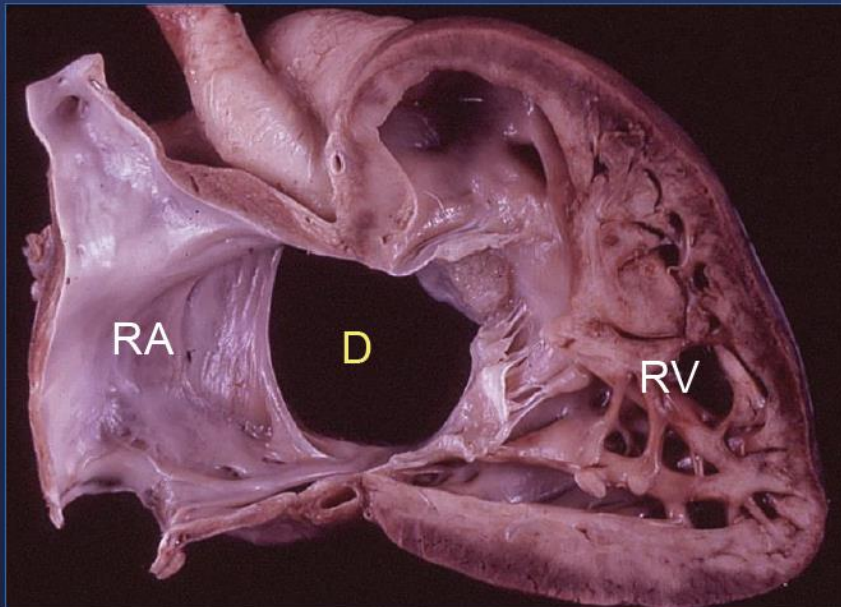
- Partieel (PAVSD)
 - ASD I
 - cleft in 'MV'
- Compleet (CAVSD)
 - ASD I
 - inlet VSD
 - common AV klep met 5 klepbladen
 - 50% trisomie 21



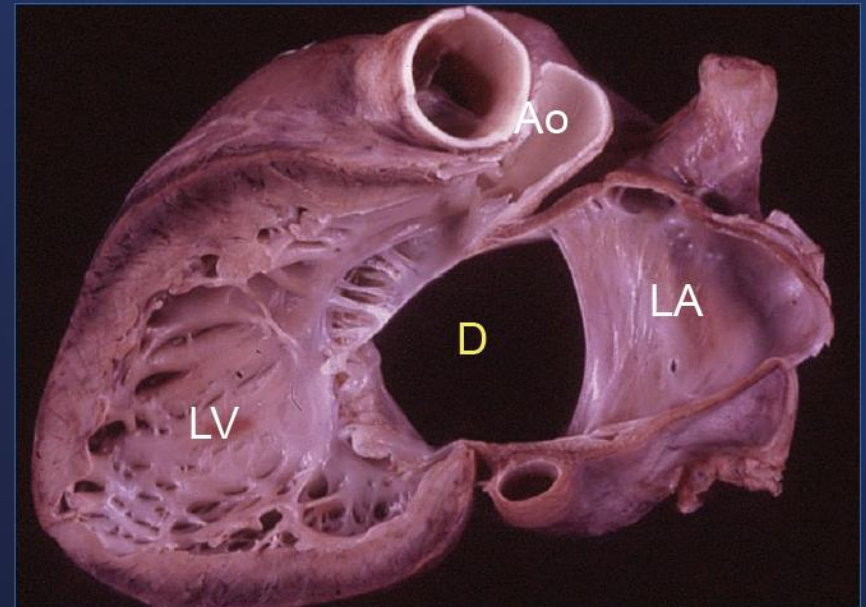
Atrioventricular Septal Defects

Complete Type - General Features

Combined Primum ASD & Inlet VSD



Large Septal Defect
(R-Sided Two-Chamber)

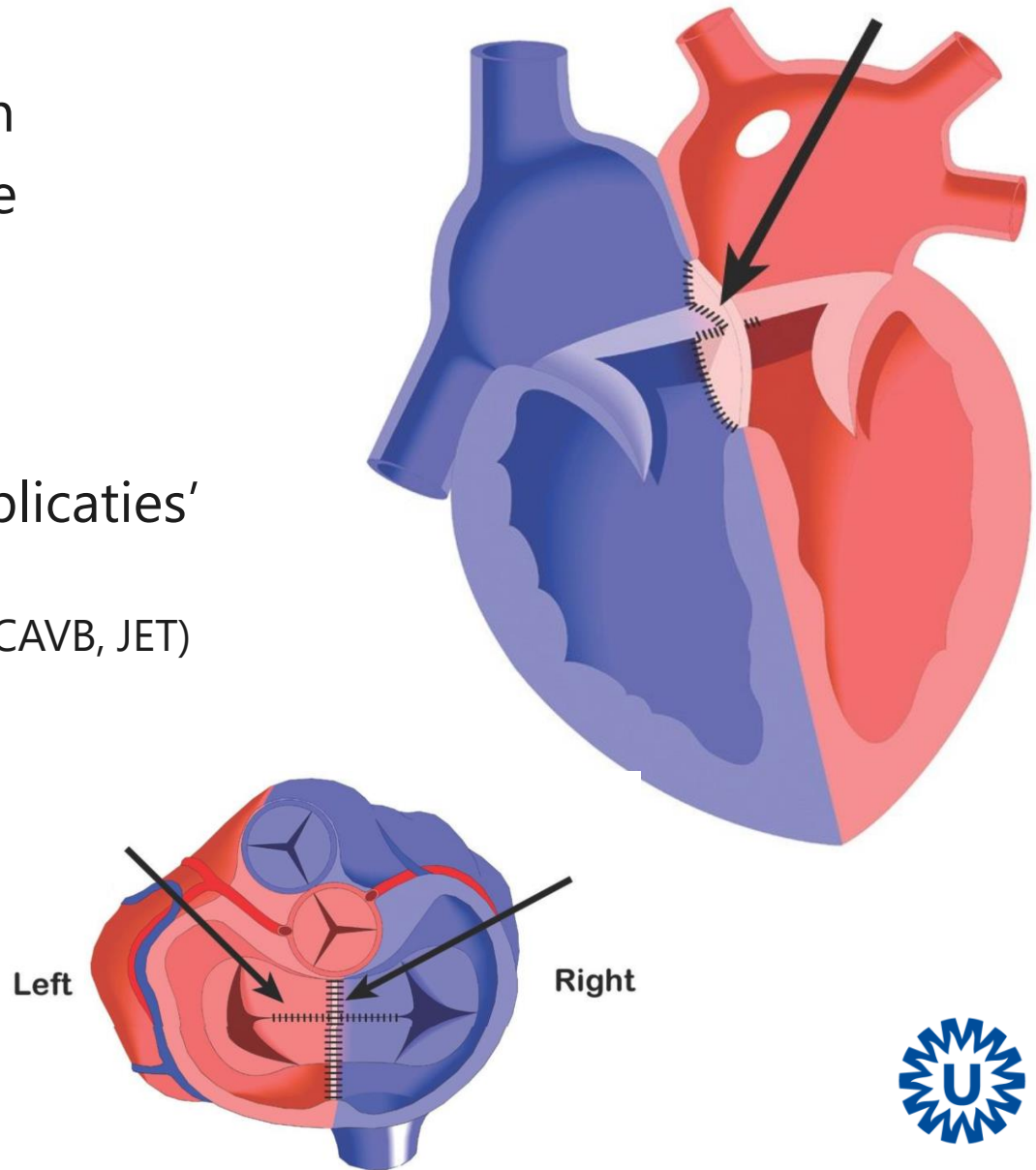


Elongated LVOT
(L-Sided Two-Chamber)

Autopsy Heart (Free Walls Removed)

Chirurgische correctie van een CAVSD

- Leeftijd 3-6 maanden
- Mediane sternotomie
- CPB, cardioplegie
- 2-patch techniek
 - Gore-Tex
- Postoperatieve 'complicaties'
 - LCOS, PHT crise
 - ritmestoornissen (CAVB, JET)
- Mortaliteit 2-5%
- 50% rest-AV klep lekkage, 10% re-op



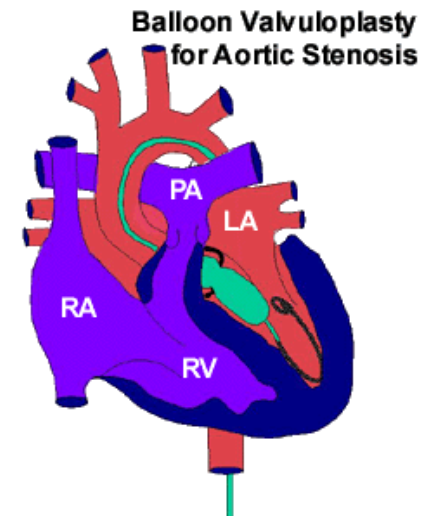
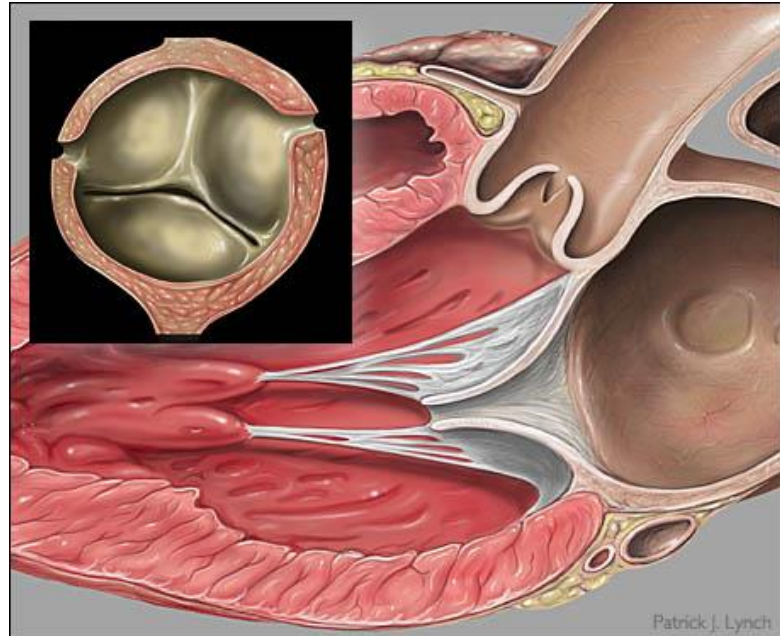
Type hartafwijking- *obstructieve afwijkingen*

- pulmonaal klep stenose (PS)
- aorta klep stenose (AS)
- coarctatio aortae (CoA)



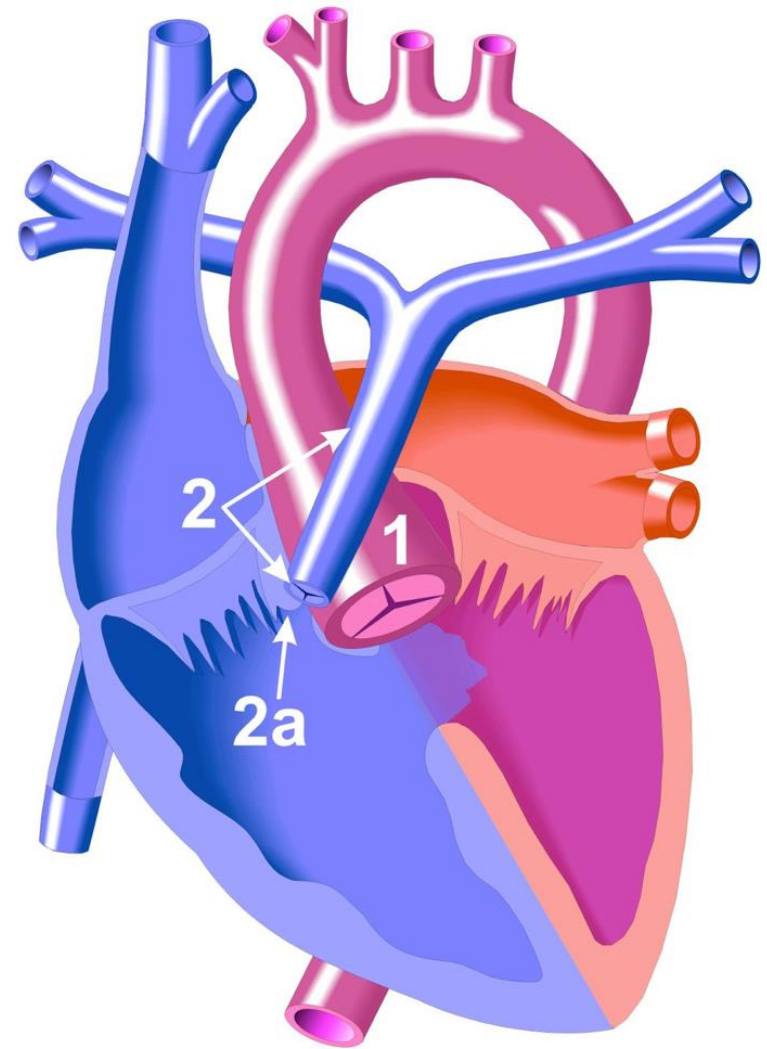
Obstructieve afwijkingen- aortaklepstenose

- 70% bicuspide aortaklep
- Kritische AS
 - neonat, dag 3, bleek, niet-plassen, hypotensie, acidose
 - soms geassocieerd met MV en LVOT afwijkingen en CoA
- Niet-kritische AS
 - oudere kinderen
 - hartruis
 - bij toename stenose: kortademig, verminderd inspanningsvermogen, syncope
 - SCD



Tetralogie van Fallot

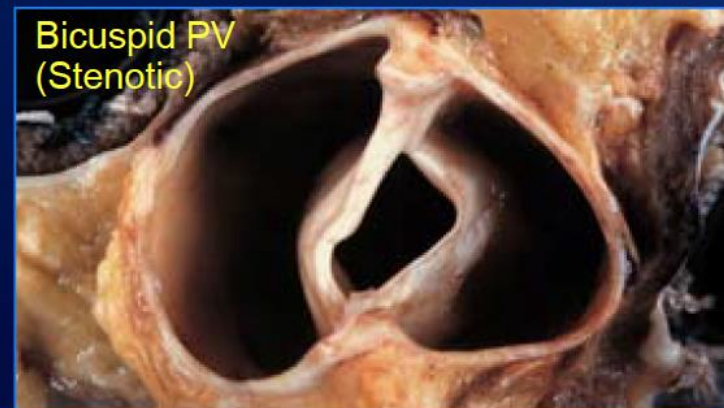
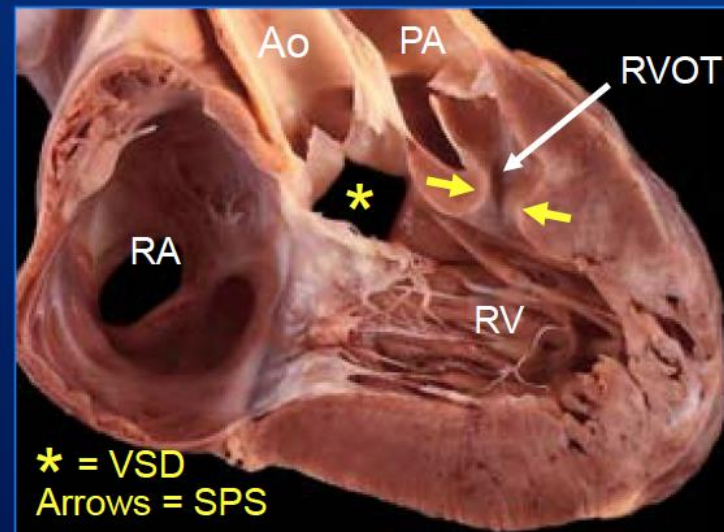
- 4 (=tetra) kenmerken
 - malalignment VSD (1)
 - overrijdende aorta (1)
 - RVOT obstructie (= subpulmonale stenose)(2a)
 - RVH
- Geassocieerde afwijkingen
 - rechter aortaboog (25%)
 - coronairanomalie (10%)
 - ASD
 - multiple VSD's
 - AVSD
 - 22q11 deletie



Tetralogy of Fallot

Characteristic Features

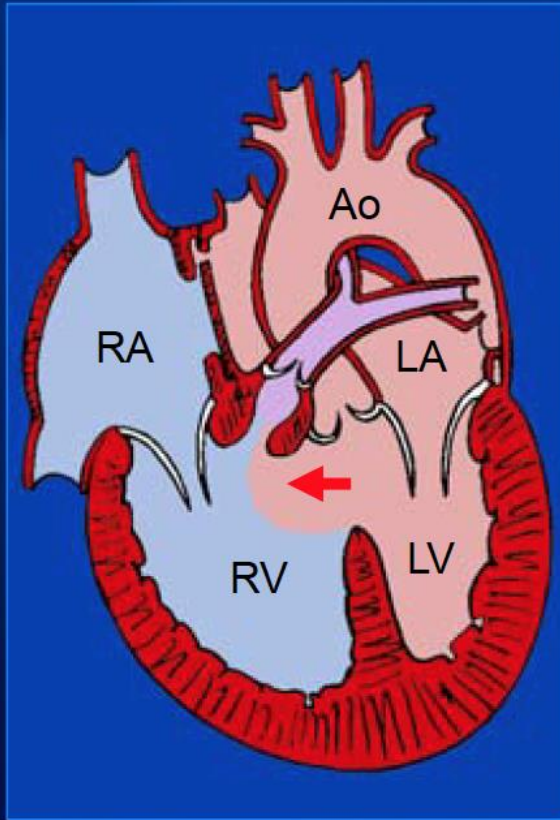
- **Components**
 - VSD (malalignment)
 - Subpulmonary stenosis
 - Overriding aorta
 - RV hypertrophy
- **RV hypertrophy**
 - Not a congenital defect
 - Due to VSD with equal pressures in RV & LV
- **Variable feature**
 - Severity of sub-PS
 - Severity of valvular PS (bicuspid in 50%)



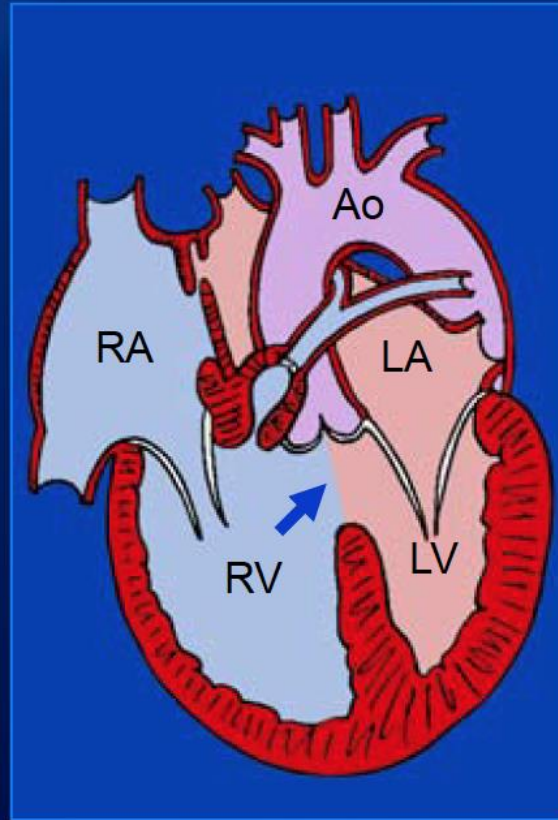
PV & Heart (RAO View)

Tetralogy of Fallot

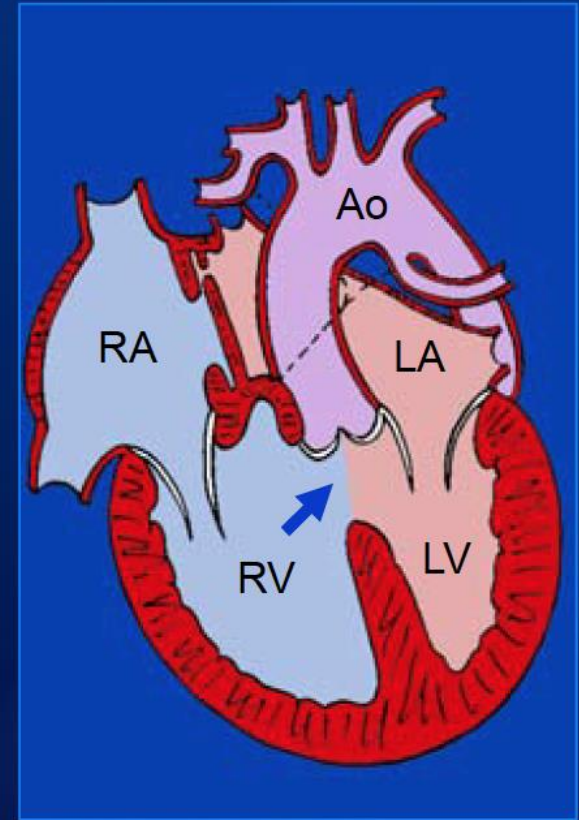
Severity of Subpulmonary Stenosis



Mild Stenosis
Pink Tetralogy



Mod-Sev Stenosis
Classic Tetralogy

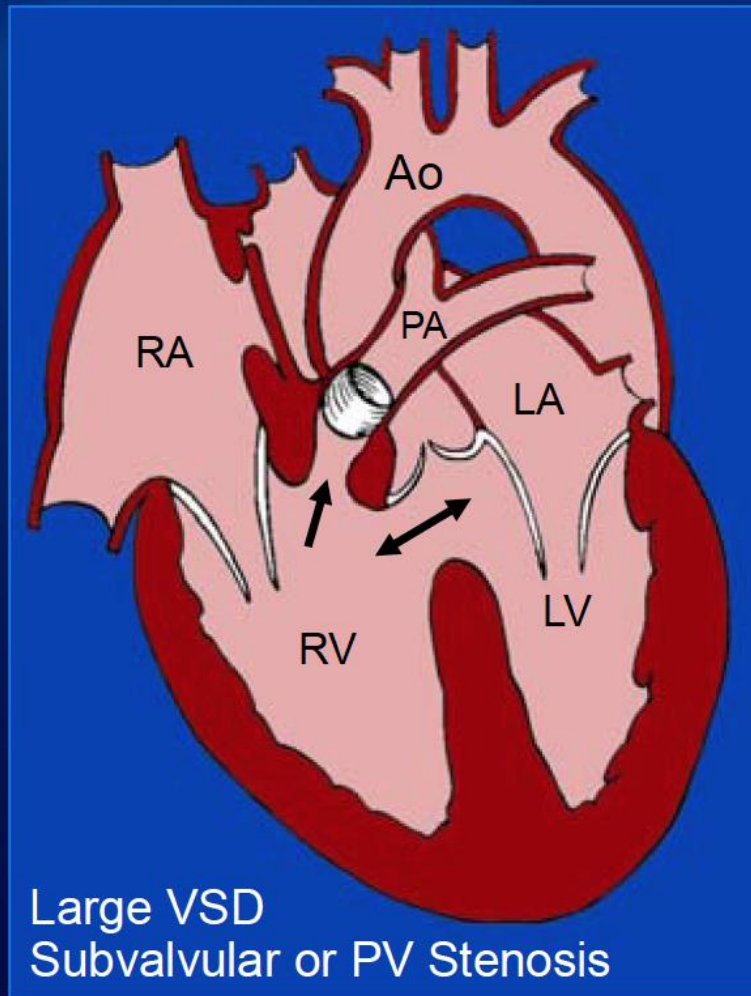


Valve Atresia
PA-VSD

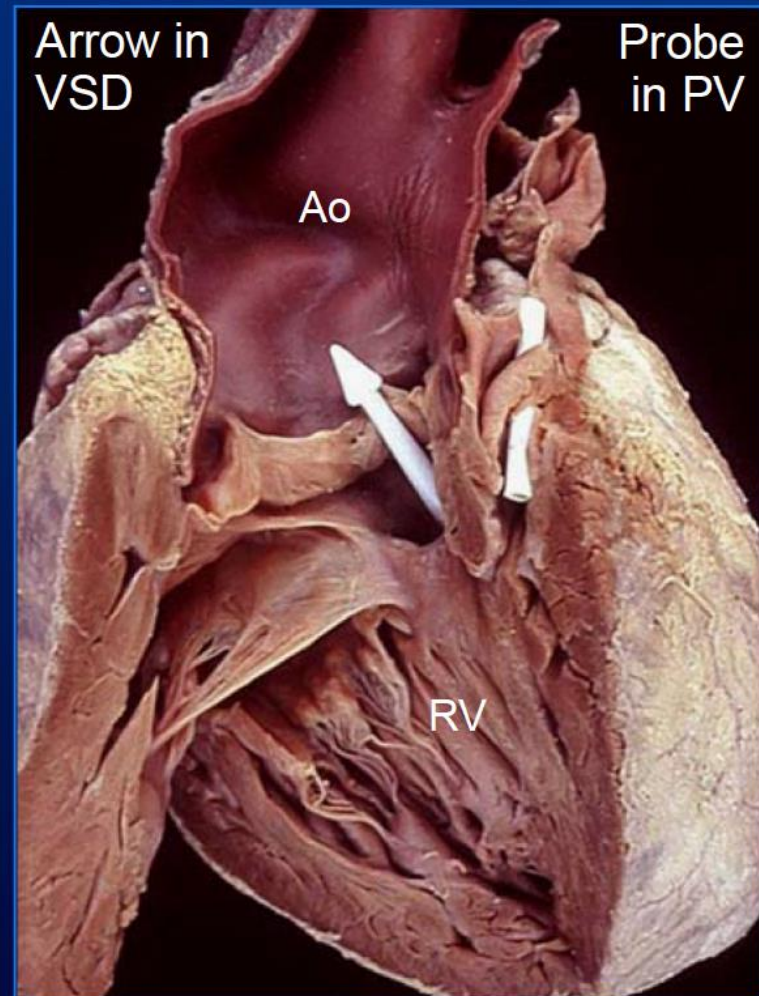
Size of PAs Is Determined By Severity of SPS

Tetralogy of Fallot

VSD & Subpulmonary Stenosis



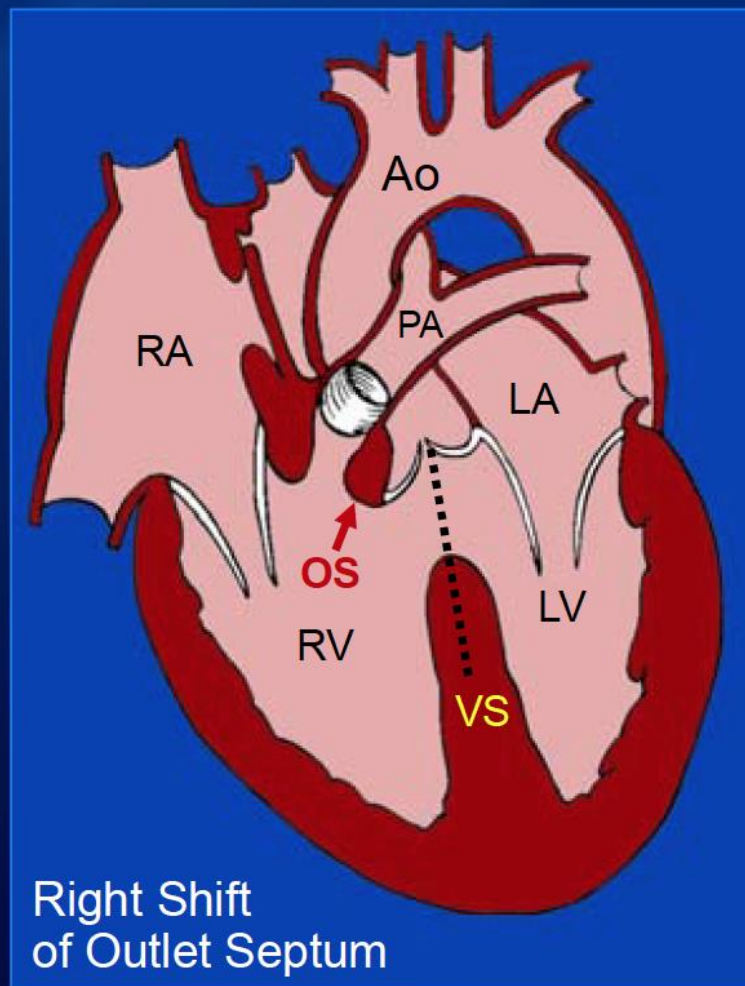
Classic Tetralogy



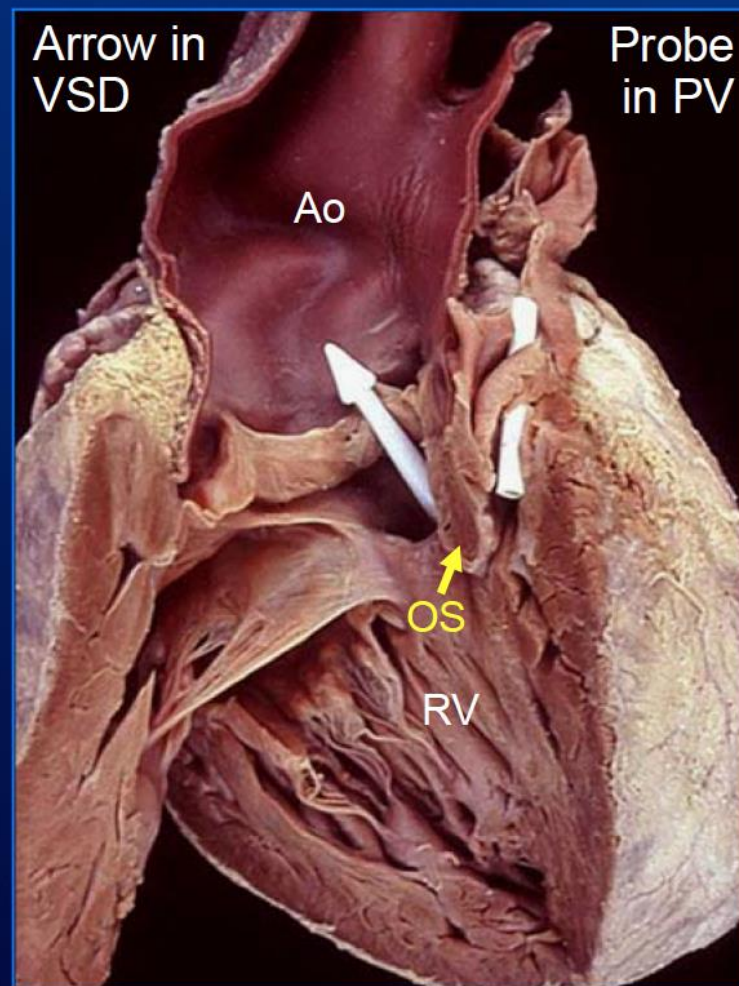
Opened RV (Ant View)

Tetralogy of Fallot

Malalignment VSD



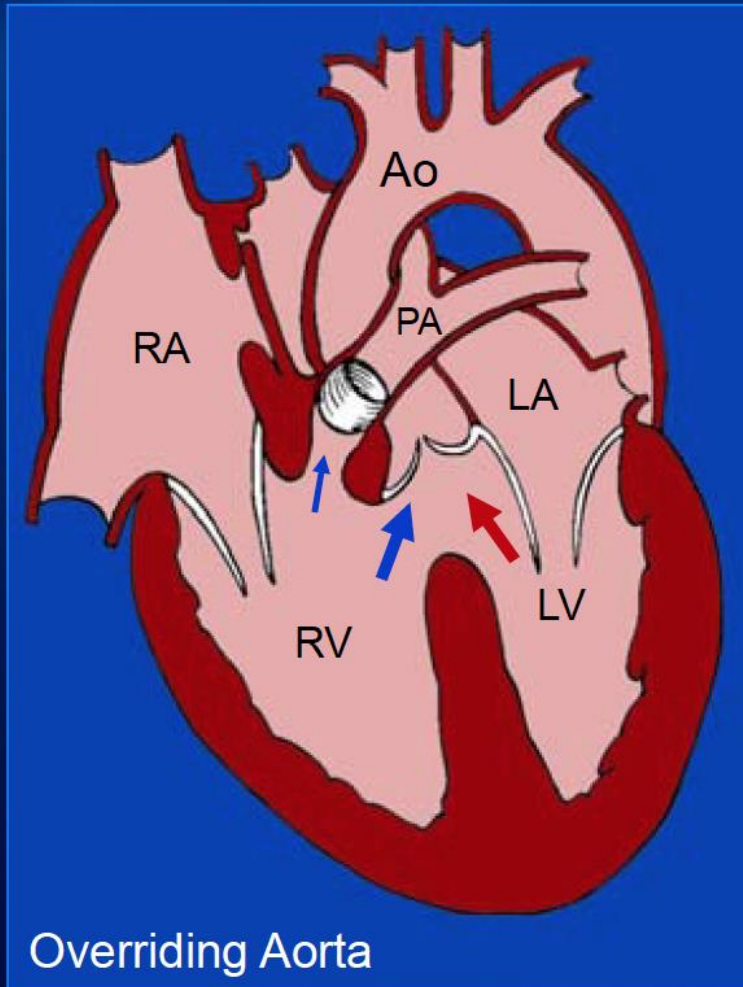
Classic Tetralogy



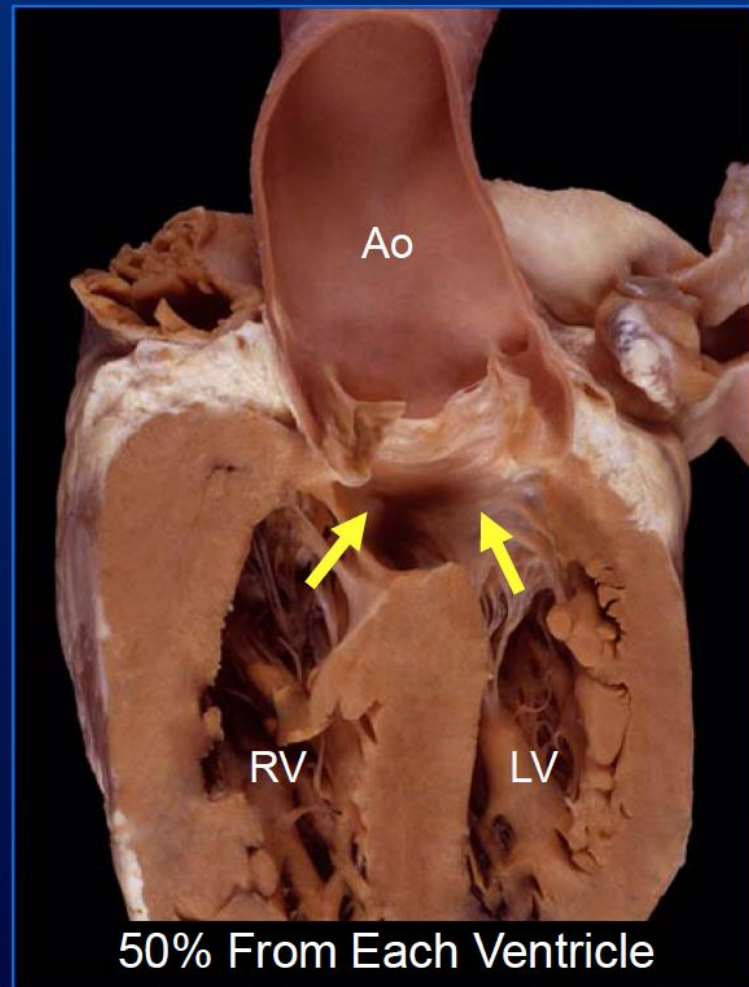
Opened RV (Ant View)

Tetralogy of Fallot

Overriding Aorta



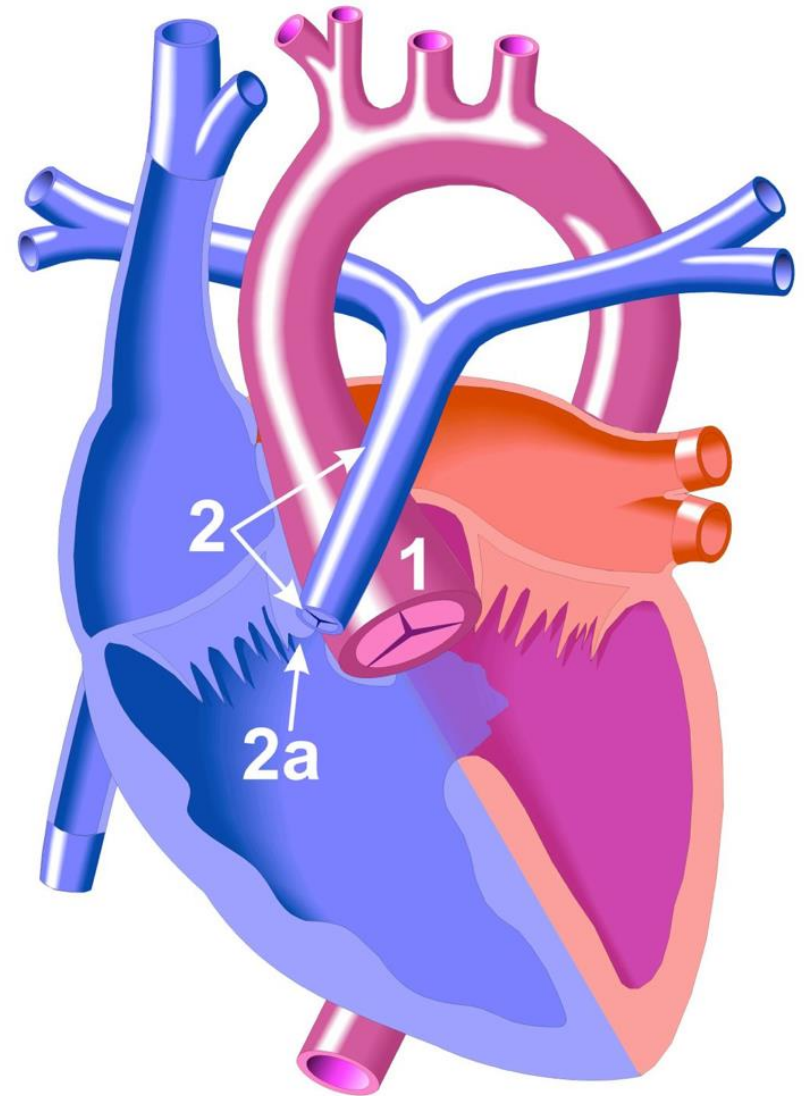
Classic Tetralogy



Overriding Aorta

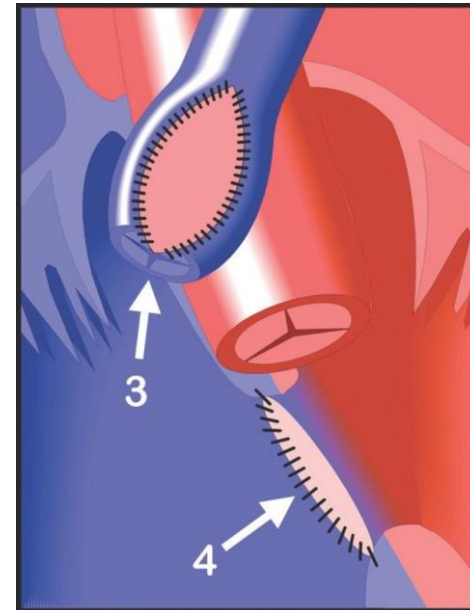
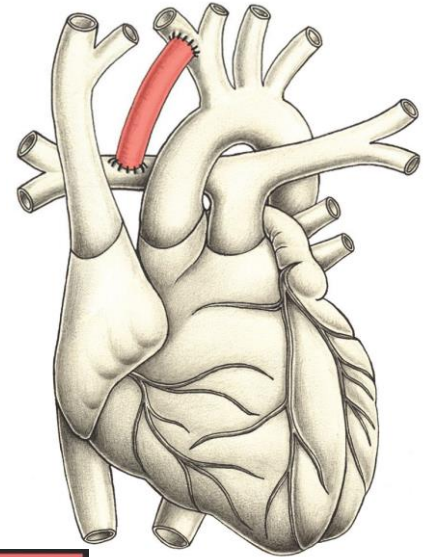
Tetralogie van Fallot

- Spells
 - typisch rond 2e-3e maand
 - uitgelokt door huilen, persen, intercurrente ziekte
 - 'spasme' RVOT > minder longdoorbloeding > eerst blauw en hyperventilatie, dan bleek/grijs, soms syncope
 - kind 'opvouwen'
 - acuut: zuurstof, fenylefrine, morfine
 - preventie: propranolol



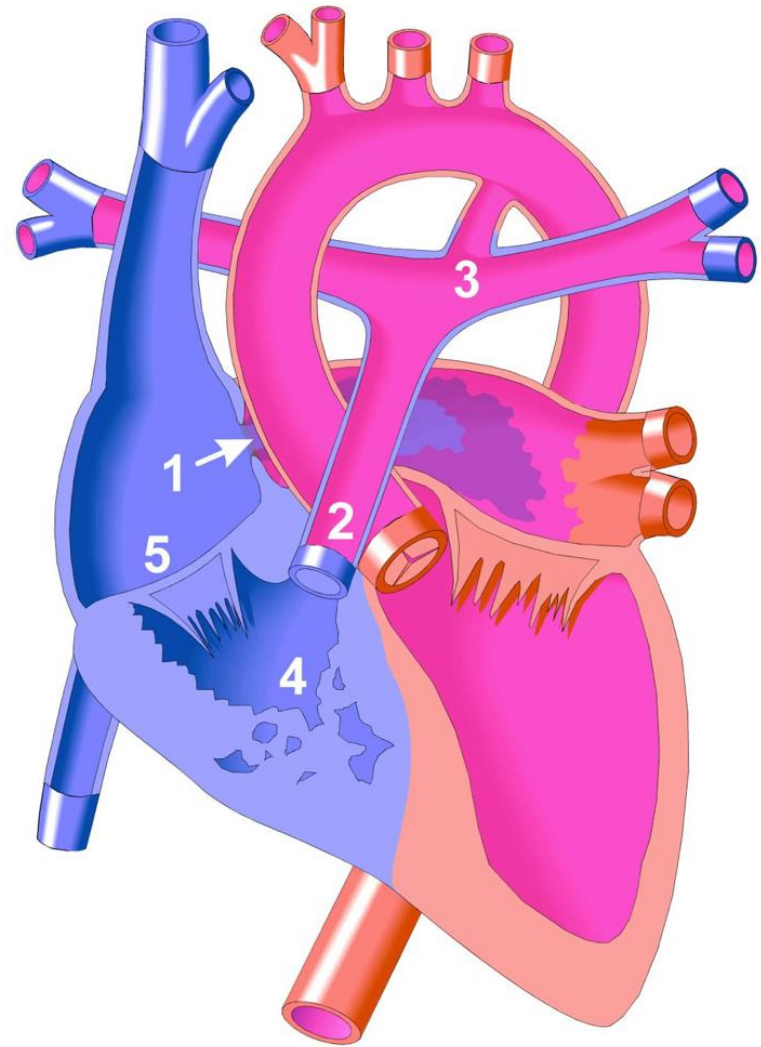
Chirurgische correctie van ToF

- Bij spell > Blalock Taussig shunt of correctie
- Correctie tussen 3e-6e maand
- Mediane sternotomie
- CPB, cardioplegie
- Incisie RA en PA (soms ook RVOT)
- Mortaliteit 2-5%
- Postoperatieve complicaties
 - LCOS
 - ritmestoornissen (CAVB, JET)



Pulmonalisatresie (PA)

- Met VSD (PAVSD) > variant ToF
- Met intact interventriculair septum (PAIVS)
 1. ASD
 2. atresie PV
 3. PDA
 4. Hypoplastisch RV
 5. Hypoplastische TV

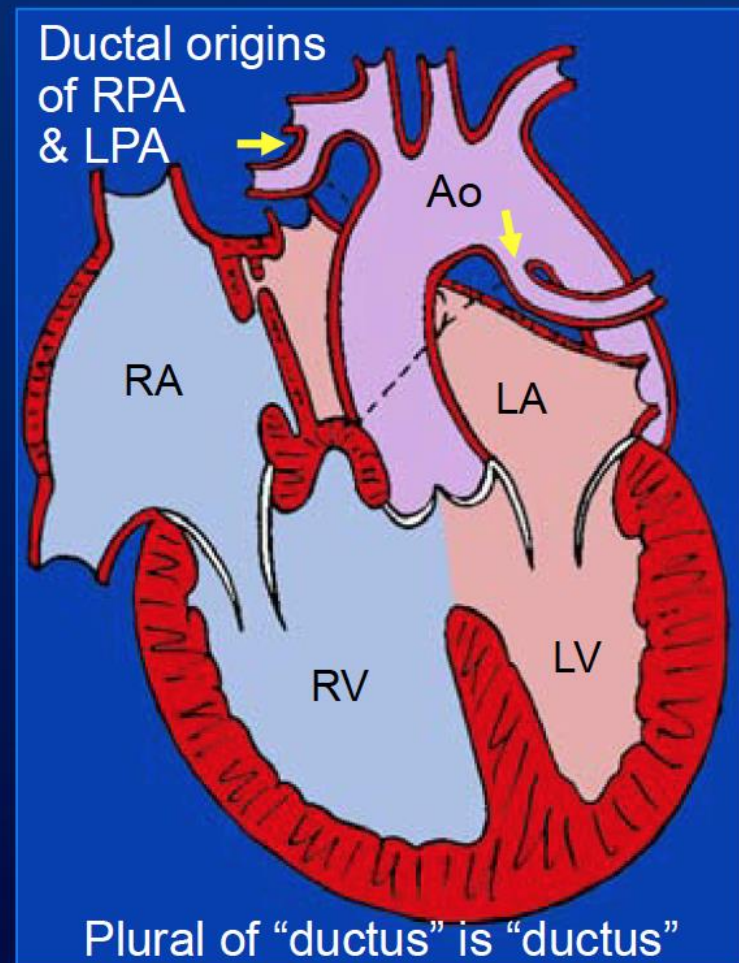


Pulmonary Atresia with VSD

Characteristic Features

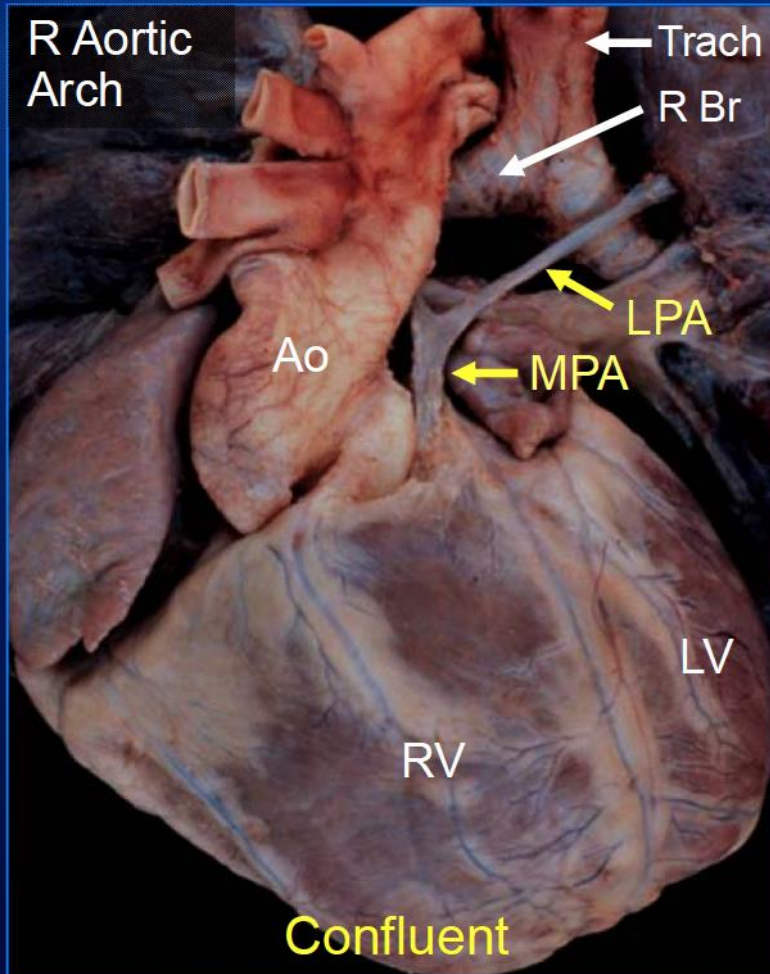
- **Components**
 - VSD (malalignment)
 - Overriding aorta
 - No RV-PA connection
 - Secondary RVH
- **Pulm arteries**

	%
Hypoplastic/cord	75
Unidentifiable	25
Confluent PAs	65
Non-confluent	35
- **Pulm blood supply**
 - PDA (R, L, or bilateral)
 - Collaterals: desc ao > subcl >> abd ao, cor

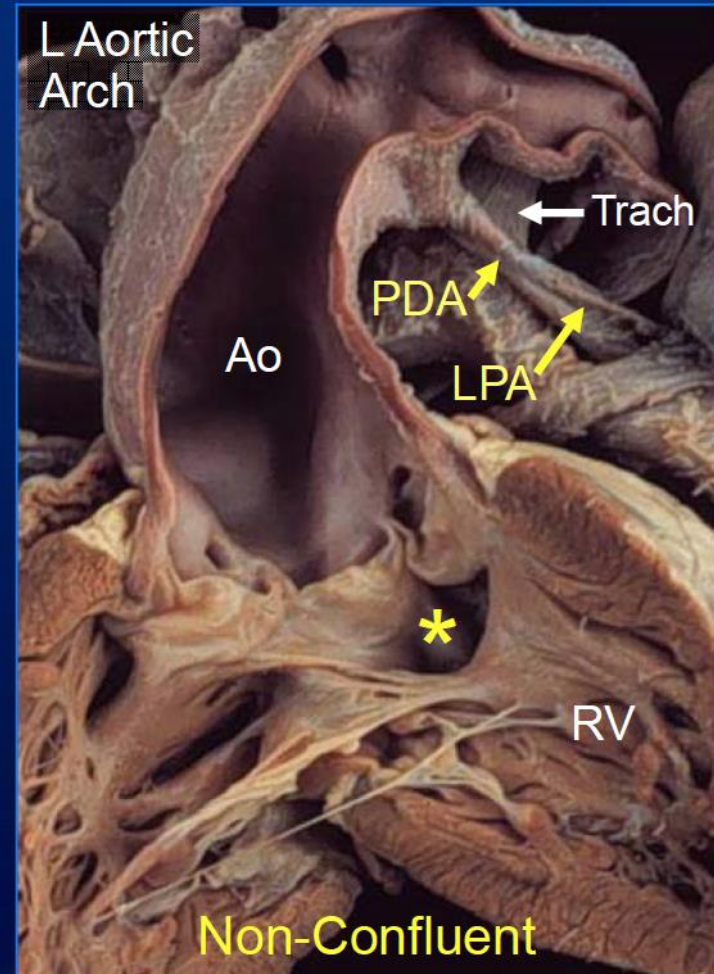


Pulmonary Atresia with VSD

Hypoplastic & Atretic Pulmonary Arteries



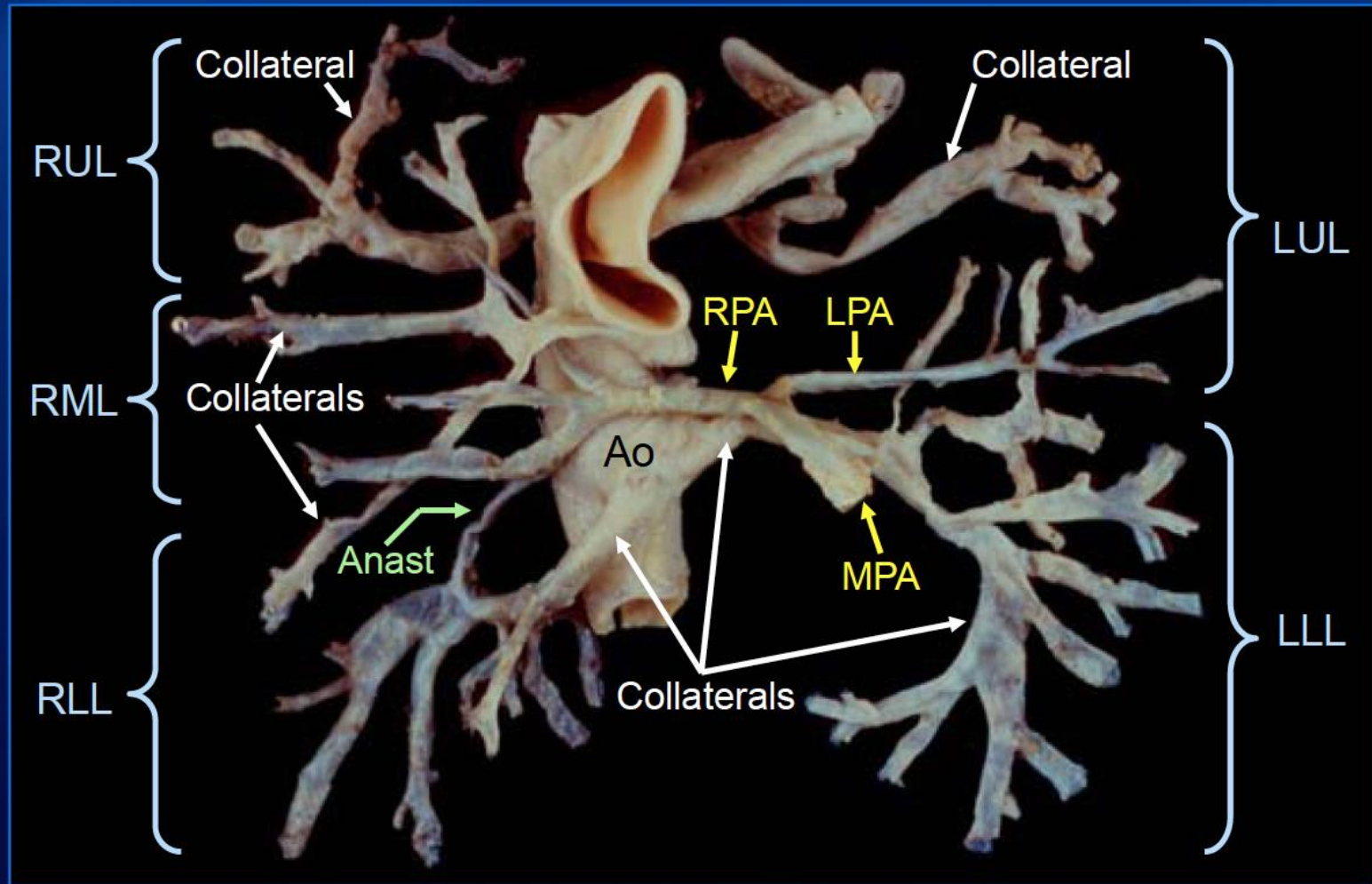
Severely Hypoplastic PAs



Atretic Proximal PAs

Pulmonary Atresia with VSD

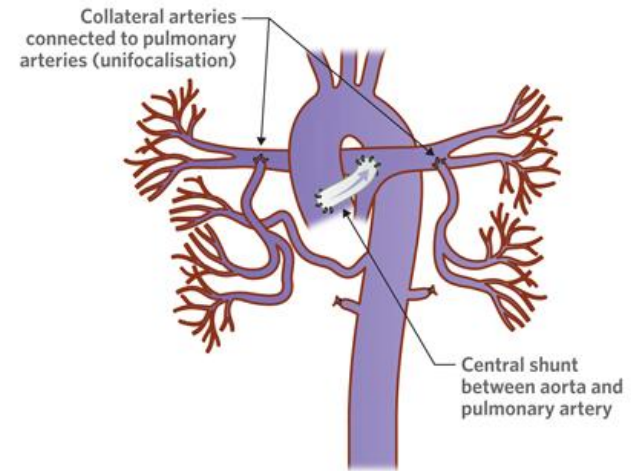
Pulmonary Arteries & Collateral Arteries



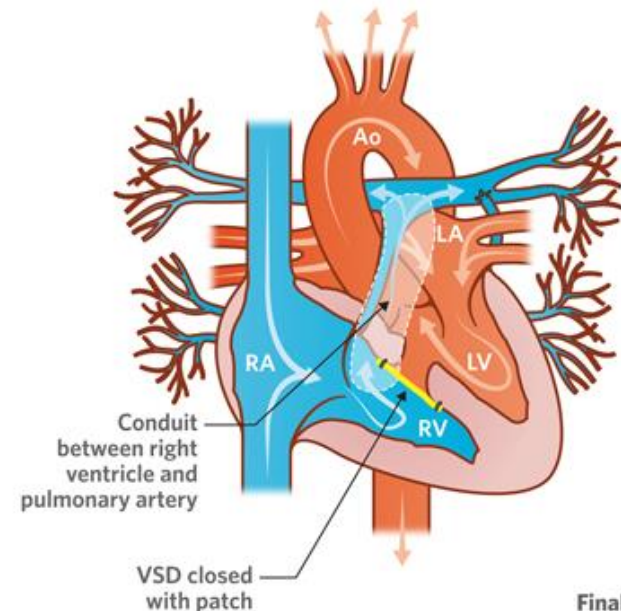
Complex Collateral Arteries (Anterior View)

Correctie van een PA met VSD

- Bij membraneuze PA
 - RF-ablatie
- Chirurgisch
 - Blalock-Taussig shunt
 - Rastelli procedure

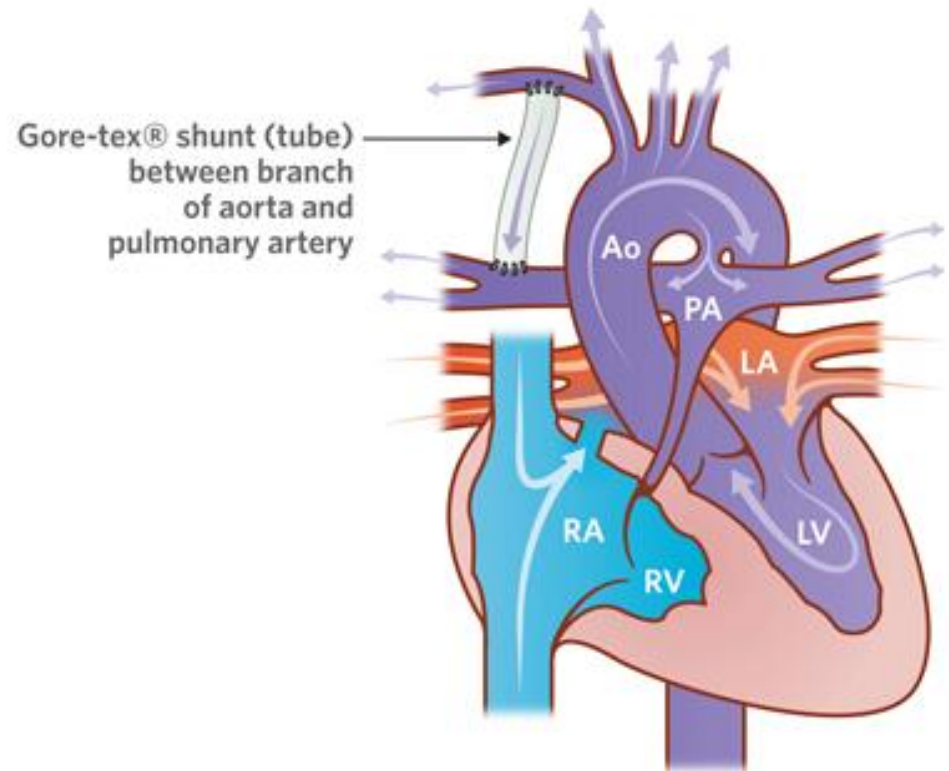


First stage of repair



Correctie van een PA met IVS

- Afhankelijk van RV
 - commissurotomy
 - patch vergroting RVOT, PA
 - (shunt)
- RV te klein
 - Fontan circulatie

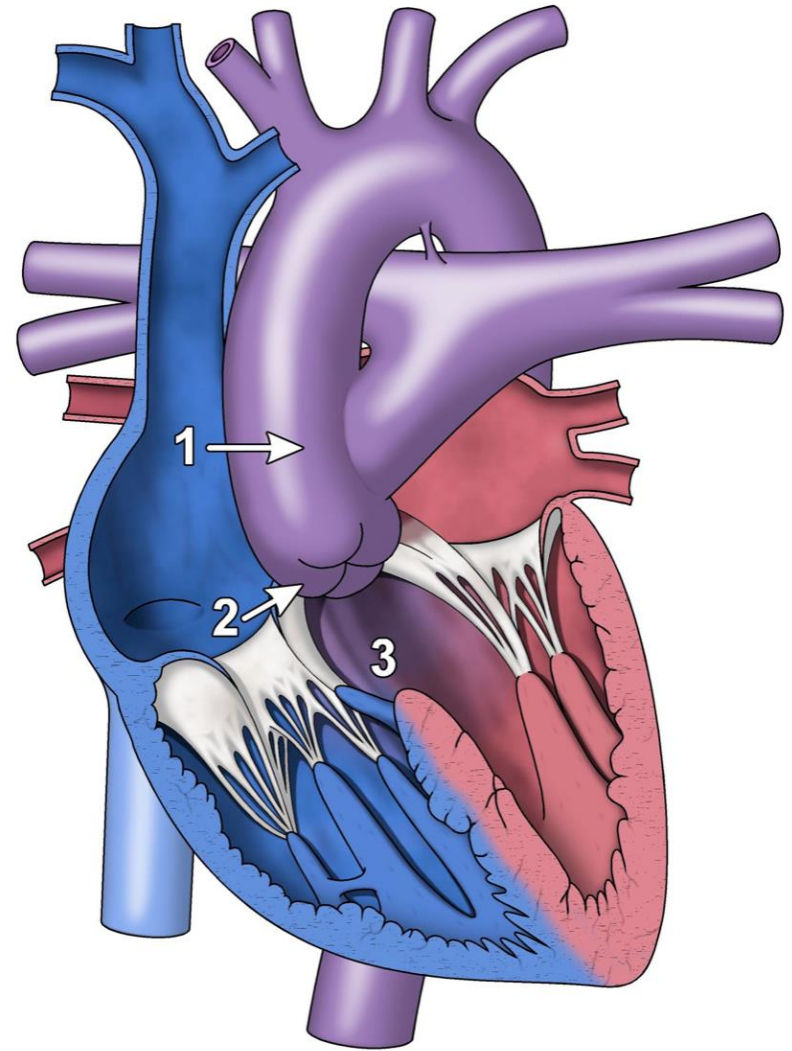


Shunt operation



Truncus arteriosus

- Verschillende types
- Type I (1)
- Afwijkende truncusklep (2)
- VSD (3)
- 22q11
- Symptomen
 - geringe cyanose
 - hartfalen



Persistent Truncal Artery

Definition & Characteristic Features

- **Definition**

Origin of ascending aorta, pulmonary arteries, and coronary arteries from a common truncal artery

- **Aortic arch** %

L aortic arch 70

R aortic arch 30

Interrupted arch 15

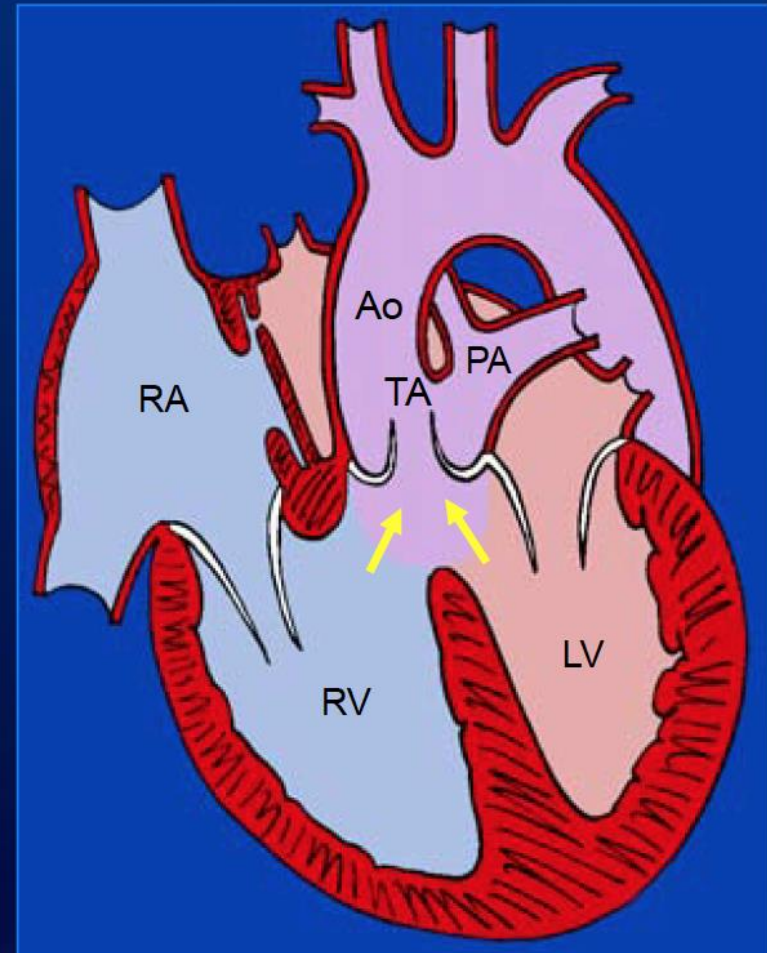
Hypoplasia/coarc 3

- **PA origin from TA** %

From main PA 60

Separate R/L PAs 25

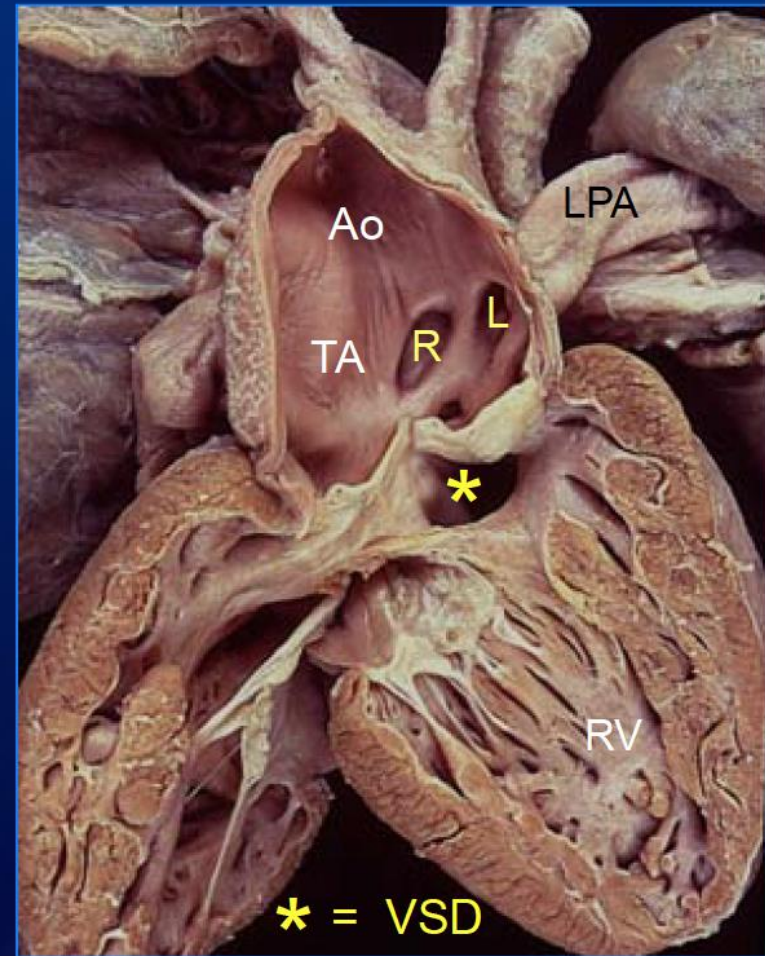
1 each: TA & aorta 15



Persistent Truncal Artery

Characteristic Features

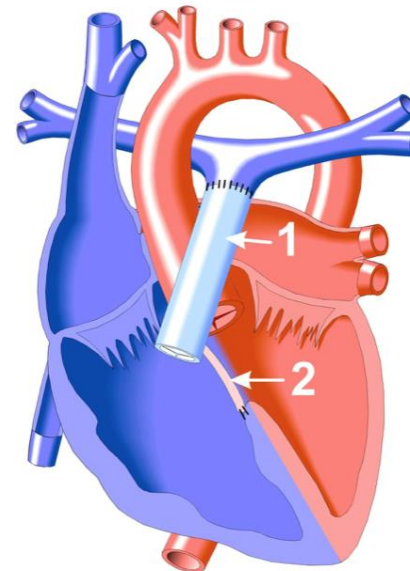
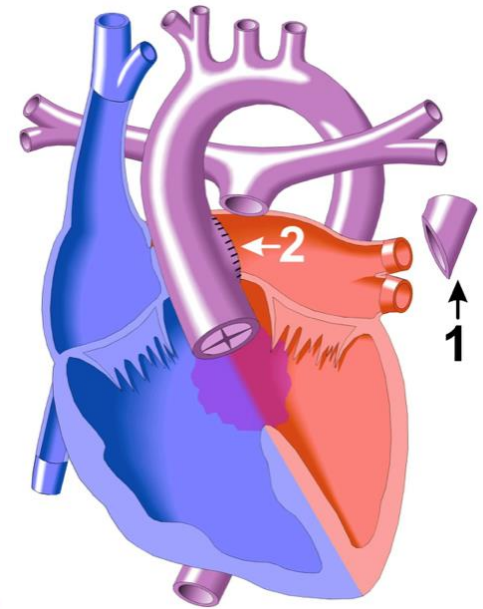
• Ductal artery	%
Absent (no remnant)	50
Present, patent	35
Present, closed	15
• Coronary arteries	%
Anom ostial origin	50
Single cor ostium	15
L cor from PA	<1
• Other conditions	%
Secundum ASD	15
L-SVC to cor sinus	10
Anom subclavian art	10
DiGeorge syndrome	15
Extracardiac anom	25



Autopsy (Opened RV)

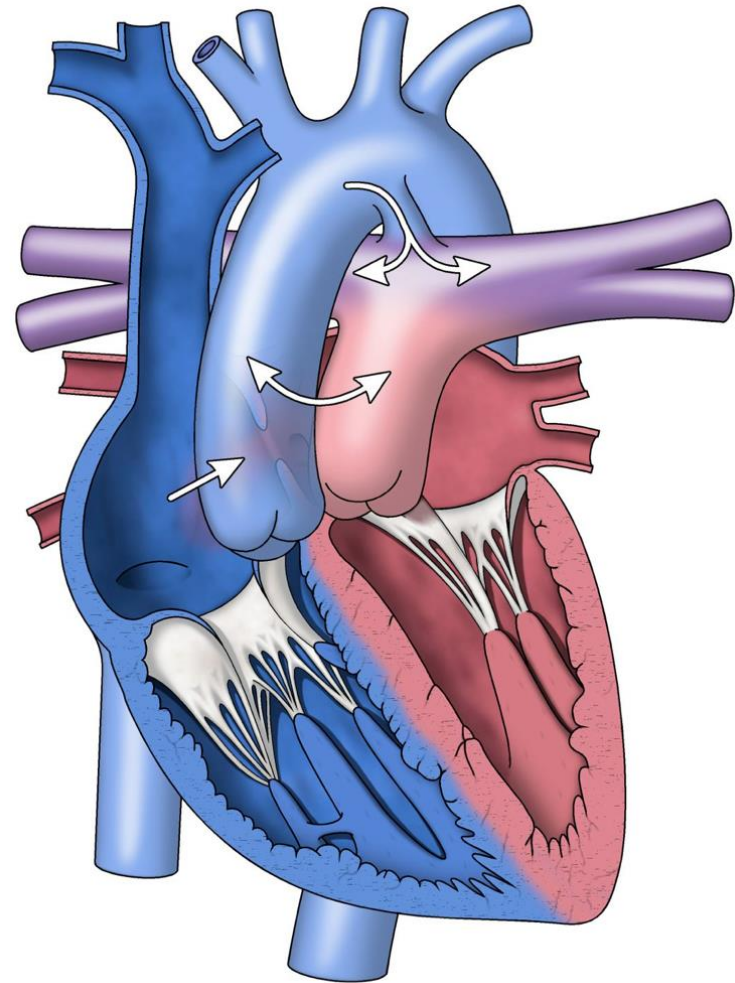
Correctie van een truncus arteriosus

- Vroege correctie binnen 1e-2e levensmaand
- Rastelli procedure
- Later soms
 - vervanging truncusklep door mechanoprothese



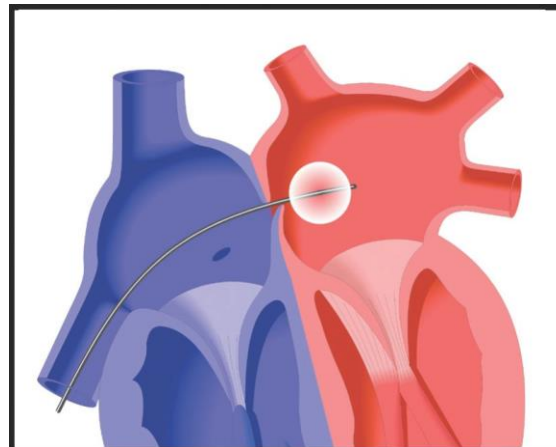
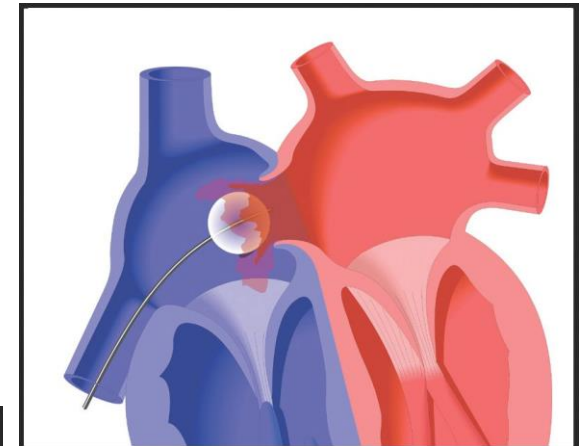
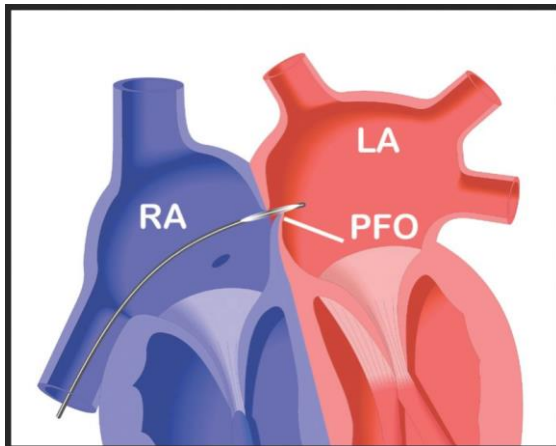
Transpositie van de grote arterieën (d-TGA)

- Na ToF vaakst voorkomende cyanotische hartafwijking
- 2/3 'simpele' TGA
- 1/3 complexe TGA
 - VSD
 - coronairanomalie
 - PS
- Symptomen
 - ernstige cyanose direct na geboorte

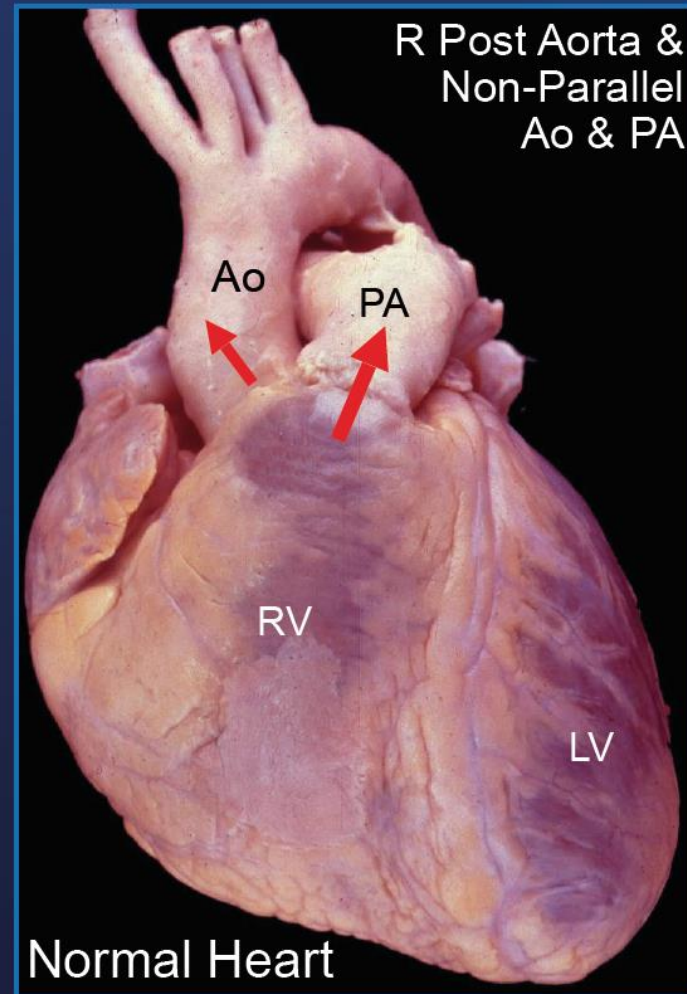
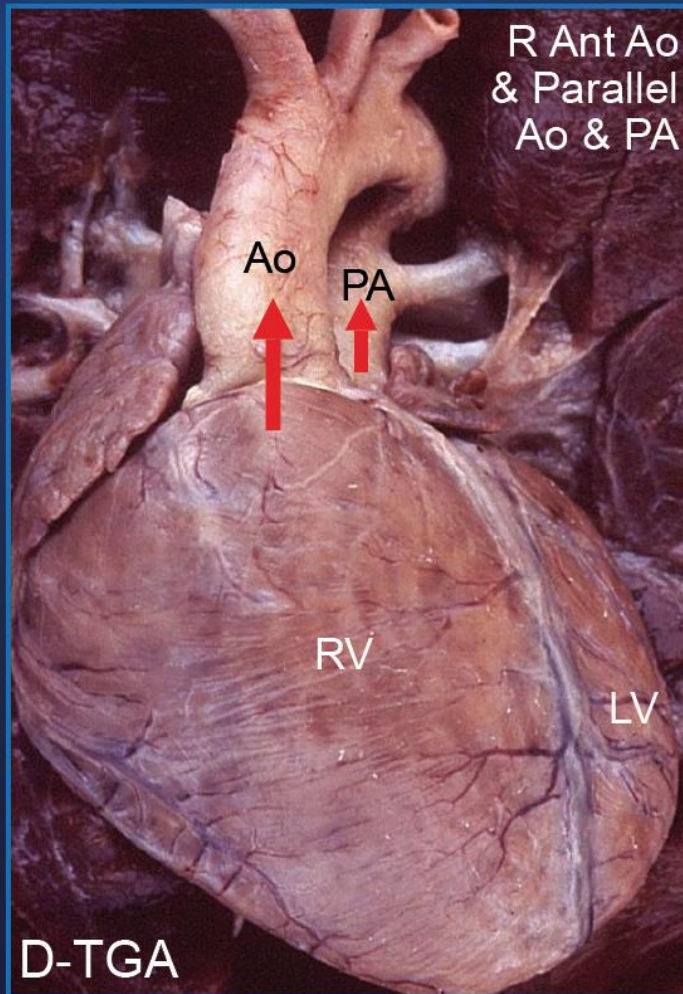


Transpositie van de grote arterieën (d-TGA)

- Atriale septostomie vlgs. Rashkind

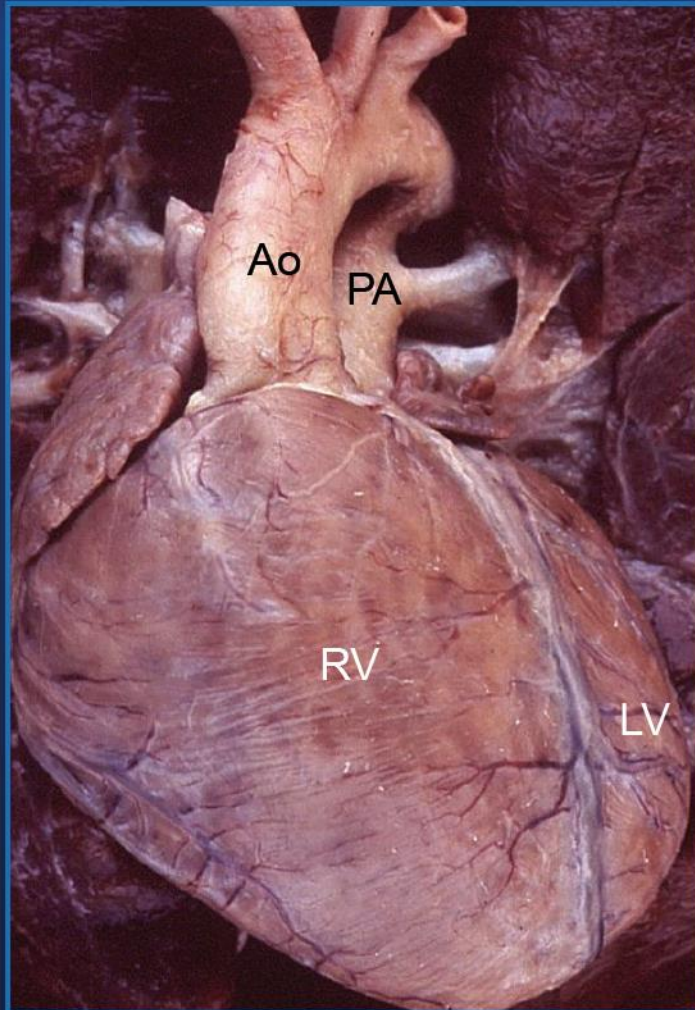


Complete Transposition (D-TGA) Features

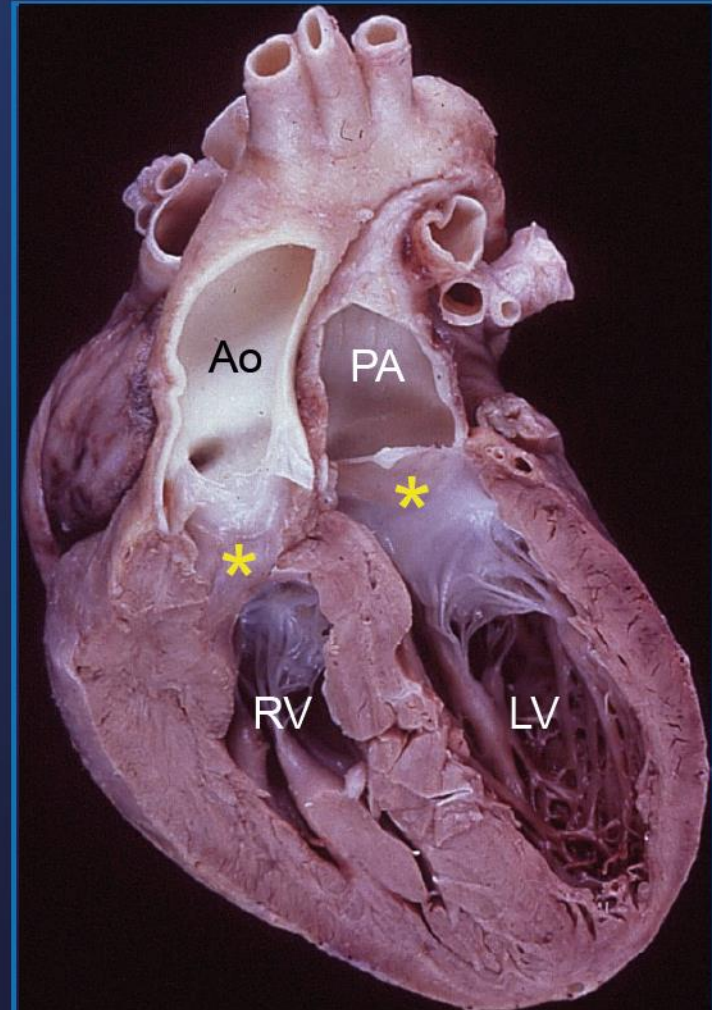


Autopsy Specimens (Anterior Views)

Complete Transposition (D-TGA) Features



Anterior View



Outflow Tracts

Complete Transposition (D-TGA)

Features

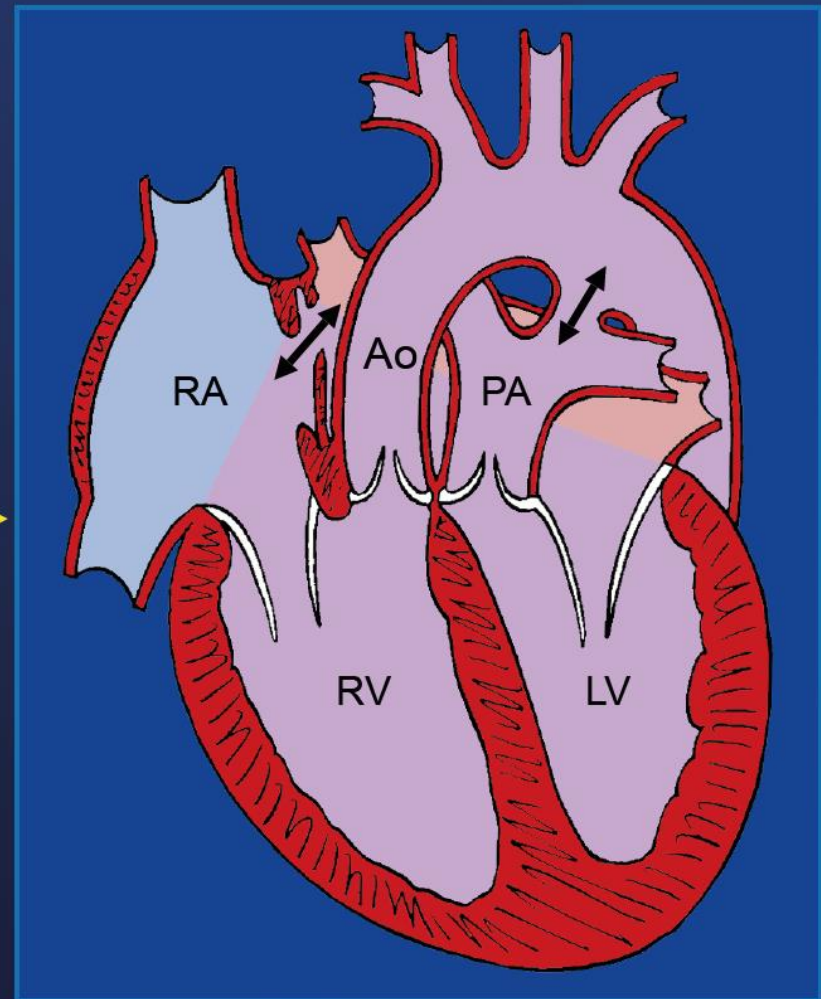
- **Four types** %

TGA + intact VS	65
TGA + VSD	20
TGA + VSD + PS	10
TGA + IVS + PS	5

- **Occurrences** %

TGA + IVS $\pm$ PS	70
TGA + VSD $\pm$ PS	30
PS in TGA + VSD	33
PS in TGA + IVS	7

- **Obligatory shunt**
 - Stable: VSD or ASD
 - Unstable: PFO or PDA



Complete Transposition (D-TGA)

Features

- **Four types** %

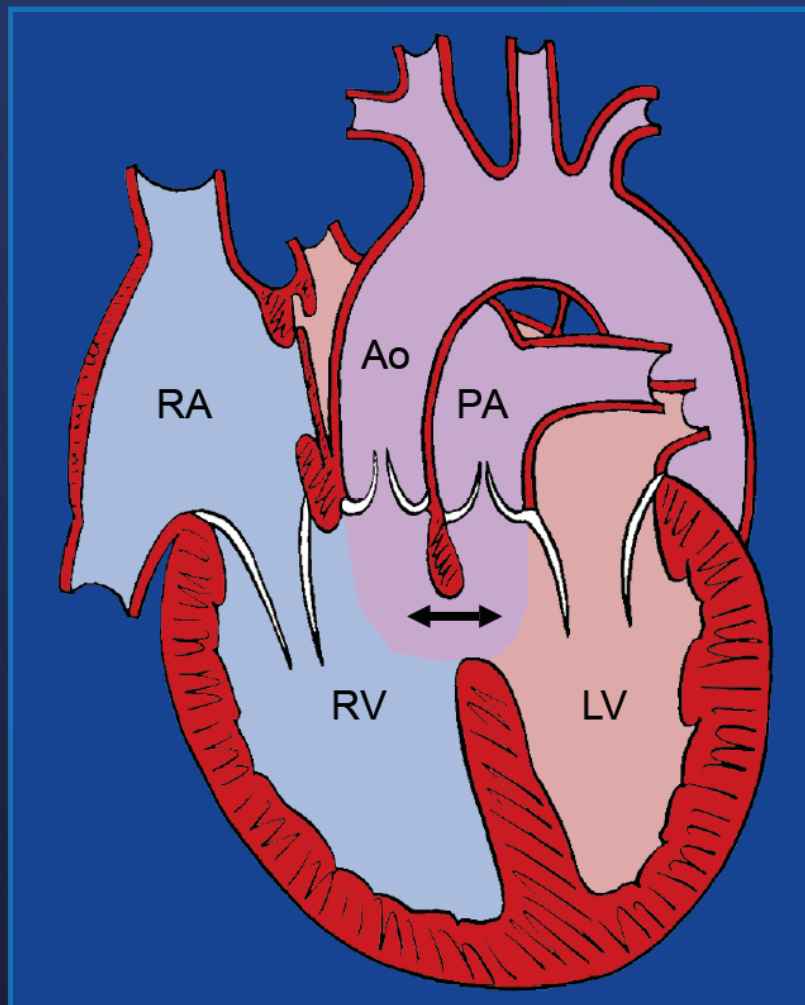
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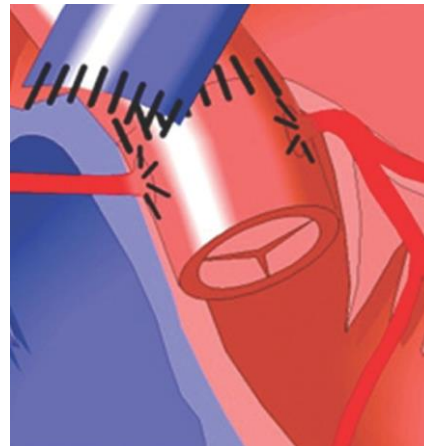
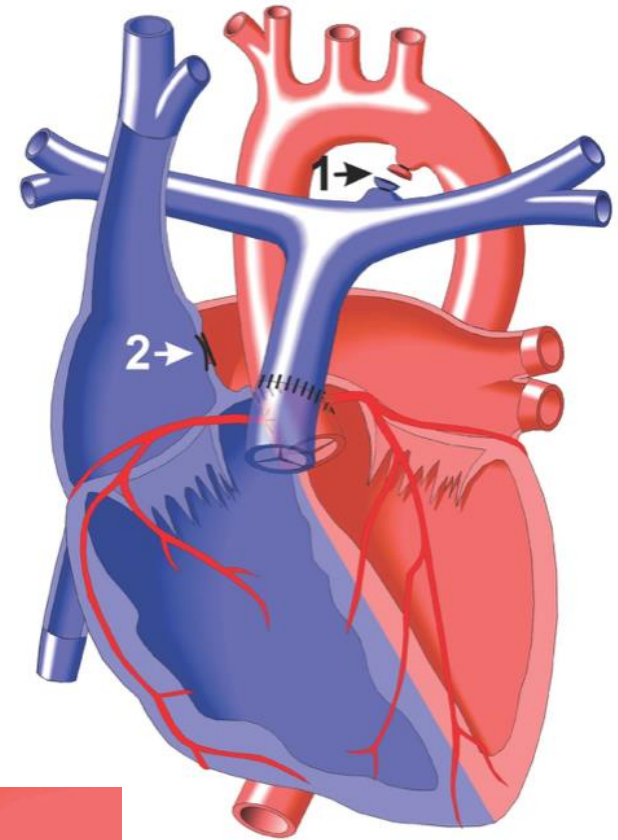
- **Obligatory shunt**

Stable: VSD or ASD
Unstable: PFO or PDA



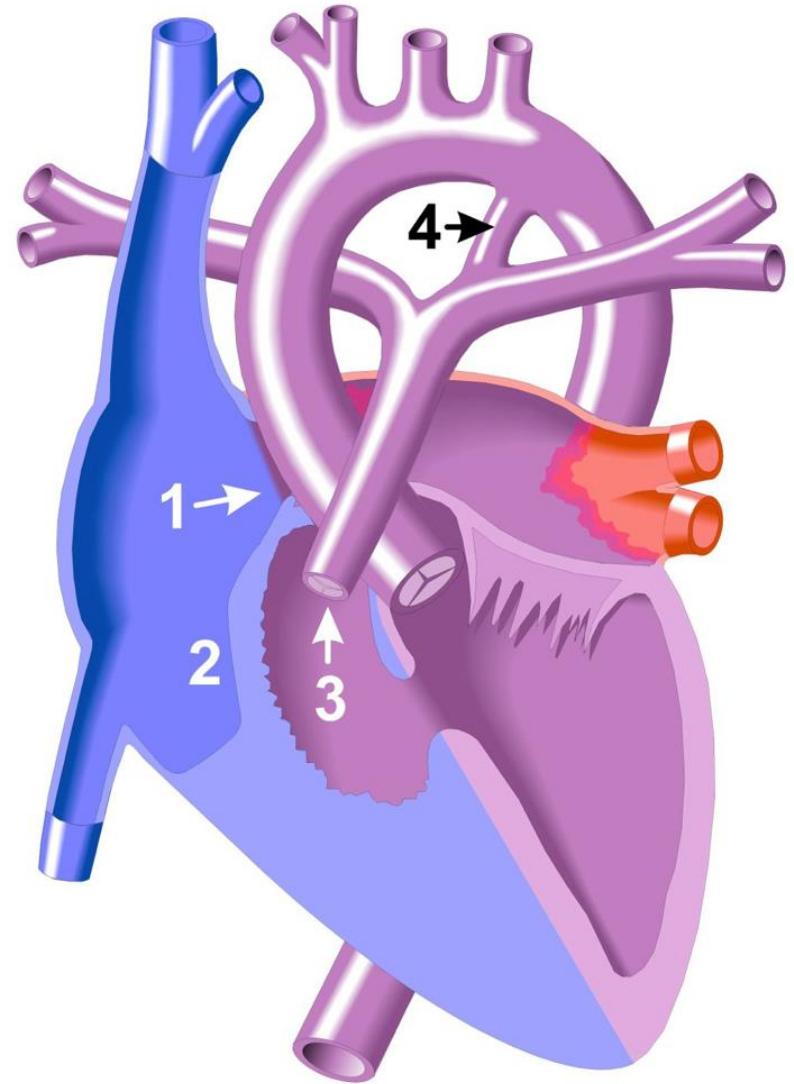
Correctie van een d-TGA

- Binnen eerste 2 (-4) weken
- Arteriële switch operatie
- Bij operatie na 2e maand
 - LV training door PA banding
- Mortaliteit
 - 'simpele' TGA <5%
 - complexe TGA $\pm 10\%$



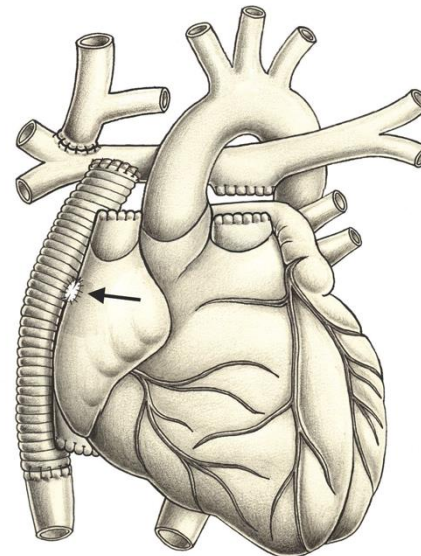
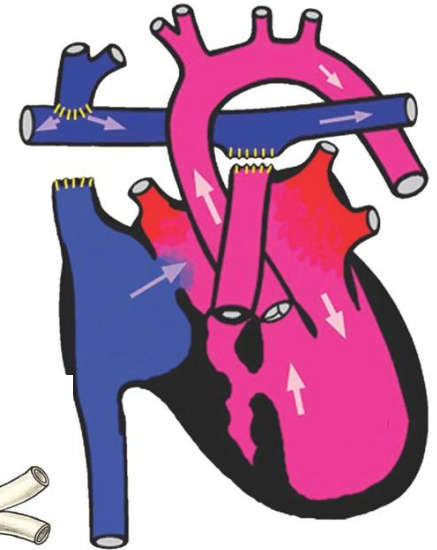
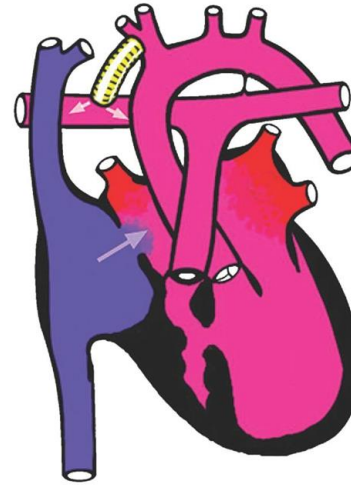
Tricuspidalisatresie (TA)

- 3 types afhankelijk positie grote arterieën
- Vaakst: TA met normale positie vd grote arterieën en PS
 1. ASD
 2. TA
 3. Pulmonalisstenose
 4. PDA



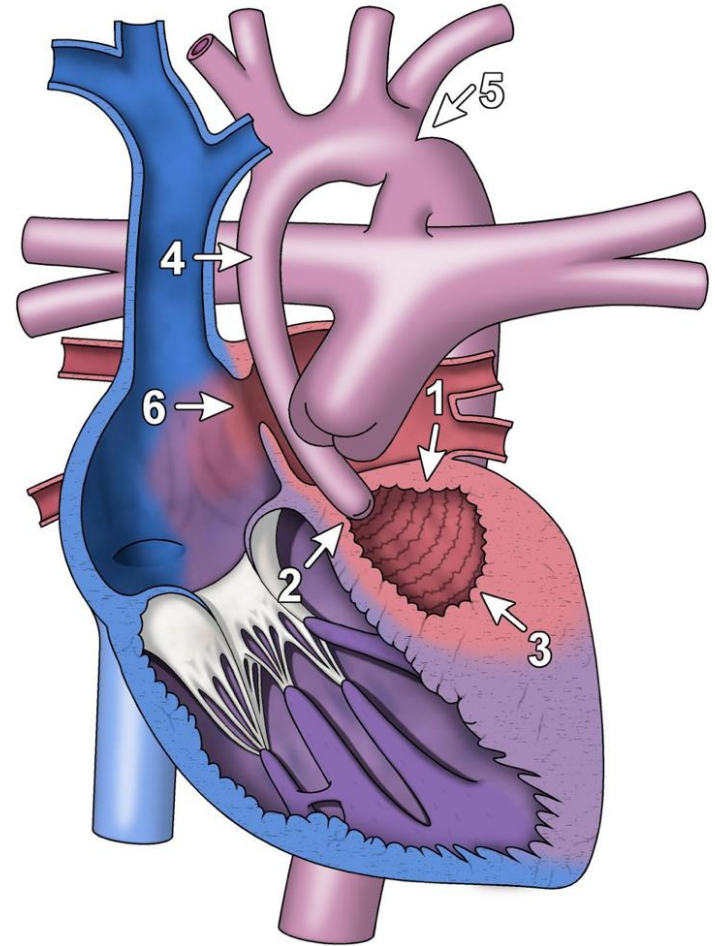
Correctie van TA

- Modified Blalock-Taussig shunt (MBTS)
- Bij TA met TGA
 - Damus-Kaye-Stansel anastomose
- 3 maanden
 - aanleggen partiële cavo-pulmonale connectie (PCPC)
- 2 jaar (12 kg)
 - aanleggen totale cavo-pulmonale connectie (TCPC, 18 mm Gore-Tex tunnel)



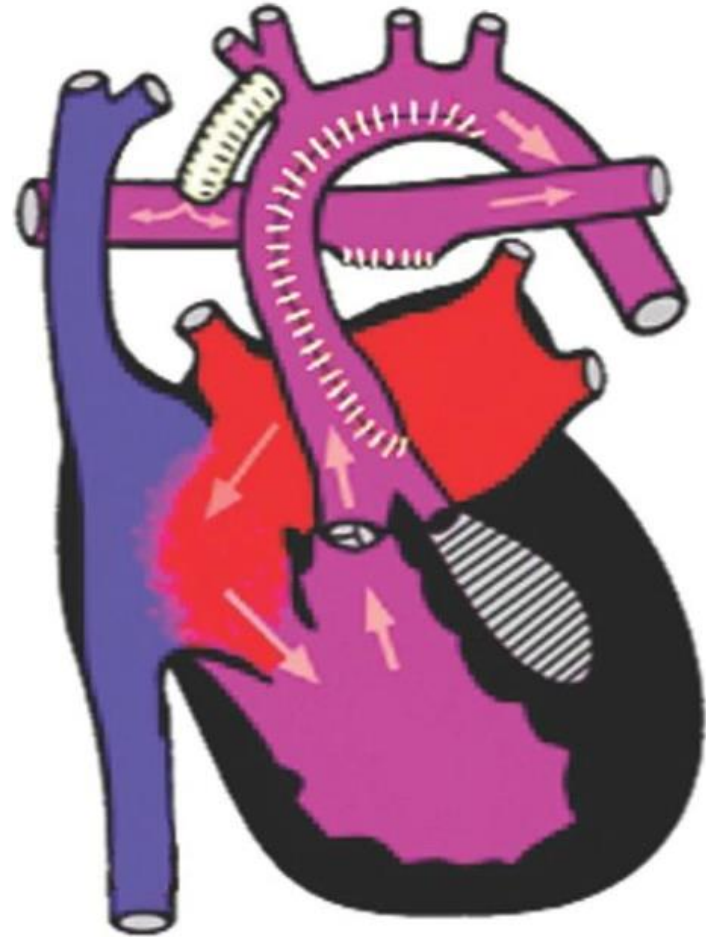
Hypoplastisch linker hartsyndroom (HLHS)

1. Atresie of stenose mitraalklep
2. Atresie of stenose aortaklep
3. Hypoplastisch LV
4. Hypoplastische aorta ascendens
5. Coarctatio aortae
6. ASD



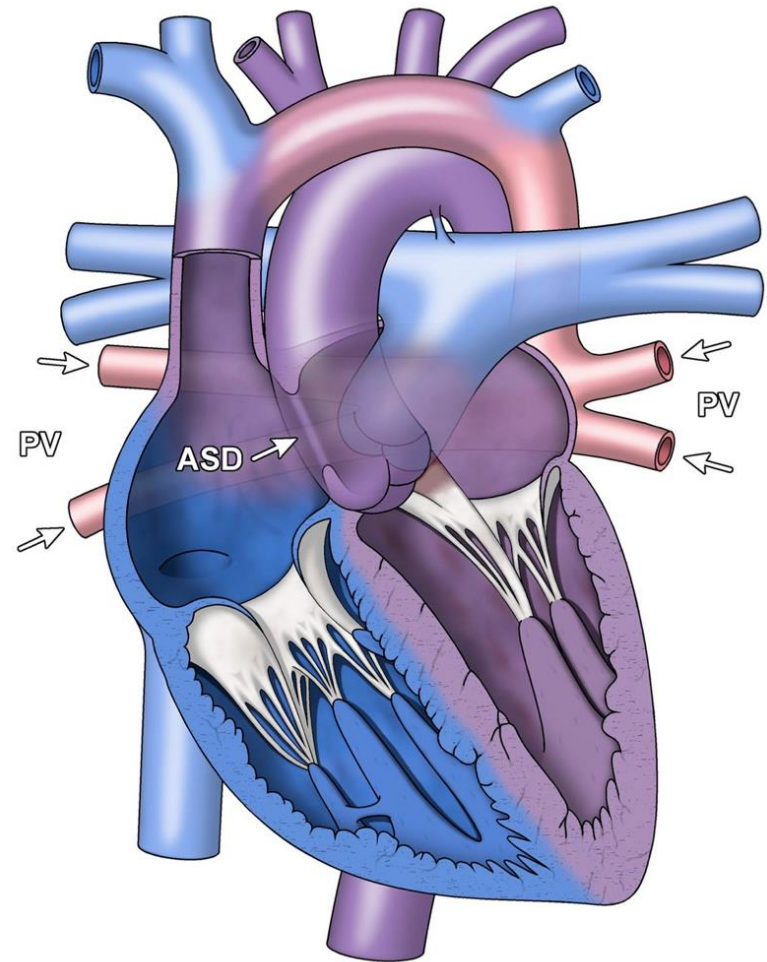
Correctie van HLHS

- Norwood 1 procedure
- PCPC
- TCPC



Totaal abnormale PV connectie (TAPVC)

- 4 types
 - supracardiaal
 - cardiaal
 - infracardiaal
 - Gemengd
- Vaak geïsoleerd
- Soms
 - TGA, ToF, PTA, HLHS, TA, CoA, AVSD
 - Heterotaxie syndroom
- Bij pulmonaalveneuze obstructie > spoed OK
- Electieve correctie binnen 1e maanden





Dank voor jullie aandacht