# Malformatii Cardiace Congenitale

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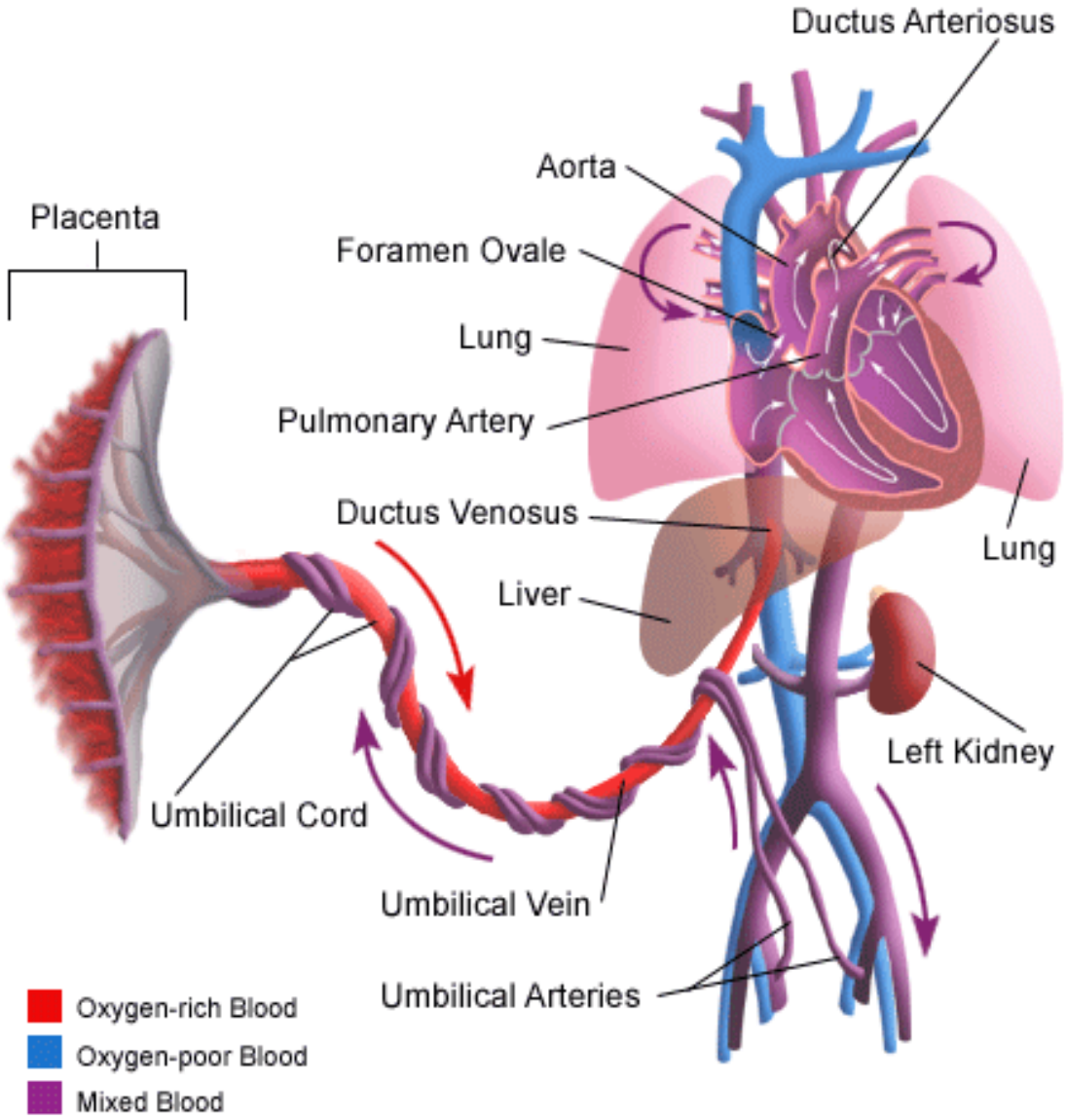
# Prevalenta

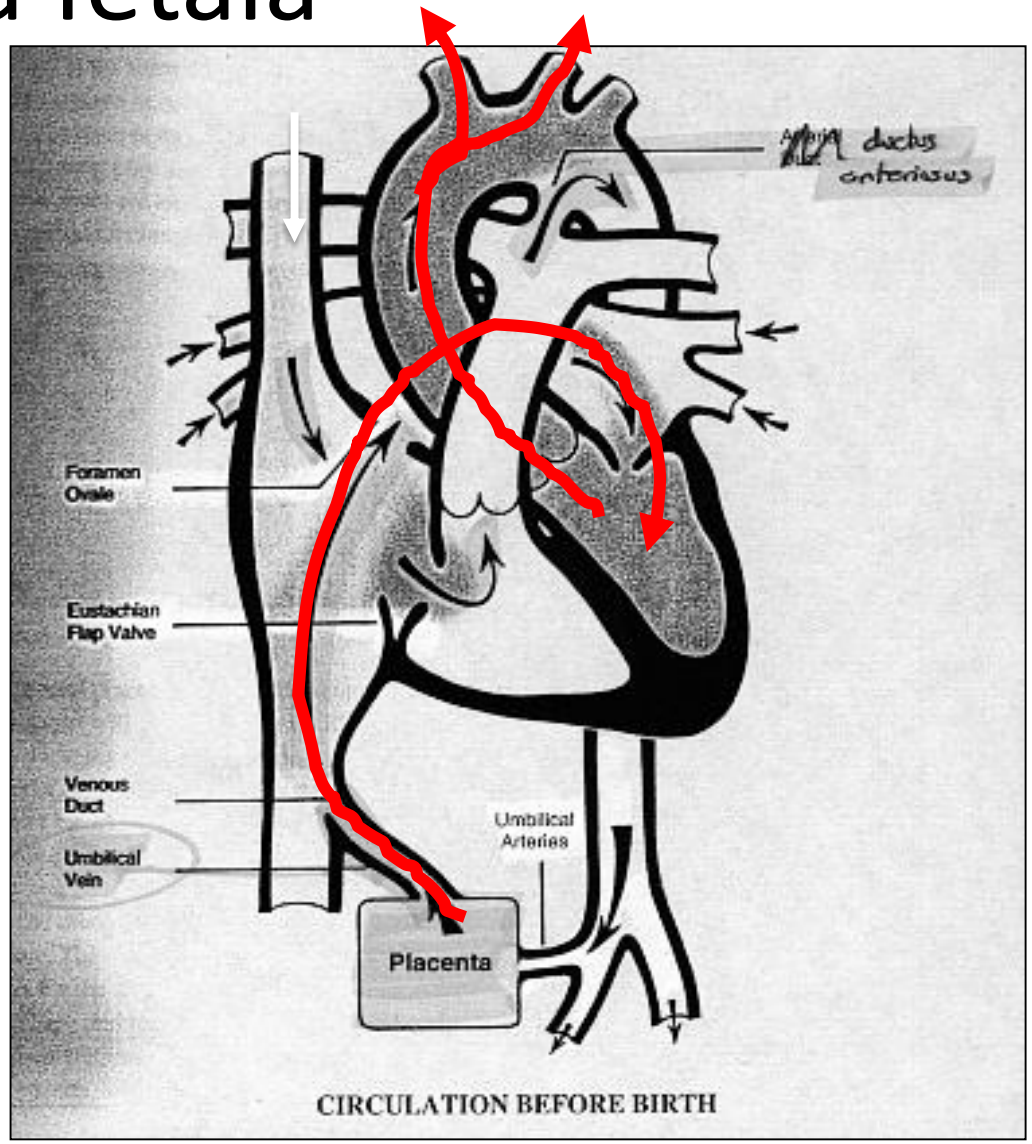
- 1/125 nasteri
- Cea mai frecventa malformatie congenitala
- Majoritatea nu au o cauza cunoscuta
- Apar in primele 8 saptamini de viata

# Fetal Circulation



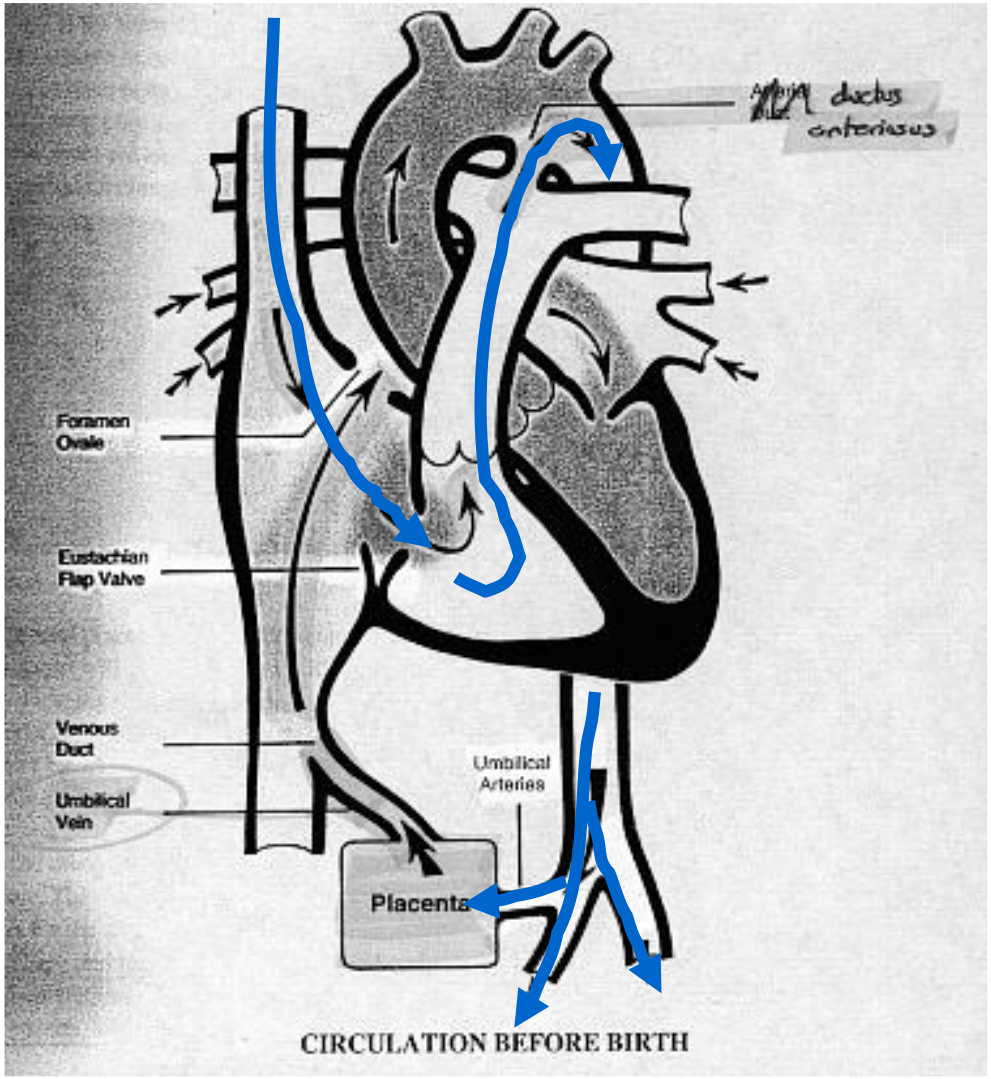
# Circulatia fetala

- Placenta
- VCI
- AD
- Prin FO
- AS
- VS
- Ao ascendenta
- Cap si mbr. sup.
- Retur prin VCS



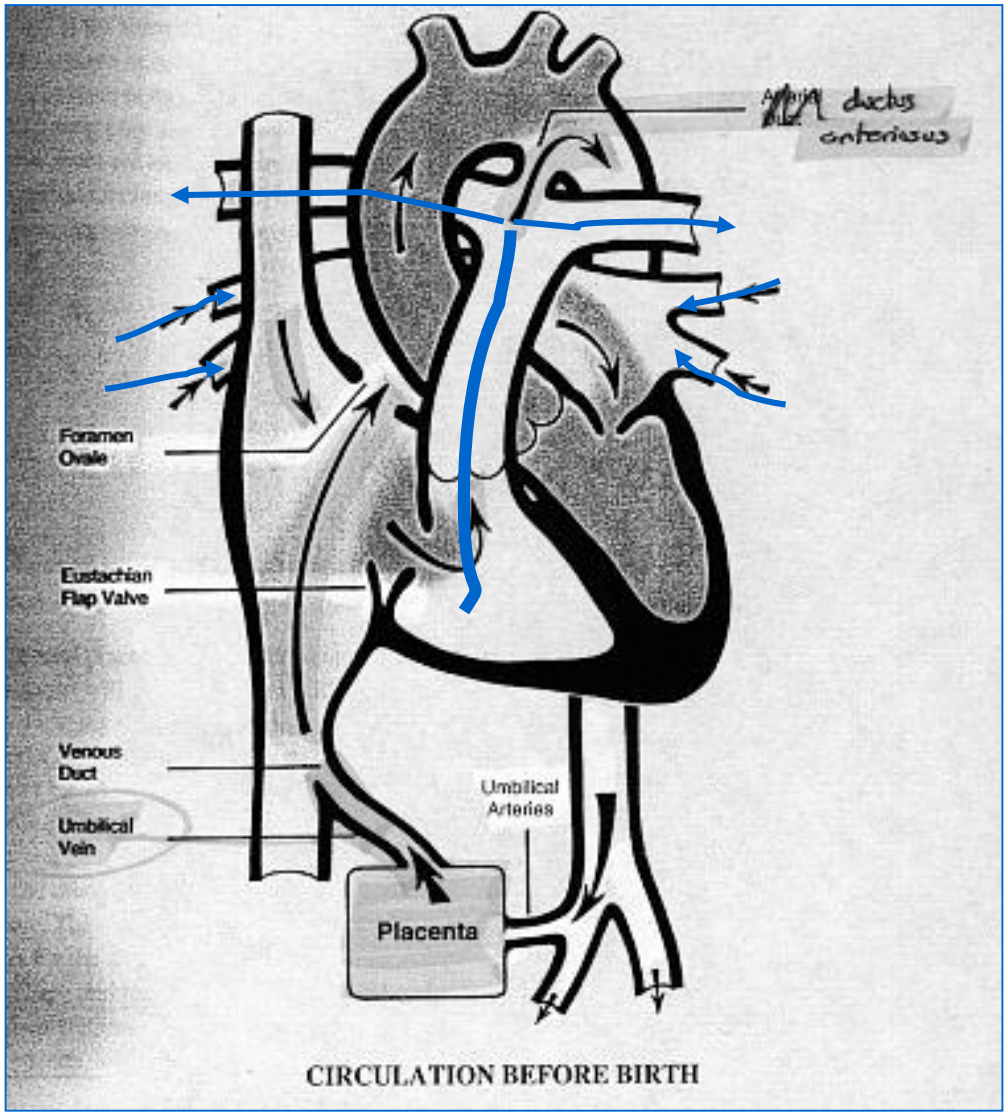
# Circulatia fetala

- VCS
- AD
- VD
- AP
- Prin CA
- Ao desc.
- Mbr. inf. si placenta



# Circulatia fetala

- Doar o mica cantitate de singe ajunge prin arterele pulmonare la plamin

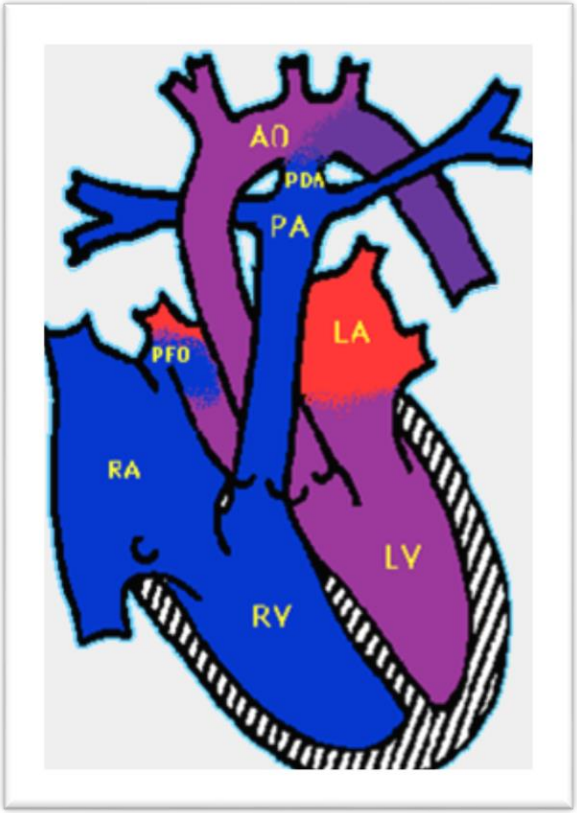


# Circulatia fetala -> neo-natala

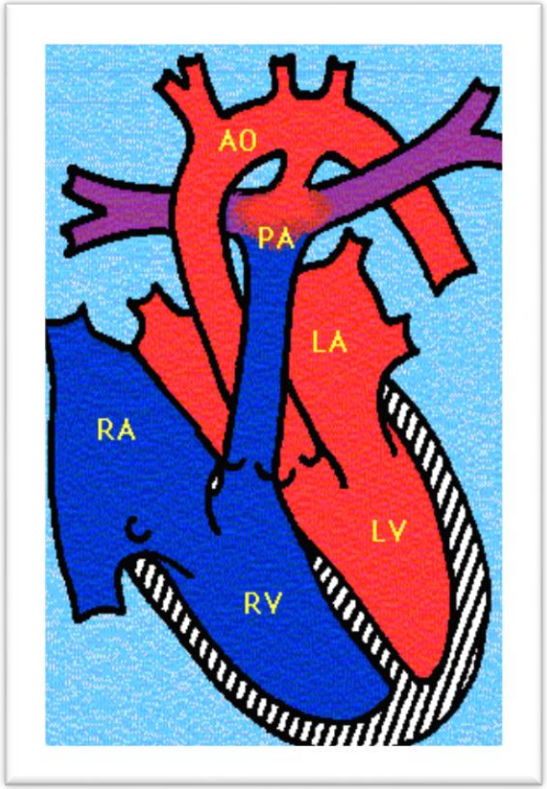
1. Crestere rezistentelor sistemice (pierderea placentei)
2. Scaderea rezistentelor pulmonare (respiratia)

- Inchiderea FO
- Inchiderea CA

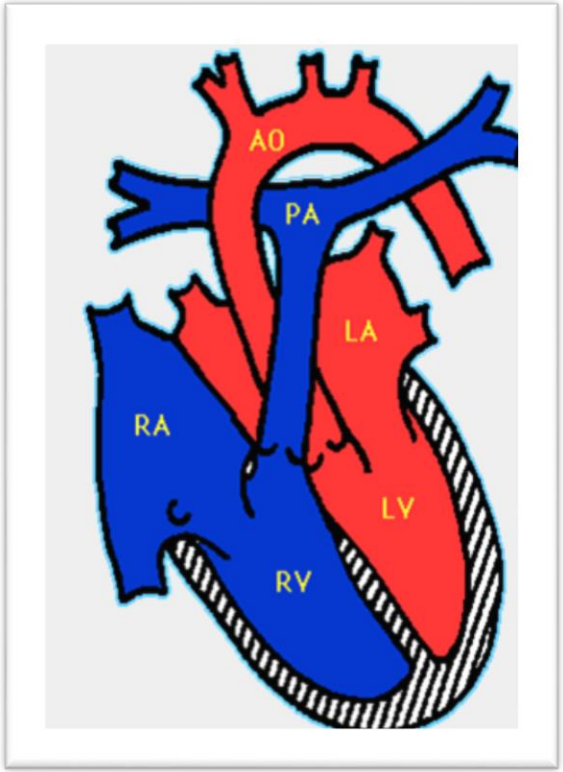
# Circulatia fetala -> neo-natala




Fetala




La 8 ore




24-72 ore

# Diferente fata de adult

- Miocard imatur
  - Procent mai mare de tesut conjunctiv (necontractil)
  - Dependenta de Ca ionizat plasmatic
- Volum bataie “fix”
- Necesar energetic mai mare
- Dominanta parasimpatica
- Lactat / glucoza ca sursa de energie (nu acizi grasi)

# Diferente fata de adult

- Consecinte:
  - DC dependent de frecventa
  - Rezerva scazuta
  - Raspuns “paradoxal” la ischemie
  - Raspuns redus la inotropice

# Valori normale

| Virsta | Greutate (kg) | TAs (mmHg) | Frecv. card. | Respiratii |
|---|---|---|---|---|
| nn | 3,5 | 60 – 100 | 110-170 | 25-60 |
| 3 -6 luni | 6 - 8 | 65 – 115 | 105-165 | 25-55 |
| 1 -4 ani | 10 - 15 | 70 – 120 | 85-150 | 20-40 |
| 6 – 10 ani | 20 – 30 | 80 – 130 | 70-135 | 15-35 |
| Adult | 70 | 95 – 140 | 60-120 | 15-25 |

# Clasificare fiziopatologica

1. Debit pulmonar crescut
2. Debit pulmonar scazut
3. Obstructive (fara șunt)
4. Severe – incompatibile cu circulatia postnatala
5. Dormante / silentioase

# Clasificare fiziopatologica

1. Debit pulmonar crescut
2. Debit pulmonar scazut
3. Obstructive (fara șunt)
4. Severe – incompatibile cu circulatia postnatala
5. Dormante / silentioase

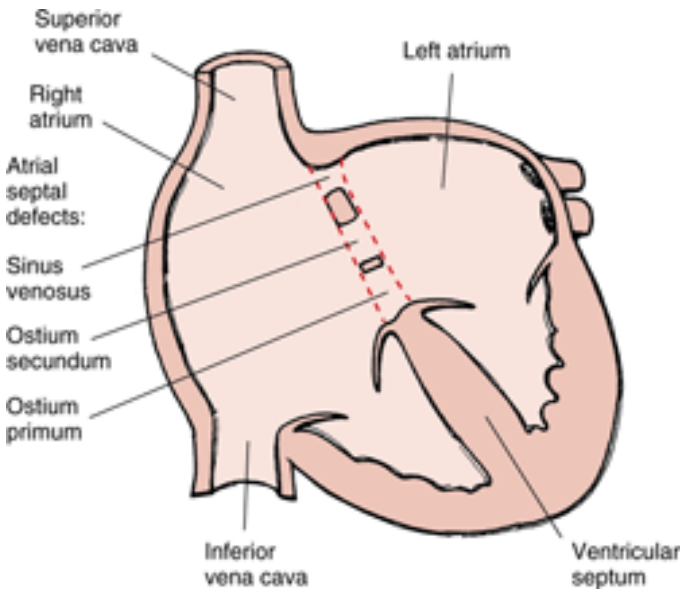
# Debit pulmonar ↑

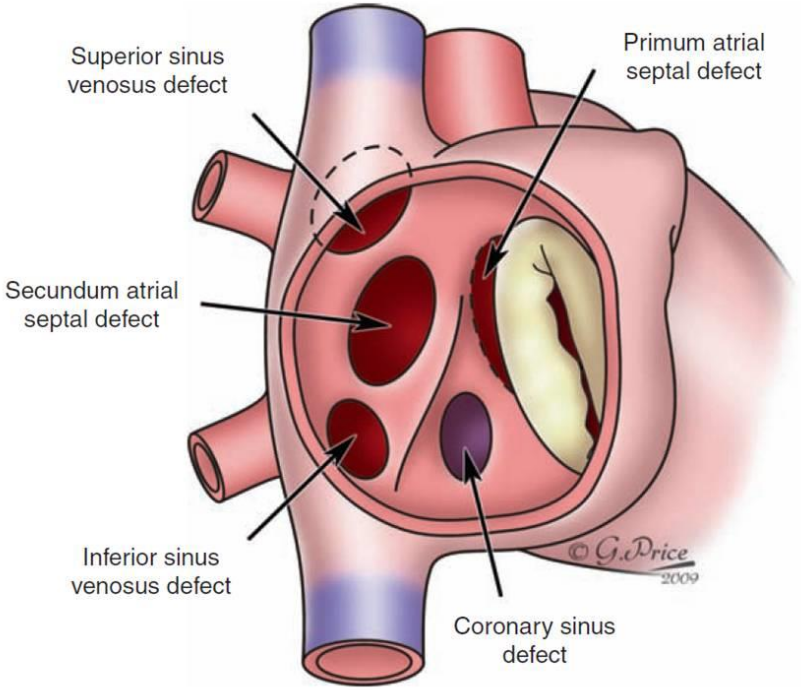
**Sediu**

- Venos
- Sept atrial
- Jonctiunea AV
- Sept ventricular
- Ao - AP

# DSA

- 10% din MCC
- OS 80%, OP 10%, sinus venosus 10%



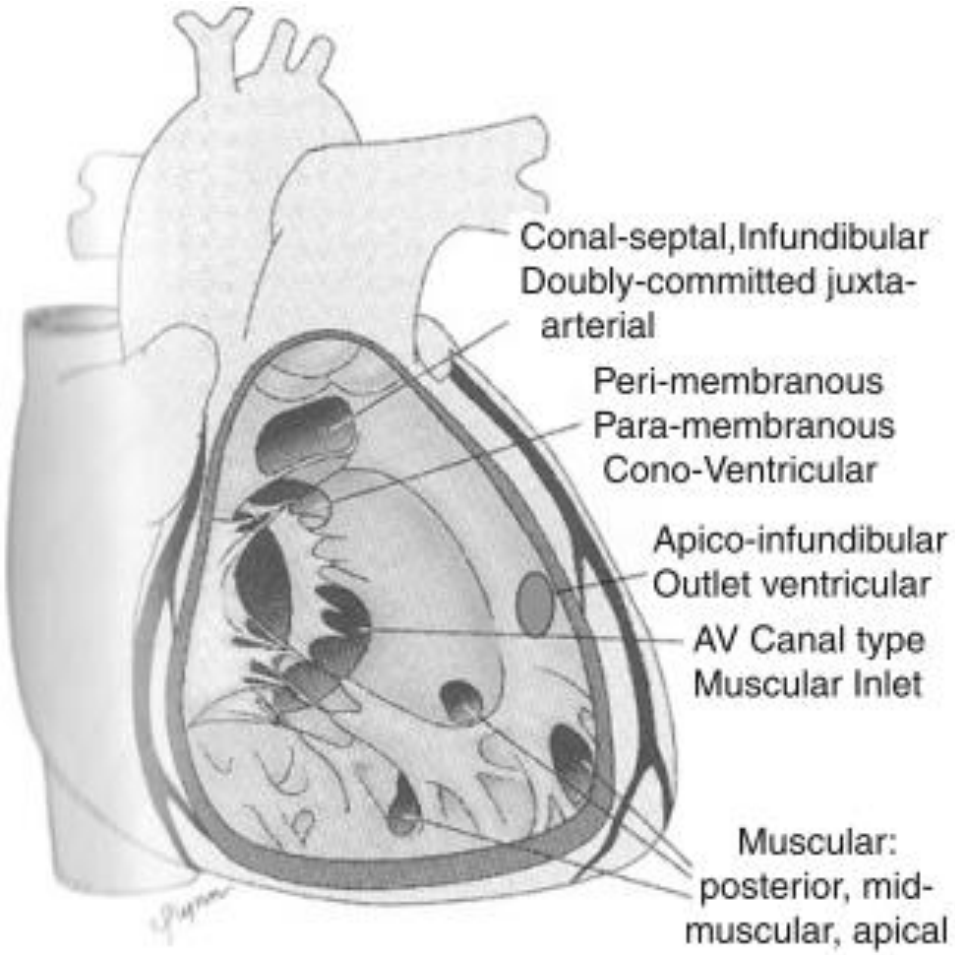
# DSA

- Majoritatea asimptomatici / impact fiziopatologic minim
- Directia suntului depinde de presiuni
- Tratament:
  - Interventional
  - Chirurgical
- Complicatii: putine

# DSV

- 30% - 40% din MCC
- Doar 30% sunt izolate
- 90% musculare
- Sunt S-D important; depinde de marime, rezistente, eventuale obstructii (Ao sau pulm.)

# DSV


Conal-septal,Infundibular
Doubly-committed juxta-arterial
Peri-membranous
Para-membranous
Cono-Ventricular
Apico-infundibular
Outlet ventricular
AV Canal type
Muscular Inlet
Muscular:
posterior, mid-muscular, apical

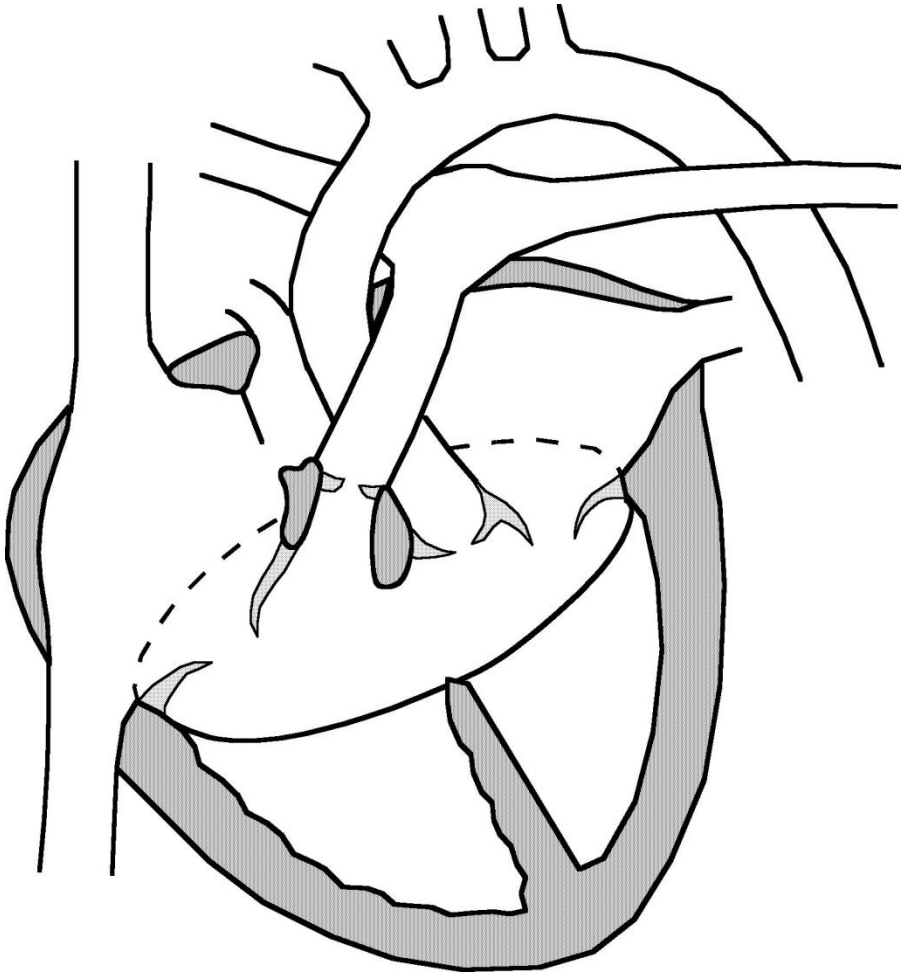
# DSV

**Management**

- Multe se inchid spontan
- Risc de HTP > 1 an (sau 6 luni Down)
- Diuretic, IEC, nutritie
- Profilaxia endocarditei (?)
- Interventional
- Chirurgical: corectie sau banding AP

# CAVC

- Valva AV unica
- Grade variante:
  - Partial
  - Incomplet
  - Complet
- OP, DSV, cleft VM



# CAP

- Patenta ductului > 8 saptamini
- 10% din MCC
- 90% izolate; f frecvent la prematur
- Fluxul dependent de : rezistente (S ; P), dimensiune
- Consecinte: hiperaflux pulmonar, HTP, supraincarcare AS/VS, furt diastolic

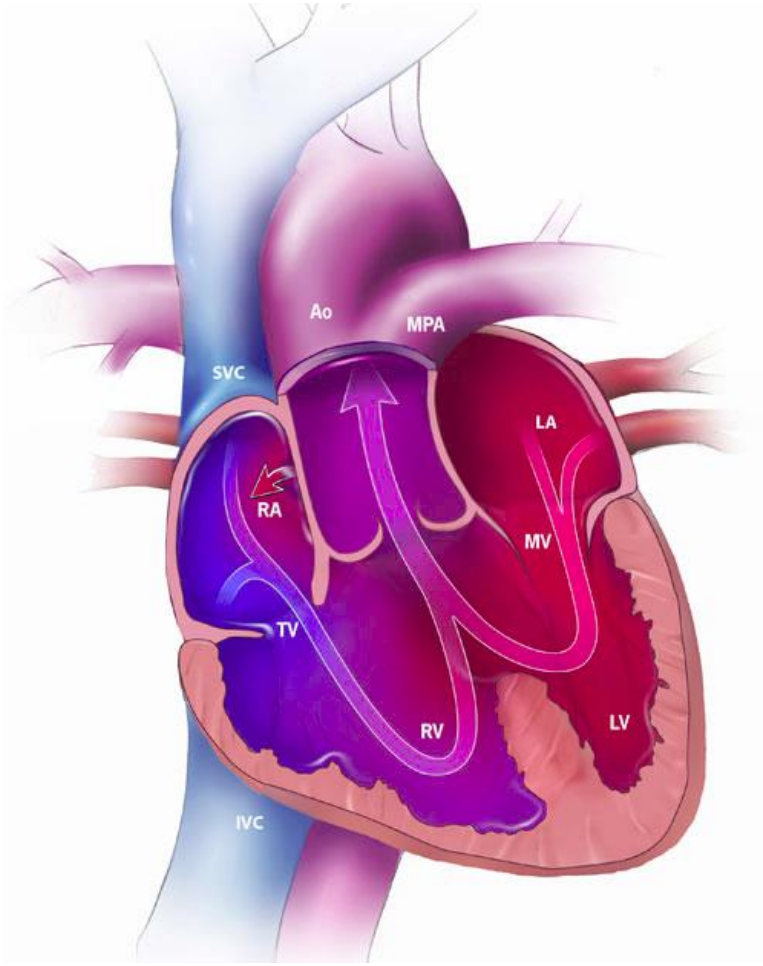
# CAP

**Terapie**

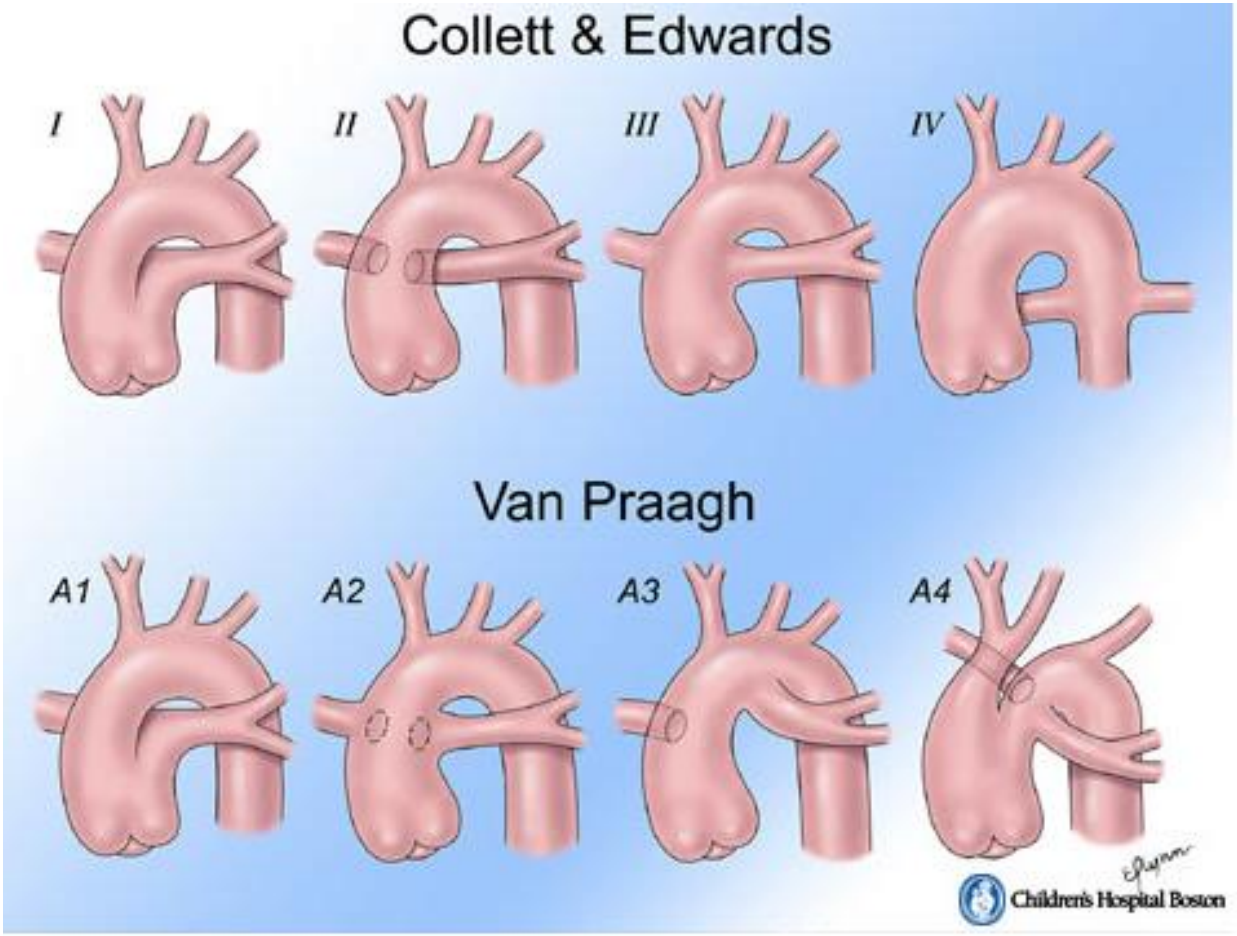
- Conservativ
- Medicamentos: Indometacin, Ibuprofen
- Interventional
- Chirurgical

# TAC

- Vas unic care emerge din ambii ventriculi – calare pe septul IV



# TAC


Collett & Edwards
I
II
III
IV
Van Praagh
A1
A2
A3
A4
Children's Hospital Boston

# TAC

- Origine comuna a celor 2 circulatii
- Saturatii egale in Ao si AP
- Presiuni egale in Ao si AP
- Debitele si Qp/Qs dependente de rezistente

# TAC

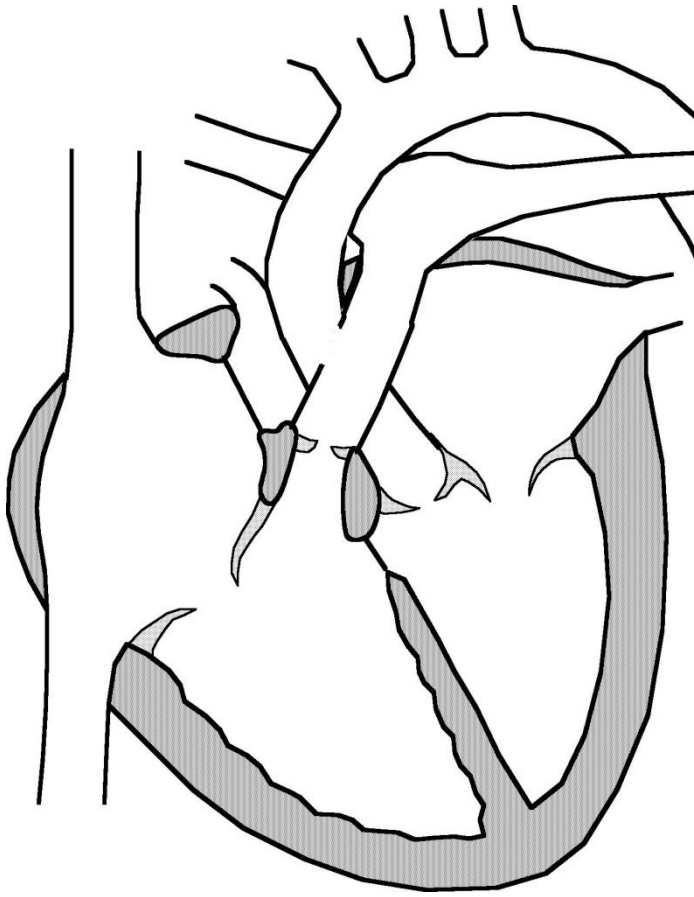
- Initial sunt cianotici, cu 75-80%
- Pe masura ce scad RVP creste fluxul in a. pulmonara si saturatia
- Suntul S-D apare si in sistola si in diastola (spre deosebire de DSV de ex.)
- Apare supraincarcarea de volum – insuficienta cardiaca
- Insuficienta valvei truncale agraveaza insuficienta cardiaca

# TAC

- Urgenta neonatala
- PREOP:  minimizarea fluxului in pulmonara (fara O2, hipoventilatie, sedare, PEEP, diuretic
- CHIRURGICAL: inchiderea DSV, separarea Ap de Ao, conduct VD-AP +/- plastia valvei Ao

# Fenestratia Ao-Pulm

- Defect de embriogeneza in separarea TAC in Ao si AP
- Comunicare intre Ao asc si trunchiul AP
- Simptomatologie de la discret la catastrofic



# Fenestratia Ao-Pulm

- Managament
- Similar TAC, VU cu circ. pulm. nerestrictiva
- Pe masura ce RVP ↓ suntul ↑
- Inclinarea balantei spre circ. sistemica
  - Sedare, curarizare
  - Hipoventilatie, PEEP
  - Diuretic
  - PGE1 (CoAo, arc Ao intrerupt)

# Debit pulmonar ↓

Obstructie in circulatia pulmonara **+** comunicare intracardiaca

Șunt dreapta – stânga -> desaturare sistemica

Cianoză

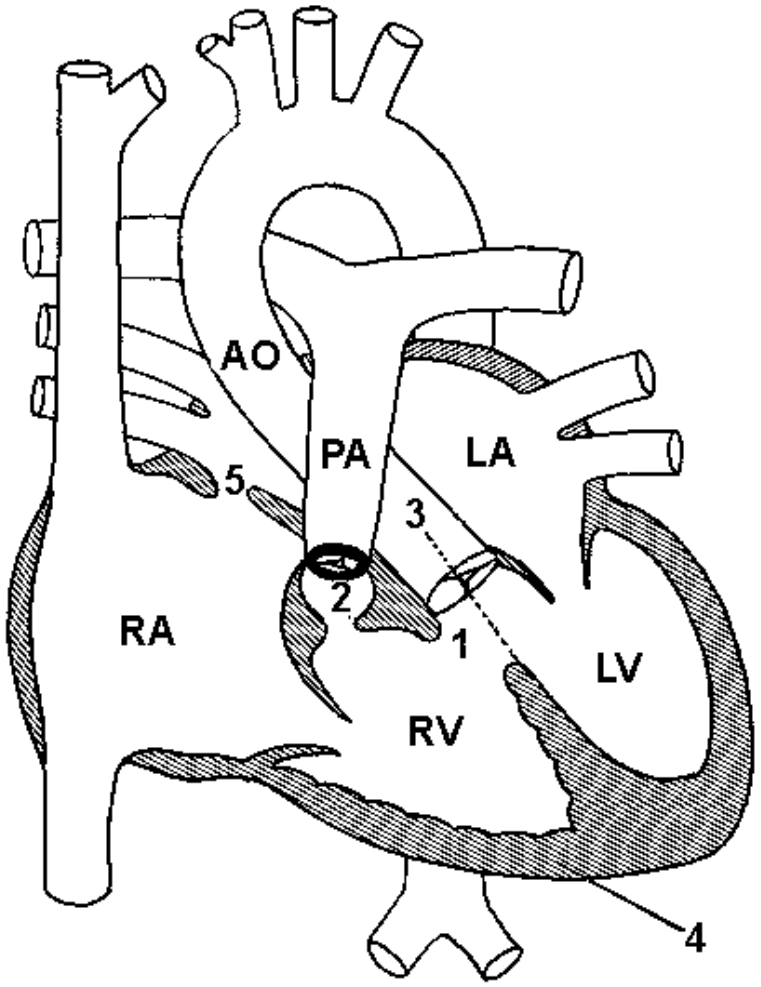
- Hipoxie cronica
- > 2,5 g% de-oxihemoglobina

# Debit pulmonar ↓

1. Stenoza pulmonara + DSA
2. Stenoza pulmonara + DSV (Fallot)
3. Atrezie tricuspida
4. Boala Ebstein
5. Ventricul unic (double-inlet) + stenoza pulmonara

# Tetralogia Fallot

1. DSV
2. SP
3. Ao “călare”
4. HVD



# Tetralogia Fallot

- Leziunea dominanta este obstructia tract ejectie VD
- Crize cianogene <- stenoza dinamica
  - Deshidratare
  - Febra
  - Agitatie
  - Anemie
  - acidoza

# Tetralogia Fallot

- Masuri
  - Volum
  - ↑ RVP (postural, farmacologic)
  - Oxigen, $NaHCO_3$
  - Beta-blocant, sedare
  - ↓ consum (febra, sedare)

# Tetralogia Fallot

POSTOP

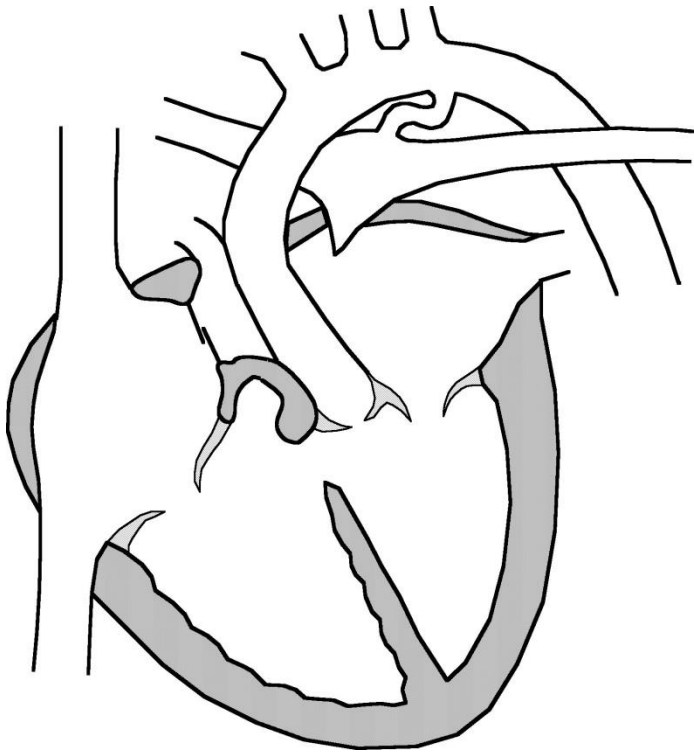
- VD cu complianta ↓↓↓ -> sdr. DC scazut
  - Inotropic lusitrop
  - PVC mare
  - ↓ RVP
  - Sustinerea TAm
  - Sincronism AV si VV
  - VM cu presiune mica

# Atrezia pulmonara + DSV

- Absenta continuitatii VD-AP
- DSV

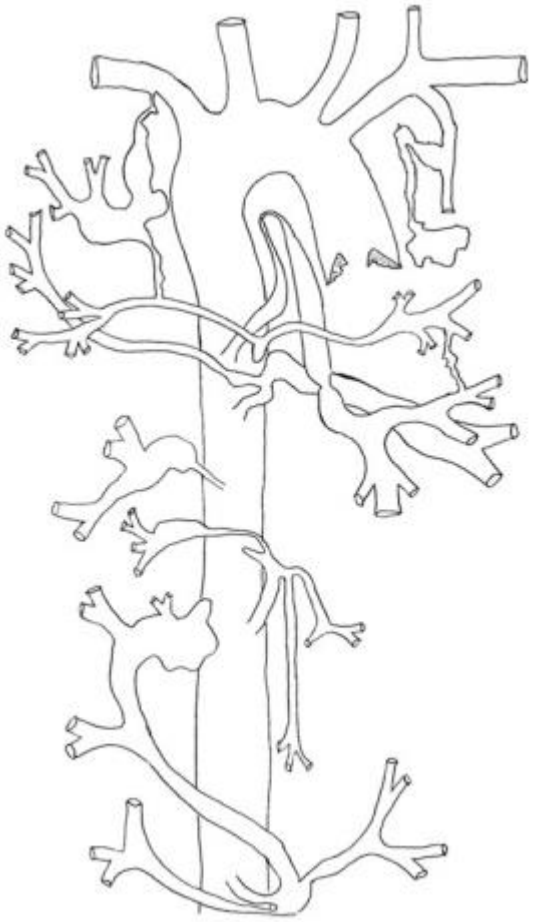
- VP – imperforata sau absenta
- AP – normala sau absenta

- CAP sau MAPCA sau colaterale intraparenchimatoase



# Atrezia pulmonara + DSV

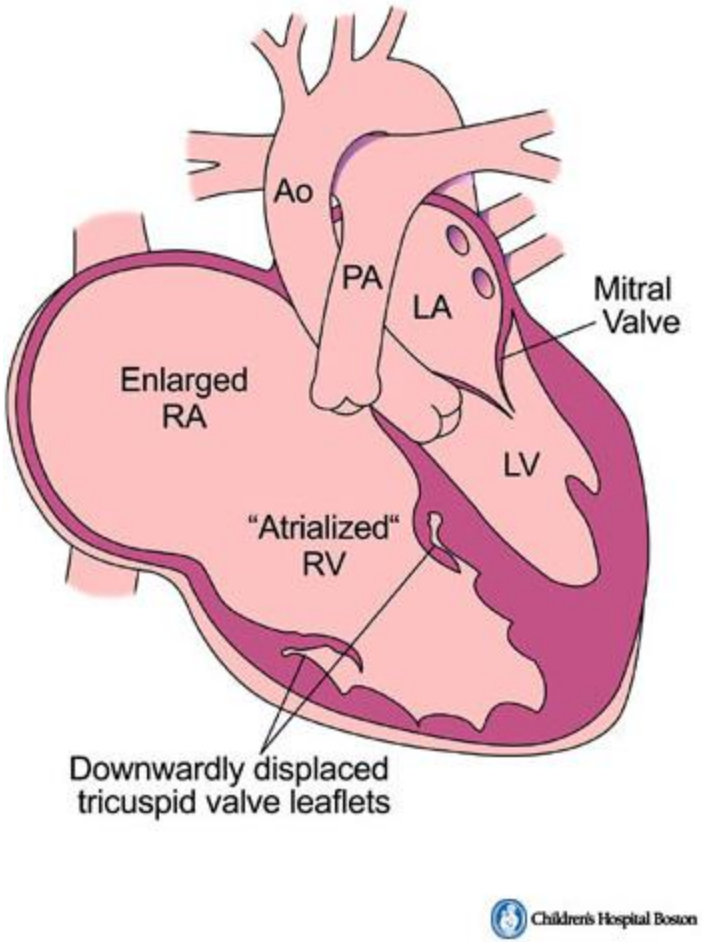
1. AP + CAP
2. AP + CAP + MAPCA
3. MAPCA +/- colaterale



# Boala Ebstein

- Insertie joasa a VT
- Aderenta de endocard
- Dilatarea portiuni atrializate
- Dilatarea inelului VT

- Asociere frecventa cu alte (DSA, SP etc.)



# Obstructive

1. Stenoza pulmonara
2. Stenoza aortica
3. Coarctatia de aorta

# Stenoza pulmonara

- Valvulara (inel, cuspe)
- Subvalvulara
  - Fixa (diafragm, VD bicameral)
  - Dinamica
- Supravalvulara

- Majoritatea sunt asimptomatice

# Stenoza pulmonara

- Presiune ↑ in VD
  - Hipertrofie
  - Ischemie +/- fibroza
  - ↑ presiunii in AD, dilatare
    - FOP
    - Aritmii
  - Disfunctie VS
    - ↓ presarcina
    - Interactiune mecanica

# Stenoza pulmonara

TRATAMENT

- Dilatare cu balon (interventional)
- Chirurgical
  - Valvotomie
  - Plastie cu petec tract de ejectie, valva, AP, ramuri

- TIC pre - postop

# Stenoza aortica

- 5% din MCC
- De 5x mai frecventa la baieti
- Asimptomatica -> critica
- Malformatie a cuspelor (mono, bi – tricuspa), sutura comisurilor
- Hipoplazia inelului
- Cu timpul apare si stenoza (fibroza) subvalv.

# Stenoza aortica

- Critica: duct dependenta; urgenta nn.
- ↑ postsarcinii
  - hipertrofie concentrica
  - Ischemie subendocardica, aritmii, ↓↓ inotropism
- Primul semn poate fi sincopa
- ↑ pres. ED -> dilatare (ireversibila)

# Stenoza aortica

MANAGEMENT

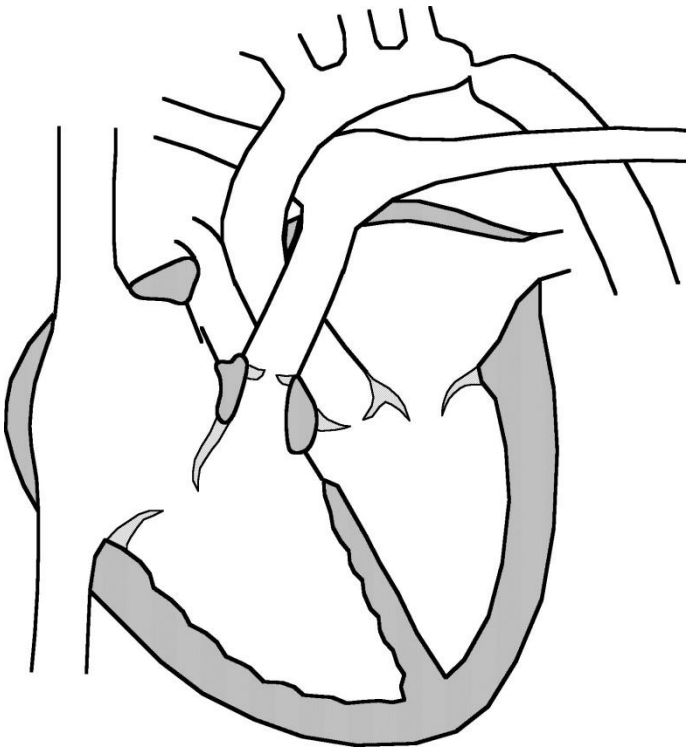
- Evaluarea VS
  - VM
  - VS hipoplazic ?
  - Miocard non-compactant, fibroelastoza
  - CoAo?
- PGE1 (necesita DSA!)

# Stenoza aortica

- Dilatare cu balon (interventional) - temporar
- Chirurgical
  - Valvotomie
  - Ross (autotransplant)
  - Ross-Kono
- Mortalitate ~ 10%

# CoAo

- Ingustarea istmului Ao
- Se poate asocia cu hipoplazia arcului, StAo, CAP
- 60% au Ao bicuspa
- 1/3 au CAP



# CoAo

- ↑ postsarcinii VS
- ↓ perfuziei in trenul inferior
- HTA in trenul superior (risc de AVC)
- ↑ pres in AS – disfunctie respiratorie

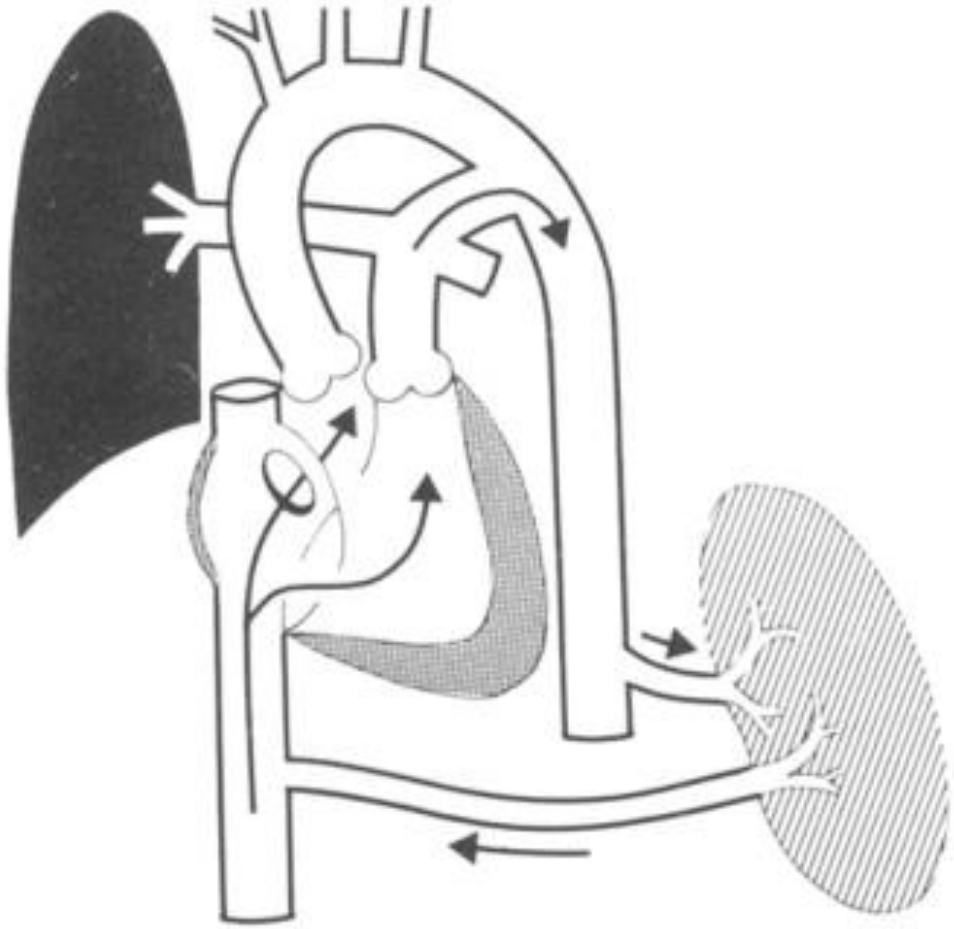
# CoAo

NN:

- PGE1
- DSA e benefic

- Interventional (?)
- Chirurgical (variate metode)

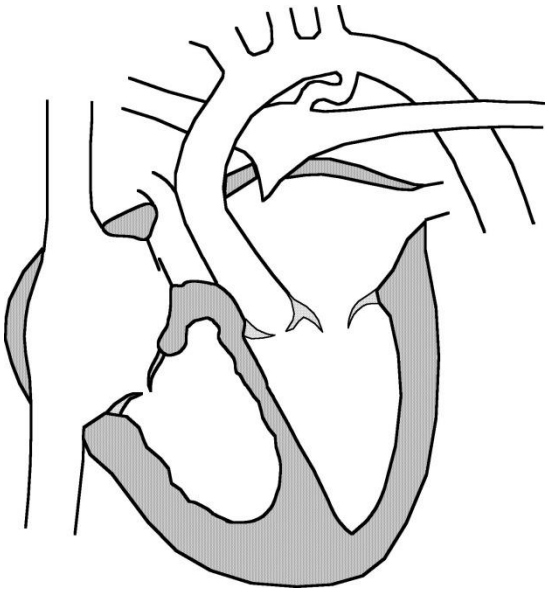
# Severe / incompatibile cu viata

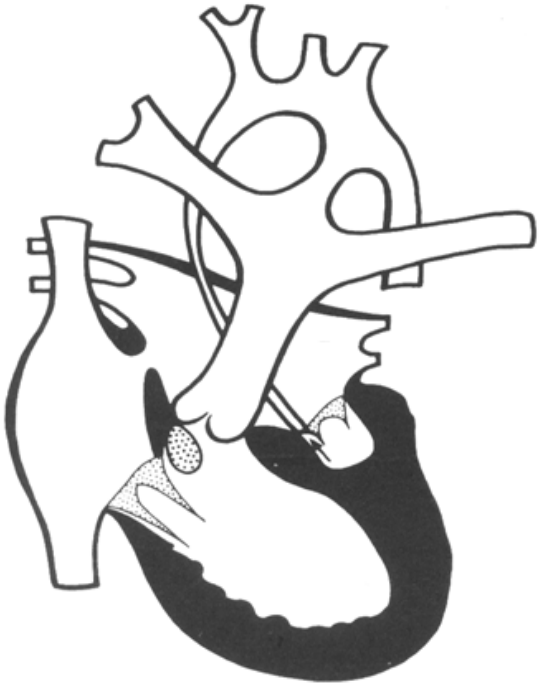


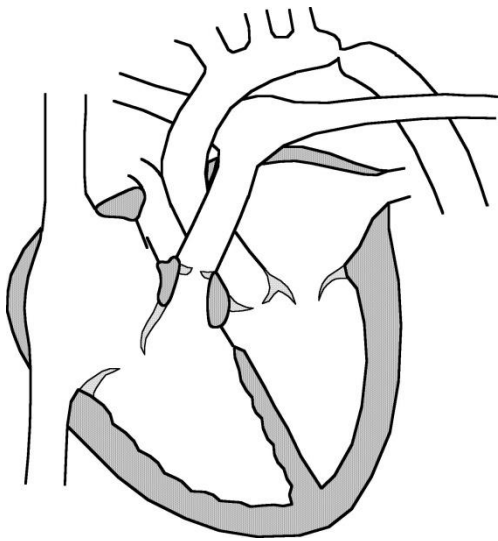
# Ductal dependente

- Atrezia pulmonara
- Atrezia aortica, mitrala, VS hipoplazic
- Arc Ao intrerupt

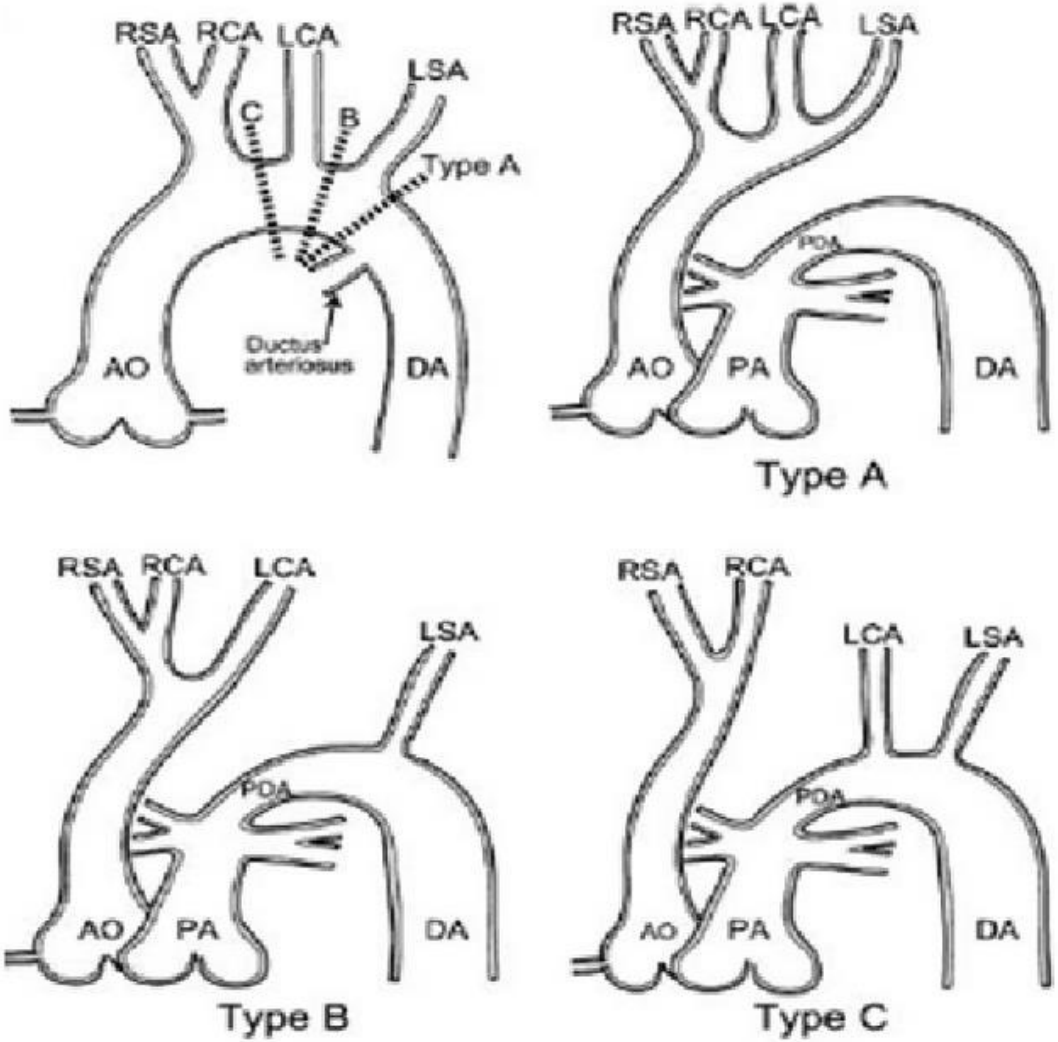
# Ductal dependente







# Ductal dependente



# Atrezia pulmonara – SiV intact

- Implica hipoplazia VD
- Afecteaza de regula si VTr
- AP e de regula bine dezvoltata

- DSA sau FOP (de regula nerestrictiv)
- Circulatia pulmonara e ductal dependenta

# Atrezia pulmonara – SiV intact

Management

- Fx de gradul de hipoplazie: 2, 1 sau 1 ½ ventricul
- NN: PGE1 / sunt SP +/- deschiderea VP
- Sugar: PCPC
- Prescolar/scolar: TCPC

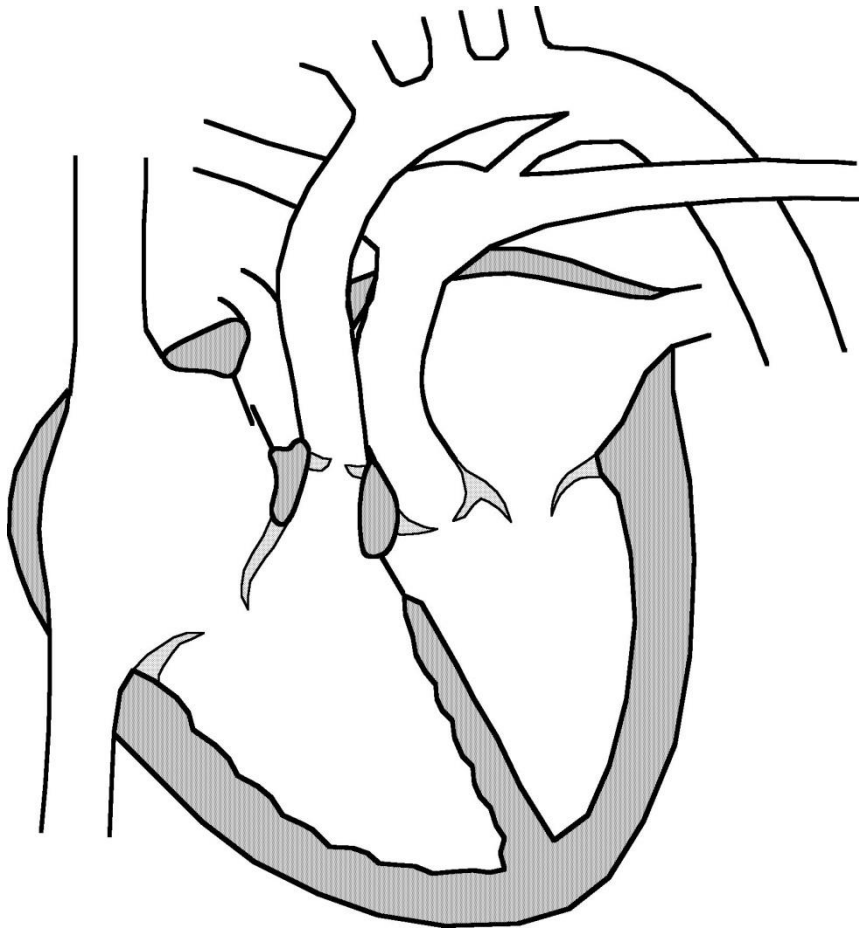
# Atrezia pulmonara – SiV intact

POSTOP

- Fluxul pulmonar !! -> SpO2 ~ 80%
  - RVP / RVS
  - Debit (volum, inotropic, TA)
  - anticoagulare
- Sustinerea VD pina la recuperare

# Circulatii paralele / TVM

- In loc de 2 circulatii in serie avem 2 in paralel
- Este obligatorie cel putin o comunicare (CAP, DSA si sau DSV)



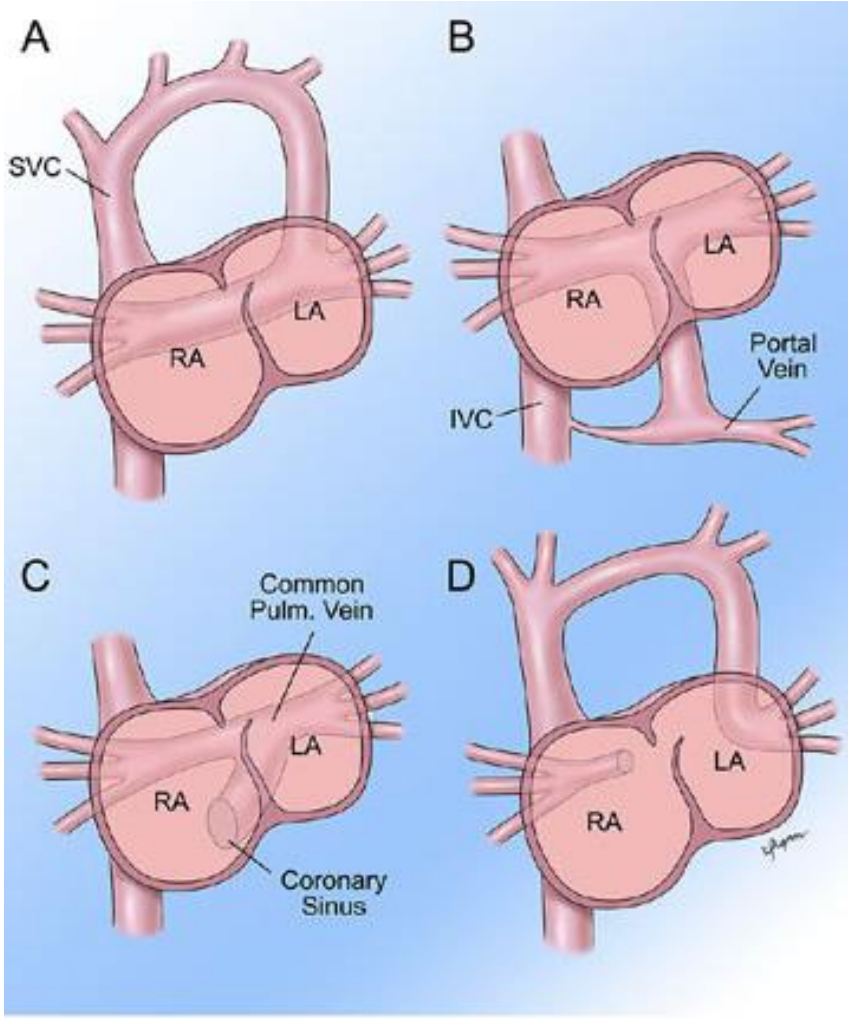
# Anomaliile de retur venos

- DVPA
- Obstructia returului venos
  - Stenoza venelor
  - Stenoza conductului
  - Cor triatriatum

# DVPA

- Total sau partial (1-3 vene)
- Prognosticul este dat de gradul de obstructie

- Supra, infra, cardiac, mixt



# DVPA

PREOP

- VM cu FiO2 1, hiperventilatie, NO
- PGE1 (in caz de obstructie a drenajului)
- Raskind
- ECMO (?)

# DVPA

- Tratament chirurgical

POSTOP

- HTP
  - Hiperventilatie, FiO2, NO
  - Sedare, analgezie
  - Inotropic inodilatator
  - Diuretic

# Cor triatriatum

- Cor triatriatum
- (Stenoza venelor pulmonare)

- Drenaj restrictiv al vv pulmonare
- HTP de tip venos
- Presarcina ↓ VS -> DC scazut

# Cor triatriatum

- Urgenta (cind e simptomatic)
- Rezolvare eminament chirurgicala

- POSTOP
- HTP restanta
- artimii

# Silentioase

Se manifesta tirziu, in viata adulta

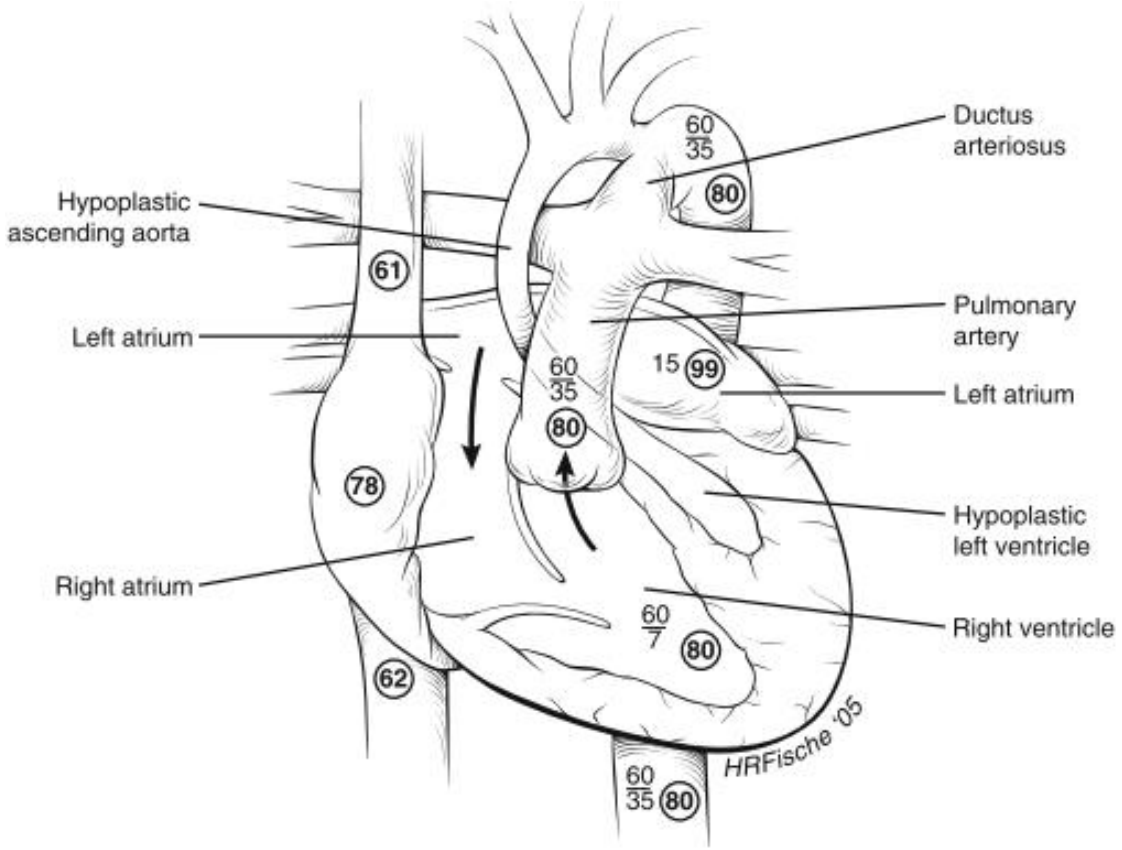
Sunt relativ benigne

- VAo bicuspa
- Anomalii coronare
- Sdr. WPW
- TVM corectata anatomic

# VS hipoplazic

- 5% din MCC

- VS
- VM
- Ao



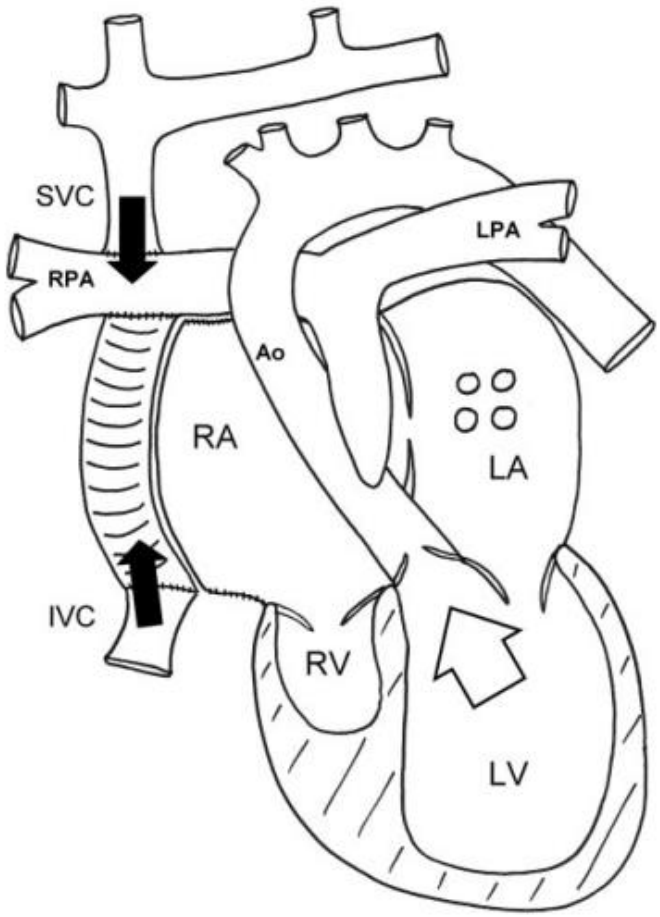
# VD hipoplazic

- Atrezia tricuspida
- Atrezia pulmonara cu SIV intact
- Double inlet ventricle
- CAVC cu ventriculi nebalansati

# Ventricul unic

- Circulatie de tip Fontan

- Returul venos -> AP
- Fara pompa in circulatia pulmonara
- Trebuie stadializata



# Faza 1 - nn

- Paliatie specifica afectiunii (Mixing total)

- Retur venos pulmonar nerestrictiv in ventricul
- Tract de ejectie nerestrictiv sistemic
- Flux limitat dar de incredere in circ. pulm. (sunt / banding)
- Qp/Qs 1:1 – 2:1
- QP/QS = (SaO2 - SvO2) / (PVO2 - SaO2)

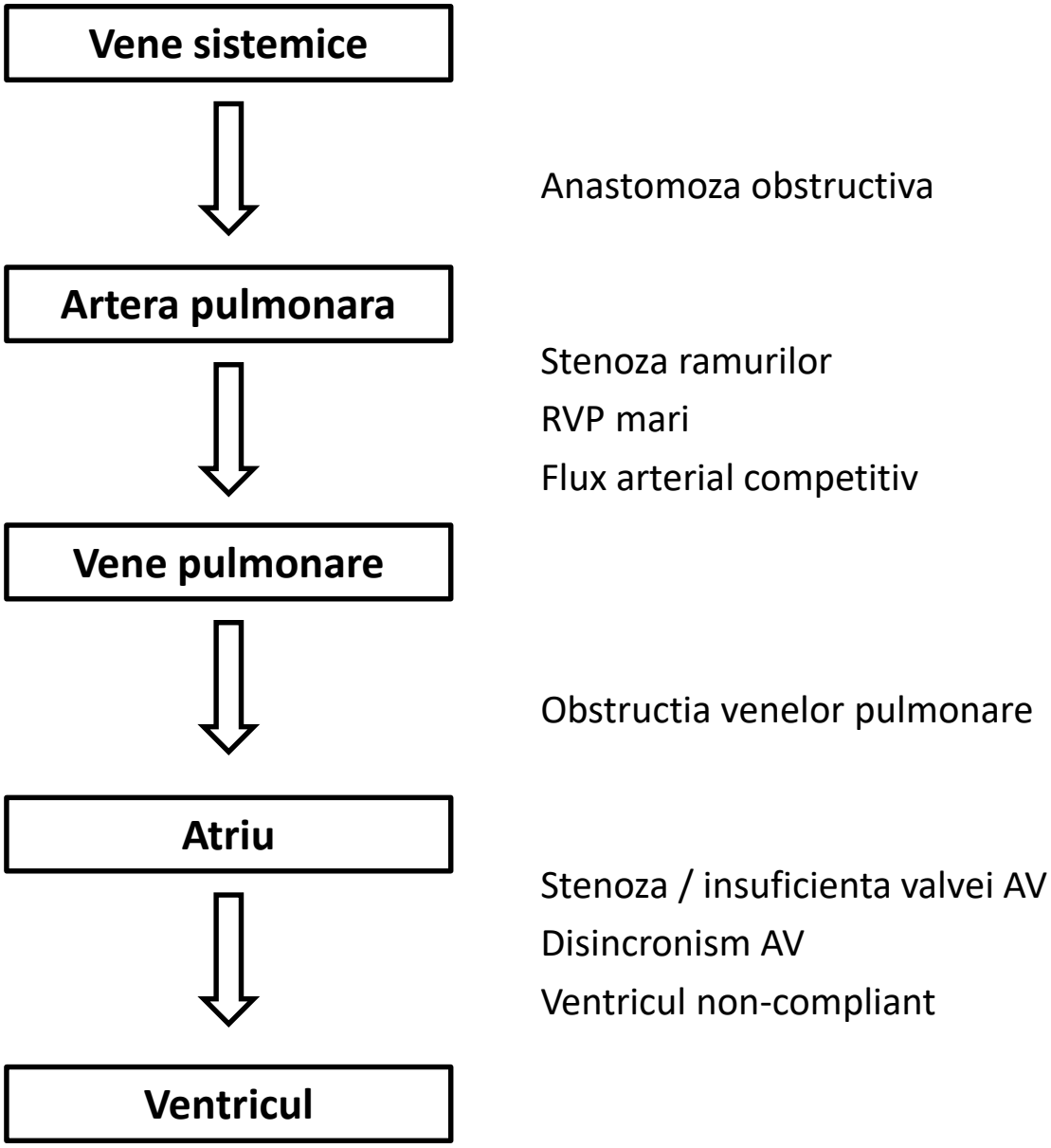
# Faza 2 – 4-6 luni

- Glenn = hemi Fontan = PCPC
- Sunt bidirectional VCS – AP
- Reduce mixingul
- Reduce incarcarea ventriculara

# Faza 3 – 2-4 ani

- Fontan
- VCS -> AP
- Circulatii separate complet, saturatii normale

- Un singur ventricul, 2 circulatii in serie
- Volum bataie normal, energie mai multa (pentru a trece prin ambele circulatii)
- Orice obstructie poate compromite


Vene sistemice
Anastomoza obstructiva
Artera pulmonara
Stenoza ramurilor
RVP mari
Flux arterial competitiv
Vene pulmonare
Obstructia venelor pulmonare
Atriu
Stenoza / insuficienta valvei AV
Disincronism AV
Ventricul non-compliant
Ventricul

# Abordare sistematica

- Evaluare completa a leziunilor
- Stabilirea fluxurilor sanguine
- Stabilirea impactului
- Elaborarea unui plan
- Imaginarea unor scenarii
- Consultati un specialist