

# AZYANOTISCHE / ZYANOTISCHE HERZERKRANKUNG

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Klinik Für Herzchirurgie



# EPIDEMIOLOGIE

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- Angeborene Herzfehler sind die häufigsten angeborenen Fehler
- 1% (40.000 / Jahr in der USA)
  - VSD
  - PDA
  - ASD
  - PS
  - AS
  - ISTA
  - Fallot – Tetralogie
  - TGA
  - HLH

In den meisten Fällen sind die Ursachen unbekannt

# EINTEILUNG

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## • **Zyanotische Fehler**

- Fallot –Tetralogie
- TGA
- UniVentrikel
- TK Atresie
- PK Atresie
- Ebstein Anomalie

Mit Shunt (R->L)

## • **Azyanotische Fehler**

- ASD
- VSD
- PDA
- Truncus arteriosus

Mit Shunt (L->R)

- AK Stenose
- ISTHA
- Gefäßbringe
- PK Stenose

Ohne Shunt

# KLINISCHE MERKMALE

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- Zyanose
- Herzinsuffizienz
- Pulmonale Hypertonie
- Wachstumsstörung

# ZYANOSE

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- Periphere Zyanose

- Normal gesättigte SaO<sub>2</sub>,  
abnormale periphere O<sub>2</sub>  
Extraktion

- Zentrale Zyanose

- Niedrige SaO<sub>2</sub>
- Rechts -> Links Shunt
- TGA

- Grad der Zyanose

- Hb
- Rechts -> Links Shunt Volumen
- SvO<sub>2</sub>

# AZYANOTISCHE HERZERKRANKUNGEN

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- **Links -> Rechts Shunt**
  - Vorhofseptumdefekt (ASD)
  - Ventrikelseptumdefekt (VSD)
  - Persistierender Duktus arteriosus (PDA)
  - Truncus arteriosus
- **Ohne Shunt**
  - AK Stenose
  - Aortenisthmusstenose (ISTHA)
  - Gefäßringe
  - Pulmonalstenose

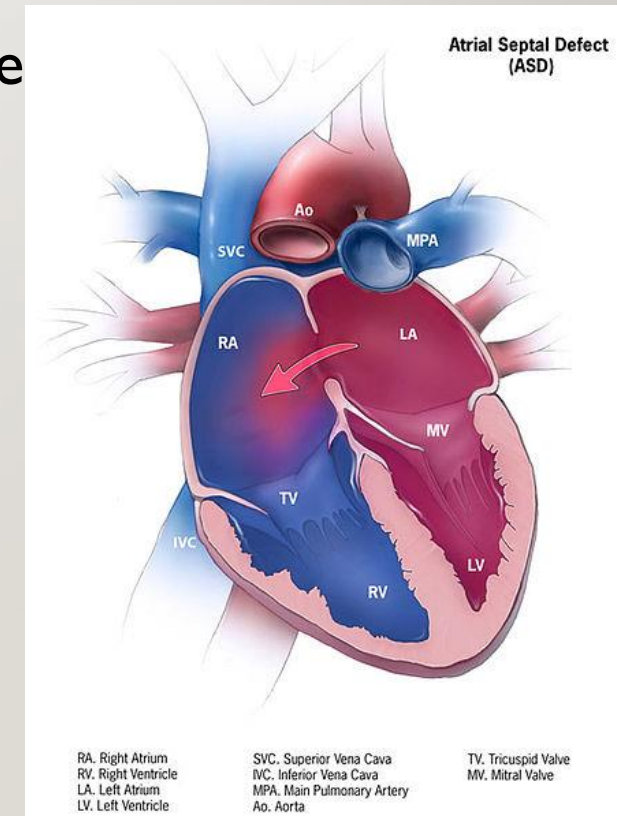
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- Links Rechts Shunt
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  - Gefäßringe
  - Pulmonalstenose

# VORHOFSEPTUMDEFEKT (ASD)

- Asymptomatisch, ggf. eingeschränkte Leistungsfähigkeit
- Systolikum über 2. ICR li
- Dyspnoe, Zeichen von Rechtsherzinsuffizienz

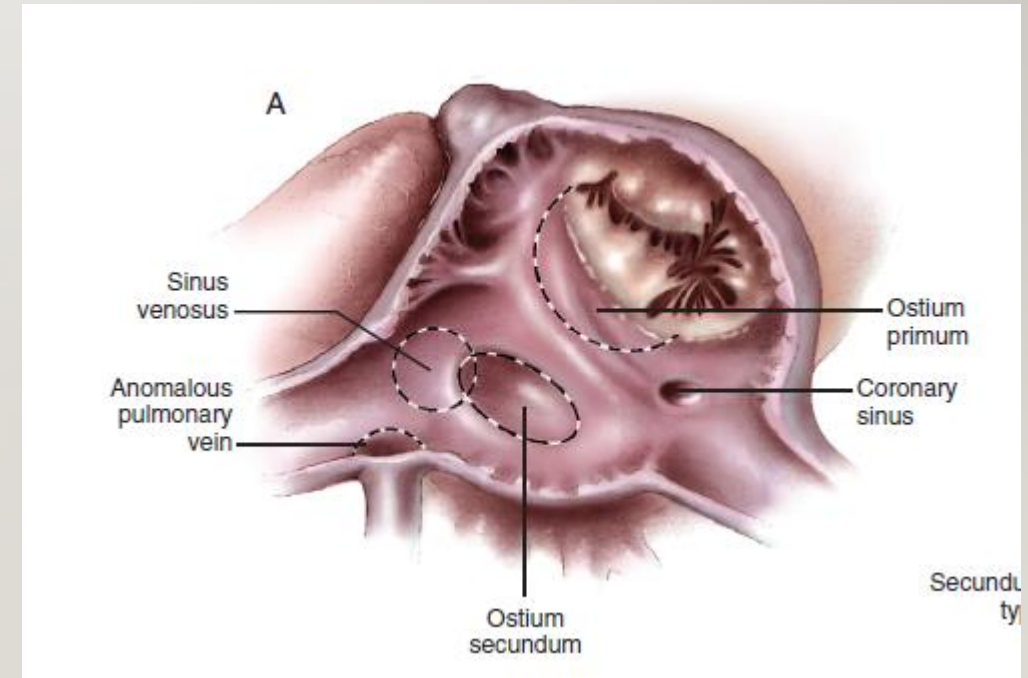




# VORHOFSEPTUMDEFEKT (ASD)

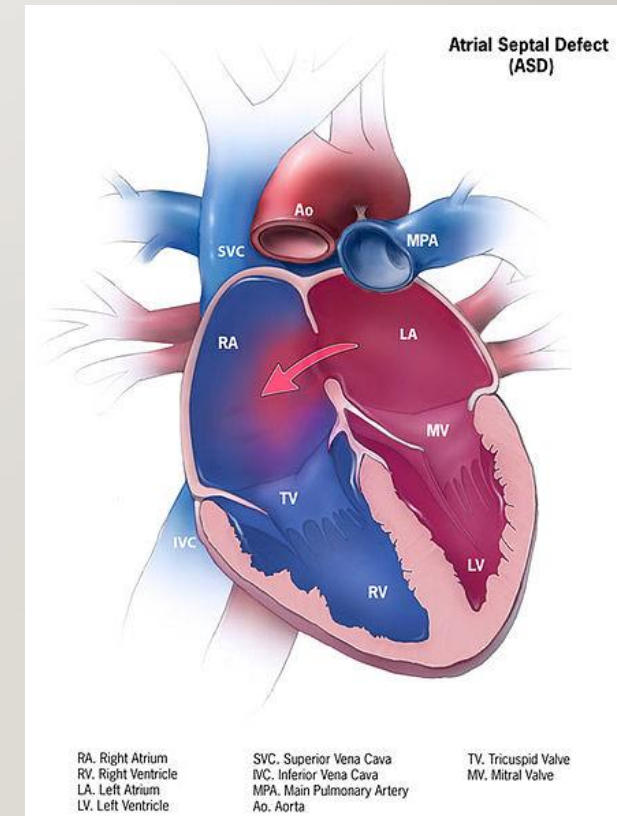
- Asymptomatisch, ggf eingeschränkte Leistungsfähigkeit
- Systolikum über 2. ICR li
- Dyspnoe, Zeichen Rechtsherzinsuffizienz

- Septum –Secundum-Defekt (>80%)
- Sinus-venosus-Defekt (5-10%)
- Koronarsinusseptumdefekt (5%)
- Septum-primum-Defekt (1%)



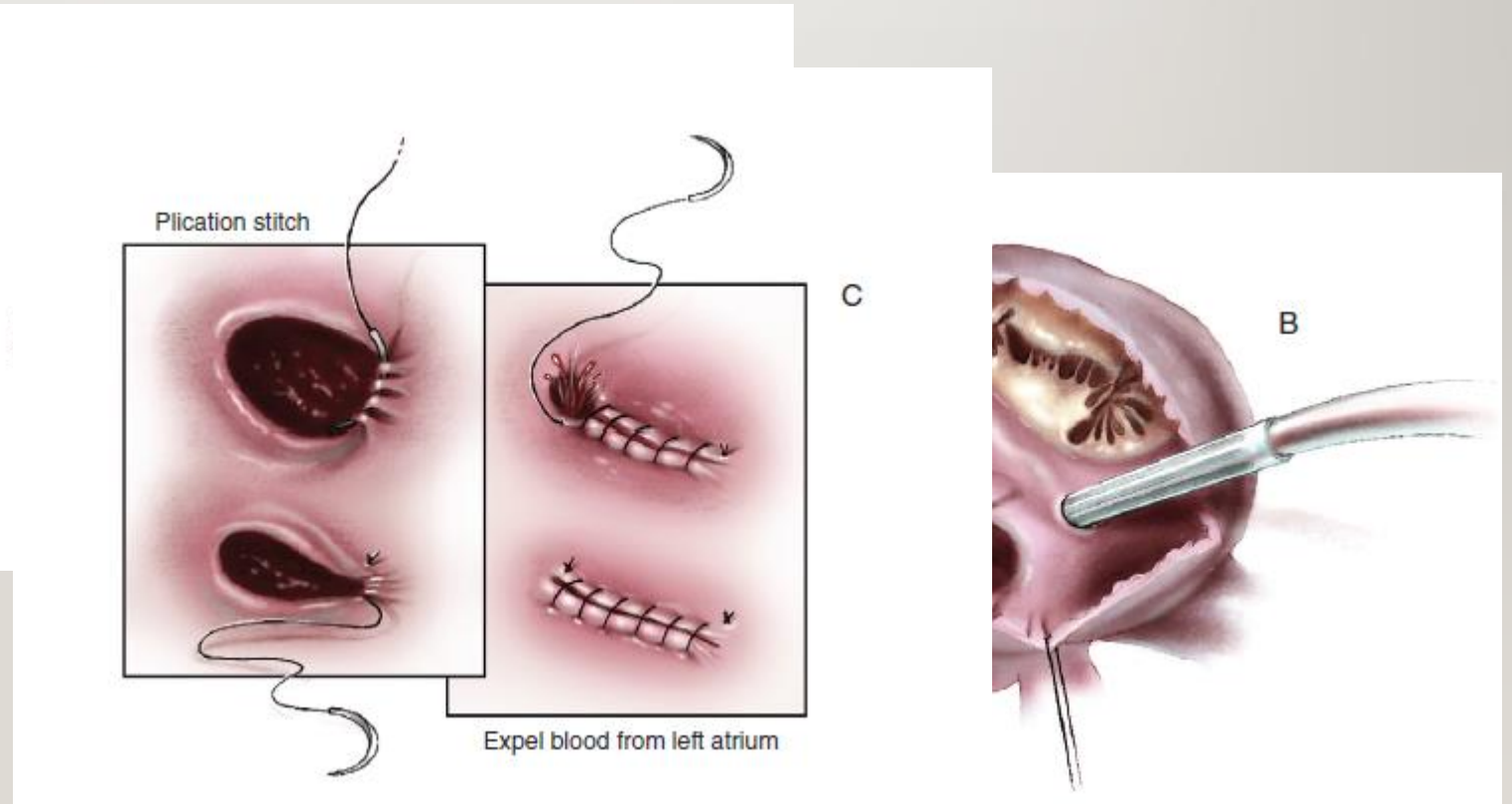
# VORHOFSEPTUMDEFEKT (ASD)

- Spontanverschlussrate : 14% - 25%
- Indikationen:
  - Herzinsuffizienz
  - Bei Rechtsherzbelastung
  - Paradoxe Embolie
  - $Q_p/Q_s > 1,3 - 1,5$
  - PA Widerstand  $> 10 \text{ WU/m}^2$

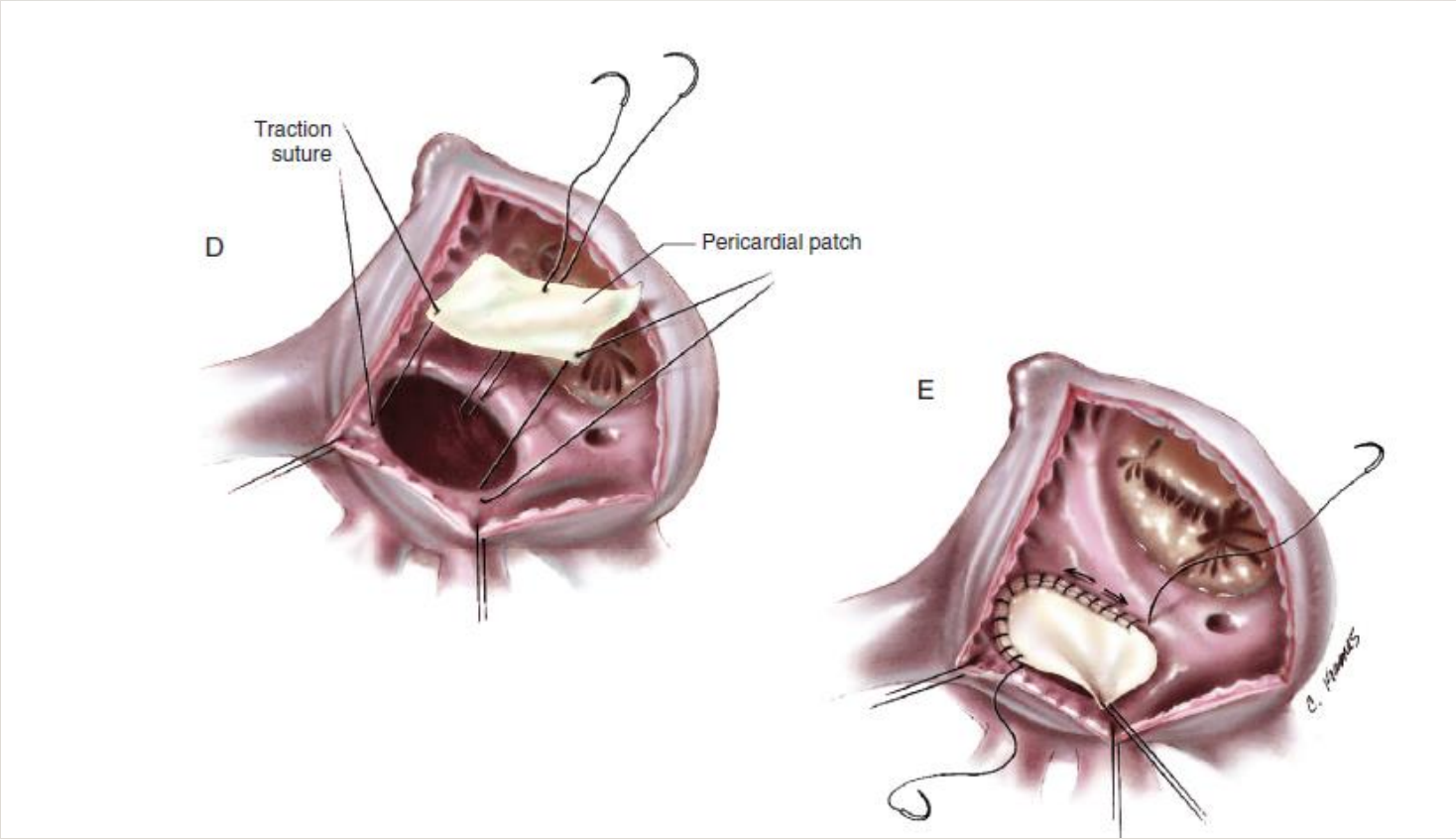


# VORHOFSEPTUMDEFEKT (ASD)

- Direkt Verschluss
- Patch Verschluss
- Ergebnisse:
  - Letalität <1%
  - Vorhof und Knotenarrhythmien
  - Normale Lebenserwartung



# VORHOFSEPTUMDEFEKT (ASD)



# AZYANOTISCHE HERZERKRANKUNGEN

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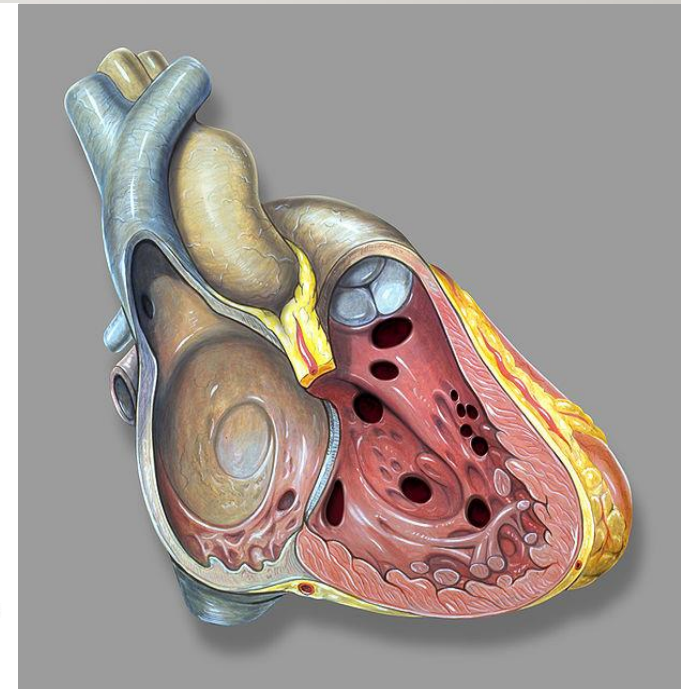
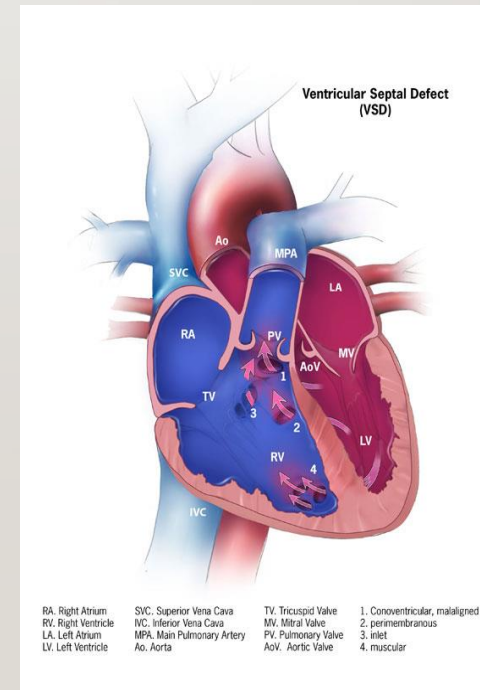
- Links Rechts Shunt
  - Vorhofseptumdefekt (ASD)
  - **Ventrikelseptumdefekt (VSD)**
  - Persistierender Duktus arteriosus (PDA)
  - Truncus arteriosus
- Ohne Shunt
  - AK Stenose
  - Aortenisthmusstenose (ISTHA)
  - Gefäßringe
  - Pulmonalstenose

# VENTRIKELSEPTUMDEFEKTE (VSD)

- Häufigsten angeborenen Herzfehler (20-25%)

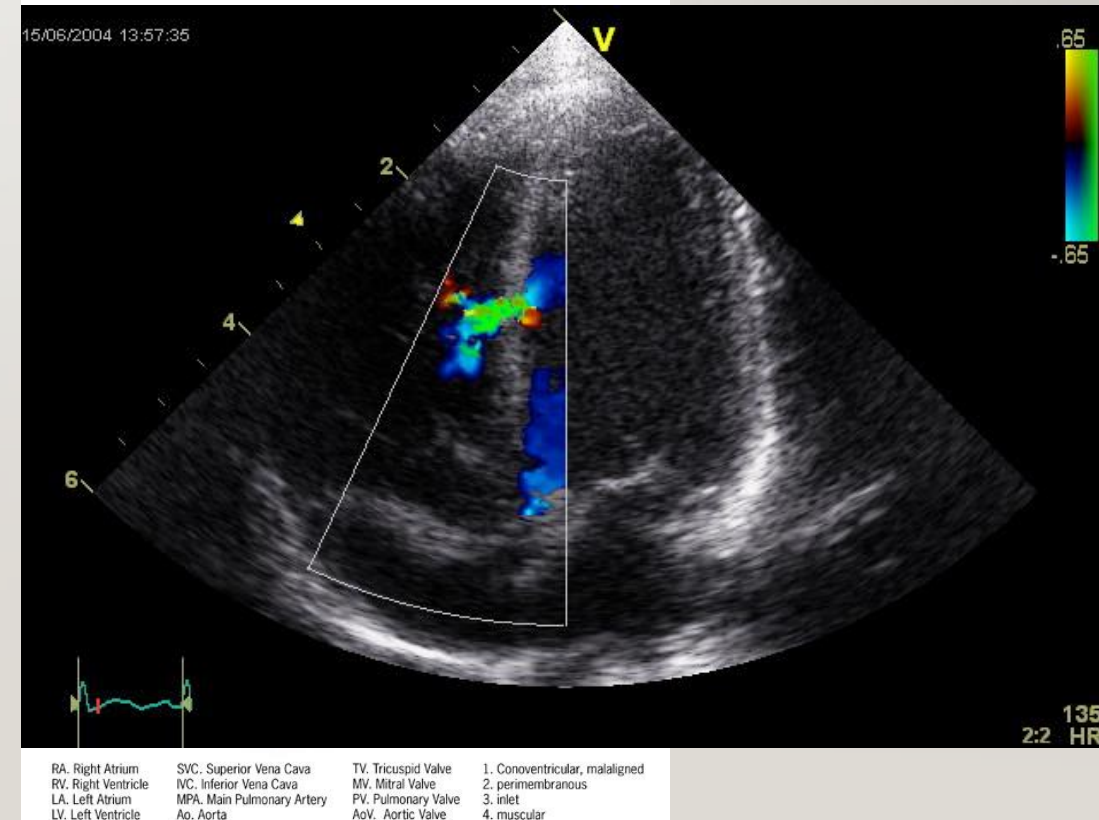
- Einteilung

- Konoventrikuläre (80%)
- AV-Kanaltyp (6%)
- Muskuläre VSD (10%)

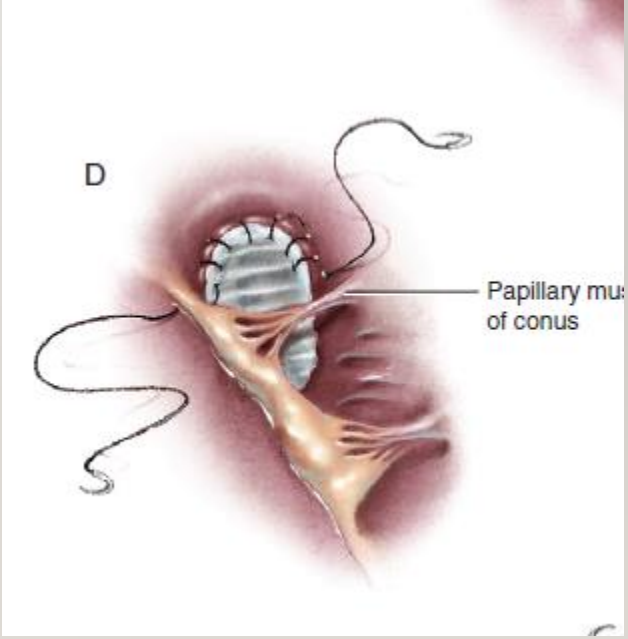
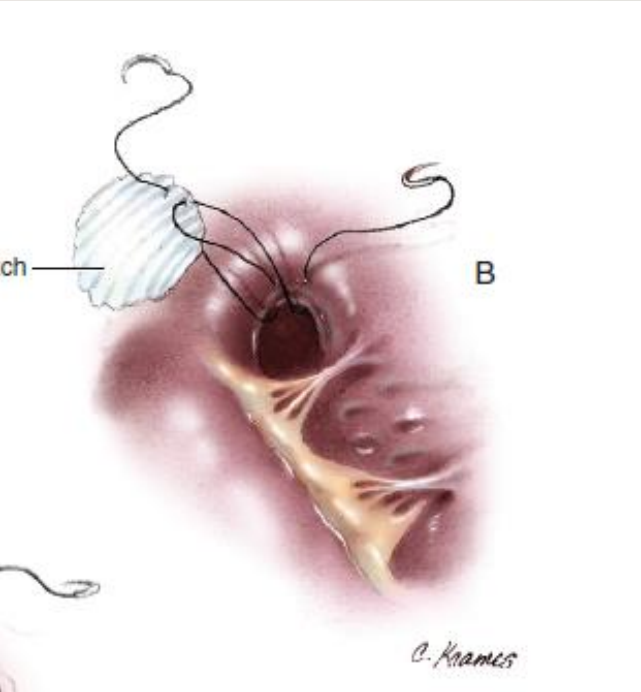
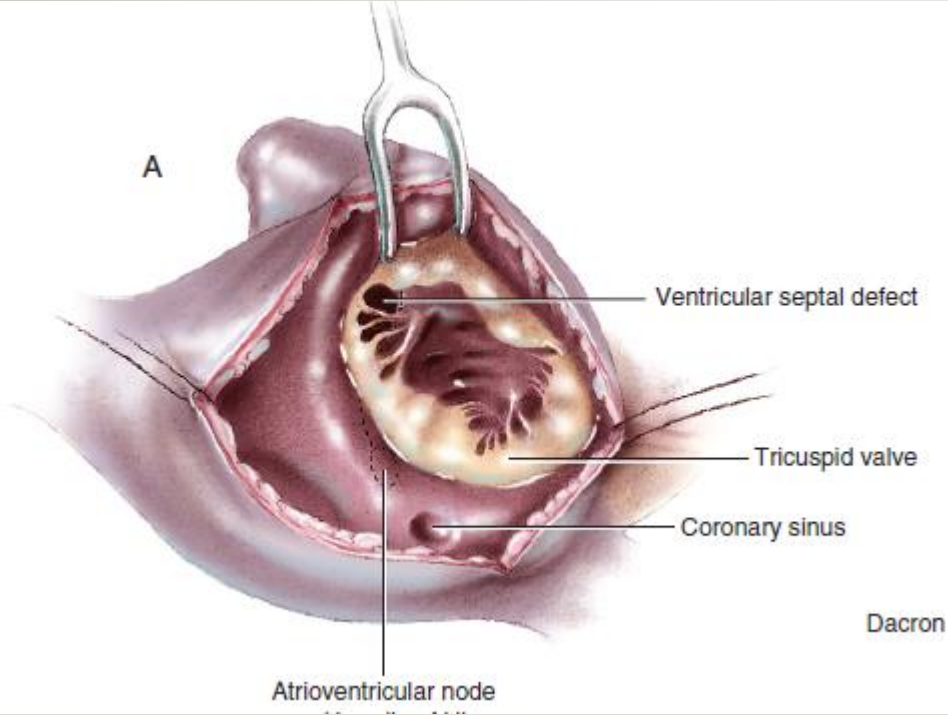


# VENTRIKELSEPTUMDEFEKTE (VSD)

- Indikation zur OP:
  - Herzinsuffizienz (OP: erste Lebenswoche)
  - $Q_p/Q_s = 1,5 - 2$  (OP: erste 3 Monate)
  - VSD > 50% der AK Fläche
  - PA Druck > 50% der systemische Druck
- Spontanverschluß: <3 Jahren
- Kontraindikation:
  - PA Widerstand > 0,7-0,5 Systemische Widerstand

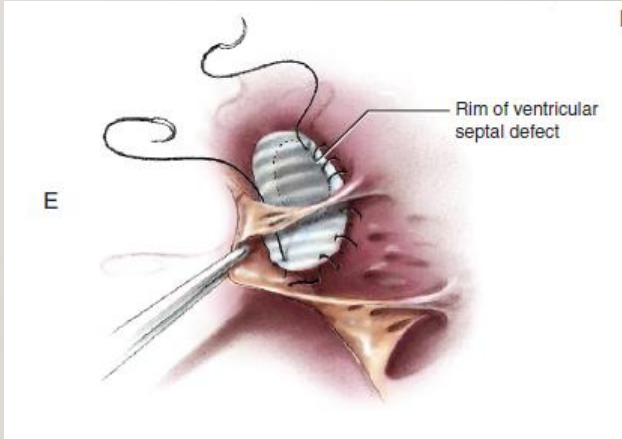
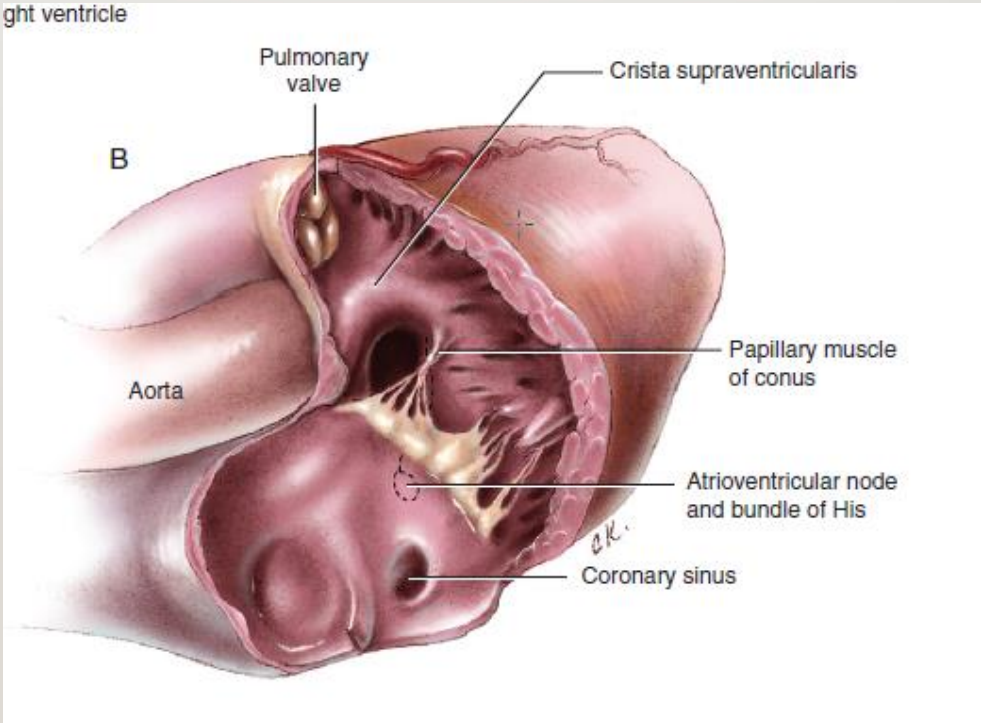
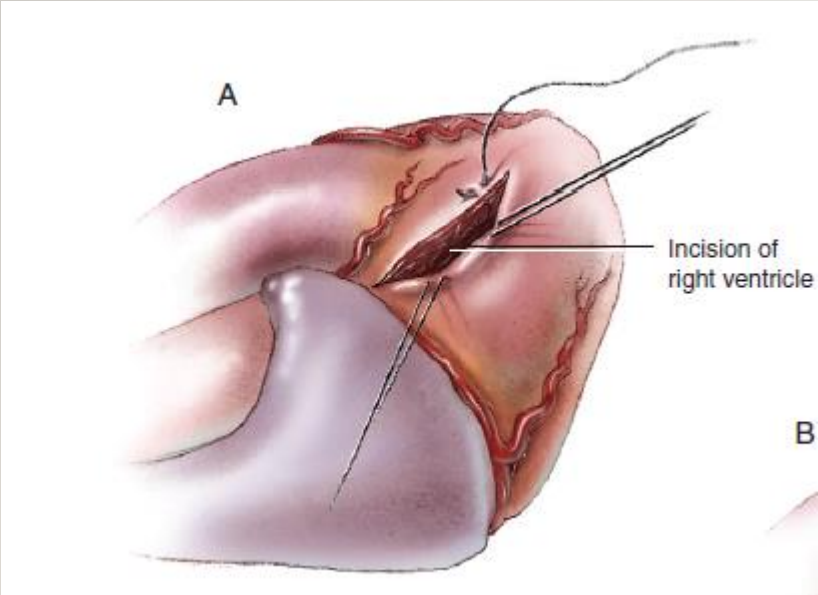


# VENTRIKELSEPTUMDEFEKTE (VSD)





# VENTRIKELSEPTUMDEFEKTE (VSD)



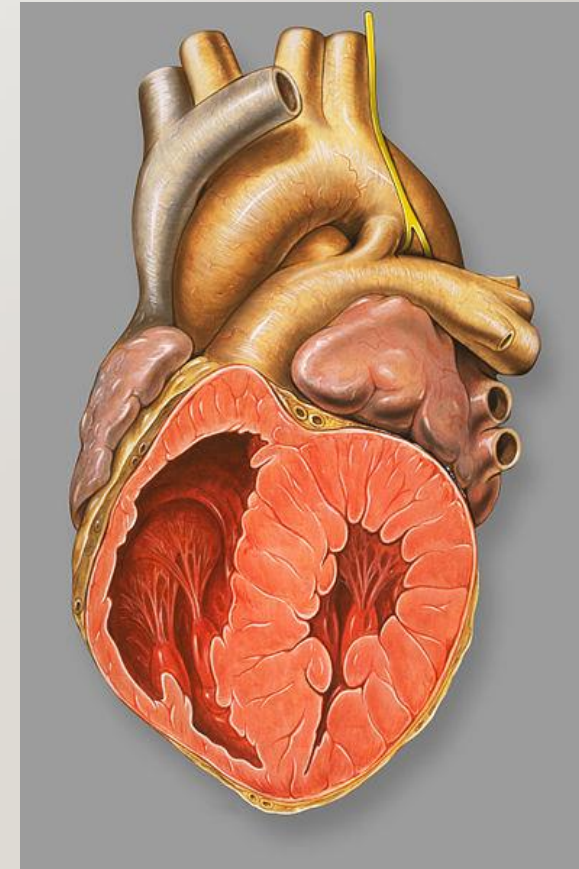
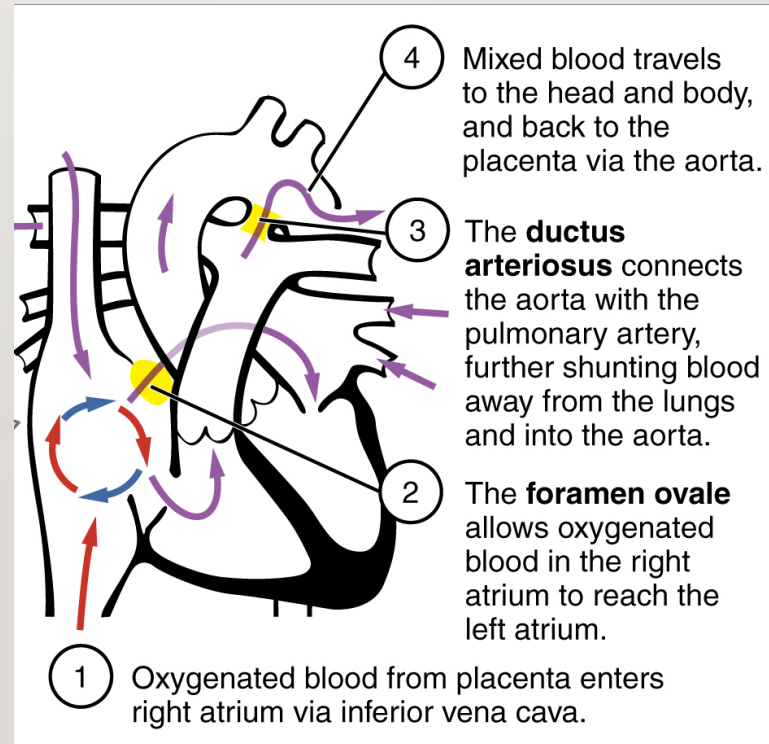
# AZYANOTISCHE HERZERKRANKUNGEN

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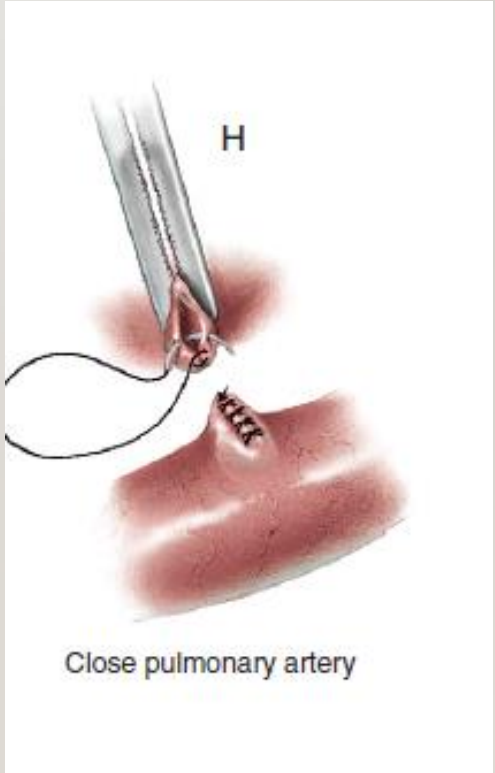
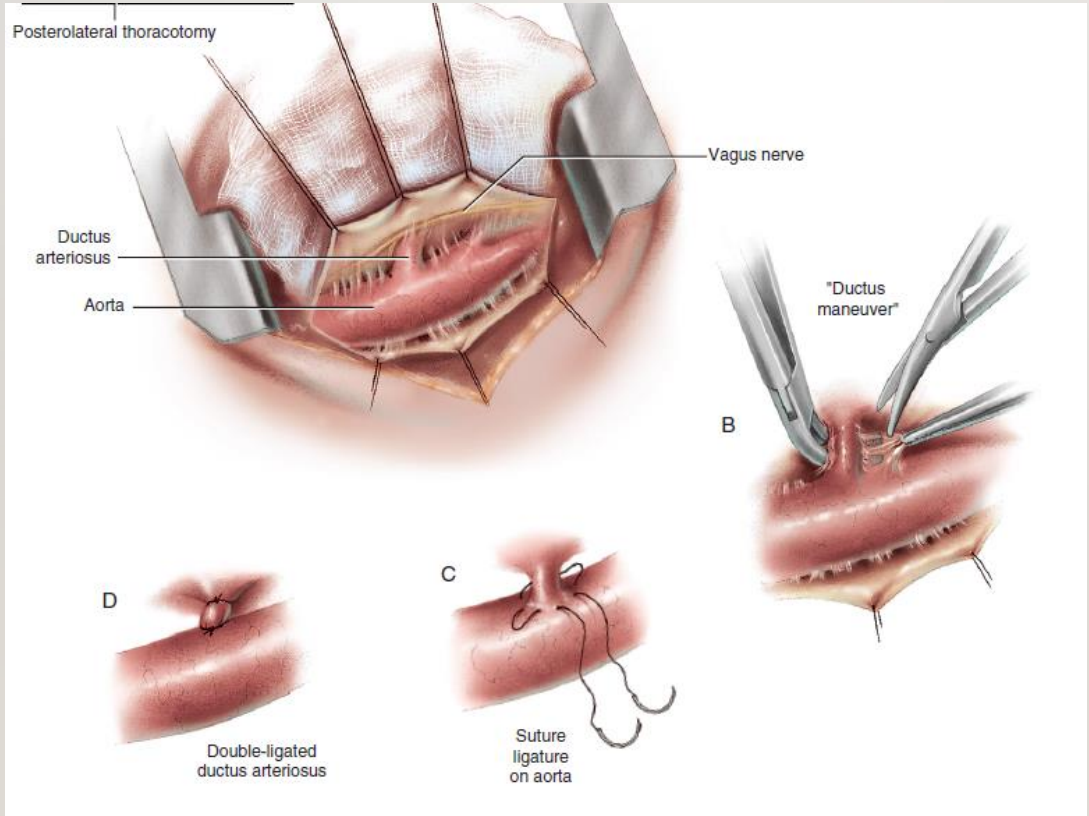
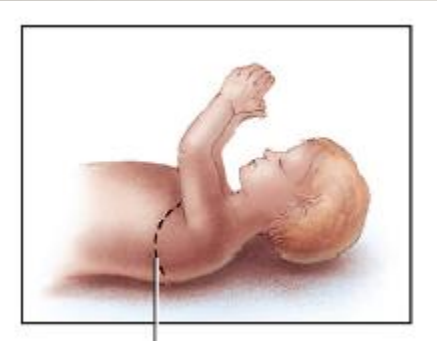
- Links Rechts Shunt
  - Vorhofseptumdefekt (ASD)
  - Ventrikelseptumdefekt (VSD)
  - **Persistierender Duktus arteriosus (PDA)**
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- Ohne Shunt
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  - Gefäßringe
  - Pulmonalstenose

# PERSISTIERENDER DUKTUS ARTERIOSUS (PDA)

- 5-10% aller angeborenen Herzfehler
- Defektgröße
  - **Klein:** Asymptomatisch
  - **Mittelgroß:** L->R Shunt
  - **Groß:** Herzinsuffizienz, pulmonale Hypertonie

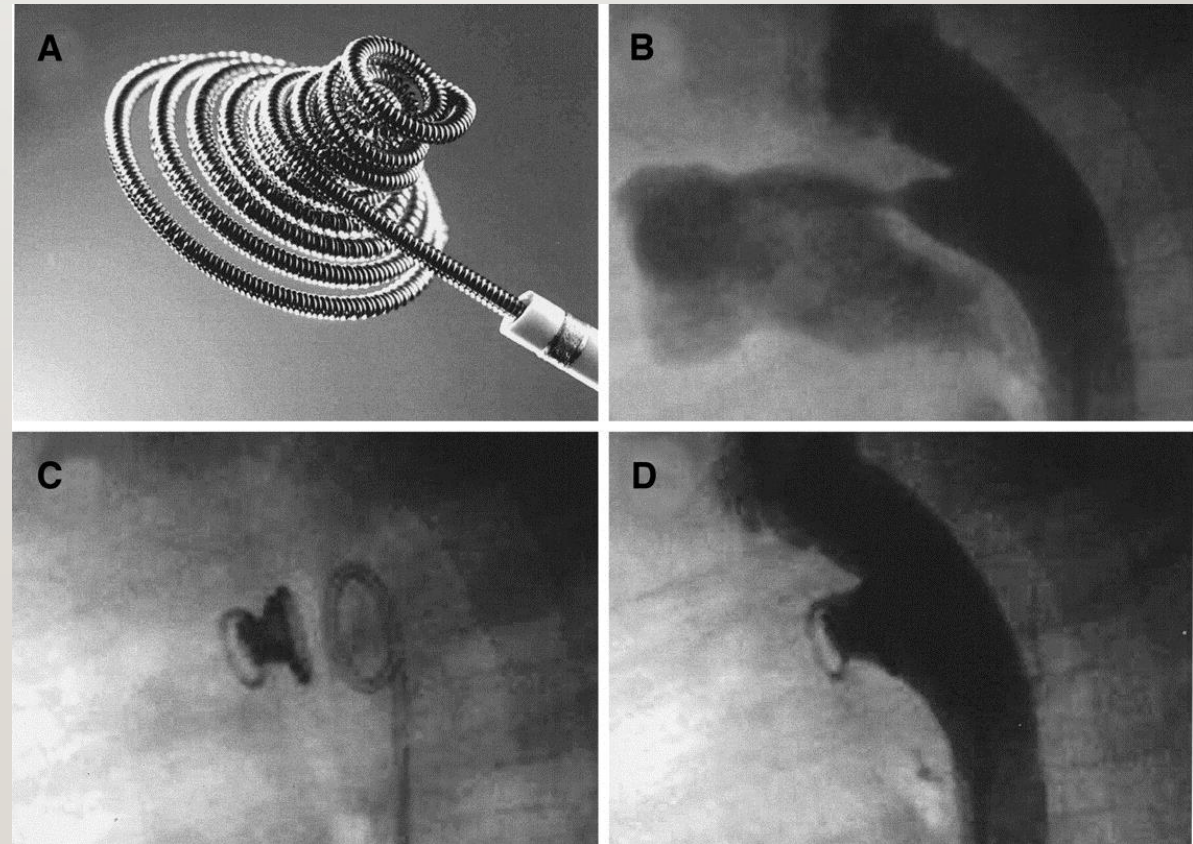
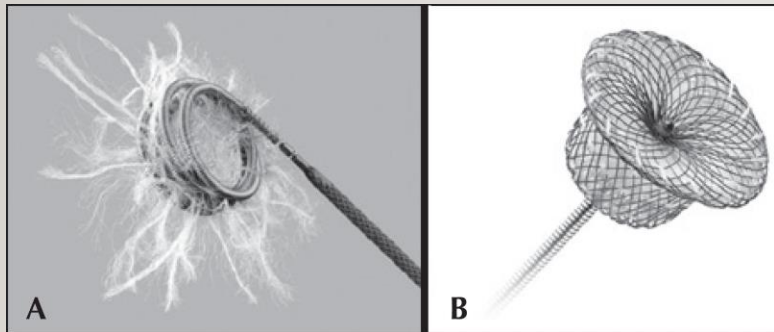


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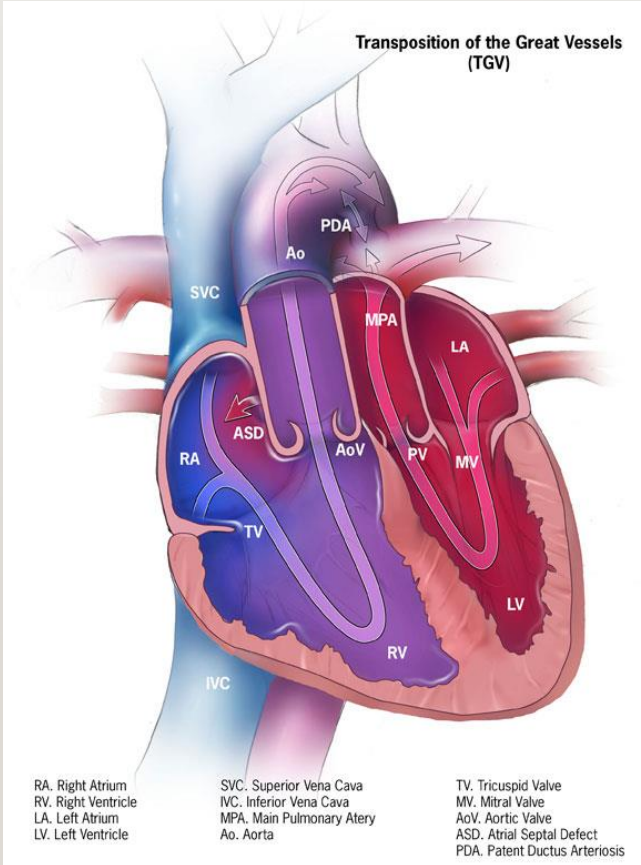
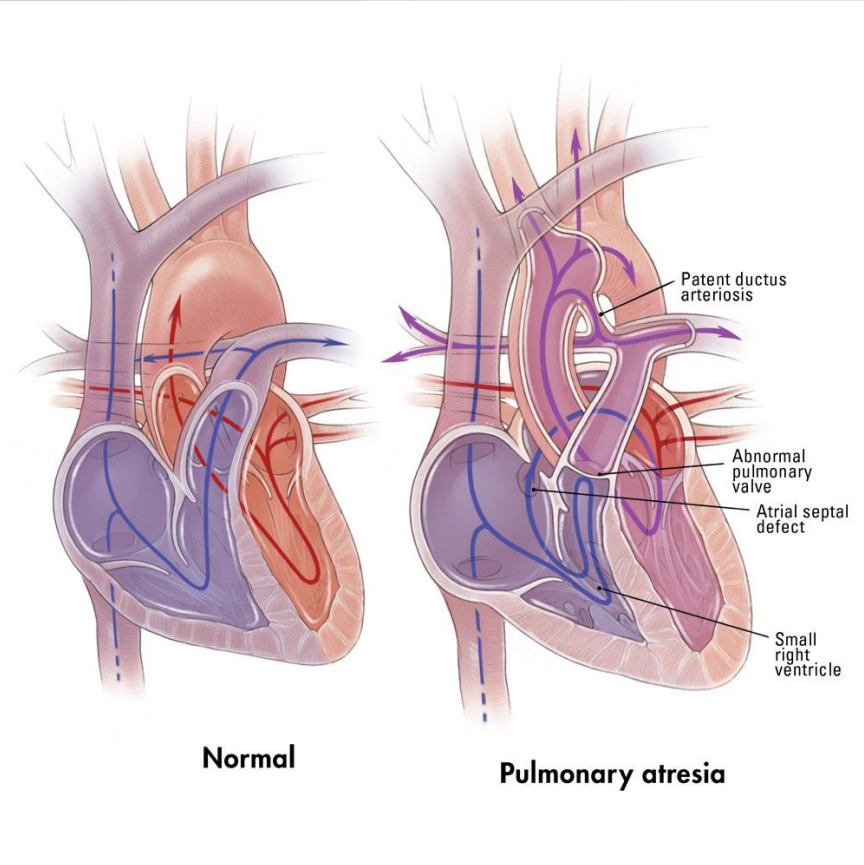


# PERSISTIERENDER DUKTUS ARTERIOSUS (PDA)

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# PERSISTIERENDER DUKTUS ARTERIOSUS (PDA)



**PGE Infusion**

# ZYANOTISCHE HERZERKRANKUNGEN

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- **Fallot –Tetralogie**
- **Transposition der großen Arterien**
- **Hypoplastisches Linksherz-Syndrom**
- TK Atresie
- PK Atresie
- Ebstein Anomalie

# ZYANOSE

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- Periphere Zyanose

- Normal gesättigte SaO<sub>2</sub>, abnormale periphere O<sub>2</sub> Extraktion

- Zentrale Zyanose

- Niedrige SaO<sub>2</sub>
- Rechts -> Links Shunt
- TGA

- Grad der Zyanose

- Hb
- Rechts -> Links Shunt Volumen
- SvO<sub>2</sub>



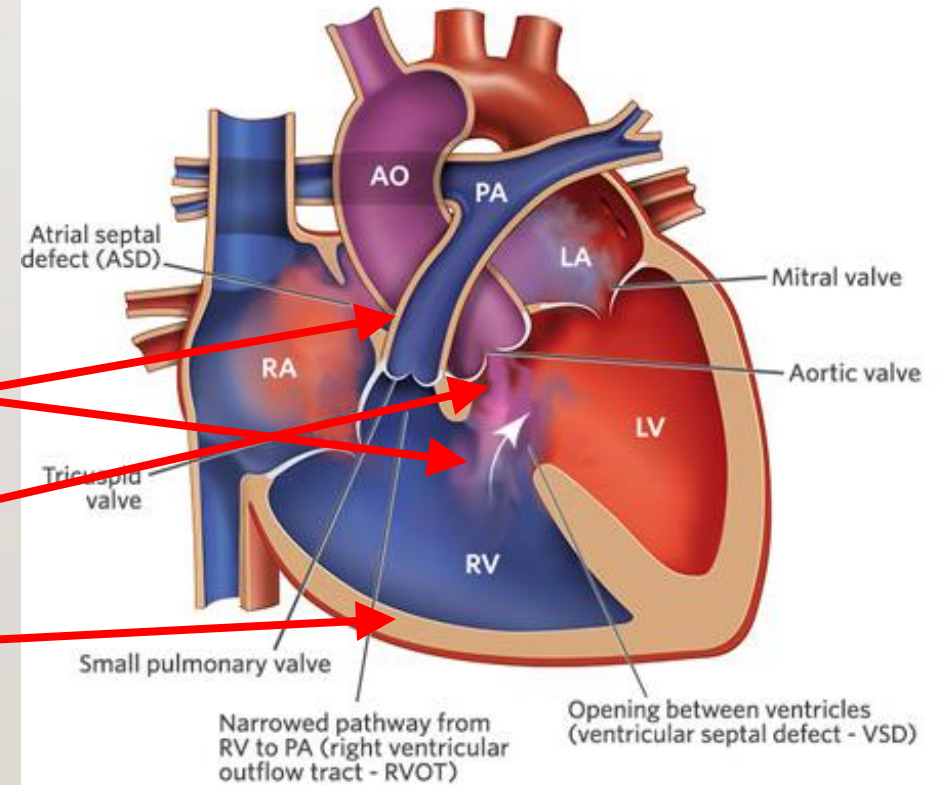
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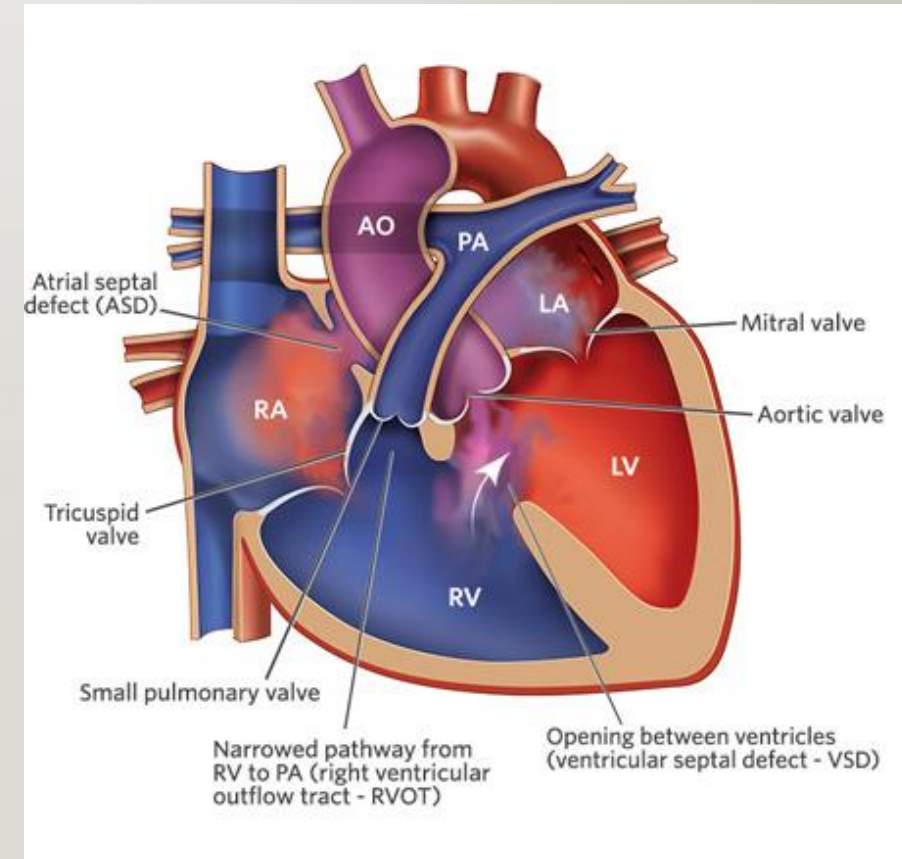
# FALLOT - TETRALOGIE

- 10 % aller angeborenen Herzfehler
- Tetralogie:
  - Malalignment VSD
  - PA Stenose
  - Dextraposition der Aorta
  - RV Hypertropie



# FALLOT - TETRALOGIE

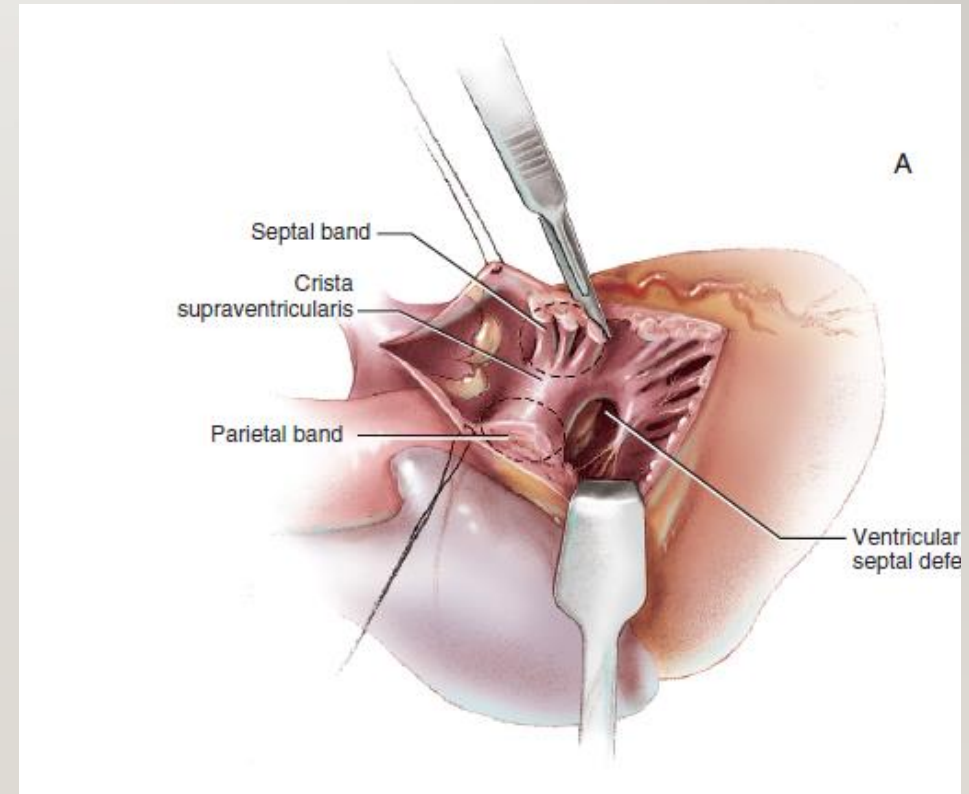
- Der VSD führt zu eine gemeinsamen Pumpkammer
- Pulmonaler Blutfluss so lange adäquat wie der Ductus arteriosus offen ist
- Nach Verschluss des Ductus fällt die arterielle Sättigung abhängig von der PA Obstruktion.



# FALLOT - TETRALOGIE

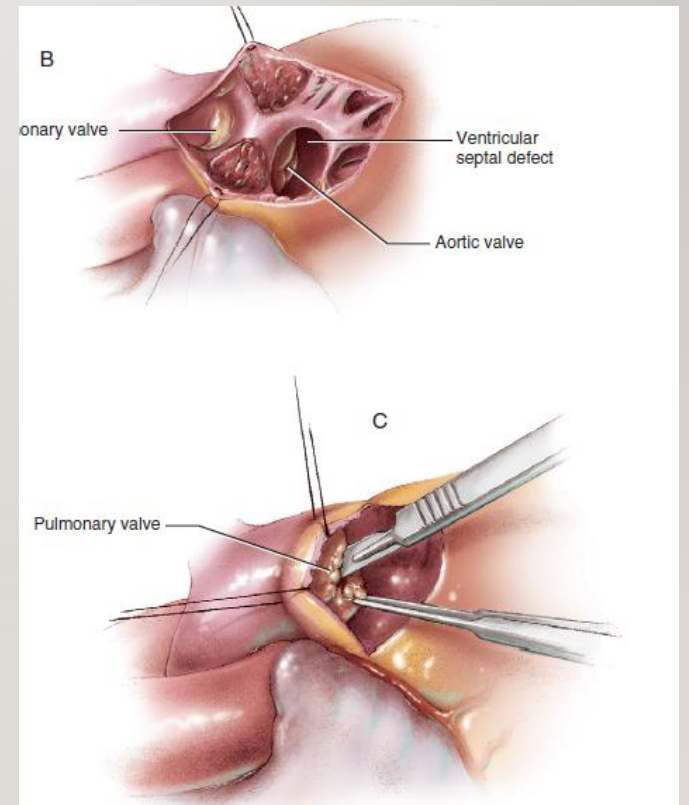
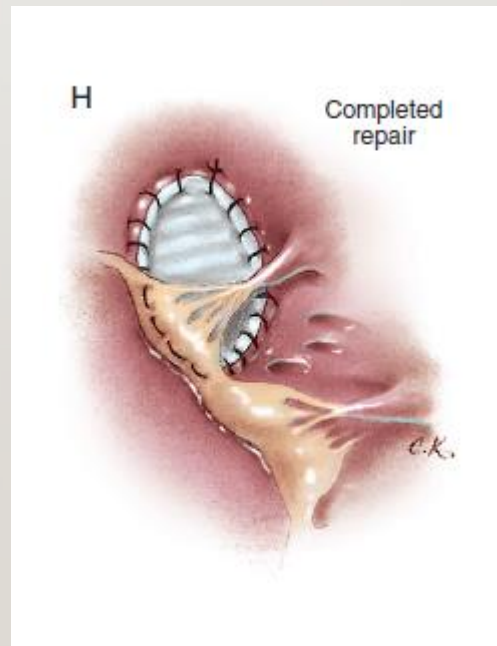
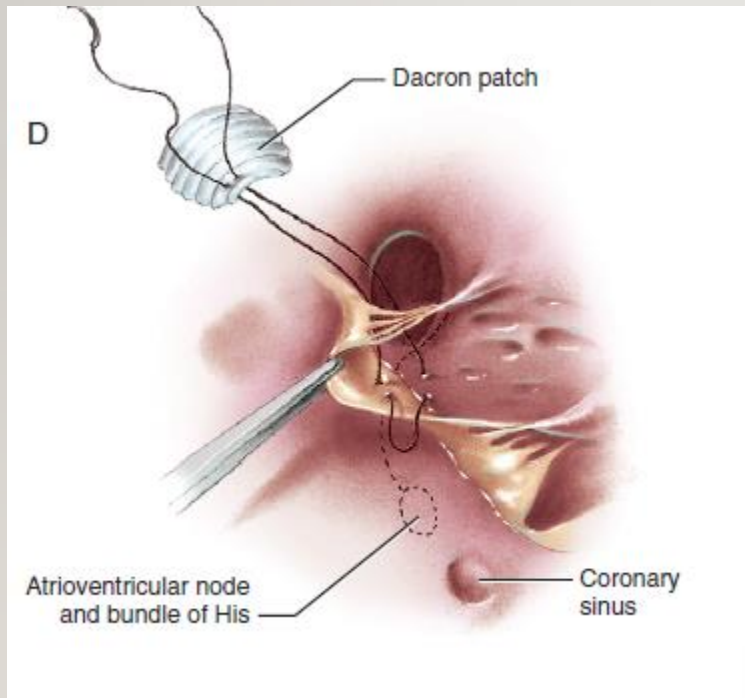
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- OP Indikation ist immer gegeben (VSD Spontanveschluß ist ausgeschlossen)
- Symptomatische Kindern: 3-4 Monat
- Asymptomatische: Später



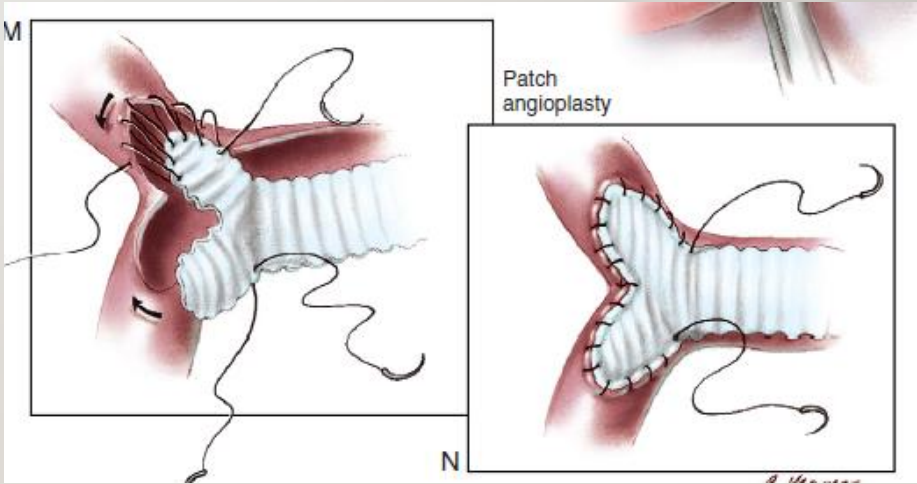
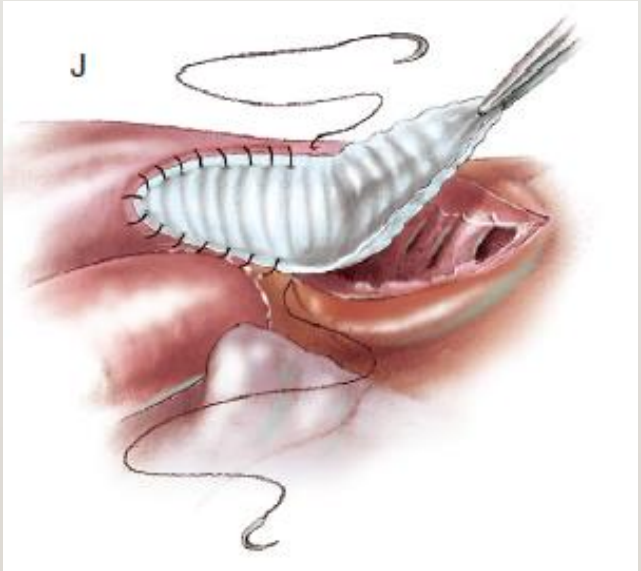
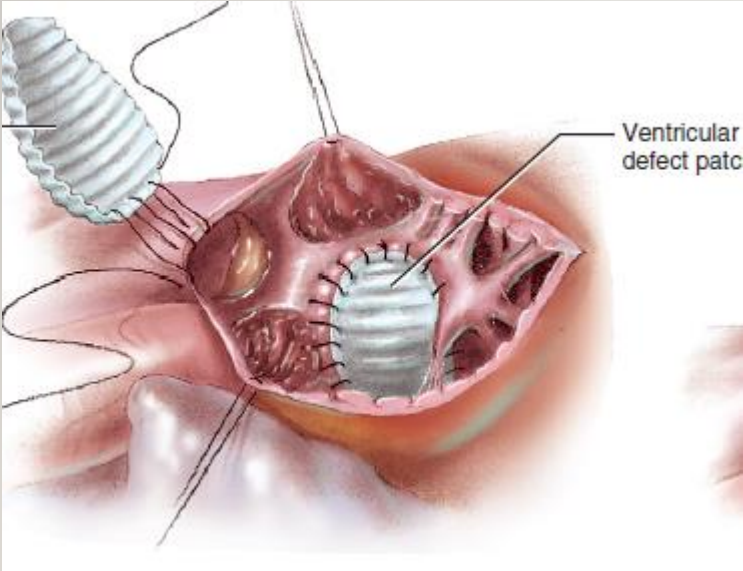
# FALLOT - TETRALOGIE

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# FALLOT - TETRALOGIE

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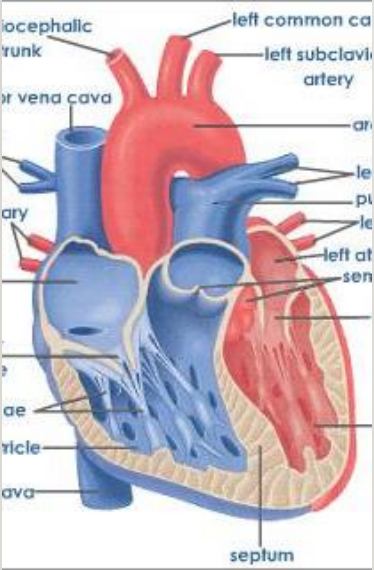
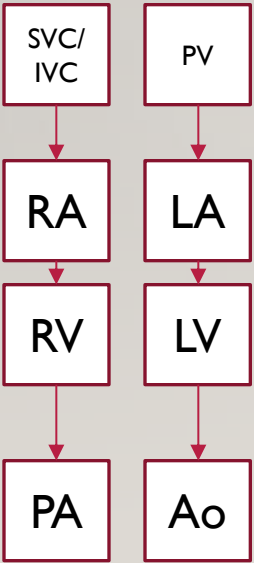
# ZYANOTISCHE HERZERKRANKUNGEN

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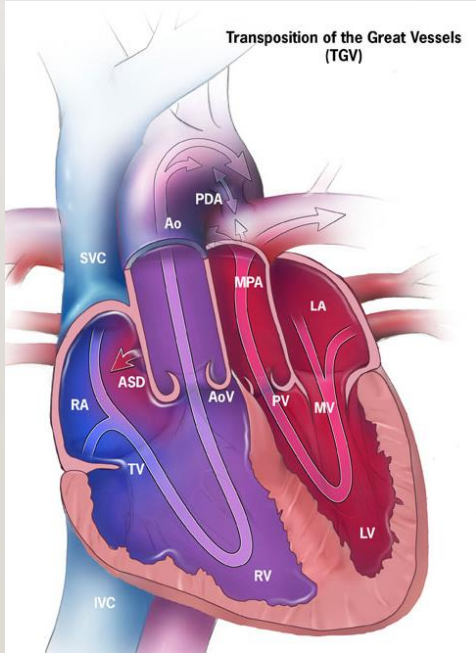
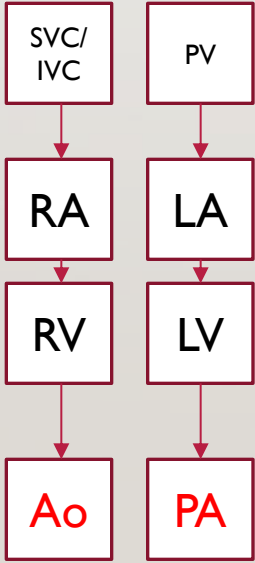
- Fallot –Tetralogie
- **Transposition der großen Arterien**
- Hypoplastisches Linksherz-Syndrom
- TK Atresie
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# TRANSPOSITION DER GROßEN ARTERIEN (TGA)

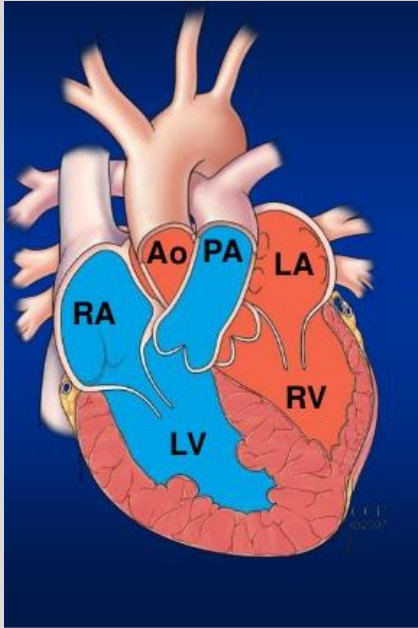
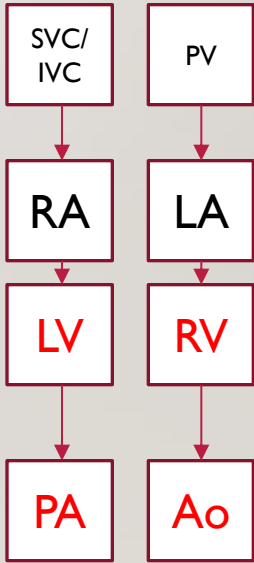
Normales Herz (S, D, S)



D-TGA (S, D, D)



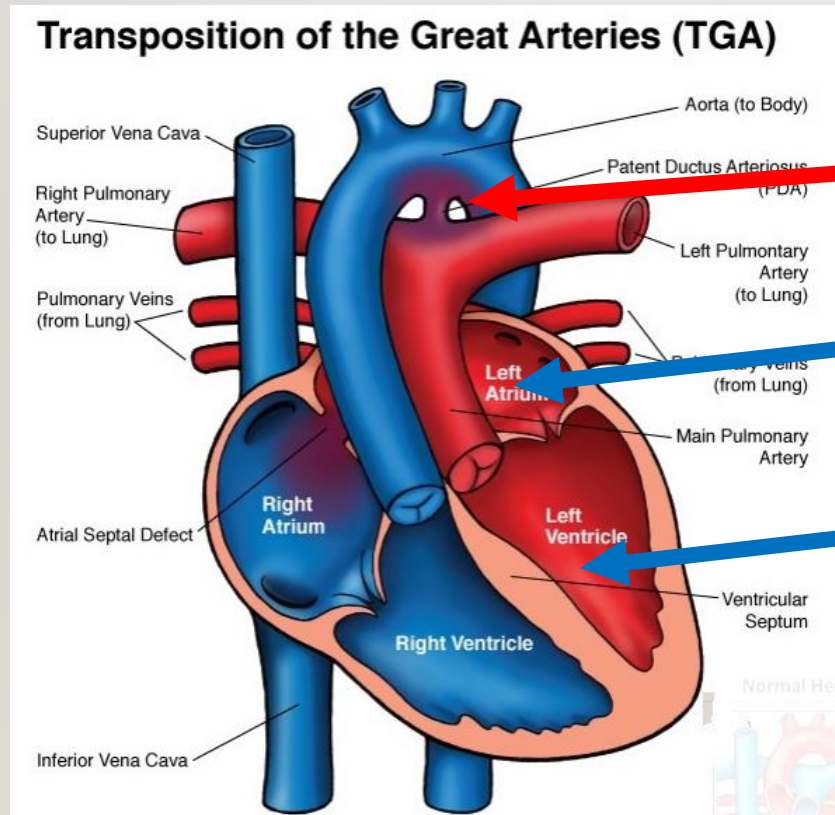
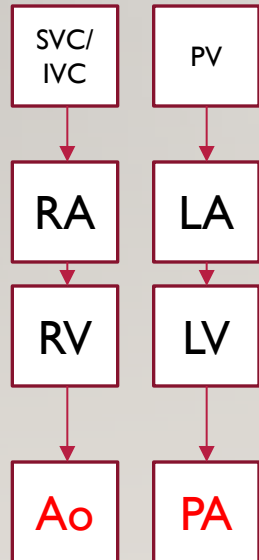
L-TGA (S, L, L)





# DEXTRO -TRANSPOSITION DER GROßEN ARTERIEN (D-TGA)

D-TGA (S, D, D)



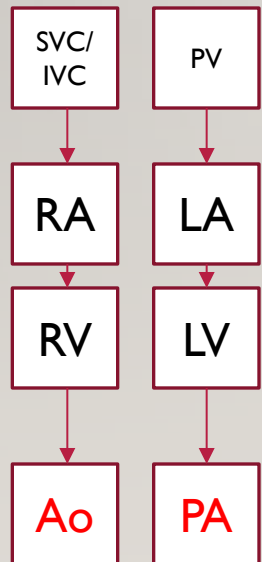
**PDA**

**ASD**

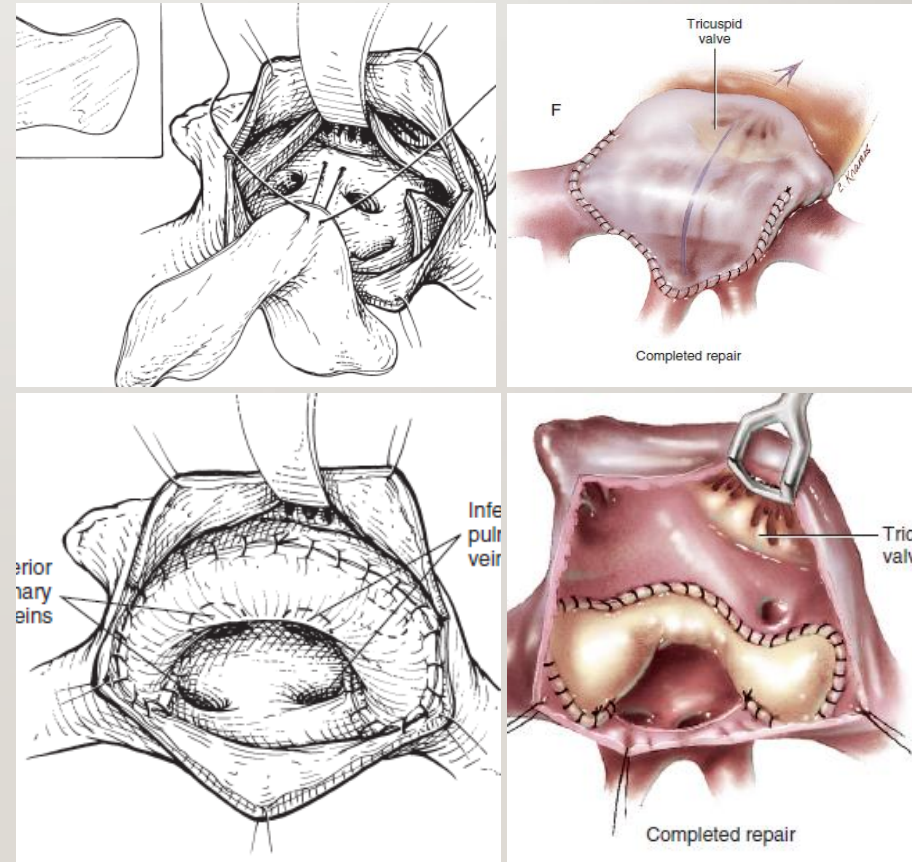
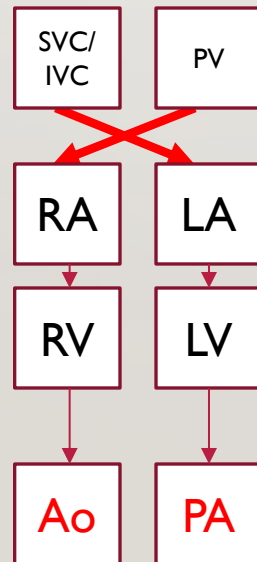
**VSD**

# DEXTRO -TRANSPOSITION DER GROßEN ARTERIEN (D-TGA)

D-TGA (S, D, D)



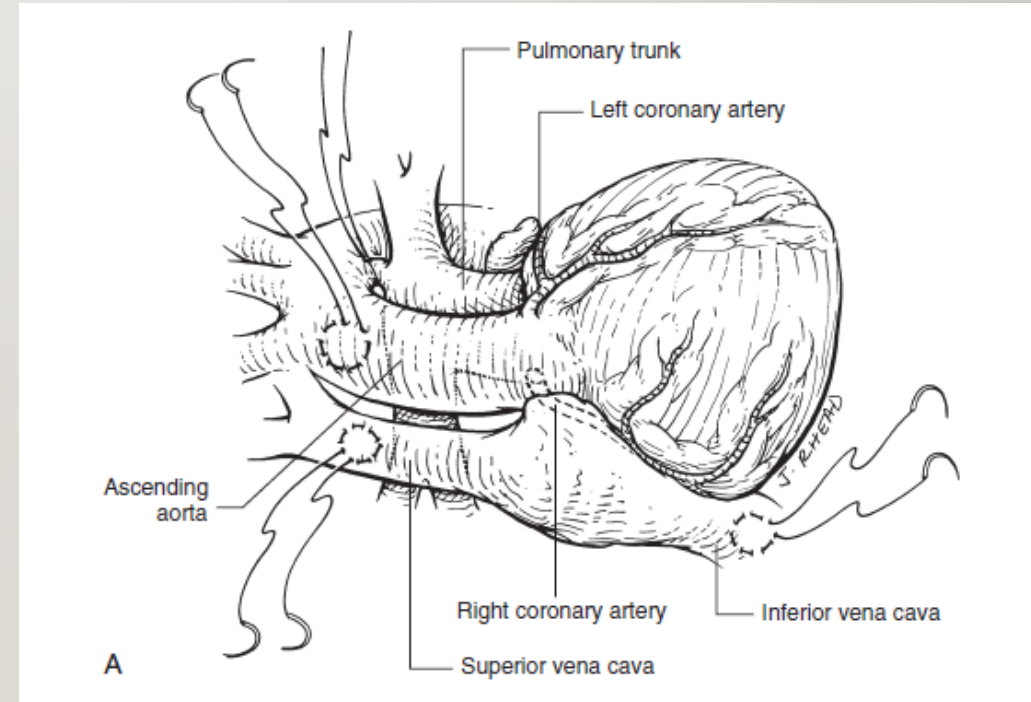
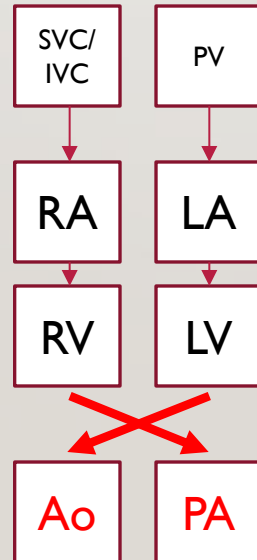
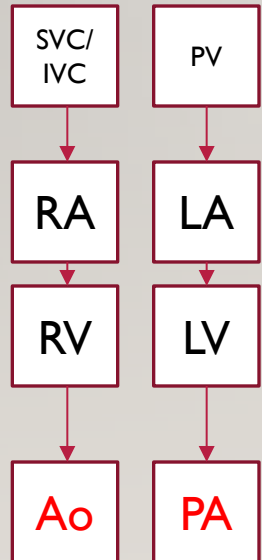
## Atrial Switch OP



# DEXTRO -TRANSPOSITION DER GROßEN ARTERIEN (D-TGA)

D-TGA (S, D, D)

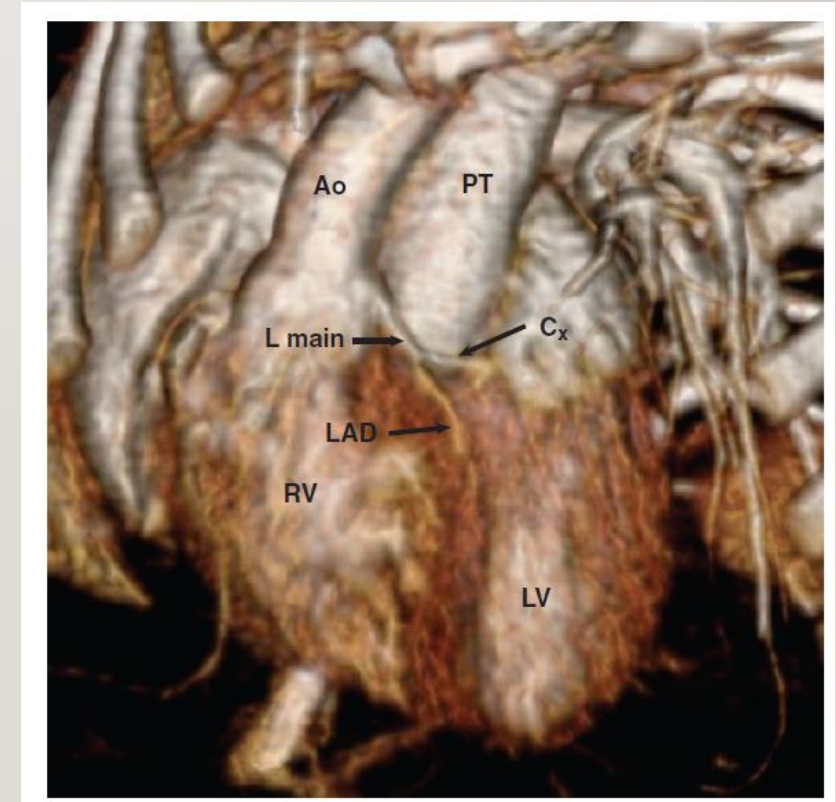
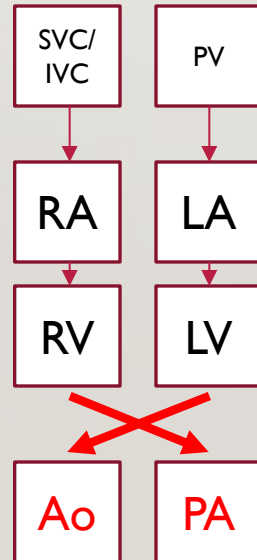
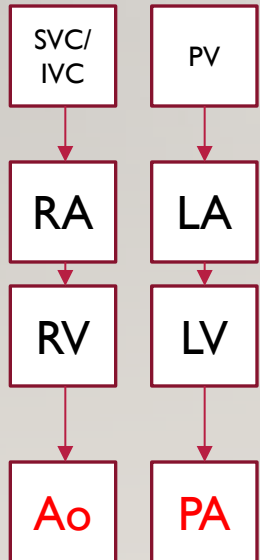
## Arterial Switch OP



# DEXTRO -TRANSPOSITION DER GROßEN ARTERIEN (D-TGA)

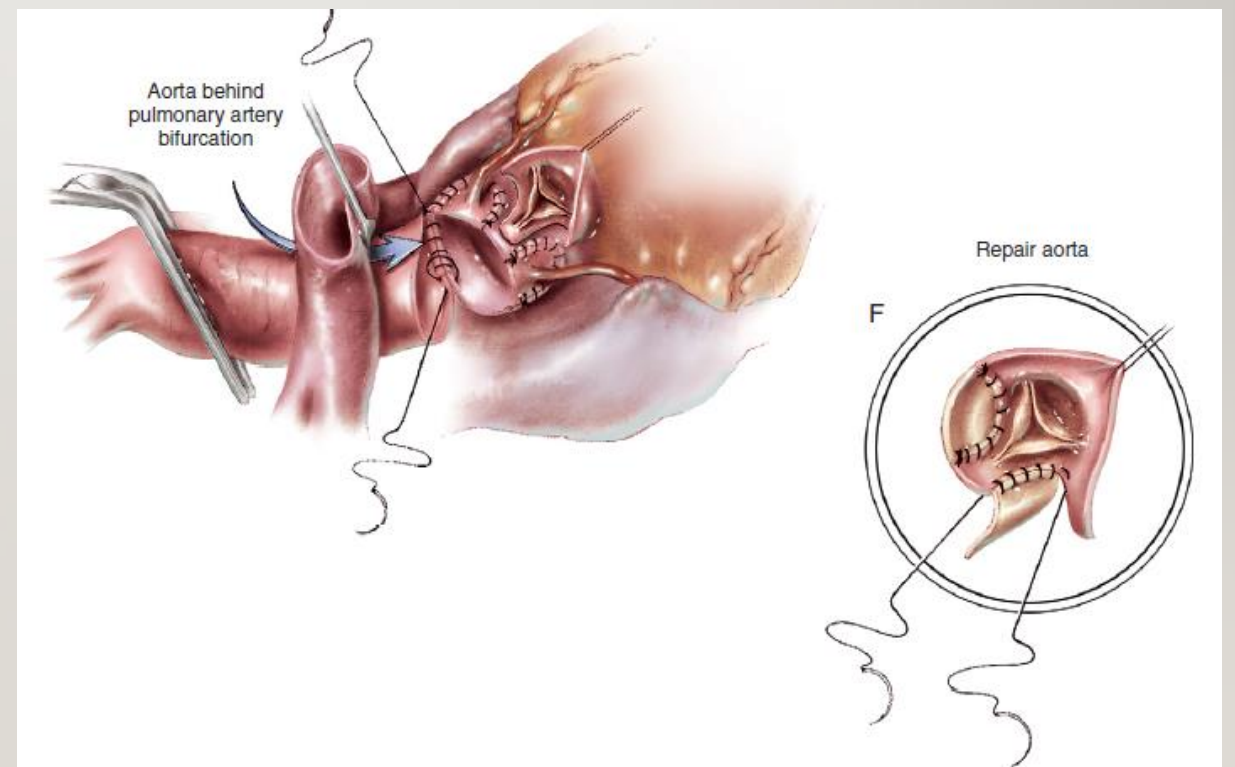
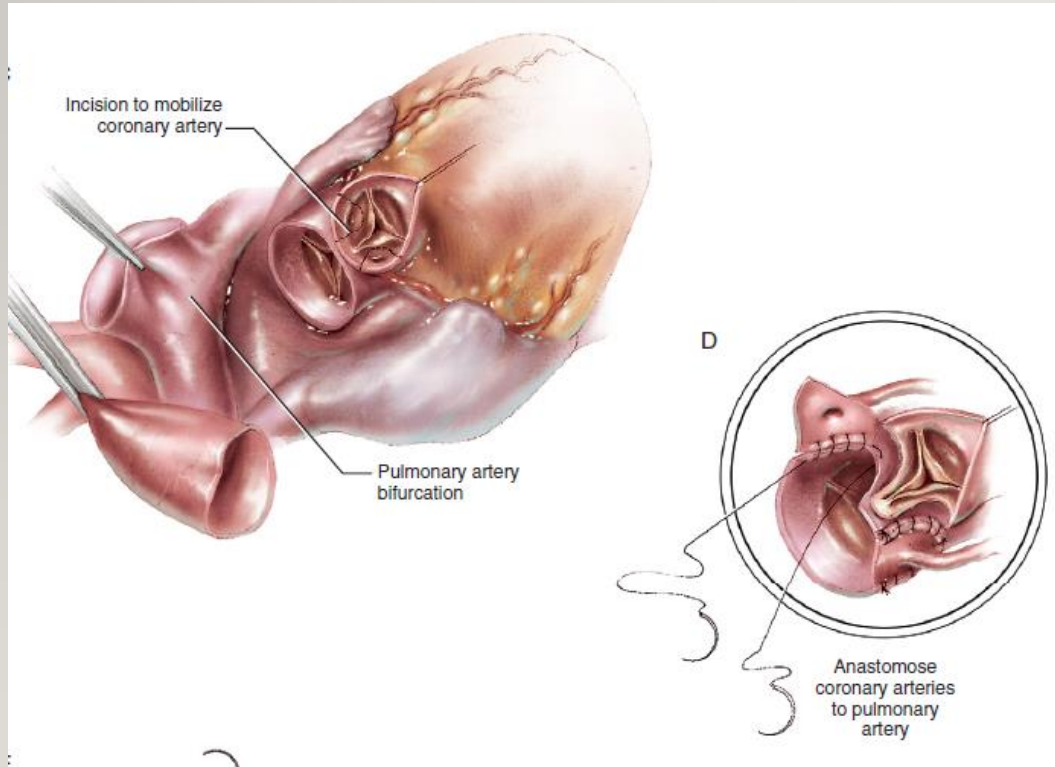
D-TGA (S, D, D)

## Arterial Switch OP



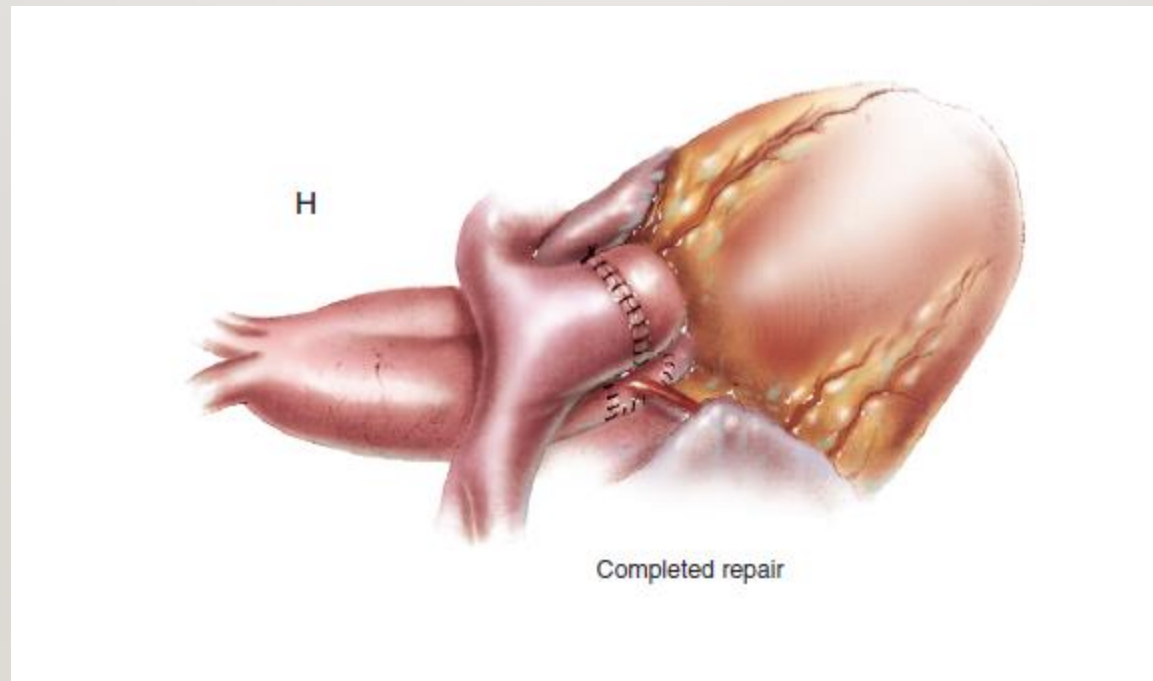
# DEXTRO -TRANSPOSITION DER GROßEN ARTERIEN (D-TGA)

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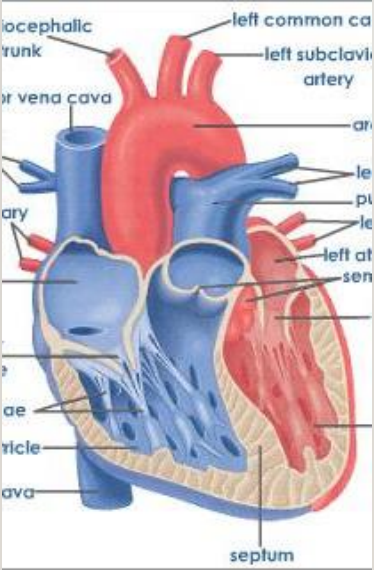
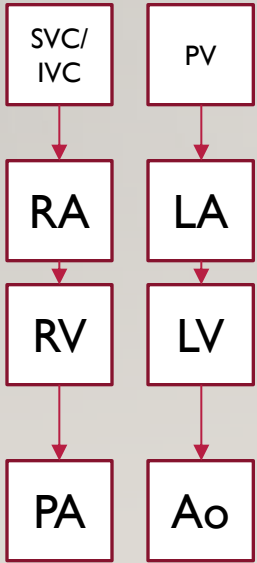
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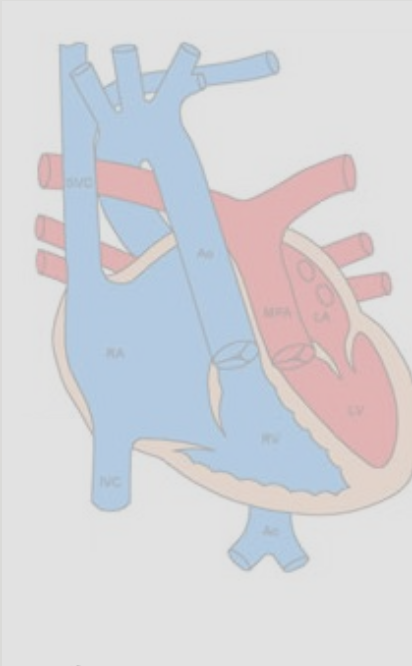
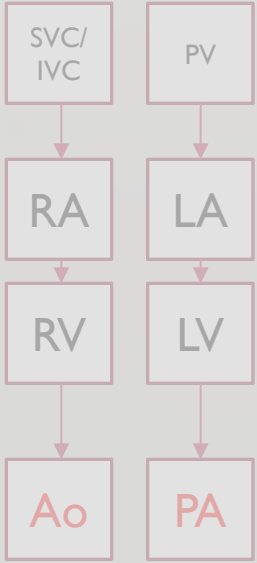


# TRANSPOSITION DER GROßEN ARTERIEN (TGA)

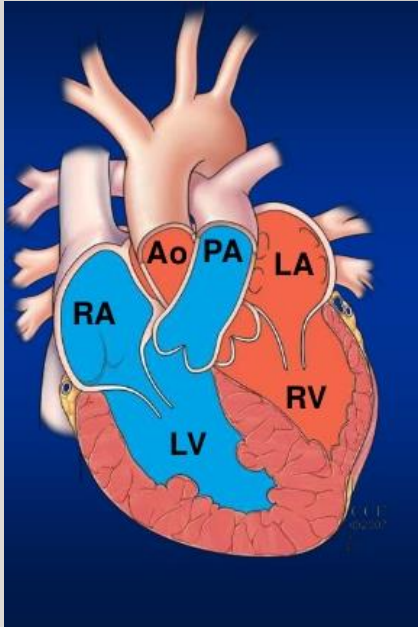
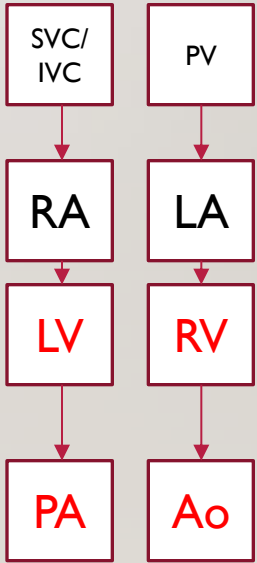
Normales Herz (S, D, S)



D-TGA (S, D, D)

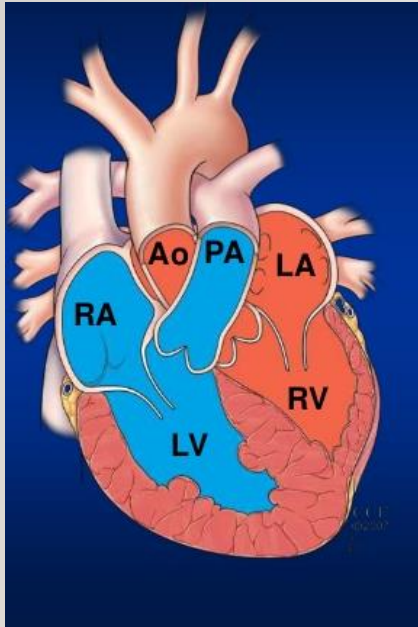
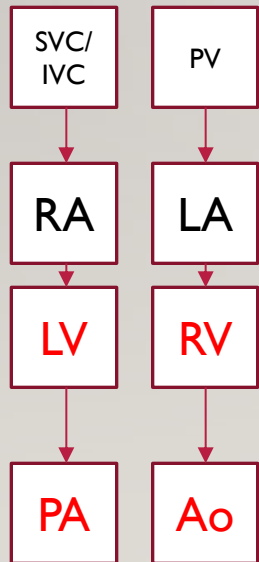


L-TGA (S, L, L)



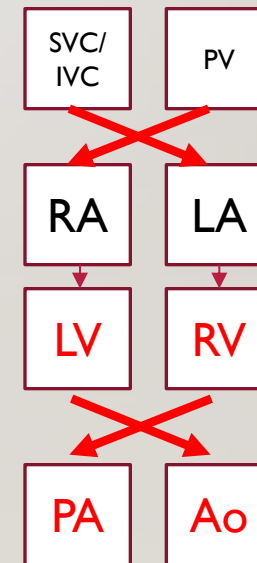
# TRANSPOSITION DER GROßEN ARTERIEN (TGA)

L-TGA (S, L, L)



- Asymptomatisch
- AV-Block, WPW
- Rechtsherzversagen

L-TGA (S, L, L)



Double  
Switch  
OP



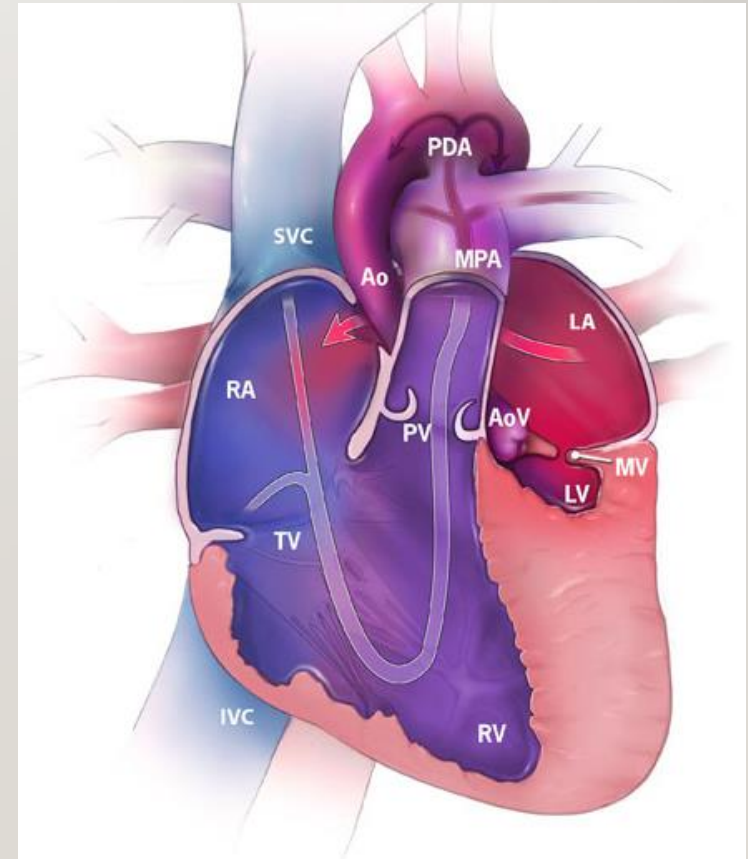
# ZYANOTISCHE HERZERKRANKUNGEN

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- Fallot –Tetralogie
- Transposition der großen Arterien
- **Hypoplastisches Linksherz-Syndrom**
- TK Atresie
- PK Atresie
- Ebstein Anomalie

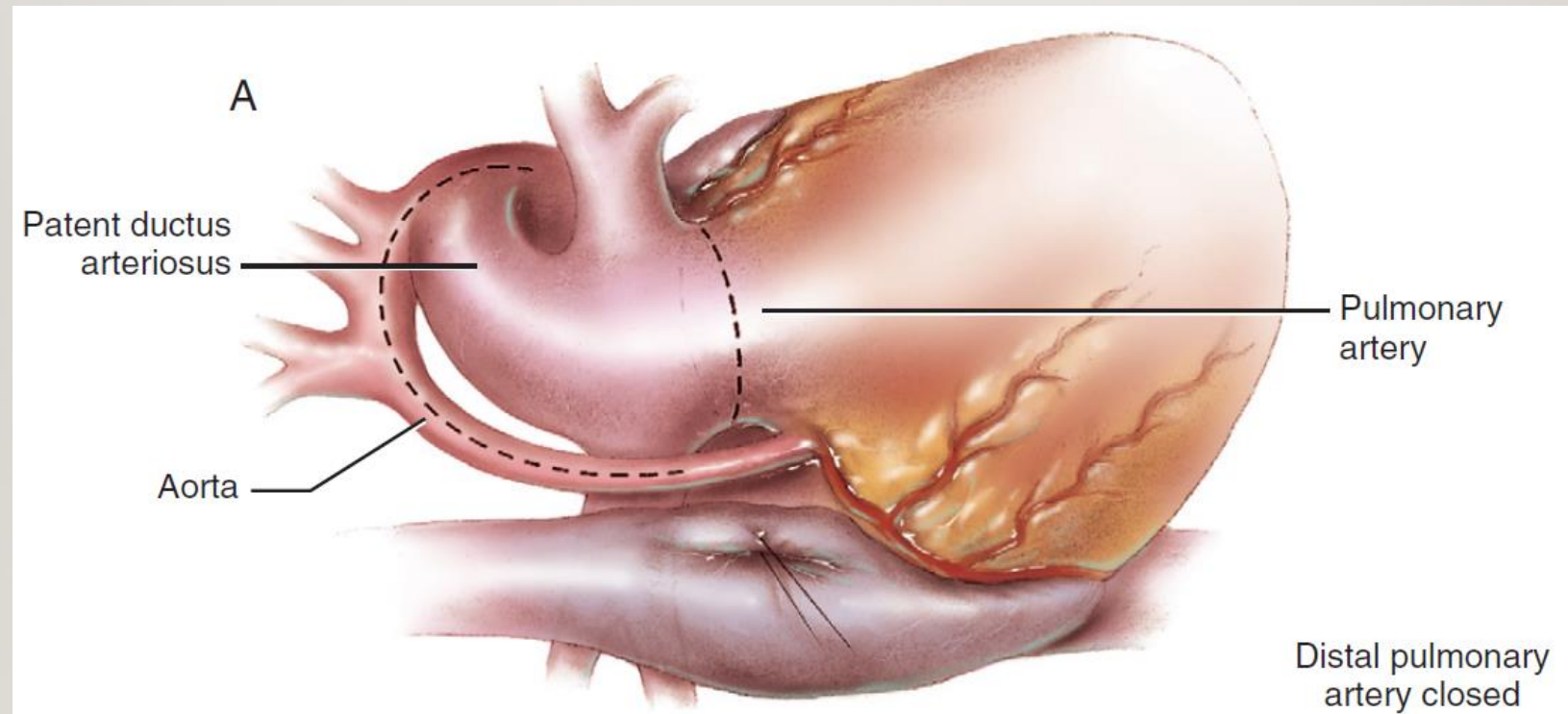
# HYPOPLASTISCHES LINKSHERZ-SYNDROM

- 1,6% aller herzkranken Säuglinge
- Häufigste Ursache für Herzinsuffizienz bei Neugeborene
- Häufigste Todesursache in der 1. Lebenswoche
- Norwood OP: wenig erfolgreich
- Initiierung neonatale Herztransplantation

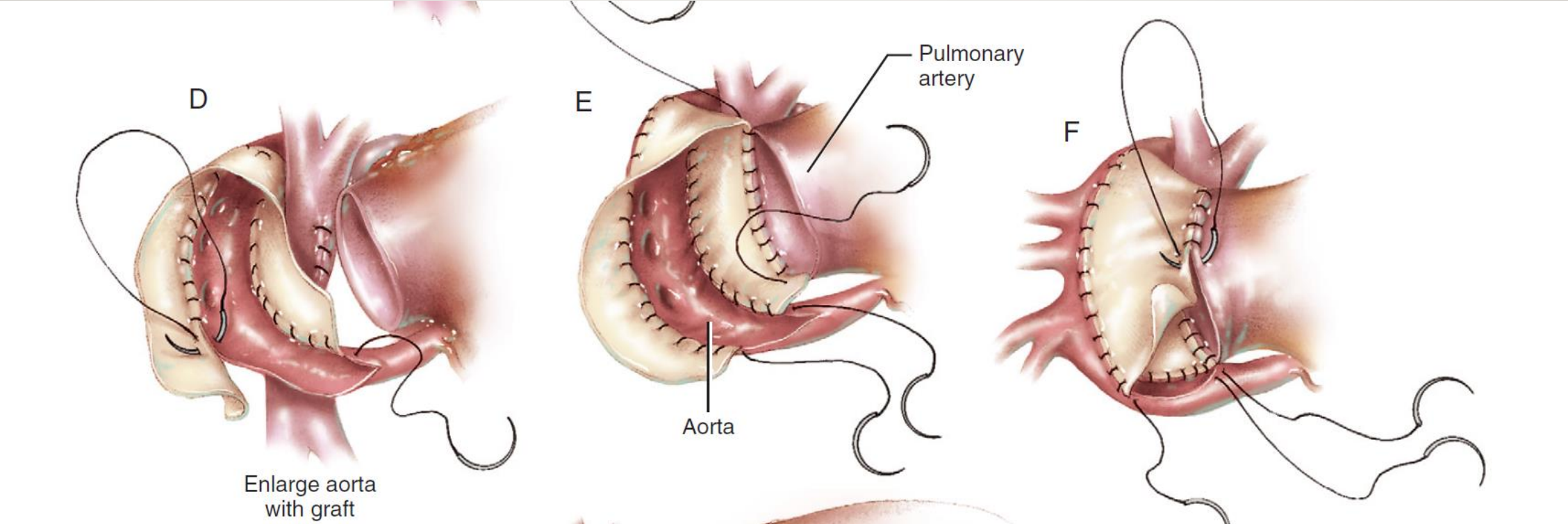


# HYPOPLASTISCHES LINKSHERZ-SYNDROM

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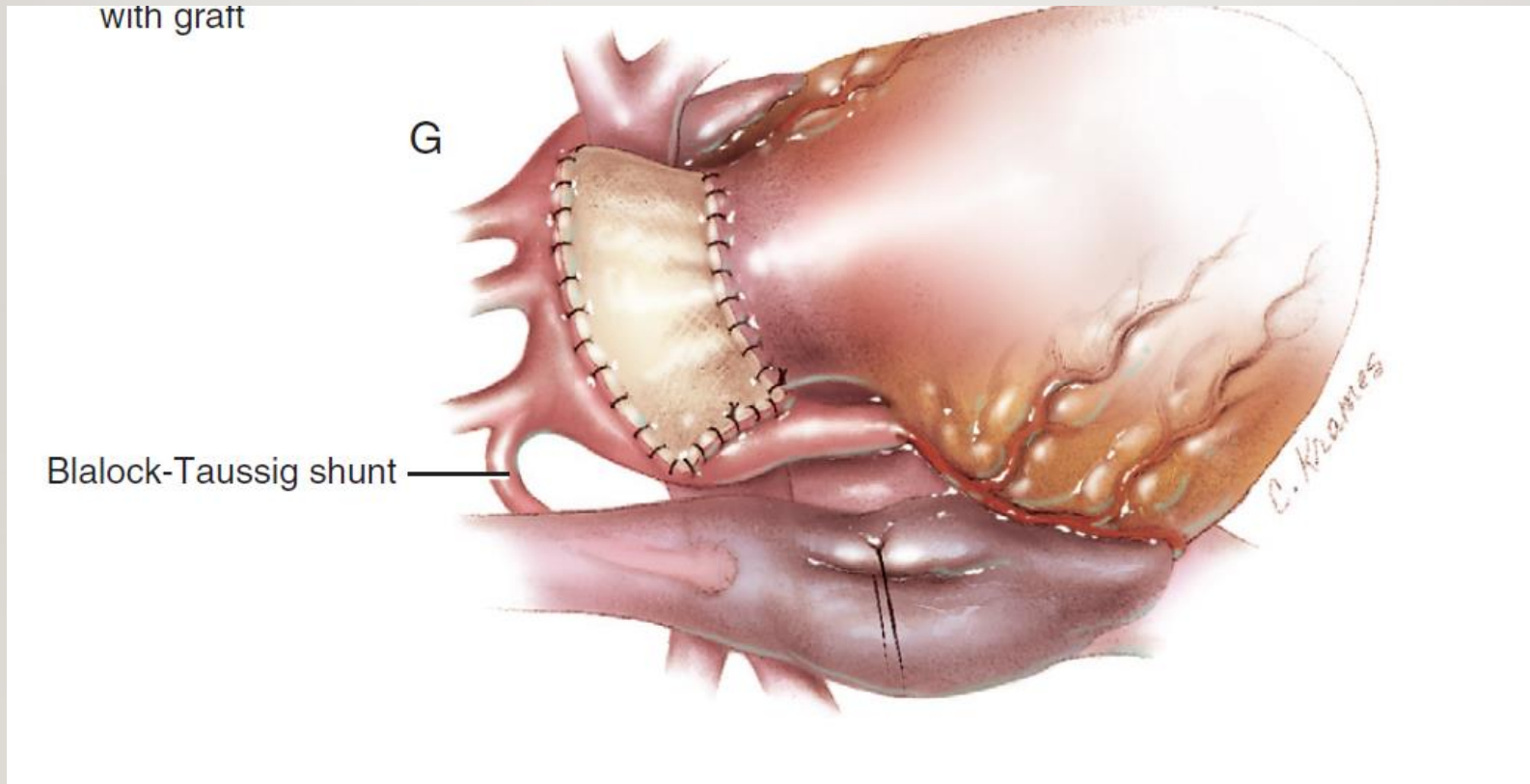


# HYPOPLASTISCHES LINKSHERZ-SYNDROM



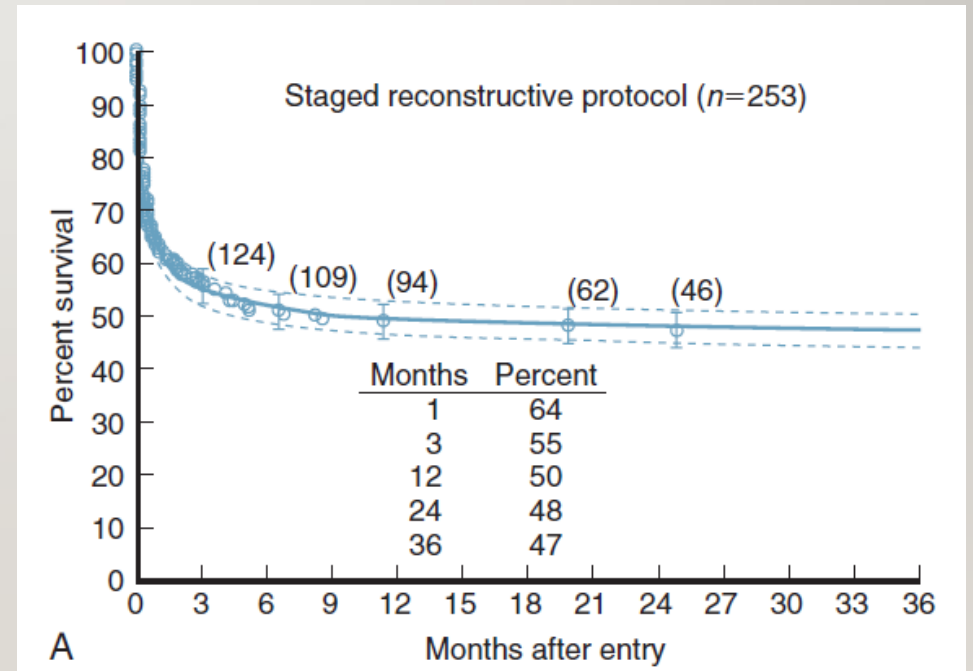
# HYPOPLASTISCHES LINKSHERZ-SYNDROM

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# HYPOPLASTISCHES LINKSHERZ-SYNDROM

- Ergebnisse:
  - Überleben:
    - I Monat: >80%
    - I Jahr: 60%



# HYPOPLASTISCHES LINKSHERZ-SYNDROM

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