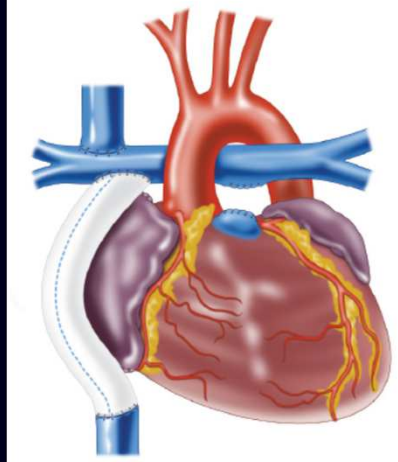
Cardiology 2018

IFIG
Scottsdale 2018

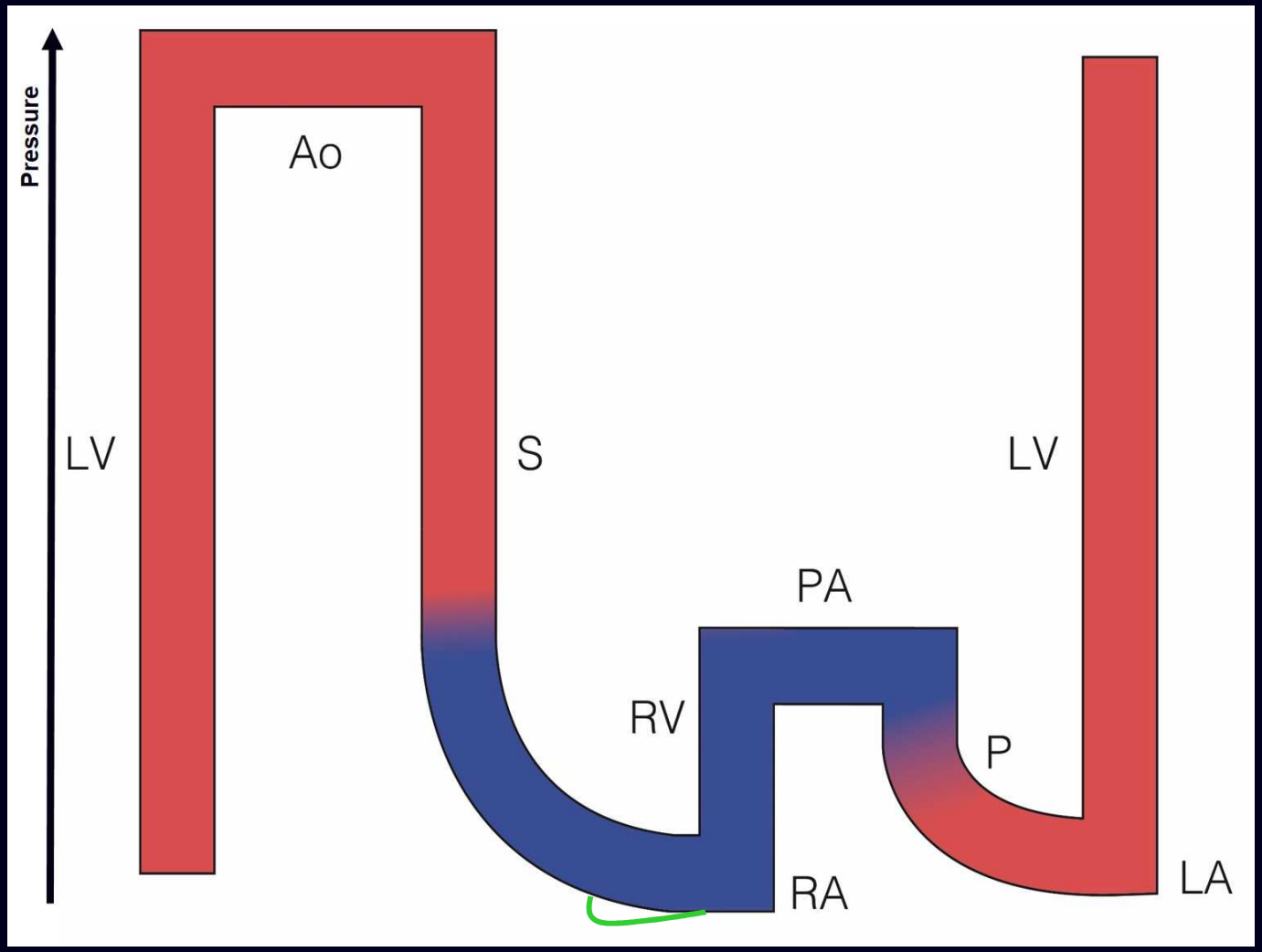


Pathophysiology and Mechanisms of Fontan Failure

No disclosures

Marc Gewillig, MD, PhD
Leuven, Belgium

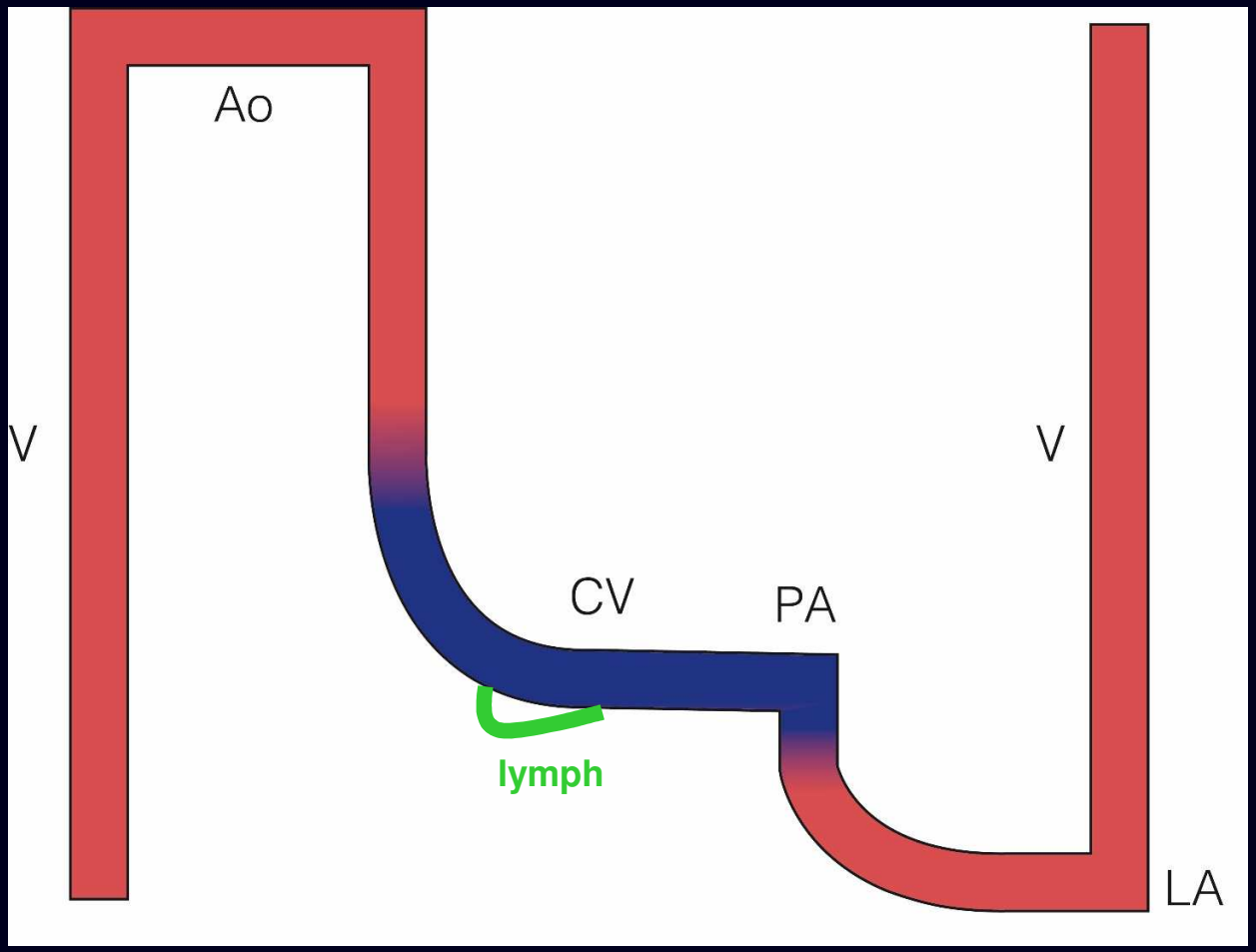
Normal biventricular circulation



- 0utput 100 – 500%
- RA pressure low
- PA pulsatility
- Ao sat > 95%

Gewillig TCS 2000

Fontan circulation: new portal system



- 0utput (50) 70 - 200%
- CV pressure high
- PA pulsatility ⇓
- Ao sat (80-) 93%
- prone to lymphatic leak

Portal system : dam

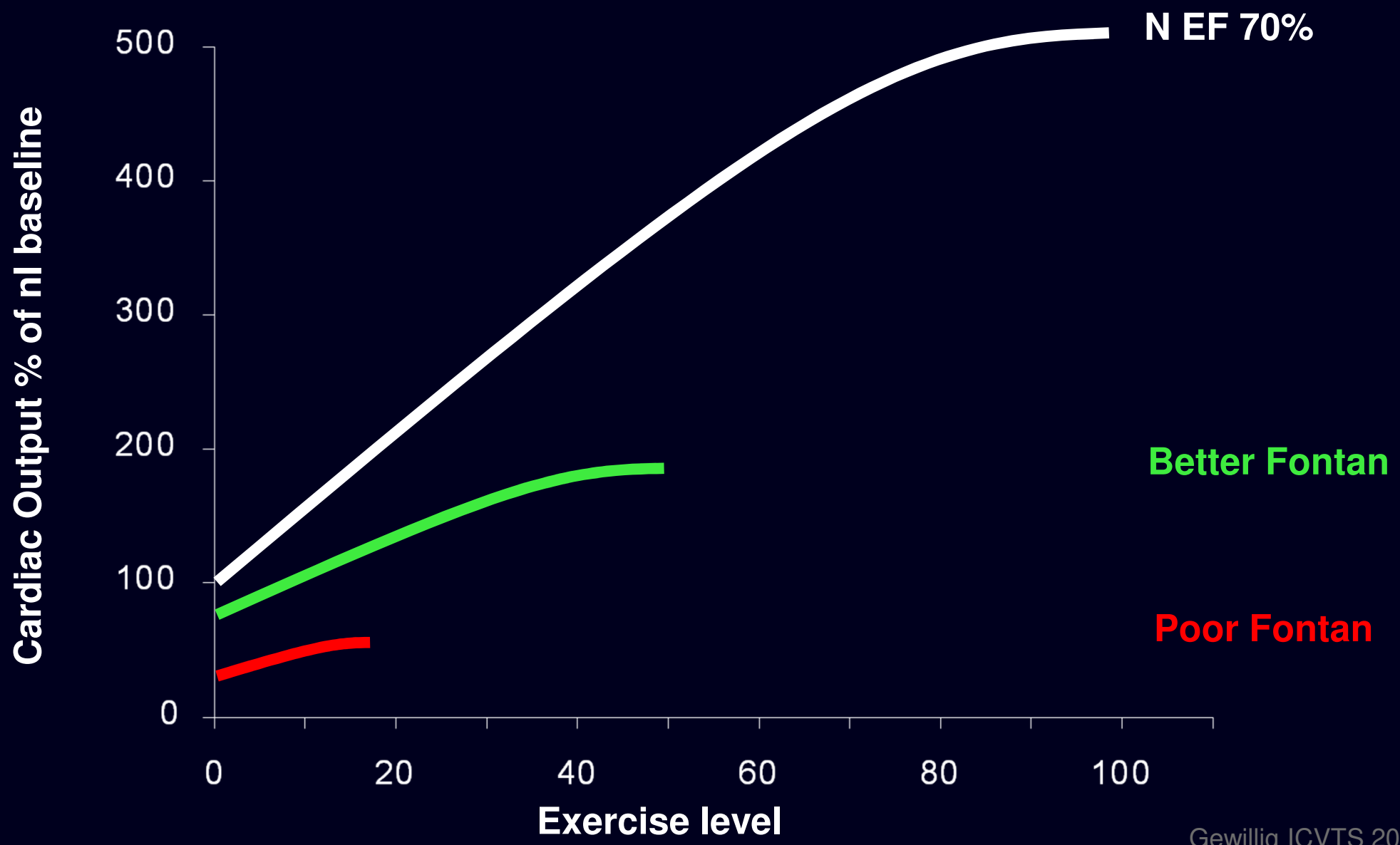
- congestion
- decreased output

Gewillig TCS 2000

Effect of a dam

- Upstream :
  - congestion
- Dam use:
  - continuous flow
  - inertion, non-pulsatile
- Downstream
  - controlled & decreased flow

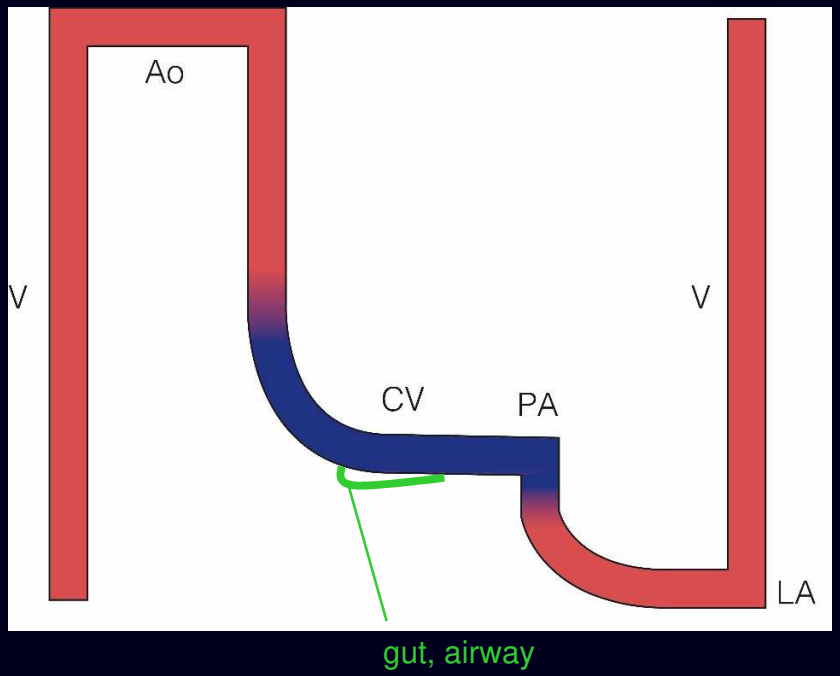
Exercise & CO: NI vs Fontan circulation



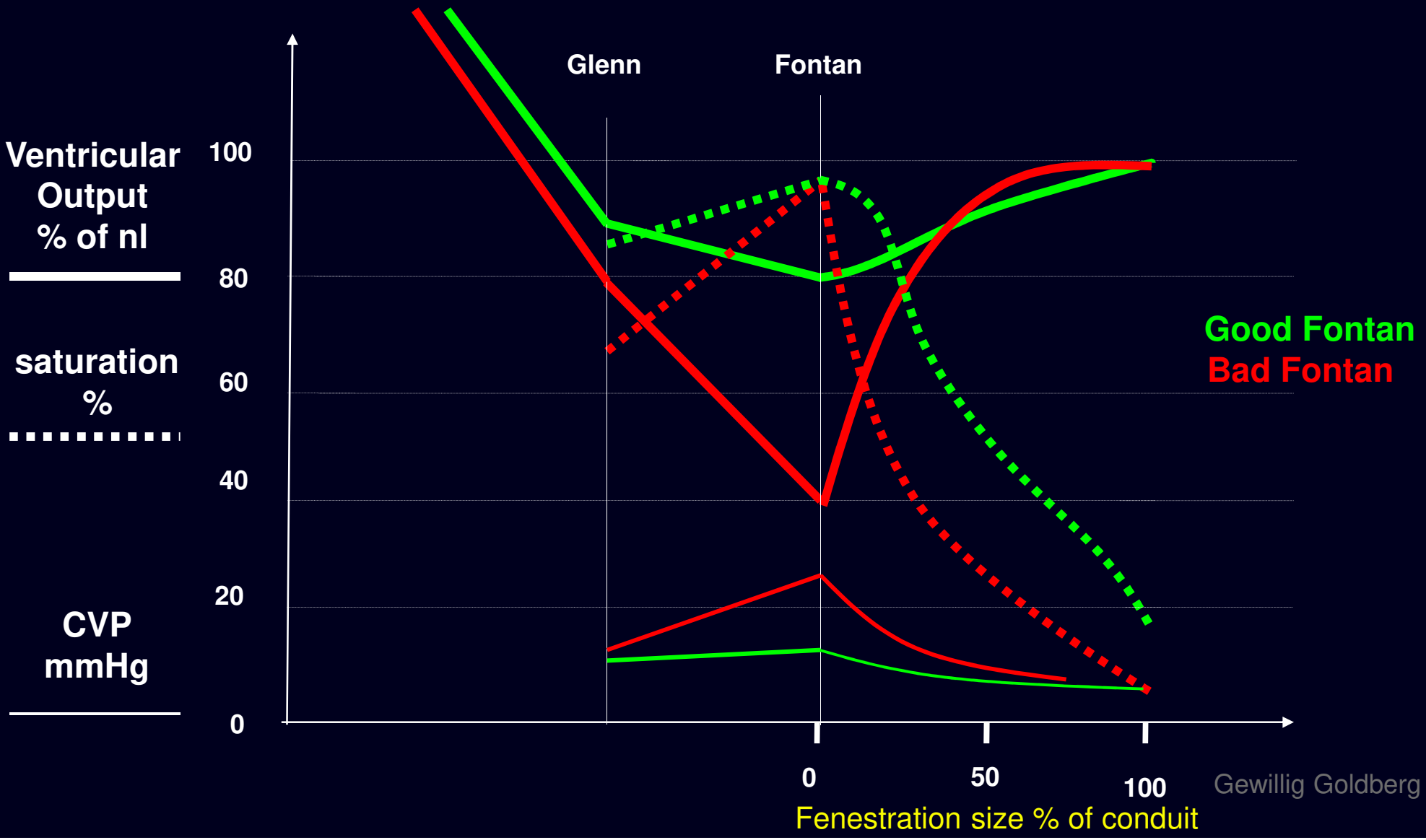
Gewillig ICVTS 2010

Fontan failure: excessive “damming”

- **Low output**
  - Exercise intolerance
  - Death
  - Pulmonary collateral flow
- **Congestion**
  - Edema, ascites
  - Liver dysfunction
  - Lymphatic failure: leak
    - PLE
    - Bronchial casts
- **Cyanosis**
  - R < L shunt
    - Intra pulmonary
    - Systemic veins < “LA”



Fontan : progressive “undoing”



Gewillig Goldberg 2014

The Fontan dilemma

Normal biventricular

No compromise

- **And full saturation**
- **And full output**
- **And no congestion**

Fontan circulation

Compromise

- **Or good saturation**
- **Or good output**

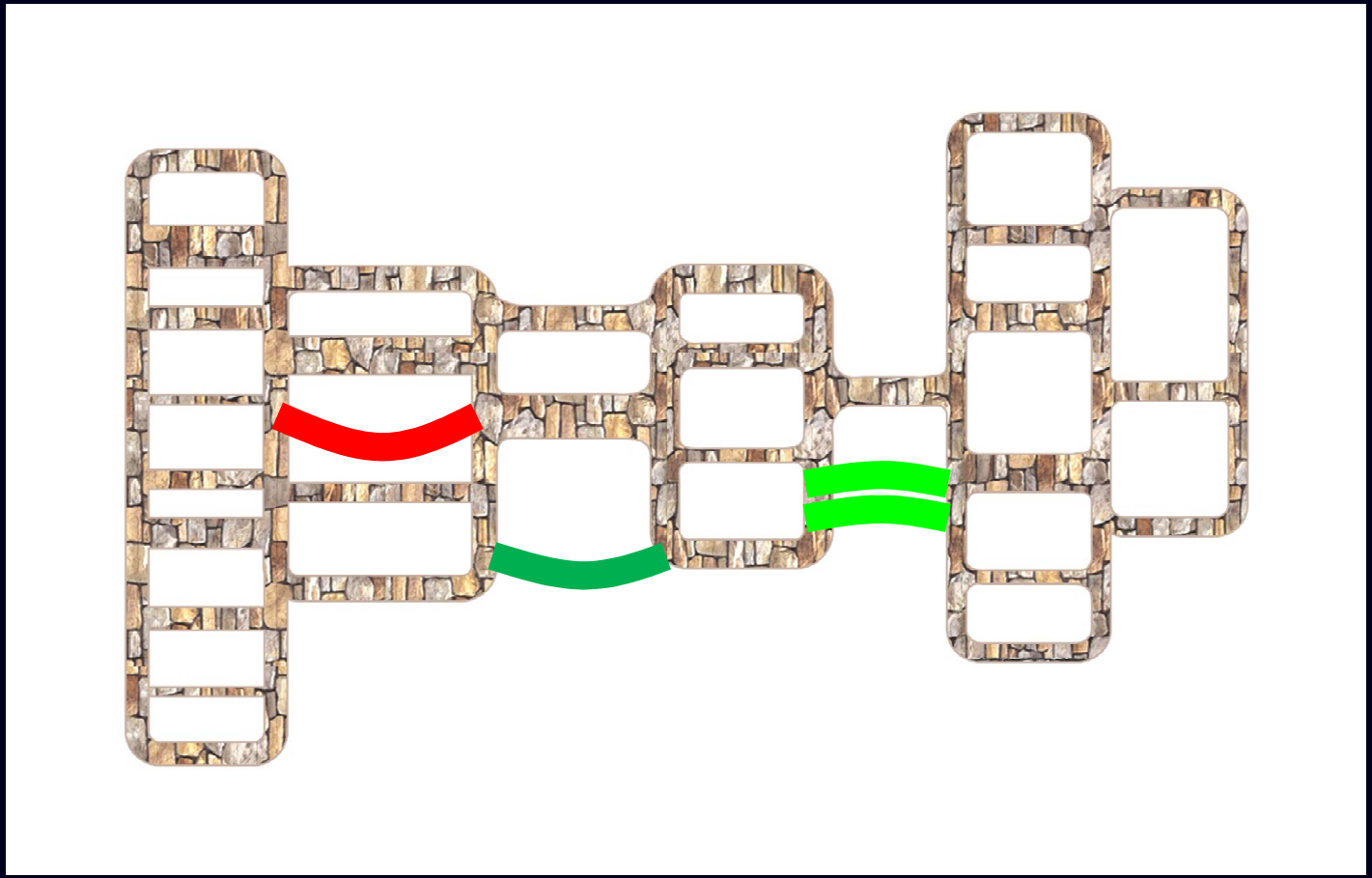
  & only moderate congestion

f° Ω of neo-portal Fontan

Multiple bottlenecks in series / steady state

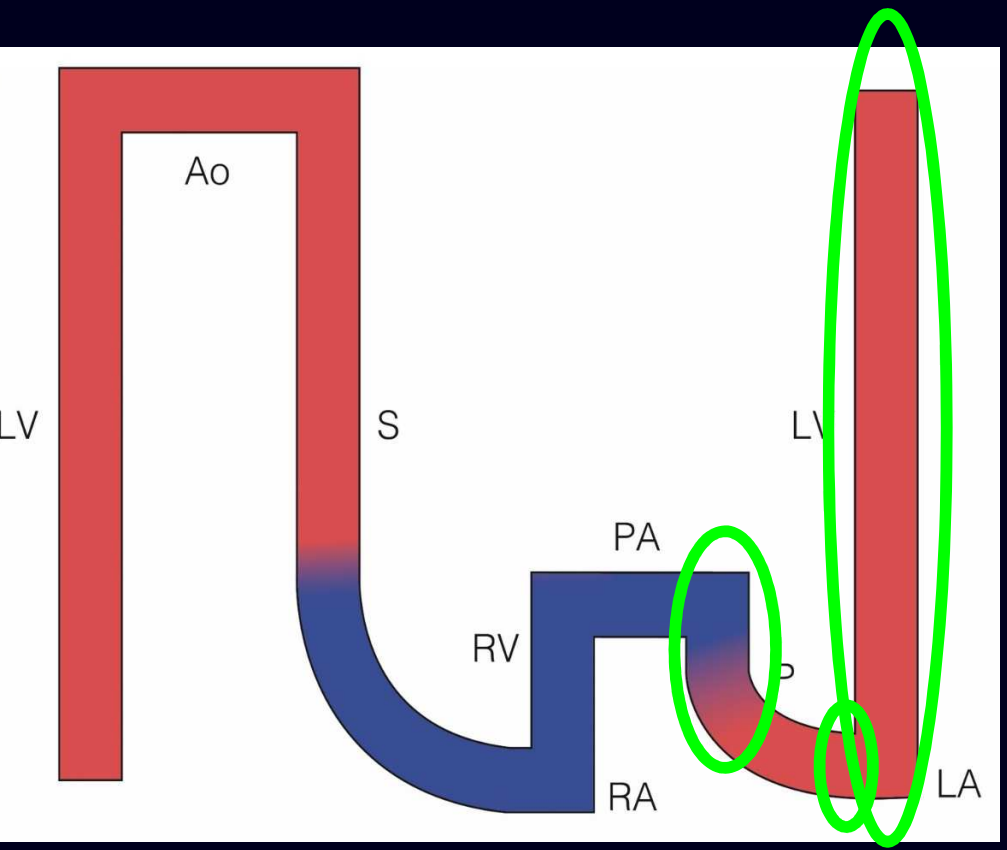
- **Multiple bottlenecks**
  - **1 critical bottleneck:**
    the only thing to work on !
  - **the other bottlenecks:**
    “irrelevant” unless shift
- **Alter effect bottleneck:**
  - **resistance**
  - **push above**
  - **pull below**

Multiple bottlenecks: go for the critical one !

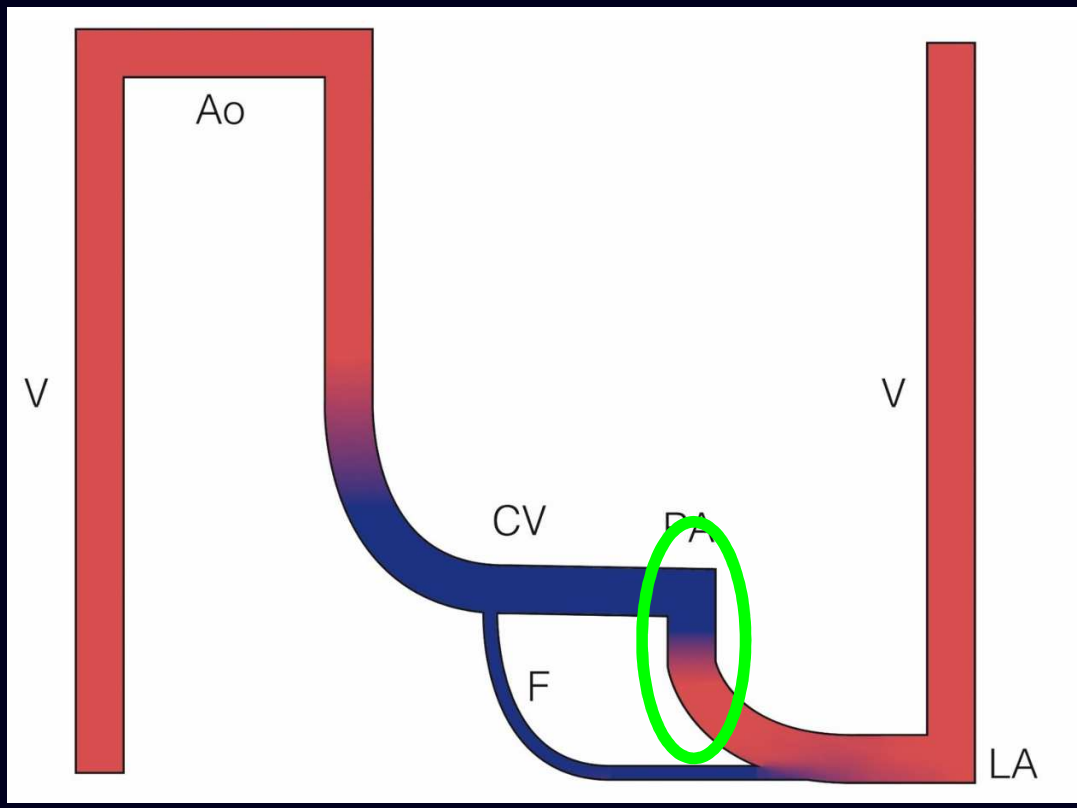


Determination of the critical bottleneck

Critical bottleneck in various circulations



- CMP
- PHT
- MS



- Fontan TCPC

Fontan : PA Flow

Pre Fontan : PA structural abnormalities

- stenosis, distortion, kink
- loss - exclusion of large vessels or micro-vessels
- hypoplasia, pulmonary vascular disease PVD
- external compression (artery – vein)

Post Fontan

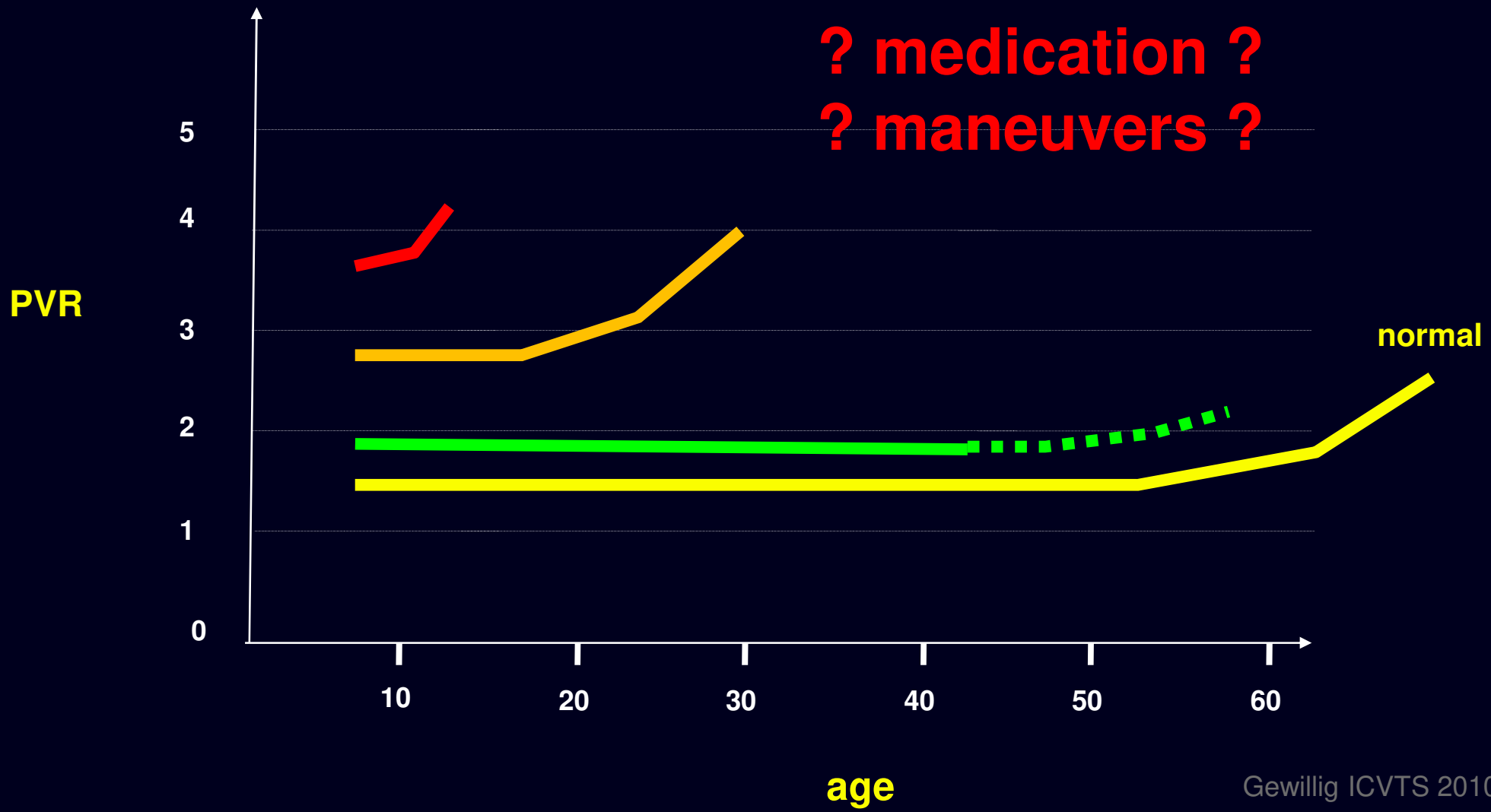
- No pulsatility, very steady
- Chronic low flow
- Absence episodes high flow – pressure, less recruitment
- Inhomogenous distribution “hepatic factor X”
- Collateral flow
- Vasoconstrictors – less vasodilators

Endothelial dysfunction

PVR ↑

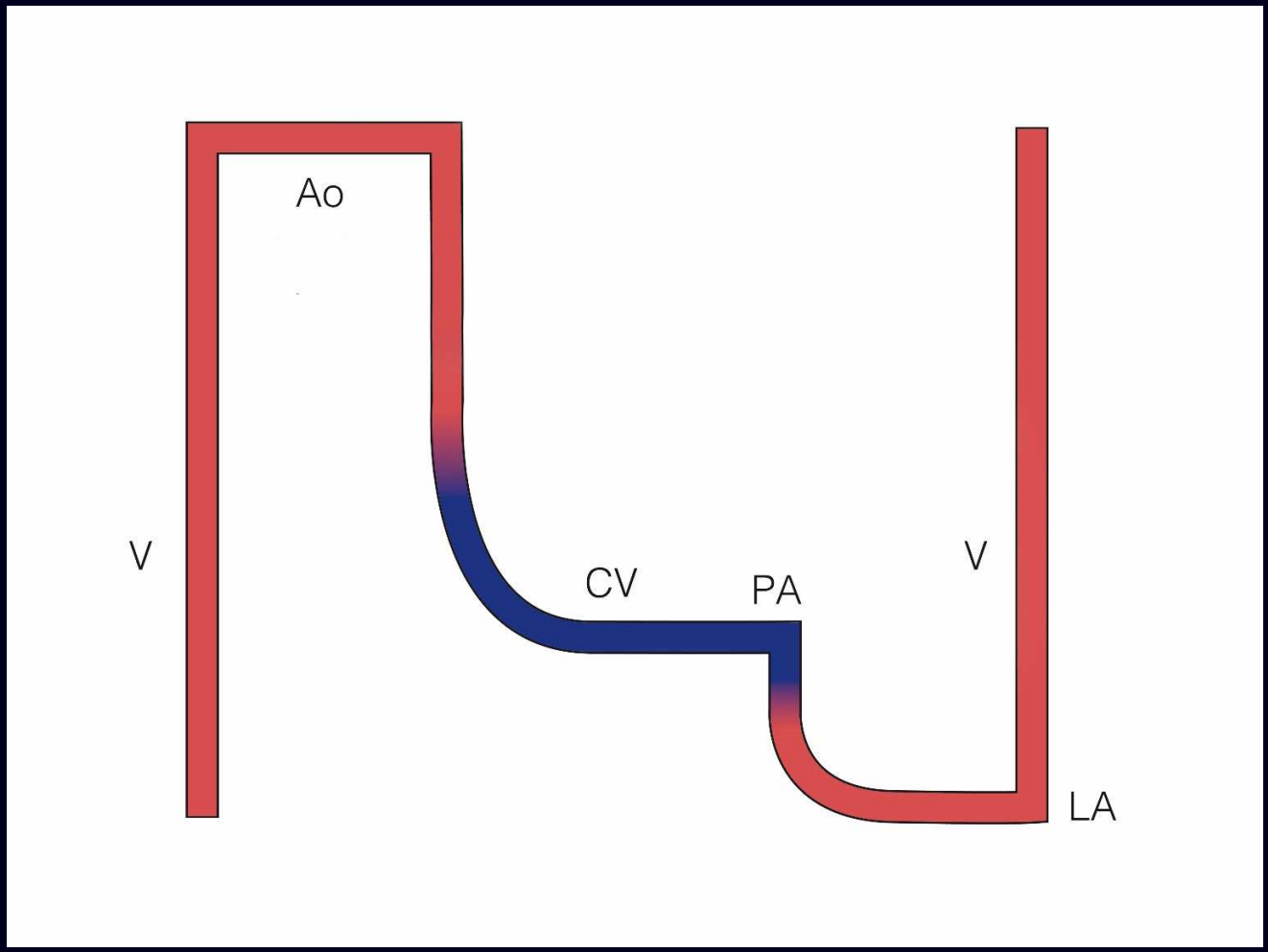
Gewillig, Heart 2016

Fontan : evolution of PVR



Gewillig ICVTS 2010

Fontan circulation: changes with time



PVR ↗ Output ↘ CVP ↗ VEDP ↗ EF ↘ SVR ↗

Gewillig Heart 2016

UVH : a ventricle with a history

Previously overloaded & stretched

fetal & initial shunt

Fontan: chronic volume deprivation

big overgrown but collapsed, strangled (? dilated)

time - failing

pressure/volume loop

Normal LV
UVH fetal
UVH shunted
UVH Fontan

New relationship V vs BSA
Loss relationship V vs BSA

Fontan : ventricle

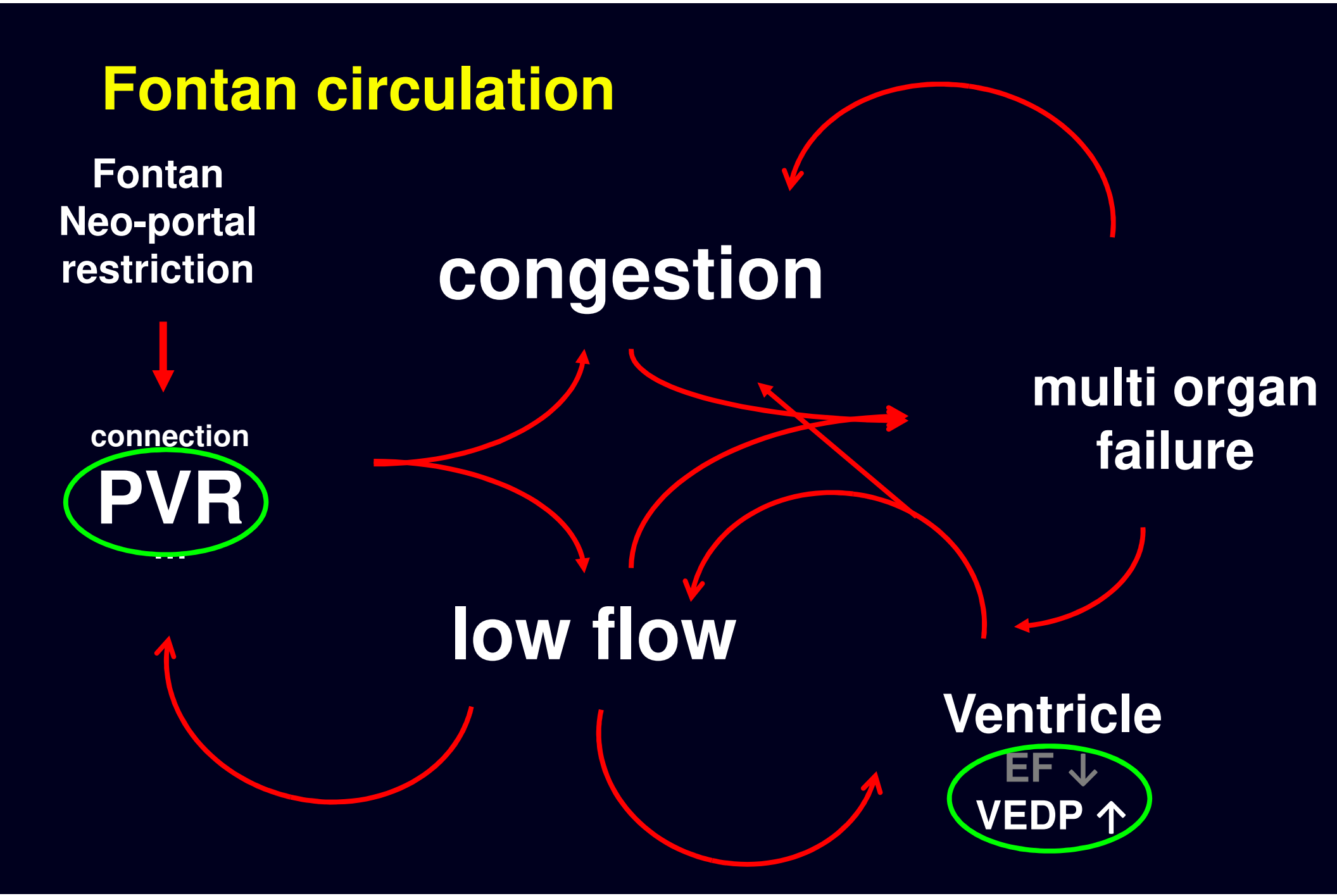
- **Pre Fontan**
  - **Overgrown:** preload ↑ ↑: fetal, initial palliation
  - **Excessive conditions:**
    - **preload ↑** shunt, MiVR
    - **afterload ↑** coarc

- **Fontan : abnormal boundary conditions**
  - **Preload: chronically deprived**
    - Chronic decreased preload
    - Exercise: no or limited stretch
    - Close to closing volume
  - **Heart rate: blunted**
  - **Afterload: decreased** (less volume against ≈ pressure)

VEDP ↑↑

EF ↓

Fontan circulation

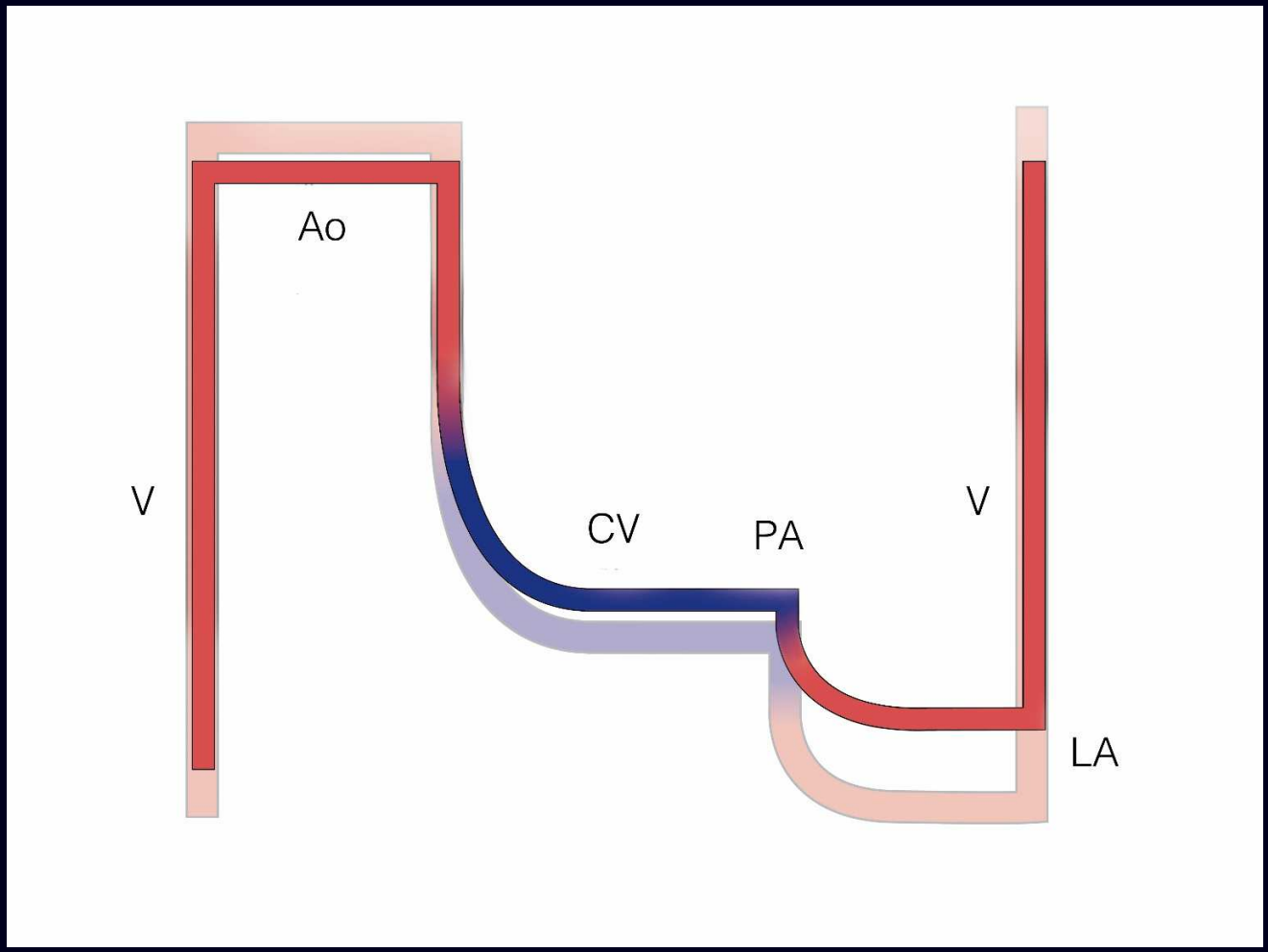


Fontan: effect of damming by portal system

- **Primary effects**
  - **Upstream: congestion**
  - **Lung: low flow with inertia (continuous non-pulsatile); oxygenation**
  - **Downstream: decreased flow**

- **Secundary effects** **Epiphenomenon: alterering will not obligatory affect primary problem**
  - **Congestion**
    - Oedema, ascites
    - Lymphatic system: PLE, bronchial cast
    - Liver fibrosis, cirrosis, CA
  - **PVR** ↑ (low flow, no pulsatility, absence high flow – high pressure, vasoactors, no recruitment)
  - **Ventricle**
    - Systole: contraction ↓
    - Diastole: suction ↓ compliance ↓ filling pressure ↑
  - **SVR** ↑ ventriculo-arterial coupling
  - **Collaterals :** veno-venous, arterio-arterial
  - **Humoral :** homeostatic reactions

Fontan circulation: changes with time



PVR ↗ **Output ↘** **CVP ↗** **VEDP ↗** **EF ↘** **SVR ↗**

Gewillig Heart 2016

Fontan : flow through critical bottleneck

1. **Pre bottleneck:**
   - Muscle pump
   - Mechanical pump CV > PA : « RVAD » (HTX)
2. **Bottleneck:**
   - Optimise Fontan connection : reconvert, streamline, conduit size
   - Dilate – stent PA stenosis, occlude collaterals
   - Dilate – stent PV « stenosis » compression
   - PVR: Pulmonary vasodilators : Mitchell 2004 at HTX: PVR 2.8 – 5.4 WU/m^2 !!!
     - Oxygen — altitude
     - NO
     - PD5-inh Ca — Sildenafil
     - Prostacyclin — Treprostinil, inhaled Iloprost
     - Endothelin A antagonist — Bosentan, Sitaxsentan, Ambrisentan
   - Respiration: asymmetric, assisted neg pressure
   - Recruitment: exercise – high flow/pressure
   - Anticoagulation
3. **Post bottleneck:**
   - Enhance ventricular suction - compliance – decrease filling pressure
     - Lusitropic drugs
     - Stretch ventricle
     - LVAD (HTX)

Fontan failure : conclusions

- **Fontan: a new critical bottleneck in circulation**
  - Wide range of functional results: from subnormal to failure
  - Expect no improvement of treatment if no effect on critical bottleneck
- **Fontan CB has & enhances interlocked downward spirals**
  - PVR
  - VEDP
- **Best way to avoid failure: optimal building blocks**
  - **Lungs** low PVR, growth by initial shunt
  - **Heart** compliance
- **Slowing & reversing the interlocked downward spirals is difficult**

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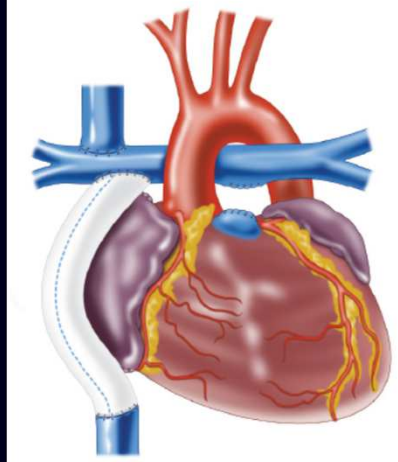

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