

Pulmonary Hypertension

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Heterogeneity of PHT in ACHD

Pulmonary Arterial Hypertension

Presence of a relevant shunt

- Atrial / ventricular or arterial
- Surgically created shunts

Prior shunt at any level
,Small' residual shunt

Impact on morbidity and mortality

Impact on Outcome

Underlying congenital heart defect



Non-restrictive communication

◆ **Absence of a shunt**
◆ **Restrictive shunt**



Cyanosis
2° Erythrocytosis

Pressure / volume overload of the ventricle(s)

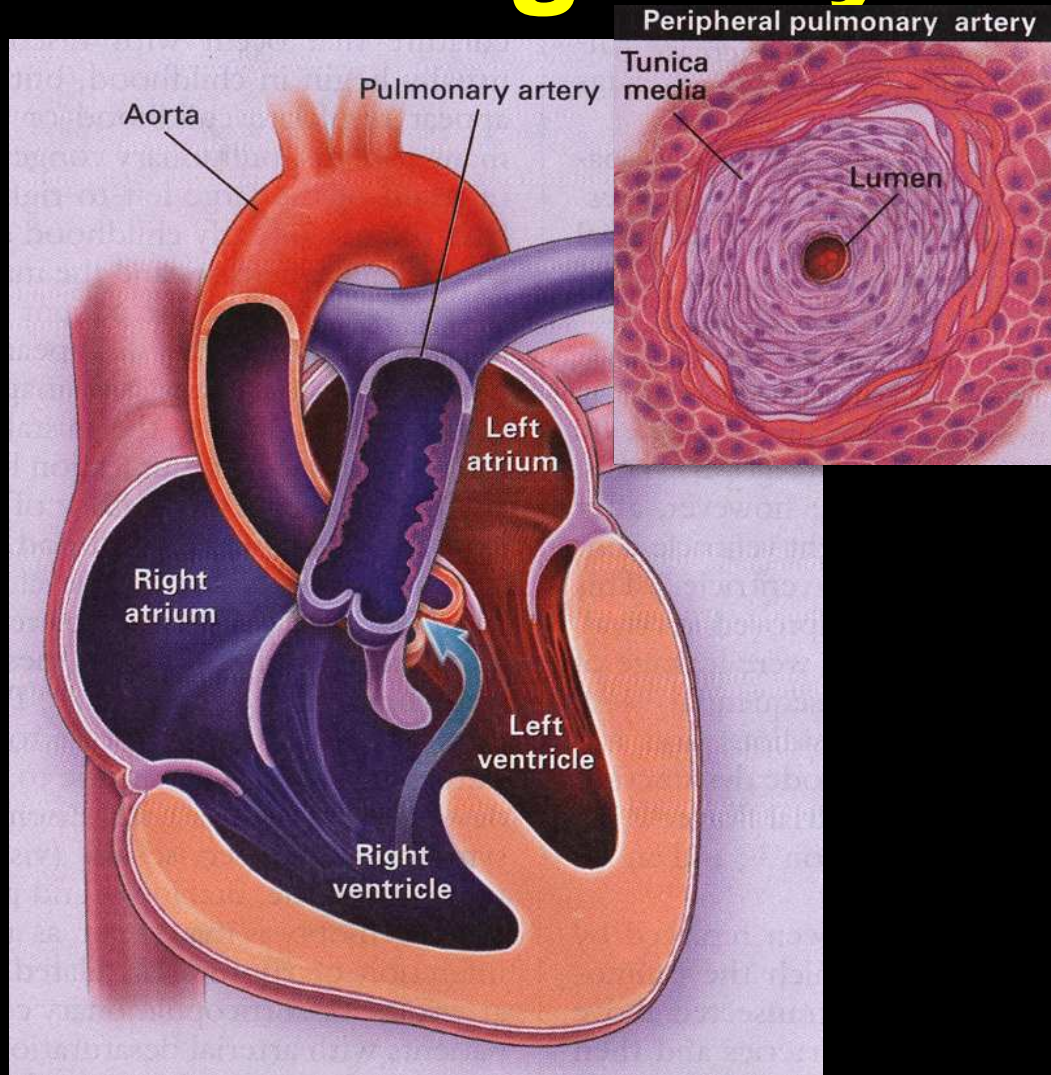
Complications

- Heart failure
- Hyperviscosity symptoms
 - Headache, fatigue....
- Infectious disease
- Bleeding
- Thromboembolic complications

Complications

- Gouty arthritis
- Risk:
 - Non-surgical procedures
 - Pregnancy
- Medication:
 - Impact on circulation

Eisenmenger Syndrome



PAHT in Adults: Multisystem-Disorder

Hematology

Heart failure

Pulmonary complications

Arrhythmias

Endocarditis

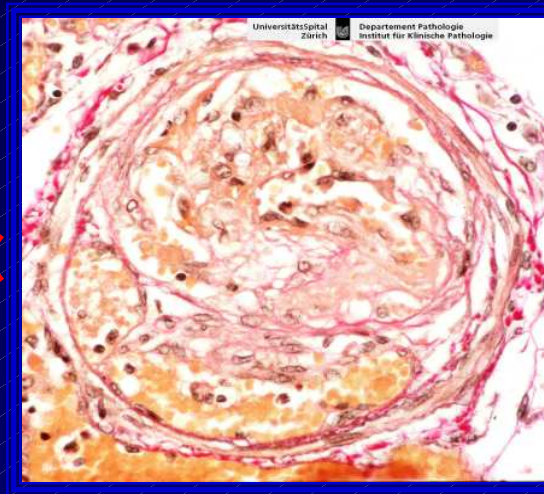
Gouty arthritis

Brain abscess

Renal complications

Hemostasis

At risk for surgery / pregnancy



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Therapies



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graph TD; A[Therapies] --> B[General Therapies]; A --> C[Specific Therapies]; A --> D[Transplantation]; B --> E["• Oxygen<br>• Diuretics<br>• Anticoagulants (?)<br>• (Digoxin)<br>• Effective contraception"]; C --> F["• Calcium Channel Blockers<br>• Prostacyclin Analogues<br>• Endothelin Receptor Antagonists<br>• Phosphodiesterase-5 Inhibitors"];
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General Therapies

Specific Therapies

Transplantation

- Oxygen
- Diuretics
- Anticoagulants (?)
- (Digoxin)
- Effective contraception

- Calcium Channel Blockers
- Prostacyclin Analogues
- Endothelin Receptor Antagonists
- Phosphodiesterase-5 Inhibitors

Therapeutic Approaches (2)

Specific Measures

Vasodilators

- Oxygen
- NO
- Ca-Channel bl.
- Prostacyclines

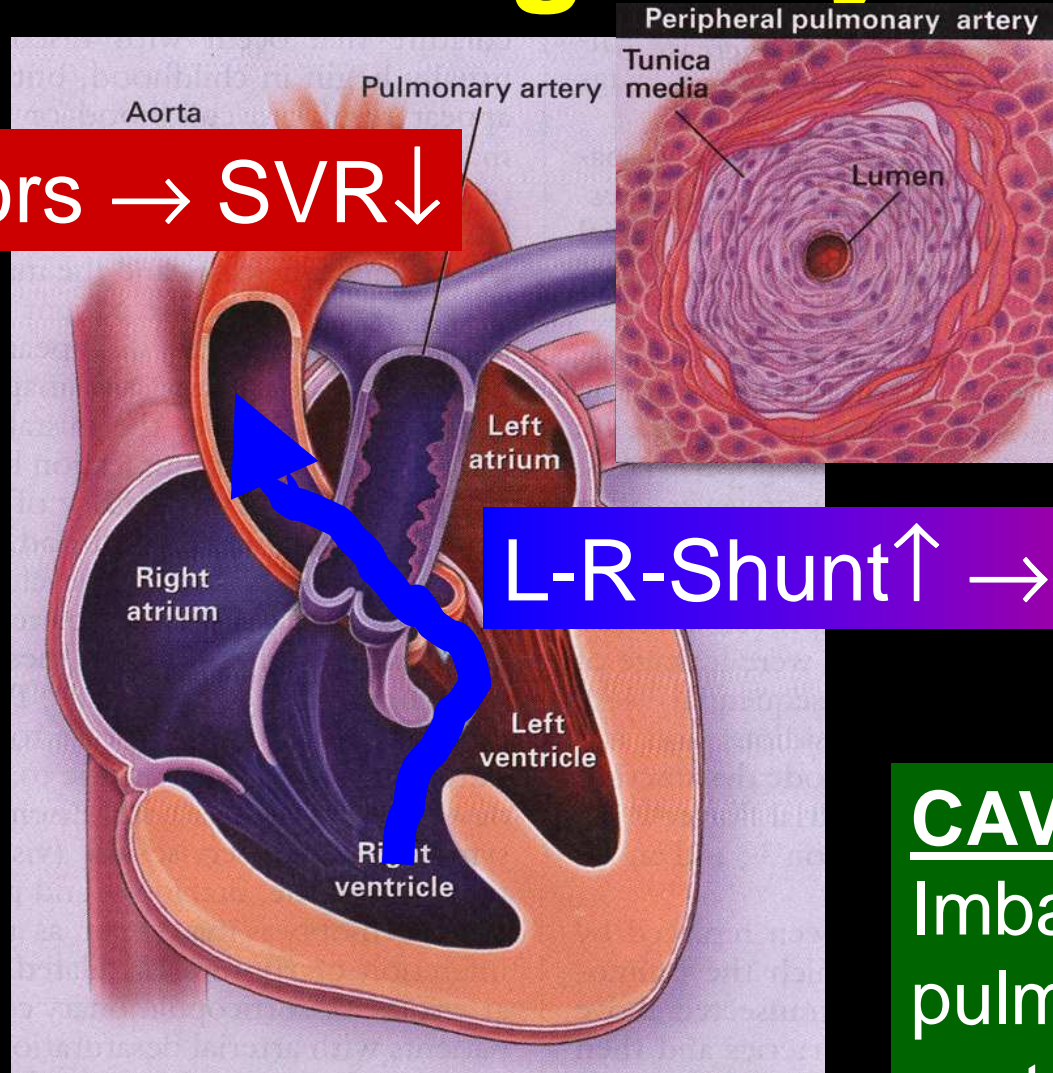
ET-Antagonists

- Bosentan
(Tracleer)

Phosphodiesterase Inhibitor (type 5)

- Sildenafil (Viagra)

Eisenmenger Syndrome



Vasodilators \rightarrow SVR \downarrow

L-R-Shunt \uparrow \rightarrow Cyanosis \uparrow

CAVEAT:
Imbalance between
pulmonary and
systemic vascular
resistance ;



- A multi-center, randomized, placebo-controlled study
- Study population: PHT due to Eisenmenger physiology
- to evaluate the effects of bosentan on oxygen saturation and cardiac hemodynamics

Primary Endpoints



- **Primary Endpoint (Safety Issues!)**

Change from baseline at week 16 in oxygen saturation at rest with room air.

- **Second Primary Endpoint (only if the first hypothesis is verified)**

Change from baseline at week 16 in pulmonary vascular resistance.

Results



- No major side effects
- Most pts felt better
- Improvement in 6 minute walk test
- No drop in SO_2
- Stable blood pressure

Conclusions



- Bosentan:
 - Safe
 - Well tolerated
- No clinical deterioration.....

MANAGEMENT

Hyperviscosity Symptoms

Dehydration

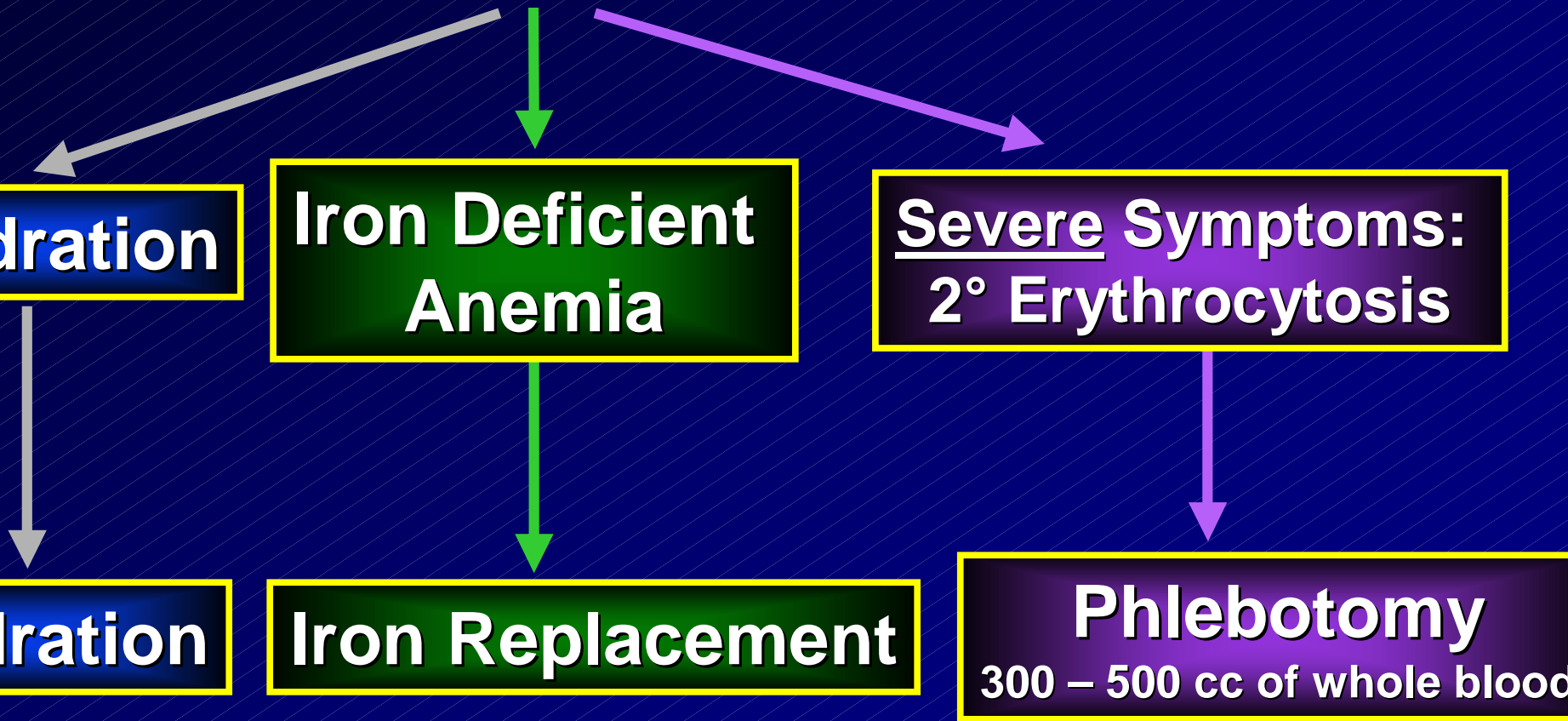
Iron Deficient Anemia

Severe Symptoms:
2° Erythrocytosis

Rehydration

Iron Replacement

Phlebotomy
300 – 500 cc of whole blood



Take Home Message

- Prevention
 - Annual flu shot
 - Pneumovax
- Adequate and rapid therapy of infections
- Change in your medication:
 - Call your congenital cardiologist

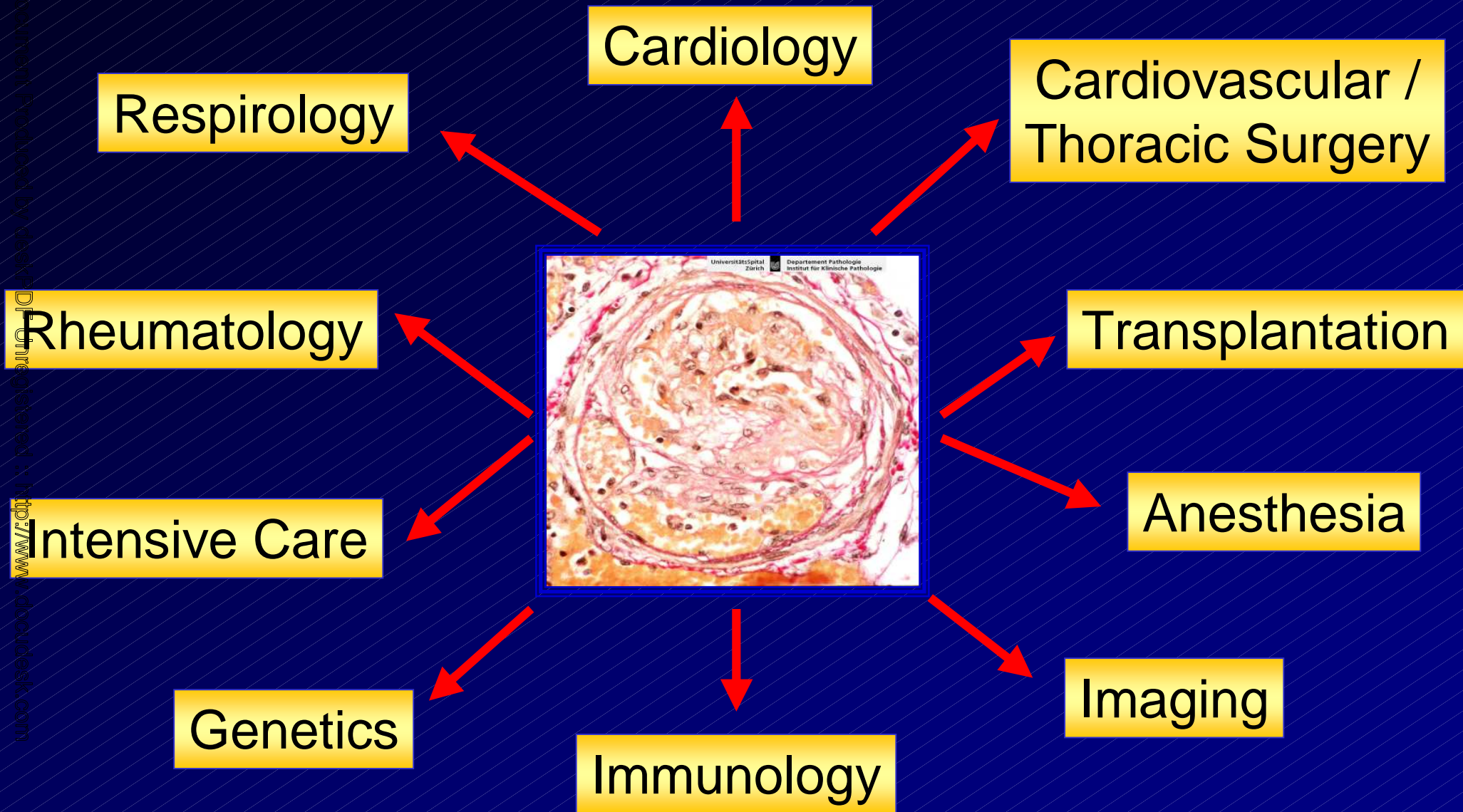
Take Home Message

- Simple surgical procedure is NOT simple - it is a challenge
 - Anesthesist
 - Postoperative period
- Use an air filter in the presence of an iv-line

Take Home Message

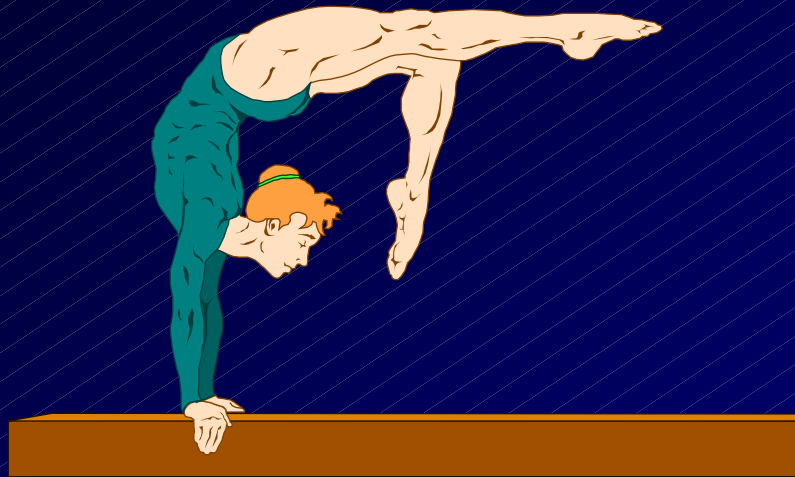
- Anticonceptional Counseling
 - Very high risk for both the mother and the fetus
- Traveling
 - Organize well in advance

Designated Centers for PAH Care



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Pulmonary HT in CHD



The **mainstay**
of care is not to
destabilize the
balanced
physiology!!