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Tetralogy of Fallot

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Agenda

- Preoperative anatomy and physiology
- Surgical approach
- Postoperative management and complications

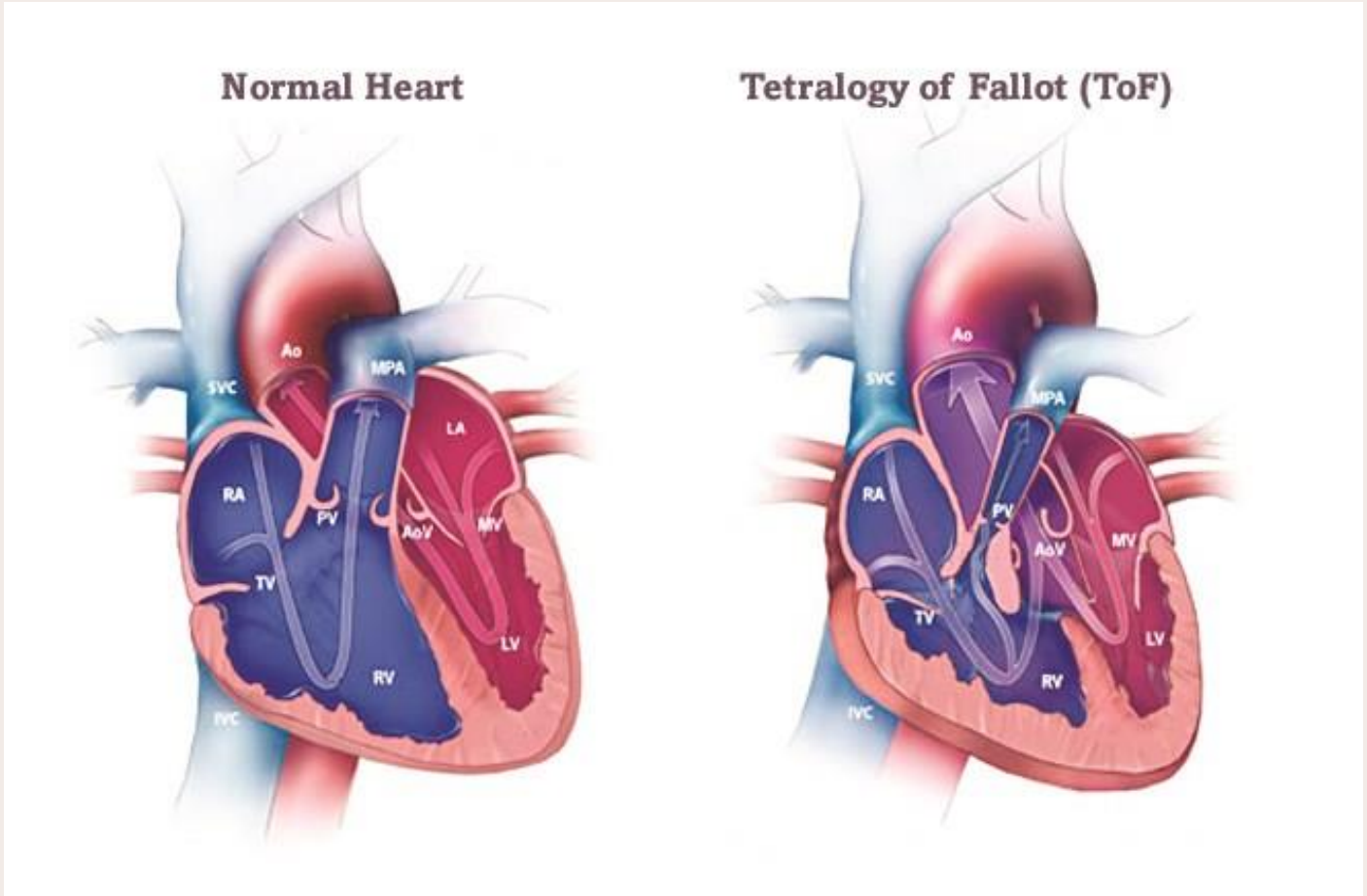
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Preoperative Anatomy and Physiology

Anatomy

- The most common cyanotic congenital anomaly
- Several variants:
  - ToF/PS ***
  - ToF/PA with and without MAPCAs
  - ToF/Absent PV
- Anteroseptal deviation of the outlet septum leads to:
  - A large, anterior malaligned VSD
  - Valvar and subvalvar RVOT obstruction
  - Aortic override
  - RV hypertrophy



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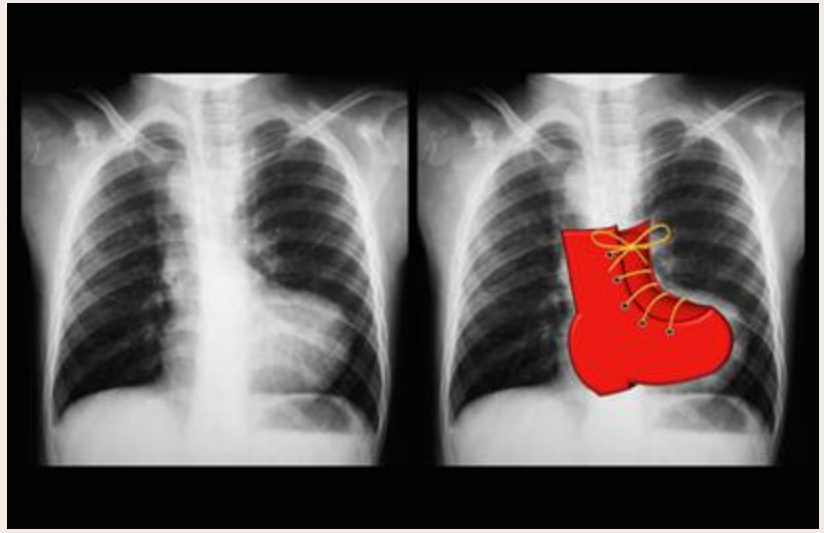
Associated Abnormalities

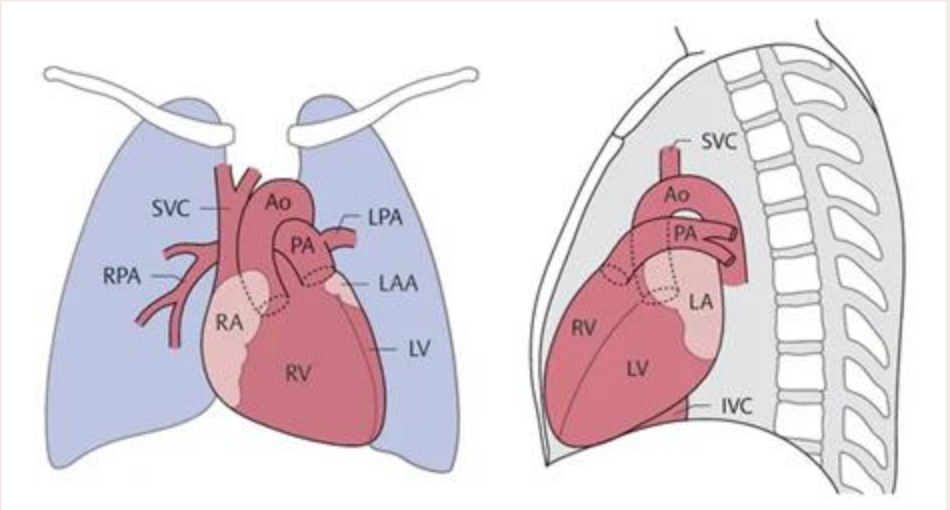
- Coronary anomalies (10-15% of patients with ToF)
  - LAD that arises from the RCA and crosses the RVOT inferiorly
- Right-sided aortic arch (25% of patients)
- TOF with complete AV canal defect
- Genetic syndromes
  - DiGeorge syndrome
  - Trisomy 21
  - VACTERL

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Preoperative Physiology

- Cyanosis
  - Degree of cyanosis is dependent on the amount of RVOTO and pulmonary valve stenosis
  - Progressive subpulmonary obstruction leads to R->L shunting, which then leads to a decrease in systemic oxygen saturation
- High-pitched systolic ejection murmur at the LUSB
- CXR: RVH produces a boot-shape appearance with decreased vascular markings in the lungs 2/2 decreased PBF
- EKG: RA enlargement, RVH





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Pre-operative Physiology

What is a hypercyanotic spell (“tet spell”)?

Acute and sustained episode of profound cyanosis caused by complete or near complete obstruction of pulmonary blood flow with worsening R ->L shunting

- Lack of systolic ejection murmur

Profound cyanosis can lead to acidosis due to poor oxygen delivery

Acidosis can reduce SVR, thereby worsening R->L shunting

Case Scenario

- 4 month old baby boy with ToF/PS admitted from home after “turning blue” when he gets mad. He is admitted in stable condition. Parents are at bedside. However, during your assessment, he starts to get quite fussy. His SpO2 drops from 90% to 55% and he looks quite cyanotic.
- What do you think is happening?
- What can you do?

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Pre-operative Physiology 2

How do we treat a tet spell?

Decrease agitation/sedation with morphine

100% FiO2

Knees to chest, squat

Volume administration

Phenylephrine if available (goal is to increase SVR)

Timing of Repair

Advantages of Early Repair

- Alleviation of cyanotic end-organ damage
- Preservation of myocardial function
- Removal of the stimulus for RV hypertrophy and fibrosis
- Reduction in the incidence of arrhythmias
- Promotion of PA growth and lung development

Repair in Developing Countries

- Higher mortality rate, 6.9-15%
  - Tanzania: 5.9%
  - Ethiopia: 12.9%
  - Pakistan: 1.3%
- Mortality rate of children from developing countries who underwent late TOF repair at a surgical center in France: 3.2%
- Expect improved outcomes over time

Preoperative Considerations

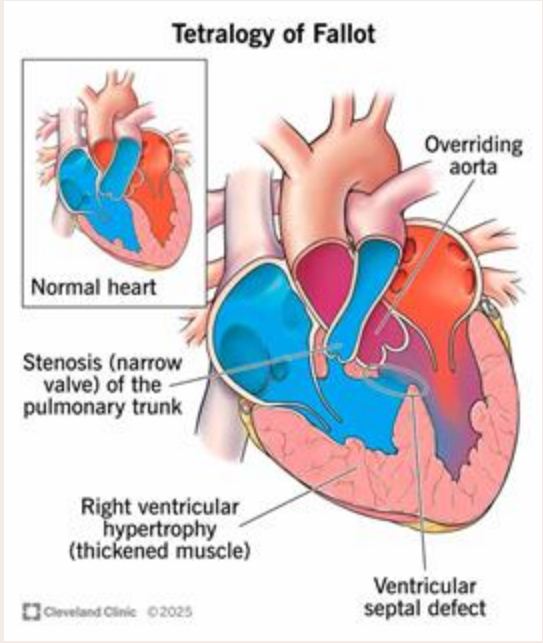
- Chronic hypoxemia and severe cyanosis
  - Median preoperative saturations <85%
  - Cyanotic end-organ damage
- Polycythemia as a response to chronic hypoxemia
  - Hct 52 +/- 11
  - Increases the risk of hyperviscosity and thrombotic events
  - Chronic glomerulopathy with decreased renal perfusion
- Growth retardation and malnutrition
  - Significant risk factor for perioperative morbidity and mortality

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Preoperative Considerations: Myocardial Dysfunction

Right Ventricular Dysfunction

- Advanced hypertrophy of the RV impairs diastolic function
- Inability of the RV to relax and FILL, which indirectly affects LV's ability to FILL and thereby provide adequate CO



Left Ventricular Dysfunction

- Impaired LV response to afterload stress in older patients
- CO= HR x SV
  - SV= EDV-ESV
  - Influenced by preload, afterload, and contractility
  - RV diastolic dysfunction impairs preload to the LV, which, in turn, affects overall CO

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Surgical Approach

Surgical Repair

Goal: Relieve RVOTO while preserving as much contractile infundibular muscle as possible

VSD patch closure via right atriotomy through the tricuspid valve

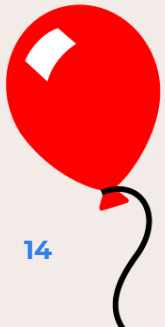
- Ventriculotomy is avoided to prevent substituting functional muscle for a noncontractile patch
- Preservation of the infundibulum -> improved function and better tolerance against PI
- Significant traction is avoided during TV inspection to prevent post op JET
- The bundle of His travels along the posteroinferior edge of the VSD and can be injured during repair

Relief of RVOTO with muscle bundle resection

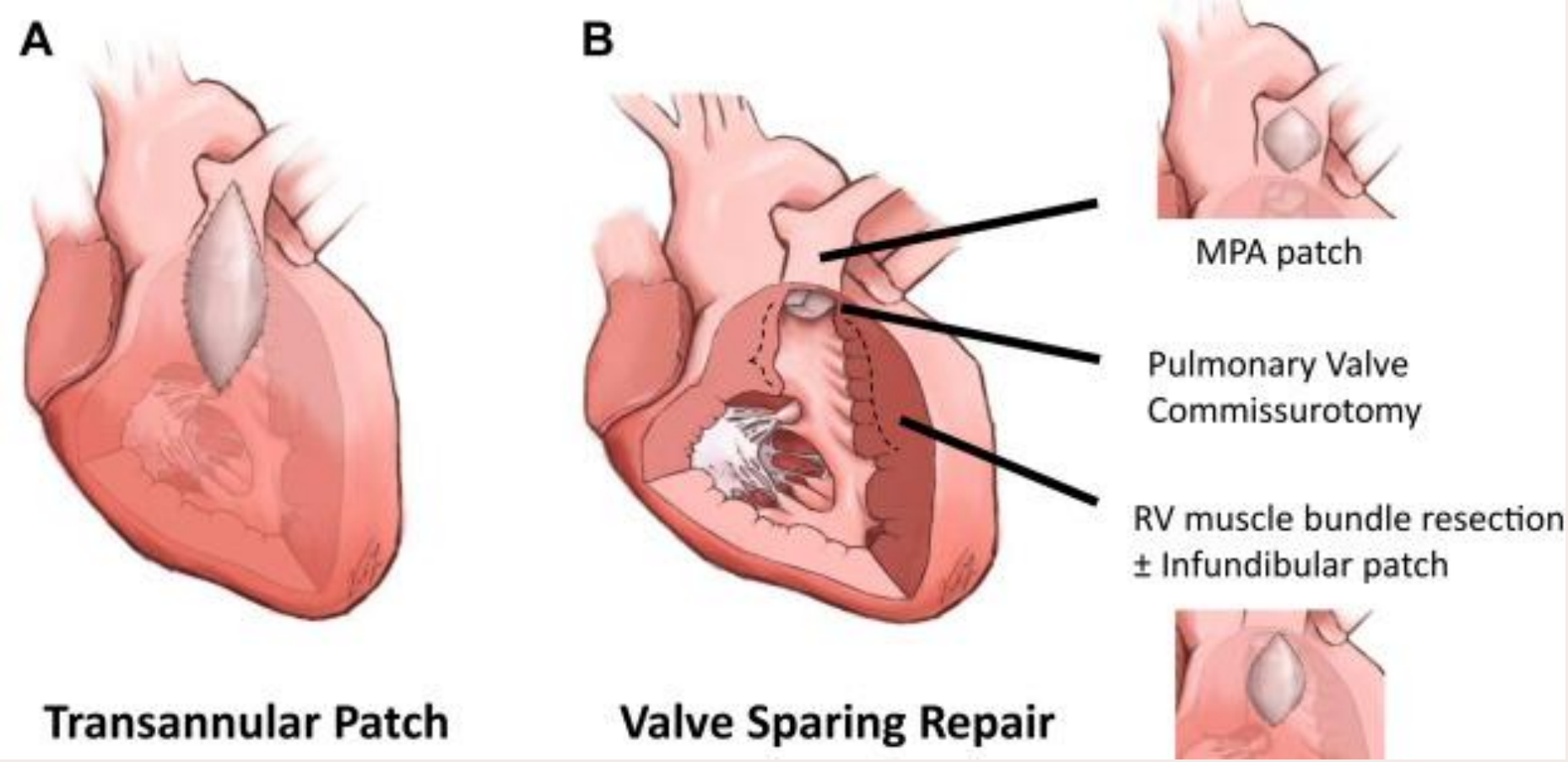
Longitudinal incision on the MPA and inspection of pulmonary valve

Adequate PV annulus = no transannular patch/ “valve-sparing” repair

Hypoplastic PV annulus => transannular incision = transannular patch repair



Surgical Repair



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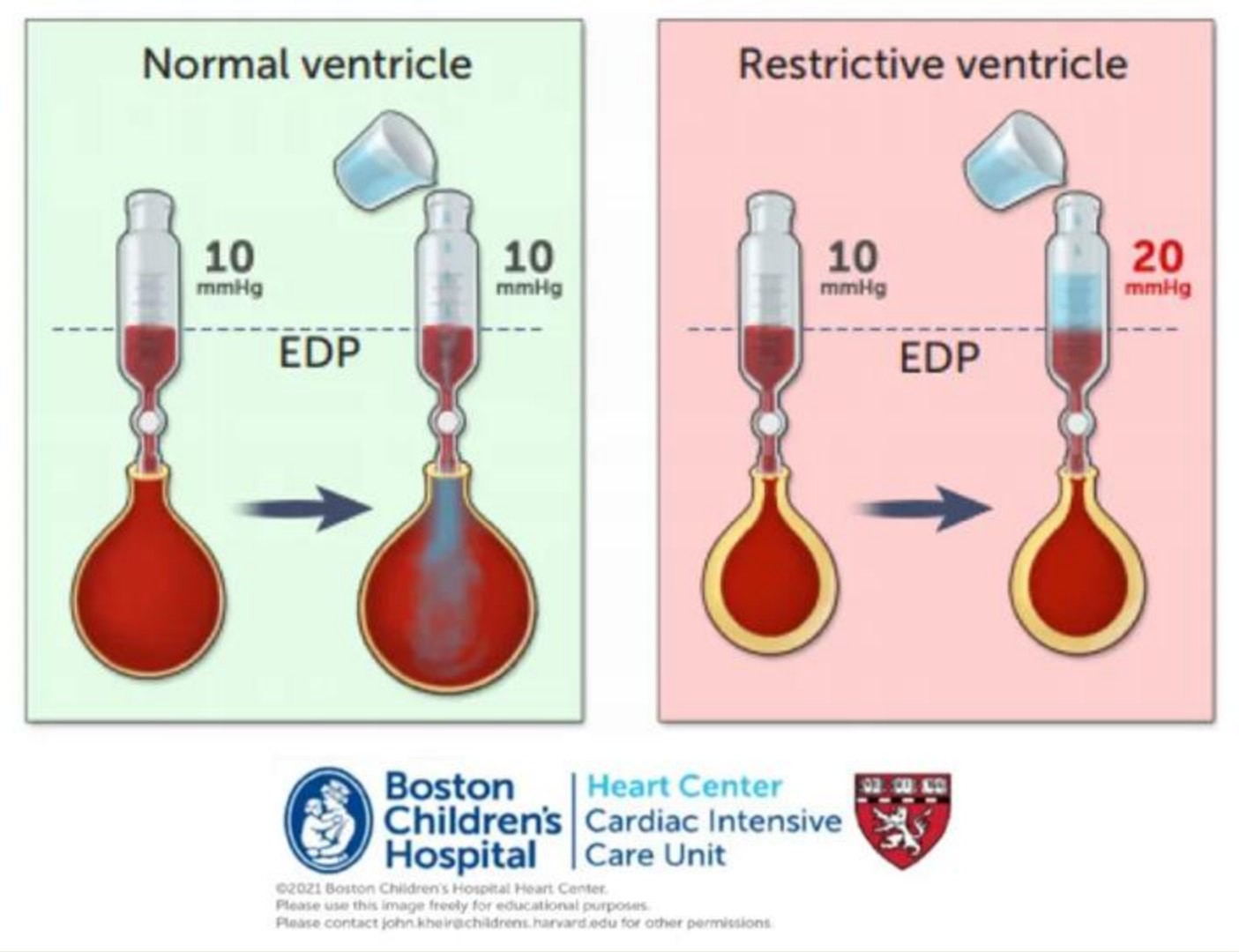
Postoperative Management and Considerations

Postoperative Considerations and Management

- Diastolic RV dysfunction due to RV hypertrophy
  - How does this affect your CVP? Is this an accurate representation of volume status?
  - Maintain adequate preload
  - Consider early extubation
  - Rate control for adequate ventricular filling time
  - Optimize RV lusitropy and afterload
  - Residual RVOTO will exacerbate RV diastolic dysfunction
- Arrhythmias
  - JET (2%)
  - CHB (0.3%)
- Residual VSD

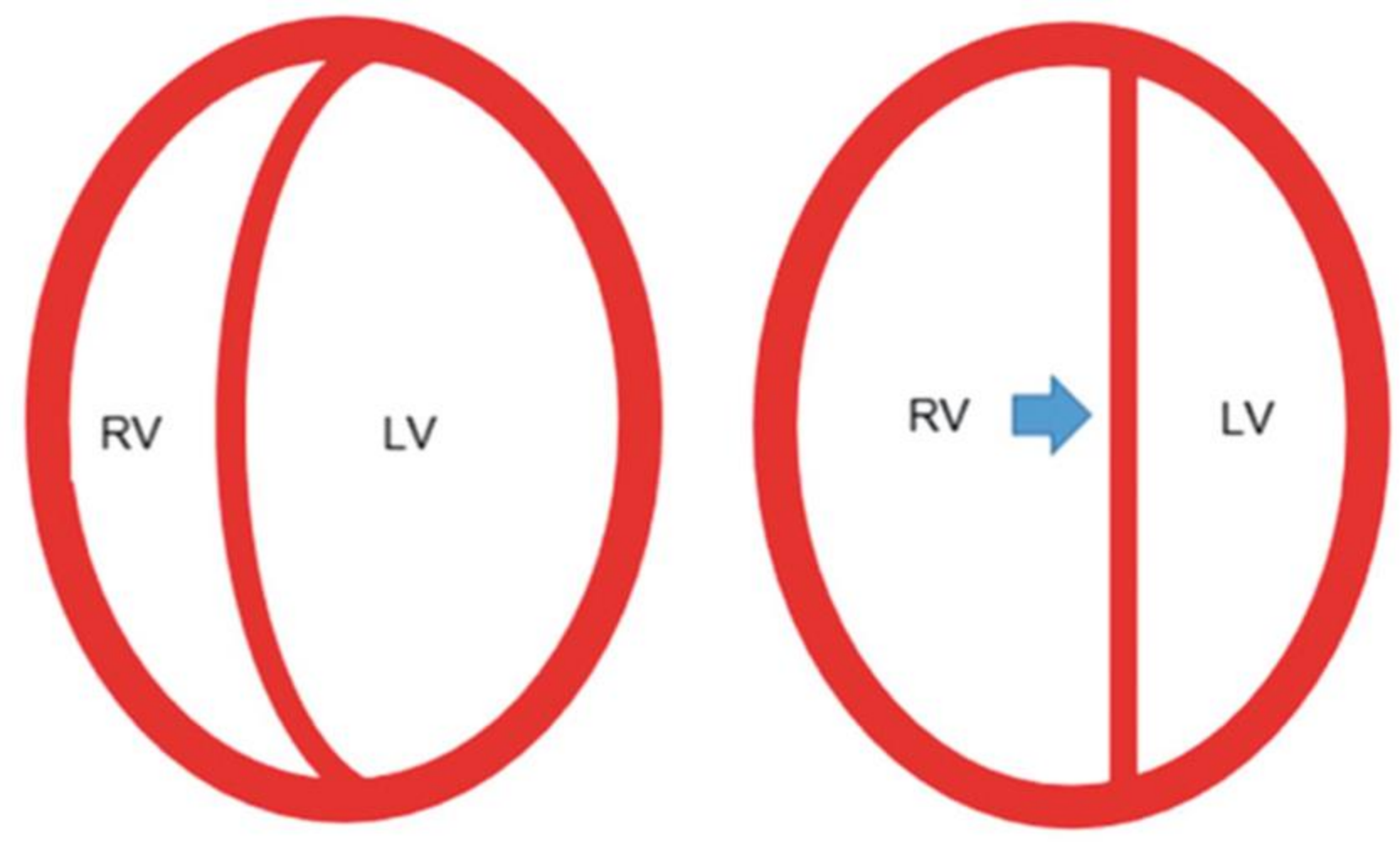
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Restrictive RV Physiology



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Ventricular Interdependence



What might happen to the interventricular septum if the patient is on milrinone?

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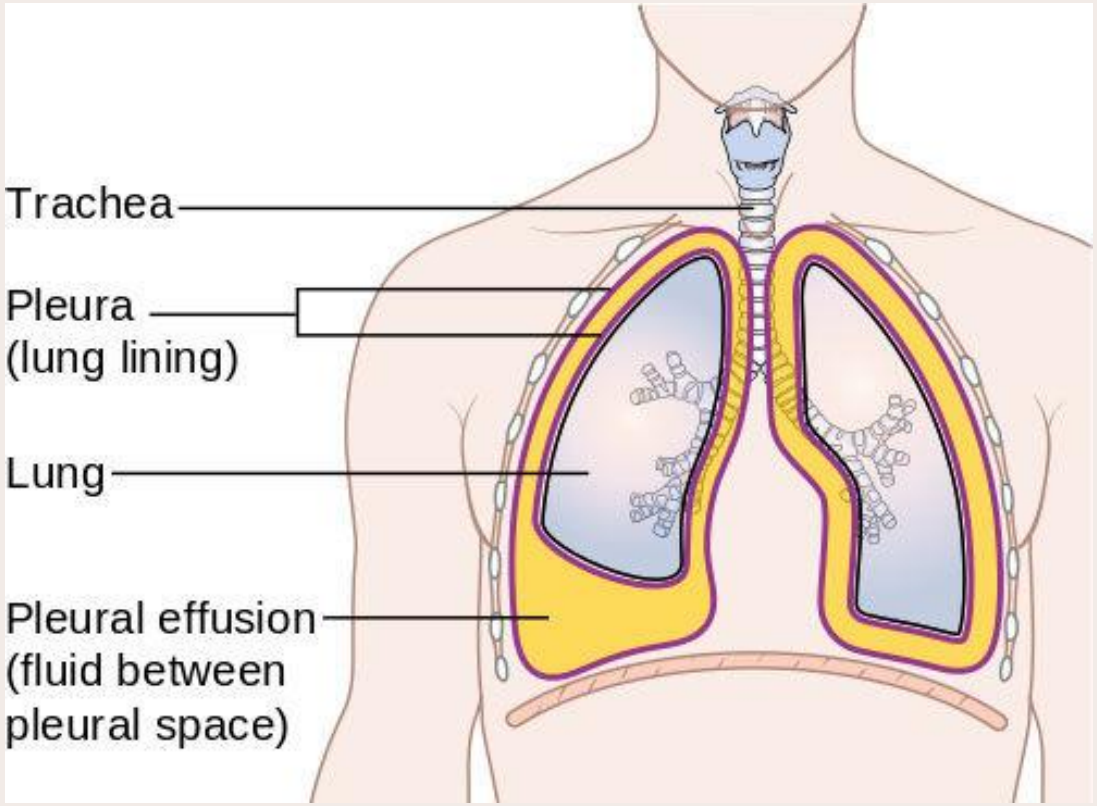
Additional Considerations

- Consider pre-op condition: chronic hypoxia, suboptimal growth, polycythemia (elevated Hgb/Hct)
- Increased risk for bleeding and thrombotic complications
- Longer inotropic and ventilator support
- Increased risk of infection (influenced by nutrition)
- Pleural and pericardial effusions

Pleural and Pericardial Effusions

- Restrictive RV physiology
- Increased central venous pressure
- Increased oncotic pressure
- Prolonged chest drainage

- Pleural effusion: fluid in the space between the lungs and chest wall
- Symptoms: shortness of breath, chest pain, cough, fluid on x-ray
- Management: diuretics, chest drain



Case Scenario Cont..

- 4 month old baby boy with TOF/PS admitted from home after “turning blue” when he gets mad. He is admitted in stable condition. Parents are at bedside. However, during your assessment, he starts to get quite fussy. His SpO2 drops from 90% to 55% and he looks quite cyanotic.
- What do you think this patient is experiencing?

Case Scenario:

- Abbreviated VS and PE:
  - HR 148, SpO2 88% (initially 68-73%)
  - CV: RRR, normal S1, single S2. Harsh 2/6 systolic ejection murmur heard loudest at the left upper sternal border when calm, decreased and almost inaudible when awake and active.

- Do you want any additional information?

Case Scenario continued..

- Echo: severe right ventricular outflow tract obstruction
- Patient desaturates to the 70s when upset. He has no IV access. What do you do?

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Case Scenario:

- Patient desaturated to the 40s with IV insertion
- What would you give this patient without established IV access?
- With IV access?
- What do you anticipate this patient may also need if he requires a lot of sedation?
- When should he go for surgery?

Case Scenario continued...

- Operative procedure:
  - Repair of tetralogy of Fallot with **preservation of the pulmonary valve annulus**, pulmonary valvuloplasty
  - A. **Closure of ventricular septal defect** (bovine pericardium (Cardiocel) via transatrial approach)
  - B. **Right ventricular myectomy**
  - Patch arterioplasty of the main pulmonary artery thin pulmonary homograft
  - No intervention on the PFO
  - Pulmonary arterioplasty: left pulmonary (pulmonary homograft)
  - Ligation and division of patent ductus arteriosus

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Case Scenario cont...

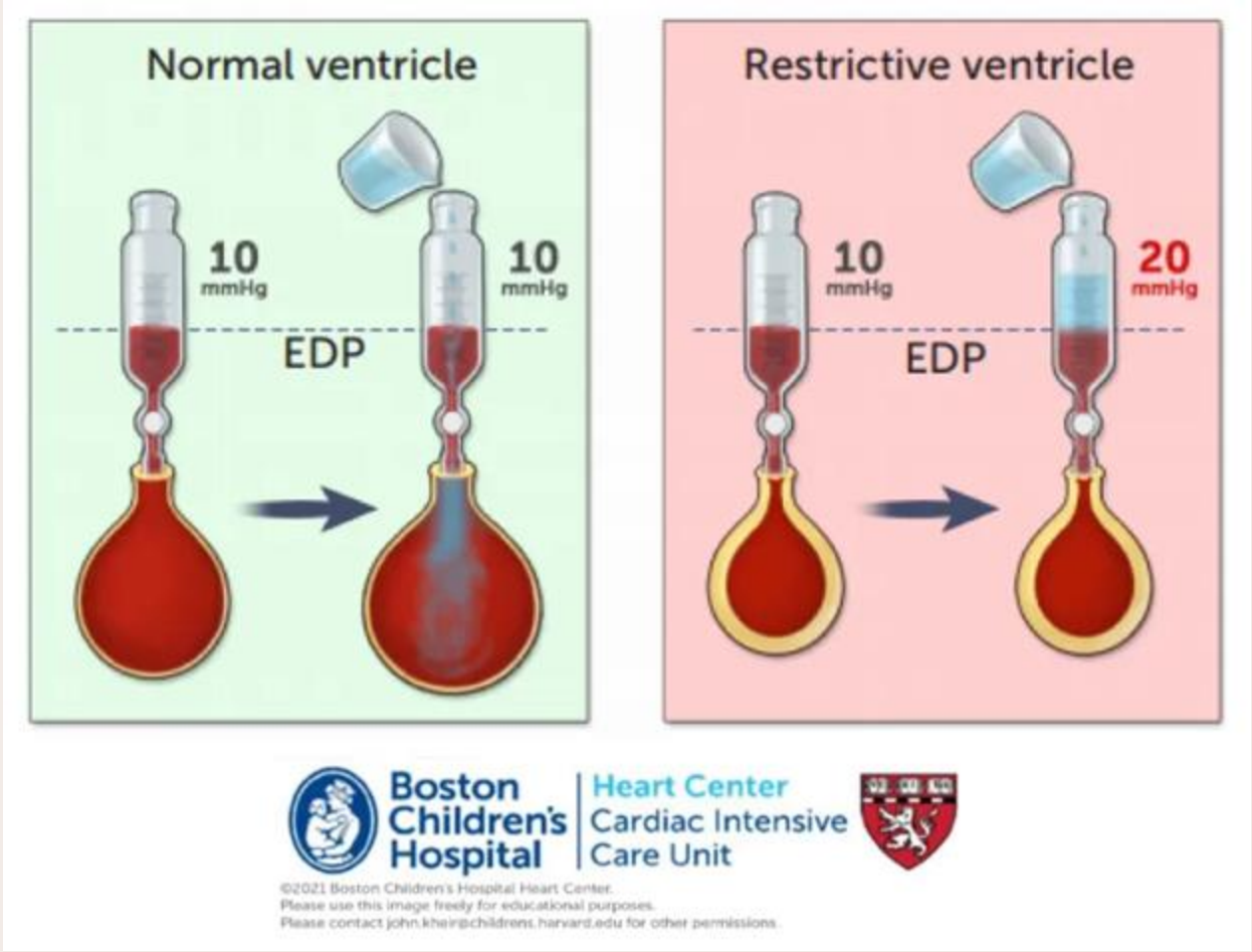
- VS: HR 140s, BP 70s/50s, CVP 15, RA 10, SpO2 92% on 60% FiO2
- PE: WWP, CRT 3 seconds
- Adrenaline 0.03, Dexmedetomadine, Fentanyl

Within the hour, patient's exam has changed. He is now pale, CRT 4-5 seconds, CVP 8, HR150s, BP 60s/40s, and SpO2 88% on 100% FiO2

- What do you think is happening?
- What other information would you like?

Case Scenario:

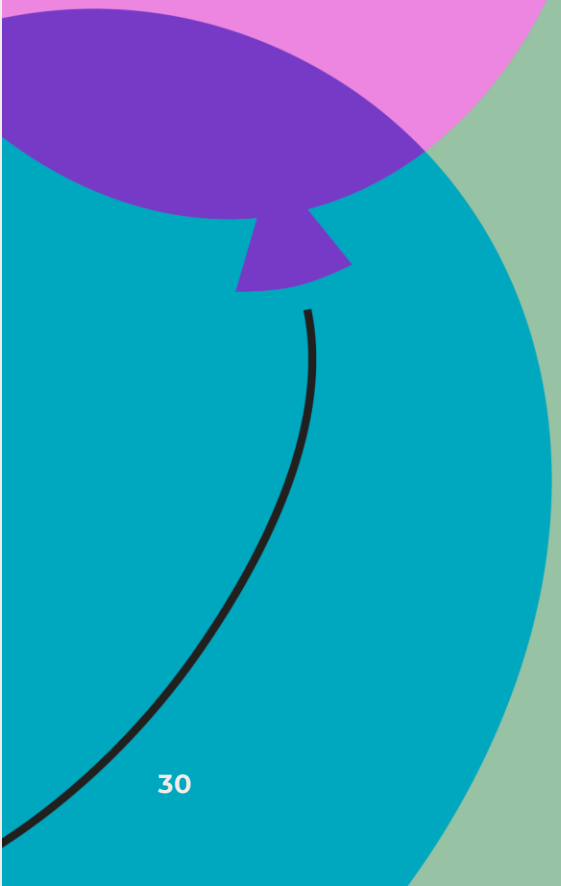
- Where will you give additional volume?
- As you give volume, the CVP trends up from 8 to 15, and the patient's BP improves.
- A few hours later, patient is hypotensive again. The provider asks you to give volume, and the CVP goes up to 18, but the patient is becoming hypotensive. Should you continue to give volume? What do you think is happening?



Case Scenario Final

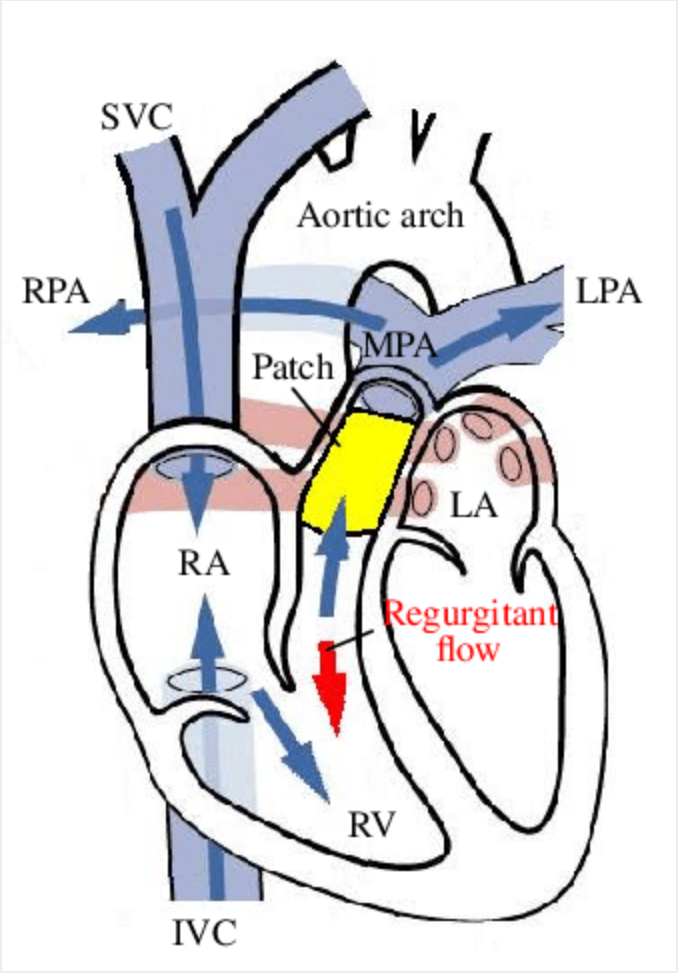
- Throughout the evening, patient continued to require volume resuscitation (CVP ~18) and high vasoactive support
- Bleeding improved
- POD 2: Able to wean vasoactives and start diuresis
- POD 8: Extubated
- POD 15: Transfer to ACCU
- POD 19: Discharged home

Long-term Considerations



Long term Complications

- Pulmonary regurgitation
- RV dilation and dysfunction
- Arrhythmias
- HF
- Need for reintervention (PV replacement)
- Residual or recurrent RVOTO
- Branch PA stenosis
- Residual VSD
- Sudden cardiac death
  - Worsening PR-> RV volume overload -> RV dilation and dysfunction -> Arrhythmias (A flutter and V fib most common) -> sudden cardiac death



Take Home Points

- Tetralogy of Fallot: 4 components all caused by anterior deviation of the conal septum
  - Subvalvar and commonly valvar PS or PA
  - Anterior malaligned VSD
  - Overriding aorta
  - RV hypertrophy
- The clinical course of ToF/PS depends on the degree of subvalvar muscular obstruction, PV hypoplasia, and concomitant RVOTO -> degree of cyanosis
- Anticipate RV diastolic dysfunction and restrictive RV physiology -> maintain adequate preload

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Questions?

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Thank you!!!