CARDIOLOGY 2023

PULMONARY HYPERTENSION

A Case Review: PH in the setting of
D-transposition of the great arteries s/p repair

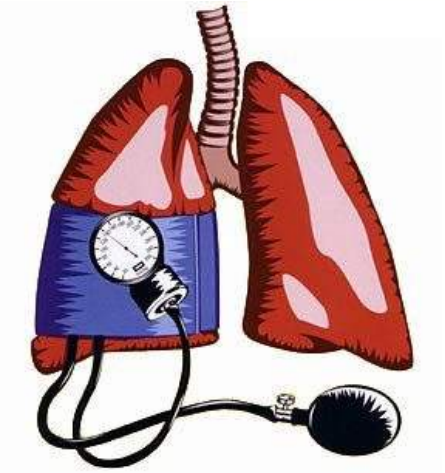
February 25, 2023

OBJECTIVES

- Share
- Review
- Reinforce

THE BASICS

- How is pulmonary hypertension defined?
- What is the etiology for pulmonary hypertension?



Pulmonary arterial hypertension (PAH) is a group of diseases characterized by a progressive increase in pulmonary vascular resistance, leading to right ventricular failure and premature death.

Simonneau G., Galie N, Rubin LJ et al. Clinical classification of pulmonary hypertension. Journal of American College of Cardiology 2004; 43 (12 Suppl S): 5S-12S.

PULMONARY HYPERTENSION ETIOLOGIES

Our focus at Cardiology 2023

1. Pulmonary arterial hypertension

- Idiopathic
- Heritable
  - BMPR2
  - ALK1, Endoglin
  - Unknown
- Drug and toxin-induced
- Associated with PAH
  - Connective tissue diseases
  - HIV infection
  - Portal hypertension
  - Congenital heart disease
  - Schistosomiasis
  - Chronic hemolytic anemia
- 1′. Pulmonary Veno-occlusive disease and/or capillary hemangiomatosis
- 1′′. Persistent pulmonary hypertension of the newborn (PPHN)

2. Pulmonary hypertension due to left heart disease

- Left ventricular systolic dysfunction
- Left ventricular diastolic dysfunction
- Valvular disease
- Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathy

4. Chronic thromboembolic pulmonary hypertension (CTEPH)

- Complication of pulmonary embolism
- Large or proximal vessel disorders
- High blood pressure, blood clots
- Reduced compliance and luminal narrowing

3. Pulmonary hypertension due to lung diseases and/or hypoxia

- Chronic obstructive pulmonary disease
- Interstitial lung disease
- Pulmonary diseases with restrictive and obstructive pattern
- Sleep-disordered breathing
- Alveolar hypoventilation disorders
- Chronic exposure to high altitude
- Developmental lung diseases

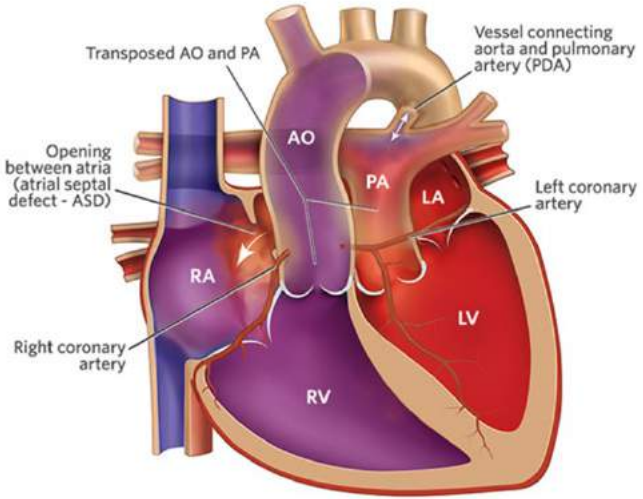
5. Pulmonary hypertension with unclear and/or multi-factorial mechanisms

- Hematologic disorders (myeloproliferative disorders, splenectomy)
- Systemic disorders (Vasculitis sarcoidosis, pulmonary Langerhans cell, histiocytosis LAM, neurofibromatosis)
- Metabolic disorders (Glycogen storage disease, thyroid disorders)
- Congenital heart disease
- Cancer-related, renal failure on dialysis

OUR PATIENT

Prenatally diagnosed with

transposition of the great arteries with an intact ventricular septum.



WHY THE DEVELOPMENT OF PH?

- Transposition of the great arteries (TGA) is reportedly associated with persistent pulmonary hypertension of the newborn (PPHN) in 1-3% of cases
- Not a definitive explanation associating TGA with PPHN
- However, consensus is with TGA there may be a sustained period of elevated pulmonary vascular resistance in utero

Hoshino, S. Somura, J. Furukawa. O. Yanagi, T. Maruo, Y. *The histological findings in transposition of the great artery with severe persistent pulmonary hypertension.* Journal of Cardiology Cases. 17 (2018) 156-162

DAY 0 OF LIFE

- Delivered and intubated
- Stabilized in NICU at OSH
- Transferred to CHOA for surgical management

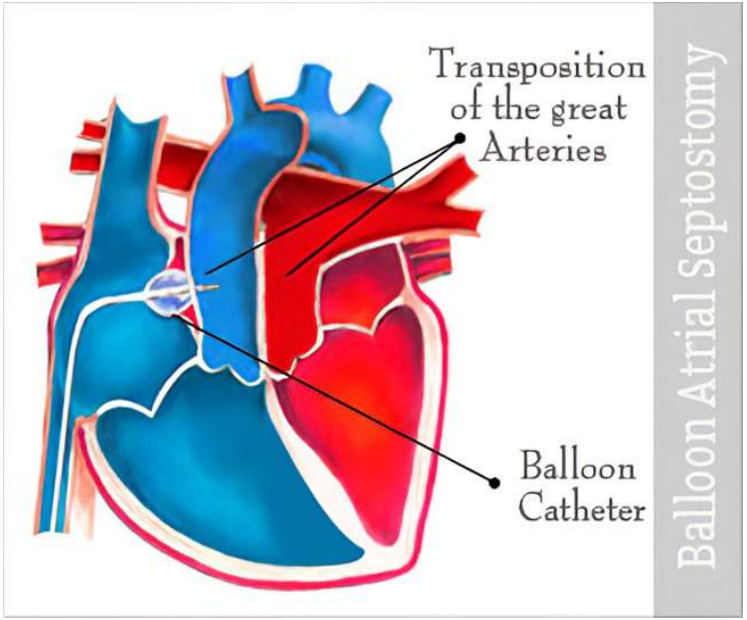
DAY 0 OF LIFE

- Due to profound and persistent hypoxemia emergently traveled to cath lab

Picture shared with permission from CHOA Cath Lab Team

DAY 0 OF LIFE: CATH

- Balloon atrial septostomy performed
- Post procedure: unrestrictive atrial level shunt
- Significant improvement in saturations
- Returned to CICU on **iNO 20 ppm**

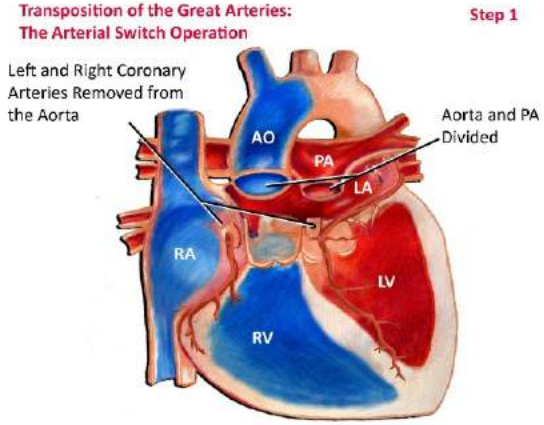


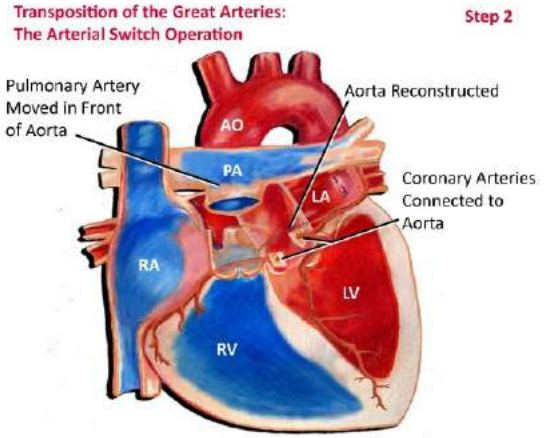
DAYS 1-3 OF LIFE: PRE-SURGERY

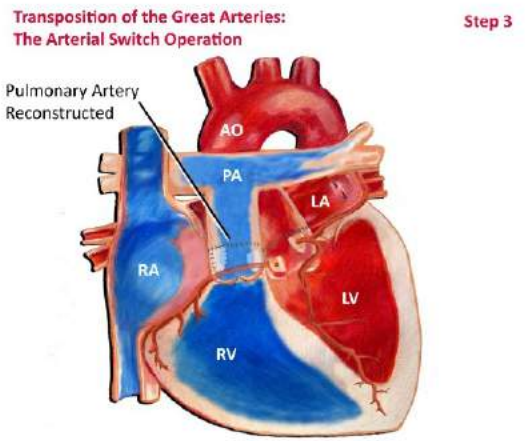
- Serial echoes performed
- Remained on **iNO 20 ppm**
- Added **milrinone and epinephrine**

There was concern for pulmonary hypertension in the pre-operative period.

DAY 4 OF LIFE: SURGICAL REPAIR

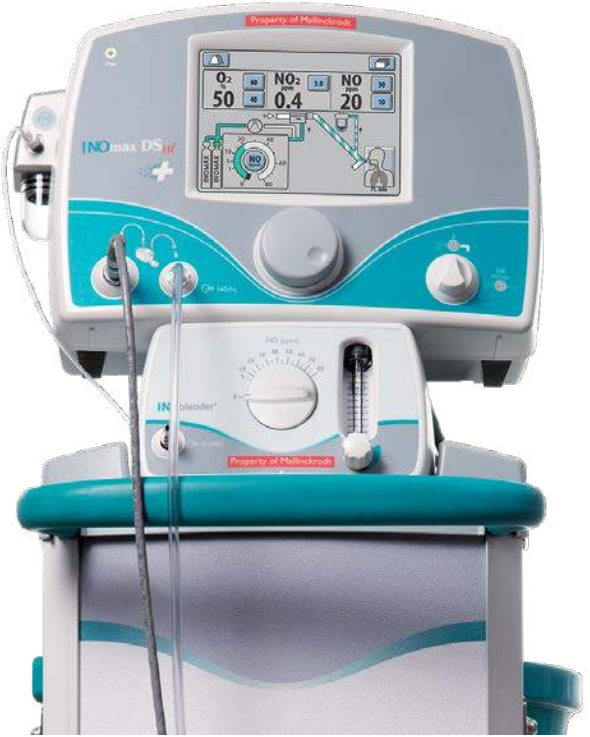






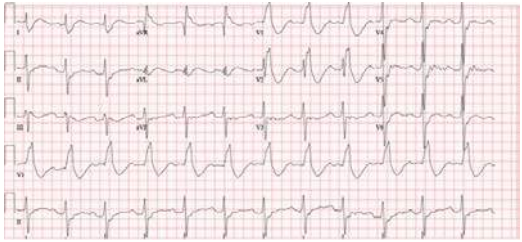
DAY 4 OF LIFE: POST OP PLAN

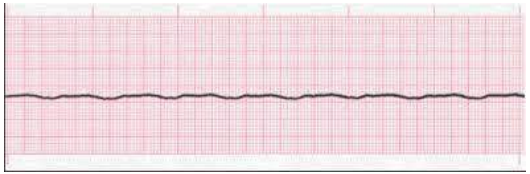
- Continue iNO at 20 ppm until chest closure
- Monitor fluid balance
- Wean ventilatory support as able



DAY 5 OF LIFE: CARDIAC ARREST

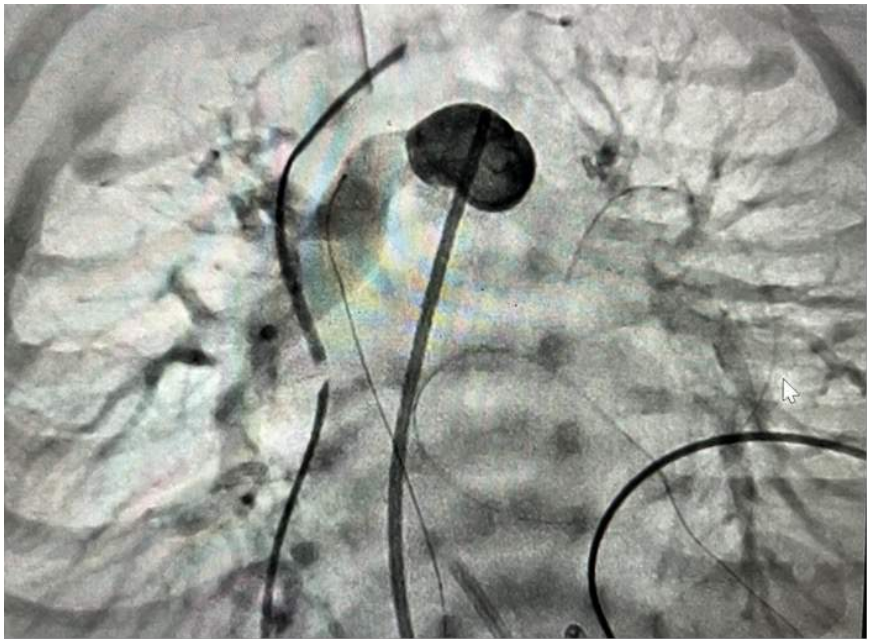
- Precipitating event - agitation with repositioning
- Widening of the QRS on EKG
- Developed asystole
- Code called





DAY 5 OF LIFE: RETURNED TO CATH LAB

- Cath confirmed
  - ✓Good repair
  - ✓normal function
  - ✓open coronaries
  - × 120% systemic RV pressure



PH CATH CRITERIA

Measures	Normal Parameters	Our Patient
mPAp	<20 mmHg	49-55
wedge	<15 mmHg	13-16
PVR	<2 wu	13.9

$$PVR = \frac{mean\ PA\ pressure\ -\ LA\ pressure}{pulmonary\ blood\ flow}$$

Sildenafil started following cath

DAY 6 OF LIFE: PH TEAM CONSULTED

Provide recommendations to lower pulmonary vascular resistance.

DAY 6 - DAY 45 OF LIFE: PH TEAM CLOSELY INVOLVED

Day 6 of life- **SQ treprostinil started**

Day 19 of life- **Bosentan started**

Day 31 of life - **weaned off iNO**

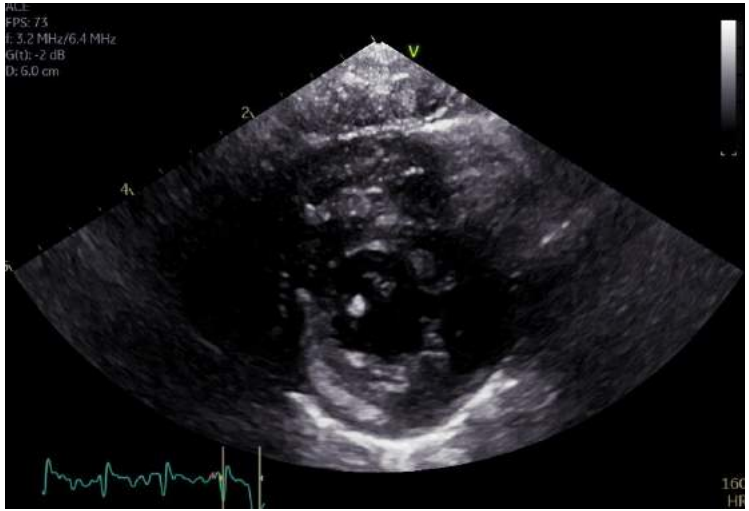
Day 35 of life - **whole exome sequence sent**

Picture shared with permission from CHOA PH Team Members

DAY 46 OF LIFE: DISCHARGE

- Discharged home on 1/4L O2, triple PH therapy and NG feeds

- Echo prior to DC; mild to moderate PH, TR ~42 mmHg, normal RV function



THE NEXT 365 DAYS: OUTPATIENT SETTING

- **Month #1** - transitioned from **sildenafil** to **tadalafil**
- **Month #2** - **Began SQ down titration**
- **Month #3** - OPMS; two episodes of penetration without aspiration
- **Month #4** - discontinued **SQ Treprostinil**

THE NEXT 365 DAYS: OUTPATIENT SETTING

- **Month #4** - adjusted diuretics
- **Month #5** - **transitioned from bosentan to ambrisentan**
- **Month #6** - OR; cleft repair and g-tube placement
- **Month #7** - OPMS; ongoing dysphagia without aspiration
- **Month #8** - **discontinued ambrisentan**
- **Month #11**- OPMS; normal study
- **Month #12** - **discontinued tadalafil**

Picture shared with permission from patient's parents

WHY THIS CASE WAS SELECTED?

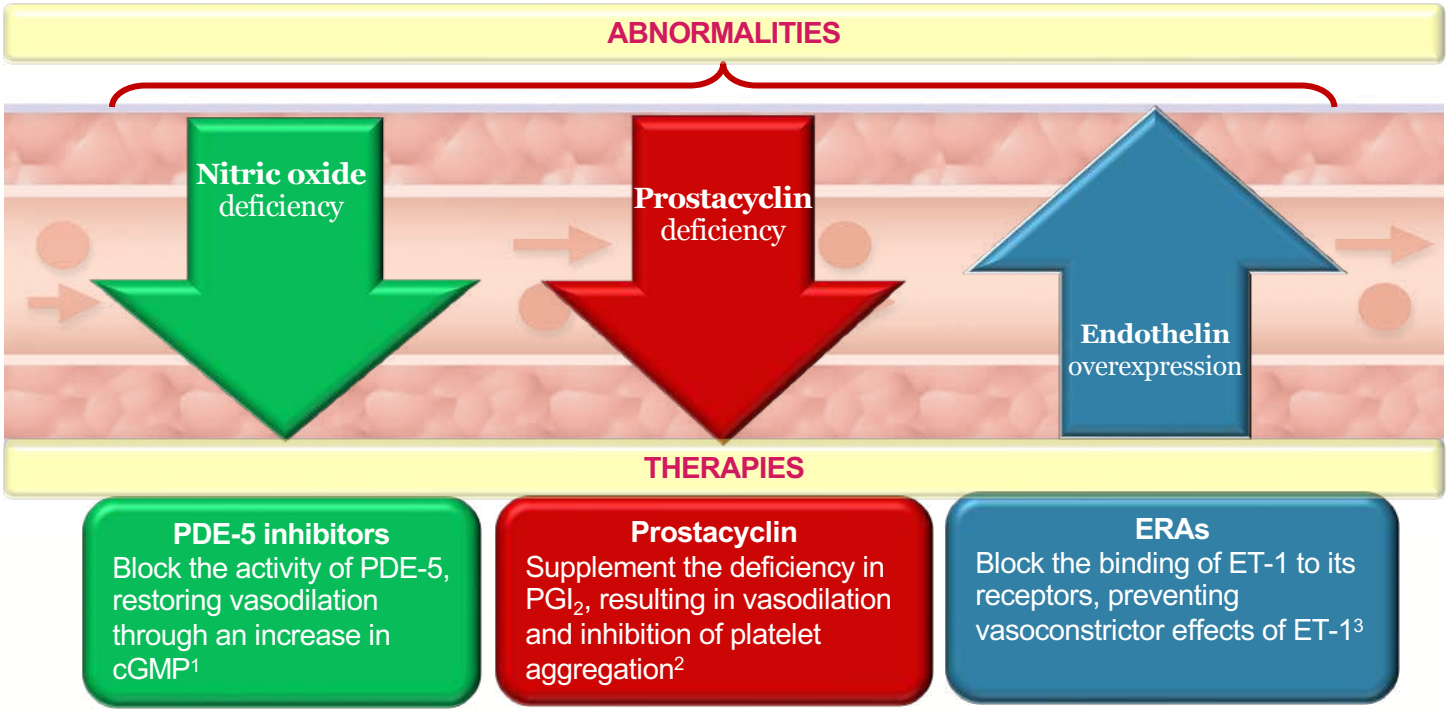
Severe Pulmonary Hypertension

Pulmonary Hypertension Resolution

REASONS FOR SUCCESS

- Early and aggressive PH Management
- Solid collaboration
- Ongoing follow up

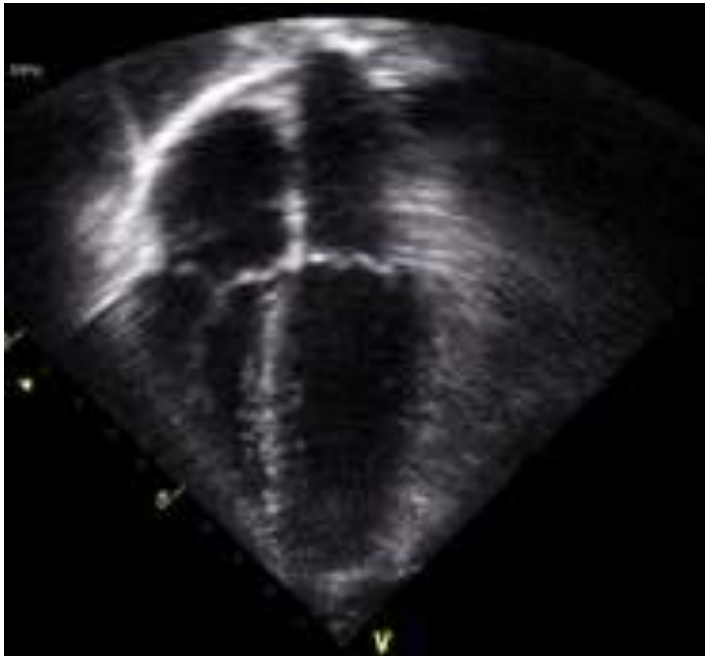
TREATMENT WAS AIMED AT INVOLVED PATHWAYS



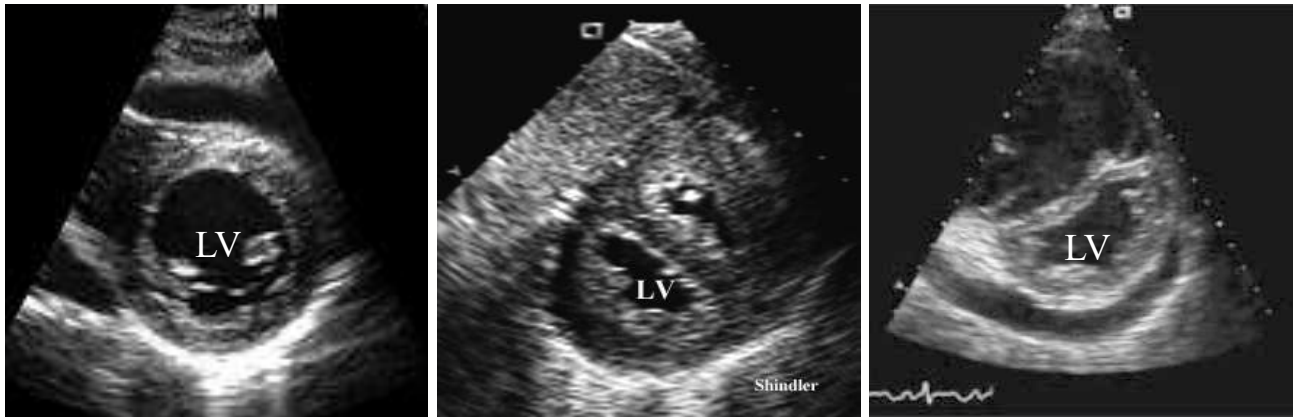
SERIAL ECHO ASSESSMENT

- RV size
- PA doppler pattern
- Septal configuration
- Biventricular function
- Directionality of shunt

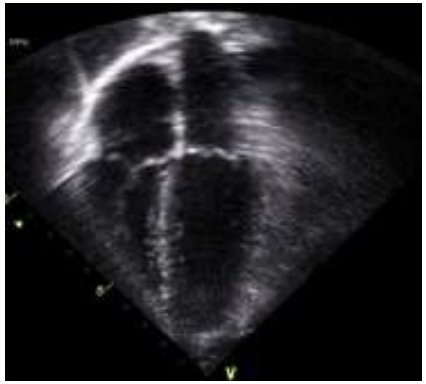
Normal 4-chamber view

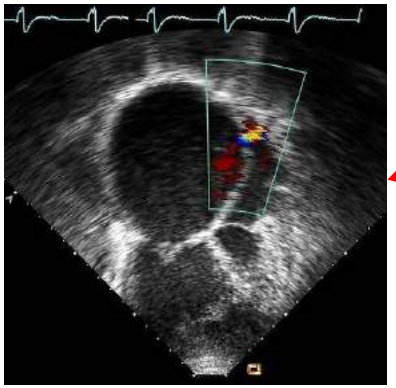


A CLOSER ECHO LOOK



NORMAL





SEVERE PH

CARDIOLOGY 2023

Children's Healthcare of Atlanta

From Echocardiography Journal: http://www2.umdnj.edu/~shindler/echo.html.

PROVIDER CONSIDERATIONS

- With all PH therapies need to fine the balance between benefits of therapy and side effects
- Remember pharmacokinetic interactions between PH therapies
- Establish realistic timeline for medication effectiveness

PROVIDER CONSIDERATIONS

- Determine optimal delivery route
- Frequently reassess optimal imaging
- **From the start, have transparent dialogue with patient, family and colleagues**

NURSING/ CASE MANAGEMENT CONSIDERATIONS

- **Advocate** for your patients and their families
- **Investigate** insurance plan well before discharge
- **Incorporate** patient and family education on a routine basis

CONCLUSIONS

- Pulmonary hypertension is a serious life threating medical condition
- Pulmonary hypertension left untreated results in high vascular resistance which places strain on the right ventricle
- The goal of pulmonary hypertension medication is to relax the vascular bed and lower pulmonary vascular resistance

Picture shared with permission from patient's parents