Overview

- Evaluation of new patient with PAH of unknown etiology
- Case studies
- Coordinating care

History – Key Questions

- Dyspnea with exertion
  - When does this occur? What activities cause symptomatology? If on RAD meds, do they work?
- Limited exercise tolerance
  - Can the child keep up with peers?
  - Worsening progression? Stroller use?
- Dizziness, chest pain, edema, cyanosis
- History of pre-syncope or syncope
- History of seizures

Making the diagnosis...

At initial presentation -
- Previously healthy child walks into PMD’s clinic presenting with symptoms of:
  - Dyspnea with exertion
  - Limited or worsening exercise tolerance

Initial Evaluation of the Child with Pulmonary Arterial Hypertension – A Nurse's Perspective

Michelle Ogawa, CPNP
**History**

- **PMH:**
  - Heart murmur
  - Arthralgia, arthritis, rash
  - Lung disease
  - Liver disease
  - Deep venous thrombosis or pulmonary embolism
  - Sleep apnea
  - Viruses (HIV risk factors, Hepatitis, EBV)
- Family history - as listed above, sudden death, FPAH

**Physical Exam Findings**

**Cardiac:**
- Loud P2
- Holosystolic murmur (tricuspid regurgitation)
- Diastolic murmur (pulmonary insufficiency)
- Right ventricular lift

**Other possible findings:**
- Edema (periorbital/foot, ascites)
- Hepatomegaly
- Cyanosis, clubbing - Eisenmenger’s

**Diagnostic Studies**

- Algorithms for comprehensive evaluation
- Modifications needed for each pediatric patient
- Anesthesia requirements may dictate when and what order some studies are completed
- Fix the “fixable” first before treating PAH

**Preliminary Studies**

- 12-lead ECG
  - Right axis deviation
  - RV hypertrophy with T-wave changes
  - Right atrial enlargement
- Chest x-ray
  - Enlarged RA and RV, dilated MPA
  - Lung parenchymal disease may be present
- Pulse oximeter
  - Oxygen desaturation possible if right-to-left shunt
### Echocardiogram
- Evaluate –
  - Cardiac structure – R/O CHD or left-sided disease
  - RV size and function
  - Estimated RV systolic pressure based on TR Jet
    - Interventricular septal flattening or bowing into LV (indirect RV pressure measurement)
  - Right atrial size
  - IVC size
  - LV size and function
- Potential for bubble study

### Ventilation/Perfusion (V/Q) Scan
- Evaluate for clots to exclude PE or CTEPH
- Age dependent – may only get Q scan

### Pulmonary Function Studies
- 6 minute walk test
- Comprehensive PFTs
- Cardiopulmonary exercise test
- Polysomnography

### High Resolution CT Scan/CT Angiography
- Evaluate lung parenchyma and presence of clots and vascular abnormalities
- Evaluate hepatic veins (abdominal CT or U/S)
Cardiac Catheterization

- Gold standard for PH diagnosis
- Right and left heart cath
  - Saturations (r/o systemic-to-pulm shunts)
  - Pressures
  - Hemodynamics – CI, PVRI, Rp:Rs
  - Vasodilator response testing
    - 100% O2, iNO 20, 40, or 80ppm OR Epoprostenol
    - Positive response for adult PAH pts:
      - mPAP decreases ≥10mmHg to pressure of ≤40mmHg with normal cardiac output

Cardiac MRI

- Evaluate for RV size, function, and output
- Completion at time of initial evaluation is age dependent

Serologic Lab Studies

- Clotting factors
- Immunologic studies
- Rheumatologic studies
- Viral Screen
- Thyroid function
- BNP / NT-proBNP
- Bone morphogenetic protein receptor (BMPR2)

Case Study #1

- 15yo male diagnosed with RAD x 3 years. Treated with Albuterol, Pulmicort and occasional steroids – limited and temporary improvement. Receiving weekly allergy shots.
- Living at altitude of 5,000 ft (1,524m)
- Previously active, taking swim lessons, but stopped at 14yo due to worsening stamina
- Now constantly fatigued, limited to walking 100yds before getting SOB, worsening chest pain, and cyanotic
- History of pre-syncope but no syncope
History

- PMH: Hospitalized as toddler for URI. No surgeries.
- ROS: RAD, seasonal allergies, otherwise negative.

Case Study #1

- Pediatrician’s findings:
  - Cardiac exam – RRR, Split S1, Cap refill<2sec
  - $O_2$ saturation 70-90% varying with activity
  - 12-lead ECG which showed RVH
  - Recent CXR was read as normal
- Differential diagnosis: CHD or PH
- Plan: Initiate supplemental $O_2$ therapy
  - Referral to local cardiologist for further work up

Cardiac Evaluation

- Echo: RV-estimated systolic pressure 148mmHg + RAP
  - (mild TR, systemic BP 118/81)
  - Severely hypertrophied
  - Moderately dilated
  - Moderately reduced systolic function
  - RA – mildly dilated
  - PI – mild to moderate
  - LV – Mildly reduced systolic function

Local cardiologist refers patient to our center…

Physical Exam

- Vital signs: HR 80, RR 28, BP 116/69, $O_2$ sat 96% on RA
- General: Thin, well-developed male in no distress.
- Respiratory: Breath sounds clear. No wheezes, rales, rhonchi.
- CV: RRR. Normal S1. S2 splits and closes physiologically. **Loud P2.** No murmurs, rubs, gallops.
- Abdomen: Soft, nondistended. No HSM.
- Extremities: Warm, well-perfused. Cap refill<2sec. +2/4 peripheral pulses. No edema, clubbing, cyanosis.
Diagnostic Studies

- **Echo:**
  - RA moderately dilated.
  - RV moderately hypertrophied, moderate to severely dilated, estimated RVSP 100mmHg + RA pressure, and moderately reduced systolic function.
  - LV mildly reduced systolic function.

- **12-lead ECG:** Sinus rhythm, rate 76, RVH, RV strain

- **CXR:** Prominence of MPA. Normal cardiac size.

- **6MW test:** 614m (2,016 ft). Notable for $O_2$ sat drop to 81% and dyspnea score by Borg scale 5 with exercise.

- **PFT:** Normal lung volumes. Normal DLCO

- **CPET:** Severe dysfunction
  - Functional capacity - $VO_2_{\text{max}}$ 34% predicted (nl>80%)
  - $VO_2_{\text{max}}$ at anaerobic threshold 21% predicted (nl>40%)
  - HR response normal, but peak was below target
  - $O_2$ saturation down to low 60s at peak exercise

- **V/Q scan – Normal perfusion and ventilation. No PE**

- **Serum labs – NT-proBNP 423pg/mL (nl<300). All other labs within normal limits.**

- **Cardiac catheterization**
  - Baseline Data: $FiO_2$ 0.30, not intubated
  - Saturations: Ao - 91% (Small PFO with right-to-left shunting)
  - Pressures: RA10, RV 109/11, LPA 111/61, m81 (suprasystemic) LPCW 14, DAo 80/50
  - Hemodynamics: CI 2.9 L/min/m$^2$; PVRi 33Wu, Rp:Rs 1.9
  - Vasodilator testing: $FiO_2$ 1.0, iNO 40ppm - no response
  - Angiography: PA wedge in both lower lobes abnormal with pruning.
Treatment

- IV prostacyclin recommended but pt & family deferred for oral therapies – sildenafil, then ambrisentan
- Warfarin started. Continue supplement O2 at night.
- Improved symptomatology but echo findings demonstrated suprasystemic RV pressure.
- IV treprostinil started 10 months after initial diagnosis.
- Follow up cath (1 year later)
  - CI 4L/min/m²; Systemic PAP, PVRi 12Wu, Rp:Rs 0.78

Case Study #2

- 6 yo female with 1.5 year history of decreased exercise tolerance and significant fatigue
  - Requesting to be carried after walking 1 block
  - Worsening “gray spells” over time
  - Dizziness with pre-syncopal episodes
  - More symptomatic at altitude and with URIs
  - Waking up tired in the morning and constantly tired throughout day

Case Study #2

- PMH: 3 months prior, pt had 2-3 day PICU stay for severe concussion after a fall from bike riding. Head CT and MRI normal.
- Family history: No significant issues. No sudden deaths.
- Cardiac Exam: RRR. Grade II/VI SEM at LLSB. Abnormal RV tap.

Cardiac Evaluation

- Echo: Severe RVH. Trivial TR. Septal motion normal, indicating low RV pressure.
- Cardiac-gated CT scan: RVH, IVS wall flat in systole and diastole. LV function was normal. Normal branching of lungs.
Cardiac Evaluation

- Cardiac catheterization:
  
  Baseline State: Room air
  - Pressures: RA 3, RV 47/4, LPA 48/16, m33, LA 4, LV 68/4, AAo 64/37
  - Hemodynamics: CI 3.9L/min/m² (Fick); PVRi 9Wu, Rp:Rs 0.53

  Acute Vasodilator Testing: FiO2 1.0/iNO 20ppm –no response

  Wedge angiogram showed no excessive pruning.

Case Study #2

- Sildenafil initiated, decreasing cyanotic spells and dizziness. Parents noted “bursts” of energy, but exercise tolerance was not improved

- Local cardiologist refers child for further PH work up

PH Work Up

At initial visit –

- 12-lead ECG: sinus, rate 73, RA enlargement, RVH

- Echo:
  - Dilated RA
  - RV mildly dilated, mildly hypertrophied, normal systolic function
  - Trivial TR
  - Normal IVS movement
  - Tiny PDA and PFO shunting left to right

Further Work Up

- Repeat echo with exercise –
  - Mild but increased TR estimating RVP 120+RAP
  - Flattened IVS
Work Up

- Q scan – normal
- Abdominal ultrasound – normal
- 6MW (skip) test – 256m (840 ft)
  - Lowest O2 saturation 93% with exercise
- PFT – Deferred – unable to complete
- Polysomnography – moderate OSA
- Blood work – NT-proBNP 137pg/mL, otherwise normal.

Treatment

- Bosentan added
- Supplemental oxygen with sleep
- Repeat cardiac cath in 6 months with likely plan to start IV treprostinil at that time.
- Continued to have pre-syncopal episodes and syncope with agitation and crying.
  - Cath – CI 4L/min/m²; PAP 1/2 – 2/3 systemic, PVRi 6.3, Rp:Rs 0.43 (previous 0.53)
  - Cath (2.5 years later) – normal CI, PVRi 4, Rp:Rs 0.2

If you don’t look…

Coordinating Care

Team Line Up

- Cardiology
- Pulmonology
- Anesthesia
- Radiology
- Cardiac catheterization lab
- ICU
- Other medical specialties

- Referring physician and other specialties
- Social Work
- Child Life
- Insurance/Admitting staff
References


Thank You!