Continuous PA Pressure Monitoring in Two Pediatric Patients with PAH

Venus Anderson, APRN-NP
Pulmonary Hypertension Coordinator
Children’s Hospital and Medical Center, Omaha NE

Disclosure

• I have no disclosures

Objectives

• Introduce CardioMEMS™ device
• Present case study of our 2 patients with CardioMEMS™ device
• Discuss RV performance and the exciting things we’ve done with the CardioMEMS™ device

Background

• The Cardio Micro-Electro-Mechanical Systems (CardioMEMS™) device is a wireless monitoring sensor implanted to measure Pulmonary Artery (PA) pressures.
• The CardioMEMS™ device received FDA approval in 2014 after data from the CHAMPION trial showed that monitoring PA pressures helped guide and improve outpatient management of (adult) heart failure preventing admissions.
• We proposed placement of the CardioMEMS™ device to directly measure pulmonary artery pressures continuously in our Pulmonary Artery Hypertension (PAH) patients to help guide treatment to improve symptoms, avoid repeat cardiac catheterizations, and offer insight to better manage their care.
CardioMEMS™ device

- Intravascular sensor implanted in the distal pulmonary artery
- Sensor 15 mm long x 3mm wide x 2mm thick
- Sensor is air tight capsule containing inductor coil and pressure sensitive capacitor

• Blood flows past the sensor creates a resonant frequency that translates to a pressure waveform
• Nitinol wire loops extend from each end of sensor for stabilization in vessel
• Typically occupies ~10% of the vessel
• Endothelialization within 3 months

CardioMEMS™ device

• Implanted during right heart catheterization
• Need vessel diameter of at least 7mm. Most adult implants are in the LPA, but can be either. We prefer RPA given possibility of reverse Potts shunt in future.
• Calibrate at time of implant (systolic/diastolic/mean PA pressure, wedge, HR, and CO)
• Pair with patient system and complete education

• No leads or batteries
• MRI approved for 1.5 or 3.0 Tesla imaging
• If patient decides they do not want to continue, sensor remains in place with no risk
• Works in conjunction with pacemakers, ICDs, and ventricular assist devices
• Technical support available to staff and families
• Contraindications:
  - Vessel diameter – given our pediatric population
  - Inability to take/tolerate ASA/Plavix

CPT billing codes for reviewing/monitoring, done every 31 days
-93297
-93299
CardioMEMS™ device

Anti-platelet:
ASA 81 mg daily, indefinitely
Plavix 75 mg daily x 30 days

Antibiotic Prophylaxis (SBE):
Recommended for 6 months after CardioMEMS device placement (Abbott does not have recommendation)

Follow up:
CardioMEMS™ device interrogated at each clinic visit with echocardiogram. Patient instructed to send daily reading from home unit in the beginning and then 2-3 x per week

Patient #1

- March, 2012, 11 year old female presented to PH team after 5 separate syncopal episodes with multiple visits with PCP and Emergency room staff over the past year.
- ER visit, an echocardiogram was obtained showing severe RV enlargement, hypertrophy, and dysfunction.
- Cardiac catheterization urgently done showing PA pressures 2 times systemic with PVRi > 30 Wu x m2. Immediate transfer to PICU on iNO and initiation of triple therapy.
- Maintained on triple therapy (PDE5i, ERA, and Prostacyclin IV) and followed closely over the next few years with some improvement on serial echocardiograms (1/2 systemic RVSP). Concerns for non-compliance with oral therapies.

Patient #1

- 2014 - echo now predicting systemic level RVSP, non compliance with oral agents admitted. Escalation of Veletri and further patient education but no improvement noted on echo.
- 2015 - repeat cardiac cath showed systemic level PA pressures despite triple therapy (mPAP 52 mmHg, PVRi 19 Wu x m2).
- IV therapy escalated, reinforcement education oral therapy completed. Increased frequency flu and echocardiography
Patient #1

• August 29th, 2018 the CardioMEMS™ device was placed to obtain daily readings of her PA pressures which were higher than anticipated.
  - Supra-systemic PA pressures, 120/60 (85)
  - Systemic pressures 81/33 (46)
  - PRVi 17.5 units x m²
• Daily CardioMEMS™ device readings with mPAP 78-99 mmHg!

Patient #1

• 9/2018 - 24 hour ambulatory blood pressure to compare systemic and PA pressures, showed PA pressures that exceed systemic BP at all times.
  - Average systolic blood pressure of 103 mmHg.
  - Highest recorded BP is 126 mmHg
• CardioMEMS™ device readings from 9/2018
  - 123/58 (83)

Patient #1

• Discussion with our CT surgical team occurred, was referred for reverse Potts shunt. Continued to have preserved RV function, normal proBNP levels and stable 6MWT.
• December 12, 2018 – Reverse Potts shunt (14mm tube graft) distal LPA to descending aorta
• During induction had significant systemic hypotension (systolic 60, MAP 30) not responsive to Epi boluses.
• Intra-op use of CardioMEMS™ demonstrated PA pressures consistently above systemic (systolic 75, mPAP 50). Urgently placed on ECMO

Patient #1

Prior to placing reverse Potts shunt, LPA needled to directly measure PA pressures and correlate to CardioMEMS™ device. Readings near identical and confirmed supra-systemic PA pressures

133/66 (90)

74/44 (56)
Patient #1

- Weaned ECMO intra-operatively without complication
- PICU course uneventful with differential cyanosis ~ 10-15 points
- Discharged post-op day #6
- F/u 2 weeks after discharge
  - proBNP normal range
  - stable chest x-ray
  - differential cyanosis 2-5 points
  - CardioMEMS™ device readings, systemic level PA pressures
- Started Remodulin wean, current rate 89 ng/kg/min. Plan to decrease by 4 ng/kg/min every other cassette change
- Daily documentation of oxygen saturations (upper/lower)

Patient #1

- Weekly to bi-weekly f/u with differential cyanosis 3-15 points
- Normal proBNP levels
- CardioMEMS™ device readings show equal pressures to systemic pressures
- RV less enlarged and LV better filled, good function, and improved TR.
- Patient report improved exercise tolerance and energy level
- Remodulin @ 34 ng/kg/min

97/54 (72)
Patient #1

CardioMEMS™ device readings trend – Patient #1

Have seen ~ 20 point decrease in mPAP post op

- Plan to complete Remodulin wean mid-March and remove central line
- 2012 – current ~ 1-2 line issues per year
- Improved quality of life without central line/pump
- Patient most excited to “go swimming”
- Without the CardioMEMS™ data, may not have moved to reverse Potts
- Improved compliance with oral therapies

Patient #2

- 16 year old male born with d-TGA, s/p balloon atrial septostomy for cyanosis DOL #1 with arterial switch at DOL #4.
- 2002, 1st year of life, evidence of 80% systemic pressures in his RV and PAs. Bosentan started as he did not respond to vasodilator challenge.
- 5 yr. of age, advanced to dual therapy with Sildenafil given lack of improvement noted on echo
- 2014, patient reported more fatigued, near syncope on a few occasions, echo suggested RV pressure ~ 90mmHg.
- Right heart cath found systemic level pressure reaching supra-systemic levels with stimulation. Started on IV Prostacyclin.

Patient #2

- Lung transplant referral made to transplant center, but due to clinical stability, no further progress made with lung transplant.
- He has been maintained on triple therapy with good compliance.
- Repeat cath, 8/2017 on triple therapy showed PVR 14 Wu x m2 with mean PA pressures 64 mmHg.
- 8/29/2018 CardioMEMS™ placed

(cath data)
mPAP 66 mmHg,
PVRi 12.68 Wu x m2
PCWP 5
Patient #2

- 24 hour ambulatory BP monitor
  Average systolic blood pressure of 105 mmHg. Highest recorded BP is 117mmHg.
  [Graph showing 24 hour ambulatory BP]
  124/59 (86)
- Discussion about reverse Potts shunt with significantly elevated PA pressures and inability to titrate therapies further due to adverse effects. Family considering this option.
- Adcirca 40 mg daily, Letairis 5 mg daily, and Remodulin 80ng/kg/min.

The Right Ventricle

- Thin walled crescent shaped ventricle, unable to adapt
- Presence of chronic elevation, RV contractility adapts to maintain
- RV contractility measured by end systolic elastance (Ees)
- RV afterload is arterial elastance (Ea)
- Ees/Ea ratio represents RV systolic performance

Ventricular-vascular coupling

\[
\text{Ees} / \text{Ea} \text{ ratio}
\]
This is known as the ventricular-vascular coupling ratio and is an independent predictor of outcomes in PAH. It is the ability of the RV to perform given the afterload.

\[\text{RV End-systolic elastance (Ees)}\]
The slope of a line through the end-systolic pressure-volume point (the left upper corner of the P-V loop) is termed End-Systolic Elastance (Ees) and is a measure of ventricular contractility.

\[\text{Arterial elastance (Ea)}\]
The slope of a line that starts on the volume axis at the end-diastolic volume, and goes through the end-systolic pressure-volume point. This line is called the Arterial Elastance line (Ea); Ea indicates ventricular afterload.

Ventricular function is defined by a pressure-volume relationship.
Exciting new direction!

Evaluate RV performance
- Cardiac cath or MRI, require sedation and/or intubation, not simultaneous
- 3D echo (3DE), non-invasive
- The CardioMEMS™ device is a non-invasive way to measure PA pressures at the same time as RV volumes from 3DE to provide data necessary to evaluate RV elastance (performance).
- This data can be an earlier marker of prognosis for our PAH patients before other RV functional assessment tools are abnormal.

Exciting new direction!

Ventricular function is defined by a pressure–volume relationship.

Exciting new direction!

Patient #1 data
- In Patient #1, we collected data at one pre-op visit, 1 month post-op and again at 2 months post-op.
- We obtained CardioMEMS™ PA pressure readings and RV volumetrics with 3DE SIMULTANEOUSLY.
- The data was analyzed and we were able to create a RV systolic pressure and volume loop.
- This kind of data is reflective of RV performance and could help to guide our therapies for our PAH patients earlier and more efficiently… This is exciting!
CardioMEMS™/3DE pressure-volume curve

<table>
<thead>
<tr>
<th></th>
<th>EDV (ml)</th>
<th>ESV (ml)</th>
<th>SV (ml)</th>
<th>EF (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-12/11</td>
<td>215.4</td>
<td>100</td>
<td>115.4</td>
<td>53.6</td>
</tr>
<tr>
<td>Post 1 month-1/18</td>
<td>206.1</td>
<td>113.5</td>
<td>92.5</td>
<td>44.9</td>
</tr>
<tr>
<td>Post 2 month-2/21</td>
<td>179.7</td>
<td>88.8</td>
<td>90.9</td>
<td>50.6</td>
</tr>
</tbody>
</table>

CardioMEMS™ device

The Road Toward Precision in PH: Personal Omics, Phenomics, and Wearables—Oh My!

Dr. Raymond Benza at Allegheny Health Network conducted a pilot study of 24 moderate-high risk adult PAH patients with the CardioMEMS device. 2/3 of the patients were on parental prostacyclin and more than half were on triple therapy.

- Discussed the safety and utility of the CardioMEMS™ HF system in patients with advanced PAH.
- Proposed that changes in PAP, CO, and ventriculo-vascular coupling detected continuously may offer an accurate and earlier prediction of clinical worsening of PAH patients.
- Utilized MRI, not 3DE (not simultaneous)

Conclusion:

What did we learn?

- The CardioMEMS™ device is reliable and we can avoid frequent cardiac catheterizations
- Easy to use at home
- Daily data increases anxiety/urgency in decision making
- Guides us to intervene sooner with objective data to support high risk surgery
- Use in conjunction with 3D echocardiography looking at pressure/volume loops for RV elastance

Conclusion:

What will we change?

- Consider CardioMEMS™ device when initiating triple therapy in our adolescent population rather than just focusing on those already on triple therapy
- More aggressive titration of therapy and palliative intervention to avoid RV failure and associated elevation of bio-markers (ProBNP)
Our PH team!

Thank you to our entire team!

Dr. Scott Fletcher
Dr. Paul Sammut
Dr. James Hammel
Dr. Jason Christensen
Dr. Kristina Rauser-Foltz
Cynthia Foster, RN, BSN
Kristine Fuller, RN, BSN
Jill Bechaz, Pharmacist
Mary Craft, RDMS, FASE
Li Ling, MD, PhD, RDMS

References:


Abraham, W. T., Stevenson, L., Bourge, R. C., Lindenfeld, J., Beaman, J., & Al-Assaf, F. (2016). Sustained efficacy of pulmonary artery pressure to guide to adjustment of chronic heart failure therapy: Complete follow-up results from the CHAMPION randomized trial. The lancet, 387(10017), 403-411. [http://dx.doi.org/10.1016/S0140-6736(15)00723-0](http://dx.doi.org/10.1016/S0140-6736(15)00723-0)


