Resolute PAS

- **CONDITION:** Coronary artery disease
- **PI:** Yale Wang, MD
- **CONTACT INFO:** Rose Peterson | Rose.Peterson@allina.com | 612-863-6051

- **DESCRIPTION:** A study to assess the continued safety and efficacy of the Resolute Onyx stent for the treatment of lesions in coronary arteries amenable to treatment with the Resolute Onyx 2.0 mm – 5.0 mm stent. The purpose of the study is to observe the continued performance of the Resolute Onyx Zotarolimus-Eluting Stent System in a real-world population.

- **CRITERIA LIST/QUALIFICATIONS:**
  - Symptomatic coronary artery disease including subjects with chronic stable angina, silent ischemia, and acute coronary syndromes including non-ST elevation and ST-elevation myocardial infarction
  - Subject is an acceptable candidate for treatment with a drug-eluting stent in accordance with the applicable guidelines on percutaneous coronary interventions, manufacturer’s Instructions for Use, and the Declaration of Helsinki.
  - Key exclusion criteria: unprotected left main disease; subjects with planned PCI of three vessel disease

- **SPONSOR:** Medtronic
Midwest Adult Congenital Center
Cardio-Pregnancy Program

- Children’s Heart Clinic
  - Surgery/EP/intervention/imaging/admin
- Minneapolis Heart Institute
  - Surgery/HF/EP/structural/imaging/admin
- Allina Health
  - Med-peds
  - Anesthesia
  - Intensivists
  - Administration
  - Pulm/Heme/ID/GI/Renal
- Minnesota Perinatal Physicians

The Changing Mortality in CHD

- 30% decrease in mortality
- Mortality shifted from children to adults
- Median age of death was 57 years (2004-2005)
- Complex CHD:
  - median age of death changed from 2 to 23 years during the study

Khairy et al. JACC 2010; 56:1149-1157
The Changing Prevalence of ACHD

prevalence of congenital heart disease (CHD) in the European Union by age group.

2000: 0.8 - 1.3 million adults with CHD in the US

% > 60 years of age 2003-2012:

- 100% increase in simple lesion admissions
- 53% increase in complex lesion admissions
- 50% of admissions were Emergency
- Most common admission diagnosis
  - CHF, Valve disease, Stroke
- High percentage of diabetes, PVD, CAD, HTN, smoking, obesity

Changing ACHD Admission Demographics

95% increase in valve procedures
57% increase in pacemakers

National Inpatient Sample Database (195,306 admissions)

Shikhar Agarwal et al. J Am Heart Assoc 2016;5:e002330
ACHD Workforce

- 2000: 0.8 - 1.3 million adults with CHD in the US (1/85 children, 1/245 adults)
- 2008: < 10% of the ACHD population is followed in specialized ACHD centers
- Over a 5 year period:
  - 50% of ACHD patients hospitalized (> 30 yo most rapidly growing segment)
  - 16% required critical care
  - 68% had at least one ER visit
- Estimated provider need: 450 ACHD specialists
  - < 1% of cardiology trainees (peds/adult)
  - 50 ACHD fellowship trainees between 1995-2005
- MHI/CHCM/CHC: recruiting for a director for 10 years until 2016
  - Kirsten Dummer program lead until 2014 – transitioned to fetal program
  - (F Wu, M Earing, A Zaidi, A Egbe, A Yetman, K Marsh, J Geradin, J Johnson, A Cedars)

Comprehensive ACHD Care Center Requirements

Certification will be required for reimbursement in the future

**ACHA ACHD Program Criteria**

**Comprehensive Care Center**

- A. ACHD Cardiologist
- B. ACHD Medical Program Director
- C. Advanced Practice Nurse/Physician Assistant
- D. Registered Nurse
- E. Cardiothoracic Surgery and Cardiothoracic Intensive Care Unit
- F. Heart Failure, Heart Transplant, Heart/Lung Transplantation
- G. Interventional Cardiac Catheterization
- H. Interventional Electrophysiology
- I. Inpatient Services
- J. Outpatient Services
- K. Transitional Services
- L. Patient-Centered Care
- M. Echocardiography
- N. Cardiac Magnetic Resonance Imaging
- O. Cardiac Computed Tomography
- P. Pulmonary Arterial Hypertension
- Q. Exercise Testing and Cardiac Rehabilitation
- R. Reproductive Services
- S. Psychology and Social Work

**Key:**

- Must have documentation at the time of official application for the ACHA ACHD Accreditation Program.
- Up to 2.5 years to establish, planning in place at the time of official application for the ACHA ACHD Accreditation Program.
Evolution of ACHD & MACC

- 2003
  - Case based collaboration
- 2003 - 2014
  - Dedicated ACHD clinics at CHC/MHI (Dummer/Carter/Samara/Lesser/Gornick)
  - All echo done at CHC, all MRI/CT MHI
- 2015
  - Established clinics at MHI, echo done at Children’s, train MHI echo techs
- 2016: build outpatient clinic (wait for a director)
  - Expand clinics at MHI (6 clinics/month), all CHD echo at MHI
  - Begin transition development – increase MHI availability/capacity
  - Angela “Lou” Reu started March 2016
- 2016: Continue outpatient expansion, build inpatient (stopped recruiting)
  - Expand outpatient clinics at MHI (12 ½ days per month, 4 full days as of October)
  - Echo coverage 5 days/week December (Nicole Hoekstra and Steph Klopper)
  - MHI scheduler (Laura Wentz)
  - Monthly working group meeting since March (2nd Tuesday of the month at ANW)
  - Monthly case review since 2016 (4th Tuesday of the month Childrens)
  - Formalize surgical collaboration MHI/CHC
  - Begin transfer visits, establish transition program – in process

Cardiopregnancy?? (I’m a ped cardiologist....)

<table>
<thead>
<tr>
<th>CHD Diagnosis</th>
<th>Prior Intervention Residual lesions</th>
<th>Diagnostics</th>
<th>Cath/EP</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>F: 23 yo</td>
<td>AS</td>
<td>Mechanical AVR</td>
<td>Echo, CTA</td>
<td></td>
</tr>
<tr>
<td>F: 38 yo</td>
<td>AV canal, Downs</td>
<td>AV Canal repair Mod-severe MR</td>
<td>Echo, Holter, MRI</td>
<td>Surgery &lt; 5 years</td>
</tr>
<tr>
<td>F: 31 yo</td>
<td>Ebsteins, WPW</td>
<td>Severe TR (45%)</td>
<td>Echo, MRI, CPST</td>
<td>EP study + ablation, Surgery &lt; 1 year MB for pregnancy</td>
</tr>
<tr>
<td>F: 31 yo</td>
<td>AS</td>
<td>Valvotomy x 2 Ross procedure Moderate conduit stenosis</td>
<td>MRI, holter, CPST</td>
<td>RV-PA conduit &lt; 5 years</td>
</tr>
<tr>
<td>F: 24 yo</td>
<td>AS/AL, s/p valvotomy</td>
<td>AS/AL, s/p valvotomy</td>
<td>MRI/echo/holter/CPST</td>
<td>Hemodynamic cath, Surgery &lt; 6 months</td>
</tr>
<tr>
<td>F: 25 yo</td>
<td>TOF, DiGeorge</td>
<td>TOF Repair, severe PI</td>
<td>Echo/holter/MRI</td>
<td>AAO replacement &lt; 5 years (AO 5.2 cm)</td>
</tr>
<tr>
<td>F: 29 yo</td>
<td>AS/COA</td>
<td>COA repair x2, Valvotomy Ross procedure</td>
<td>Echo/holter/MRI</td>
<td>AAO replacement &lt; 5 years (AO 5.2 cm)</td>
</tr>
<tr>
<td>F: 26 yo</td>
<td>AV Canal, Downs</td>
<td>AV canal repair Mod-severe MR</td>
<td>Echo/holter</td>
<td></td>
</tr>
<tr>
<td>F: 24 yo</td>
<td>PA/VSD</td>
<td>RV-PA conduit Severe PI</td>
<td>Echo/holter/MRI/CPST</td>
<td>RV-PA conduit &lt; 6 months</td>
</tr>
<tr>
<td>F: 22 yo</td>
<td>COA, Turners</td>
<td>s/p Coa balloon/stent</td>
<td>Echo/CTA q 5 yrs</td>
<td></td>
</tr>
<tr>
<td>F: 29 yo</td>
<td>VSD (small)</td>
<td>35 wks pregnant</td>
<td>Echo</td>
<td></td>
</tr>
<tr>
<td>F: 35 yo</td>
<td>ASD (3.5 cm)</td>
<td>18 weeks pregnant</td>
<td>Echo/Holter/TEE</td>
<td>Hemodynamic cath, Surgery 6 months post partum</td>
</tr>
</tbody>
</table>
Pregnancy in women with CHD

• 43% of women never counseled about contraception
• Use of contraception in ACHD
  • 36% of women were using appropriate contraception for CHD lesion
  • 20% of women use contraception contraindicated for their CHD lesion
    • ie: no estrogen in single ventricle patients
• 48% never told of pregnancy is high risk by their physician
  • pediatricians don’t talk about this stuff, particularly when parents in the room
• 45% of pregnancies were unexpected
• UK – cardiac disease is the most common cause of death in pregnancy
  40% of death attributed to substandard care

Lindley, ObstetGynecol 2015 Aug;126(2):363-9
Thorne, Heart 2006, Oct; 92 (10) :1520-1525
Vigl, AM J Cardiol. 2010 Nov 1; 106(9)1317-21

![Causes of pregnancy-related death in the United States: 2011-2012](CDC-Gov)
Most Common Noncardiac Reason for admission in ACHD

2908 patients CONCOR Registry 2001-2006
8916 admissions (61% cardiac)
4926 interventions (50% cardiac)

The emerging burden of hospital admissions of adults with CHD. Verheugt & Mulder, Heart 2010, Volume 96, Issue 10

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REPRODUCTIVE SERVICES
- P/P encouraging all female patients to have gynecological care
- P/P for discussion of birth control and sexual function as it relates to CHD
- P/P for discussion with all female CHD patients to provide pre-pregnancy counseling/family planning
- P/P for discussion of counseling related to sexual dysfunction
- Documented availability of high risk obstetric care
  - ACHD documented plan for delivery in collaboration with maternal/fetal medicine and anesthesia
  - ACHD providers must have consulting privileges in ACHD OB unit

GENETIC COUNSELING AVAILABILITY

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ACHD and Cardiopregnancy

My Vision

- Stepwise expansion volume
- Stepwise expansion in complexity

Instead......

- Highly complex patients
- Rapid growth of the program
- We have collectively cared for these patients at a very high level with novel therapies
- We have incredible potential that doesn’t exist elsewhere

This program highlights the best of MHI/Children’s/MPP & MotherBaby/ANW/Allina

- Med-Peds
- Adult and Peds Cardiology (EP/Intervention/HF/ICU/Imaging)
- Surgery (peds CV/adult CV/vascular)
- ICU
- Minnesota Perinatal
- Anesthesia
- Nursing and support staff

ACHD Program through 3 case reviews

- Cardiopregnancy/Structural Heart Intervention (Children’s/MPP/MHI/ANW)
  - 23 year old woman with severe AS/MS secondary to renal disease
  - presented 18 weeks gestation
  - MPP & mother baby, OB & Cardiac anesthesia/Med-peds/renal/psychiatry/Cardiology(general/structural)

- Heart Failure/EP/Surgery (Children’s/MHI/ANW)
  - 31 year old male with single ventricle heart disease & mechanical aortic valve
  - flutter/arrest/echo/obstructed mechanical valve

- Structural Heart Intervention (Children’s/MHI/ANW)
  - 44 year old male with tricuspid and pulmonary valve disease
  - Class III-IV heart failure, not a transplant candidate
  - Hospitalists/MHI surgery, CHC/MHI intervention, Advanced HF, ACHD
Background: ACHD and Maternal Pregnancy Risk

Prior cardiac event (heart failure, transient ischemic attack, stroke before pregnancy or arrhythmia).
Baseline NYHA functional class III or IV.
Left heart obstruction (mitral valve area <.2 cm²; aortic valve area <.5 cm²; peak LV outflow tract gradient >30 mmHg by echocardiography).
Reduced systemic ventricular systolic function (ejection fraction <50%).

Mortality: 0.0% (95% CI 0.0%–1.1% in 75% of patients). (AVSNA research)'

History of arrhythmias.
Baseline NYHA functional class IV.
Left heart obstruction (aortic valve peak gradient 166 mm Hg).
Reduced left ventricular ejection fraction.
Left ventricular systolic dysfunction.
Reduced left ventricular systolic dysfunction.
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WHO Class I
Uncomplicated, small or mild
- Pulmonary stenosis
- Patent ductus arteriosus (PDA)
- Mitral valve prolapse
Successively repaired simple lesions
- Ventricular septal defect (VSD)
- Atrial septal defect (ASD)
- Anomalous pulmonary venous drainage
- PDA

WHO Class II (restricted to patients previously well and uncomplicated)
- Uncomplicated ASD or VSD
- Repair Tetralogy of Fallot (without sequelae)
- Most arrhythmias

WHO Class II-III (depending on the individual)
- Mild left ventricular impairment
- Hypertrophic cardiomyopathy
- Native valve disease or tissue heart valve replacement not considered in class I
- Marfan syndrome without aortic dilation
- Aorta <4.5 mm associated with bicuspid aortic valve disease
- Repair aortic coarctaion

WHO Class III
- Mechanical valve replacement
- Systemic right ventricle
- Fontan circulation
- Cystic heart disease (unrepaired)
- Other complex congenital heart disease
- Marfan syndrome aortic dilation >4.5 mm
- Aortic dilation 4.5–50 mm associated with bicuspid aortic valve disease

WHO Class IV (pregnancy contraindicated)
- Pulmonary hypertension
- Severe systemic ventricular dysfunction (LV EF <30%, NYHA III or IV)
- Previous peripartum cardiomyopathy with residual impairment of left ventricular function
- Severe mitral, severe symptomatic aortic stenosis
- Marfan syndrome aortic dilation >4.5 mm
- Aortic dilation >50 mm associated with bicuspid aortic valve disease
- Native severe aortic coarctation

Pregnancy OB Complications by WHO Category
ROPAC: 2700 pregnancies 2008-2014

Incidence and predictors of obstetric complications in women with structural heart disease. BMJ. Van Hagen et al
Maternal Adverse event by ACHD Diagnosis

What about the Baby???

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Fetal Outcomes in Maternal ACHD

213 pregnancies/203 women

10% maternal events
(mortality, arrhythmia, vascular event, need for urgent intervention 6 months post partum, endocarditis)

37% offspring events
(fetal death, neonatal death, premature delivery < 37 weeks, SGA, RDS, infections, NICU admission, CHD, or IVH)

Case#1: Cardiopregnancy/Structural Heart Intervention (Children’s/MPP/MHI/ANW & Allina)

- 23 year old woman
- ESRD on dialysis (s/p transplant with graft failure years prior)
- Secondary hyperparathyroidism
- Renovascular hypertension
- Chronic pain and bipolar disorder, h/o suicide attempts and depression/anxiety, opioid dependency
- Anemia of chronic renal failure
- Exertional shortness of breath and syncope (Class III-IV symptoms)
- Presented 18 weeks pregnant with severe MS and severe AS
  - By report counseled that pregnancy was not high risk
Diagnostic Evaluation

- **TTE and TEE (6/19/2017):**
  - Severe AS: Peak velocity 4.98, mean gradient 59 mmHg, valve area 0.9 cm².
  - Severe MS: Mean gradient of 14.8 at hr 82 bpm (TTE) and mean gradient of 10 mmHg (TEE)
  - Good LV fxn, LVH, mild TR with normal PA pressure estimates

- **CT (6/20/2017):**
  - Annular area 397 mm², perimeter 72 mm,
  - Coronary ostia height 15 mm left, 14 mm right
  - Recommendation: 23 mm Sapien with 2.3% oversizing vs 26 mm Evolut

- **RHC: 6/21/2017 RA3, RV 36/8, PA 31/12, mean 20 PCW 12**
Symptomatic Severe AS in Pregnancy

- Mechanical valve
  - Bypass/Surgery
  - Anticoagulation: VKA or therapeutic Heparin
  - Hemorrhage/Valve thrombus
  - Long term valve

- Bioprosthetic Valve
  - Bypass/Surgery
  - Anticoagulation: Aspirin
  - Repeat surgery < 10 years

- TAVR
  - No Bypass
  - Anticoagulation: Aspirin and Plavix
  - Repeat Surgery?

Cardio-Pulmonary Bypass in Pregnancy

- Literature review of cases from 1991-2013
- Most common indication is for mitral or aortic valve replacement/repair
- Maternal mortality similar to non-pregnant patient
- 18.6% fetal mortality (highest in earlier trimesters)

<table>
<thead>
<tr>
<th>Variable</th>
<th>Survived</th>
<th>Died</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maternal age (years)</td>
<td>26.5±6.1</td>
<td>28.6±5.4</td>
<td>0.8380</td>
</tr>
<tr>
<td>Week of gestation at onset of cardiac symptoms</td>
<td>23.4±6.3</td>
<td>27.5±7.4</td>
<td>0.3047</td>
</tr>
<tr>
<td>Week of gestation at cardiac surgery</td>
<td>24.2±8.4</td>
<td>19.0±7.2</td>
<td>0.0873</td>
</tr>
<tr>
<td>Week of gestation of patients who underwent cardiac surgery under CPB prior to delivery</td>
<td>17.0±7.7</td>
<td>17.3±10.3</td>
<td>0.8893</td>
</tr>
<tr>
<td>Time of delivery (week of gestation)</td>
<td>36.5±6.1</td>
<td>28.1±7.3</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Aortic pressure (mmHg)</td>
<td>70.4±6.6</td>
<td>72.3±11.1</td>
<td>0.1935</td>
</tr>
<tr>
<td>Flow rate (a/kg min)</td>
<td>3.3±1.2</td>
<td>3.9±1.8</td>
<td>0.4645</td>
</tr>
<tr>
<td>Temperature (°C)</td>
<td>32.7±4.0</td>
<td>31.0±3.6</td>
<td>0.0895</td>
</tr>
<tr>
<td>Cardiopulmonary bypass time (min)</td>
<td>86.5±43.5</td>
<td>104.3±58.6</td>
<td>0.8876</td>
</tr>
<tr>
<td>Cross-clamping time (min)</td>
<td>40.8±35.1</td>
<td>44.9±23.0</td>
<td>0.7011</td>
</tr>
</tbody>
</table>

Anticoagulation in Pregnancy

- **Warfarin**
  - Best for maternal valve safety
  - Worst for baby (> 5 mg)
    - Crosses placenta
    - 6-12 wks embropathy
    - Fetal hemorrhage

- **Heparin (UFH or LMWH)**
  - Worst for maternal valve/thrombosis
  - Systemic maternal side effects (osteoporosis)
  - Best for baby (no effects)
    - Does not cross placenta

TAVR – Dr. Mooney

- Aortic balloon valvuloplasty
- 23 mm Sapien S3 device deployed
- 1+ AI noted, perivalvular area, augmented balloon inflation – resolved
- Post procedure Aortic velocity 2.59 m/sec, mean gradient 13mm Hg and EOA 1.5 cm²
- LBBB x 2 days, resolved prior to hospital discharge
- Now only one WHO Class IV contraindication for pregnancy + one class III
- Admitted in preterm labor at 23 weeks (thank you LOU!!!)
- IM to Mother baby to CVICU to mother baby
- Vaginal delivery at 26 weeks, 1.9 kg baby boy (in NICU)
Other Pregnant ACHD Patients 2017

- 13 ACHD patients followed since January 1, 2017
  - 5 delivered (1 premature, 4 term deliveries)
  - 6 currently pregnant
  - 1 transferred care to Mayo
  - 1 miscarriage
- ACHD Diagnosis
  - AS/TAVR x1
  - AS/MS and TAVR x1
  - AV Canal
  - ASD
  - Tetralogy of Fallot
  - Aortic Coarctation
  - Fontan
  - D-TGA, s/p atrial switch with systemic RV

ACHD and Cardio-Pregnancy

- Cardiopregnancy is an essential component of ACHD care
- ACHD is only a subset of a Cardiopregnancy Program
- Programmatic approach to all high risk cardiac disease in pregnancy
  - Arrhythmia (fetal and maternal)
  - Valve disease (non congenital)
  - Aortopathy
  - Heart Failure
  - Coronary artery disease/dissection
  - Womens Health
    - Pre-eclampsia – 2-8% of pregnancies (Increased risk of heart disease 2-4x, similar to smoking)
- Collaboration between MPP & mother baby, MHI &ANW
CardioPregnancy Program Vision

- Transition visit in late adolescence focused on contraception/pregnancy risk/pregnancy outcome
- Ob/Gyn Care & Contraception for WHO 3&4: MOMS clinic at mother/baby
- Pre-conception counseling: maternal risk of pregnancy AND fetal outcomes
  - Combined perinatal/cardiology visit
  - Medication review and changes when necessary prior to conception (ACE/Statins/Warfarin are teratogenic)
  - Pre pregnancy intervention when needed to reduce maternal and fetal pregnancy risk (valve lesions/aortopathy)
  - Neonatology consult for high risk patients
- Pregnancy Care
  - Appropriate perinatal & cardiology care for cardiac condition – needs to be patient centered
  - Cardiology care determined by WHO category
    - At least one visit during pregnancy, most will have visit each trimester or monthly
  - Fetal echocardiography (CHC team)
  - Neonatal consult for patients at high risk of prematurity or fetal diagnosis of CHD
  - Delivery plan (where: Mother baby or cardiac ICU? OB nurse to cardiac ICU or cardiac nurse to OB/Childrens CVCC/MHI CVCC)
    - C-section VS vaginal delivery/assisted second stage VS routine delivery
    - Monitoring
    - Anesthesia (neuraxial vs general, cardiac vs OB anesthesia)
    - Anticoagulation
    - Post partum
- Cardiopregnancy “Level 1” equivalent (maternal cardiac arrest – 5 minutes to get the baby out)
  - Cardiac OB catastrophe plan
    - Surgical: two patients, who operates first – aortic dissection at 30 weeks
    - Cardiac arrest: cardioversion/ECMO/Delivery
    - TAVR valve thrombus: Low dose thrombolytics

Case #2: Heart Failure/EP/Surgery (Children’s/MHI/ANW/Allina)

- 33 year old male
- Single Ventricle, Followed by ACHD (followed by Dr. Singh, now by Dr. Carter)
- Physiological equivalent of tricuspid atresia
- Atrio-pulmonary Fontan 1988, s/p revision to lateral tunnel 1999
- AI, s/p mechanical AO valve replacement 1999 (25 mm St. Judes)
- SN dysfunction, s/p epicardial pacemaker
- Atrial tachycardia
- Meds: Coumadin (non-compliant, valve occlusion 2010 treated with TPA)
  - Enalapril
  - PCN (asplenia)
  - Amiodarone
- Social: works, has his own home, 4 year old daughter
- BMI 17, weight 100 lbs, sats 93%
Asplenia, Right AV Valve Atresia, VSD, PA, L-malposed AO and anomalous PV

**SURGICAL**
- 1984: BT shunt x2
- 1988: RA-PA Fontan
- 1999: Fontan Revision AO valve
- 2006: Valve Thrombus TPA

**EP**
- 1984: SND
- 1988: Pacer
- 1999: EPS: Atrial tachy, ventricular rate > 300 bpm
- 2010: Sotolol
- 2012: Epicardial pacer
- 2015: For fractured atrial lead 5th sternotomy

HEART FAILURE, EP, VALVE DISEASE.......

Clinic Visit prior to Admission

- Albumin 4.4 (no Protein Losing Enteropathy (PLE))
- AFP: normal (screen for hepatocellular carcinoma)
- BNP 38
- HGB 17.2

**Baseline echo:**
- Low normal single ventricle function
- Moderate common AV valve insufficiency
- Well functioning mechanical aortic valve

**Pacer check:**
- AAI lower rate 60 bpm.
- Sinus at 71 bpm. Atrial paced 18%.
- 2 episodes of atrial tachycardia, 17 seconds total duration
- Malfunctioning RV lead
Risk Factors for Bad Things in Fontan

• Age > 30
• Gender: Male
• Single Ventricle (Heterotaxy)
• SND/flutter/chronotropic incompetence
• Pacemaker
• Prosthetic Aortic Valve
• Wasting/Frailty
• Moderate decrease in ventricular function
• Moderate AV valve regurgitation
• Medical noncompliance


• Fairview Ridges ER
• Exertional fatigue for two days
• Ran out of amiodarone
• INR 1.4 on admission
• Outside Chest CT: read as a saddle embolus – started on heparin and transferred to ANW
• Atrial tachycardia on arrival (7/8)
• Dr. Chu on in the Cardiac ICU
• Dr. Carter on call for MACC
• Amiodarone prior to cardioversion
  • Nausea/vomiting – PEA
  • Emergent VA ecmo – Cardiac ICU
• Echo:
  • moderate AV valve regurgitation
  • EF 40%
  • aortic valve functioning normally
• Weaned from ECMO 2 days later
  • Cardiac arrest
  • ECMO Restarted and to Cardiac ICU
• TTE Echo:
  • Moderate AV valve regurgitation
  • Moderate-severely decreased function
  • Probable obstructed mechanical AO Valve
• TEE
  • L-malposed Aorta
  • AoV not well visualized

- Rhythm control in adults with CHD and IART or atrial fibrillation
  - Identify and treat precipitating factors
  - Consider catheter ablation
  - Systemic ventricular hypertrophy or
    - Systemic ventricular dysfunction?
      - No
          - Flecainide
          - Propafenone
          - Sotalol
          - Amiodarone
          - Dofetilide
      - Yes
          - Moderate CHD
          - Complex CHD
          - Moderate or subpulmonary ventricular dysfunction?
            - No
              - Amiodarone
              - Dofetilide
            - Yes
              - Amiodarone
              - Dofetilide

- DOPI: 3.5 mcg/kg/min
- Epinephrine: 0.2 mcg/kg/min
- Nitroprusside: 0.5 mcg/kg/min

- Metabolism
  - Sodium: 135 mEq/L
  - Potassium: 4 mEq/L
  - Calcium: 1.5 mEq/L
  - HCO3: 26 mEq/L
<table>
<thead>
<tr>
<th>Guidelines</th>
<th>Functional status</th>
<th>Therapeutic strategy</th>
</tr>
</thead>
<tbody>
<tr>
<td>ACC AHA 2014</td>
<td>NYHA Class III, IV Mobile or large clot burden (≥0.8 cm³)</td>
<td>Emergency surgery</td>
</tr>
<tr>
<td></td>
<td>Right sided valves NYHA Class I, II</td>
<td>Fibrinolytic therapy</td>
</tr>
<tr>
<td>ESC 2012⁹</td>
<td>Critically ill patients</td>
<td>Emergency surgery</td>
</tr>
<tr>
<td></td>
<td>Severe comorbidities or unavailability or right-sided valves</td>
<td>Fibrinolytic therapy</td>
</tr>
<tr>
<td></td>
<td>Nonobstructive PHVT if complicated by embolism or persists despite anticoagulation</td>
<td>Surgery</td>
</tr>
<tr>
<td>ACCP 2012⁹</td>
<td>Left sided PHVT and large thrombus area (≥0.8 cm³)</td>
<td>Surgery</td>
</tr>
<tr>
<td></td>
<td>Left sided PHVT and small thrombus area (&lt;0.8 cm³)</td>
<td>Fibrinolytic therapy</td>
</tr>
<tr>
<td>Society for heart valve disease 2005⁹¹</td>
<td>All patients Failed thrombolysis or contraindications</td>
<td>Fibrinolytic therapy Surgery</td>
</tr>
<tr>
<td>PHVT: Prosthetic heart valve thrombosis, NYHA: New York Heart Association</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
NO ONE WANTED TO WRITE THIS ORDER

- Given thrombolytics x 2 doses
  - No change in valve gradient
  - Improvement in ventricular function
  - Bleeding at the groin site and dropped Hgb
- Dr. Ben Sun:
  - minimally invasive AO valve replacement
  - 23mm bioprosthetic tissue valve
  - Hemothorax post-op, MRSA pneumonia
- Weaned from ECMO POD #2
- Discharged to LTAC on 3 weeks of antibiotics
- Digoxin, Carvedilol started for atrial arrhythmia
- ECHO on discharge: moderate MR, LV EF 40%
- Now home
Case 3: Structural Heart Intervention *(Children’s/MHI/ANW)*

- 44 year old man with TOF & Noonan syndrome
- Complete TOF repair with transannular patch (age 1)
- RVOT aneurysm resection (age 5)
- Pulmonary homograft x 2 (age 16 and 24)
- TVR bioprosthetic x 2 (age 16 and 24)
- TVR mechanical (age 33)
- Prednisone and transfusion-dependent Diamond Blackfan anemia
  - Recurrent CHF with Hgb 6-7
- Chronic atrial fibrillation
- Class IV HF symptoms with frequent hospitalizations
• CMR
  • LVEF – 42%
  • RVEF – 50%
  • Pulmonary Insufficiency 43%
  • RVEDVI – 183 ml/m2 (63-111);
  • RVESVI – 92 ml/m2 (18-46)
• High risk surgical candidate
  • 5 prior sternotomies
  • 5 prior valve replacements
  • Transfusion dependent/chronic steroid
• Not a transplant candidate

HEART FAILURE, EP, VALVE DISEASE........

Drs Sorajja, Vezmar, Baker, Khsettry
Perventricular placement of a Edwards 29 mm XT valve

Seen early October: no hospitalization since procedure in April
Palliative Care/Heart Failure team/Med Peds (Children’s/MHI/ANW/Allina)

- 45 year old male with TA, Fontan, chronic PLE x 15 years/Afib
  - Waterston shunt, RA-PA fontan, revision to lateral tunnel, pacemaker for SS syndrome, epicardial pacser, AV node ablation secondary to permanent flutter
  - Admitted with cellulitis, diastolic dysfunction & ascites, renal insufficiency, Lasix infusion
  - Weight 375, not a transplant candidate
  - Discharged to hospice

- 29 year old woman: Downs, Eisenmengers, Baseline saturations 80’s on dual pulm vasodilator therapy
  - Admitted with pseudomonas sepsis, epi/vasopressin intubation, sats 50s, made DNR

- 45 year old man: d-TGA, s/p atrial baffle, severe RV dysfunction and atrial arrhythmias
  - not a transplant candidate due to alcohol and drug addiction, non compliant
  - CT: RVEDV 681/336 m/m2, RV EF 15%
  - Died at home

- Thank you palliative care team (Leslee Macho) and med-peds (Erin Novak)

MACC Outpatient: Current State

- MACC dedicated scheduler: July 2017, Laura Wentz
- Outpatient clinic at MHI since January 2016
  - 2014: 254 patient visits
  - July 2016-July 217: 445 patient visits
  - 12 ½ day clinic days per month in current quarter + full day Wednesday (October 1st)
  - Triage nurse support and clinic nurse support (EP) – September 2017
- 2 MHI echo techs trained since 2016 (Nicole Hoekstra, Steph Klopper)
  - CHD echo 5 days weekly starting November 2017
  - 3rd echo tech to be trained starting December (Molly)
  - Congenital echo template (report generator works for two ventricles, 4 valves)
- PA: Angela (Lou) Reu (March 2016)
- Abbott Pre-op clinic for all patients July 2017
- Coumadin clinic MHI since September 2017
- Monthly clinical case conference since January 2015 (4th Tuesday monthly – Children’s – 10 cases tomorrow)
  - Med peds, anesthesia, ICU, surgeons, interventionalist, MPP, peds and adult CV, EP nurses, palliative care, heart failure
- Monthly ACHD Working group – first meeting March 2017
- ACHD consult service call system: March 2017
- Transition Program – implementation 2017/first quarter 2018 (now have MACC clinic availability)
MACC Surgery – Current State

• 35 MACC surgical cases in the past 12 months
  • Median age 30 (range 20 – 62 years old)
    • 18 cases MHI
      • (PV replacement, AVR, coronary unroofing)
    • 17 cases Children’s (Dr. Overman, Moga)
      • (Konno, composite aortic graft, fontan revision, AVR, generator replacement, ASD)
  • Dr. Moga (operating with DiBardino, Flavin, Askew, Farivar, Kshetry)
    • Scrub in on all CHD cases at MHI: September 2017
    • Agreed to be primary surgeon at MHI within 1 year (declared August 9th, 2017)

MACC Advanced Imaging– Current State

Jana Lindberg/David Caye

- 18 Years old
- 50% of CHD MRI
- 30% of CHD CT
SYSTEMIC RV

Left Atrium

RV AO

L‐TGA, failing systemic RV

Atrio‐Pulmonary Fontan

25 year old, Coarctation

53 year old, Coarctation

D‐TGA, atrial baffle, lead in RV

DORV, s/p Rastelli conduit stenosis

s/p Aortic Valvotomy

s/p Asd closure with RVE

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Radiation Safety in Children With Congenital and Acquired Heart Disease

A Scientific Position Statement on Multimodality Dose Optimization From the Image Gently Alliance

- Guidelines
- ACC AUC
- CT Course
- Core Lab
- Registry

- CTDIvol 0.23
- DLP 2.9
Cardiopregnancy—Current State

3 patients for pre-pregnancy counseling
Fontan coming next week

MACC Transition: Current State

- Transition materials finalized
  - Educational program beginning age 12-14
- Now have MHI clinic availability
- Implement transfer visit during late adolescence by Jan 2018
- Implement full transition program over two years
  - 253 patients age 12-13
  - 286 patients age 14-17
  - 222 patients age 18 and older

The proportion of CHD patients in contact with the healthcare system by age and by CHD complexity

Andrew S. Mackie et al., Circulation. 2009;120:302-309

The proportion of CHD patients in contact with the healthcare system by age and by CHD complexity

Bradley S. Morrow et al., Circulation. 2012;126:1143-1172
Next Steps

- Moving from individual commitment to formalized institutional commitment
  - LOI from 2012 being updated between Children’s and Allina
  - Ongoing discussions regarding legal/billing structures (as complex as the patients....)
- Intervention and surgery: procedural support to programmatic support
- Develop redundancy and depth to existing programmatic components
- Develop non-cardiac specialist support
  - Hematology/Respiratory/ID/Nephrology/GI
  - Nursing/Respiratory therapy, etc
- Develop data and quality improvement mechanisms

Thank You

- Lou Reu
- Pam Glick
- Nancy Newell
- Judy Edwards
- Jana Lindberg
- Dave Caye
- Jill Sandstrom
- Tamara Langeberg
- All EP nurses
- Laura Wentz
- Cassie – MN perinatal
- Laurie Johnson
- Rachel Hinsch
- Megan Hoover
- Kim Adomaly
- Dave Tetzloff
- Tim Wood
- Pete Melchert
- Erin Novak
- All med/peds
- Josh Huelster
- Dave Williams
- Matt Sundblad
- Leslee Macho
- Nicole Hoekstra
- Steph Kloepper
- Pacemaker staff
- Paul Reineke
- Phil Zeccardi
- Tom Talley
- Andrew Sweeney
- Bill Wagner
- Alex Campbell
- JoEllyn Moore
- Matt Chu
- John Lesser
- Paul Sorajja
- Mike Samara
- Barry Cabuyay
- Kasia Renowitz
- Peter Zimbwa
- Dave Burton
- Chris Carter
- Vascular Surgeons
- Mario Goessel
- Coumadin clinic RNs
- Chuck Gornick
- Bill Katsiyaniss
- John Lesser
- Mike Mooney
- Dan DiBardino
- Frank Moga
- David Overman
- Saied Farivar
- Tom Flavin
- Ben Sun
- Carl Mudy
- Kirsten Dummer
- CHC fetal docs
- David Dassenko
CHD Care from fetal diagnosis to palliative care at the end of life.

Fetal Outcome in ACHD/Cardiac Disease

WHO Category and Fetal Outcome

ACHD diagnosis and Fetal Outcome

Fetal CHD risk by Parental CH Diagnosis

Completed Pregnancy by ACHD Diagnosis

Willem Drenthen et al. JACC 2007;49:2303-2311
Expert Reviews of ObstetGynecol 2010