Why build and aortopathy clinic for pediatric and adult multidisciplinary care?
MAD Symposium June 24, 2017
• **Recommendation:** The clinic should diagnose and manage Marfan syndrome and related conditions across age ranges, from prenatal through adult. The transition from medical management in childhood to adulthood is especially important and a clear plan must be in place for all patients (National Marfan Foundation).

• **Vision:** Provide patient and family-centered multidisciplinary care across the lifespan for patients with genetically triggered aortopathies, including Marfan syndrome, and their at-risk family members.
PubMed Search

- Pediatric and Adult Subspecialty clinic - 0
- Family subspecialty clinic - 0
- Transition of care pediatric to adult – 1668
  - Models of comprehensive Multidisciplinary Care of Individuals in the United States with Genetic Disorders (Grosse, PEDIATRICS, 2009)
    - Chronic care model in primary care
    - Disease specific comprehensive clinic
Physician perspective

Marfan syndrome
Loeys-Dietz

Familial thoracic AA (FTAA)

Bicuspid aortic valve with ascending AA

- FBN1/TGFβ, others
- MYH11, ACTA2
- MYLCK, Others
- NOTCH1, TIMP3
- Others

Adapted from HC Dietz, 2014
Family perspective: 32 father with history of aortic dissection at age 34. Family has one child.

Genetic testing
Depends on father’s testing
Father with history of aortic dissection at age 35

- Pediatrician
- Outside Cardiologist
- Genetics
- Imaging
- Adult cardiology
- Pediatric cardiology
- Genetic testing with father (index case)
- Additional imaging where appropriate
- Adult and pediatric cardiology consultation
- Research protocols offered if eligible
- Ophthalmology, Pulmonology, Surgery, Ortho etc
Future State Clinic Sessions

**Morning**
- **Peds Combined – Cards + Genetics, GC**
  - 3 New Ped Combo – 90 mins
  - 1 Rtn Ped Combo – 60 mins
- **Peds Combined – Cards + Genetics, GC**
  - New Ped Combo – 90 mins
  - Rtn Ped Combo – 60 mins
- **New Ped Combo – 90 mins**
- **Rtn Ped Combo – 60 mins**
- **Adult Cards**
  - 1 New Adult Cards – 60 mins
  - 6 Rtn Adult Cards – 30 mins
- **Social Work, Genetic Counselor, Research Coord, Surgeon Consult, etc.**

**Afternoon**
- **Peds Cards**
  - 1 New Peds Cards – 60 mins
  - 6 Rtn Peds Cards – 30 mins
- **Adult Combined – Cards + Genetics, GC**
  - New Adult Combo – 90 mins
  - Rtn Adult Combo – 60 mins
- **3 New Adult Combo – 90 mins**
- **1 Rtn Adult Combo – 60 mins (end of PM session)**
- **Genetic Counselor, Research Coordinator**

**Rooms**
- **A**
  - 8 appts, 4 pts
- **B**
  - 7 appts, 7 pts
- **C**
  - 7 appts, 7 pts
- **D**
  - 8 appts, 4 pts

Session – 8 hours
AM: 11 patients
PM: 11 patients
Goals of a family centered approach

- Identification of at risk family members
- Targeted genetic testing
- Improved patient education
- Improved coordination of care between subspecialties.
- Transition built into the program
- Improved compliance and surveillance
- Decreased morbidity and mortality