Partnering with the Primary Care Provider in the Care of the Child with Complex Heart Disease

- Lowell Frank, MD
- Children’s National Health System
Case presentation

- Born at 38 weeks gestation, routine prenatal care
- Fetal echo → HLHS
- Delivered at local hospital, APGARs 8,9
- UA/UV lines placed; PGE started
- Scheduled transport to CNMC CICU

- Norwood-Sano on DOL #4
- Chest closed on POD #2
- Extubated on POD #6
- Transferred to HKU on POD #14
- RA on POD #18
- Required NG feeds 2 add’l weeks
Discharge summary

- 6 weeks old
- Full PO, 24 kcal/oz formula
- SpO2: 75-80%
- RR 45-70
- Brain MRI

- Meds
  - Aspirin 20 mg daily
  - Lasix 4 mg PO bid
  - Vitamin D
  - Zantac
Now what?
Overview

- Provide a roadmap for children in the single ventricle pathway
- Brief review of anatomy and physiology
- Define the stages of “new normal”
- Associated pediatric issues
  - Growth and nutrition
  - Vaccinations
  - Neurodevelopmental screening
  - ADHD and stimulants
Hypoplastic left heart syndrome

Systemic venous blood

Pulmonary venous blood

Hypoplastic left ventricle

Cardiac output
HLHS s/p Norwood-Sano

- Atrial septectomy
- Reconstructed neoaorta
- 5 mm Sano shunt
What is normal?

- Vital signs
- Follow-up
Interstage: growth and nutrition

- JCCHD NPC-QIC
- Home monitoring
- Team members at CNMC
  - Who coordinates?
Interstage: growth and nutrition

Cross, Future Cardiology, 2012

Cross, Cardiol Young, 2013
Interstage: vaccinations

- Diminished response to inactivated vaccines post-CPB
- Delay 6 weeks from surgery (or ECMO)
- Synagis
- Later vaccine issues – aspirin
  - Influenza
  - Varicella
Case continued…

- Monthly follow-up
- Weaned off lasix by 2 months of age
- Outpatient cardiac cath at 4.5 months of age, good hemodynamics
- Bidirectional Glenn (stage 2) at 6 months of age
- Discharged home on POD #8 with sats 80-85%
Bidirectional Glenn

SVC to PA anastomosis

Pulm veins

Takedown of shunt
The new normal, part 2

- Initially irritable; resolves
- Much more stable physiology
- Saturations 75-85%
- Normal respirations
- Improved feeding
- Less frequent follow-up
- Minimal medications
  - ASA + issues + style!
Cardiac neurodevelopment

AHA Scientific Statement

Neurodevelopmental Outcomes in Children With Congenital Heart Disease: Evaluation and Management
A Scientific Statement From the American Heart Association
This statement has been approved by the American Academy of Pediatrics.

Marino et al., Circulation 2012
Cardiac neurodevelopment

Prevalence of Neurodevelopmental Impairment

- Severe Impairment
- Mild or Combined Disabilities
- No Disabilities

Complexity of Congenital Heart Disease
Cardiac neurodevelopment

SURVEILLANCE

- Developmental surveillance at all well child visits.
- For children with CHD this includes attending to:
  - RISK stratification
    - Are one of the risk factors present? Does the child need formal follow up?
  - Asking the right questions
    - Consider using screening tools to help
  - Documentation of milestones, observations

Courtesy Jacqueline Sanz, Ph.D. Co-Director, CANDO Program Neuropsychologist
Cardiac neurodevelopment

SCREENING and REFERRAL

- Per AAP recommendations:
  - At 9, 18, 30, 48 months
  - Autism screening at 18 and 24 months
  - Continue to watch for “ADHD” and other behavioral/emotional problems
- Use AAP recommended tools, for example:
  - Ages and Stages, Brief Infant Toddler Social Emotional Assessment, Pediatric Symptom Checklist, Vanderbilt (for ADHD), CHAT (for Autism)
  - For Autism, see Johnson CP, Myers SM Pediatrics, 2007, 120: 1183-1215

Courtesy Jacqueline Sanz, Ph.D. Co-Director, CANDO Program Neuropsychologist
Case continued...
Fenestrated Fontan

IVC to PA anastamososis

Fenestration

Pulm veins
The new normal, part 3
ECG screening for stimulant meds

• Probably not useful
  ▪ Reported causes of death not something a baseline ECG screens for!
  ▪ There may not be an actual increased risk!