

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

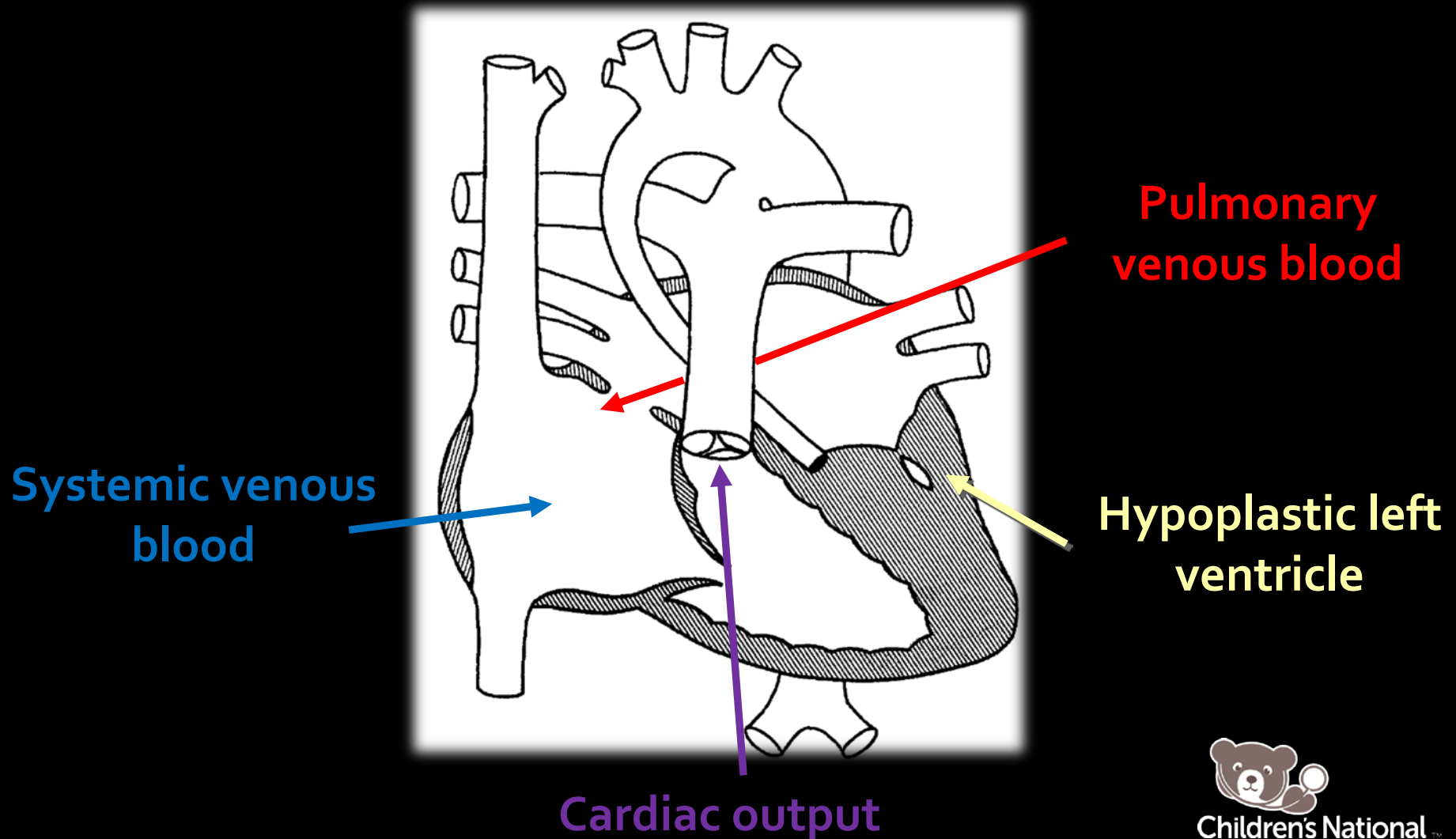
- 6 weeks old
- Full PO, 24 kcal/oz formula
- SpO₂: 75- 80%
- RR 45-70
- Brain MRI
- Meds
 - Aspirin 20 mg daily
 - Lasix 4 mg PO bid
 - Vitamin D
 - Zantac

Now what?

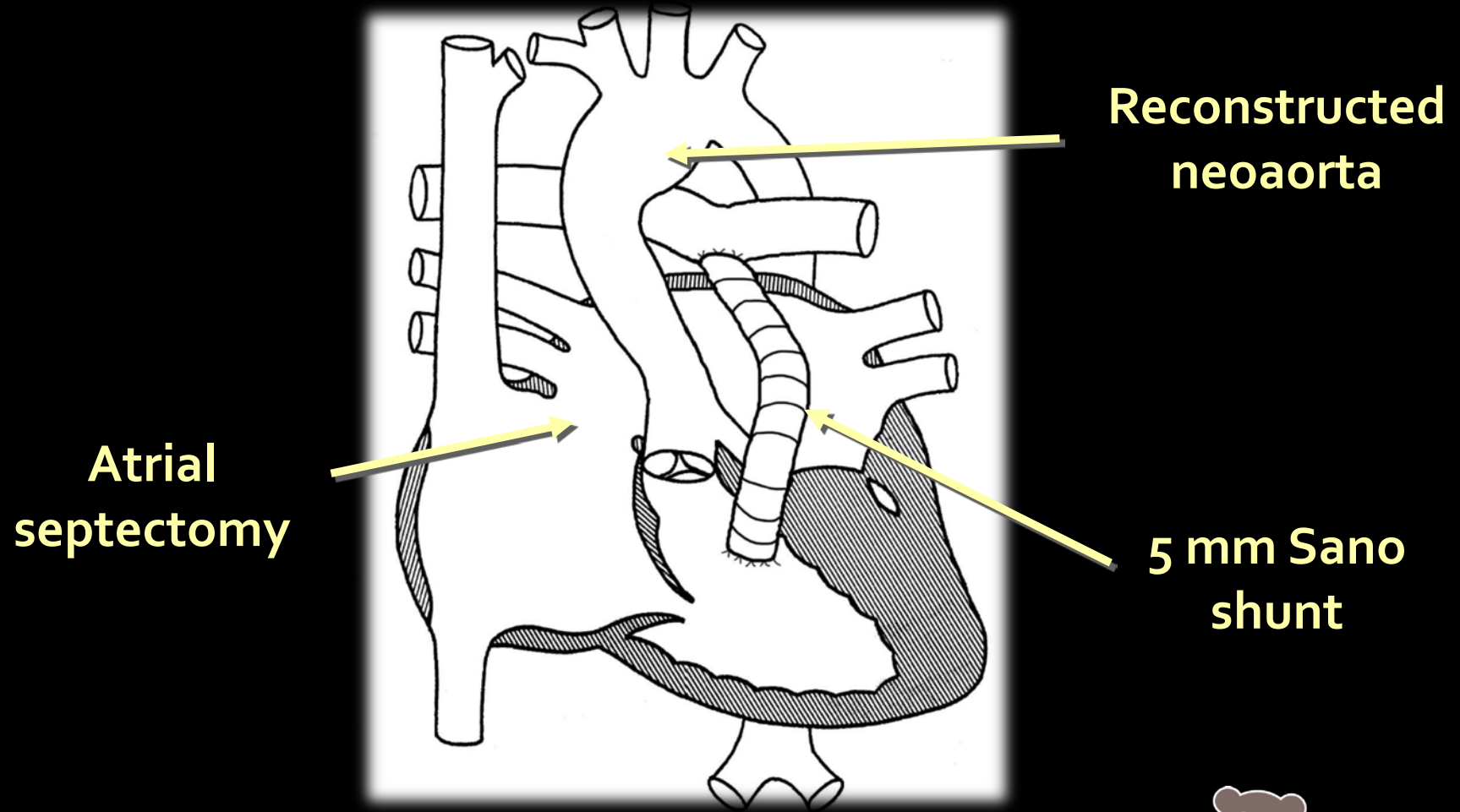
Overview

- 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



HLHS s/p Norwood-Sano



What is normal?

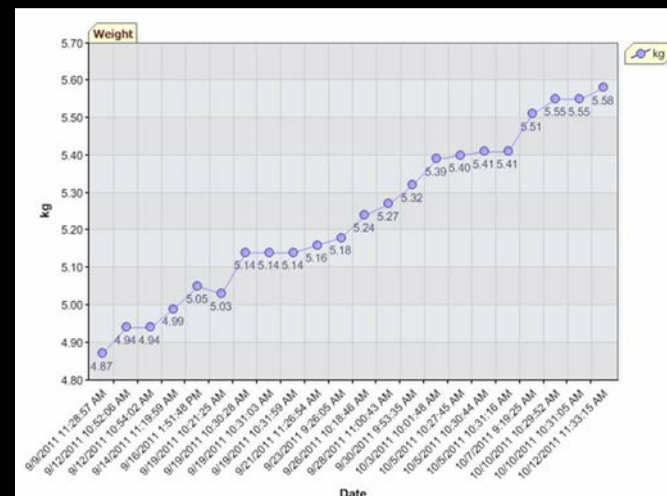
- Vital signs
- Follow-up

Interstage: growth and nutrition

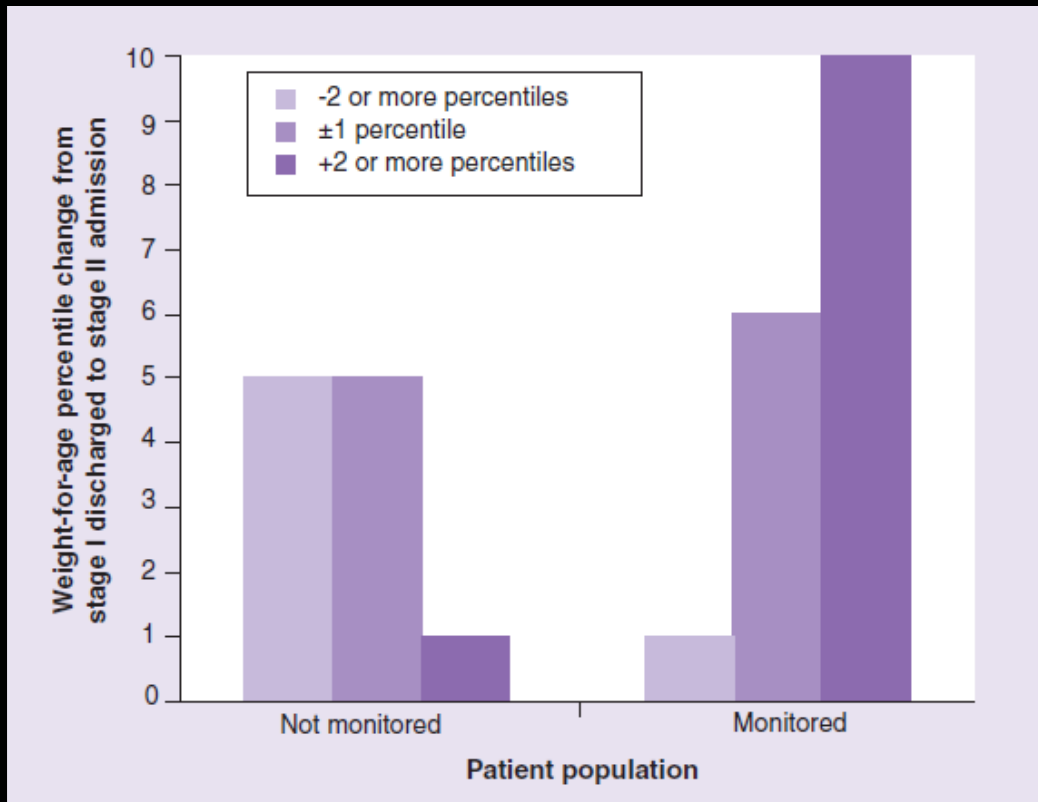
- JCCHD NPC-QIC
- Home monitoring
- Team members at CNMC
 - Who coordinates?

Questions session on 11/1/2010

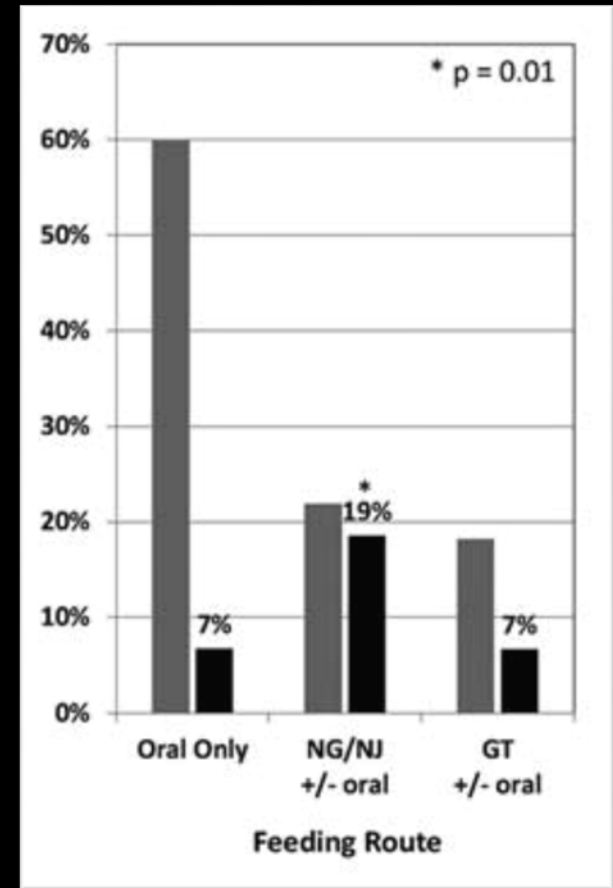
Question	Answer	Answers	Triaged
Is your child sleeping more than usual?	No	Yes No	
How much formula/breast milk has your child eaten in the past 24 hours? (cc's)	710		
Have a great day! Please call if you have any questions or concerns.	-		
Has your child developed a fever (greater than 100.4 degrees)?	No	Yes No	
Is your child sweating more than usual when eating?	No	Yes No	
Is your child taking a longer time to eat than usual?	No	Yes No	
Is your child turning more blue than usual?	No	Yes No	
If you answered yes to any of these questions, please page My Baby's Heart @ 202-259-6600. When you hear the first beeps, enter 2194. After the second beeps, enter your call back number with area code.	-		



Interstage: growth and nutrition



Cross, Future Cardiology, 2012



Cross, Cardiol Young, 2013



Children's National™

Interstage: vaccinations

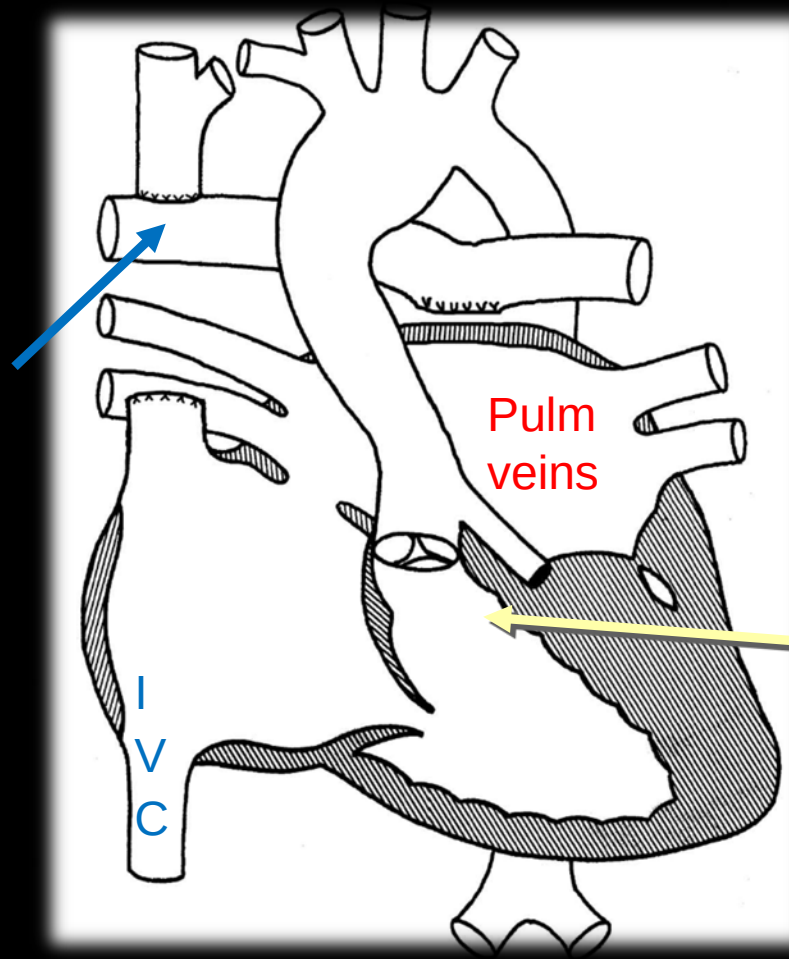
- Diminished response to inactivated vaccines post-CPB
- Delay 6 weeks from surgery (or ECMO)
- Synagis
- Later vaccine issues – aspirin
 - Influenza
 - Varicella

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

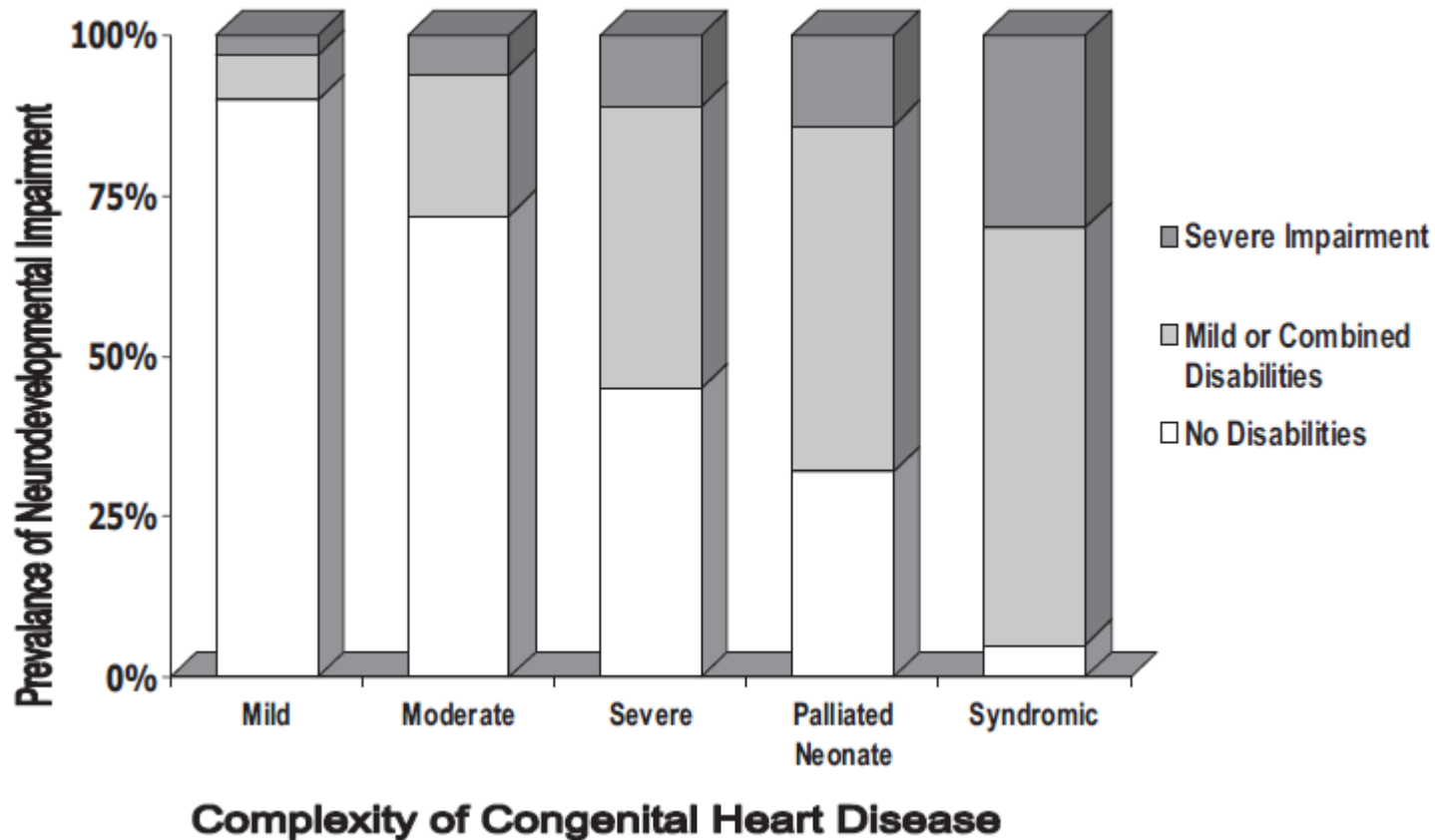
Neurodevelopmental Outcomes in Children With Congenital Heart Disease: Evaluation and Management

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



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

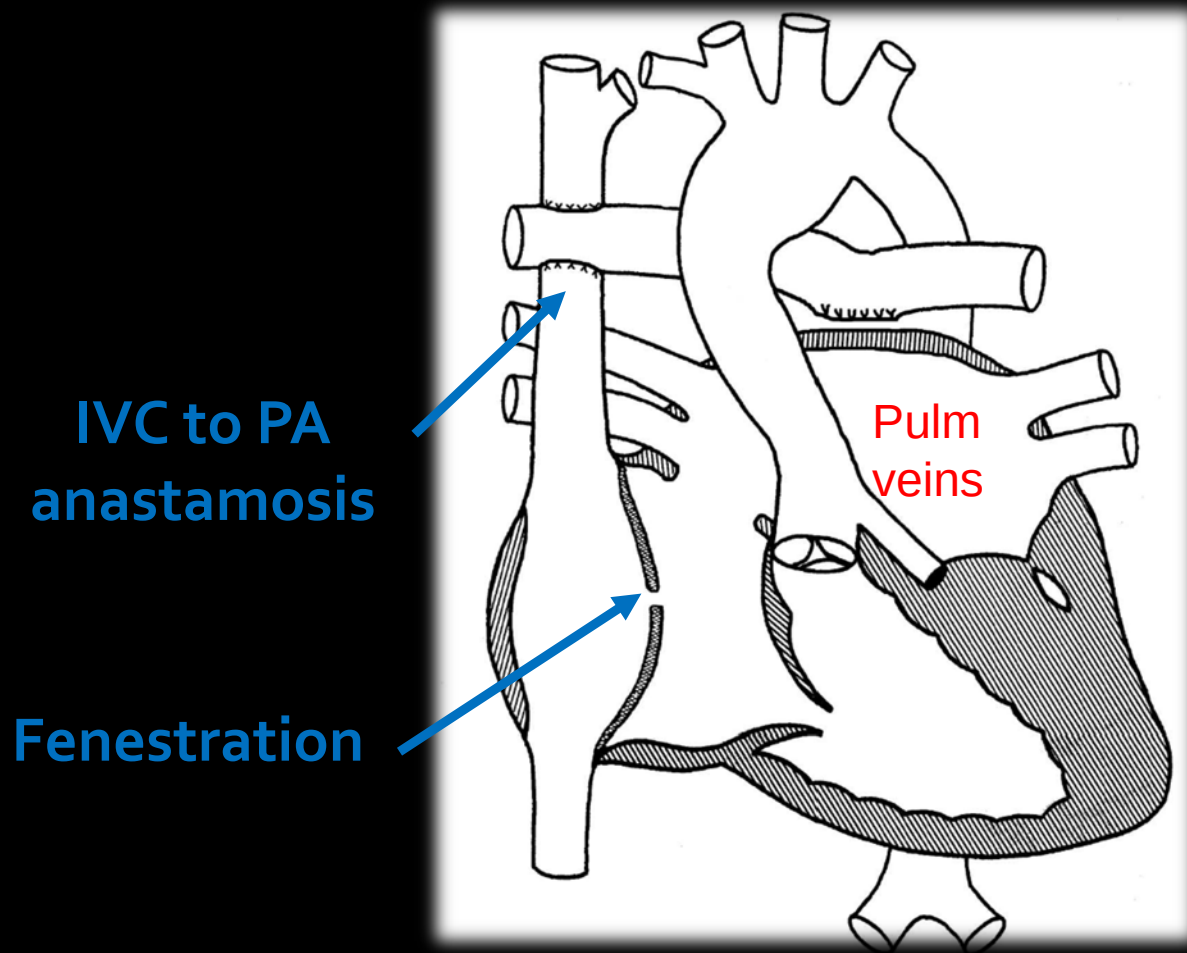
Cardiac neurodevelopment

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

Case continued...

Fenestrated Fontan



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!