

Establishment of the Model for a Specialized Fontan Clinic: Challenges and Benefits

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Why establish the program?



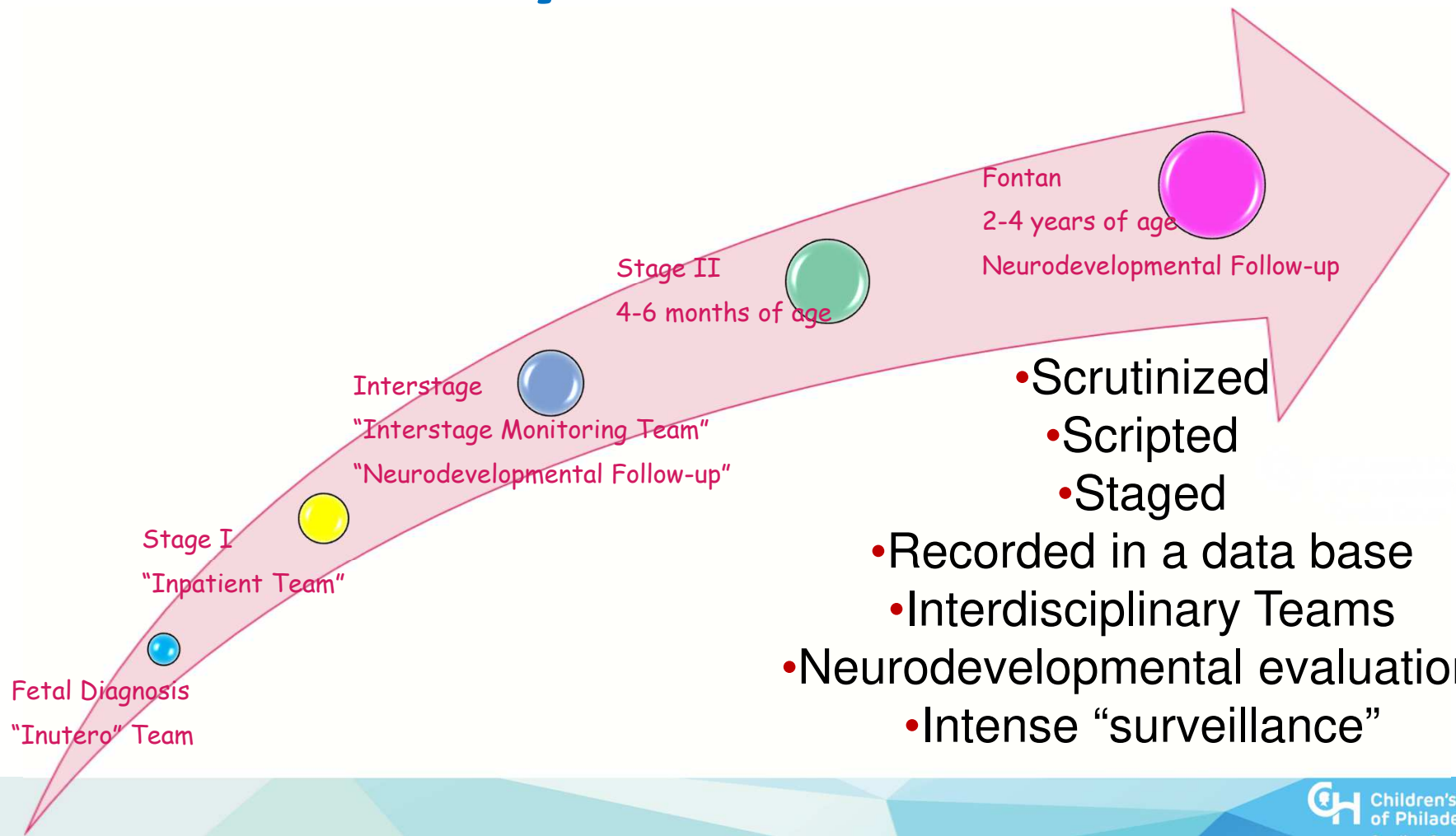
A photograph of a massive, curling ocean wave crashing onto a sandy beach. The wave is a vibrant turquoise color with white foam at the top. The sun is shining brightly in the upper right corner of the frame, creating a lens flare effect. The sky is a deep blue with scattered white clouds. In the distance, a line of dark green trees marks the edge of the beach. A small, dark figure of a person is visible on the sand near the base of the wave.

Growing group of Fontan survivors...

Single Ventricle Survivorship

- Survival to adulthood is expected and numbers are growing
- Multiple centers expertly care for single ventricle patients
- Diagnosis is now a fetal event; as early as 14 weeks gestation
- Neurodevelopmental issues are not uncommon, they are expected
- Interstage monitoring is practiced almost universally and makes a difference!
- Parents and patients are increasingly our partners and should be
- Survivors graduate from high school, go to college, require birth control, get married, get tattoos, become pregnant...
- Anxiety, depression, especially in adolescence and young adulthood, is an issue
- Survivors require life long care that includes adult health care providers but “good” transition to adult care is still a challenge
- Many of our patients doing “well” in childhood...but unravel in early adulthood

“Pathway” to Fontan



A wide-angle photograph of a dirt road stretching into the distance under a blue sky with scattered white clouds. The road is flanked by green grass and a fence line is visible on the left. The text is overlaid in white, bold font.

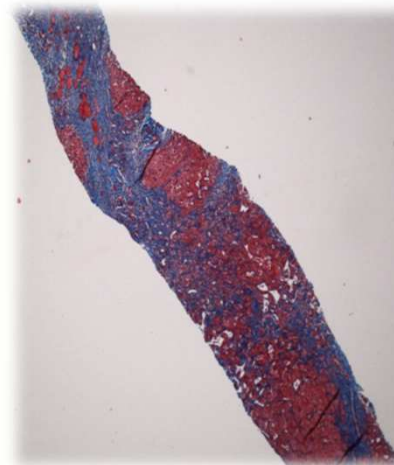
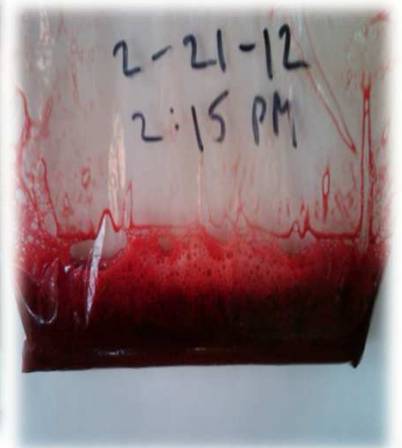
**no defined or agreed upon
“routines”**

no “stages”

no “script”

no “team” of specialists

**when it is least expected and without warning...
even in our “well” Fontans...**

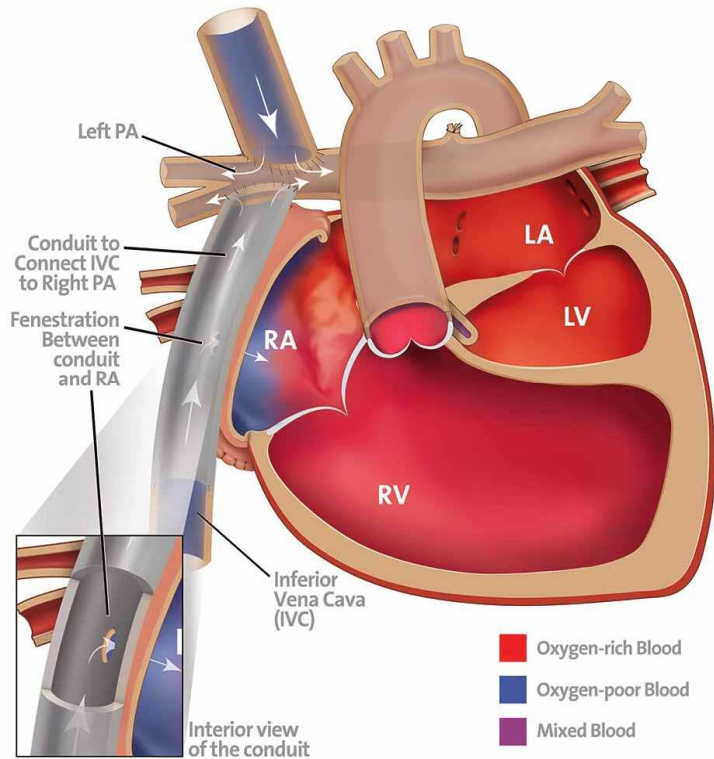


There can be significant challenges

- Lymphatic issues: Protein Losing Enteropathy and Plastic Bronchitis
- Hepatic fibrosis, cirrhosis, carcinoma
- Hemoptysis
- Thromboembolism
- Systolic and Diastolic Dysfunction
- Valve issues
- Rhythm issues
- Immune Issues
- Growth and Bone Issues
- Cognitive and Behavioral Issues
- Mental Health concerns
- Need for transplant...not just the heart...VAD first?
- Need for palliative care



This is not “normal” physiology...ever



- Blood is “routed” to and through lungs...
 - Passive flow, not pumped there, no pulsatility
- Elevation in systemic and central venous pressure
- Impact on ventricular filling and function
- Liver congestion
- Elevated hepatic venous pressure
- Increased lymphatic production but impedance to flow

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

Inconvenient and Disruptive Fontan truths

- Survivors, even those who are “well”, are an “at risk” group.
- Fontan physiology is **NOT** normal and it impacts **multiple** other organ systems both acutely and over time
- Exact mechanism(s) behind complications like plastic bronchitis and PLE have not been well understood thus treatment options thus far have been problematic but lymphatic system derangements are the likely etiology for both
- Risks cannot be screened for by echo, exercise testing, ecg's, and holters alone...not just the heart that is impacted
- We now measure time and age by “years out from the Fontan”
- Risk for comorbidities increases with time from the Fontan

This is the definition of Chronic Illness

The World Health Organization: Chronic Diseases

- Fontans have a health condition that is persistent and long lasting in nature
- Fontans have a *disease* that lasts longer than three months
- It may get slowly worse over time...
- It may lead to death...
- It may cause permanent changes to the body...
- It may impact their quality of life...

Mandate for Chronic Illness Care

“Encourage the coordination of medical care with the development of patient centered medical homes and accountable care organizations”

Robert Wood Johnson Foundation

Chronic Conditions: Making the Case for Ongoing Care(2004)

We have an opportunity to create a medical home for our Fontan patients...



Cardiology
Hepatology
Endocrinology
Immunology
Others:

- Lymphatics
- Pulmonary
- Psychology
- Neurology
- Hematology
- Nephology



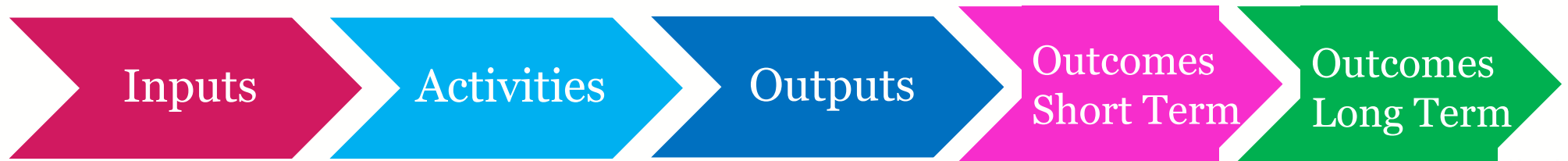
Single Ventricle Survivorship Program (SVSP)
~Started in 2011

- Almost 500 Fontans have been seen in the program since 2011

How did we establish SVSP?

How Did We Get Started?

A Logic Model for SVSP



Personnel,\$,Expertise What will you do What will you produce Short term benefit Long term benefit

Long Term Outcome Goal

- Provide a medical home for single ventricle patients after the Fontan in a multidisciplinary clinic providing comprehensive management leading to improved survival, wellness, and quality of life for Fontan survivors

Short term outcomes

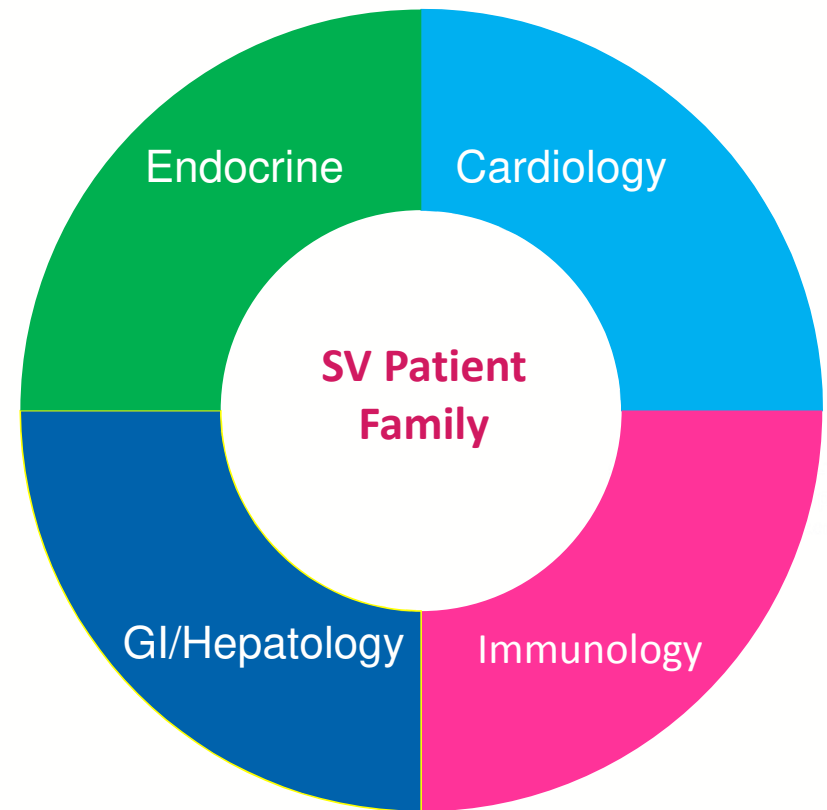
- Surveillance of all Fontan survivors through comprehensive and multidisciplinary evaluation at determined touchpoints in time in a single clinic setting
- Evaluation & management of complications such as protein-losing enteropathy, plastic bronchitis, hemoptysis, liver pathology, and failing Fontan circulation
- Education of the patient and family: the Fontan, the future, the liver, exercise, healthy self care
- Facilitation of transition to adult care
- Coordination of transplant evaluation for “failing” Fontans
- Determine ***appropriate and cost effective*** testing protocols, timing of testing, and intervals for follow-up of patients
- Identify “at risk” patients before symptoms so we can intervene early

Outputs: What Have We Produced?

- Single Ventricle Survivorship Multidisciplinary Clinic
- Establishment of a medical home for Fontan patients
- Database
- Patients returning to CHOP
- New referrals
- Second opinions
- Research
- Increased knowledge of liver, growth, immune effects after Fontan
- Advocacy

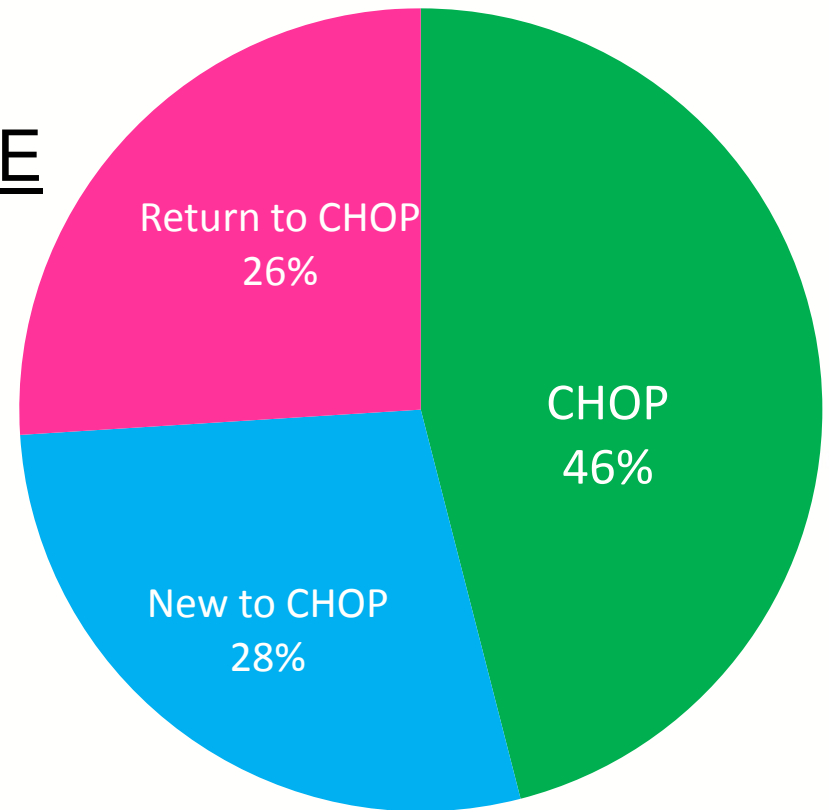
Output: The Multidisciplinary Clinic

- Clinics weekly with cardiology
 - Multidisciplinary clinic monthly; twice monthly if a 5th Thursday
 - 4 specialists, 4-5 patients
- Specialists rotate to the patients
- Coordinator manages flow in clinic
 - Testing before; day or days before
 - Pre clinic team review
 - Post clinic team wrap up
 - Follow-up with family/referring MD



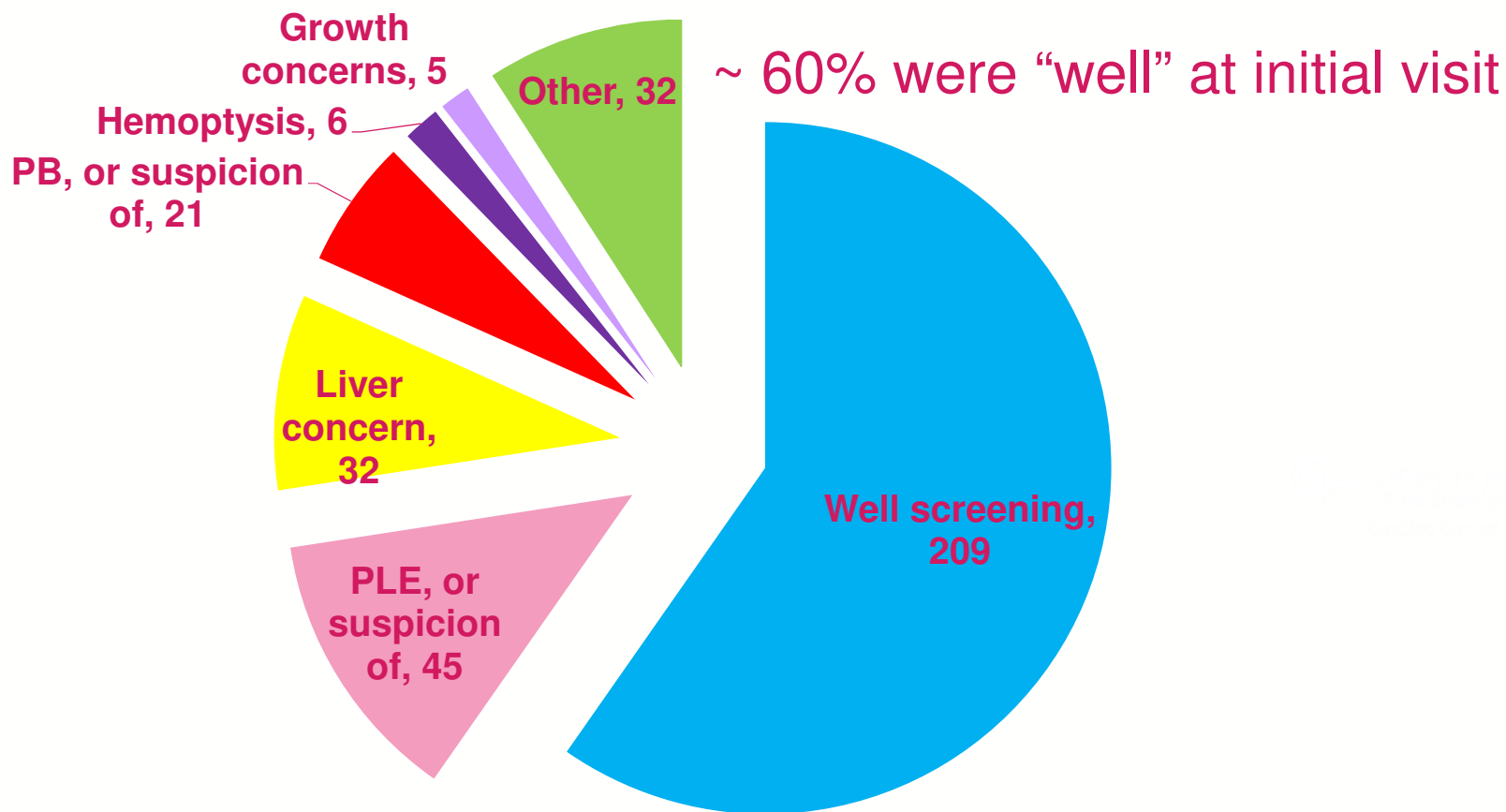
Where are patients from?

- The program DOES NOT REPLACE regular follow-up with cardiologist
- Patient can be seen in the program even if surgery was elsewhere

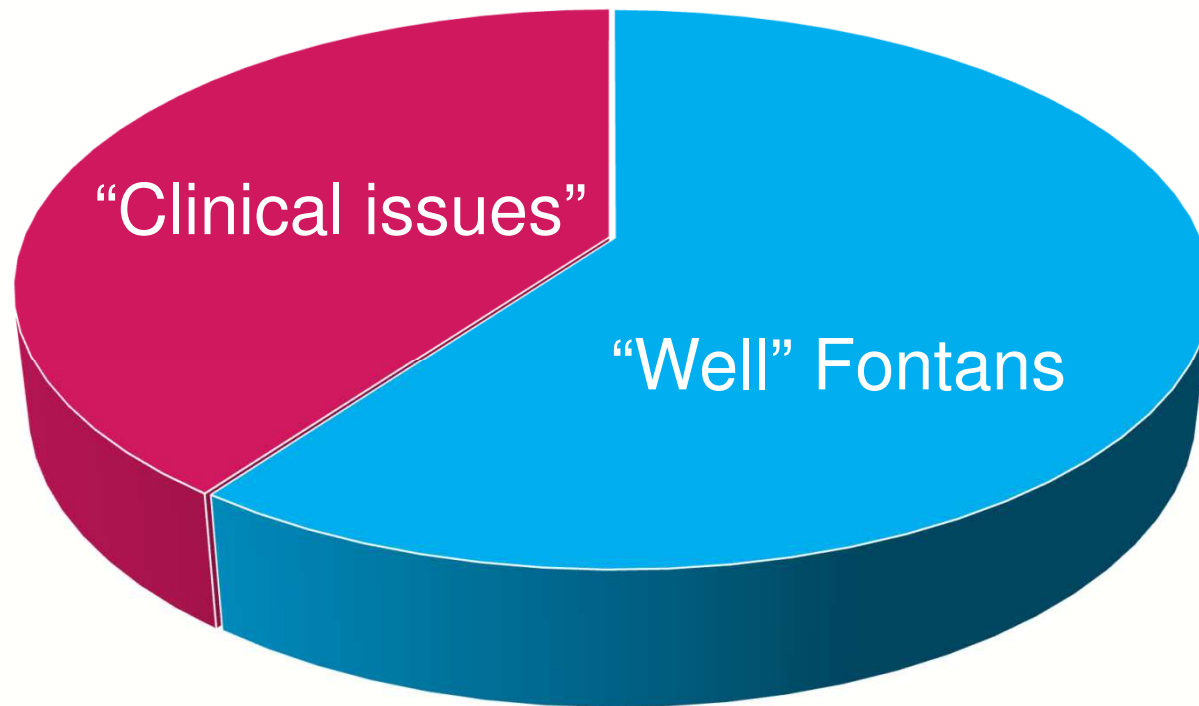


Reason for Evaluation/Indication for Referral to SVSP

Total n= 356 patients



Most patients in SVSP “well”...for now



Activities: What testing are we doing?

- Echo
- Exercise Stress Testing
- Abdominal Ultrasound with Doppler; possible US or MRI elastography based on age
- DEXA scan
- Labs looking at liver, growth, bone health, and immune system
 - BNP, CBC with diff, Comp Metabolic, GGT, PTH, Vitamin D25OH, PT/INR, IGG, Cystatin C
- Cardiac MRI “if” valve leakage, function, cyanosis, branch PA concerns and if can do it unsedated.
- Avoid, if possible, sedation and anesthesia
- Since 2012: Comprehensive evaluation with MRI, Cardiac Cath, percutaneous liver biopsy in patients who are over 10 years out from the Fontan...but not until we see patient in clinic first

Testing is coordinated before the clinic day; results in clinic

Resources needed

- The RIGHT specialists with committed time from their division
- Space
- Financial counselor for insurance issues
- Nurse Practitioner
- Administrative Support
- Data Base Management
- Clinic Coordinator
- Nutrition support
- Support from ECHO, Exercise, US, Dexa, lab staffs
- Funding for salaries. Where will it come from? Institution or philanthropy?
- Institution “buyin” that this is important

Benefits

- Breadth of patients
- Depth of Data
 - GGT, PTH, ALC, IGG, Hgb, LFT's, Medication burden, Anticoagulation, Growth/Bone/Puberty/Exercise capacity/Liver and Spleen US/ Cath and liver biopsy evaluation
- Breadth and Depth give us Context to interpret results, determine research needs, when to intervene and worry and when not to
- Increased knowledge
 - Liver
 - PLE
 - PB
 - Hemoptysis
 - The Lymphatics
- Research produced: Liver, Growth, Immune issues
- Parental and Patient empowerment and education

Unexpected barriers

- Assumed parents would love this...but not all did
- Assumed our colleagues would support this...but not all did
- Assumed we would need to advertise...they came like a tidal wave...
 - If you build it they will come!
- Needed to educate and collaborate with US, IR, Lab
- Needed to educate and get buy in from MA's doing vital signs and the registration desk
- Quickly outgrew resources, space, and clinic time/provider commitment
- Catch up with patients...getting the older child and adolescent in so we can move to the younger patients

Unintended Consequences

- Fear
- Anxiety
- Reactionary Medicine
- Cost

Challenges

- Personnel
 - Need the right providers; HUGE commitment
 - Database Manager
- Billing
 - This is expensive. This is clinical care. We are not conducting research we are caring for patients.
 - Shared billing? or Separate billing for providers
- Insurance
 - Have to have a financial counselor to sort through insurance. Critical to do this. Prior authorization.
- Space
 - Space to see patients; space for providers to discuss patients; space for trainees
- Registration/Medical Assistants
- EPIC
- Data Base Manager
- FUNDING for additional interprofessional involvement: Social work, Nutrition, Psychology
- Balance between research and clinical care

Top 3 Themes for all Multidisciplinary Clinics at CHOP ~January 2018

Challenges

- Staffing – funding/availability
- Communication/coordination
- Clinic flow/efficiency

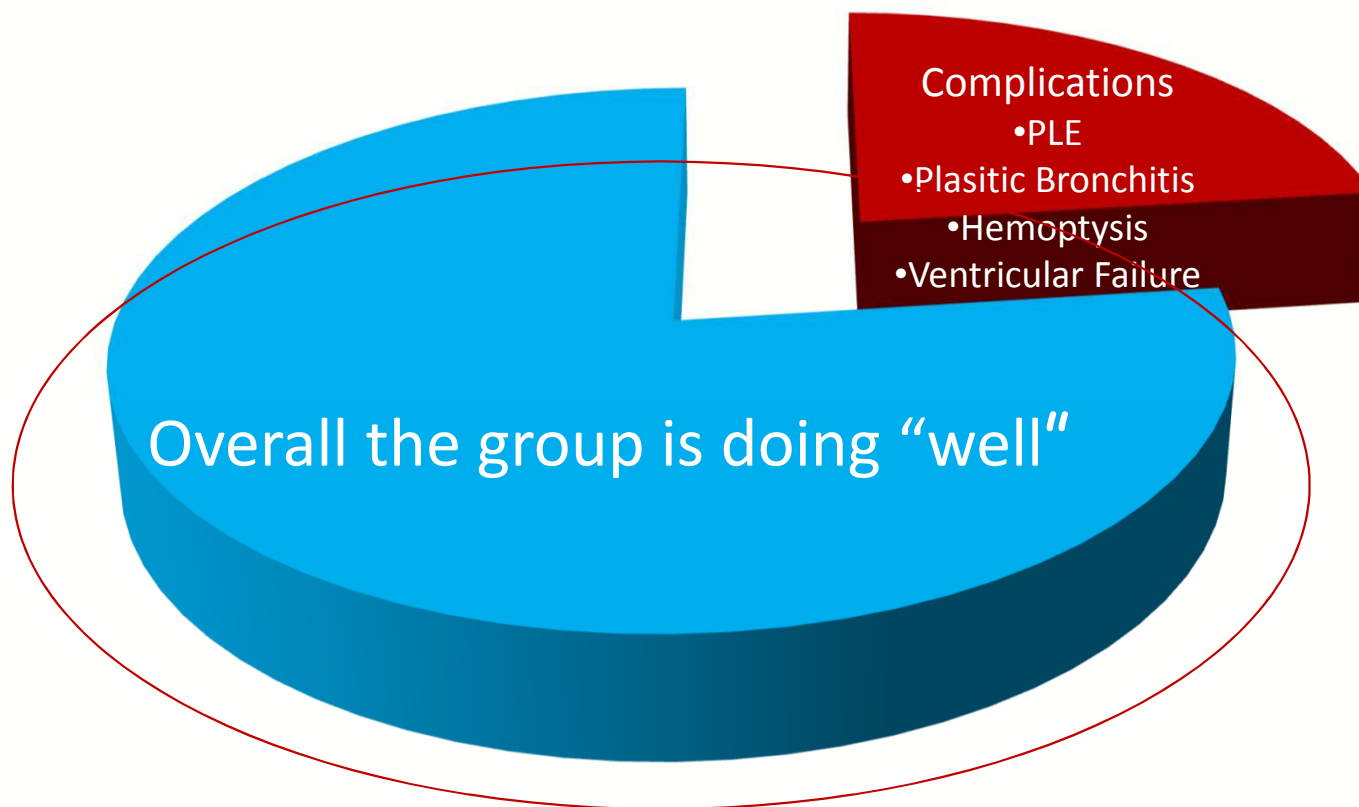
Successes

- Teambuilding/collaboration
- Patient/family support
- Institutional support

Shared Resource Opportunities

- Technological support
- Social work
- Billing/financial counseling

Fontan Survivors in SVSP



Children's Hospital
of Philadelphia
Cardiac Center

We Have Opportunities!!

- Make the Fontan the best it can be for each patient
- Optimize tolerance of this physiology past age 20-30 years
- Identify complications early and try to rescue the failing Fontans
 - PLE, Plastic Bronchitis, Hemoptysis, Function
 - Stabilize, Reverse, and Buy good quality time
- Provide creative, innovative, cost effective care
- Promote a wellness lifestyle
- Encourage healthy diets and intentional physical activity
- Provide education about avoidance of risky behaviors
- Assist patients to embrace their health potential/self care
- Conduct Research
- Improve the scope of our screening; social work, psychology

Time to Pivot?

Children's Hospital
of Philadelphia
Cardiac Center

The *FORWARD* Project

Fontan
Rehabilitation,
Wellness,
Activity and
Resilience
Development
Project



EXERCISE:

- Develop individualized cardiac rehab/exercise strategies
- Lower extremity strengthening
- “personal trainer” prescriptions

MENTAL HEALTH:

- Develop patient and family specific assessments and treatment strategies
- Stress/anxiety reduction
- Improve QOL

NUTRITION:

- Nutritional consultation
- Assess and optimize caloric intake
- Assess and optimize vitamin & essential elements intake

EDUCATION:

- Develop educational modules for patient and family through all stages of care (prenatal, surgery, post-op, childhood, adolescent, adult)

Remember...What Should We Do With a Problem?

“problems” should be looked at as challenges
the bigger the challenge
the greater the opportunity

SVSP CHOP Team

Cardiology: Jack Rychik, MD and David Goldberg, MD

GI-Hepatology: Liz Rand, MD , Henry Lin, MD, Edisio Semeao, MD

Endocrinology/Bone Health: Edna Mancilla, MD

Lymphatics: Yoav Dori, MD and Max Itkin, MD

Pulmonary-Plastic Bronchitis: Sam Goldfarb, MD

Pathology: Pierre Russo, MD

Immunology-PLE: Jennifer Heimall, MD and Kate Sullivan, MD

Heme: Leslie Raffini, MD

Renal: Kevin Meyers, MD

Neurology: Dan Licht, MD

Neuropsychology: Lyla Hampton

IR: Anne Marie Cahill, MD, Marc Keller, MD

Radiology: Kassa Darga, MD

APN/Program Manger: Katie Dodds, CRNP

Clinic Coordinator/Administrative Assistant: Judy Sposato

Financial Counselor: Bryan Phillips

Social Work, Child Life, Nutrition, Staff in US/Dexa/Lab/MRI/IR