

Fontan Outcomes Network Update

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Disclosures:

No financial conflicts

Gratitude for philanthropic support of SVP

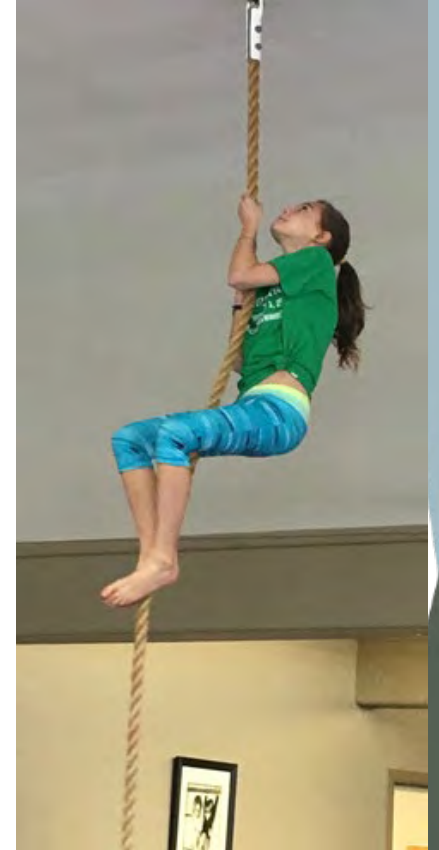
I believe in **healthcare for all** Americans

I believe in building **networks** not walls

I have learned a lot about **living wholeheartedly**
from people born with half a heart

Landscape of Fontan work

- Lots going on!
- US & international collaborations
- Lots of progress AND lots of unknowns
- Lots of “green grass and opportunity” for whole community- > clinicians, patients, and families



Paradigm Shift in Single V Lifelong Care

- **Thrive not just survive**
- **Optimize functional outcomes, rather than treating once failing physiology has already developed**
 - Anticipatory, longitudinal care for early identification and treatment of morbidity
- **Multi-center, collaborative research, QI, and advocacy**







Journey to FON



PHN
Fontan

IFIG, June 2016

FON Design
Aug 2017

*A whole life
with half a heart*



AHA
FALD

AHA Fontan
Care



Australia New Zealand Fontan Registry

1. [Hepatic and renal end-organ damage in the Fontan circulation: A report from the Australian and New Zealand Fontan Registry.](#)
Wilson TG, **d'Udekem** Y, Winlaw DS, Cordina RL, Celermajer DS, Wheaton GR, Bullock A, Gentles TL, Weintraub RG, Justo RN, Grigg LE, Radford DJ, Hardikar W, Cheung M, Cain TM, Rao P, Alexander SI, Ayer J, Verrall C, Du Plessis K, Chapman J, Rice K, Barry J, Zannino D, Iyengar AJ; Australian and New Zealand **Fontan Registry**.
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2. ["Will she live a long happy life?" Parents' concerns for their children with Fontan circulation.](#)
du Plessis K, Peters R, King I, Robertson K, Mackley J, Maree R, Stanley T, Pickford L, Rose B, Orchard M, Stewart H, **d'Udekem** Y.
Int J Cardiol Heart Vasc. 2018 Mar 9;18:65-70. doi: 10.1016/j.ijcha.2018.02.008. eCollection 2018 Mar.
PMID: 29876506 **Free PMC Article**
[Similar articles](#)
3. ["How long will I continue to be normal?" Adults with a Fontan circulation's greatest concerns.](#)
du Plessis K, Peters R, King I, Robertson K, Mackley J, Maree R, Stanley T, Pickford L, Rose B, Orchard M, Stewart H, **d'Udekem** Y.
Int J Cardiol. 2018 Jun 1;260:54-59. doi: 10.1016/j.ijcard.2018.01.098.
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4. [Clinical Outcomes in Adolescents and Adults After the Fontan Procedure.](#)
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Poh CL, **d'Udekem** Y.
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6. [Twenty-Five Year Outcomes of the Lateral Tunnel Fontan Procedure.](#)
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7. [Involvement of patients and parents in research undertaken by the Australian and New Zealand Fontan Registry.](#)
d'Udekem Y, Forsdick V, du Plessis K.
Cardiol Young. 2018 Apr;28(4):517-521. doi: 10.1017/S1047951117001494. Epub 2017 Aug 17.
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8. [Hospital Inpatient Costs for Single Ventricle Patients Surviving the Fontan Procedure.](#)
Huang L, Schilling C, Dalziel KM, Xie S, Celermajer DS, McNeil JJ, Winlaw D, Hornung TS, Radford DJ, Grigg LE, Bullock A, Wheaton GR, Justo RN, Blake J, Bishop R, Du Plessis K, **d'Udekem** Y.
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9. [Hospital costs and cost implications of co-morbid conditions for patients with single ventricle in the period through to Fontan completion.](#)
Huang L, Dalziel KM, Schilling C, Celermajer DS, McNeil JJ, Winlaw D, Gentles T, Radford DJ, Cheung M, Bullock A, Wheaton GR, Justo RN, Selbie LA, Forsdick V, Du Plessis K, **d'Udekem** Y.
Int J Cardiol. 2017 Aug 1;240:178-182. doi: 10.1016/j.ijcard.2017.04.056. Epub 2017 Apr 20.
PMID: 28456482
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10. [Evolution of Left Ventricular Size in Late Survivors of Surgery for Hypoplastic Left Heart Syndrome.](#)
Bjurbom M, Iyengar AJ, Moenkemeyer F, Konstantinov IE, Brizard CP, **d'Udekem** Y.
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12. [Common atrioventricular valve failure during single ventricle palliation†.](#)
King G, Gentles TL, Winlaw DS, Cordina R, Bullock A, Grigg LE, Alphonso N, Radford DJ, Zannino D, Buratto E, **d'Udekem** Y.
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STATE OF THE ART CLINICAL CARE OF THE FONTAN CIRCULATION –

A MULTI-INSTITUTIONAL SURVEY OF PRACTICE VARIABILITY IN IDENTIFIED FONTAN PROGRAMS ACROSS THE U.S.

DiMaria, M et. al. Pediatric Cardiology 2018

Affiliated with
 University of Colorado
Anschutz Medical Campus

 **Stanford**
Children's Health

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Surveillance:

- Very little scientific data to tell us:
 - What to check
 - How often
 - What to do when we find a problem
- Need collaboration to redefine quality outcomes

- *Younoszai, A., et. al., World Congress of Pediatric Cardiology 2017*

- *DiMaria, M et. al. Pediatric Cardiology 2018*

-

American Heart Association (AHA)

- **Scientific Statement on the Care of the Fontan**
 - Compendium of current knowledge about Fontan circulation- 20 international expert authors; addresses both child and adult
 - Embargo
 - "Coming Soon"for the past 2 years
- **Concept of Surveillance Matrix by domains & age**

Surveillance Matrix: Domains and Ages

	School Entry (4-5 y)	Early School Age (6-8y)	Childhood (9-12y)	Early Teen (13-15 y)	Late Teen (16-18)
Cardiac- Imaging, CPET, rhythm					
Neurodev Hepatic Lymphatic Endo Renal					
Nutrition Care Coordination					
Resiliency Transition					

- **Fontan Udenafil Exercise Longitudinal Trial (FUEL)**

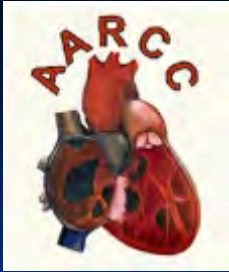
- What: A six-month randomized, placebo control trial in adolescents after Fontan
- Outcomes: Exercise, ventricular function, vascular function
- Who: 400 subjects at 30 sites in the United States, Canada and South Korea
- Status: *Recruitment Complete!*

- **FUEL Open Label Extension (FUEL OLE)**

- What: A 12-24 month safety trial for new subjects or FUEL participants
- Outcomes: Safety, efficacy (exercise, ventricular function, vascular function)
- Who: 300 subjects at 28 sites in the United States and Canada
- Status: *Recruitment ongoing*

- **FUEL Fontan Associated Liver Disease Ancillary Study (FUEL FALD)**

- What: An evaluation of the impact of udenafil on liver stiffness
- Outcomes: MR and ultrasound elastography and biomarkers of liver fibrosis
- Who: Up to 100 subjects who are also participating in FUEL OLE
- Status: *Recruitment ongoing*



Total of eight grants submitted:

1. **The Systemic Right Ventricle: Risks and Outcomes of Congestive Heart Failure.” Craig Broberg and TGA subgroup, AHA**
2. **Cardiovascular Outcomes of Pregnancy in Turner Syndrome: A Multi-Center Retrospective Case-Control Study. Jasmine Grewal and pregnancy subgroup, University of British Columbia**
3. **The additive effects of inspiratory muscle and skeletal muscle training in the Fontan population, Anitha John, Salil Ginde, Fred Wu, Elisa Bradley and the Fontan subgroup, PHN**
4. **A Multi-Institutional Neurocognitive Discovery Study (MINDS) in ACHD, Scott Cohen, Ali Zaidi and the neurodev. subgroup, PHN**
5. **Predictive Value of Hepatic Fibrosis Scores in Patients with Fontan Circulation. Fred Wu and Fontan subgroup, CHF**
6. **Fontan Outcomes Study To Improve Transplant Experience and Results: The FOSTER Study, Matt Lewis, CHF**
7. **Characterizing Hepatic Fibrosis Scores in Patients with Fontan Circulation. Fred Wu and Fontan subgroup, Saving Tiny Hearts**
8. **Engaging the Fontan population to improve Physical Activity Participation. Anitha John and the Fontan subgroup, PCORI**



Stanford Single Ventricle Scientific Summit

— April 29-30, 2019 —

Bringing together distinguished
scientists, bioengineers, and clinicians
from around the world to focus on the
issue of preservation of ventricular
function in single ventricle patients



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FONTAN OUTCOMES NETWORK UPDATE

*Carole Lannon, Diane Pickles, Jack Rychik,
Gail Wright*

*NPCQIC Spring Learning Session:
May 4, 2019*

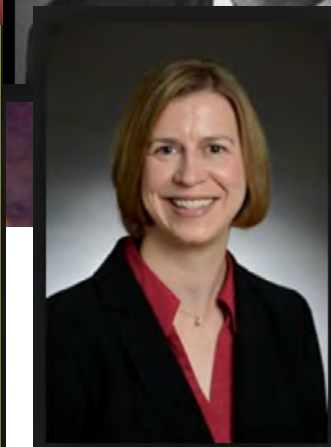
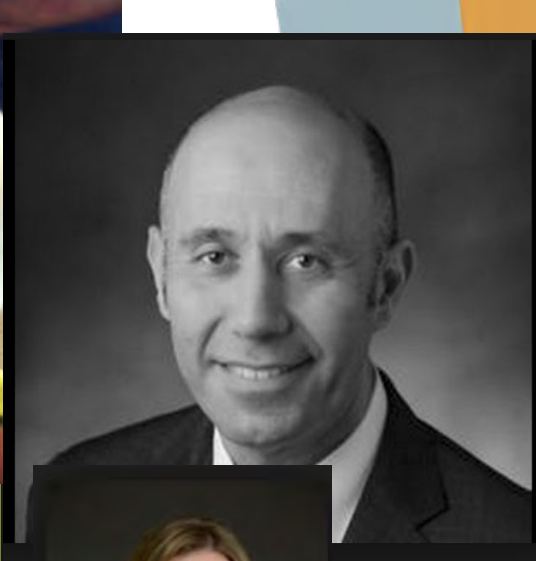
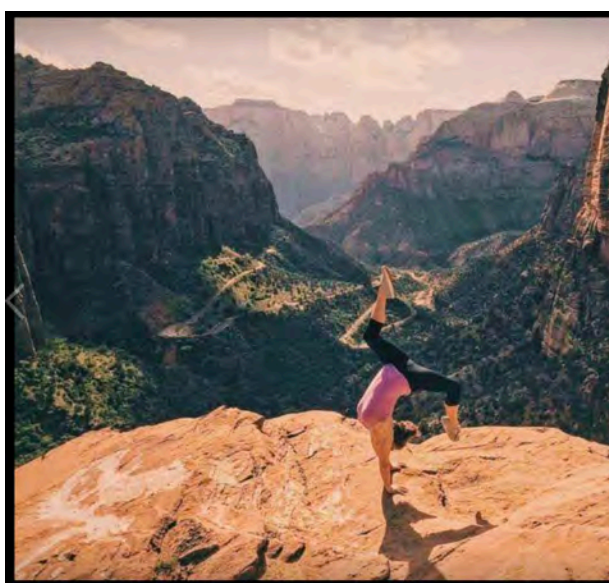
WHY: *THE VISION*

It's not good enough....

- To enable our single ventricle patients to survive but not ensure that they thrive
- To wait for failed physiology and then treat the failure rather than work to prevent the failures
- To accept neurodevelopmental deficits and social emotional problems as part of the disease
- To live with the fear and uncertainty that patients and families currently face each day



FONTAN OUTCOMES NETWORK



Fontan Outcomes Network:

- ***Our vision:*** to dramatically improve outcomes for individuals with Fontan physiology
- ***Our mission:*** to optimize longevity and quality of life for individuals with Fontan physiology and **their families** *by improving their physical health and functioning, neurodevelopment, and emotional health and resilience.*
 - *We will accomplish this through*
 - *Development of a Learning Health System co-produced with patients, parents, clinicians and researchers*
 - *Dissemination of “best practices” among clinical sites*
 - *Discovery of new therapies*

Quality of Life

Better quality of life for patients, siblings and caregivers

Longevity

Longer lives for patients

Resilience & Emotional Health

- Good quality of life
- Effective & healthy coping strategies
- Good social connections and support
- Minimal/manageable psychological distress

Neurodevelopment

- Increased life skills and executive functioning
- Advancing to college or career/vocational training and other key milestones
- Increases in adaptive functioning measures

Physical Health & Functioning

- Increased survival without transplant
- Increased/maintenance of exercise capacity
- Decreased major morbidities

You only see obstacles when you take your eyes off the goal.

WHAT: CURRENT APPROACH & STATUS

Key Driver Diagram: Fontan Outcomes Network

Global Aim:

Optimize longevity & QoL of individuals with Fontan circulation & optimize QOL of their families

Outcome AIMS:

Quality of Life

Longevity

← **Optimize Resilience & Emotional Health**

← **Optimize Neurodevelopment**

← **Optimize health and wellness by preserving functional status**

Fontan Outcomes Network

System Drivers

System Driver Measures

Outcome AIM(s)

QOL

- Increase quality of life (scored by validated tool) of patients, siblings and caregivers

Longevity:

- Increase average length of life

Optimize health and wellness of individuals with Fontan circulation by preserving functional status

Achieve ongoing developmental surveillance including early identification and interventions to support neurodevelopmental needs, so that each individual meets his / her full potential*

* full potential= success in family, school, work, and relationships

Emotional Health & Resilience of Individuals with Fontan Circulation and their Families

- Increase in % of patients who maintain exercise capacity at 90% of initial maxvO2 (usually obtained at 10-12 years of age)
- Increase in % of patients who survive without transplant
- Decrease in % of patients who experience (?1 component) of Major Morbidities Bundle

- Improved life skills / executive functioning consistent with individual's abilities:
 - Increase % of individuals graduating high school and advancing to college or career/vocational training
 - Increase score on standard measure of adaptive functioning (ABAS-3 selected items) in all domains

- Increase % of individual and family members who report good QOL
- Increase in the % of individuals and family members who report they have effective and healthy coping strategies
- Increase in the % of individuals and family members who report good social connections/ support
- Increase the % of individuals who report minimal/manageable psychological distress

Fontan Outcomes Network: guided by data

➤ Registry

- Fontan patients: ALL Ages (across lifespan, not just pediatric)
- Enrollment: at time of Fontan or 1st clinic visit after registry live
- Start with consensus-driven, prioritized, core data
- Ongoing collection of key data elements
 - Across key driver domains: Physical health and functioning, resilience and emotional health, neurodevelopment
 - Clinic visits and prompt for annual visit follow up
 - Patient/parent reported outcomes

Fontan Outcomes Network: “Co-production”

➤ Network

- Collaboration among patients, parents, multidisciplinary clinicians, and researchers informs prioritization of outcomes
- Request to data vendor will include incorporation of existing data from other CNU registries
- Sustainable and scalable infrastructure
- Bridging with ACHD, Heart Failure, community cardiologists, and other CNU registries

HOW & WHEN:
WE'RE GETTING THERE!
& *NEXT STEPS*

Design Recap- Accomplishments to Date:

- ✓ Aligned on Vision, Mission and Governance
- ✓ Developed and refined global aims and theory/approach for improvement
- ✓ Obtained startup funding
- ✓ Developed and refined outcome and process measures
- ✓ Established data workgroup with reps to synthesize efforts from all 3 workgroups
- ✓ Prioritized/defined data elements for registry forms
- ✓ Drafted forms for workgroups to review
- ✓ Drafted *Central* IRB protocol

Where We Are Headed

	Apr – Jun 19	Jun – Sep 19	Oct – Dec 19
Meetings	<ul style="list-style-type: none">➤ NPC-QIC Virtual Learning Session (5/3, 5/4)		<ul style="list-style-type: none">➤ NPC-QIC Learning Session (11/2, 11/3)
Data Elements and Registry	<ul style="list-style-type: none">➤ Workgroups Finalize DRAFTs of Data Forms	<ul style="list-style-type: none">➤ Pilot Care Centers & families test paper forms➤ Compile feedback from Centers & modify forms➤ Vendor Selection Process	<ul style="list-style-type: none">➤ Contract with vendor to begin registry build
IRB Protocol/Legal	<ul style="list-style-type: none">➤ Refine IRB Draft	<ul style="list-style-type: none">➤ Update IRB Protocol with Vendor when selected➤ Draft Data Use Agreement or BAA	<ul style="list-style-type: none">➤ Initiate DUA/BAA/IRB approval at FON Participating Care Centers
Design Workgroups	<ul style="list-style-type: none">➤ Operational definition for outcomes & process measures		



FONTAN OUTCOMES NETWORK

Launch 2020!

Fontan Outcomes Network: Optimizing Outcomes

➤ *Envision the Future:*

- ✓ Have data on over 1000 Fontan pts in 2-3 years
- ✓ Beginning to reliably track outcomes locally & nationally
- ✓ Can start to understand the individual trajectory
- ✓ Building foundation for data driven care strategies and clinical trials
- ✓ Evidence-informed advocacy for mental health & school services

Fontan Outcomes Network: *A Paradigm Shift in Single V Lifelong Care*

- **Thrive not just survive**
- **Optimize functional outcomes**
 - Anticipatory, longitudinal care for early identification and treatment of morbidity
- **Multi-center, collaborative research, QI, and advocacy**

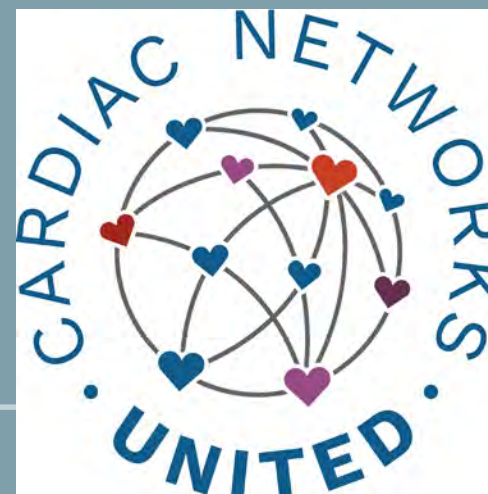
How You Can Engage with this Work

➤ **Organize teams/Engage stakeholders:**

- Consider clinicians, patients and parents who would be a part of the FON team at your center
- Share with stakeholders that FON is coming soon (patients & parents, clinician colleagues, Heart Center leadership)

➤ **Receive periodic FON email updates-** email info@npcqic.org to be included

➤ **Encourage teen/young adult patients** to participate in patient run Fontan session in November (Alicia Wilmoth, Tom Glenn, Meg Roswick Didier)



**ICAN'T,
BUT
WE CAN.**

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NATIONAL PEDIATRIC CARDIOLOGY
Quality Improvement Collaborative