Decades of Heart: Partnering with Families for Lifelong Pediatric Cardiac Care

Derek Human
Parent & Grandparent
Executive Medical Director

What to expect

Appreciate	Appreciate the advancements in pediatric cardiology and how they have shaped care over the decades.
Understand	Understand the crucial role families play in the treatment and long-term well-being of children with heart conditions.
Receive	Receive practical advice on fostering strong, enduring partnerships that ensure the best outcomes for children with cardiac conditions.

What are you getting?

- A history – my journey in Cardiology
- The changing aspects of Cardiac Care
- Caring about the patient & family
- Learning from my patients
- Communicating with each other

Sixty years ago: Africa to Oxford

40 years ago, a cardiology trainee in Cape Town

- What tools did we have?

  - Chest X-ray
  - ECG
  - Clinical Acumen

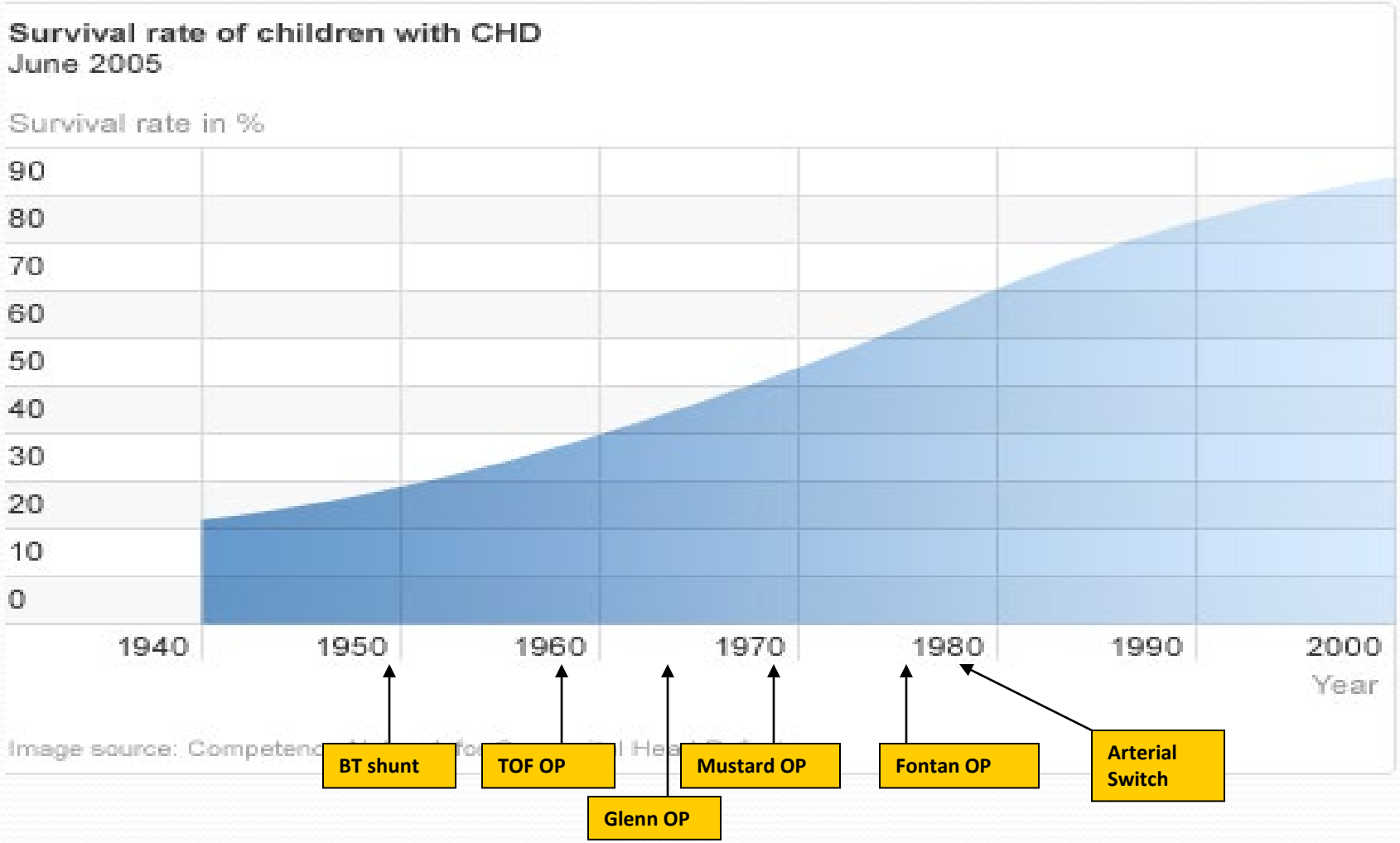
  - Cardiac Catheterisation

Long ago and far away:

Red Cross WM Children's Hospital

Groote Schuur Hospital

Survival Rates of Children with CHD



http://www.kompetenznetz-ahf.de/en/congenital-heart-defects/

Tools which have changed the Outcome

Pre-natal and early diagnosis

Non-invasive Imaging

- Echocardiography: angiography rarely needed pre-op
- Cross-sectional imaging: CT and MR

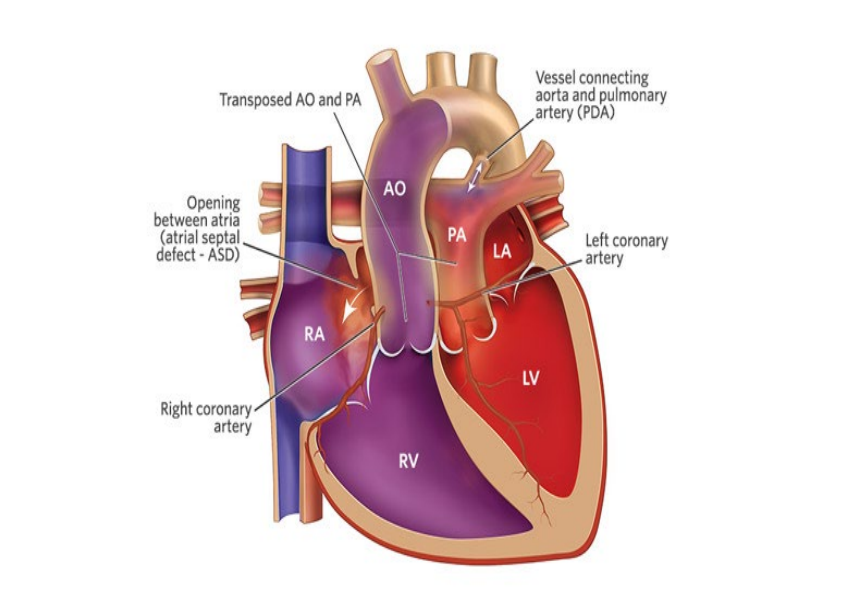
Neonatal surgical techniques

- Cardiopulmonary bypass strategy
- Cardiac Anesthesia: early extubation
- CICU management
  - Prostaglandin, Nitric Oxide, Milrinone, Dexamedetomidine

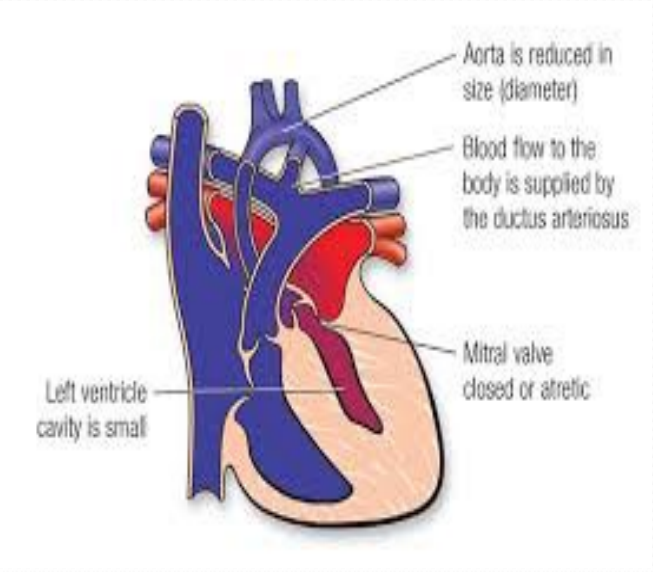
The changing aspects of Cardiac Care

A Tale of Two Conditions

Transposition



Hypoplastic L Heart



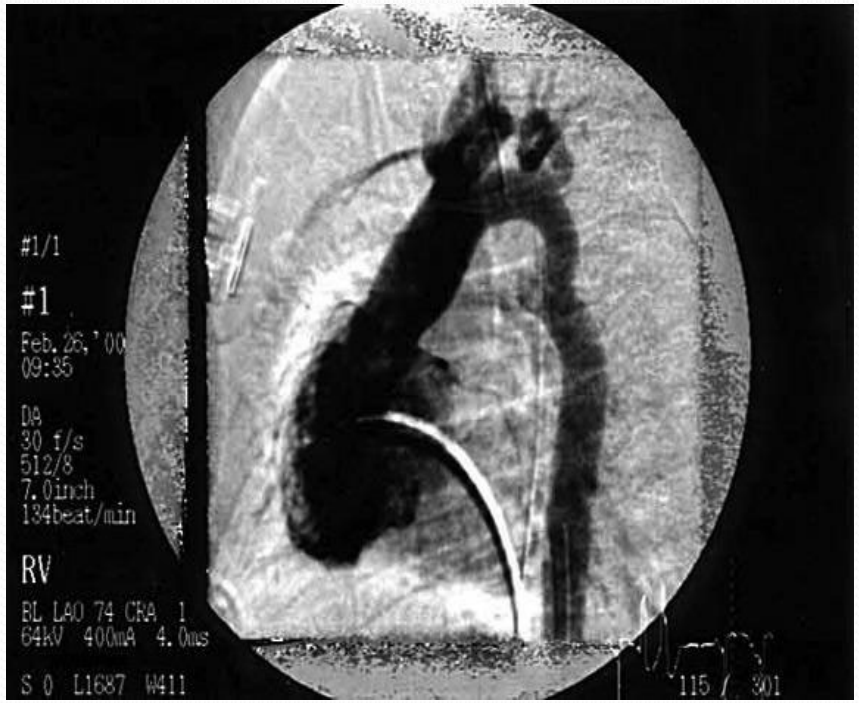
Transposition circa 1980

Acutely ill infant

- Hypoxic and acidotic

Urgent cardiac catheterization

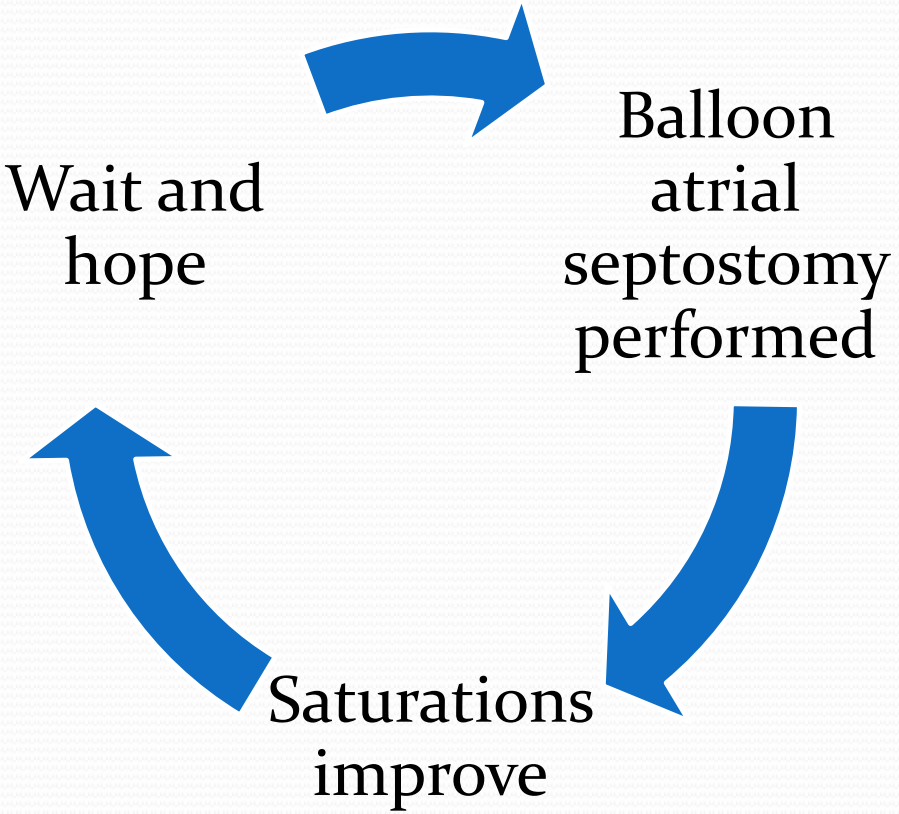
Intubate & ventilate

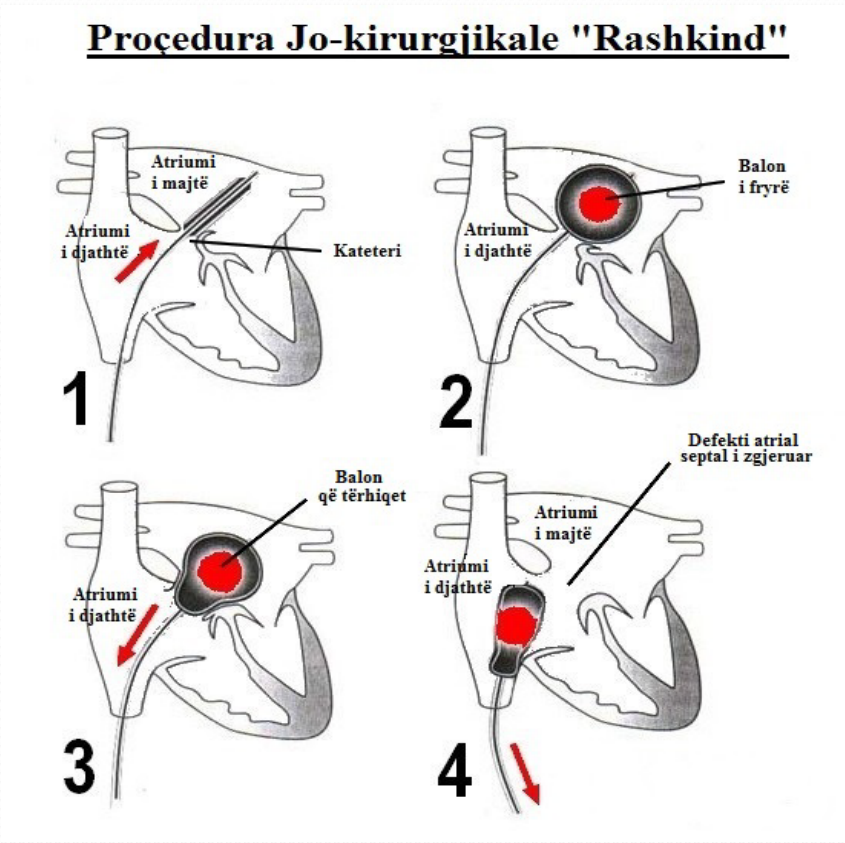


The cardiac cath enables the first minimally invasive procedure because…

- “ A technique for producing an atrial septal defect without thoracotomy or anesthesia is presented. It can be performed rapidly in any cardiac catheterization laboratory”….1966

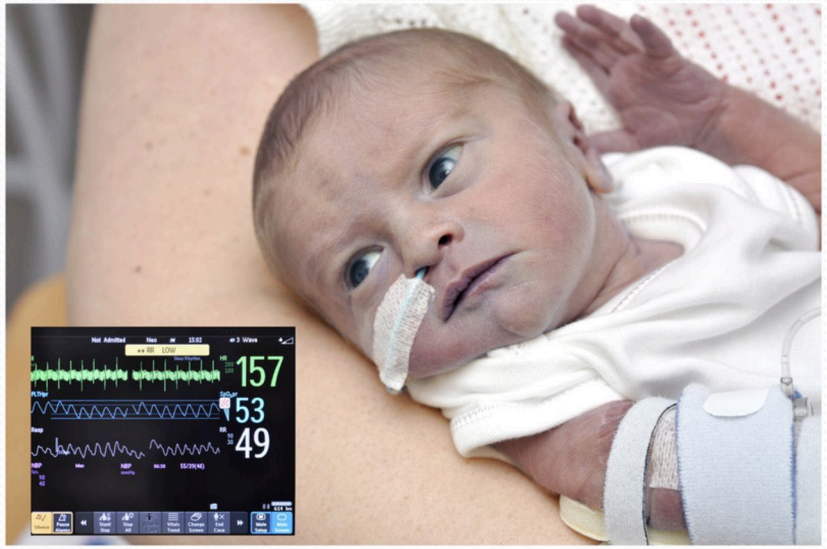
Rashkind BAS: avoiding need for thoracotomy





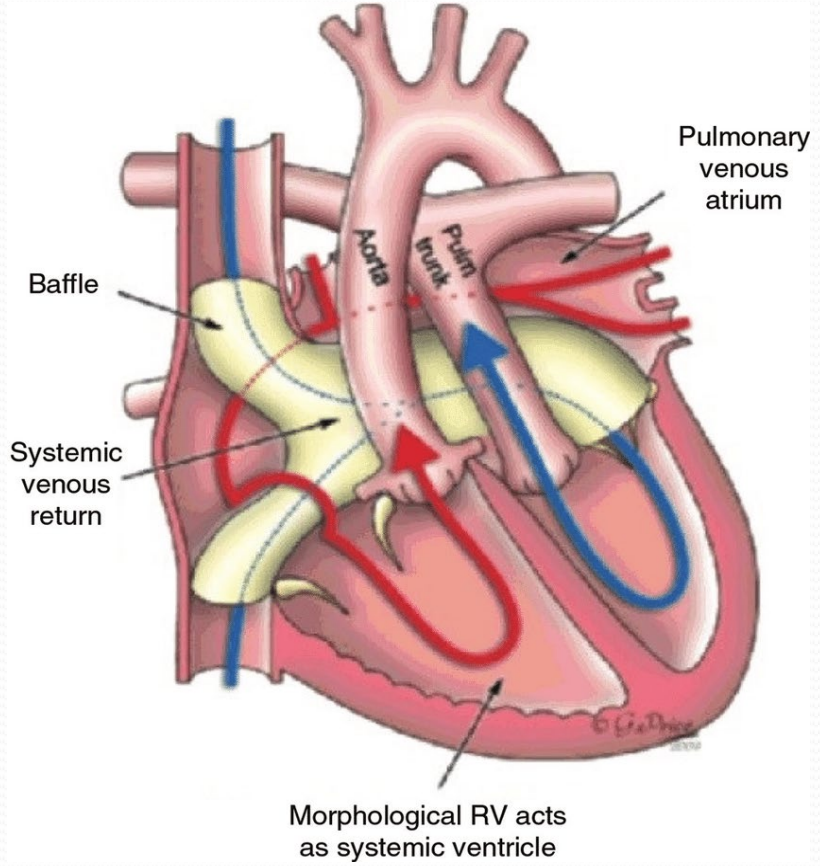
Transposition circa 1980

- Septostomy done
- Neonatal open heart not done
- Repair at 3 months…
- N=188 admissions to GLH, Auckland NZ
  - 3 died before BAS
  - 1 persisting hypoxia
  - 6 late cerebral deaths
  - 1 procedural death
  - 3 died with infections
- 10%-20% Mortality rate awaiting the repair



© OMI 00278894.jpg Date Taken: 03/08/2015

Senning or Mustard: “Atrial Switch”



550 Mustard operations: 1963-1998

Results

From 1963 to 1998, we performed 550 Mustard operations. The arterial switch replaced the Mustard operation for patients with an associated VSD after 1978 and for neonates with isolated TGA, after October 1988.

Survival of the first 547 patients is shown in Figure I. Both early and late survival after a Mustard operation is better for patients with isolated TGA[10-11] compared with survival for children with TGA and associated lesions, usually a VSD[12] (Fig II). The association of poor outcome with associated VSD persists into adulthood.

Fig I. Actuarial survival (Kaplan-Meier) in 547 patients after a Mustard operation at the University of Toronto. Survival 25 years after operation is 64% n = 25, the number of patients entering each time period indicated on the horizontal axis.

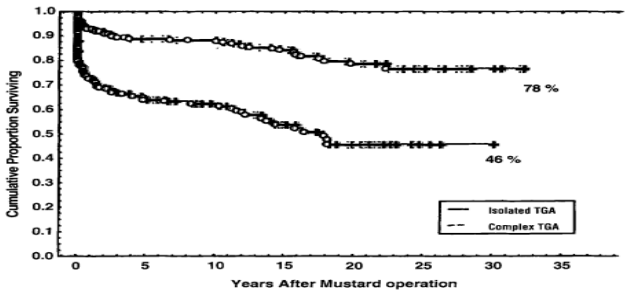


Fig II. Survival of patients with isolated transposition after a Mustard operation (n = 375) is 78% at 25 years. For patients with complex transposition (n = 172), early and late survival is significantly lower (P .001).

Mustard Repair: ongoing hazards

Mechanical	• Baffle stenosis and obstruction
Functional	• Systemic (right ventricle) Dysfunction • Tricuspid valve insufficiency
Electrical	• Atrial Tachycardias • Sinus node dysfunction
Psychosocial	• Developmental Delay • Stroke

Atrial Switch Repairs:

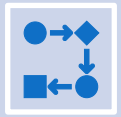
A unique disease complex

Creating survivors with unique Cardiac problems

These Adult CHD complications will never be easy

- Small numbers
- Unusual anatomy
- Outside guidelines

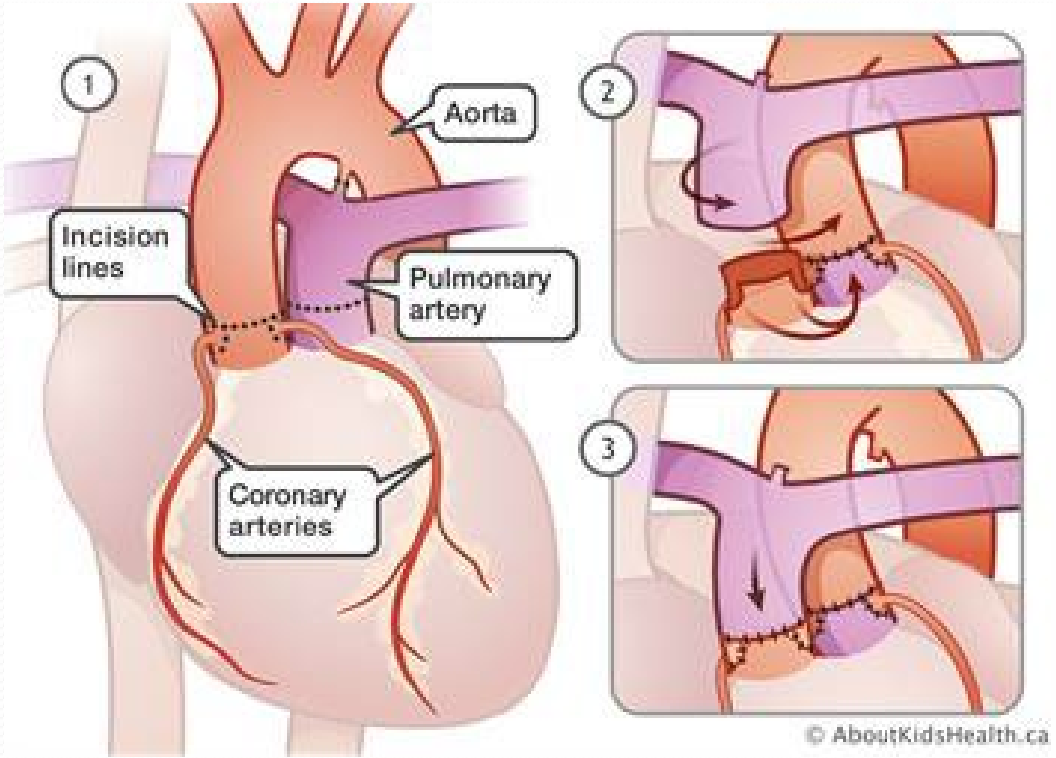
Survival 85% @ 5y…..65% @ 30y



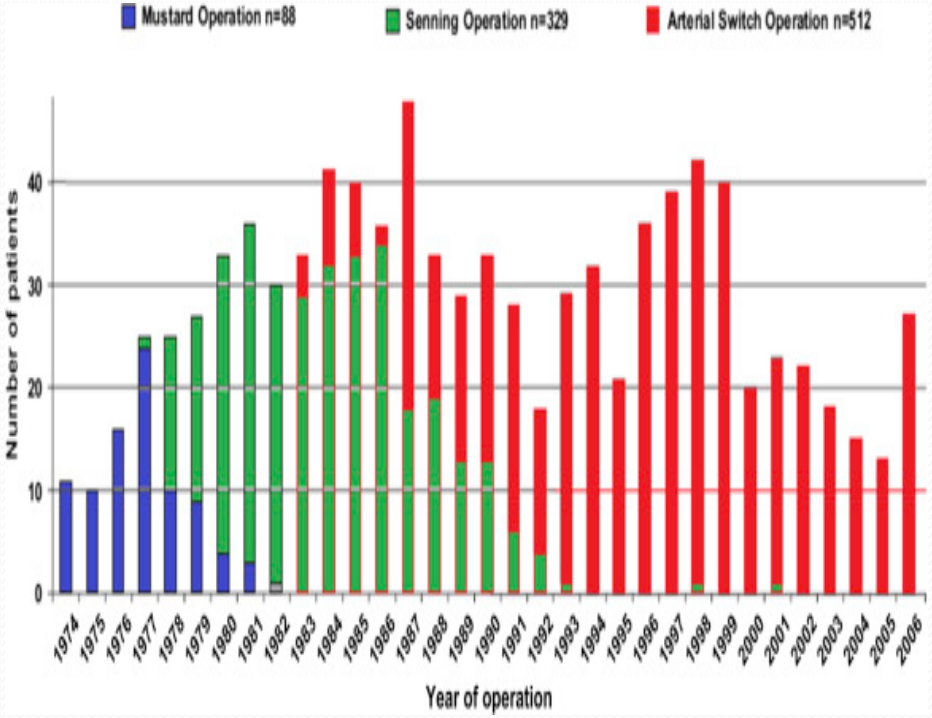
Ideally prevent this entire disease complex….

The Arterial Switch Operation:

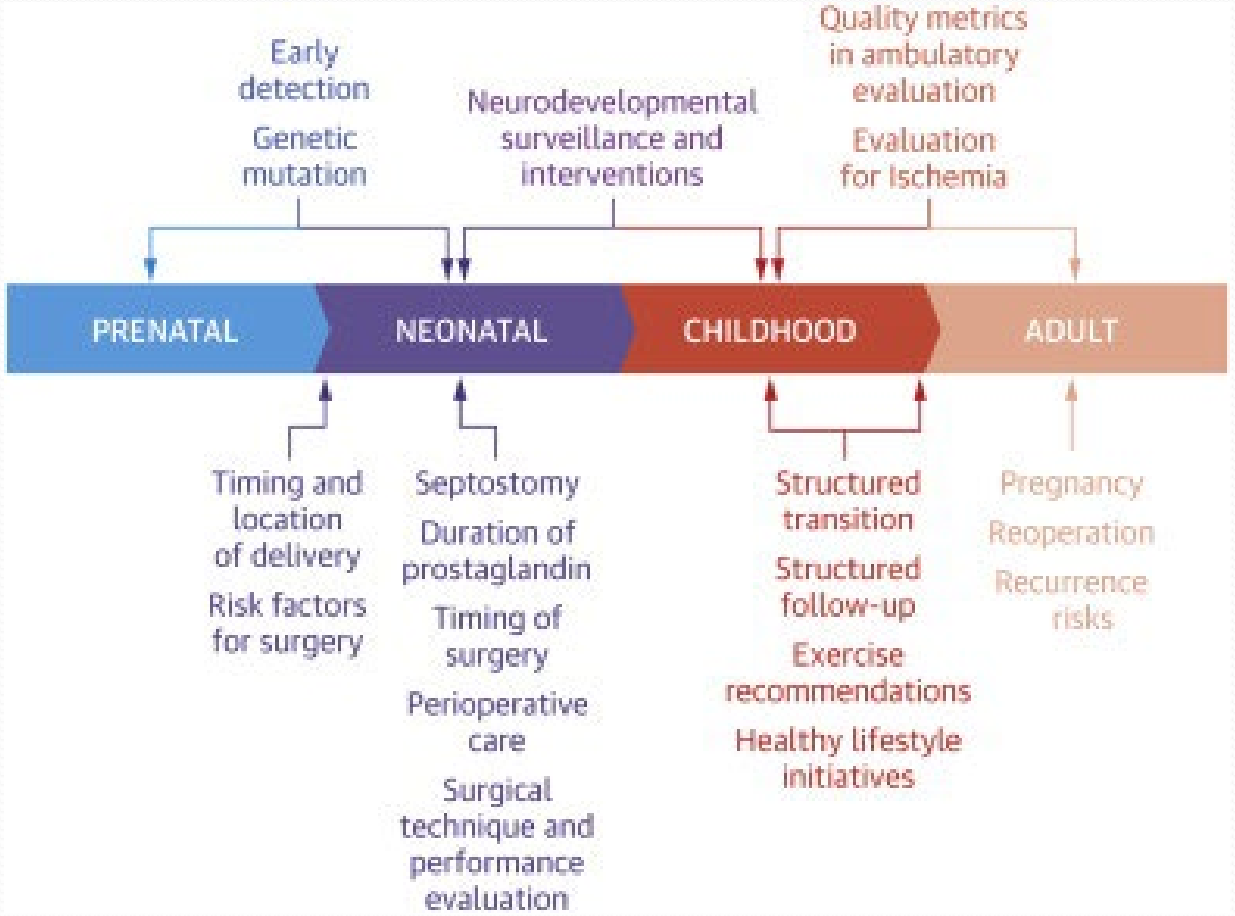
An anatomical repair



I believe 1000 times, the tears of defeat to not having fought…



Transposition: Timeline of Care

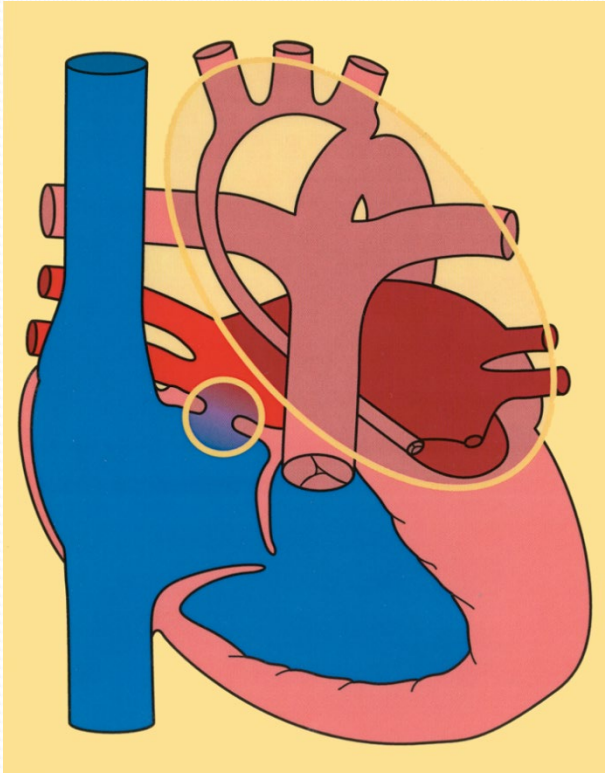


Follow up in PACH Clinic: 2020

- Over 90% of patients survive to adulthood
- Rates of inactivity & obesity have soared
- Exercise Prescription, not Restriction
- MRI provides the ideal window to predict good long-term outcomes

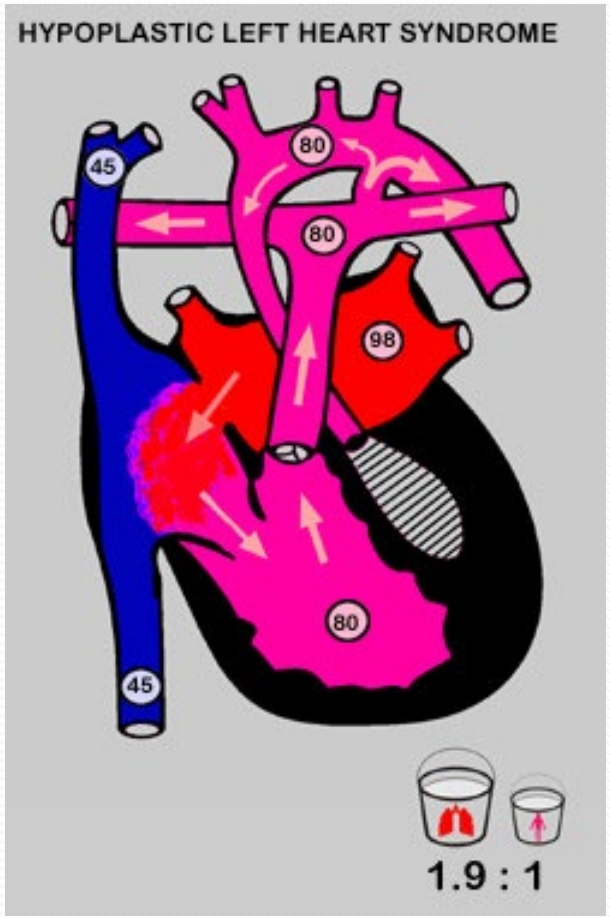
What about the very complex? Hypoplastic Left Heart

- Single Ventricle
- Unbalanced Flow
- Inadequate Systemic Circulation

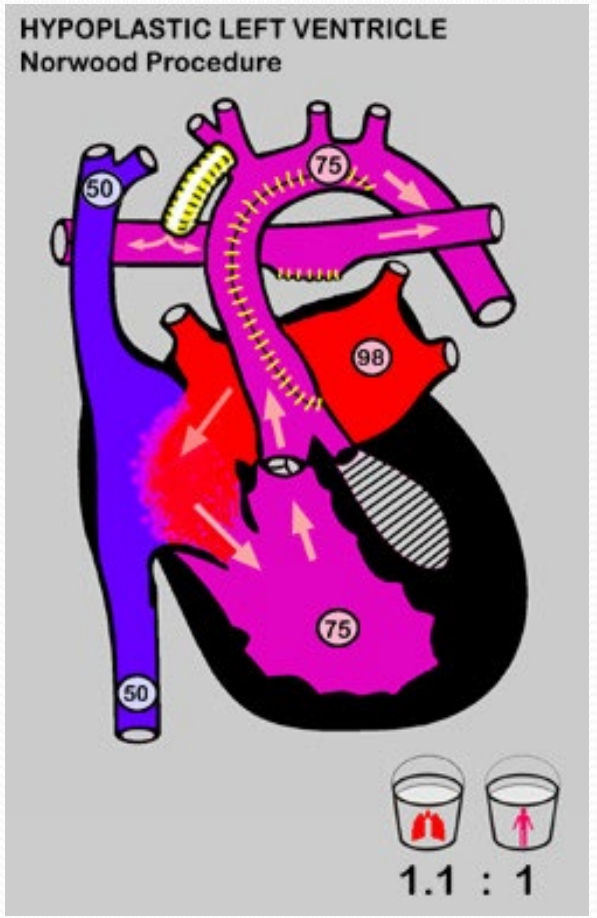


Hypoplastic Left Heart Syndrome

Treatment with Norwood Procedure



Dr William Norwood

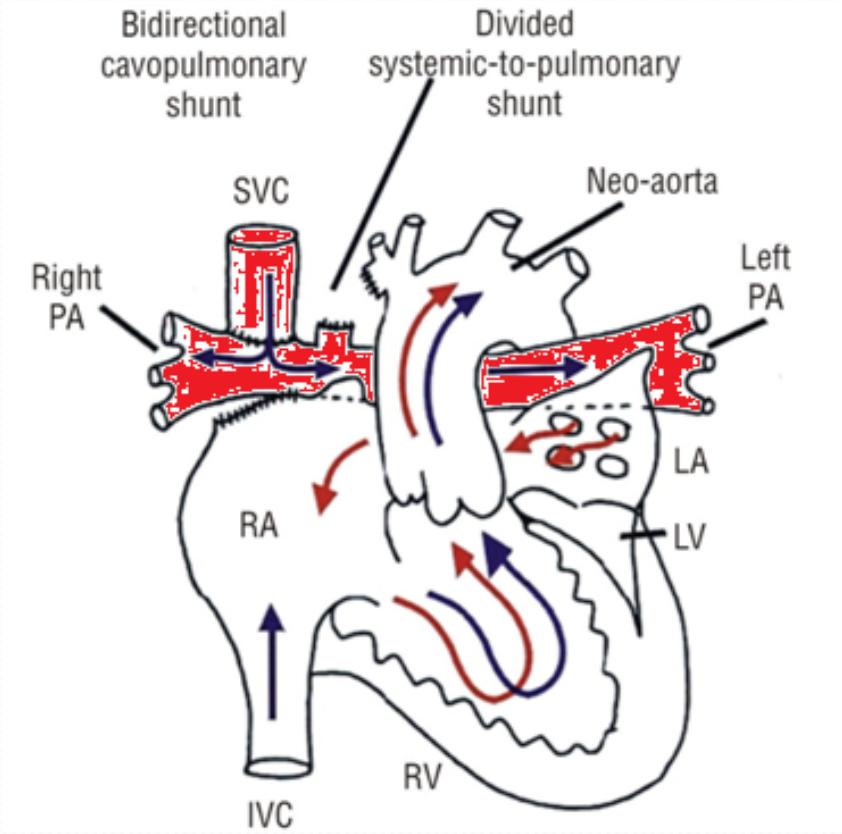


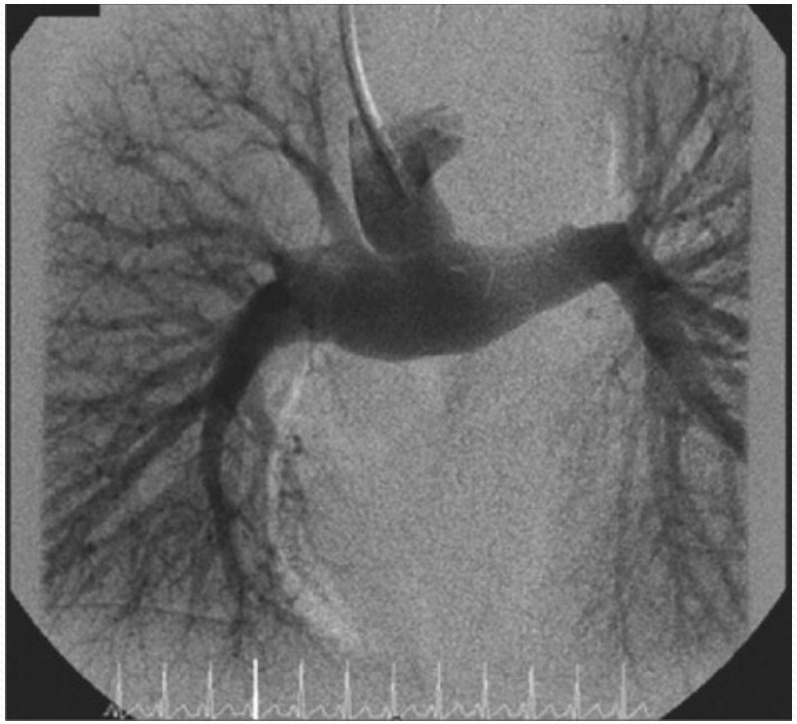
The Fragile Infant

Inter stage mortality ~15%

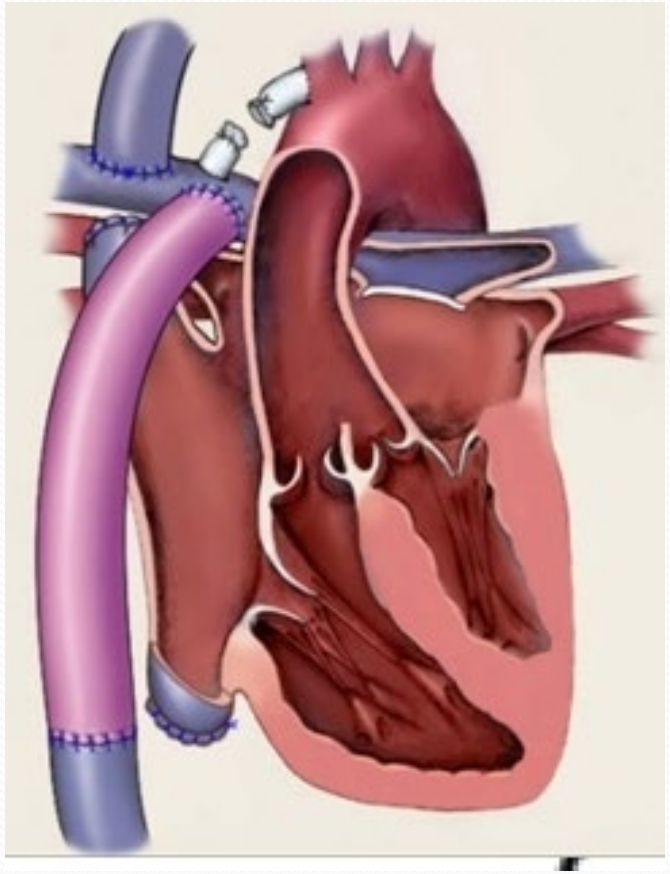
Decreased coronary flow
Shunt obstruction
Arrhythmia
Right ventricular dysfunction
Arch obstruction
AV valve regurgitation
Acute intercurrent illness
- Hypovolemia
- Hypoxemia

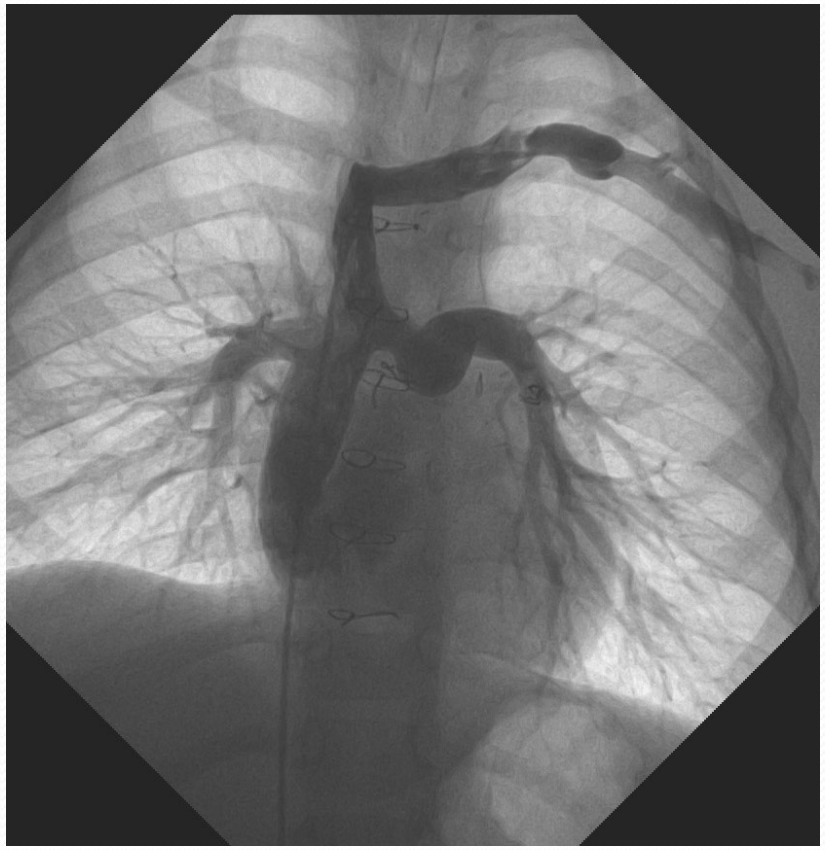
Stage 2 – Bidirectional Glenn





Stage 3 – Fontan state:
Total Cavopulmonary Connection





Fontan circulation patients
A unique disease complex

- Francis Fontan: the initial procedure 50 years ago
- 70,000 survivors worldwide
- Likely to double over 20 years
- Wide variety of anatomical substrates
  - Single Ventricle Physiology
- 3 Major and many minor Surgical variations
- Unlike the atrial switch…this disease complex is not going away

Living in the Fontan state

Non-pulsatile Pulmonary blood flow

Elevated systemic venous pressure

Mild desaturation, elevated hemoglobin

Potential Thrombosis & Embolism

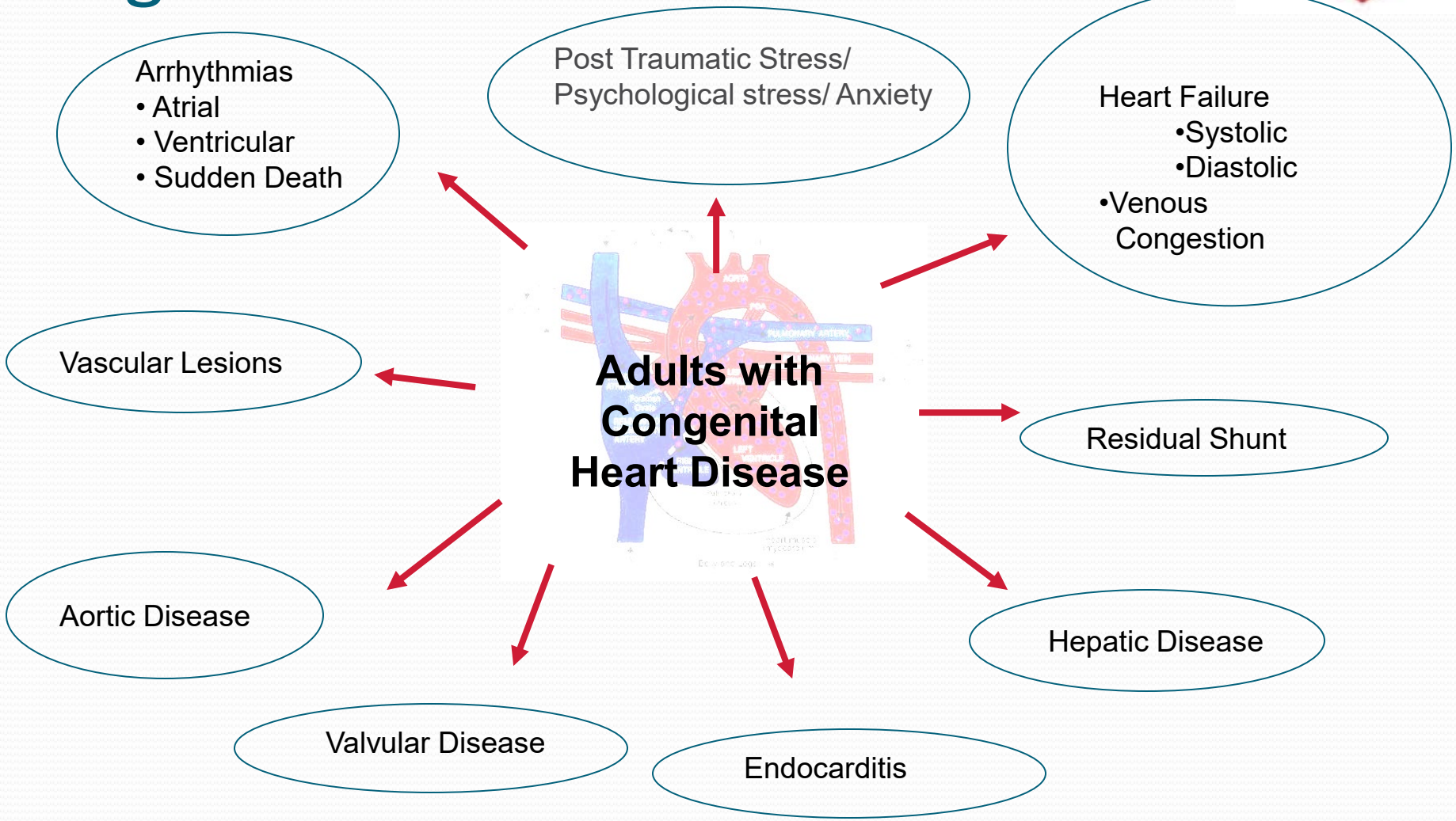
Perfect Single Ventricle function

- No A-V Regurgitation
- No Outlet Obstruction
- No Arrhythmia
- Normal filling hemodynamics

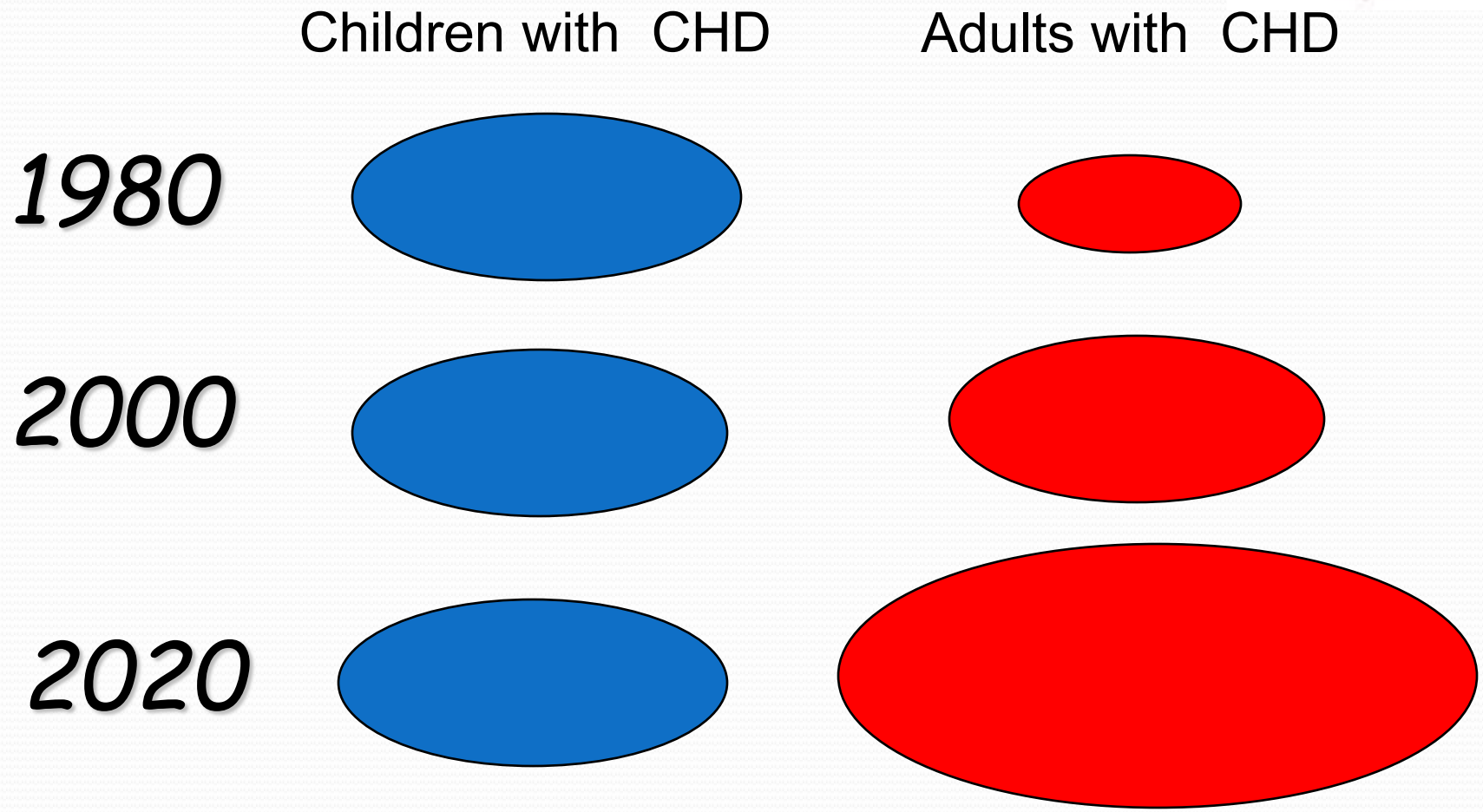
92% Survival to 15 years

85% Survival to 30 years

Living in the Fontan State:



Population shift



Slide courtesy of Gary Webb

How should we manage this unique disease complex?

Today we are in the early phase of characterizing this circulation

Understanding why some patients do well, and others not, is in its infancy

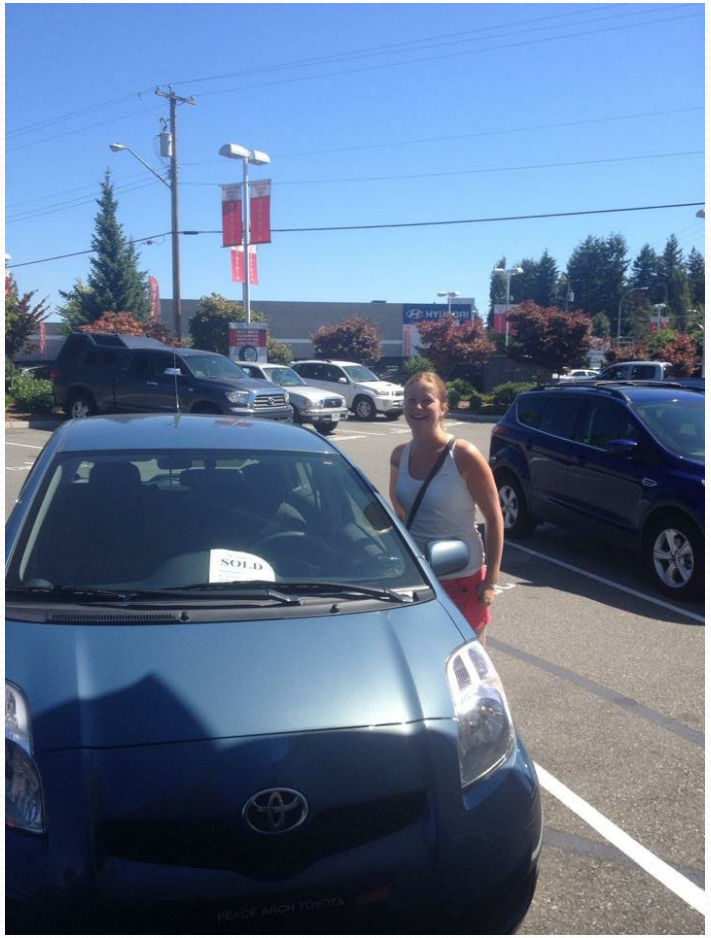
We have not started to evaluate the impact of interventions

- What is the place for Mechanical Circulatory Support?
- For Transplantation?

Regular, structured follow up in a dedicated clinic is just the start of proper care for this population:

Virani Pacific Adult Congenital Heart Clinic: V-PACH

Psychosocial Challenges
Living with a chronic health condition



- 3 Positive Themes
  - Happy in themselves
    - Proud, mature, special
  - Focus on possibilities
    - Managing restrictions
  - Committed to life
    - Living with uncertainty
    - Making the most of life

Surfing the Giant Waves

Managing the future challenges

Caring about and learning from, the patient & the family

Why do this?

Historical context

Monitoring outcomes:

- Care that is Safe and Efficient and Effective

Establishing programmes:

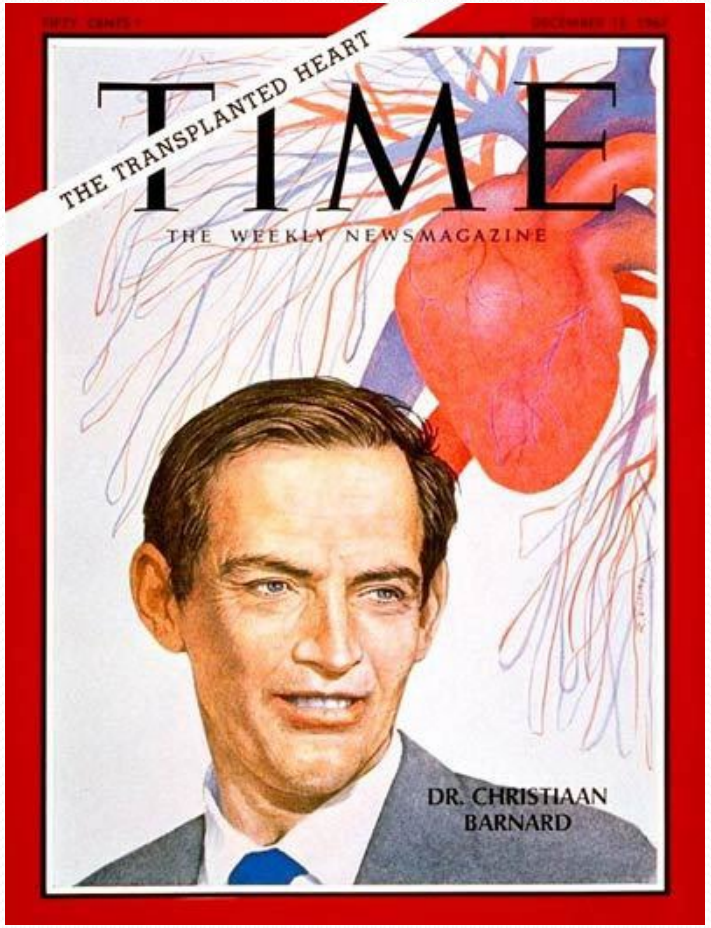
- Care that is Accessible and Equitable and Eco-friendly

Communicating:

- Care that completes the circle of person and family:
- Holistic, compassionate, dignified and in partnership

Quality in Pediatric Cardiac Care

World’s most famous cardiac surgeon?



Not a Discussion between Equals

Outcomes in Tetralogy
Red Cross Children's Hospital

Dr Chris Barnard:
Eminence-based practice

- Repair at any age
- Results are the same
- Results are excellent

Dr Derek Human:
Evidence-based practice

- Results are not the same
- Worse if infant repair
- Results are significantly different
  - <5% mortality vs.
  - 16% mortality

Virtue Ethics

(In God we trust, the rest bring data)

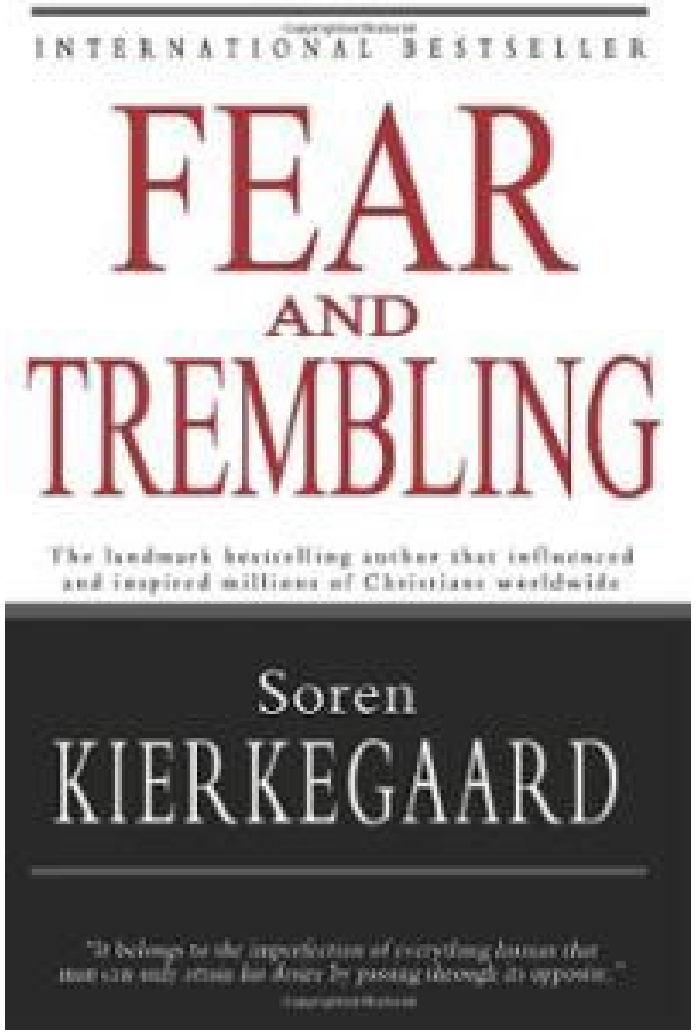
- Conscience
- Courage
- Wisdom & Data
- Doing the right thing enables ethical behavior

The Bristol Royal Infirmary Inquiry

- Professor Ian Kennedy: 1998-2001
- Increased mortality in Cardiac Surgery, continued for over 5 years
  - 577 Witnesses
  - 900,000 pages
  - 196 Recommendations
- The most significant change we call for is in the culture of the NHS
  - This is an account of people who cared greatly about human suffering, were dedicated & well-motivated
  - Sadly, some lacked insight, & their behavior was flawed

Quality in Pediatric Cardiac Care
Flight Plan approach introduced 2018

- Tracking all errors or events that occur during a Cardiac Surgical admission
- NP records from daily rounds, summary to QA team each month
- QA team reviews EMR & Surgical list for consistency
- Monthly ‘Flight Plan’ report to the Cardiac Sciences Team.



BCCH continues to monitor events every month, with the whole team

- Mortality remains low
- ICU stay and Length of Stay in hospital are well below norms
- Event rates are low overall, and compare well to published data
- Complex mix of cases
  - Very few out of Province referrals
- Challenge to identify most high-risk cases and prevent the cascade of reinforcing errors
  - Event rates have not changed over the past several years

Caring about, and learning from, the patient & the family

Why do this?

Establishing programmes:

- Care that is Accessible and Equitable and Eco-friendly

Communicating:

- Care that completes the circle of person and family:
- Holistic, compassionate, dignified and in partnership

INNOVATION AND EFFICIENCY

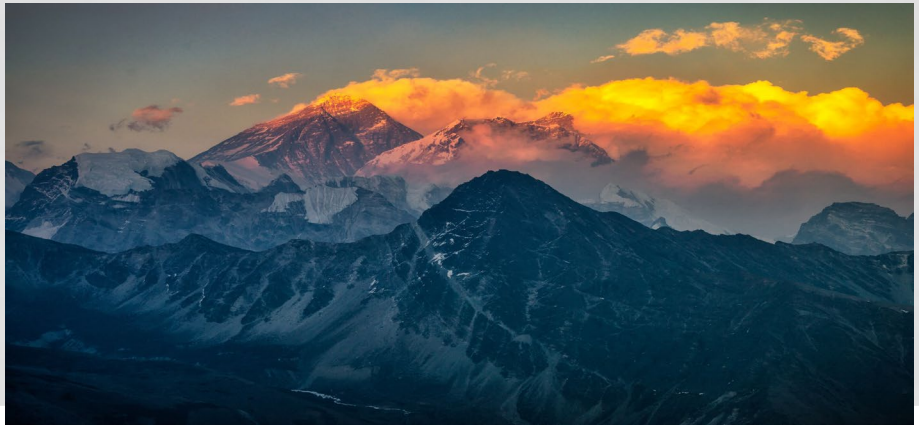
A PROVINCIAL PROGRAM FOR PEDIATRIC CARDIOLOGY

PRESENTED BY:
MARY SPENCER, RN MSN , AND
DR. DEREK HUMAN, FRCPC

Cardiology Partnership Program 1994

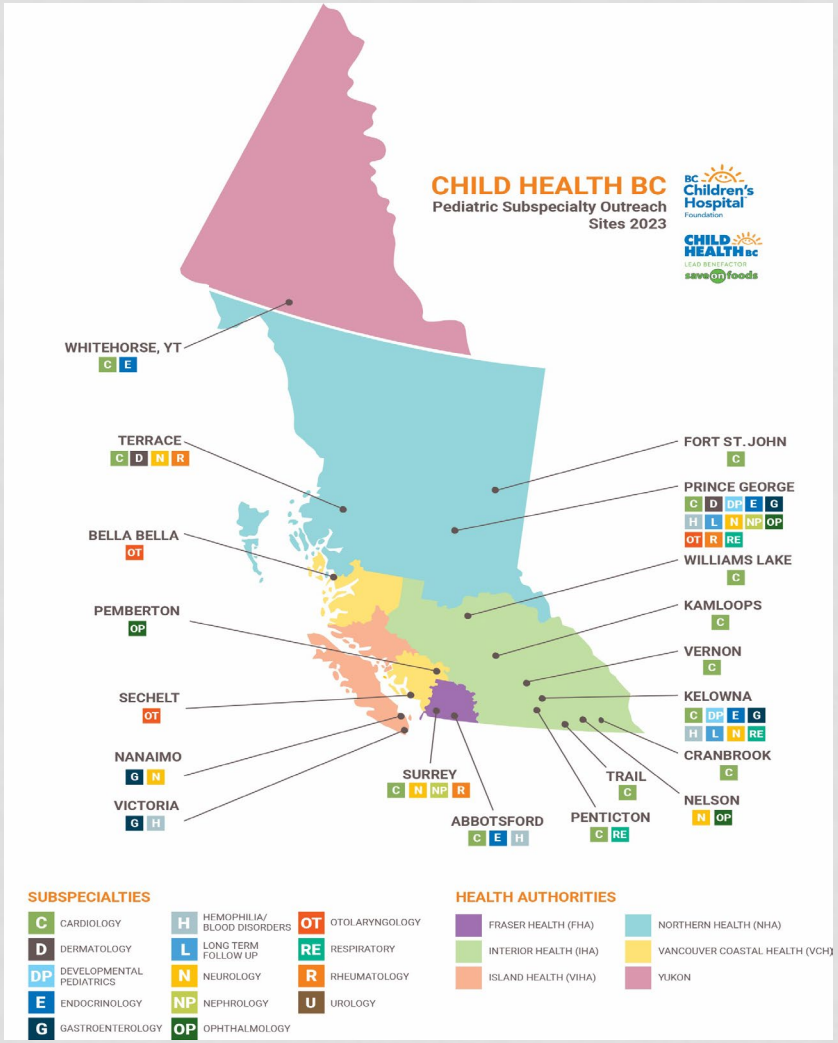
PROGRAM OBJECTIVES

- To increase Pediatric Cardiology consultant access
- To strengthen cardiology care and support for children and families
- To strengthen community-based pediatric services
- To optimize resource use
- To provide a forum for mutual education



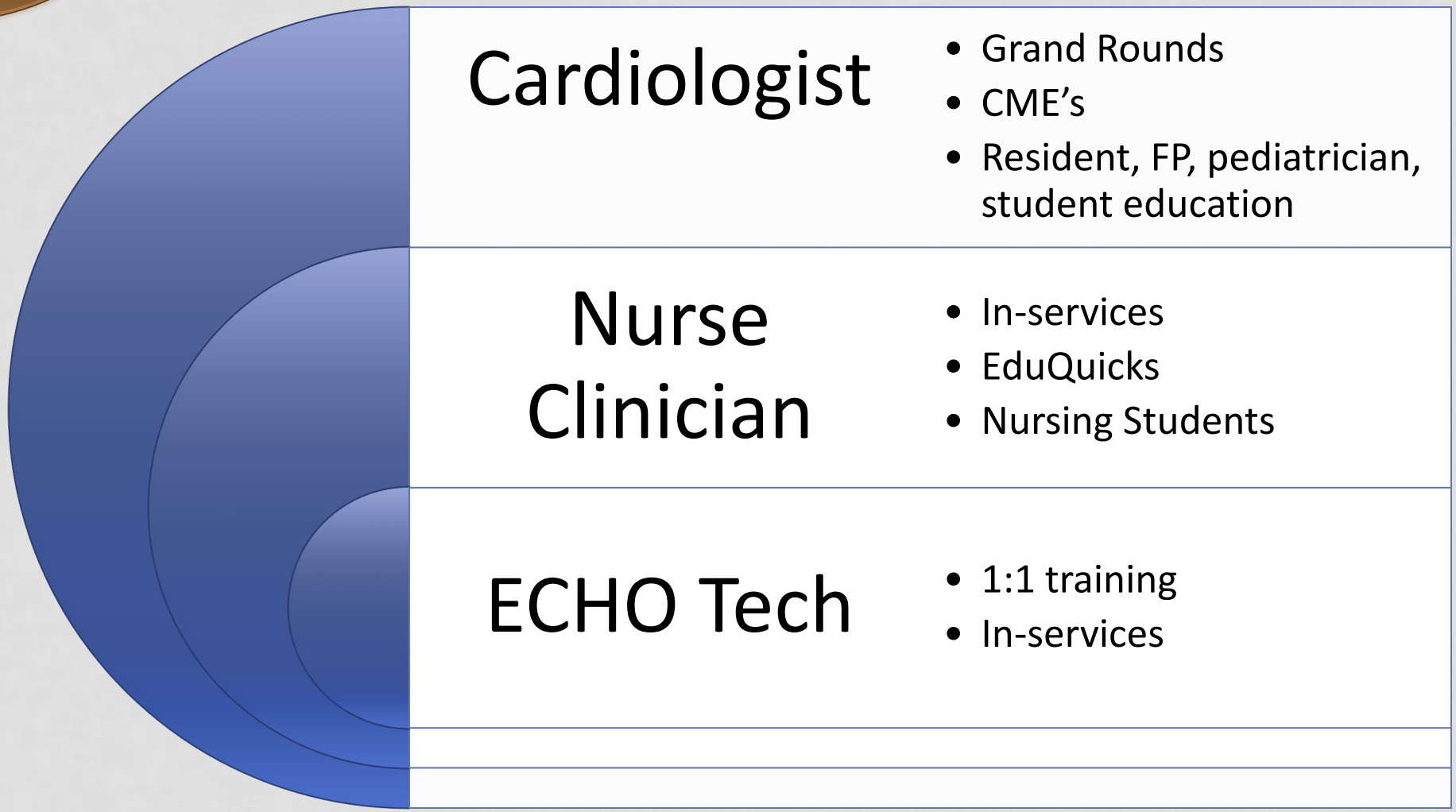
BC by Health Authority

BRITISH COLUMBIA AND YUKON

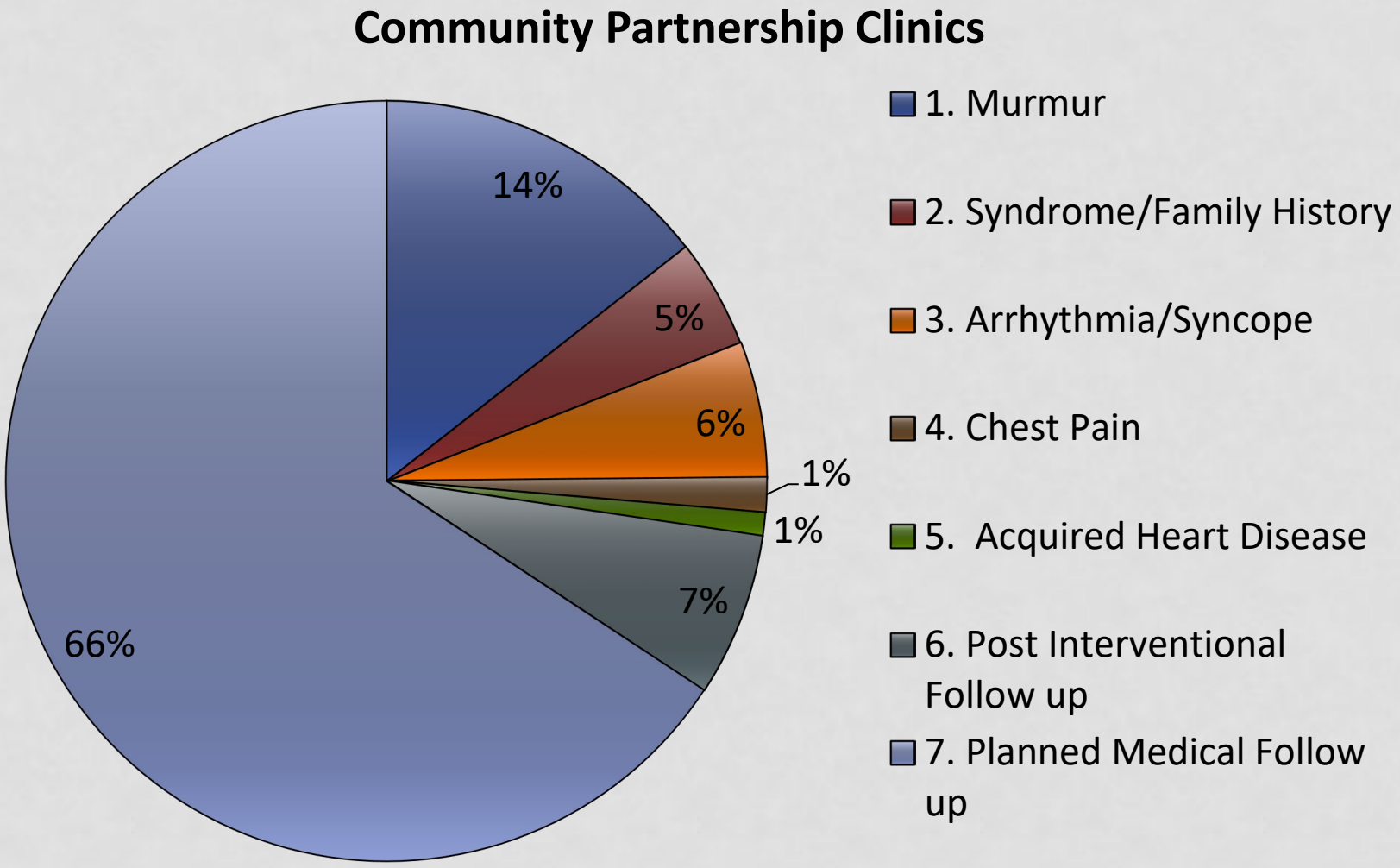


The Clinical Team

TEAM ROLES IN EDUCATION



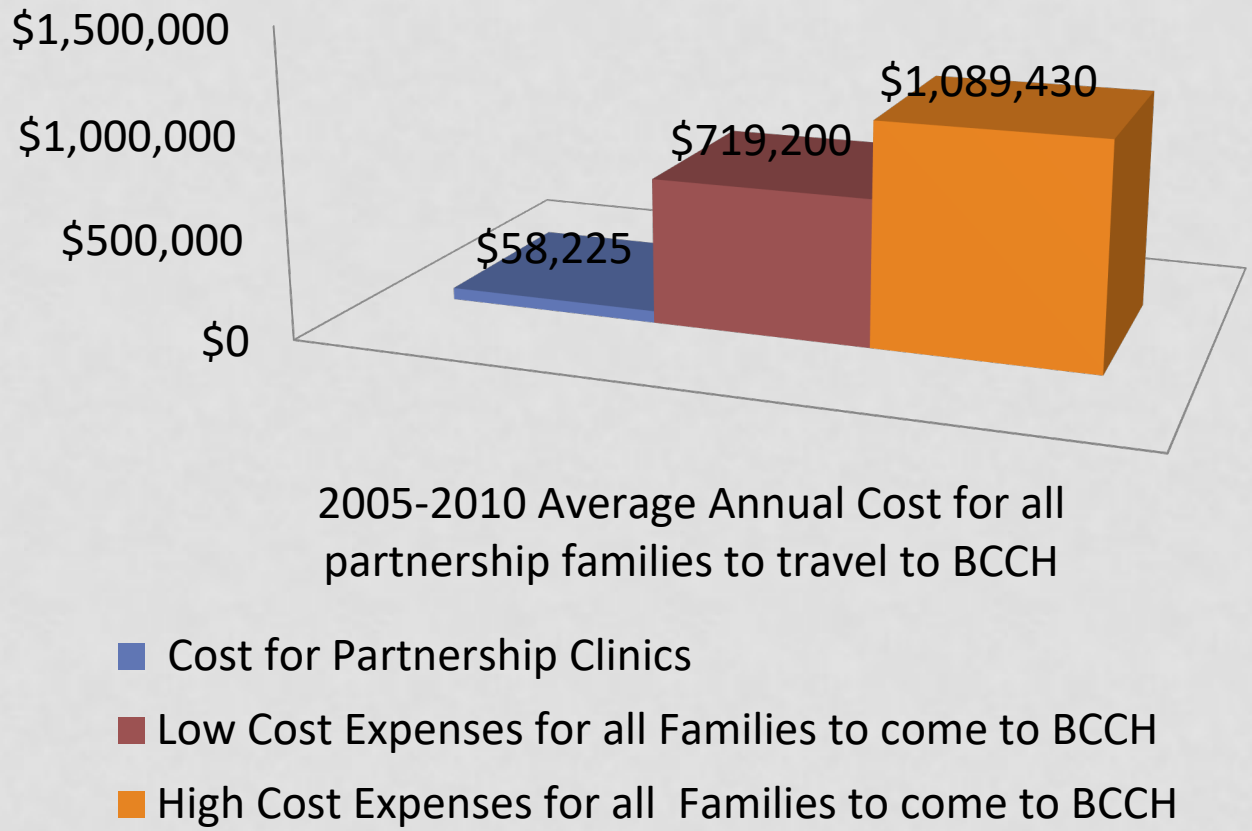
REASON FOR REFERRAL



THE BENEFITS

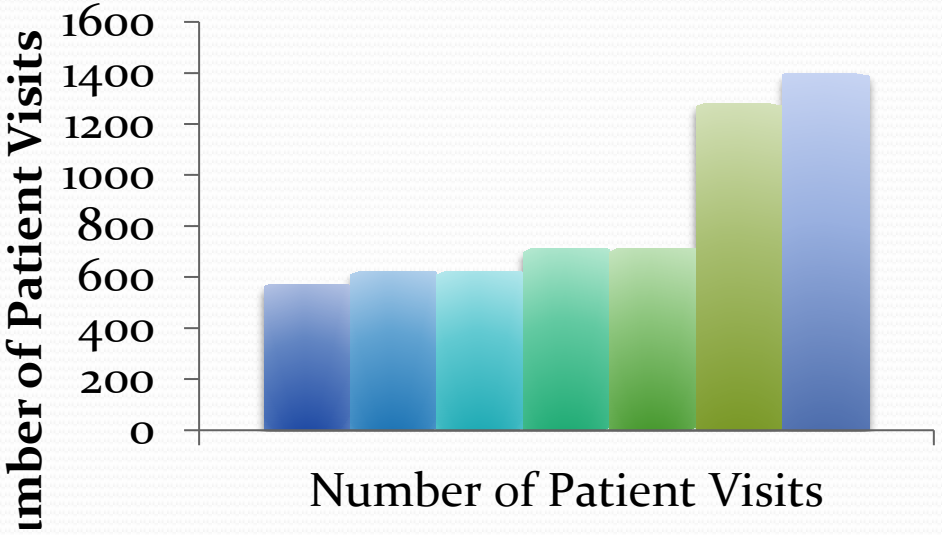
Health Care Professionals	• Networking and communication • Develop expertise
Families	• Decreased cost and travel • Follow up • Decreased stress
Communities	• Efficient • Environmentally friendly. • Efficient NPO use

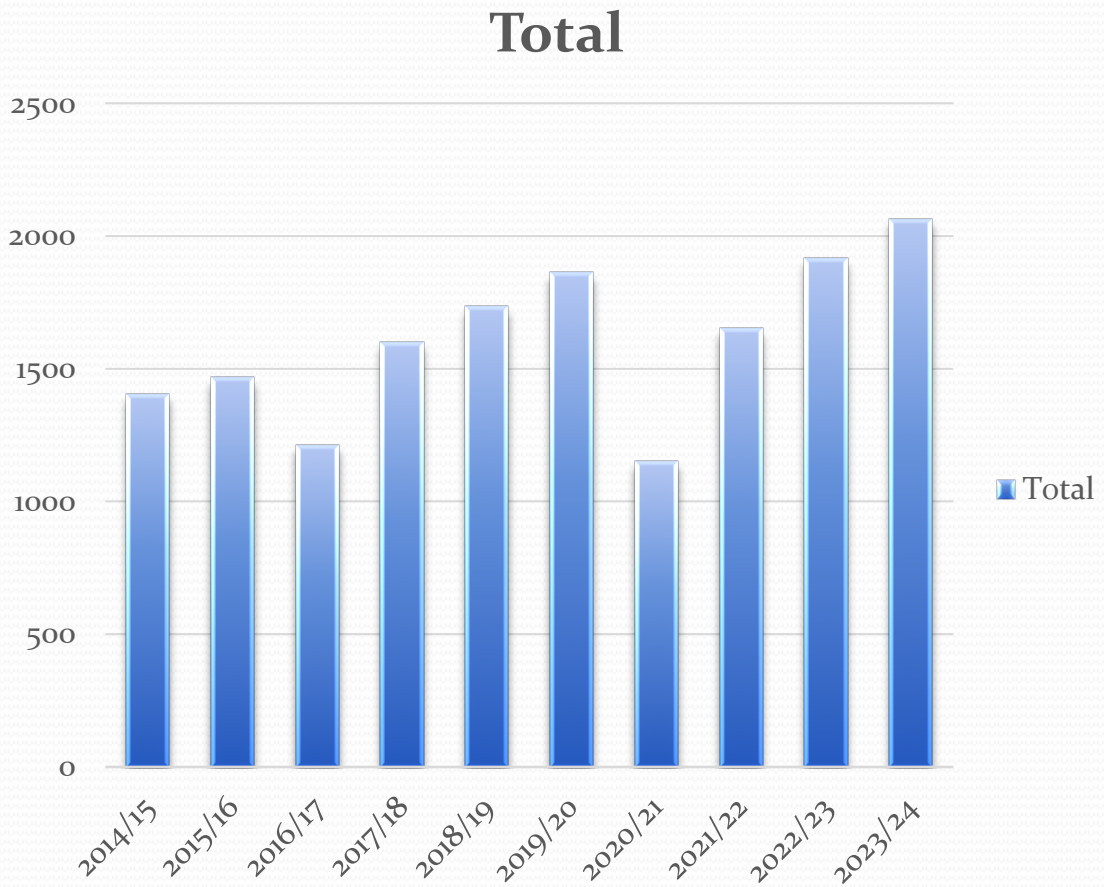
A LOOK AT COST FOR FAMILIES



Growth of Partnership

- 115 patients seen in 1994
- Surrey clinic added 2013





Partnership is just one example of the programs we have developed to deliver excellent & accessible care

- Telehealth for inherited arrhythmias
- WelTel for follow up of Transplant patients
- Antenatal care plans for unique diagnostic problems
- Expanding the V-PACH program to run clinics in the Interior and Northern BC
- Embracing virtual consultations and remote echo reading and reporting

Caring about, and learning from, the patient & the family

Why do this?

Communicating:

- Care that completes the circle of person and family:
- Holistic, compassionate, dignified and in partnership

Consent and understanding

SHARED DECISION-MAKING (SDM)

This is a collaborative process that

Engages health care professionals and patients in making health decisions and

Is fundamental for informed consent and patient-centered care.

Unhelpful Communications: Impact on Families

- A mismatch between perception of information needs and the manner information is provided creates:
  - Emotional distress
  - Illness management challenges
  - Distrust in health provider's professionalism or competence
- Extreme psychological distress associated with:
  - Too much information
  - Inconsistent information among/between professionals
  - Overabundance of negative or discouraging information
  - Gaps in information provided

Supportive Communication

"Tell Me More"

The 3 Level conversation

1) Informational: What is happening?

Tell me more about what information you need about options for additional testing related to a possible genetic syndrome.

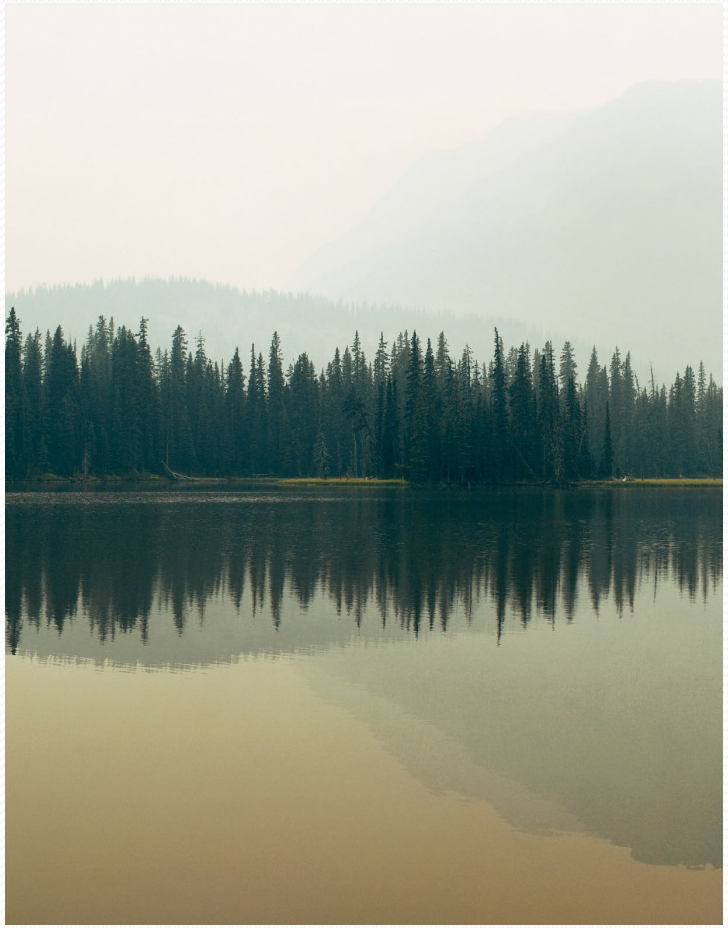
2) Emotional: How do I feel about this?

Tell me more about how you are feeling about what we have discussed about the medical treatments and options we have discussed.

3) Self-Identity: What does this mean to me and my family?

Tell me more about how Sarah's need for surgery is impacting you and your family?

Coercion and Consent: FNHA 2023



Realizing my Superpower

- *As a care provider, prioritizing and advocating for informed consent is one of your superpowers.*
- *You bring with you a wealth of knowledge and experience that lights the path for others.*
- *By illuminating the path and walking with a patient in true partnership, you create inspiring change within healthcare, and especially that individual's life*

Realizing my Superpower

Thank you for bringing your most curious self to appointments, for bringing your most attentive mind, and your best intentions for each individual patient with their own complex personal history.

The work you do in a good way sends ripples of change farther than any of us know.

— **Toni Winterhoff** (Ey Clan ey) is a Xa'xsta member living and working on Sto:lo territory. Toni works at the FNHA as a Healthy Children Specialist.

I am the guide to this new country

Entering the health care system can sometimes feel like going to another country. The roads are different, the customs are confusing, and you don't necessarily understand the language being spoken around you and to you.

Consent forms are within that same discordance. They are often created with complex medical terms and are there for the protection of those working within the health care system, not considering the understanding of the people receiving care.

It is the responsibility of providers to provide clarity. In Canada, a great deal of work still needs to be done to provide that clarity in a formalized way.

—Dr. Unjali Malhotra, Medical Officer, Women's Health, First Nations Health Authority

Using my super-power

Reflect on inherent power imbalances in patient-provider relationships and uphold patients' full decision-making power and autonomy over their health.

- Reflect on the values, assumptions and belief structures you bring to interactions with your patients, especially those from a different culture.

- Take the time to get to know the patient and their community, being genuine, listening respectfully, being attentive and acknowledging the patient’s lived experience, and providing the time for the patient to tell their story.

- Ask questions and provide input and feedback.

The Challenge of Patient-led Decision-Making

Ultimately, people own their own health and wellness journeys and patients must be the final decision-maker.

- SHARED DECISION-MAKING (SDM) is a collaborative process that engages health care professionals and patients in making health decisions. It is fundamental for informed consent and patient-centered care.
- PATIENT-LED DECISION-MAKING is a type of shared decision-making in which the patient, not the provider, has the decisional responsibility. The provider shares their expert knowledge with the patient, and the patient makes their own informed decision about their care.
  - *Kon AA. The Shared Decision-Making Continuum. JAMA, 2010; 204 (8):903-4.*

Mark and his family as teachers:
Starts in Saskatoon in 1989

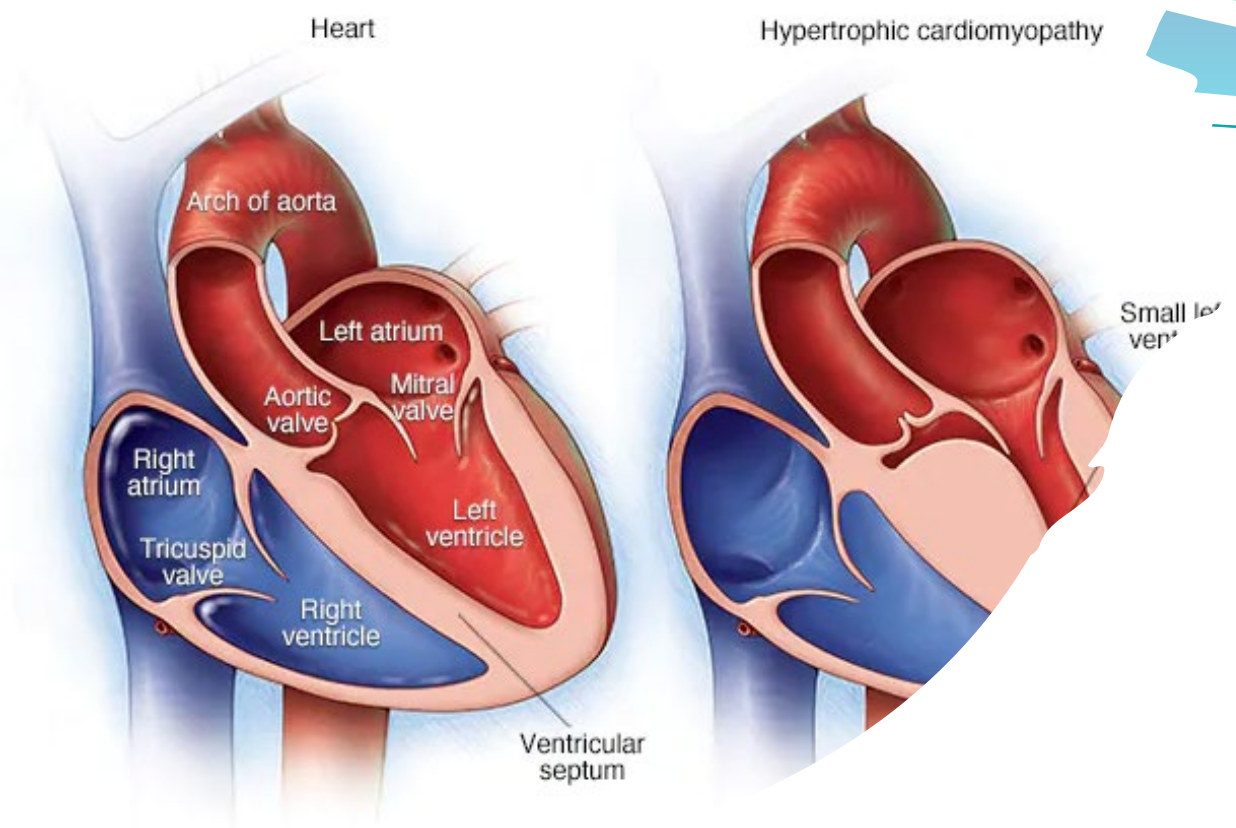
- HLH diagnosed antenatally
  - …this condition is not compatible with life…
- 2 Experimental options
  - Transplantation in California
  - Surgical palliation in Boston – Dr Norwood

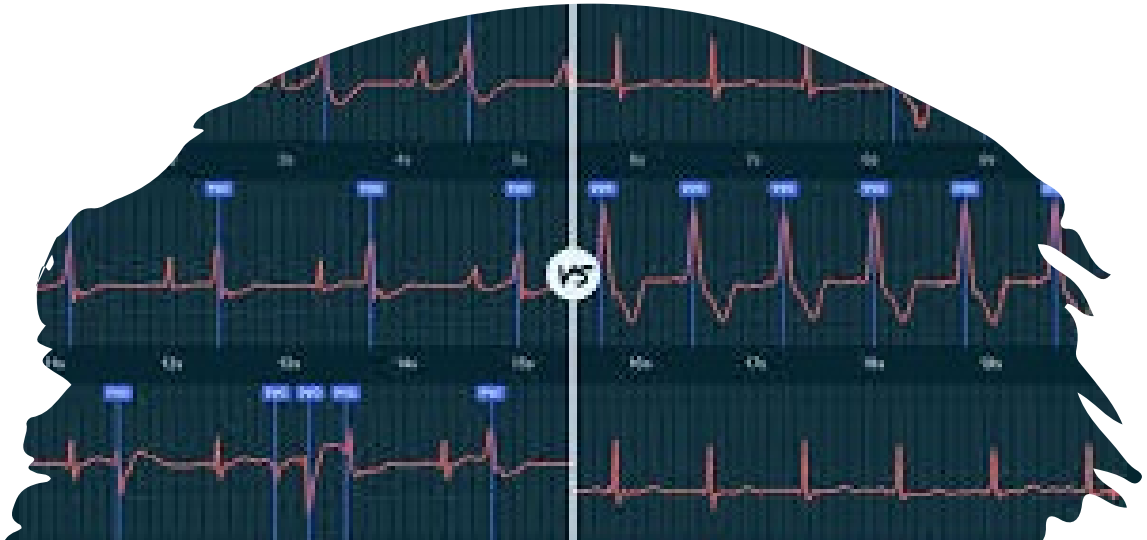
Making a decision… based on limited information

- *Anfechtung, the ultimate personal struggle*
- ***In the Infinite Resignation, there is peace and rest***
- You go with your intuition, and faith in the course chosen
- Is this a test, of how much we love our baby?

Learning and understanding from the experts: the family caring for a complex patient

- **SHARED DECISION-MAKING (SDM)** is a collaborative process that engages health care professionals and patients in making health decisions. It is fundamental for informed consent and patient-centered care.





Diagnosed at birth:

Hypertrophic Cardiomyopathy & Noonan Syndrome

- High dose beta blocker therapy
- Ventricular ectopy burden 15%
- Risk of Sudden unexpected death
- Neurological complications:
  - Hydrocephalus
  - Optic glioma
  - Multiple brainstem lesions

JUNE 2019 PARENTS TOLD ME ABOUT THIS LETTER IN JACC: COULD THIS HELP HIM?

A recent publication from University of Montreal entitled:

Hypertrophic Cardiomyopathy in Noonan Syndrome Treated by MEK Inhibition was published in the Journal of the American College of Cardiology, 2019, Volume 73, issue 17.

Lead author was Dr. Gregor Andelfinger; I have corresponded with him regarding the use of trametinib in this patient.

Success in only 2 patients at this stage, much younger than our patient

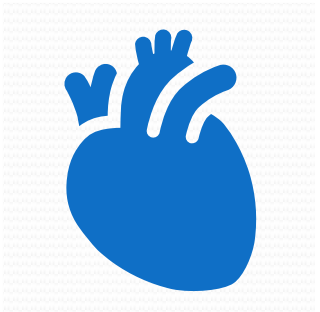
Application made for Innovative therapy and funding at BCCH

June 2024: no major medication problems

What next?

He is one of 35 patients on this medication, publication now accepted with over 20 contributors from around the world:

Impact of MEK inhibition on childhood RASopathy-associated hypertrophic cardiomyopathy



Cardiac condition is stable

Ventricular ectopy resolved

Happy and active

No progression of his neurological lesions

Hope:
Facilitate uncertainty management

In the absence of full and certain knowledge, individuals come to value information as medical opinion.

Unlike clinicians, a parent's concern is not about accurate calculation of predictive values in relation to population data, but about gathering the resources to enable them to cope and make the most appropriate decisions for their family.

Thank you and questions