Follow-up

Importance

Pitfalls

Roadblocks
Long, long ago, in a land far, far away...

Eisenmenger syndrome

- Unrepaired congenital heart disease
Prince slays dragon... prince marries beautiful princess...

1970s – 1980s

• Explosive growth in knowledge
• Intense transdisciplinary collaboration
• Unsettled nomenclature
• Surgery moving toward the very young
Migration of Expertise to Extremes of Age
...and they lived happily ever after?

A word of warning

- Jane Somerville
- Grown Up Congenital Heart Disease
Discontinuity of Follow-Up
Discontinuity of Care
Malalignment of Care
Just a Coarctation!

Just a mechanical problem

- Operation #1
  Patch graft, 1960s, age 5

- Operation #2
  Pseudoaneurysm at patch, 1960s

Recoarctation & calcification

- Operation #3
  Ascending to abdominal aorta conduit, 1970s
Just a few problems...

Thyroid cancer (1980s)
  - Radiation to neck & superior mediastinum

Paroxysmal atrial fibrillation

Hypertension

Then (nearly age 50)...
  - Pulse disappeared from left arm (2012)
  - This prompted investigation
Findings

Gradient

• Ascending aorta 200 mmHg
• Descending aorta 100 mmHg

Left subclavian artery occluded
Subaortic stenosis
Bicuspid aortic valve
Ascending aortic aneurysm
Stenotic head vessels
Big heart! Calcified conduit!
Operation #4

Arrested

- → cardiopulmonary bypass

40-year-old graft

- Disintegrated with multiple tears, calcified
- Blood channel within pseudo-aneurysm capsule
- Circulatory arrest for repair

2 senior cardiovascular surgeons
Reoperation #4

Repairs

- Old graft replaced
- Bicuspid aortic valve repaired
- Subaortic membrane peeled out
Is this the end of the story?

Likely future events

- Aortic valve replacement
- Ascending aorta replacement, with branch grafts to head vessels
- Possibly endografting of bypassing conduit
Bridging the Chasm
The CHSS Model

CHSS cooperative centers

- Across continent

Nature of studies

- A few rare lesions
- Time zero at or near birth
- Capture all management strategies: none, medical, cardiologic, surgical

Emphasis

- Long-range outcome / appropriateness
Roadblocks
Sinkholes

Medical records data
Sinkholes

Data sinkholes

• Fragment longitudinal health record
• Obstruct portability for data sharing (interoperability)

Result

• Lack of long-term outlook data after heart repairs
“Constructive” Solutions

Universal Healthcare Identifier

• Essential for linking data sources

Pilot examples

• Registry and Surveillance in Hemoglobinopathies
• Muscular Dystrophy Surveillance & Tracking & Research Network
• Autism spectrum disorder using global unique identifiers (GUIDs)
"Constructive" Solutions

Interoperability

• It is coming
• It seems not to be easy
Bridges to Nowhere

Lost to follow-up
Roadblocks: HIPAA & IRBs
IRB Roadblock

Follow-up hindered

• Controlled from institutions remote in time and place from patient

• % of patients followed plummets
What CHSS has Done

Central IRB

• In most institutions, IRBs have permitted a notice to be given to parents to contact a central IRB for consent to be followed

Central follow-up

• About 60% follow-up if done centrally
• About 10% follow-up if by institutions
Lessons Learned

Follow-up for clinical events and status is vital

- But it will always be criticized as reflecting “old” treatment

In the U.S., even vital status is tough to discover

- Only Google / NSA knows who is alive and who is dead
Lessons Learned

Life-long follow-up is best done by a central mechanism

• But it is expensive and tangled up with restrictions
Lessons Learned

Much really interesting data is longitudinal

- Repeated surveillance (like echos)
- Repeated health-related QoL
- Often requires central adjudication

Challenge is obtaining the images / data and extracting the information
Lessons Learned

Parents / patients generally love to be followed regularly

- A connection when medical care may be fragmented
- Aids reconnection

Parent / patient incentives help

- $25 Amazon gift cards
Lessons Learned

Federated data

- An ideally federated medical record system
- With longitudinal discrete data
- And a reliable means to share them

Would make follow-up fairly automatic

- It would improve medical care, research, and surveillance efforts
Parked Slides
Importance of Long-Term Follow-up

CHSS Perspective & Lessons Learned
Gaps from Sinkholes

No population-based data!

- Need prevalence data across the lifespan—neonates, infants, children, adolescents, adults

Why needed?

- Characterize disease burden: morbidity, mortality, healthcare use & costs, non-healthcare costs
Longitudinal Outcomes

Never corrected disease
Uncorrected components of disease
Secondary effects of disease & its treatment
Health sequelae of treated disease
Superimposed unrelated adult conditions
Long-Term Measures

Clinical hard data
  • Death
  • Reinterventions
  • Pacemakers, etc

Functional health status
  • Exercise capacity, e.g.

Quality of life
  • Self-reported surveys