Risk Factors in ACHD Redo Surgery: Strategies to Optimize Outcomes

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NO DISCLOSURES
• Congenital Heart Disease is the most common form of Congenital defect

• Most forms of CHD can not be considered curable and have residual sequelae

• Transition of care from childhood to adulthood is highly variable, many lost to follow-up

• In those with previous surgery, many face multiple reoperations with increasingly higher risk
In the past, Adults with Congenital Heart Disease were uncommon and hard to spot
With advances in medical, surgical, and intensive care interventions, an estimated 9 out of 10 children born with CHD in the United States will survive into Adulthood.
Changing picture of congenital Heart Disease in the USA

Williams RG et al JACC 2006;47(4):701-7
1,000,000 adults are living with Congenital Heart Disease in the United States.
Barriers to Access Care for ACHD Patients

• Failure to transition from pediatric to adult Care

• Insufficient education of patients and caregivers regarding disease nature and follow-up

• Inadequate system of management of patient’s cognitive or psychosocial impairment

• Lack of sufficient numbers of specialty clinics and regional centers

• Inadequate access to or availability of insurance
Loss of follow-up from age 6 to 22 years among the entire Quebec CHD cohort

I may give advice nobody follows
How can we optimize the outcomes for surgical procedures in ACHD patients?
"I'm sorry, Mr. Funucci, but we've decided to award the ceiling project to Michelangelo."
WHO SHOULD DO THE SURGERY?
Risk factors for adverse events after surgery for ACHD

- History of CVA
- Chronic Lung Disease
- Prolonged CPB time
- Multiple Procedures
- Renal Dysfunction
- Male Gender
- Aortic surgery
- Heart failure status
- Surgery by non-Congenital Cardiac Surgeon

National Practice Patterns for Management of Adult Congenital Heart Disease

Karamlou et al  Circulation. 2008;118:2345-2352
National Practice Patterns for Management of Adult Congenital Heart Disease

• Nationwide Inpatient Sample 1988-2003
• 12 congenital heart disease diagnostic groups for both children and adults undergoing cardiac procedures
• Pediatric patients more likely to have CHS perform procedures (68%) ACHS(GUCH) > likely to have non-CHS perform procedures (95%)
• Operations by Pediatric Heart Surgeons decreases in-Hospital death
What is the ideal setting for ACHD?

Our ACGME Congenital Surgery training program requirements still have not acknowledged any need for special training in ACHD
OUR STRATEGY
KNOW THE OPERATIONS AND RISKS
PULMONARY VALVE REPLACEMENT, ASD, and RVOT CONDUIT OPERATION MOST COMMON
Changing Practice of Cardiac Surgery in ACHD

• Patients operated during 2 different time periods- 1990-1994 and 1998-2002 single GUCH unit
• Simple operations (ASD’s) significantly decreased 45% to 27%
• Repeat operations increased from 25% to 50%

WJ Brawn and group, Heart 2004,91:207-212

BIRMINGHAM CHILDREN’S, UK
Survival of ACHD correlates with # of Surgical Interventions Zomer, Circulation. 2011;124:2195-2201

Inverse Relationship
AGING AND THE ACHD PATIENT

• MANY OF THESE PATIENTS ARE GENERALLY UNWELL HAVING NEGLECTED THEMSELVES FOR YEARS, OFTEN THINKING THEY WERE CURED

• THE ADDITION OF ACQUIRED HEART DISEASES LIKE CAD AND THE MULTISYSTEM PROBLEMS LIKE DIABETES COPD, OBESITY, AND CIRRHOSIS THAT OCCUR WITH TIME AMPLIFY THE PROGNOSIS SIGNIFICANTLY
Morbidity increases with duration of surgery as well as number of operations.
Risk Factors and Early Outcomes of Multiple Reoperations in Adults with Congenital Heart Disease

- Subsequent sternotomy showed increased early mortality, yet neither sternotomy number nor cardiac injury was an independent predictor of early death.
- Early mortality was reduced with increased ejection fraction.
- Urgent operation, longer bypass time, and single ventricle diagnosis were independent risk factors for early death.

MAKE THE OPERATION AS SAFE AS POSSIBLE!
KNOW THE EXACT ANATOMY
Pre-operative Evaluations: what studies are useful?
Balance cost vs knowledge gained?

• Echocardiogram: presence of shunts, function
• CT angiogram: relationship of cardiac structures to each other and the sternum
• MRI: right ventricular function
• Cardiac Catheterization: pressures, resistance
• Ultrasound of peripheral vasculature: which vessels are open for emergent cannulation
• TEE: bubble study for intra-cardiac shunts
USEFUL CT SCANS
Pre-Operative Planning

• “Pre Flight” Planning: KNOW EXACTLY WHAT YOU ARE GOING TO DO, anticipate problems (inadvertent cardiac or aortic injury), outline the operative sequence: efficiency, not speed alone, is the key

• This is crucial to the outcome since the length of CPB is a significant risk factor for early mortality
Risks of Redo Sternotomy

- 602 redo sternotomies
  - 67% second sternotomy, 28% third sternotomy, 4% fourth sternotomy, .8% fifth sternotomy, .2% sixth sternotomy
  - Hospital survival 98% 590/602
    - None of the 12 deaths secondary to redo sternotomy
    - 2 cases received transfusion secondary to injury
    - Only 4/602 had femoral cannulation (2 planned)
  - Sternal infection 0.5%

1/3 had 3 or more sternotomies

Be sure that the operation is necessary, but don’t delay if it is!
Indications have changed for Re-intervention in Adults with prior TOF repair

- PVR
- Symptomatic patients
  - Exercise intolerance
  - Signs of heart failure
  - Syncope due to arrhythmia
- Asymptomatic patients 2 or more criteria below
  - Decrease in objective exercise capacity
  - Progressive RV dilation: end diastolic volume index >150 ml/m2 RV/LV, end diastolic volume ratio >2
  - Progressive RV or LV systolic dysfunction: RVEF <47%, LVEF <55%
  - Progressive TR (at least moderate)
  - RVOTO with RV systolic pressure >80 mmHg or large aneurysm
  - Sustained atrial/ventricular arrhythmias with severe RV dilation, QRS duration > 140 ms
  - Significant Residual VSD or ASD
Surgical PVR is low risk, low mortality

Options for PV replacement
PVR Technique
CRYOABLATION ADDED IF RVOT INDUCIBLE VENTRICULAR TACHYCARDIA
RV to PA conduits are a different Matter

- Higher risk of cardiac injury and early mortality: 3 times the risk compared to PVR alone
- Diagnoses include PA/TOF, TGA, TA, DORV, ccTGA, septated univentricular hearts
- Suggest covering just the conduit with polytetrafluoroethylene pericardial membrane at the first operation
PTFE Pericardial Membrane
Use of Pericardial Substitute (PTFE)

• My advice: don’t use it routinely
• Why? Leaves a waxy scar over the entire mediastinum under the membrane which makes it harder to identify individual structures
• Indications:
  RV to PA conduits- place only over the conduit
  Cover large, anterior aortas, Ross procedures, LVAD’s
High Risk Re-sternotomy Technique

• If there is high risk for emergent peripheral CPB, cannulate prior to sternotomy, i.e. Aortic aneurysm eroding into sternum.

• Use of “Time Out” to make sure all members of the team are on the same page. Communication is crucial. All cannulae, blood, and equipment should be in room and ready.

• For planned peripheral cannulation for re-sternotomy prefer percutaneous femoral venous and axillary arterial cannulation with or without graft.
Direct Axillary Artery Cannulation
EDWARDS QUICKDRAW VENOUS CANNULA

25 French (8.3mm) diameter
The Congenital Interventional Cardiologist

Professor Neil Wilson

AVOID AN OPERATION
Medtronic Melody Valve
Contegra bovine jugular vein valve sewn into small metal frame

2 sizes ID: up to 20mm and 22mm
Medtronic Melody Valve

In the pulmonary position
Consecutive Percutaneous Valve-in-Valve Replacement Late After Ross Procedure: A Novel Approach in an Adult With Congenital Heart Disease

Dominik M. Wiktor,¹,² MD, Joseph D. Kay,¹,² MD, and Michael S. Kim,¹,²* MD
Melody Valve in failed tricuspid valve position
Bilateral Melody Valves in Pulmonary Position
HYBRID LAB

Interventional Cardiologist and Surgeon work in concert
LIMIT THE OPERATION
Hybrid Approach for pulmonary valve

(A) Angiographic measurements of the MPA prior to plication. (B) Measurements of the MPA post-plication. Notice that the diameter of the MPA has been reduced from about 33 to 23 mm. (C) MPA with stent in place to create a landing zone for the Melody valve. Diameter now measures about 20 mm. (D) Angiography with the Melody valve in place showing no significant regurgitation.
NEWER VALVES may obviate the need for a Surgeon once more

Edwards Sapien XT valve

Tri-leaflet bovine pericardial valve

Available in larger sizes 26mm and 29mm
Venous P-valve sizes up to 35mm

Dr. Gareth Morgan
UCH and CHC ACHD Interventionalist
“Sir, please calm down. Our automated surgeon does not respond well under stressing loads.”
IN CONCLUSION

An experienced, dedicated multi-disciplinary team including ACHD specialized Cardiologists, Adult and Congenital Heart Surgeons, Cardiac Intensivists, Anesthesiologists, and Medical specialists including renal, hepatic, infectious disease, and nutrition, both adult and pediatric, working together is the best paradigm to optimize outcome.
Thank You