1

Surgical Management of Adults with Congenital Heart Disease: Importance of a Congenital and Aortic Partnership

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Seattle Children's
HOSPITAL · RESEARCH · FOUNDATION

UW Medicine

2

Disclosures

- **None**

3

Advances in Care for Patients with CHD

- Significant medical and surgical advances
  - Improving surgical techniques
  - ICU and anesthetic care
  - Transcatheter interventions
- Survival to adulthood:
  - *15% →90%*

Brida, M *et al. Int J Cardiol CHD.* 2020

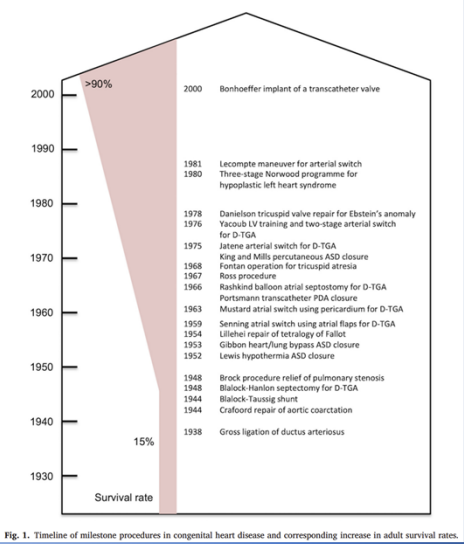
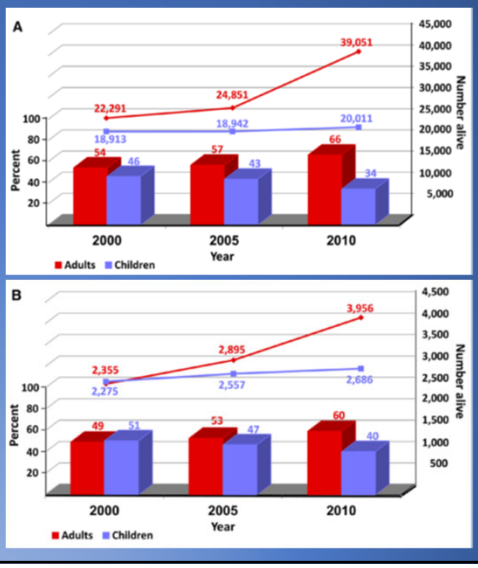


Fig. 1. Timeline of milestone procedures in congenital heart disease and corresponding increase in adult survival rates.

4

Advances in Care for Patients with CHD

- Increase in adult patients
  - More adults than children starting in 1990s
  - Now more than double
- Many of the patients with complex disease



Marelli, AJ *et al*. *Circulation*. 2014

5

ACHD Population in United States

Category and Age Group	CHD Severity/Race-Ethnicity	Estimated US Prevalence per 1000 (95% Confidence Interval), %	Estimated No. of Individuals (95% Confidence Interval)*
CHD severity			
All ages	Overall	7.85 (7.79–7.92)	2 425 000 (2 405 000–2 444 000)
	Severe	0.92 (0.90–0.94)	283 000 (277 000–290 000)
Children	Overall	13.21 (13.03–13.39)	980 000 (966 000–993 000)
	Severe	1.66 (1.60–1.73)	123 000 (119 000–128 000)
Adults	Overall	6.16 (6.10–6.22)	1 444 500 (1 431 000–1 459 000)
	Severe	0.68 (0.66–0.70)	160 000 (155 000–165 000)
Race-ethnicity			
Children	Non-Hispanic white	13.31 (13.12–13.49)	620 000 (612 000–629 000)
	Non-Hispanic black	12.69 (12.50–12.88)	133 000 (131 000–135 000)
	Hispanic	13.26 (13.08–13.45)	227 000 (224 000–230 000)
Adults	Non-Hispanic white	6.36 (6.29–6.42)	1 104 000 (1 094 000–1 115 000)
	Non-Hispanic black	5.63 (5.56–5.69)	155 000 (153 000–156 000)
	Hispanic	5.58 (5.52–5.65)	186 000 (184 000–188 000)

Gilboa, SM *et al*. *Circulation*. 2016

6

ACHD Population in United States

- Often compared to tsunami
- Patients lost to follow-up after aging out of pediatric practice
- Not enough cardiologists training in ACHD
  - Poor reimbursement
  - Lack of exposure
- ACHD care limited to larger academic centers

7

Standardizing Care

- Creation of accredited ACHD centers of excellence
- Extensive requirements
  - Only 51 centers in 26 states
  - Adjacent to large pediatric centers
- Many ACHD clinics that don't meet all requirements

8

Comprehensive Care Center Requirements

ACHA ACHD PROGRAM CRITERIA Comprehensive Care Center
A. ACHD Cardiologist
B. ACHD Medical Program Director
C. Advanced Practice Nurse/Physician Assistant
D. Registered Nurse
E. Cardiothoracic Surgery and Cardiothoracic Intensive Care Unit
F. Heart Failure, Heart Transplant, Heart/Lung Transplantation
G. Interventional Cardiac Catheterization
H. Interventional Electrophysiology
I. Inpatient Services
J. Outpatient Services
K. Transitional Services
L. Patient-Centered Care
M. Echocardiography
N. Cardiac Magnetic Resonance Imaging
O. Cardiac Computed Tomography
P. Pulmonary Arterial Hypertension
Q. Exercise Testing and Cardiac Rehabilitation
R. Reproductive Services
S. Psychology and Social Work
T. Cardiac Anesthesia

9

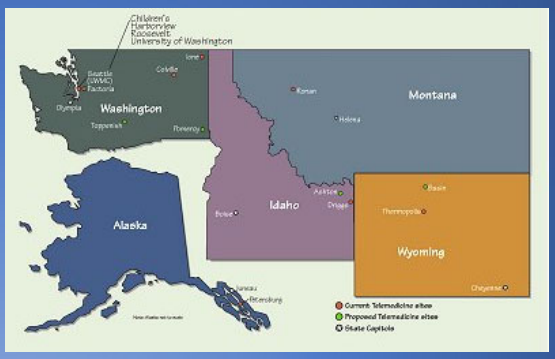
CCC CT Surgery Requirements

- Surgical director certified by ABTS in congenital heart surgery
- Minimum of two surgeons
  - 24/7 availability for consultation or emergency surgery
- Expertise in mechanical support and cardiac transplant
- All surgical patients discussed in multidisciplinary format
  - Includes ACHD cardiology, interventional, EP, imaging, anesthesia, ICU

10

UWMC ACHD Program

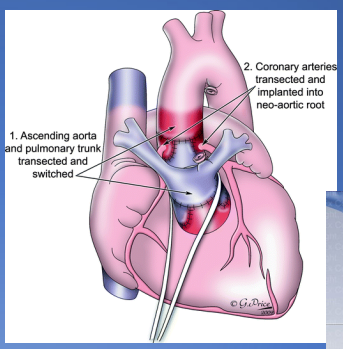
- 7 ACHD cardiologists
- 2 ACHD interventional
  - 1 from Children's
- 3 EP
- 3 HF/Tx cardiologists
- 1 dedicated anesthesiologist
- CTICU
- Two surgeons

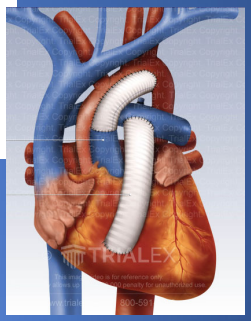


11

What Type of Surgeon?

- Pediatric Congenital Surgeon
  - Primarily at children's hospital, may come to adult hospital for adult cases
  - Very little enthusiasm in congenital community
  - More interested in neonatal and infant repairs
  - Adult cases not appealing





12

What Type of Surgeon?

- Adult Surgeon With Congenital Background
  - Common early on (1990s-2000s)
  - More contemporary surgeons less comfortable
    - Less congenital training
  - Less favorable outcomes

13

National Practice Patterns for Management of Adult Congenital Heart Disease

Operation By Pediatric Heart Surgeons Decreases In-Hospital Death

Tara Karamlou, MD, Brian S. Diggs, PhD, Thomas Person, MD, Ross M. Ungerleider, MD, MBA, and Karl F. Welke, MD, MS

- National Inpatient Sample
  - 1988-2003
  - ~40,000 ACHD operations
  - Compared "Adult" to "Pediatric"
- Pediatric surgeons >75% case volume
- Major outcome death
  - Minor outcomes LOS, cost

Variable	Weighted Frequency±SD
Patients	
Mean±SD age, y	50.0±1.8
Female, n (%)	22 732±739 (56.2±0.6)
Patient diagnosis, n (%)	
HLHS	40±14 (0.099±0.035)
l-TGA	195±37 (0.481±0.089)
d-TGA	236±40 (0.582±0.097)
DORV	195±37 (0.481±0.089)
Other transposition	80±21 (0.198±0.051)
PAVSD	73±21 (0.181±0.051)
ToF	1013±81 (2.50±0.19)
TAPVR	130±24 (0.322±0.060)
Coarctation of the aorta	1625±104 (4.02±0.24)
VSD	5307±212 (13.12±0.46)
Ostium primum ASD	993±74 (2.45±0.17)
Ostium secundum ASD	30 574±1135 (75.56±0.72)

Karamlou T, *et al*. *Circulation*. 2008

14

National Practice Patterns for Management of Adult Congenital Heart Disease

Operation By Pediatric Heart Surgeons Decreases In-Hospital Death

Tara Karamlou, MD, Brian S. Diggs, PhD, Thomas Person, MD, Ross M. Ungerleider, MD, MBA, and Karl F. Welke, MD, MS

Table 4. Multivariable Factors Associated With In-Hospital Mortality (Table view)

Variable	Odds Ratio	95% CI	*P*
Pediatric heart surgeon as dichotomous variable			
Nonpediatric heart surgeon	4.50	2.12–9.53	<0.0001
Female gender	1.32	1.05–1.64	0.0150
Diagnosis of VSD	2.71	1.75–4.20	<0.0001
Ostium primum ASD	0.18	0.06–0.55	0.0026
Coarctation of the aorta	0.19	0.09–0.41	<0.0001
Ostium secundum ASD	0.18	0.12–0.29	<0.0001
Congestive heart failure	6.65	2.90–15.24	<0.0001
PVD	1.89	1.17–3.07	0.0099
Renal failure	2.79	1.43–5.44	0.0027
Complicated diabetes mellitus	3.31	1.68–6.52	0.0005
Annual percentage of GUCH+pediatric heart surgery operations as a continuous variable			
Higher percentage of GUCH+pediatric heart operations	0.65	0.43–0.99	0.047
Female gender	1.2	0.99–1.6	0.053
Ostium primum ASD	0.23	0.08–0.65	0.006
Coarctation of the aorta	0.23	0.20–0.53	<0.001
Ostium secundum ASD	0.21	0.14–0.31	<0.001
Congestive heart failure	6.40	3.41–12.0	<0.001
Renal failure	3.23	1.81–5.8	<0.001

Karamlou T, *et al. Circulation*. 2008

15

ACHD Surgical Population

- Evolution over last 30 years
  - Introduction of transcatheter options
    - ASD
    - Pulmonary valve
    - Aortic valve
    - Coarctation stenting
  - Improvement in ablative techniques

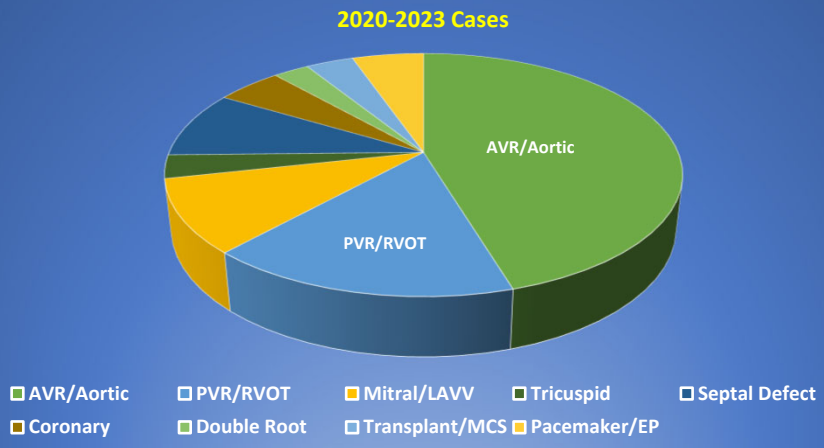
Table 2
Original primary anatomic diagnoses, classified according to EACTS congenital database [15].

Anatomic diagnosis	Number (%) (*n* = 963)
Septal defects	332 (34.5%)
-Atrial septal defect	196 (20.4%)
-Ventricular septal defect	57 (5.9%)
-Atrio-ventricular septal defect, partial	56 (5.8%)
Left heart lesions	242 (25.12%)
-Aortic stenosis, valvar	162 (16.8%)
-Aortic stenosis, subvalvar	28 (2.9%)
Right heart l	194 (20.1%)
-Tetralogy of F	123 (12.8%)
-Ebstein's disease	31 (3.2%)
Thoracic art	98 (10.2%)
-Aortic coarctation	43 (4.5%)
-Aortic coarctation + aortic valve stenosis	34 (3.5%)
Transposition of the Great Arteries (TGA)	37 (3.8%)
-TGA, IVS, including LVOTO	17 (1.8%)
-TGA, VSD, including LVOTO	12 (1.4%)
Single ventricle	35 (3.6%)
Electrophysiologic	16 (1.7%)
-Arrhythmia, heart block, congenital	16 (1.7%)
Pulmonary venous anomalies	7 (0.7%)
-Partial anomalous pulmonary venous connection	5 (0.5%)
-Cor triatriatum	2 (0.2%)

Putman LM, *et al. Eur J Cardiothorac Surg*. 2009

16

ACHD Surgical Population-UW 2020-23



17

UW ACHD Program 2020

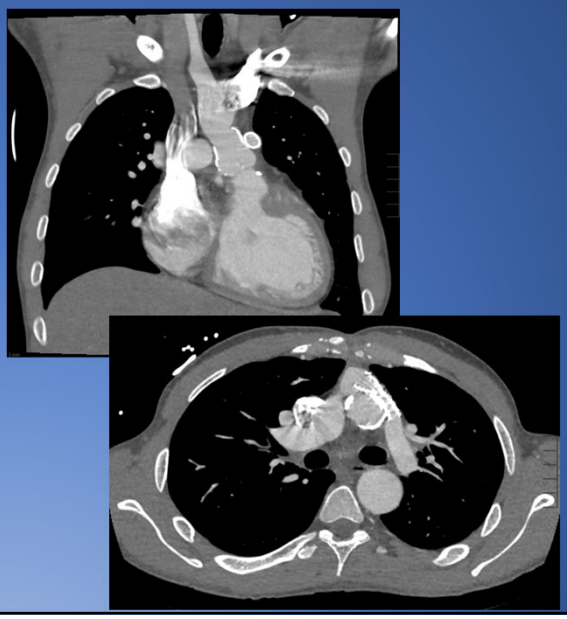
Ed Verrier MD
Professor of Surgery
~30 years ACHD experience

Chris Burke MD
Asst. Professor of Surgery
0 years ACHD experience

18

March 2020

- "So…I have a case I was hoping you could help me with…"
  - 26 yo M, hx of d-TGA
  - Six previous sternotomies
  - 3 previous aortic homografts
  - Stent in L pulmonary artery
  - Aortic endocarditis with severe AI

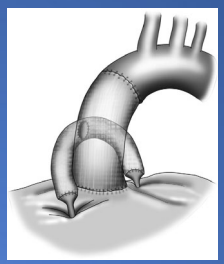


19

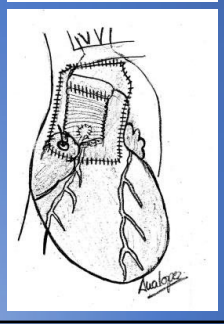
March 2020-Conduct of Operation

- R axillary cannulation
- Redo Sternotomy
- Takedown of LeCompte
- Aortic Root replacement (19mm St. Jude)
- Cabrol reconstruction of coronary arteries
- Reconstruction of LPA with tube graft
- Temporary closure with Cabrol patch

- Many more procedures in 2020



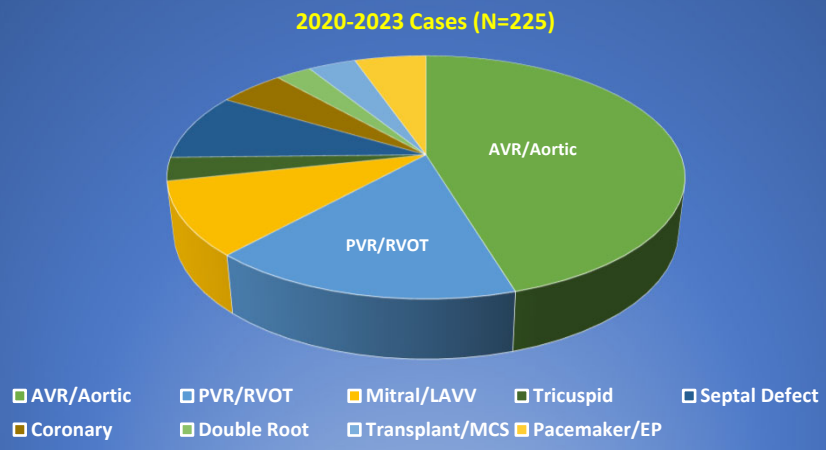
Cabrol Graft



Cabrol Patch

20

Ideal Surgeon for ACHD Population



21

Ideal Surgeon for ACHD Population

- Congenital anatomy, physiology, operations
  - Right sided lesions, septal defects
- Transplant/MCS
- Advanced aortic surgical and transcatheter techniques
  - Aortic valve repair
- Coronary
- Understanding of transcatheter technology
- Basic understanding of EP/ pacemakers

Congenital Surgeon

Aortic Surgeon

? Surgeon (Congenital)

22

Referral Pattern-UW

Burke Clinic

- Aortic Valve disease
- Ascending aortic aneurysm
- Aortic arch pathology

Mauchley Clinic

- Pulmonary valve/RVOT pathology
- Septal Defects
- Tricuspid valve/Ebstein's
- Sequelae of AVSD repair
- Anomalous coronaries
- Transplant evaluations

23

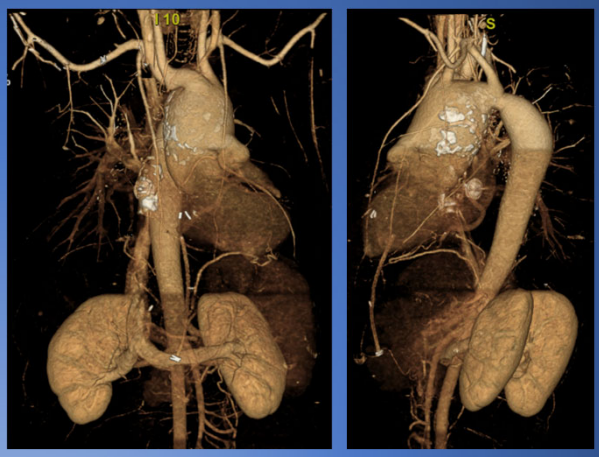
Combined Cases

- Ross procedure
- Aortic valve repair
- Double root replacement
- Aortic root/ascending in combination with complex anatomy
- Complex LVOT obstruction
- Unusual coronary anomalies

24

Case #1

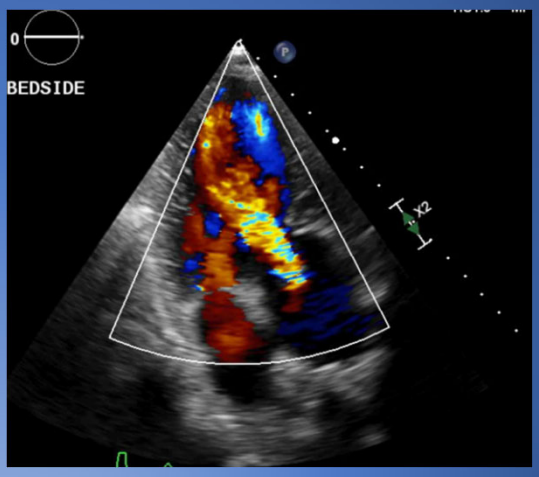
- 25 yo M with DILV, L-TGA, coarctation.
- Surgical palliation with:
  - Norwood procedure (DKS with aortic arch reconstruction, shunt)
  - Bidirectional Glenn shunt
  - Extracardiac fenestrated Fontan
- Followed for ascending aneurysm
  - Now ***7.1cm***, severe neo-AI, intermittent chest pain, mild decrease in LV function



25

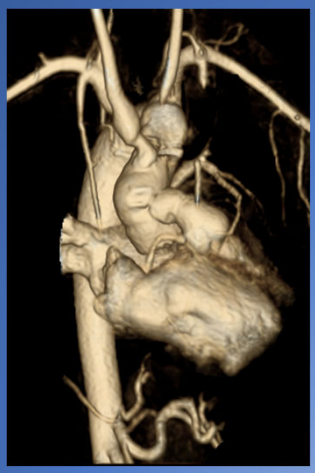
Case #1-Surgical Approach

- R axillary artery cannulation, L anterior thoracotomy for LV vent, venous cannulation SVC and femoral vein
- Cool to 18℃, X-clamp, ostial cardioplegia
  - Native aortic root separated from DKS
  - Zone 2 arch replacement with branched graft
  - Bentall (On-x 27/29) to neo-aortic root
  - Native aortic root anastomosed to Bentall graft



26

Case #1-Final Result (MRA)



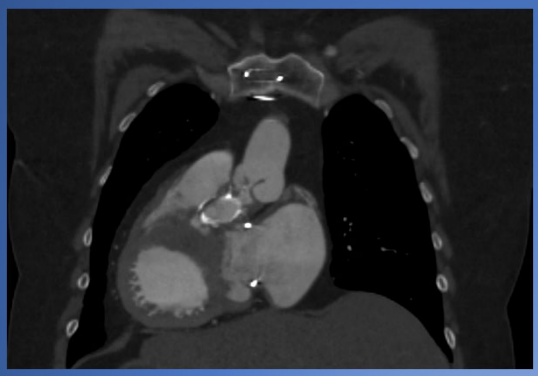
27

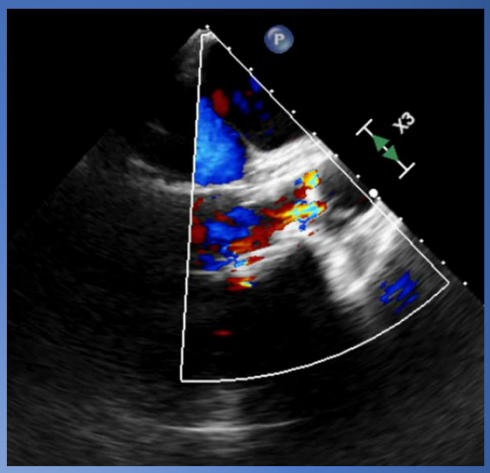
Case #2

- 46 year old with heterotaxy syndrome, situs inversus with mirror-image dextrocardia, TAPVR, DORV, and partial AVSD.
  - TAPVR, DORV, partial AVSD all repaired as a child
  - Revision of ASD patch and TV annuloplasty as young adult
- Progressive heart failure symptoms related to severe LVOT obstruction and aortic insufficiency
  - VSD patch calcified
  - Long, narrow LVOT

28

Case #2

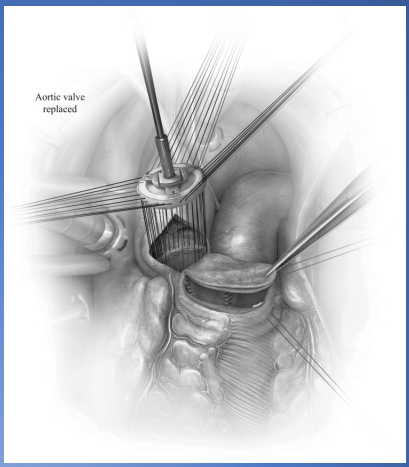




29

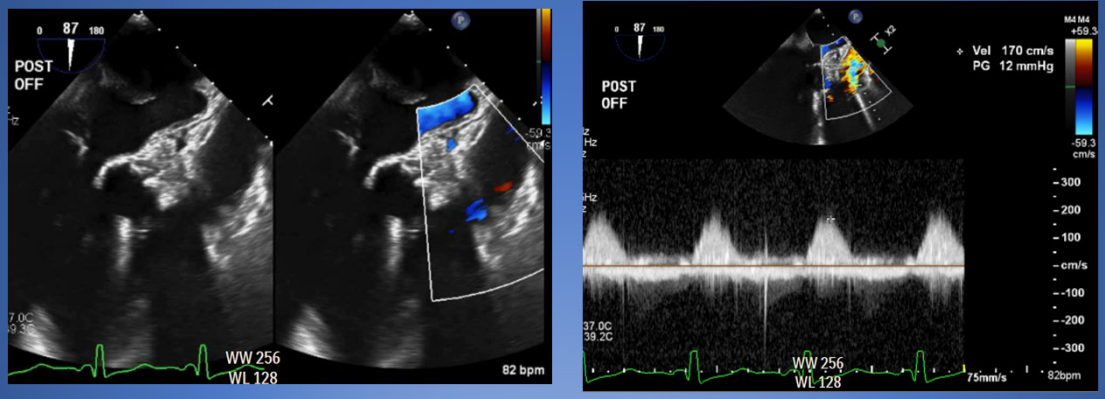
Case #2-surgical approach

- Redo sternotomy, aortic/RA cannulation
- Ostial cardioplegia
- Resection of AV leaflets and inspection of LVOT
- Incision into RV, near complete removal of VSD patch
- Dacron Konno patch to replace VSD patch and enlarge annulus
  - 23 mm St Jude mechanical valve



30

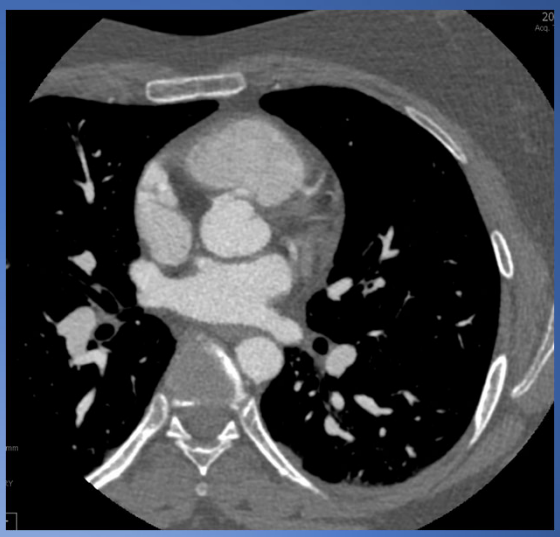
Case #2-Final Result



31

Case #3

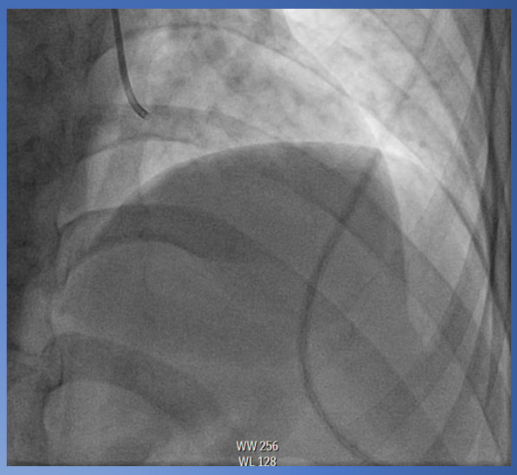
- 32 yo F with Turner's syndrome
  - 2 successful IVF pregnancies
  - Intermittent chest pain after delivery of second child
  - ST changes on ETT
  - Imaging noted intraseptal LM off of R coronary sinus
- Referred for surgery



32

Case #3

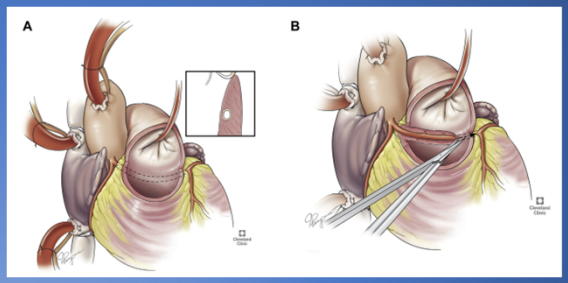
- Normal LM ostium
- No evidence of compression at rest
- Left dominant system



33

Case #3-Surgical Approach

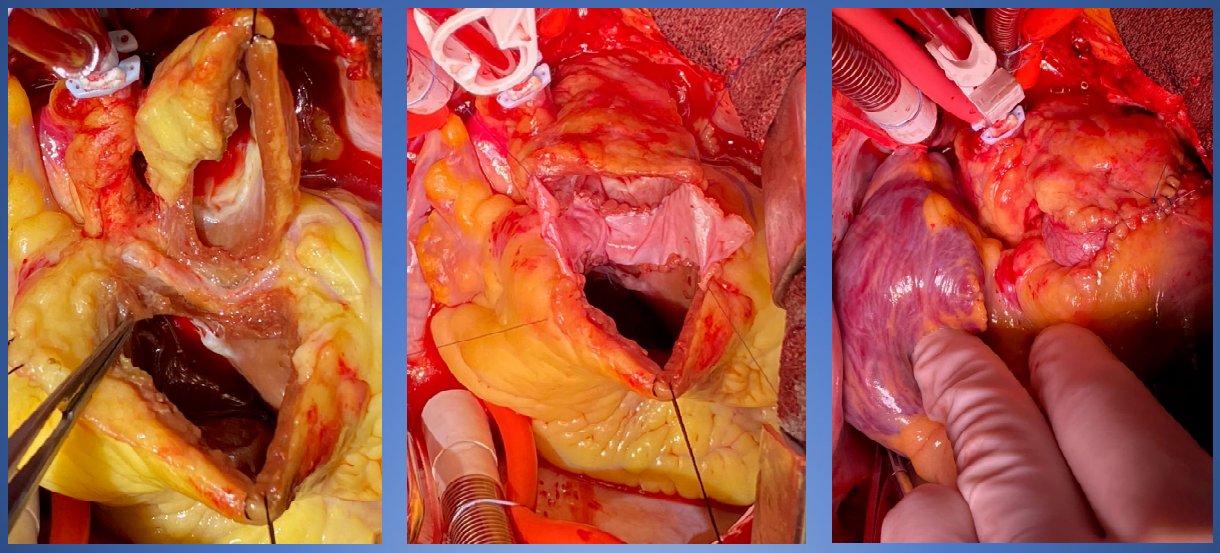
- Transconal Unroofing as described by Najm *et al*.
- Reconstruction of RVOT with autologous pericardial patch



Najm *et al. Ann Thorac Surg*. 2019

34

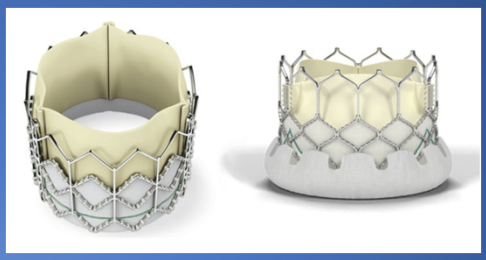
Case #3-Operative Pictures

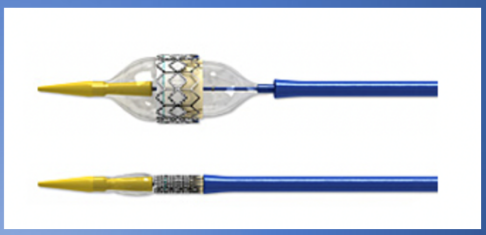


35

Aortic Valve Replacement-Young Patients

- Aortic valve disease common in ACHD population
  - Often present in 20s/30s
- Reluctance to take coumadin
  - Family planning
  - Active lifestyle
- Promise of TAVR
  - More biologic valves

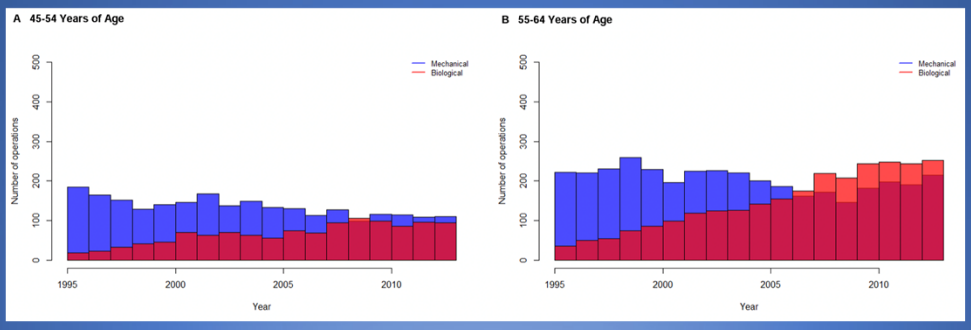




36

Mechanical or Biologic Prostheses for Aortic-Valve and Mitral-Valve Replacement

The NEW ENGLAND JOURNAL of MEDICINE



Goldstone *et al. N Engl J Med.* 2017

37

Mechanical or Biologic Prostheses for Aortic-Valve and Mitral-Valve Replacement

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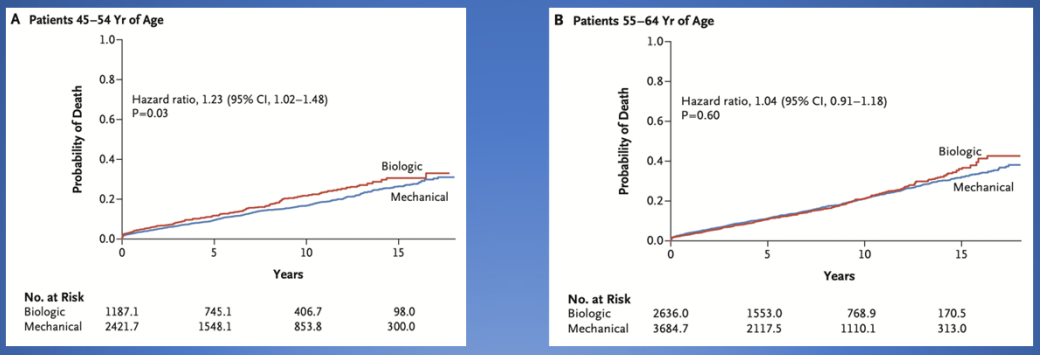


30-40% Mortality at 15 Years!

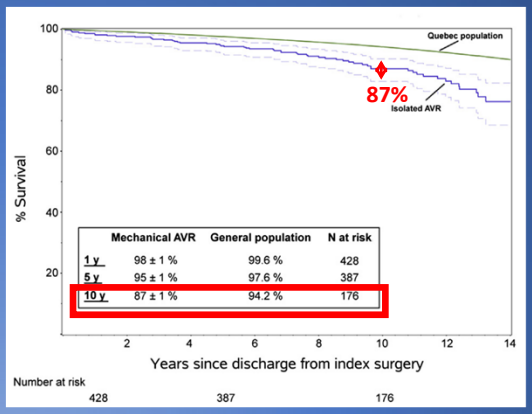
Goldstone *et al. N Engl J Med.* 2017

38

Long-term outcomes after elective isolated mechanical aortic valve replacement in young adults

The Journal of Thoracic and Cardiovascular Surgery • Volume 148, Number 4

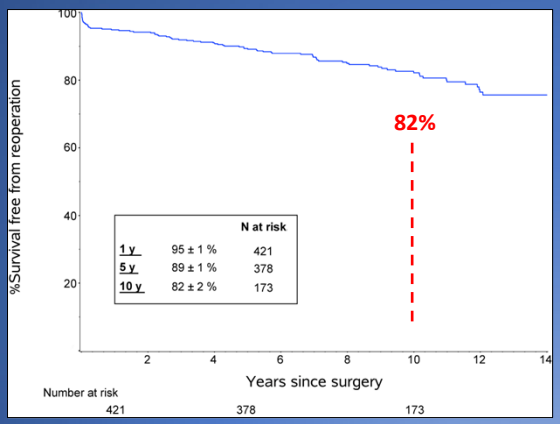
- 1997-2006: 469 isolated mAVR <65 years
- Exclusion: concomitant procedures, CAD, reop, emergencies, active endocarditis
- Mean age: 53.2 ± 9.2



Bouhout *et al. J Thorac Cardiovasc Surg*. 2014

39

Survival Free From Reoperation

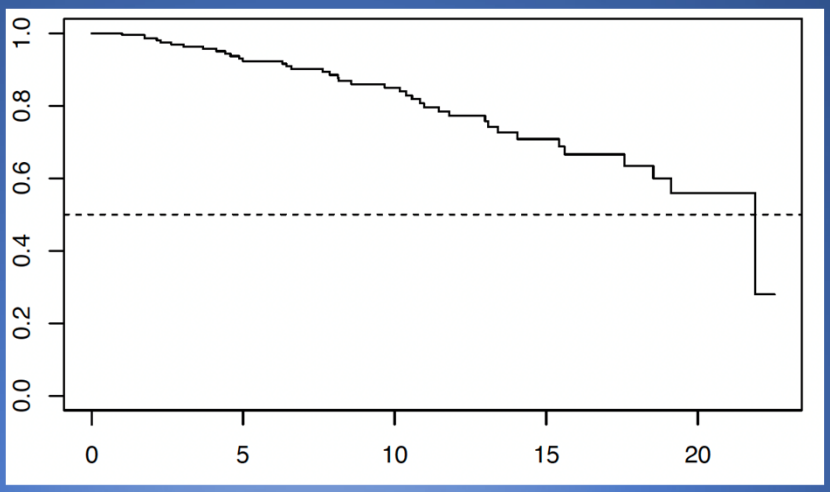


At 10 Years, 1 in 5 Patients is Dead or has had a Reoperation!!!

Bouhout *et al. J Thorac Cardiovasc Surg*. 2014

40

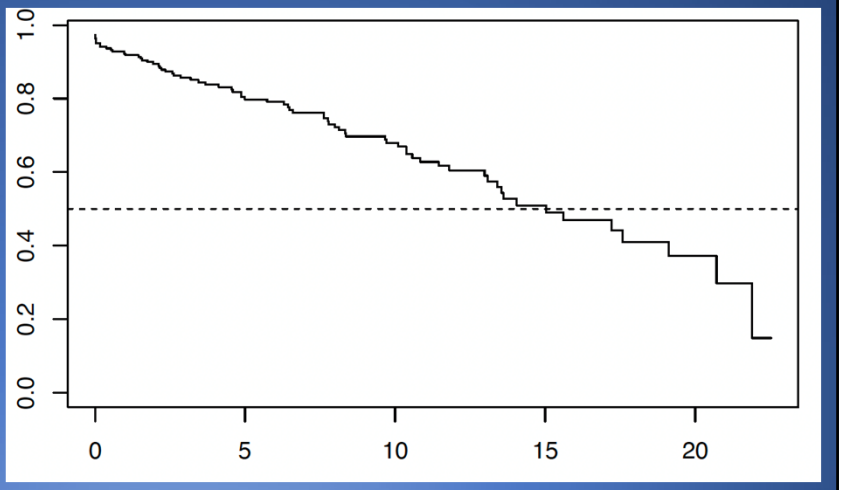
Freedom From Mortality



29% mortality at 15 years!

41

Freedom From Adverse Event



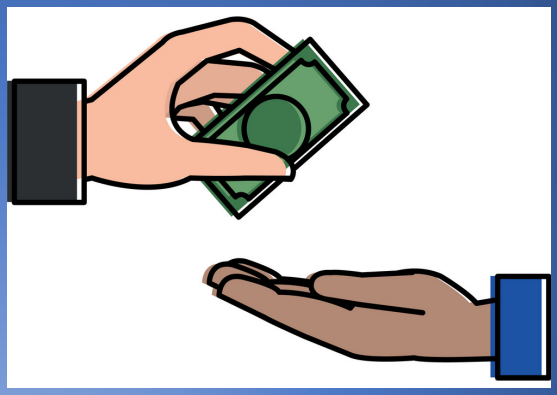
Adverse event	Frequency
Major Hemorrhage	11.8%
GI/Abdominal Bleed	8.2%
Intracranial Bleed	4.5%
Reintervention	4.5%
Embolism	4.5%
Endocarditis	2.3%

50% dead/major event at 15 years!

42

Cost of AVR in Young Patients

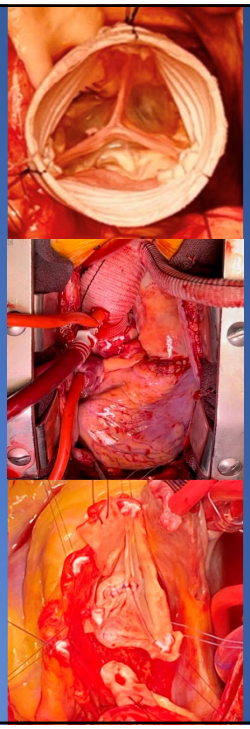
- Young and middle aged AVR patients pay a price in life-years or complications regardless of valve type
- *Can we do better in this population?*



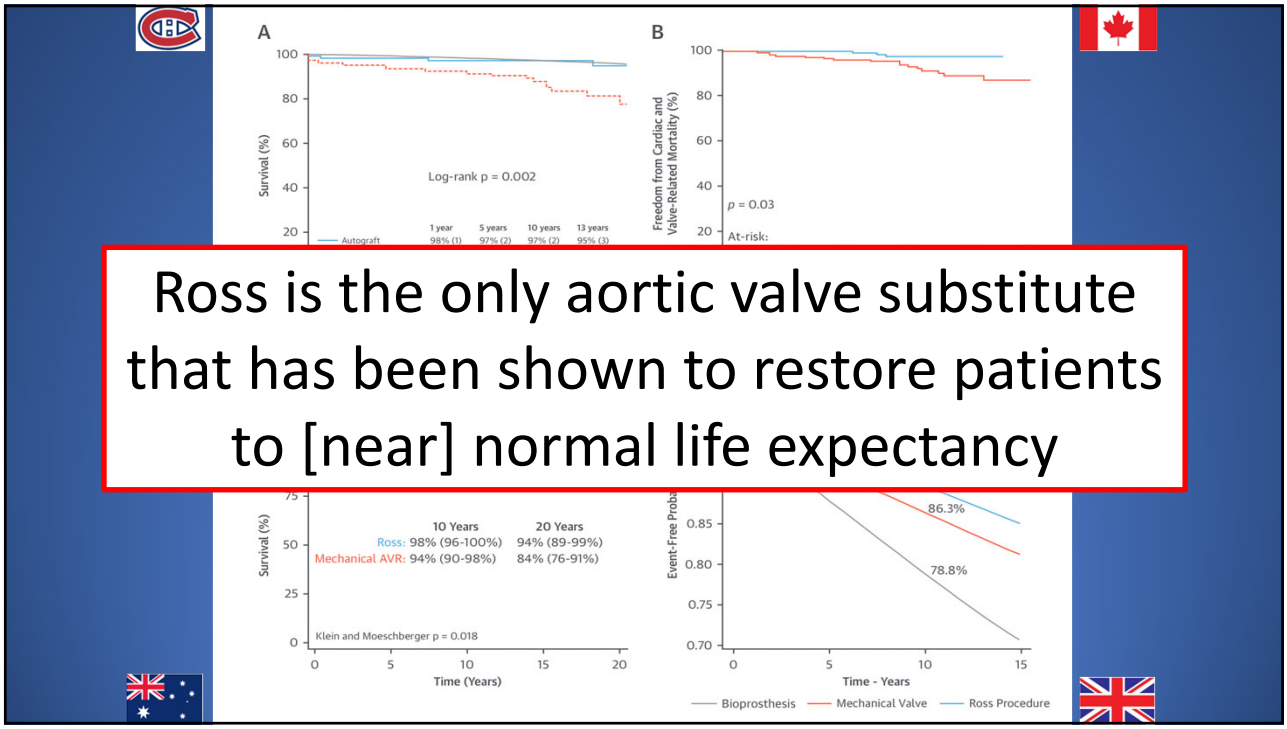
43

The "Living Aortic Valve Complex"

- Better hemodynamics
- Less infection/endocarditis
- No anticoagulation
- *Improved survival???*



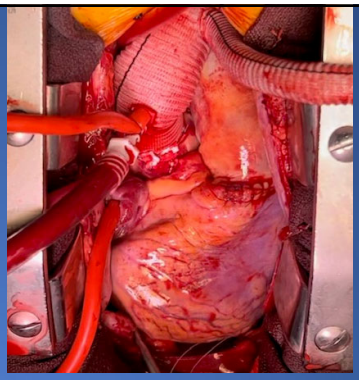
44

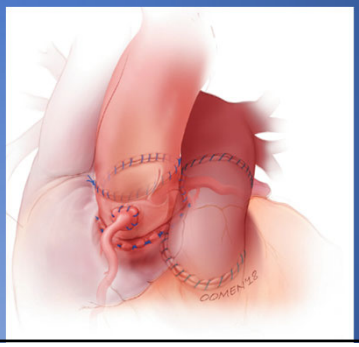


45

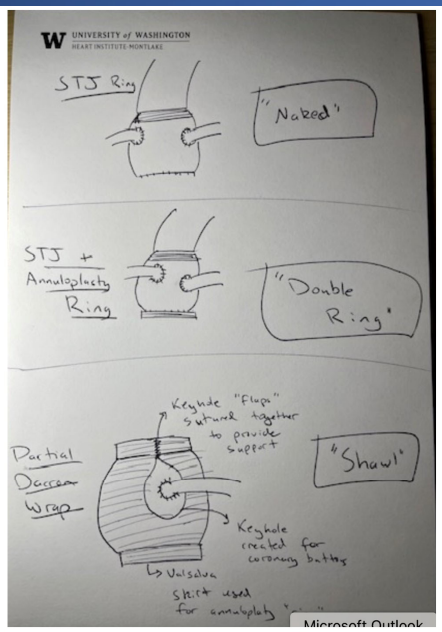
UW Ross Program

- First patient in June 2020
- Total of 90 patients to date
- Excellent clinical and valve outcomes
  - 1% mortality
  - 1% stroke
  - 98% freedom from 2+ AI





46



ALL Ross patients receive an STJ ring. Purpose: 1) Support autograft distal suture line and 2) Prevent future STJ dilation

AS predominant→ "Naked" technique (El-Hamamsy style running implant with addition of STJ Dacron ring)

AI predominant, annulus > 27 mm, aortic annulus > 2 mm greater than pulmonary annulus → "Double Ring" technique (external Dacron ring at annulus and STJ)

AI predominant with dilated (40-50mm) or aneurysmal* (> 50mm) root→ "Shawl" (Valsalva skirt anchoring as external annuloplasty, graft "keyholed" for coronary buttons and reattached to provide "partial" Dacron support)

*very rare

47

Outcomes-UW Ross program

- 5.6% reintervention
  - 1 take-back for bleeding
  - 1 pseudoaneurysm at 6 months
  - 2 balloon dilation of homograft

Table 3. Short- And Long-Term Outcomes after Ross Procedure

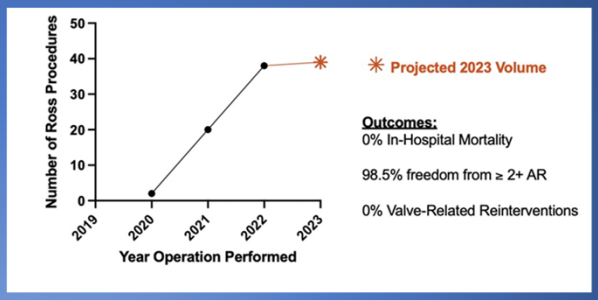
Characteristic	N = 71
Short-Term	
Tracheostomy	0 (0%)
New CVVH/HD	1 (1.4%)
Permanent Pacemaker Insertion	5 (7.0%)
Reoperation	2 (2.8%)
ICU Transfusion	4 (5.6%)
ICU Length of Stay	3 (+/-2)
Atrial Fibrillation Requiring Intervention	12 (17%)
Pneumonia	4 (5.6%)
Stroke (within 24 hours)	0 (0%)
TIA	0 (0%)
Prolonged inotropes (>48 hours)	5 (7.0%)
Wound infection	1 (1.4%)
In-hospital Mortality	0 (0%)
Long-Term	
Mortality During Follow-Up	1 (1.4%)
Valve-Related Reintervention	0 (0%)
Any Reintervention	4 (5.6%)
Pregnancy	2 (2.8%)

/ n (%); Mean (SD)
CVVH/HD: Continuous veno-venous hemofiltration/Hemodialysis
TIA: Transient Ischemic Attack
ICU: Intensive Care Unit

48

Lessons Learned

- Delicate between rapid programmatic growth and optimal outcomes
- Dedicated and specialized team essential
  - Congenital and aortic partnership
- "Buy-in" from all involved
  - ICU, outpatient→BP control
- Follow-up essential



49

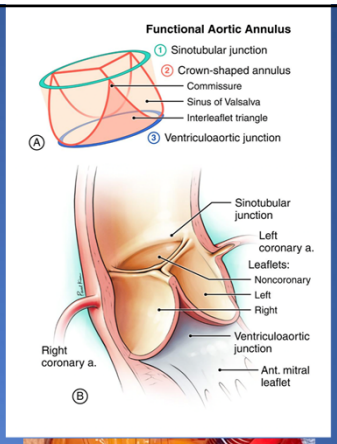
Aortic Valve Repair

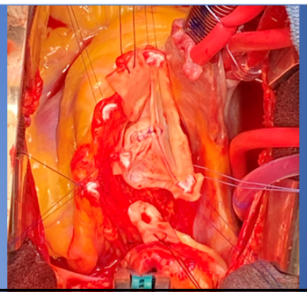
- Reserved for patients with primary AI
- Commonly associated with root aneurysm→ VSRR
- **Most BAV AI patients can be repaired**
- Rapid area of growth and innovation

50

Repair Techniques

- AI comes from pathology within→ **annulus, STJ, or leaflets**
- Repair techniques address:
  - Annuloplasty
  - STJ ring
  - Leaflet repair/plication

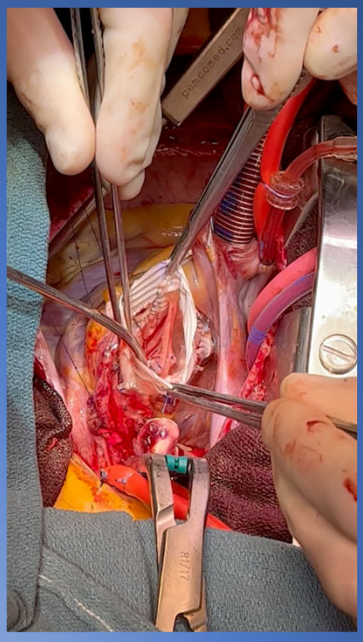




51

Case #1-AVr

- 21 yo M with severe BAV AI and 4.9 cm root
- David V with BAV repair
- Post→ no AI, MG 7 mmHg

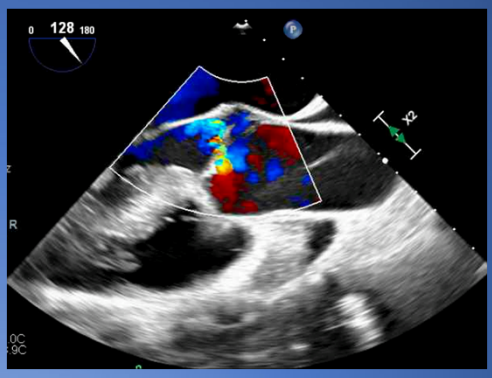


52

Case #2-AVr

- 40s M with root aneurysm, TAV, moderate AI
- Hx of bleeding-concerned about anticoagulation

TAV with eccentric AI jet!!



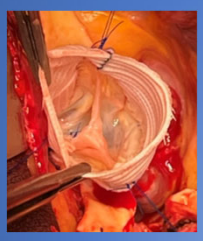
53

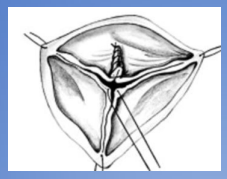
Case #2-AVr

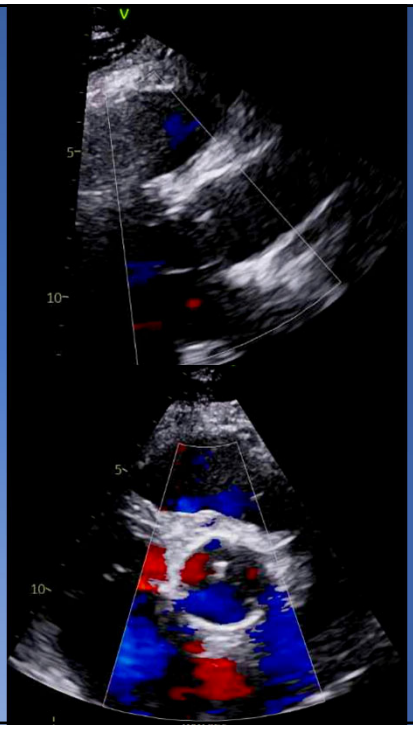
Elongated NCC

Prolapse of RCC

David V with asymmetric commissural reimplantation and plication of RCC

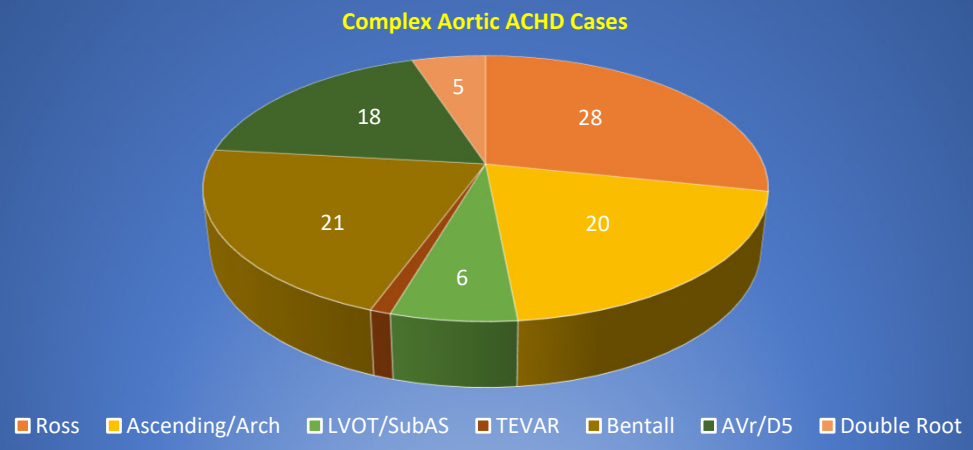






54

Aortic ACHD Cases 2020-2022 (N=75)



55

Final Thoughts

- ACHD population continues to grow and will need surgical and transcatheter intervention
- Surgery on the aortic valve or ascending aorta is common
- The expertise of a surgeon familiar with complex aortic techniques is critical for the success of an ACHD program
- Partnership between a congenital surgeon and aortic surgeon offers the most comprehensive care for this complex population

56

57