Children's Hospital Colorado
Heart Institute

HEART
INSTITUTE

Risk Factors in ACHD Redo Surgery: Strategies to Optimize Outcomes

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ACHD 2017 Symposium, Nov 30-Dec 2, Queenstown, New Zealand

NO DISCLOSURES

Adult Congenital Heart Disease: Facts

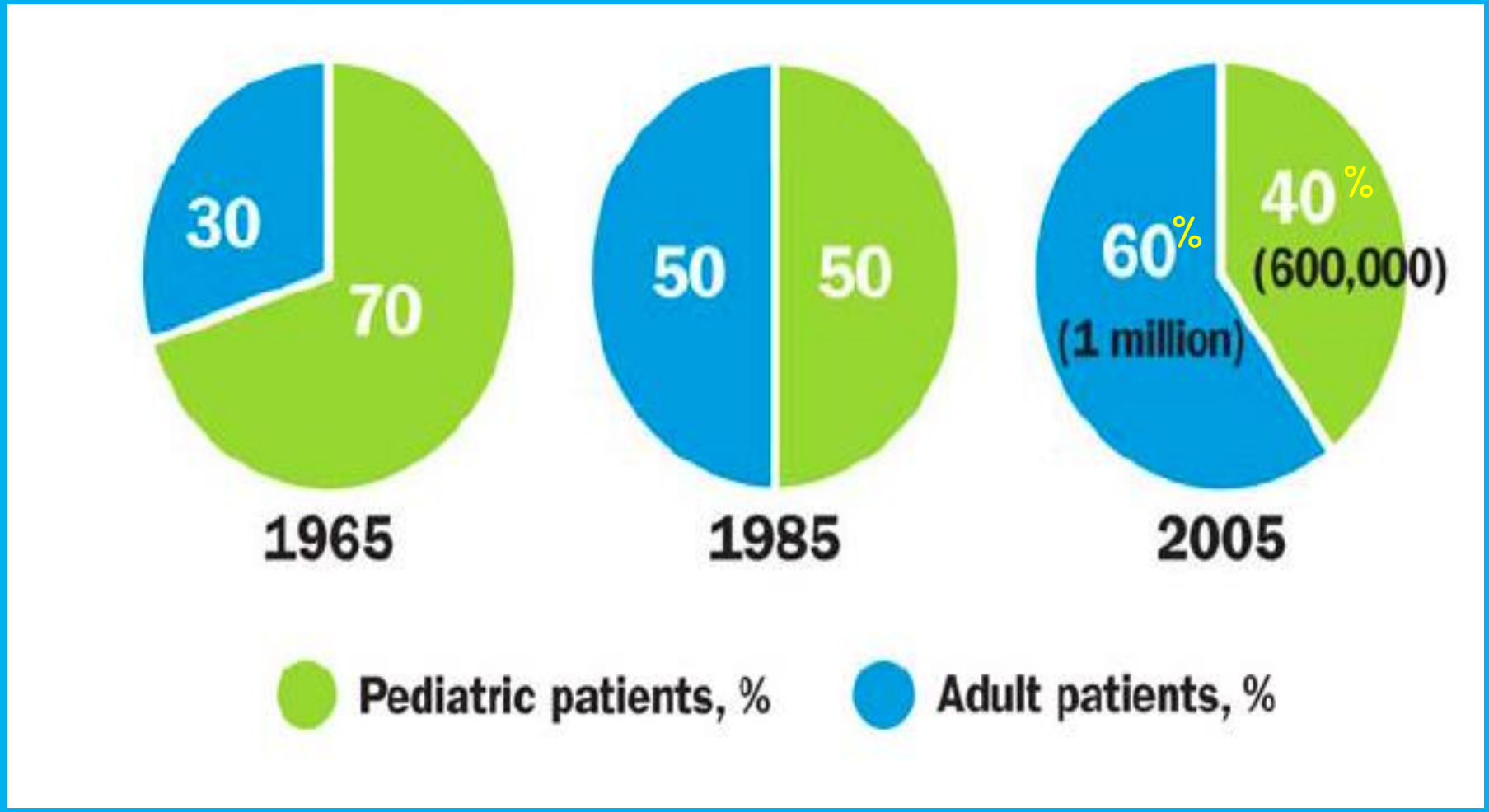
- Congenital Heart Disease is the most common form of Congenital defect
- Most forms of CHD can not be considered curable and have residual sequelae
- Transition of care from childhood to adulthood is highly variable, many lost to follow-up
- In those with previous surgery, many face multiple reoperations with increasingly higher risk

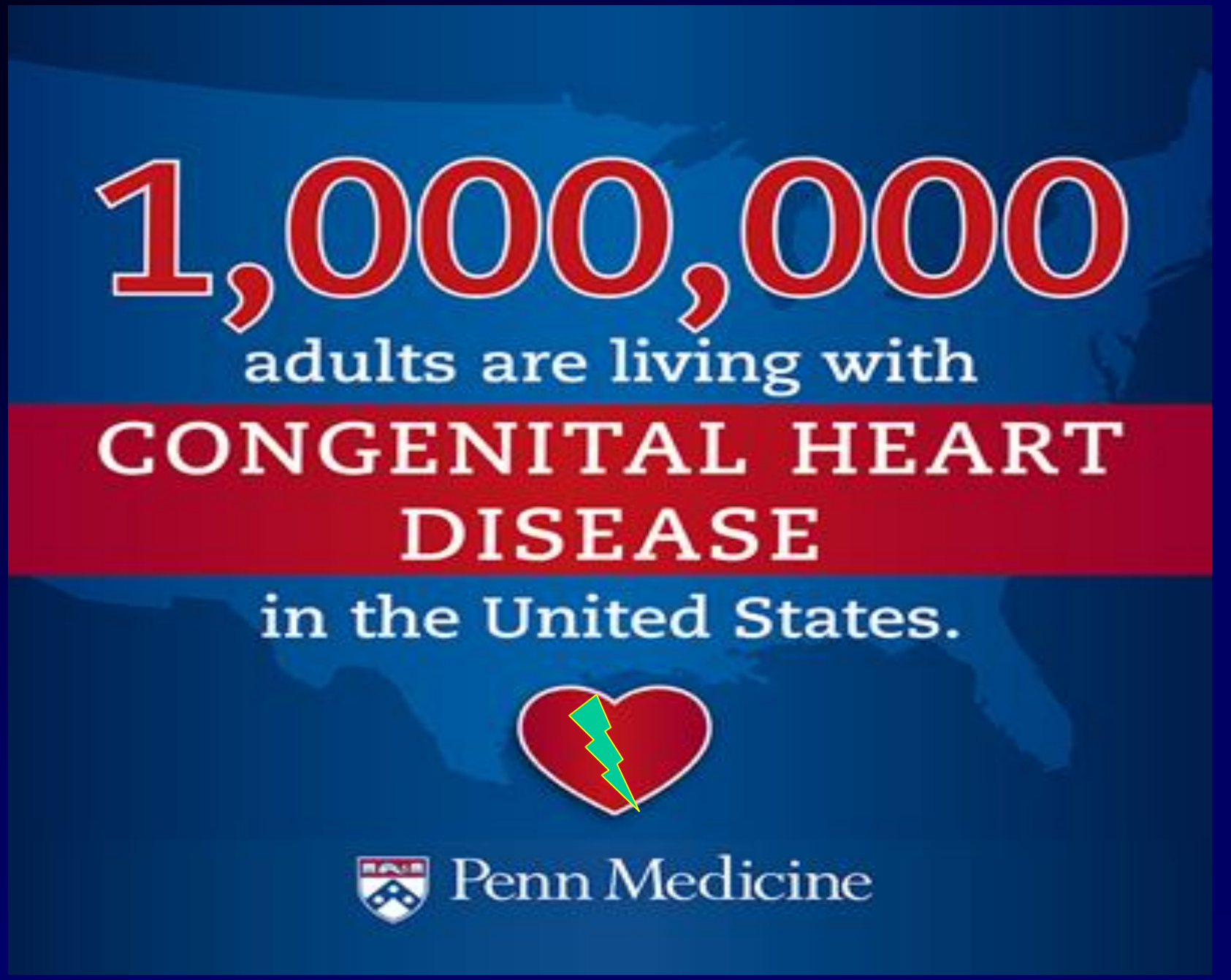
In the past, Adults with Congenital Heart Disease were uncommon and hard to spot

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

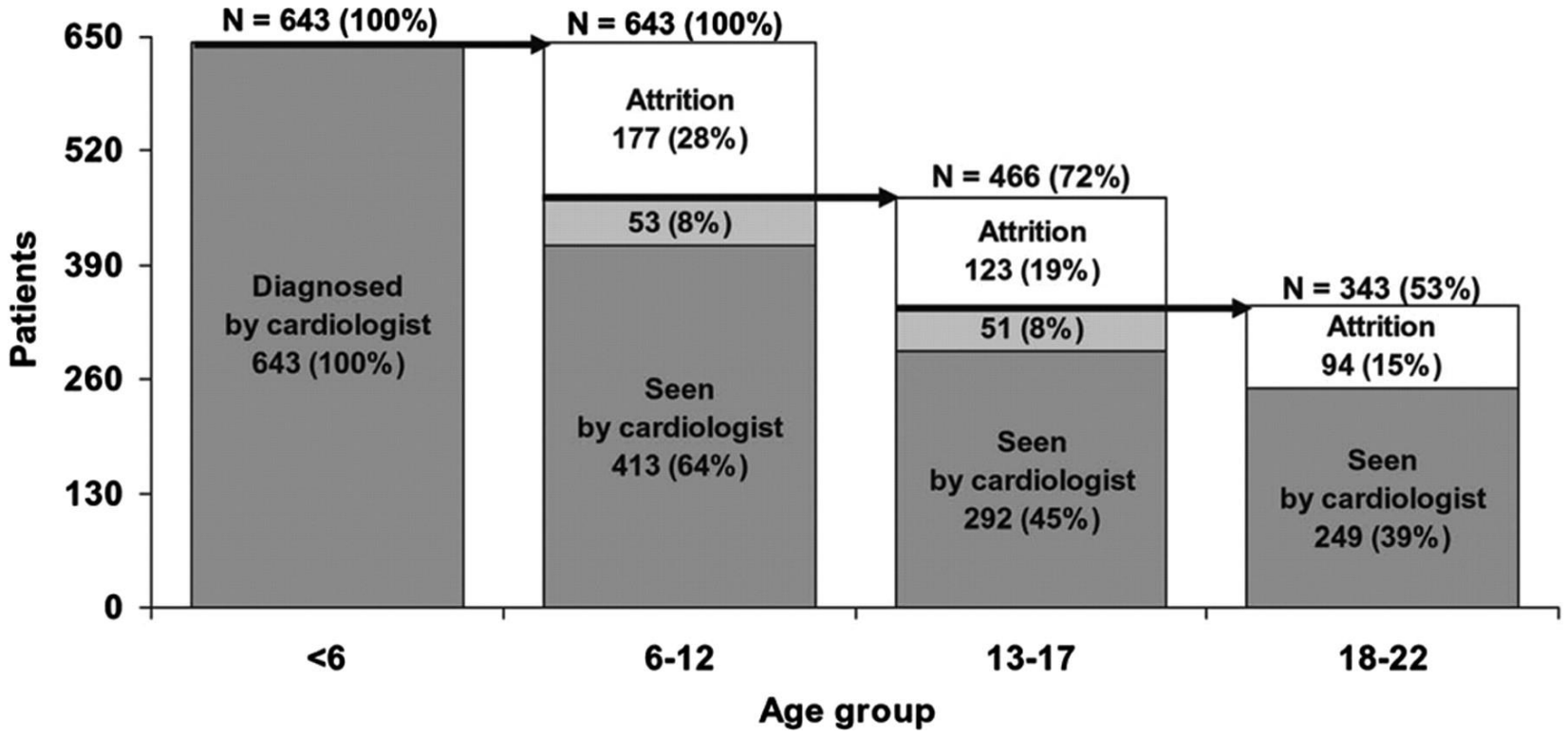



1,000,000
adults are living with
CONGENITAL HEART DISEASE
in the United States.
Penn Medicine

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

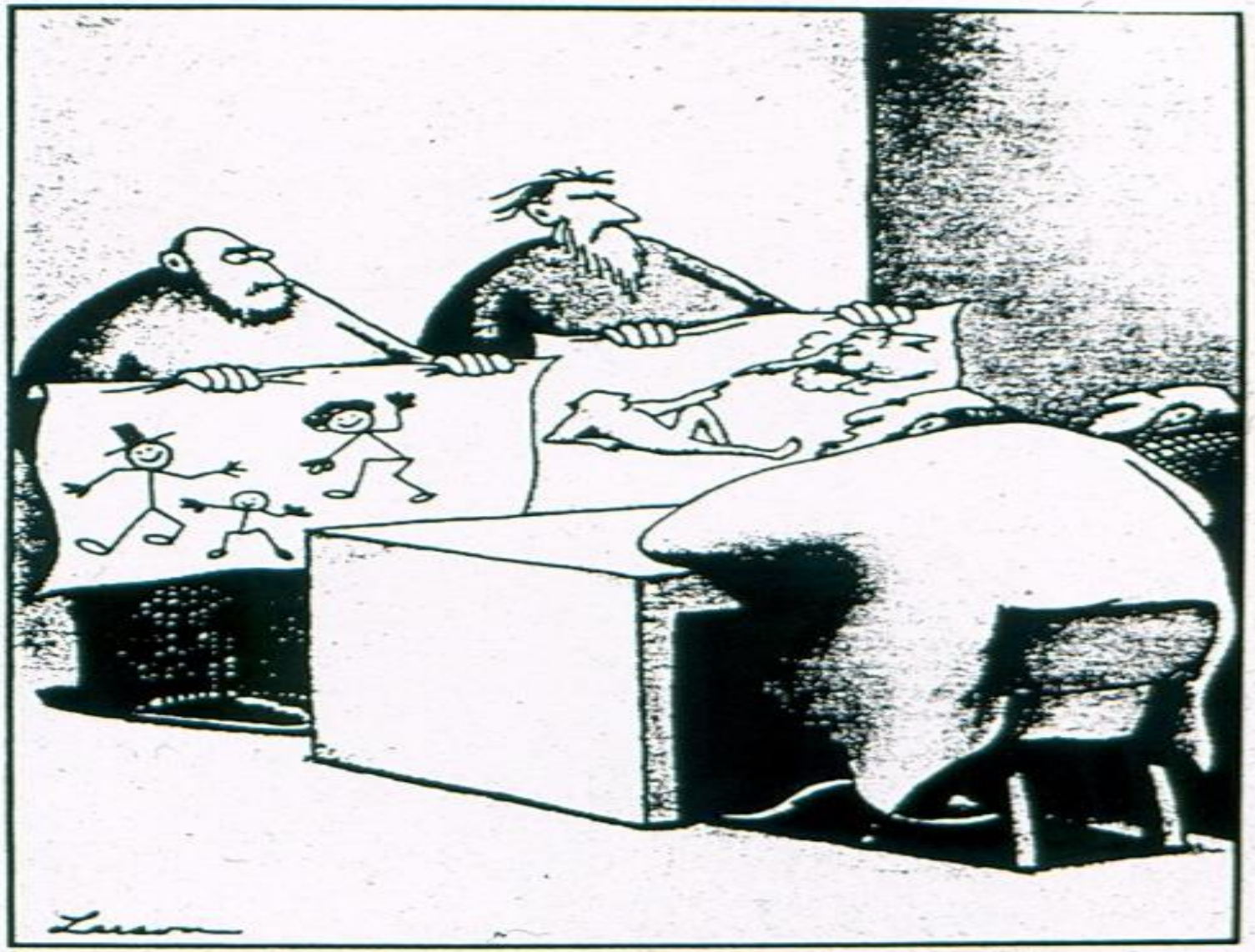
Loss of follow-up from age 6 to 22 years among the entire Quebec CHD cohort



Mackie A S et al. Circulation 2009;120:302-309

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 ★

Kogon,B Annals Thorac Surg 2013;95:1377-82

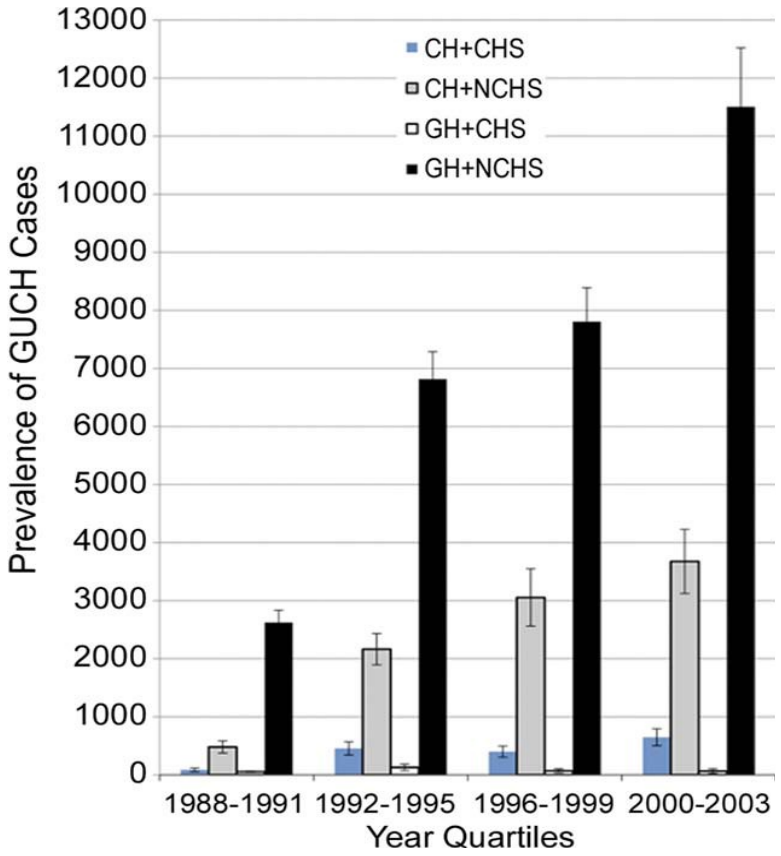
National Practice Patterns for Management of Adult Congenital Heart Disease

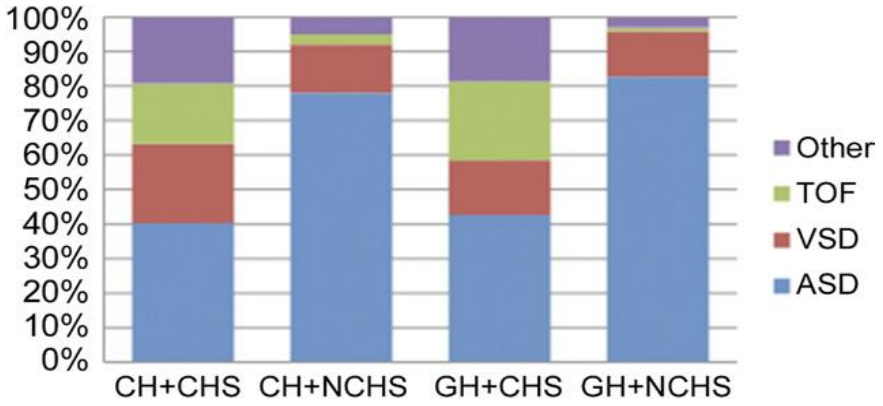
Karamlou et al Circulation. 2008;118:2345-2352

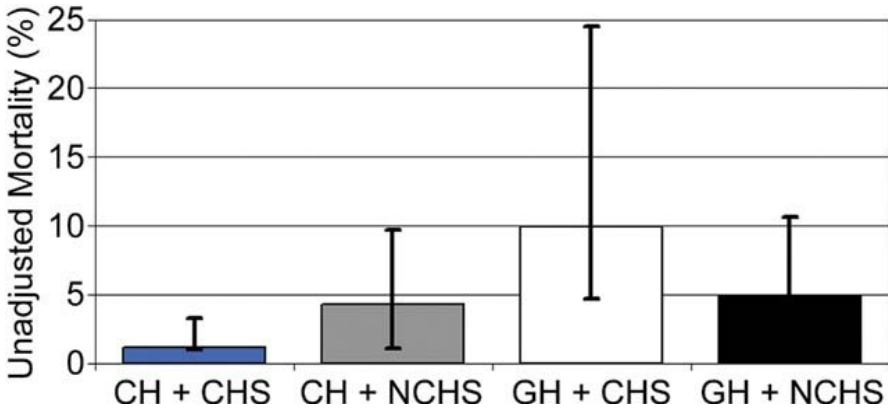
National Practice Patterns for Management of Adult Congenital Heart Disease

- 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?



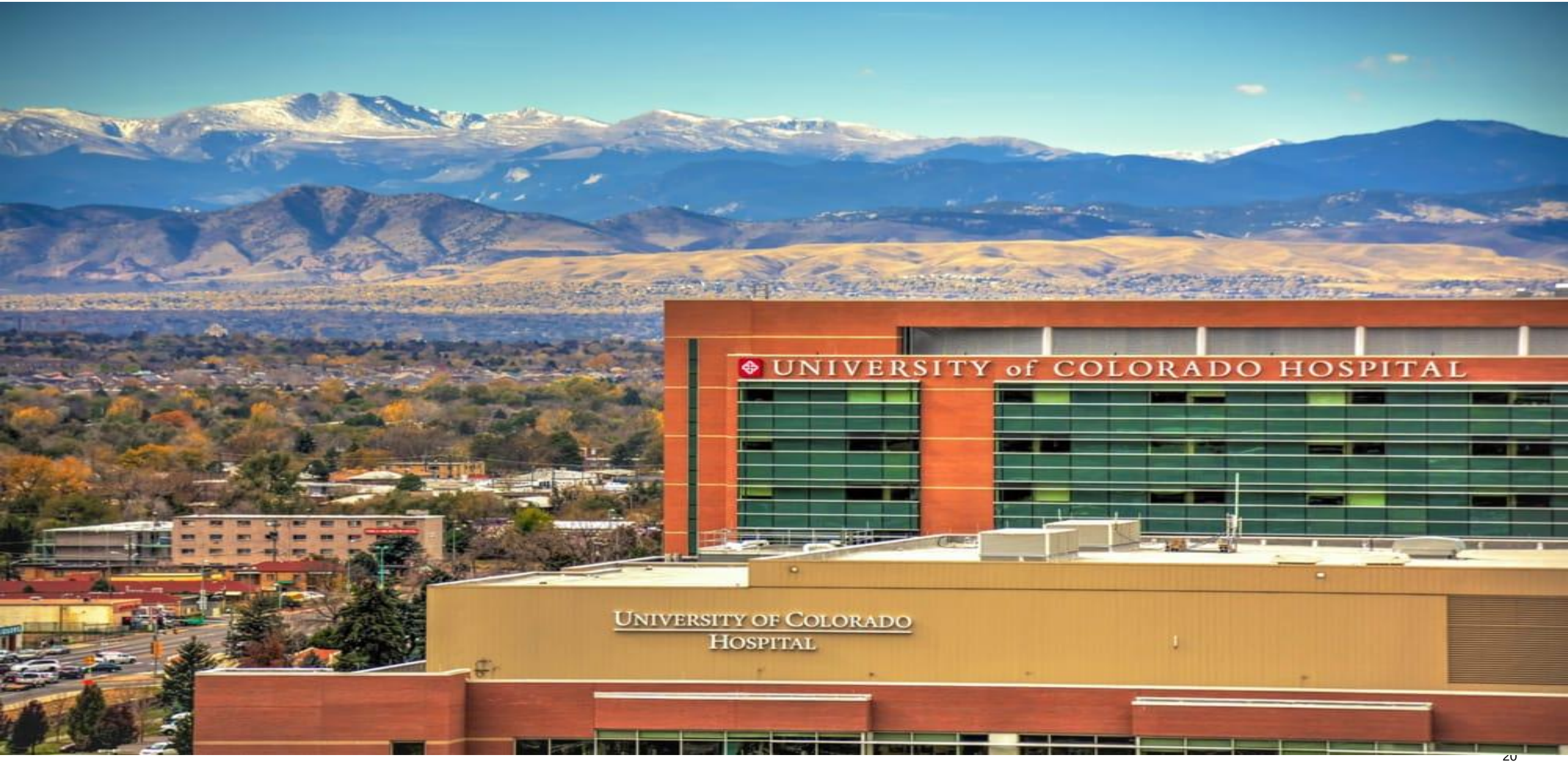




Karamlou, Ann Thorac Surg 2010;90:573–9

Our ACGME Congenital Surgery training program requirements still have not acknowledged any need for special training in ACHD

OUR STRATEGY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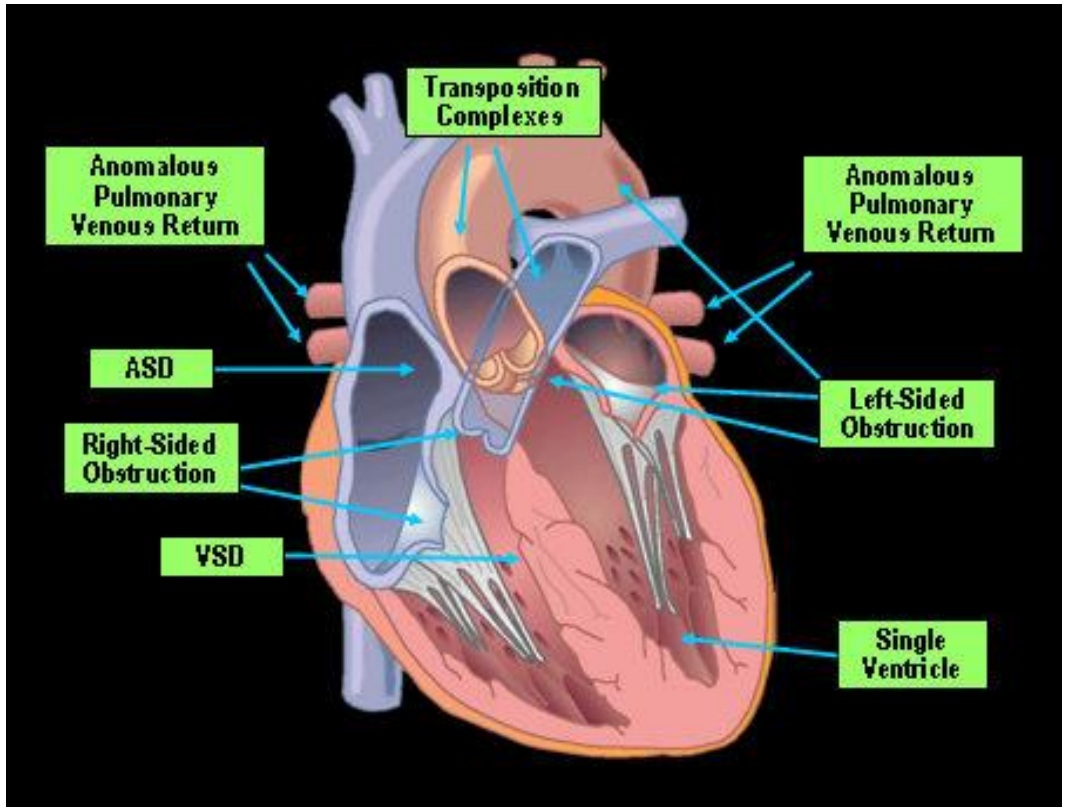
HOSPITAL


TH
KNOW THE
OPERATIONS
AND RISKS

ACH Surgery Mortality: STS Database Analysis

Mascio,C, *J Thorac Cardiovasc Surg* 2011;142:1090-1097



- Overall (n = 5265) 109 (2.1%)
- Pulmonary valve replacement (n = 574) 4 (0.7%)
- ASD repair (n = 365) 0 (0%)
- Conduit operation (n = 328) 8 (2.4%)
- Aortic aneurysm repair (n = 136) 3 (2.2%)
- Mitral valvuloplasty (n = 135) 1 (0.7%)
- Ross operation (n = 108) 2 (1.9%)
- Fontan revision/conversion (n = 105) 11 (11%)

PULMONARY VALVE REPLACEMENT, ASD, and RVOT CONDUIT OPERATION MOST COMMON

Changing Practice of Cardiac Surgery in ACHD

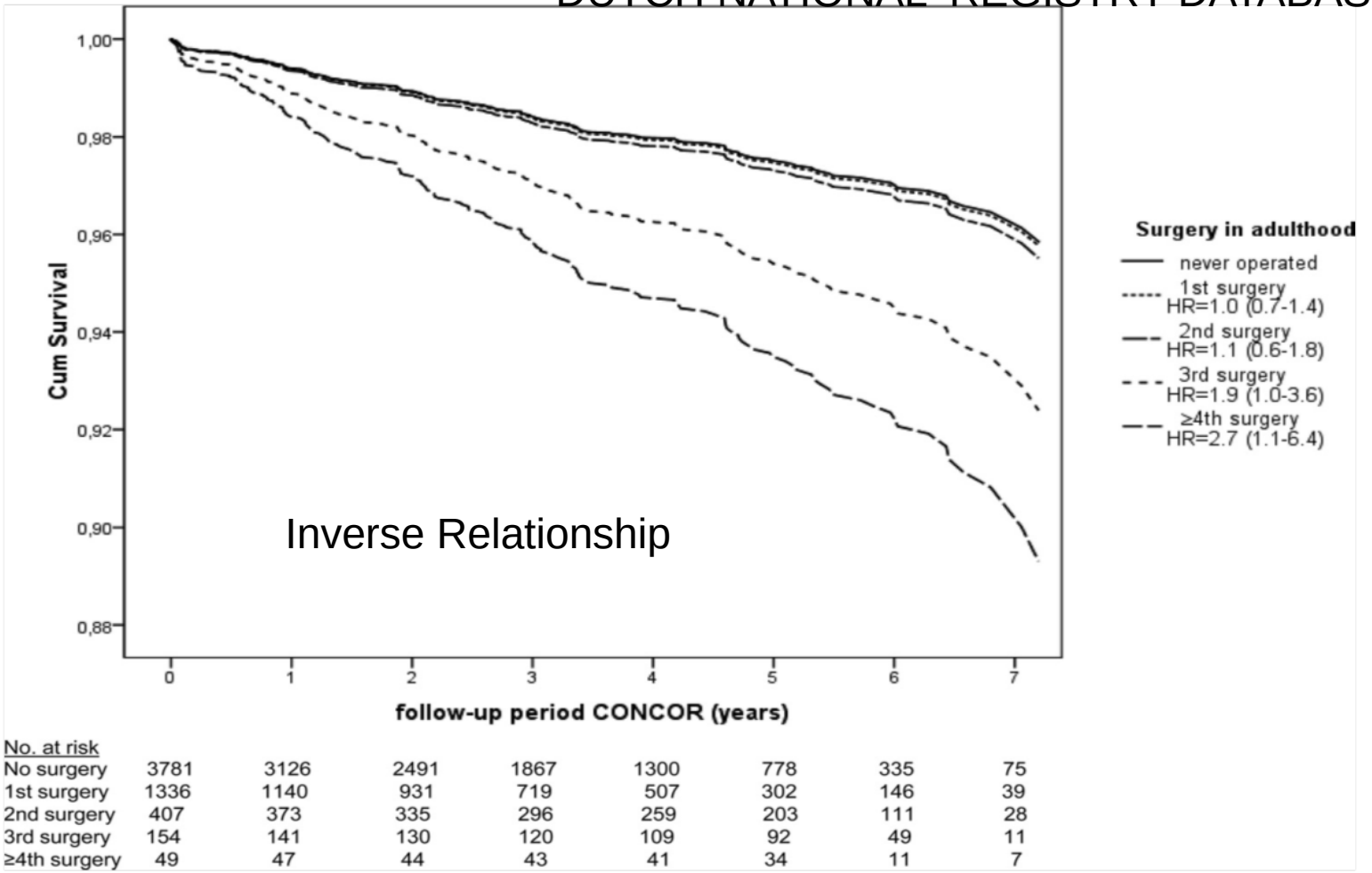
- Patients operated during 2 different time periods- 1990-1994 and 1998-2002 single GUCH unit
- Simple operations(ASD's) significantly decreased 45% to 27%
- Repeat operations increased from 25% to 50%

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

DUTCH NATIONAL REGISTRY DATABASE



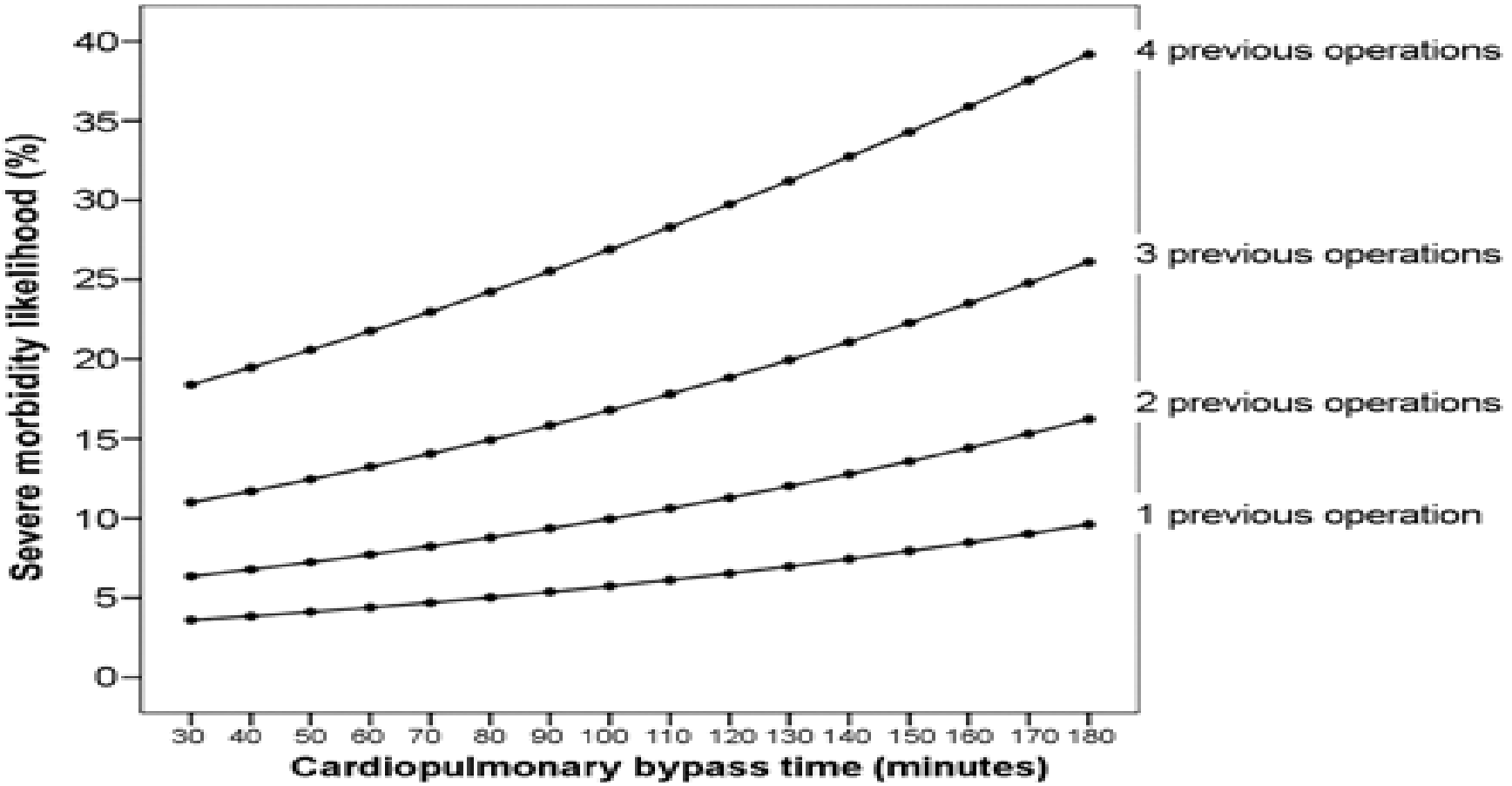
No. at risk								
No surgery	3781	3126	2491	1867	1300	778	335	75
1st surgery	1336	1140	931	719	507	302	146	39
2nd surgery	407	373	335	296	259	203	111	28
3rd surgery	154	141	130	120	109	92	49	11
≥4th surgery	49	47	44	43	41	34	11	7

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



Giamberti *Ann Thorac Surg* 2009;88:1284-1289

Milan, Italy

Risk Factors and Early Outcomes of Multiple Reoperations in Adults with Congenital Heart Disease

Dearani, j Ann Thorac Surg 2011;92:122-30

- 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


BLUE SKY AVIATION

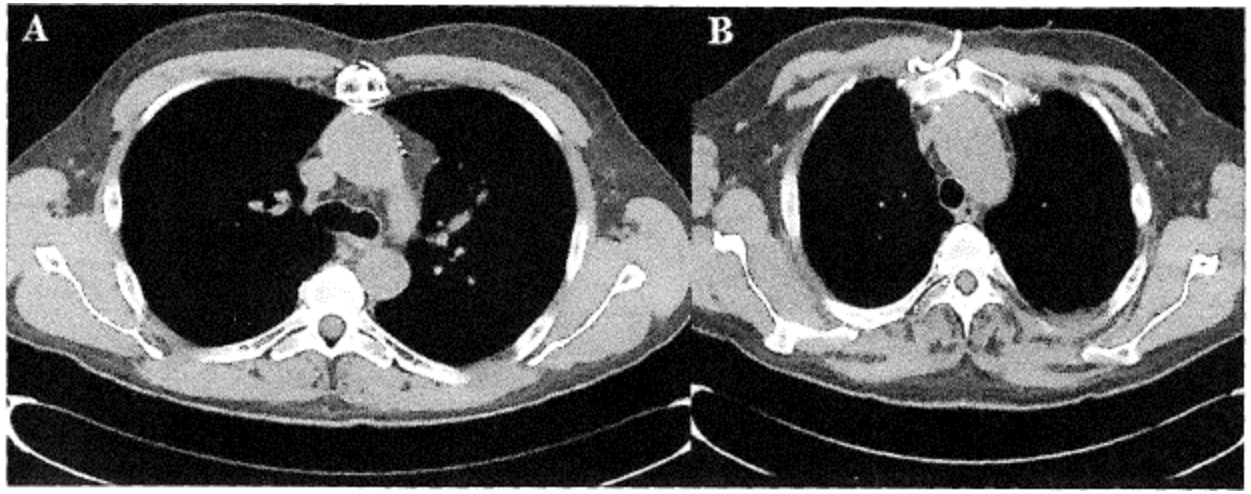
MAKE THE OPERATION AS SAFE AS POSSIBLE !

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

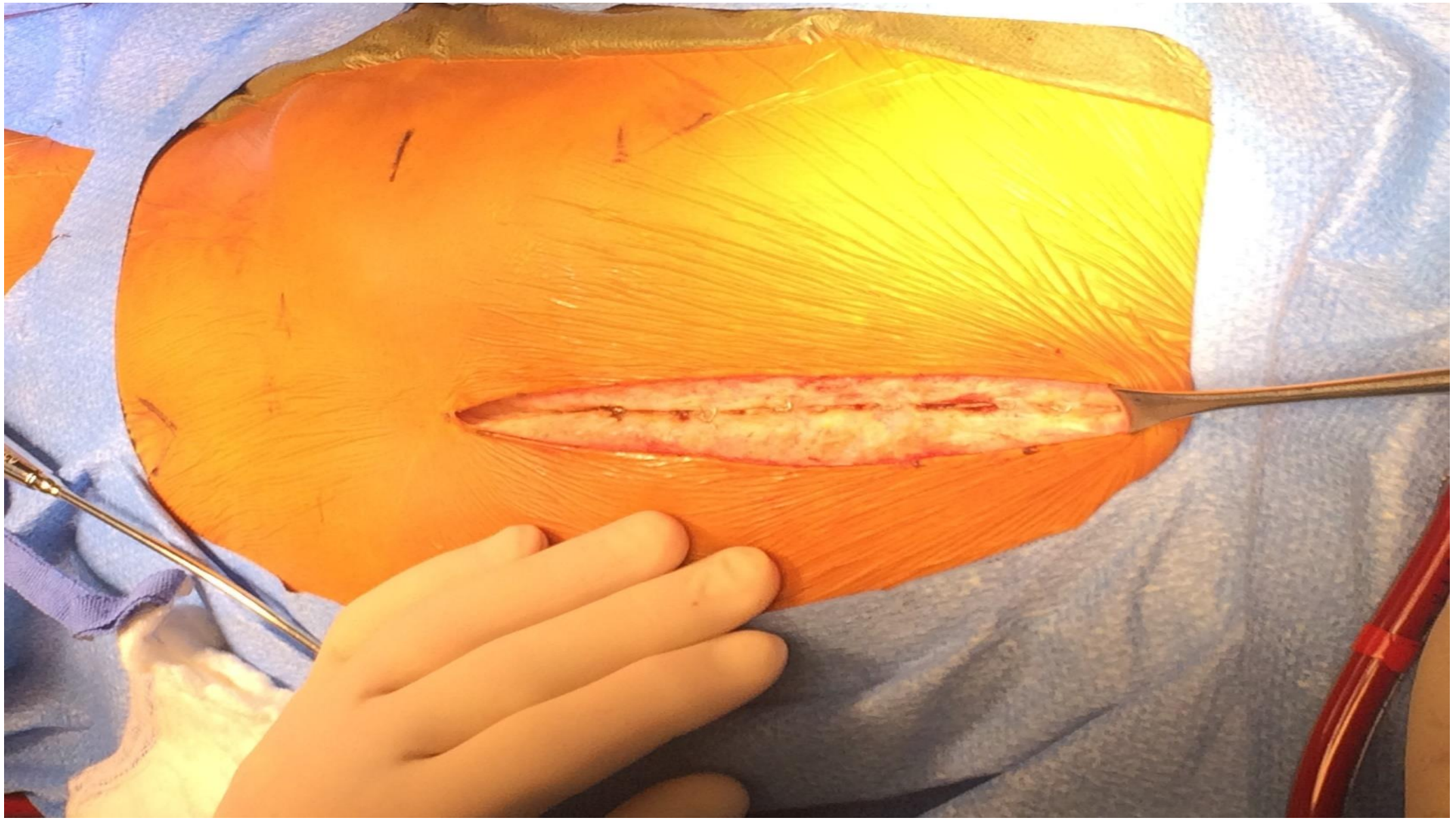
- This is crucial to the outcome since the length of CPB is a significant risk factor for early mortality

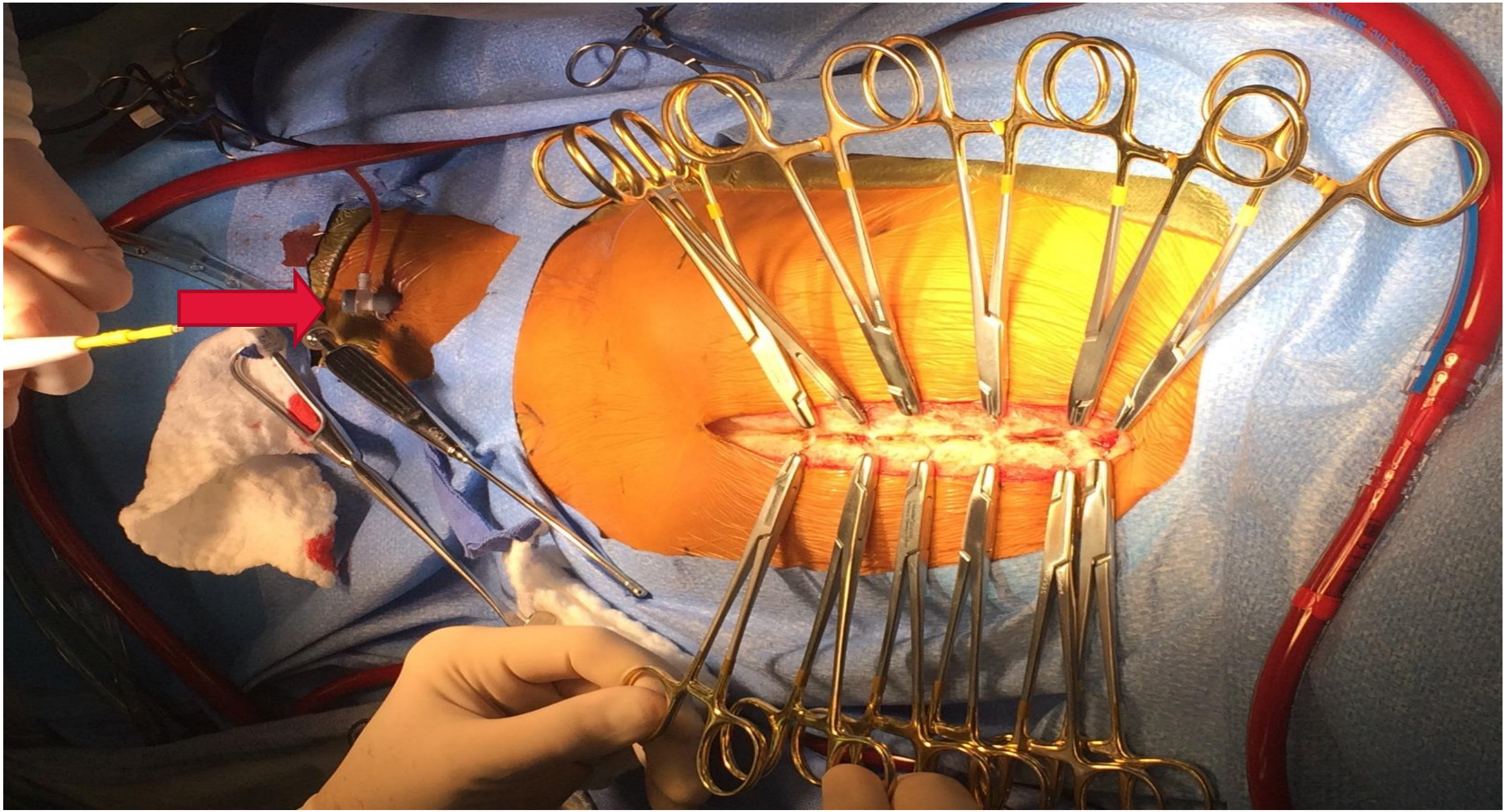
Risks of Redo Sternotomy

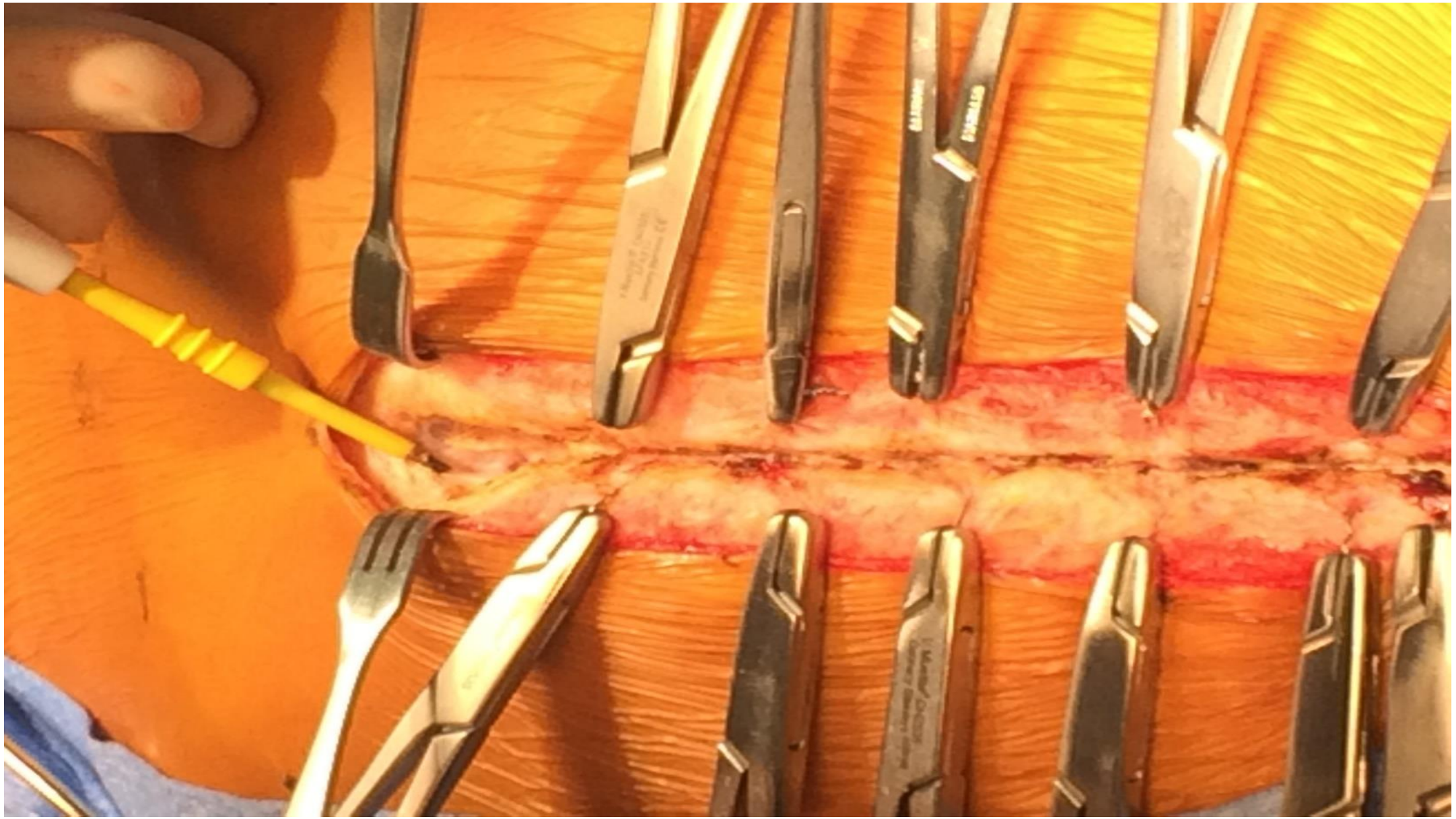
- 602 redo sternotomies
  - 67% second sternotomy, 28% third sternotomy, 4% fourth Sternotomy, .8% fifth sternotomy, .2% sixth sternotomy
- Hospital survival 98% 590/602
  - None of the 12 deaths secondary to redo sternotomy
- Major injury in 2/602: minor injury in 4/602.
  - 2 cases received transfusion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

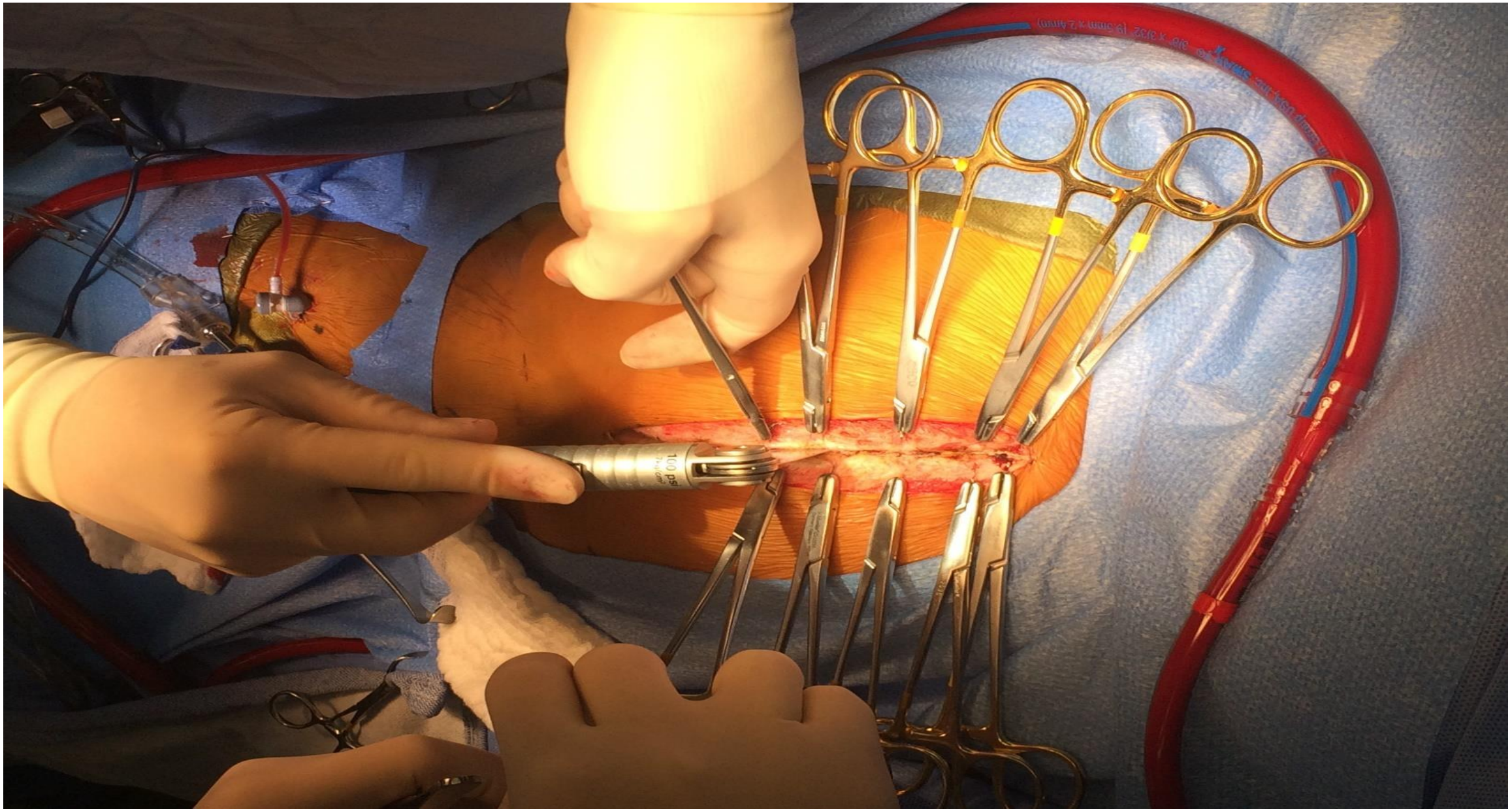
1/3 had 3 or more sternotomies

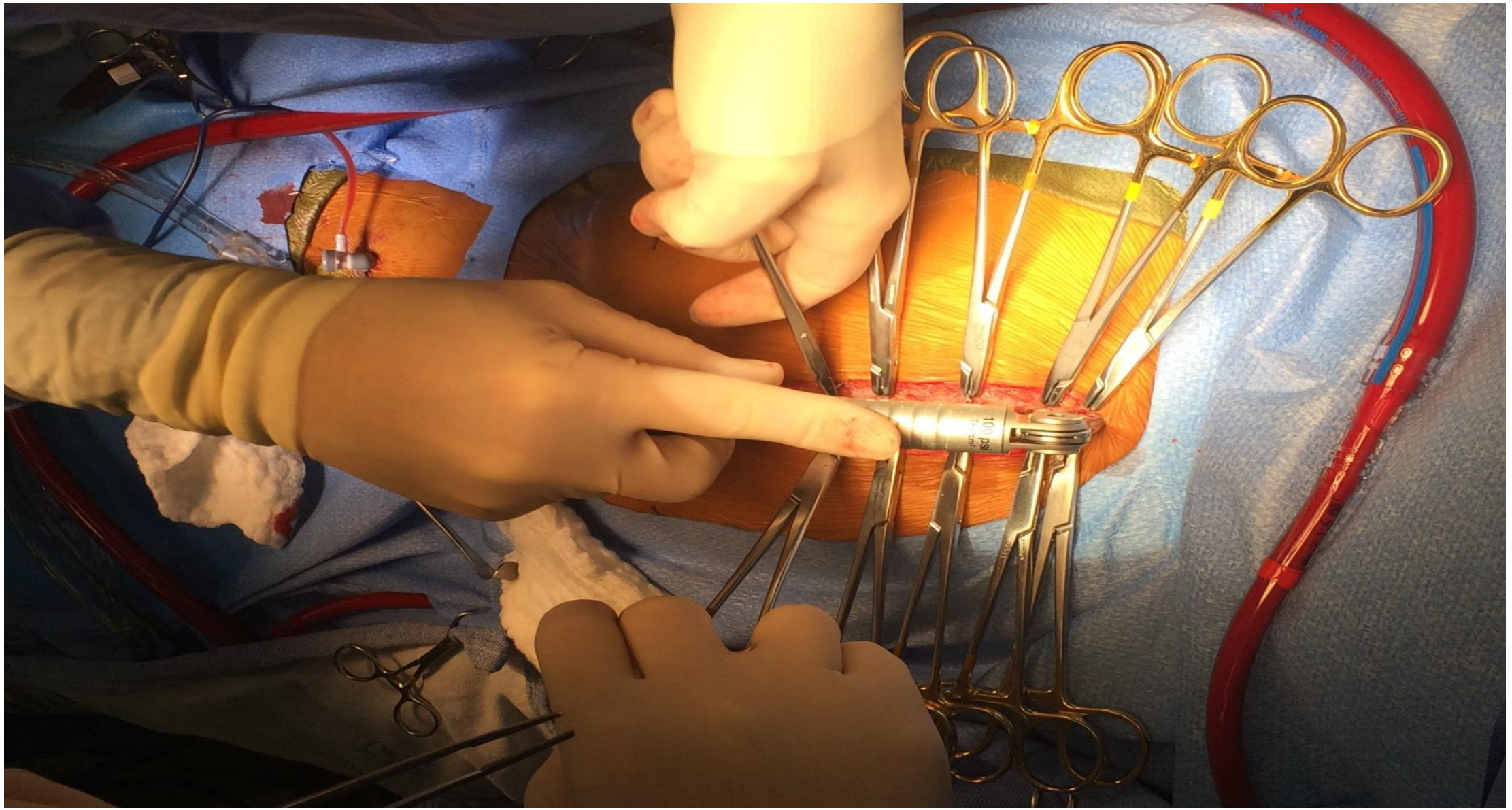
Morales, et al. Ann Thorac Surg 2008;86:897–902

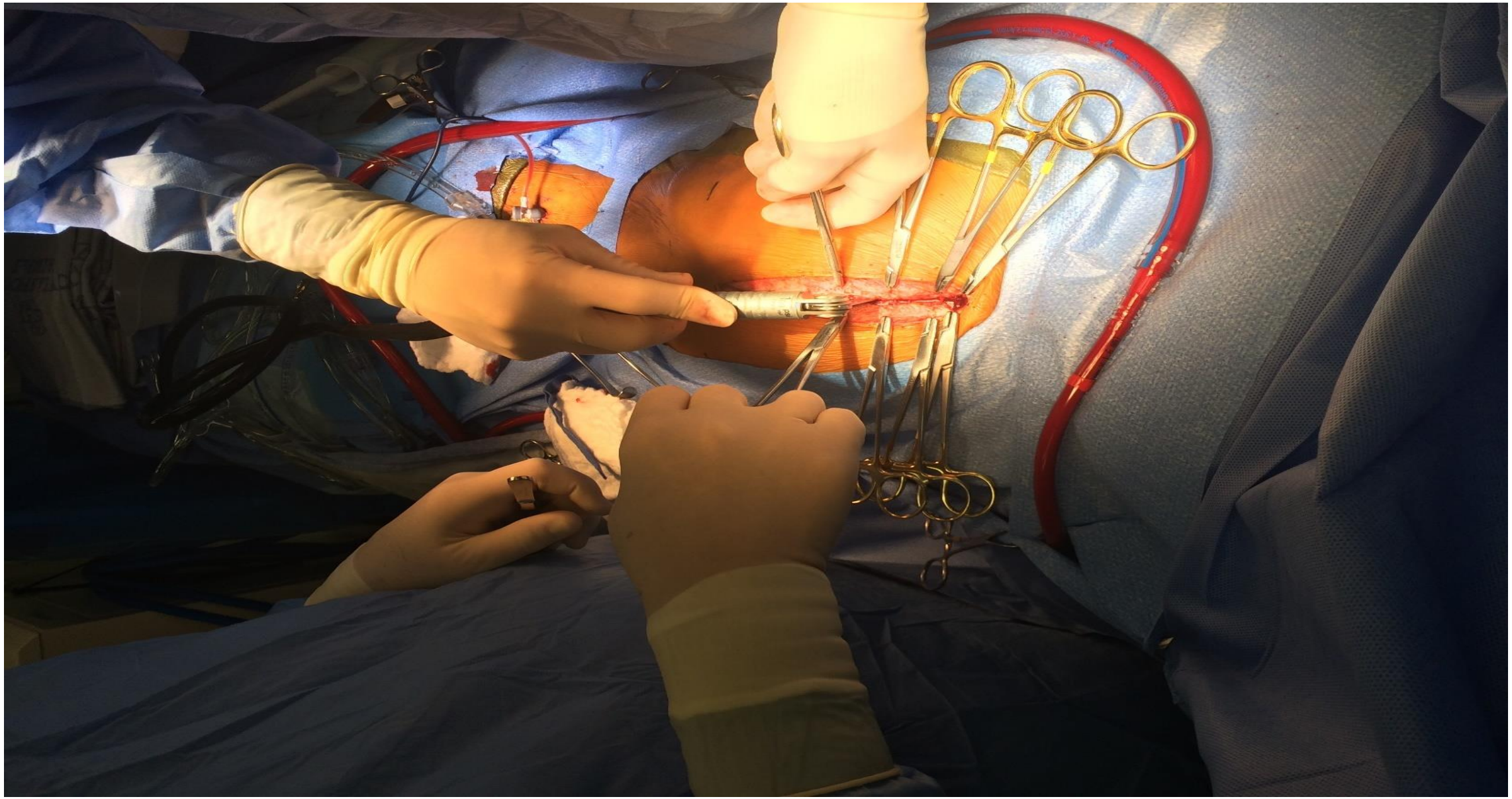


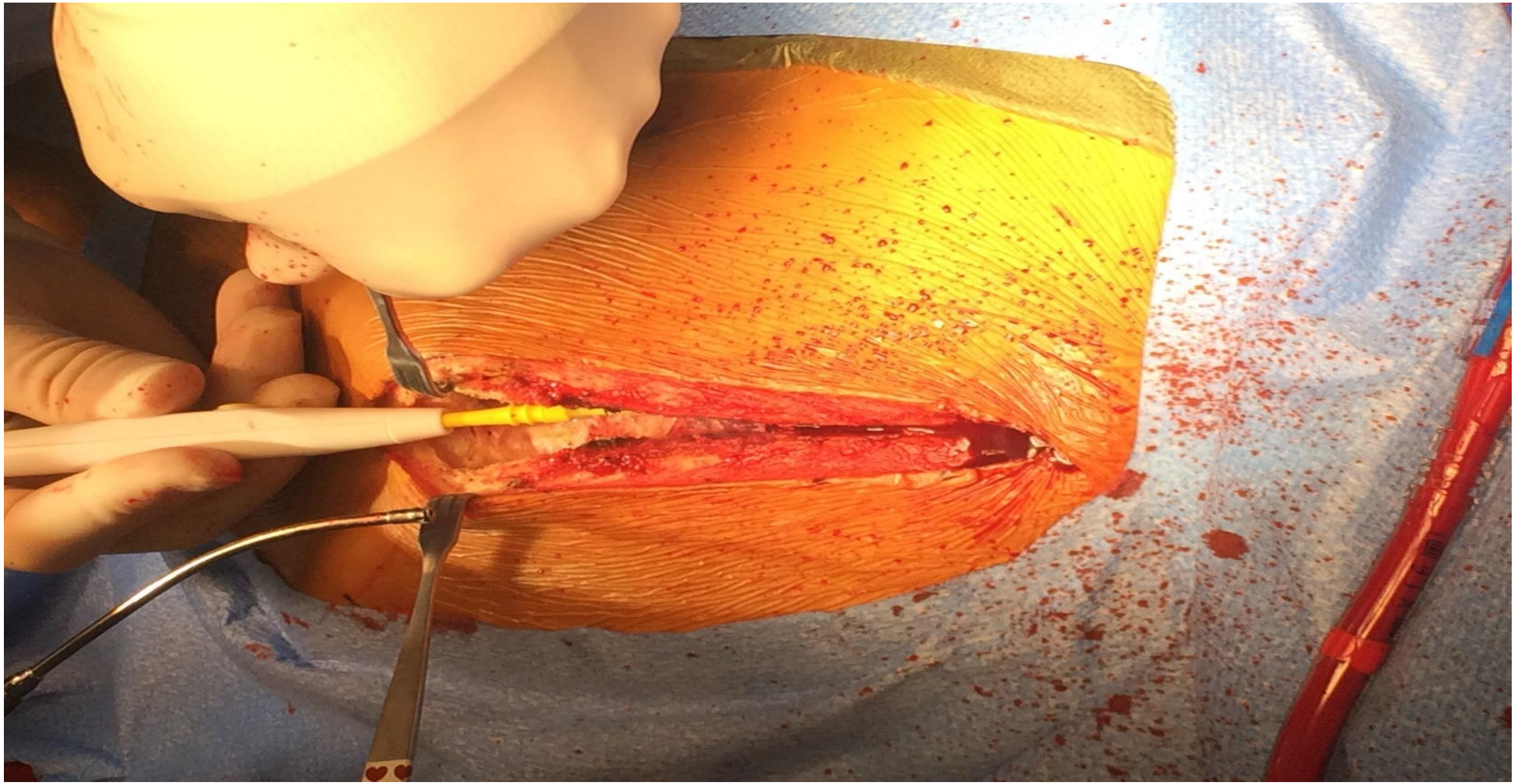


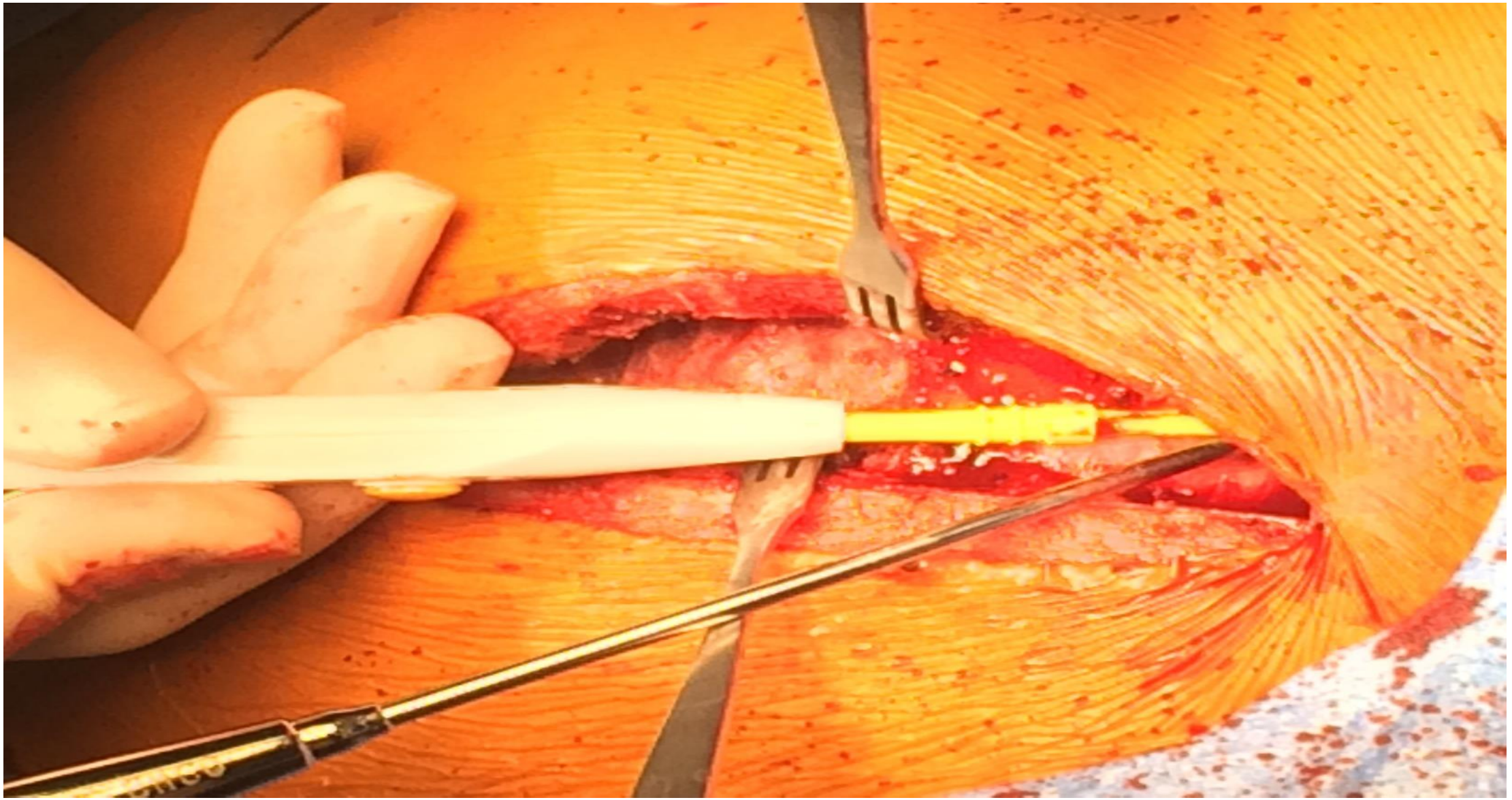












Be sure that the operation is necessary, but don't delay if it is!

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

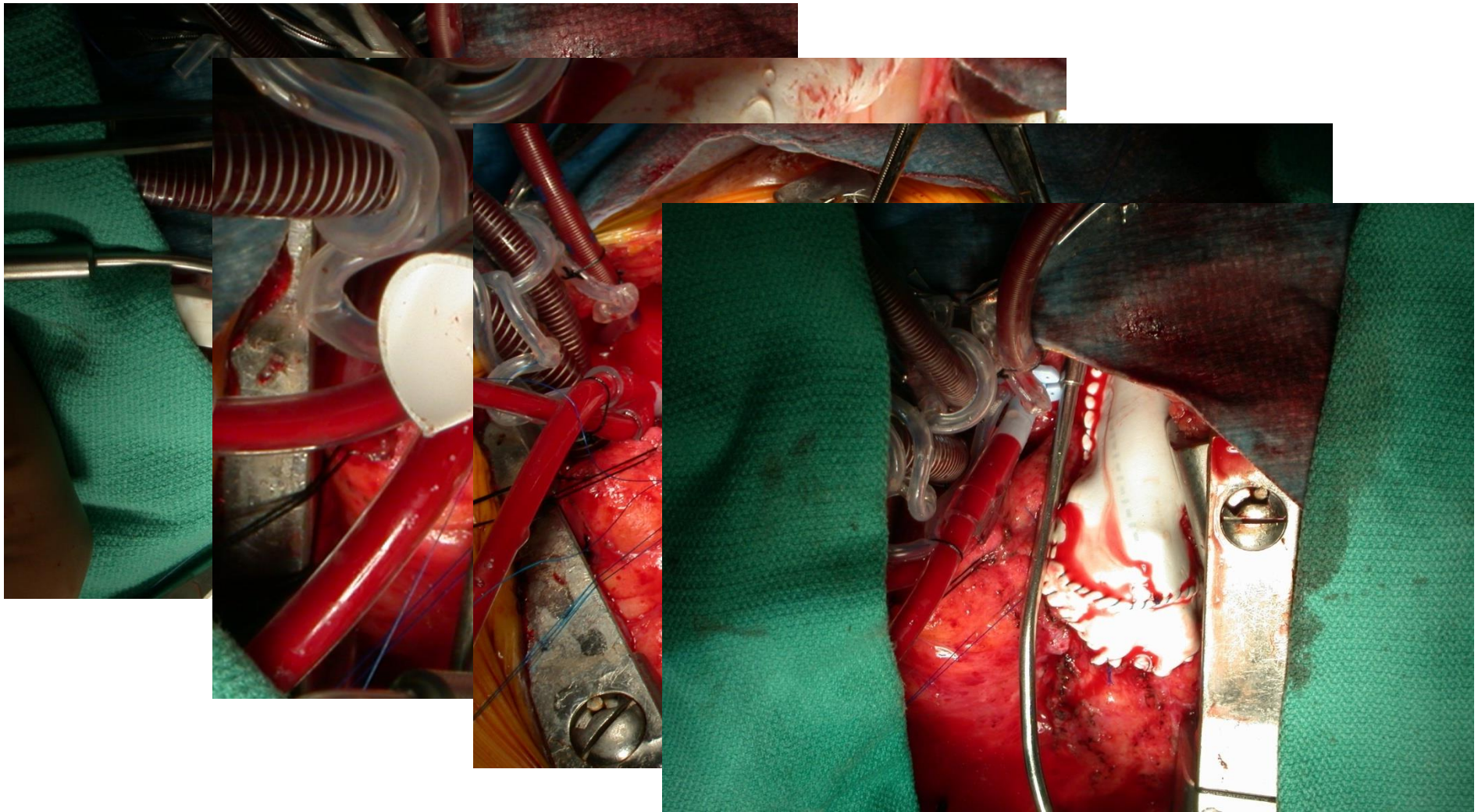
Table 2 Perioperative and late mortality of pulmonary valve replacement after TOF repair

Institution	Year	Number of Patients	Operative Death	Average Length of Follow-Up (years)	Late Death or transplant
SUNY, Syracuse [142]	1985	11	0	1	0
Children's Memorial Hospital, Chicago [143]	1997	49	1		
University of Toronto [101]	1997	85	1	5.8	3
Mayo Clinic [87]	2001	42	1		
Children's Hospital, Atlanta [144]	2002	100	1	4.9	1
Leiden University, The Netherlands [97]	2002	26	0	1.5	1
New England Med Center, Boston [105]	2003	36	0	5	1
University of Zurich, Switzerland [100]	2005	39	0	1.25	0
Multicenter, The Netherlands [145]	2006	158	0	4.2	2
University of Toronto [99]	2007	82	0	8.8	2
University Medical Center, Rotterdam [14]	2008	17	0	6.4	0
International Society of Congenital Heart Disease [107]	2008	93	0	3	2
Great Ormond Street, London[94]	2008	71	0	1	0
Emory University [146]	2009	107	3		
Children's Hospital Boston [88]	2009	77	0	2.8	6
Children's Hospital, Atlanta [147]	2010	42	0	2.2	0
		1035	0.68%		2.2%

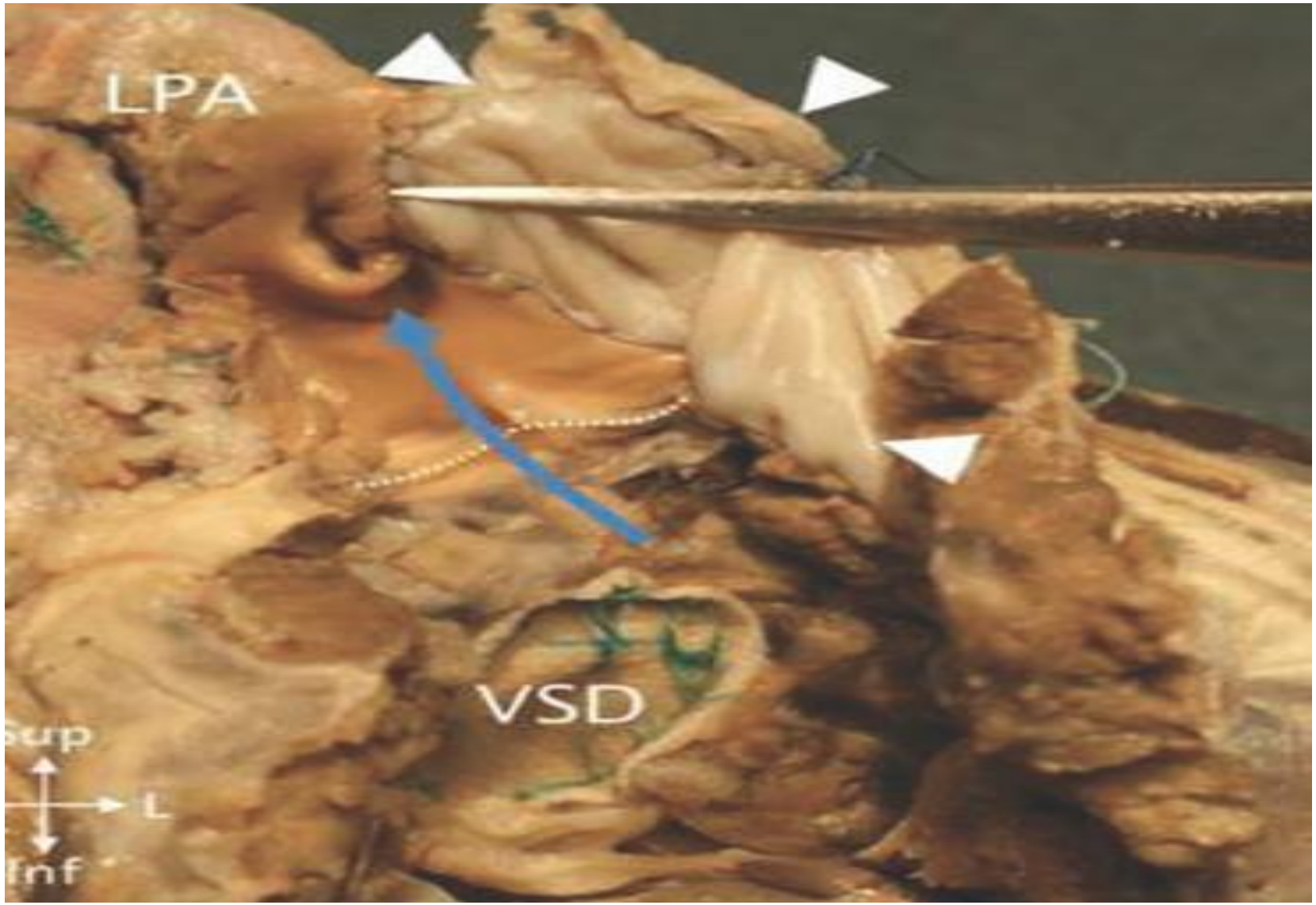
Geva T, Journal of Cardiovascular Magnetic Resonance 2011.

Options for PV replacement

PVR Technique



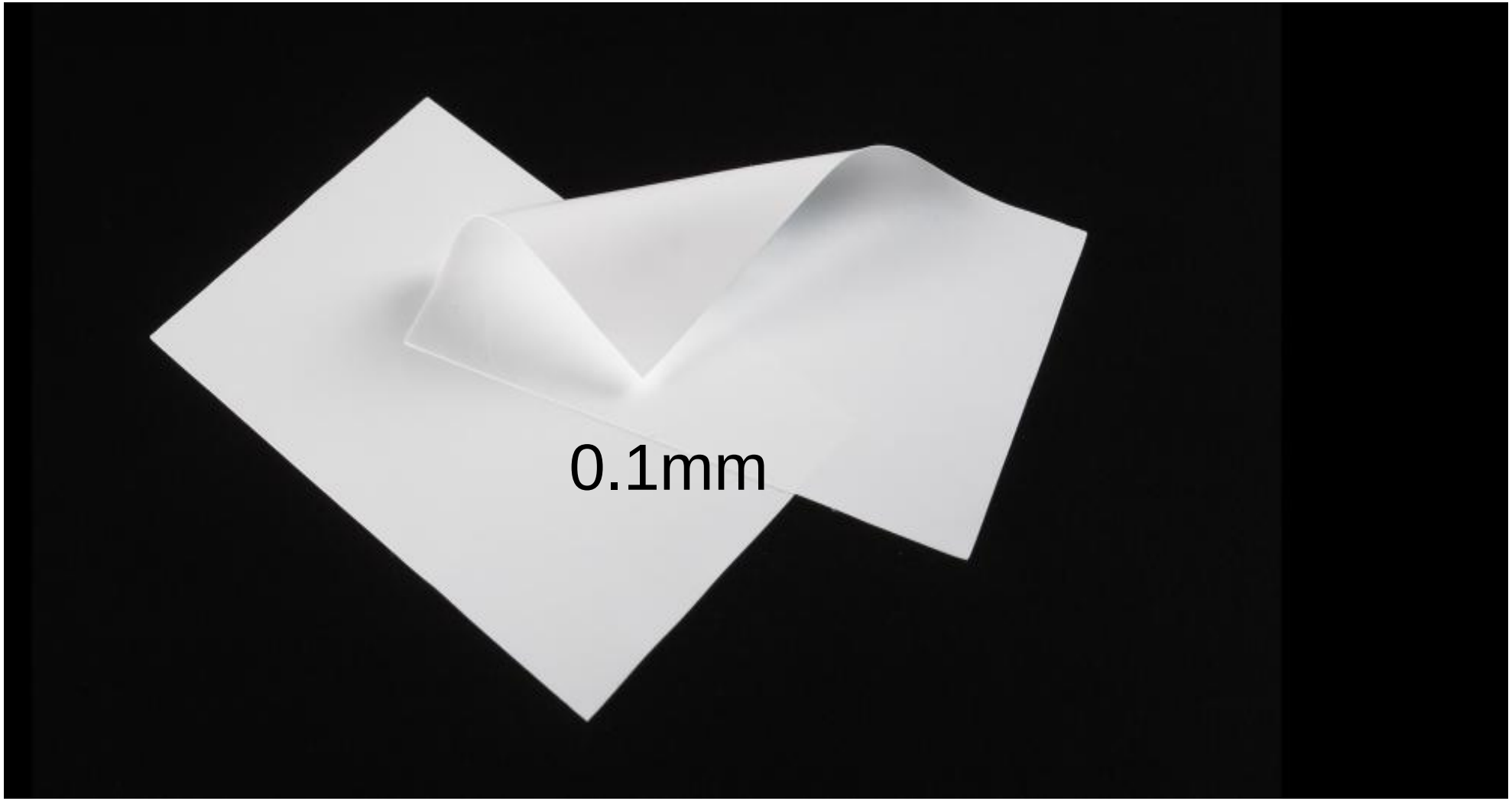
CRYOABLATION ADDED IF RVOT INDUCIBLE VENTRICULAR TACHYCARDIA



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)

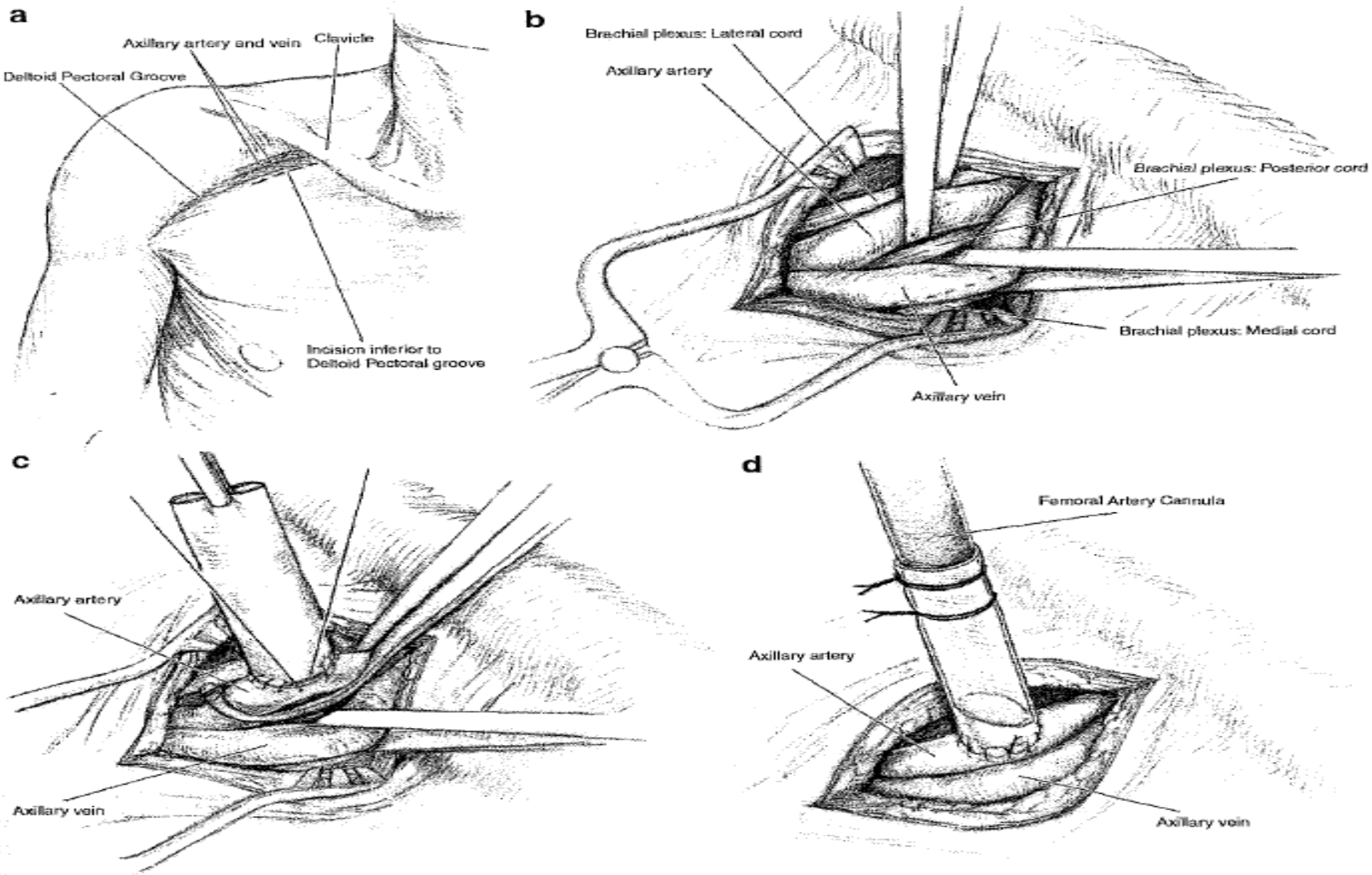
- My advice: don't use it routinely
- Why? Leaves a waxy scar over the entire mediastinum under the membrane which makes it harder to identify individual structures
- Indications:

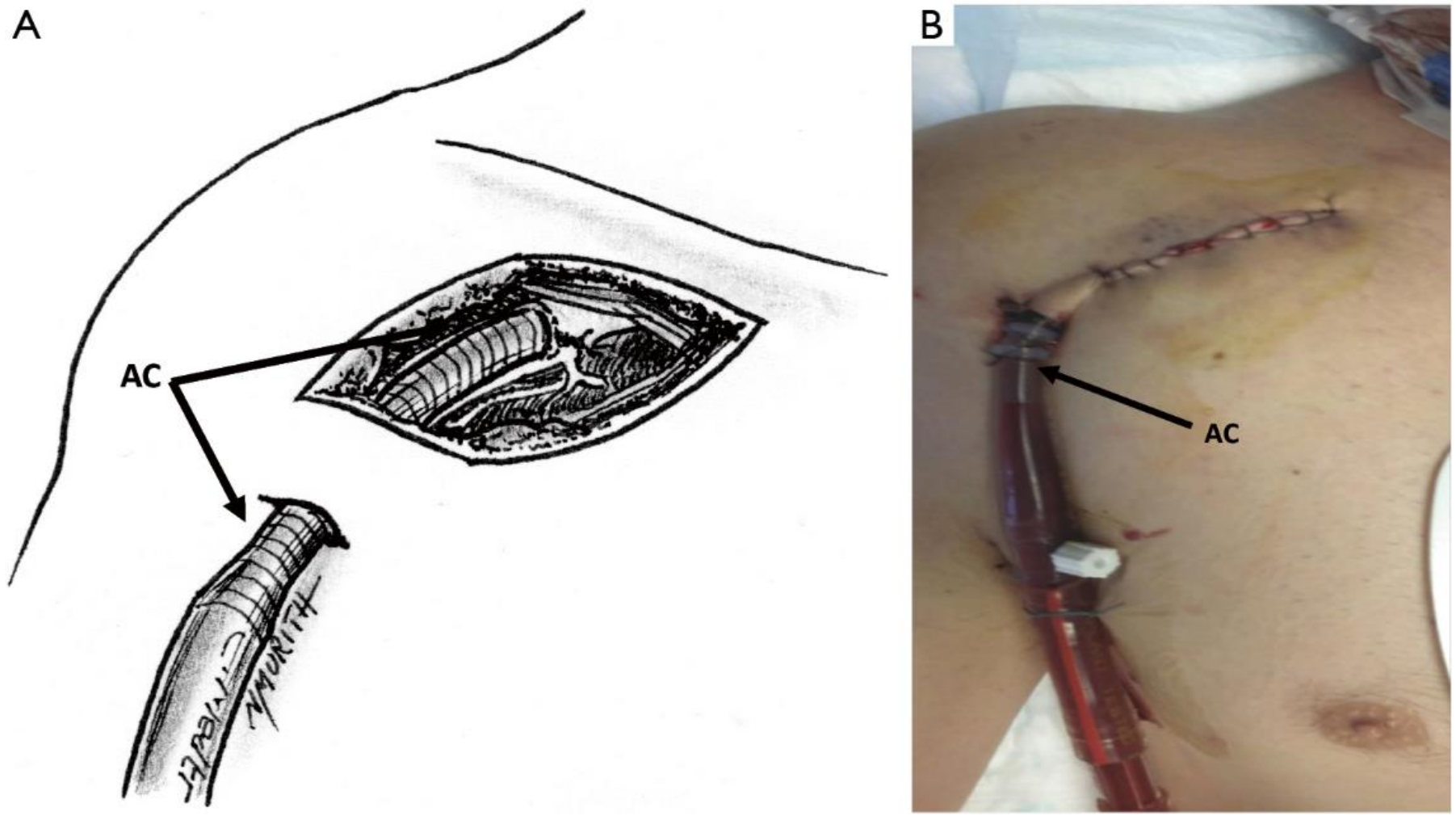
  RV to PA conduits- place only over the conduit

  Cover large, anterior aortas, Ross procedures, LVAD's

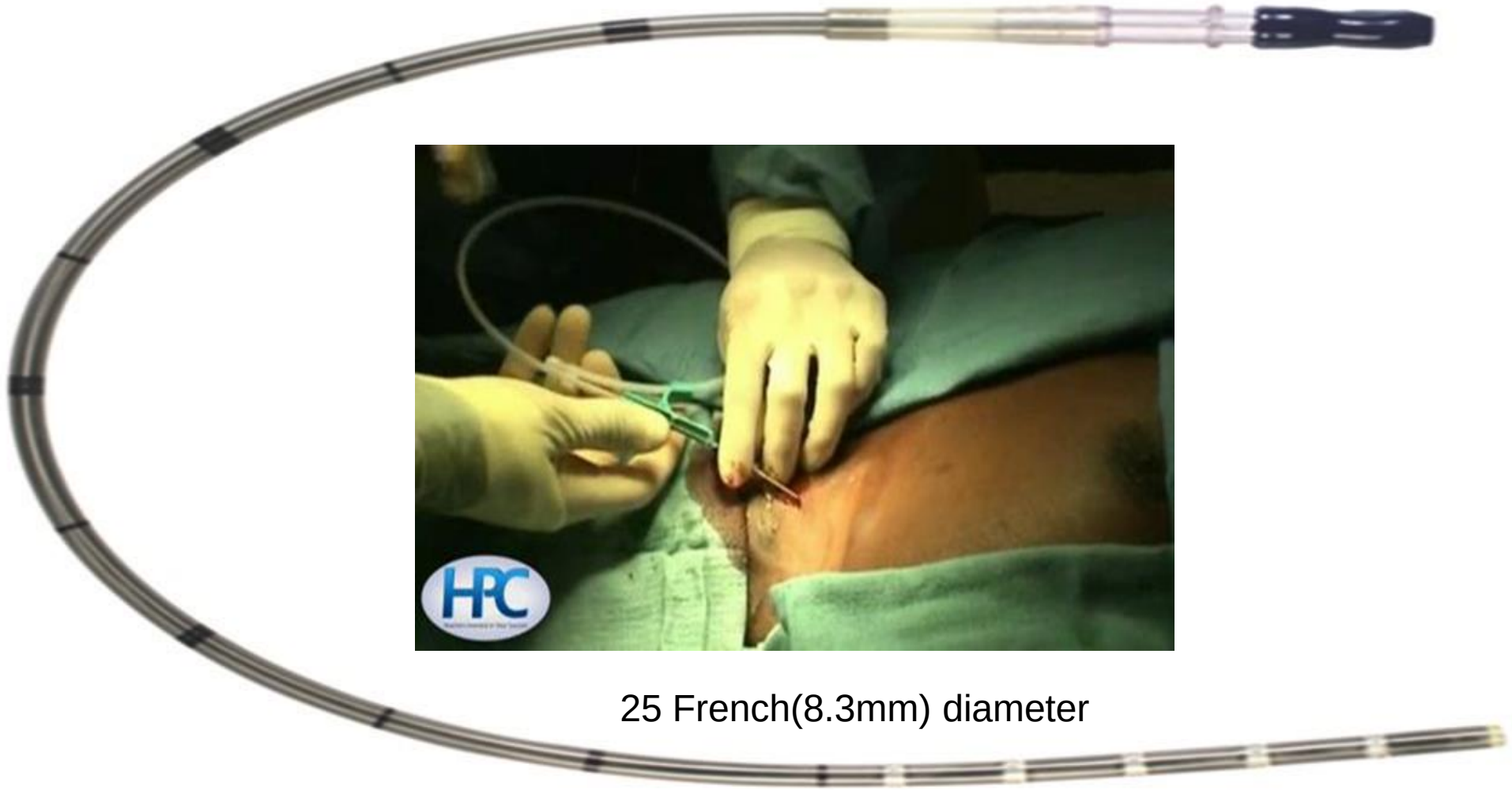
High Risk Re-sternotomy Technique

- 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.


a
Axillary artery and vein
Clavicle
Deltoid Pectoral Groove
Incision inferior to
Deltoid Pectoral groove
b
Brachial plexus: Lateral cord
Axillary artery
Brachial plexus: Posterior cord
Brachial plexus: Medial cord
Axillary vein
c
Axillary artery
Axillary vein
d
Femoral Artery Cannula
Axillary artery
Axillary vein



Direct Axillary Artery Cannulation



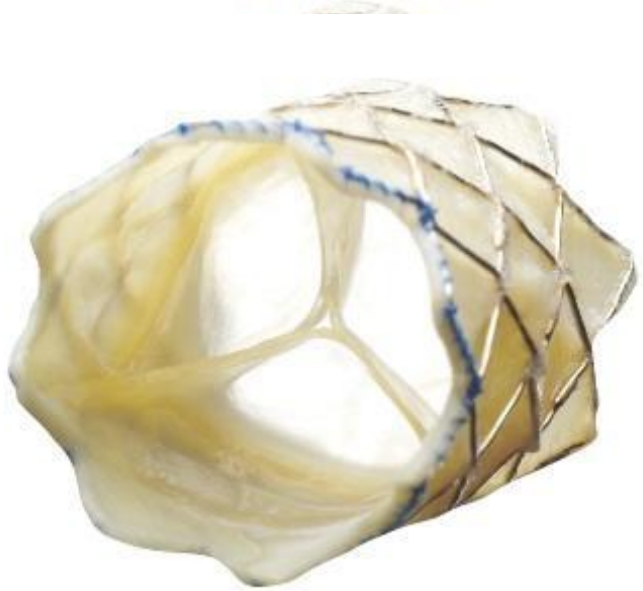
25 French(8.3mm) diameter

EDWARDS QUICKDRAW VENOUS CANNULA

The Congenital Interventional Cardiologist

Medtronic Melody Valve

Contegra bovine jugular vein valve sewn into small metal frame



2 sizes ID: up to 20mm and 22mm

Medtronic Melody Valve

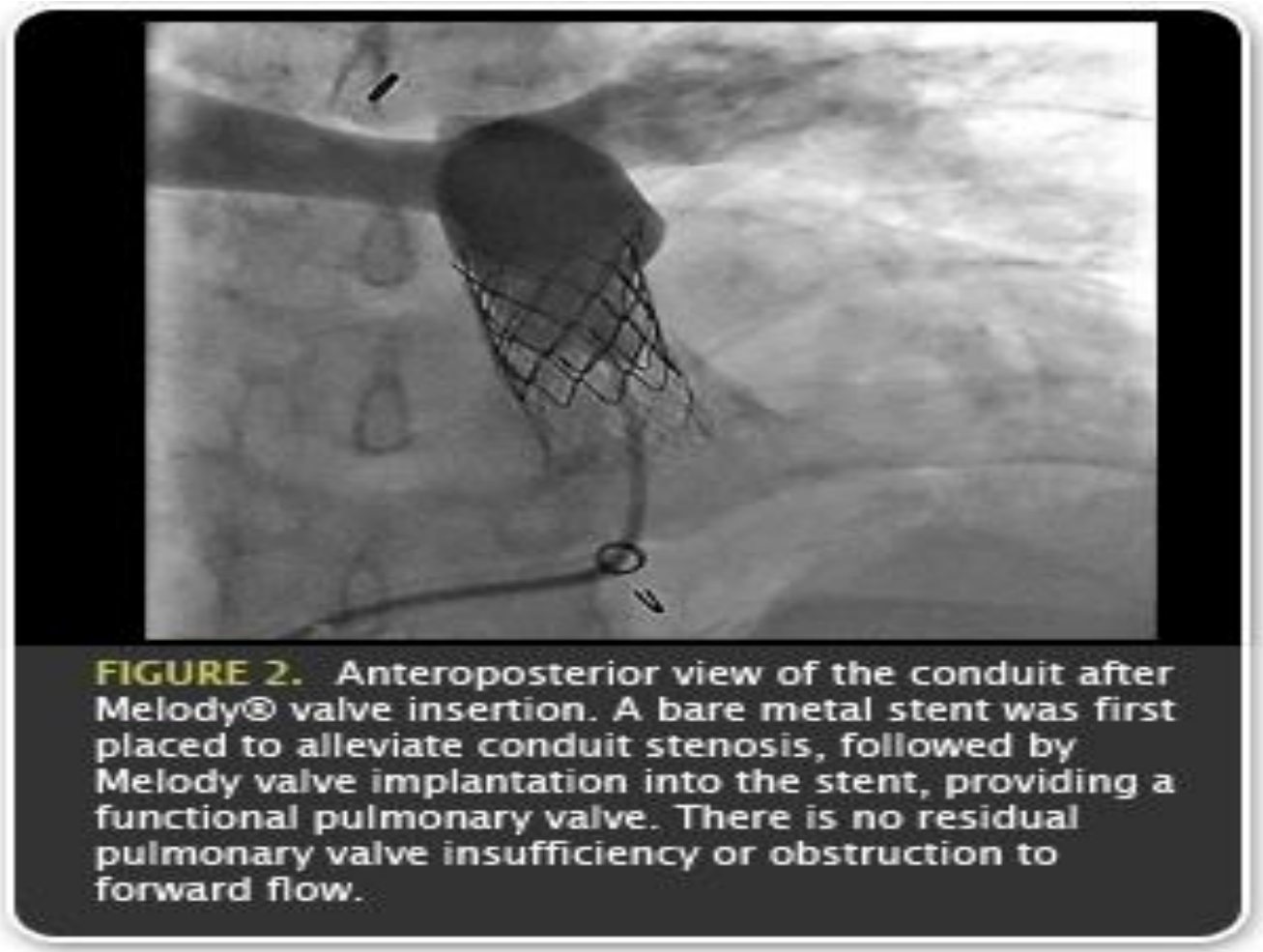


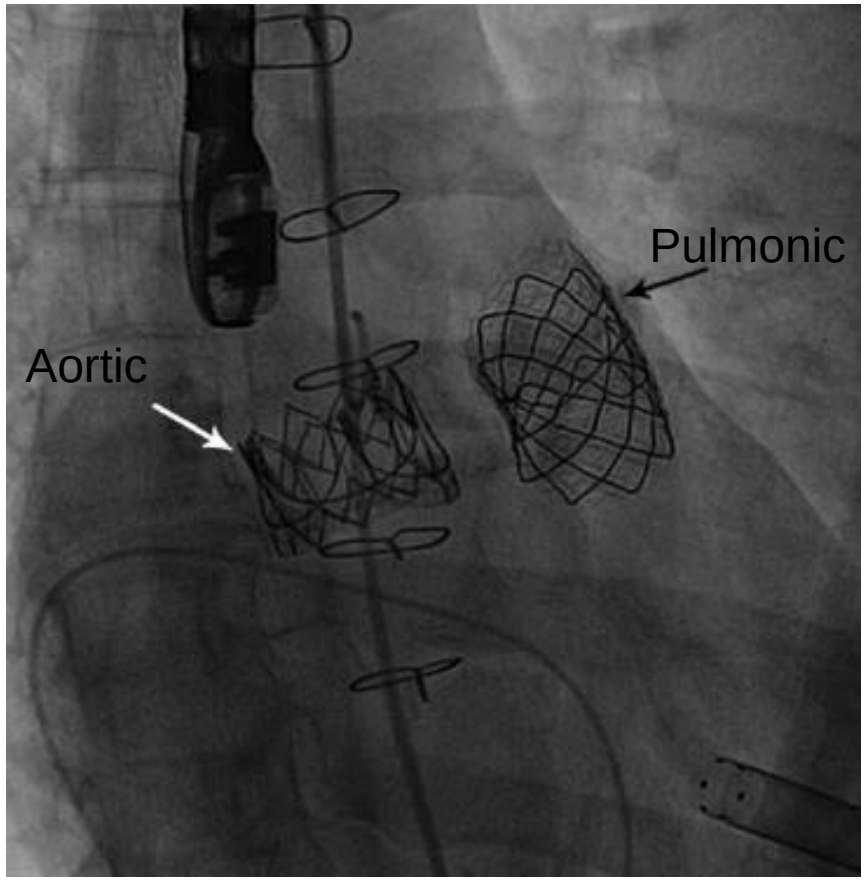
FIGURE 2. Anteroposterior view of the conduit after Melody® valve insertion. A bare metal stent was first placed to alleviate conduit stenosis, followed by Melody valve implantation into the stent, providing a functional pulmonary valve. There is no residual pulmonary valve insufficiency or obstruction to forward flow.

In the pulmonary position

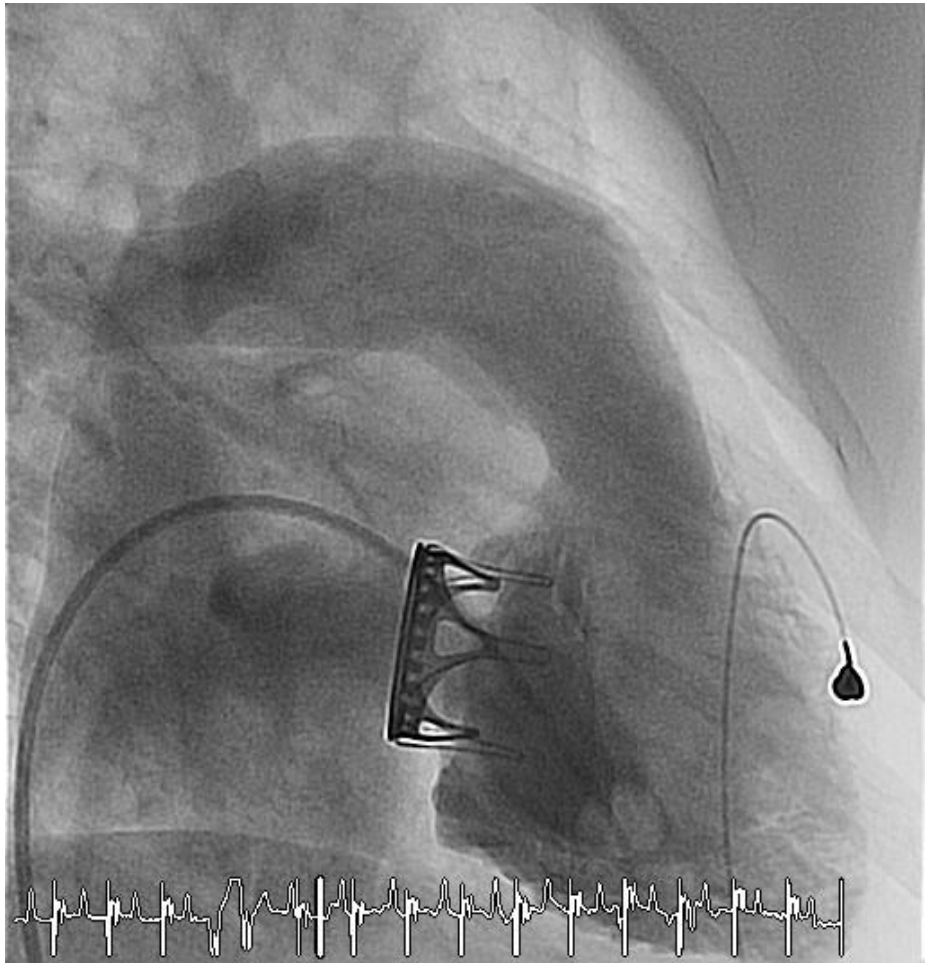
Catheterization and Cardiovascular Interventions 86:1092–1096 (2015)

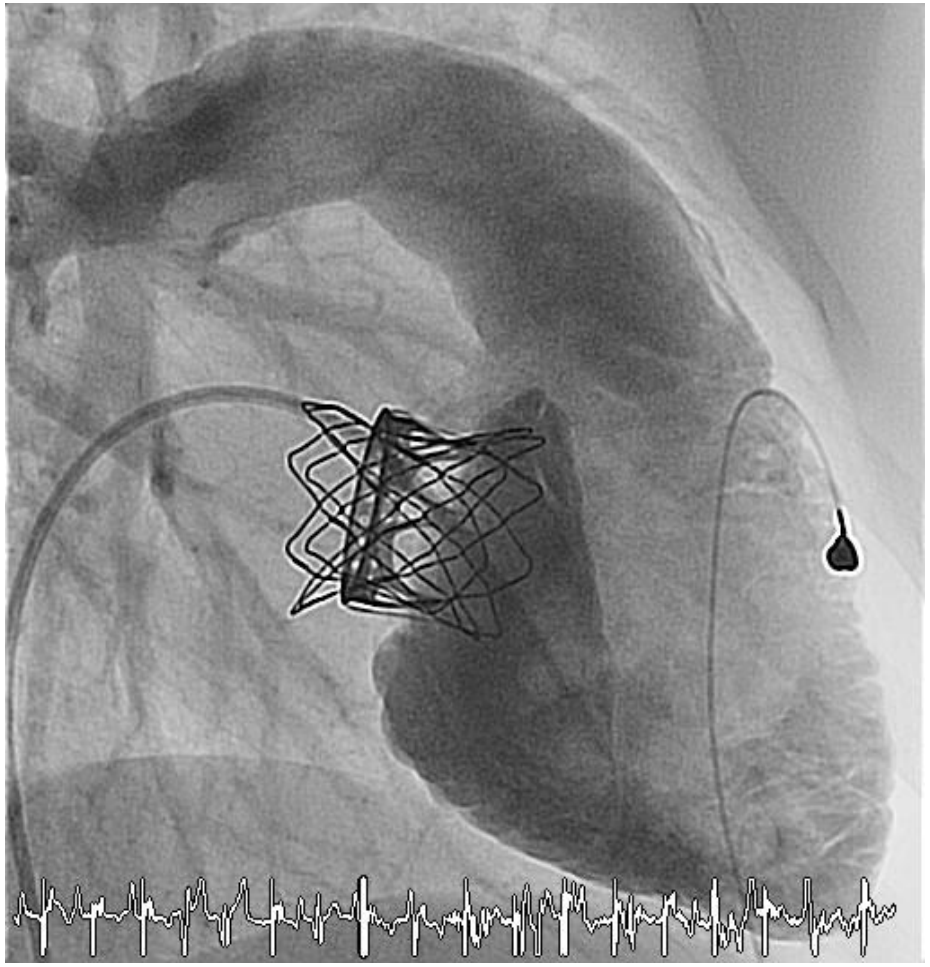
Consecutive Percutaneous Valve-in-Valve Replacement Late After Ross Procedure: A Novel Approach in an Adult With Congenital Heart Disease

Dominik M. Wiktor,[1,2] MD, Joseph D. Kay,[1,2] MD, and Michael S. Kim,[1,2*] MD

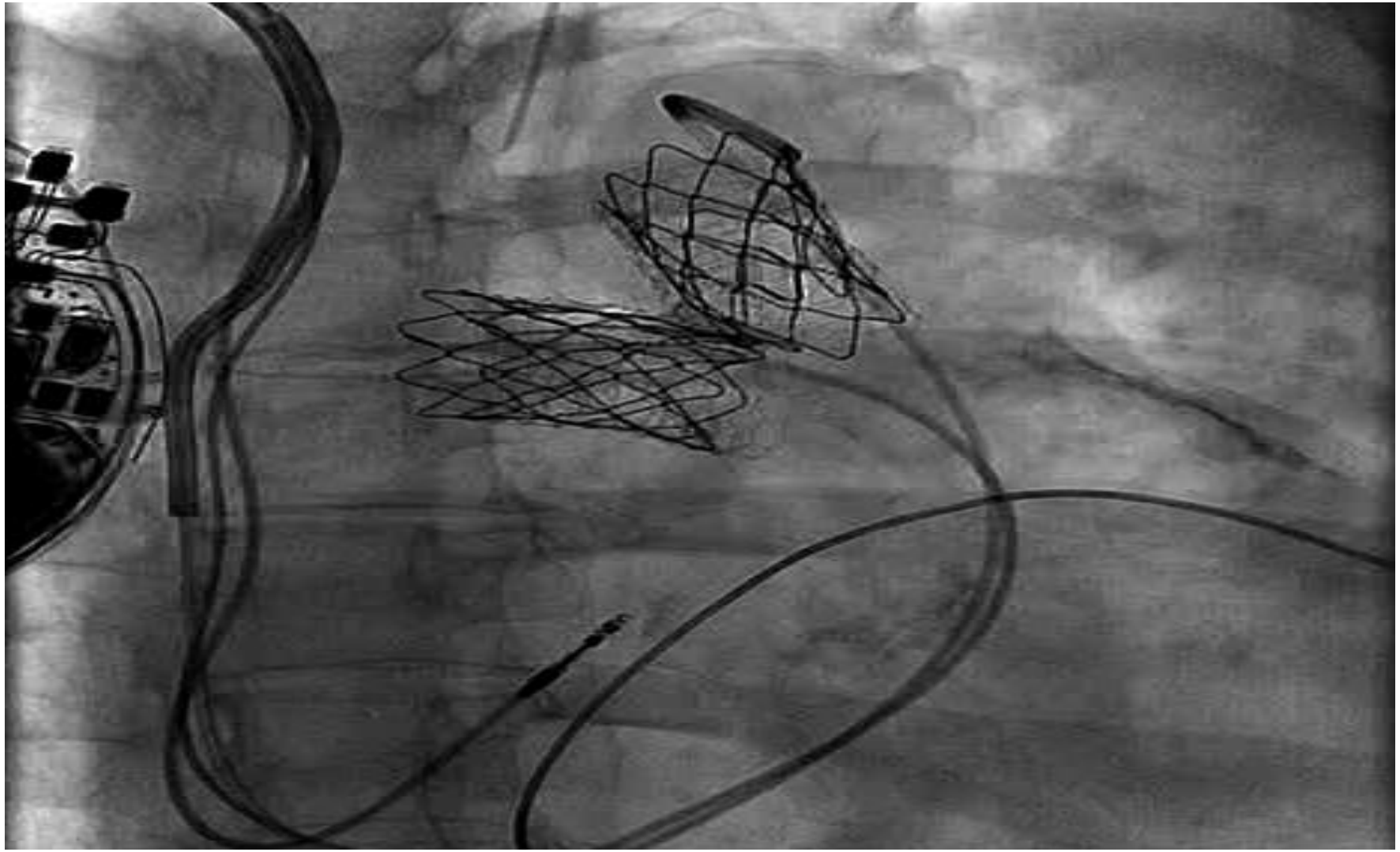


Melody Valve in failed tricuspid valve position

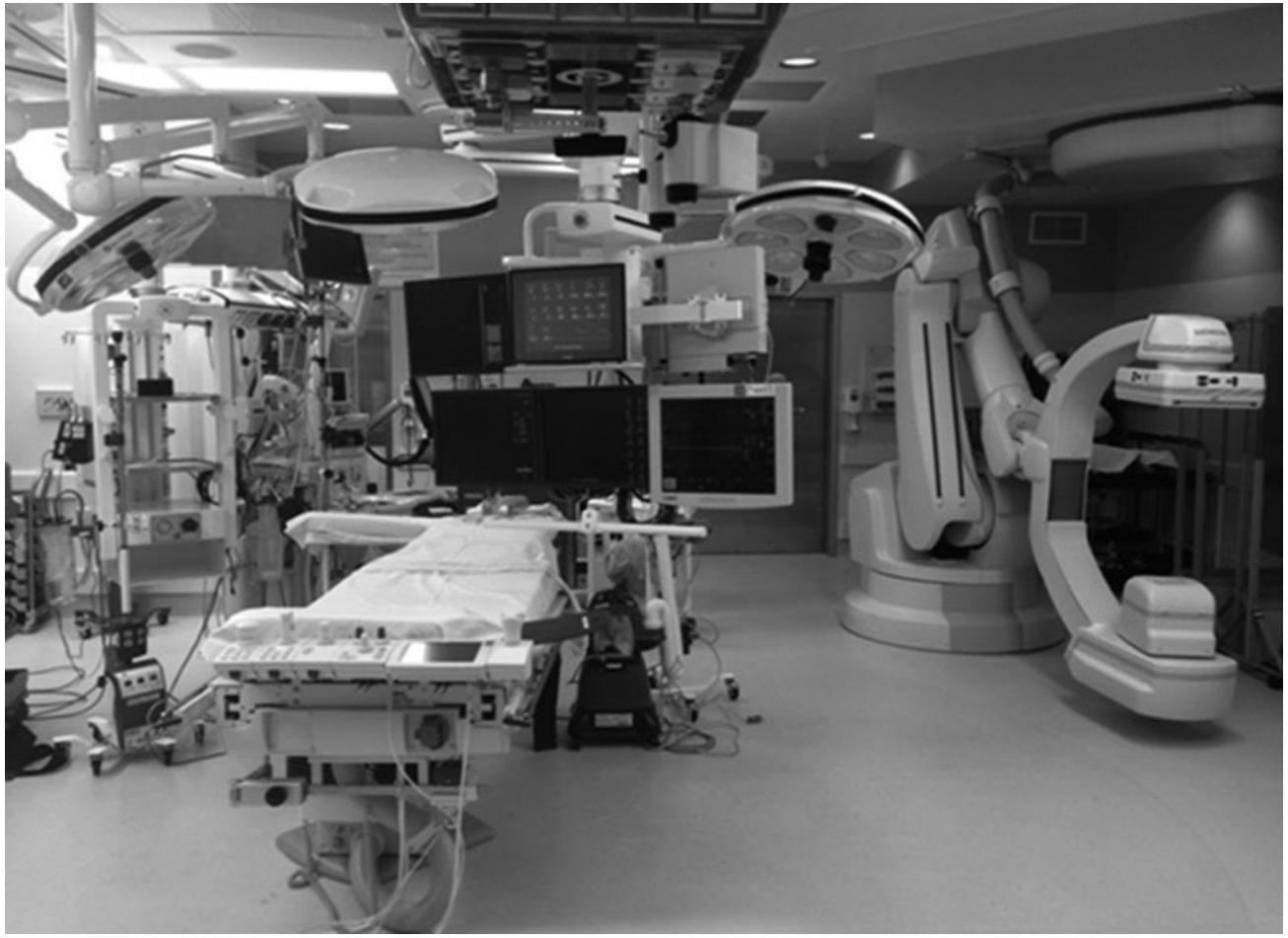




Bilateral Melody Valves in Pulmonary Position

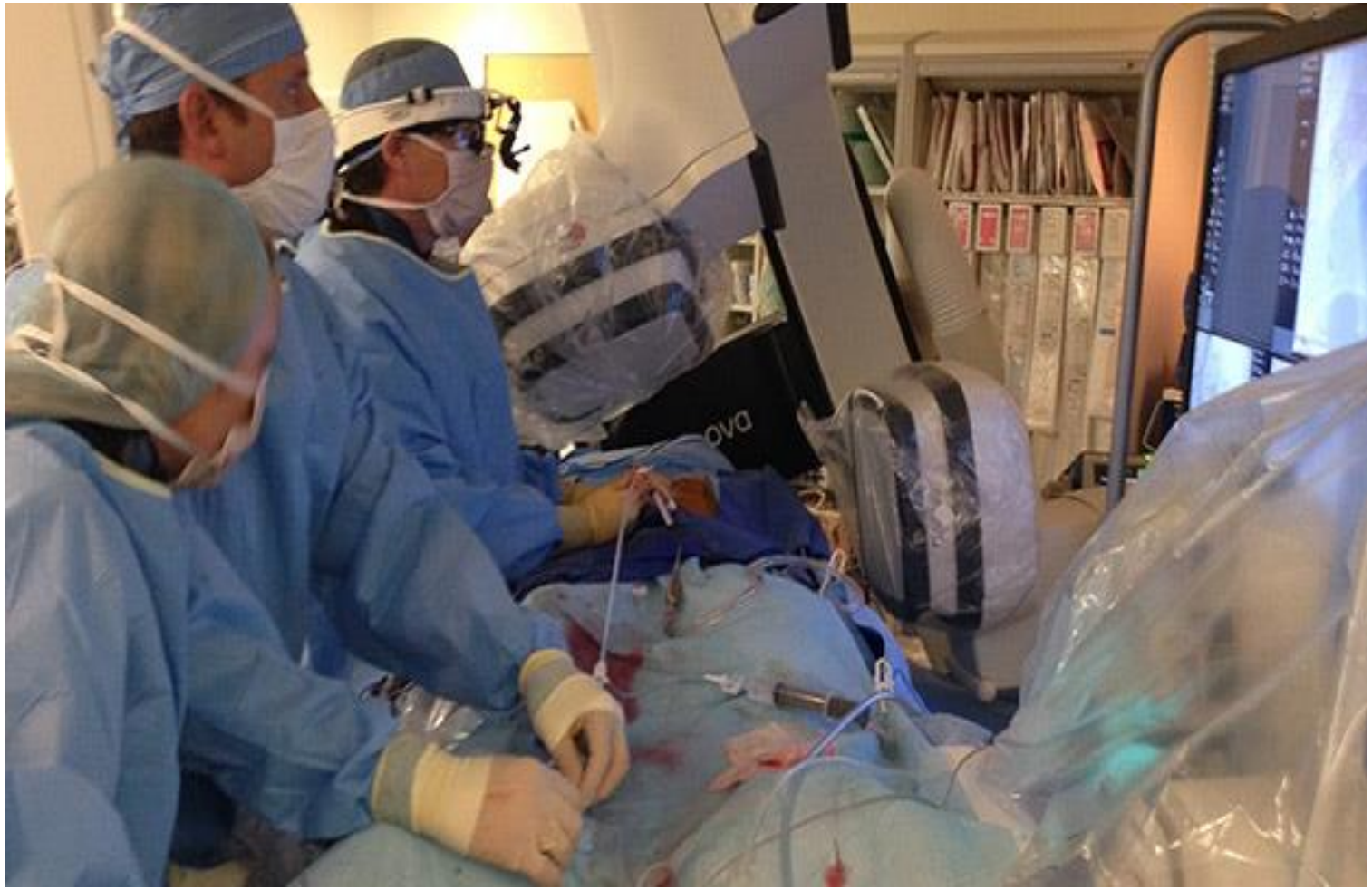


HYBRID LAB

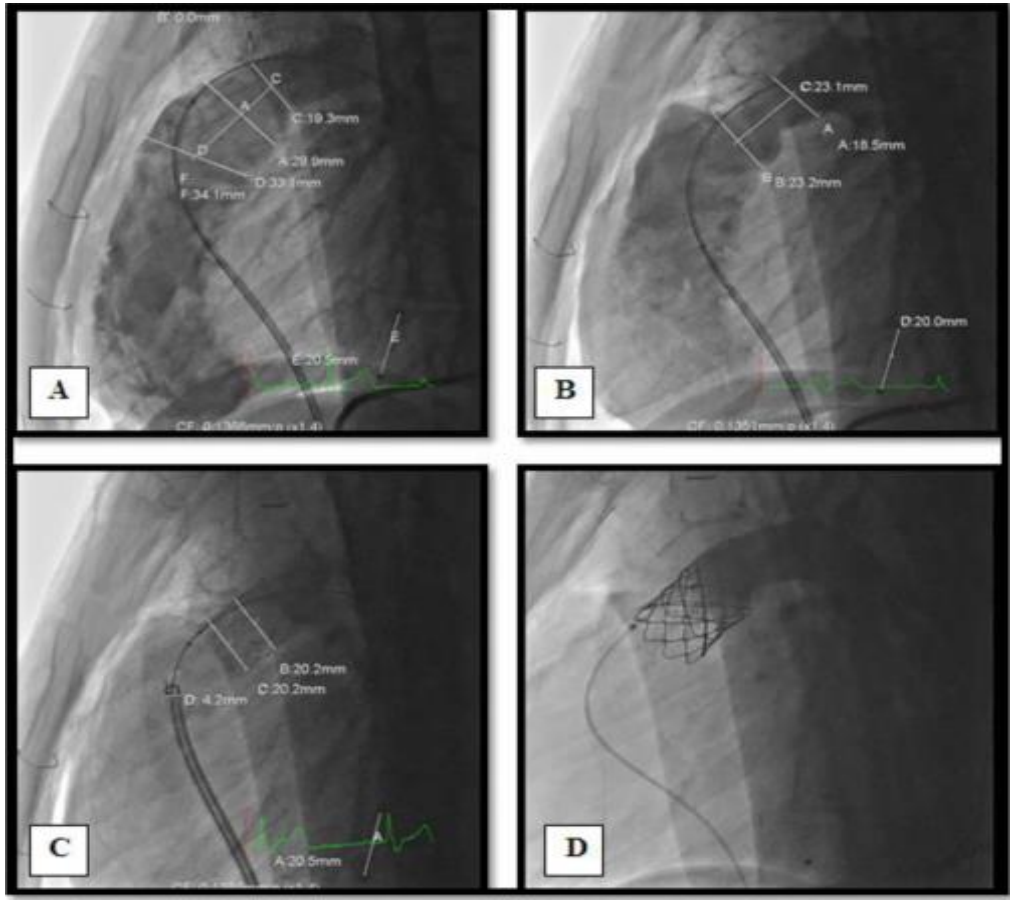


Interventional Cardiologist and Surgeon work in concert

LIMIT THE OPERATION



Hybrid Approach for pulmonary valve



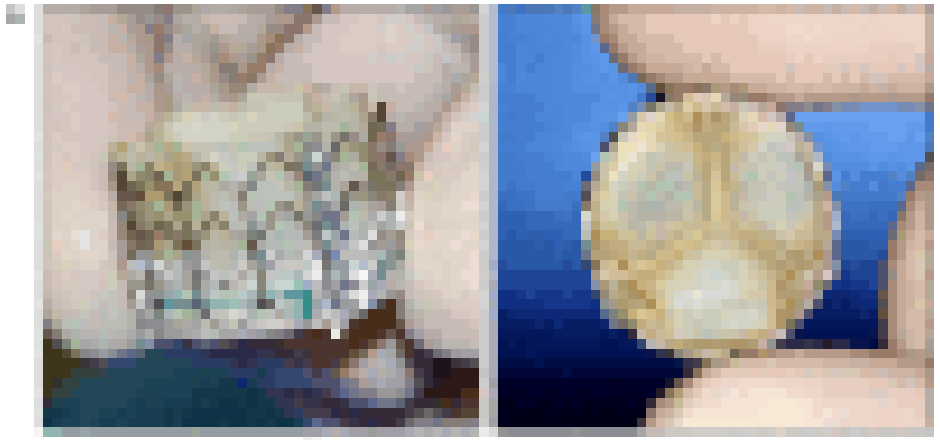
(A) Angiographic measurements of the MPA prior to plication. (B) Measurements of the MPA post-plication. Notice that the diameter of the MPA has been reduced from about 33 to 23 mm. (C) MPA with stent in place to create a landing zone for the Melody valve. Diameter now measures about 20 mm. (D) Angiography 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

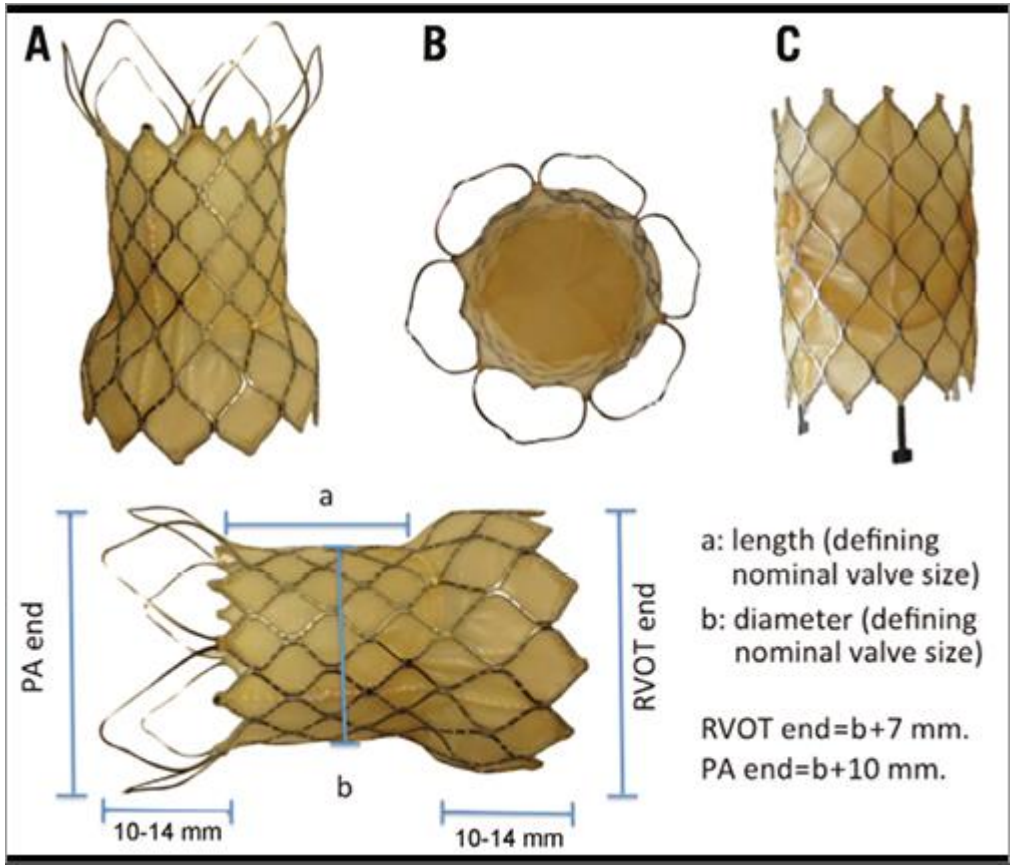
Tri-leaflet bovine pericardial valve

Available in larger sizes 26mm and 29mm



Dr. Gareth Morgan

UCH and CHC ACHD Interventionalist

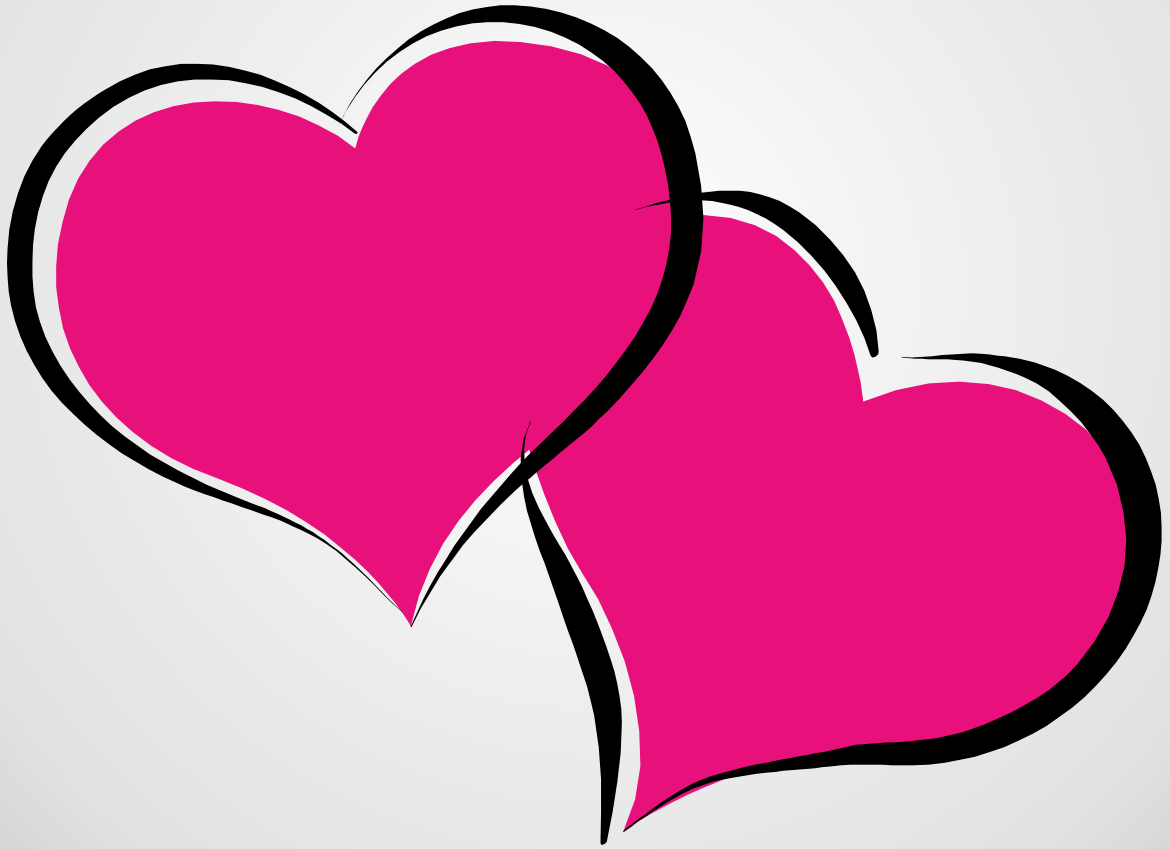


Venous P- valve sizes up to 35mm

"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

QUESTIONS ?