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Mechanical Support in the Failing Fontan-Kreutzer

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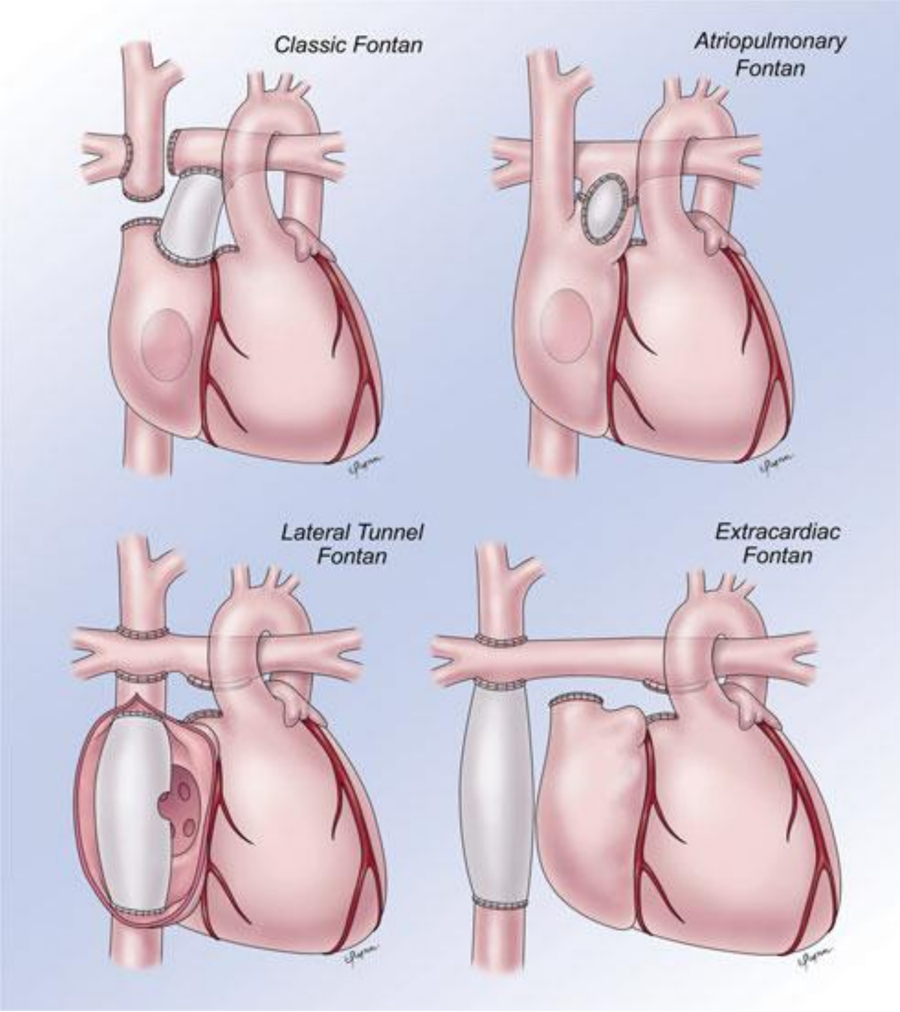
Consultant: W.L. Gore

Fontan-Kreutzer Failure

Early vs. Chronic

Mode of Failure:
Pump vs. Non Pump

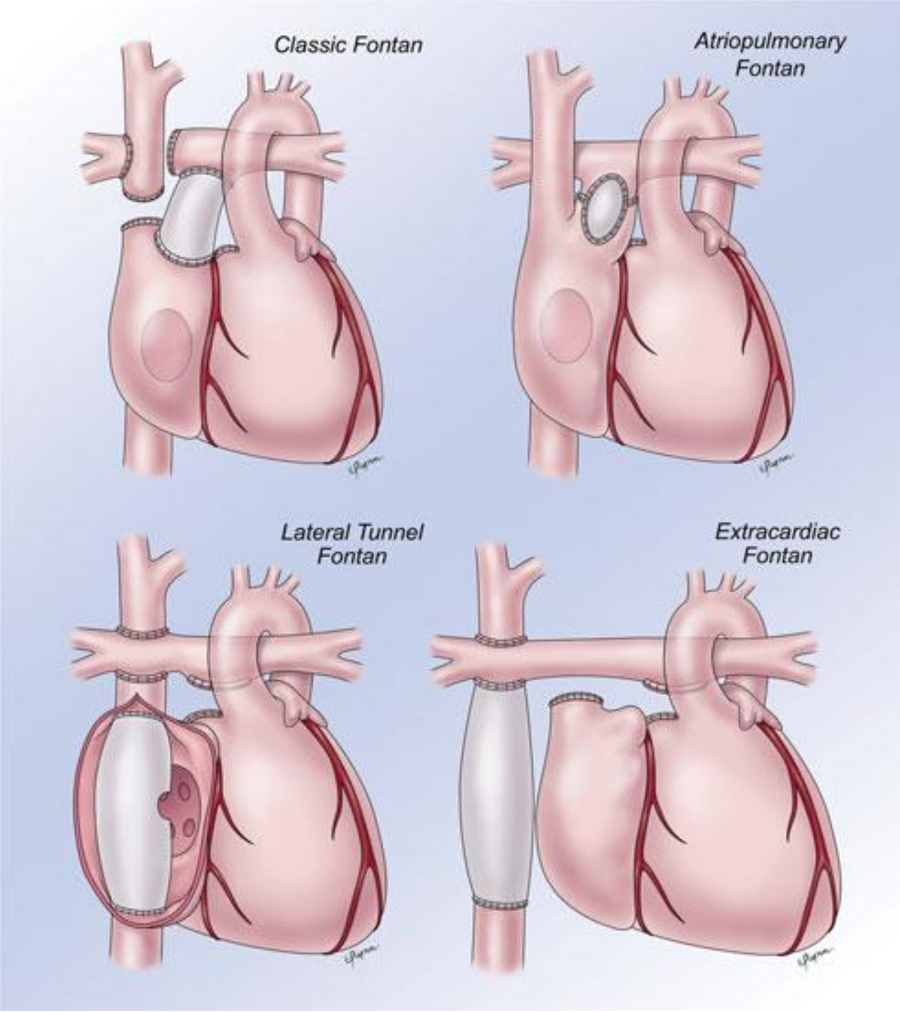


Valente AM, Landzberg MJ, Powell AJ: Adult congenital heart disease. Edited by Libby P. In Essential Atlas of Cardiovascular Disease. New York, New York: Springer; 2009:231–246

Fontan-Kreutzer Failure

Mode of Failure:

Pump



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Indications for Mechanical Support

- Failed maximal medical therapy with no surgical options
- Inability to wean from cardiopulmonary bypass
- Cardiogenic Shock
- Decreased organ perfusion
- Intractable Arrhythmias

Bridge to Transplant or Destination Therapy

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VAD: Surgical Challenges

Relative **Contraindications**

- Patient is NOT a Transplant Candidate
  - Exception – Pulmonary Hypertension: Appropriate for some patients
- Anatomical Diagnosis
- Neurologic Concern
  - Intracranial Hemorrhage, Severe Neurologic Impairment
- Isolated Pulmonary Dysfunction
- Sepsis
- Multisystem Organ Failure

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Mechanical Assist Devices for the Fontan-Kreutzer

action ADVANCED CARDIAC THERAPIES IMPROVING OUTCOMES NETWORK

Cardiac Dysfunction:

1. Severe systolic dysfunction
    - Less than 35% by echocardiogram
    - Less than 30% by MRI
2. Moderately depressed dysfunction when accompanied by >moderate or severe systemic AV valve regurgitation
3. Significant growth derangement or failure to thrive
4. Decreasing exercise tolerance by patient report as measured on sequential exercise testing
5. Significant electrophysiologic abnormalities including arrhythmia or aborted sudden **death**

Fontan Pathway Dysfunction:

1. Symptomatic chronic fluid overload despite increasing diuretic therapy
2. Occurrence of chronic ascites or pleural effusion refractory to therapy
3. Major hemodynamic disturbance resulting in symptoms including diastolic failure or symptomatic cyanosis

Lymphatic Dysfunction: PLE or plastic bronchitis requiring multiple admissions in 12 months

Extracardiac Dysfunction: Hemoptysis, liver disease or chronic kidney disease

Heart Transplantation in Children after a Fontan Procedure: Better than People Think

Kirk R. Kanter

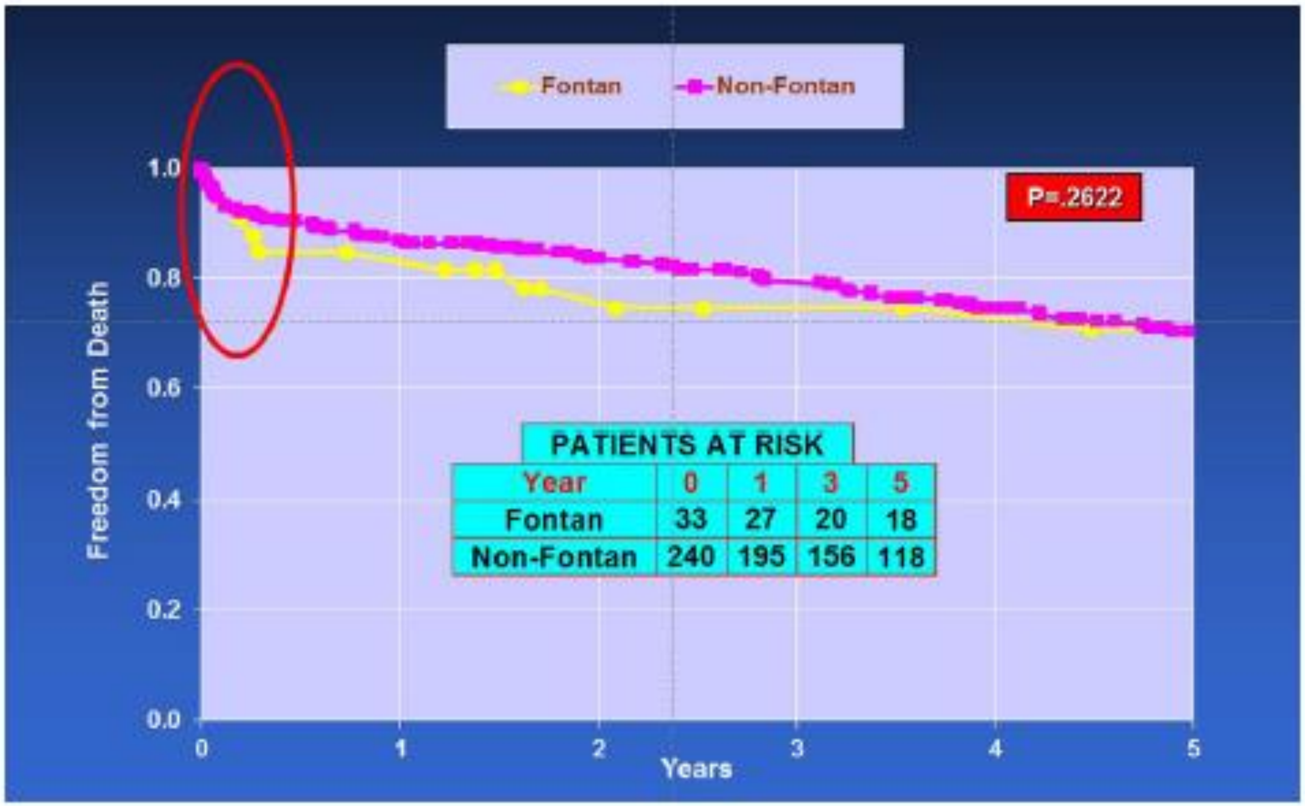

Figure 1 Kaplan-Meier actuarial freedom from death stratified by patients who had a previous Fontan procedure (closed boxes) and those who did not (open circles). The survival estimates are not statistically different ($P = .2622$). The oval emphasizes the increased attrition in the first 6 months after transplantation in the Fontan patients.

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 19:44-49

Is Four Stage Management the Future of Univentricular Hearts? Destination Therapy in the Young

Robert D.B. Jaquiss, and Hamza Aziz

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 19:50-54

Table 4 Comparison of Transplant Versus Fontan with Durable RVAD

	Transplant	Fontan + RVAD
Resting cardiac output	Normal	Normal
Exercise cardiac output	Normal	Normal
Hepatic venous pressure	Normal/slightly elevated	Normal
Chronic anticoagulation	No	Yes
Power cord	No	Probably
Immunosuppression risks	Yes	No
Diabetes, hypertension, renal failure	Yes	No
Additional surgery needed?	Yes (re-do transplant)	Yes (re-do VAD or transplant)

Abbreviations: RVAD, right ventricular assist device; VAD, ventricular assist device.

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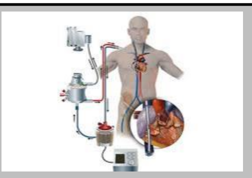
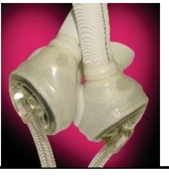
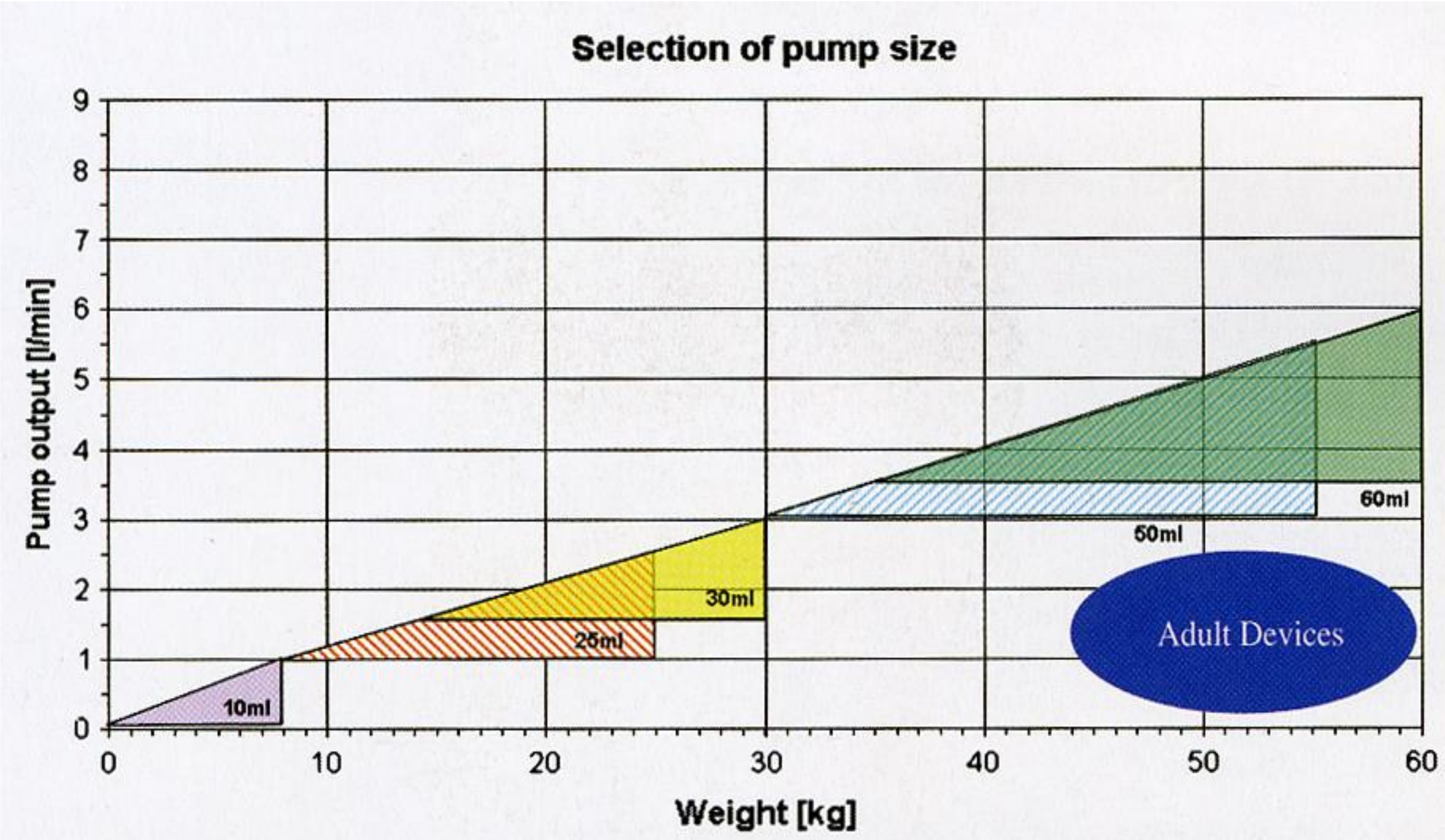
Mechanical Assist Devices Choices at CHOP

Device		Position	Pulsatile	Discharge	Destination
ECMO		Paracorporeal	N	N	N
Pedimag		Paracorporeal	N	N	N
Berlin		Paracorporeal	Y	N	N
Heartware		Intracorporeal	N	Y	Y
HM3 Thoratec		Intracorporeal	N	Y	Y
Syncardia		Intracorporeal	Y	Y	Y

Mechanical Assist Devices Choices at CHOP

Device		Position	Pulsatile	Discharge	Destination
ECMO		Paracorporeal	N	N	N
Pedimag		Paracorporeal	N	N	N
Berlin		Paracorporeal	Y	N	N
Heartware		Intracorporeal	N	Y	Y
HM3 Thoratec		Intracorporeal	N	Y	Y
Syncardia		Intracorporeal	Y	Y	Y

Options for Mechanical Support at CHOP
Patient Size



Options for Mechanical Support at CHOP
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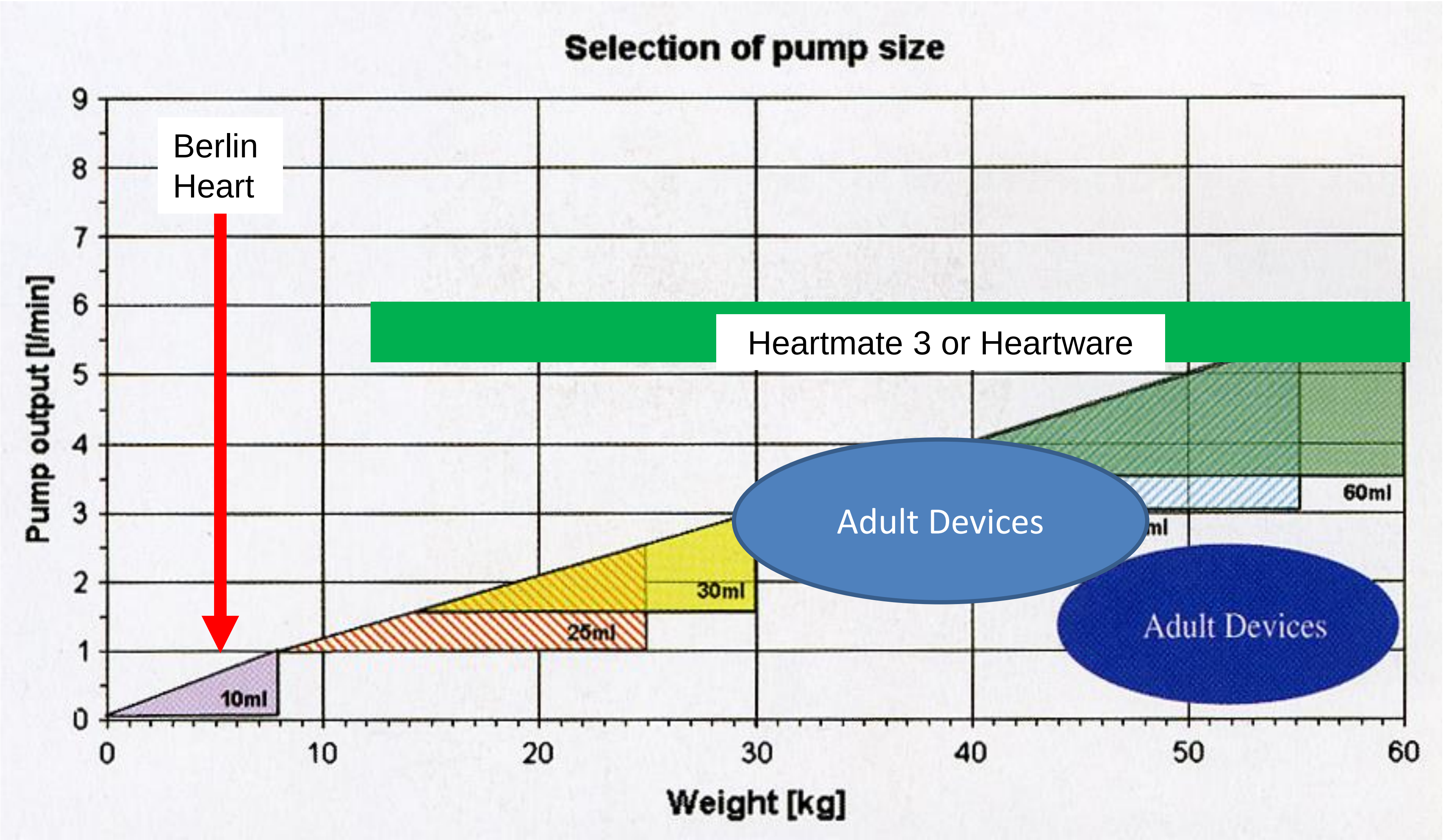

VAD: Surgical Challenges

Basic Principles of Mechanical Assistance

Acute

- Simple, rapid employment
- Decompression of myocardium
- Maintenance of end-organ perfusion
  - Elimination of edema
  - Improvement in nutrition
- Long-term reliability
- Portability
  - Ability to rehabilitate

Chronic

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Surgical Challenges – Implantable VAD

- Consideration of chest or abdominal DOMAIN
    - Chest closure challenging. Need for a bioprosthetic membrane to approximate chest or abdomen (fascial closure)
- Consider AV valve resection
- Pre-peritoneal vs Intra-abdominal placement
- Make drive line tunnels LONG
- TEE use after insertion is a must!
    - No residual PFO, Obstruction of inflow cannula by septum
    - Obstruction of Right Atrial Conduit
    - Ventricular position

Table 1
Ventricular assist devices in Fontan patients.

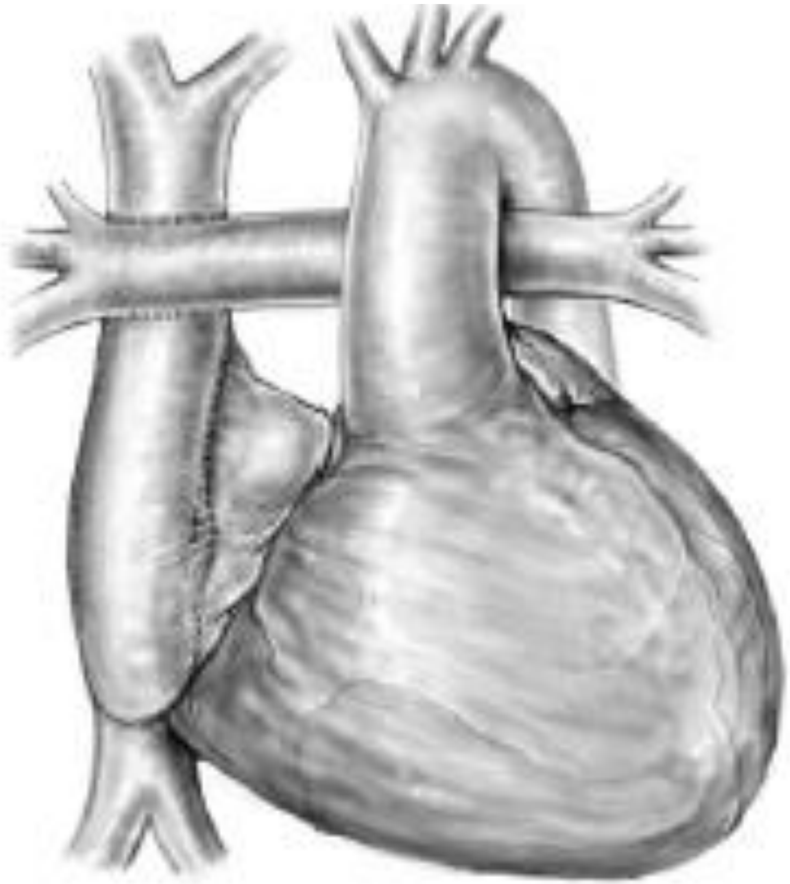
Progress in Pediatric Cardiology 34 (2012) 27–30

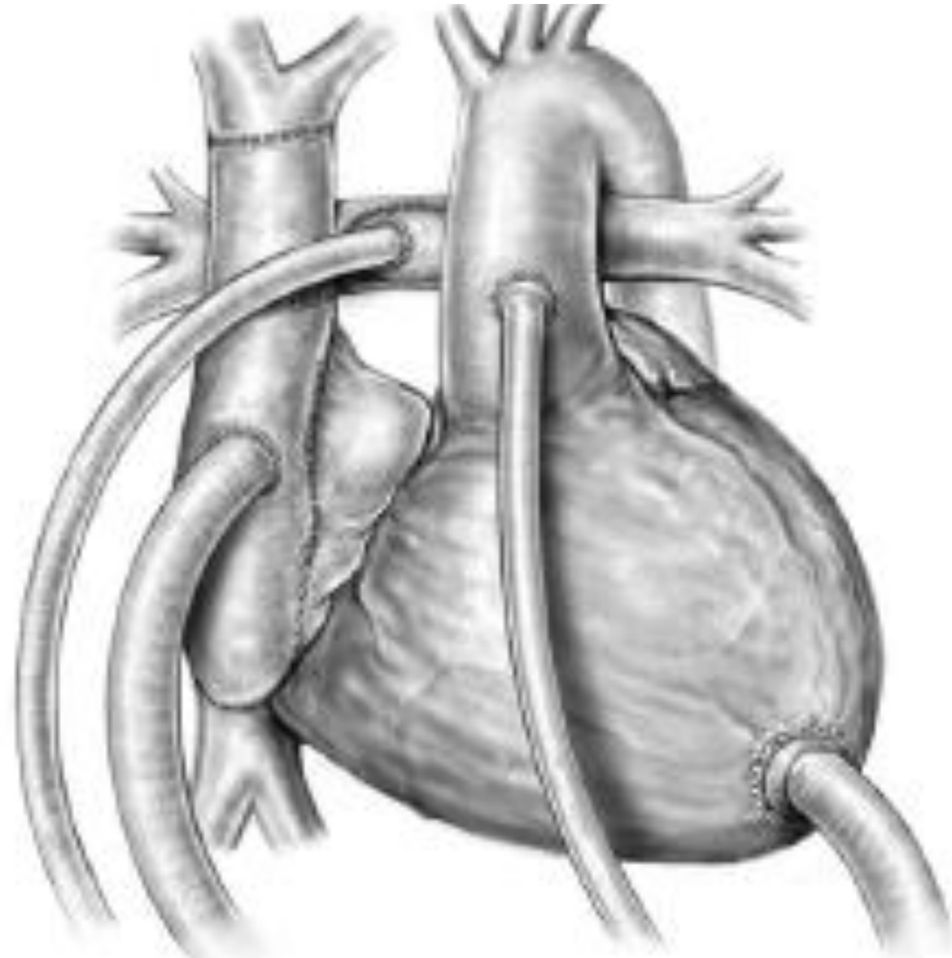
Author	Diagnosis	Age at VAD	Type of VAD	Outcomes
Fraser et al. 2011	HLHS	15 years	Heartmate II	VAD to TXP at 71 days
Carderelli et al. 2009	HLHS	18 months	Berlin Heart	Cardiac recovery, VAD explanation at 6 mos
Russo et al. 2008	Tricuspid atresia	14 years	Centrifugal VAD, Heartmate	VAD to TXP
Calvaruso et al. 2007	Mitral atresia	10 years	Berlin Heart	VAD to TXP at 7 days
Newcomb et al. 2007	DILV, TGA, subpulmonary stenosis	25 years	Thoratec	VAD to TXP at 5 mos
Nathan et al. 2006	HLHS	4 years	Berlin Heart	VAD to TXP at 28 days, died 8 days post-txp
Frazier et al. 2005	Tricuspid atresia	14 years	Heartmate IP LVAS	VAD to TXP at 45 days

VAD: Surgical Challenges – Single Ventricle

Successful implantation of a Berlin heart biventricular assist device in a failing single ventricle

Meena Nathan, MD, Christopher Baird, MD, Francis Fynn-Thompson, MD, Christopher Almond, MD, MPH,
Ravi Thiagarajan, MD, MPH, Peter Laussen, MBBS, Elizabeth Blume, MD, and Frank Pigula, MD, Boston, Mass





VAD: Surgical Challenges – Single Ventricle

Creation of a Self-Made Total Artificial Heart Using Combined Components of Available Ventricular Assist Devices. Tjan TDT et al. *Thorac Cardiov Surg* 2008: 56; 51-9.

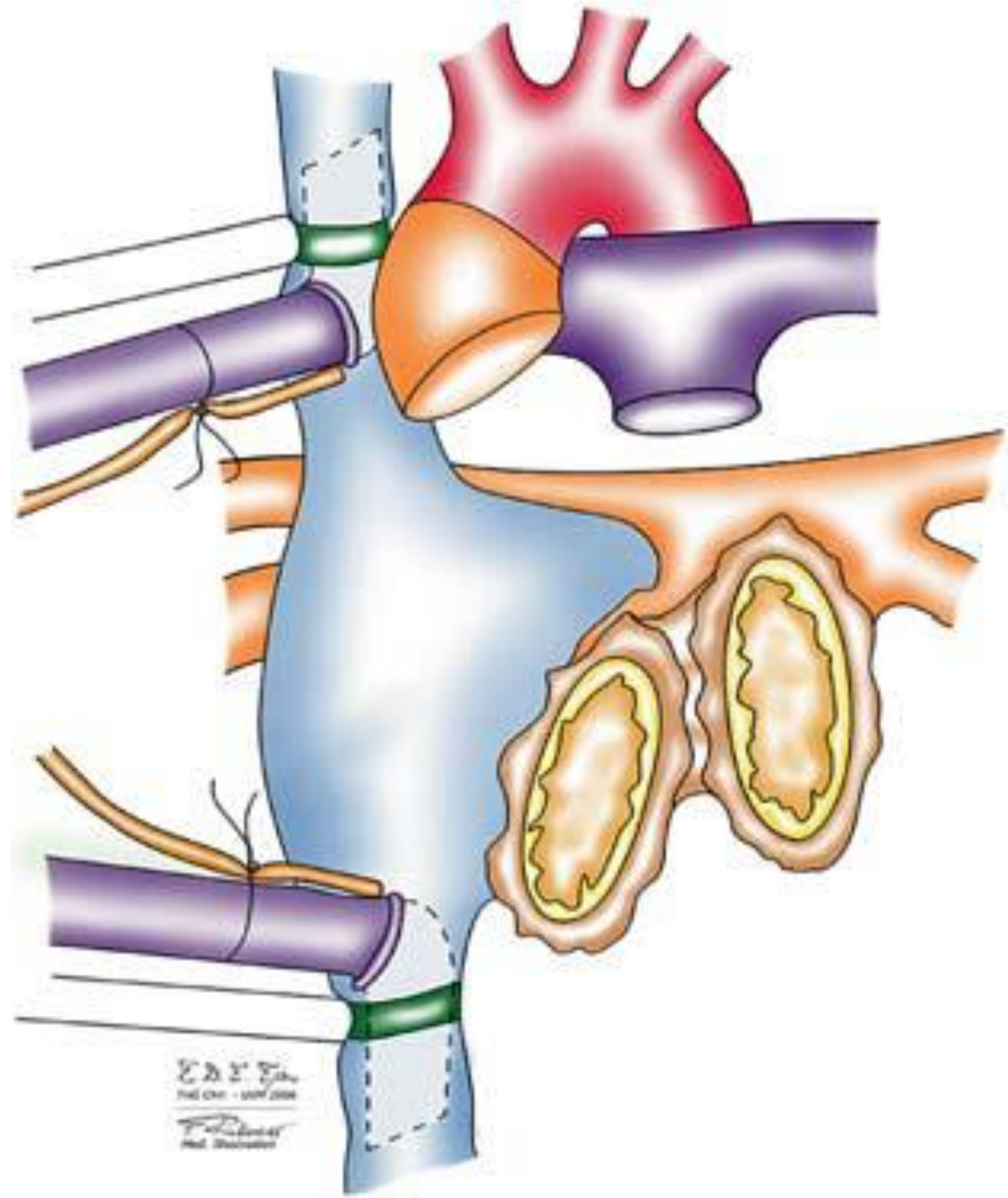


Figure 1. The left and right ventricles were resected leaving a 5mm residual just distal from the AV valve annulus.

VAD: Surgical Challenges – Single Ventricle

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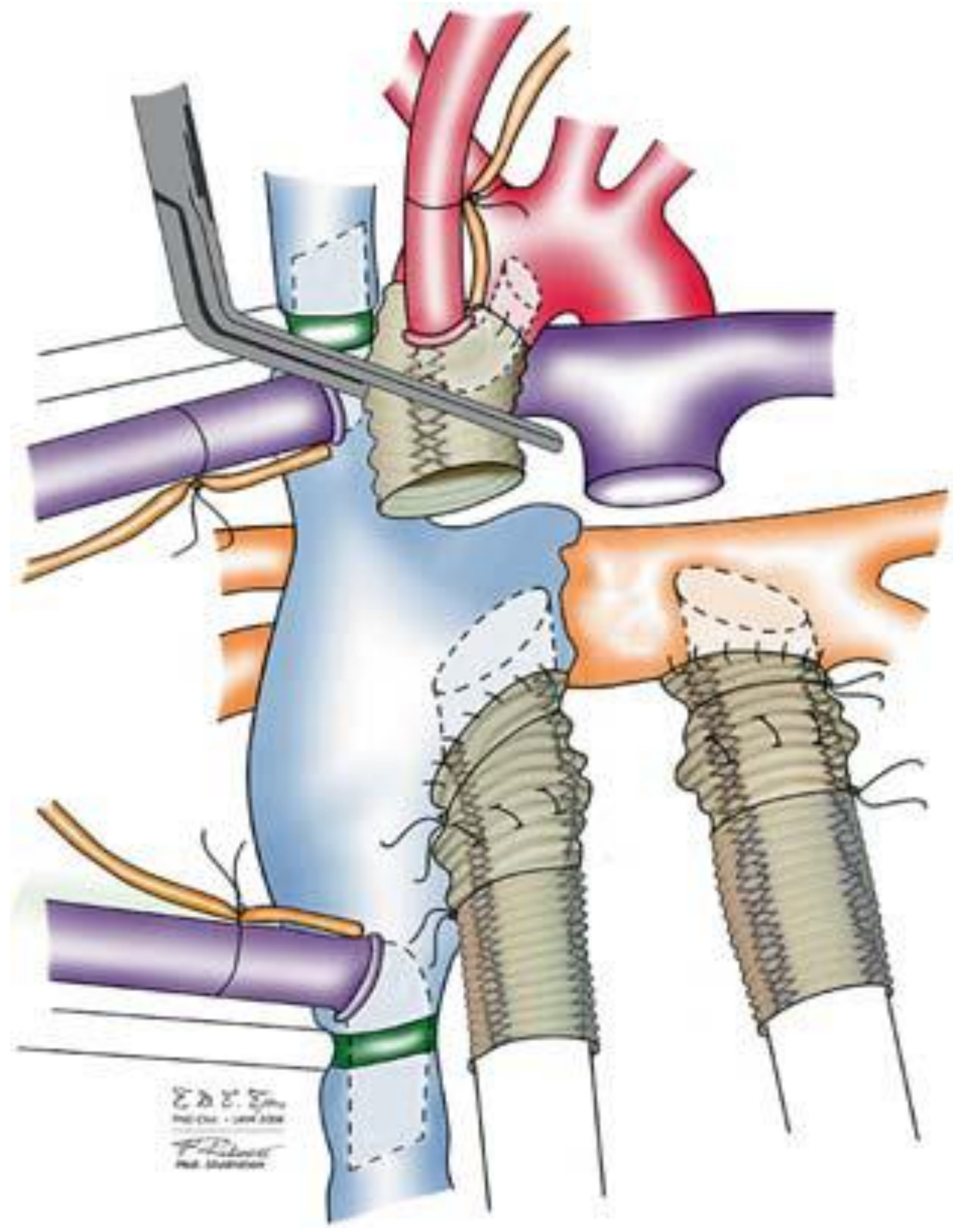

Figure 3. The inflow cannula from the Berlin-Heart EXCOR system introduced in each atria through the cuffs.

VAD: Surgical Challenges – Single Ventricle

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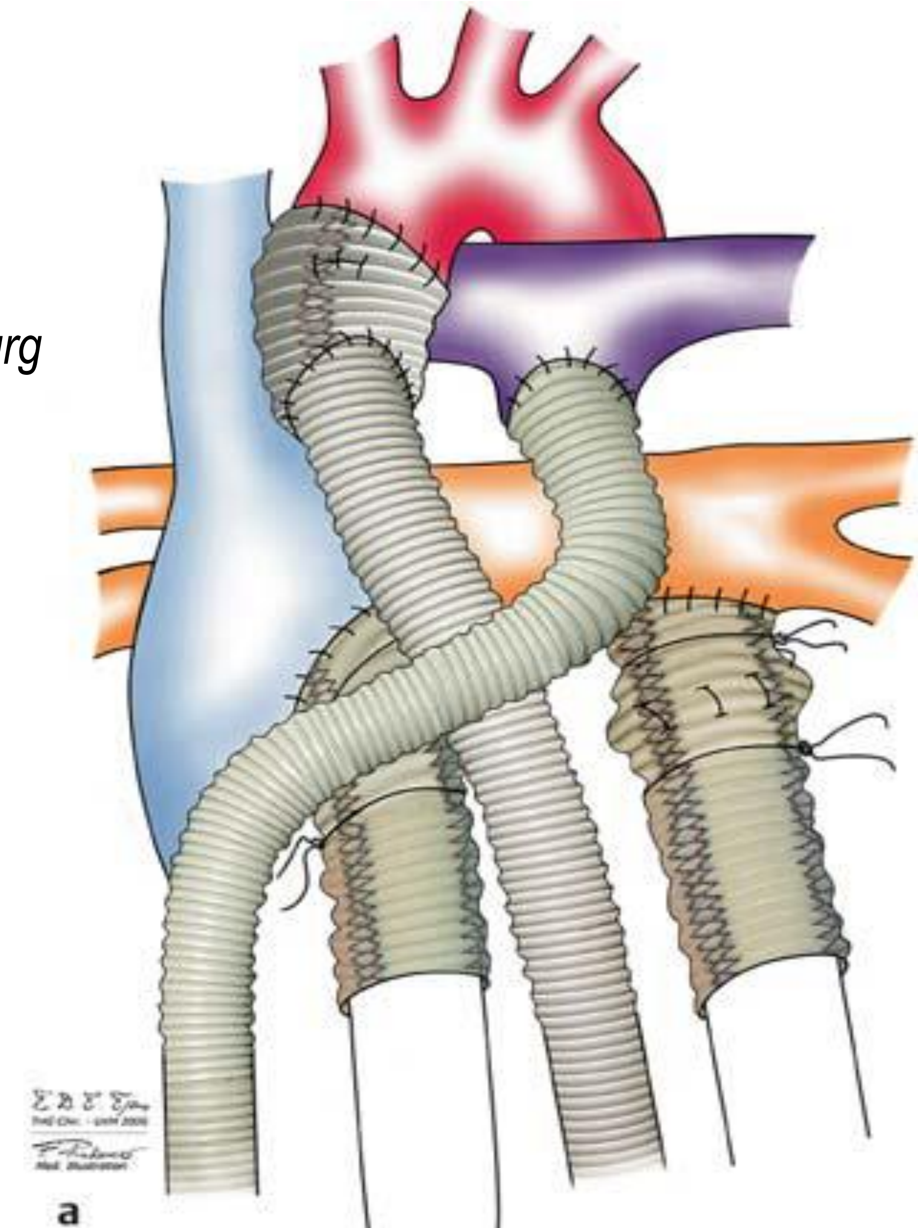
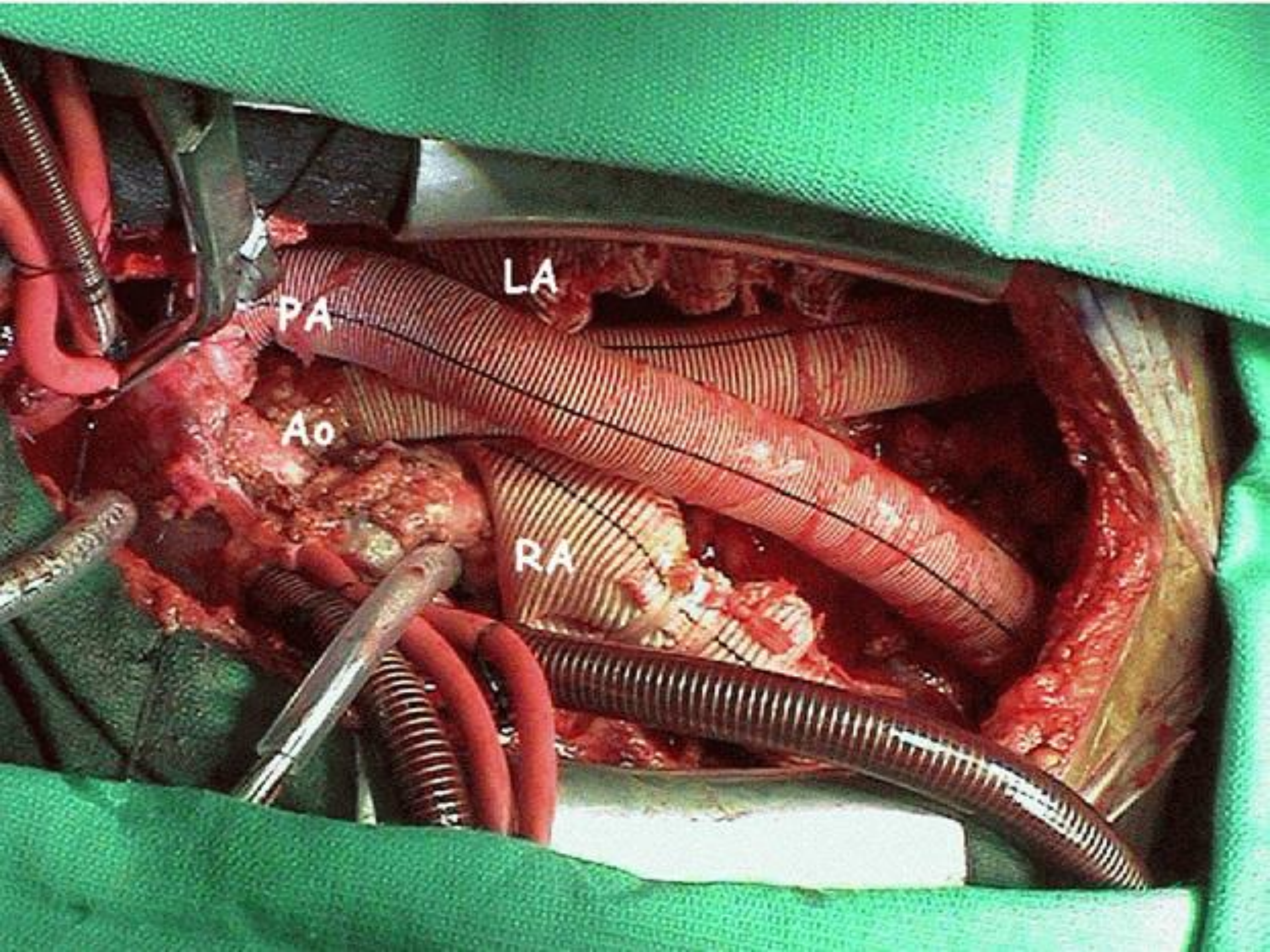
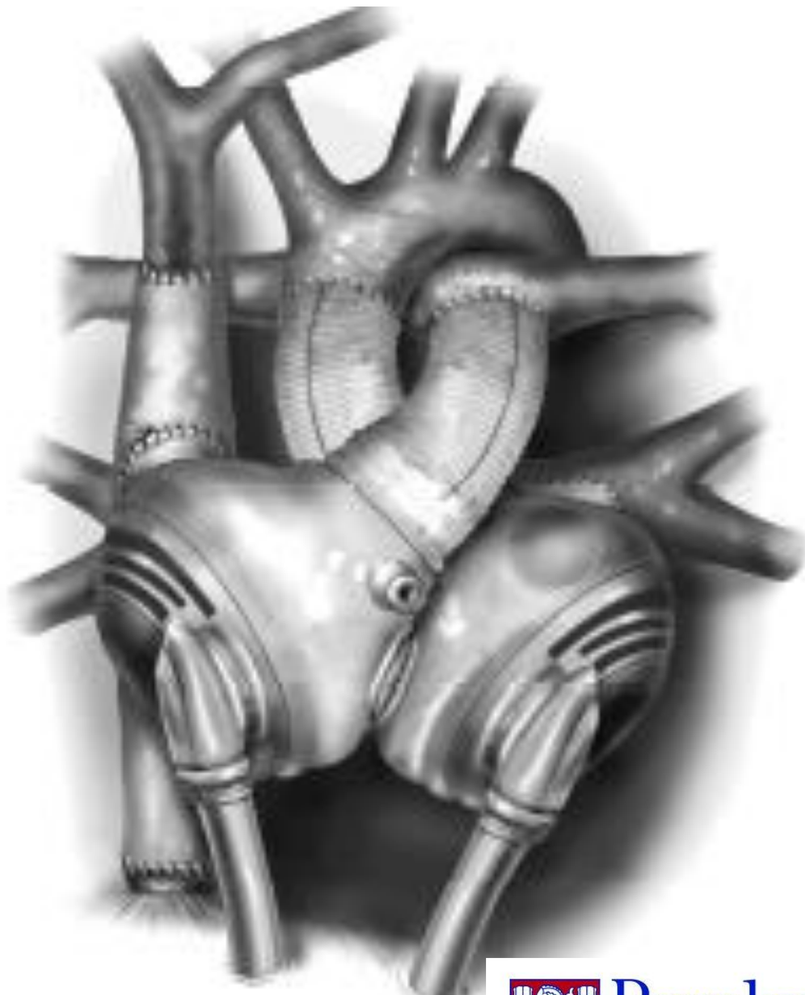


Figure 4. Connection of the outflow conduit from the Thoratec system with the pulmonary artery and the ascending aorta.


LA
PA
Ao
RA

SynCardia TAH



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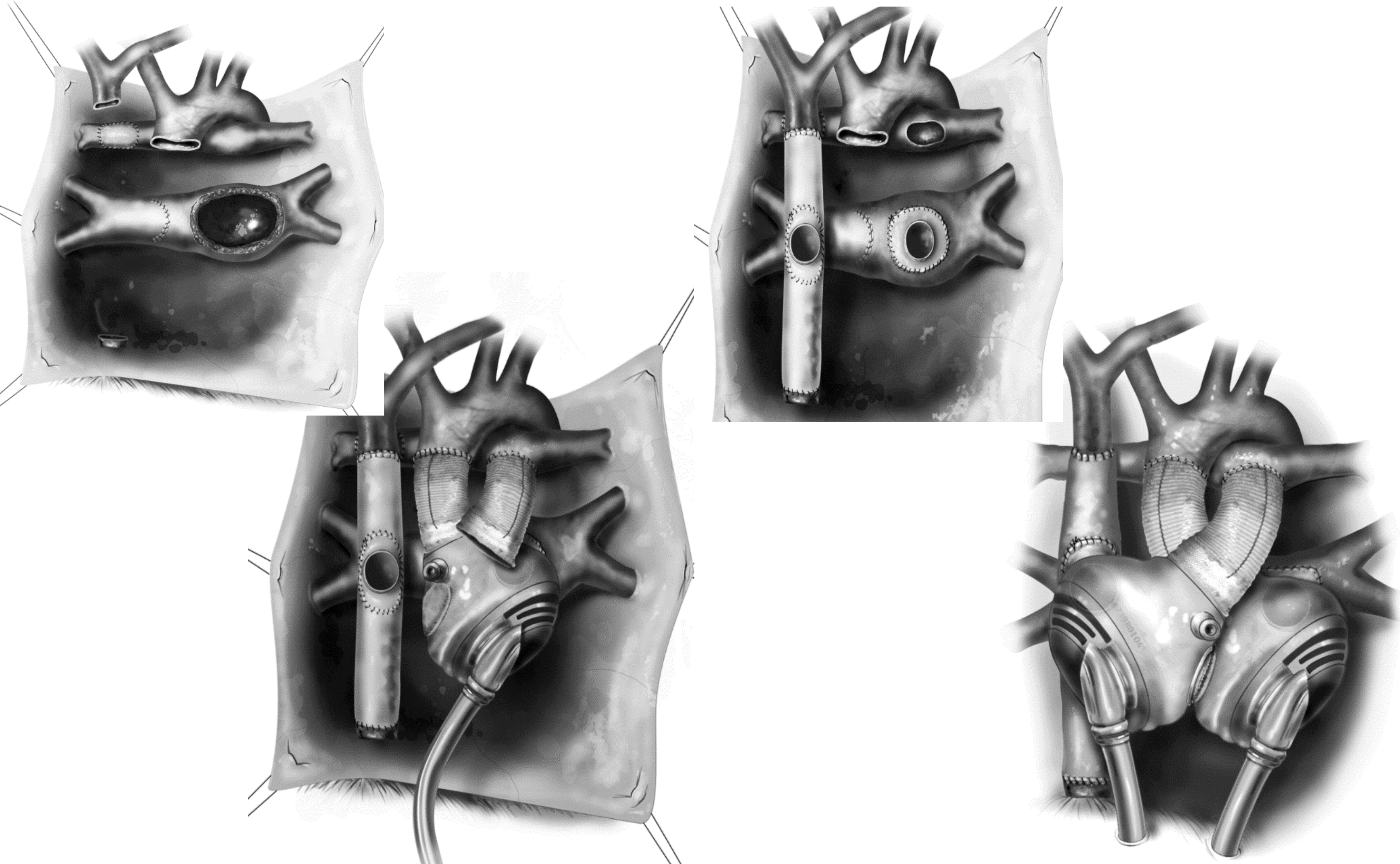

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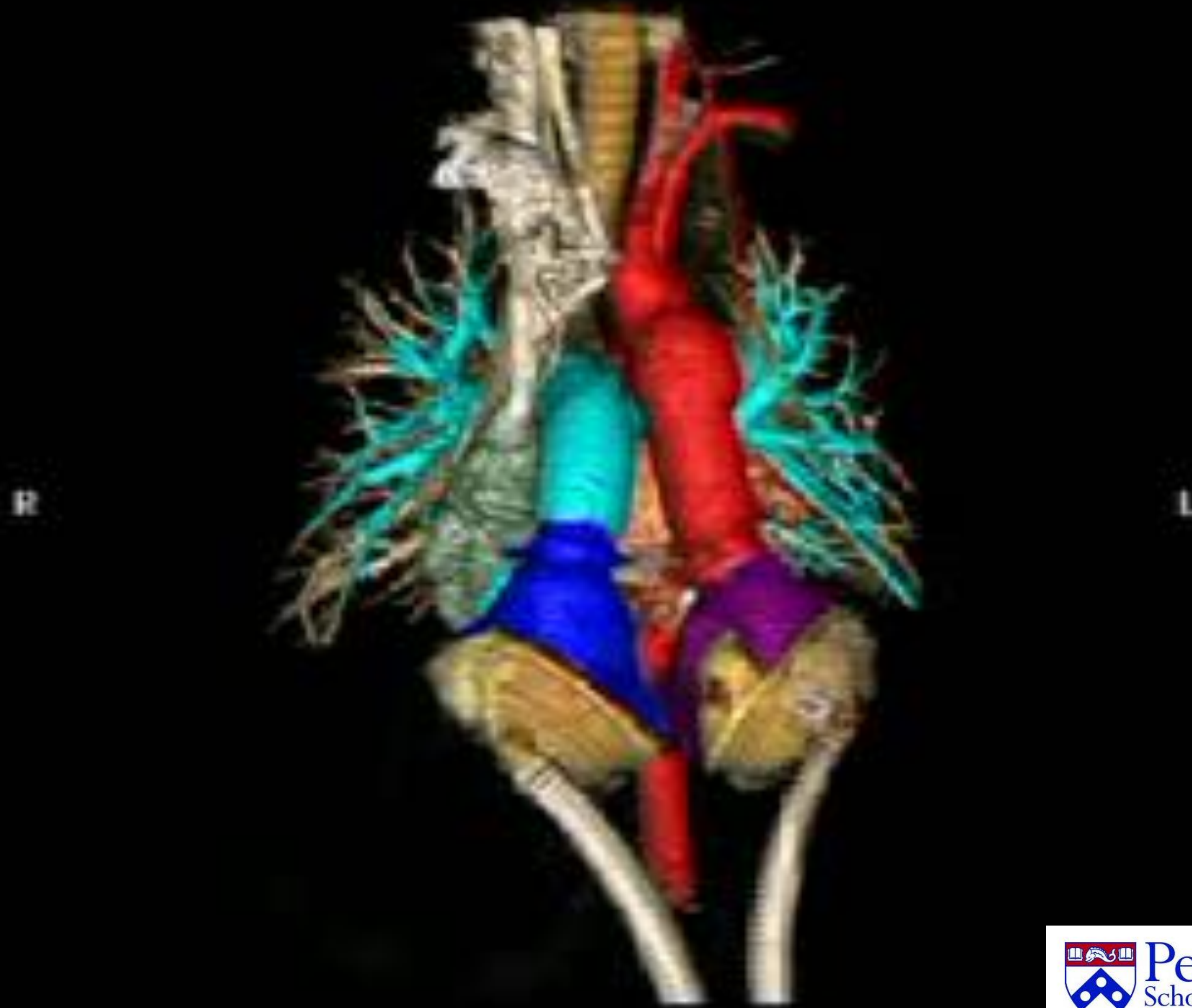
R
L

HeartWare

HeartWare Systems, Framingham, MA

- Long-term device
- Centrifugal
- Continuous flow
- 10L/min flow
- BSA as low as $0.5m^2$
- Discharge to home
- Small
  - Pericardial space

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An overview of mechanical circulatory support in single-ventricle patients

Jacob R. Miller[1], Timothy S. Lancaster[1], Connor Callahan[2], Aaron M. Abarbanell[3], Pirooz Eghtesady[3]

Transl Pediatr 2018;7(2):151-161

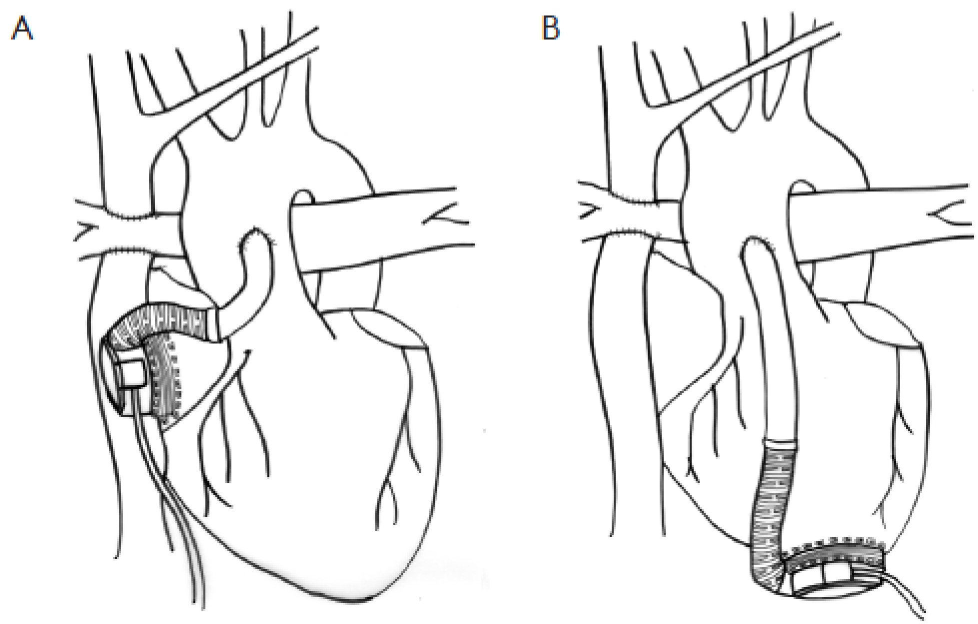

Figure 3 HeartWare HVAD placement in patients with a failing Fontan. (A) The HVAD inflow cannula is placed within the ventricular apex with the outflow cannula into the neoaorta; (B) the HVAD inflow cannula is placed into the atrium with the outflow cannula into the neoaorta.

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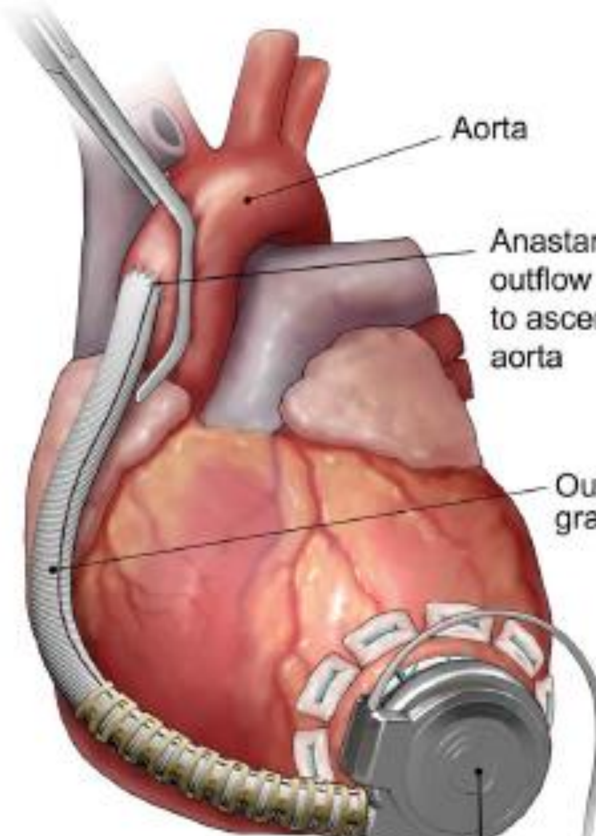
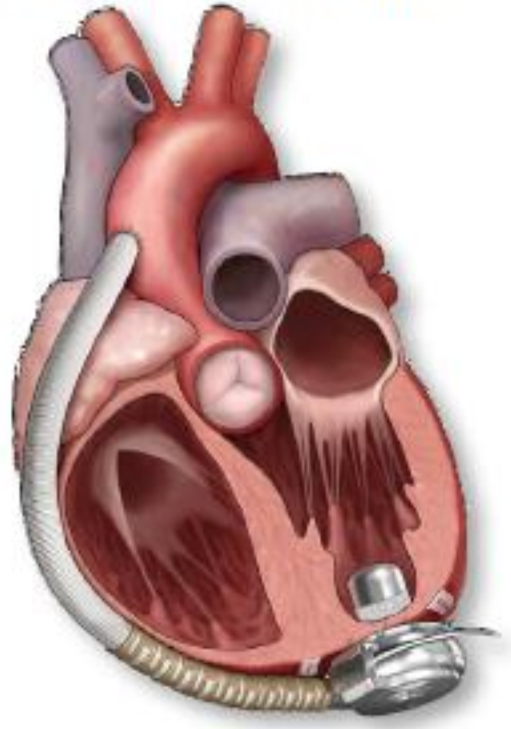


Pump Placement



CAUTION: ALWAYS ensure the inflow cannula position is pointed toward the mitral valve and parallel to the interventricular septum to optimize HVAD Pump operation.

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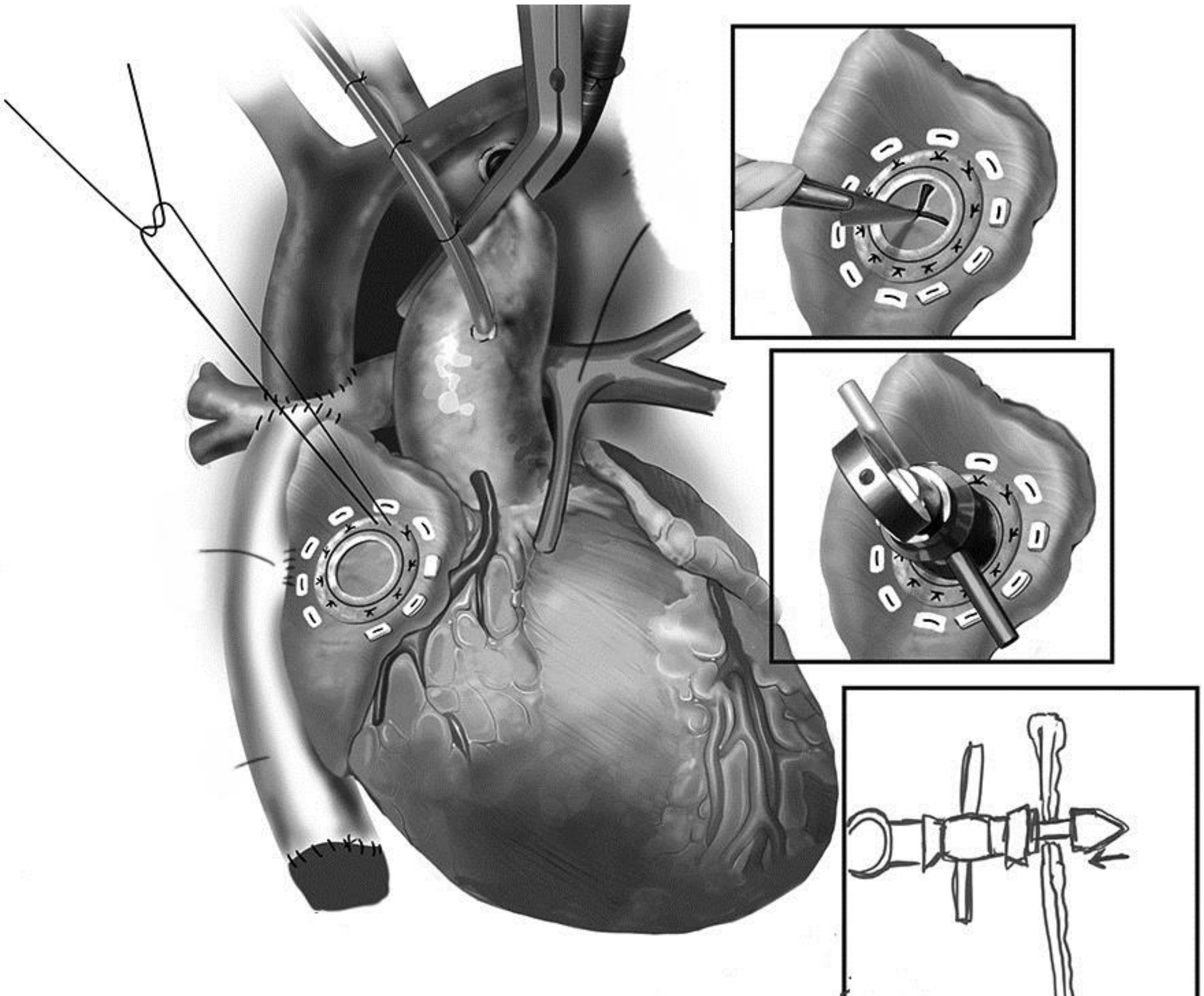

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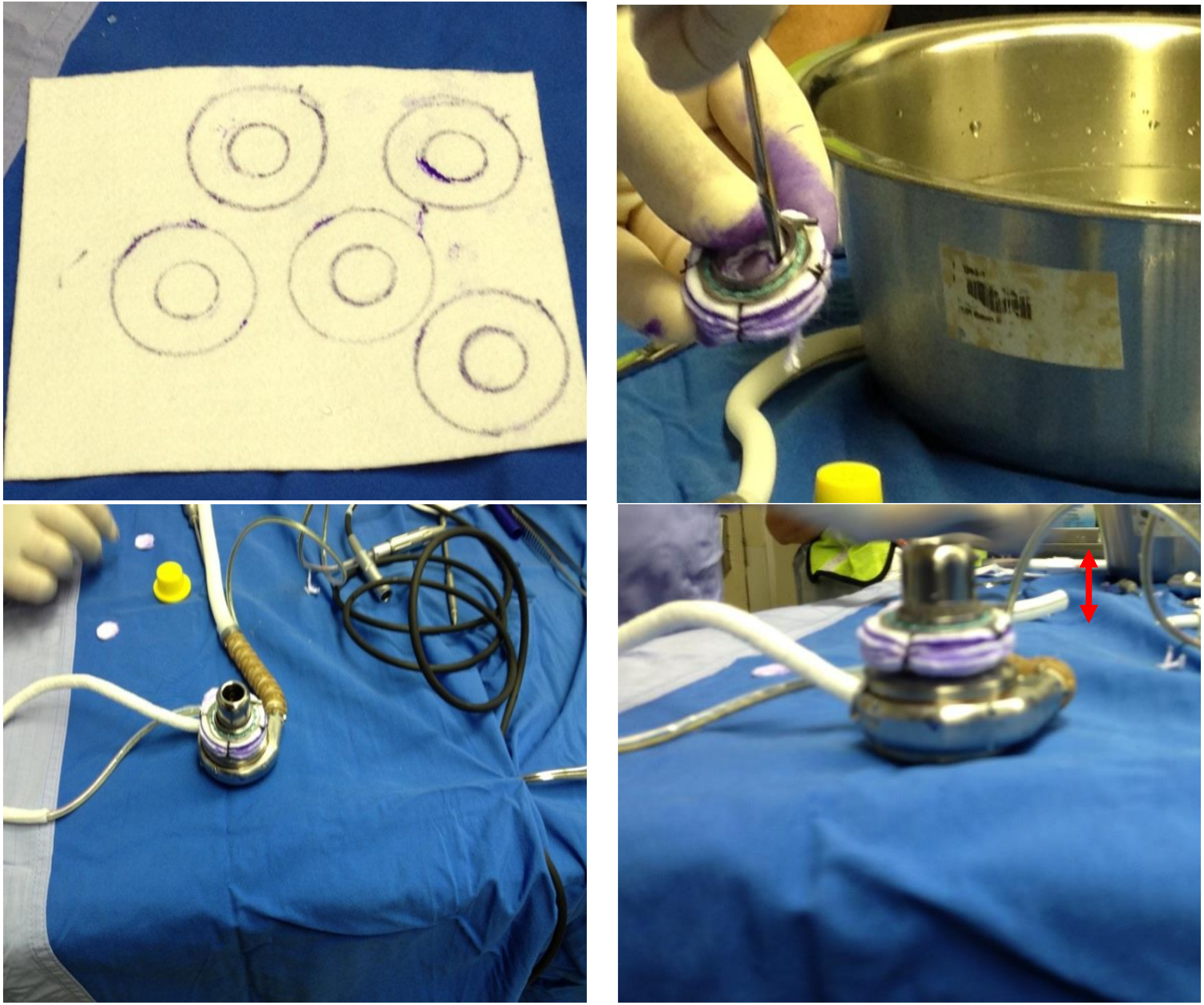

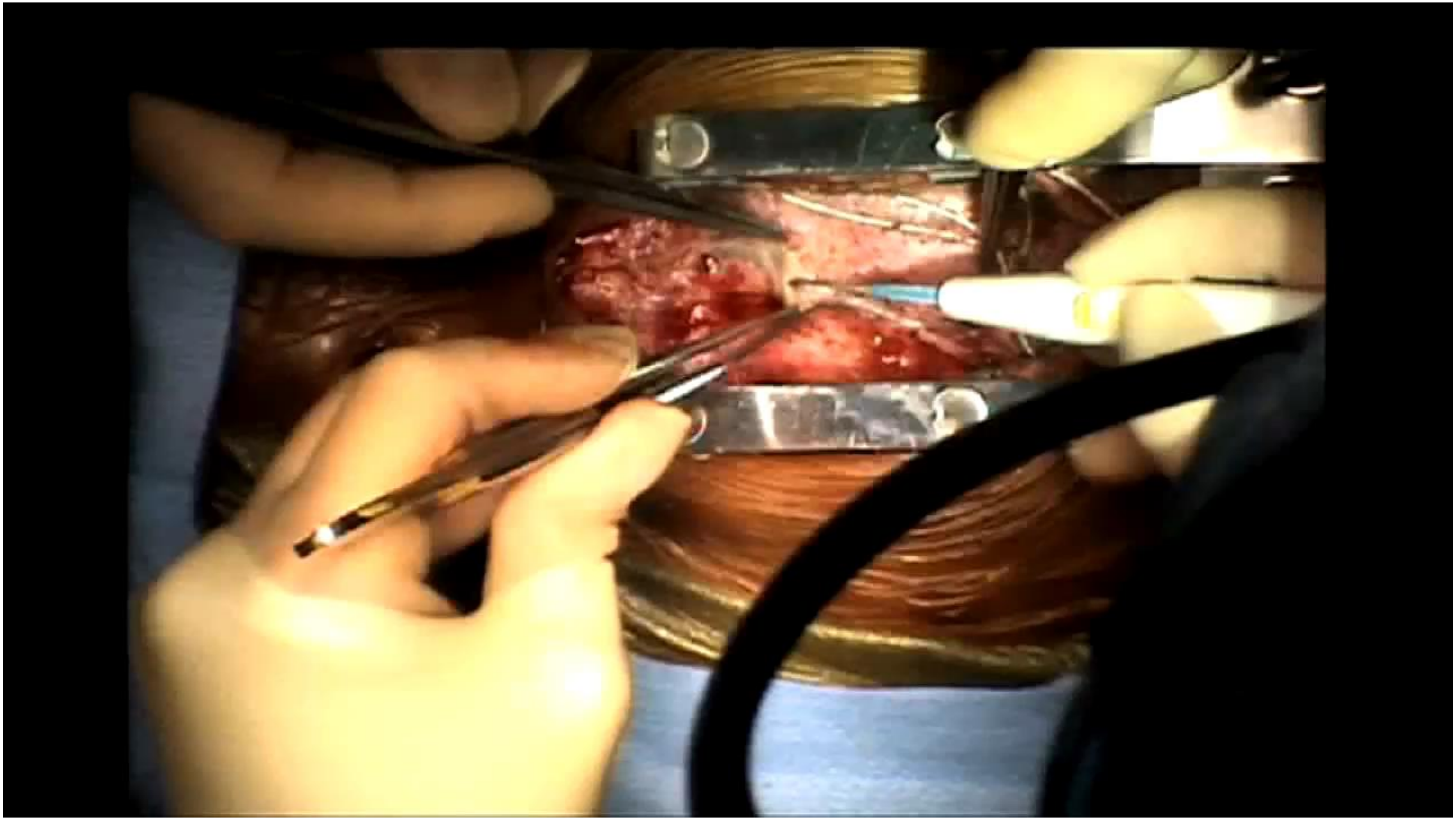

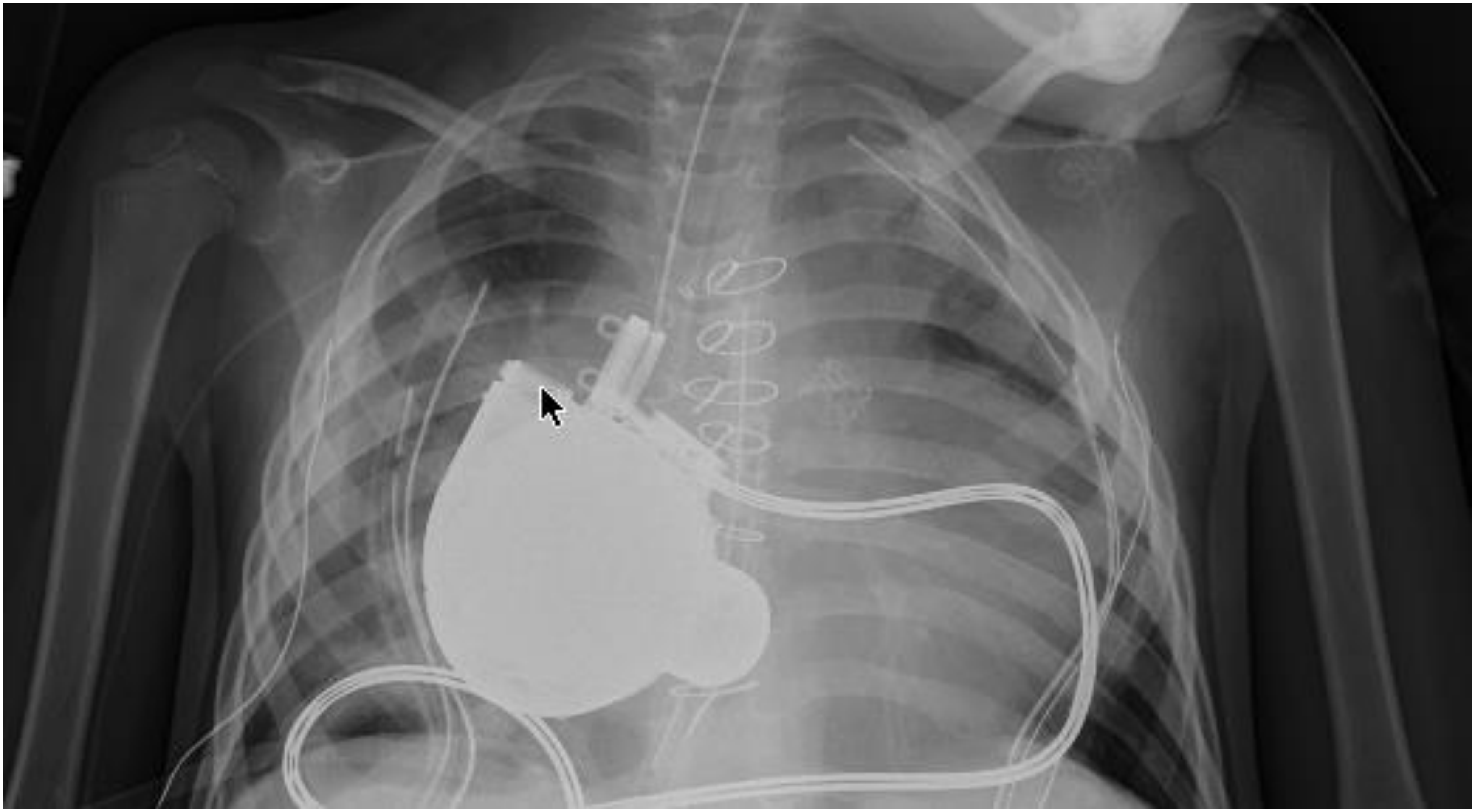

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Cavopulmonary Assist: (Em)powering the Univentricular Fontan Circulation

Mark D. Rodefeld,[a] Steven H. Frankel,[b] and Guruprasad A. Giridharan[c]

Table 2 Bioengineering Considerations for the Ideal Cavopulmonary Assist Device

1. 3-way or 4-way flow capability with a single pump/impeller
2. Percutaneous endovascular support which functions in the existing TCPC pathway and maximizes native endothelium exposure (non-synthetic pathways)
3. Optimization of passive TCPC flow patterns - whether functioning or not
4. Partial support in a location where there will be no myocardial recovery to assume function of the pump (*therefore, it is not a VAD*)
5. Pressure in the TCPC is low (10 to 12 mmHg); therefore thrombogenicity potential is high
6. Pressure boost required is very low (optimum, 6 to 8 mmHg; range, 0 to 30 mmHg)
7. Low preload and afterload dependence (a high degree of fluid slip):
   A. Prevent upstream vena caval suction collapse
   B. Prevent downstream perfusion lung injury
8. Ability to address/modify split differential vena caval inflow:
   A. SVC predominant flow in neonates/infants (SVC/IVC: 60/40)
   B. IVC predominant flow in adults (SVC/IVC: 30/70)
9. No fluid reservoir for the pump inlet to draw from
10. No barrier to recirculation should be required
11. The Fontan venous pathway must remain unobstructed during full support, during weaning, in the event of device failure, and after removal of the device

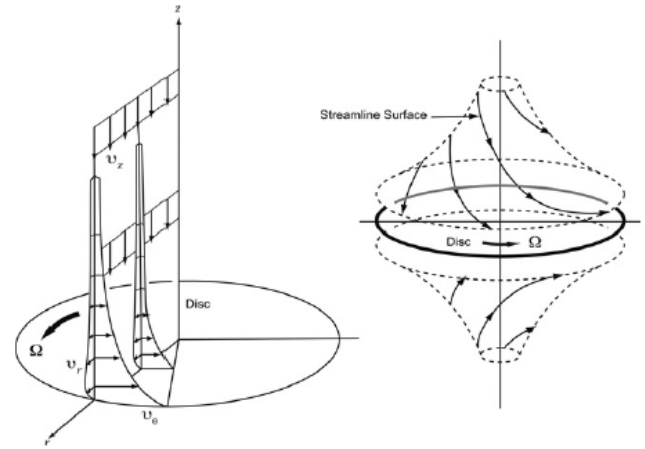

Figure 2 Von Karman viscous pump. *Left*, Fluid is induced to rotate by disc rotation, resulting in radial outflow. The outgoing fluid is replaced by inflow from the axial field. *Right*, On both sides of the disk, this results in opposed axial (vena caval) inflow and orthogonally opposed (pulmonary arterial) outflow.

Table 3 Therapeutic Potential of Cavopulmonary Assist

Existing paradigm:
- Adult failing Fontan
  - Bridge-to-recovery
  - Bridge-to-transplant
- Stage-2 and -3 repair
- Stabilization after repair
- Destination therapy: the "biventricular Fontan"
- Static percutaneous implant
- Passive flow optimization

New paradigm:
- Combined Stage 2-3
  - One-stage Fontan conversion
- Combined Stage 1-2
  - Norwood (no shunt) + Glenn
- Combined Stage 1-3
  - Neonatal Fontan
- Support for other hybrid approaches
  - Percutaneous Fontan completion

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