Solutions for TGA, VSD and LVOTO

Victor O Morell, MD

Eugene S Wiener Professor and Chair

Pediatric CT Surgery

Children's Hospital of Pittsburgh

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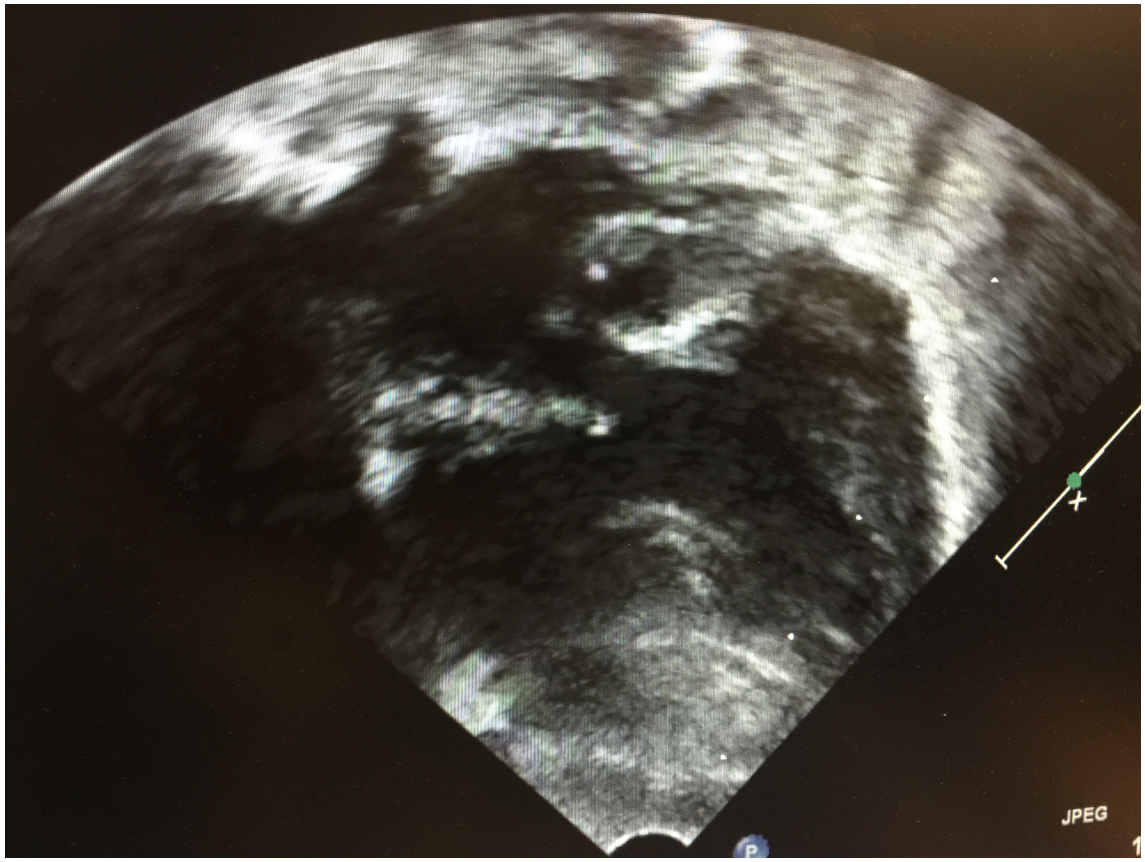
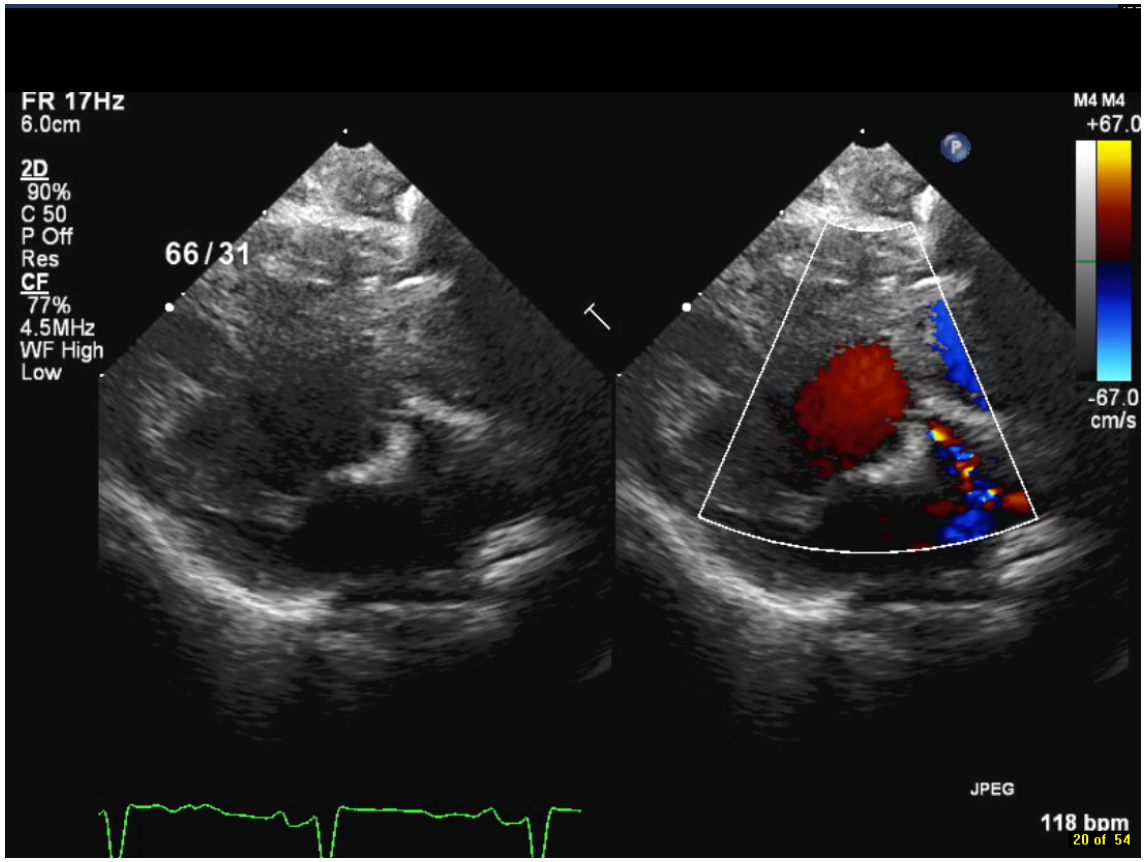
Disclosure

- **Xeltis Pulmonary Valve Conduit**

Heterogeneous Group of Patients Important to define:

- **LVOTO**
  - PV annulus size (Z-score)
  - Morphology of the PV
  - Subvalvar obstruction
- **VSD type**
  - Location/size
- **RV size**
- **AVV straddling**
- **Great vessels spatial relationship**
- **Coronary anatomy**





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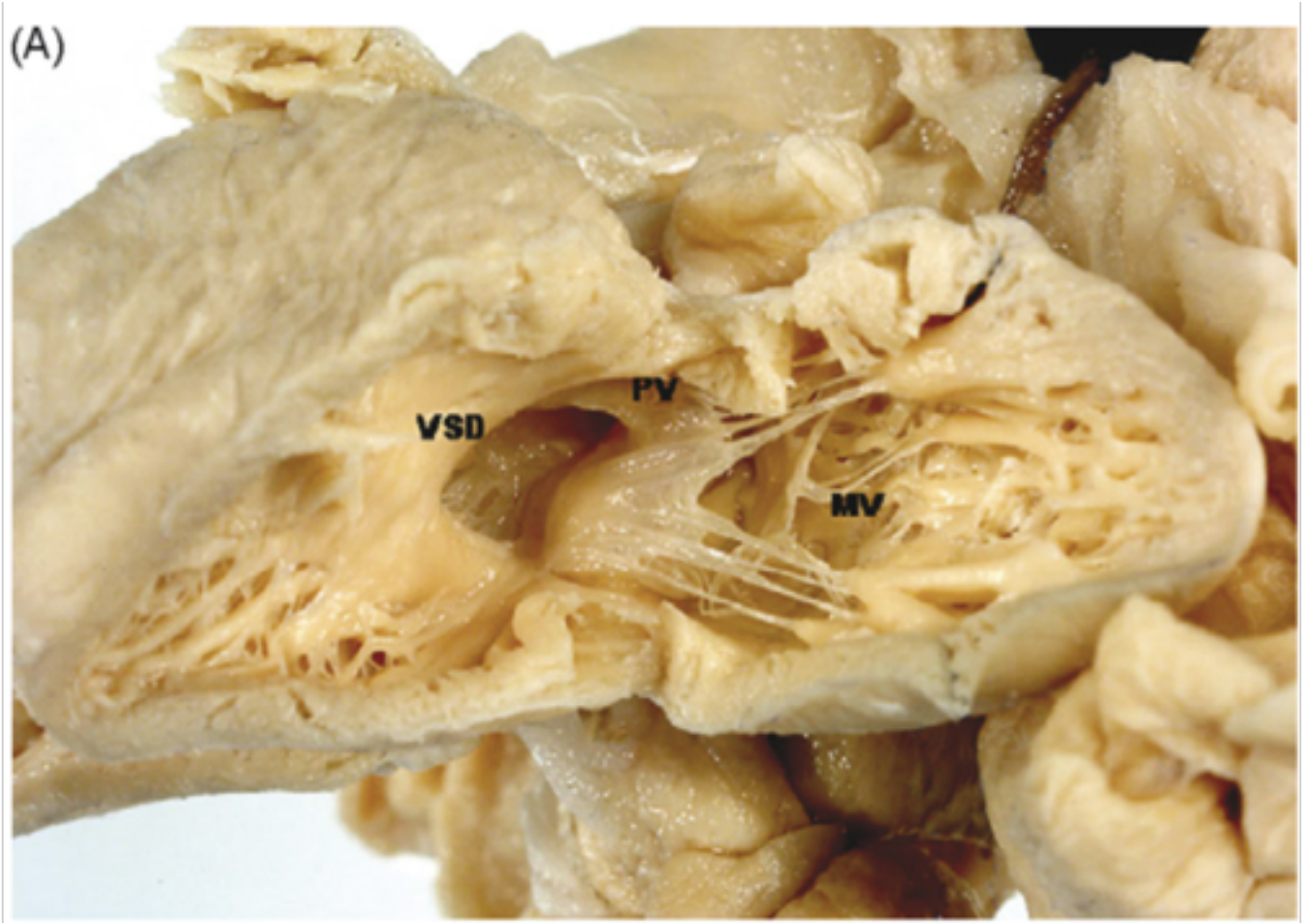
Anatomy
VSD type



Hazekamp et al, 2007

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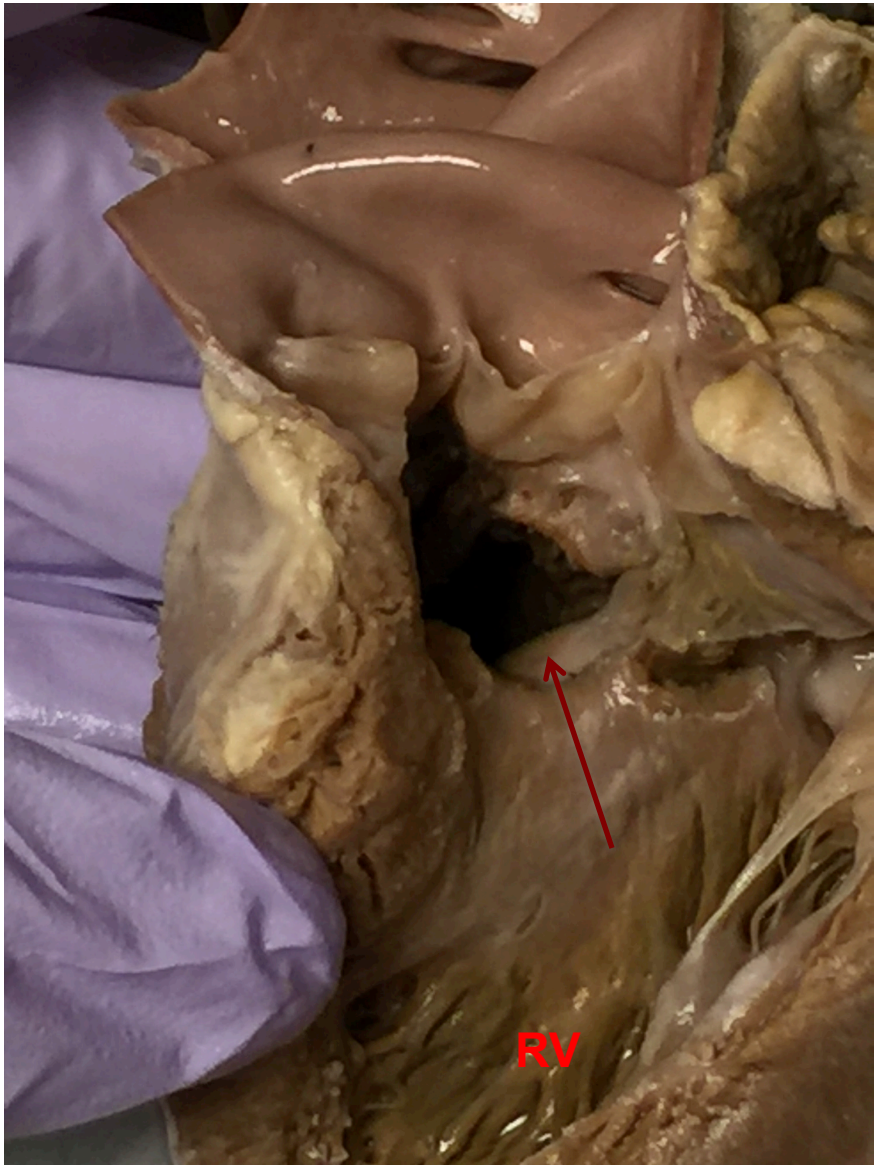
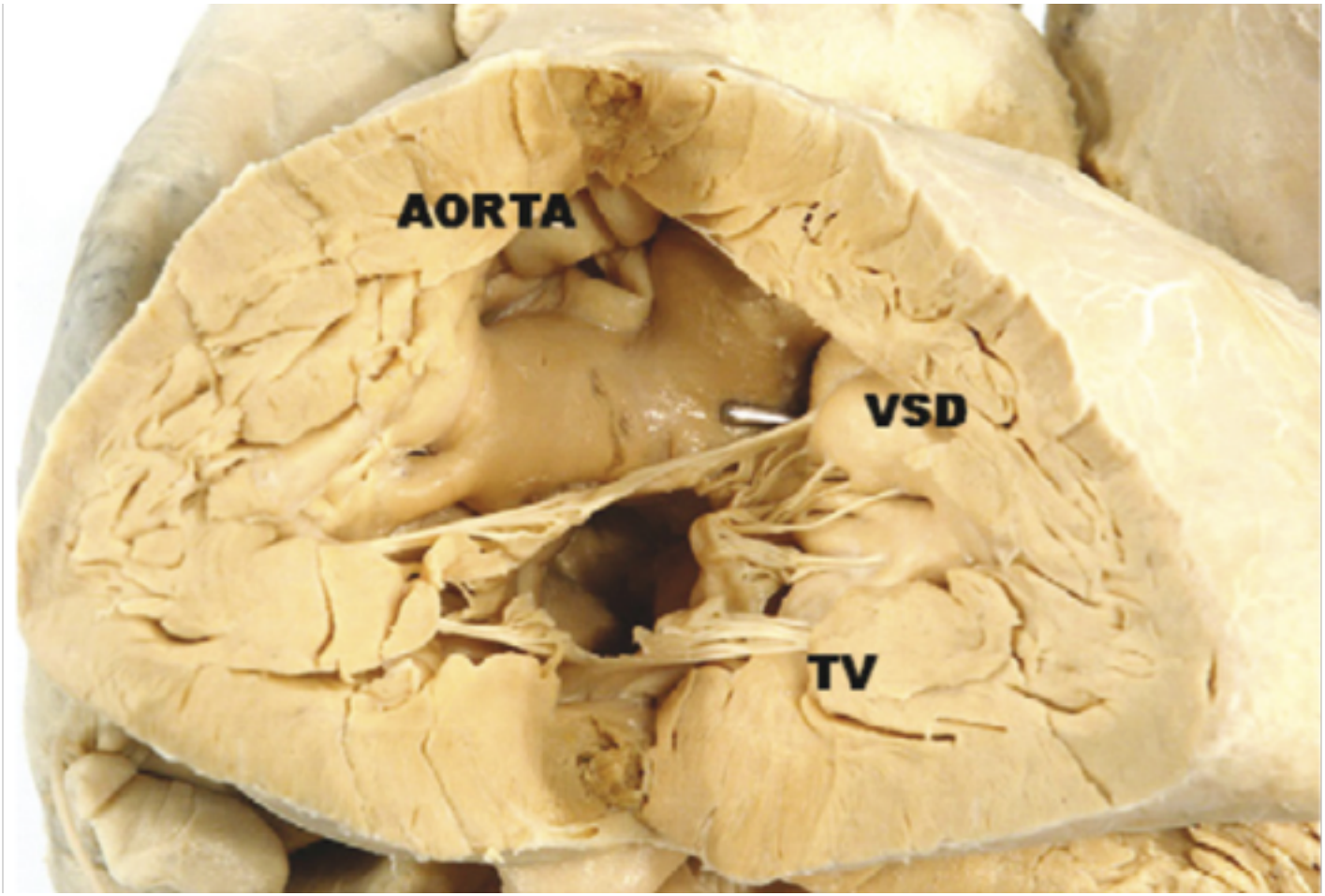


Anatomy
RV volume



Hazekamp et al, 2007

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Anatomy
AVV: Straddling

Hazekamp et al, 2007

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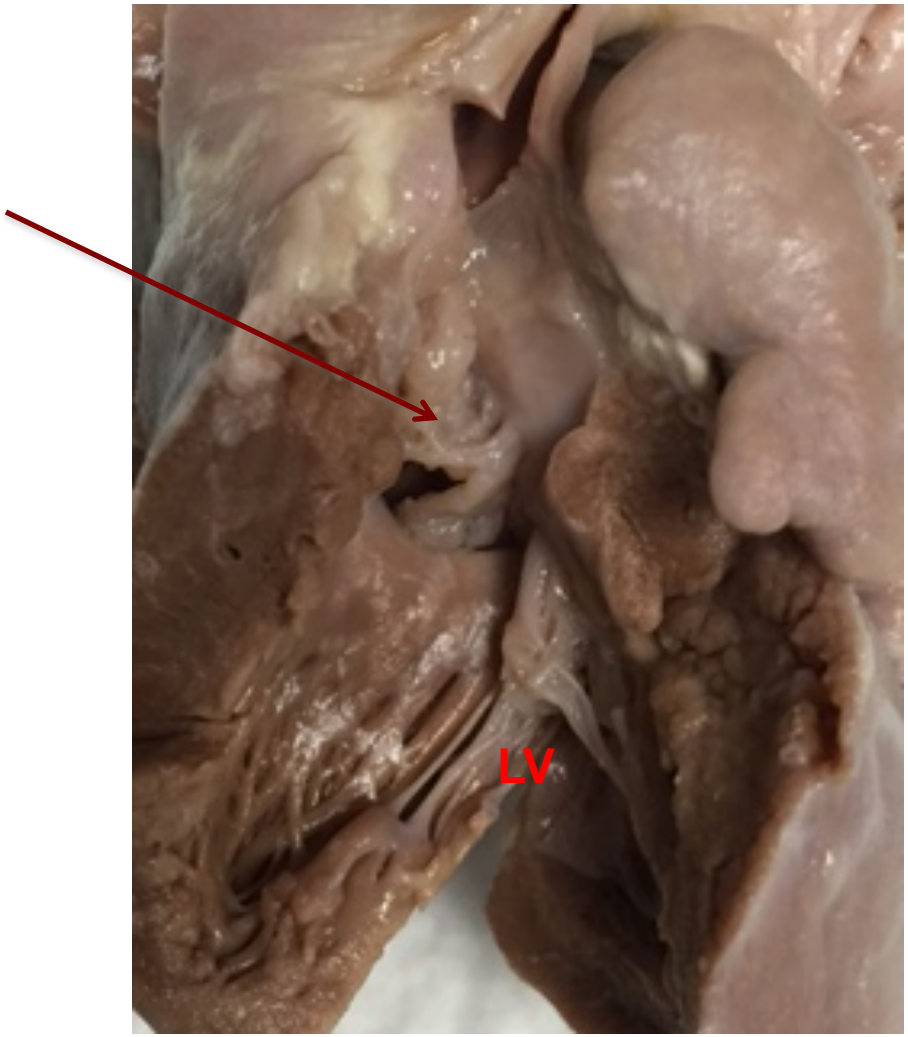
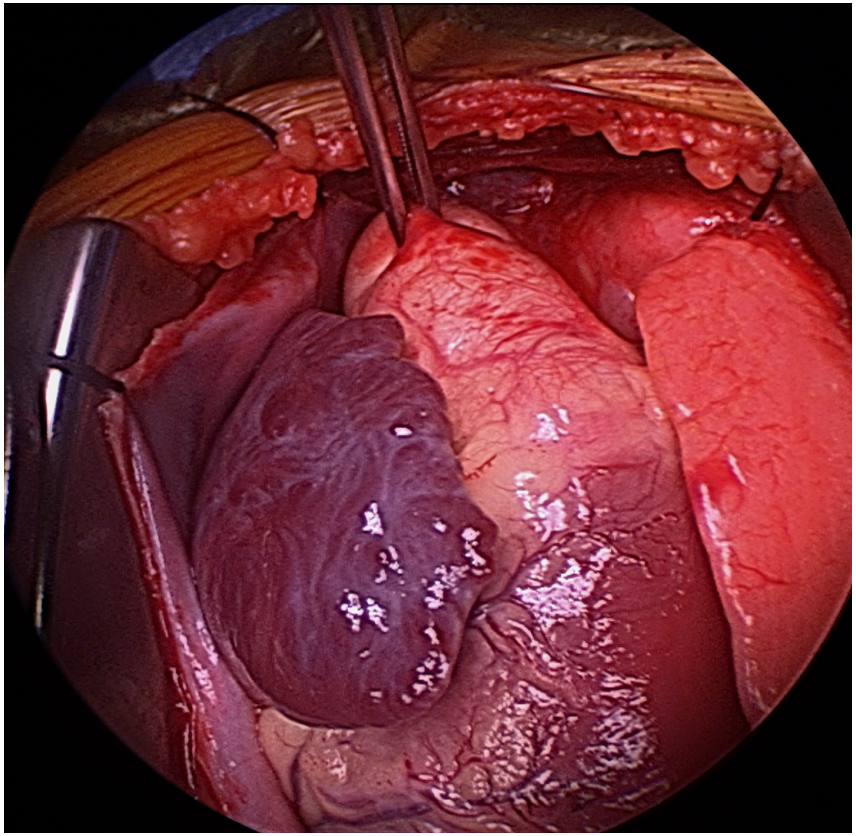
Abnormal TV attachments

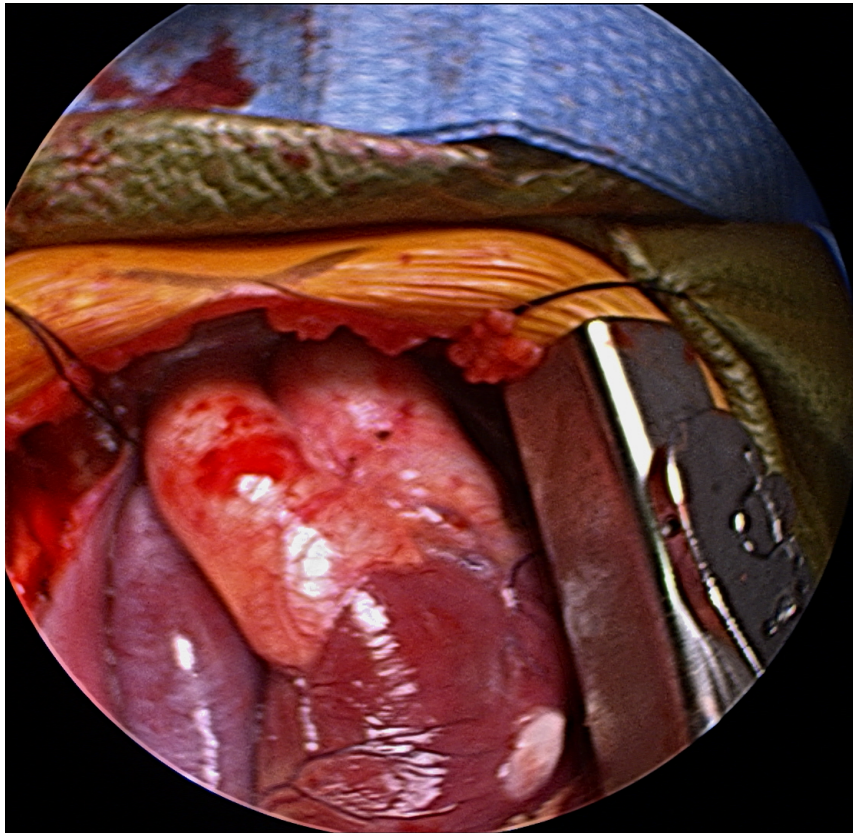


Great Vessels

AP



Side by Side



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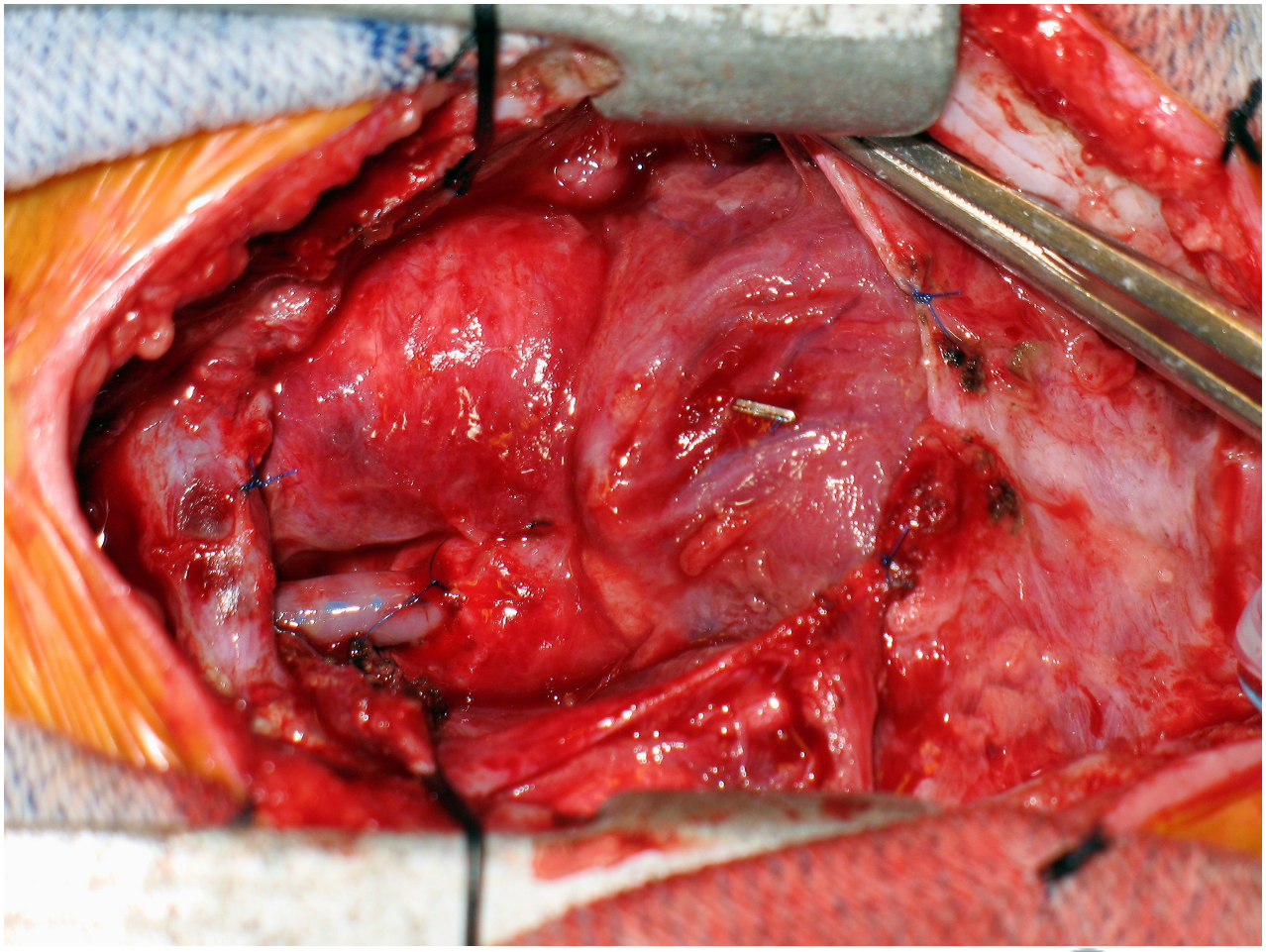
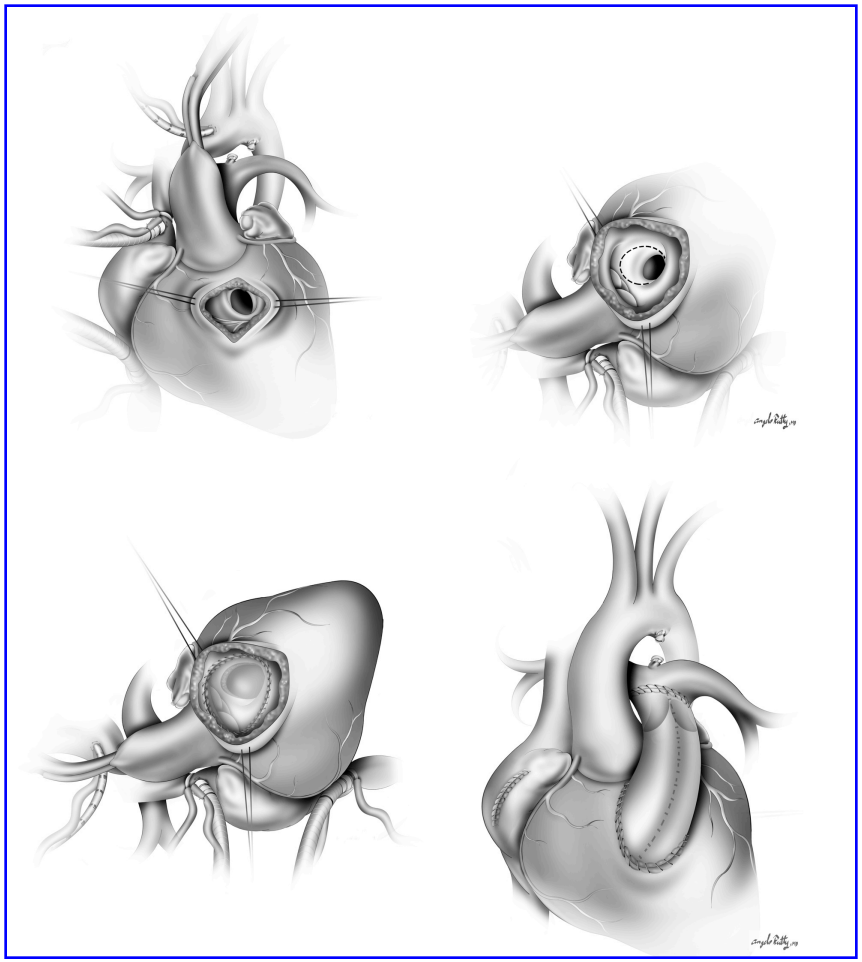
Coronary Arteries

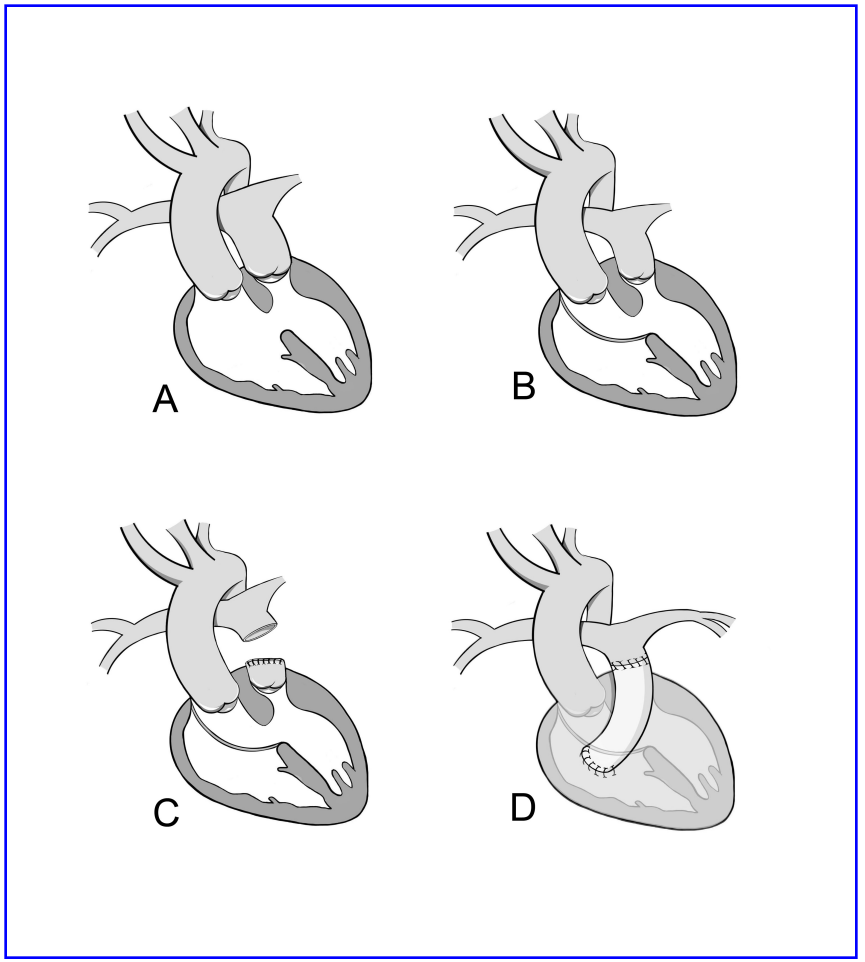


Surgical Options

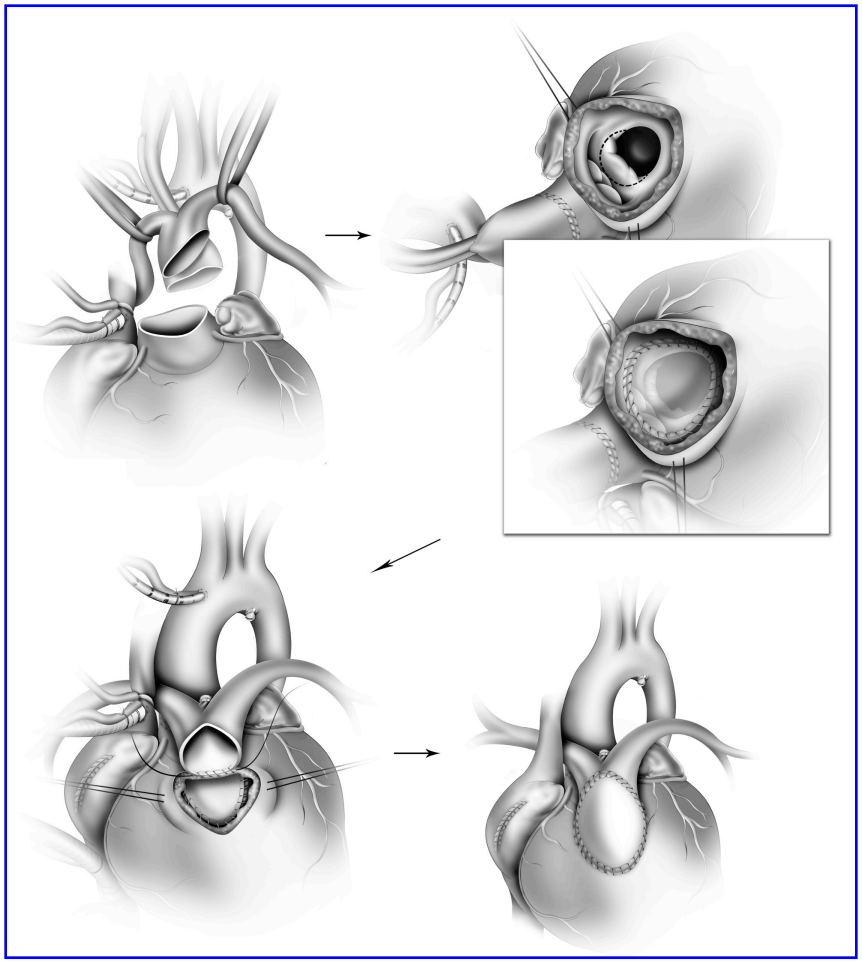
1. Rastelli Repair (1969)
2. Aortic translocation/Bex-Nikaidoh Procedure (1980/84)
3. Lecompte Intraventricular Repair (REV Procedure) (1988)
4. ASO/VSD closure/resection of LVOTO
5. Single Ventricle

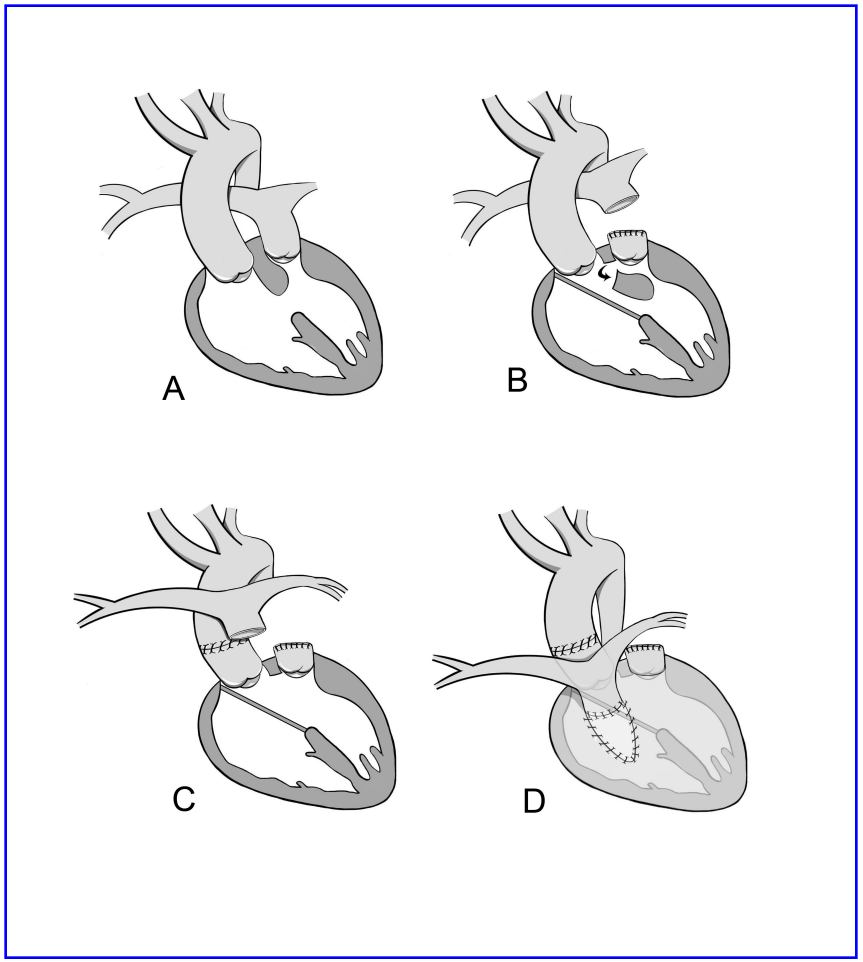
Rastelli



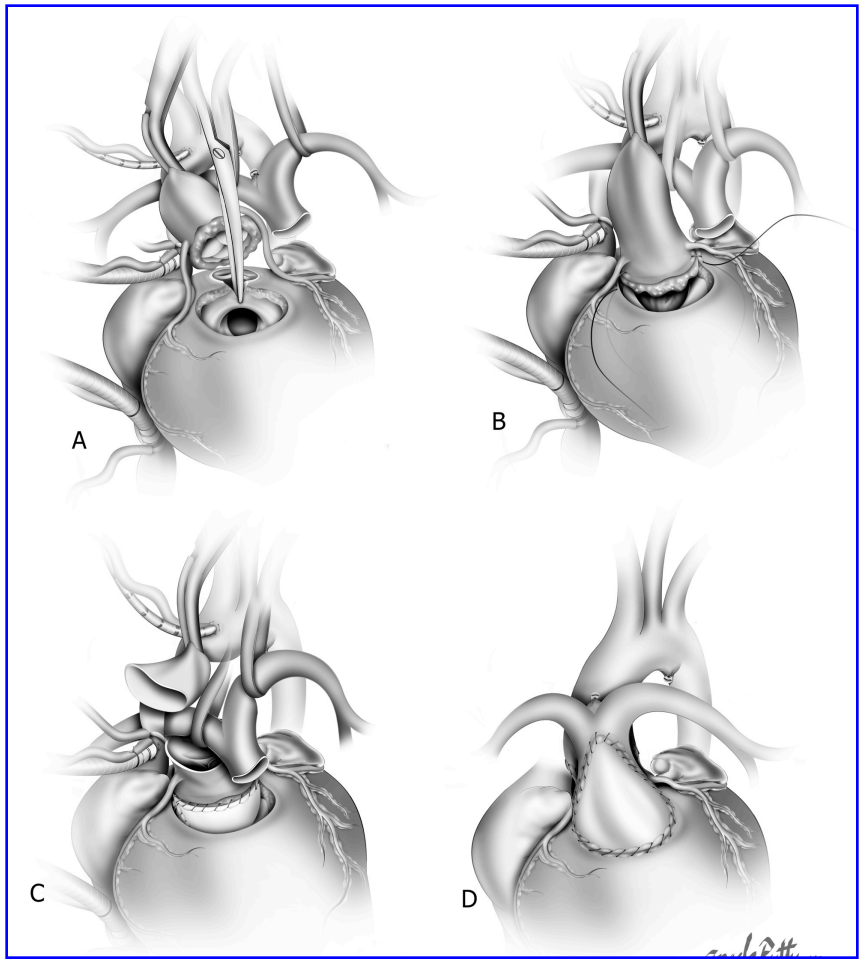


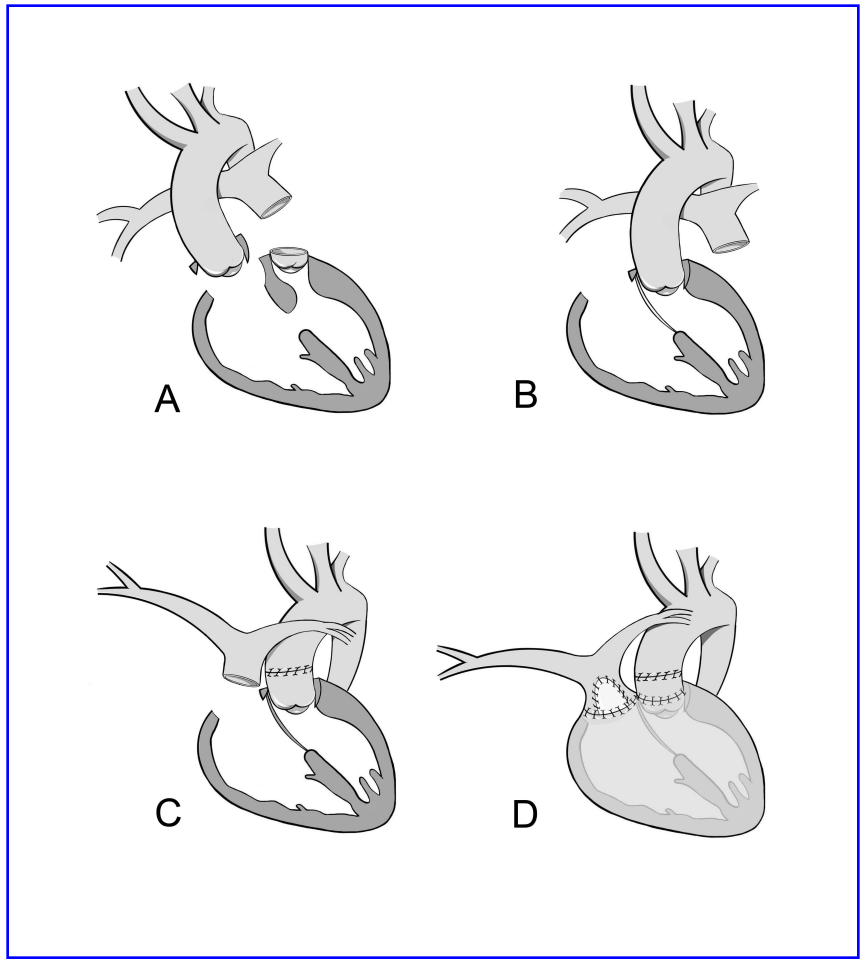
REV



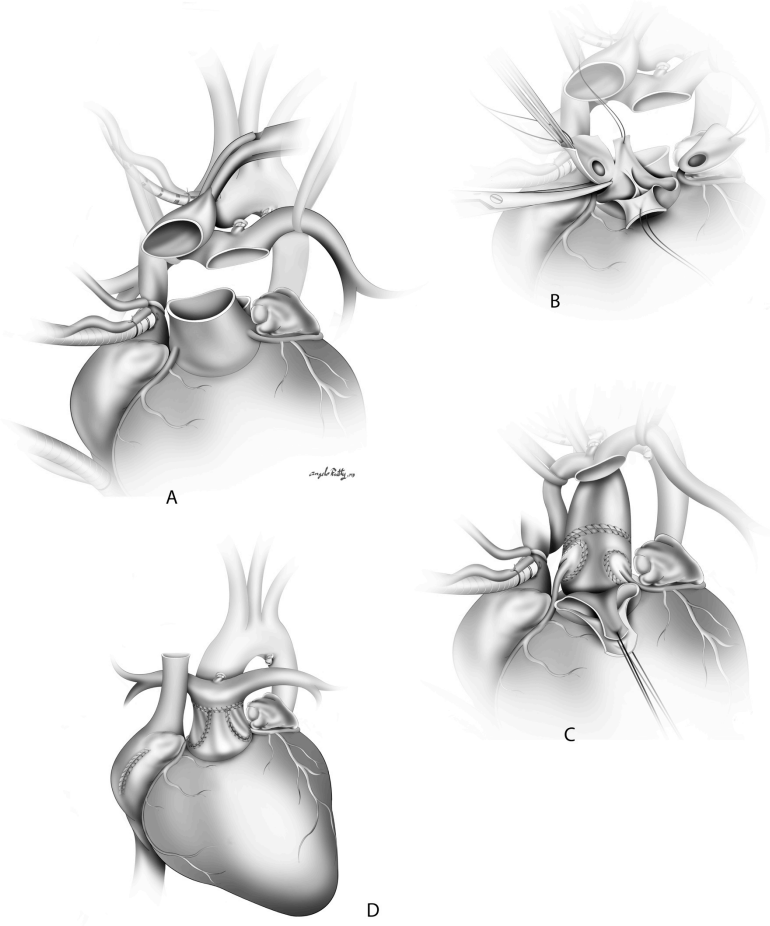


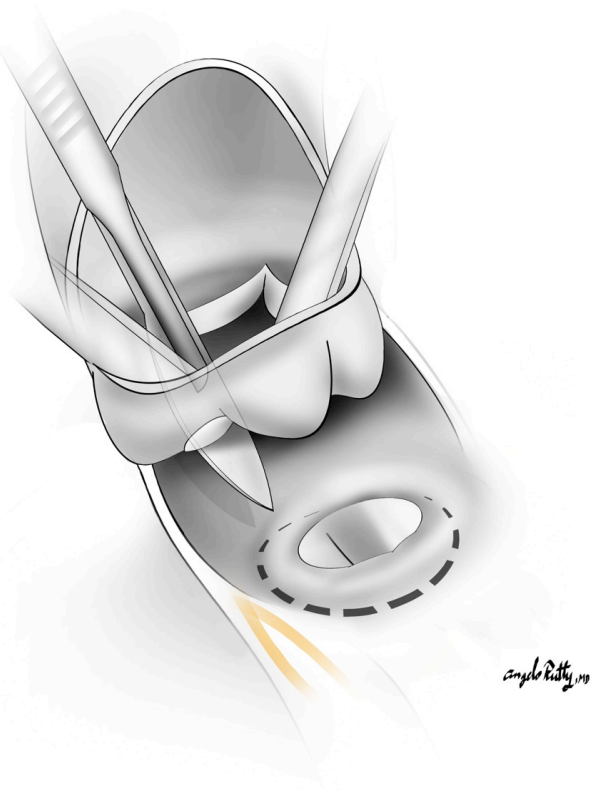
Bex-Nikaidoh





ASO with LVOT resection





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STS Congenital Heart Surgery Data Summary
All Patients

Participant 50051
STS Period Ending 06/30/2017

Duke Clinical Research Institute

Table 4: Primary diagnosis by anomaly, Last 4 Years(Jul 2013 - Jun 2017) - cont.

Primary Diagnosis	Participant		STS	
	N	% of All	N	% of All
Single ventricle, Other	3	0.2%	365	0.3%
Single Ventricle + Total anomalous pulmonary venous connection (TAPVC)	2	0.1%	180	0.1%
TRANSPOSITION OF THE GREAT ARTERIES				
Congenitally corrected TGA	5	0.3%	122	0.1%
Congenitally corrected TGA, IVS	0	0.0%	91	0.1%
Congenitally corrected TGA, IVS-LVOTO	0	0.0%	25	0.0%
Congenitally corrected TGA, VSD	3	0.2%	232	0.2%
Congenitally corrected TGA, VSD-LVOTO	0	0.0%	196	0.2%
TGA, IVS	26	1.5%	2,073	1.7%
TGA, IVS-LVOTO	1	0.1%	29	0.0%
TGA, VSD	8	0.5%	1,367	1.1%
TGA, VSD-LVOTO	3	0.2%	359	0.3%

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STS Congenital Heart Surgery Data Summary
All Patients

Participant 50051
STS Period Ending 06/30/2017

Duke Clinical Research Institute

Table 5: Primary procedure by anomaly, Last 4 Years (Jul 2013 - Jun 2017) - cont.

Primary Procedure	Participant				STS		
	Overall		Mortality				
	N	% of All	N	%	N	% of All	% Mort.
Mustard	0	0.0%	-	-	18	0.0%	0.0%
Atrial baffle procedure, Mustard or Senning revision	2	0.1%	0	0.0%	29	0.0%	3.4%
Rastelli	3	0.2%	0	0.0%	203	0.2%	3.0%
REV	0	0.0%	-	-	9	0.0%	11.1%
Aortic root translocation over left ventricle (Including Nikaidoh procedure)	4	0.2%	0	0.0%	76	0.1%	2.6%
TGA, Other procedures (Kawashima, LV-PA conduit, other)	0	0.0%	-	-	5	0.0%	20.0%

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Pediatr Cardiol 2015
DOI 10.1007/s00246-015-1092-x

ORIGINAL ARTICLE

D-Transposition of the Great Arteries with Ventricular Septal Defect and Left Ventricular Outflow Tract Obstruction (D-TGA/VSD/LVOTO): A Survey of Perceptions, Preferences, and Experience

Mohammed K. Al-Jughiman · Maryam A. Al-Omair · Glen S. Van Arsdell · Victor O. Morell · Marshall L. Jacobs

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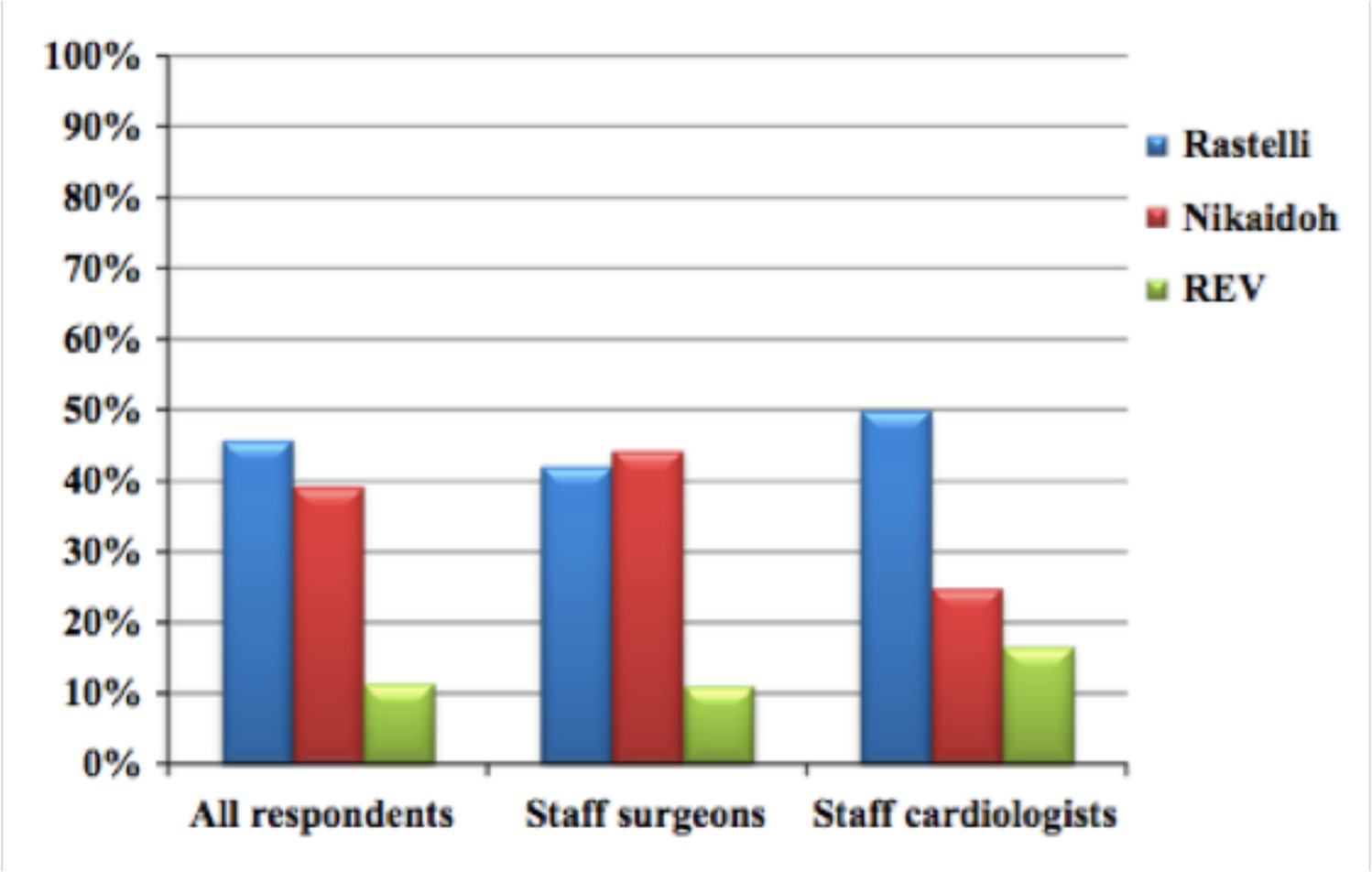
Procedure that should be considered the first option



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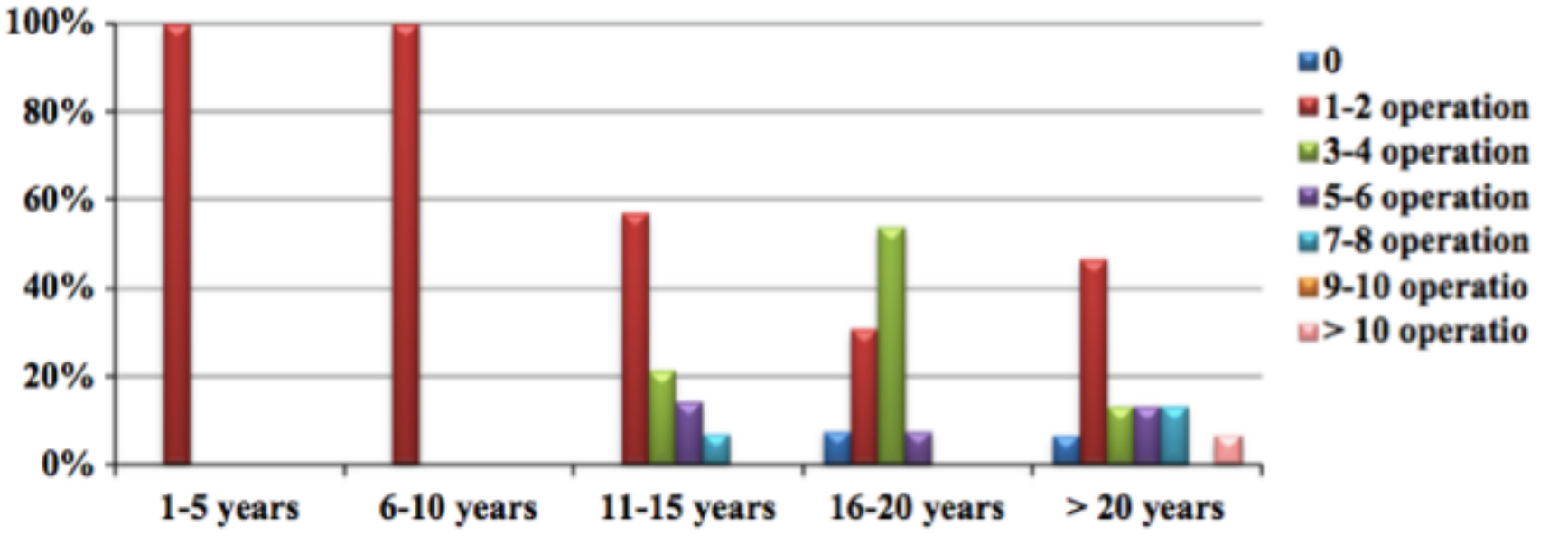
Total number of Rastelli, Nikaidoh, and REV performed per year on average stratified by surgeons experience in the field



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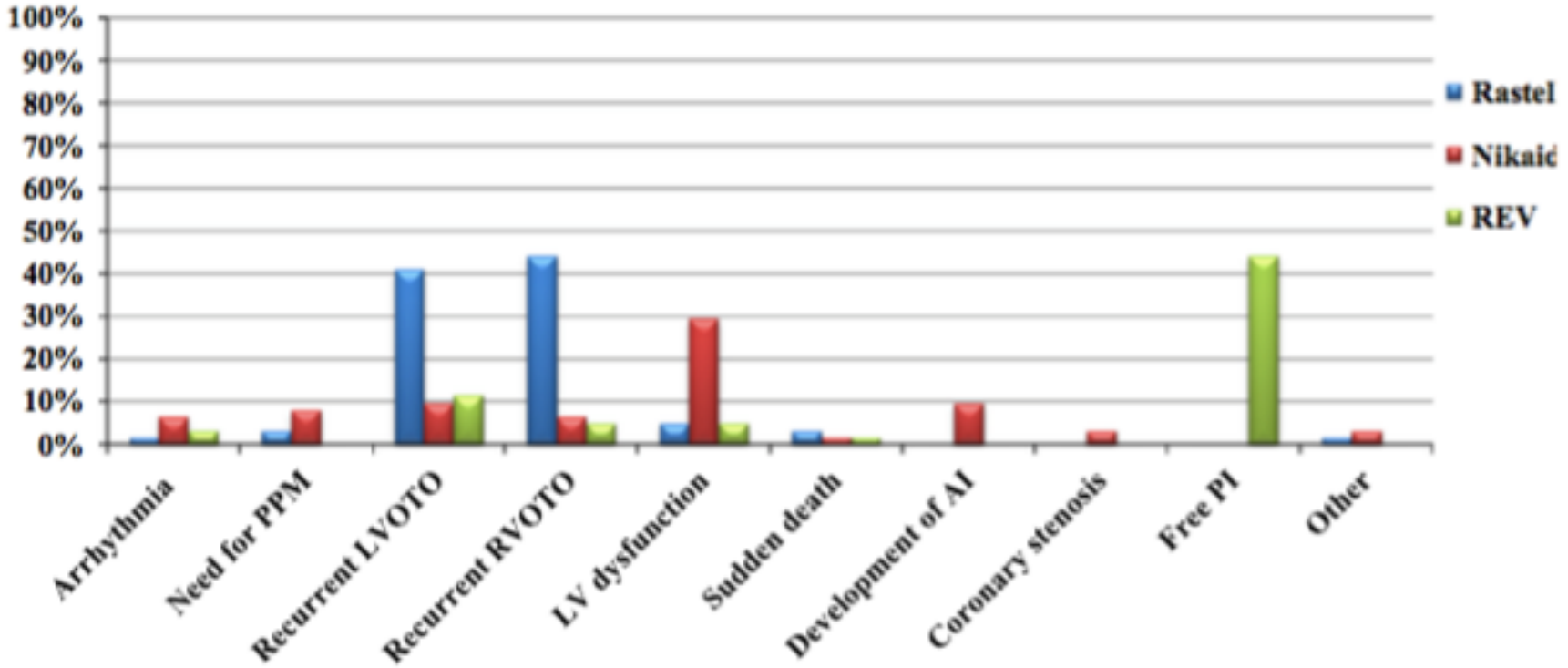
What worries clinicians the most after each procedure?



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Anatomy

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European Journal of Cardio-thoracic Surgery 31 (2007) 879–887

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

www.elsevier.com/locate/ejcts

The optimal procedure for the great arteries and left ventricular outflow tract obstruction. An anatomical study☆

Mark Hazekamp*, Francisco Portela, Margot Bartelings

Departments of Cardiothoracic Surgery and Anatomy, Leiden University Medical Center, Leiden, The Netherlands

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Hazekamp et al, 2007

Table 2
Anatomical characteristics and suggestions for surgical repair in specimens with TGA/VSD and LVOTO

Number	Basic anomaly	VSD	LVOTO	Best procedure	Alternative procedure	VSD enlargement necessary with Rastelli?	Comment	Position great arteries	Coronary anatomy
181	TGA/VSD/LVOTO	Perimembranous subpulmonary	Obstructive, dysplastic, bicuspid pulmonary valve; annular hypoplasia	Nikaidoh	REV/Rastelli	Yes	REV/Rastelli will result in a small RV	Aorta right anterior	1LCx-2R
435	TGA/VSD/LVOTO	Perimembranous subpulmonary	Obstructive, dysplastic, bicuspid pulmonary valve; annular hypoplasia; straddling mitral valve; cleft anterior mitral leaflet	Univentricular pathway	–		Resection of the outlet septum is necessary but will result in detachment of the mitral valve	Aorta anterior	1LCx-2R
884	TGA/VSD/LVOTO	Perimembranous subpulmonary	Straddling mitral valve; cleft anterior mitral leaflet	Univentricular pathway	–		Relief of LVOTO cannot be performed without damage to the mitral valve	Aorta anterior	1LCx-2R
3337	TGA/VSD/LVOTO	Perimembranous subpulmonary	ALM; posteromedian muscle; conus	ASO with VSD closure and resection of obstructive muscle tissue	–	n.a.	–	Side by side	1RL- 2LCx
3570	TGA/VSD/LVOTO	Perimembranous subpulmonary	Anteroseptal twist; hypertrophy of the LV anterior wall	ASO with VSD closure and resection of obstructive muscle tissue	–	n.a.	–	Aorta right anterior	1LCx-2R
6134	TGA/VSD/LVOTO	Perimembranous subpulmonary	Anterior mitral valve rotation and malattachment of the anterior papillary muscle; incomplete cleft anterior mitral leaflet	Senning with VSD closure and residual LVOTO	Univentricular pathway	n.a.	Both Nikaidoh and REV/Rastelli impossible as the LVOTO cannot be relieved without damage to the mitral valve	Aorta right anterior	1LCx-2R
6198	TGA/VSD/LVOTO	Perimembranous subpulmonary	Bicuspid obstructive pulmonary valve; anterior mitral malattachment	Nikaidoh	REV/Rastelli	?	REV/Rastelli will result in a small RV	Aorta right anterior	1L-2CxR
5554	TGA/VSD/LVOTO	Perimembranous subpulmonary	Bicuspid pulmonary valve; fibrous ridge; anterior mitral malattachment	Nikaidoh	–	n.a.	REV/Rastelli impossible due to mitral malattachment on outlet septum	Aorta right anterior	1LCx-2R
3646	TGA/DORV/LVOTO (Taussig–Bing)	Perimembranous subpulmonary	Accessory mitral valve tissue	ASO with VSD closure and resection of accessory tissue	–	n.a.	–	Side by side	1R-2LCx

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Hazekamp et al, 2007

Table 2
Anatomical characteristics and suggestions for surgical repair in specimens with TGA/VSD and LVOTO

Number	Basic anomaly	VSD	LVOTO	Best procedure	Alternative procedure	VSD enlargement necessary with Rastelli?	Comment	Position great arteries	Coronary anatomy
3268	TGA/VSD/LVOTO	Muscular subpulmonary	Aneurysmatic tissue of membranous septum	ASO with VSD closure and resection of aneurysmatic tissue	–	n.a.	–	Aorta right anterior	2LCxR (single coronary ostium)
3371	TGA/VSD/LVOTO	Muscular subpulmonary	Bicuspid dysplastic pulmonary valve; malalignment outlet septum	ASO with VSD closure and resection of outlet septum; shaving of the pulmonary valve	–	n.a.	–	Aorta right anterior	1LCx-2R
3856	TGA/VSD/LVOTO	Muscular subpulmonary	Pulmonary annulus hypoplasia; obstructive conus	Nikaidoh	REV/Rastelli	Yes	REV/Rastelli will result in a small RV	Aorta right anterior	1L-2LCxR
4538	TGA/DORV/LVOTO (Taussig–Bing)	Muscular subpulmonary	Fibrous ridge; ALM	ASO with VSD closure and resection of fibromuscular tissue	–	n.a.	–	Side by side	1RL-2LCx
4731	TGA/VSD/LVOTO	Doubly committed	Unicommissural pulmonary valve; malalignment outlet septum	Nikaidoh	REV/Rastelli	Yes	REV/Rastelli will result in a small RV	Aorta left anterior (dextrocardia)	1LCx-2R (quadricuspid aortic valve)
4402	TGA/VSD/LVOTO	Doubly committed	Bicuspid pulmonary valve; annular hypoplasia	Nikaidoh	REV/Rastelli	No	Both Nikaidoh and REV/Rastelli possible	Aorta right anterior	1LCx-2R
6679	TGA/VSD/LVOTO	Perimembranous subaortic anterior	Obstructive conal tunnel; bicuspid pulmonary valve	REV/Rastelli	Nikaidoh seems not possible	No	REV/Rastelli will give straight connection LV-Ao	Side by side	1RL-2Cx
5760	TGA/VSD/LVOTO	Perimembranous subaortic	Malalignment outlet septum	ASO with VSD closure and partial resection of outlet septum	–	n.a.	–	Side by side	1RL-2Cx

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ORIGINAL ARTICLE

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Current mid-term outcome with an integrated surgical strategy for correction of d-transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction†

Benjamin Bierbach[a], Claudia Arenz[a], Phillip Suchowerskyj[b], Sylvia Schroth[a], Jadwiga Blaschczok[a], Boulos Asfour[a], Martin Schneider[a] and Viktor Hraška[a,*]

[a] Department of Cardiothoracic Surgery, German Pediatric Heart Center, Sankt Augustin, Germany
[b] Department of Cardiology, German Pediatric Heart Center, Sankt Augustin, Germany

* Corresponding author. German Pediatric Heart Center, Arnold Janssen Strasse 29, 53757 Sankt Augustin, Germany. Tel: +49-2241249601; fax: +49-2241249602; e-mail: v.hraska@asklepios.com (V. Hraška).

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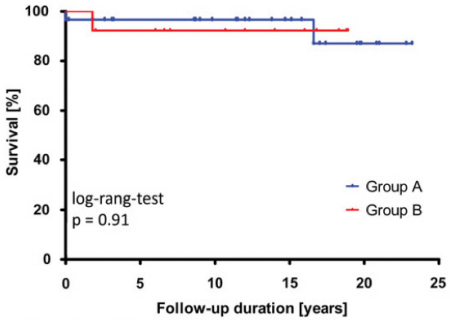
Surgical management protocol

In light of the frequent complications associated with placement of right ventricular to pulmonary artery conduits, we opted for repair on arterial level whenever the LVOTO could be addressed by resection of obstructive tissue and/or valvular repair. In cases with complex multilevel LVOTO, we performed intraventricular re-routing.

We aimed to allocate the appropriate surgical strategy for the individual patient based on the diagnosed intra- and extracardiac anatomy. The treatment algorithm was based on the anatomical substrate of the LVOTO, size, haemodynamic characteristic and position of the most relevant VSD, AV valve anatomy and the coronary artery pattern (suitability for coronary transfer or root translocation).

Decision-making algorithm:

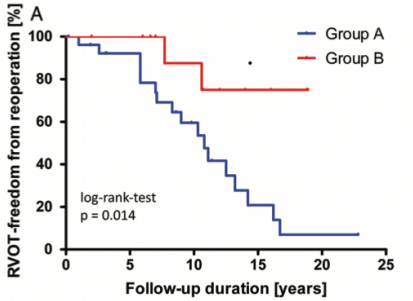
- transoesophageal echocardiography in theatre,
- evaluation of the coronary anatomy after sternotomy,
- evaluation of the size and the position of the VSD through the tricuspid valve,
- transection of both great arteries,
- evaluation of the RVOT and the VSD through the aortic valve,
- evaluation of the LVOTO through the PV,
- if ASO is contemplated, the PV repair is performed first,
- LVOTO is resected through the PV,
- final evaluation of the LVOT through PV (−2*z* measured by Hegar dilators are accepted).



Patients at risk:

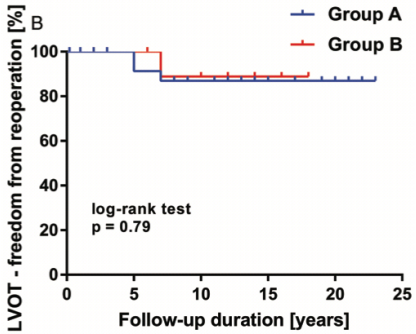
Group A	25	21	13	4
Group B	12	9	6	3

Figure 1: Mortality after intraventricular rerouting (Group A) and repair on arterial level (Group B).



Patients at risk:

Group A	20	10	4
Group B	12	8	3



Patients at risk:

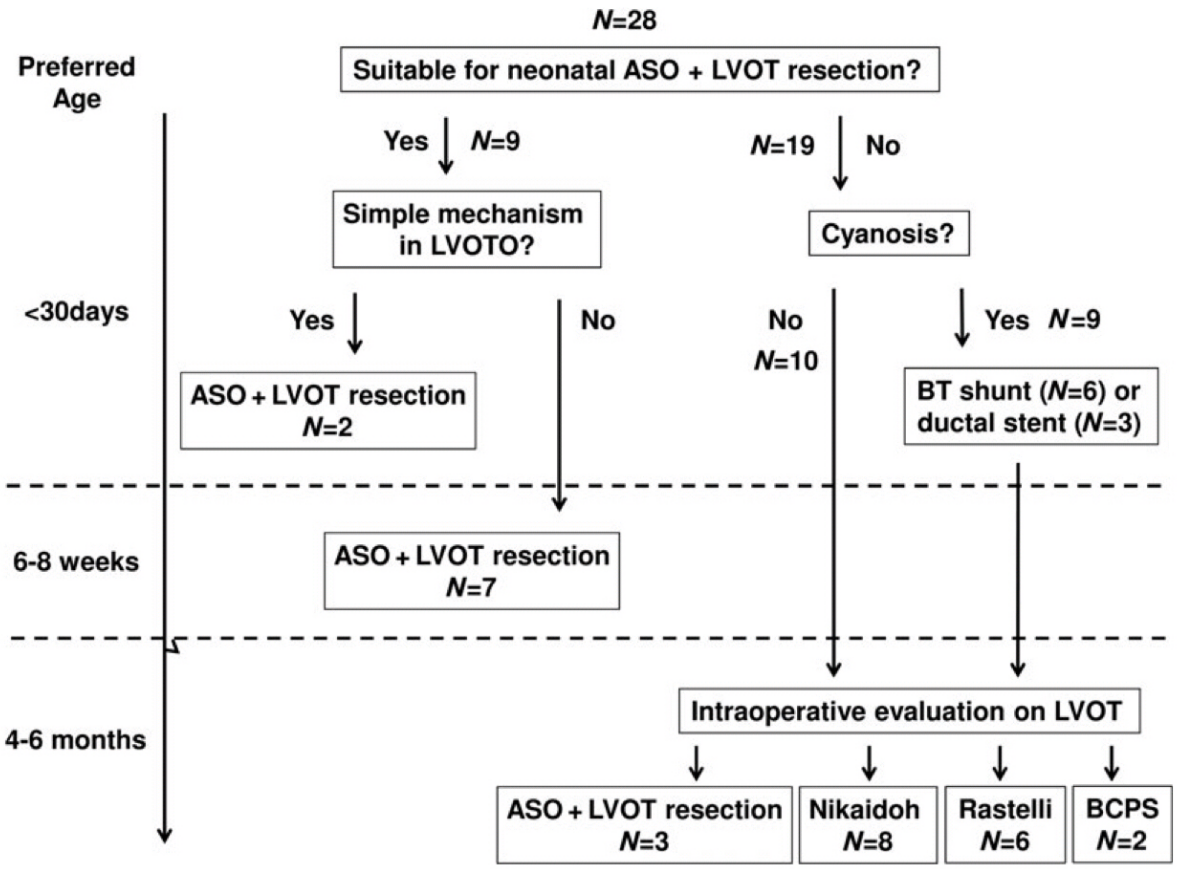
Group A	**30**	**24**	**12**	**4**
Group B	**12**	**7**	**5**	

European Journal of Cardio-Thoracic Surgery 44 (2013) 1085–1094
doi:10.1093/ejcts/ezt283 Advance Access publication 25 June 2013

ORIGINAL ARTICLE

Anatomical factors determining surgical decision-making in patients with transposition of the great arteries with left ventricular outflow tract obstruction†

Osami Honjo‡, Yasuhiro Kotani‡, Tara Bharucha, Luc Mertens, Christopher A. Caldarone, Andrew N. Redington and Glen Van Arsdell*



Outcomes After Anatomic Repair for D-Transposition of the Great Arteries With Left Ventricular Outflow Tract Obstruction

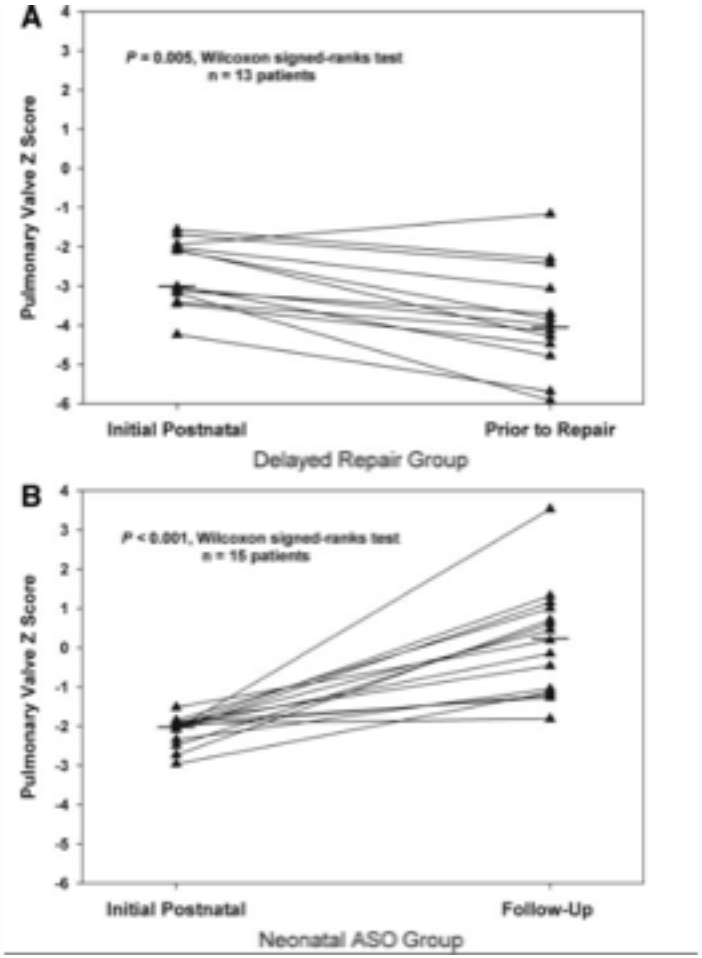
Sitaram M. Emani, MD; Rebecca Beroukhim, MD; David Zurakowski, PhD; Frank A. Pigula, MD; John E. Mayer, MD; Pedro J. del Nido, MD; Tal Geva, MD; Emile A. Bacha, MD

Background—D-transposition of the great arteries (TGA) with left ventricular outflow tract obstruction (LVOTO) may be treated with arterial switch operation (ASO) with or without LVOT intervention, as well as non-ASO anatomic repairs, such as aortic translocation or Rastelli procedure. We evaluated midterm results of repair for TGA/LVOTO at our institution.

Methods and Results—Eighty-eight patients with TGA/LVOTO who underwent anatomic repair were retrospectively reviewed. LVOTO was defined as pulmonary valve (PV) z-score ≤ -2.0 or LVOT gradient ≥ 20 mm Hg in the presence of anatomic subvalvar stenosis. Risk factors for LVOT reintervention were determined by logistic regression. There was no hospital mortality and 1 late mortality. Patients undergoing Rastelli procedure were more likely to require surgical reintervention for LVOTO compared to the other groups ($P=0.015$). Patients undergoing ASO alone had a higher rate of late LVOT reintervention compared to those who had concomitant ASO/LVOT intervention ($P=$NS). In those undergoing Rastelli, a larger PV z-score was a predictor of LVOT reintervention ($P=0.012$). PV z-scores significantly decreased before repair in patients undergoing delayed repair ($P=0.005$); however, they increased significantly after neonatal ASO ($P<0.001$).

Conclusions—Patients with TGA/LVOTO who undergo Rastelli repair have a high rate of LVOT reintervention. Higher preoperative PV z-score is a risk factor for reintervention in this group. Patients with mild/moderate LVOTO undergoing ASO alone without LVOT intervention may have an increased risk of LVOT reintervention. In neonates who are candidates for ASO, delay of repair is associated with diminution in size of PV, which may subsequently reduce their suitability for ASO. (***Circulation***. **2009;120[suppl 1]:S53–S58.**)

Emani et al, 2009

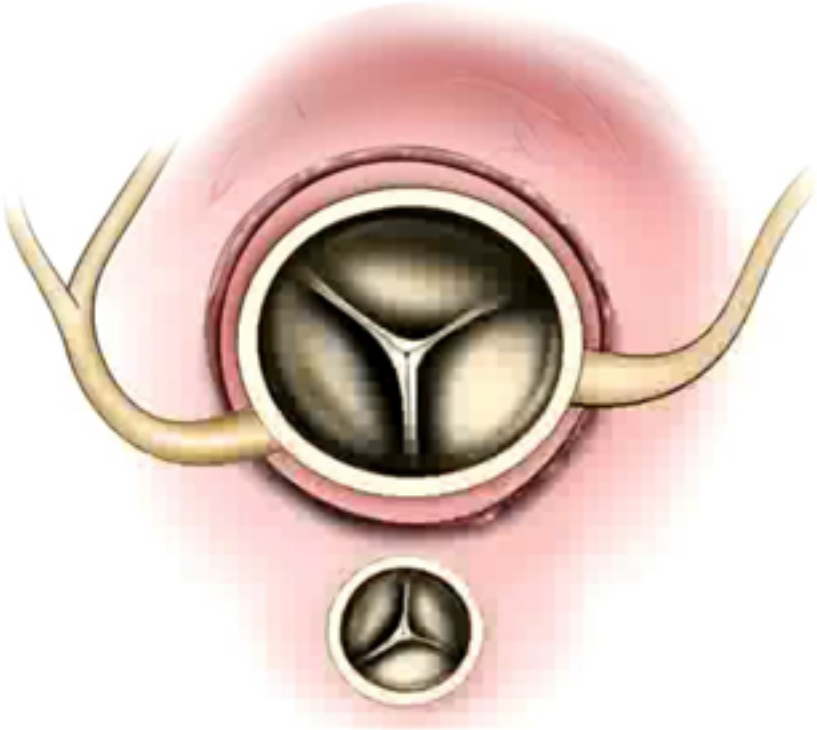


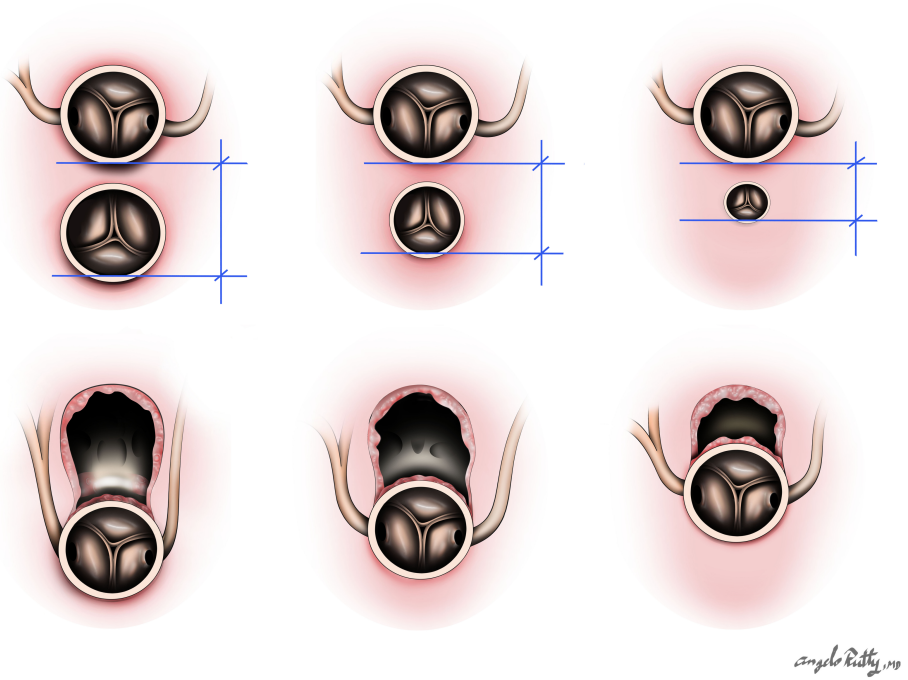
Children's Hospital of Pittsburgh of UPMC

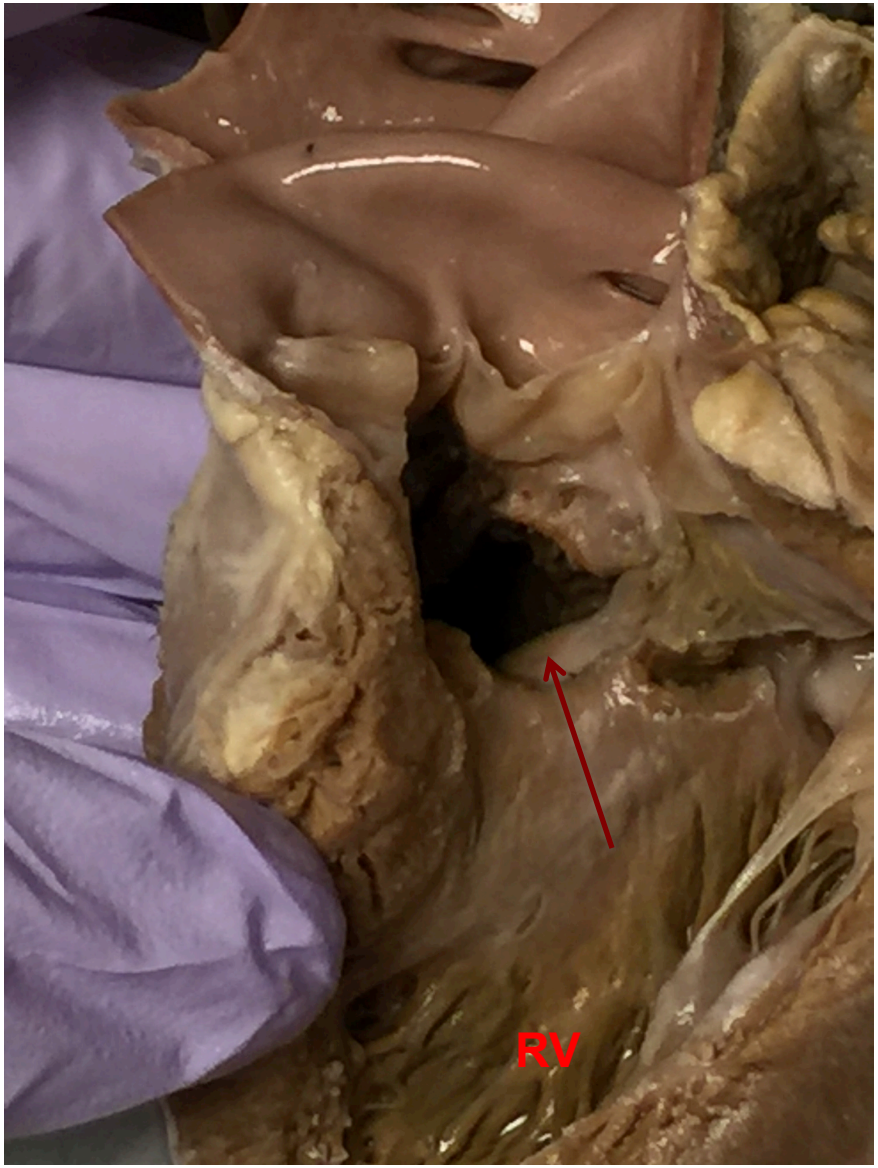
Emani et al, 2009

- **Most patients with PV z-scores of >3 and resectable LVOT obstruction should be considered candidates for an ASO/LVOT resection**
- **For PV annular z-score <3, unicommissural PV, or unresectable LVOTO, the aortic translocation is preferred.**
- **Palliation with a Blalock-Taussig shunt should be avoided in favor of neonatal repair if the patient is deemed a candidate for ASO.**

Nikaidoh or Rastelli?







doi:10.1510/icvts.2009.223982

INTERACTIVE CARDIOVASCULAR AND THORACIC SURGERY

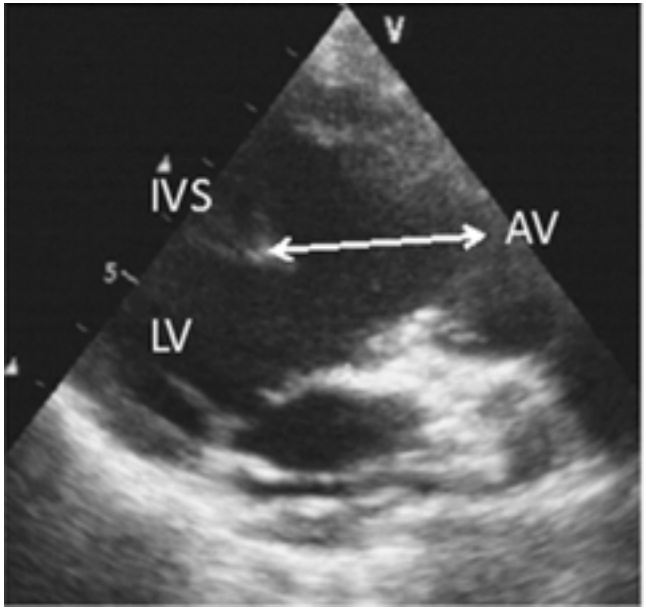
Interactive CardioVascular and Thoracic Surgery 10 (2010) 900–905

www.icvts.org

Institutional report - Congenital

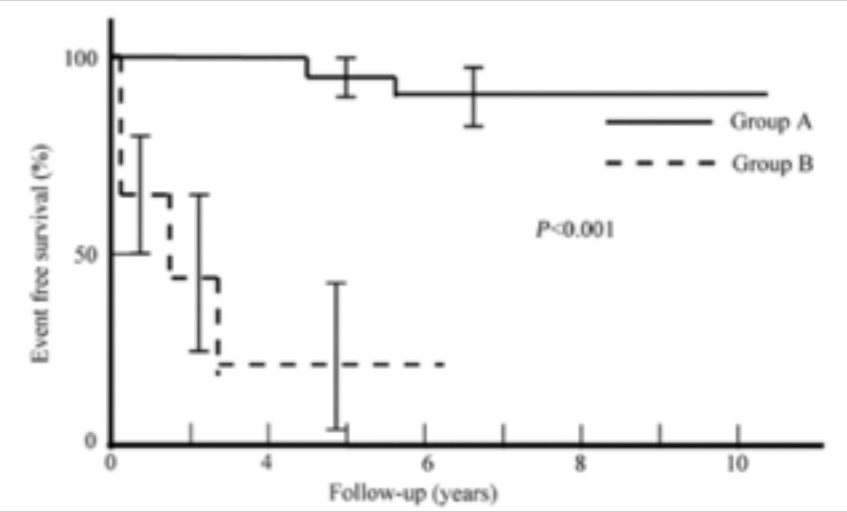
The impact of the length between the top of the interventricular septum and the aortic valve on the indications for a biventricular repair in patients with a transposition of the great arteries or a double outlet right ventricle☆

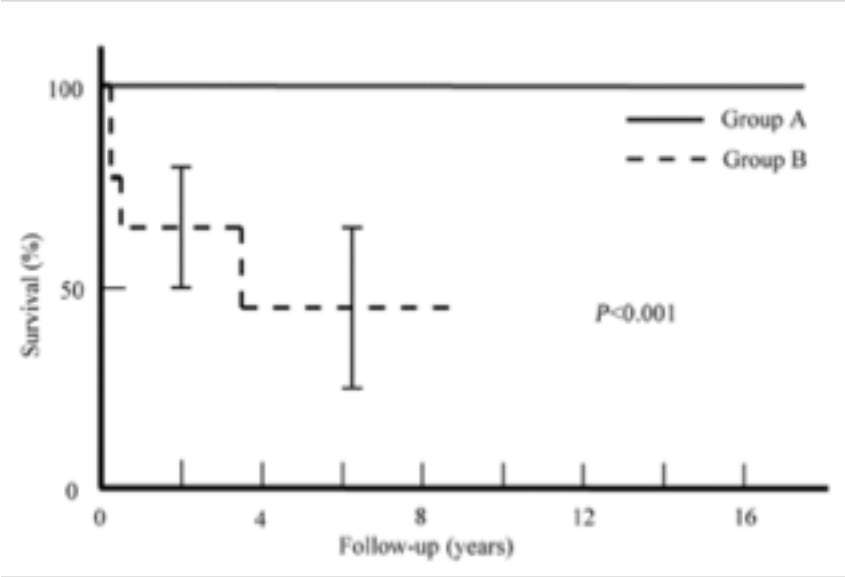
Yasuhiro Fujii[a,*], Yasuhiro Kotani[a], Masami Takagaki[a], Sadahiko Arai[a], Shingo Kasahara[a], Shin-ichi Otsuki[b], Shunji Sano[a]



Fijii et al, 2010

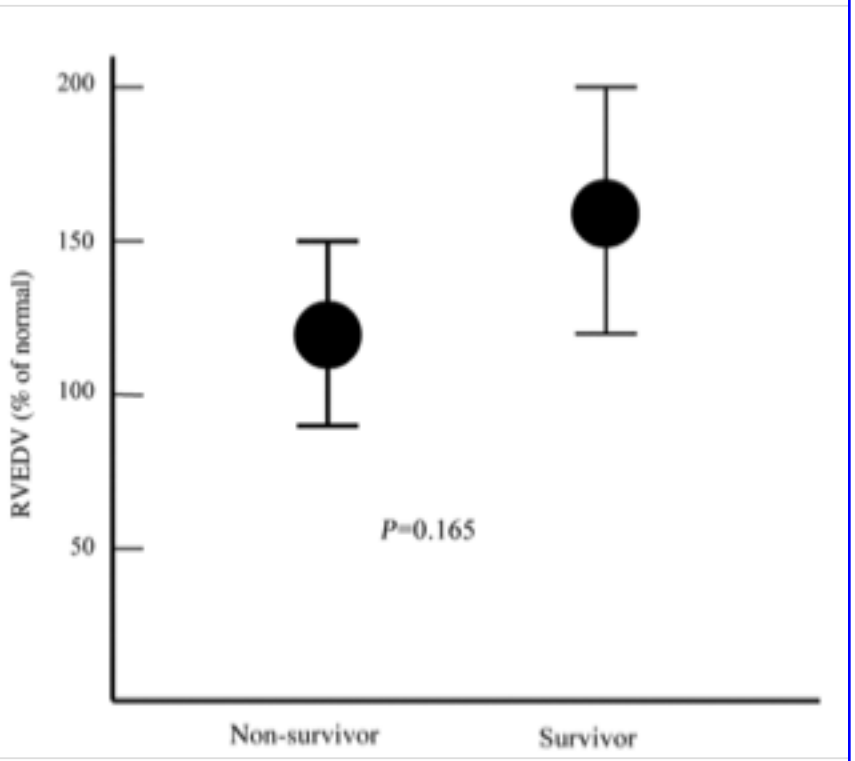
Group A; IVS–AV length of <80% of normal LVDd.
Group B; IVS–AV length of >80% of normal LVDd.





Children's Hospital of Pittsburgh of UPMC

Fijii et al, 2010

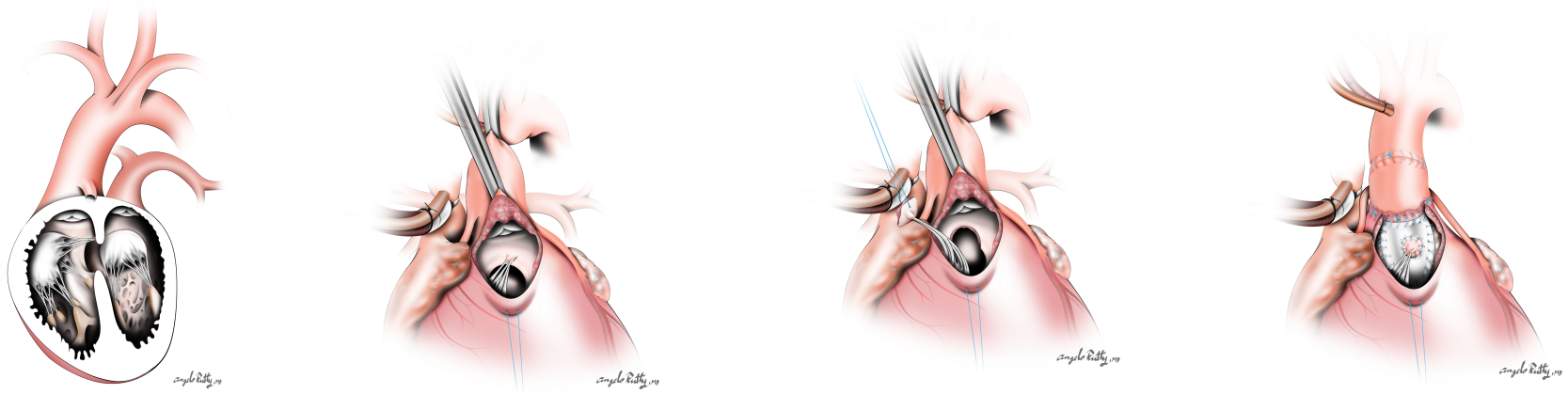


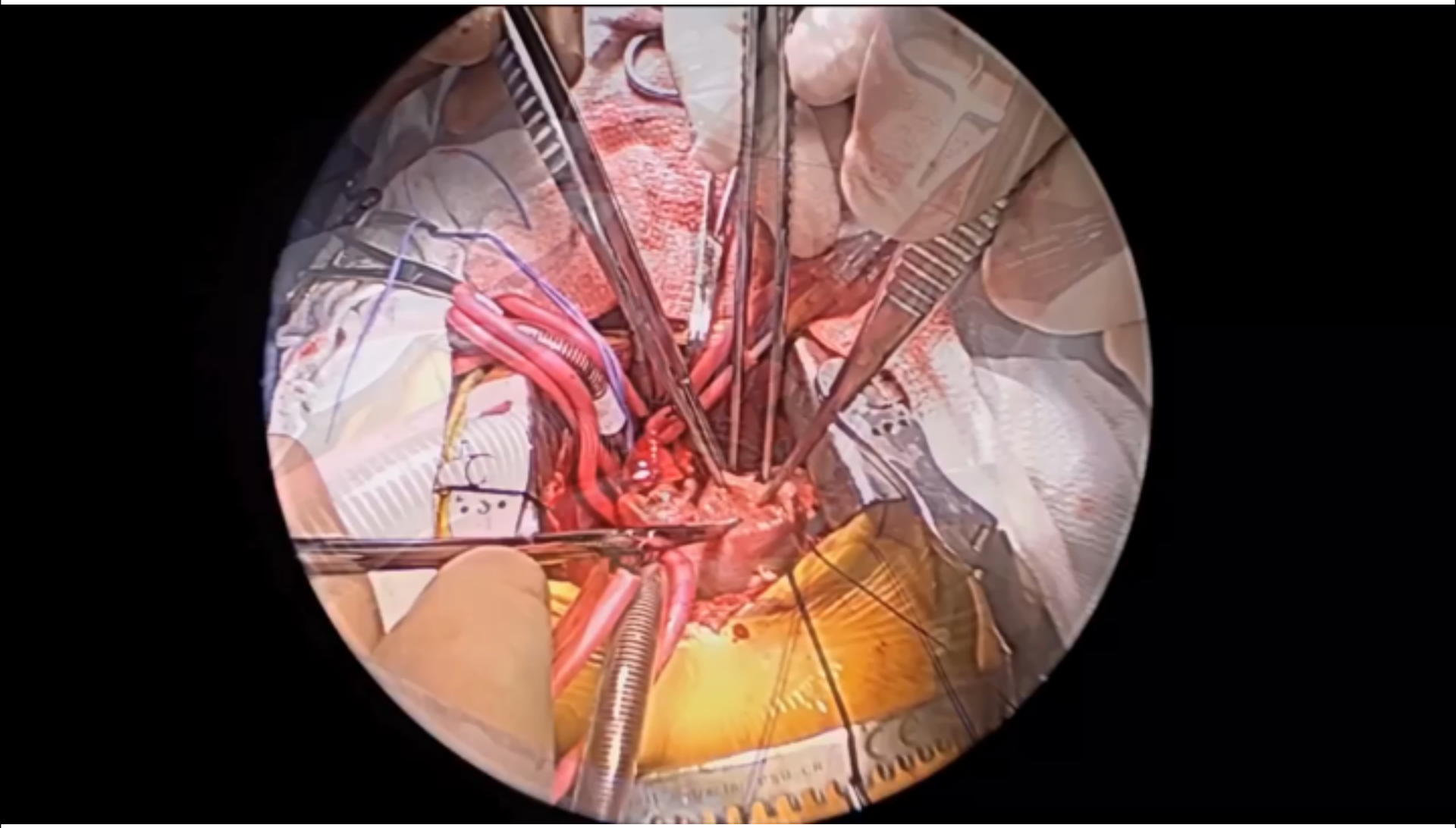
- **A large RV allows the placement of a sufficiently larger rerouting patch in the RV without impairing the function of the RV**

Fijii et al, 2010

- **In conclusion, *the IVS–AV length was found to be a significant risk factor for mortality* and postoperative cardiogenic events. However, *a larger RVEDV may somewhat compensate for the risk of mortality*. The IVS–AV length should be taken into consideration when selecting the optimal surgical procedures for these patients.**

Abnormal AVV attachments





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ELSEVIER

European Journal of Cardio-thoracic Surgery xxx (2006) xxx–xxx

EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

www.elsevier.com/locate/ejcts

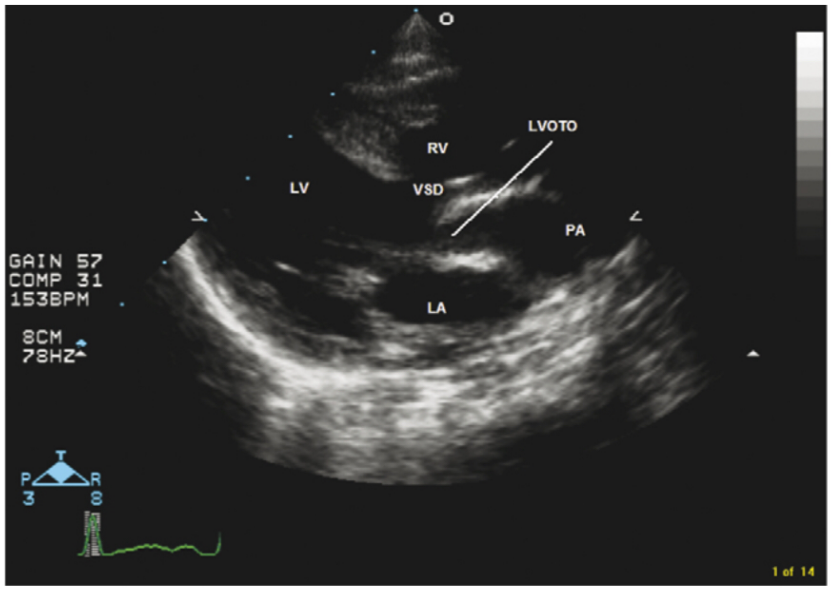
Case report

Aortic translocation for the management of transposition of the great arteries with a ventricular septal defect, pulmonary stenosis, and hypoplasia of the right ventricle

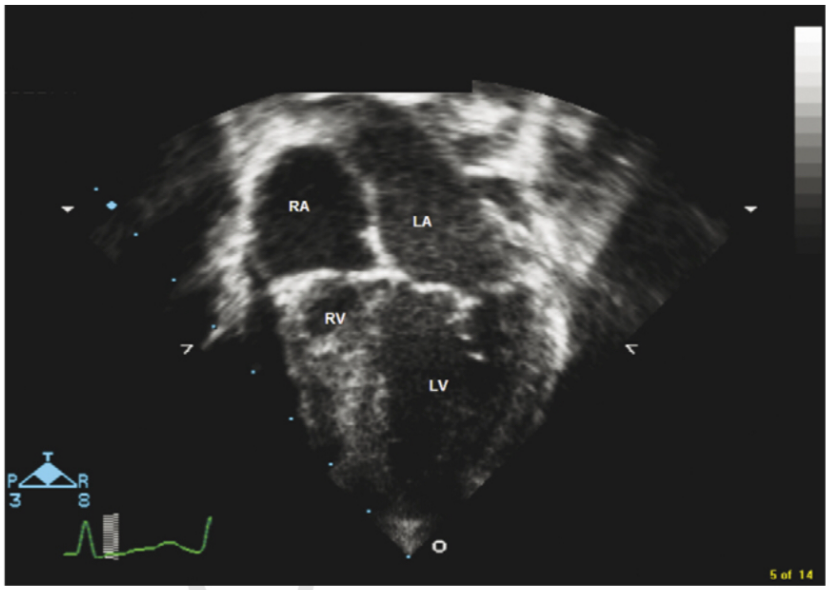
Victor O. Morell *, Peter A. Wearden

Section of Pediatric Cardiothoracic Surgery of the Heart, Lung and Esophageal Surgical Institute, University of Pittsburgh Medical School, Children's Hospital of Pittsburgh, Pittsburgh, PA, United States

Received 19 September 2006; received in revised form 13 November 2006; accepted 20 November 2006



(A)



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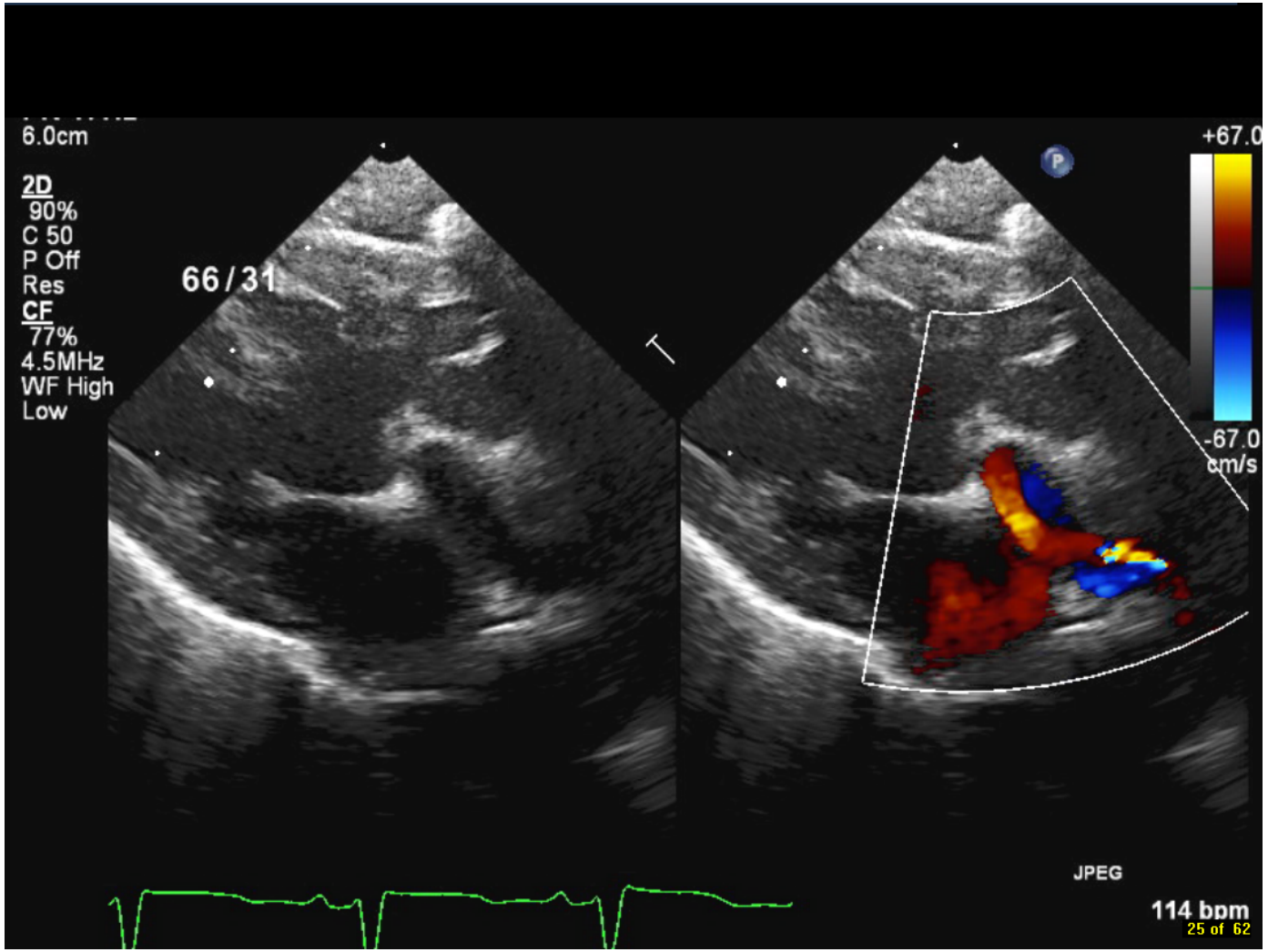

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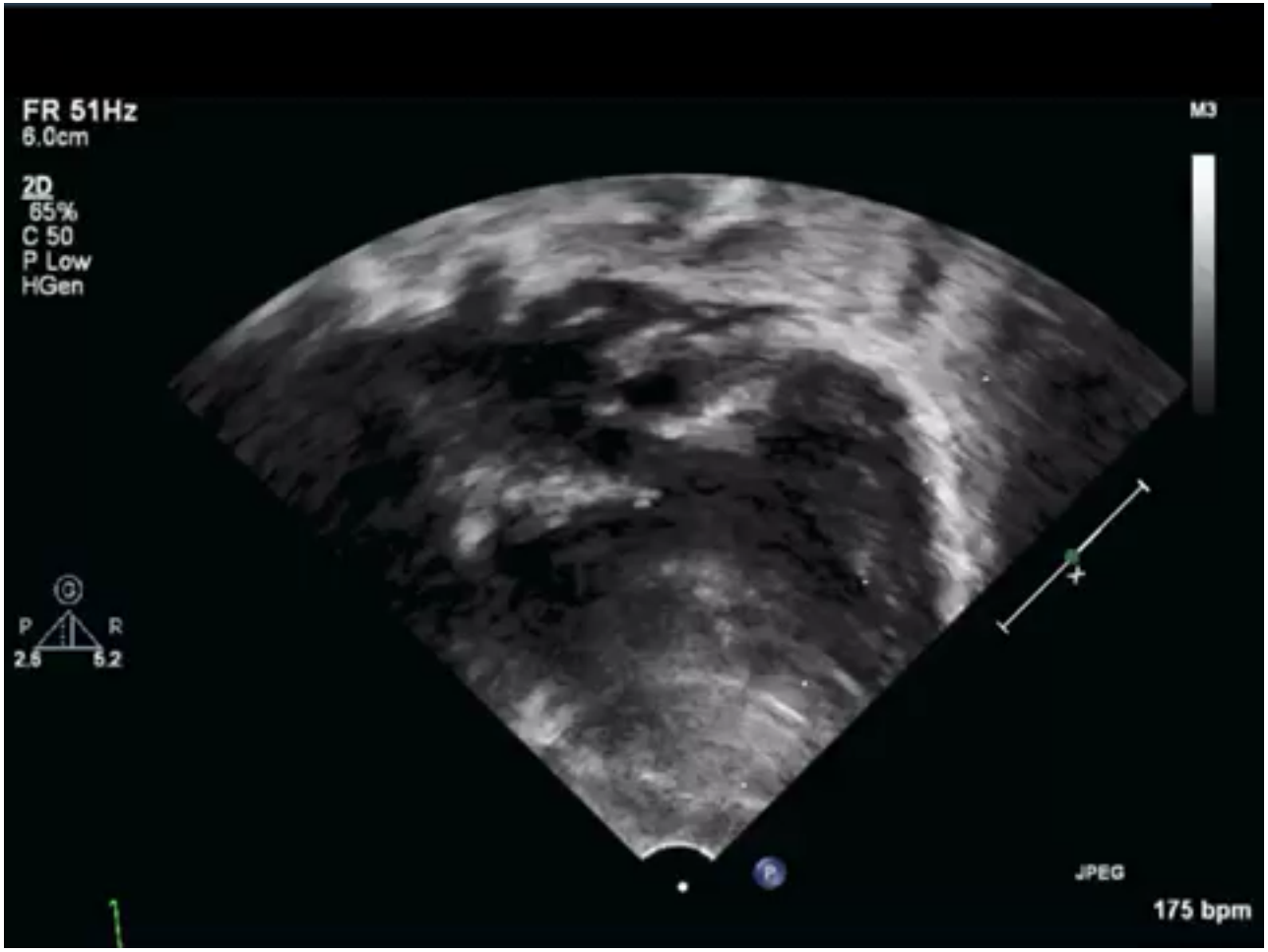

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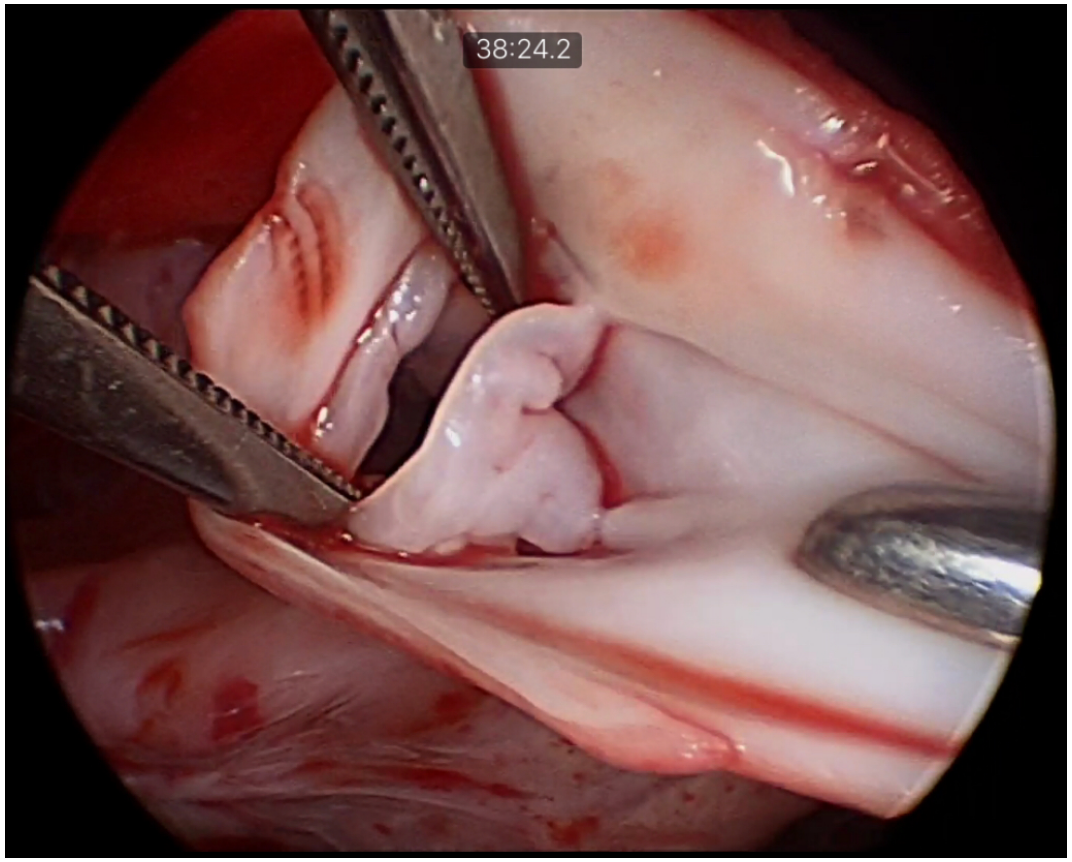

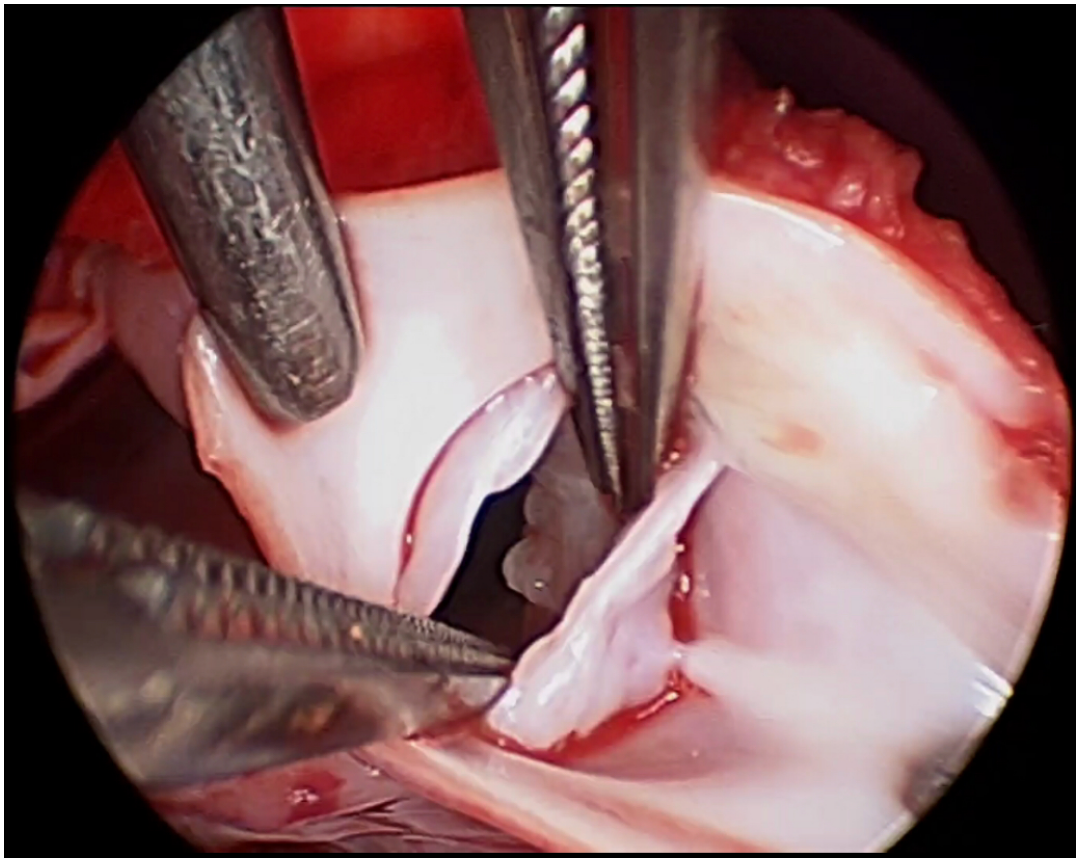

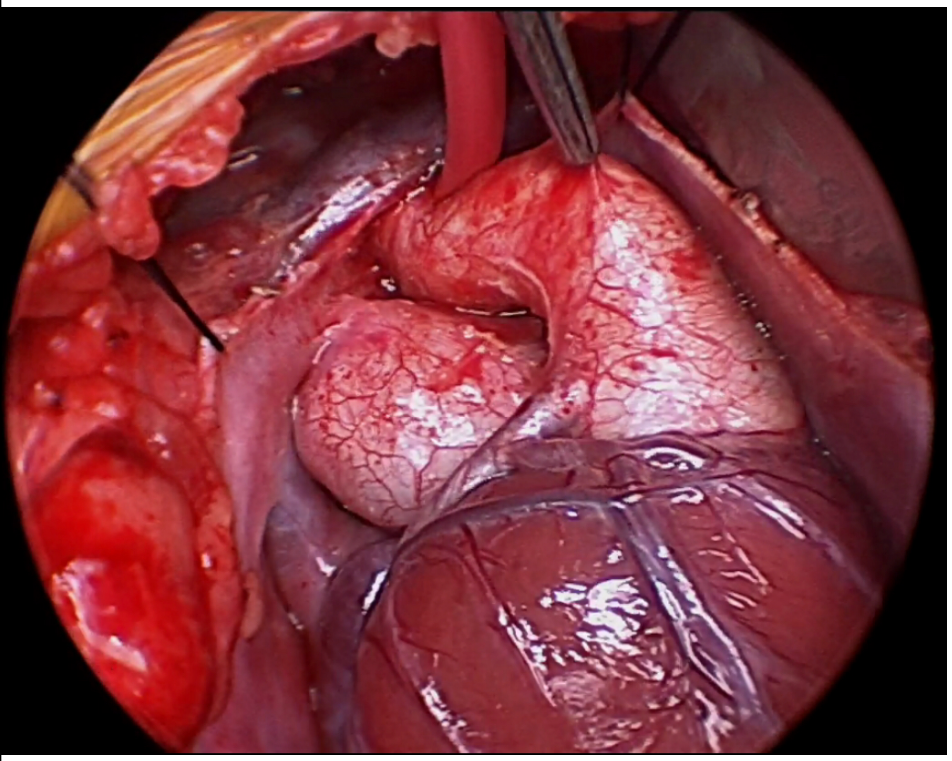

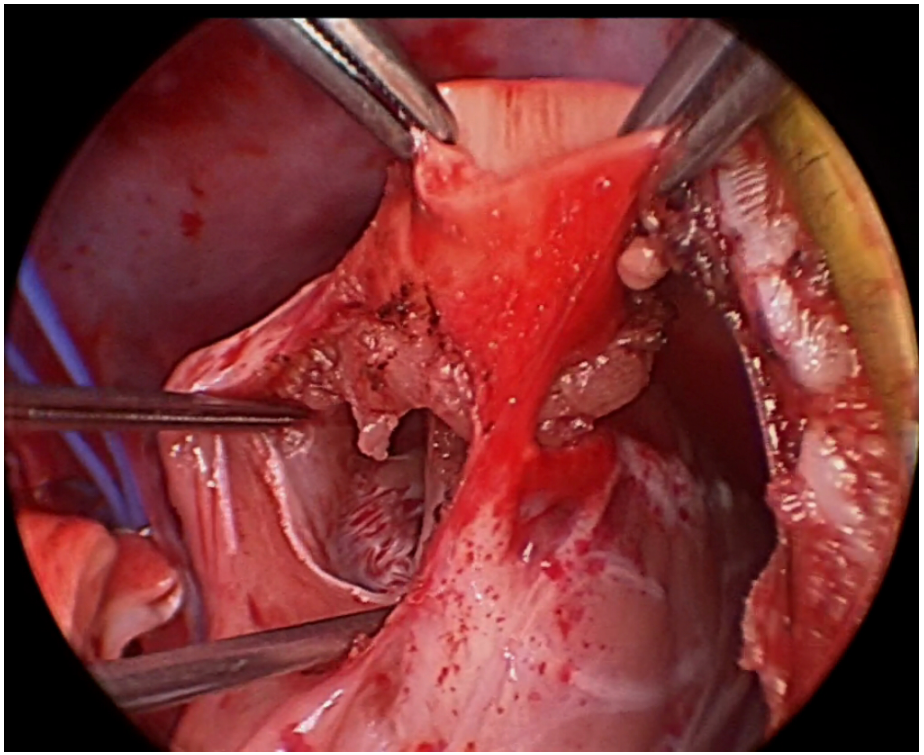

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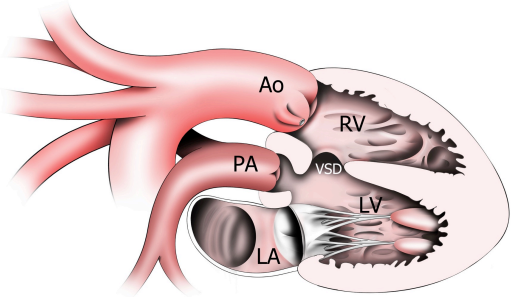
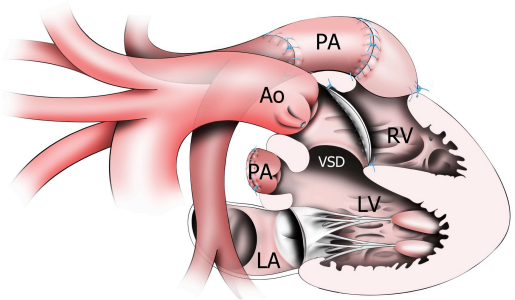
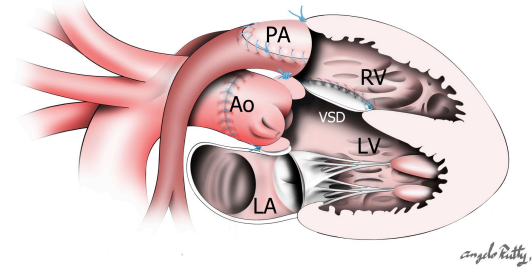
LVOT



TGA/VSD/PS



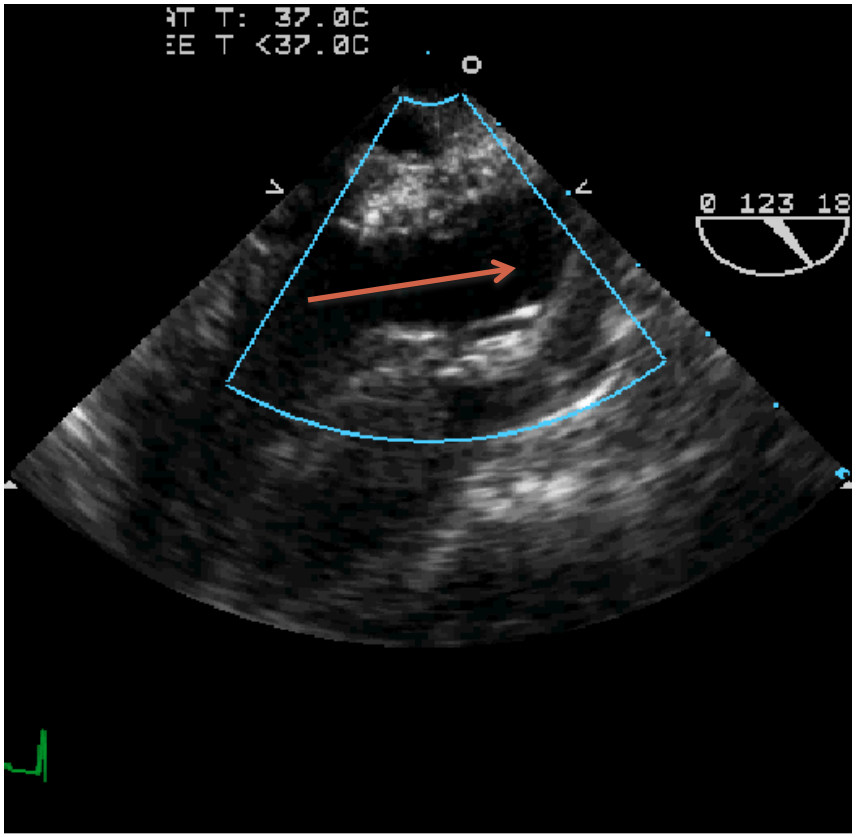
Rastelli



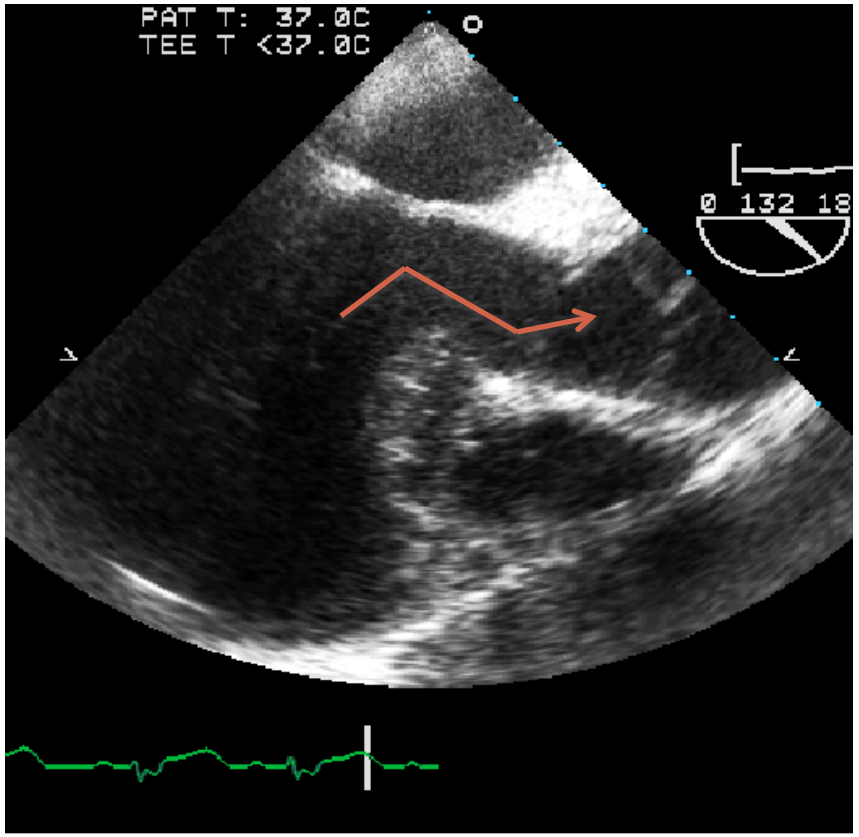
Nikaidoh

LVOT

Nikaidoh



Rastelli



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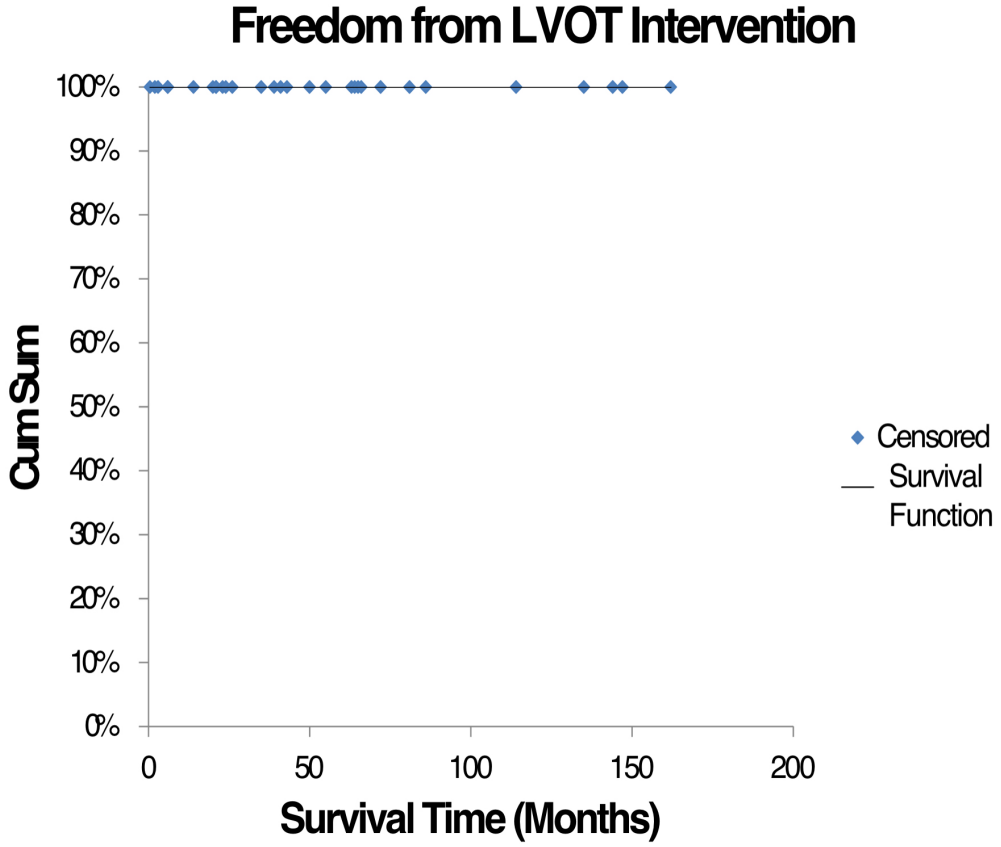
Strategy for biventricular outflow tract reconstruction: Rastelli, REV, or Nikaidoh procedure?

JTCVS 2008

Sheng-Shou Hu, MD, PhD, Zhi-Gang Liu, MD, PhD, Shou-Jun Li, MD, Xiang-dong Shen, MD, Xu Wang, MD, Jin-ping Liu, MD, Fu-Xia Yan, MD, Li-qing Wang, MD, and Yong-qing Li, MD

"Postoperative echocardiography ***demonstrated physiologic hemodynamics in the left ventricular outflow tract and normal heart function in the Nikaidoh*** group. ***Abnormal flow pattern in the left ventricular outflow tract was noted in both REV and Rastelli groups***. There were no late deaths or reoperations in any group during follow-up.



*Unpublished data

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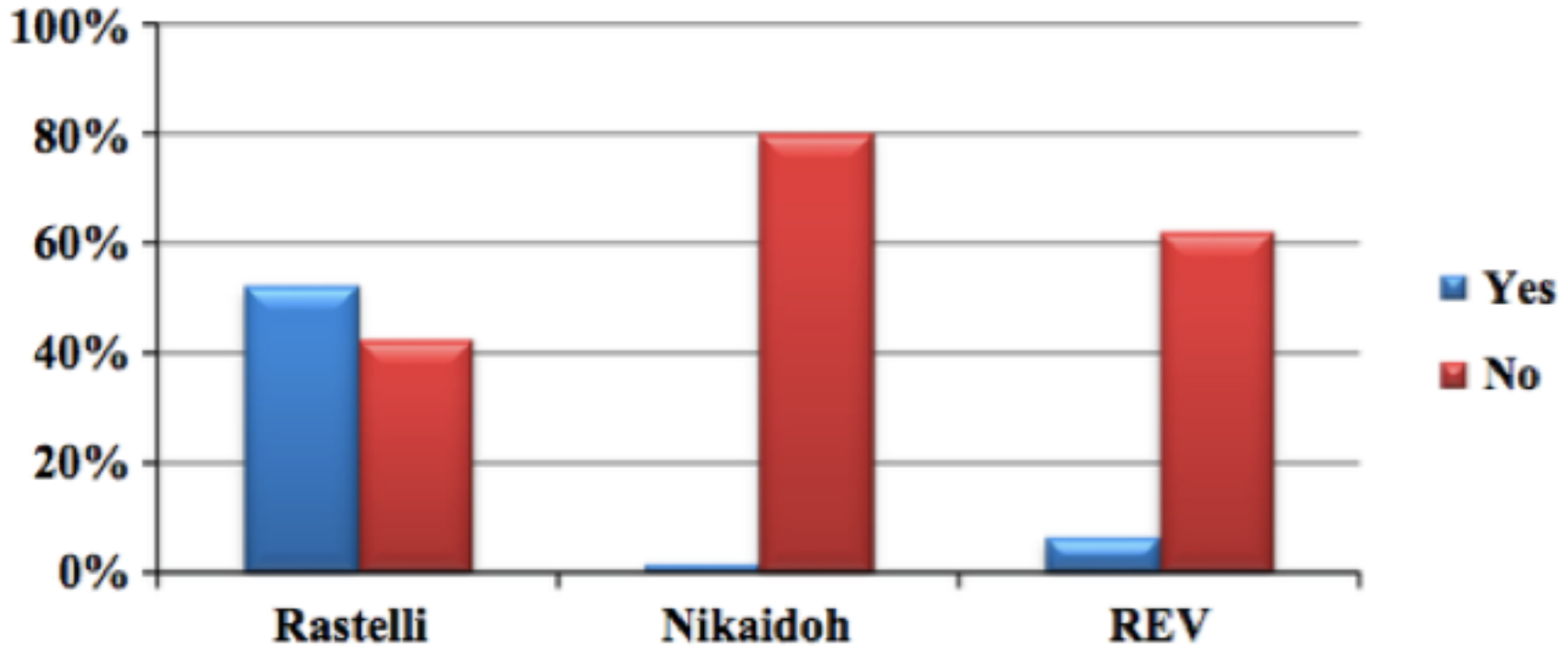
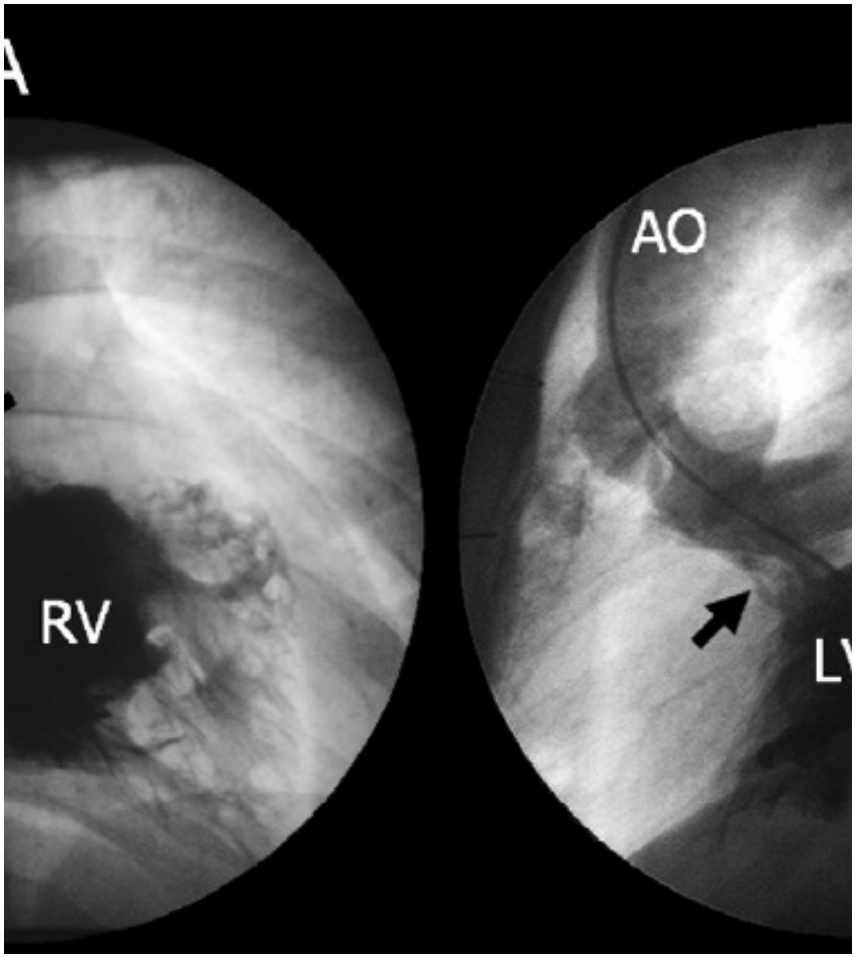
Have any of your patients developed recurrent LVOTO?



LVOTO



- “ Awareness of LVOTO has led us to frequently include VSD enlargement (resection of the anterosuperior margin) concomitant with the Rastelli procedure”

Kreutzer et al.,J Thorac Cardiovasc Surg 2000;120:211-23

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ATS 2009

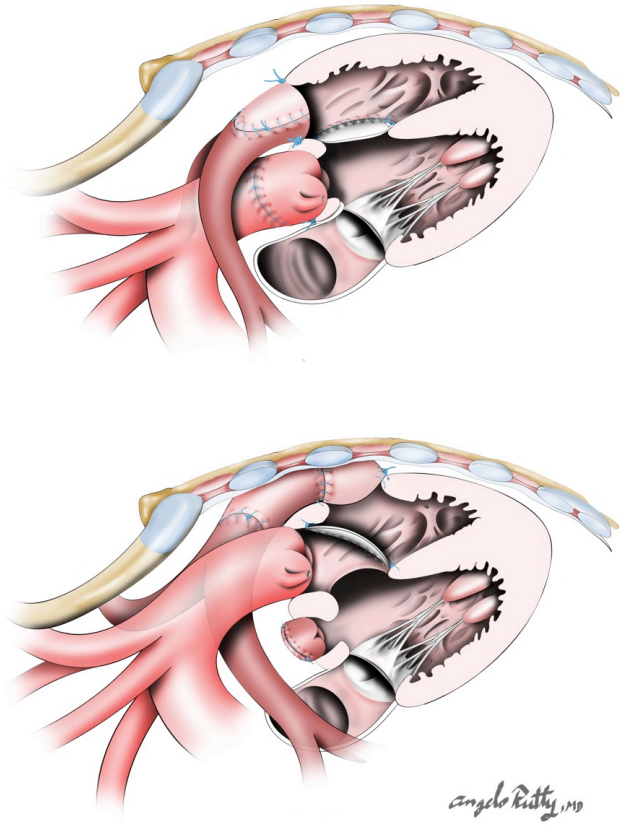
The Rastelli Procedure for Transposition of the Great Arteries: Resection of the Infundibular Septum Diminishes Recurrent Left Ventricular Outflow Tract Obstruction Risk

Bahaaldin Alsoufi, MD, Abid Awan, MD, Ahmad Al-Omrani, MD, Mamdouh Al-Ahmadi, MD, Charles C. Canver, MD, Ziad Bulbul, MD, Avedis Kalloghlian, MD, and Zohair Al-Halees, MD

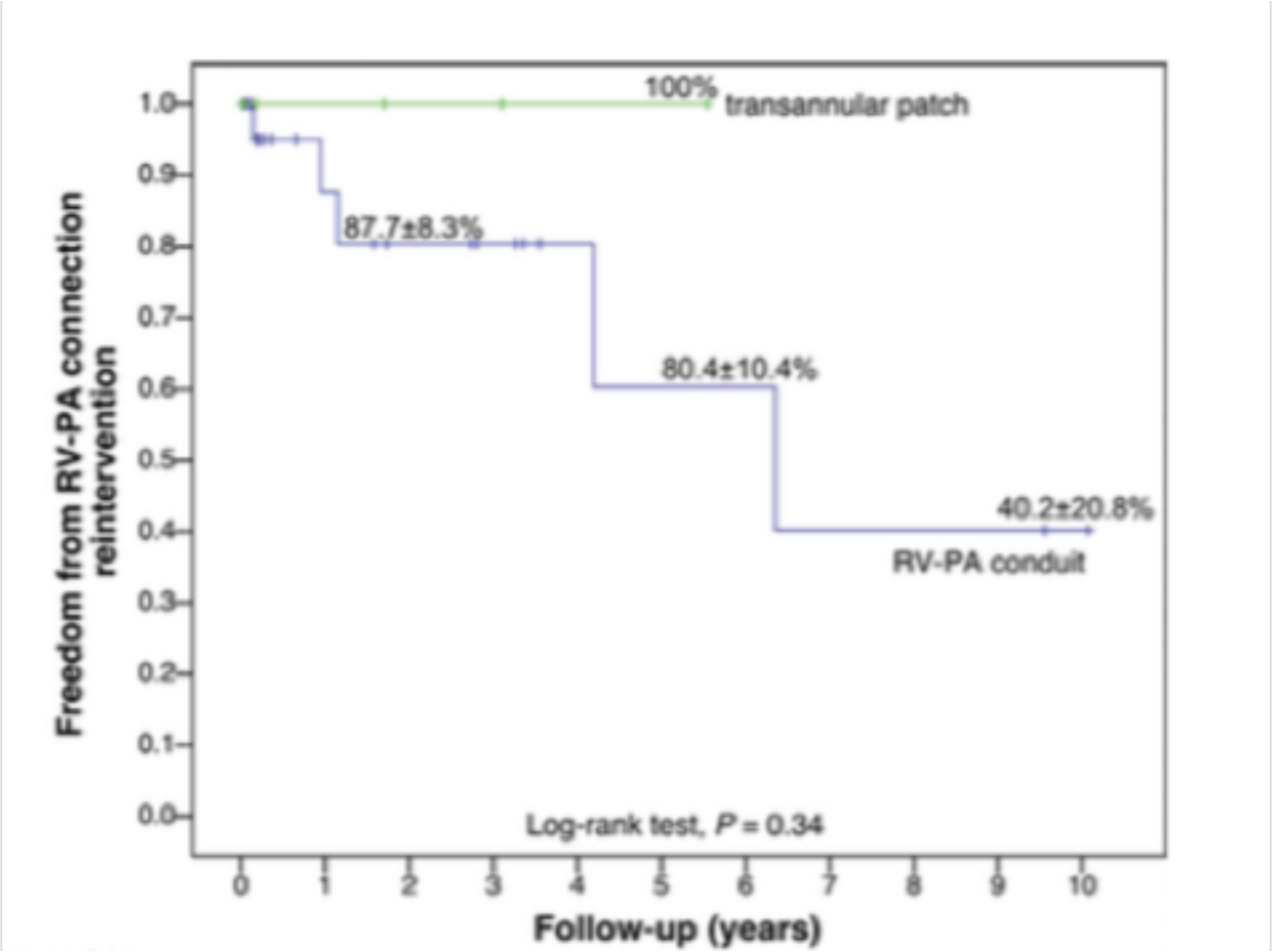
King Faisal Heart Institute, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia

Children's Hospital of Pittsburgh of UPMC

RVOT



Raju et al, 2015
Nikaidoh



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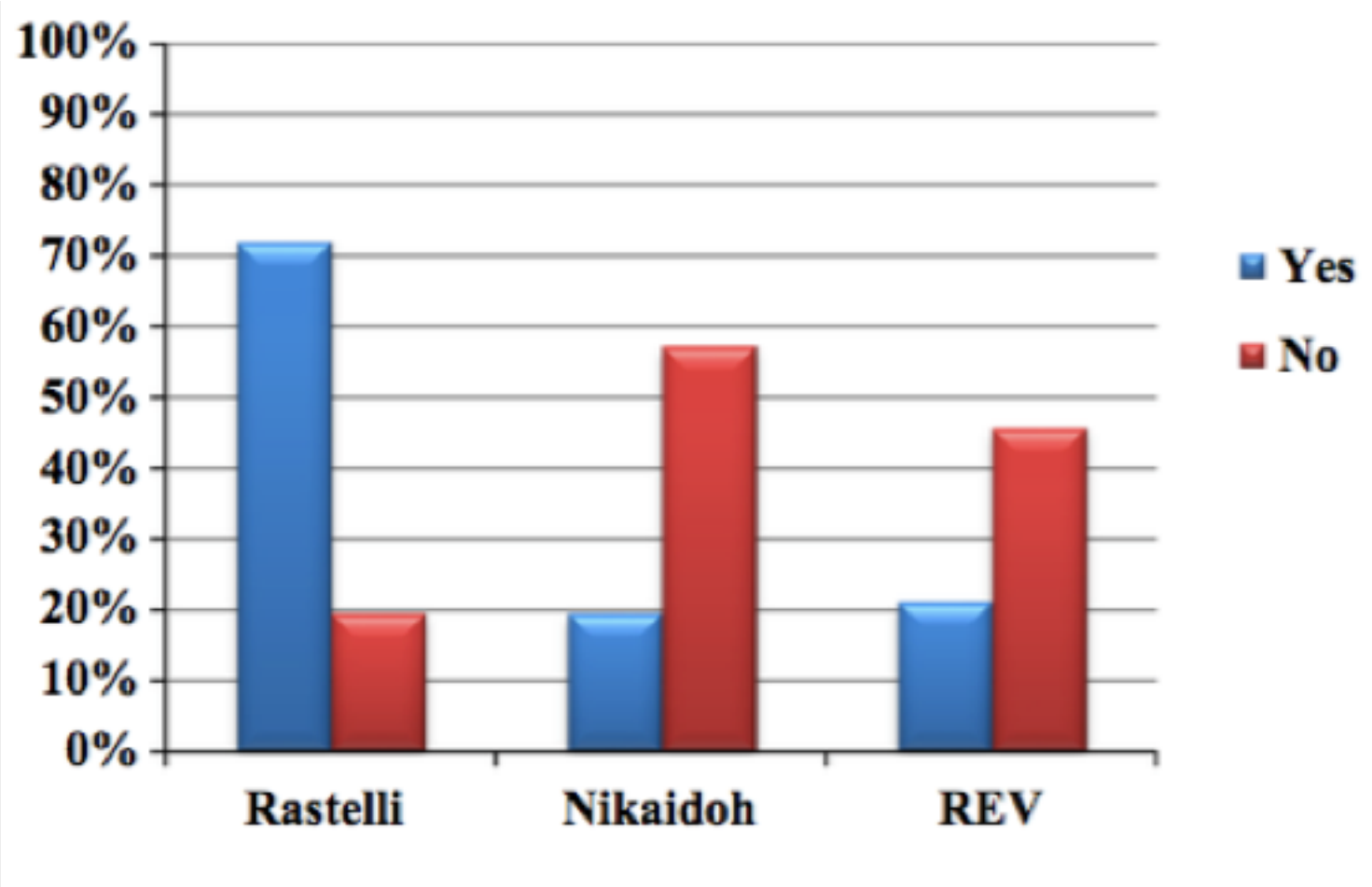
Have any of your patients developed recurrent RVOTO?



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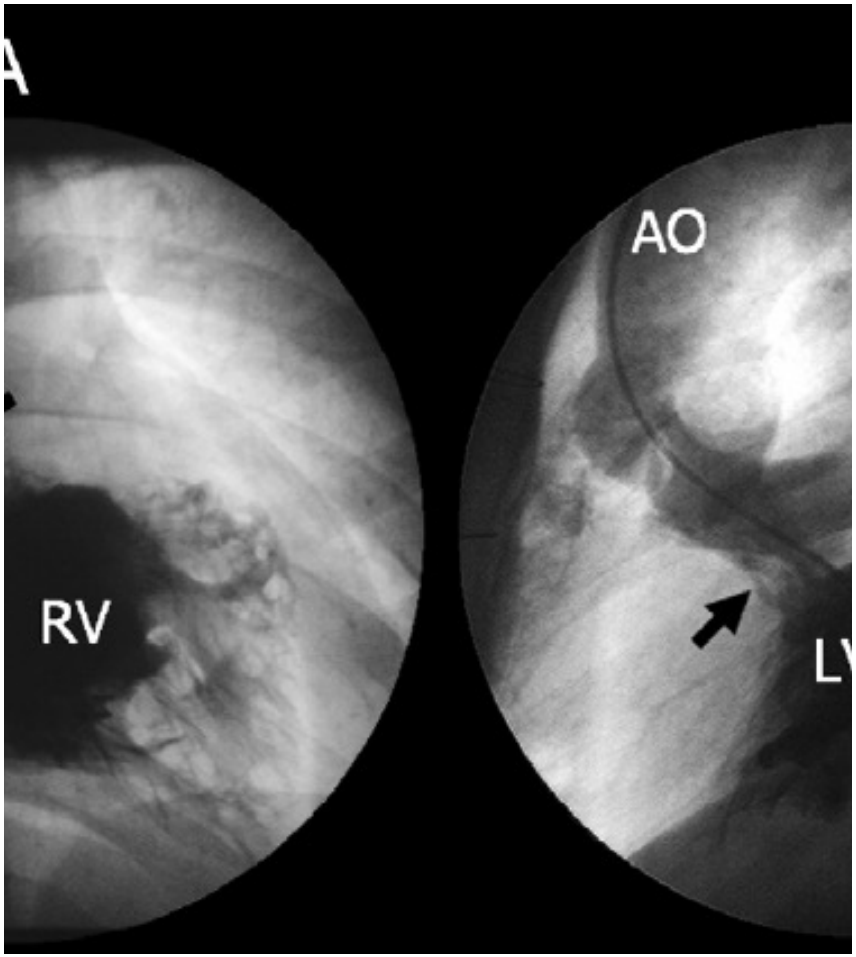
RVOTO



- **The only technique of RVOT reconstruction with good late results was <u>direct anastomosis</u>**

Kreutzer et al.,J Thorac Cardiovasc Surg 2000;120:211-23

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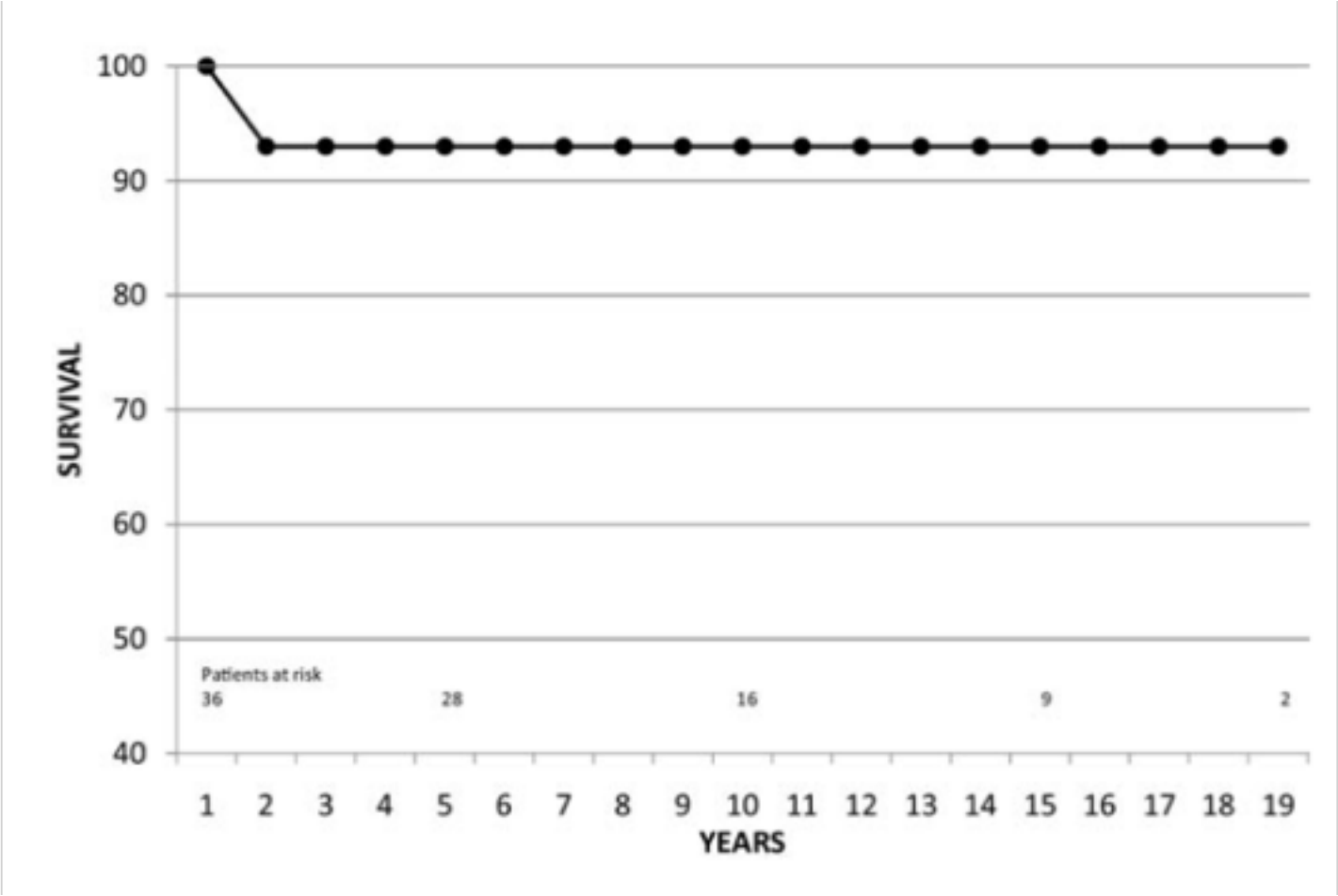
Brown et al, 2011
Rastelli



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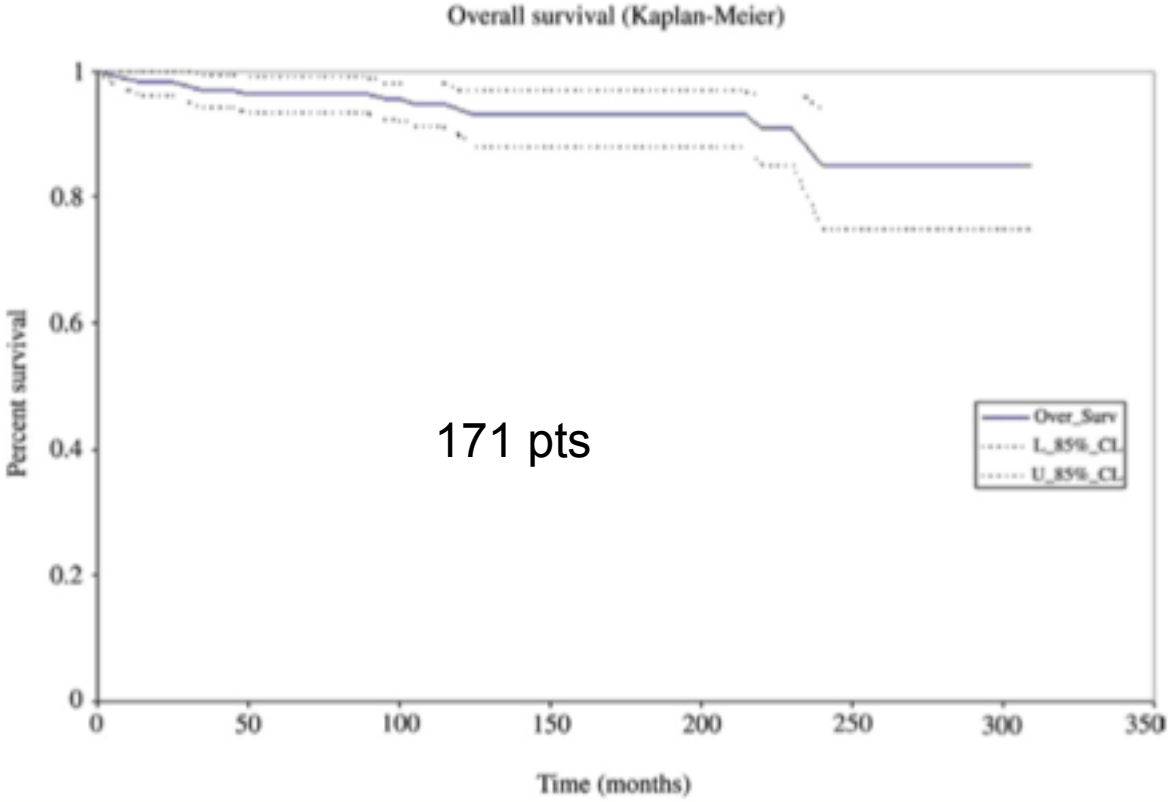
doi:10.1510/mmcts.2007.003046

MULTIMEDIA MANUAL OF CARDIOTHORACIC SURGERY
MMCTS

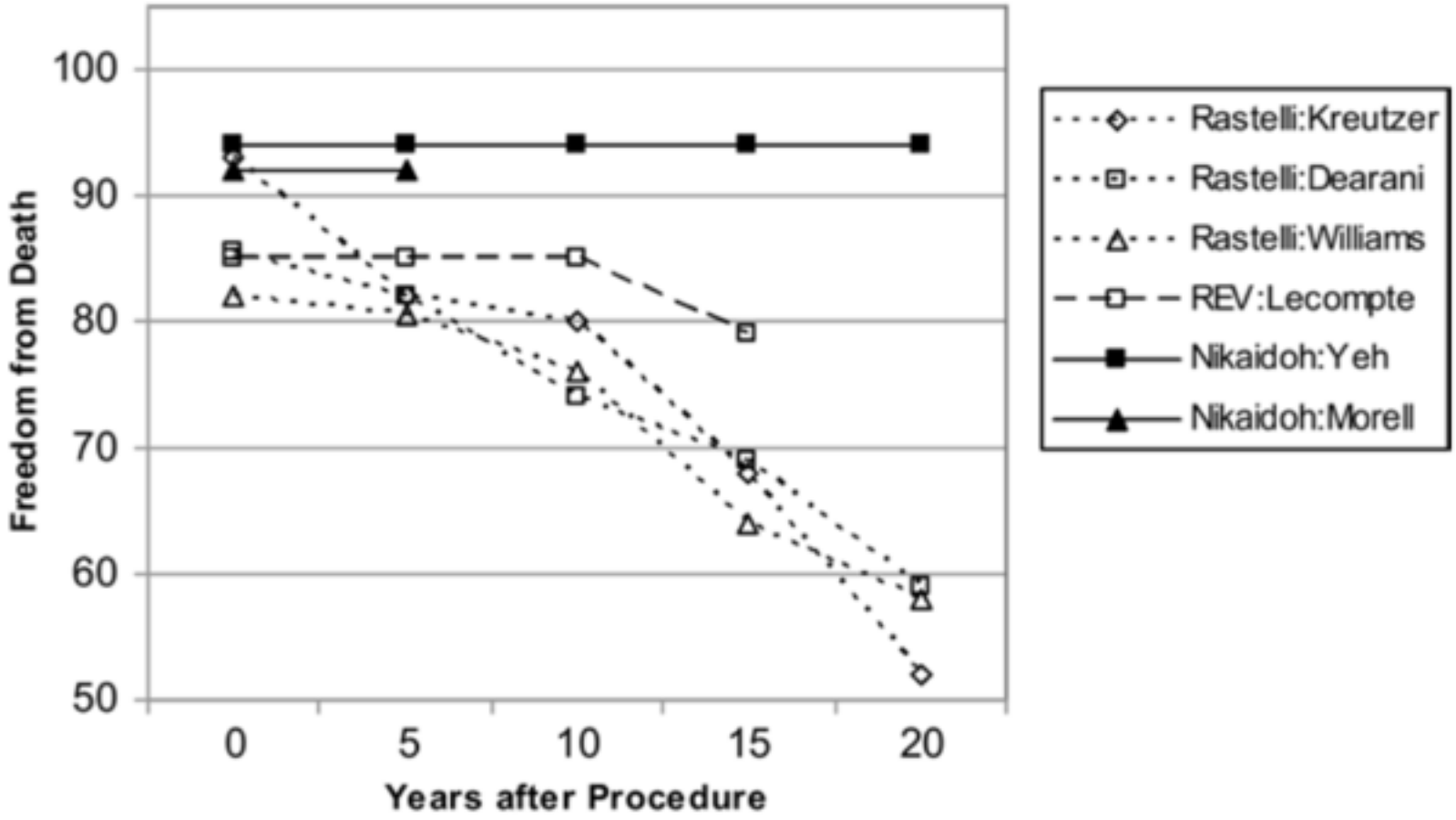
Surgery for malposition of the great arteries: the REV procedure

Duccio Di Carlo[a,*], Yves Lecompte[b], Biagio Tomasco[c], Laurence Cohen[b], Pascal Vouhé[d]



Graph 1. Overall survival (Kaplan–Meier) after the REV operation time = months postoperatively.

Yeh, et al 2007



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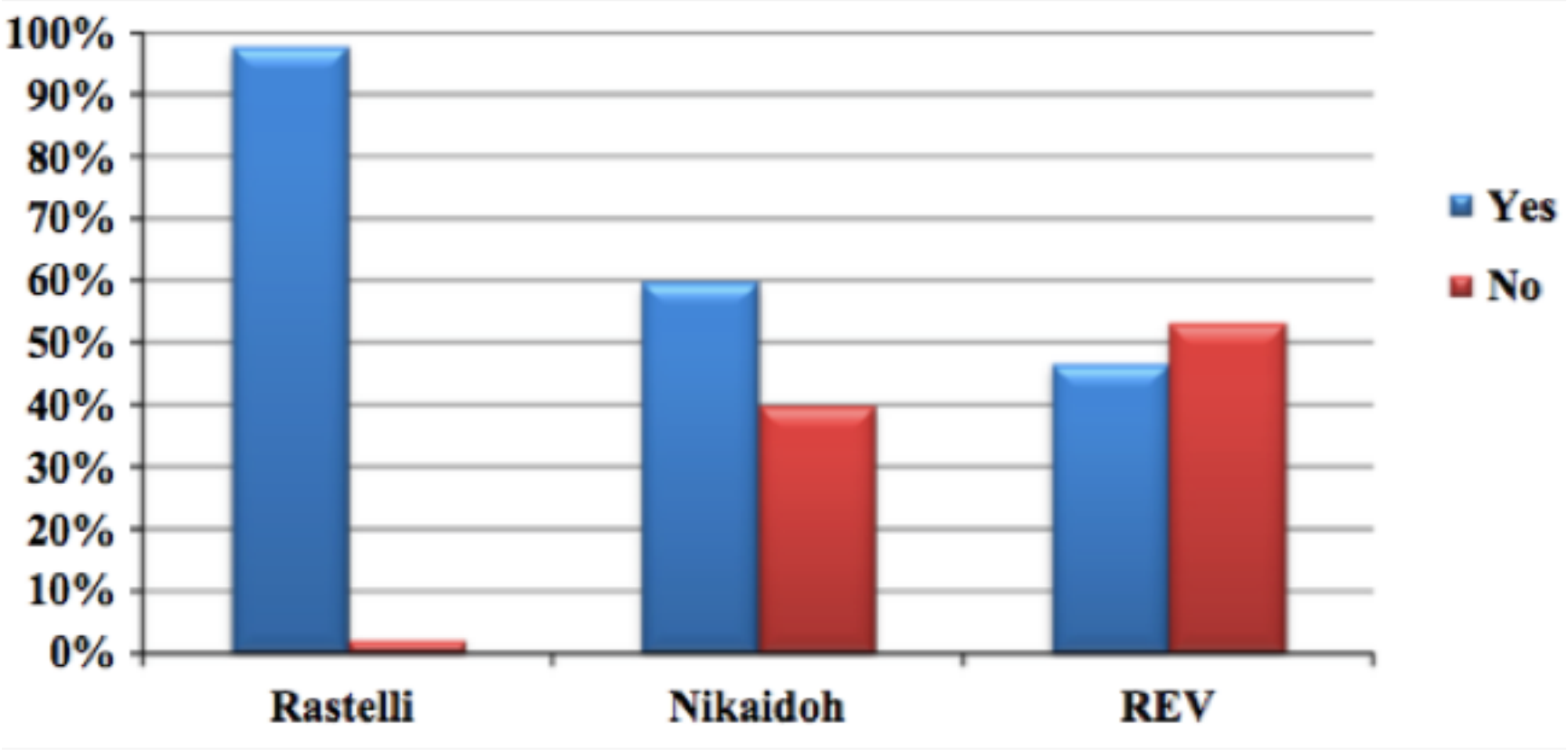
As a staff surgeon
have you done any Rastelli, Nikaidoh, or REV?



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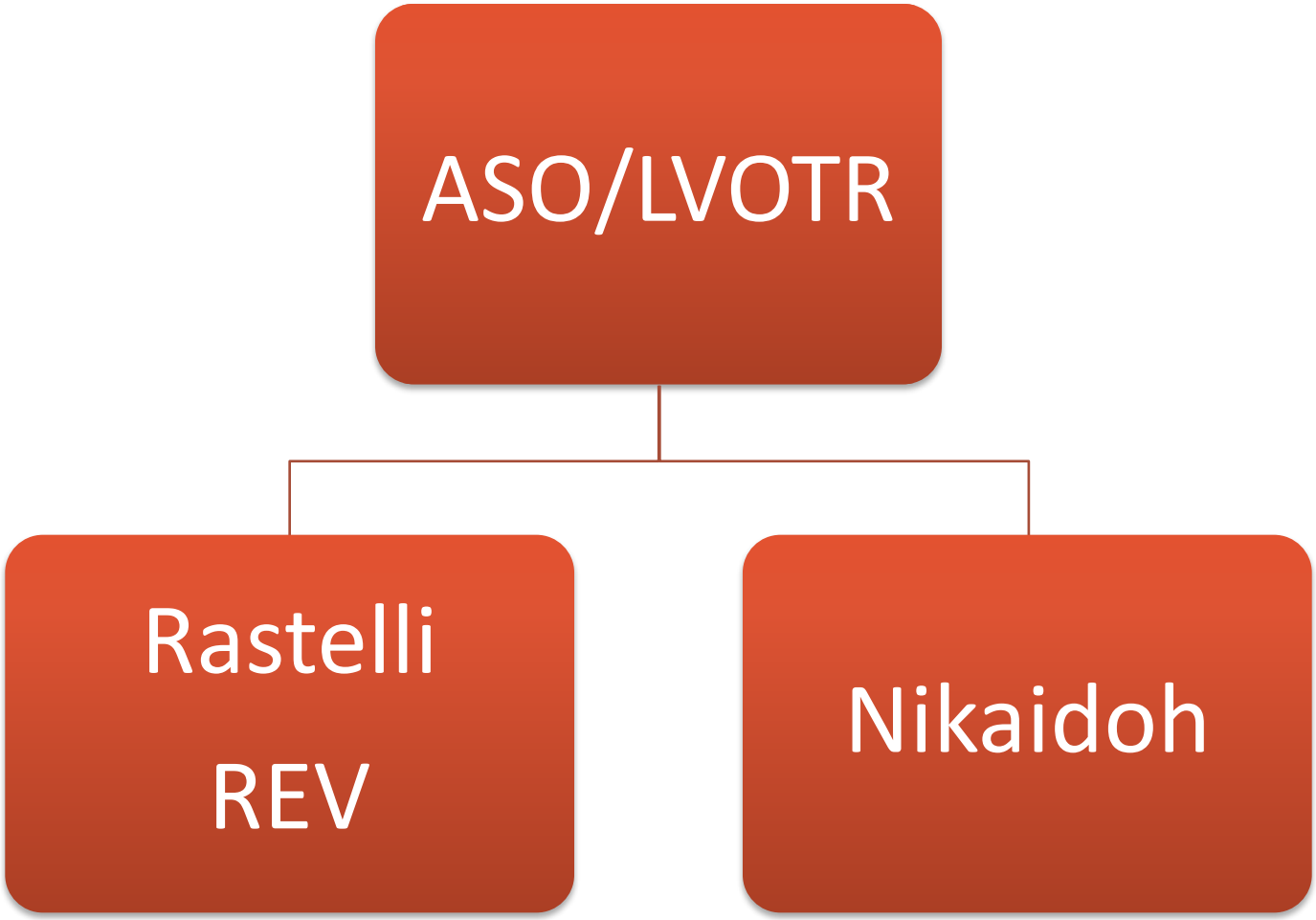
Conclusions

- **In patients with TGA/VSD/PS good outcomes can be achieved with any of the multiple surgical procedures**
  - The most important factor is choosing the correct technique for the "anatomy"
- **Not "one" procedure ideally fits "all patients"**
  - Surgeons need to be familiar with all techniques

Conclusions



Conclusions

- **ASO/LVOTR**
  - Resectable LVOTO
  - PV annulus >-2 to -3
  - “reasonable PV”
  - Avoid BT shunt

Conclusions

- **Rastelli/REV**
  - VSD close to Aorta
    - Short intraventricular tunnel
    - Preserve RV volume
  - Important to resect infundibular septum
    - Reduce recurrent LVOTO

Conclusions

- **Nikaidoh**
  - For patients with complex anatomy
    - Inlet, restrictive, multiple VSD
    - Significant distance from VSD to Ao
    - Abnormal AVV attachments
    - Hypoplastic RV

Conclusions

- **RVOT**
  - Avoid Conduits
  - Direct RV to PA connection significantly decreases RVOT reoperations

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