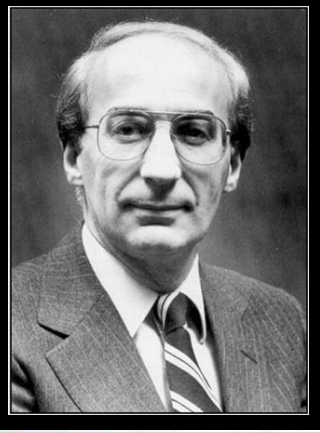
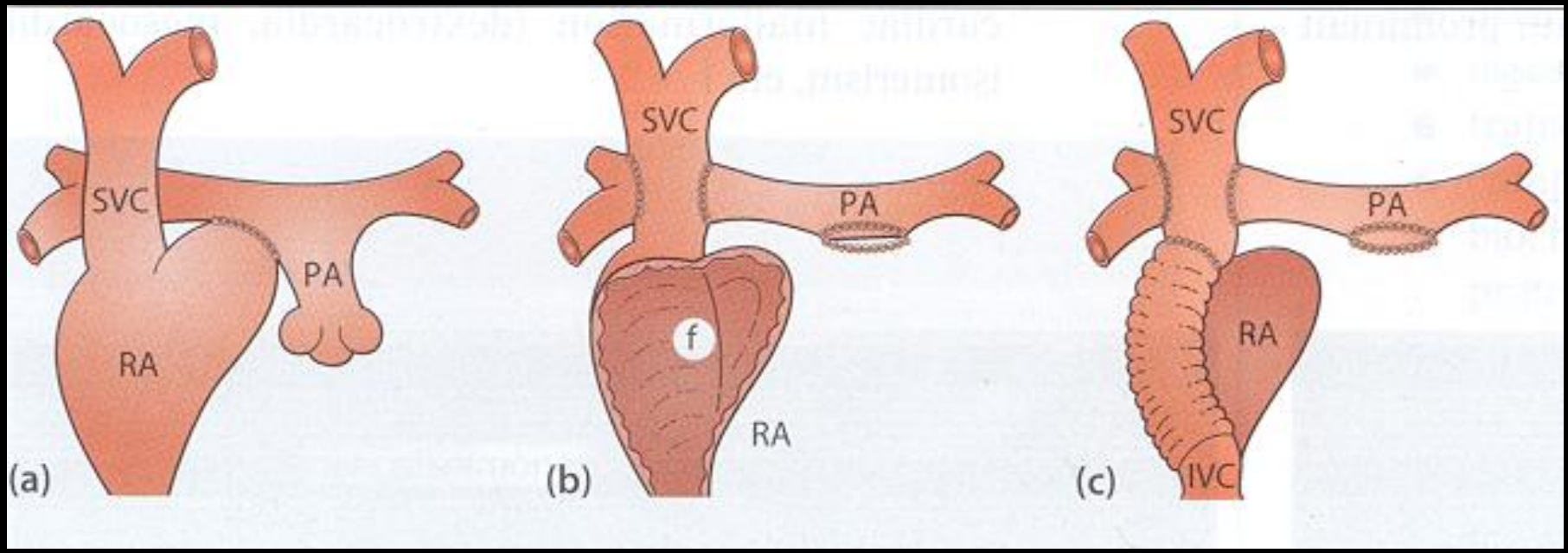
Catheter Ablation in the Fontan

Approaches in Atriopulmonary, Lateral Tunnel and Extracardiac

Mark McGuire
Royal Prince Alfred Hospital
Prince of Wales Hospital
Sydney Australia

Fontan Procedures



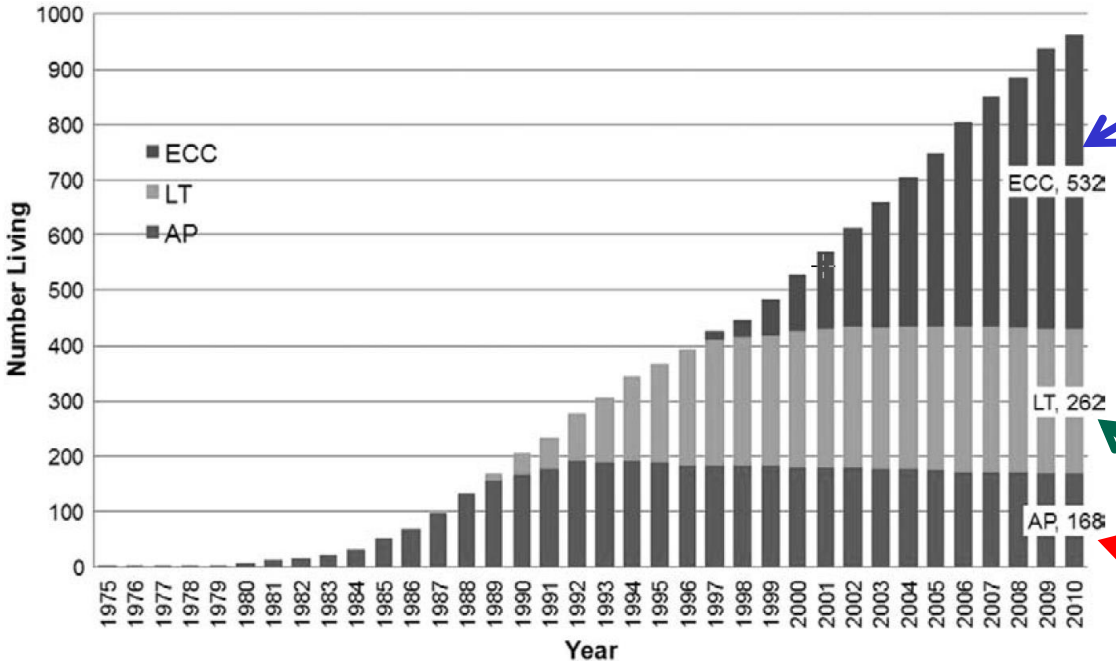


Atriopulmonary Connection 1971

Lateral Tunnel (TCPC) 1987

Extracardiac 1990

Types of Fontan Australia & New Zealand (n=1006)



Extracardiac Conduit (n:532) 53%

Figure 1. Distribution of the techniques used in the growing population of Fontan patients alive in Australia and New Zealand. AP indicates atriopulmonary connection; ECC, extracardiac conduit; and LT, lateral tunnel.

Lateral Tunnel (n:262) 26%

Atrio-pulmonary (n:168) 17%

d'Udekem Y et al Circulation 2014;130:[suppl 1]S32-S38

Risk of Arrhythmia Post Fontan (n=520)

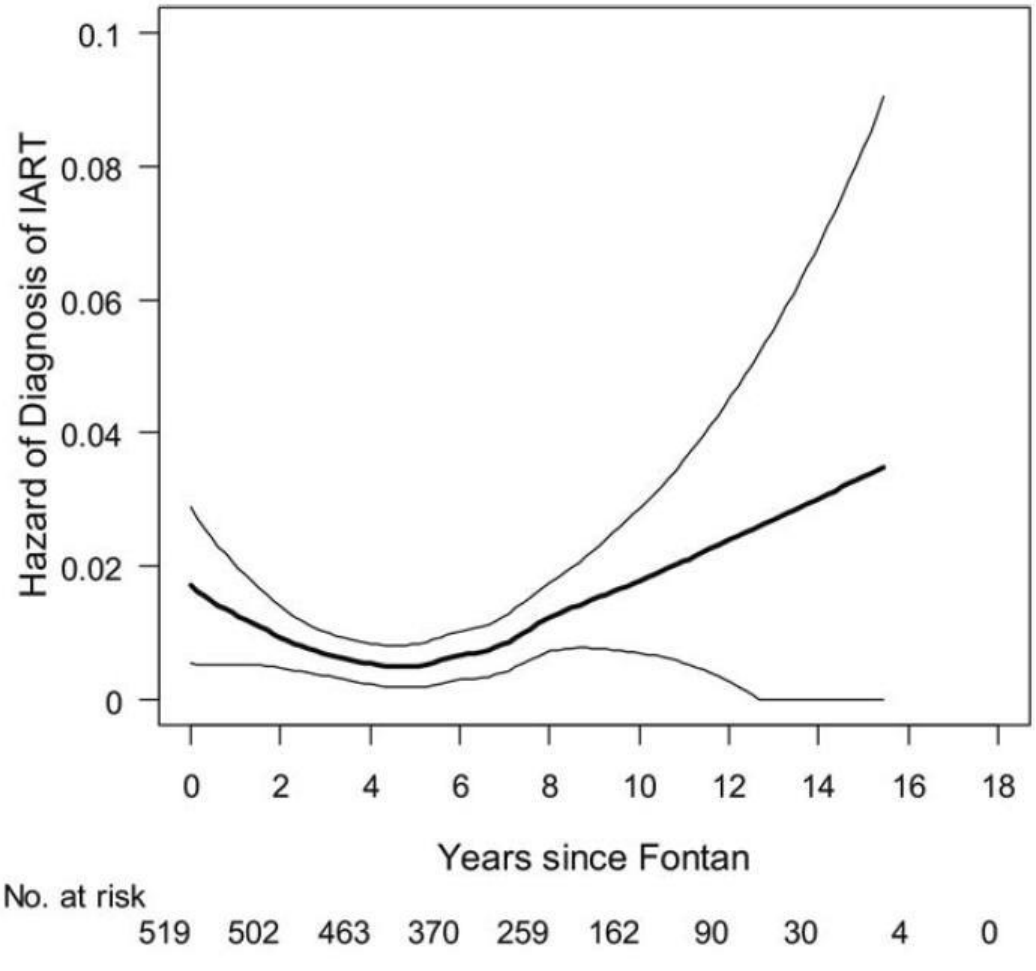


Figure 1 **Hazard of IART Following the Fontan Operation, With 95% Confidence Bands**

Date of diagnosis was missing for 1 patient, thus the number at risk initially is 519. IART = intra-atrial re-entrant tachycardia.

Stephenson EA et al
J Am Coll Cardiol
2010;56:890–6

Risk of Arrhythmia Post Fontan (n=520)

Risk Depends on Type of Procedure

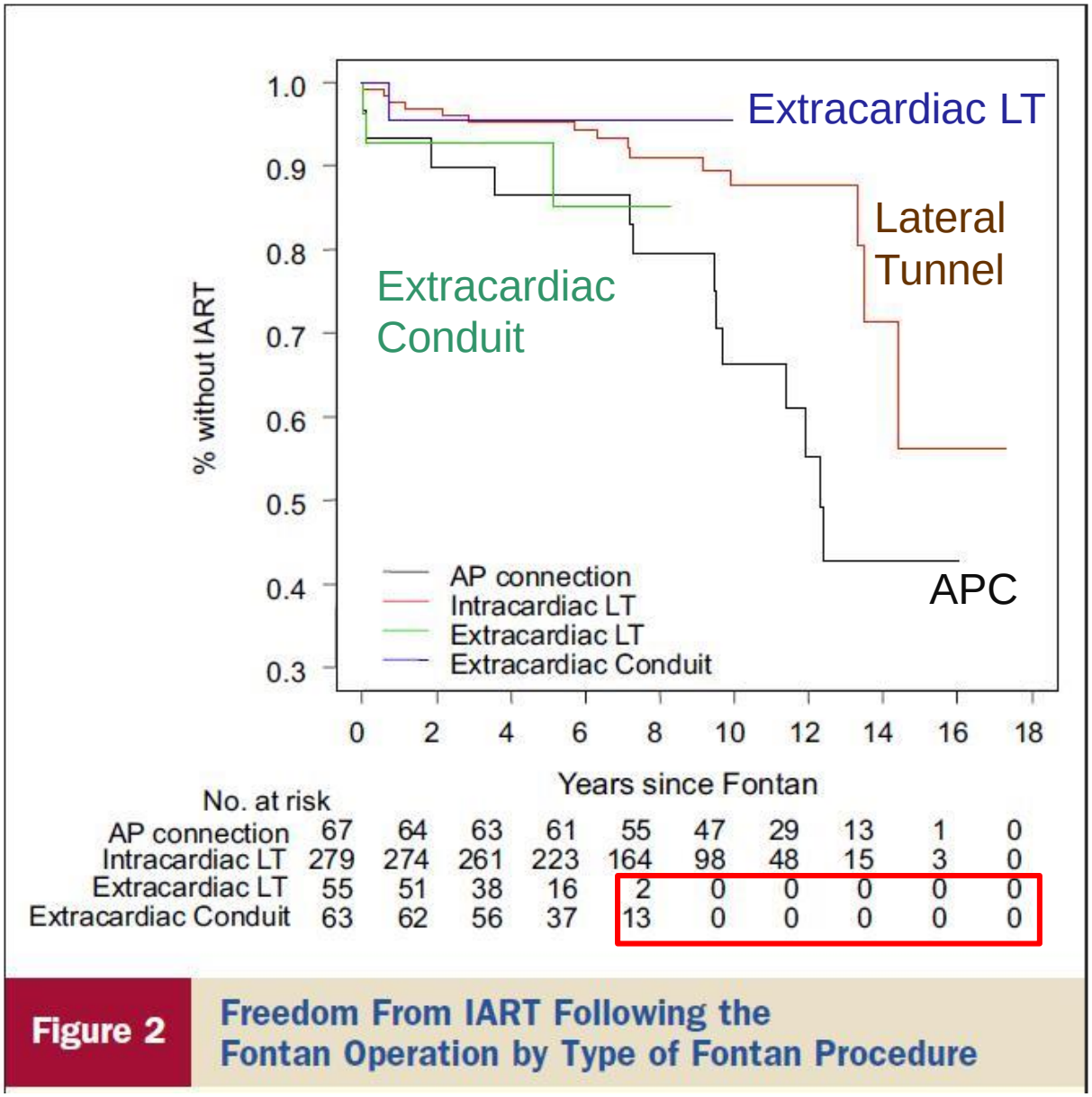
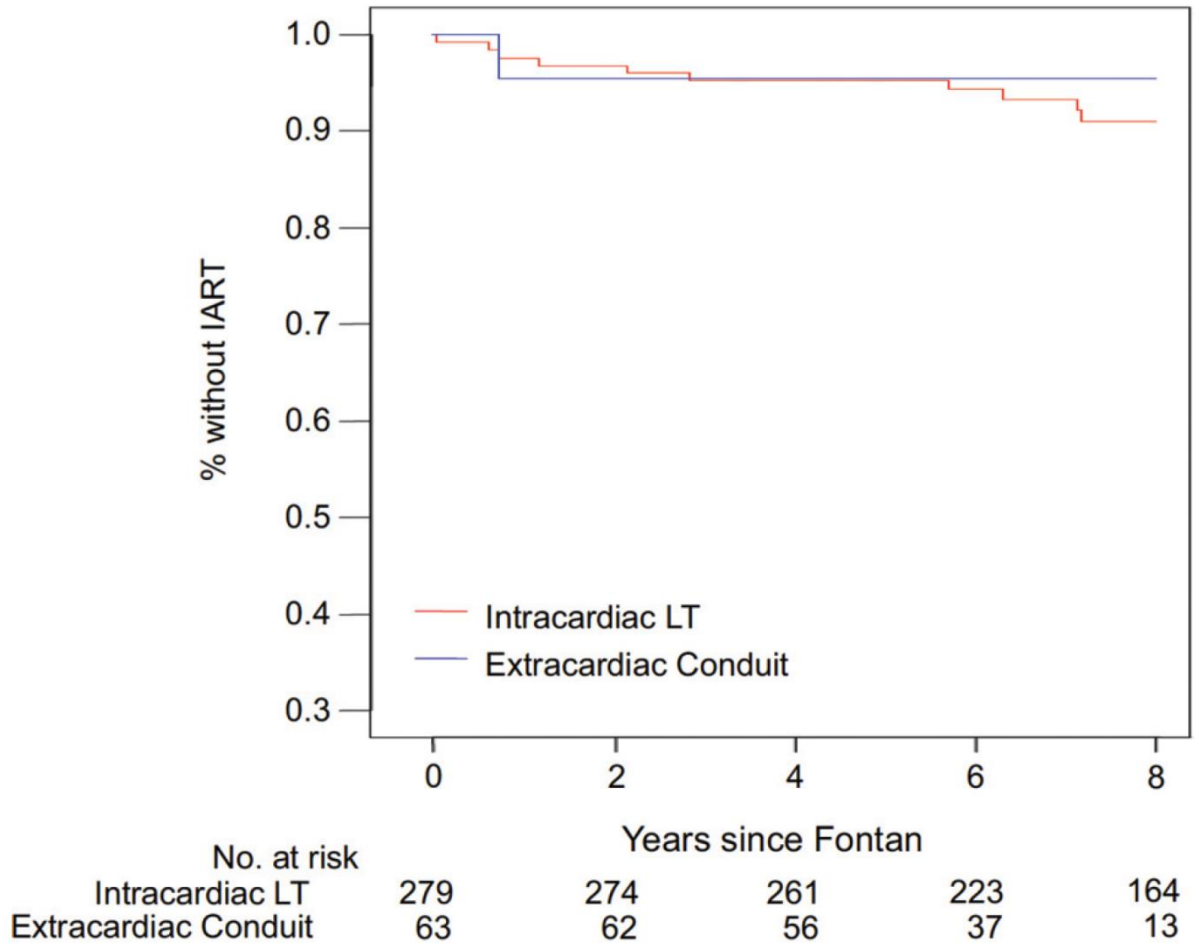


Figure 2 Freedom From IART Following the Fontan Operation by Type of Fontan Procedure

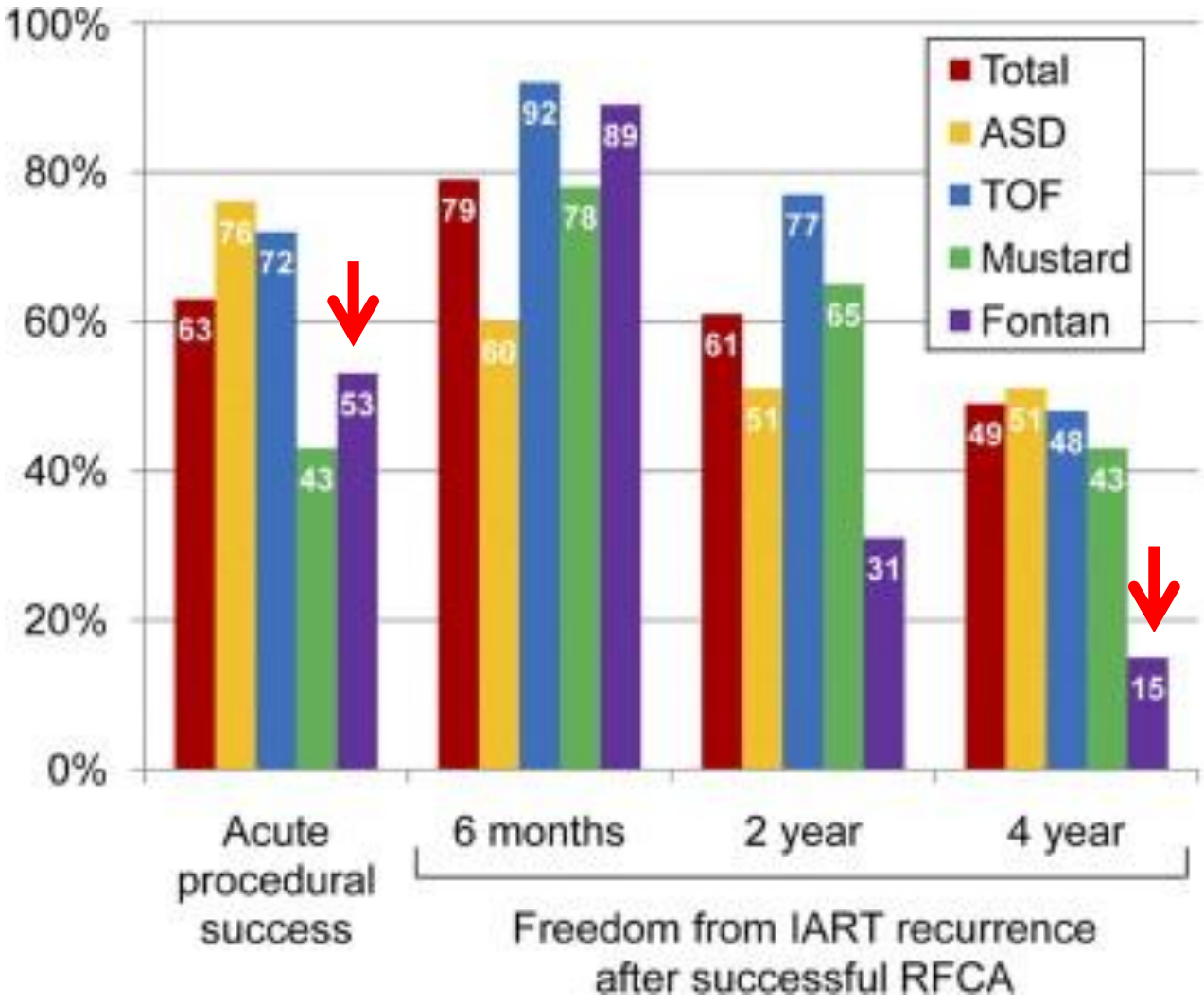
Stephenson EA et al
J Am Coll Cardiol
2010;56:890–6

ADJUSTED INCIDENCE ATRIAL ARRHYTHMIAS
No difference between LT & ECC P=0.22



Stephenson EA et al
J Am Coll Cardiol 2010;56:890–6
Modified by Khairy P

Acute Procedural Success and Freedom From Arrhythmia Recurrence After Successful Ablation



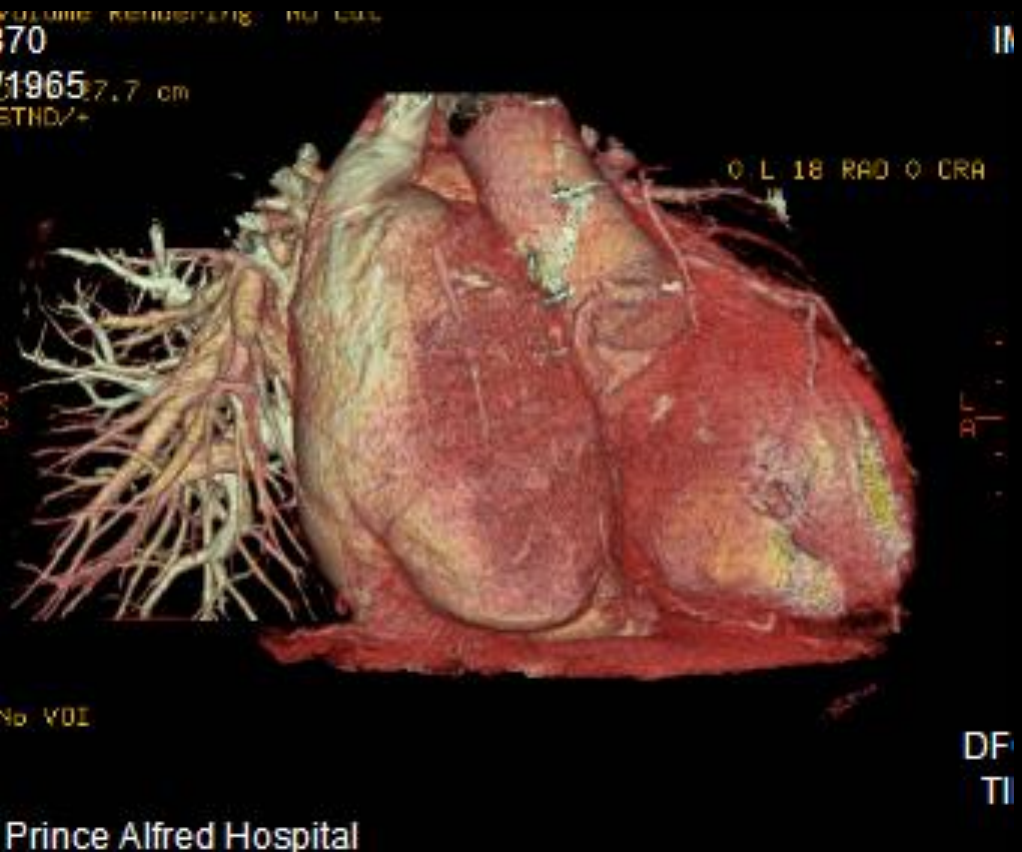
Yap S-C et al JACC 2010;56:1589

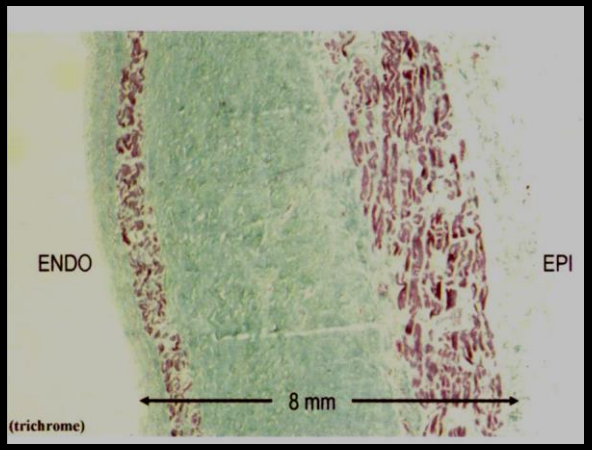
CATHETER ABLATION IN THE FONTAN PATIENT

Challenges

- Multiple reentrant circuits
- Thick walled chambers
- Large chambers: catheter contact
- Low flow (coagulum formation)
- Limited access (LT, ECC, Patch over TV, ilio-femoral stenosis)
- Difficulty identifying site of conduction system
- Heart block: urgent thoracotomy
- Tachycardia poorly tolerated
- Stable timing reference difficult
- Cannulating CS

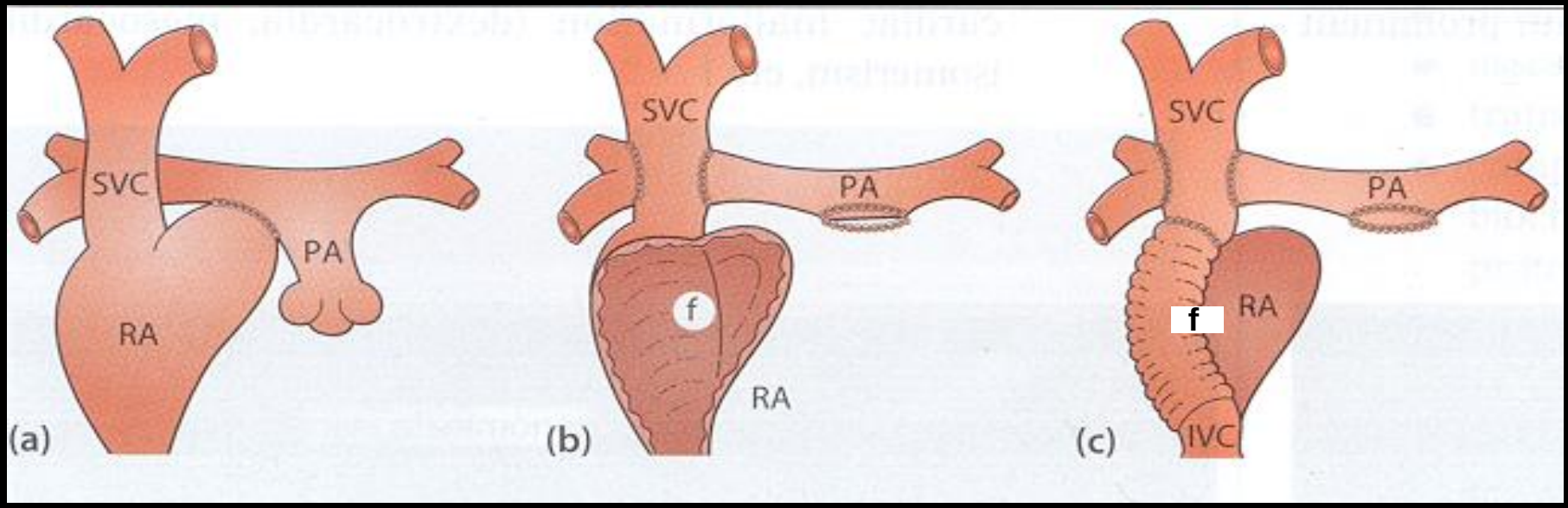
Right Atrium Post Fontan





EP Walsh

Fontan Procedures – Specific Issues



APC

Chamber size +++
Wall Thickness +++
No. tachycardias +++
Baffle puncture usu. not required
Baffle puncture difficulty +

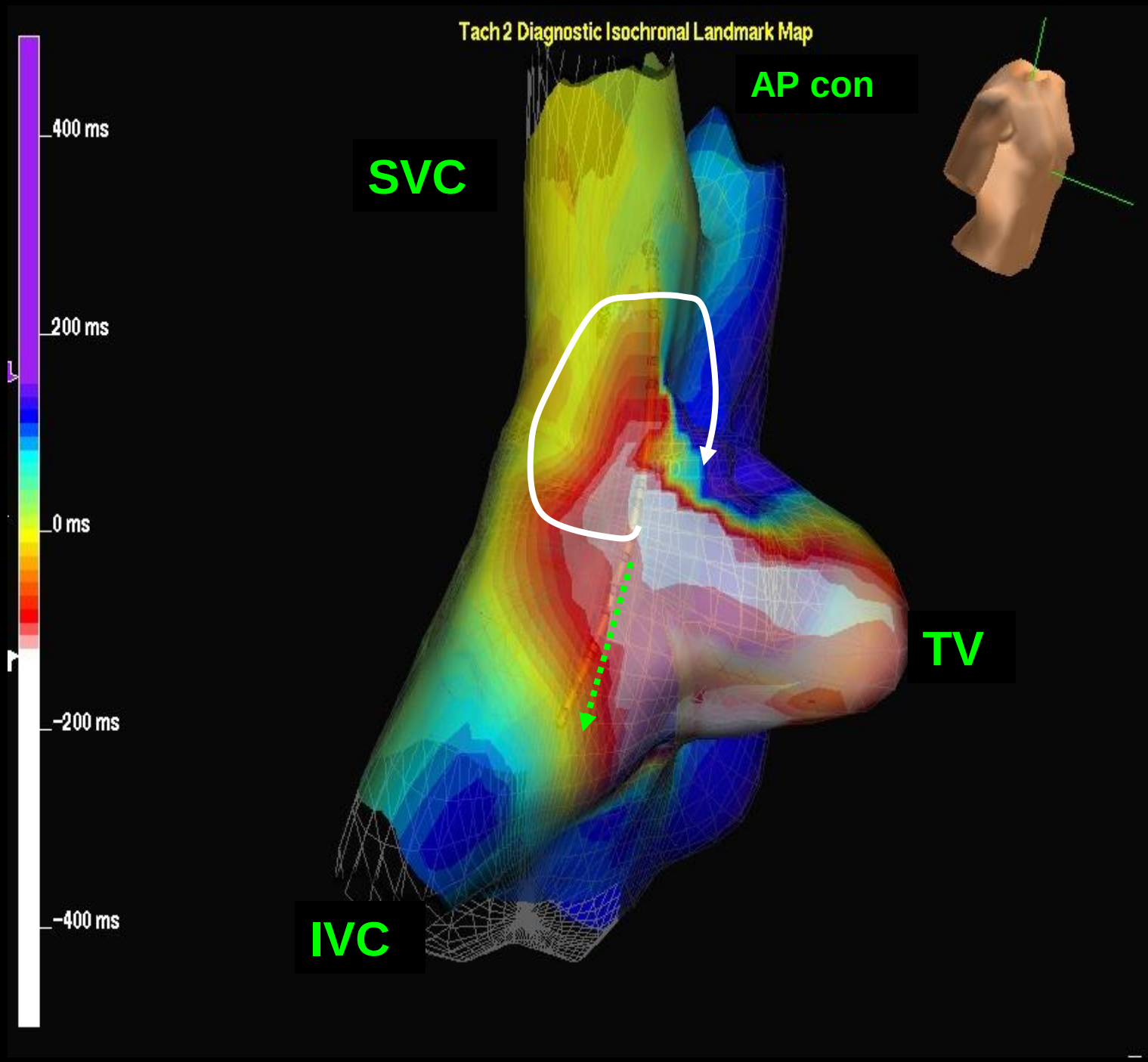
Lateral Tunnel

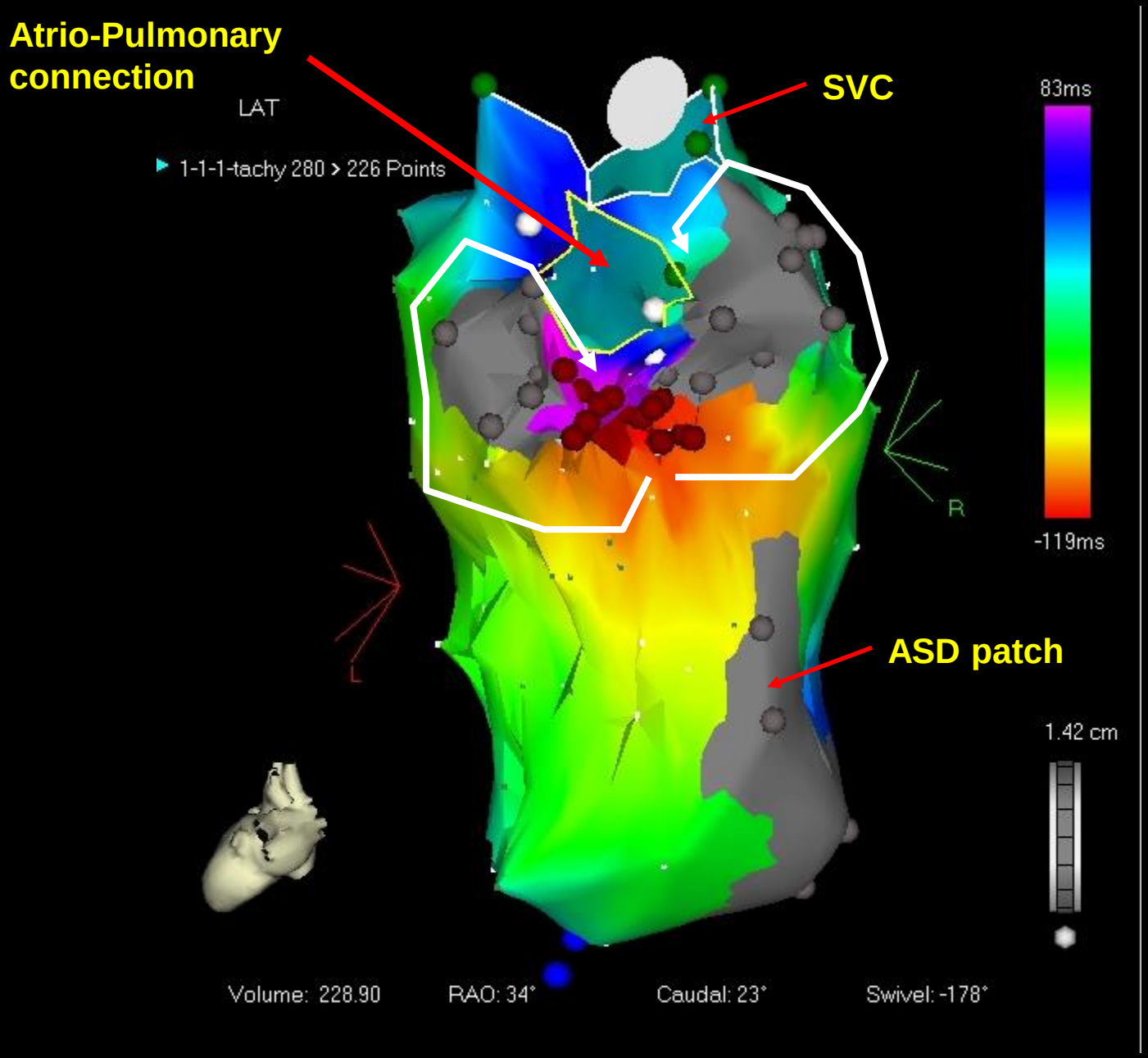
Chamber size +
Wall Thickness ++
No. tachycardias ++
Baffle puncture usu. not required
Baffle puncture difficulty +
Fenestrations often present

ECC

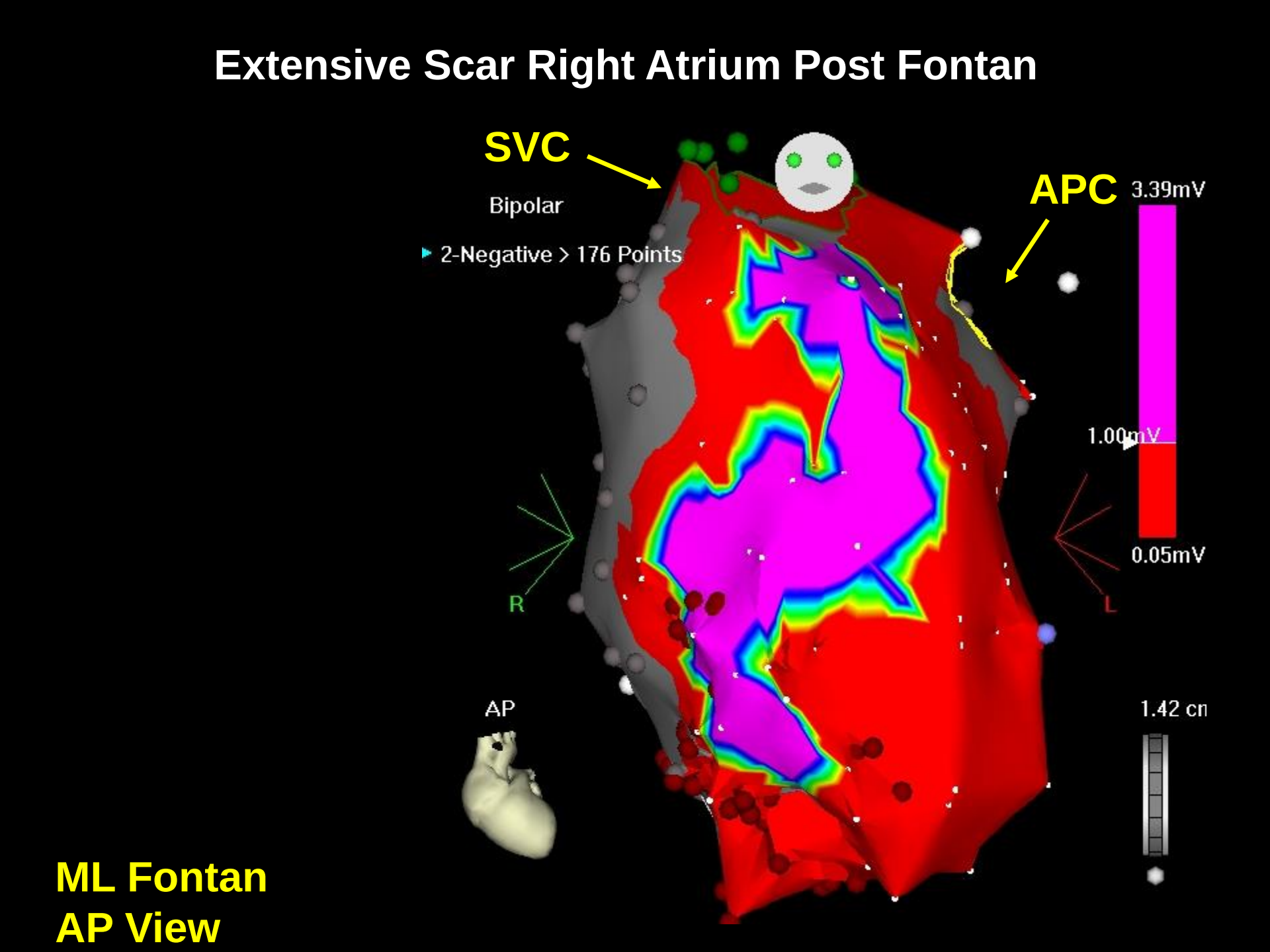
Chamber Size +
Wall Thickness +
No. tachycardias +
Conduit crossing always required
Conduit puncture difficulty +++

Mr DP 22
Fontan
Tricuspid atresia

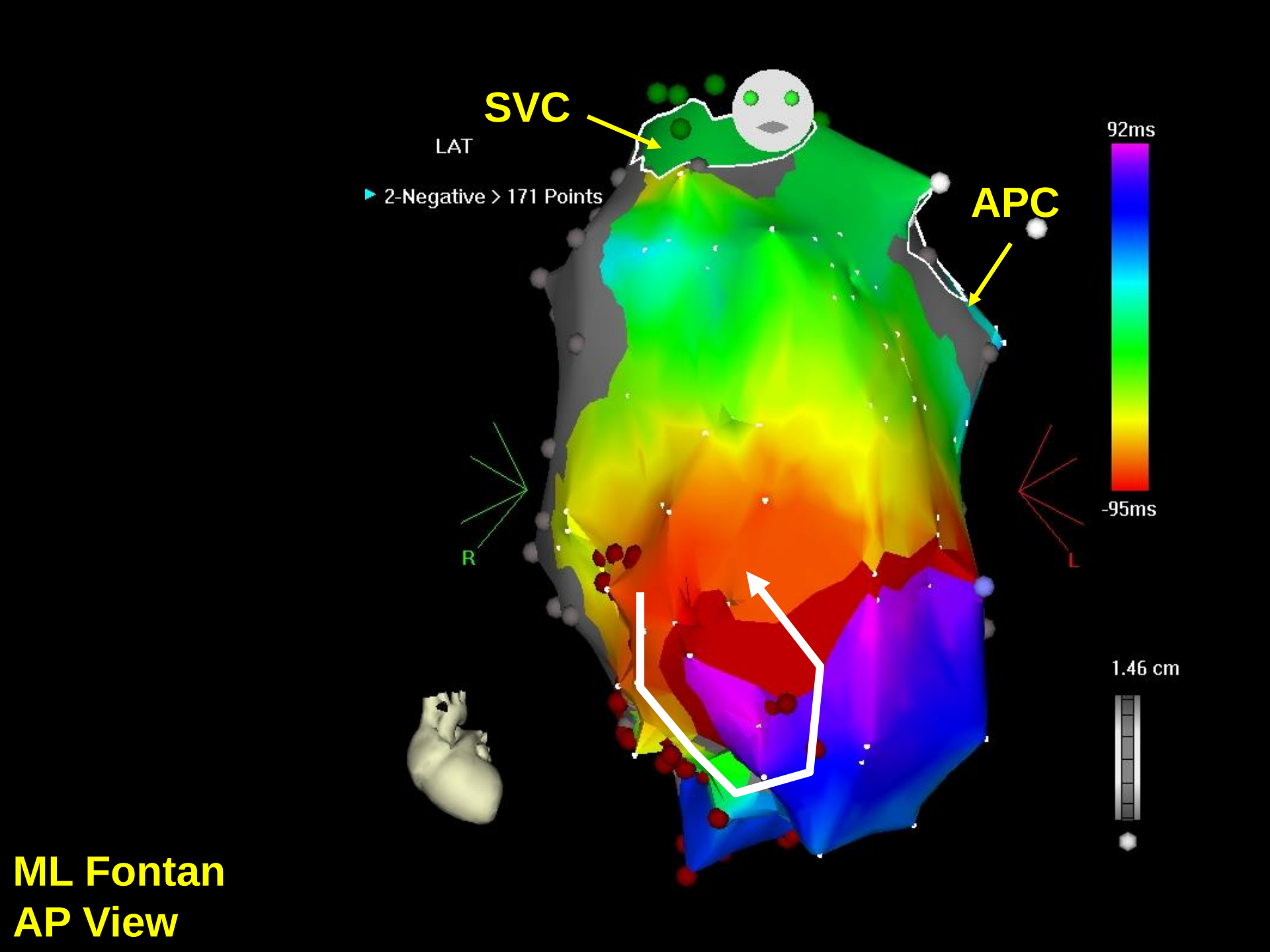


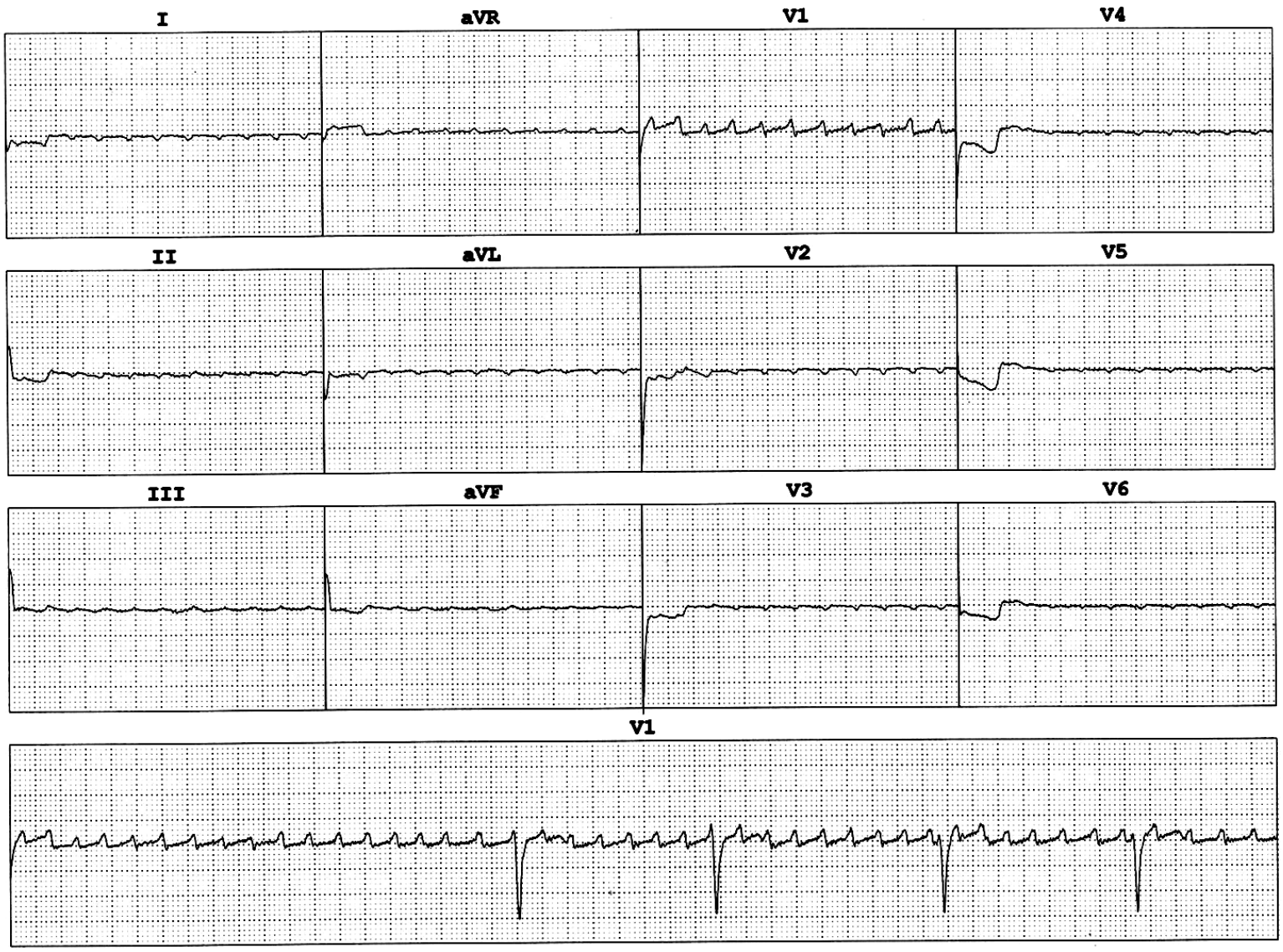

Atrio-Pulmonary connection
LAT
1-1-1-tachy 280 > 226 Points
SVC
83ms
-119ms
R
ASD patch
1.42 cm
Volume: 228.90
RAO: 34°
Caudal: 23°
Swivel: -178°

Extensive Scar Right Atrium Post Fontan



ML Fontan
AP View


SVC
APC
LAT
2-Negative > 171 Points
92ms
-95ms
R
L
1.46 cm
ML Fontan
AP View


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
V1

MRI in Extracardiac Fontan

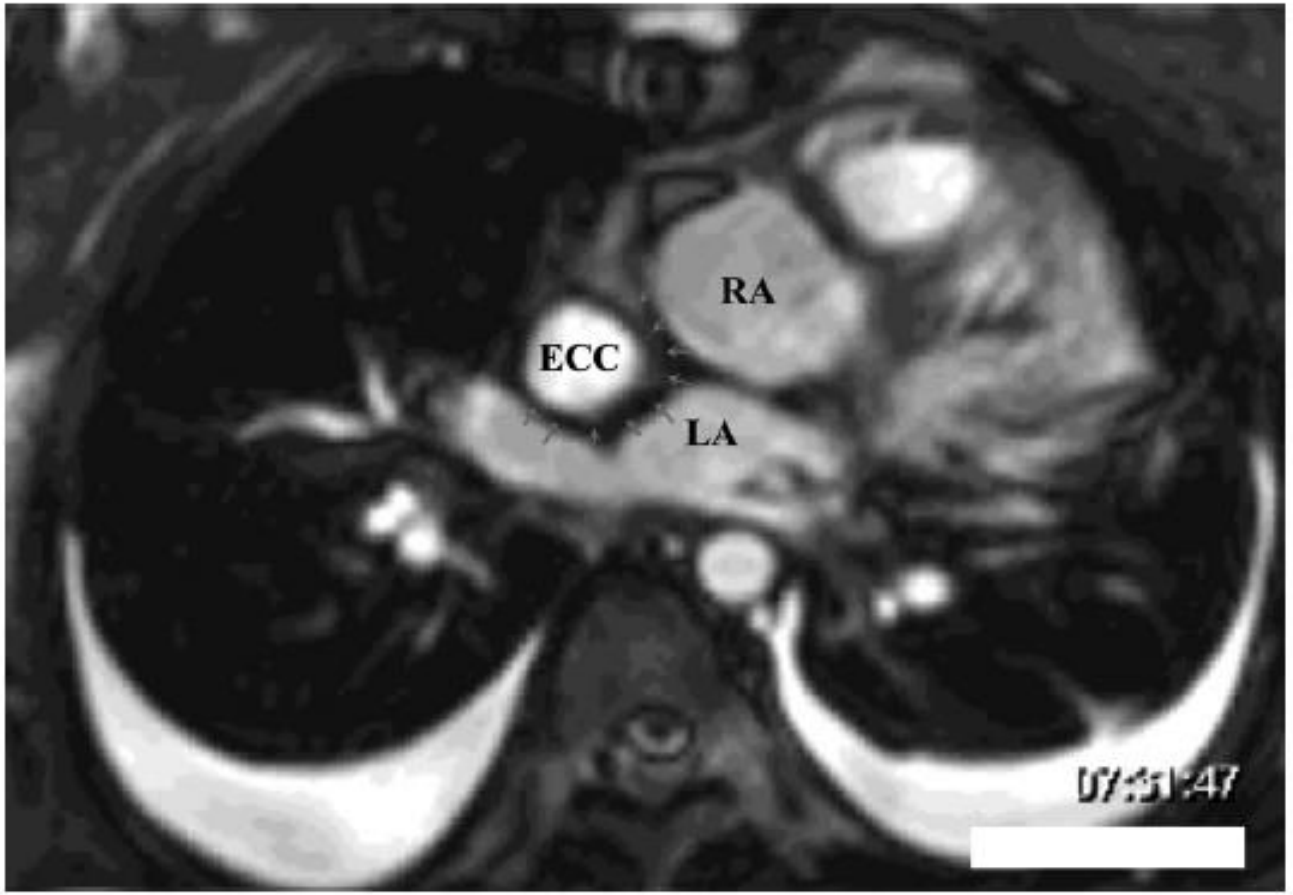
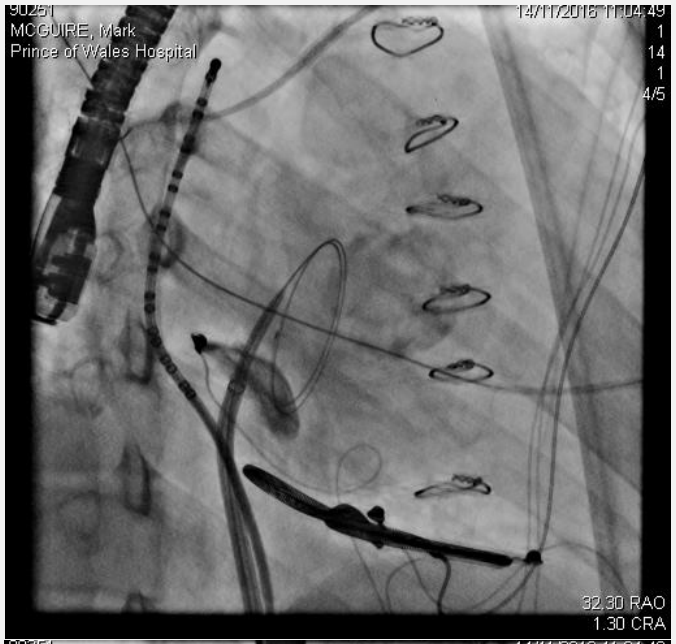


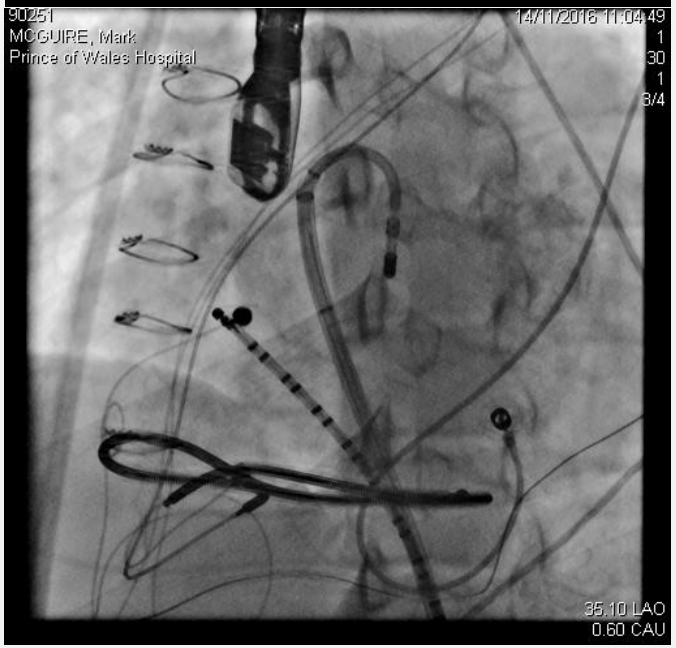
Figure 6. Relationship between extracardiac conduit and pulmonary venous atrium. An axial view of a MRI scan is shown in a patient with an extracardiac conduit (ECC). The arrows mark the space between the ECC and neopulmonary venous atrium, composed of the right (RA) and left (LA) atria.

Khairy P , Circulation 2012;126:2516

Transconduit Puncture: female 33, situs inversus, laevocardia, mitral atresia, ECC

35 atm.

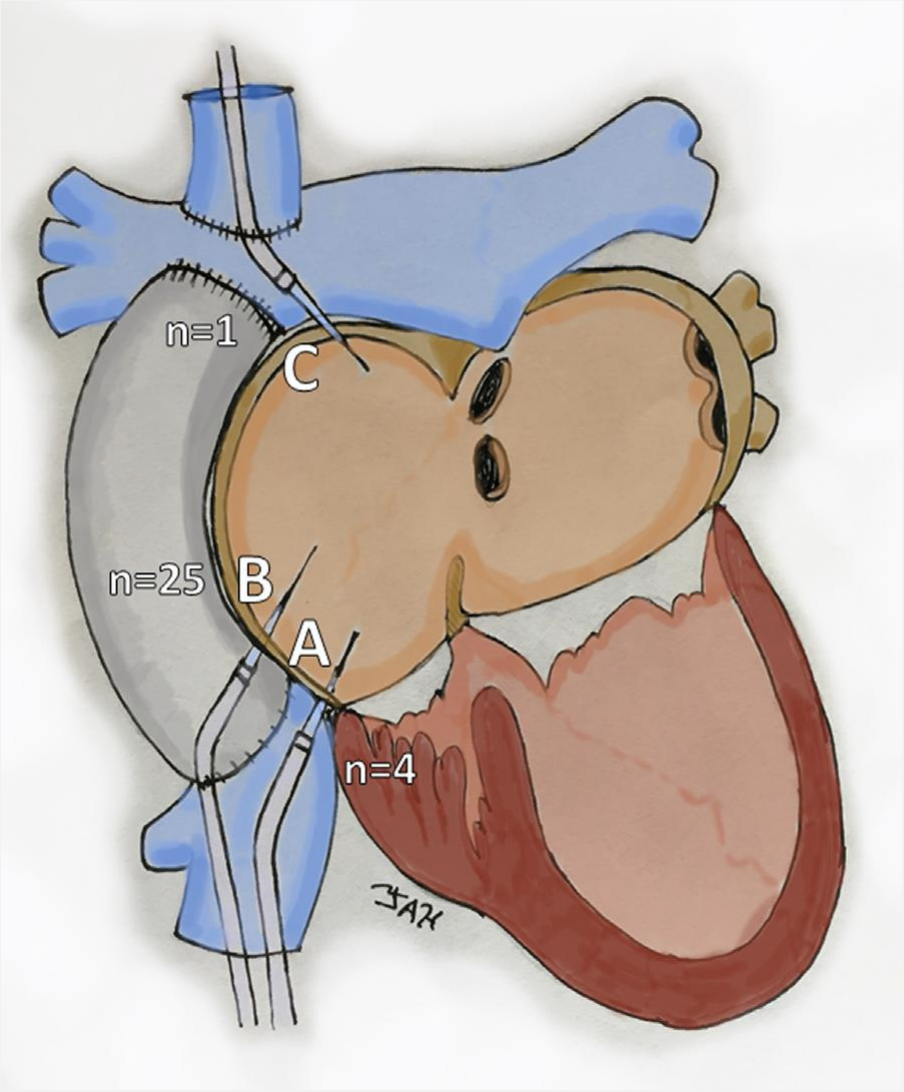




TRANSCONDUIT ACCESS FOR ABLATION IN EXTRACARDIAC CONDUIT

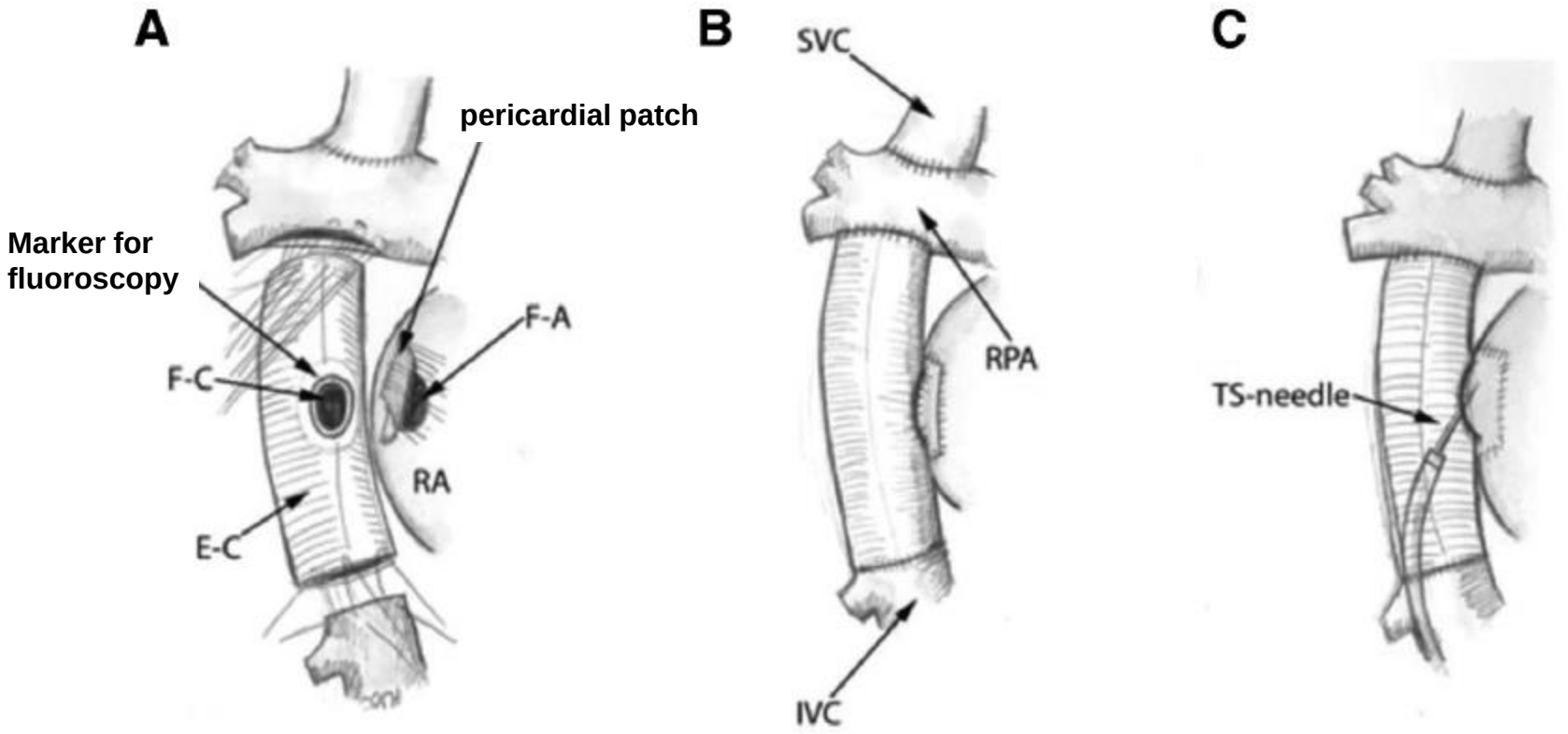
Multi-centre (9)
N: 36 patients
Primary ECC:24; Conversion 12

Procedures: 46
Access to atrium:
Puncture: 63%, Fenestration: 26%
Acute success: 83%
No complications due to conduit puncture
Arrhythmia recurrence: 17% @ 0.4 yrs



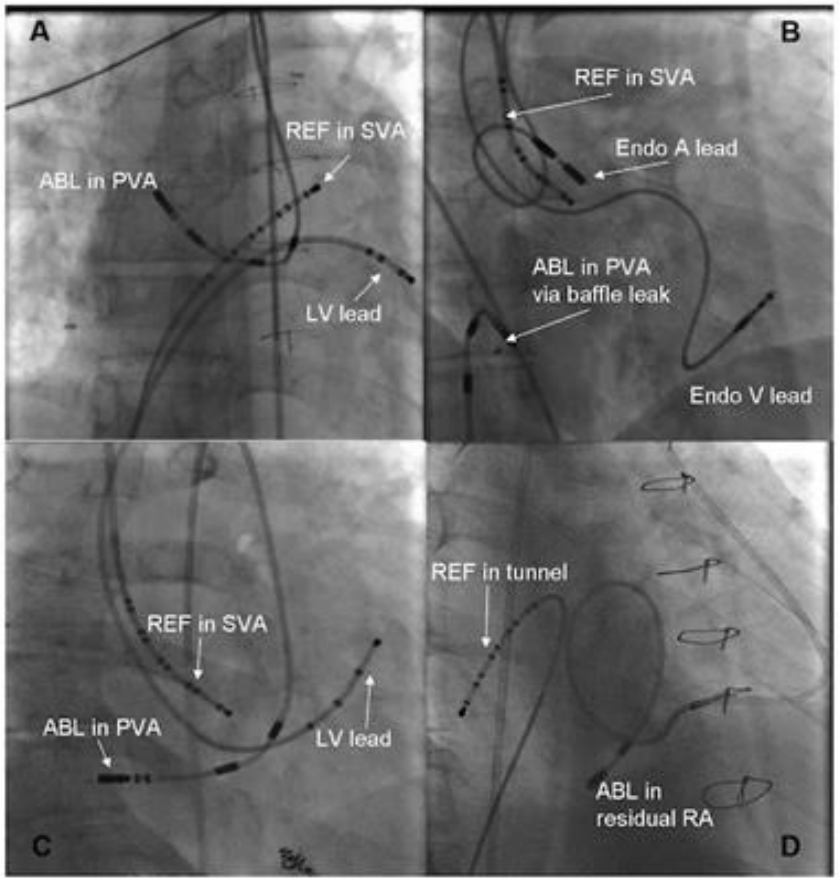
Moore JP et al Heart Rhythm 2016;13:1891-1897

Trapdoor Modification to Extracardiac Fontan



Aboulhosn J et al, Congenit Heart Dis 2010;5:430

Robotic Magnetic Navigation in Congenital Heart Disease



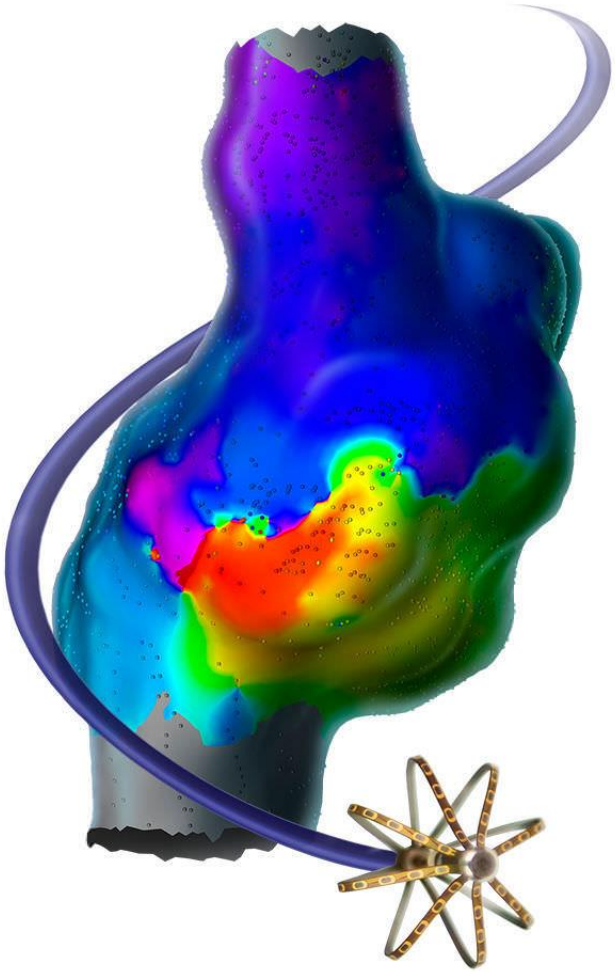
- **Very expensive**
- **Others haven't had same results**
- **Several centres have abandoned**

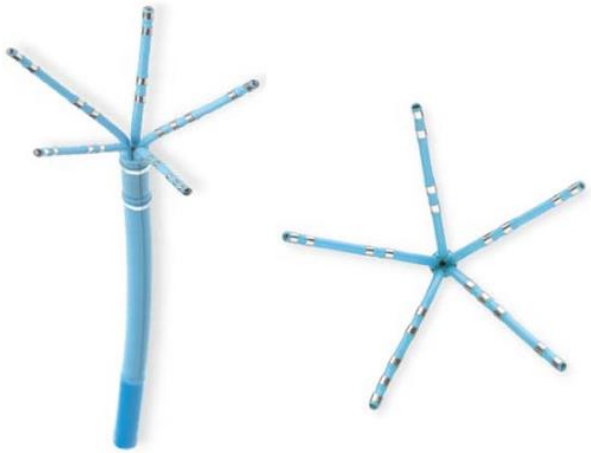
N= 13

No recurrence in 10/13, mean FU 201 days

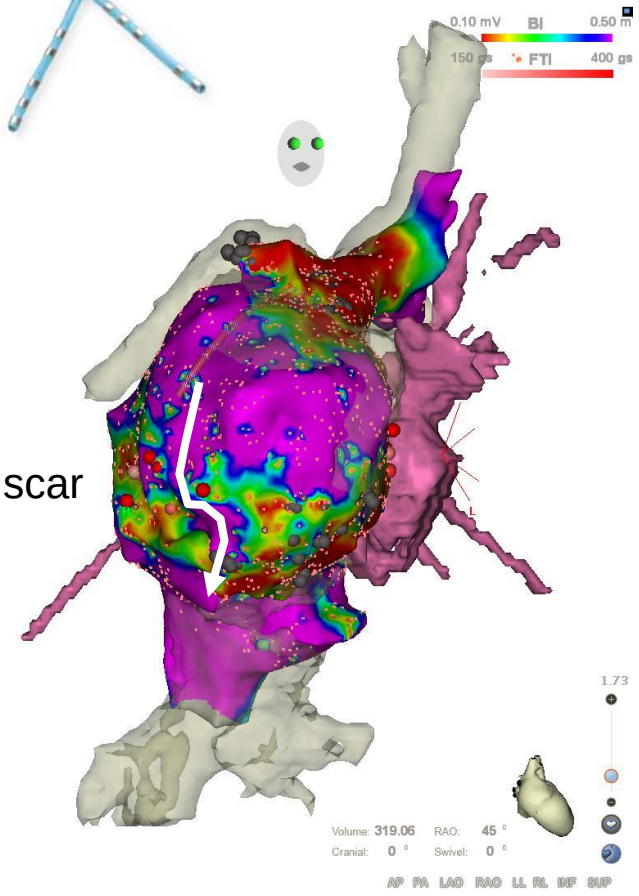
Ernst S et al, Circ Arrhythm Electrophysiol. 2012;5:131-139

High Resolution Simultaneous Multipoint Mapping





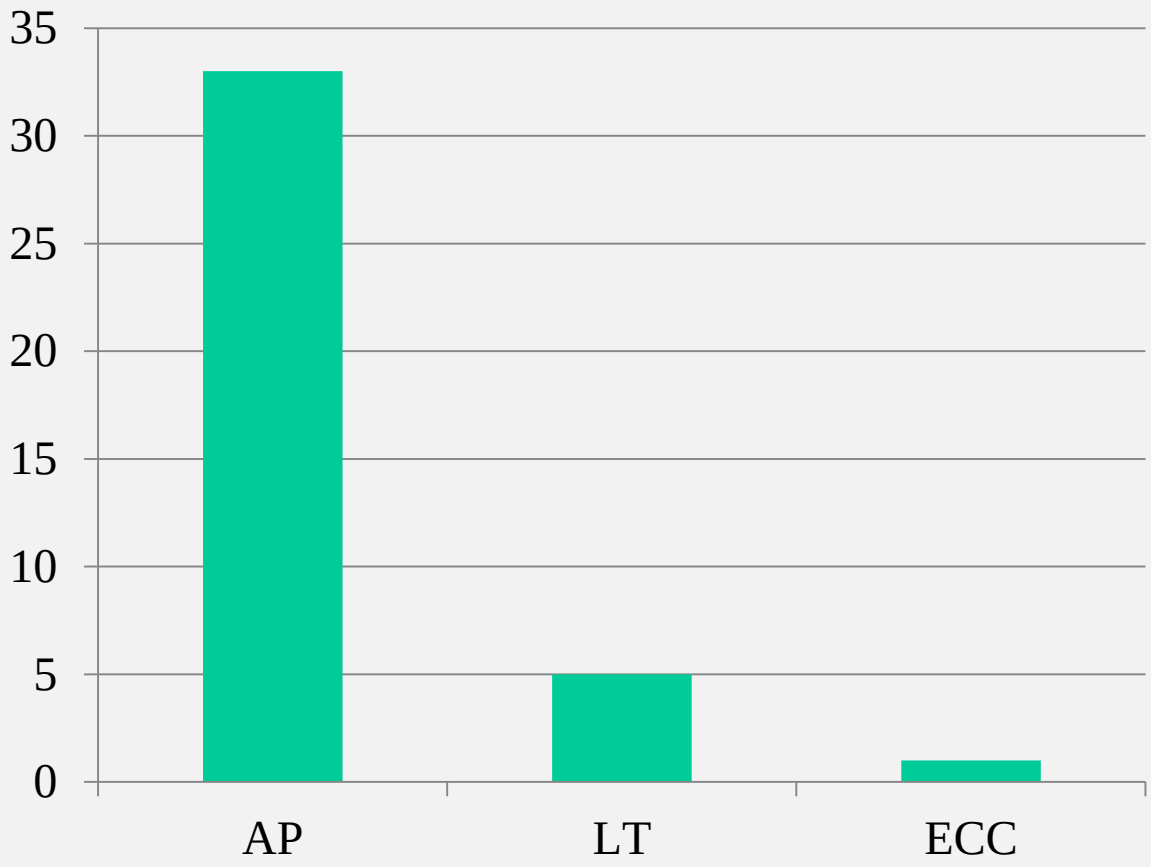
Channel through scar



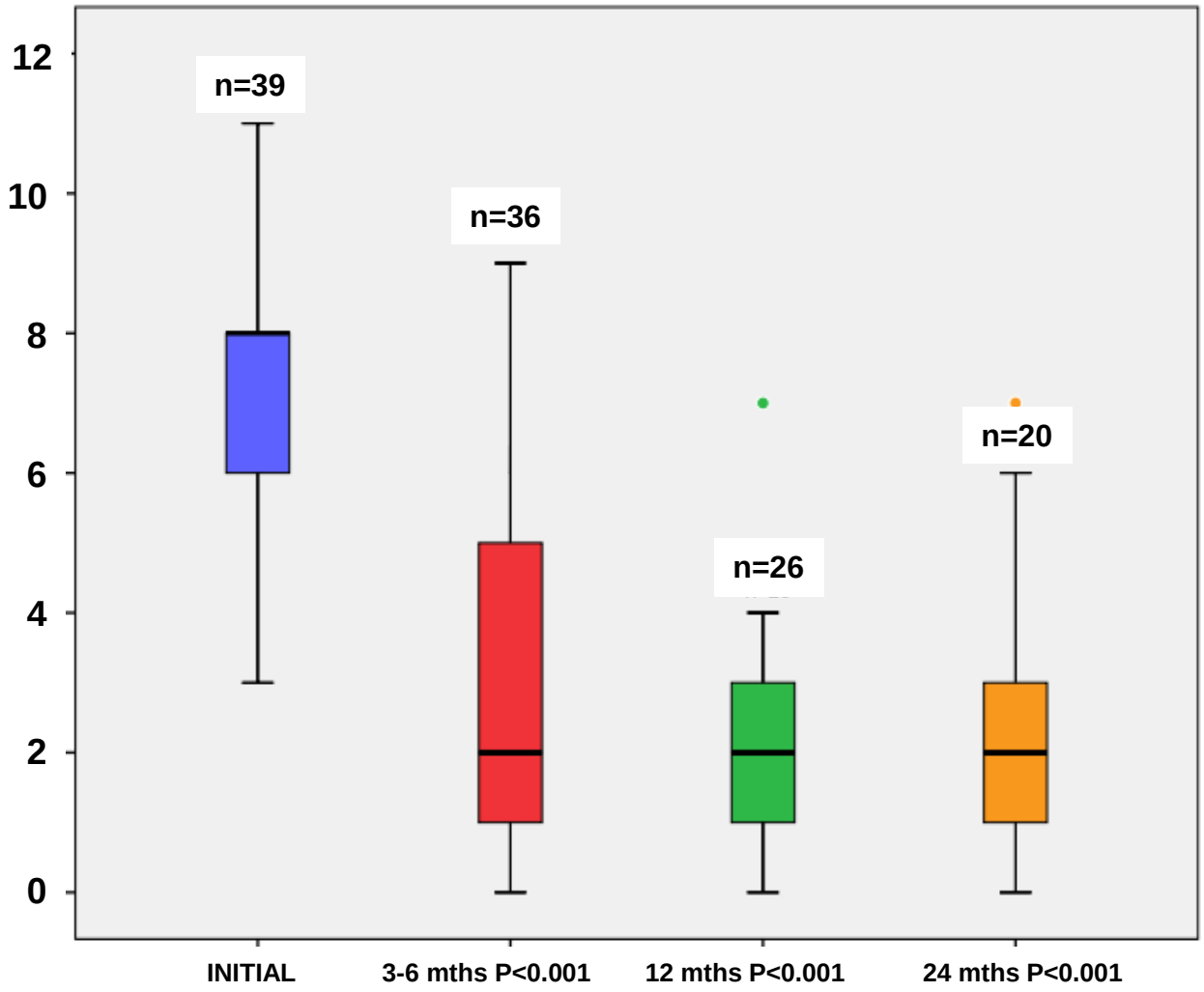
Catheter Ablation in Fontan

- 39 consecutive ablations in 28 Fontan patients, 2000-2017
- Mean age 33+/- 7 years
- Mean 2.8 tachycardias per case (range 0-10)
- Ablation strategy:
  - Targeted via activation / entrainment mapping in 15 (38%)
  - Substrate-based approach in 11 (28%)
  - Targeted + substrate in 11 (28%)
  - Slow pathway ablation for concurrent AVNRT in 2 cases
- Mean follow up 4.2 years

Ablation: Fontan Types



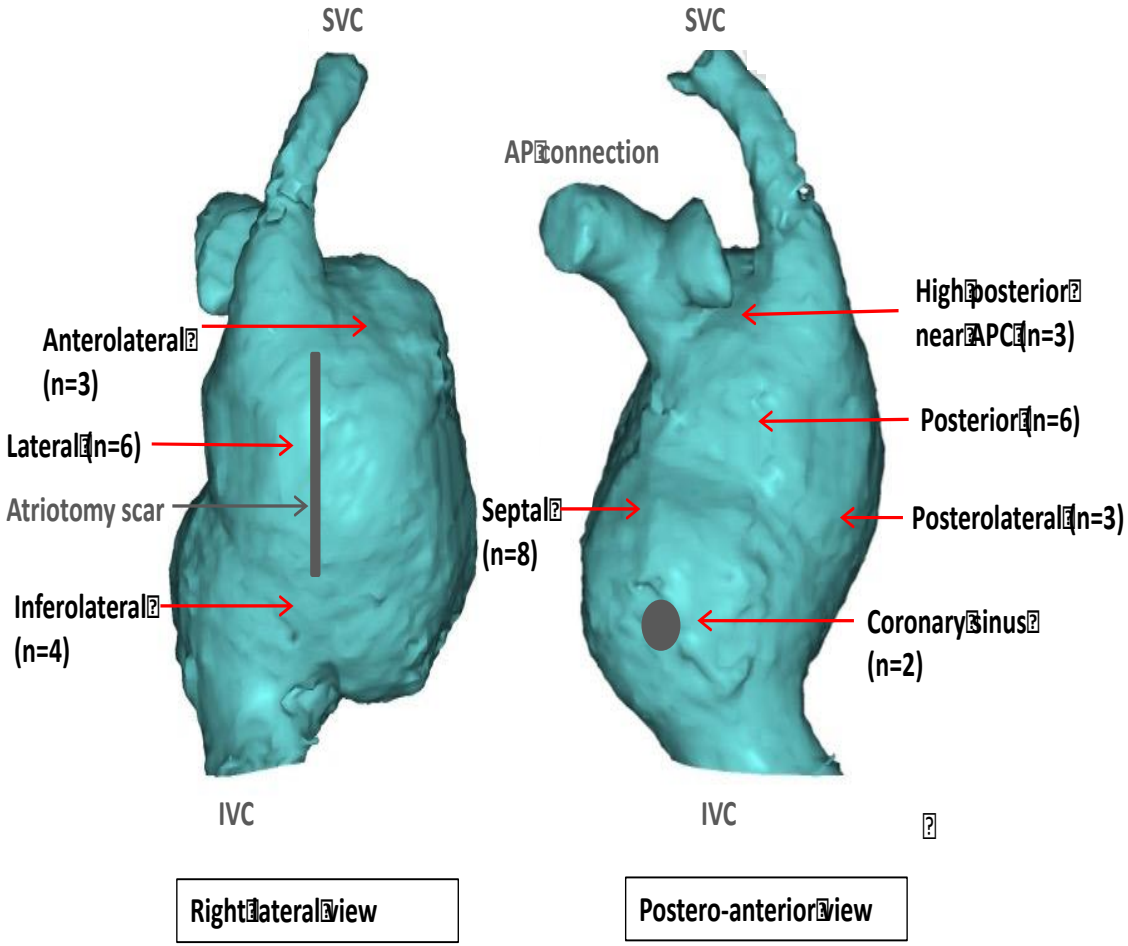
Clinical Arrhythmia Severity Score (CASS)* Following Ablation



* Triedman JK et al JACC 2002

Moore B et al 2017

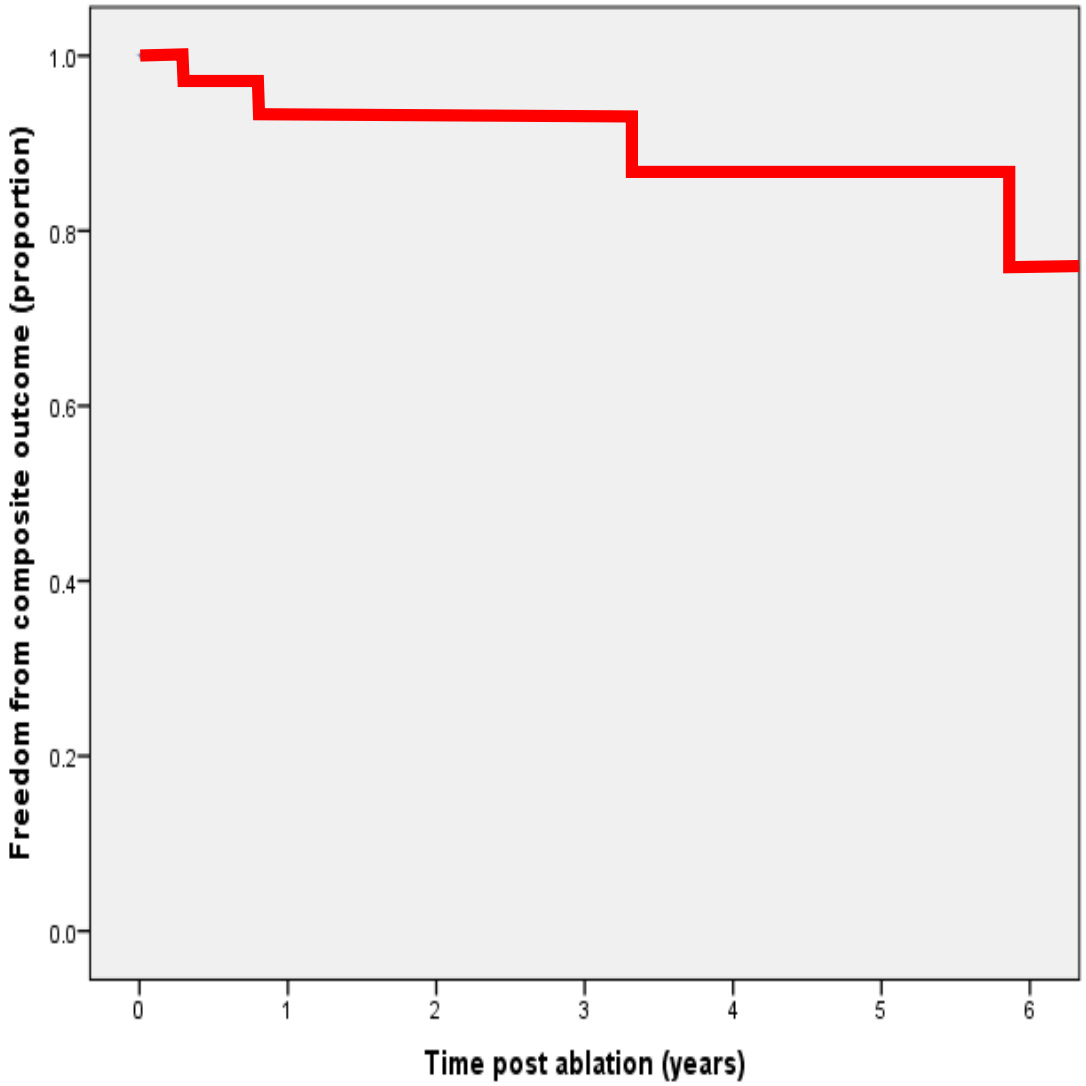
Location of the critical isthmus



Acute Procedural Success & Complications

- “Complete” success in 23/39 cases (59%)
- “Partial” success in 10/39 cases (26%)
- “Unsuccessful” in 6/39 cases (15%)
- Suspected clinical arrhythmia ablated in 79%
- Complications
  - 4 femoral vascular complications (3 required minor operative intervention)
  - 2 cases where temporary pacing was required post procedure for transient bradycardia (1 temporary pacing wire, 1 transcutaneous)
  - No strokes or pericardial effusions / tamponade

Freedom from Death, Transplant, Conversion Surgery Post-Ablation



CATHETER ABLATION IN THE FONTAN PATIENT

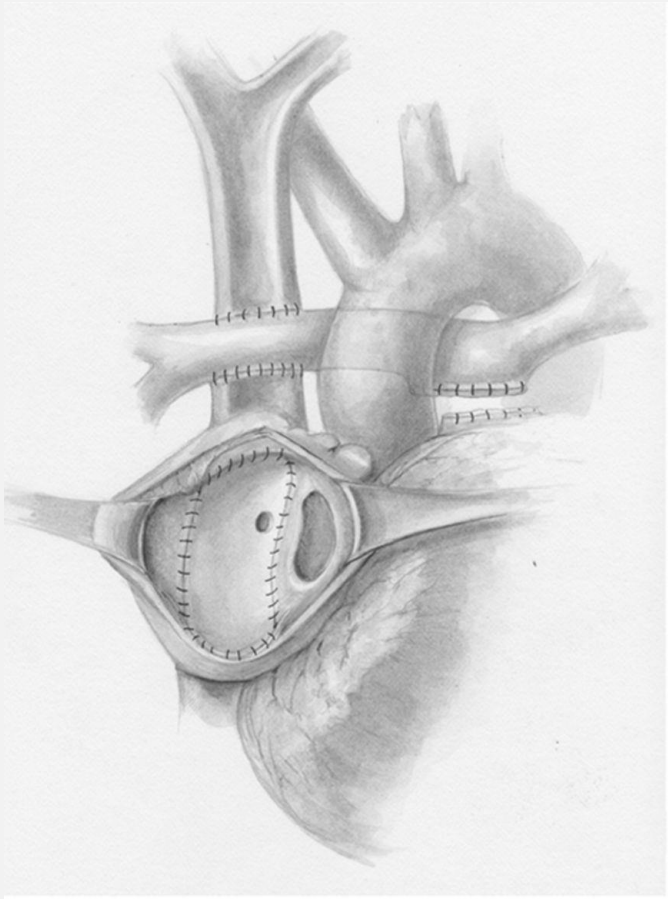
Summary

- Difficult procedure (median proc. time 300 mins)
- Most patients benefit (acute success: 50-80%) – improvement sustained for several years
- Evolving technology likely to improve acute success rate
- Complication & mortality rate low
- Palliative rather than curative: recurrence rate significant
- Should it be always be considered before revision surgery in patients with good haemodynamics?

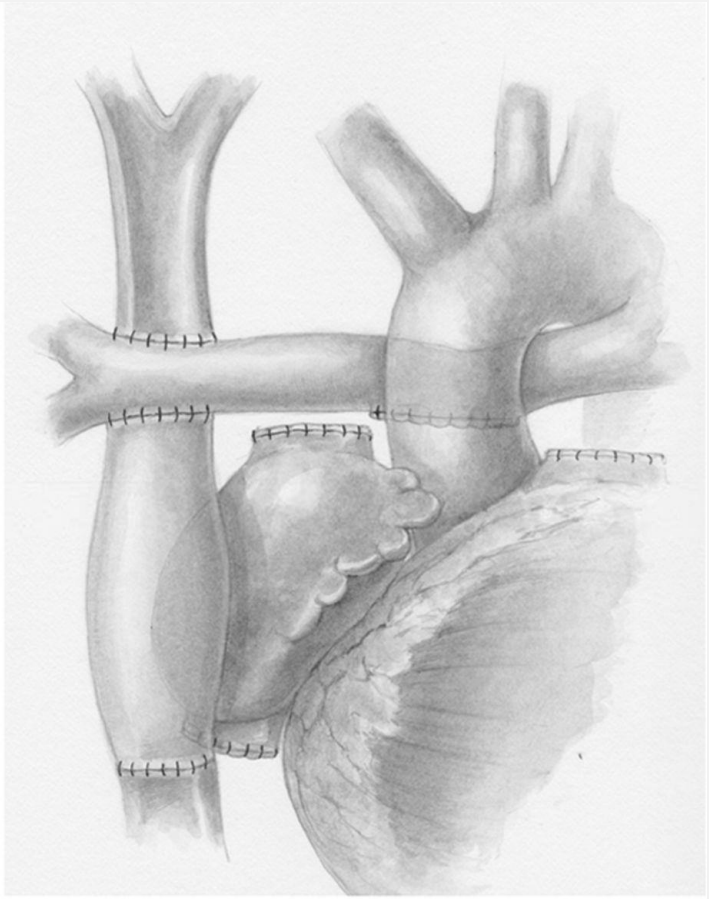
Clinical Arrhythmia Severity Score*

Category	Score	Category	Score
Documented Arrhythmia		**Frequency of Cardioversion**	
none	0	no DCCV	0
non-sustained	1	one DCCV	1
sustained	2	Pacemaker conversion	2
incessant	3	2 or more DCCV	3
Arrhythmia Severity		**Antiarrhythmic Drugs**	
asymptomatic	0	none or digoxin only	0
palpitations only	1	class II or IV	1
syncope/CHF/thrombosis	2	class I or III	2
cardiac arrest	3	amiodarone toxicity	3

*Triedman JK, Alexander ME, Love BA, Collins KK, Berul CI, Bevilacqua LM, Walsh EP. Influence of patient factors and ablative technologies on outcomes of radiofrequency ablation of intra-atrial re-entrant tachycardia in patients with congenital heart disease. Journal of the American College of Cardiology. 2002 Jun 5;39(11):1827-35.



Lateral Tunnel Fontan



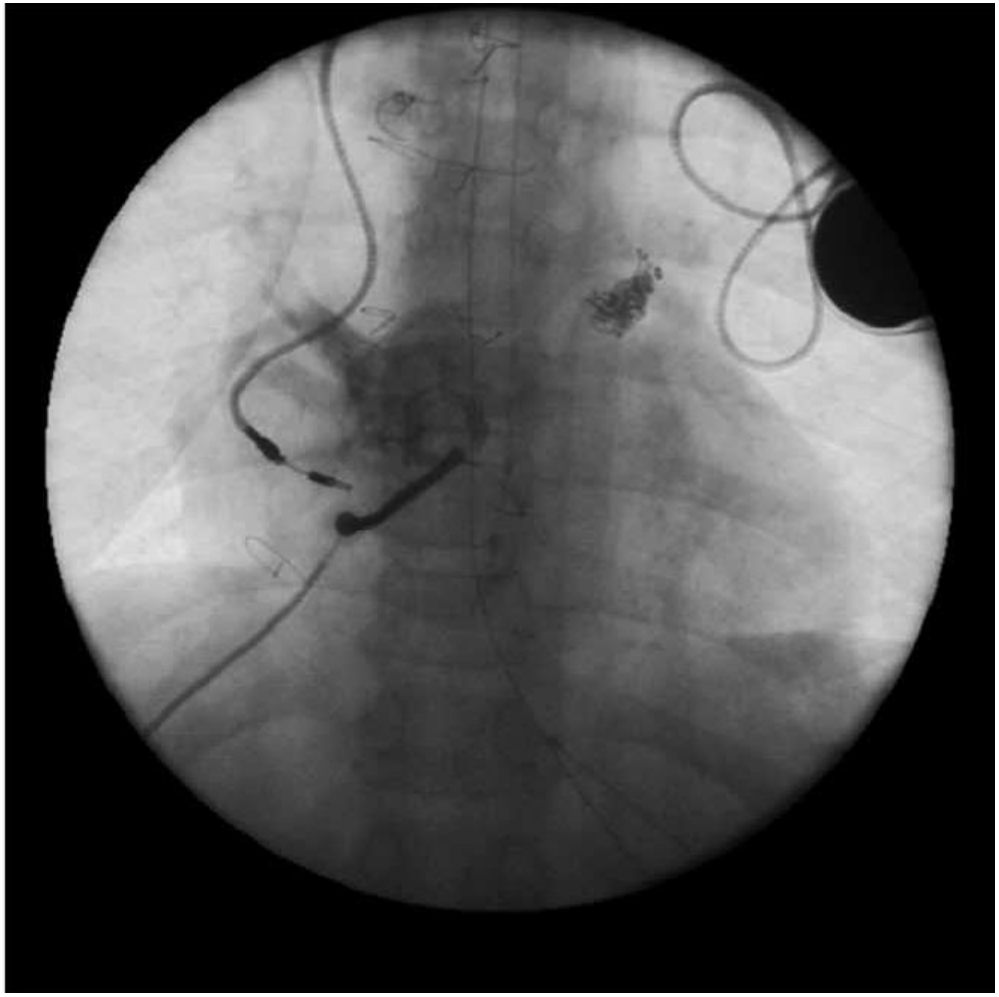
Extracardiac Fontan

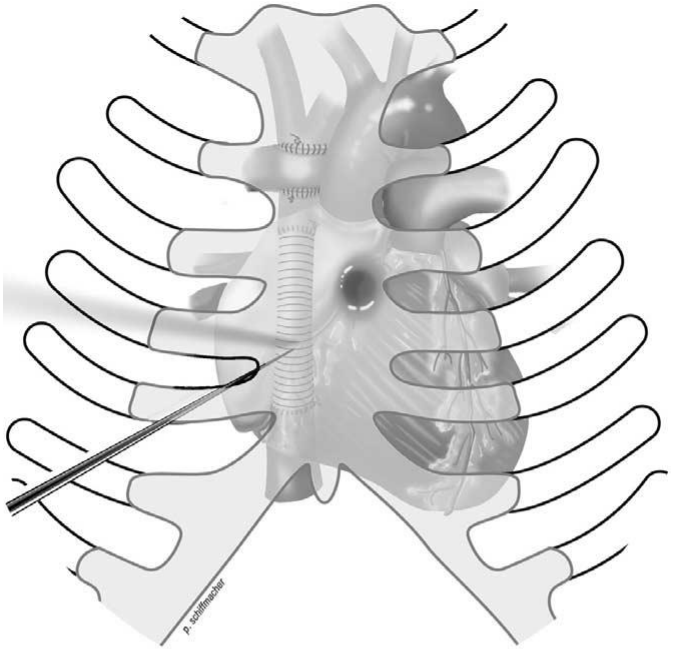
Kogon B et al Circulation. 2012;126:2511-2515

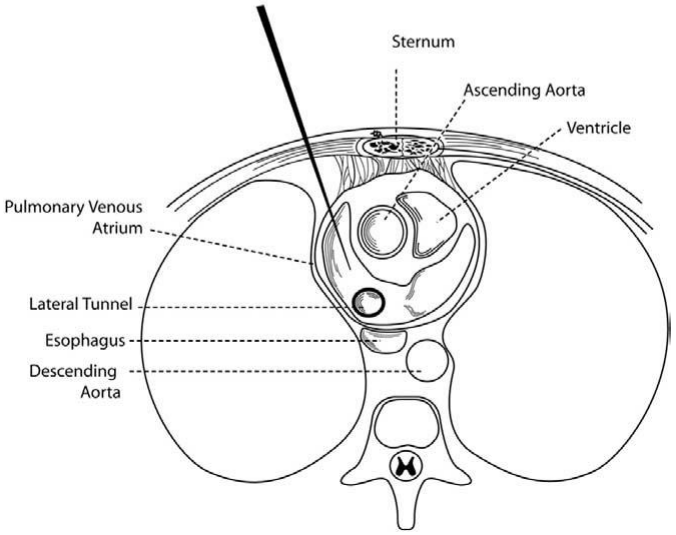
Outcomes

- 1) Acute procedural success
  - “Complete” = all induced arrhythmias ablated (none inducible end of case)
  - “Partial” = some, but not all, induced arrhythmias ablated (≥ 1 inducible end of case)
  - “Unsuccessful” = no arrhythmias able to be ablated
- 2) Complications
- 3) Clinical arrhythmia severity score at baseline, 3-6, 12 and 24 months
- 4) Freedom from death, transplant or conversion surgery composite

Transthoracic Puncture For Access to PV Atrium Lateral Tunnel Fontan

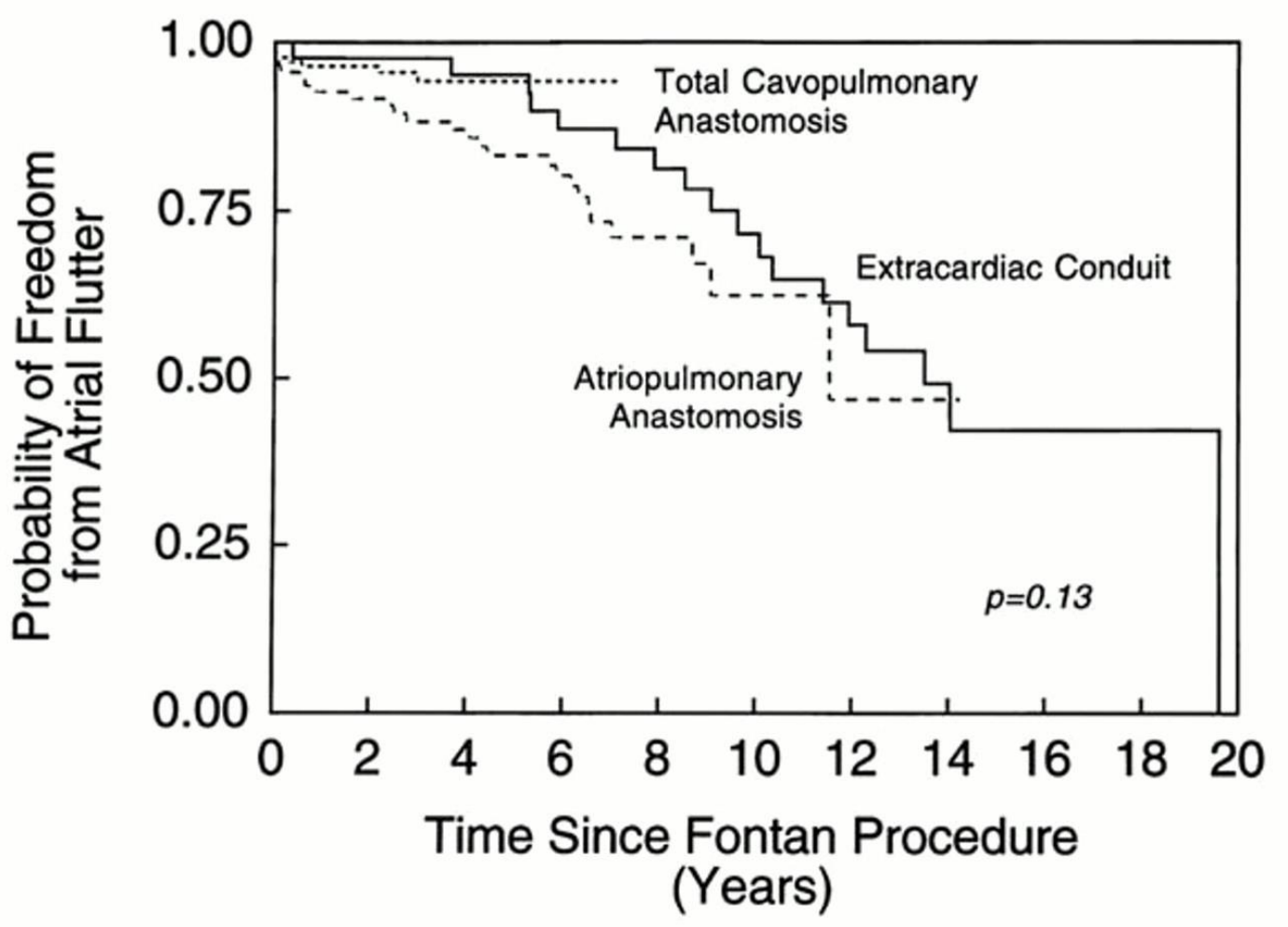






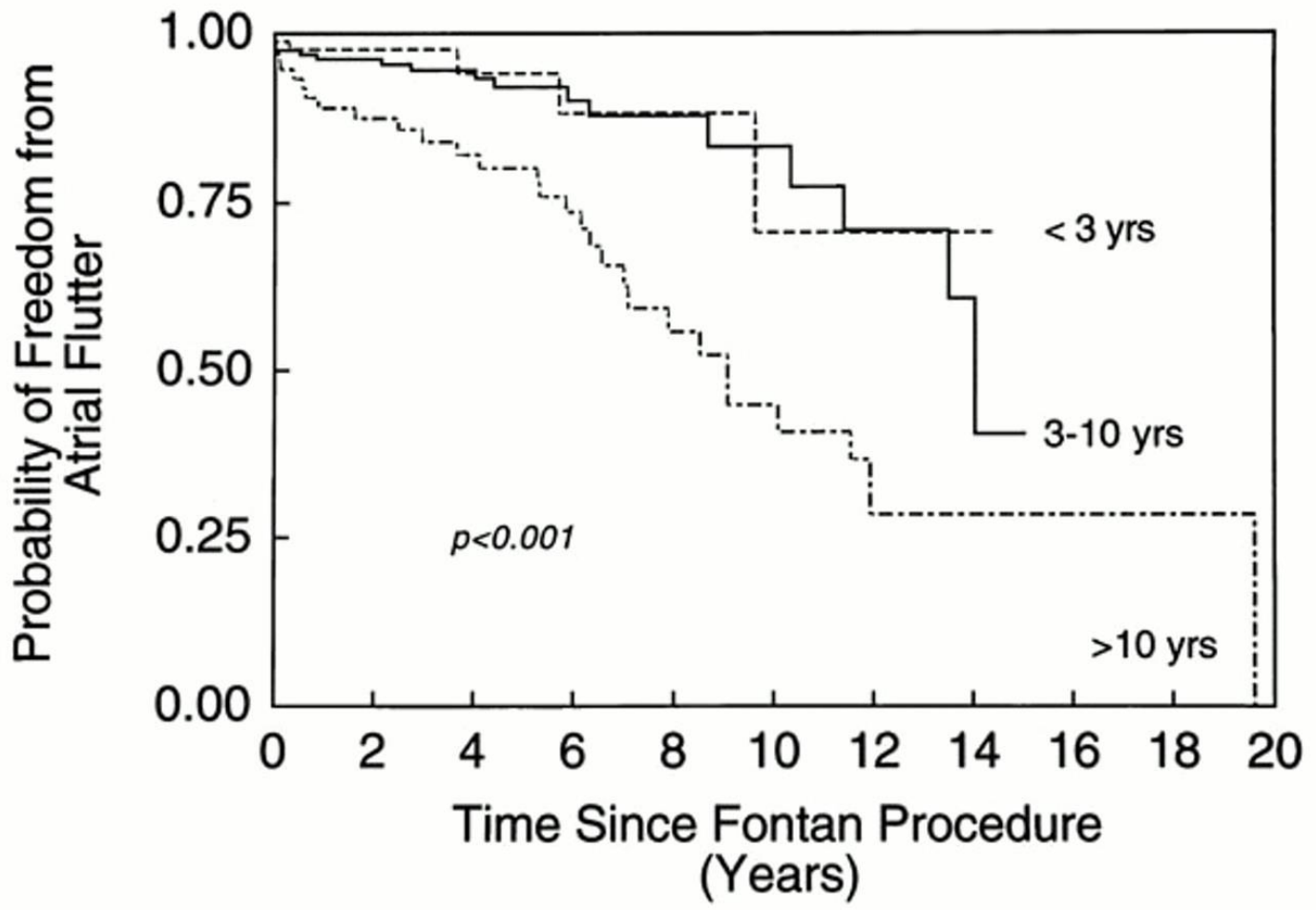
Nehgme, RA (Heart Rhythm 2006;3:37– 43)

ATRIAL FLUTTER AFTER FONTAN (N:334)
Risk Depends on Procedure Type



Fishberger S. B. et al.; J Thorac Cardiovasc Surg 1997;113:80-86

ATRIAL FLUTTER AFTER FONTAN



Fishberger S. B. et al.; J Thorac Cardiovasc Surg 1997;113:80-86

REPAIRED CONGENITAL HEART DISEASE
A Perfect Storm For Arrhythmia

Substrate

- Scarring and fibrosis
- Surgical incisions and suture lines
- Atrial dilatation
- Myocyte stretch
- Hypertrophy
- Cellular hypoxia

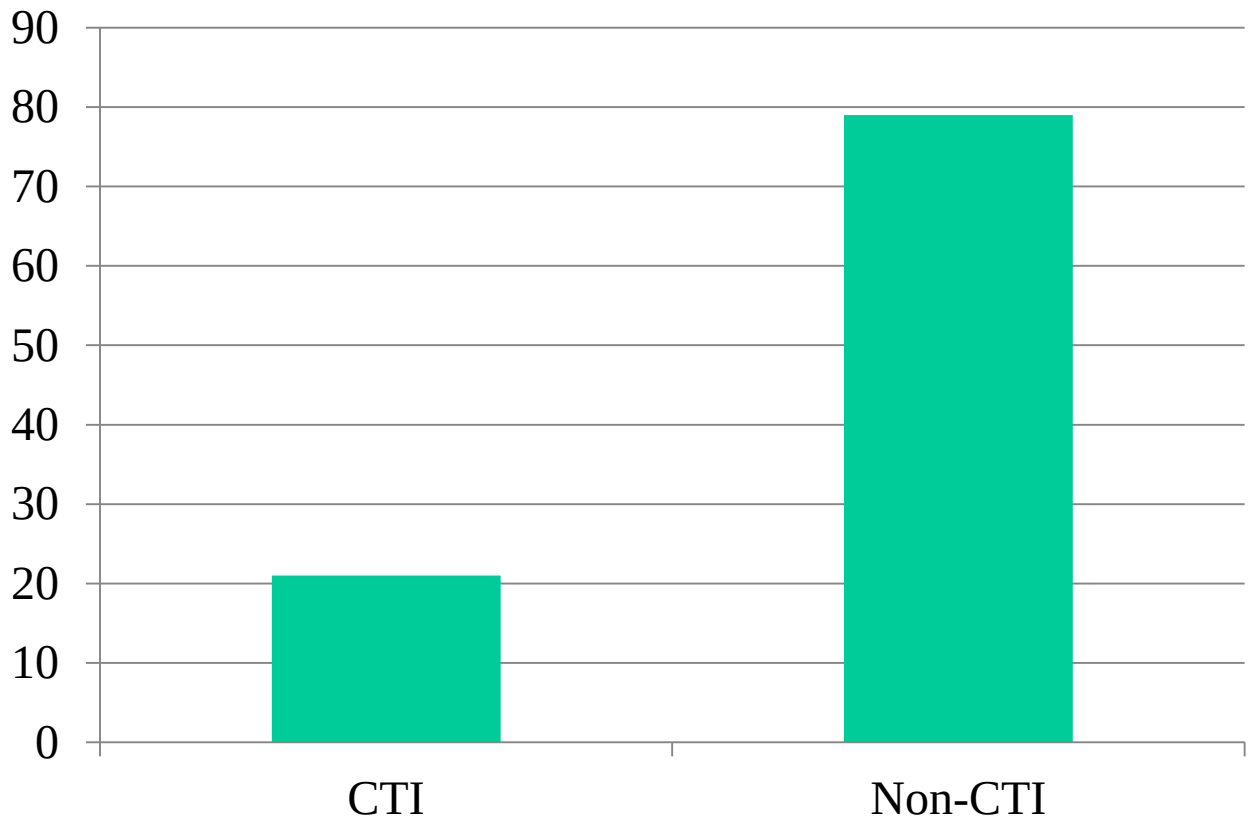
Preparation For Ablation

- Obtain surgical report
- Imaging: CT, CMRI, TTE, TEE, venography
- Assess hemodynamic status
- Plan vascular approach: femoral or subclavian / jugular?
- Plan chamber access:
  - Retrograde aortic
  - Fenestration
  - Puncture baffle
  - Transthoracic puncture
  - Surgical assistance – hybrid access
- Plan for heart block & need for pacing in Fontan

Equipment for Ablation in Fontan and Atrial Switch

- 3D mapping
- Deflectable sheaths
- Irrigated ablation catheters
- Contact force?
- “Adequate” number catheters / electrodes
- TEE
- ICE?
- Magnetic navigation?

Site of IART in Fontan Patients
N= 19



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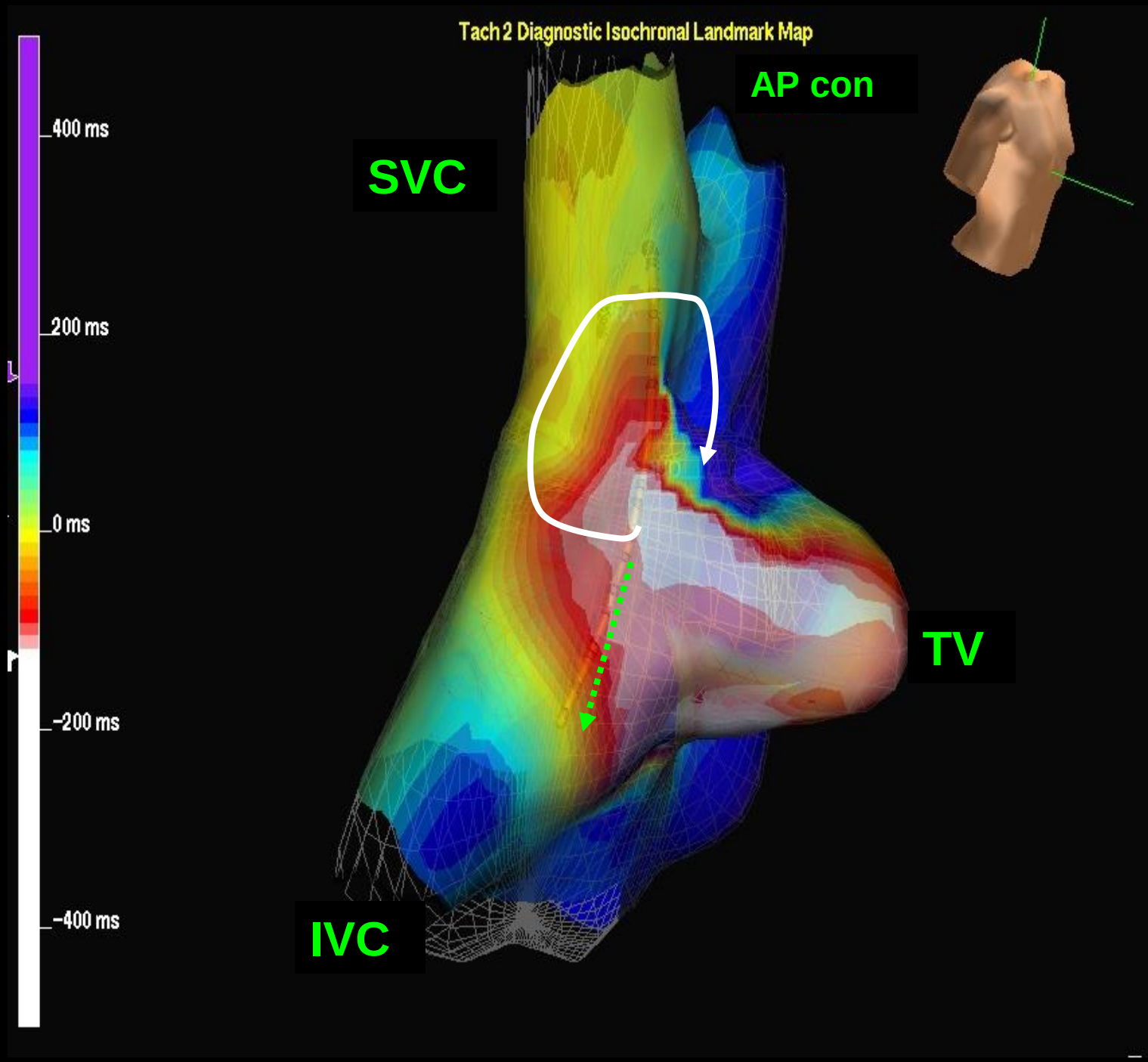
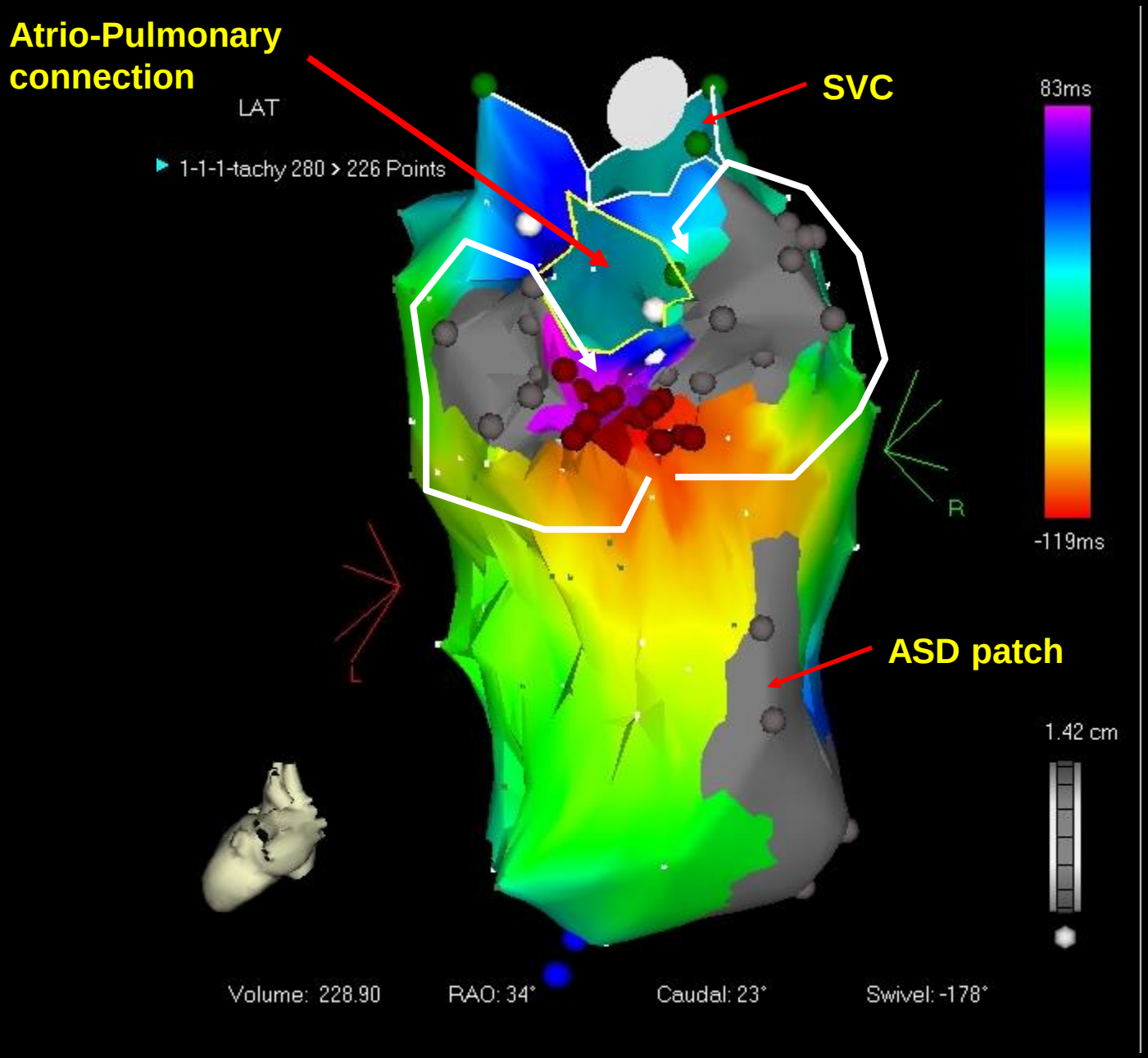
Mr DP 22
Fontan
Tricuspid atresia

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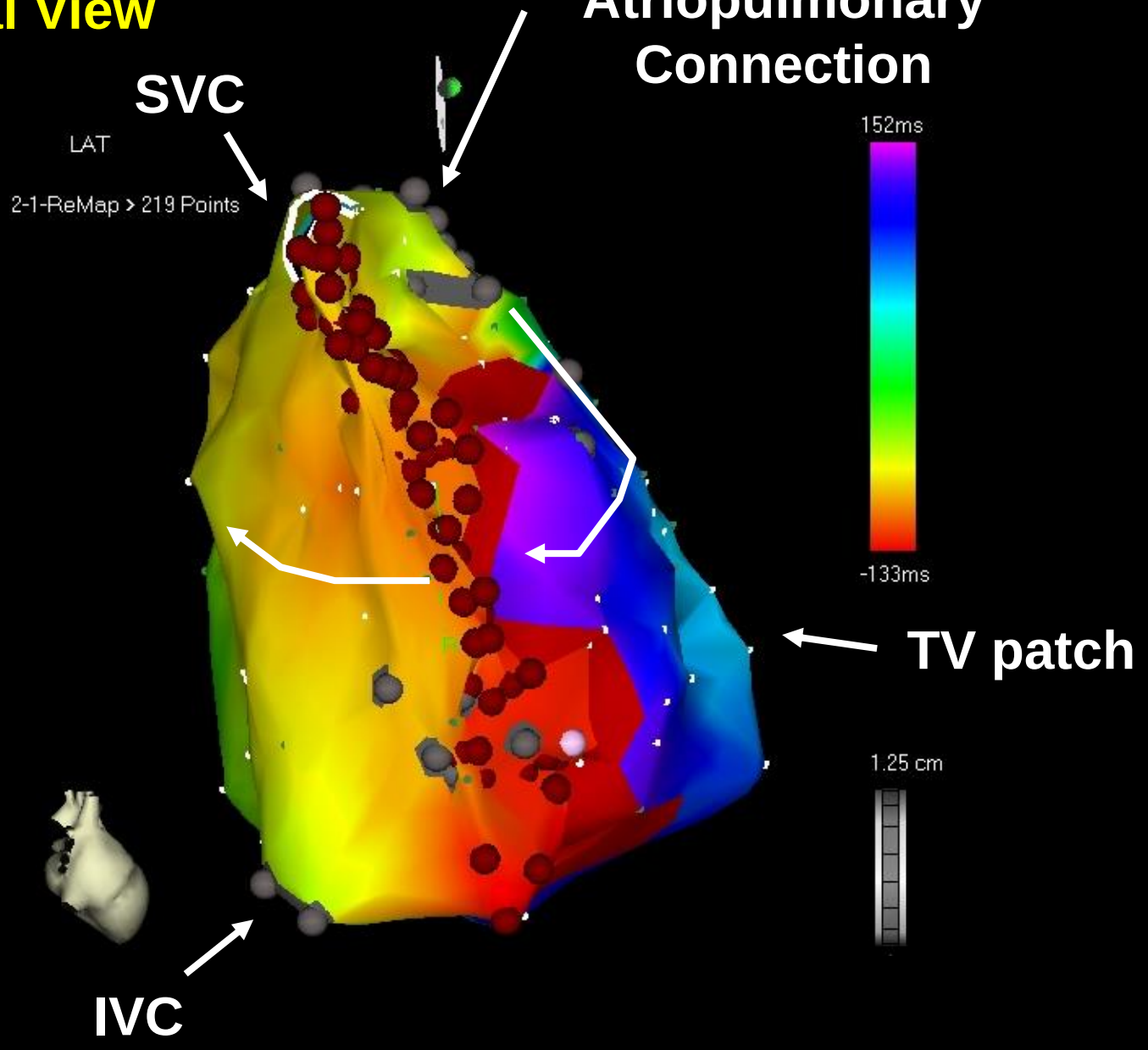
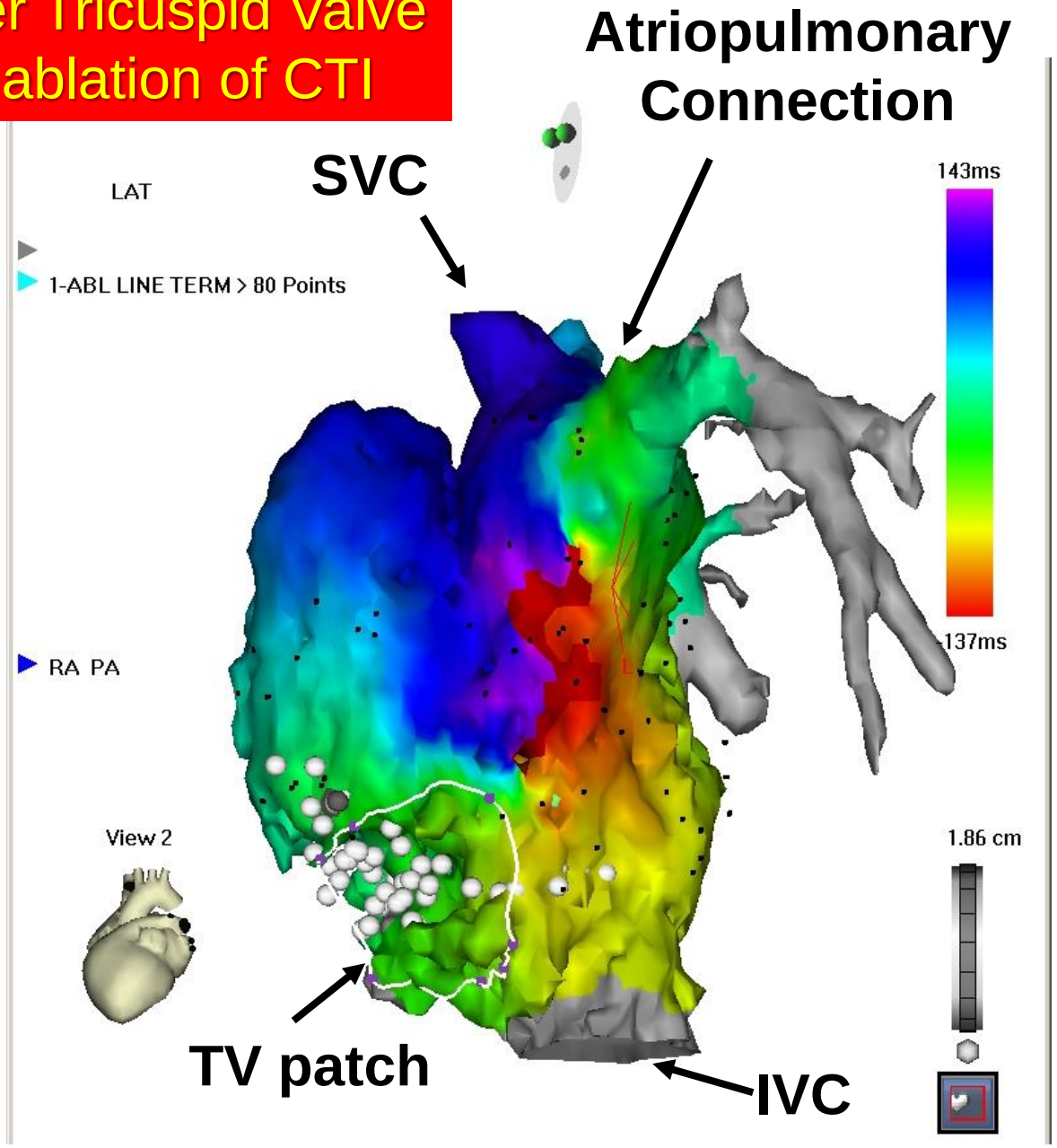
SB Fontan
Right Lateral View



Patch Over Tricuspid Valve Hindering ablation of CTI

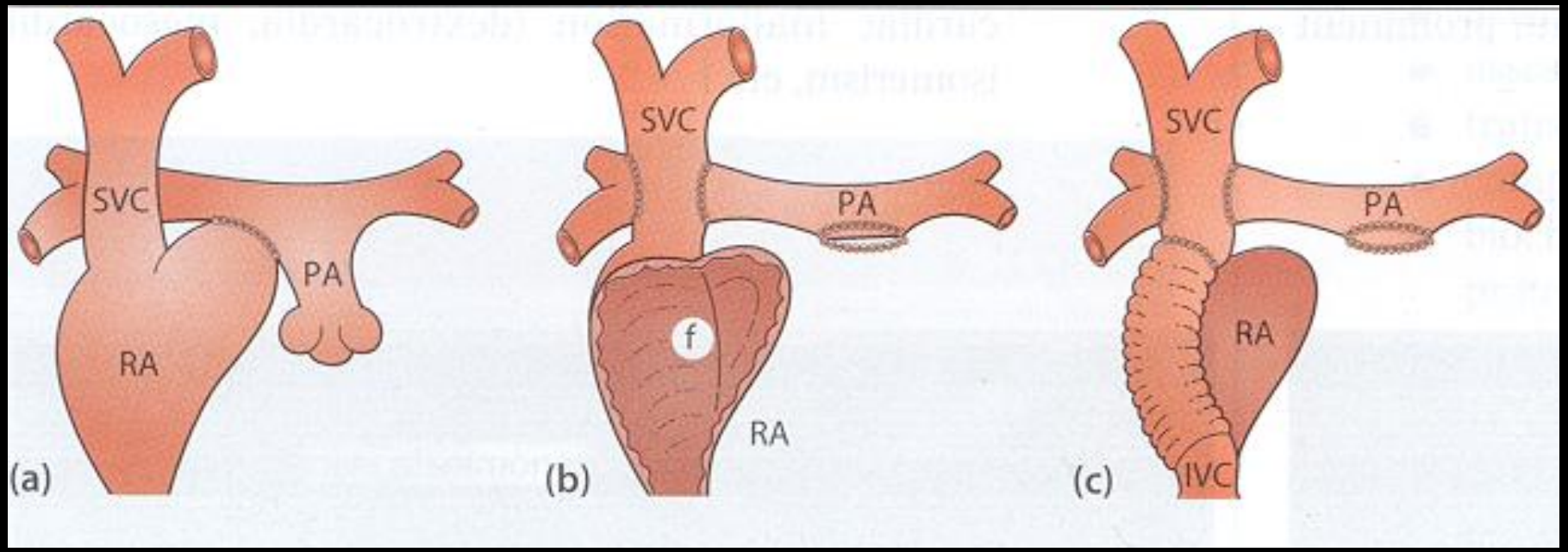


Ablation Adult Congenital Heart Disease

Tips

- Use entrainment judiciously
- Check frequently that Tc has not changed
- Anaesthetist (anesthesiologist)

Fontan Procedures



Atriopulmonary Connection c.1968

Lateral Tunnel (TCPC) 1987

Extracardiac 1990

Risk of Arrhythmia Post Fontan (n=520)

Risk Depends on Type of Procedure

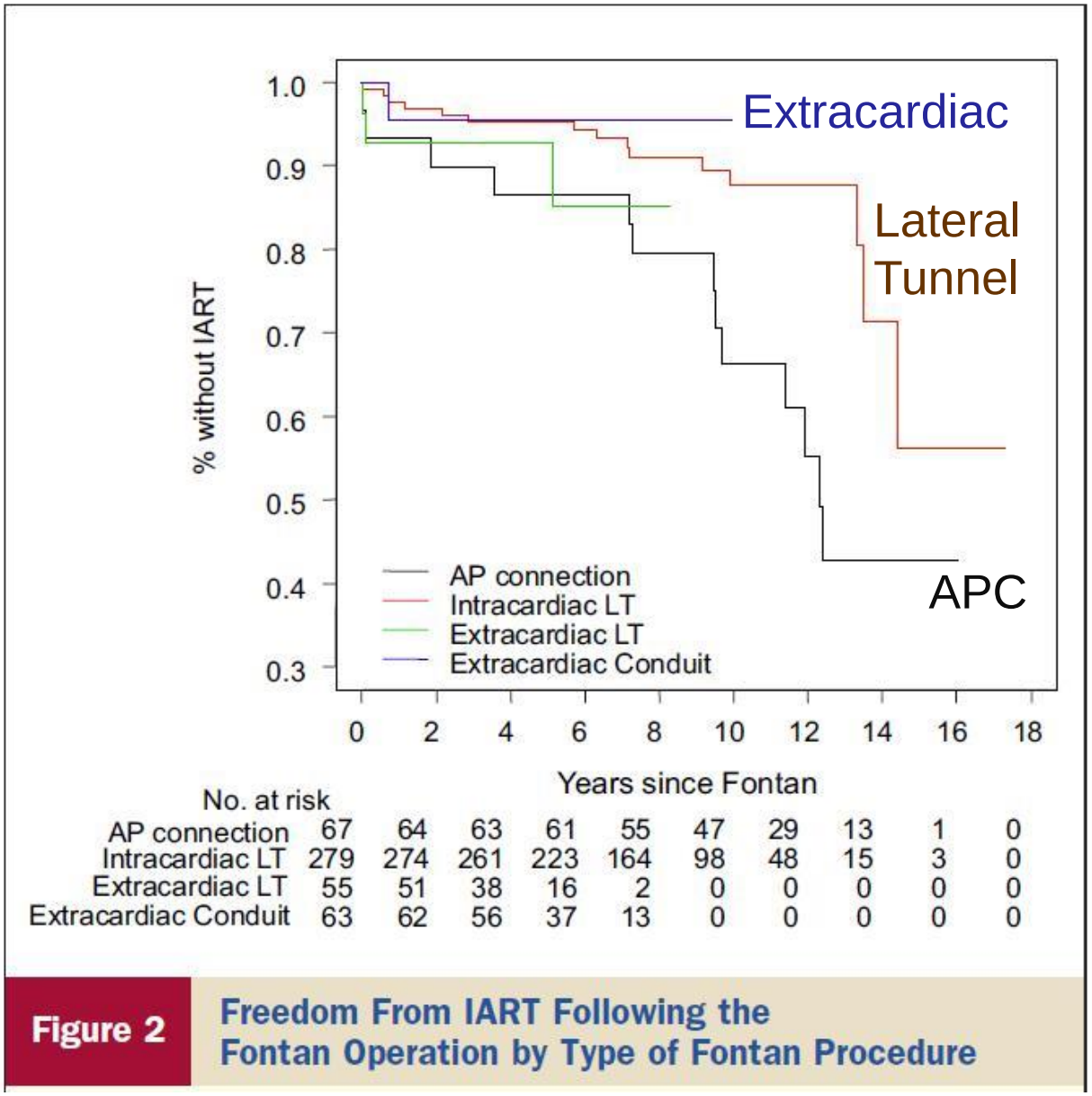


Figure 2 Freedom From IART Following the Fontan Operation by Type of Fontan Procedure

Stephenson EA et al
J Am Coll Cardiol
2010;56:890–6

Baffle Puncture

Baffle Materials

- Pericardium
- Cardiac flap
- Synthetic: PETE (Dacron™), PTFE (Teflon™, Gore-Tex™)

Methods

- Direct puncture with trans-septal needle / sheath
- Adjunctive radiofrequency energy
- Proprietary RF needle
- Balloon angioplasty to enlarge puncture

Tip*: Very stiff mitral valvuloplasty wire may aid access

Adverse Events Associated with Trans Baffle Access

Boston Children's Hospital

Puncture n=54, Baffle leak/fenestration n= 20

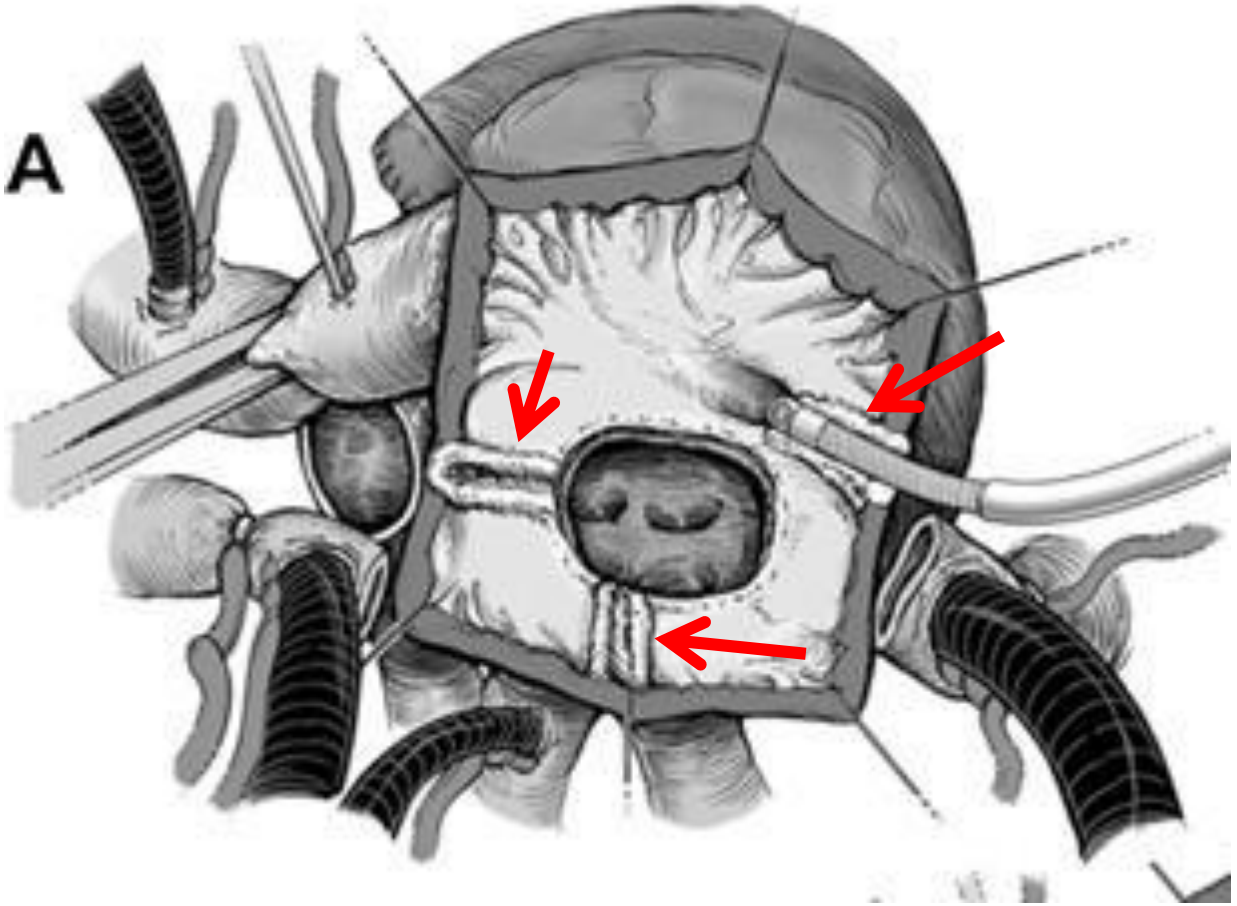
Table 6. Adverse Events

Severity	Adverse Event	TBA	No TBA	*P* Value
Catastrophic	Death	2	0	—
Major	Shunts and cyanosis	2	0	—
	Permanent AV block	1	0	—
	CS thrombosis	0	1	—
	Retroperitoneal bleed	1	0	—
Moderate	Pseudoaneurysm	1	0	—
	Acute kidney injury	0	3	0.05
	Hemoptysis	1	0	—
All	Total>minor adverse events	8	4	1

AV indicates atrioventricular; CS, coronary sinus; TBA, transbaffle access.

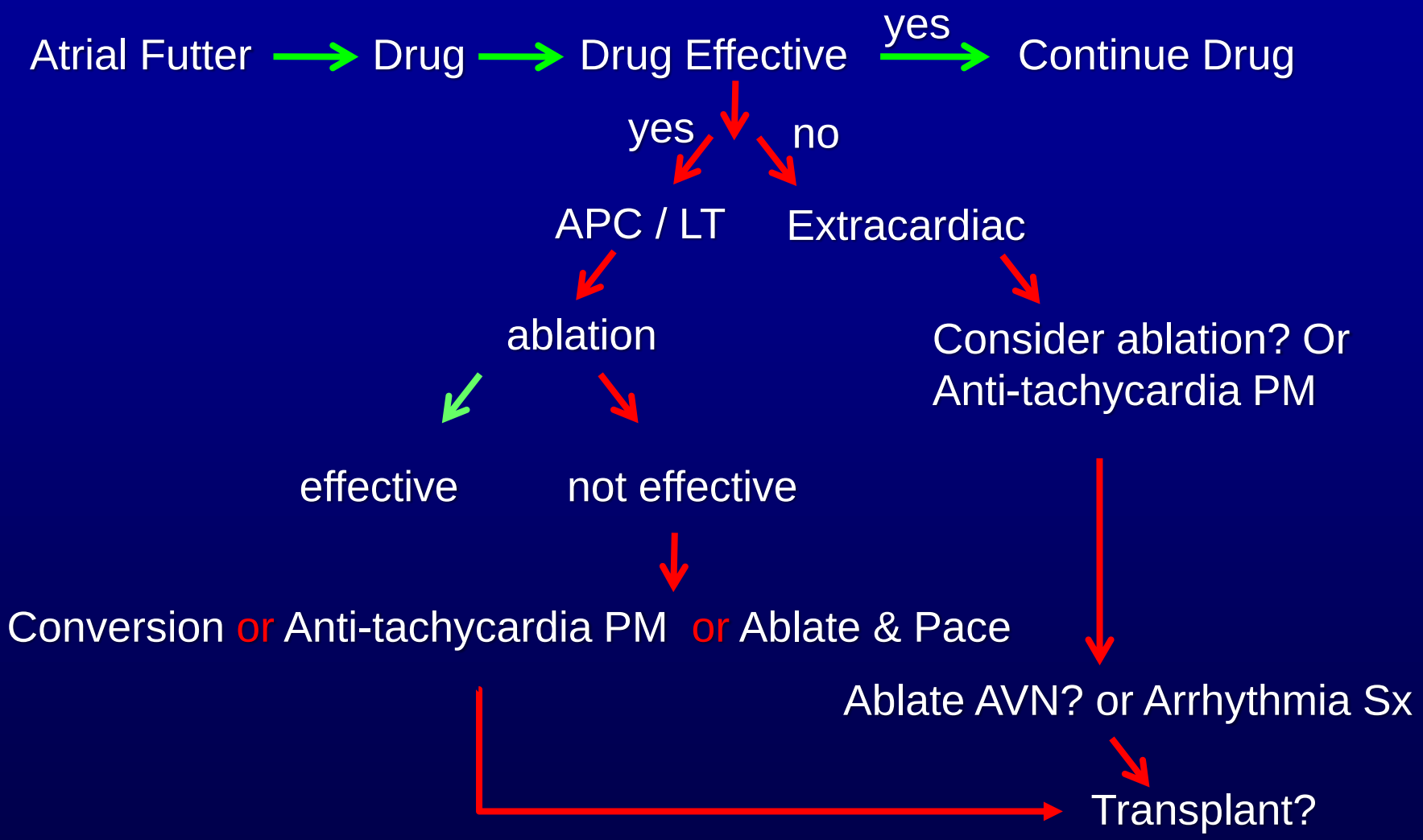
Correa R et al: J. Am Heart Assoc 2013;2:e000325

Right Atrial Maze Procedure During Fontan Conversion



Mavroudis C, Eur J Cardiothorac Surg 2008;34:1034-1040

Arrhythmia After Fontan with Good Haemodynamics



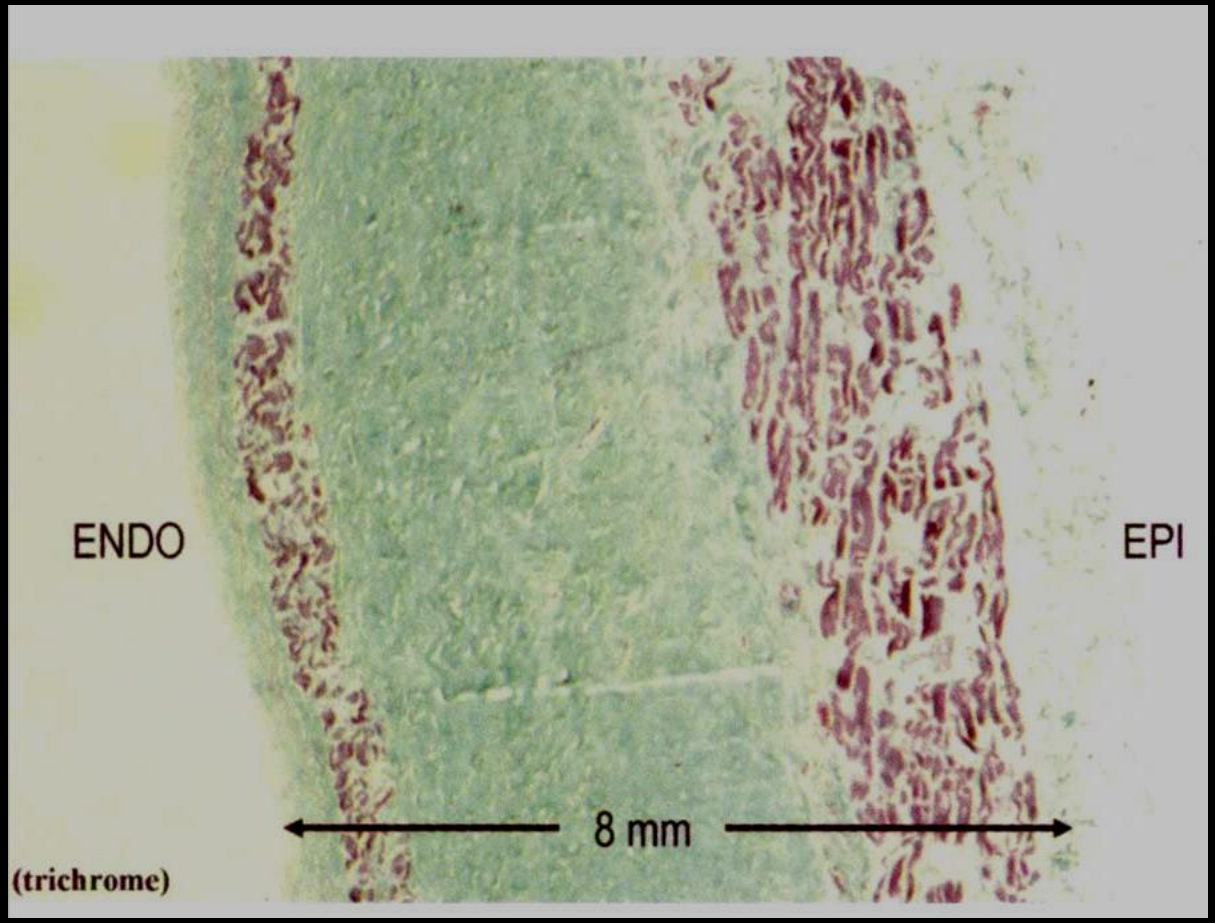
ABLATION POST FONTAN

- Challenging
- Preparation & planning key to success & avoiding bad outcomes
- Modern equipment/technique facilitates ablation
- Quality of life significantly improved
- Significant risk of late arrhythmias

TITLE

TITLE

Right Atrial Wall Post Fontan



EP Walsh

Extracardiac Fontan

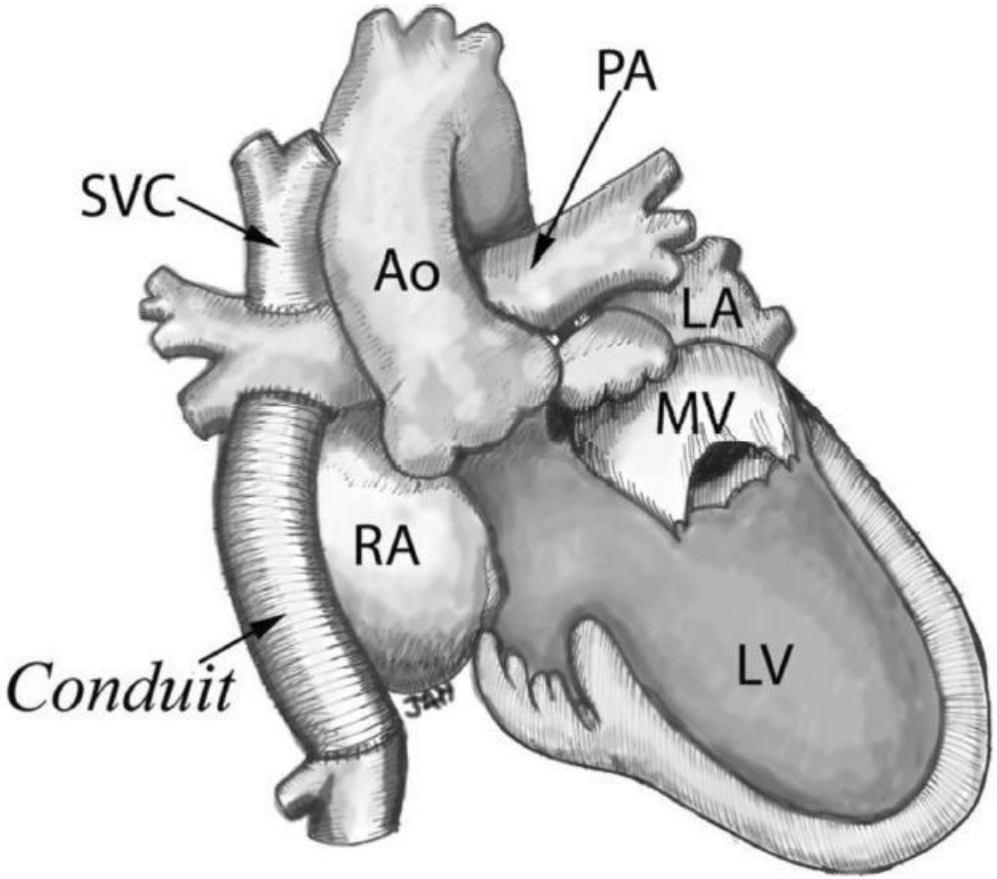
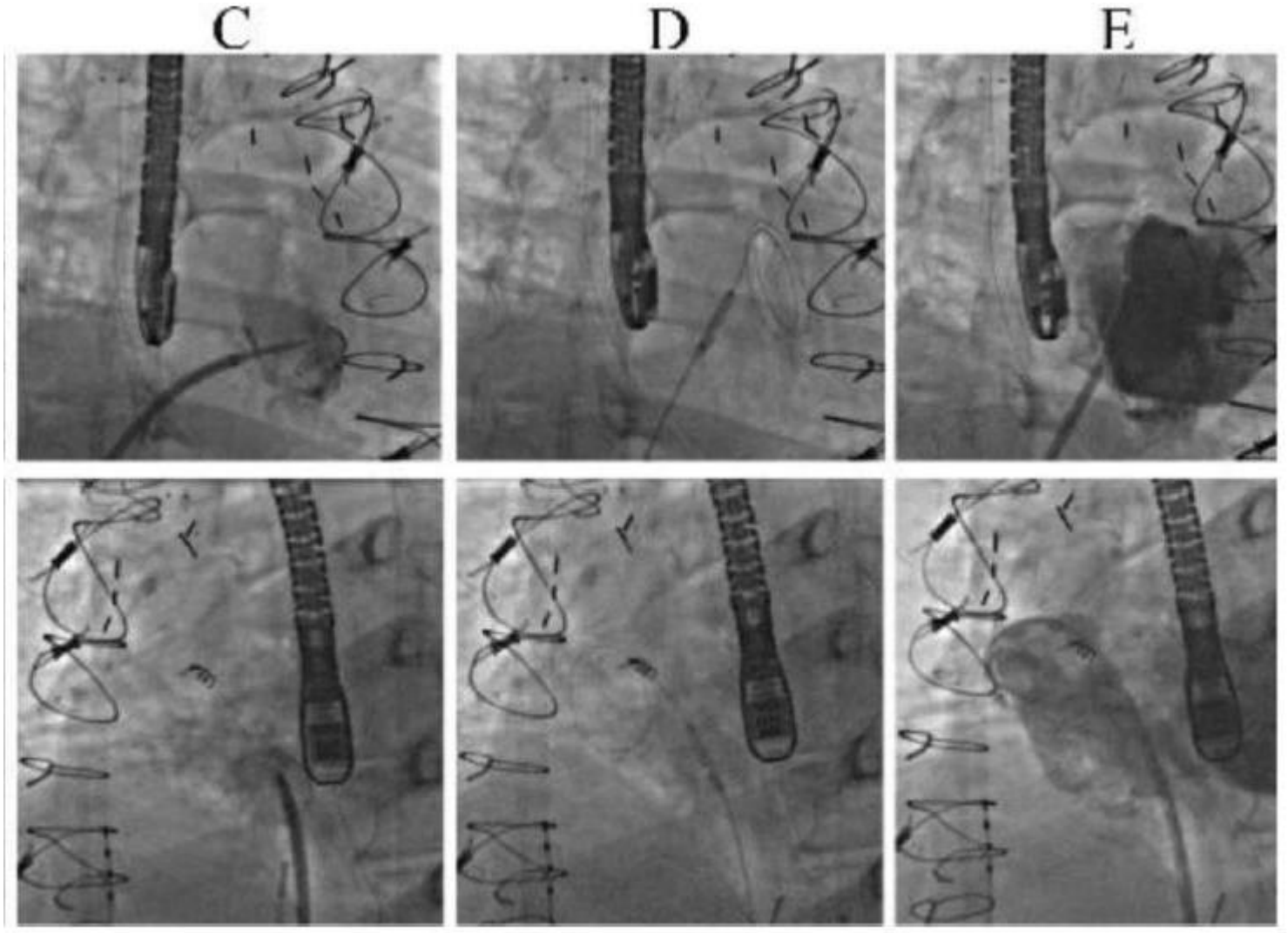


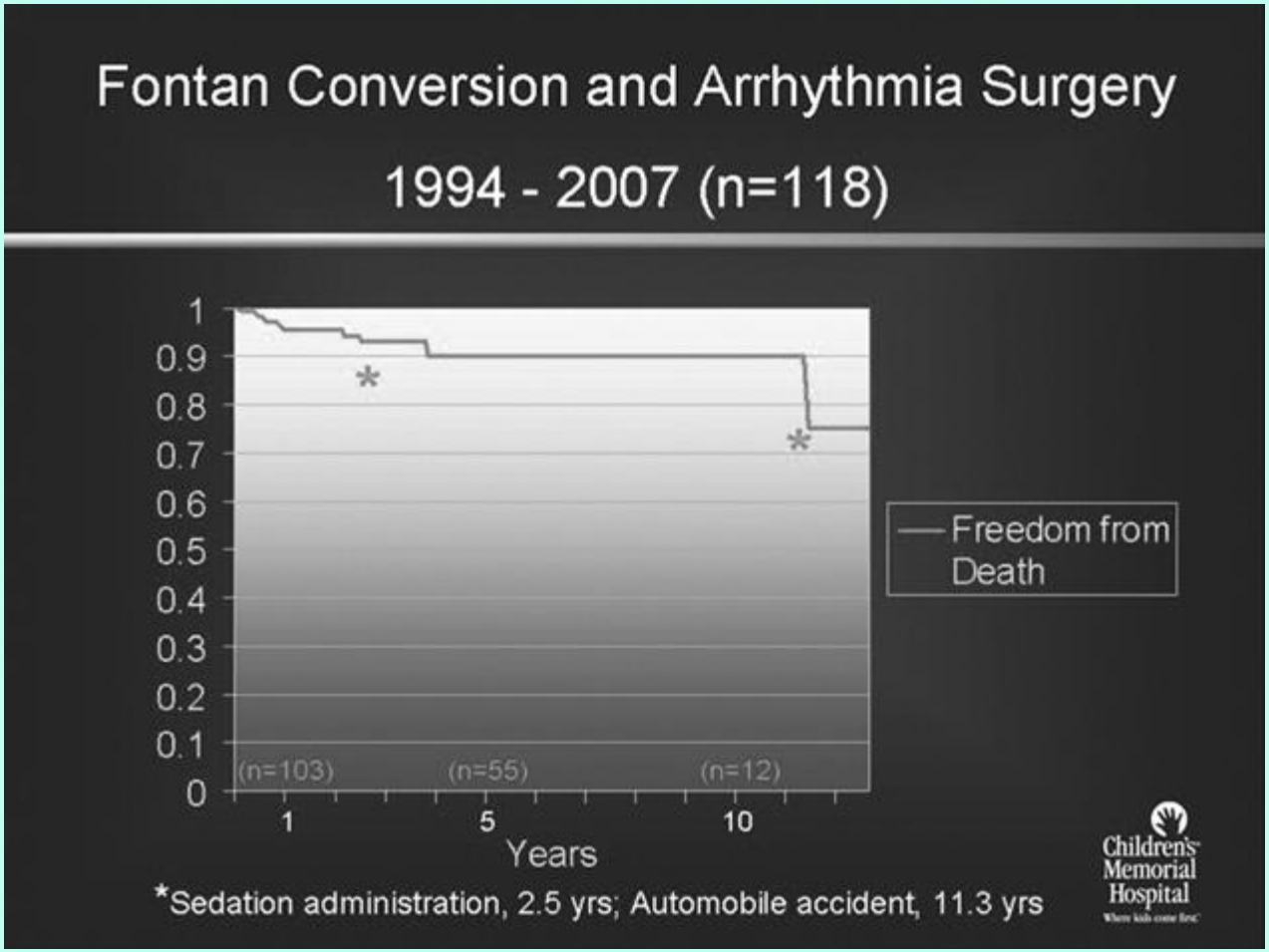
Figure 1. Rendering of post-surgical anatomy
There is tricuspid valve atresia with hypoplastic right ventricle and well-developed systemic left ventricle (LV). Great arteries are D-transposed with the aorta (Ao) anteriorly dextraposed and the banded pulmonary artery (PA) posteriorly levoposed. Superior vena cava (SVC) is connected to PA via bidirectional Glenn shunt. Extracardiac conduit connects inferior vena cava to PA. Note close proximity of extracardiac conduit to "right atrial" portion (RA) of pulmonary venous atrium. The "left atrium" (LA) and mitral valve (MV) are labeled.

Dave, A: Heart Rhythm. 2010 ; 7(3): 413–416

Extracardiac Fontan: Access To RA : Conduit Puncture



Dave, A: Heart Rhythm. 2010 ; 7(3): 413–416



Mavroudis C, Eur J Cardiothorac Surg 2008;34:1034-1040

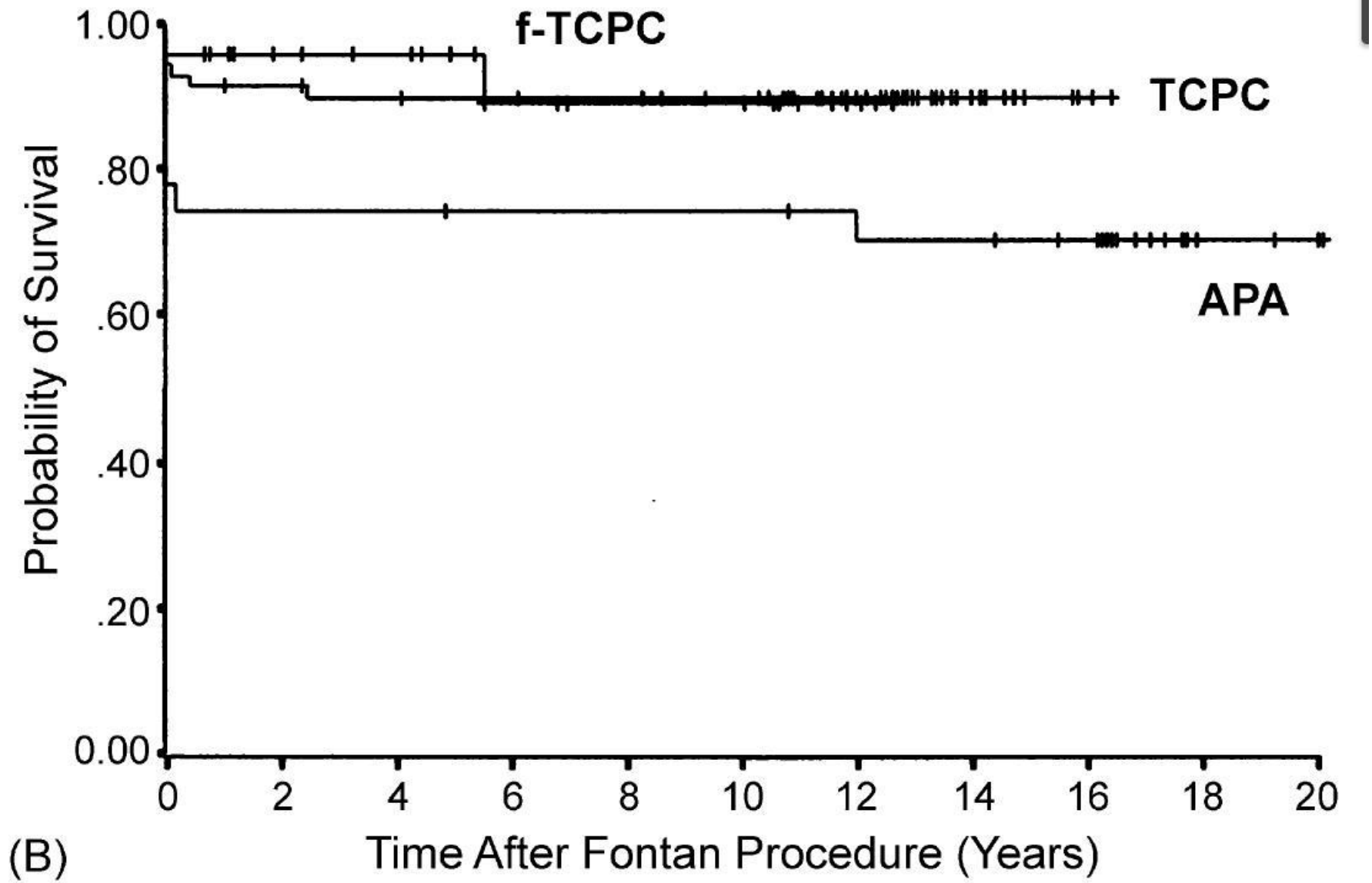
ATRIAL ARRHYTHMIAS AFTER SURGICAL REPAIR

Lesion / Repair	Incidence
Fontan[1,2]	41-50%
Mustard / Senning[3]	30%
TOF[4]	34%

1: Fishberger S. J Thoracic Cardiovasc Surg 1997;113:80
2: Ghai A. JACC 2001;37:585
3: Flinn CJ. NEJM 1984;310:1635
4: Roos-Hesselink. Circulation 1995;91:2214

Fenestration Effect on Survival?

Ono et al: Eur J Cardiothorac Surg 2006;30:923



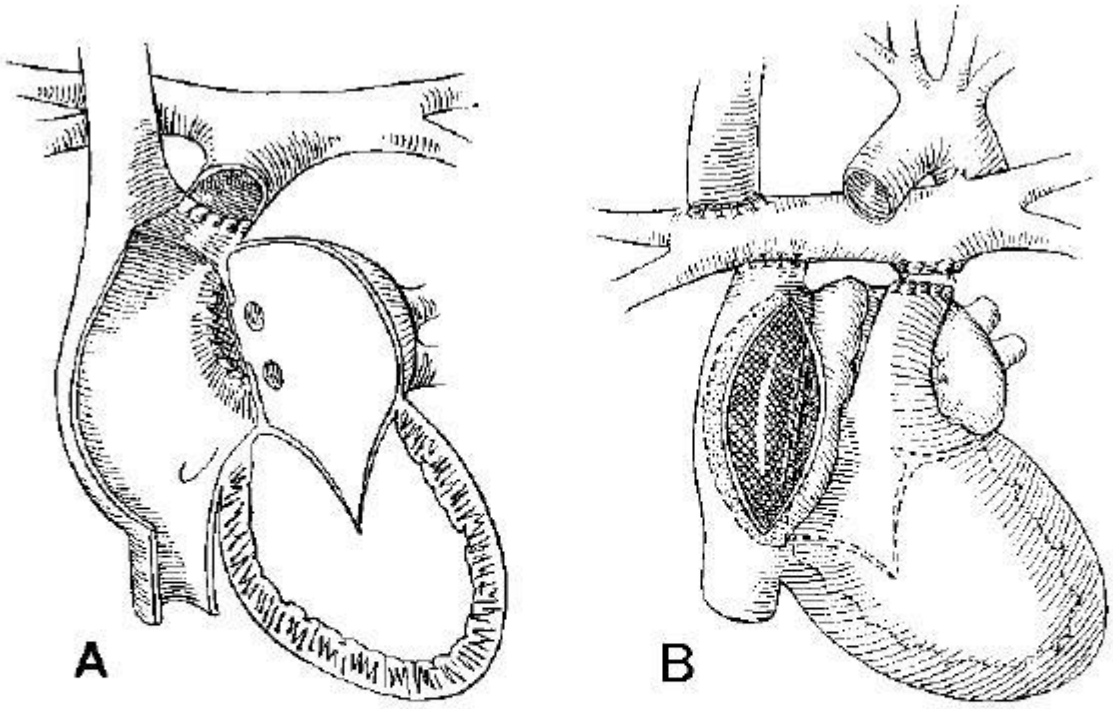


Figure 1. Two types of modified Fontan connections are shown. A, Atriopulmonary connection. B, Total cavopulmonary connection. See text for description. (Figure is used with permission of the Mayo Foundation.)

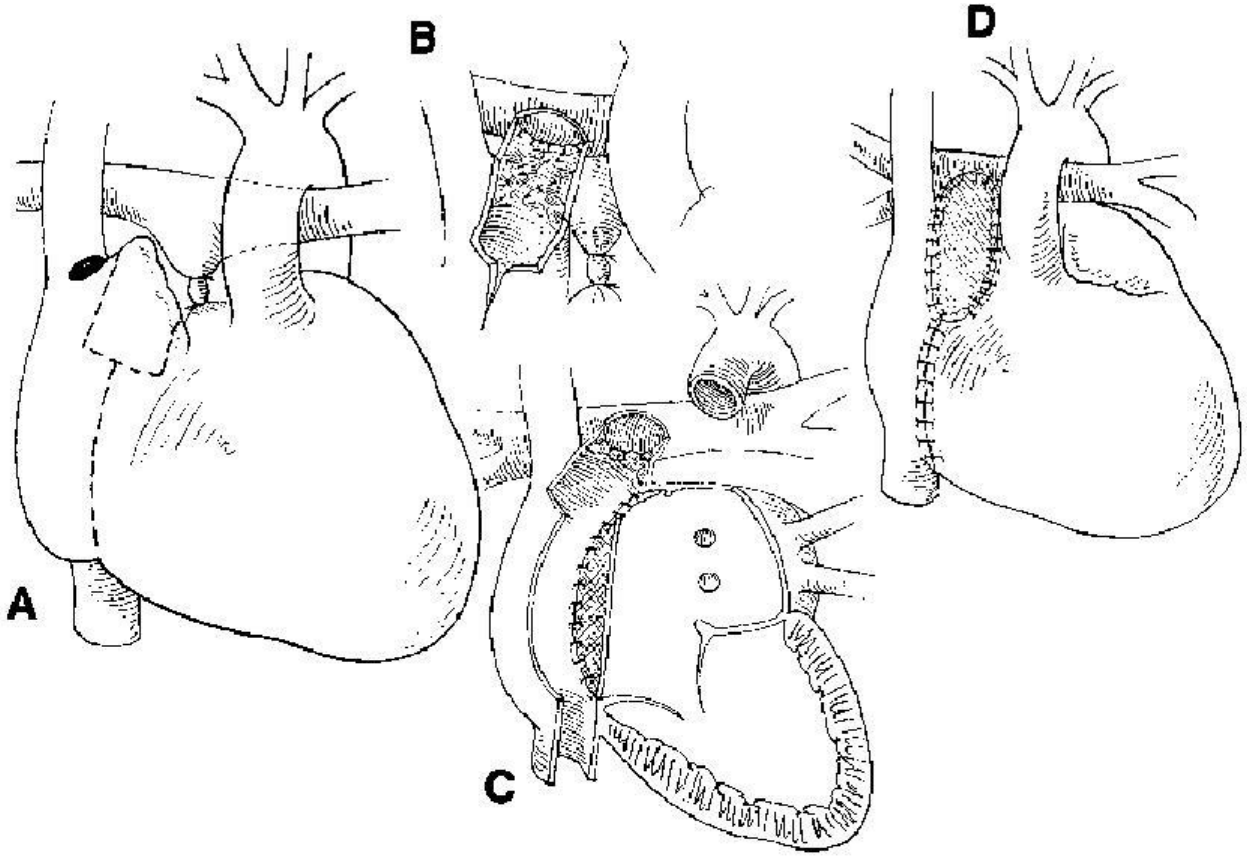


Figure 2. Surgical technique for APC-lat modification. A, Incisions in the free wall of right atrium and atrial appendage. B, Anastomosis of flap of atrial appendage to pulmonary artery. C, Intra-atrial baffle inserted to direct blood from inferior vena cava to atrial appendage–pulmonary artery connection. D, Pericardial patch to complete the connection. (Figure is used with permission of the Mayo Foundation.)

Extracardiac Fontan: Access To RA : Conduit Puncture

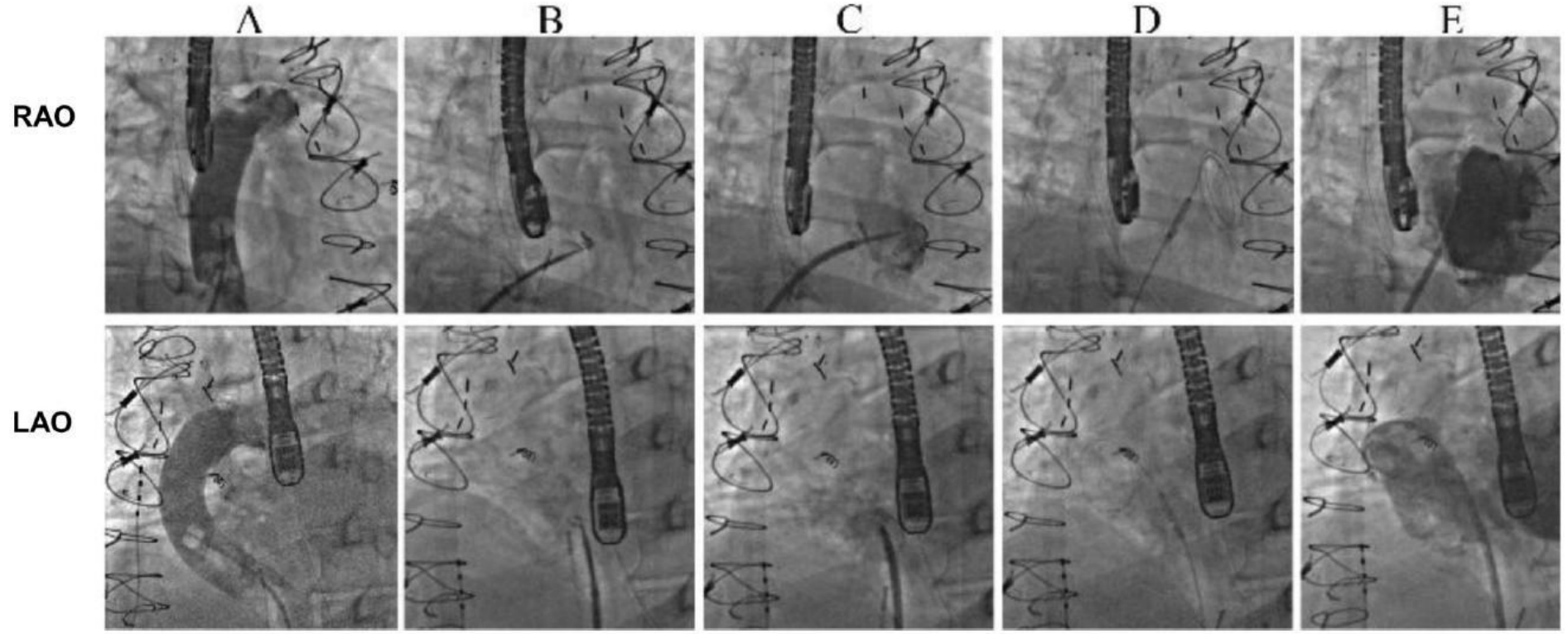


Figure 2.
Fluoroscopic images showing (A) still-frame from conduit angiogram, (B) transseptal needle with contrast staining of conduit prior to crossing, (C) dilator of transseptal sheath crossing conduit into pulmonary venous atrium, (D) Toray valvuloplasty wire coiled in atrium with angioplasty balloon inflated across conduit crossing, and (E) still-frame from angiography of pulmonary venous atrium.

Dave, A: Heart Rhythm. 2010 ; 7(3): 413–416

REPAIRED CONGENITAL HEART DISEASE
A "Perfect Storm" For Arrhythmia

Substrate

- Scarring and fibrosis
- Surgical incisions and suture lines
- Atrial dilatation – bigger chambers
- Myocyte stretch
- Hypertrophy & heart failure
- Cellular hypoxia

ARRHYTHMIAS POST FONTAN

"Late" Arrhythmias

Bradycardia:

- Sinus node dysfunction
- Intra-atrial block

Tachycardia:

- Intra-atrial reentry (atrial flutter)
- Standard SVT (AVNRT & accessory pathways)
- Atrial fibrillation (following AV valve regurgitation)
- Ventricular arrhythmias with ventricular failure

ARRHYTHMIAS POST FONTAN

Bradycardia

- Sinus node dysfunction but also intra-atrial block
- Probable cause: sinus node fibrosis +/- ischaemia
- Incidence: highest LT, intermediate APC, lowest for extracardiac

Fontan Conversion – Arrhythmia Surgery– Pacemaker Implantation

Epicardial Leads

- Placement difficult because of adhesions
- Bipolar – required for anti-tachycardia pacemakers
- Both sides of ventricular mass for CRT?

Pacing Ventricle Via Coronary Sinus - APC Fontan

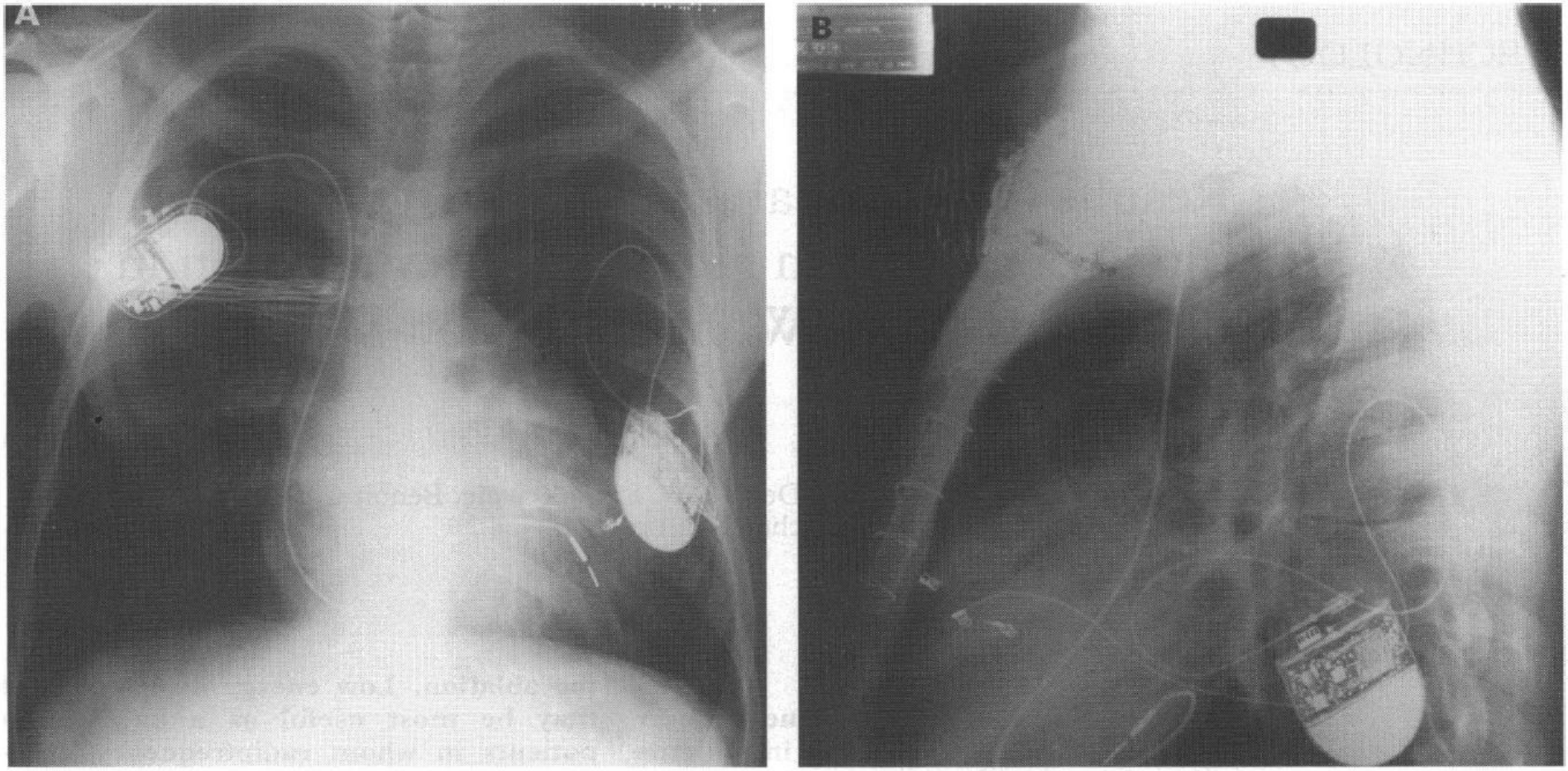
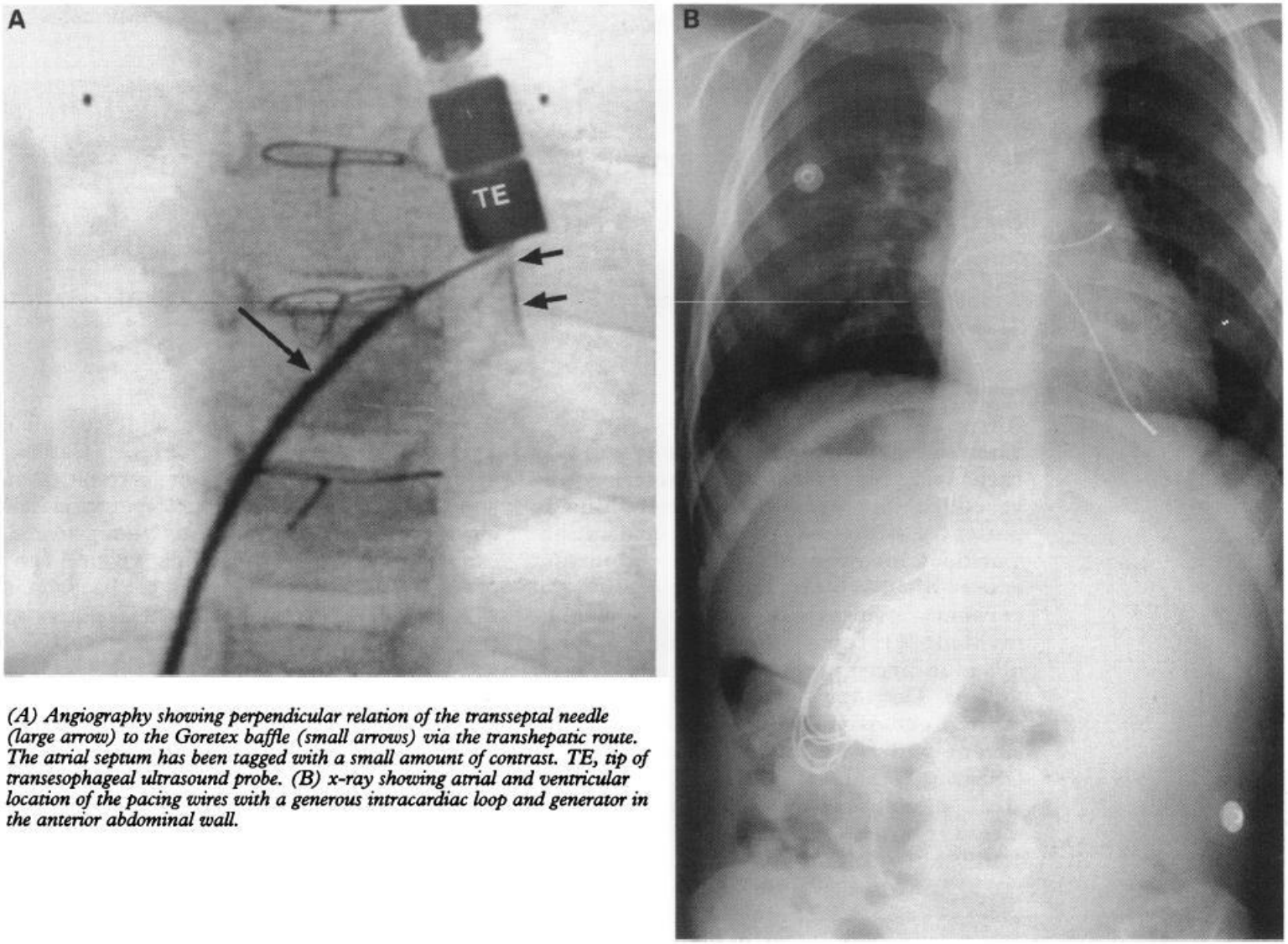


Figure 2 Chest radiographs in posteroanterior (A) and lateral (B) projections, after successful ventricular pacing through the coronary sinus. The redundant epicardial system is still in situ.

Blackburn M (Br HeartJ 1993;70:578-579)

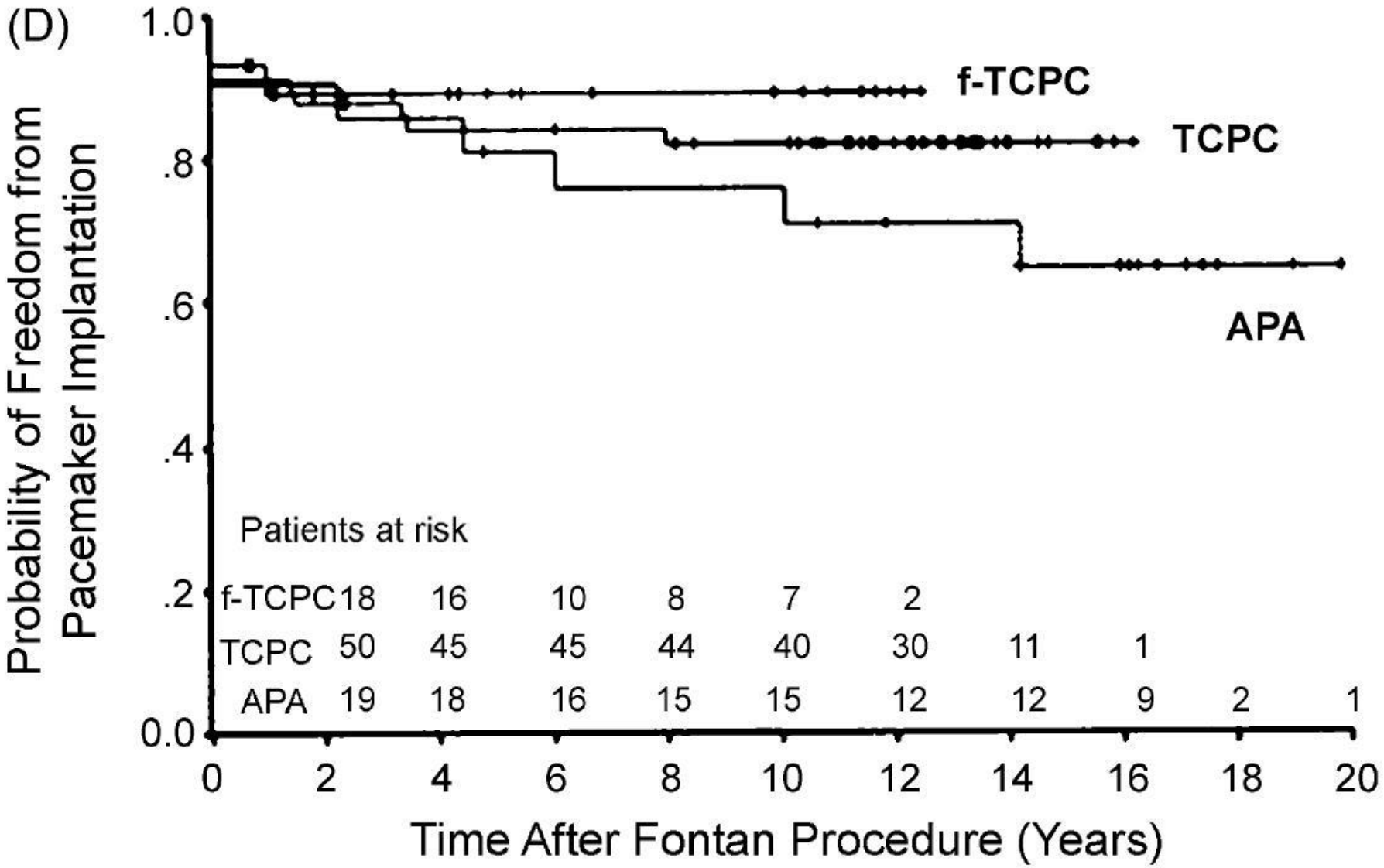
Pacing Via Hepatic Veins After LT Fontan



(A) Angiography showing perpendicular relation of the transseptal needle (large arrow) to the Goretex baffle (small arrows) via the transhepatic route. The atrial septum has been tagged with a small amount of contrast. TE, tip of transesophageal ultrasound probe. (B) x-ray showing atrial and ventricular location of the pacing wires with a generous intracardiac loop and generator in the anterior abdominal wall.

Fenestration (lateral tunnel) No Effect on Need for PPM?

Ono et al: Eur J Cardiothorac Surg 2006;30:923



TACHYCARDIA POST FONTAN

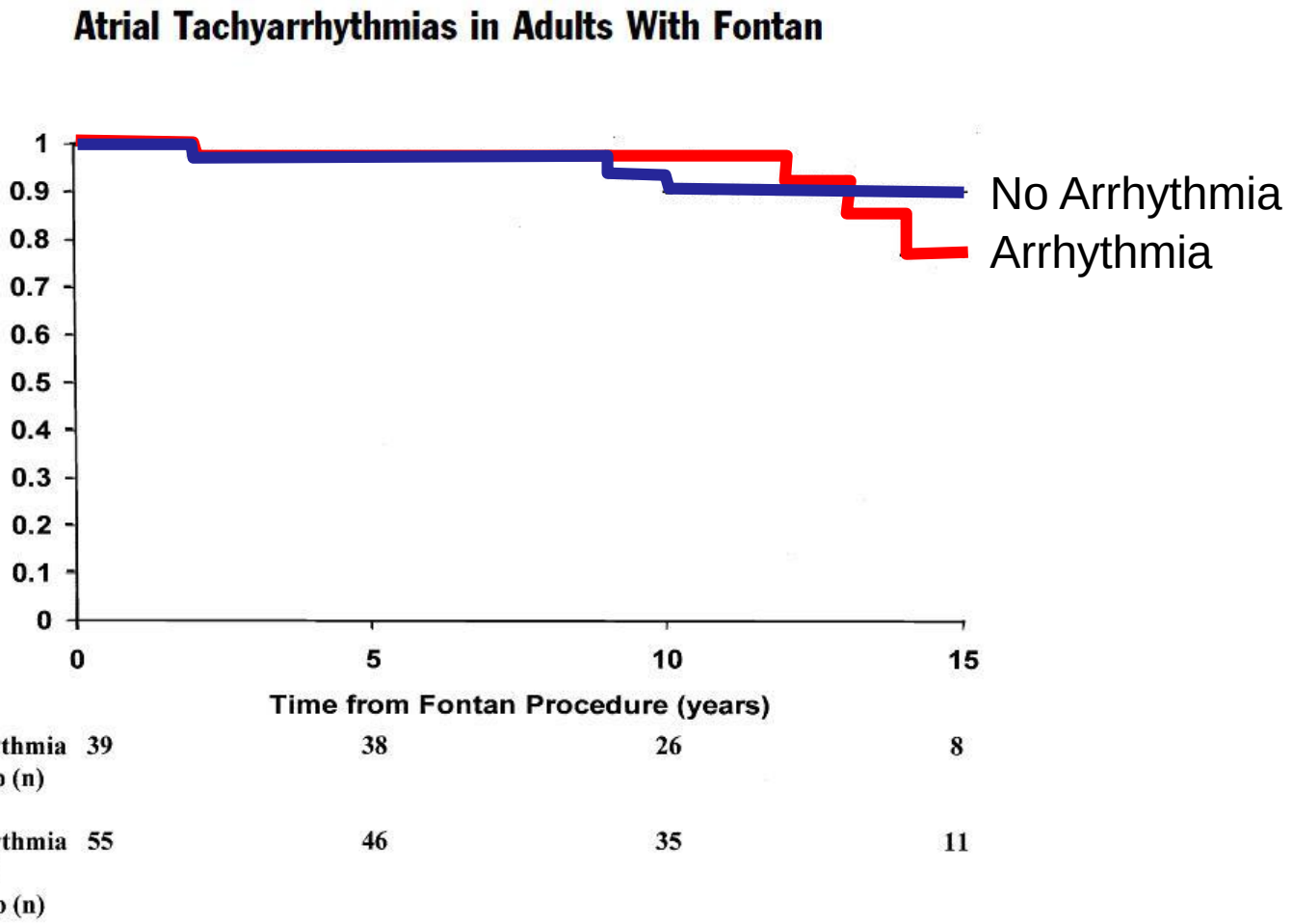
Atrial flutter / fibrillation / SVT

- 17 % after 5 years[1]
- 16 % after 5 years[2]
- 12.5% after 8.9 years[3]
- 10.6 % after 3.7 years[4]
- 41 % after 11 years[5]
- 7.3% after 8.6 years[6]

1. Durongpisitkul K: Circulation 1998;98:1099
2. Fishberger SB: J Thorac Cardiovasc S 1997;113:80
3. Girod DA: Circulation 1987;75:605
4. Gwellig : Br Heart J 1992;67:72
5. Ghai A: J Am Coll Cardiol 2001;37:585–92
6. Stephenson EA: J Am Coll Cardiol 2010;56:890

Effect of Arrhythmia On Survival Post-Fontan?

Ghai et al (Toronto) JACC 2001:37:585



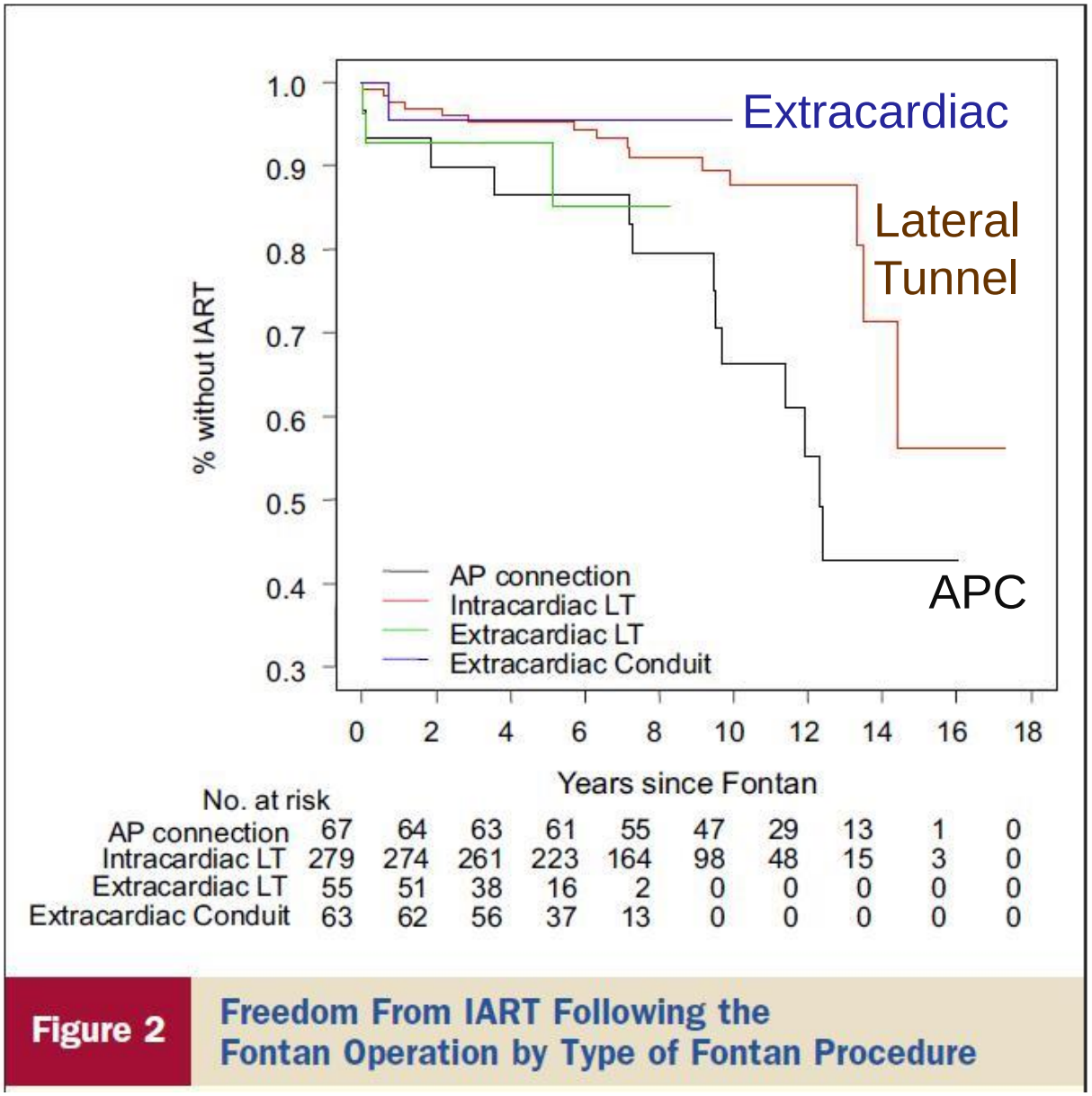
ATRIAL FLUTTER AFTER FONTAN

Risk Factors

- Procedure type (APC vs LT vs extracardiac)
- Fenestration and or collaterals
- Age at surgery
- AV valve regurgitation

Risk of Arrhythmia Post Fontan (n=520)

Risk Depends on Type of Procedure



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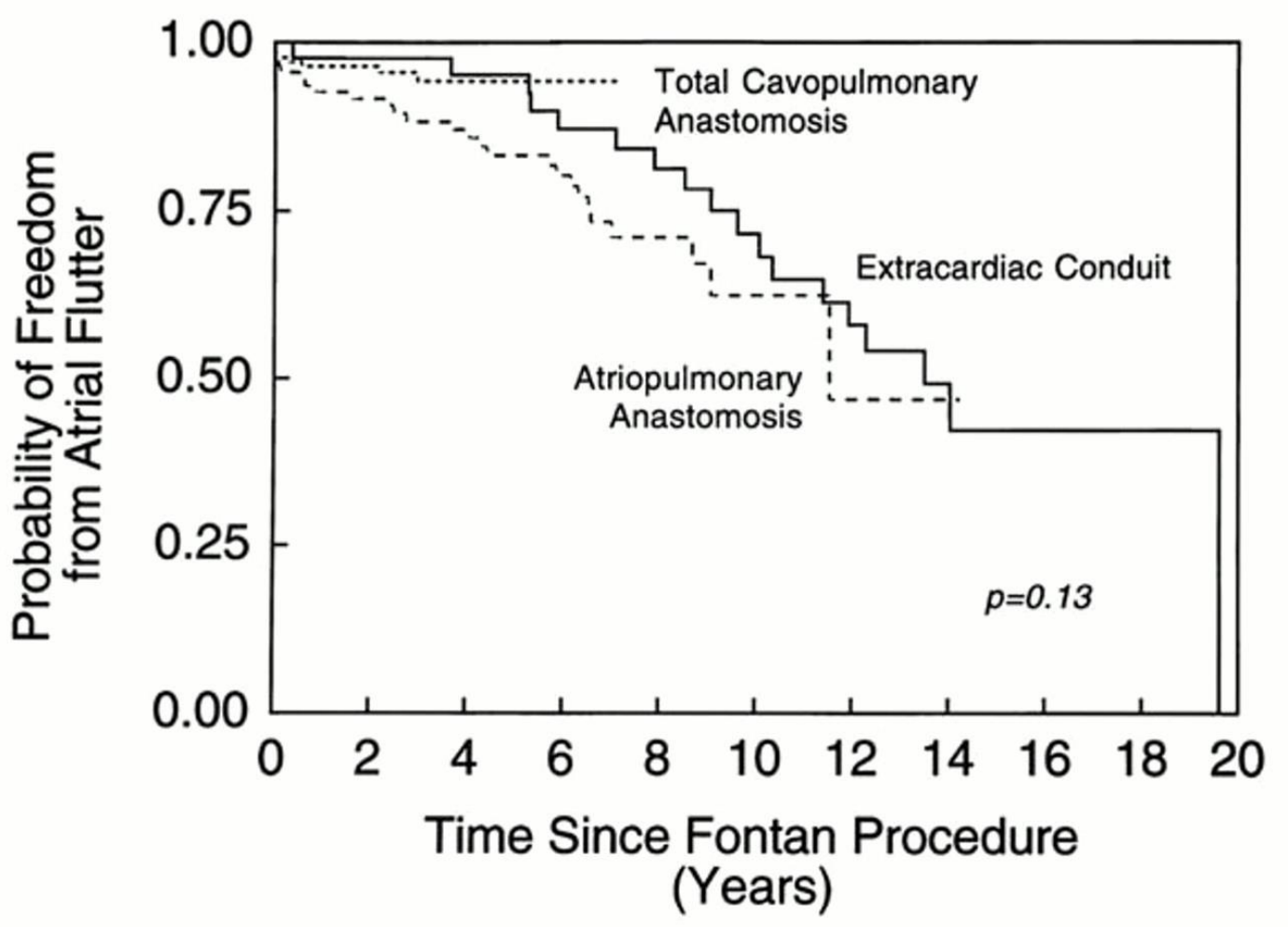
ATRIAL FLUTTER AFTER FONTAN
Risk Depends on Procedure Type



Fishberger S. B. et al.; J Thorac Cardiovasc Surg 1997;113:80-86

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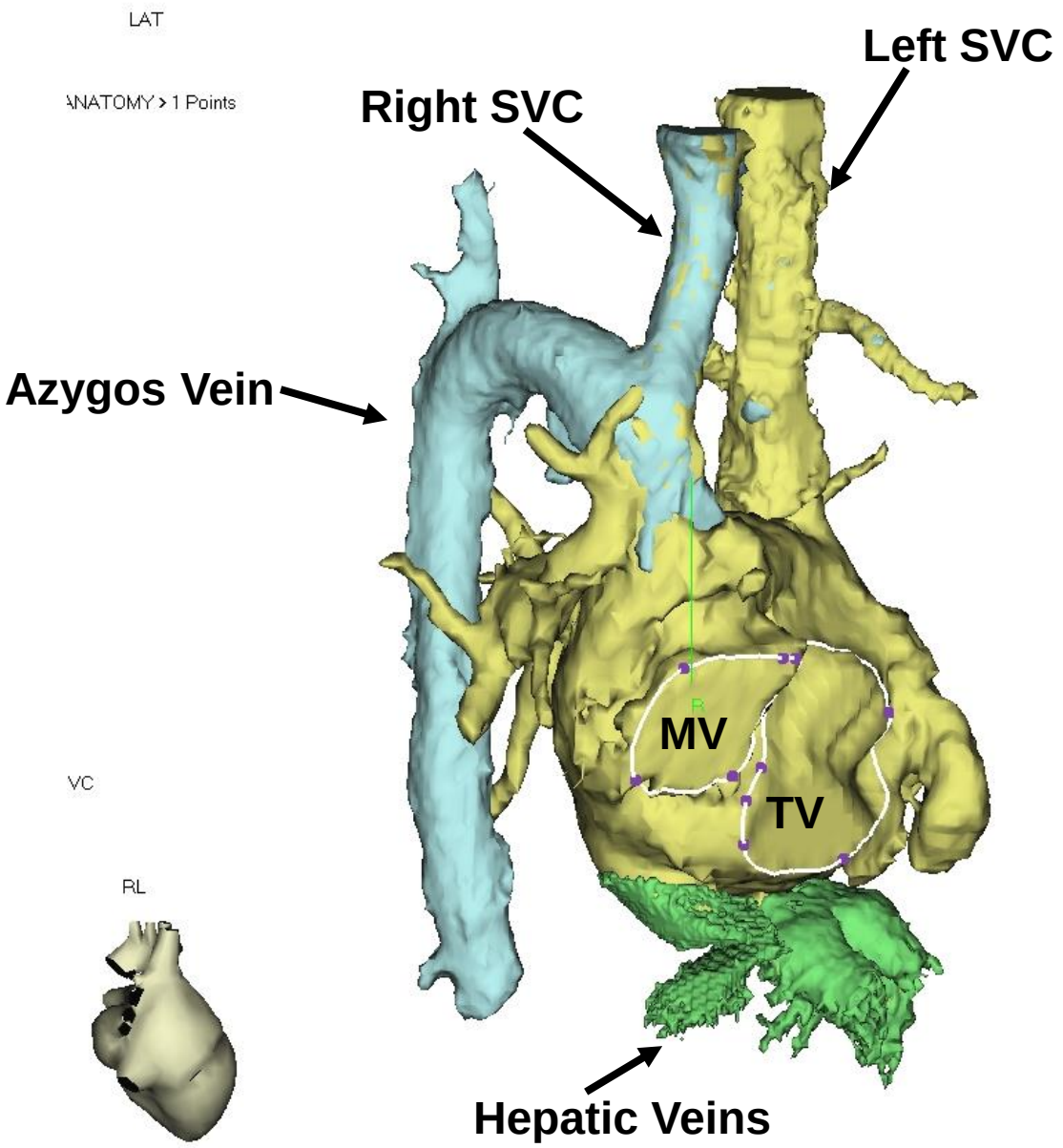
3: Flinn CJ. NEJM 1984;310:1635
4: Roos-Hesselink. Circulation 1995;91:2214

Importance of CT/MRI scan

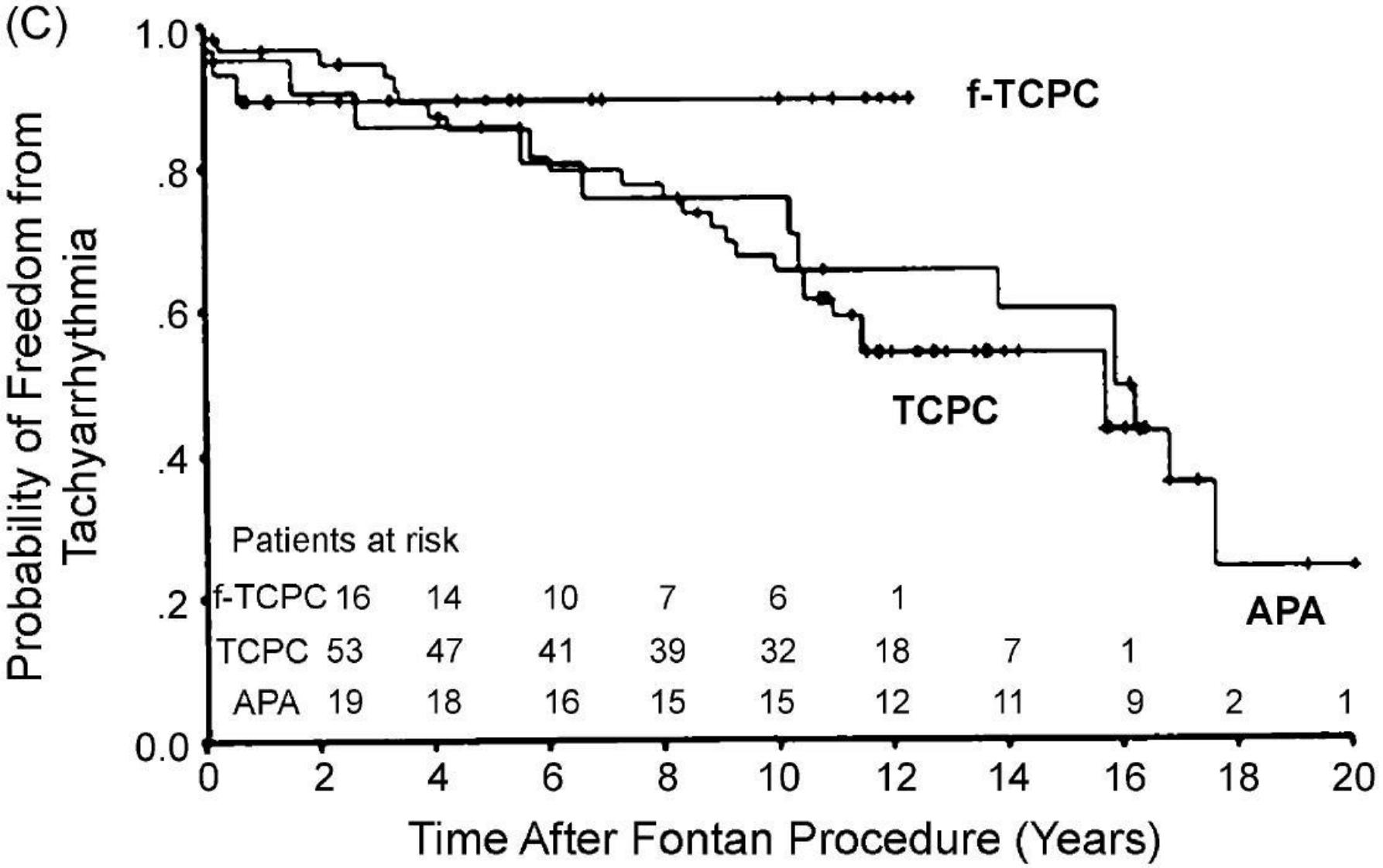


Dextrocardia
Interrupted IVC

RAO View

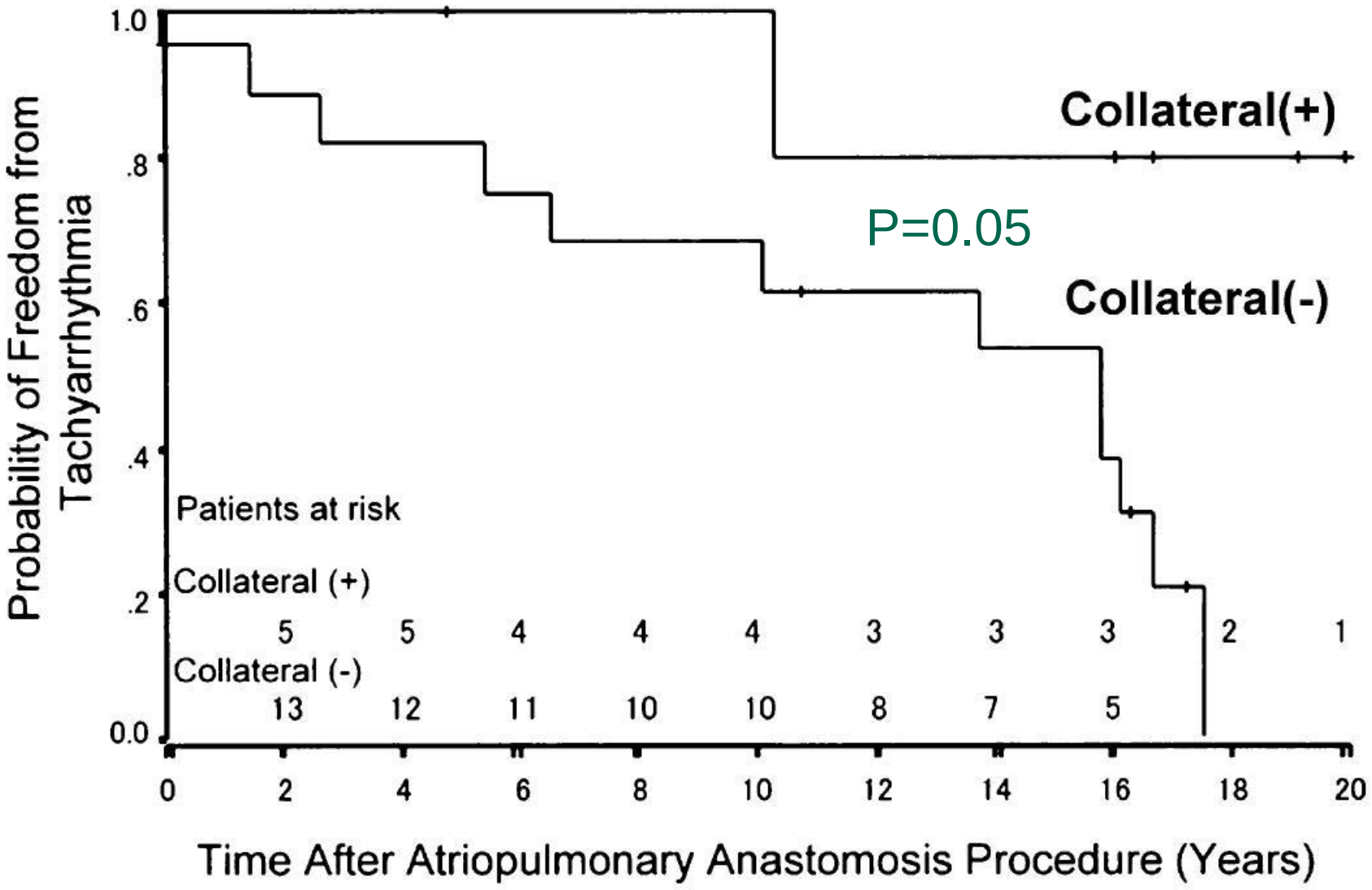
Fenestration (lateral tunnel) Decreases Late Arrhythmias?

Ono et al: Eur J Cardiothorac Surg 2006;30:923

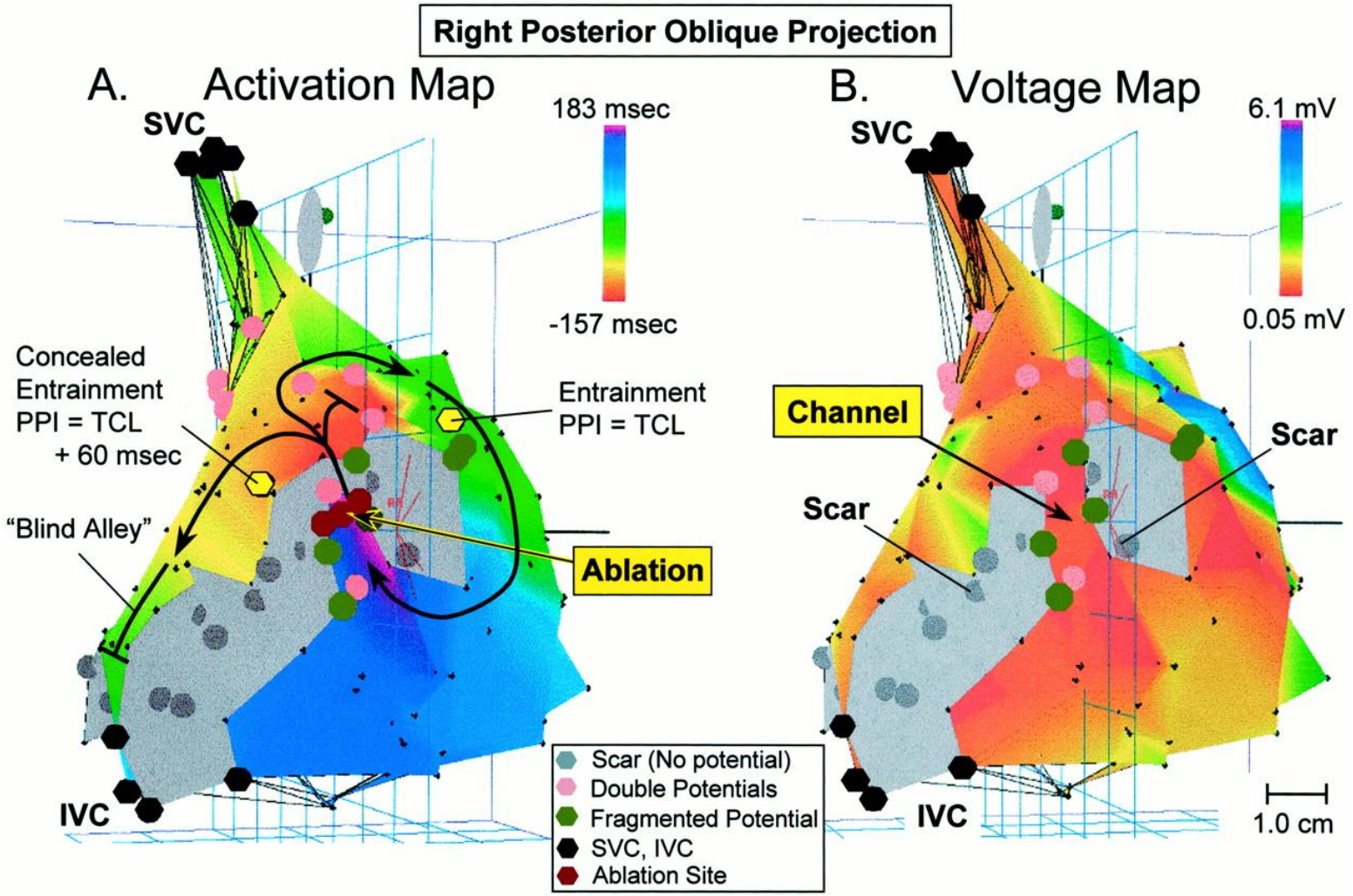


Effect of Collaterals on Tachyarrhythmias?

Ono et al: Eur J Cardiothorac Surg 2006;30:923



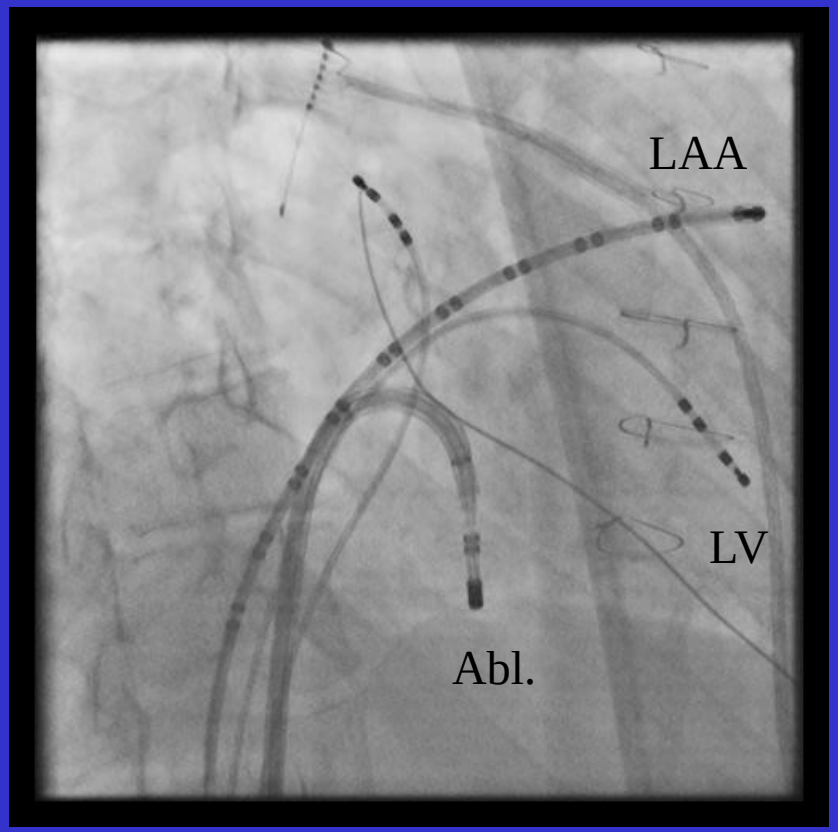
A, Activation map during MacroAT in ASD patient 1 shows continuous activation around smaller upper dense scar (gray area with gray tags) and line of double potentials (pink tags) and through channel (width 1.6 cm) between scars.



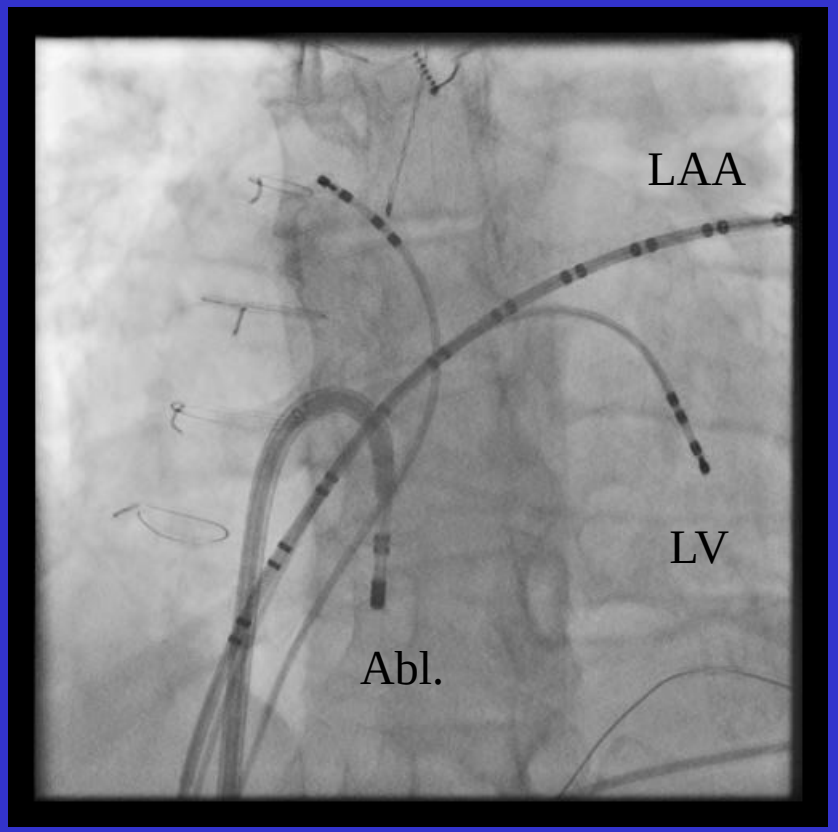
Hiroshi Nakagawa et al. Circulation. 2001;103:699-709

Mustard: trans-baffle approach to right (PV) atrium

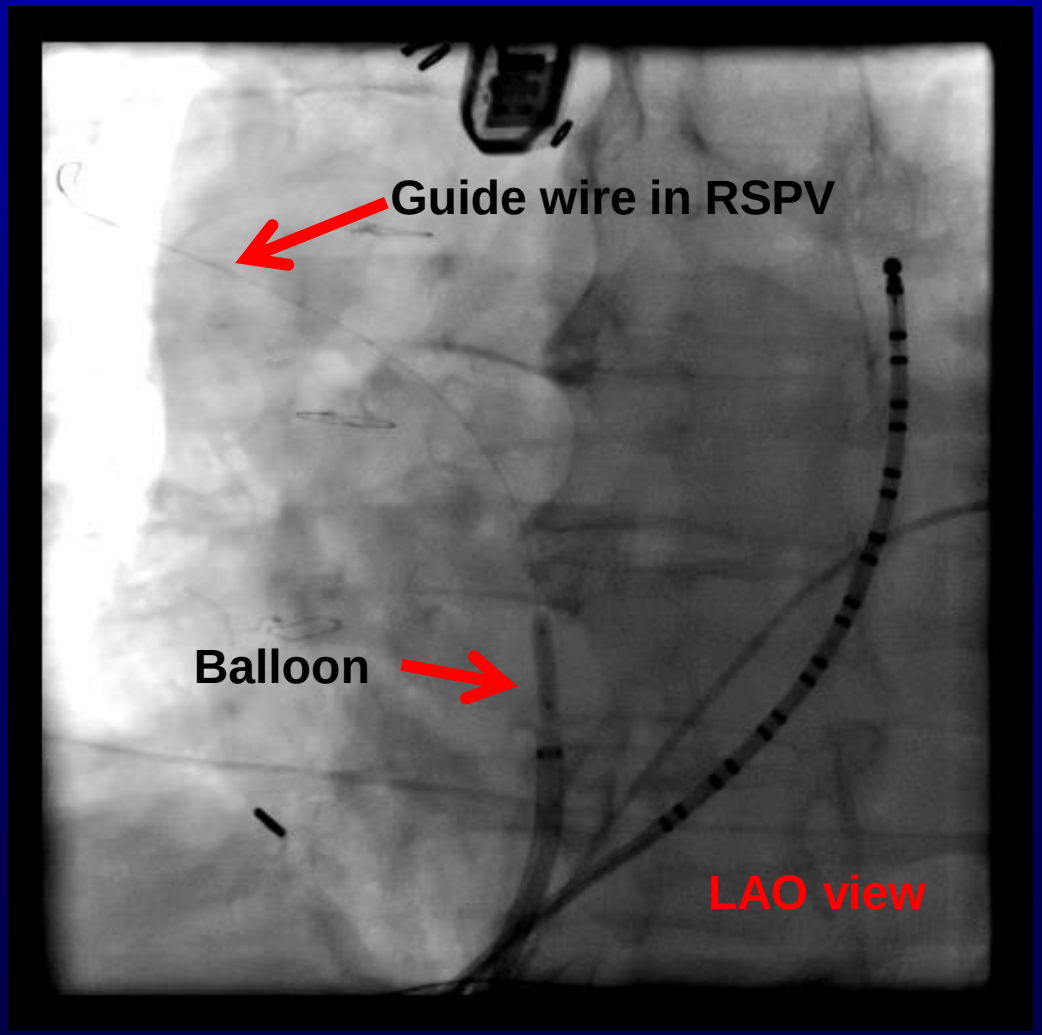
RAO

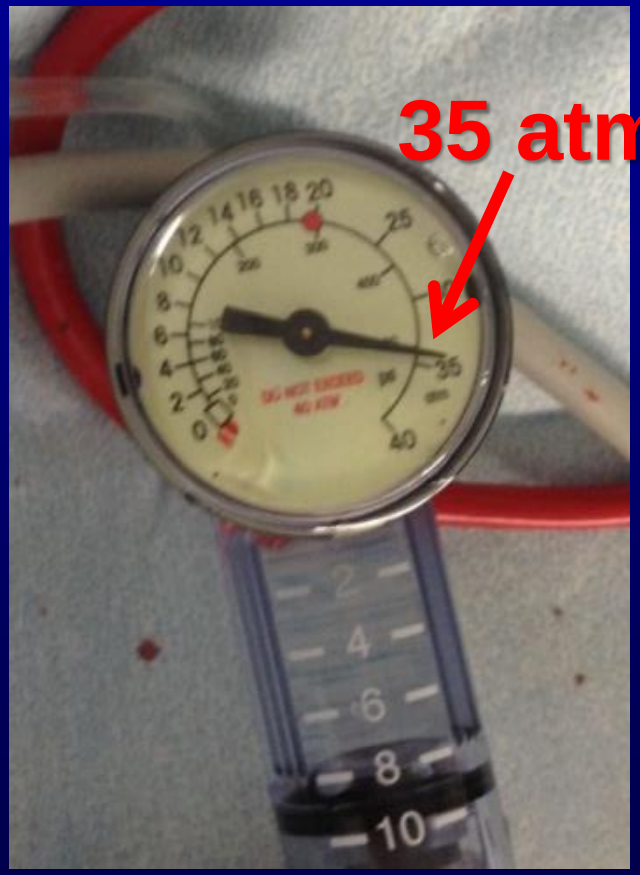


PA

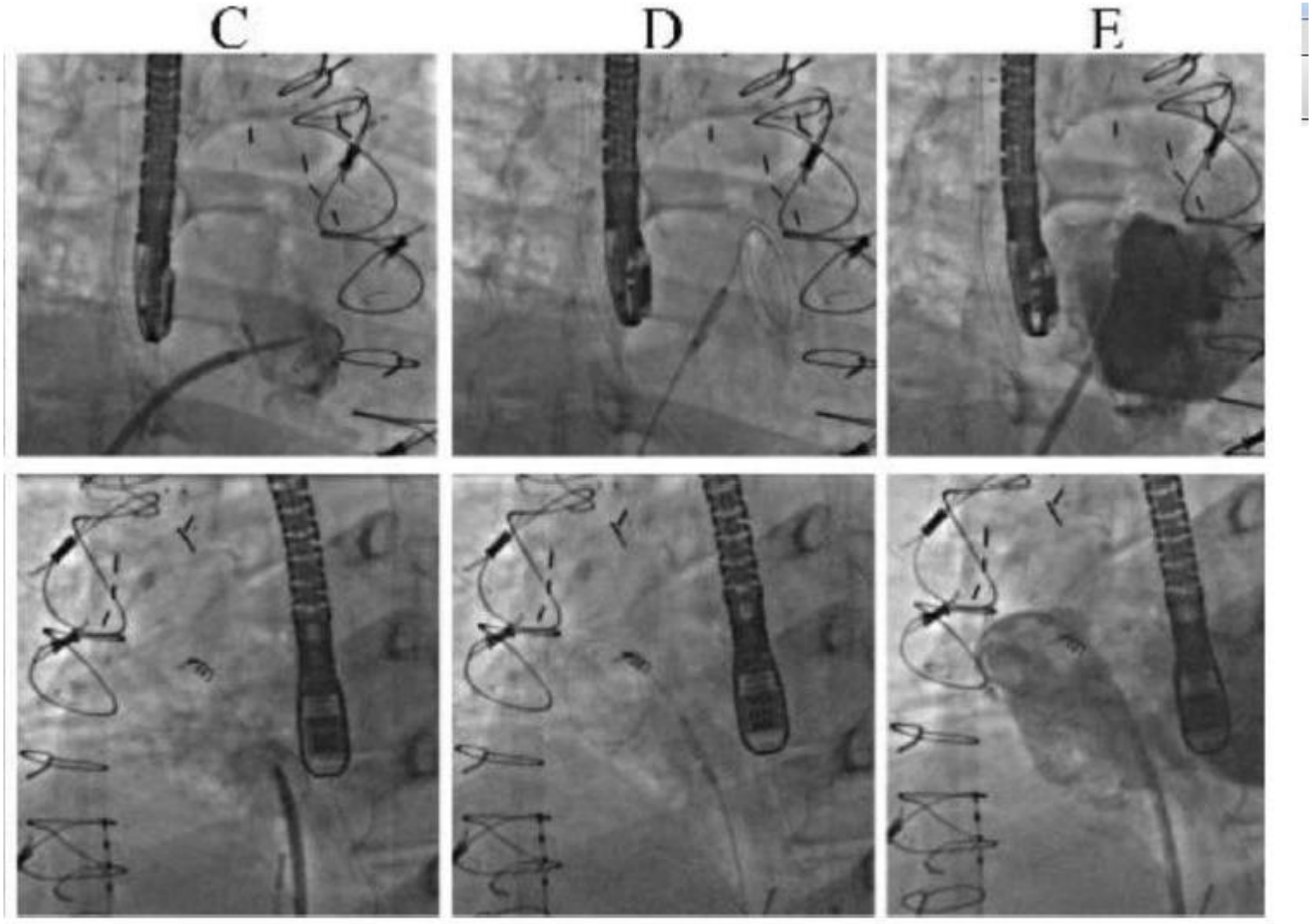


BAFFLE PUNCTURE
In Mustard/Senning & Fontan Patients



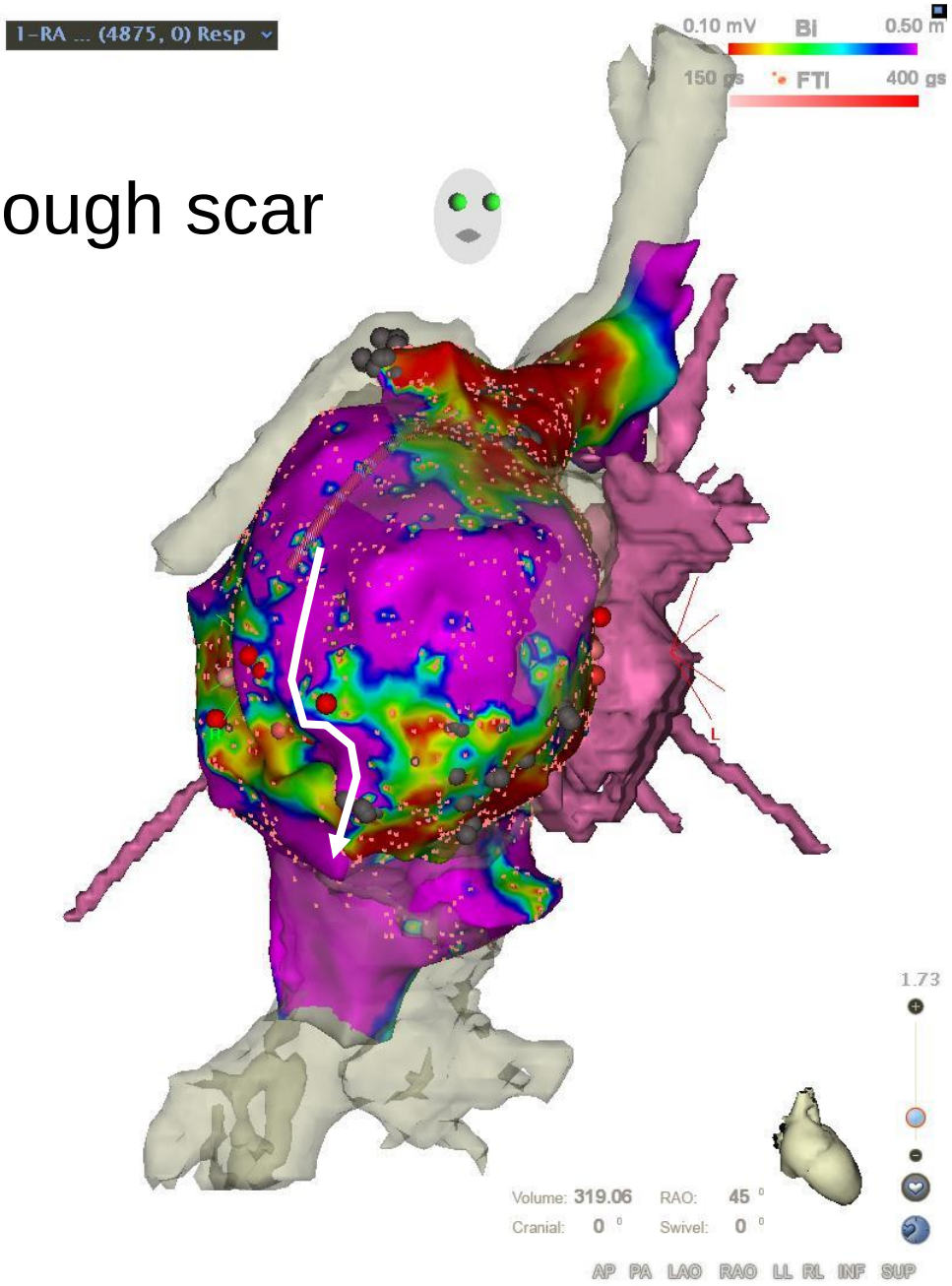


Extracardiac Fontan: Access To RA : Conduit Puncture



Dave, AS: Heart Rhythm. 2010 ; 7(3): 413–416

Channel through scar

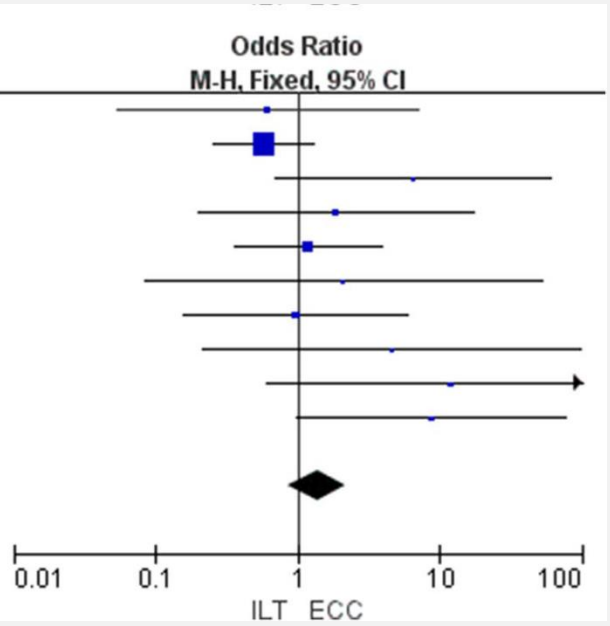


Meta-analysis Late Arrhythmias
LT vs ECC

C Late arrhythmias

Study or Subgroup	ILT Events	ILT Total	ECC Events	ECC Total	Weight	Odds Ratio M-H, Fixed, 95% CI
Azakie 2001	1	43	2	54	5.9%	0.62 [0.05, 7.06]
Balaji 2014	9	602	17	669	54.2%	0.58 [0.26, 1.32]
Bossers 2015	4	46	1	69	2.5%	6.48 [0.70, 59.92]
d' Udekem 2007	4	105	1	48	4.5%	1.86 [0.20, 17.11]
Fiore 2007	11	113	4	48	17.3%	1.19 [0.36, 3.93]
Hakacova 2008	1	60	0	41	2.0%	2.09 [0.08, 52.64]
Lee 2007	2	67	3	98	8.1%	0.97 [0.16, 5.99]
Nakano 2004	2	88	0	79	1.7%	4.60 [0.22, 97.19]
Nürnberg 2004	3	29	0	45	1.2%	12.02 [0.60, 241.80]
Sarkis 2011	7	26	1	25	2.5%	8.84 [1.00, 78.22]
Total (95% CI)		**1179**		**1176**	**100.0%**	**1.37 [0.85, 2.21]**
Total events	44		29			

Heterogeneity: Chi² = 12.27, df = 9 (P = 0.20); I² = 27%
Test for overall effect: Z = 1.30 (P = 0.19)



Li D et al Pediatr Cardiol 2017;38:873-880