



Transcatheter ablation of arrhythmias associated with congenital heart disease

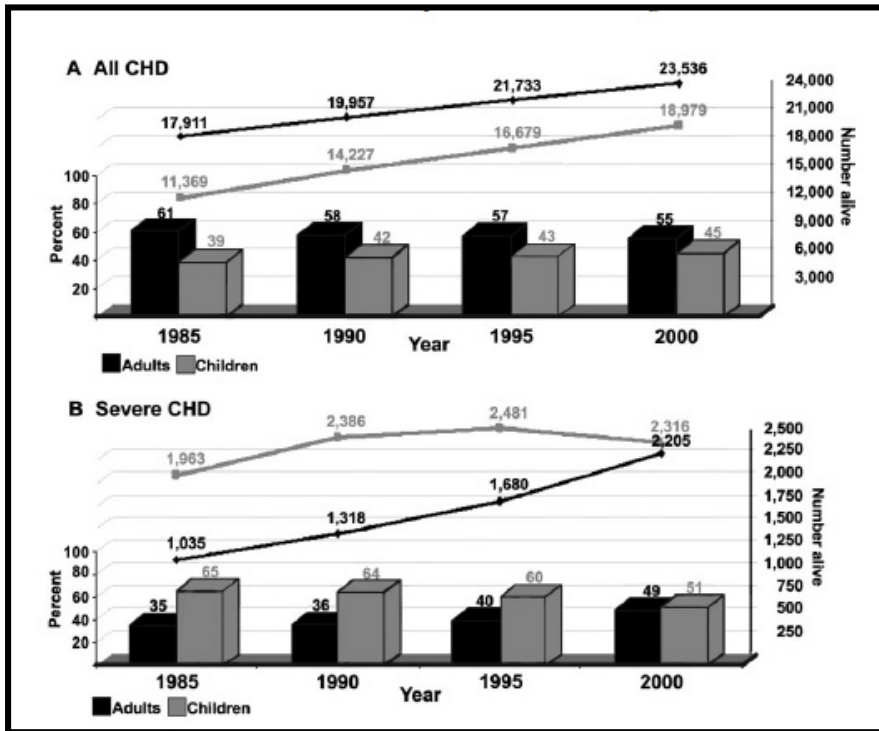
Dr. Marco Scaglione

marco.scaglione.at@gmail.com

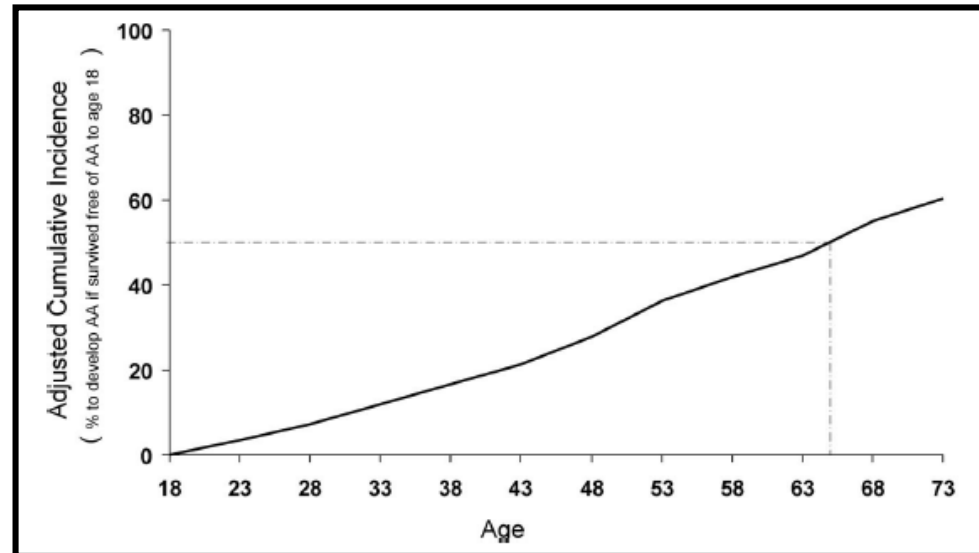
*Cardiovascular Division
Cardinal Massaia Hospital – Asti
University of Turin - Italy*

Atrial Arrhythmias in Adults with CHD: Age Distribution

“Advances in pediatric cardiovascular care have resulted in more adults with CHD”



Marelli AJ et al; Circulation 2007



Bouchardhy et al; Circulation 2009



Interventional Electrophysiology in Patients With Congenital Heart Disease

Relative Risk for Specific Arrhythmias in Common Congenital Heart Defects

	IART	AF	WPW	VT/SCD	SA Node Dysfunction	Spontaneous AV Block	Traumatic AV Block
VSD	+			+			+
ASD	+	+					
TOF	++			++			+
AS		+		++			+
D-TGA (M&S)	+++			++	+++		
CAVC	+					+	++
SING V (F)	+++	+		+	+++		
L-TGA	+		++	+		++	+++
Ebstein's anomaly	++		+++	+			

AF indicates atrial fibrillation; WPW, Wolff-Parkinson-White syndrome; SCD, sudden cardiac death; SA, sinoatrial; VSD, ventricular septal defect; ASD, atrial septal defect; TOF, tetralogy of Fallot; AS, aortic stenosis; M&S, after the Mustard or Senning operation; CAVC, common AV canal defect; SING V (F), single ventricle after the Fontan operation; + + +, high risk; + +, moderate risk; and +, slight risk.

P. Walsh Circulation 2007



2008 AHA/ACC Guidelines

Class I

1. Complete and appropriate noninvasive testing, as well as clear knowledge of the specific anatomy and review of all surgical and procedural records, is recommended before electrophysiological testing or device placement is attempted in ACHD patients. (*Level of Evidence: C*)

cardiovascular picture, particularly repairable hemodynamic issues that might favor a surgical or catheter-

tachycardia (VT). (*Level of Evidence: C*)

2. Pacemaker implantation can be beneficial in ACHD

Catheter ablation procedures for ACHD patients should be performed at centers where the staff is experienced with the complex anatomy and distinctive arrhythmia substrates encountered in congenital heart defects. (*Level of Evidence: B*)

5. Epicardial pacemaker and device lead placement should be performed in all cyanotic patients with intracardiac shunts who require devices. (*Level of Evidence: B*)



Role of Catheter Ablation in Postoperative Arrhythmias

Evolution of Ablation Techniques for IART

Dates	Technique	Acute Success	Recurrence Rates
92-95	Entrainment/ 4-mm tip	60-75%	>50%
95-98	"Cooled" tip	80-90%	30-50%
99-00	3D mapping	80-90%	30-50%
00-05	Demonstrate block	90-100%	25-30%
05-	Advanced map, cooled tip, cryo	90-100%	???

USEFULNESS OF MAPPING SYSTEMS

J. Philippe Saul PACE 2008





Samantha, che vive a Ronco Scrivia (Genova) con il marito e il figlio Mattia, quattro anni, è operata in un'impresa di pulizie dopo aver lavorato in una sartoria. Tra poco metterà al mondo il secondo bambino, Marco, stesso nome del cardiologo che l'ha operata mentre era incinta.

la situazione era diventata drammatica. **Il cuore non ce la faceva più a pompare a quella velocità.** Era sfinito, dilatato al massimo, sul punto di cedere.

A quel punto ho realizzato che in tutti quegli anni, anche se non me ne rendevo conto, la tachicardia atriale ectopica aveva danneggiato il mio cuore, costretto a battere come se io stessi sempre correndo. **Rischio il trapianto.**

Al sesto mese di gravidanza, i medici hanno deciso di sottopormi d'urgenza a una terapia di beta-bloccanti nel tentativo di arginare i battiti, sopra i 160 al minuto. Tentativo fallito. **L'ultima possibilità, problematica in gestazione per via dell'uso dei raggi X, era l'intervento di ablazione.**

Al San Martino di Genova mi hanno indirizzata ad Asti, all'ospedale Cardinal Massaia, dove dispongono di un macchinario all'avanguardia per la cura dell'aritmia e dove sarebbero stati in grado di operarmi senza far ricorso a trasfusioni di sangue, che io rifiuto essendo testimone di Geova.

Dopo gli accertamenti (elettrocardiogramma, risonanza, ecocardiogramma), sono entrata in sala

operatoria. Tesisissima, anche se il cardiologo, Marco Scaglione, mi aveva rassicurato che avrebbe ridotto al minimo l'uso di raggi X. C'è riuscito in un tempo da Guinness: appena 41 secondi.

I battiti si sono calmati e Marco nascerà tra poco

Per me è stata una magia. Ho assistito in diretta all'intervento, in anestesia locale. Ho avvertito la sonda entrare nell'arteria femorale e salire fino a raggiungere il cuore. Ho visto il catetere entrare nello schermo e ho avvertito un leggero calore. Aveva bruciato il focolaio che dava origine all'aritmia.

Il cuore si è calmato subito. **Da 160 al minuto, i battiti sono scesi a 90.**

«Adesso riparte», mi sono detta. Invece no, si è placato. L'operazione è riuscita. Finalmente, dopo dieci anni di corsa, posso tirare il fiato! Fuori dalla sala operatoria, ho ricevuto la telefonata di mio figlio: «È ancora rotto il cuore, mamma?». Sono scoppiata in lacrime: «No, Mattia, il cuore di mamma lo hanno riparato. Anche Marco, che è nel pancione, sta molto bene». Presto lo conosceremo. >>>

Samantha Crimiti

L'impresa del cardiologo: l'ablazione transcateretere di Samantha è uno dei pochi casi al mondo su donne incinte

Nel mondo sono stati eseguiti poco più di dieci interventi di ablazione transcateretere per risolvere l'aritmia (cioè l'alterazione del ritmo cardiaco) su donne in gravidanza come Samantha Crimiti. Nel suo caso è stato stracciato il record di velocità nell'impiego dei raggi X: appena 41 secondi, per non danneggiare il feto, contro i sette minuti e mezzo del più breve tempo prima d'ora raggiunto in un intervento di uguale complessità. Il merito va a Marco Scaglione (marco.scaglione@ok.rcs.it), primario di cardiologia del Cardinal Massaia di Asti.

■ **L'ARITMIA.** «La tachicardia atriale ectopica, da cui era affetta la paziente, è un disturbo dell'impianto elettrico del cuore,

dove una regione inizia a comportarsi come una pila autonoma, innescando un'aritmia, un ritmo eccessivo», spiega Scaglione. «Lo sforzo di battere a una frequenza accelerata può provocare la tachicardia-miopatia, cioè uno sfianamento del cuore, che si dilata, e una riduzione della funzione dei ventricoli, addetti a pompare il sangue. La situazione può diventare così grave da richiedere un trapianto».

■ **L'INTERVENTO.** La soluzione è l'ablazione transcateretere, risolutiva nel 90% dei casi circa. In anestesia locale, con un sondino introdotto in corrispondenza dell'inguine (attraverso la vena o l'arteria femorale) e spinto fino al cuore, si va a bruciare con la

radiofrequenza, il circuito elettrico che genera l'alterazione del ritmo cardiaco. Ma l'intervento richiede l'uso di raggi X, necessari per illuminare il percorso del catetere.

■ **ASTI È ALL'AVANGUARDIA.**

Il centro aritmologico di Asti è stato tra i primi a dotarsi di una tecnologia d'avanguardia, chiamata Carto 3. «È un sistema che riesce a ricostruire in tempo reale la mappa in tre dimensioni dell'interno del cuore, anatomica ed elettrica, permettendo anche la visualizzazione di parte dei cateteri», dice Scaglione. «Integrando queste informazioni con le immagini della risonanza o della Tac, il chirurgo si orienta con precisione, limitando l'uso dei raggi X».

Marco Scaglione

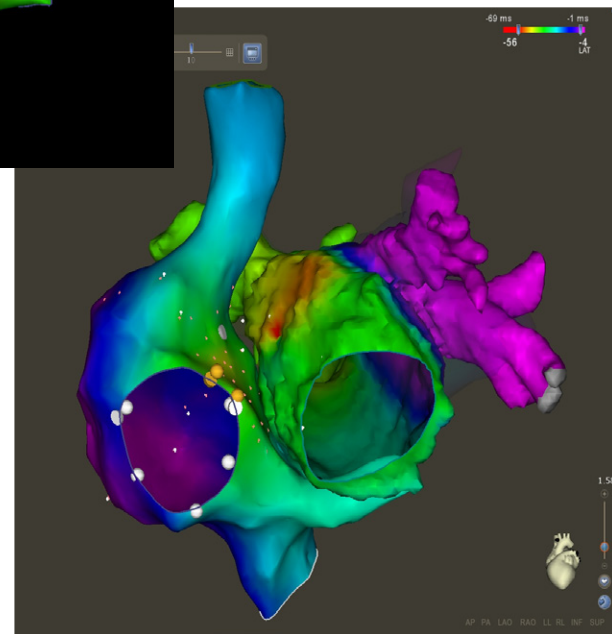
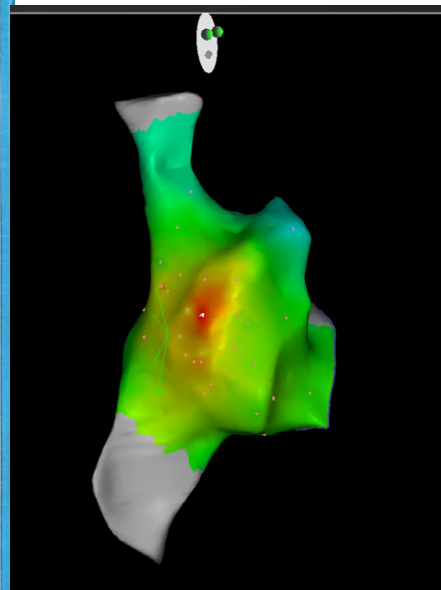


cardiologo

Piemontese, 46 anni, è responsabile del centro aritmologico dell'ospedale di Asti, struttura di riferimento nazionale dove vengono effettuate circa 500 ablazioni all'anno. Insegna anche alla scuola di specializzazione in cardiologia dell'Università di Genova.

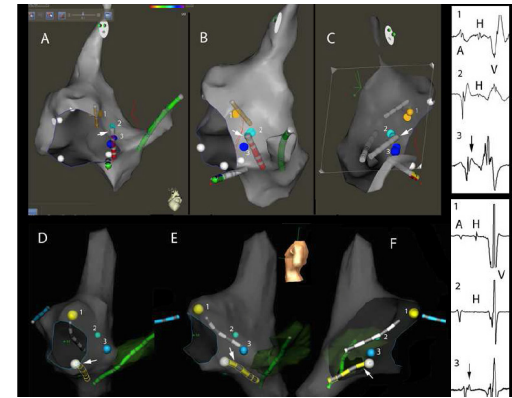
luglio 2010

ABLATION IN PREGNANCY ZERO FLUORO 2 cases



Single Center Experience of Fluoroless AVNRT Ablation Guided by Electroanatomic Reconstruction in Children and Adolescents

Fluoroless ablation was feasible in **19/21**, (2 patients 50 s and 45 s)



Zero-Fluoroscopy Ablation of Accessory Pathways in Children and Adolescents: CARTO3 Electroanatomic Mapping Combined with RF and Cryoenergy

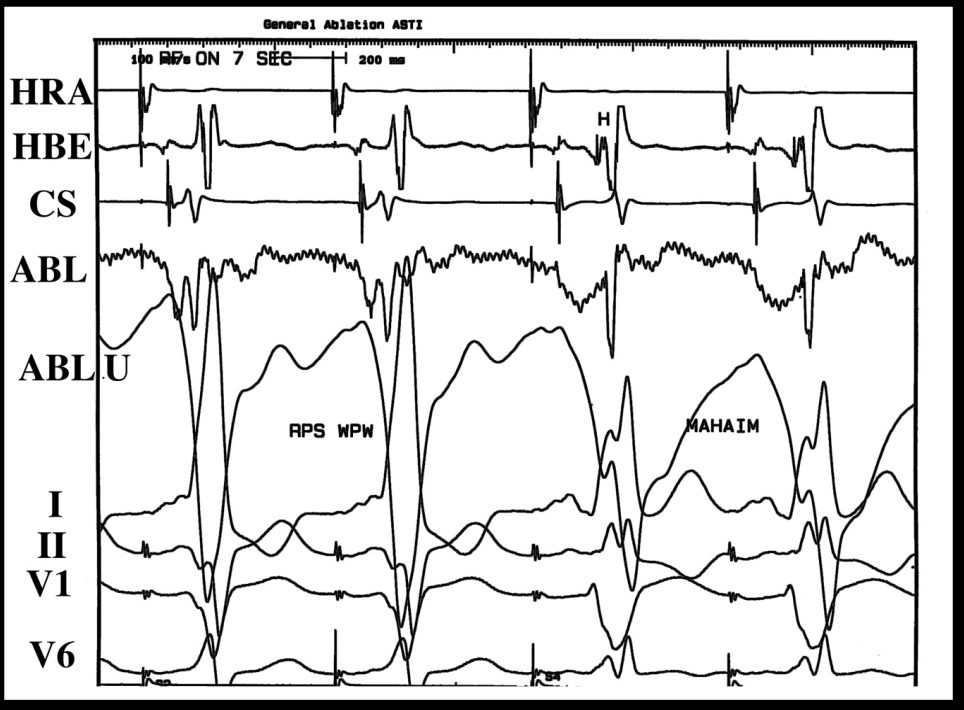
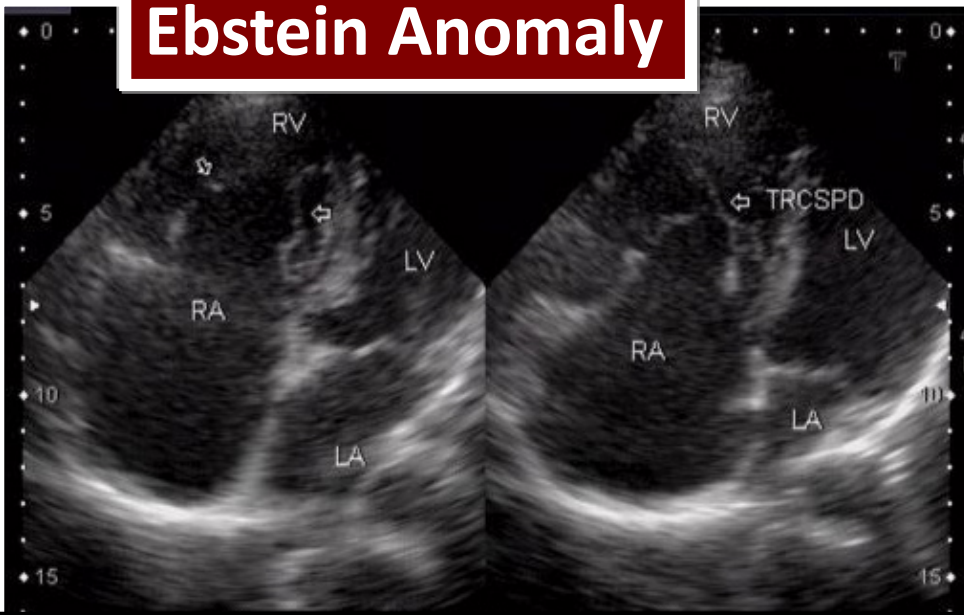
Fluoroless ablation in 100 % of the patients



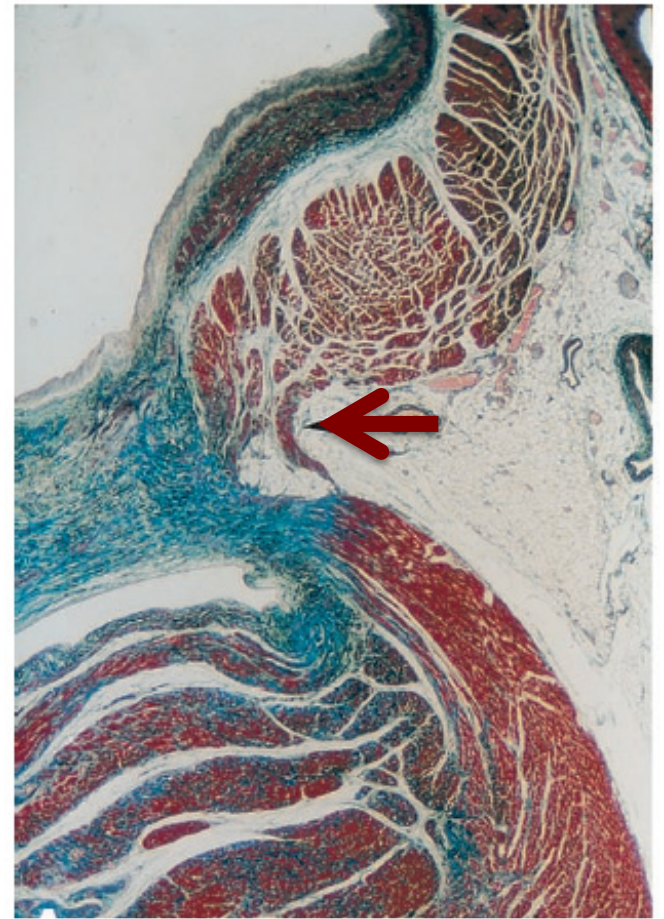
Scaglione M et al, PACE 2013, PACE 2015



Ebstein Anomaly



Embriogenesis



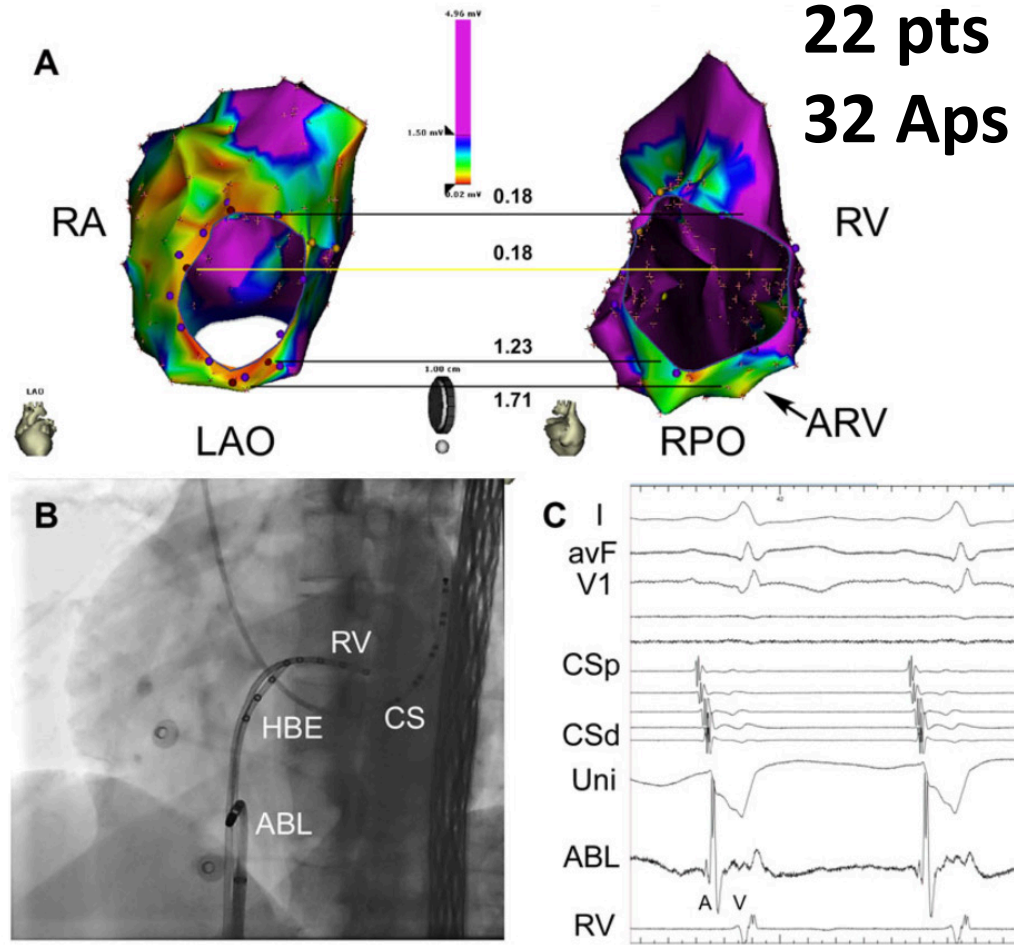
**MULTIPLE
ACCESSORY PATHWAYS**



Frequency of Fractionated Ventricular Activation and Atrial/Ventricular Electrogram Amplitude Ratio at Successful Ablation Target of Accessory Pathways in Patients with Ebstein's Anomaly

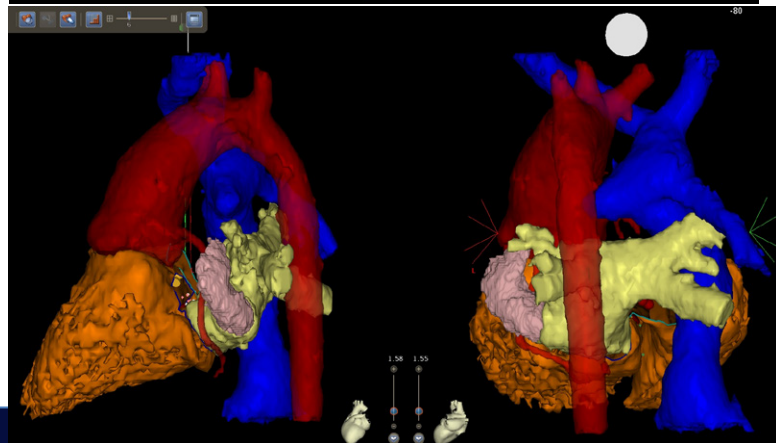
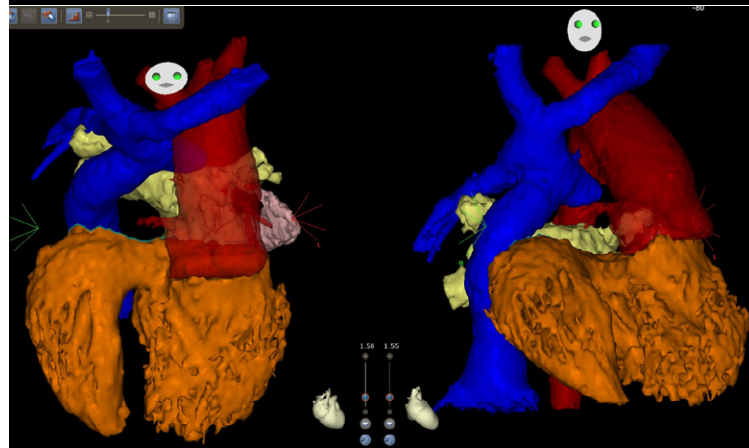
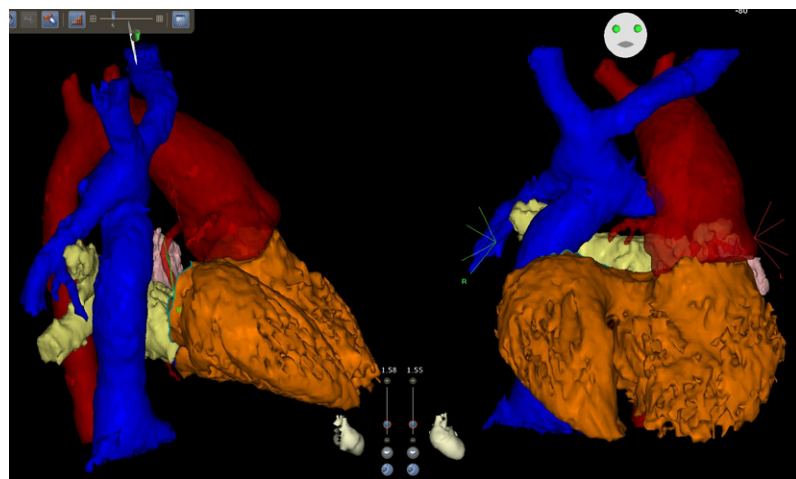
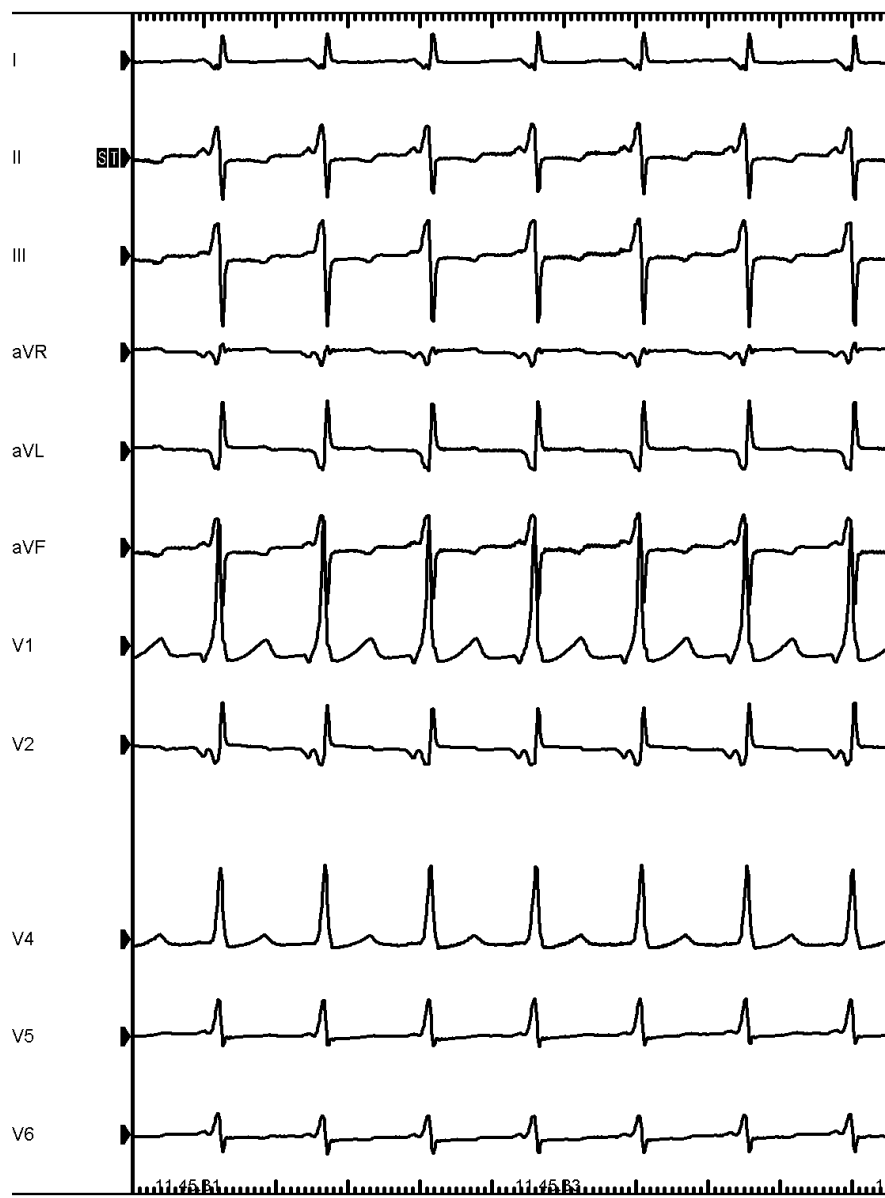
Guo et al JCE 2015

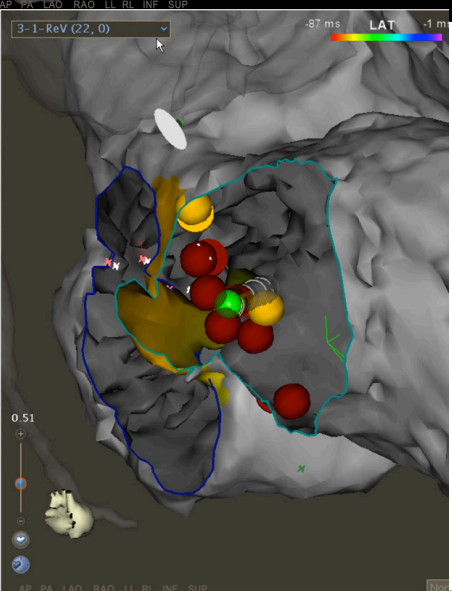
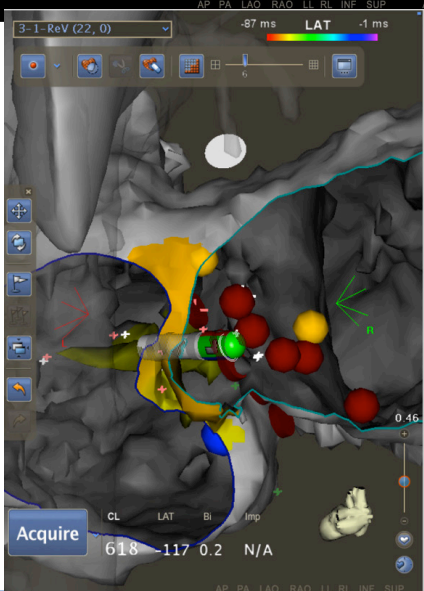
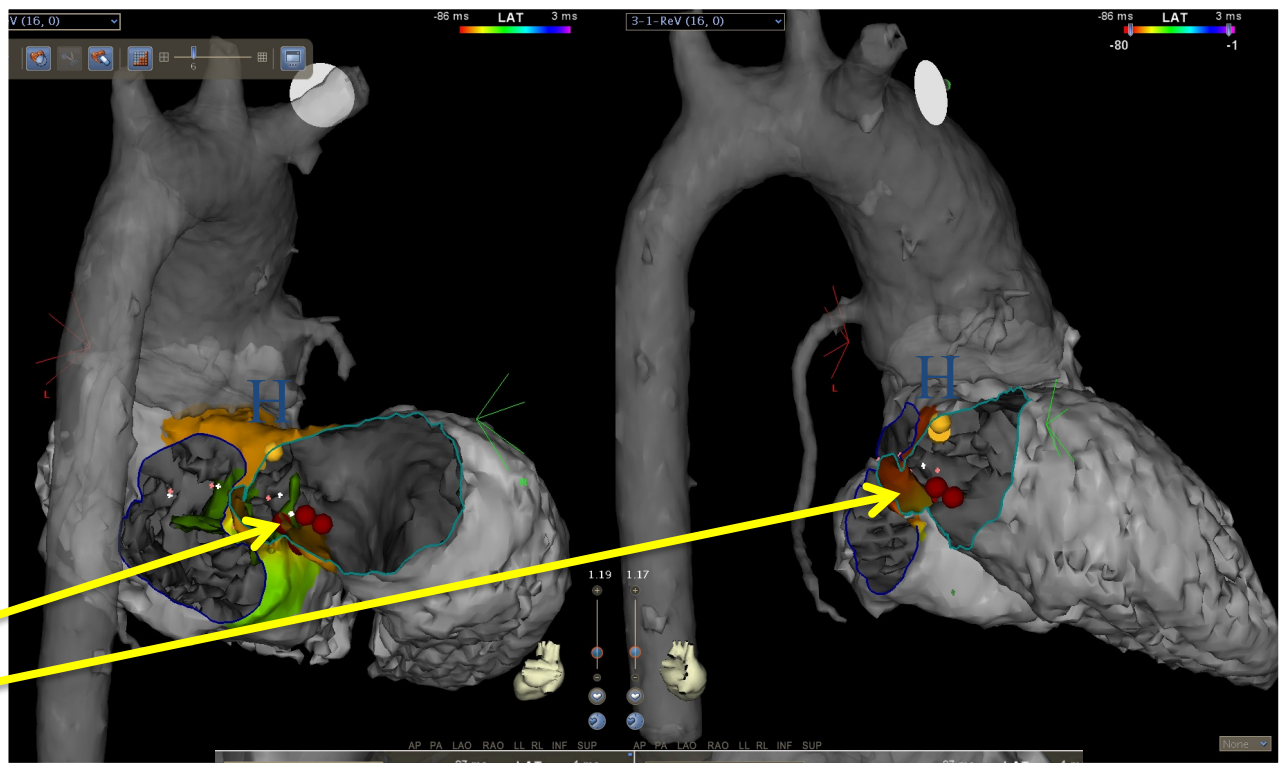
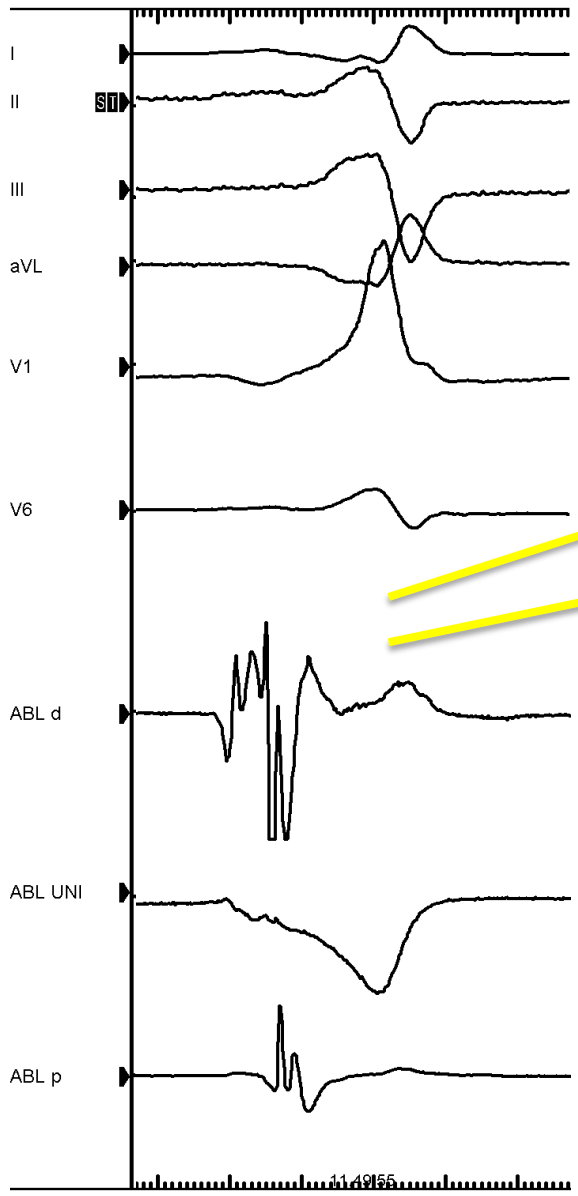
22 pts
32 Aps RIGHT SIDED

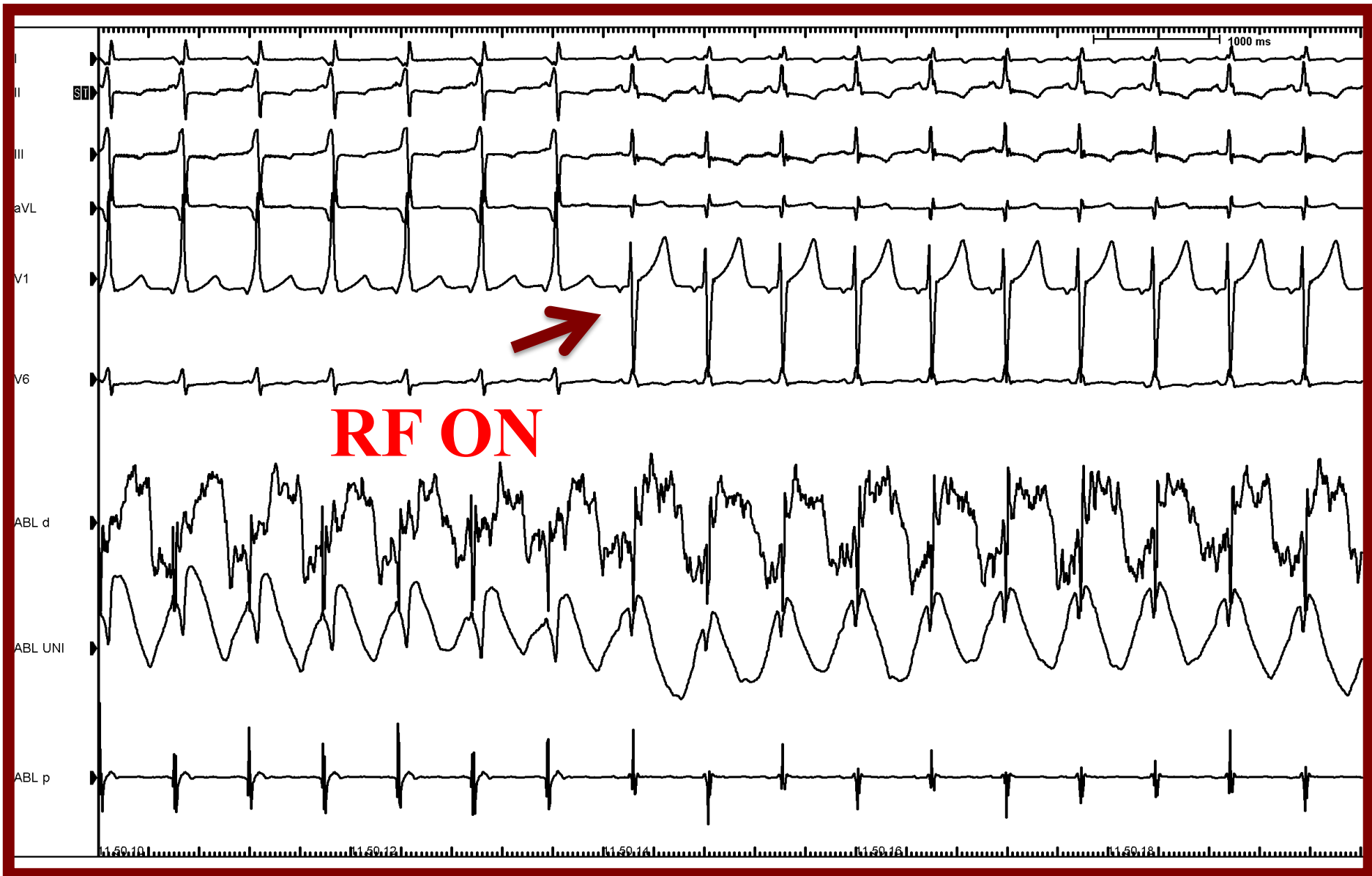


Challenging anatomy





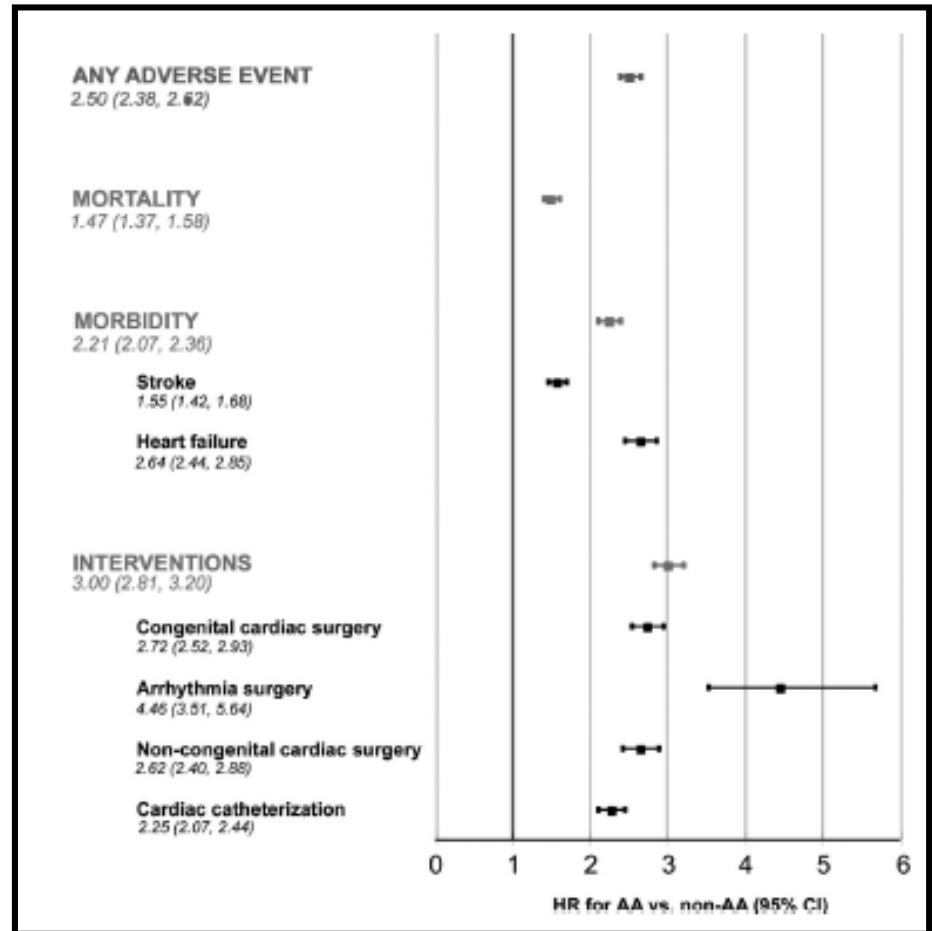




Atrial Arrhythmias in Adults with CHD: *Impact on Prognosis*

*Atrial arrhythmias conferred a 2.5-fold higher risk of adverse events with a near 50% increase in mortality**

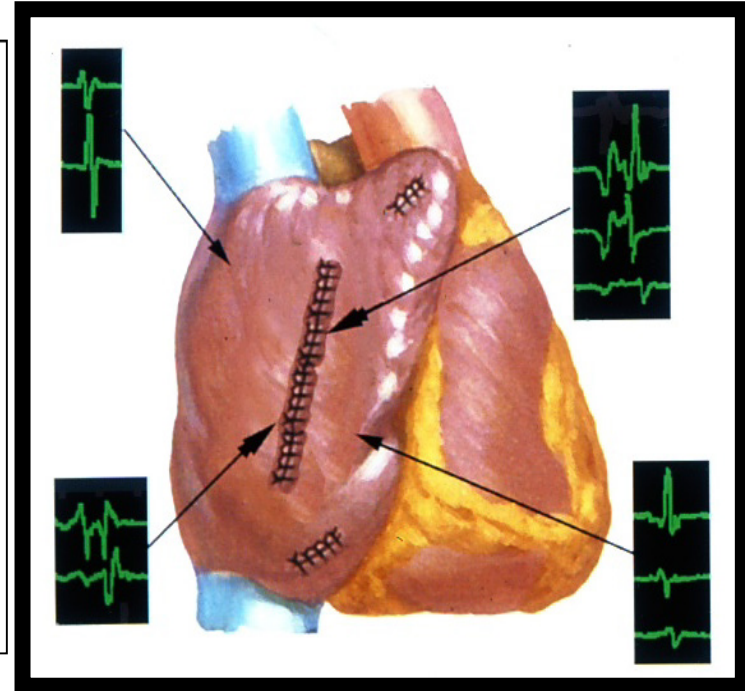
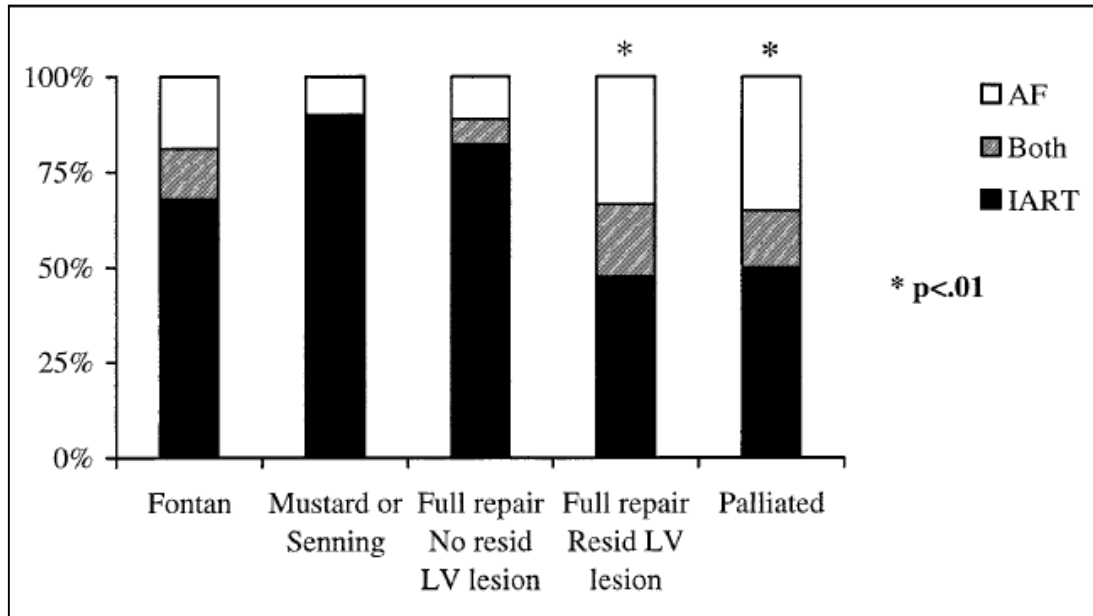
Pts with CHD who developed atrial arrhythmias had a > 50% ↑ stroke risk and 2-3 fold ↑ HF risk and occurrence of cardiac interventions



Bouchardhy et al; Circulation 2009; 120:1679-1686



Prevalence of AF and IART in Congenital Heart Disease



- ✓ Up to a third of patients with CHD and atrial tachycardia who require cardioversion may have AF
- ✓ Patients with significant and unrepaired lesion may be relatively more prone to AF

Kirsh JA et al, *Am J Cardiol* 2002;90:338-340



Update on Interventional Electrophysiology in Congenital Heart Disease

Evolving Solutions for Complex Hearts

Ablation for AF in CHD

As mentioned, AF is emerging as a more significant problem as patients with CHD attain older ages.¹² There is now a small but growing literature describing catheter ablation of AF for this population. Most cases have involved fairly straightforward lesions, such as atrial septal defects,^{45,46} although a few reports of more complex anatomy also exist. One item that may set these cases apart from more conventional AF ablation in a structurally normal heart is the challenge of achieving left atrial access across patches and septal occlusion devices, but there is now ample evidence that this can be accomplished safely.^{23,45,46} Catheter ablation for AF will almost certainly become a more common intervention for CHD in the near future.

Sherwin et al; *Circ Arrhythm Electrophysiol.* 2013



Very long-term results of electroanatomic-guided radiofrequency ablation of atrial arrhythmias in patients with surgically corrected atrial septal defect

Scaglione et al EUROPACE 2014

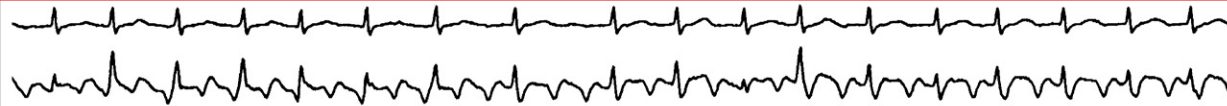
Table 1 Baseline characteristics

Males/females	18/28
Patients' mean age (years)	49 ± 13
Patients' mean age at surgery (years)	25 ± 13
Defect type	
Ostium secundum ASD	41/46
Ostium primum ASD	5/46
Known correction modality	28/46
Autologous/synthetic patch	17/28
Continuous suture	10/28
Combined approach	1/28
Second surgical intervention	6/46
Surgical reintervention on ASD	3/6
Percutaneous closure of ASD	1/6
Botallo duct closure	1/6
Right outflow tract reconstruction	1/6



Very long-term results of electroanatomic-guided radiofrequency ablation of atrial arrhythmias in patients with surgically corrected atrial septal defect

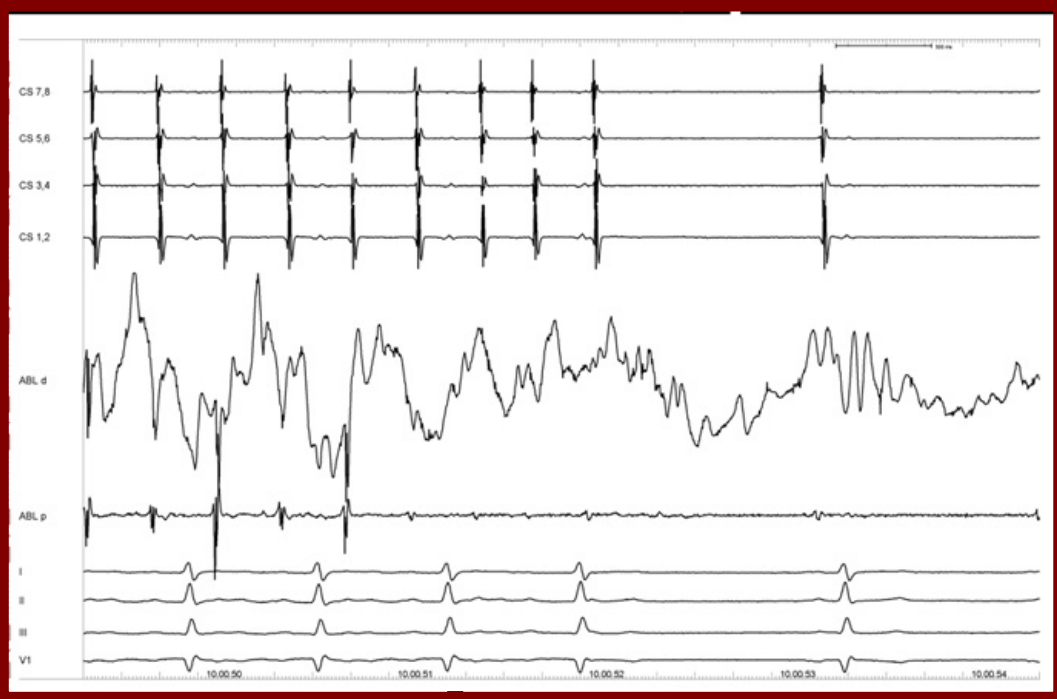
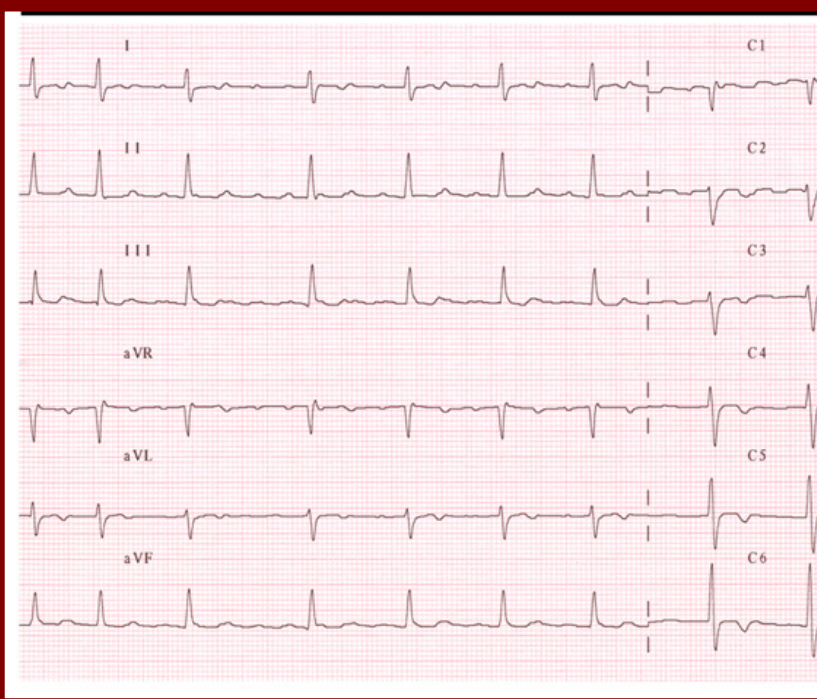
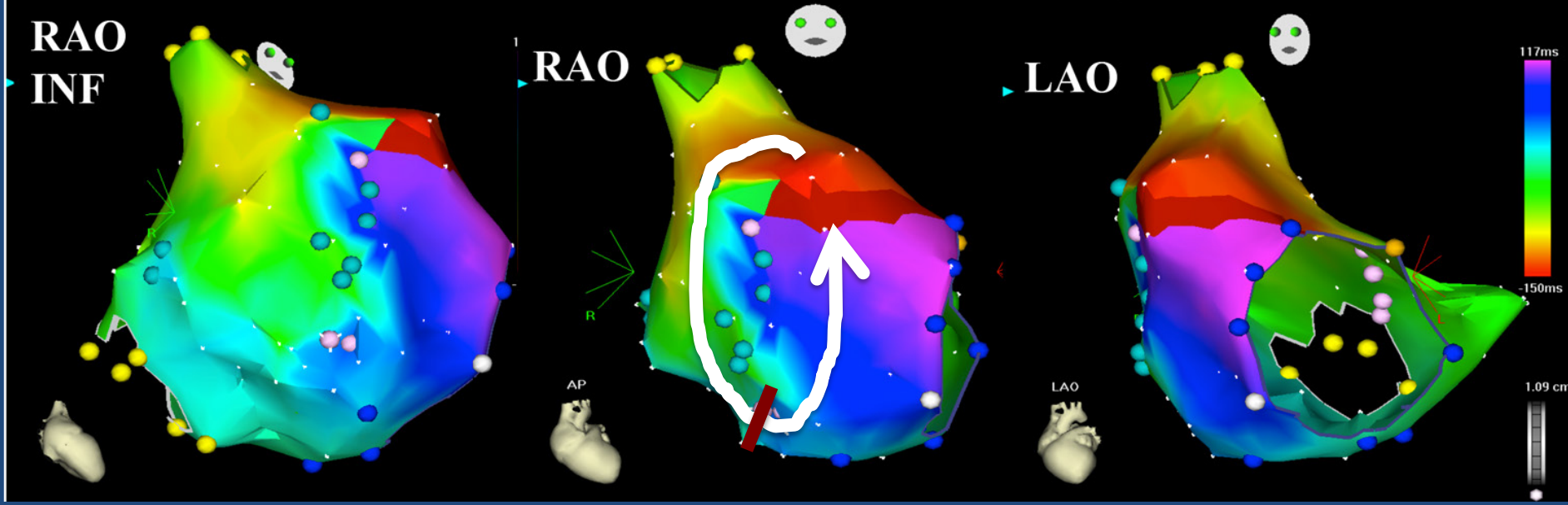
Scaglione et al **EUROPACE 2014**

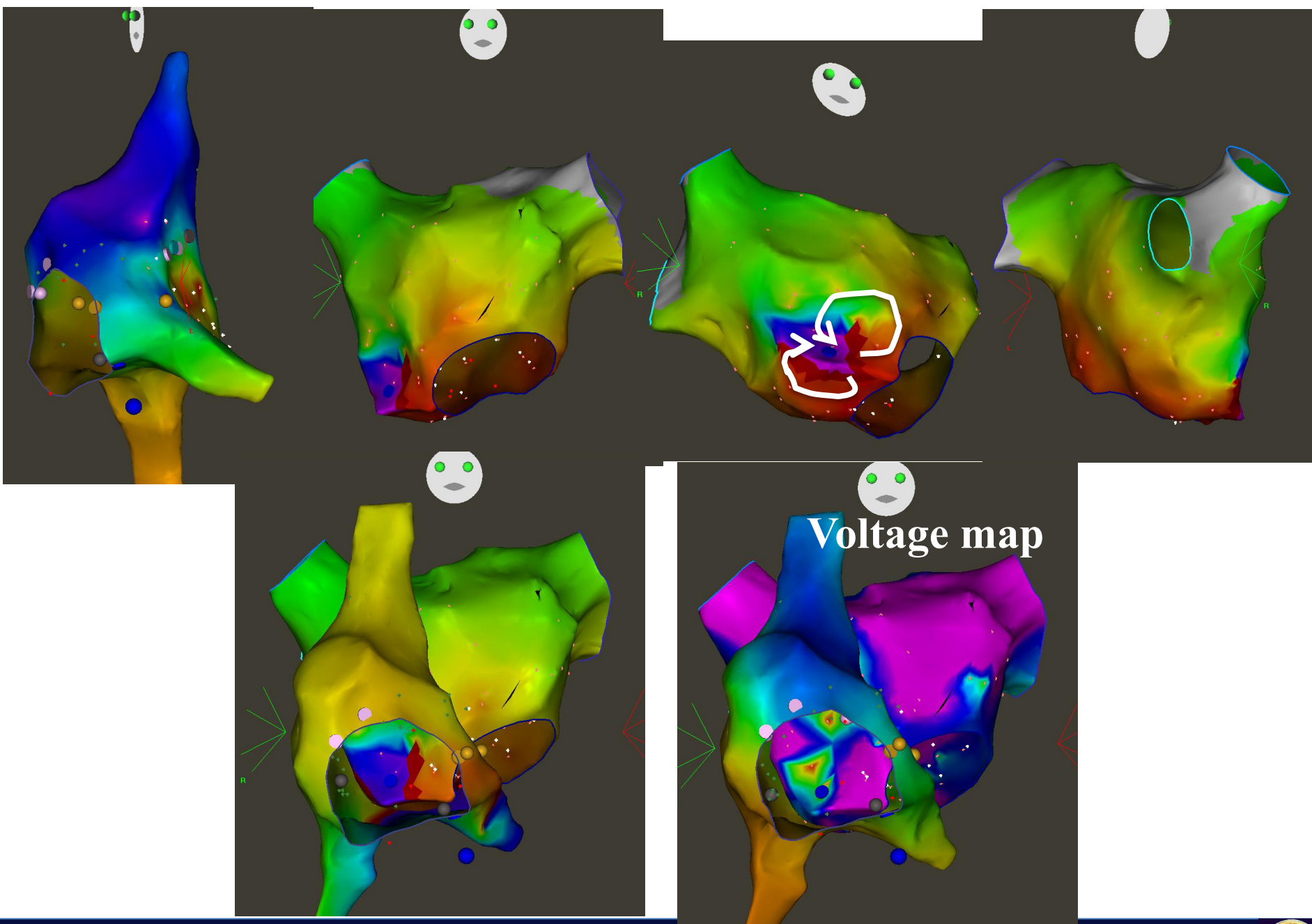


Arrhythmia's onset after surgery (years)	19 ± 12
Symptoms	40/46
Arrhythmia	
Typical atrial flutter	22/46
Atypical atrial flutter	16/46
Atrial tachycardia	8/46

AF COEXISTED IN 40%







Voltage map



Right atrium activation map in SR

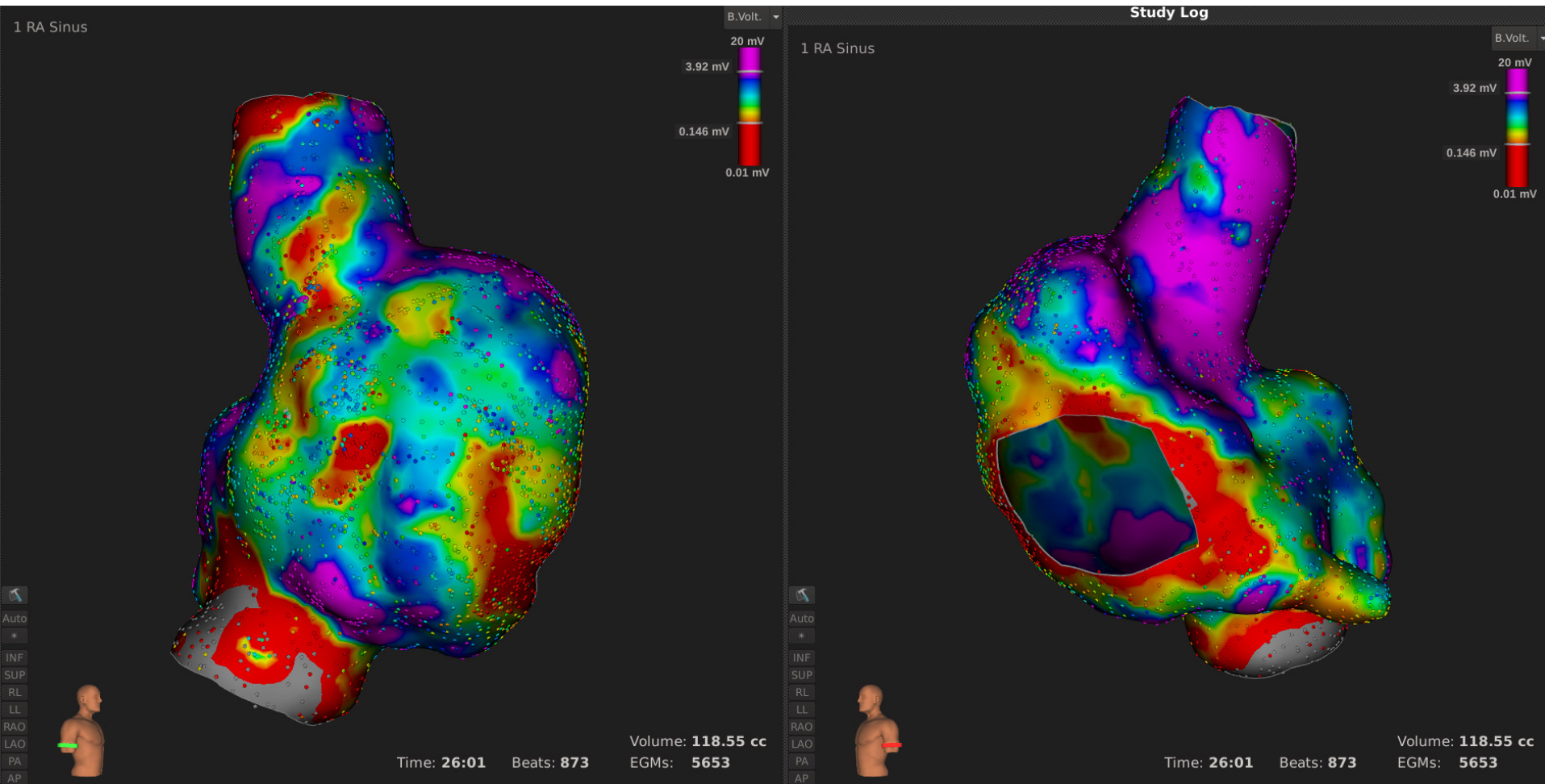


Right atrium activation map

Lines of block



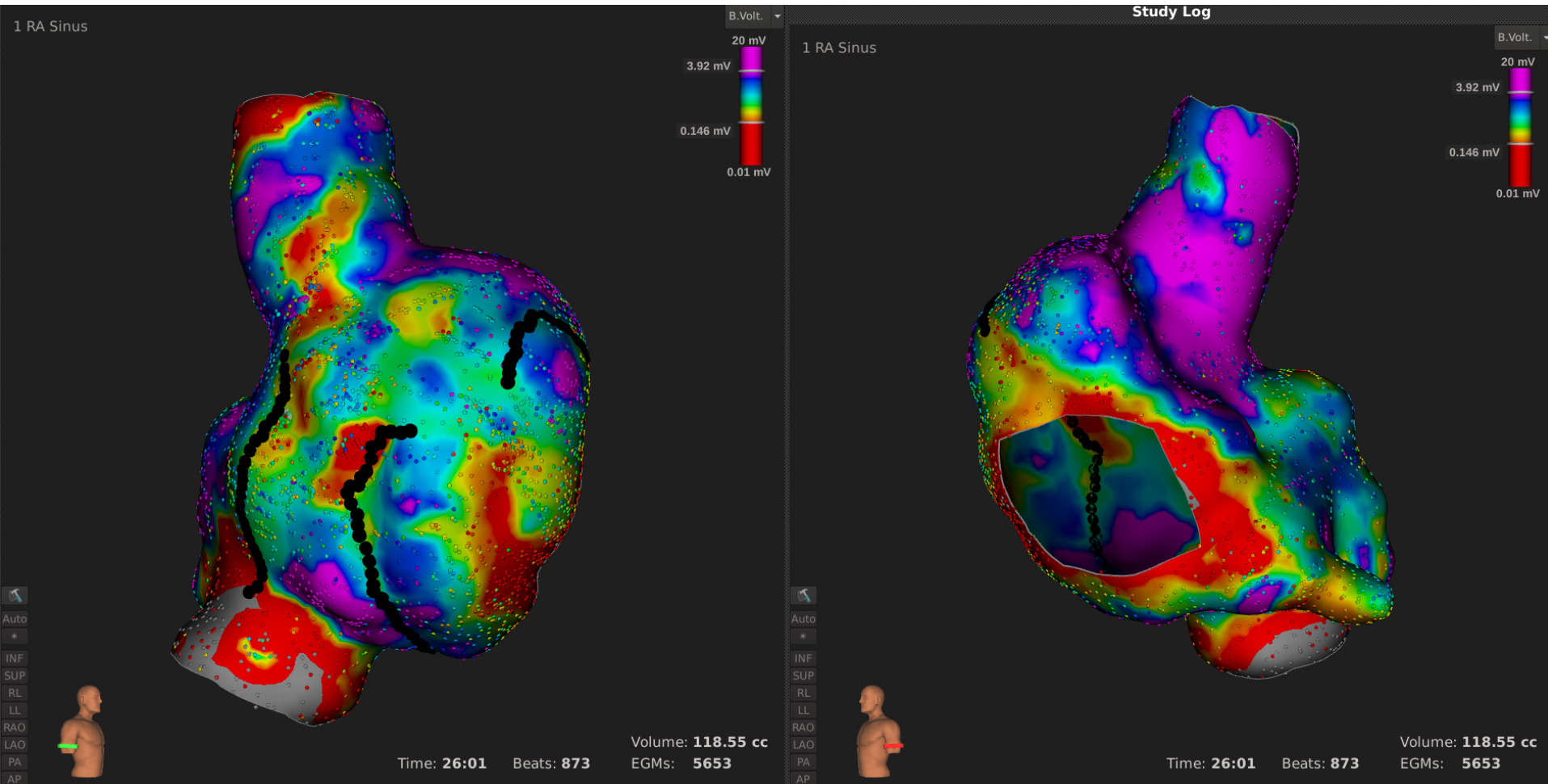
Right atrium voltage map



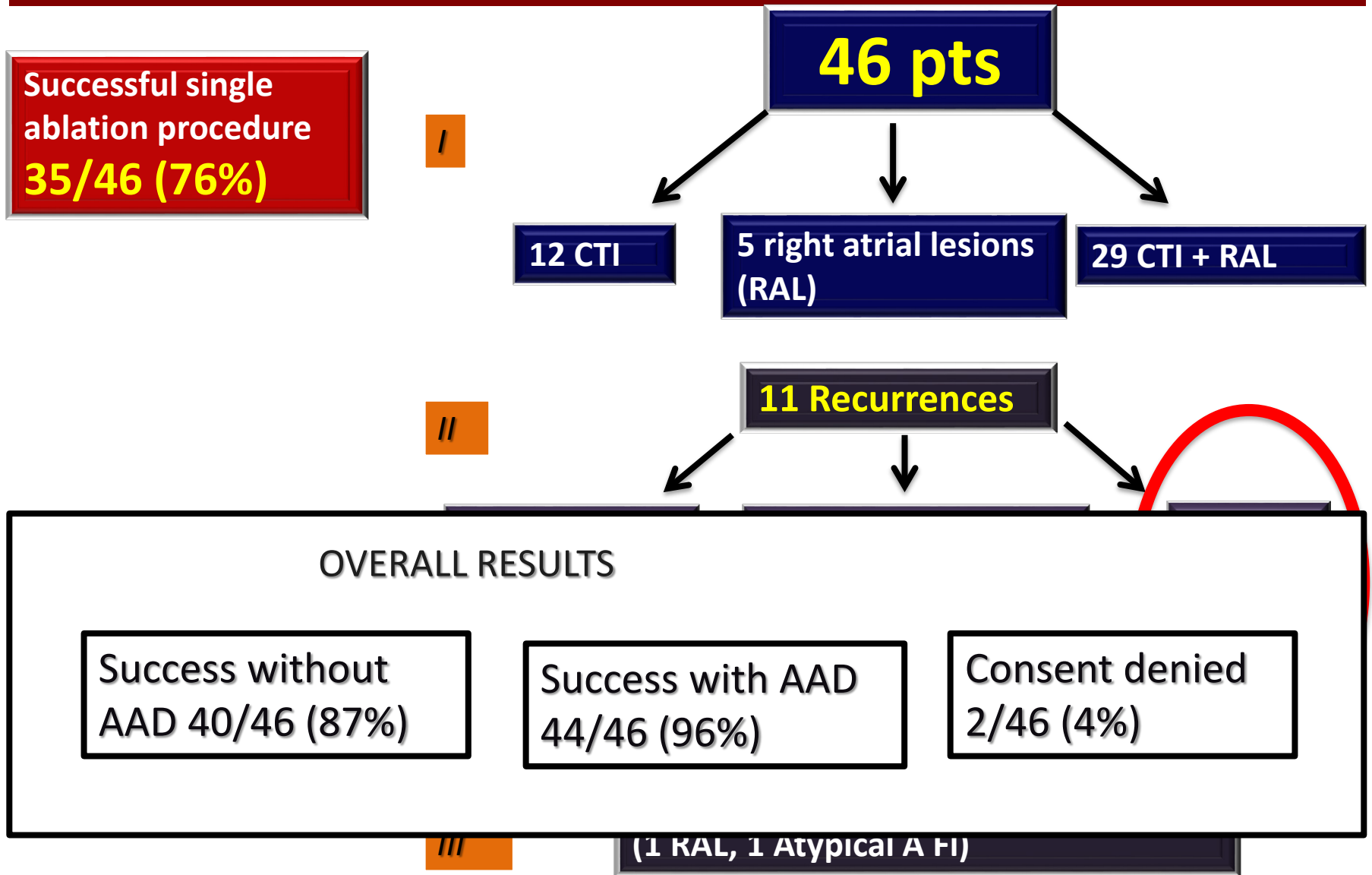
Right atrium voltage map

Lines of block

Detailed map allowed to tailor the ablation strategy



Long Term Ablation Results (FU 7 ± 3.8 y)





Europace (2014) **16**, 1800–1807
doi:10.1093/europace/euu076



CLINICAL RESEARCH
Electrophysiology and ablation

Very long-term results of electroanatomic-guided radiofrequency ablation of atrial arrhythmias in patients with surgically corrected atrial septal defect

...in recent years right sided arrhythmias ablation in GUCH has been managed fluoroles.

Scaglione et al EUROPACE 2014

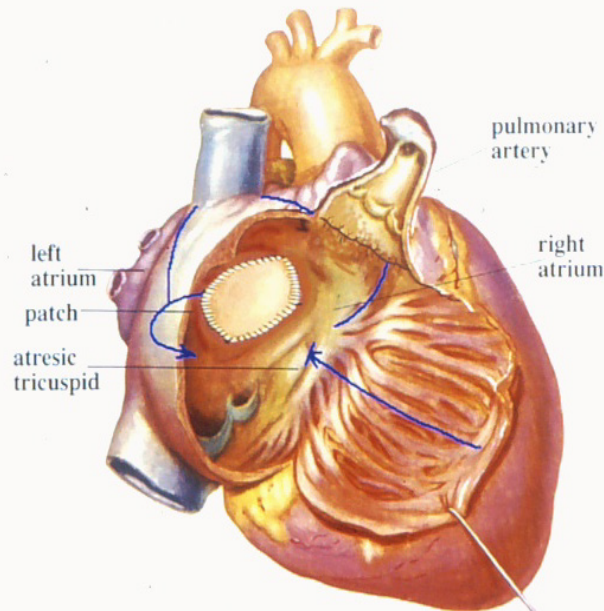


RF Ablation after Fontan intervention

Author	Journal	Year	N° pts	Type of arrhythmias	Success rate	Recurrences rate	Complications rate
Triedman	JACC	1997	22	IART	73%	52%	NA
Collins	AmJC	2000	43	IART	72%	NA	NA
Betts	Circul	2000	5	IART	60%	60%	NA
Hebe	Pediatr Cardiol	2000	11	IART	54%	27%	NA
Triedman	JACC	2002	63	IART	66-79%	52%	NA
Yap	JCE	2012	11	IART	45%	50%	NA
Anguera	AmJC	2015	8	FLA AT	88%	41%	5.4%



modified Fontan operation



ASTI population

31 pts (macroreentry)

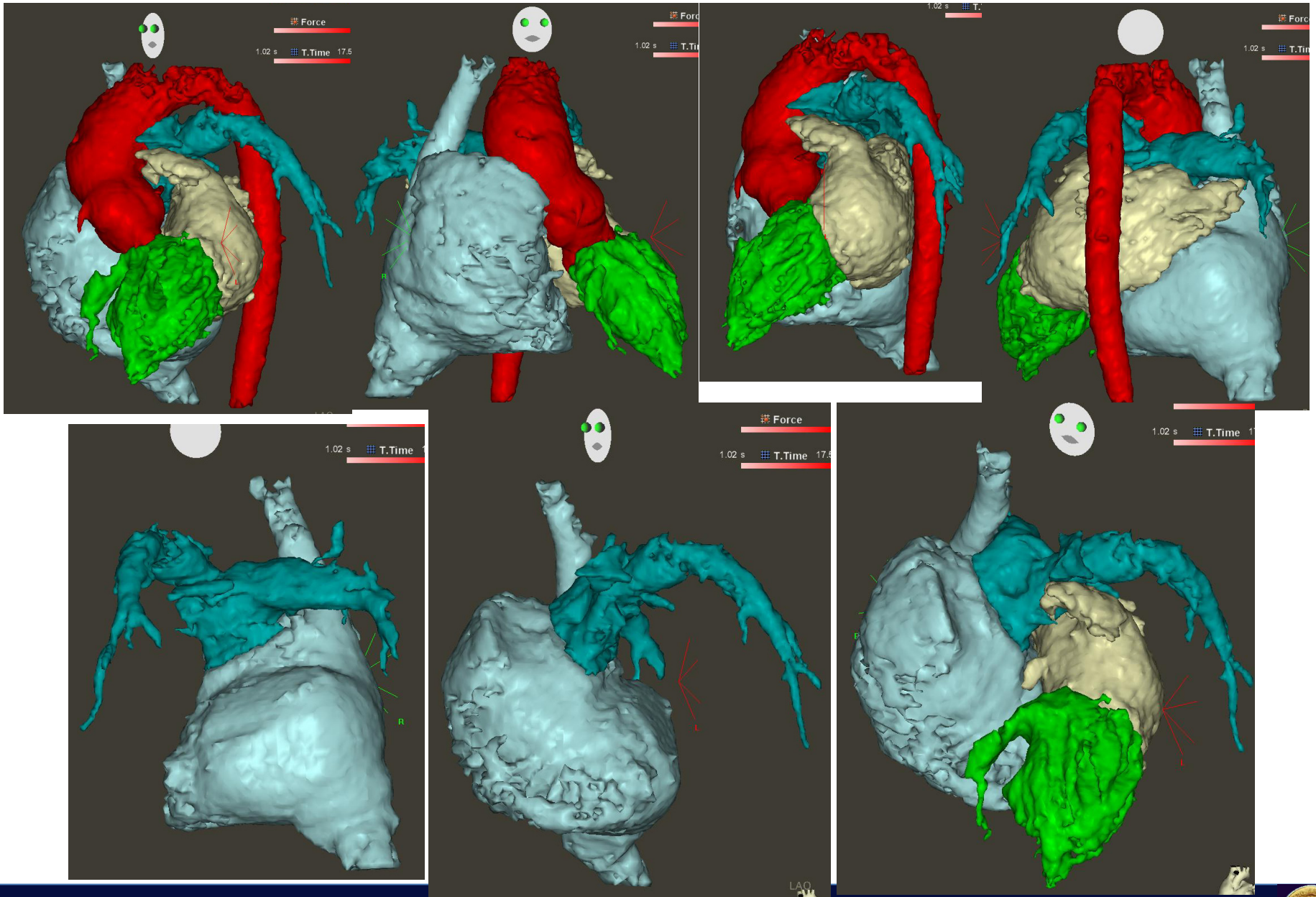
97 % acute success

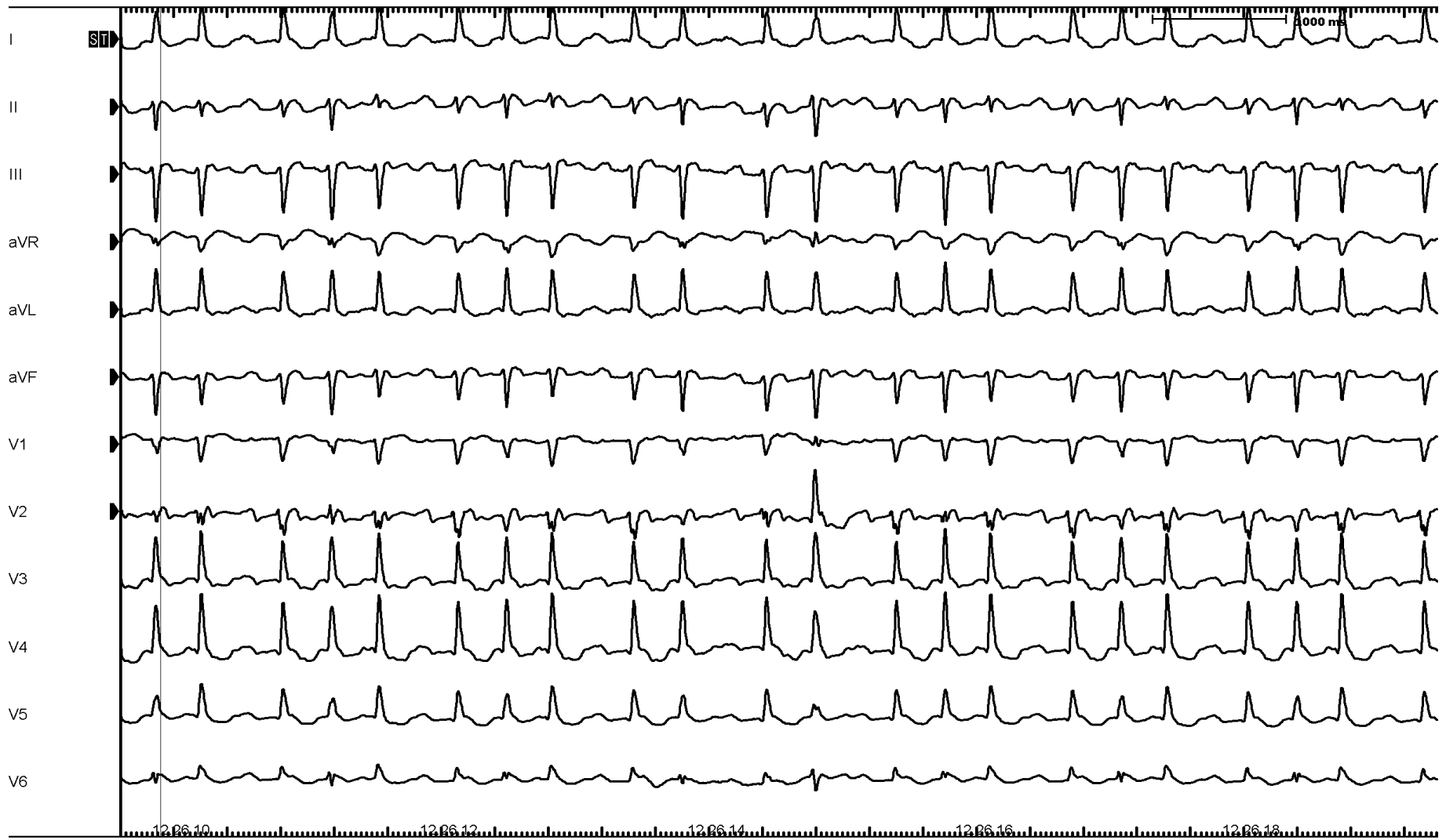
24% recurrences (1° Procedure)

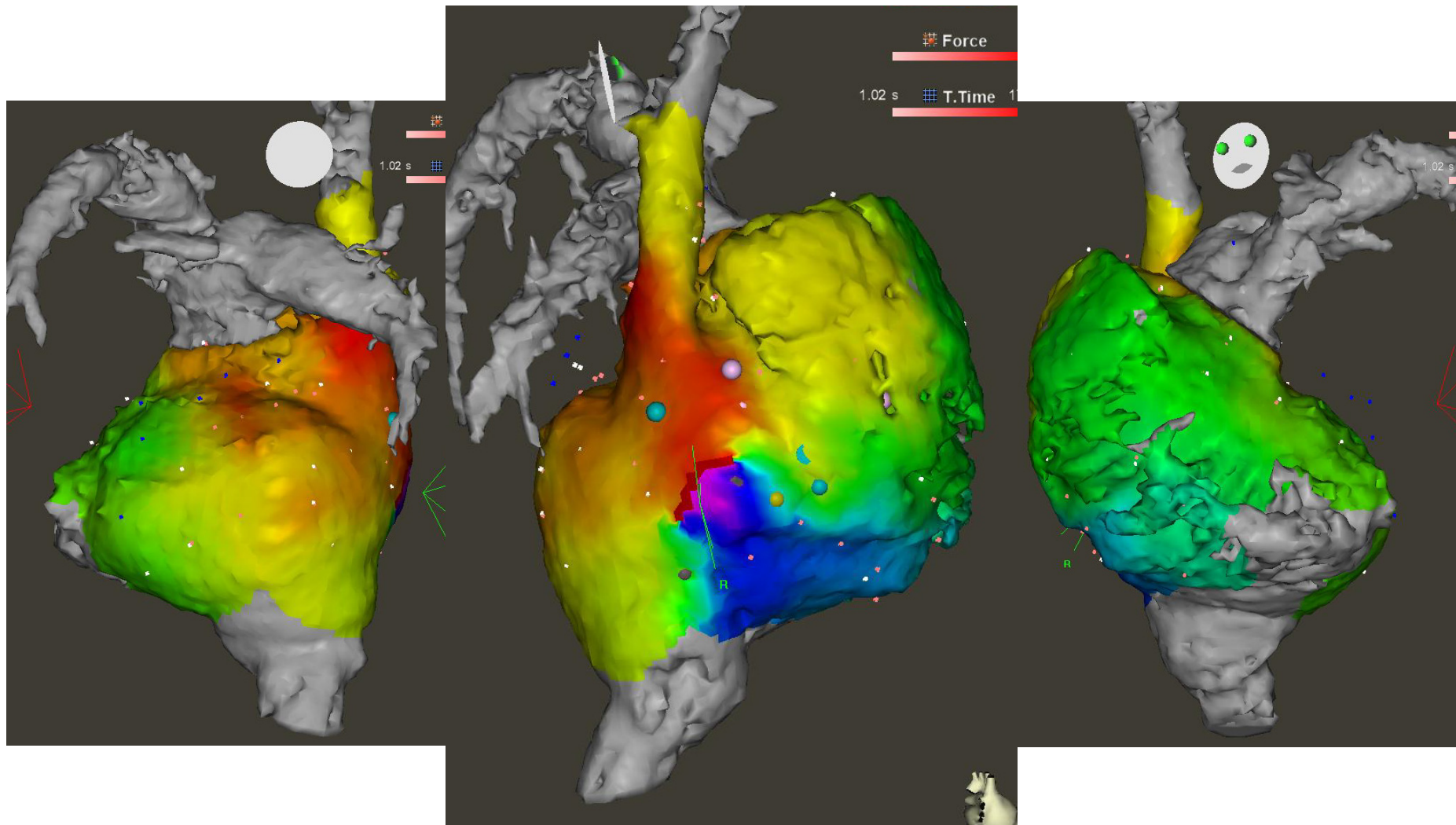
No complications

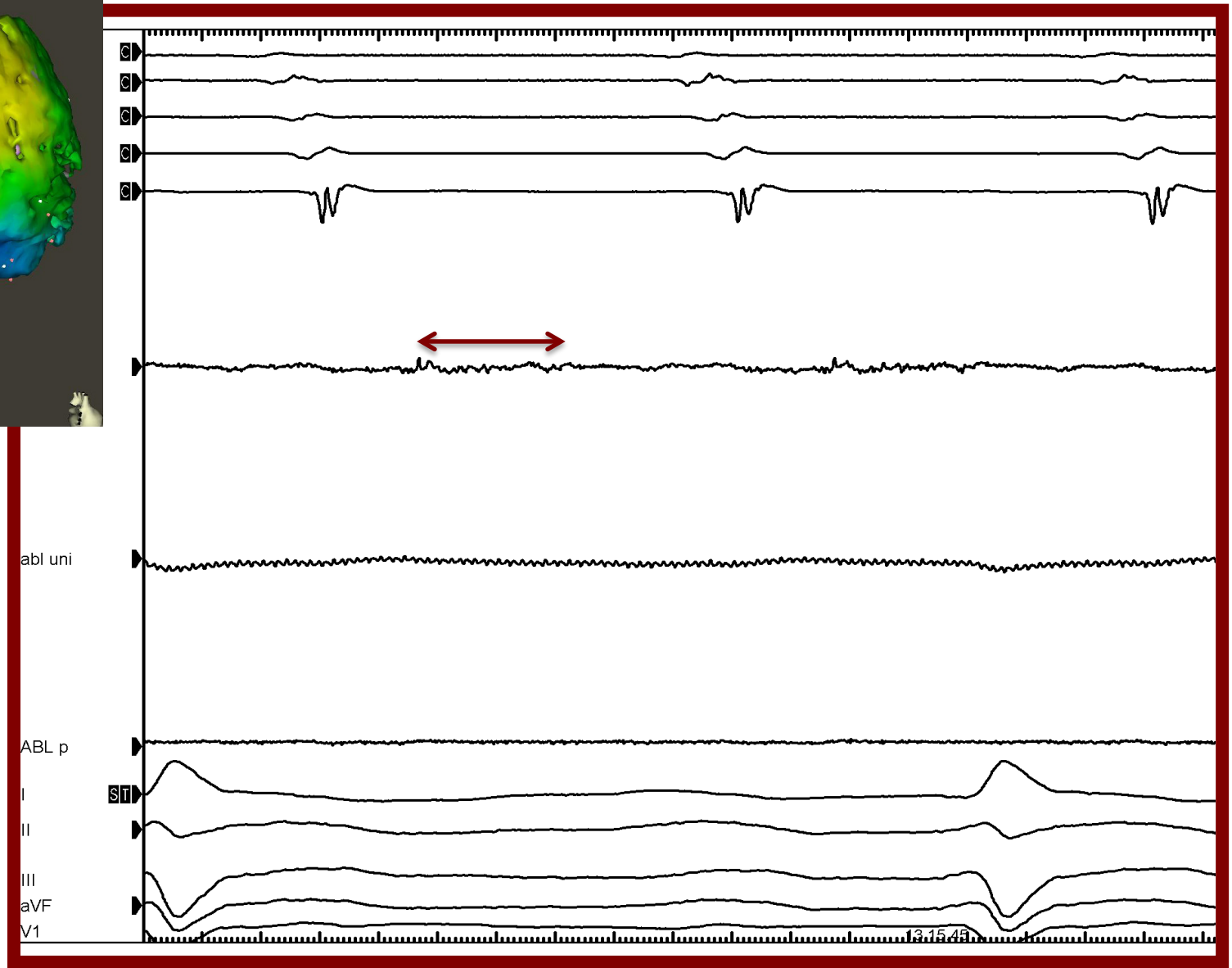
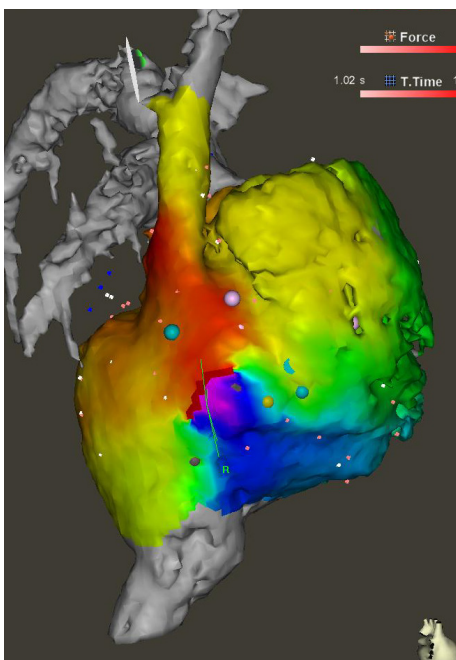


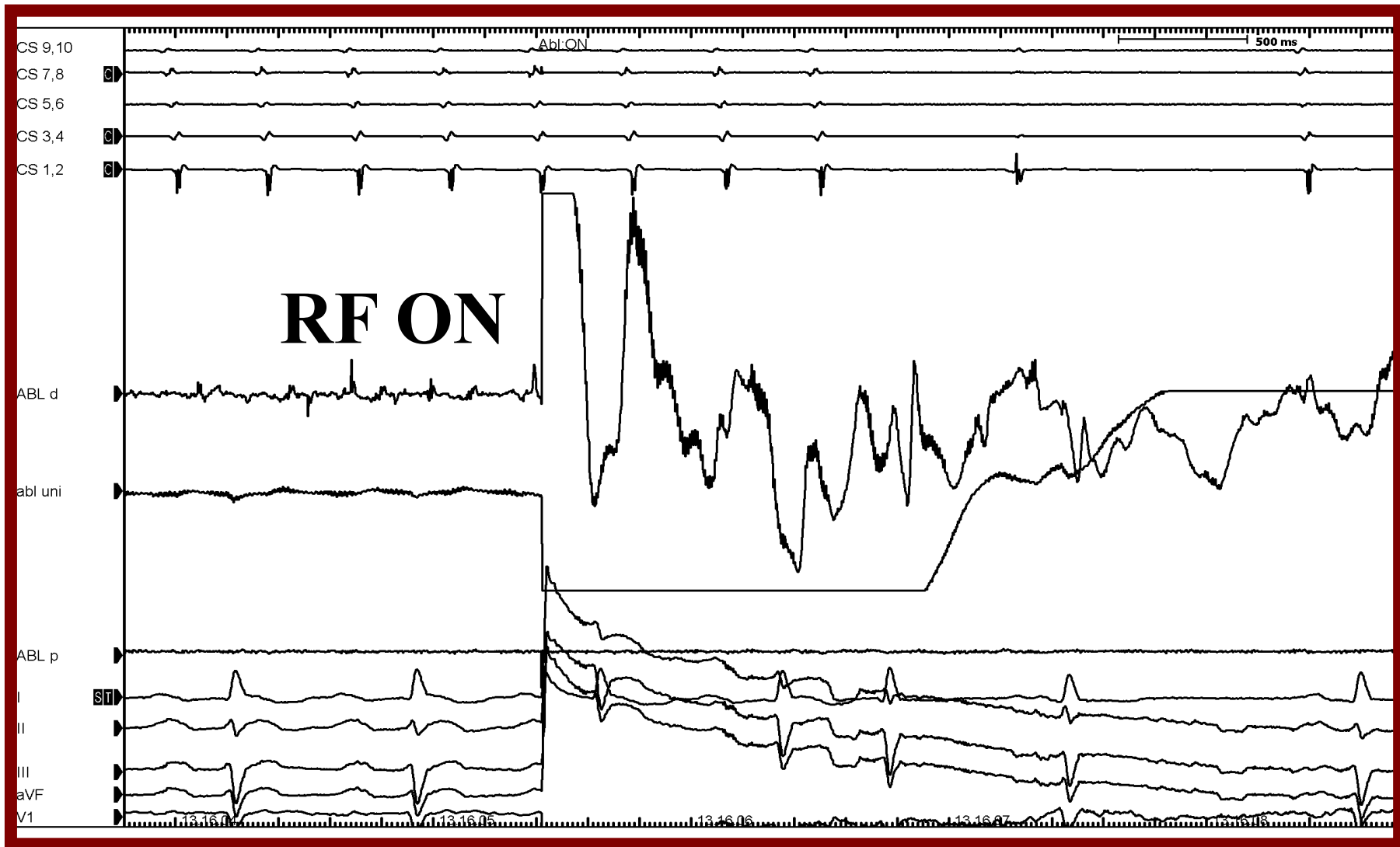
Fontan





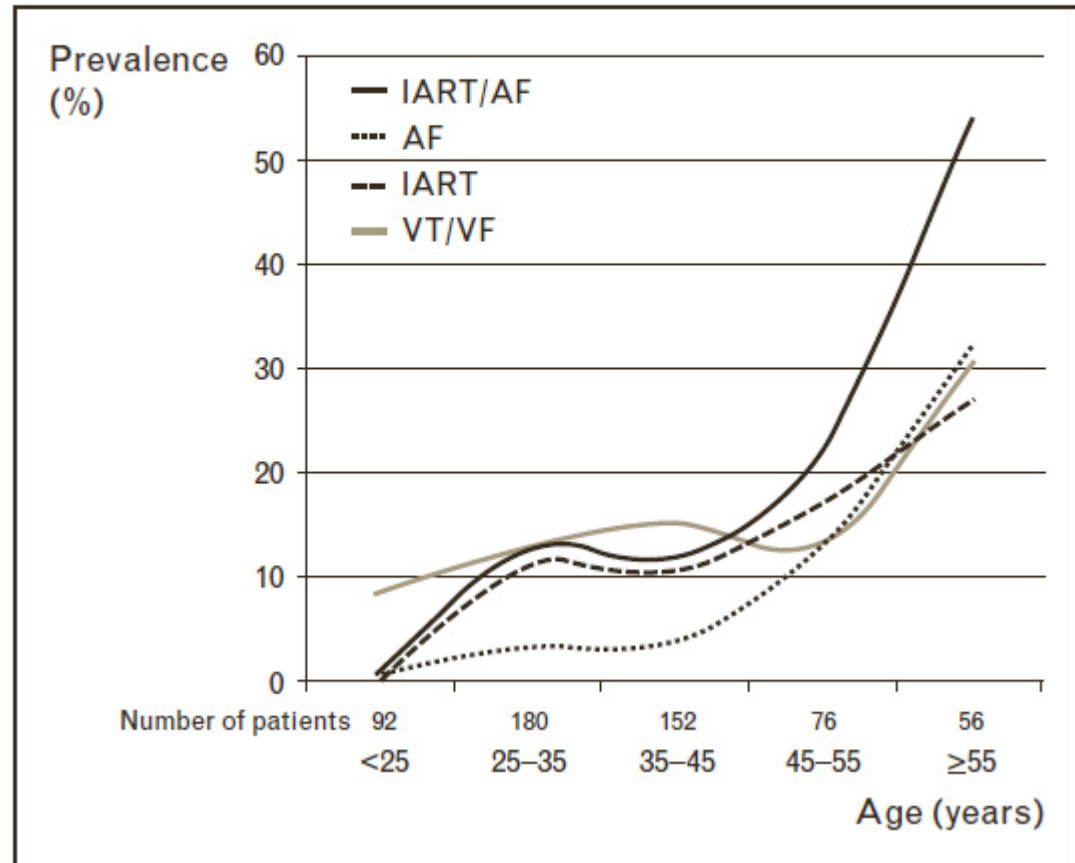
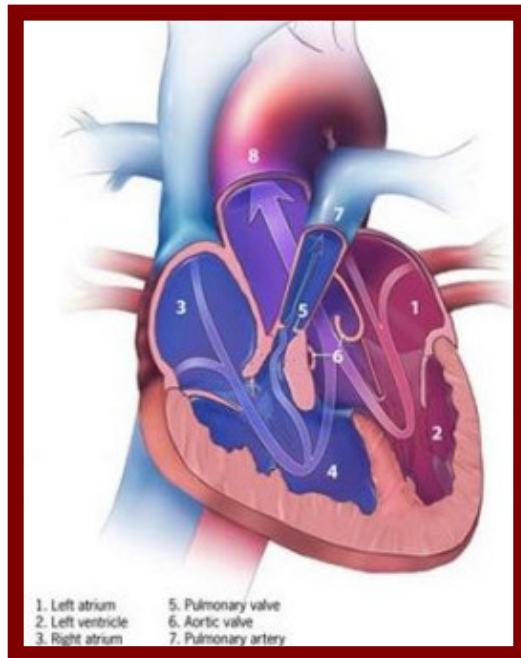






Management of arrhythmias in patients with tetralogy of Fallot

Figure 1 Prevalence of tachyarrhythmias in surgically repaired tetralogy of Fallot according to age



Le Gloan L., Current Opinion in Cardiology, 2011

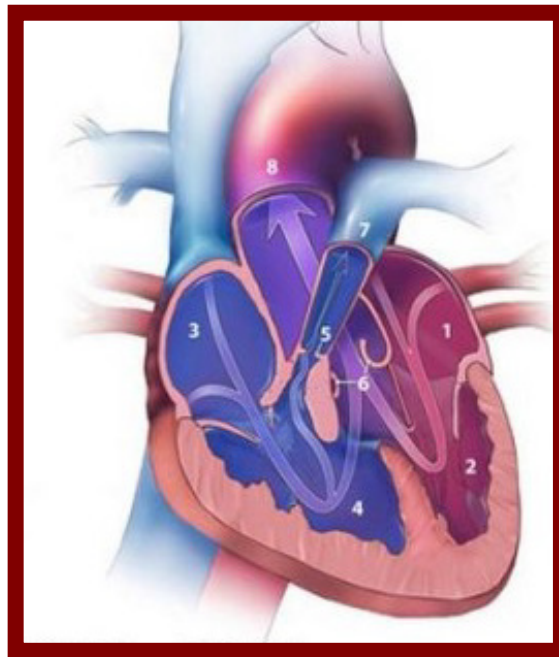


ATRIAL ARRHYTHMIAS

RF Ablation in repaired ToF

Author	Journal	Year	N° pts	Type of arrhythmias	Success rate	Recurrence rate	Complications rate
Triedman	JACC	1997	8	IART	73%	NA	38%
Collins	AmJC	2000	10	IART	71%	18%	NA
Triedman	JACC	2002	37	IART	66-79%	36%	16%
Anguera	AmJC	2015	15	FLA (CTI- dep/non) AT	88%	41%	5.4%





ASTI population

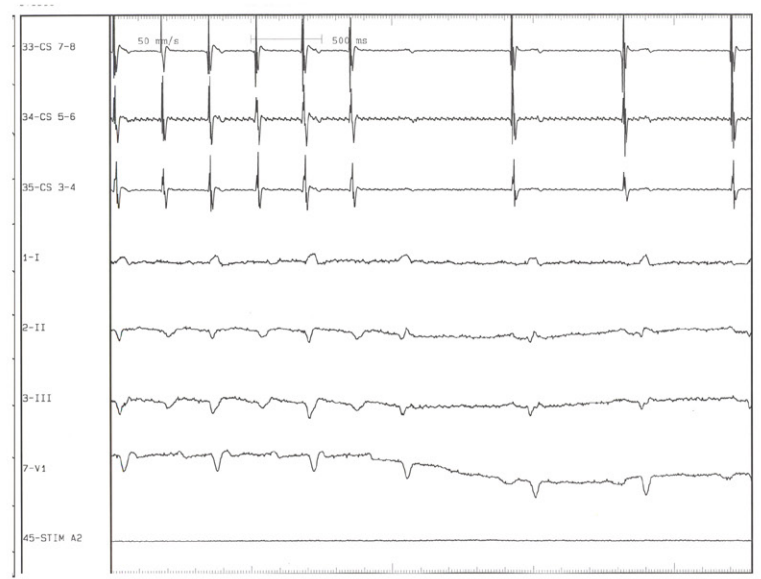
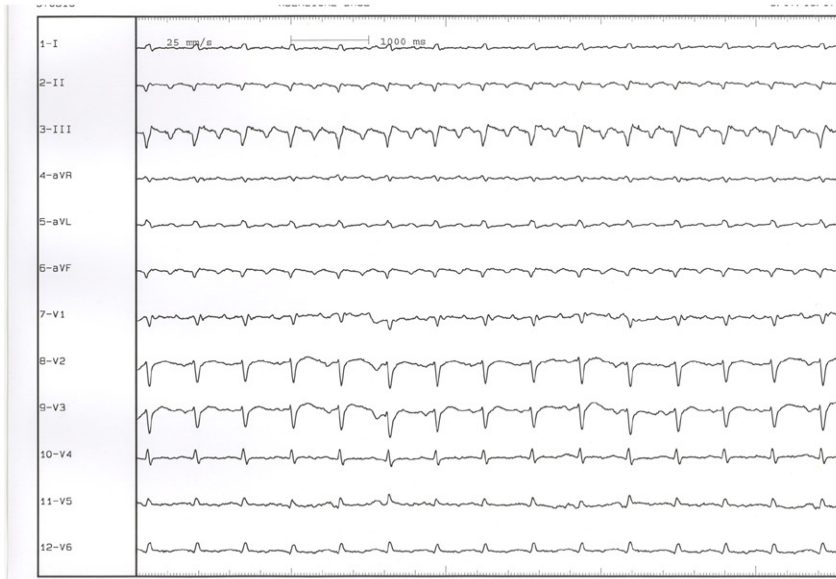
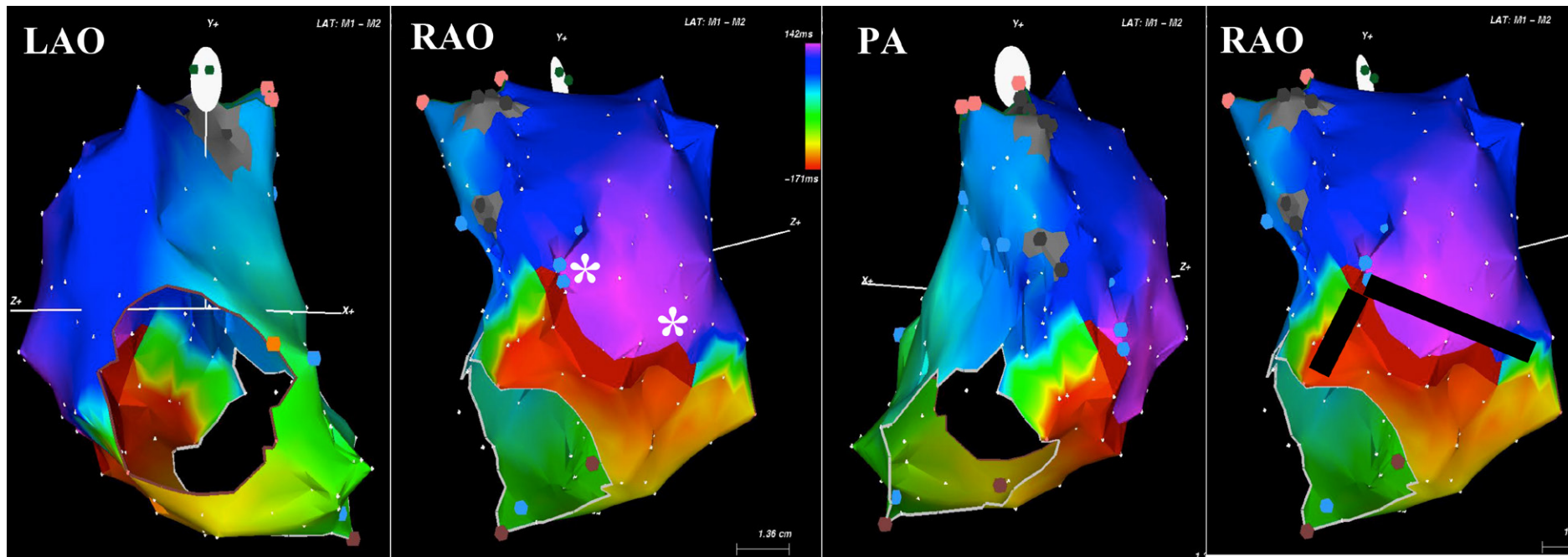
28 pts

97 % acute success

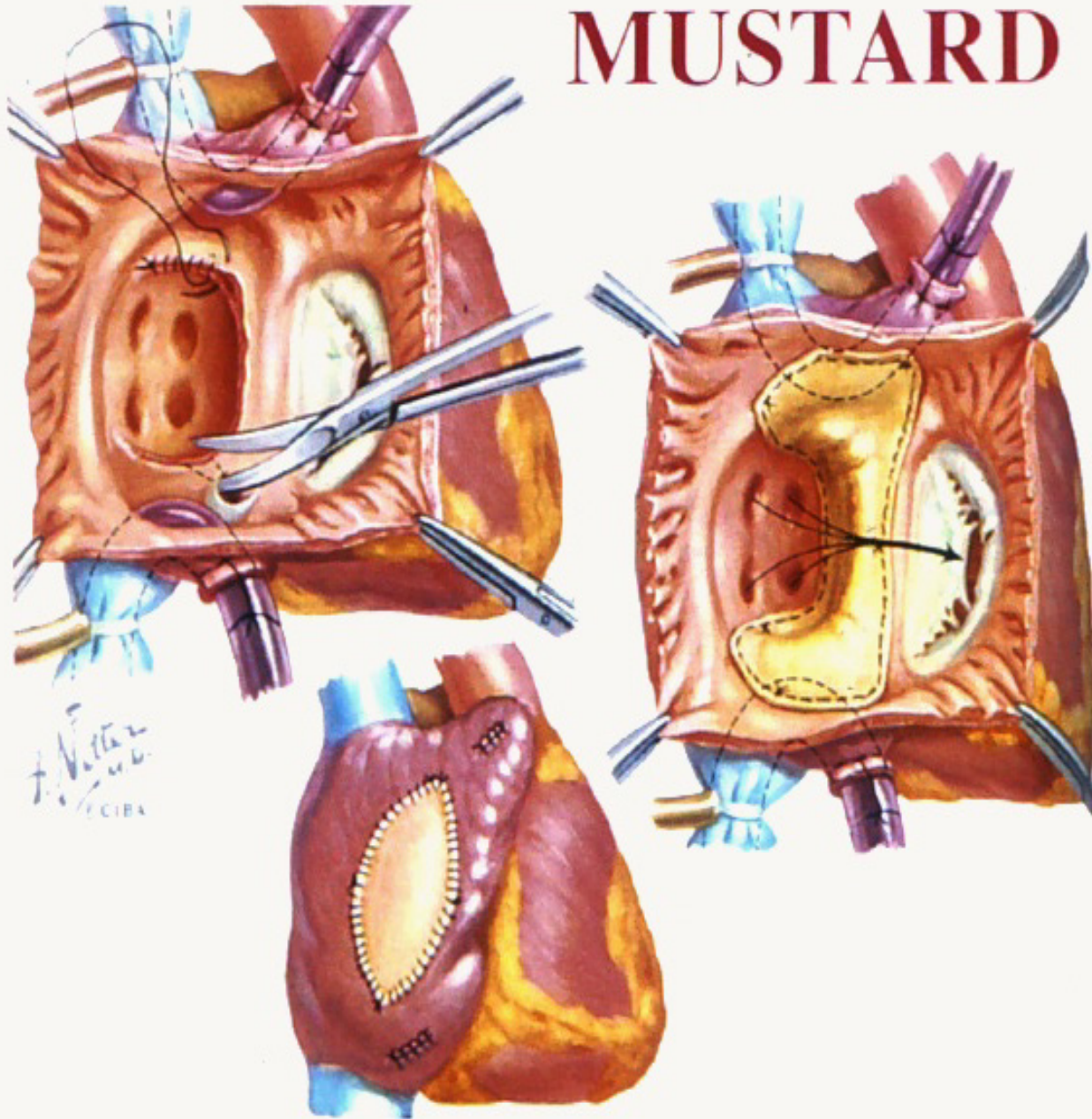
18% recurrences (1^o Procedure)

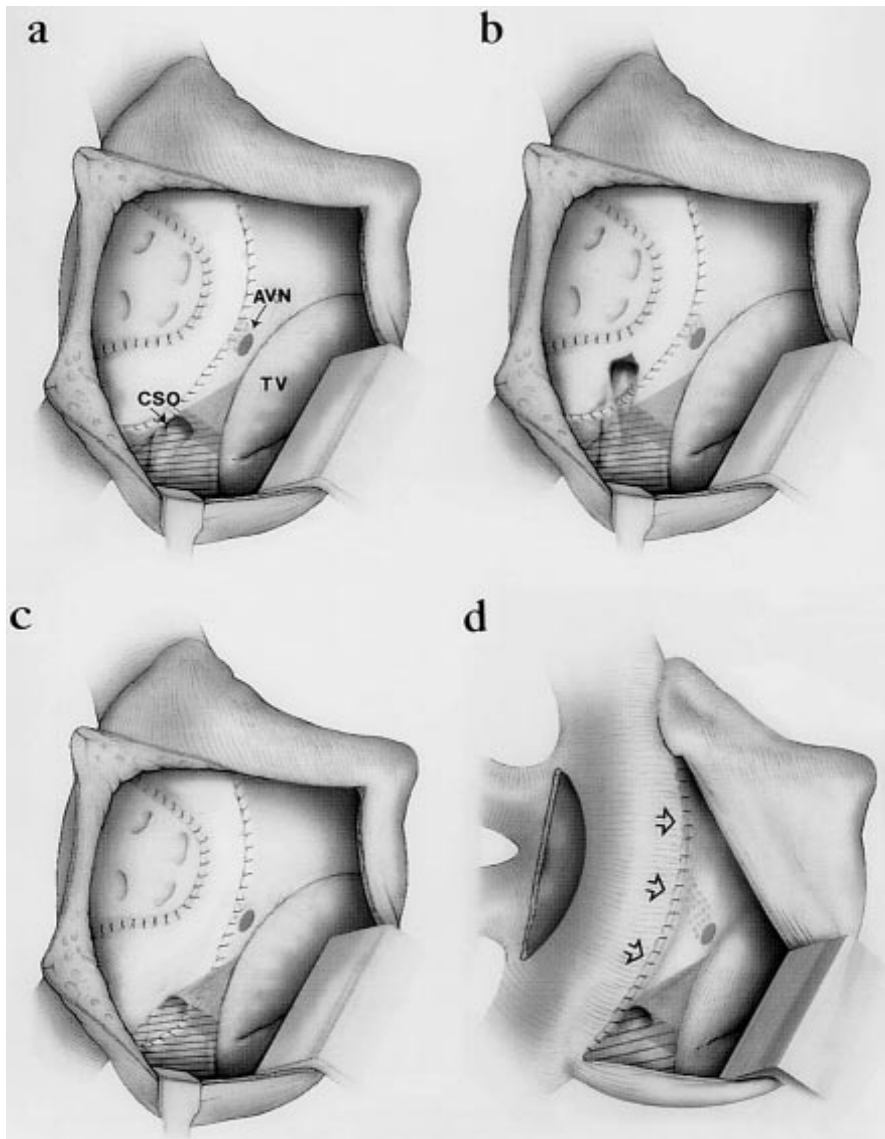
No complications



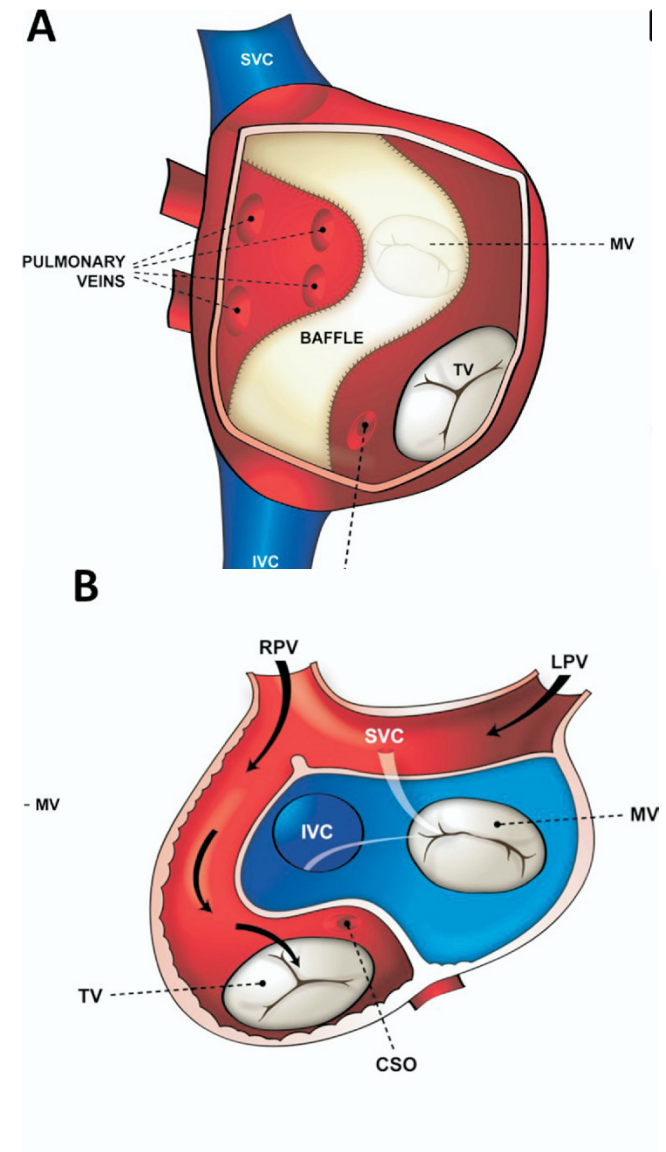


MUSTARD





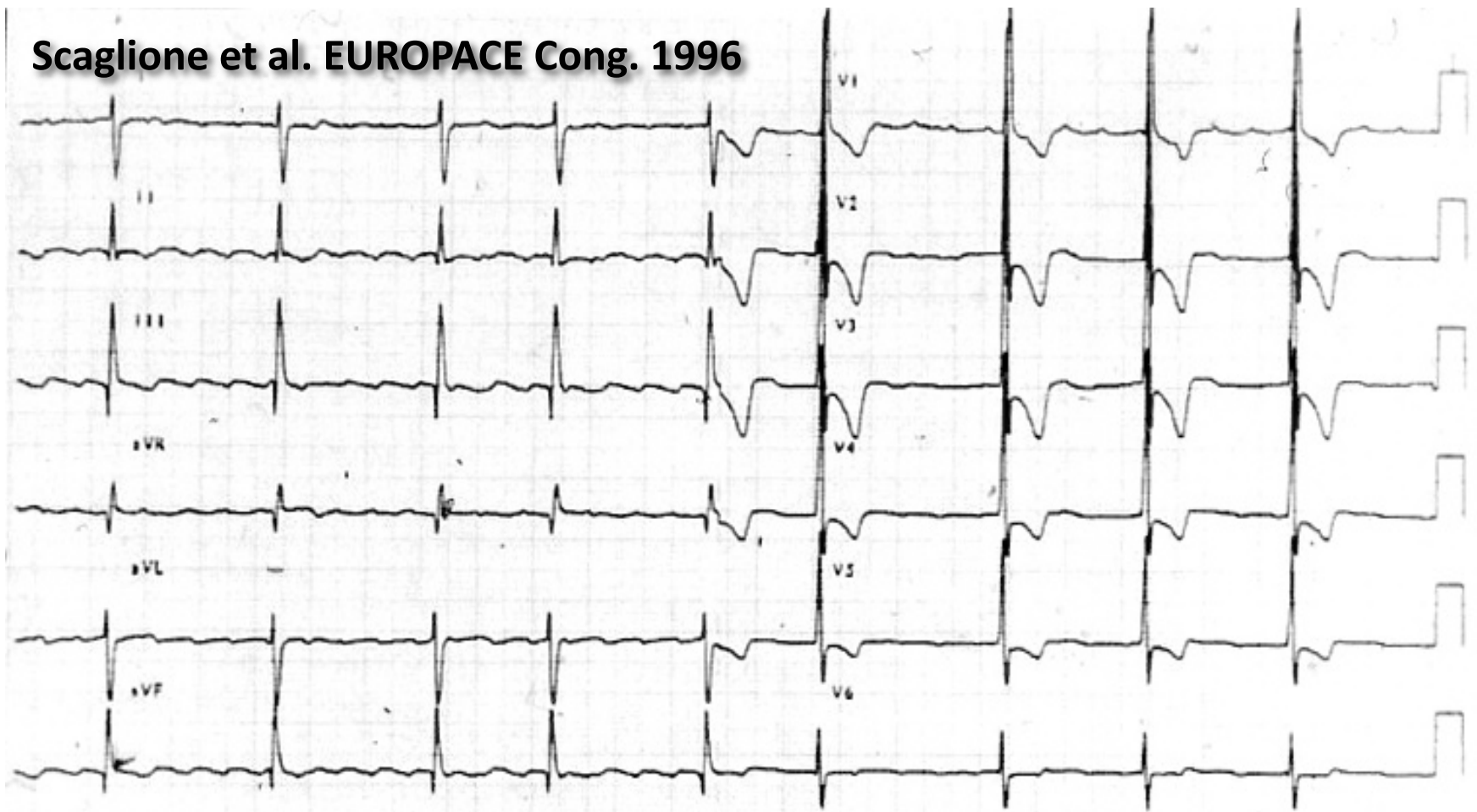
Kanter et al JACC 2000



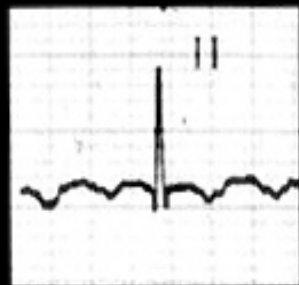
Khairy et al Heart Rhythm 2009



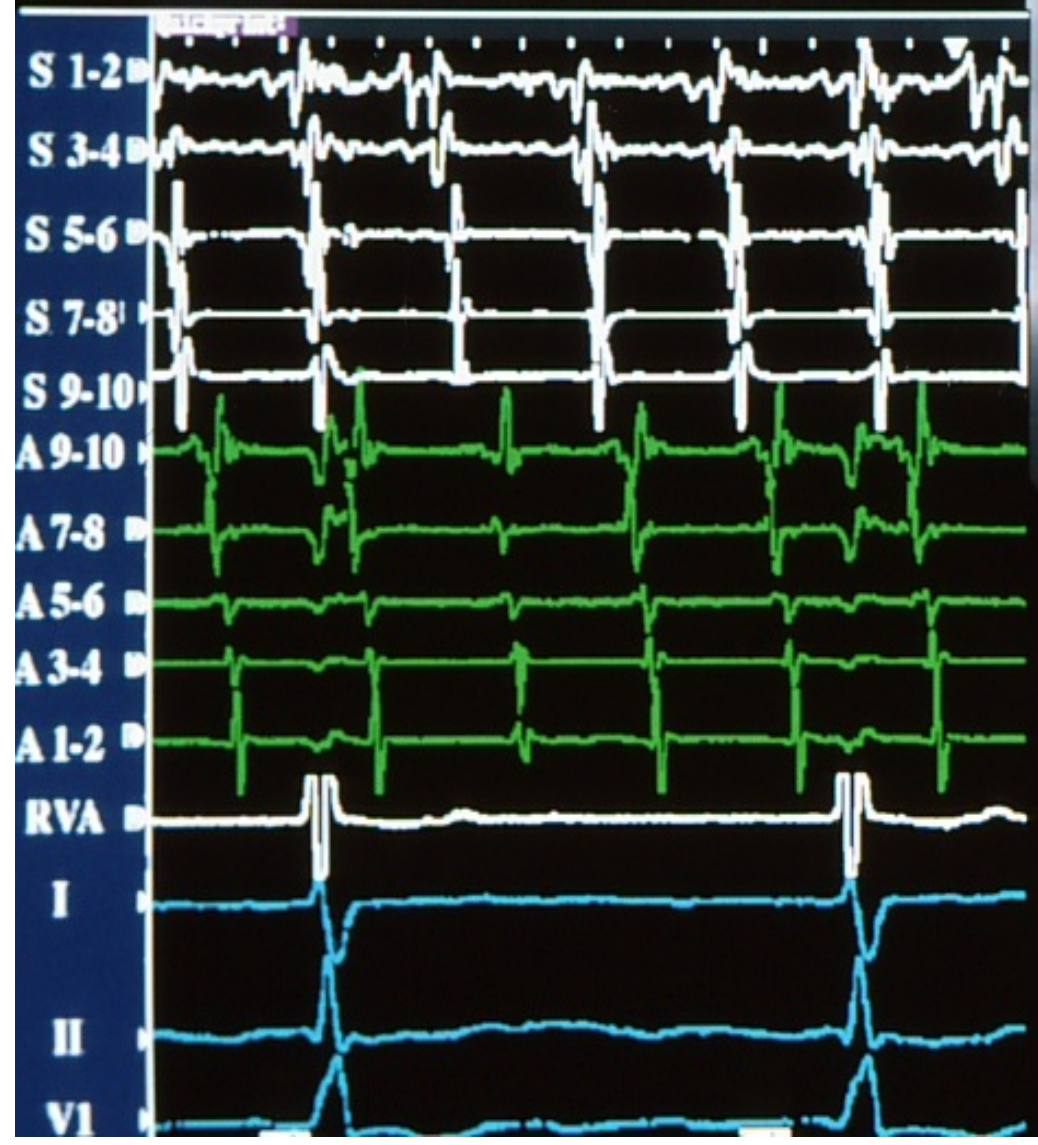
Scaglione et al. EUROPACE Cong. 1996



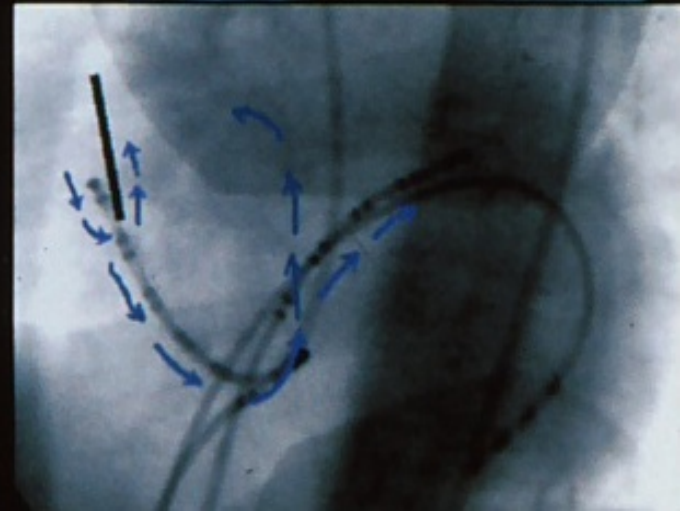
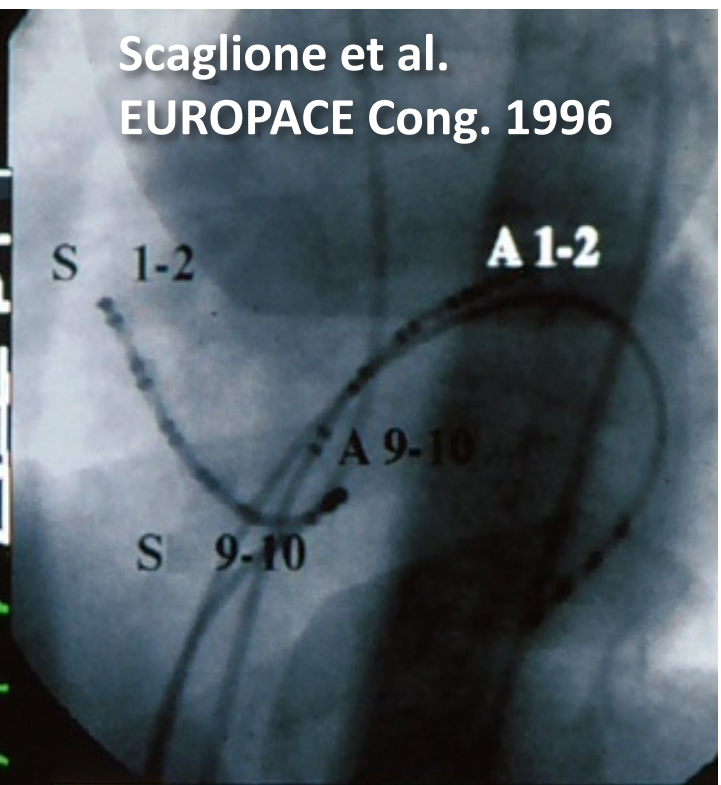
Mustard

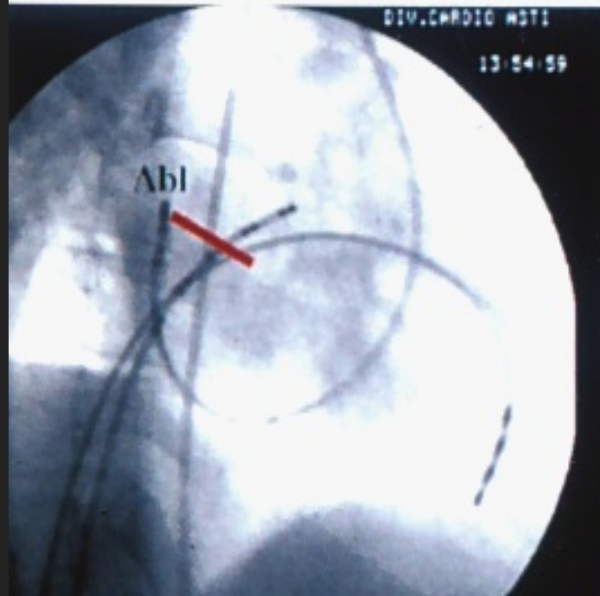
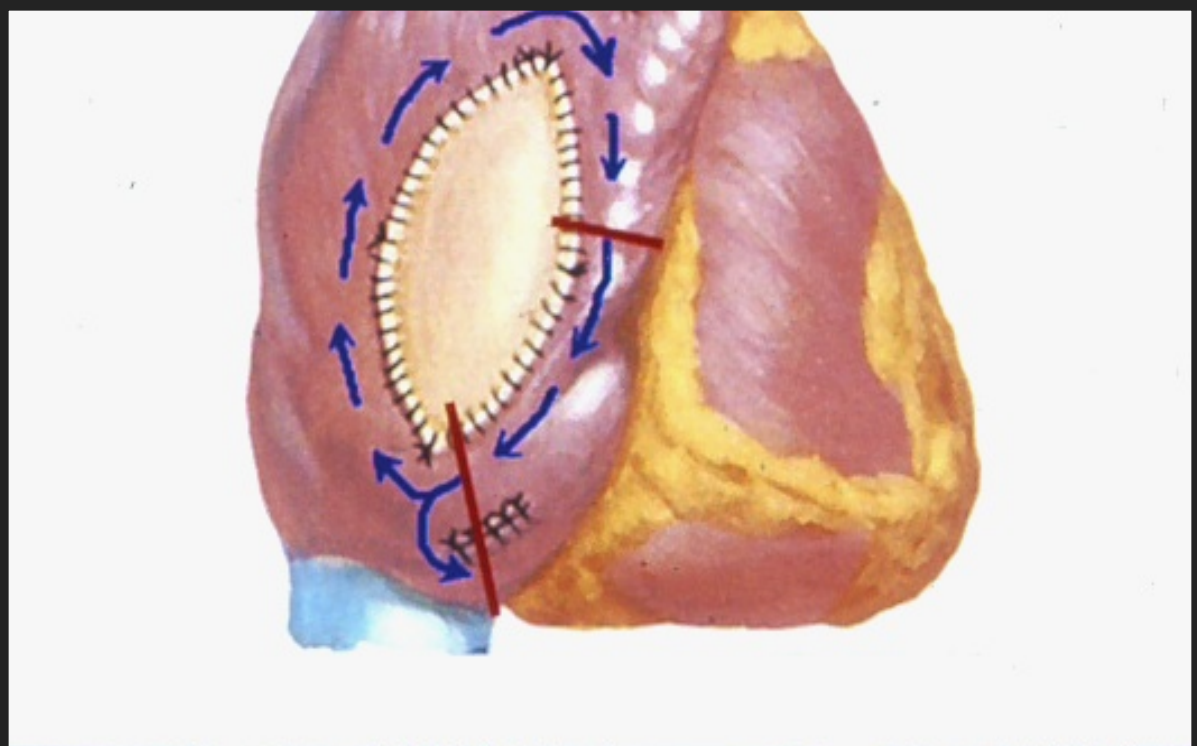


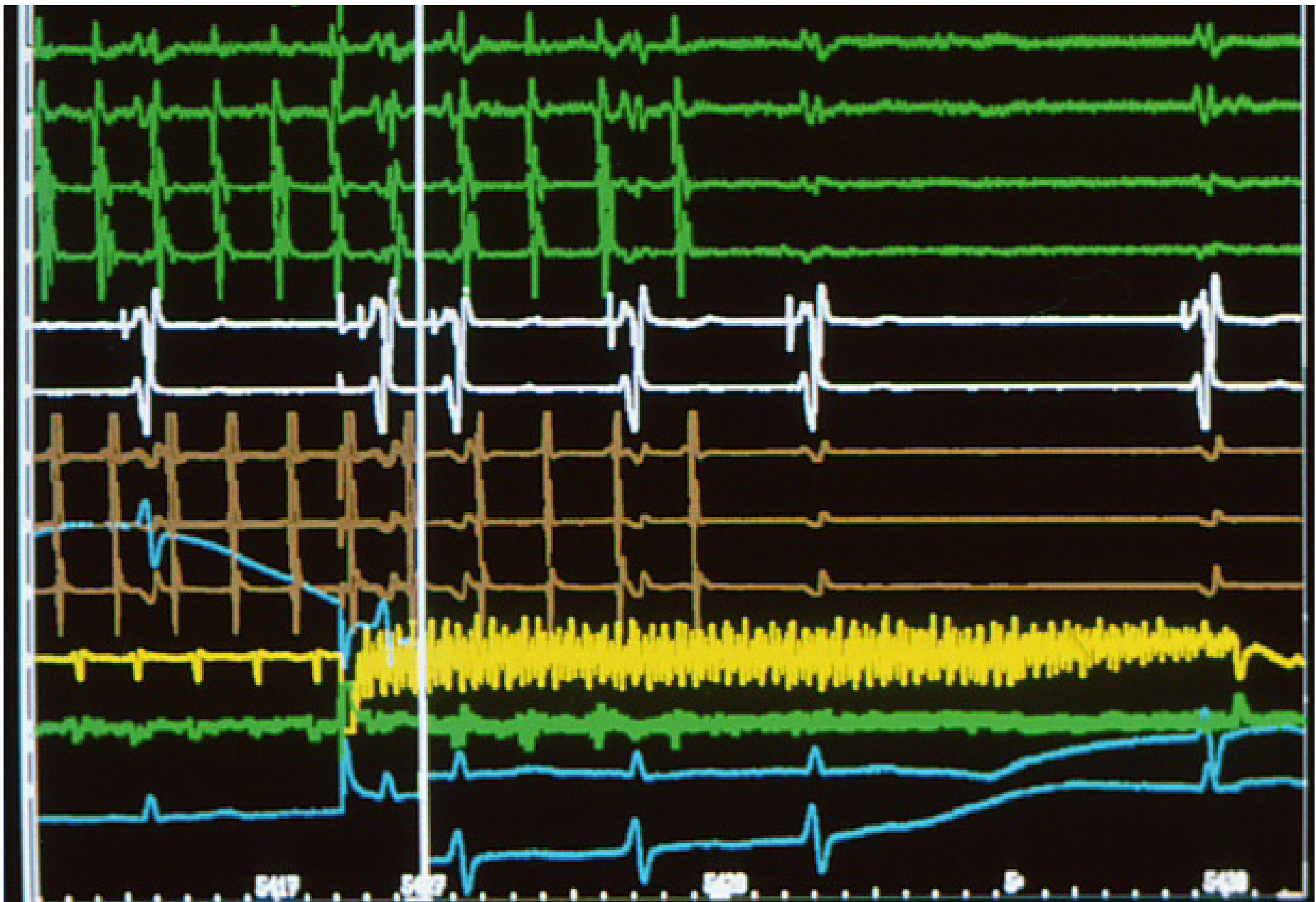
LAO 30° ENDOCARDIAL ACTIVATION



Scaglione et al.
EUROPACE Cong. 1996







The safety and efficacy of trans-baffle puncture to enable catheter ablation of atrial tachycardias following the Mustard procedure: A single centre experience and literature review [☆]

Literature describing cases of trans-baffle puncture following atrial switch surgery.

Study (ref. #)	Year	Electrophysiological procedure?	No. of trans-baffle punctures	No. of successful punctures	Intra-operative imaging	Complications
El-Said et al. [[]]	2000	No	16	15	Fluoroscopy only	None
Perry et al. [[]]	2003	Yes	2	2	Fluoroscopy only	None
Schwagten et al. [[]]	2009	Yes	1	1	Fluoroscopy + TOE	None
Peichl et al. [[]]	2009	Yes	3	3	Fluoroscopy + ICE	None

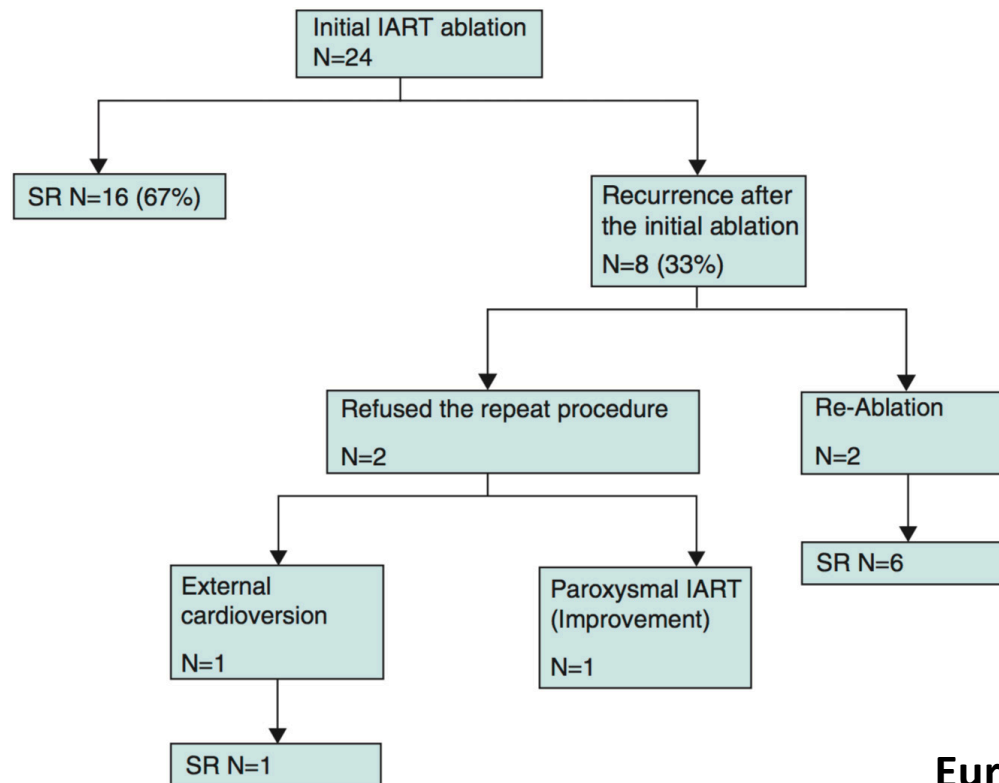
Procedure number	Patient number	Age (years)	Major cardiac co-morbidity	Number of ATs ablated	Number of ATs requiring PVA ablation	ATs (cycle lengths, ms)	Termination of all ATs with ablation	Complications
1	1	35	Mechanical TVR, VSD	3	1	Focal PVA (272–290) Focal SVA (250) MRE with SVA isthmus (246)	Yes	None
2	2	38	Biventricular dysfunction, PHT	1	1	Typical CTI flutter (320)	Yes	None
3	3	36	Severe RV dysfunction, ICD	1	1	MRE with PVA isthmus (560)	Yes	None
4	3	37	Severe RV dysfunction, ICD	2	2	Focal PVA (225) Focal PVA (350)	Yes	None
5	4	41	VSD, subpulmonary stenosis	1	1	Typical CTI flutter (280)	Yes	None
6	5	18	Nil	1	1	Typical CTI flutter (350)	Yes	None
7	6	53	Severe RV dysfunction, ICD (via persistent left SVC)	1	1	CTI dependent bi-atrial MRE (380)	Yes	None
8	7	58	Severe RV dysfunction, ICD	1	1	Typical CTI flutter (310) PVI & roof line for AF	Yes	None
9	8	39	Moderate RV dysfunction	1	1	Typical CTI flutter (280)	Yes	None

Jones et al Int J Cardiol 2013



Transbaffle catheter ablation of atrial re-entrant tachycardia within the pulmonary venous atrium in adult patients with congenital heart disease

Ulrich Krause*, David Backhoff, Sophia Klehs, Heike E. Schneider, and Thomas Paul



Europace 2016



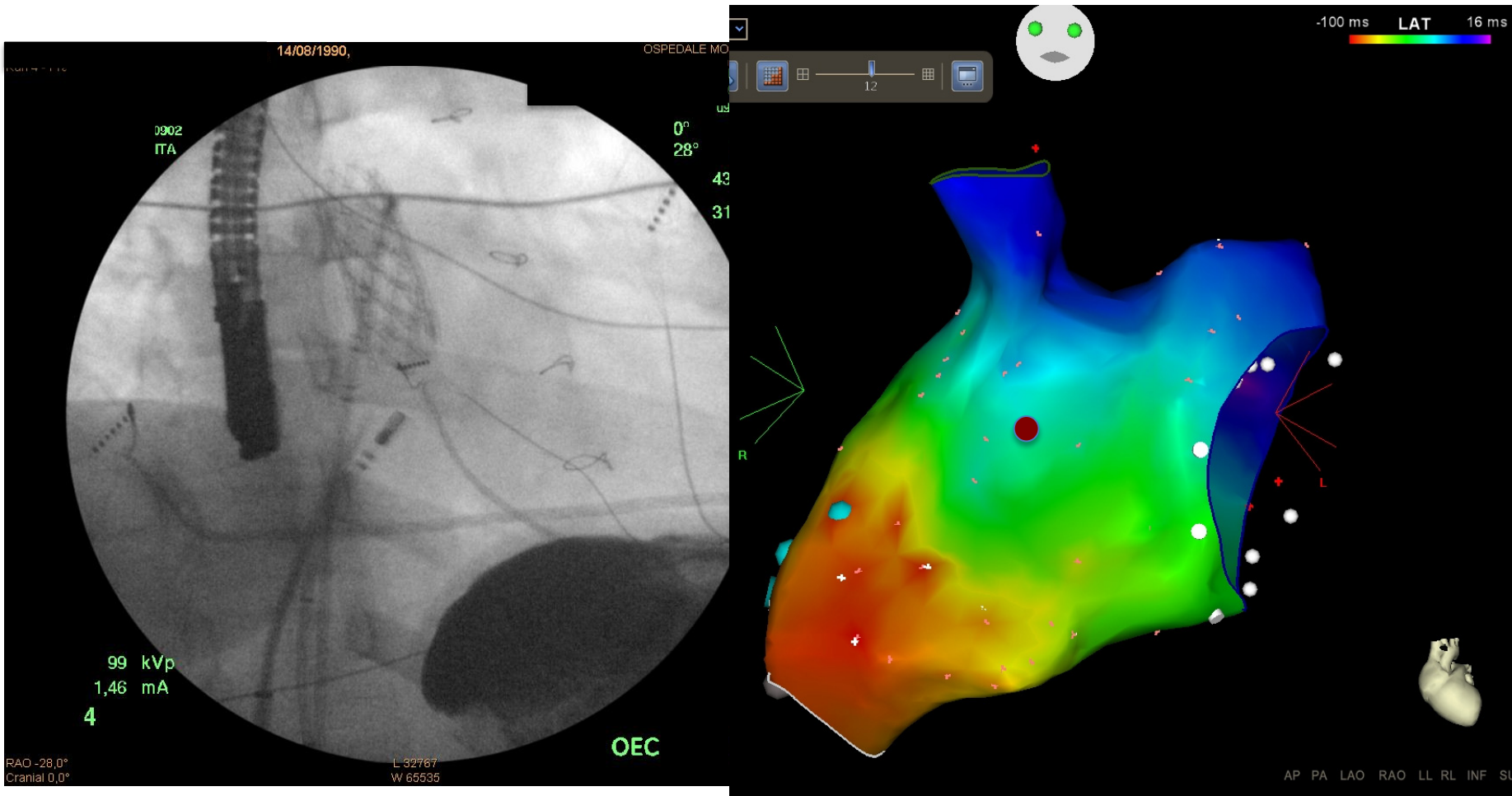
Transbaffle Mapping and Ablation for Atrial Tachycardias After Mustard, Senning, or Fontan Operations

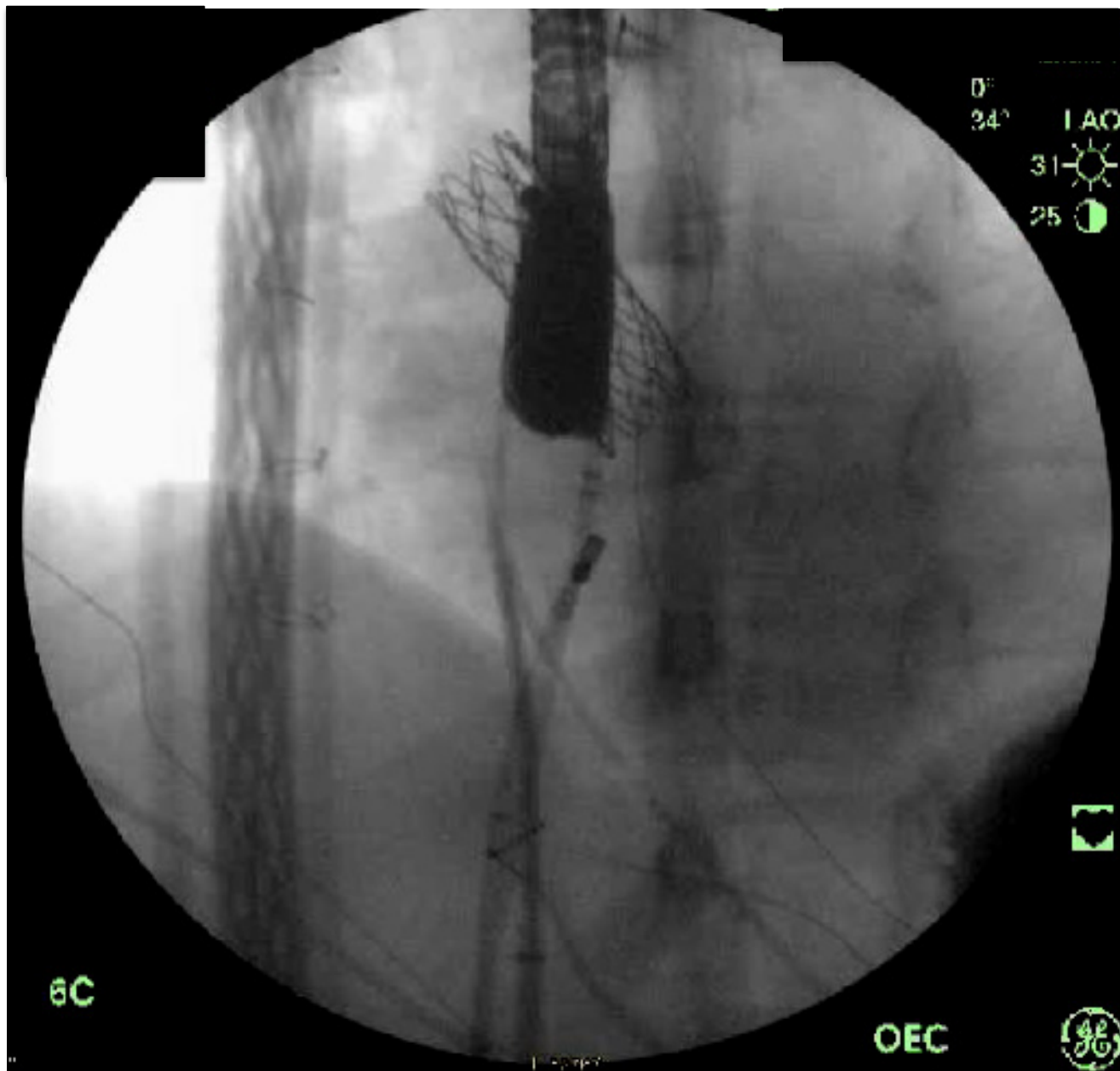
J Am Heart Ass 2013

Rafael Correa, MD; Edward P. Walsh, MD; Mark E. Alexander, MD; Douglas Y. Mah, MD; Frank Cecchin, MD; Dominic J. Abrams, MD, MRCP; John K. Triedman, MD

Severity	Adverse Event	TBA	No TBA	<i>P</i> 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



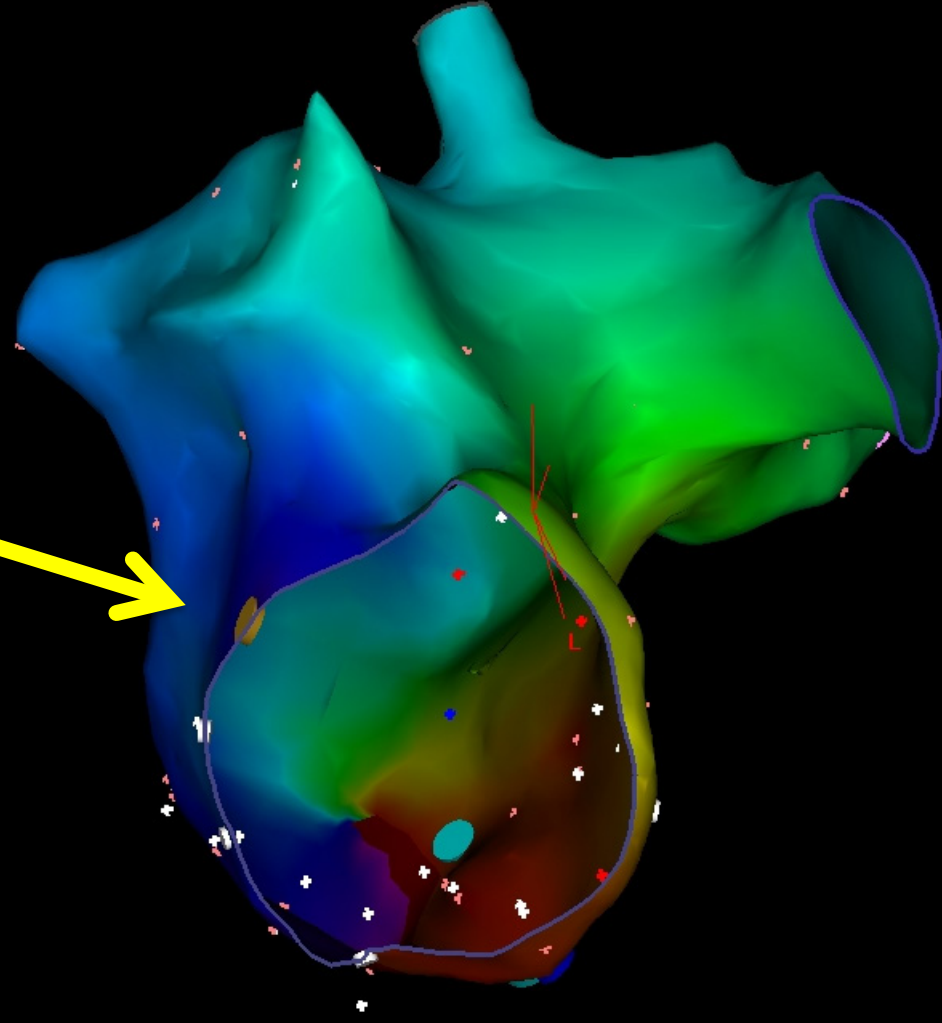
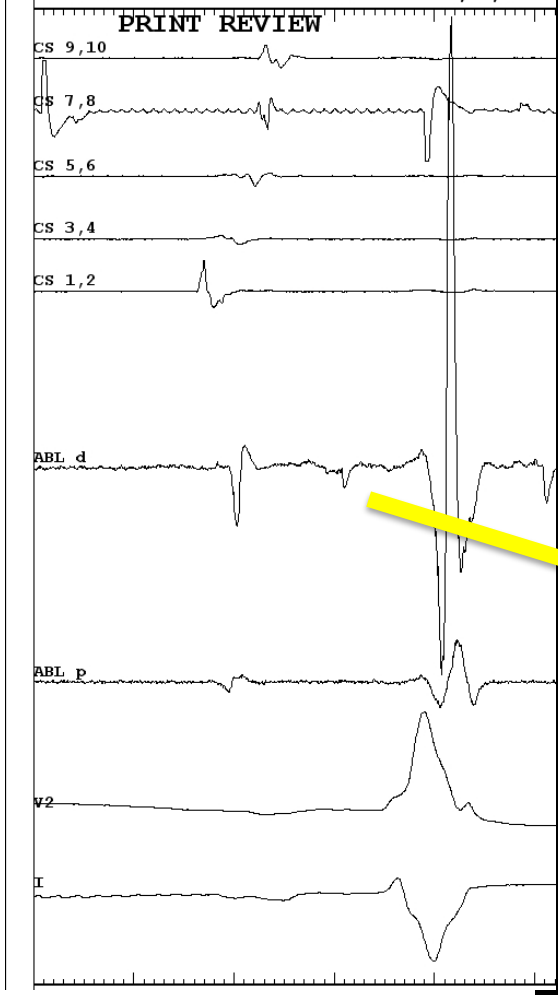
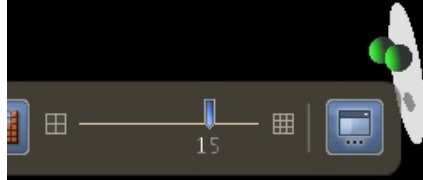




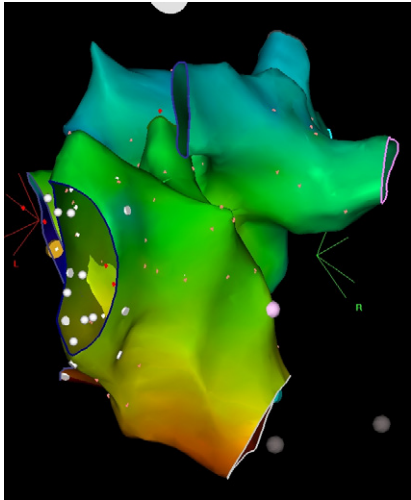
RIGHT ATRIUM

LAO

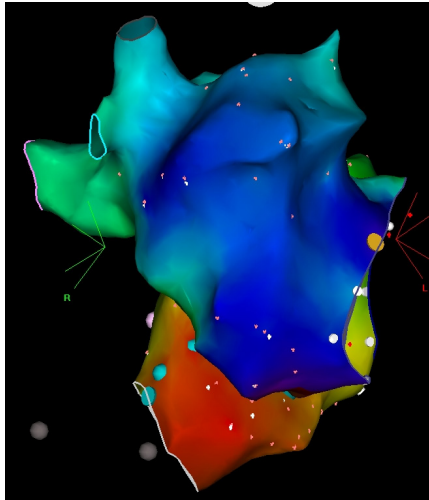
-91 ms LAT 108



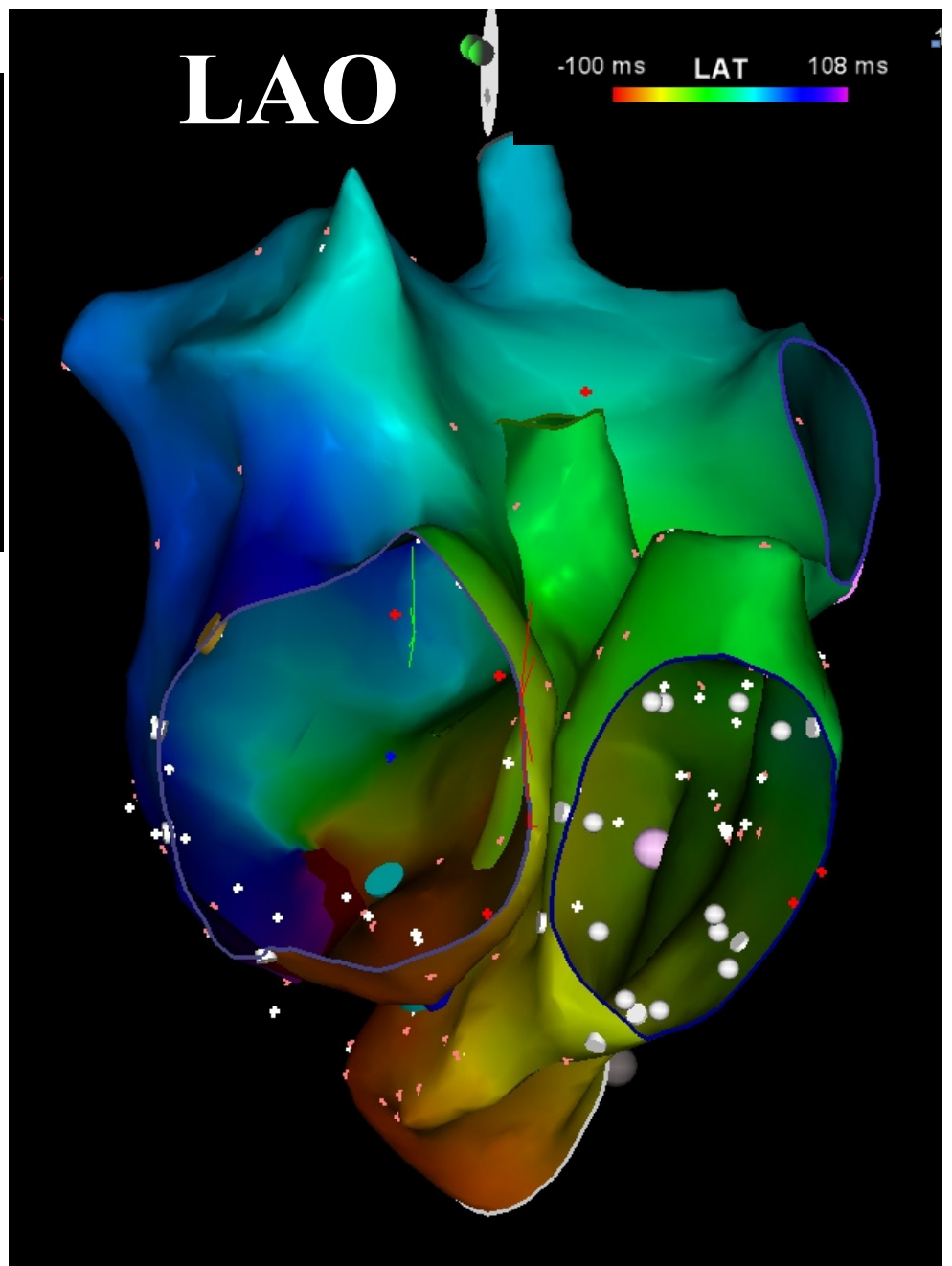
LL



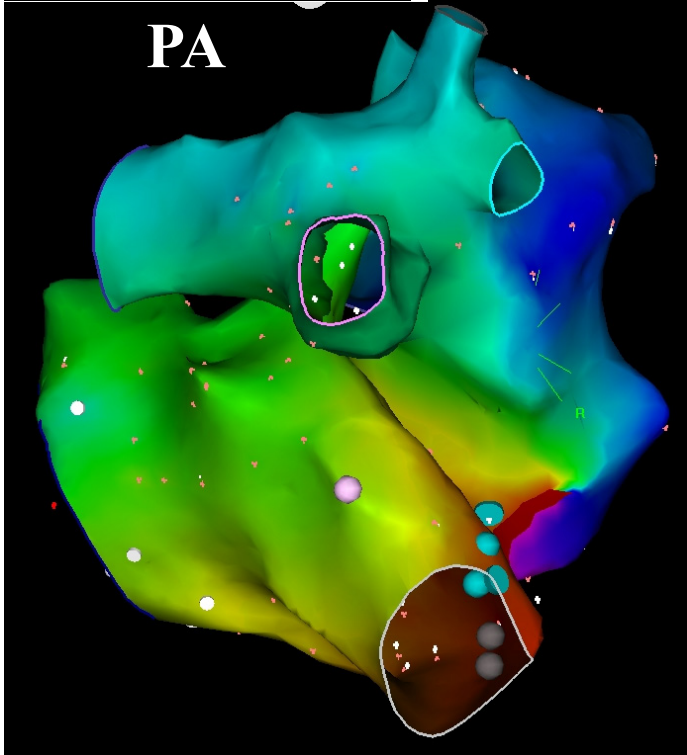
RAO



LAO



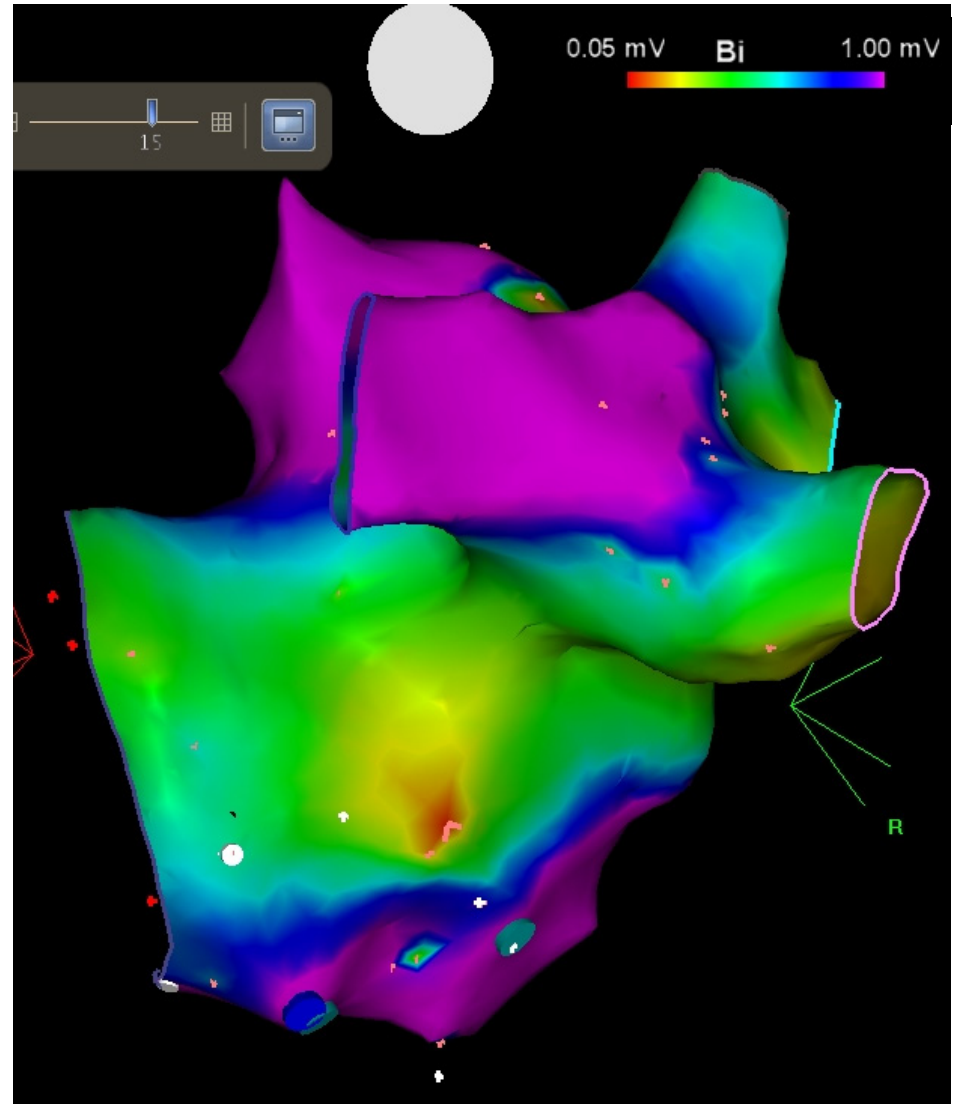
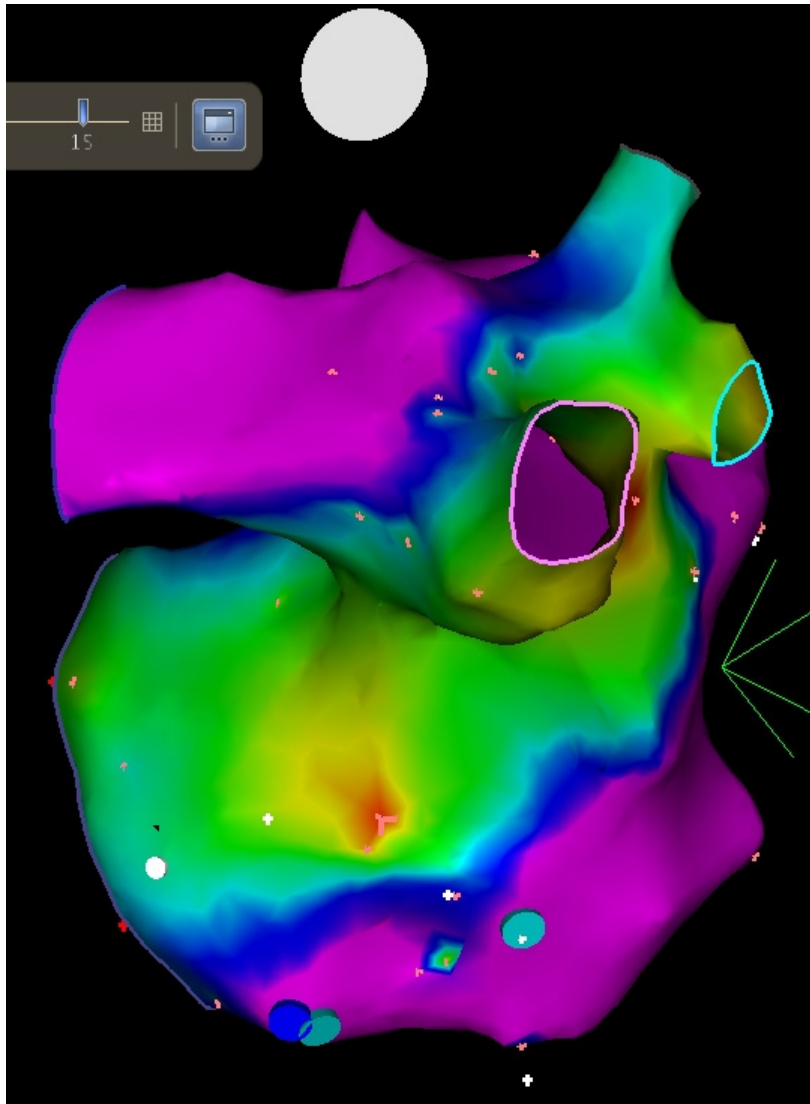
PA

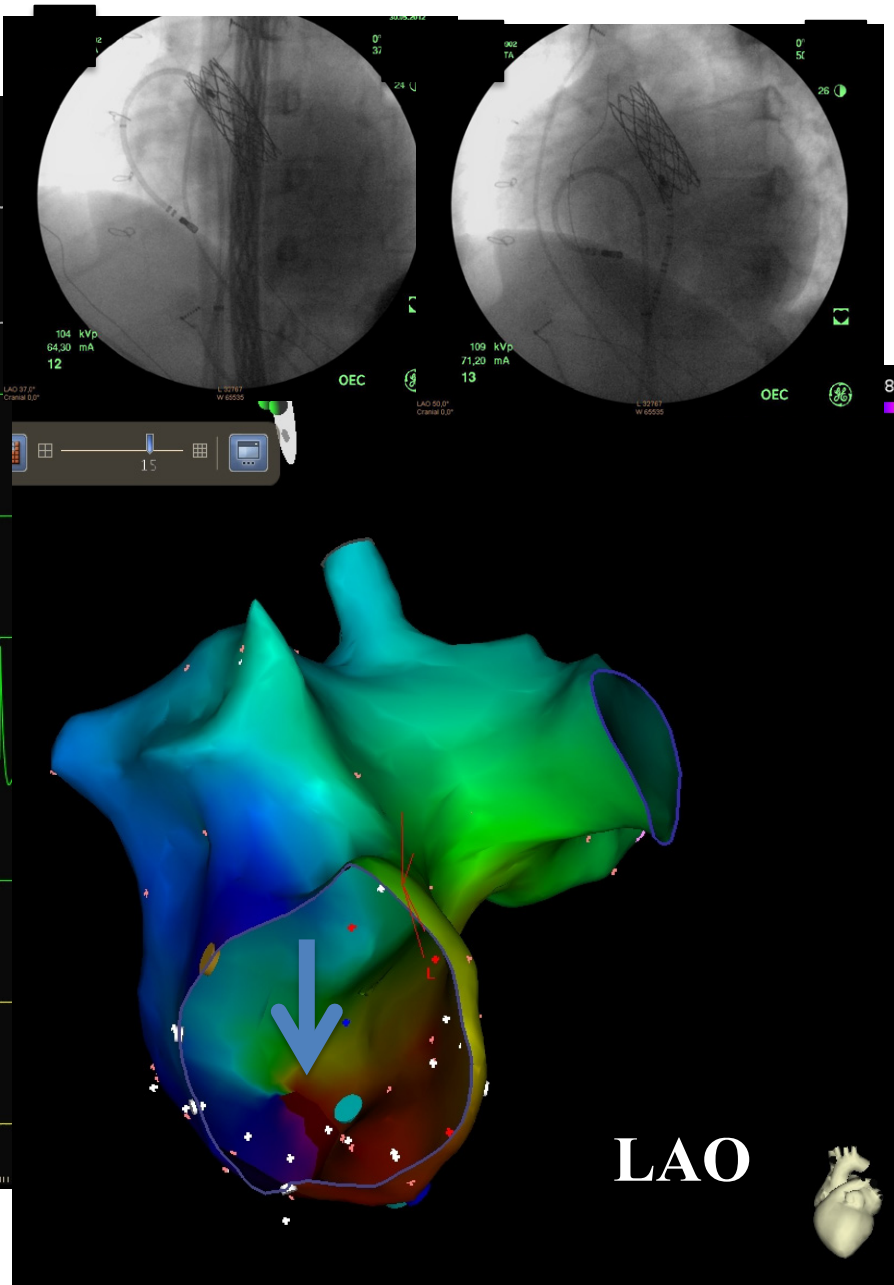


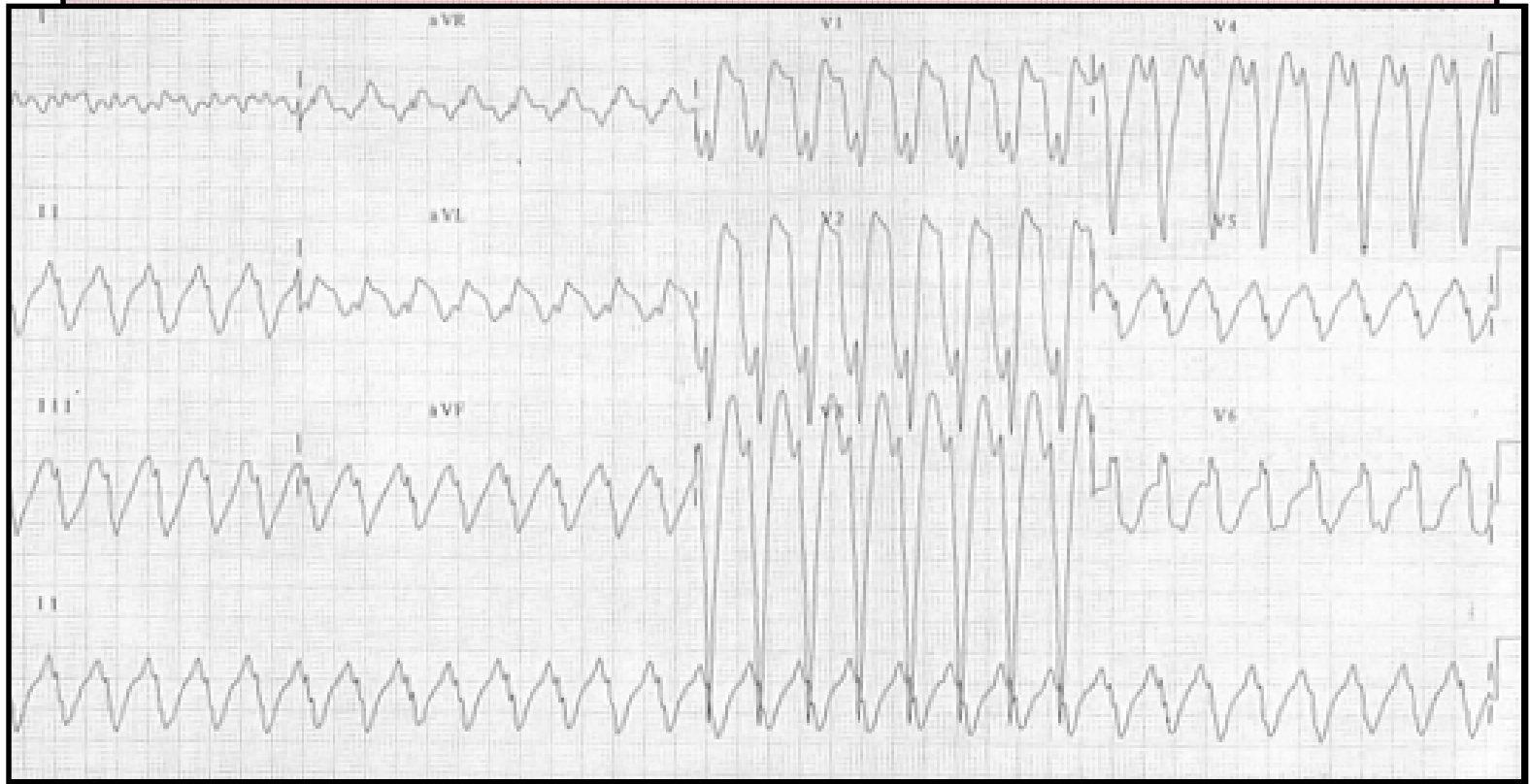
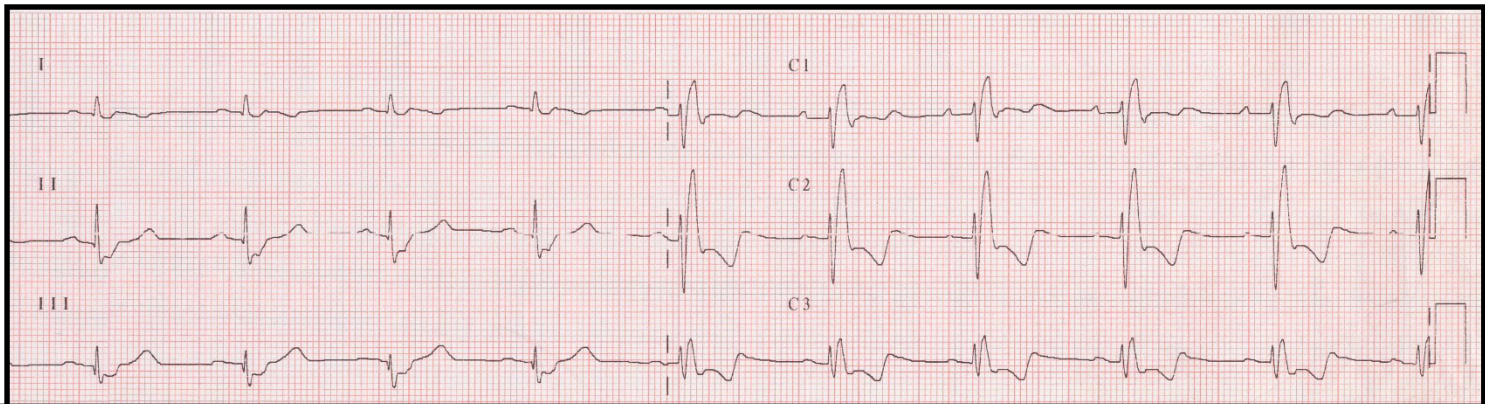
PA

VOLTAGE MAP

LL







ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

- SCD is of particular concern in GUCH patients.
- Five defects with greatest known risk:
 - repaired Tetralogy of Fallot,
 - TGA with atrial switch,
 - congenitally corrected TGA,
 - Aortic Stenosis,
 - Univentricular hearts.
- Unexplained syncope is a warning event requiring careful evaluation of arrhythmia.
- Although various risk factors have been defined, algorithms for risk assessment and indications for ICD implantation have so far not been well established.



ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)



General Indications for EP and ICD Implantation in GUCH

- ICD implantation is indicated in survivors of cardiac arrest after exclusion of reversible causes.
- Patients with spontaneous sustained VT should undergo invasive haemodynamic and EP evaluation. Recommended therapy includes catheter ablation or surgical resection to eliminate VT. If that is not successful, ICD implantation is recommended.
- Invasive haemodynamic and EP evaluation is reasonable in patients with unexplained syncope and impaired ventricular function. In the absence of a defined and reversible cause, ICD implantation is reasonable.
- EP testing may be considered for patients with ventricular couplets or non-sustained VT to determine the risk of sustained VT.

Class ^a	Level ^b
I	B
I	C
IIa	B
IIb	C

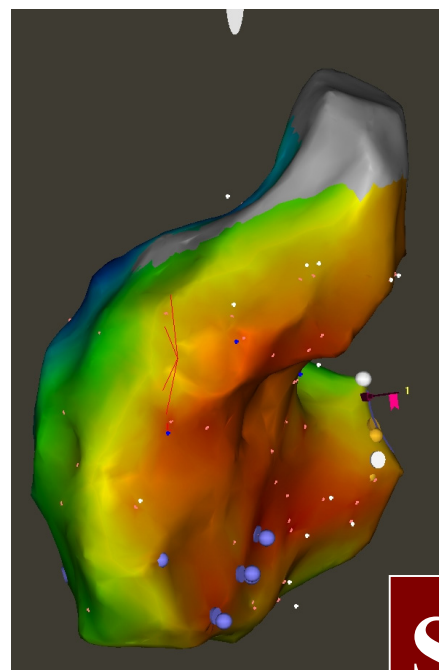


VENTRICULAR TACHYCARDIA

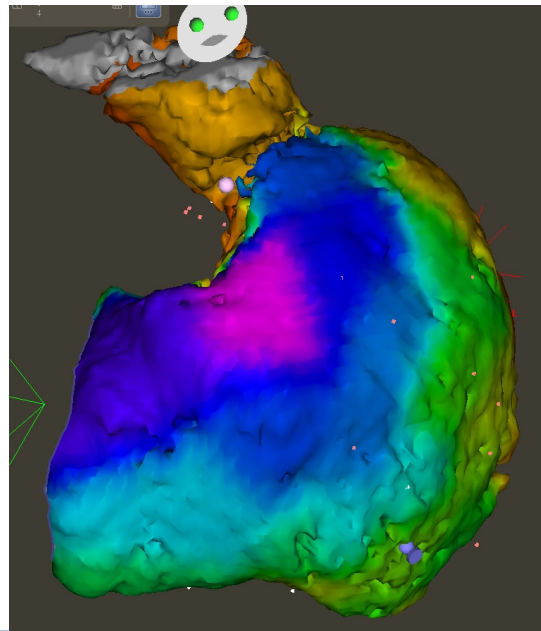
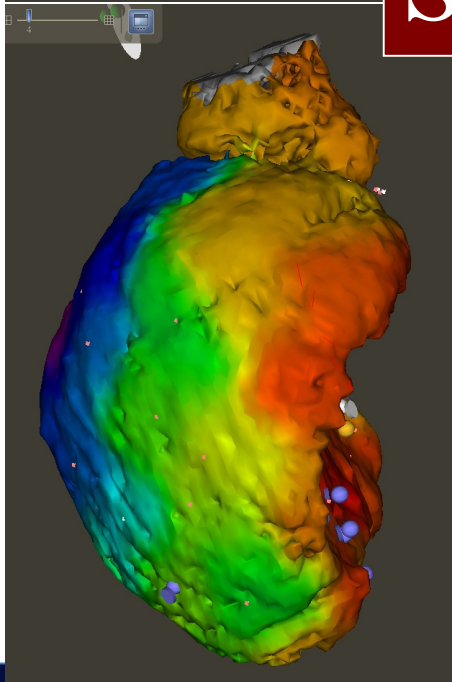
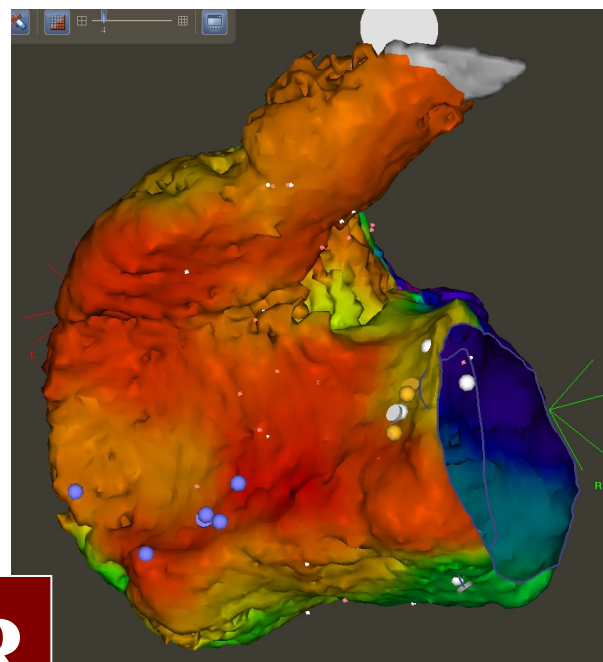
RF Ablation in repaired ToF

Author	Journal	Year	N° pts	Type of arrhythmias	Success rate	Recurrences rate	Complications rate
Horton	JCE	1997	2	VT	100%	0%	0%
Fukuhara	PACE	2000	1	VT	100%	0%	0%
Hebe	Pediatr Cardiol	2000	8	VT	58%	25%	NA
Kapel	Circ Arrhythm Electrophys	2014	28	VT	100%	0%	0%

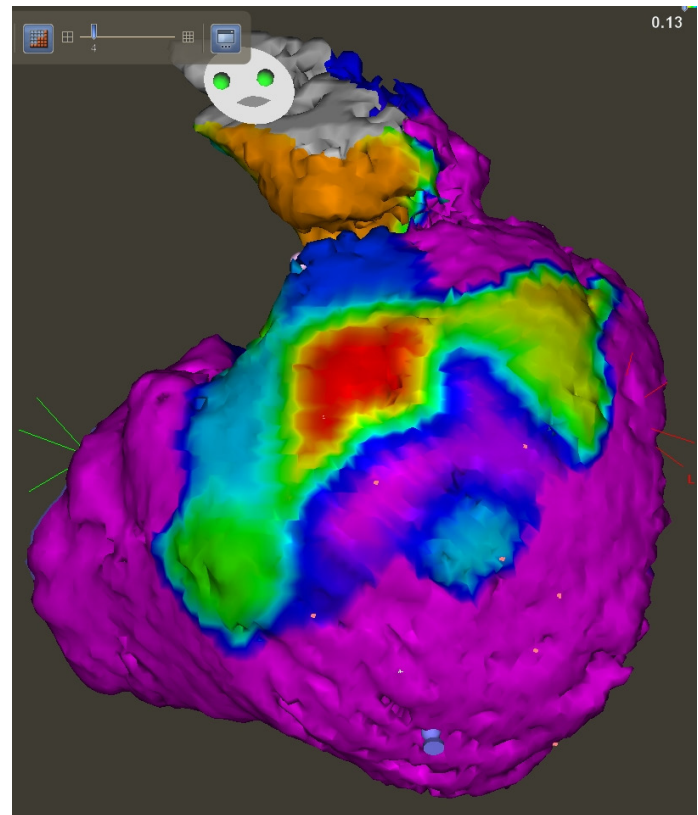




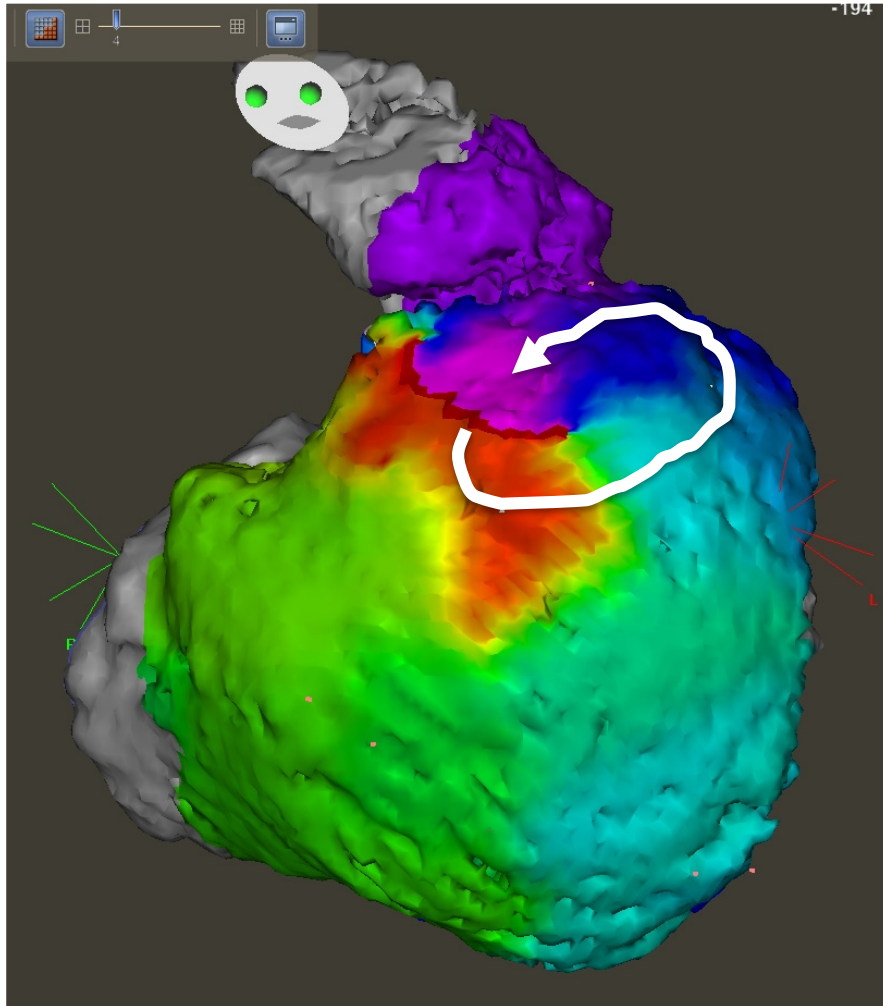
SR



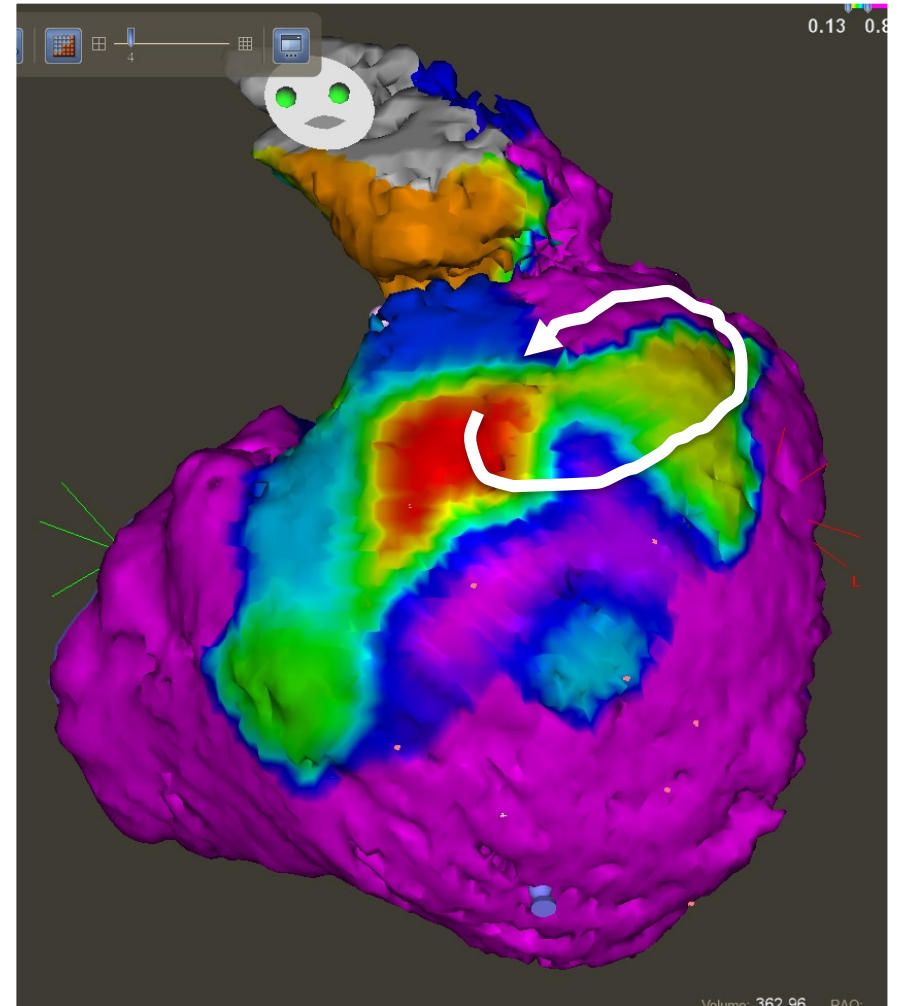
Voltage map ventriculotomy

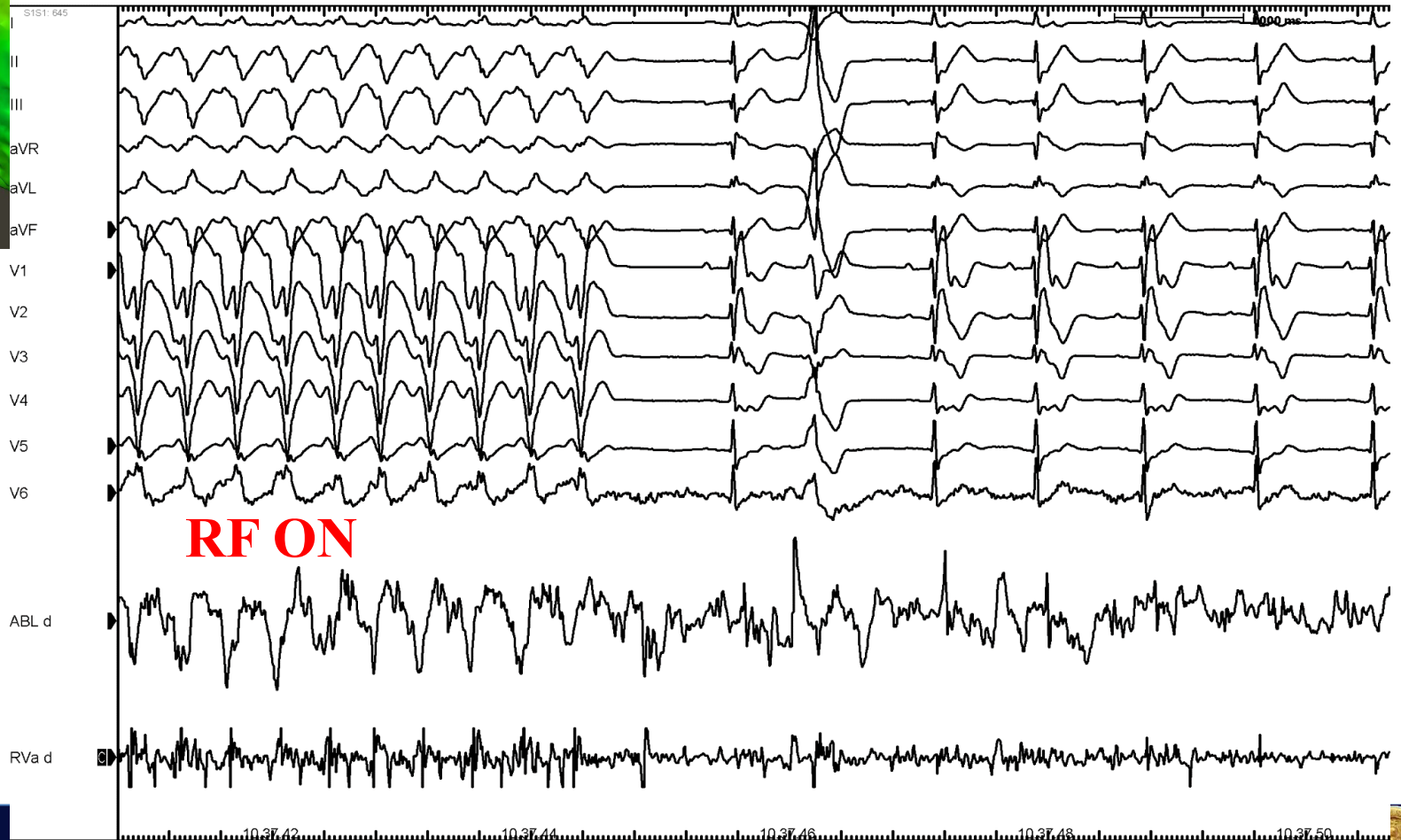
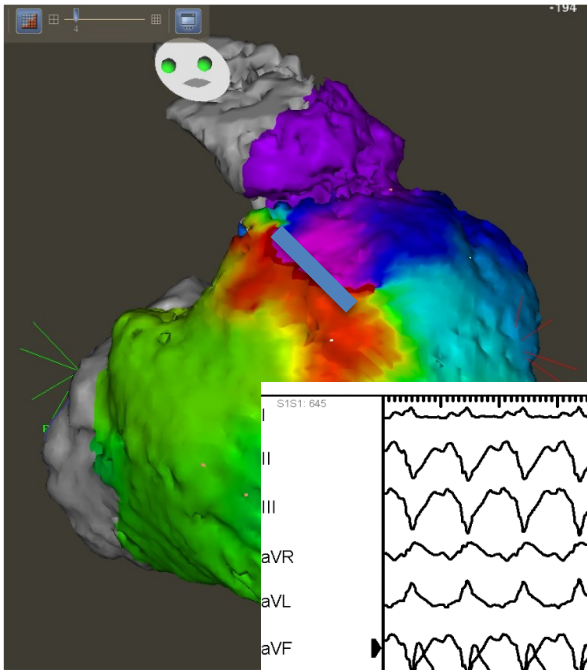


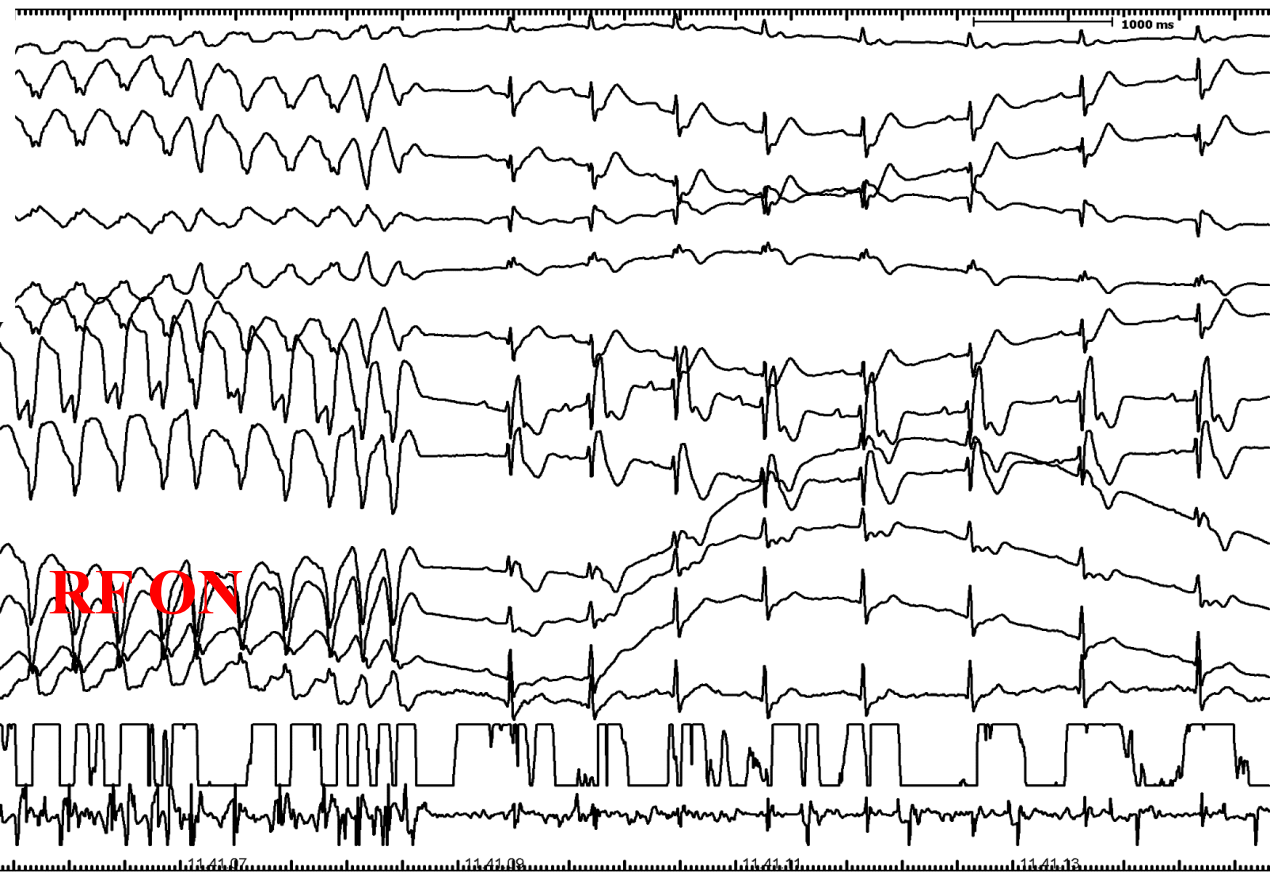
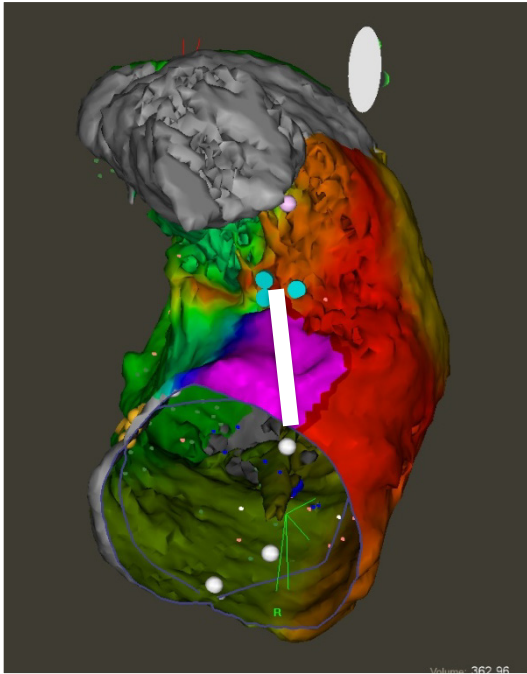
VT ACTIVATION MAP

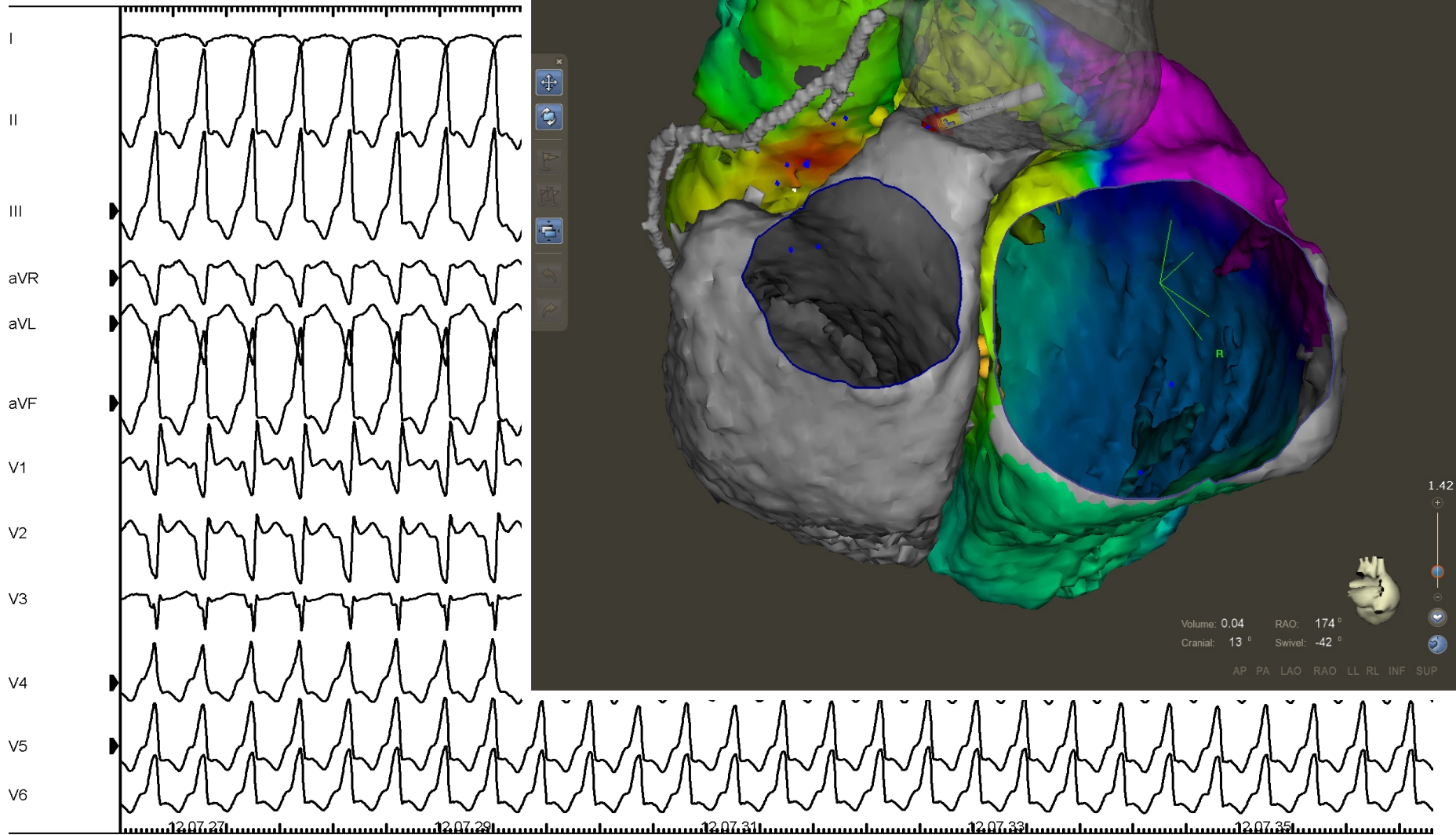


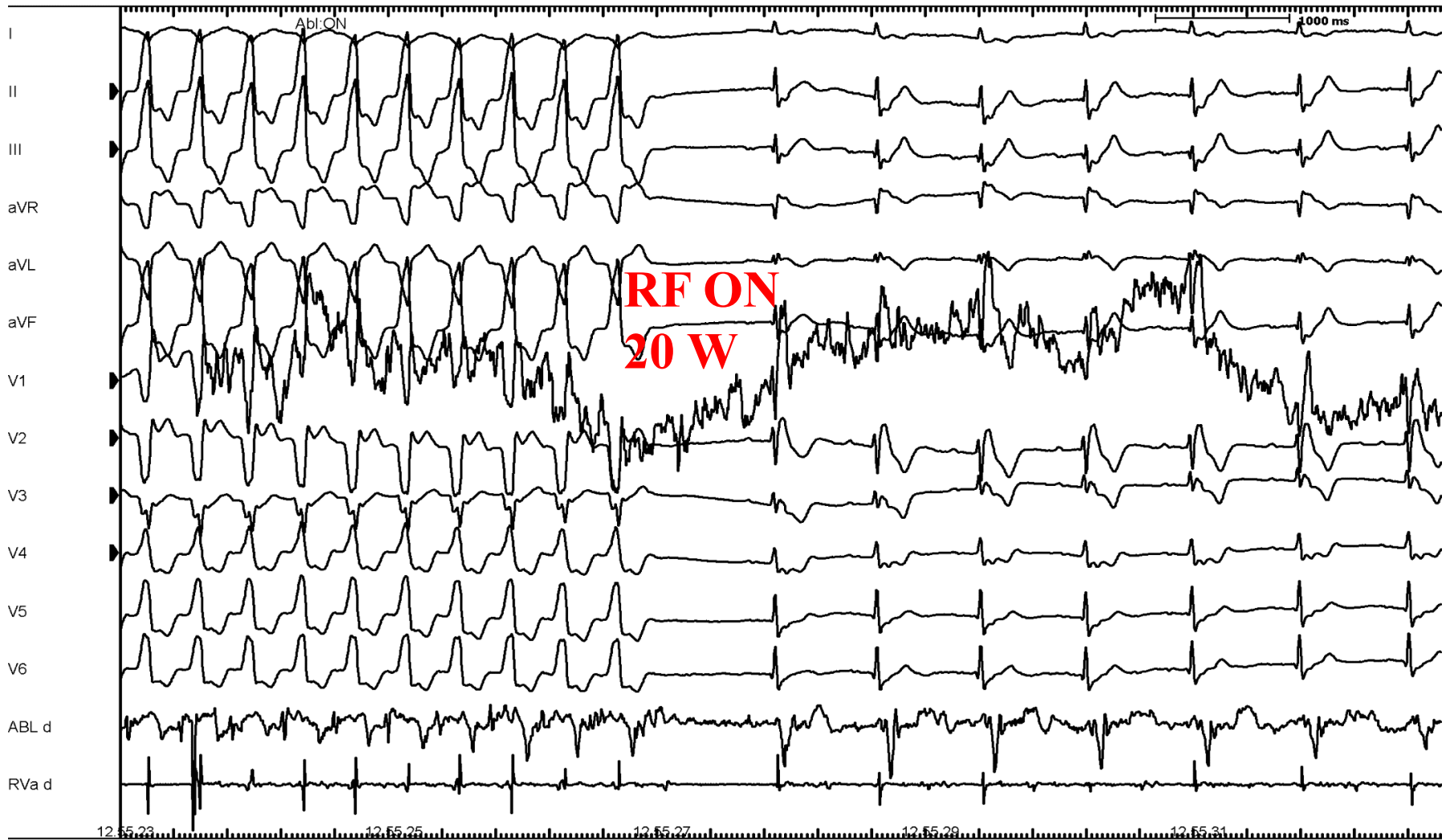
SR BIPOLAR MAP











No more inducible



ARRHYTHMIAS IN CONGENITAL HD

...are part of the natural history.

...often are related to surgical repair.

...since they may be a negative prognostic indicator/impair haemodynamic, they should be treated.



TREATMENT OF ARRHYTHMIAS IN CONGENITAL HD

...is a challenge.

Catheter ablation may be a curative option both in atrial and ventricular arrhythmias.

ICD implantation should be evaluated in particular situations



Requirements for electrophysiological treatment of arrhythmias in congenital HD

Electrophysiological experience in complex anatomical evaluation and complex arrhythmia ablation.

Anesthesiologist support.

Electroanatomical mapping system.

Radiology department availability (MRI/CT).

Transesophageal echo lab.





GUCH NETWORK

STAR ALLIANCE

