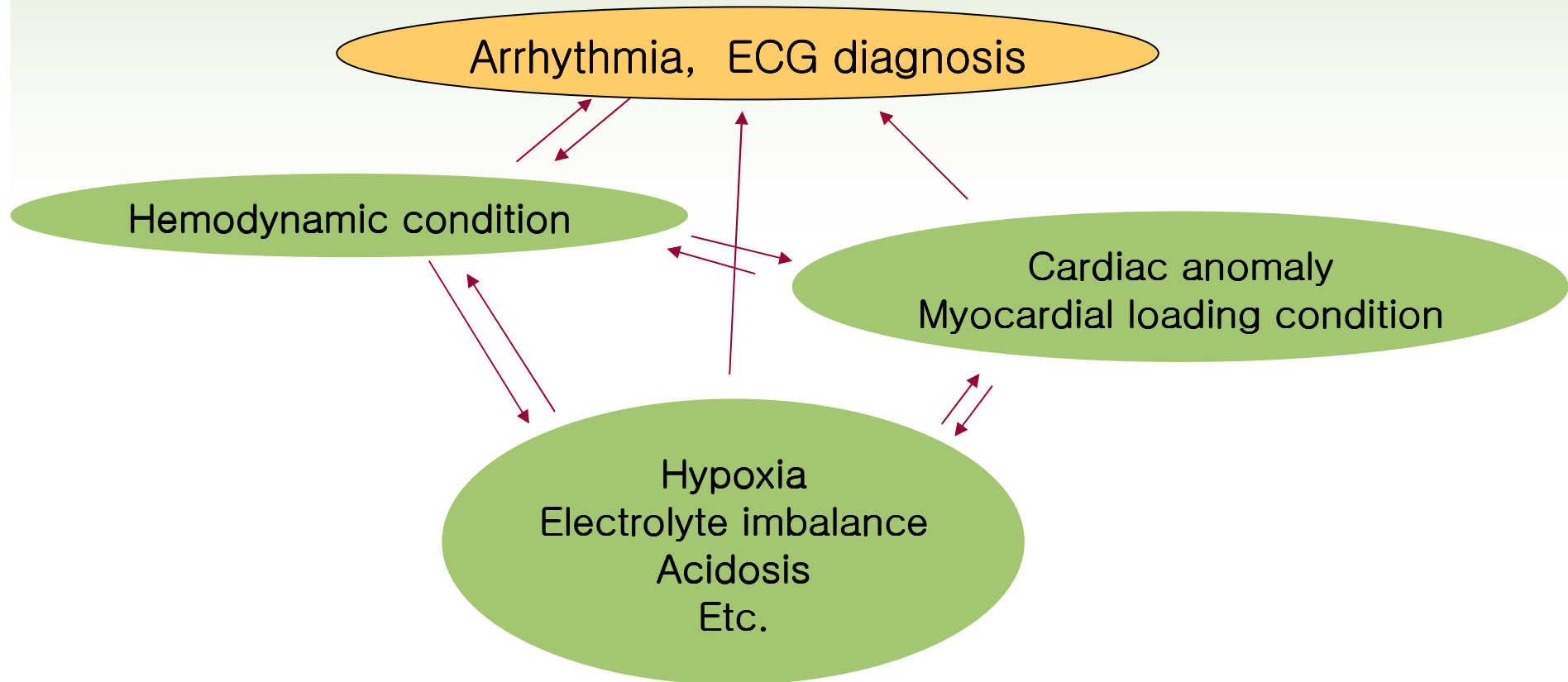


Management of Atrial arrhythmia after surgery of congenital heart disease

Seoul National University Children's Hospital
Bae Eun-Jung

Treatment of tachycardia

- Attention to big cardiovascular picture



Case 1. 6y/F

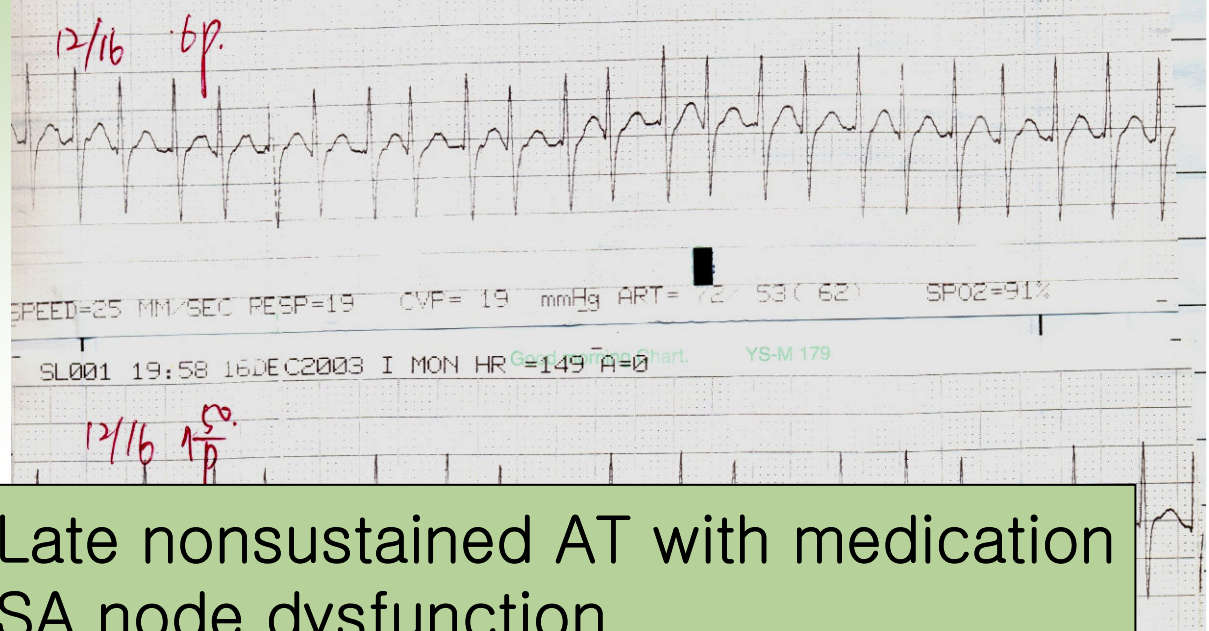
- Initial presentation with cyanosis (11m of age)
- Right isomerism.
{A.D.D} Left Ao arch.
Common inlet RV , rudimentary LV
TAPVR to innominate vein with mild obstruction.
DORV with PS ,Mild AVVR
- TAPVR repair and BCPS (15m, 6.8 kg)
- progressed AVVR to severe degree
- Fenestrated lateral tunnel Fontan op and common AVV repair (38m of age , Bwt 10.4kg)

Postoperative Condition

Intermittent palpitation, Exercise intolerance FC II-III

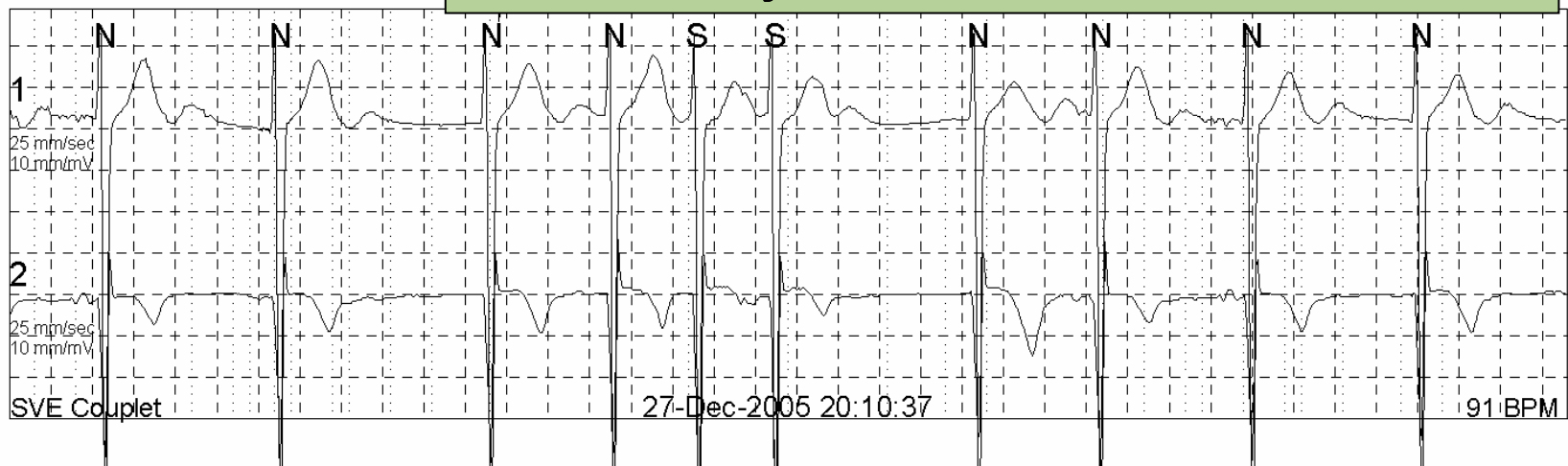
Medication: Sotalol, warfarin, digoxin, diuretics

Early post-Fontan
2003



Holter, 2005

Late nonsustained AT with medication
SA node dysfunction



Clinical :

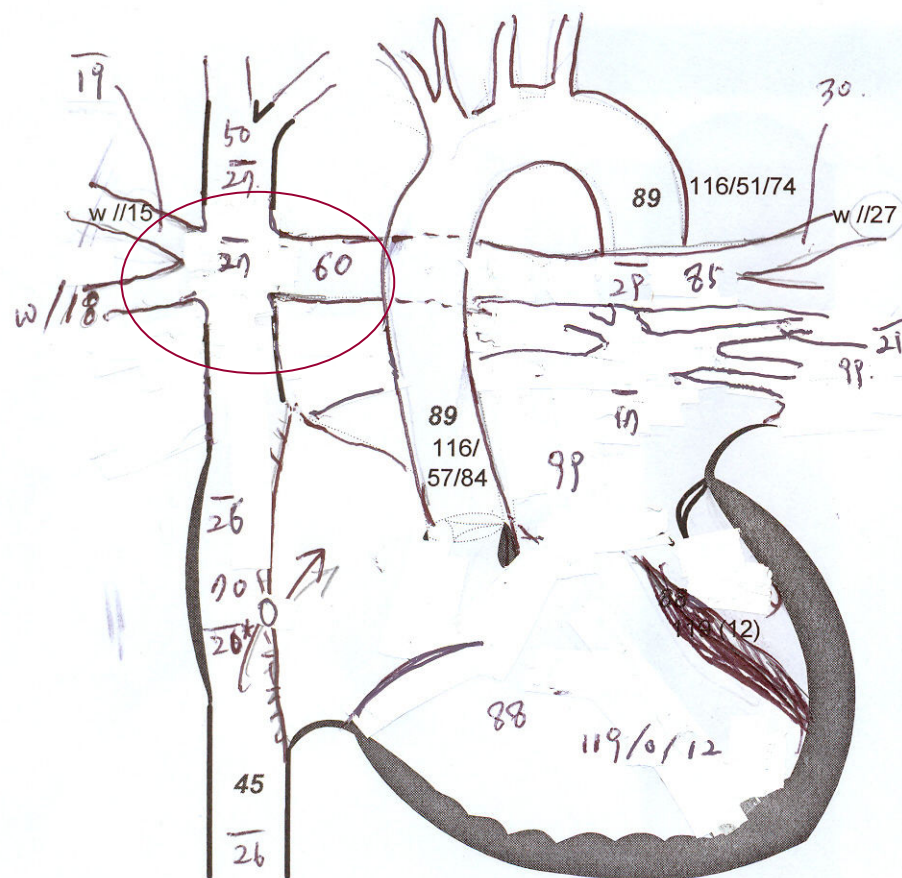
Cath 2006, 3 y after Fontan op

Comment :

wt: 12.0 kg Ht: 95 cm BSA: 0.57 m2

Complication:

Hgb: 12.4 HR: VO2:



* with oxygen

Fontan pathway mean : 26mmHg
 with oxygen 20 mmHg
 distal RPA : 22, RLPA : 18 mmHg
 LUPA 30 , LUPAW : 27 mmHg
 RUPA : 19, RUPAW : 15 mmHg
 LUPV : 21 RLPV : 17 mmHg

Recommendation/Plan

Diagnosis/Intervention

{A.D.D} left arch.
 S/P Fontan op. *1/2 TAPVR (supracardiac) type*
 CAVSD, hypoplastic LV at left posterior.

DORV.

No Fontan pathway stenosis.

AVVR : mild,
 elevated SVC, IVC, Fontan pathway, and both PA pressure

no significant PV *stenosis*
 underdeveloped LPA peripheral bed
 numerous arterial collaterals to LPA bed

Fontan pathway, SVC flow to RPA

RPA: overflow

elevated LVEDP, LAP

PG through fenestration : 8mmHg

EPS>

baseline: mainly sinus node
 dysfunction with accelerated junctional
 rhythm

no inducible junctional tachycardia

nonsustained atrial flutter fibrillation

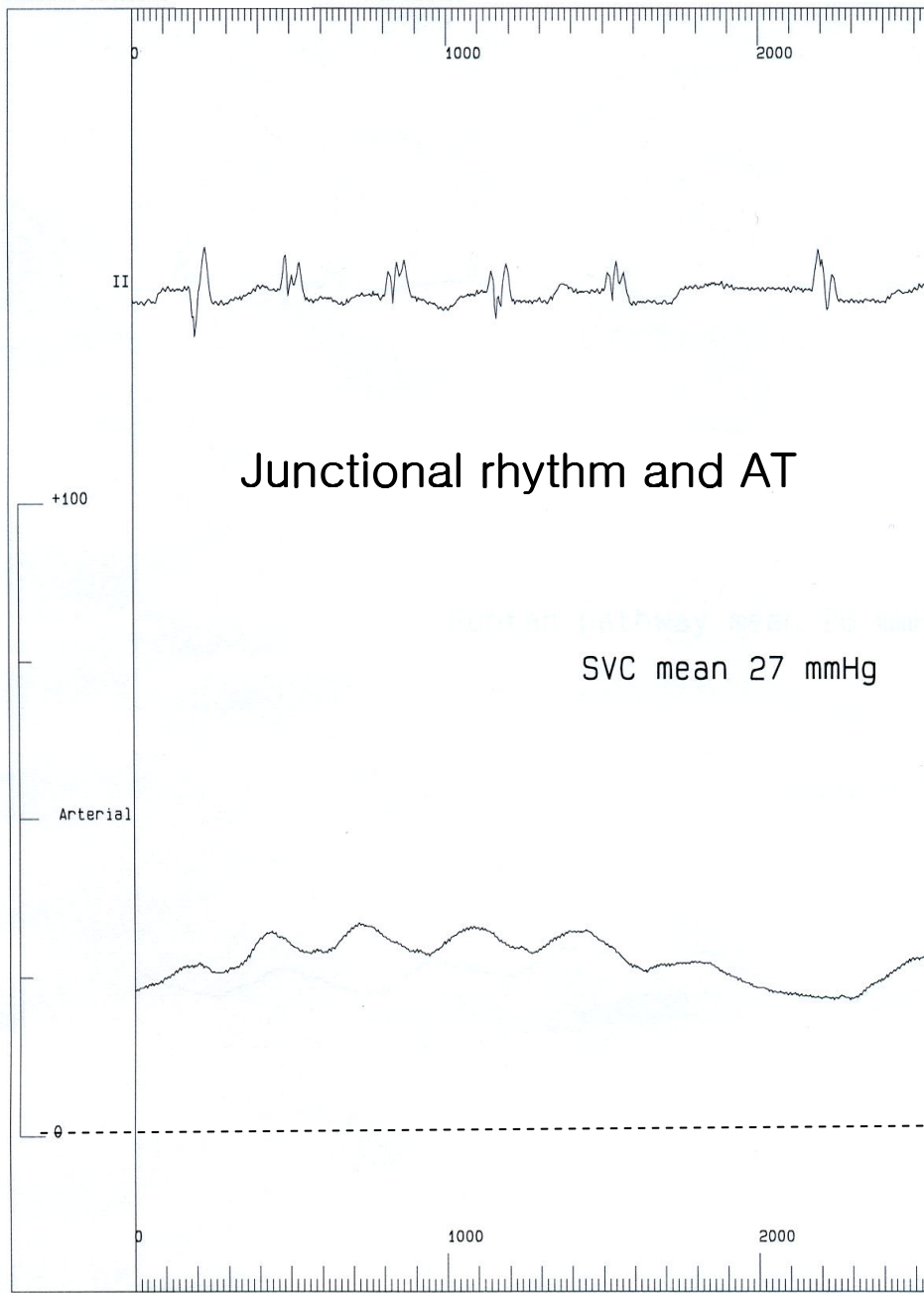
was induced

no VA conduction during V pacing

* mean CVP with sinus rhythm, oxygen:
 14 mmHg

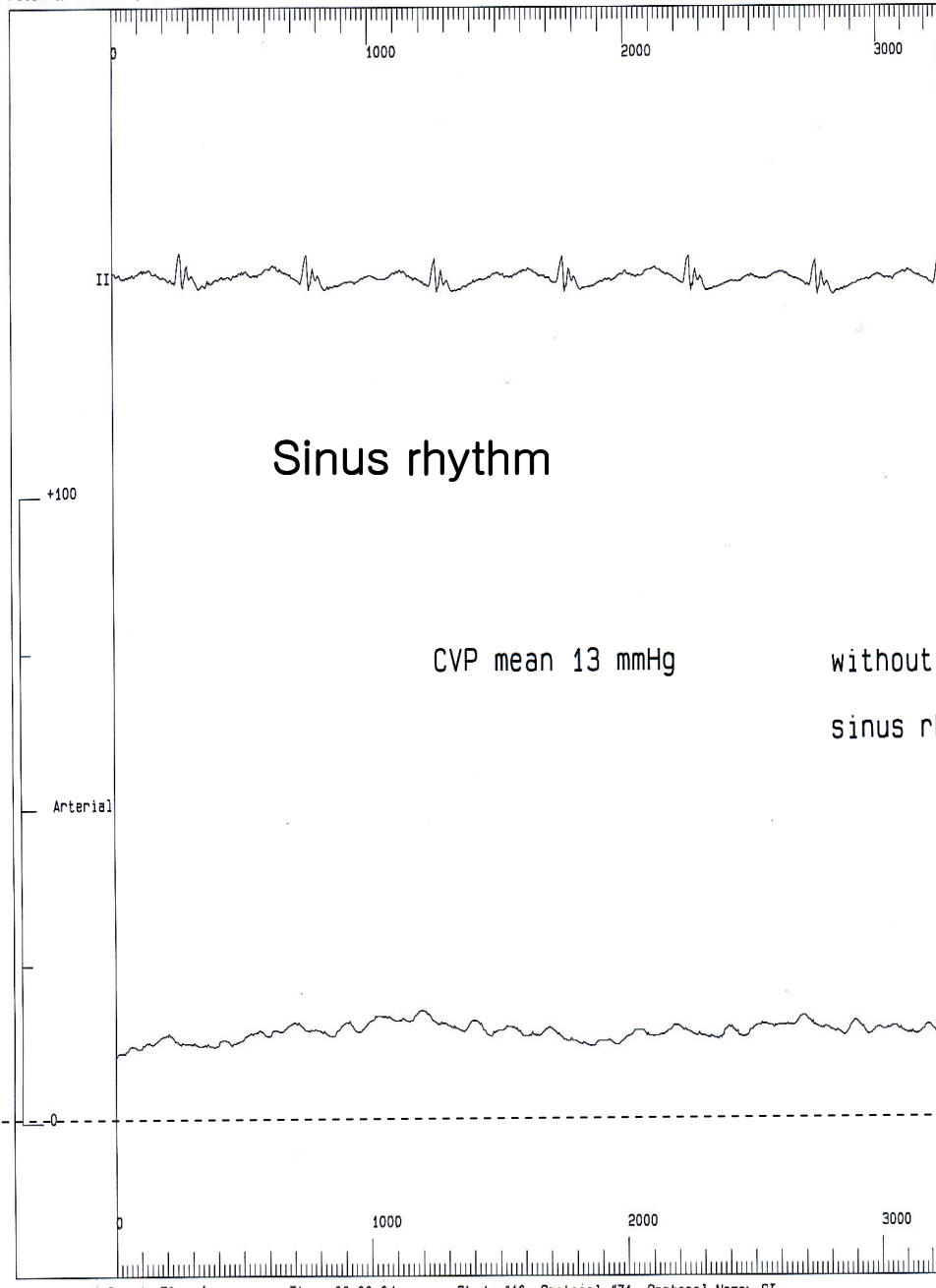
Patient: 73495574,

ID: 200602

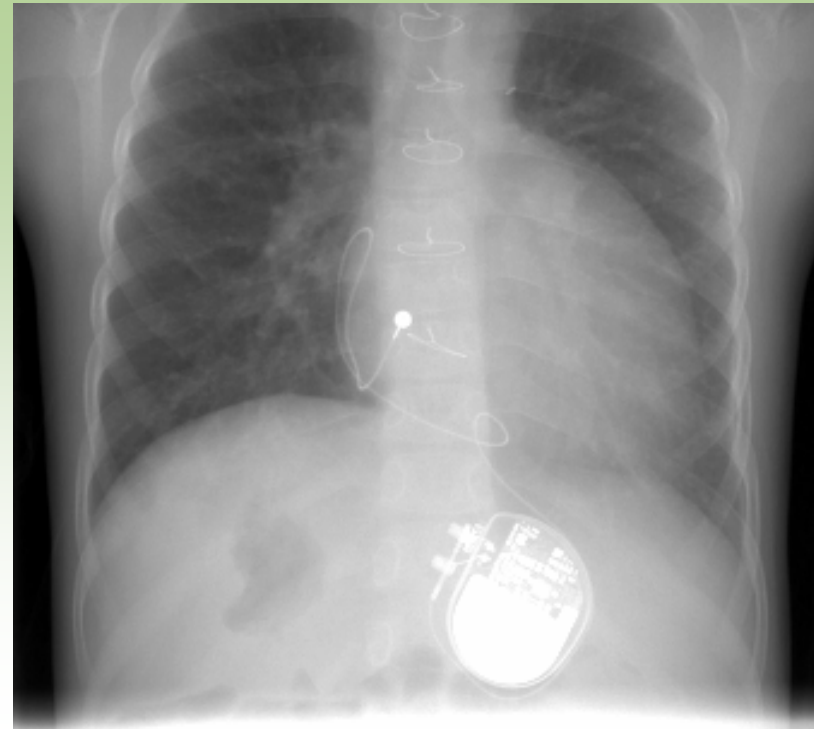


Patient: 73495574,

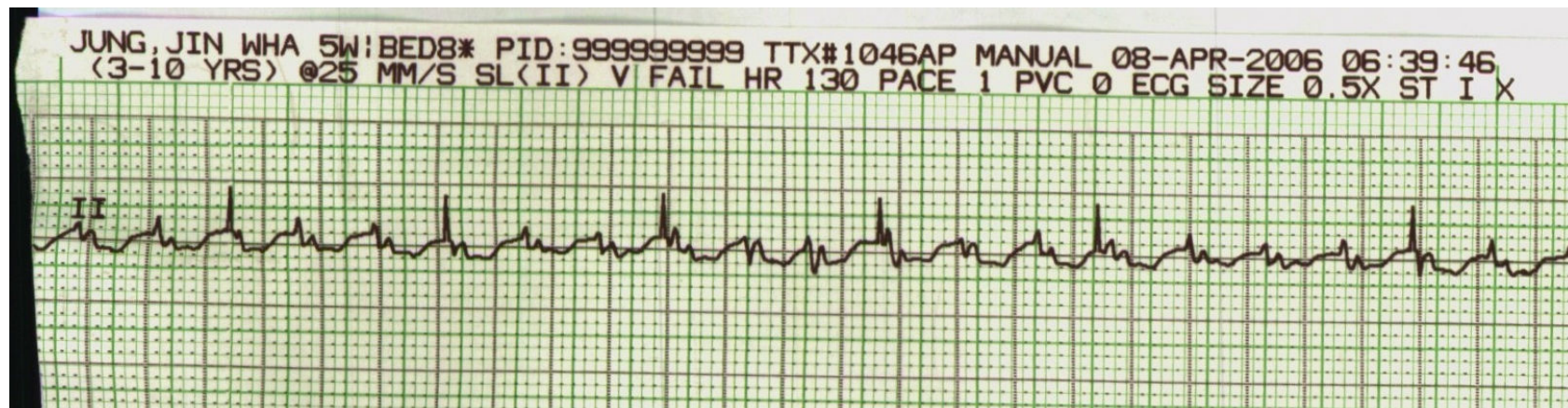
ID: 200602



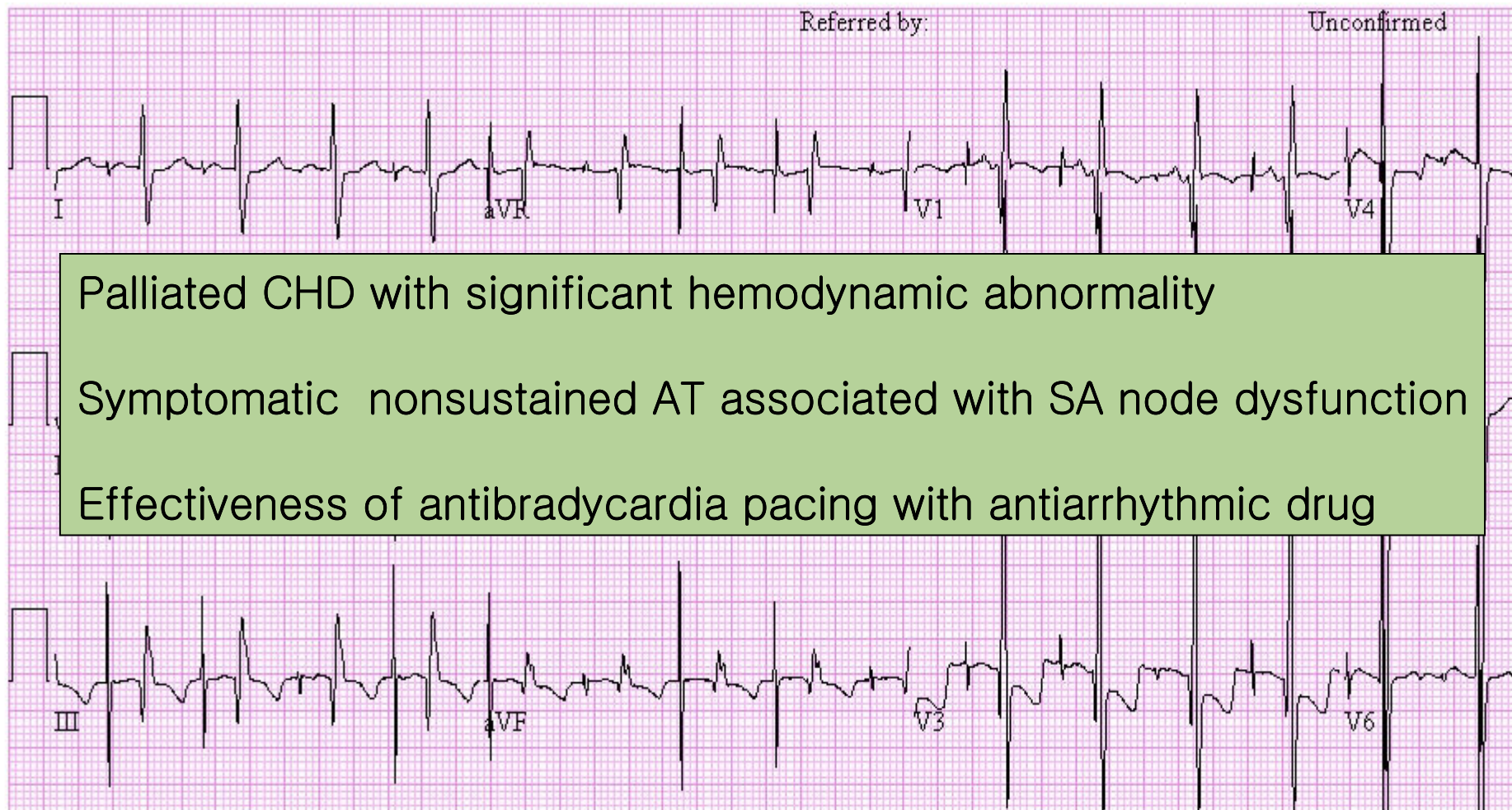
AAIR pacemaker



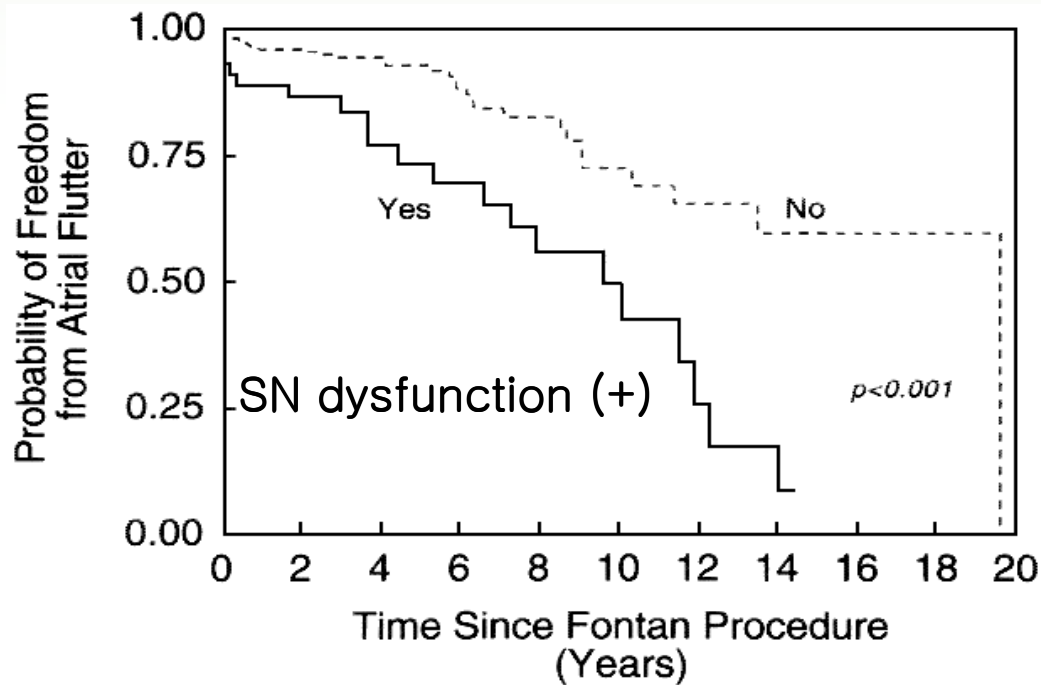
- Postop. tachycardia → sotalolol



AAIR pacemaker; Antibradycardia pacing
& Solatol 3.5mg/kg/day #3
→ Nearly absent palpitation for 1 y
improved exercise tolerance

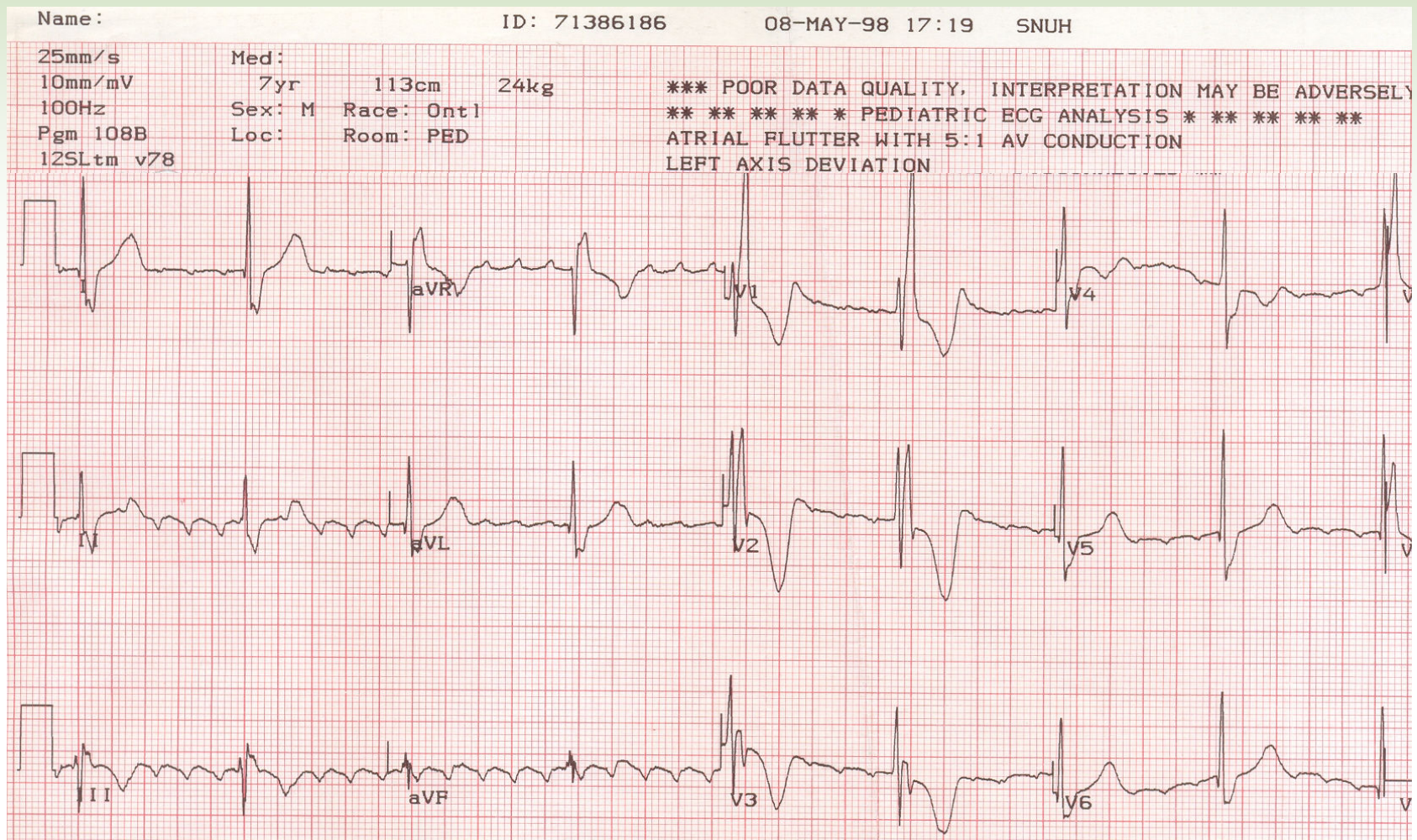


Sinus node dysfunction increases the incidence of late AT



(Fishberger SB, JTCS 1997)

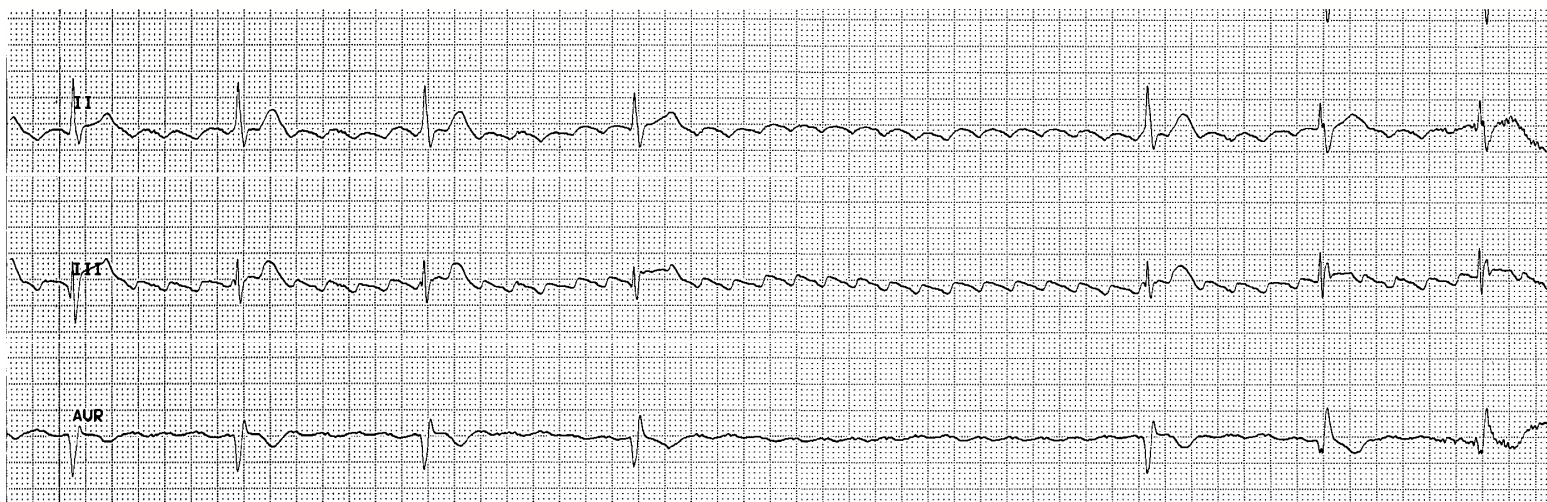
Case 3. S/P VSD closure ('93), Palpitation ('95); atrial flutter
→ Digoxin for 3 y , recurred AFL ('98)
→ Add sotalol (1.5-2mg/kg/d) for 3 y
2002.9& 02.10; Visited ER d/t atrial flutter → DC cardioversion



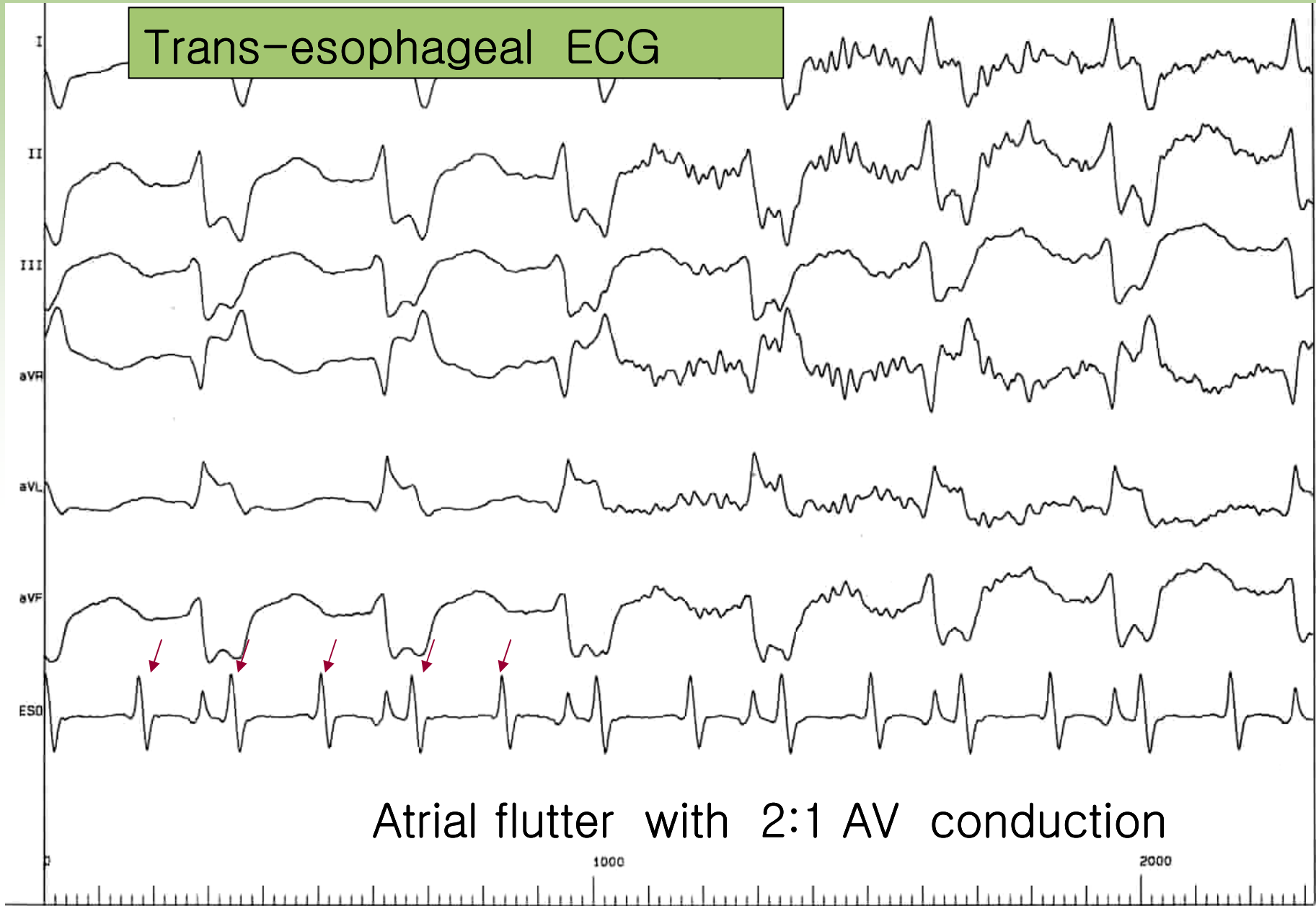


After adenosine injection

Atrial flutter

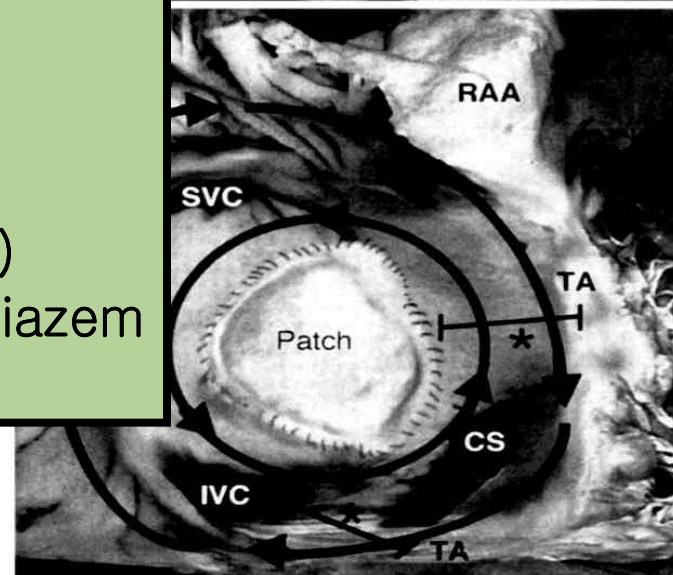
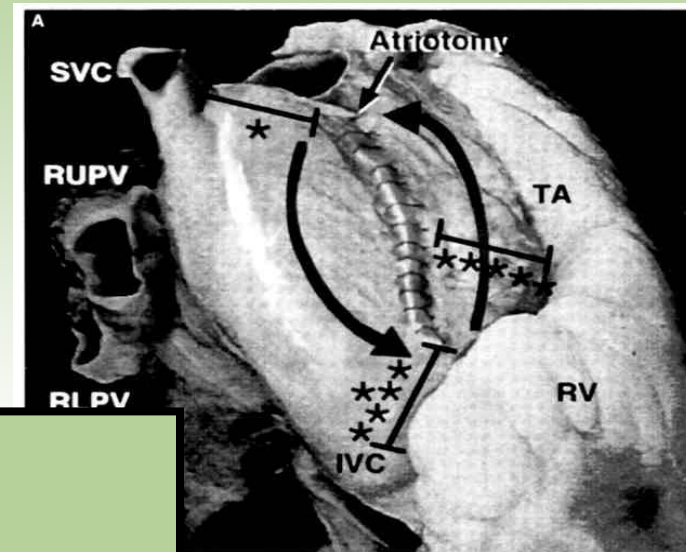
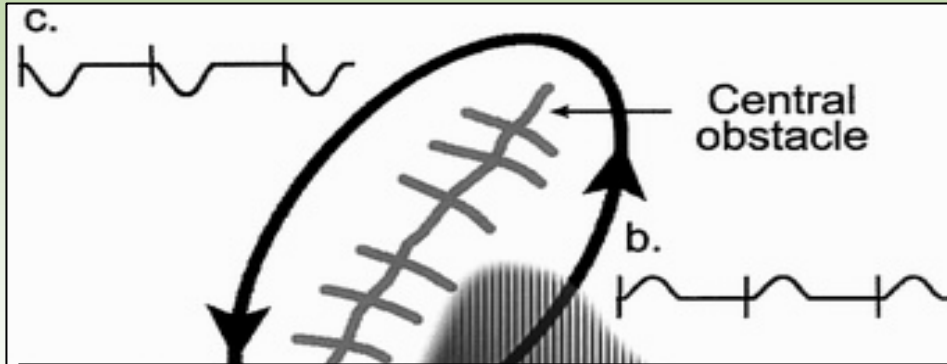


Trans-esophageal ECG



Atrial flutter with 2:1 AV conduction

Circuit of atrial flutter



Acute Mx of AFL

DC cardioversion; 0.5–1J/kg

Anti-tachycardia pacing

- Esophageal lead
- Pacemaker telemetry

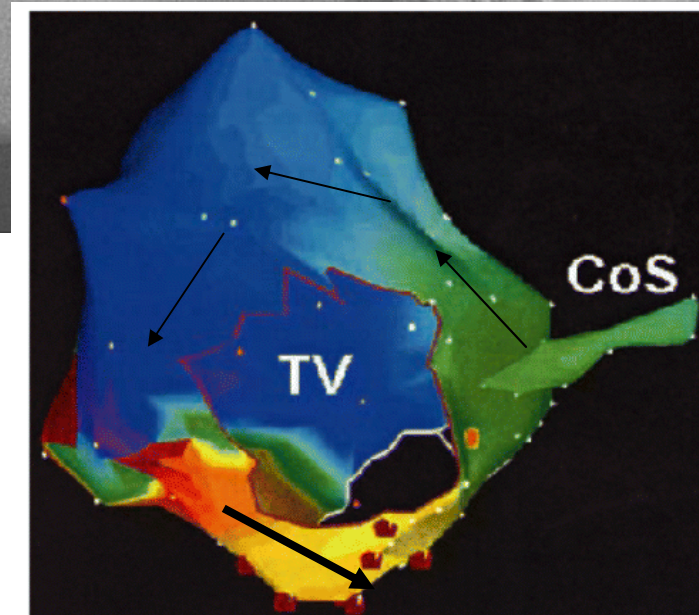
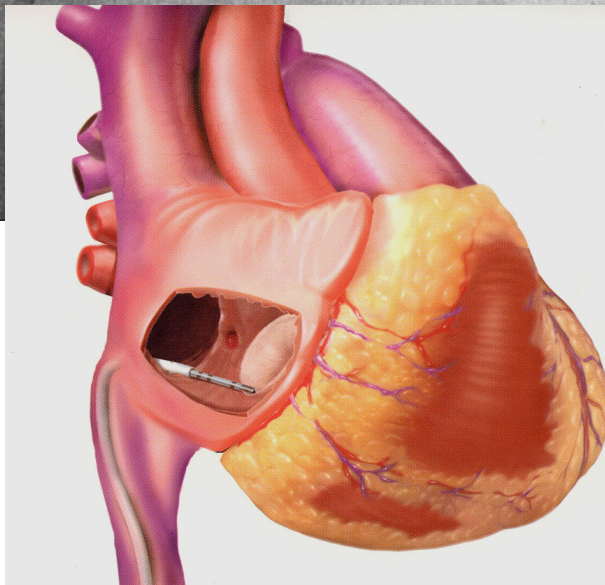
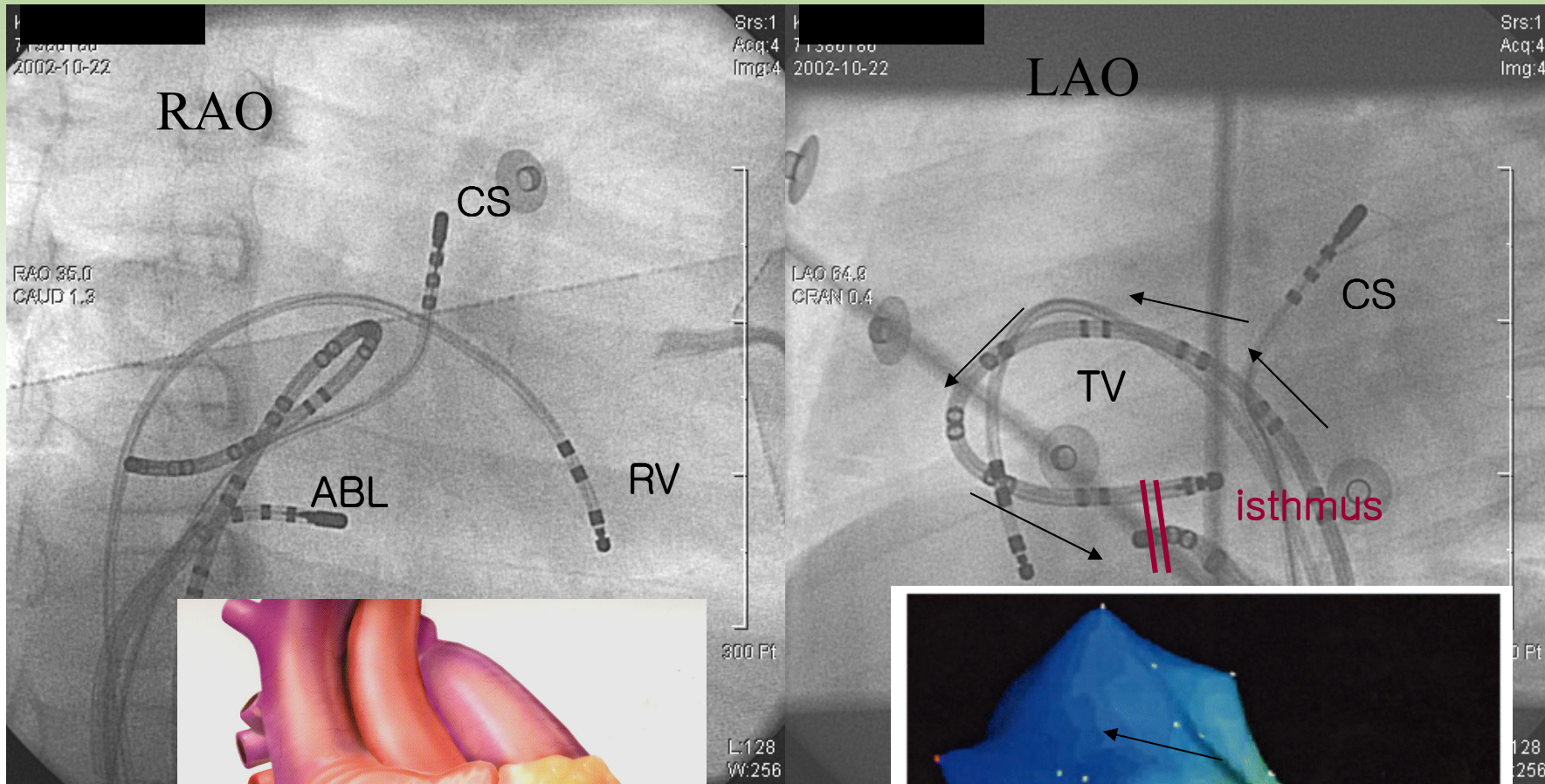
Single dose of antiarrhythmic drug (AAD)

Rate control ; digoxin, beta blocker, diltiazem

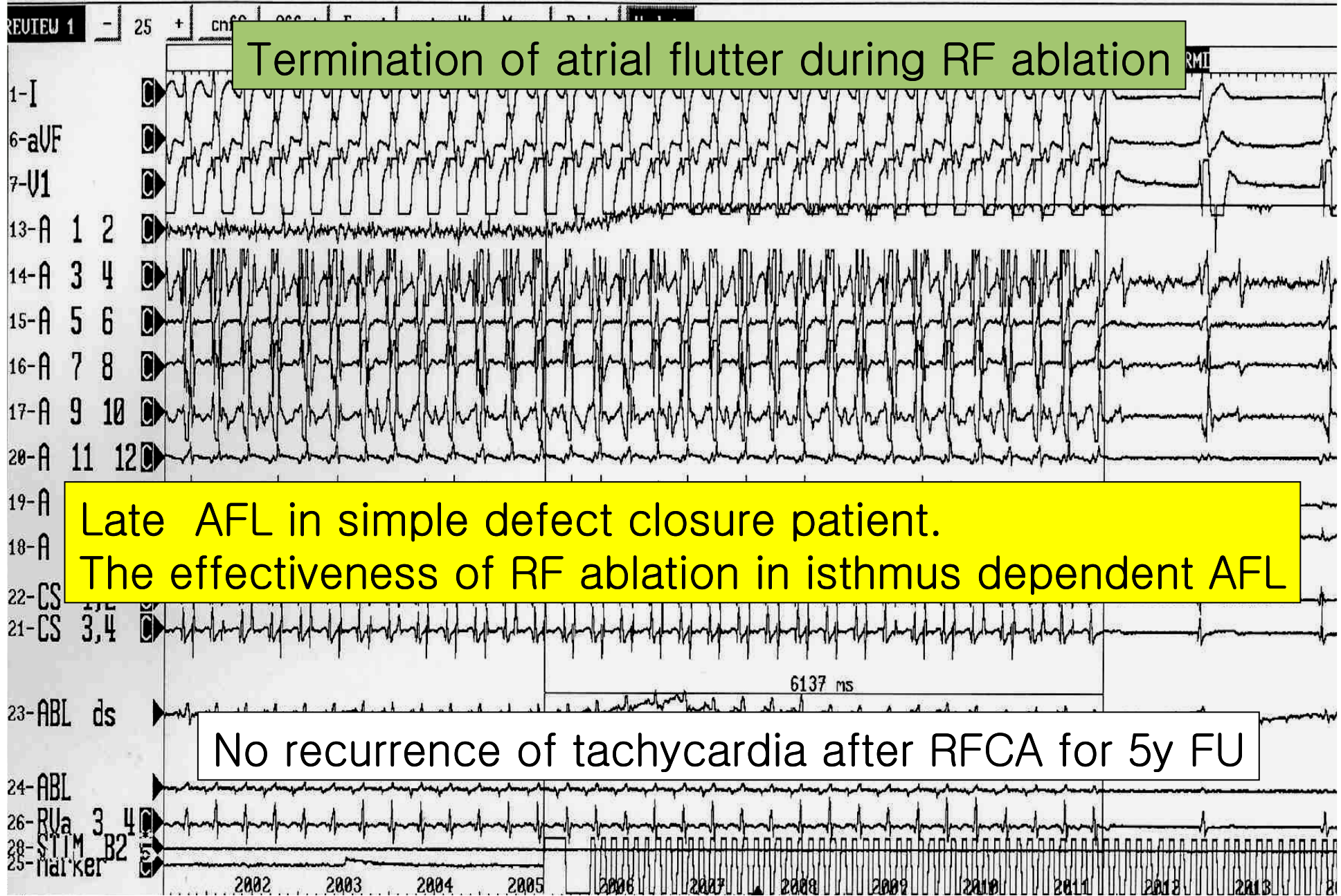
(Kalman JM et al.circulation.1996)

2002, Radiofrequency Ablation

Periannular atrial flutter



Termination of atrial flutter during RF ablation



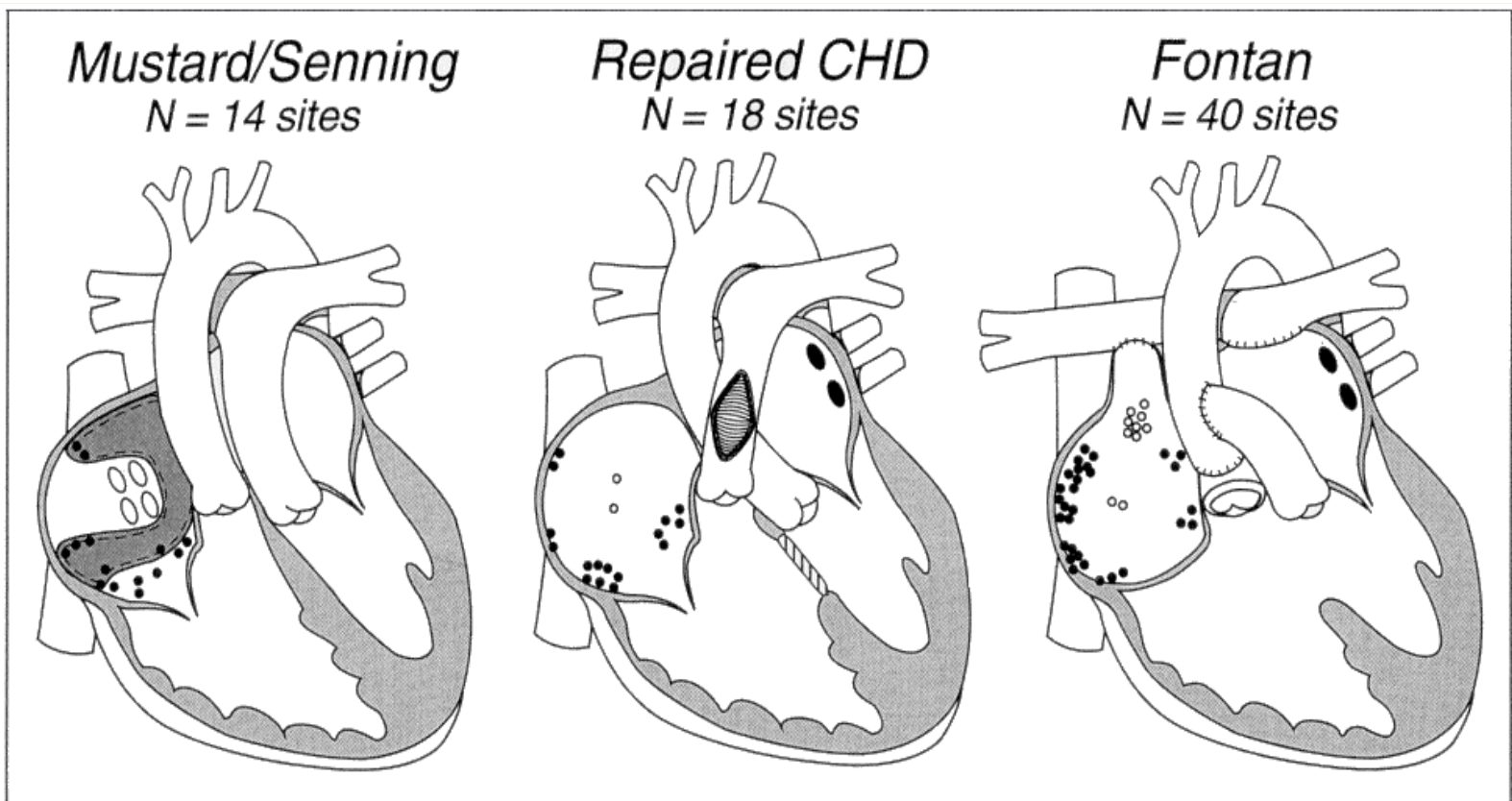
Late AFL in simple defect closure patient.
The effectiveness of RF ablation in isthmus dependent AFL

No recurrence of tachycardia after RFCA for 5y FU

Importance of atrial flutter isthmus in postoperative IART

(Van Hare ; circulation 2000)

- ◉ 19 postoperative IART cases
21 IART circuits
- ◉ Disease
 - Fontan 1(0/1), TOF 4(3/4), TGA 4(3/4), septal defect 6(5/6)
 - Others 5
- ◉ 15/21 ; isthmus dependent IART
 - Success 93.3% isthmus flutters
90.4% total



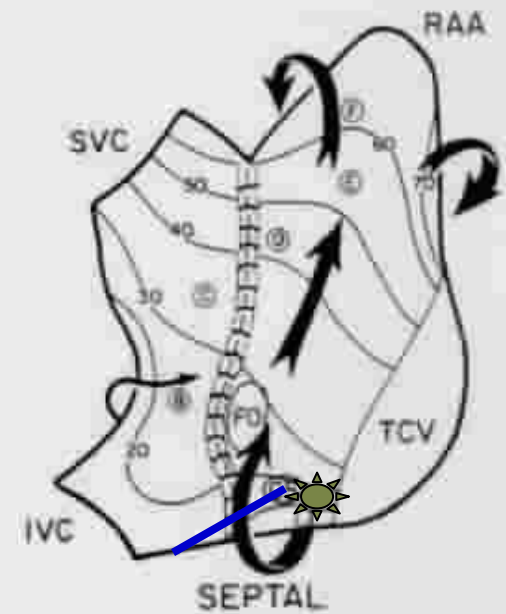
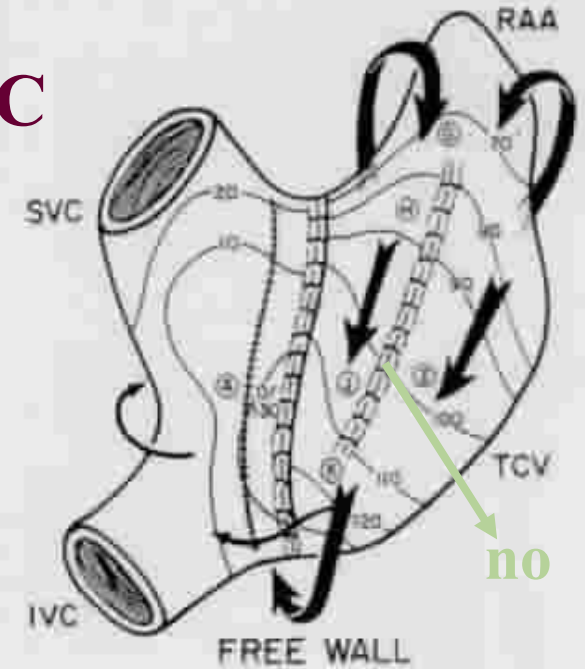
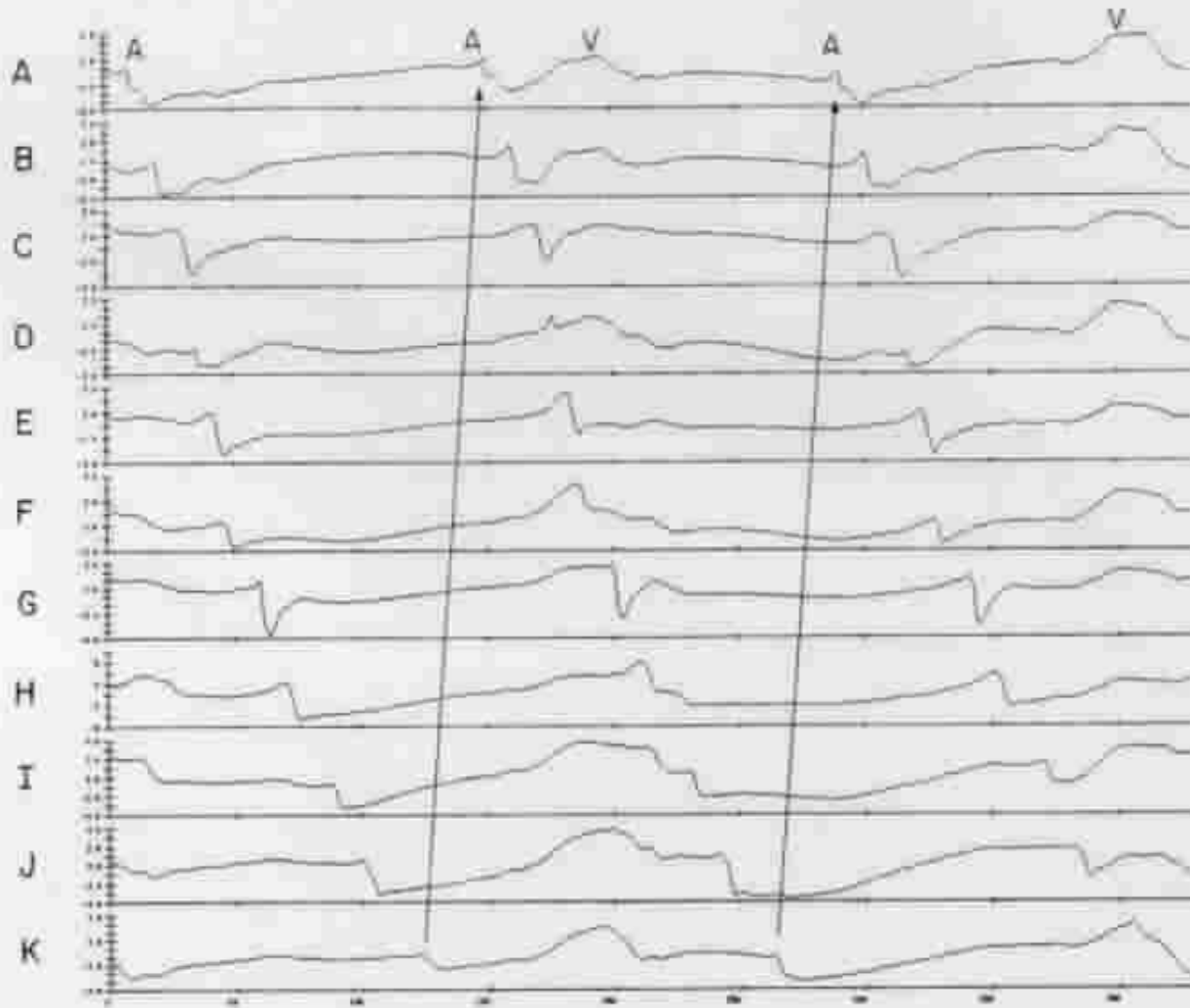
(Collin K.AJC.2000)

	RF Sites	RAVV/IVC Isthmus	Lateral RA Wall	Anterior RA Wall	Atrial Septum	Posterior RA
Mustard/Senning	14	8 (57%)	6 (43%)	0	0	0
Repaired CHD	18	12 (67%)	4 (22%)	2 (11%)	0	0
Fontan*	40	6 (15%)	21 (53%)	10 (25%)	3 (7%)	0

*Fisher's exact test, significant at $p = 0.001$.

IVC = inferior vena cava; RA = right atrium; RAVV = right atrioventricular valve.

Atrial reentrant tachycardia in TCPC



RIGHT ATRIUM

The effect of preventive isthmus cryoablation during lateral tunnel Fontan operation

- SNUCH experience (Kwon BS et al, 1st PCCS)

-From Feb. 1997 to Dec. 2003

Elective isthmus cryoablation

16 patients (M:F = 9:7)

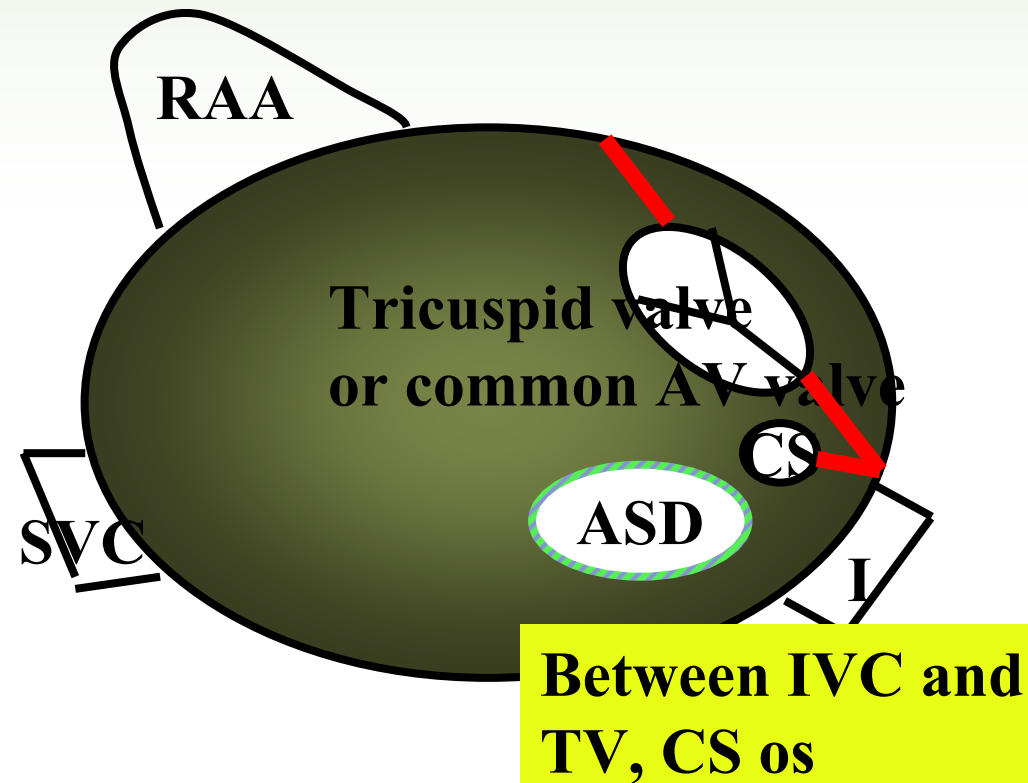
-EP study

From 2001 to 2006

59 patients s/p Fontan

M : F = 40 : 19

10.5 ± 5.0 (range:3-26) yrs



Comparison of Inducible AFL with no AFL (II)

	No AFL (%)	AFL (%)	P-value
BDG	19/ 36 (53)	10 /23(44)	0.430
Other IC repair	23 /36 (64)	15 /23 (65)	0.970
Fontan op			0.032
LT	23 /36 (64)	16 /23 (70)	0.712
APC	1 /36 (2.8)	4/23 (17)	0.070
ECC	10/36 (28)	2/23 (8.7)	0.103
Cryoablation	15/36 (42)	1/23 (4.3)	0.002
Age at BDG	1.07 ± 0.64	1.36 ± 0.92	0.378
Age at Fontan	3.17 ± 4.15	3.99 ± 2.47	0.395
ASD	17/36 (47)	7/23 (30)	0.279
Atrial septectomy	14/36 (39)	9/23 (39)	0.985
ASD repair	0/36 (0)	4/23 (17)	0.019
PAB	6/36 (17)	2/23 (9)	0.464
Pul a. reconstruction	15/36 (42)	7/23 (30)	0.422

Risk Factor of Inducible AFL

	No. (%)	OR	95% C.I.	P-value	
Cryoablation (-)		15.71	.90-129	0.011	
Other IC repair		1.06	0.36-3.17	0.917	
BDG		0.61	0.21-1.77	0.367	
Age at BDG		1.72	0.53-5.54	0.367	
Fontan op	ECC	1.00		0.070	
	APC	5 (8)	15.0	1.65-136.17	0.016
	LT	42 (71)	3.22	0.60-16.27	0.176
Age at Fontan		1.07	0.92-1.24	0.403	
ASD	34 (58)	0.489	0.16-1.74	0.204	
Septectomy	23 (39)	1.01	0.35-2.95	0.985	
ASD repair	4 (6.8)	-	-	NS	
PA banding	8 (14)	0.476	0.87-2.59	0.391	
PA recon	22 (37)	0.61	0.20-1.86	0.386	

Multivariate Analysis of Risk Factors

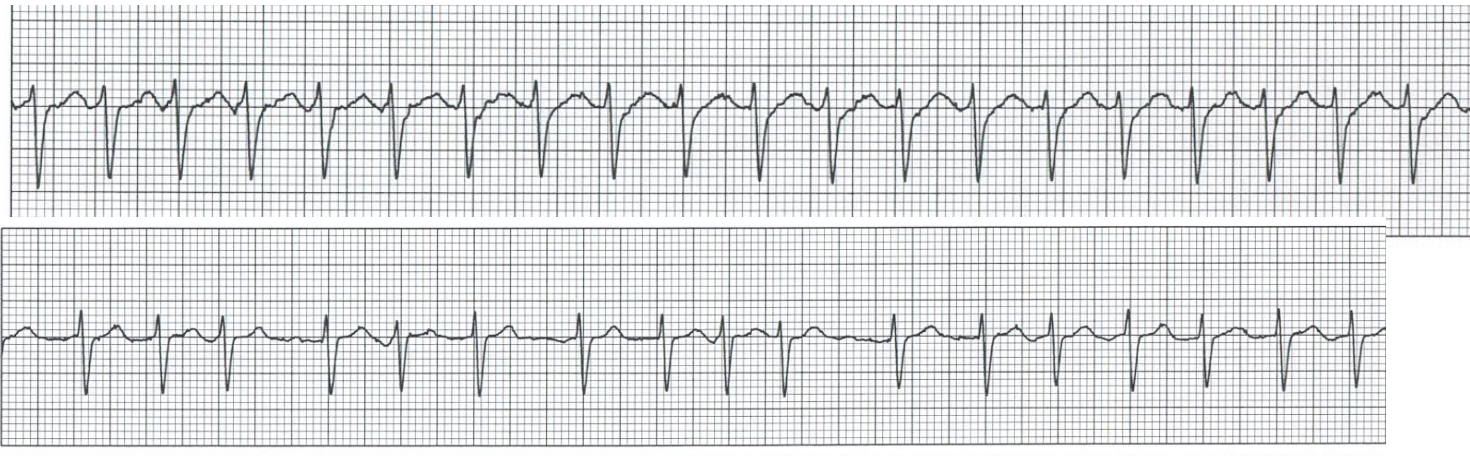
	No. (%)	OR	95% C.I.	P-value
Cryoablation	16/59(27)	2.90	0.27-31.09	0.380
F/u duration		1.43	1.04-1.95	0.026
Age		2.09	1.11-3.92	0.022
Age at EPS		0.53	0.27-1.05	0.069

IART (intraatrial reentrant tachycardia) AFL (atrial flutter)

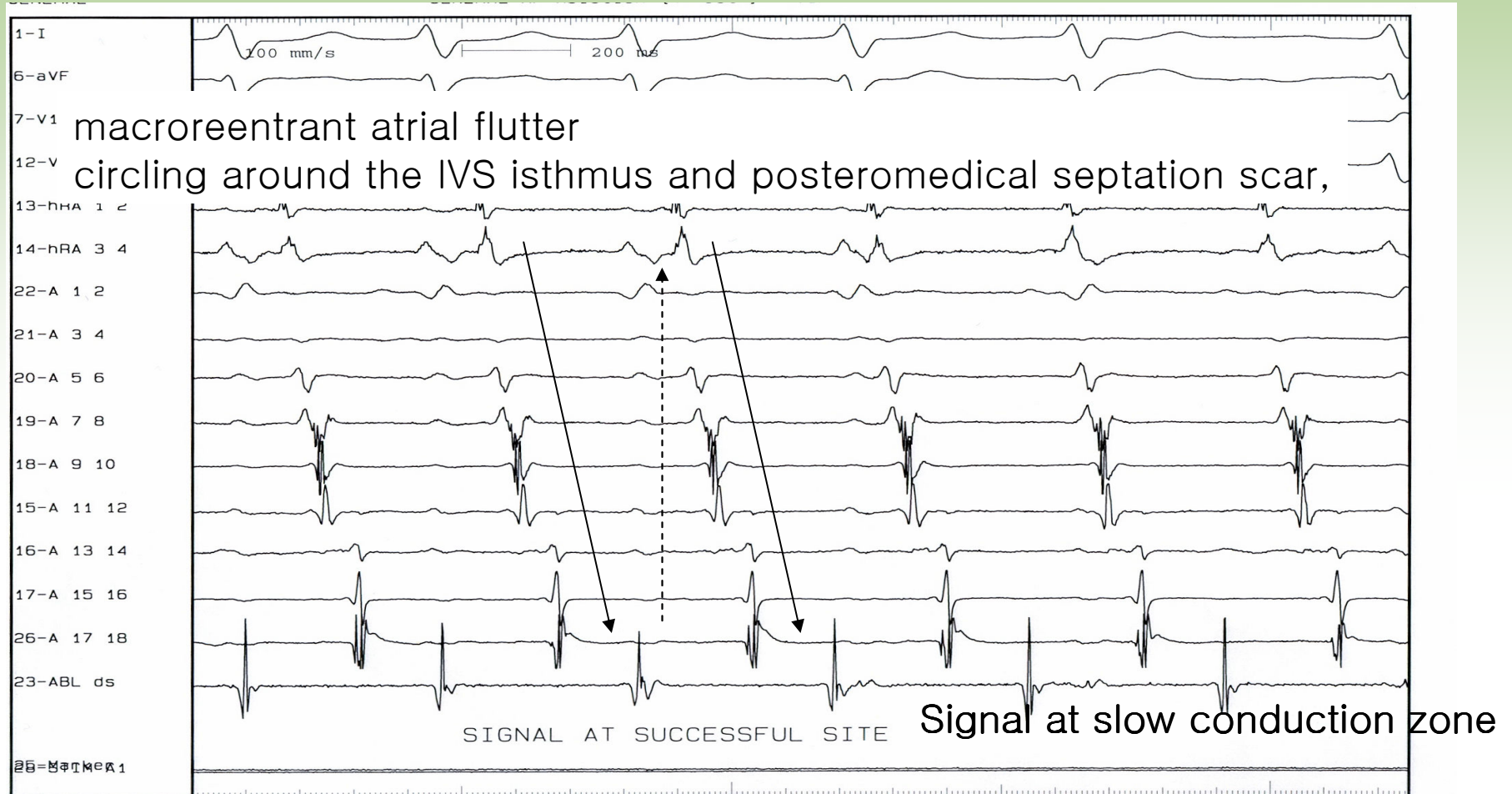
- ◎ 14-29% after Fontan type repair
- ◎ 10-15% after Mustard or Senning op.
 - (Gewilling circulation 1991, Garson , JACC 1985)
 - Mortality ; 17%, sudden death 10%
380 AFL , mean FU 6.5 y (JACC 1985, Garson)
 - Risk of thromboembolism
 - Hemodynamic deterioration
 - Myocardial damage

Case 4. 20 y/ F

- {S.D.D} double inlet RV , DORV , PS
- s/p atriopulmonary connection Fontan op (1989)
- Recurrent symptomatic tachycardia for 4 years in spite of antiarrhythmic medications
(digoxin, beta blocker → sotalol,)
admitted monthly



Radiofrequency catheter ablation



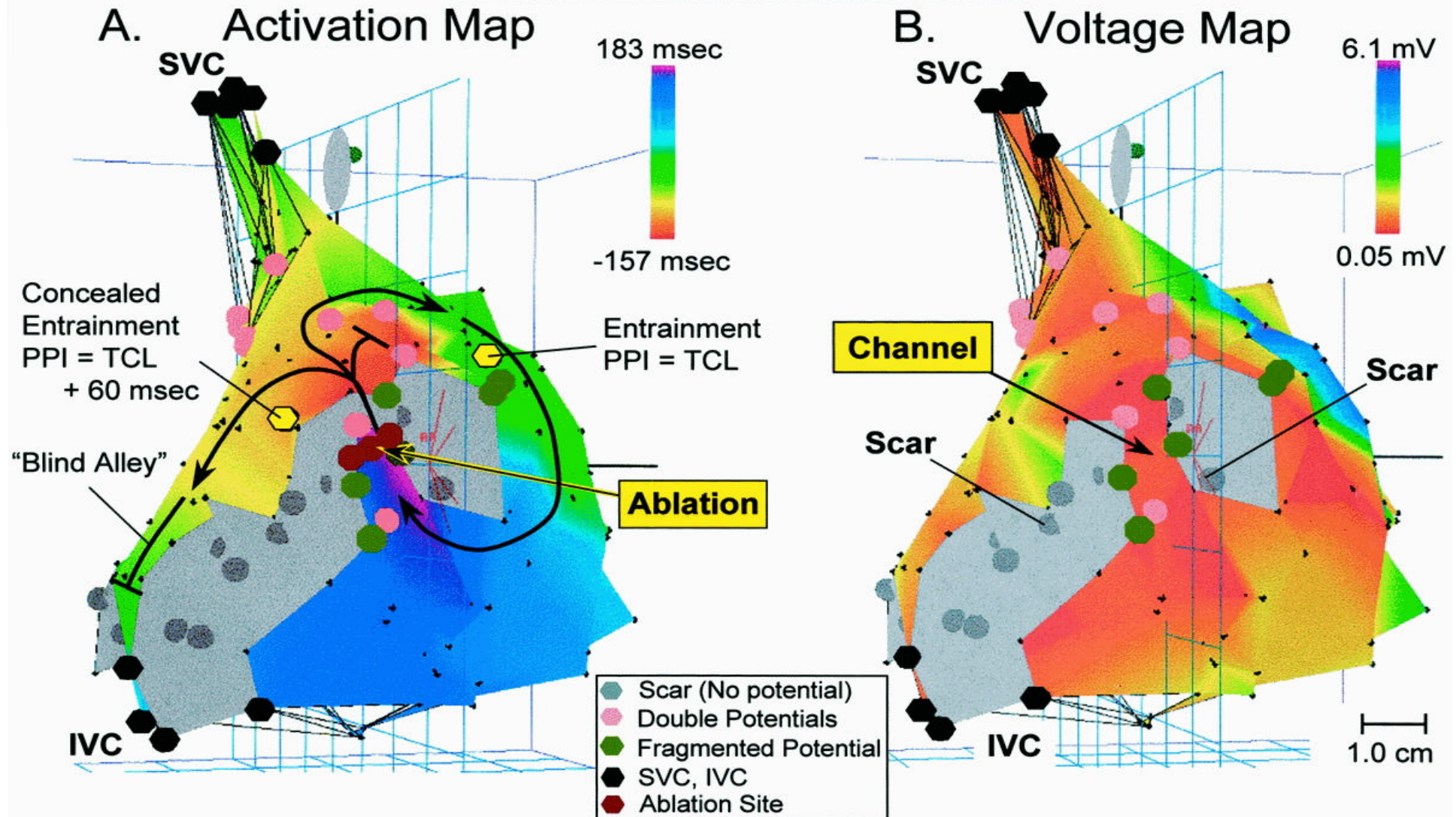
Initial acute success

Recurred 2 m later ,

on sotalol currently , self limited palpitation 0-2/yr for 3 y FU

Advanced spatial referencing technique CARTO system

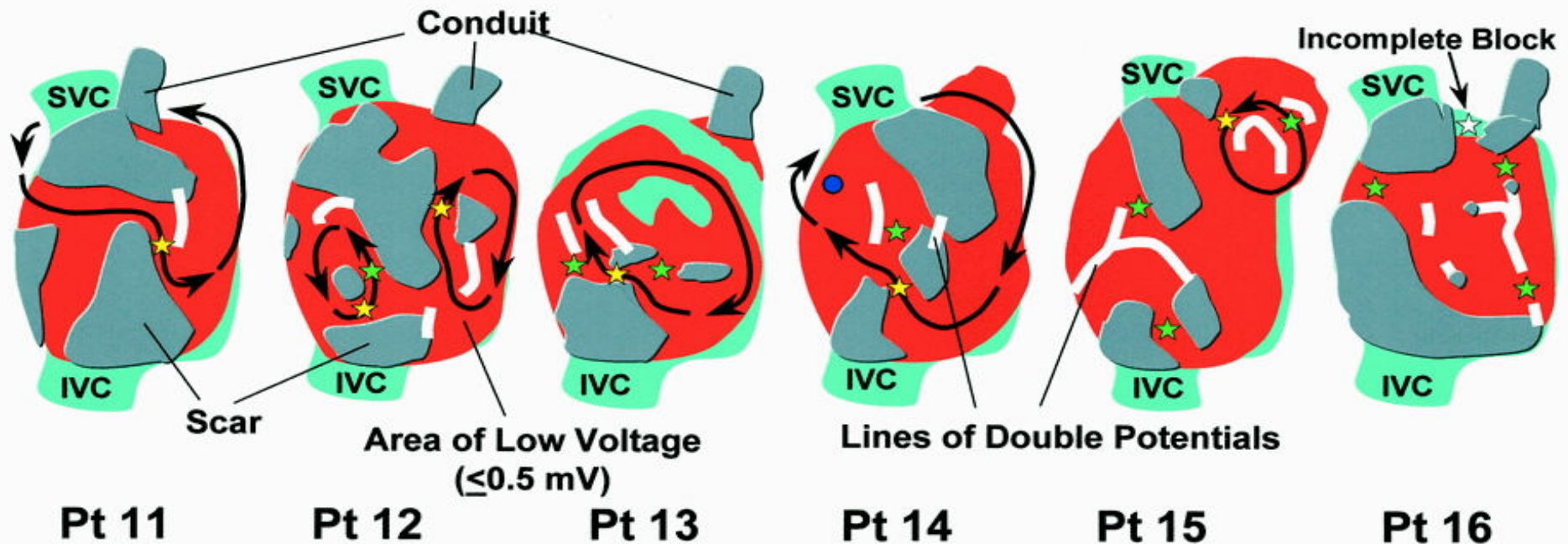
Right Posterior Oblique Projection



Heterogeneous scars and circuits

(Nakagawa H et al. Circulation 2001)

c Fontan Procedure

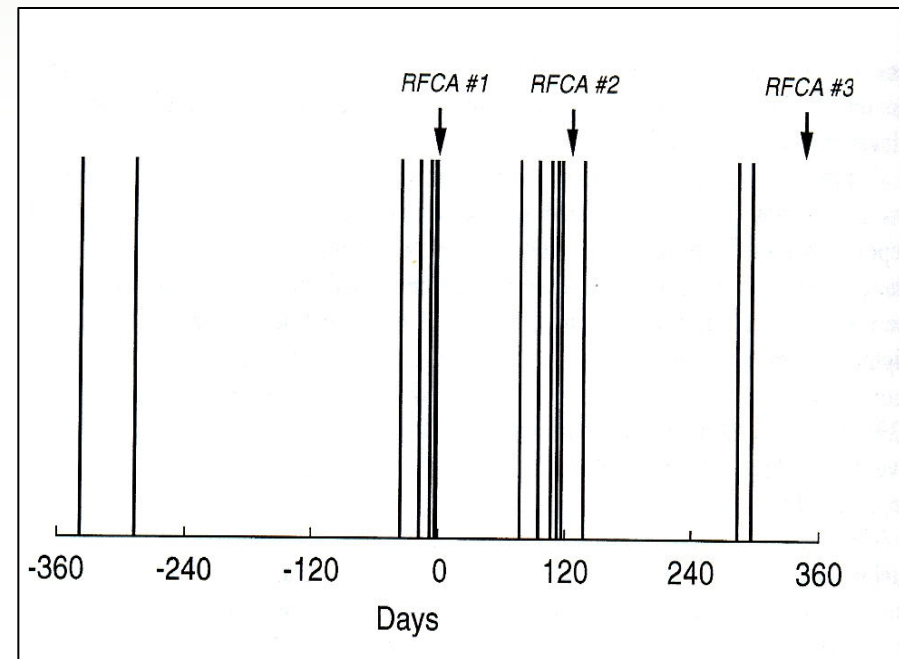
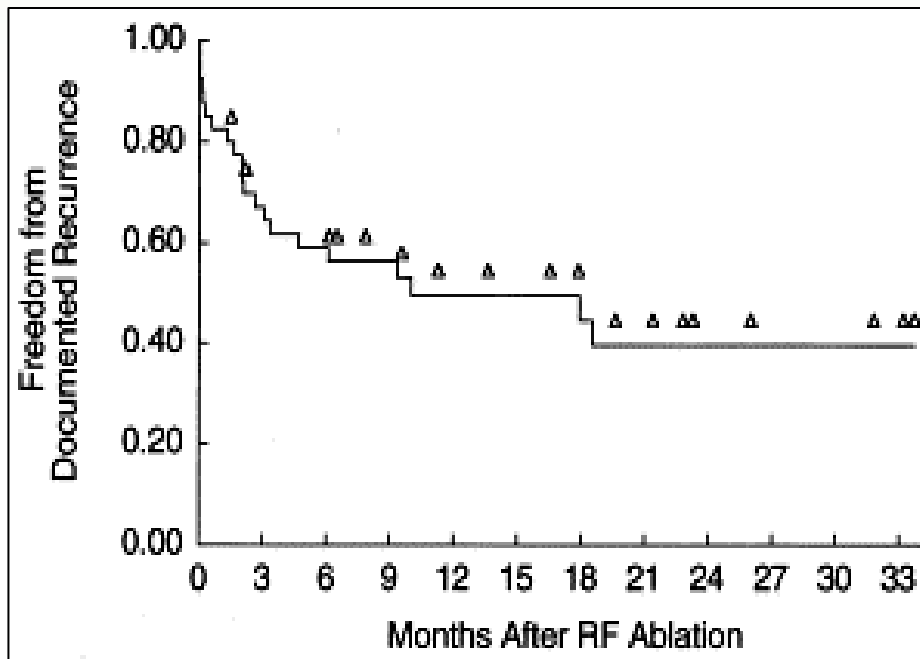


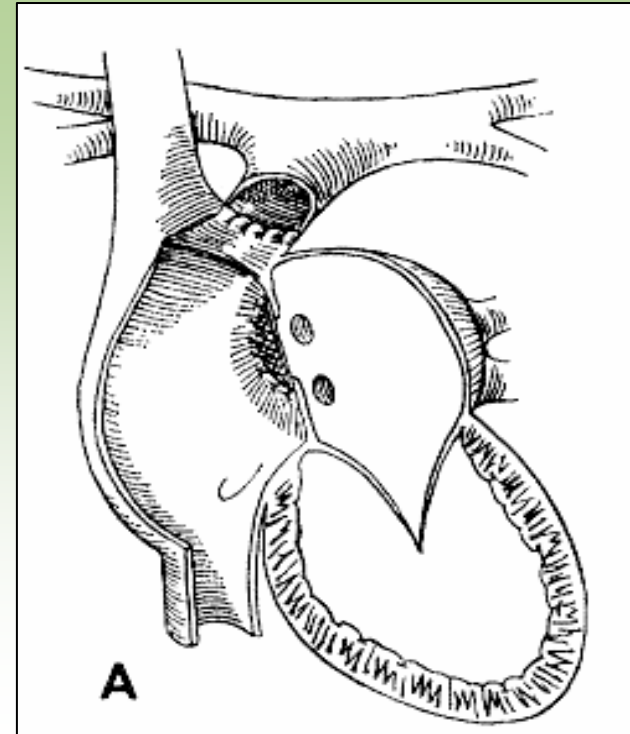
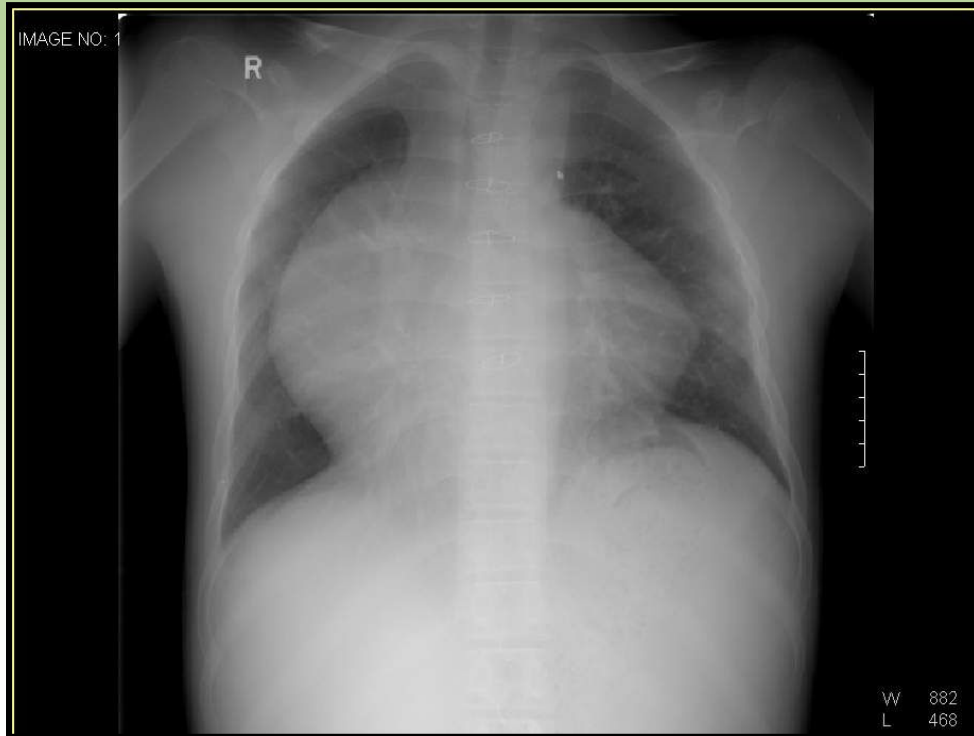
Radiofrequency catheter ablation

Overall success rate; 50-85%

Recurrence rate; 40-50% in a year

(Triedman JK, JACC 1997)





Case 6. 16y/m, Tricuspid atresia

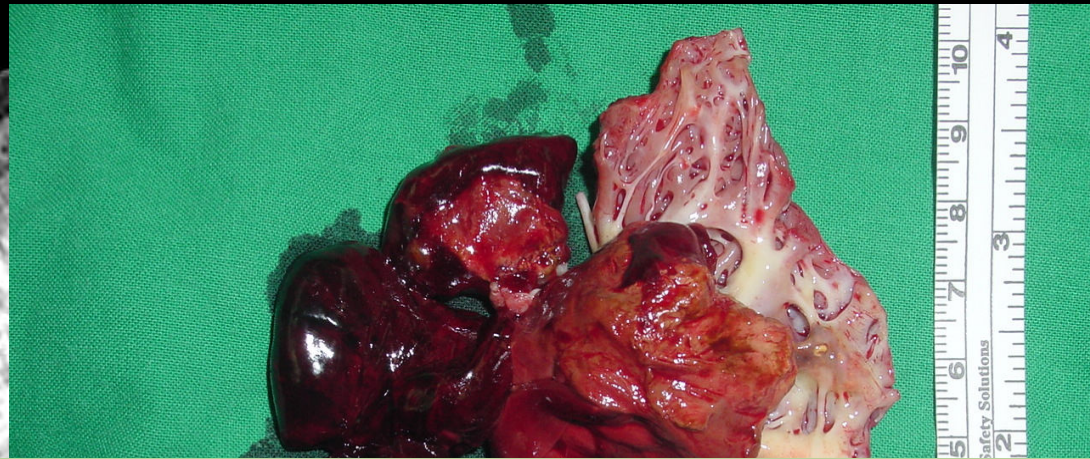
S/p Atriopulmonary connection

lost FU for 10 y and visited ER

- tachycardia (atrial flutter), progressive edema, dyspnea

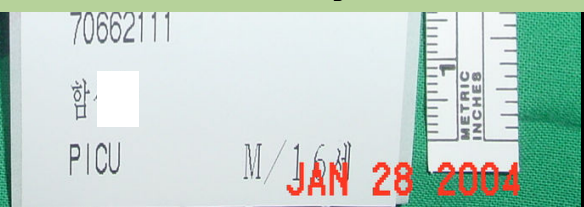
IMAGE NO: 235
LightSpeed Ultra
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FFS
Se 2
Im 234
1.3thk



No recurrence of atrial flutter for 3 y
Warfarin, enalapril, low dose of diuretics
NYHA FCII, full time job , mild liver dysfunction

Fontan con
RA reducti
RA maze o



TP -153.7
TI 446
kVp 100
mAs 688
GT 0
2004-01-05
11:28
Chest CT (Pulmonary ar

50mm

W 350
L 40

Atrial flutter, thrombosis, hemodynamic outcome

systemic AVVR, biatrial enlargement

RA thrombosis, heart failure

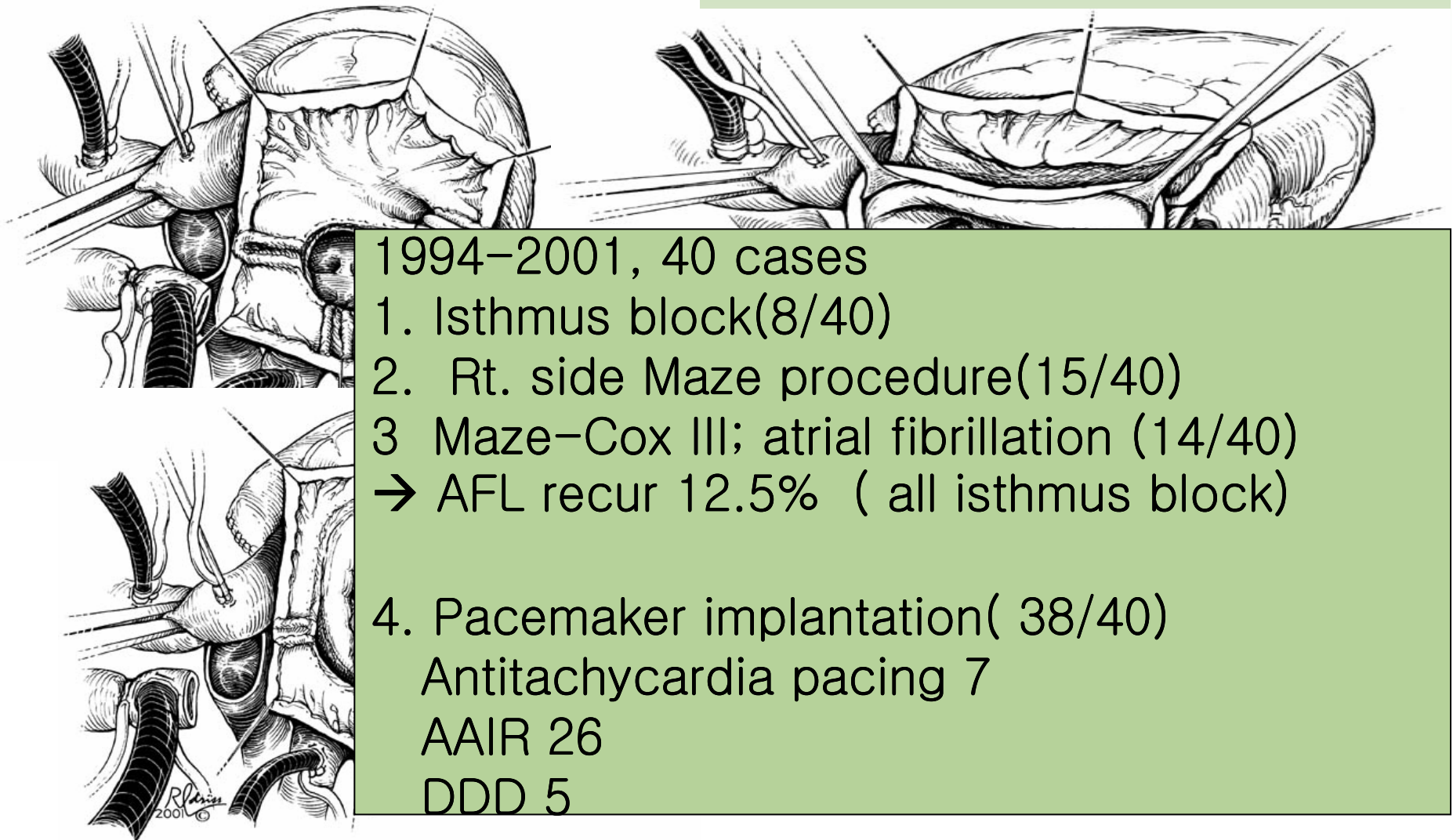
(Ghai et al., JACC 2001)

Clinical Outcome	Arrhythmia Group	Arrhythmia-Free Group	p Value
Heart failure	18 (46)	7 (13)	0.0036
Right atrial thrombus	12 (31)	2 (4)	0.0062
Pulmonary embolus	2 (5)	5 (9)	0.37

Chamber Dimension	Arrhythmia Group		Arrhythmia-Free Group		p Value
	n	Dimension or Volume (mean \pm SD)	n	Dimension or Volume (mean \pm SD)	
RA-ML	29	59 \pm 19 mm	35	49 \pm 13 mm	0.019
RA-SI	29	60 \pm 18 mm	35	51 \pm 15 mm	0.037
RA Volume	29	139 \pm 149 ml	35	76 \pm 54 ml	0.040
LA size	32	44 \pm 10 mm	44	37 \pm 9 mm	0.002

Conversion Fontan and arrhythmia surgery

Mavroudis C, Deal BJ et al. JTCS 1998 , JTCS, 2001, Circulation 2002



1994–2001, 40 cases

1. Isthmus block(8/40)

2. Rt. side Maze procedure(15/40)

3 Maze–Cox III; atrial fibrillation (14/40)

→ AFL recur 12.5% (all isthmus block)

4. Pacemaker implantation(38/40)

Antitachycardia pacing 7

AAIR 26

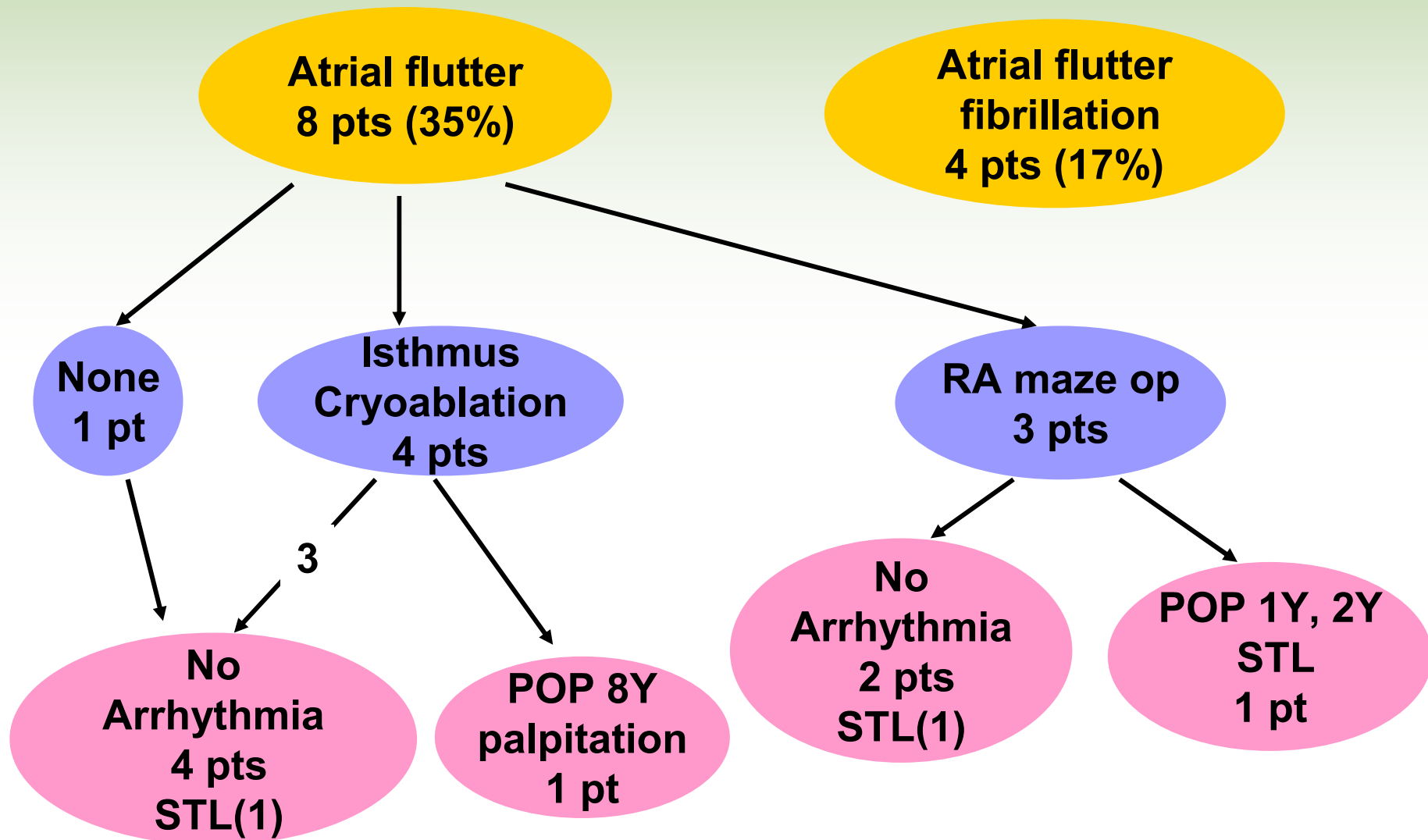
DDD 5

Fontan conversion and arrhythmia surgery

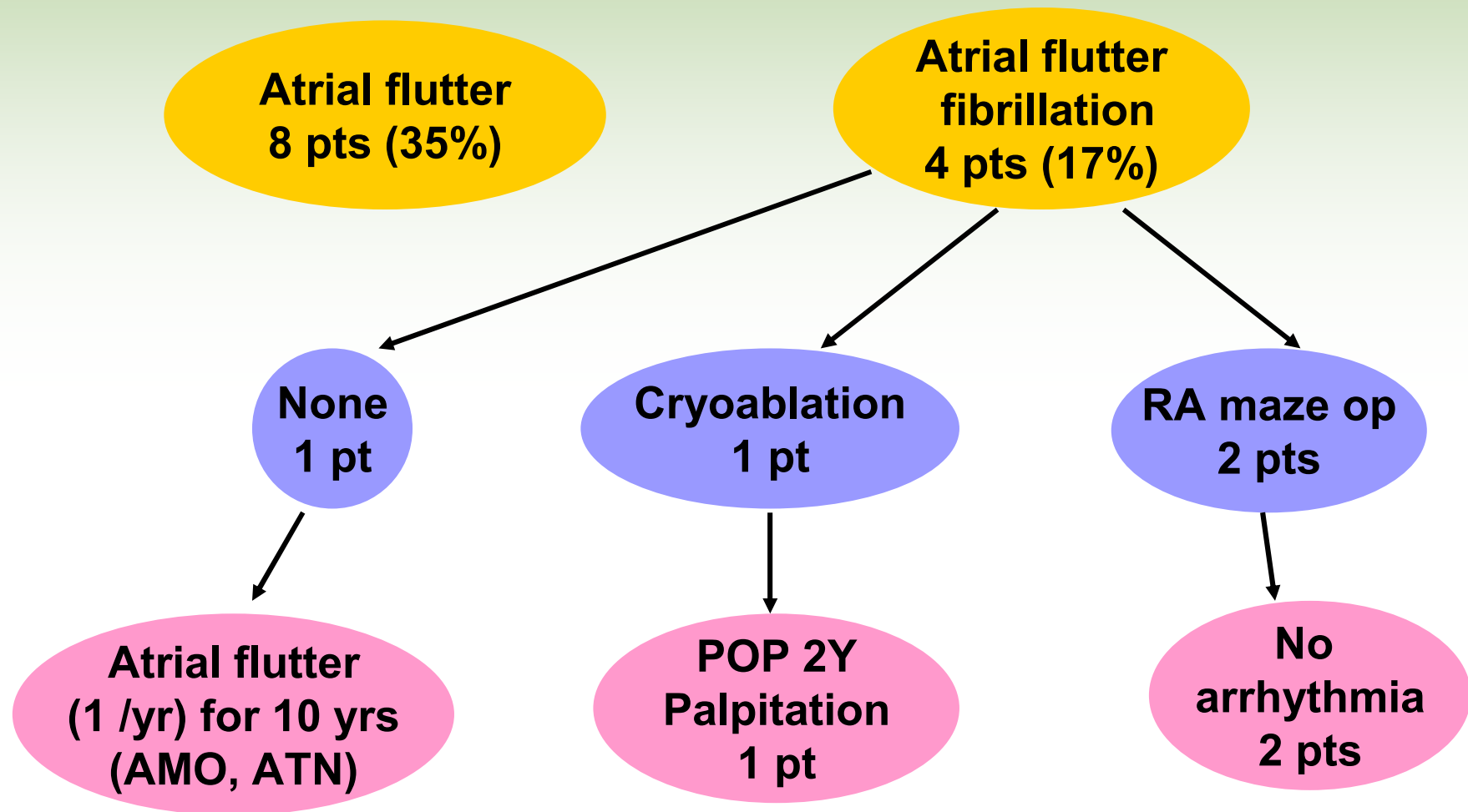
SNUCH experience (Kim WH, 2005 EJCS)

- Fontan conversion ; 23 / 392 patients
 - Fontan failure 11
 - Intractable Arrhythmia ; 13
- Atrial flutter : 12/23 (52%)
Atrial fibrillation: 4/23 (17%)
Sinus node dysfunction : 13 /23 (57%)
- Arrhythmia surgery:
 - Isthmus cryoablation :13/23 (57%)
 - Right-sided maze : 5/23 (22%)
- Permanent pacemaker insertion :14/23 (61%)
- OP mortality; 2 (severe PLE)

Arrhythmia

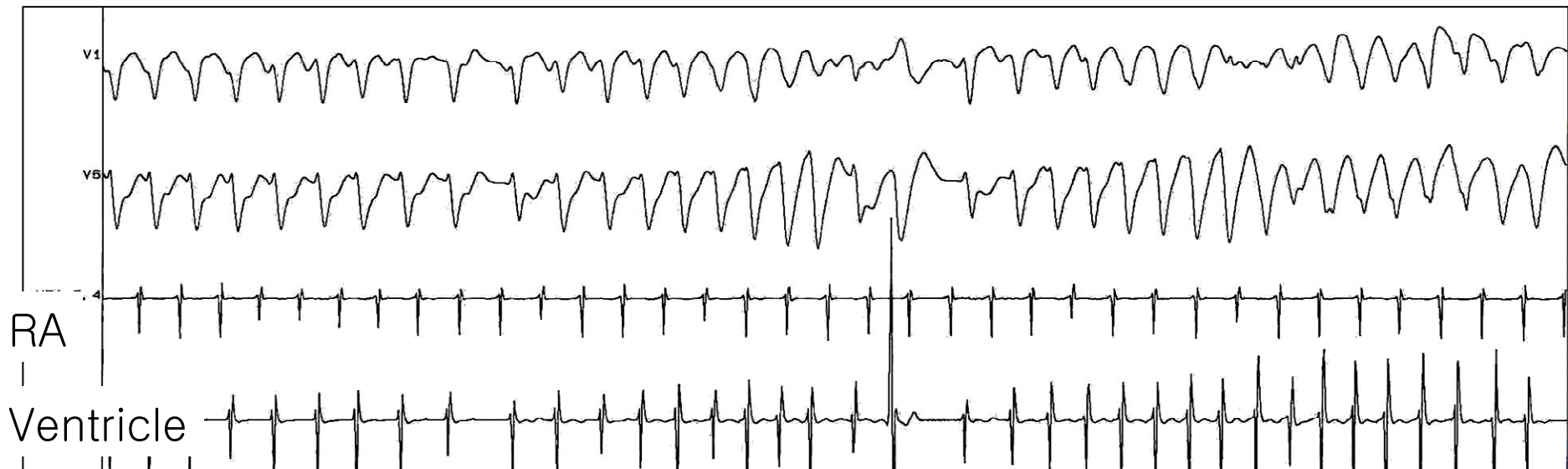


Atrial flutter → RA maze
Atrial fibrillation → Cox –Maze III



Case7. 22/F

- Dx> Single inlet RV, Dextrocardia,
- s/p 90.6 APC Fontan op. & AV valvuloplasty
- Lost FU for 6 y
- Visted ER d/t seizure-like episode
- EPS; induced atrial flutter with rapid ventricular conduction



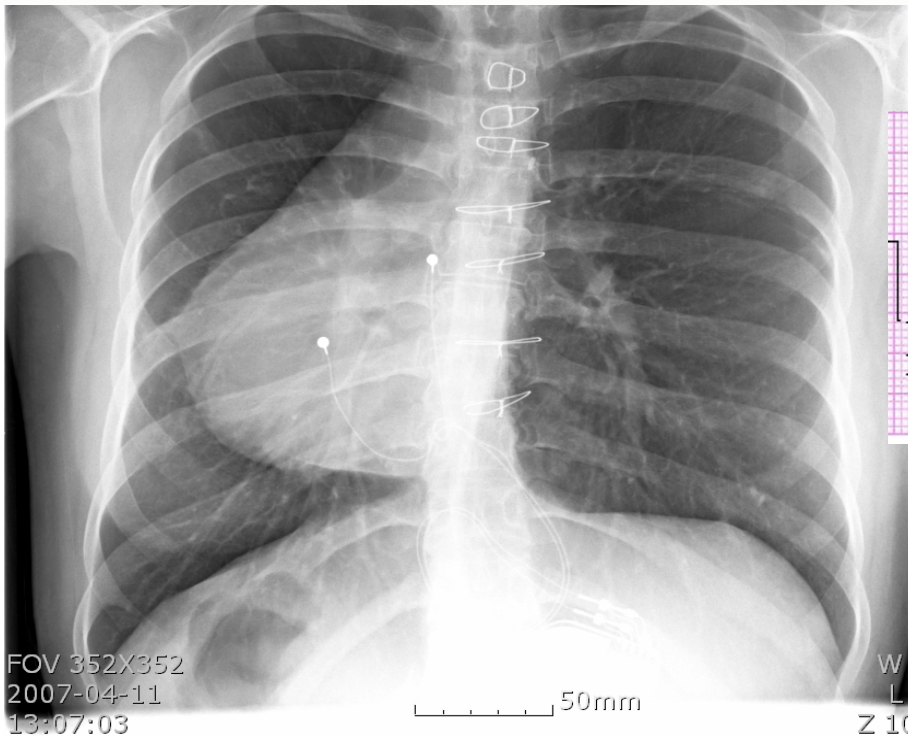
AFL → degenerated to polymorphic VT, VF

Sotalol for 6m

FU EPS ; still induced AFL with 1:1 AV conduction

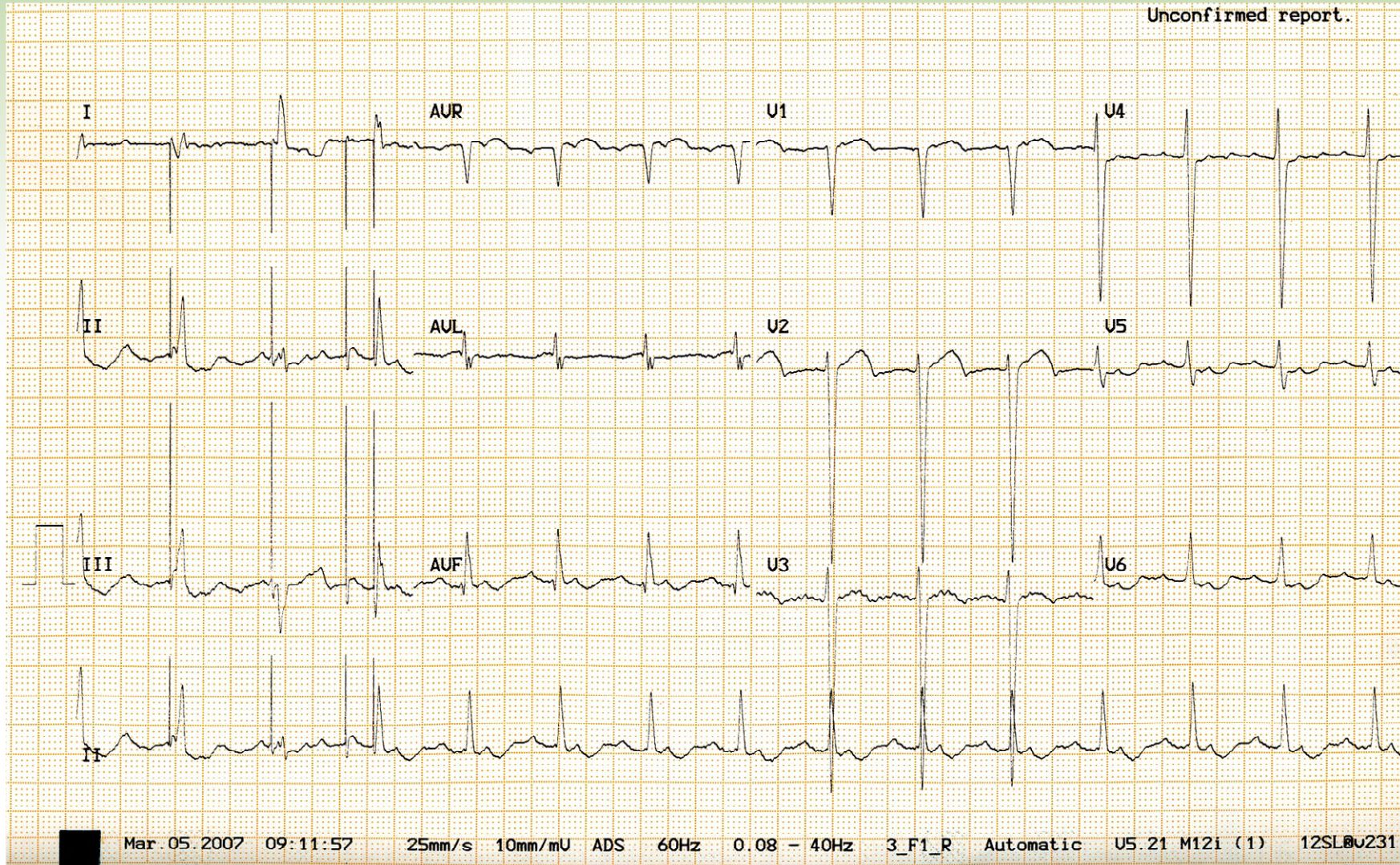
→. Redo Fontan. Rt. side Maze op.

- Fontan conversion : Extracardiac conduit TCPS
 - Arrhythmia surgery : Rt. side maze (cryoablation)
- Single AV valve valvuloplasty & annuloplasty
- Permanent pacemaker (DDDR epicardial),
basal rate 90–80/min



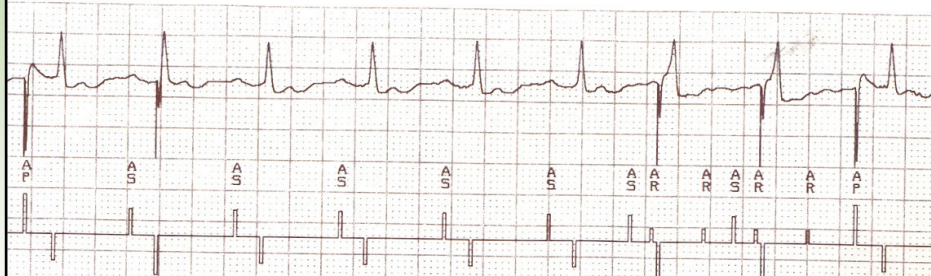
First degree AV block

Recurred atrial flutter, slower rate, 2:1 AV conduction
Mild palpitation

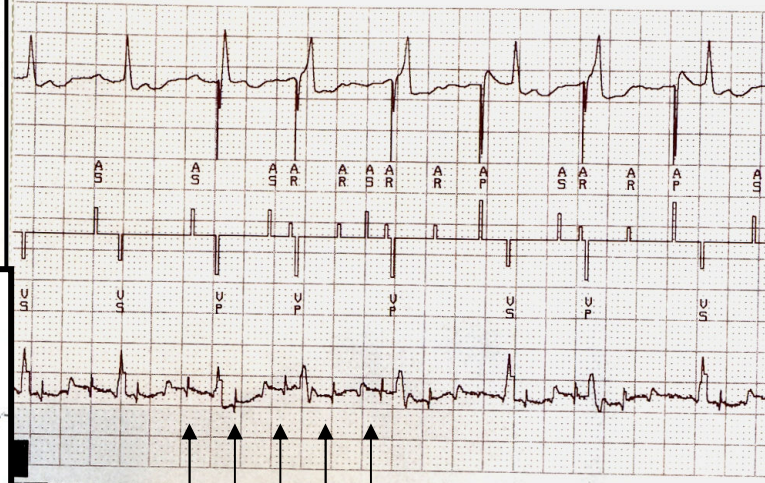


Antitachycardia pacing via telemetry

Burst A 210 msec



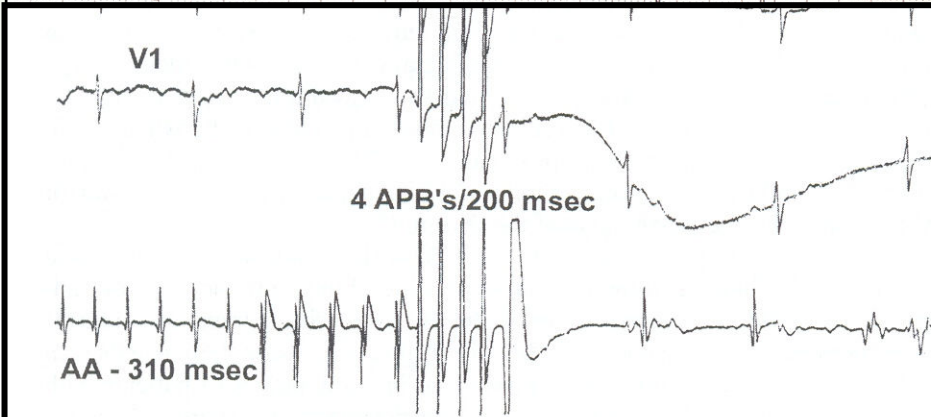
Burst A 240 msec



V1

4 APB's/200 msec

AA - 310 msec



Terminated AFL

TCL 300 msec

Burst A 210 msec (via A00 pacing)

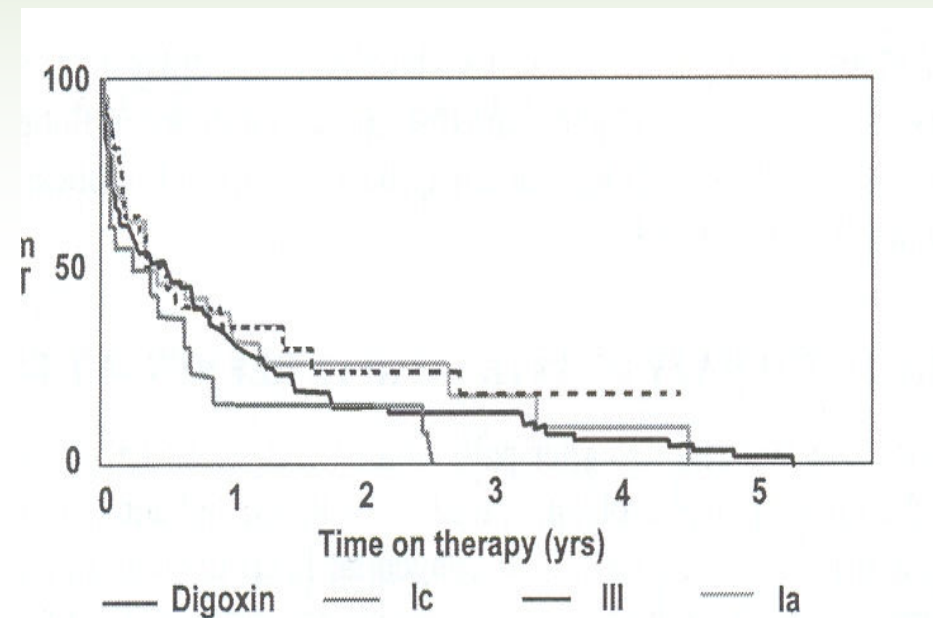


Tx of intraatrial reentrant arrhythmia

- Antiarrhythmic medication ;class III, Ia
- antibradycardia pacing \pm permanent overdrive pacing
- antitachycardia pacing
- RF ablation
- operation : conversion of atriopulmonary connection to TCPC; atriectomy; Maze operation in RA
- drugs (anticoagulation, DGX, diuretics, ACE inhibitor)

Chronic Mx of IART

- Antiarrhythmic medication
 - class III; sotalol, amiodarone, ibutilide
 - beta blocker, digoxin
 - procainamide



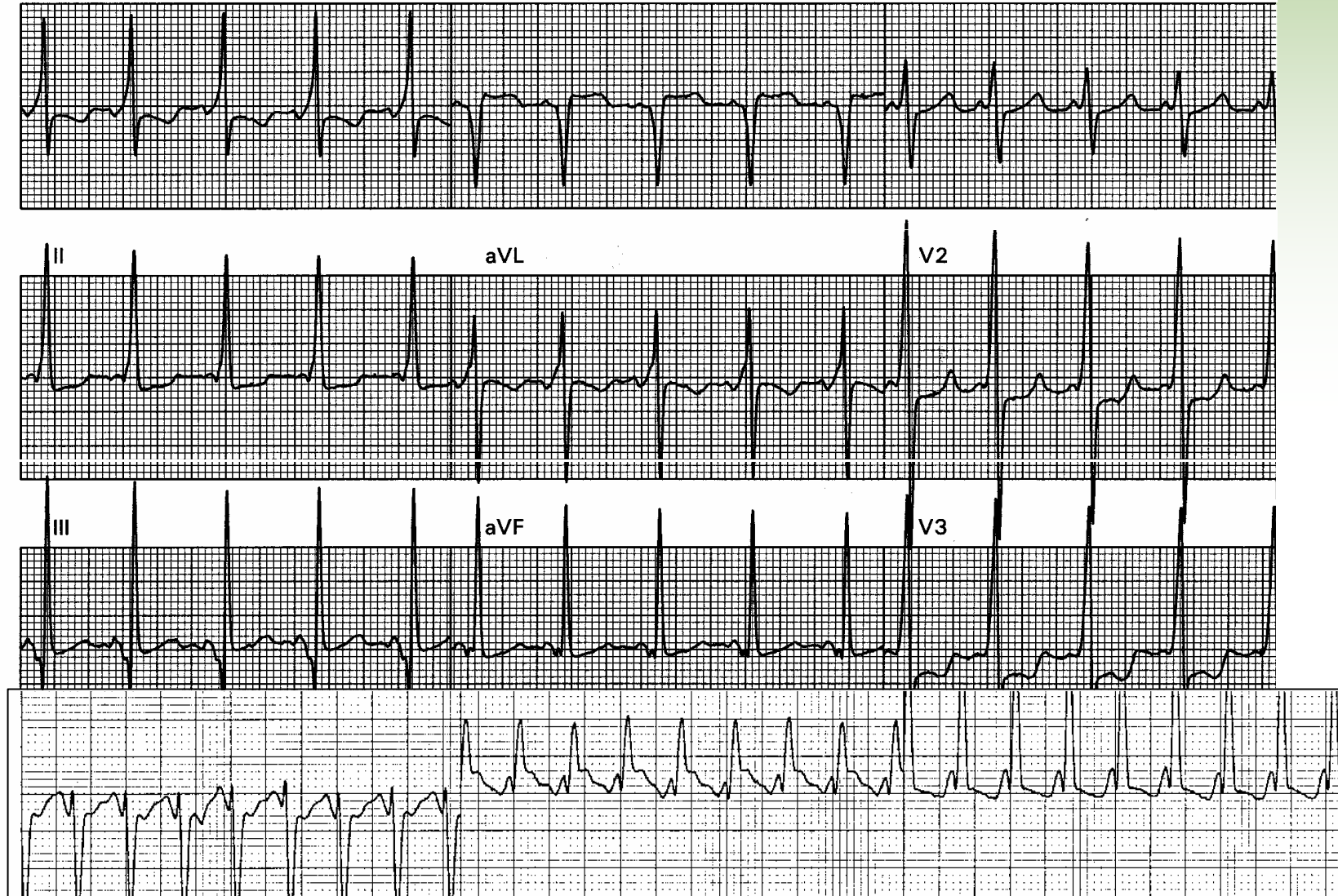
adverse effect;

- aggravation of SN dysfunction,
- paradoxical inc of ventricular conduction
- Other side effect of drug itself

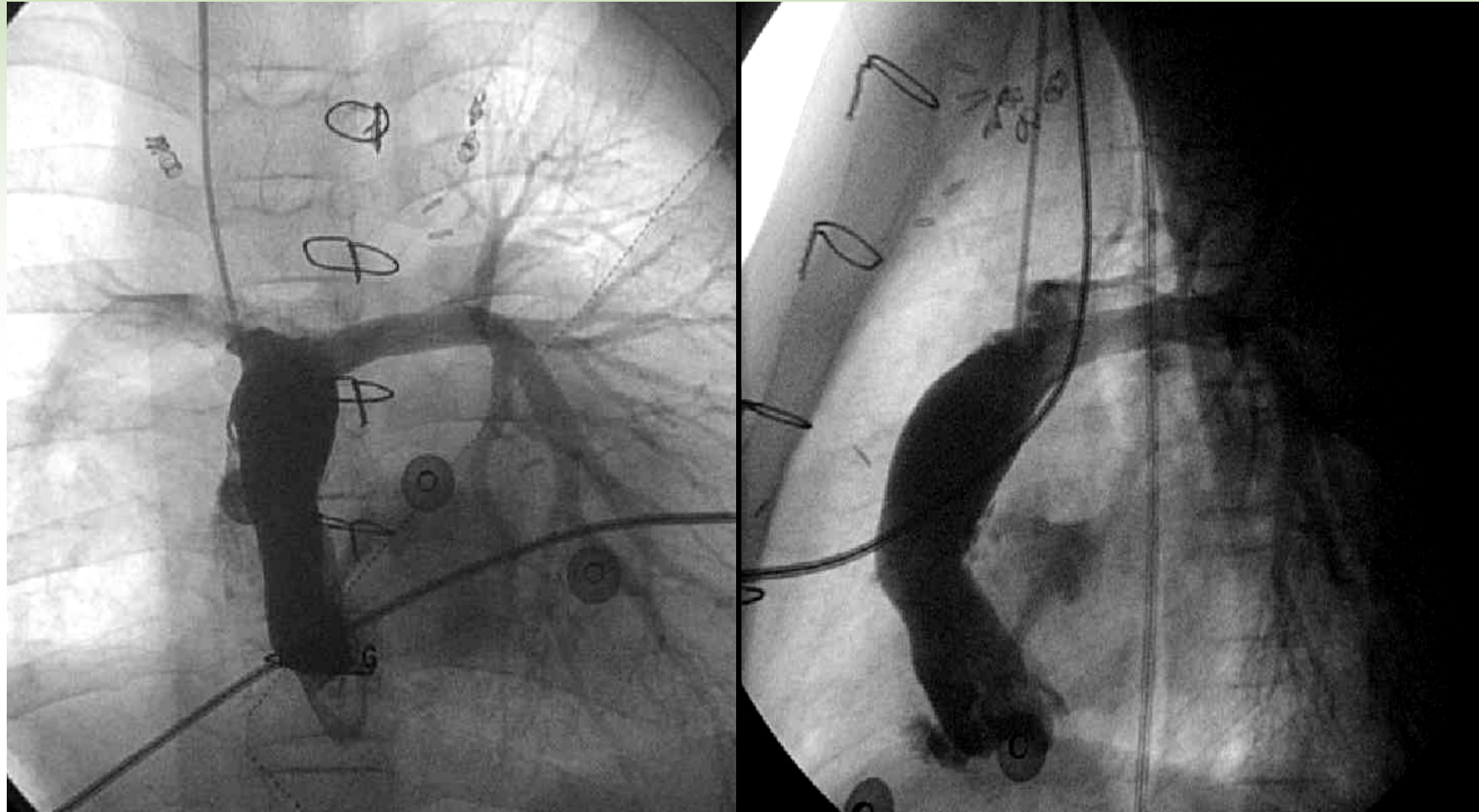
Other supraventricular tachycardias

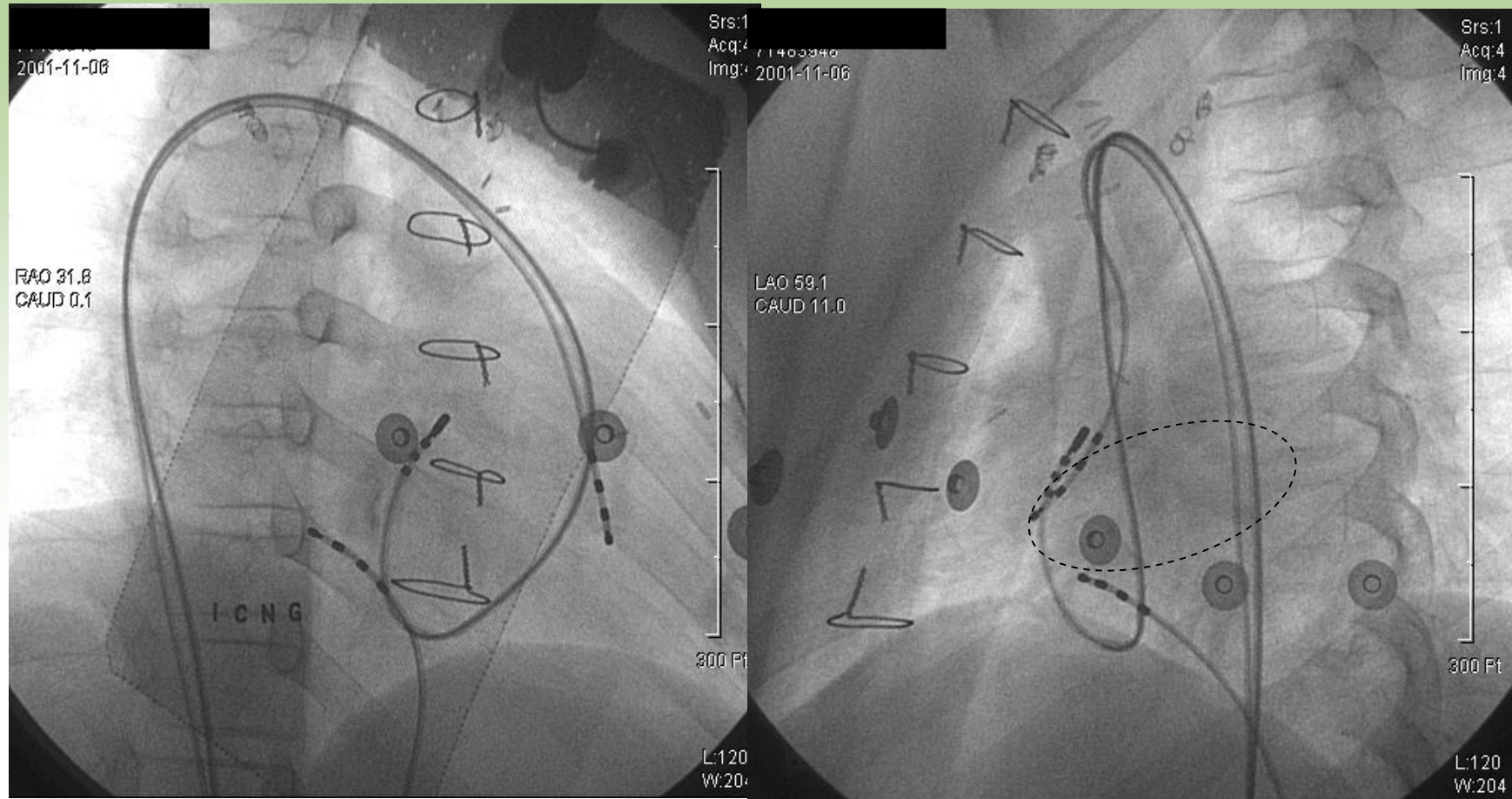
- Atrioventricular accessory pathway
WPW syndrome
 - Rare spontaneous resolution of WPW in congenital heart anomaly
- Twin AV node related tachycardia
- Focal junctional tachycardia
- AV nodal reentrant tachycardia

Case. 11y/F, s/p Fontan common inlet RV, DORV



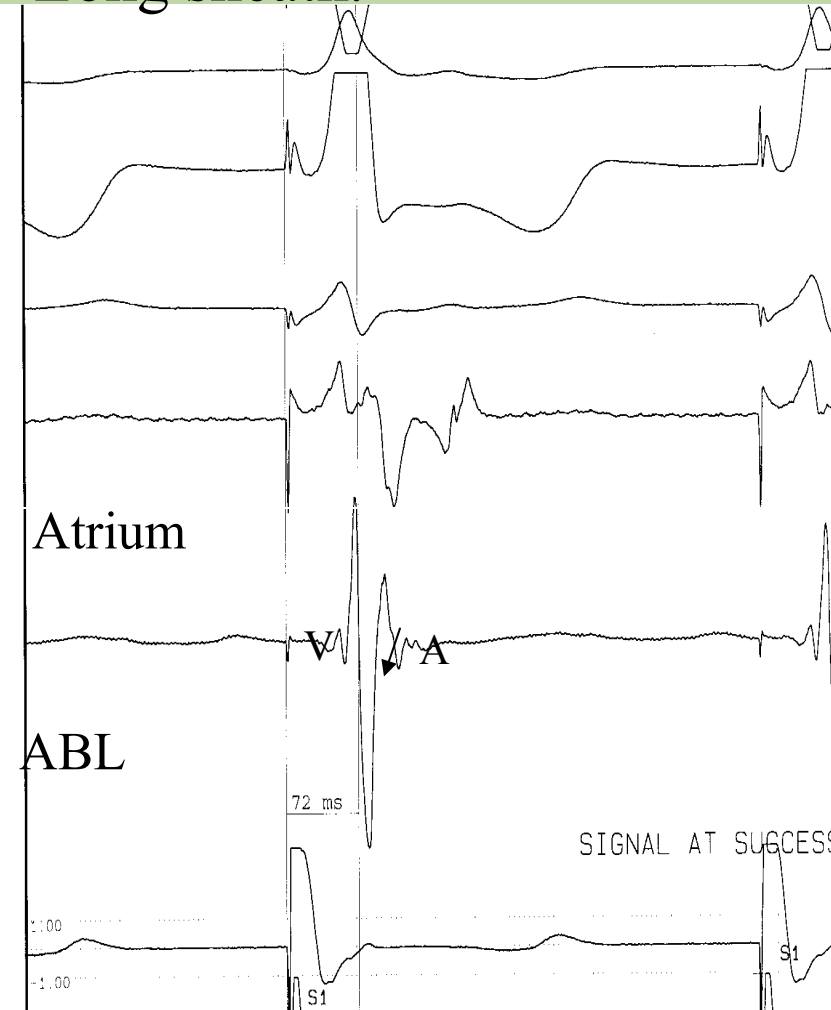
S/p Lateral tunnel Fontan operation





Retrograde atrial approach, 2 arterial accesses
Poor ablation catheter stabilization
Unstable V pacing through retrograde V pacing

3rd session, antegrade approach Long sheath.

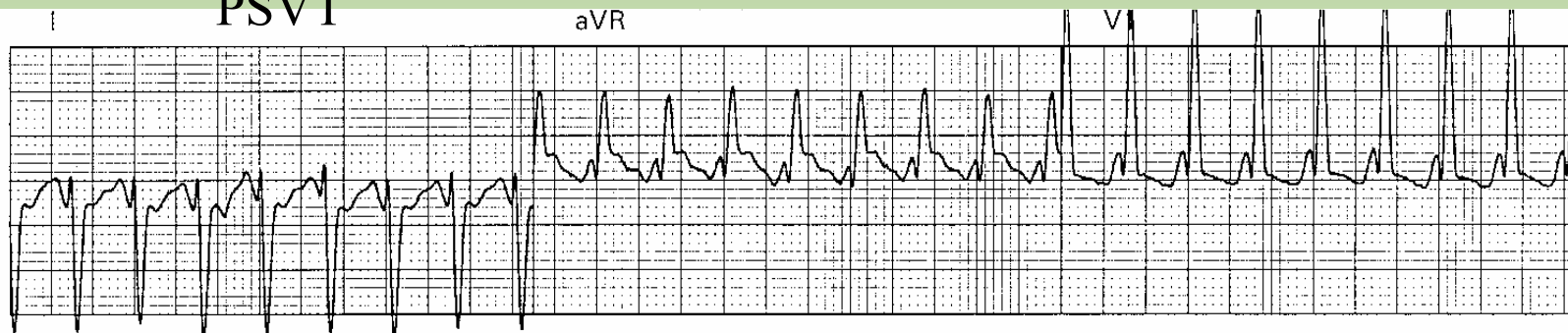


Symptomatic tachycardia or delta wave
→ EPS before Fontan completion
Accessory pathway → RF ablation before Fontan completion

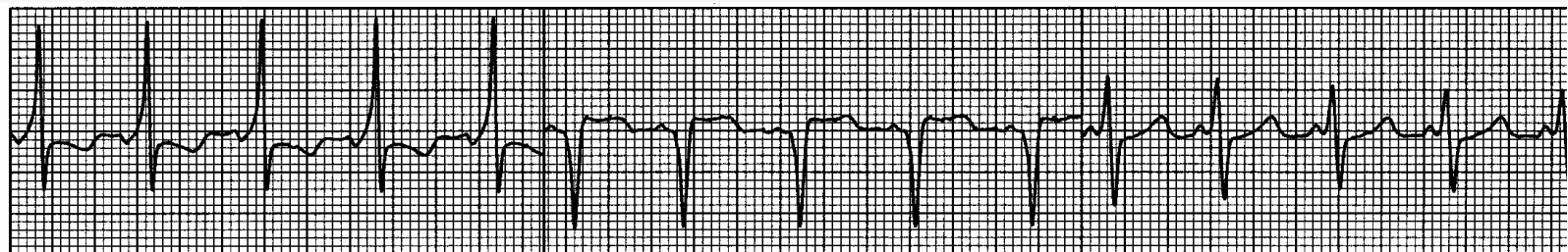
PSVT

aVR

V1



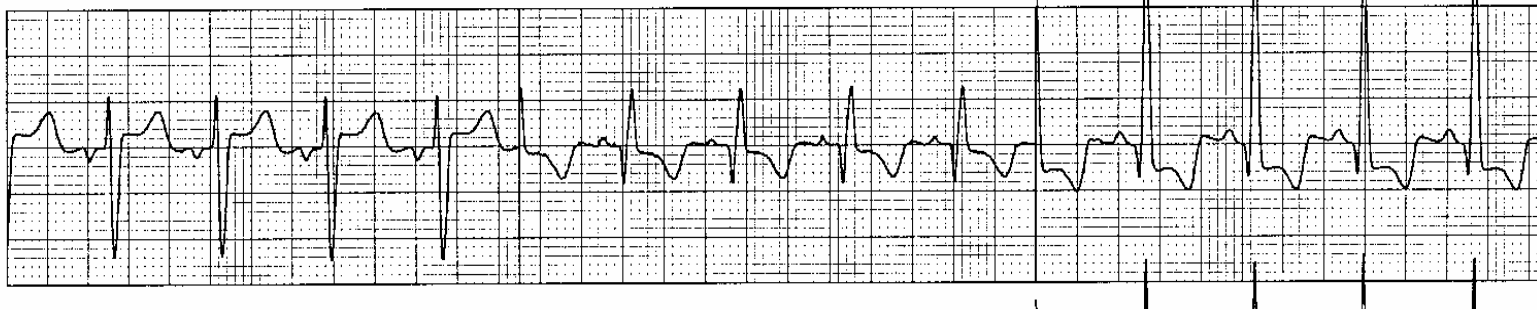
Before RFCA



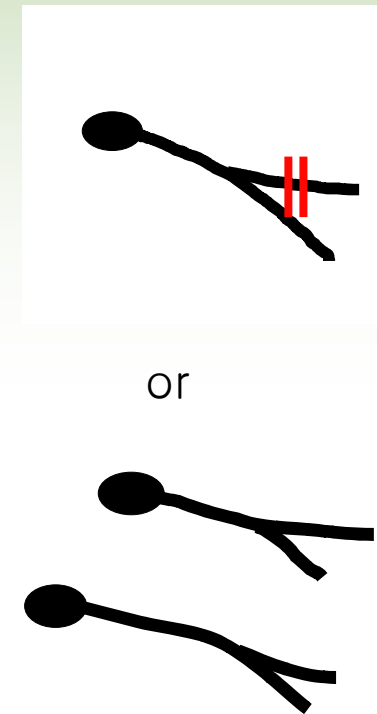
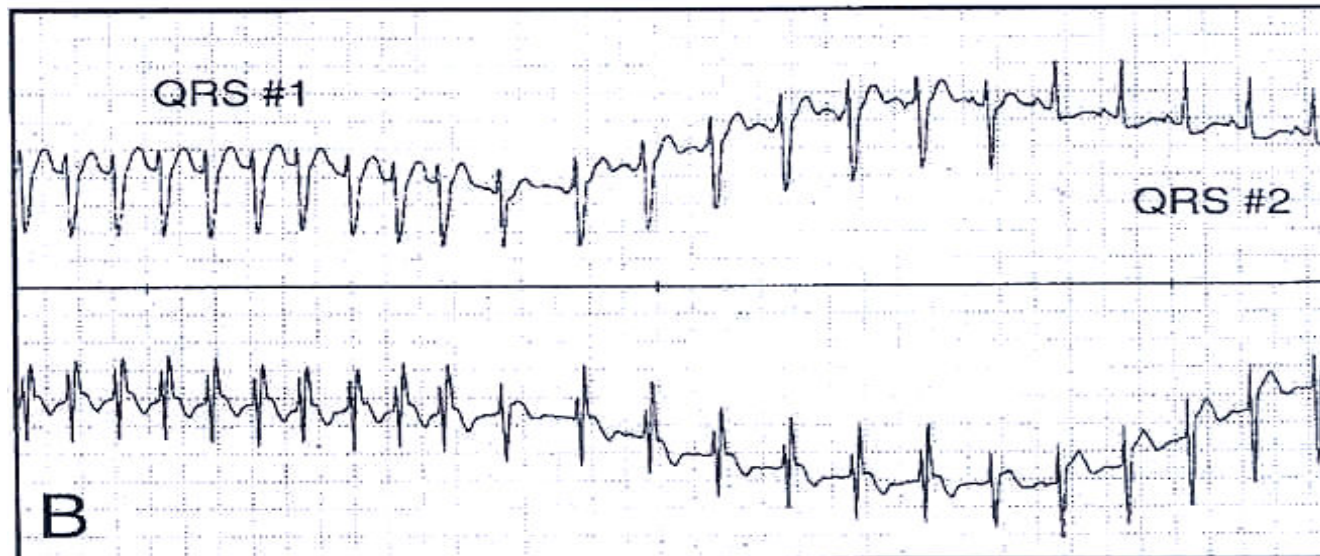
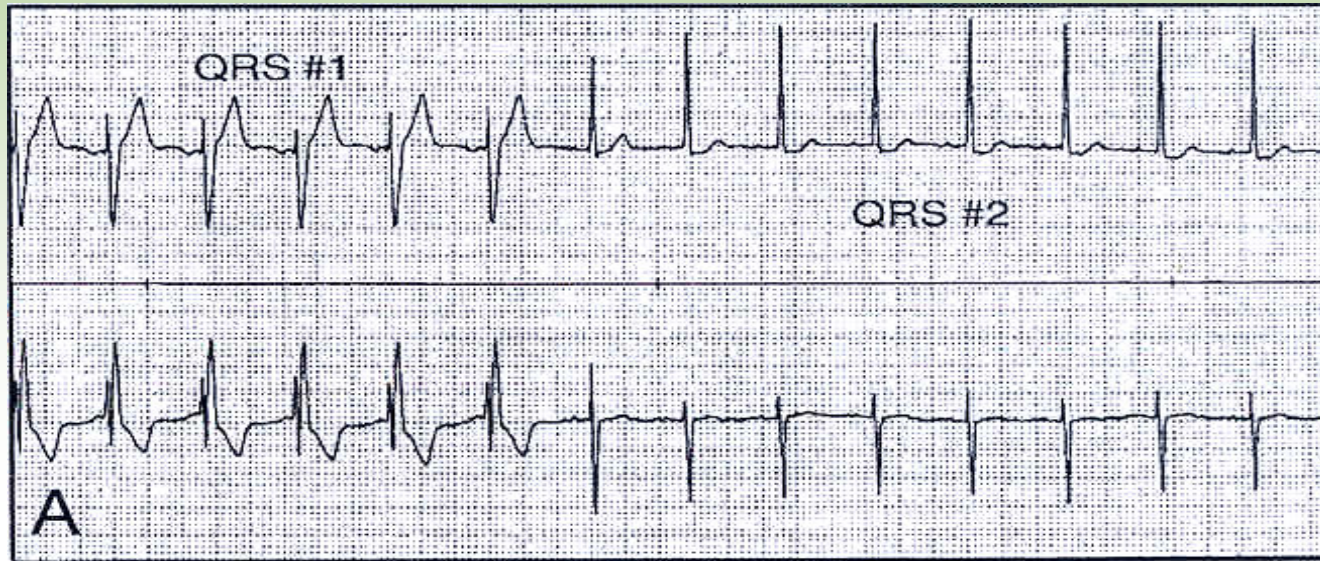
After RFCA

aVR

V1

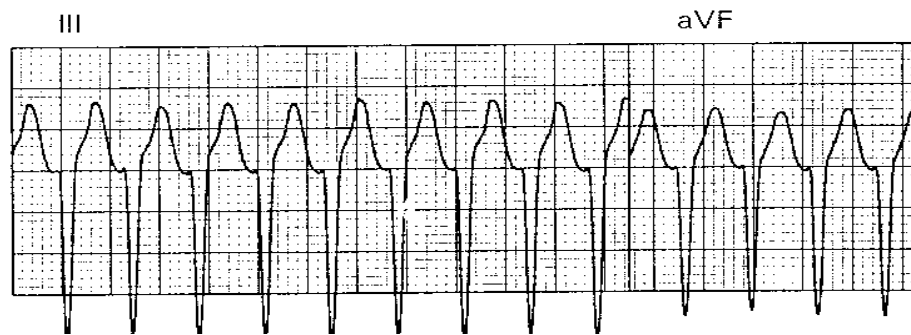
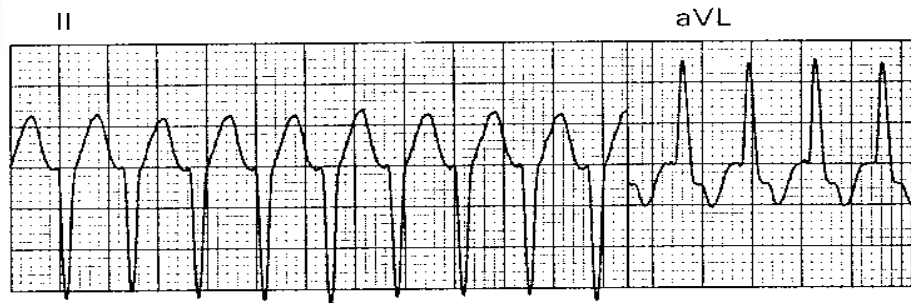
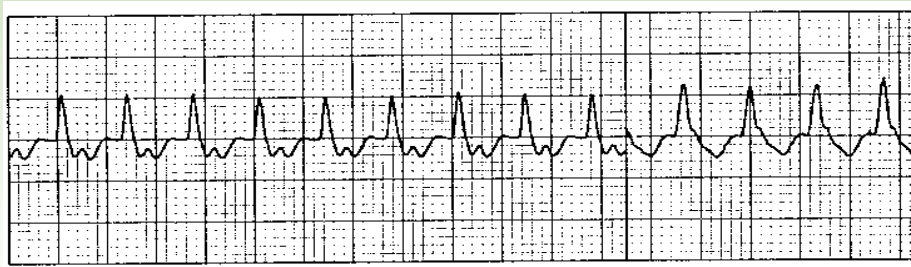


Changed QRS without delta wave In complex congenital heart disease



(Epstein M et al, 2001)

Case, 4y/m



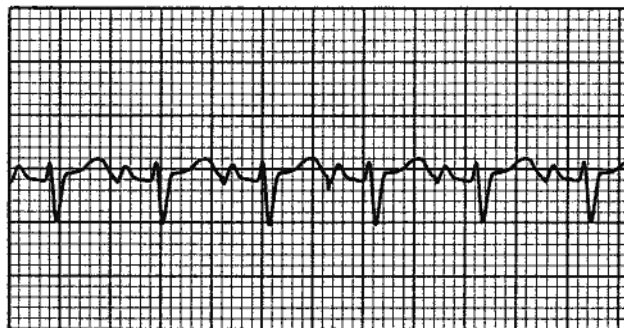
Right isomerism
Common inlet RV
DORV PS
S/p BCPS,

Adenosine sensitive PSVT for 3 y

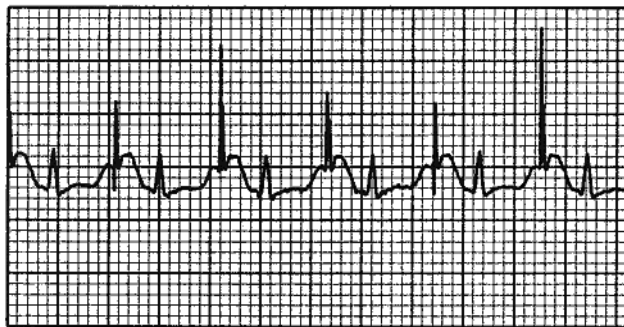
EPS

No preexcitation
decremental VA conduction
VA conduction was blocked by
adenosine
Induced SVT by V pacing

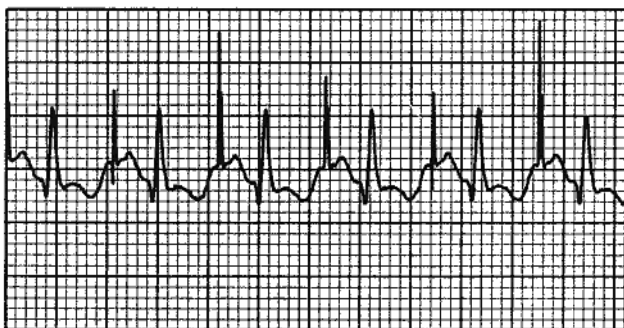
I High atrial pacing



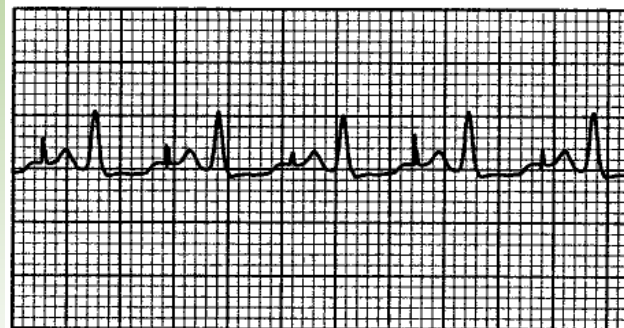
II



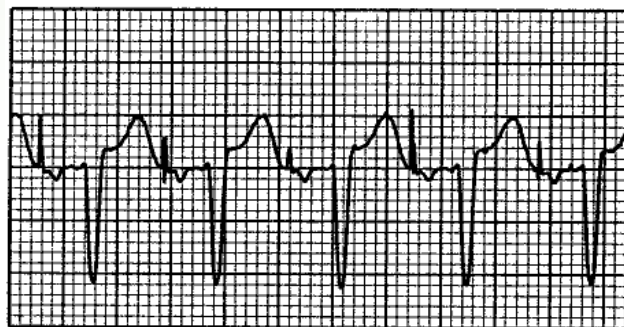
III



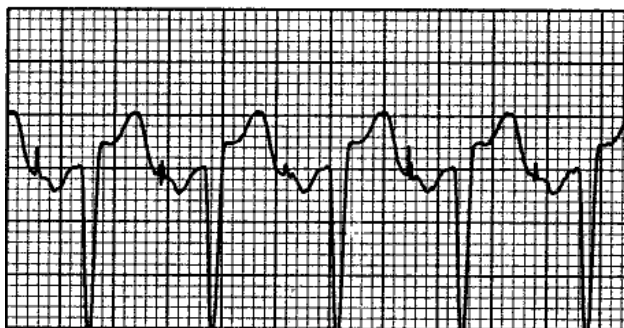
I Low atrial pacing



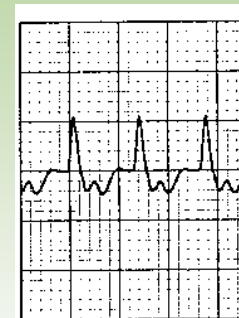
II



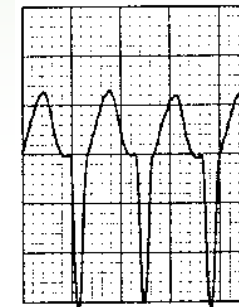
III



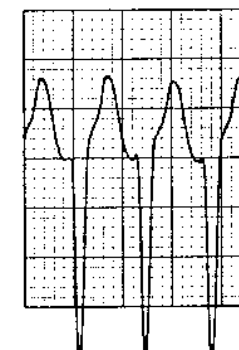
PSVT



II



III

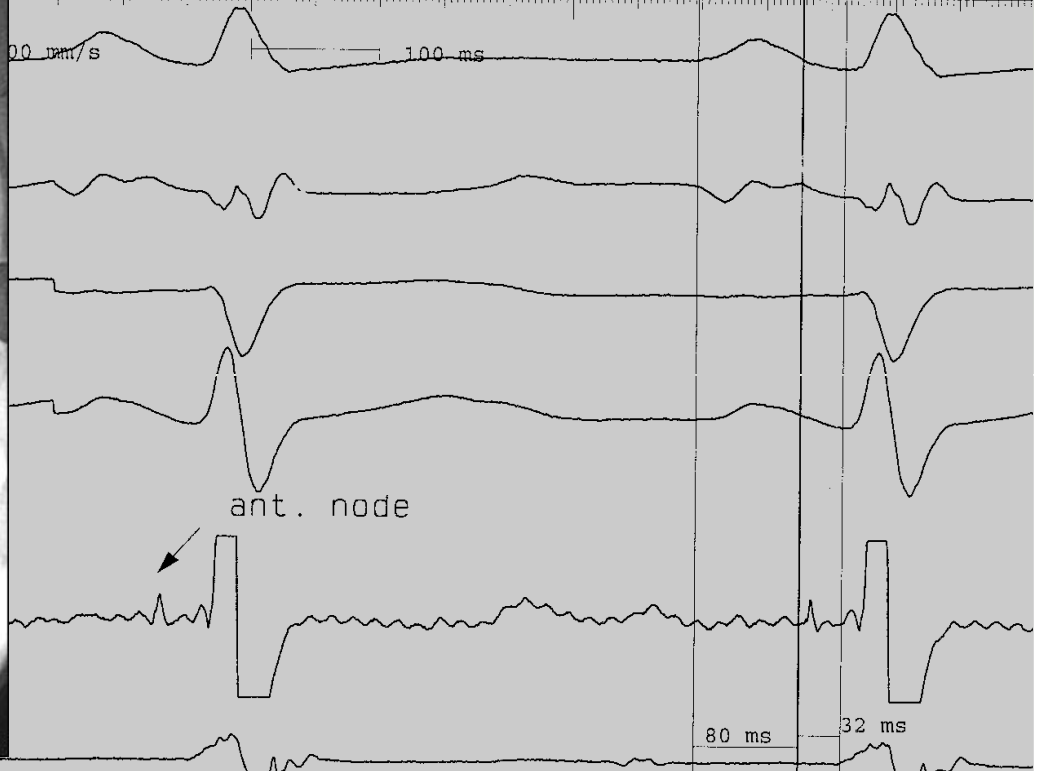


600 J00 HOE (M)
7335745-6
2001-07-26

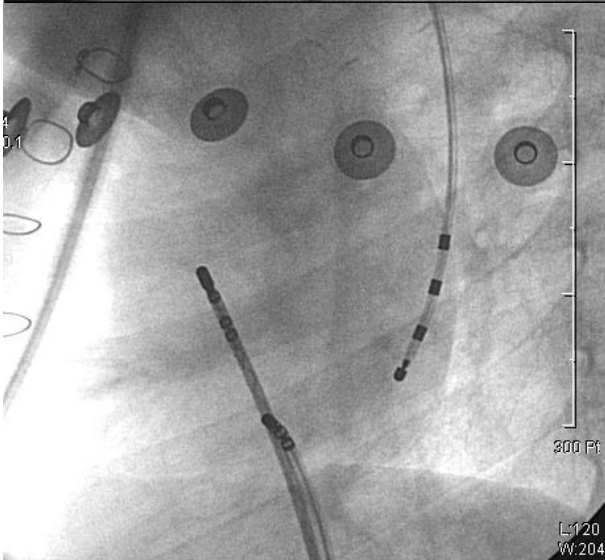
RAO 18.5
CRAN 15.7

Srs:1
Acq:9
Img:9

GENERAL RF Ablation [4+ cath] 4.1!



L:120
VV:204

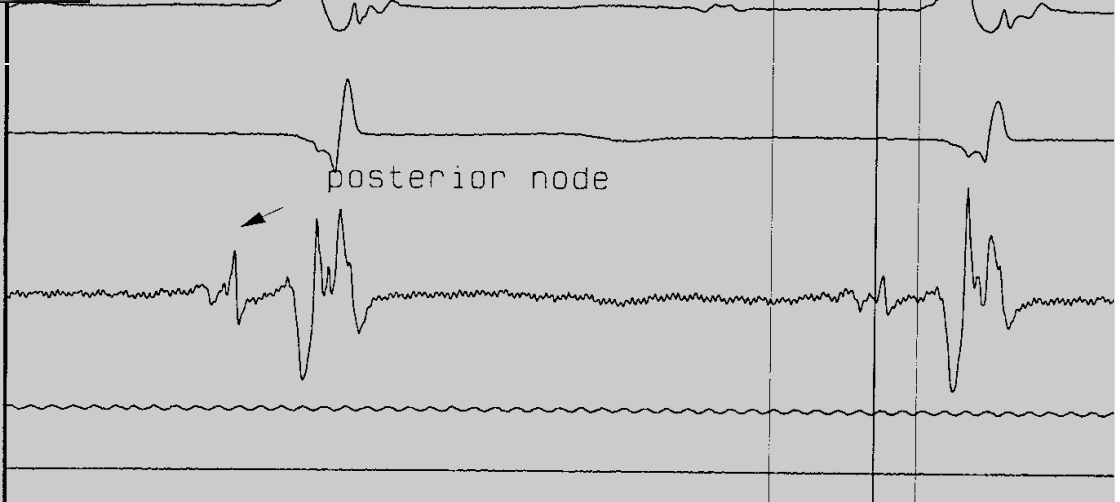


23-ABL ds

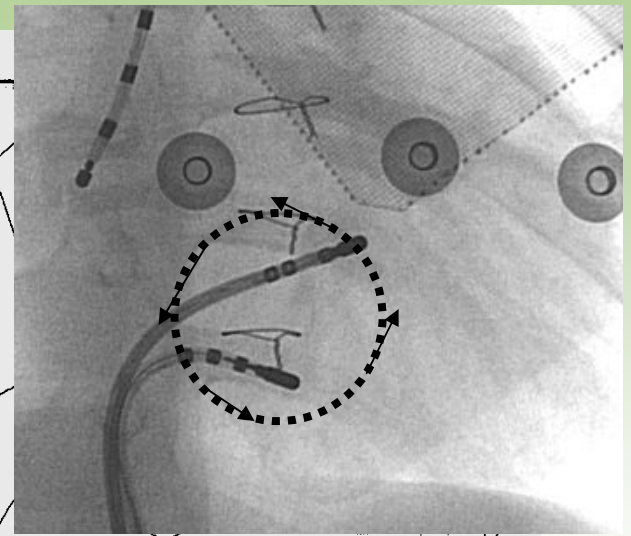
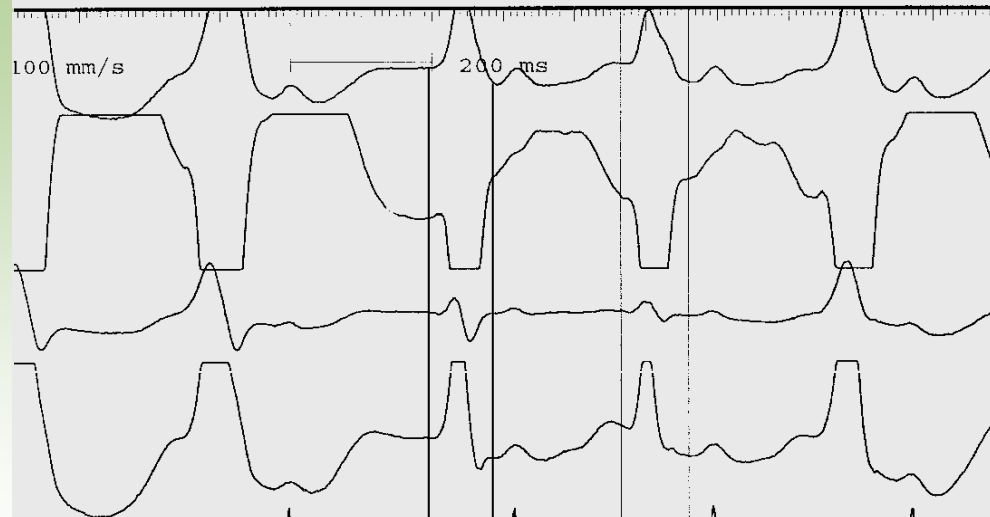
24-ABL

26-RVa 3 4

28-STIM A1



GENERAL RF Ablation [4+ cath] 4.1!



High atrium

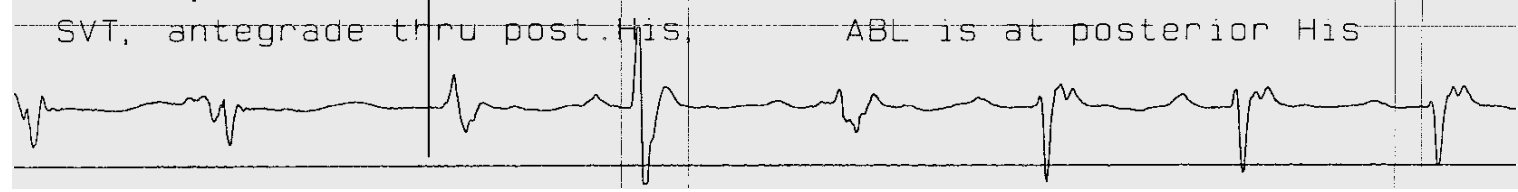


Low septum



SVT, antegrade thru post.His.

ABL is at posterior His

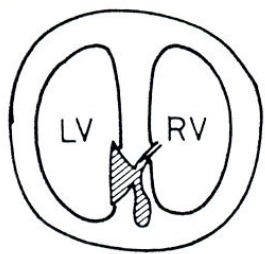


The cardiac conduction system in situs ambiguus

(Dickinson et al. circulation 1979;59:879)

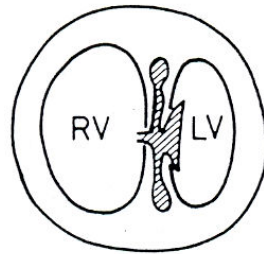
Mönkeberg's sling

13 cases of situs ambiguus



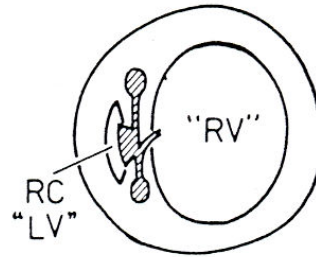
3 cases

Left Isomerism

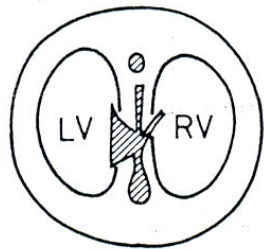


5 cases

L-loop

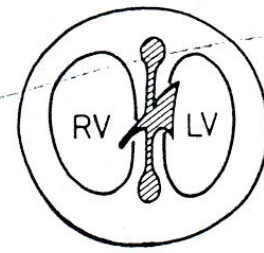


1 case

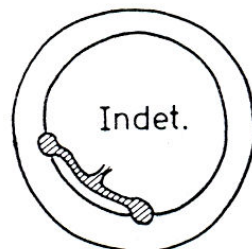


1 case

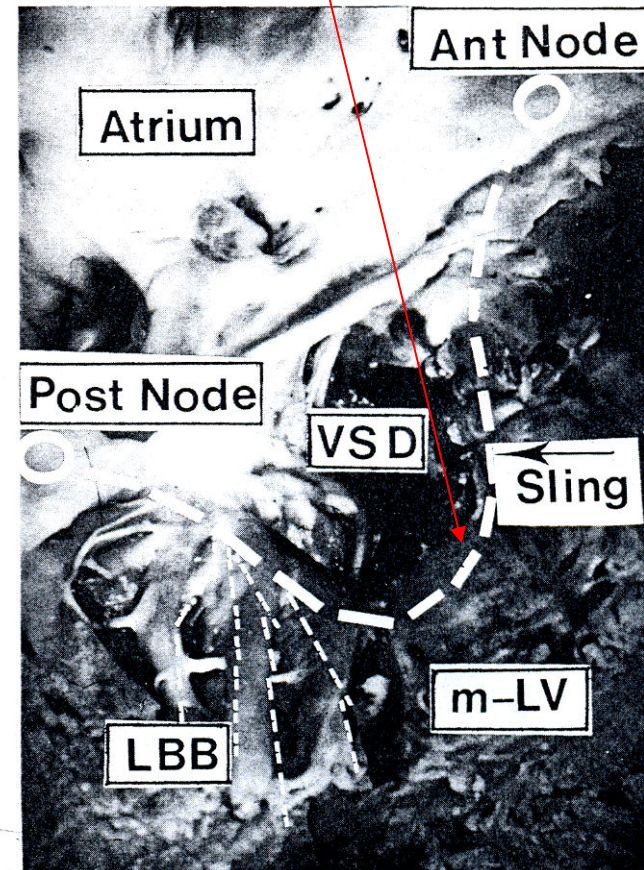
Right Isomerism



1 case

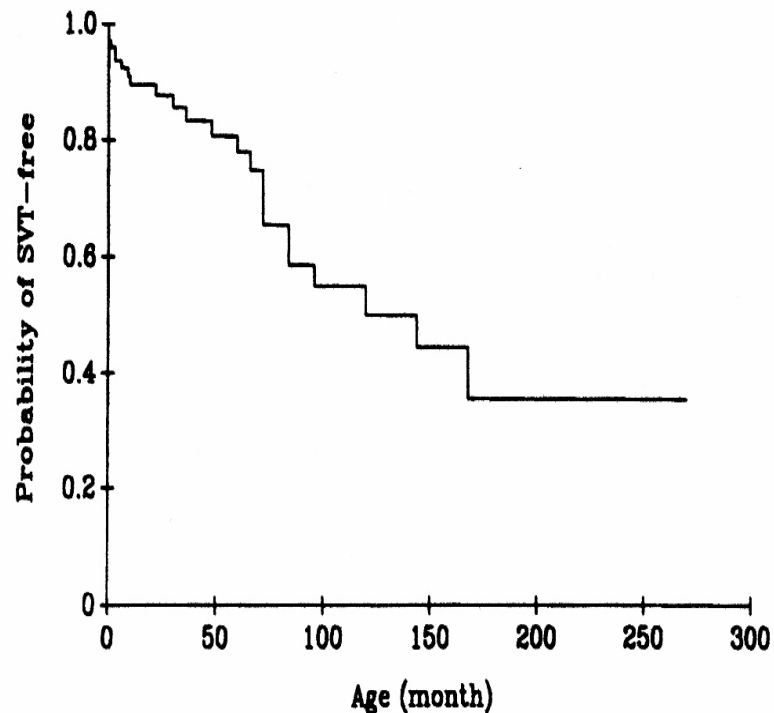


2 cases



SVT in right isomerism

- Very high incidence (24.8%) of SVT in right isomerism (Wu MH et al 1998, JACC)



1987–1996

Total 101 right isomerism patients
(Fontan survivors 7/19)

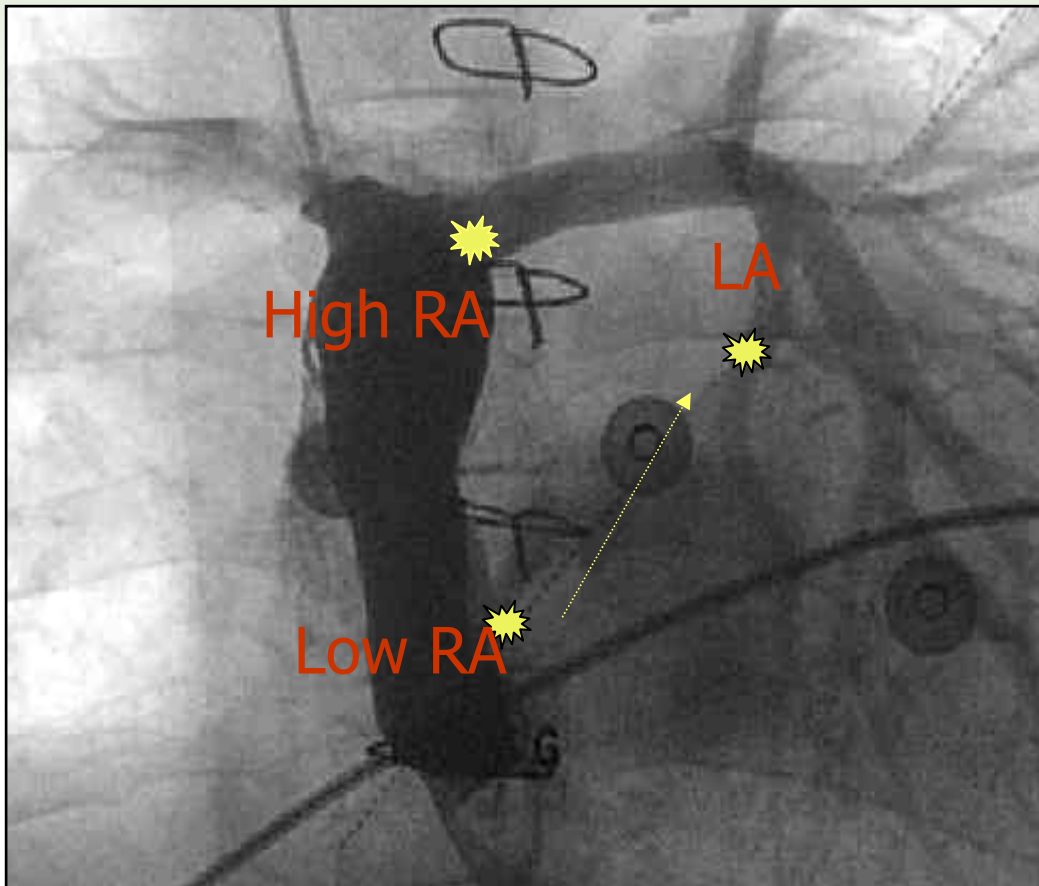
Median FU ; 38mo

Documented SVT(not AFL); 24.8 %
7 EPS; twin node tachycardia
Unknown exact mechanism of SVT

Actuarial curve for the probability
of being free from SVT

Twin AV Node and Induced Supraventricular Tachycardia in Fontan Palliation Patients (Bae et al , PACE 2005)

3 or more different atrial sites pacing



EPS on 52 patients
s/p Fontan op.
(Jan 2001–Oct 2003)

high RA and low RA directly
high LA and low LA
via fenestration, CS
or esophagus

Changes of QRS complexes as an evidence of a twin AV node

- Multisite atrial pacing

QRS change; 10/52 (19%)

Right isomerism; 9/20(45%)

AV discordance ; 1/8(13%)

**Other functionally univentricular heart ;
0/22(0%)**



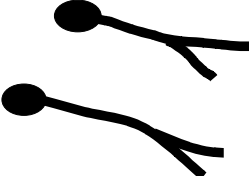

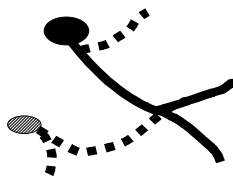
- Risk factor for twin AV node

- **heterotaxy syndrome(p<0.001)**

- **complex AVSD (p=0.007)**

(Bae et al , PACE 2005)

Categorization of AV node in complex univentricular heart

	2QRS paced	2QRS ECG	Two HBE	Presented in this study	HBE-retro, preceding V during AVRT	AVRT	JT with AV dissociation
	-	-	-	+	-	-	possible
	+	+	+	+	+	possible	possible
	+	+	+	+	-	possible	possible
	-	-	+	-	+	possible	possible
	+	±	±	+	+	possible	possible
Mahaim-like							

(Bae et al , PACE 2005)

Arrhythmia in post CHD surgery

Careful FU is needed

Surgical scar –related

RA dilatation ;

- s/p Fontan, valve or ventricular dysfunction
- atrial flutter (intraatrial reentry)
- non-reentrant atrial tachycardia

Preventive

Modification of surgery;

less scar, less corridor, less distension,
sinus node preservation.

Pacemaker implantation ; control bradycardia

Therapeutic

- Antiarrhythmic medication
- Catheter ablation with considerable effect
- Surgical correction;
arrhythmia surgery, Fontan conversion

Heterotaxy syndrome

- High incidence of bradyarrhythmia and tachyarrhythmia
- More important in Asian population
- Associated with
inherent anomaly of SA node,
AV node and conduction system

Twin AV node may be an important cause of tachyarrhythmia
– For SVT in Fontan patients,
we need to think of basic anomaly again.