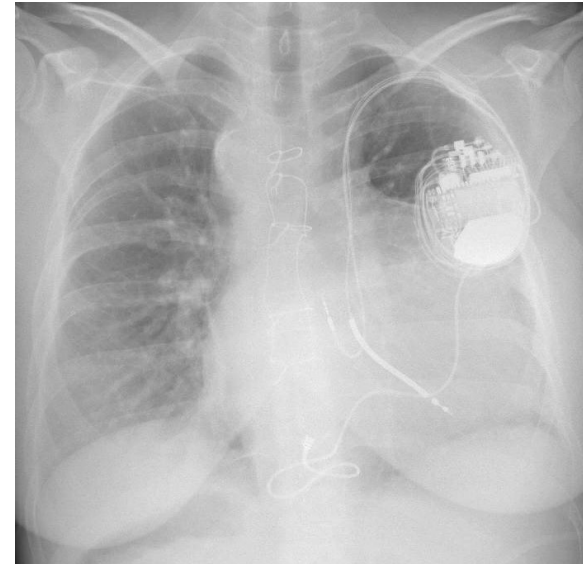
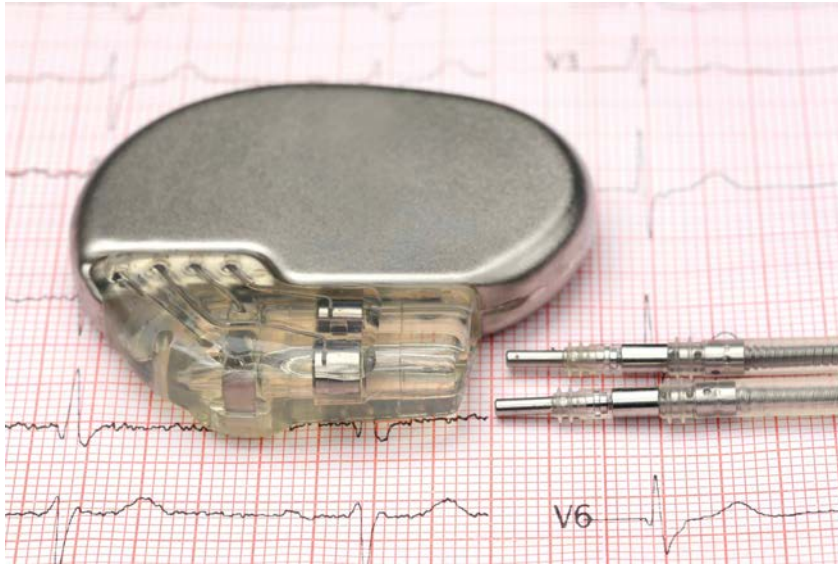


# CARDIAC PACING IN PEDIATRIC AND CONGENITAL PATIENTS

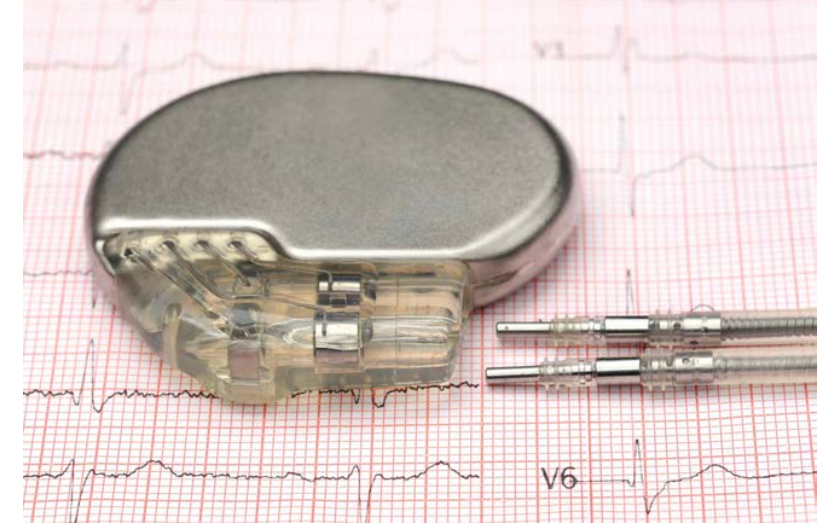


Marie WILKIN / Victor WALDMANN

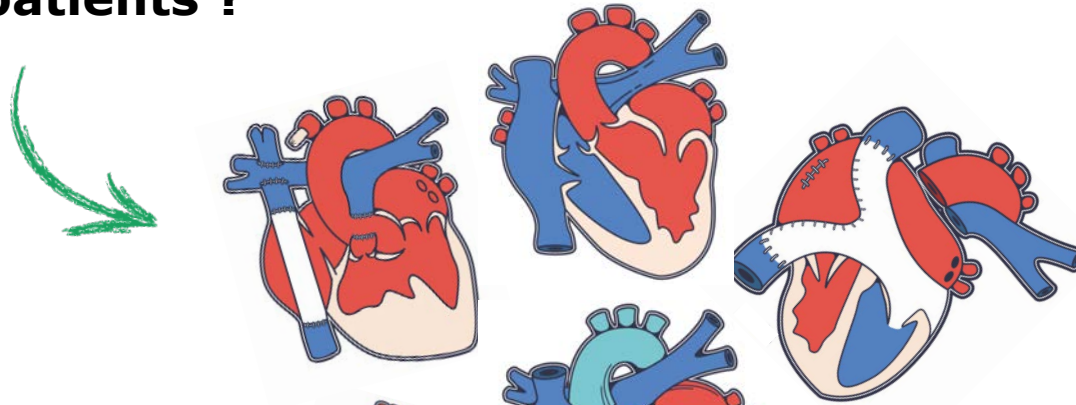
*Hôpital Européen Georges Pompidou — Hôpital Necker*



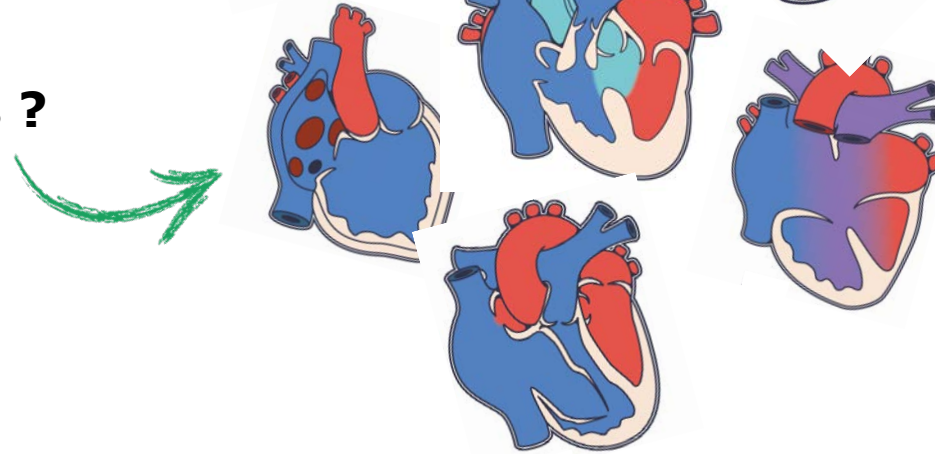
# PACEMAKER IN PEDATRIC & CHD PATIENTS



**Which patients ?**



**Indications ?**



**Complications ?**

**How ?**



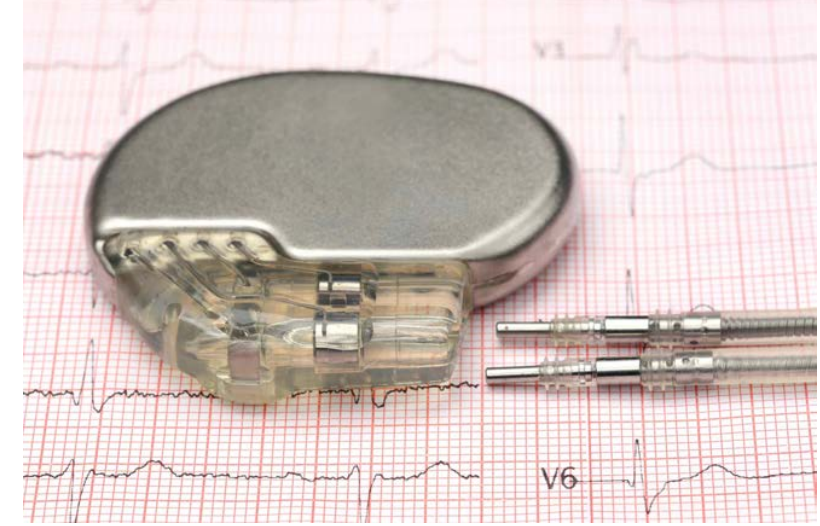
**Cardiac Resynchronization ?**



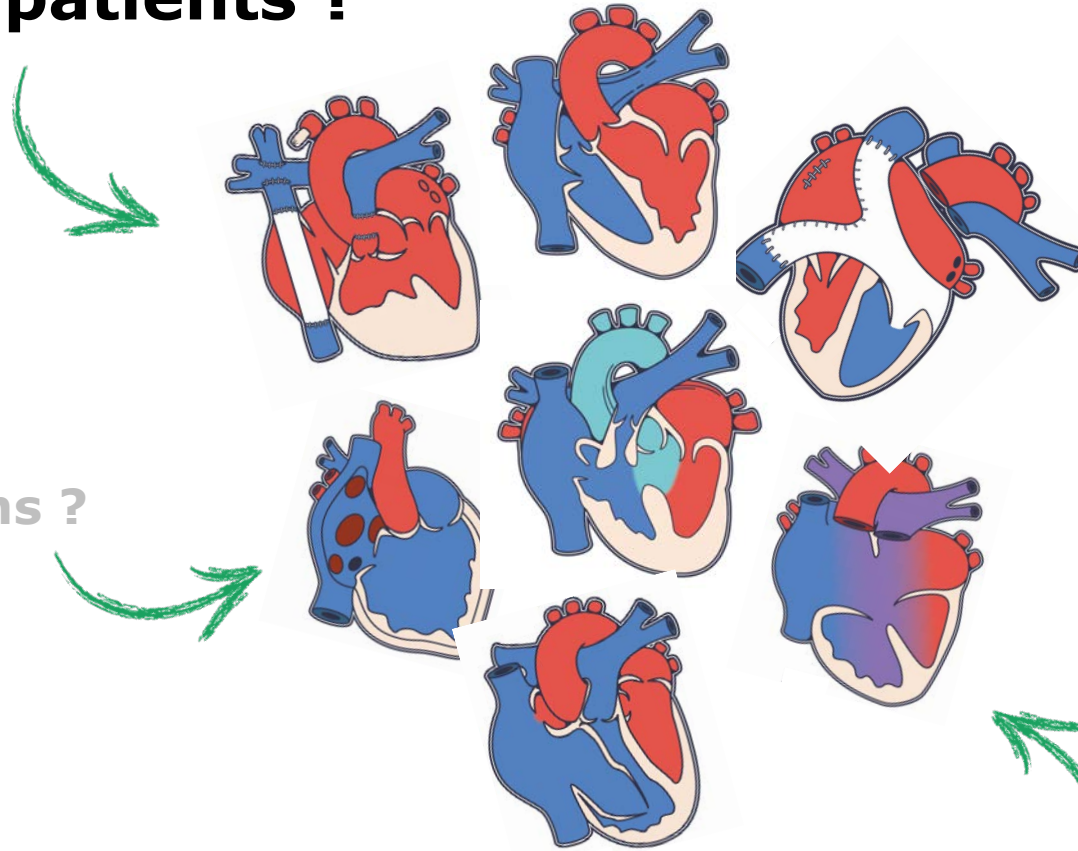


# PACEMAKER IN PEDATRIC & CHD PATIENTS

Which patients ?



Indications ?

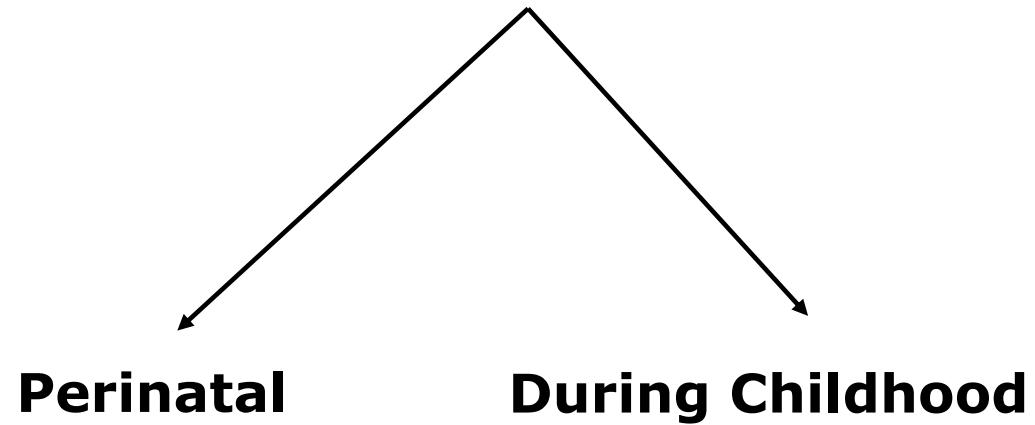


Complications ?

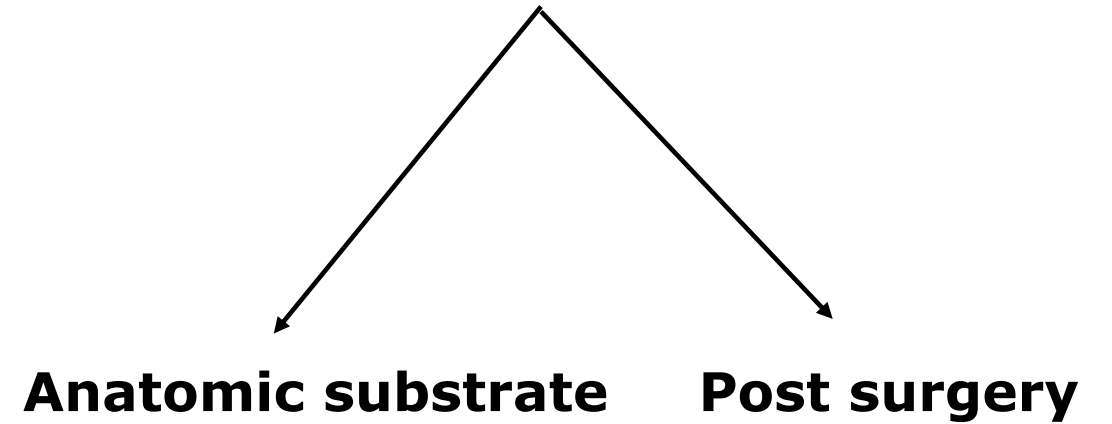
How ?

Cardiac Resynchronization ?

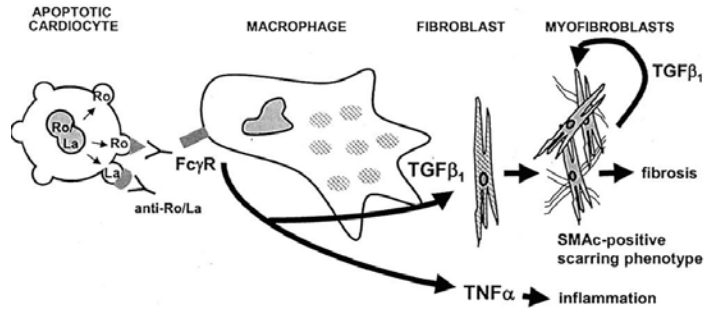
## Structurally Normal Heart



## Congenital Heart Disease



## Autoimmune congenital AV block



- Ac Anti SSA/SSB
- 1/10 000 births
- Mortality 15-20%
- AV block risk = 2-3%
- Recurrence = 20%

## Progressive AV Block

*Progressive Cardiac Conduction Disease (PCCD)*

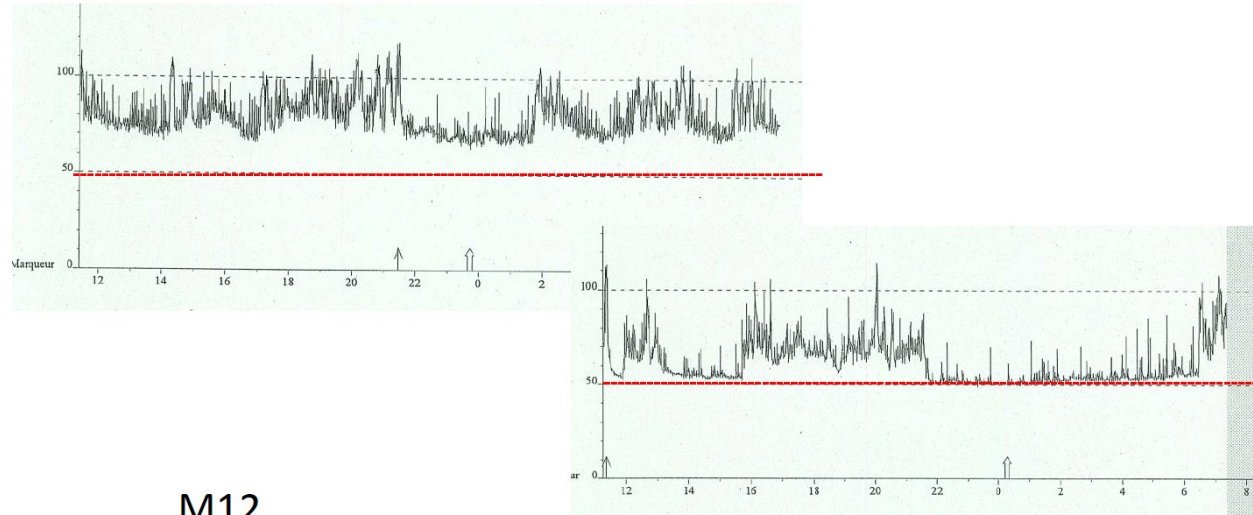
- Unknown cause ++
- SCN5A, SCN1B
- NKX2.5 (+ ASD)

## Other causes

- LQTS
- Isolated sinus node dysfunction
- Infectious/toxic causes
- Myopathies and cardiomyopathies

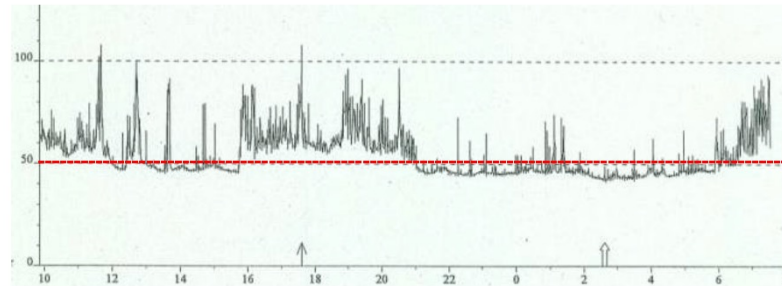
# AUTOIMMUNE AV BLOCK

Naissance

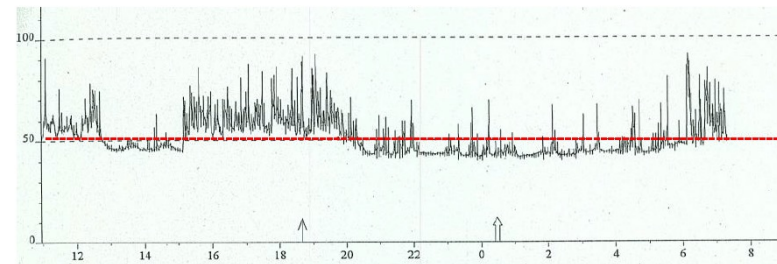


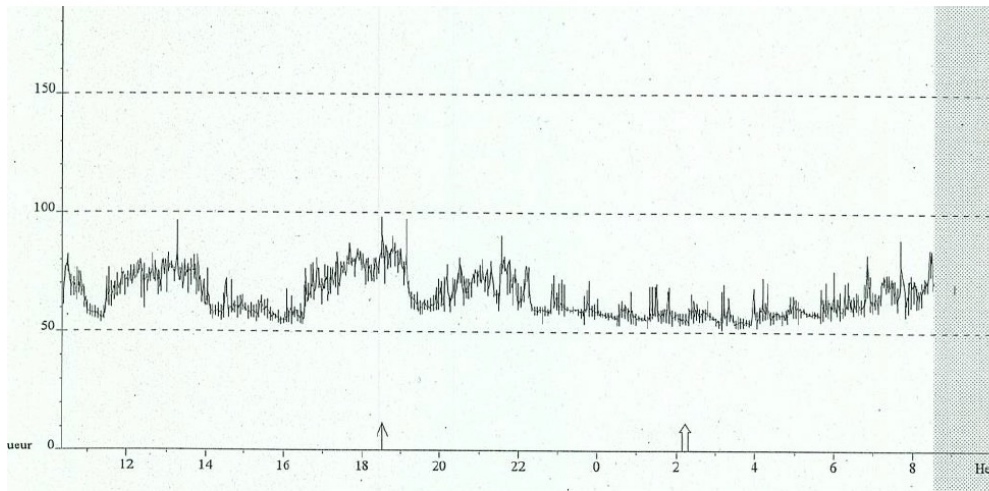
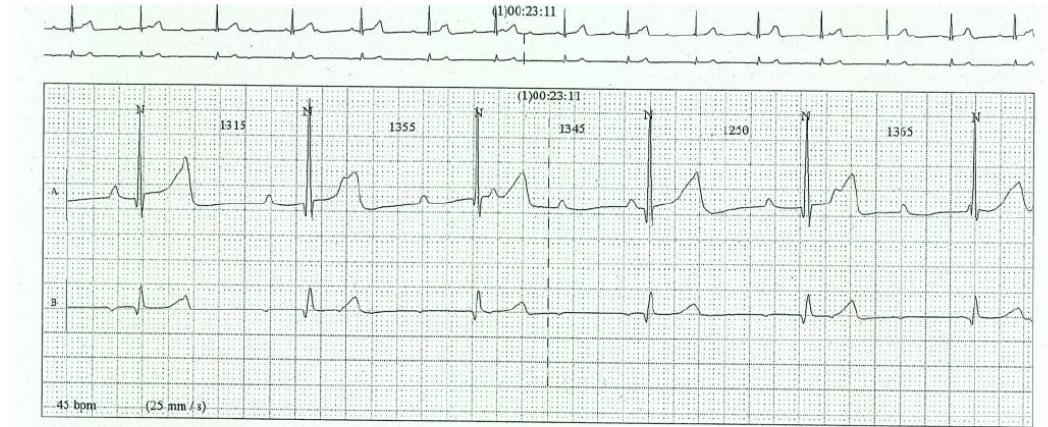
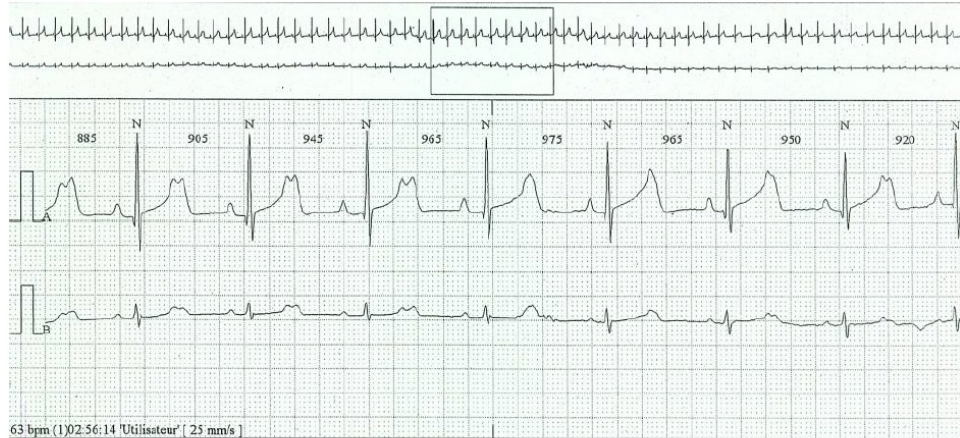
M6

M12

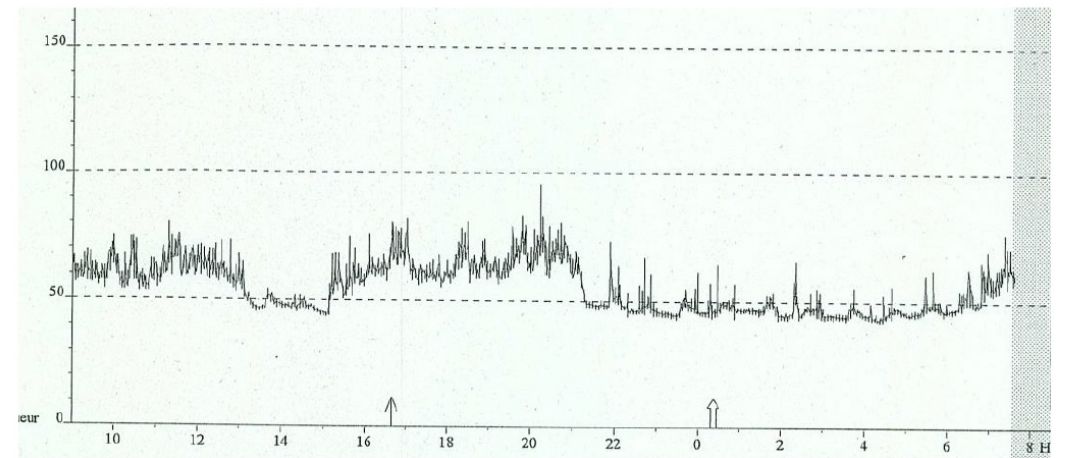


M18



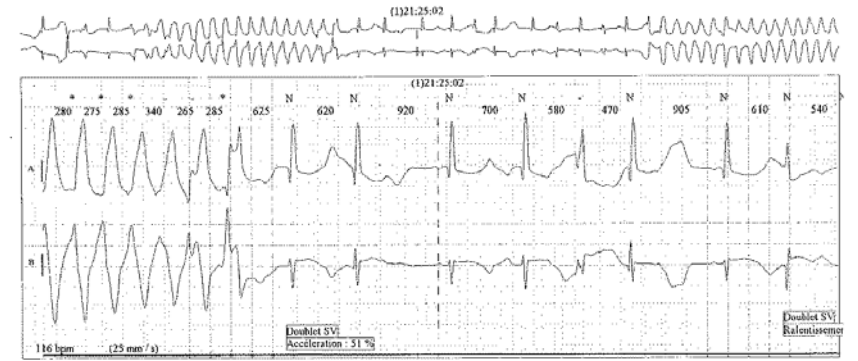
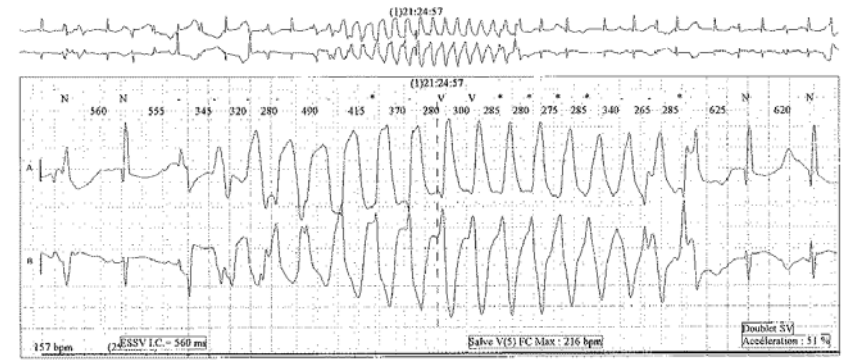
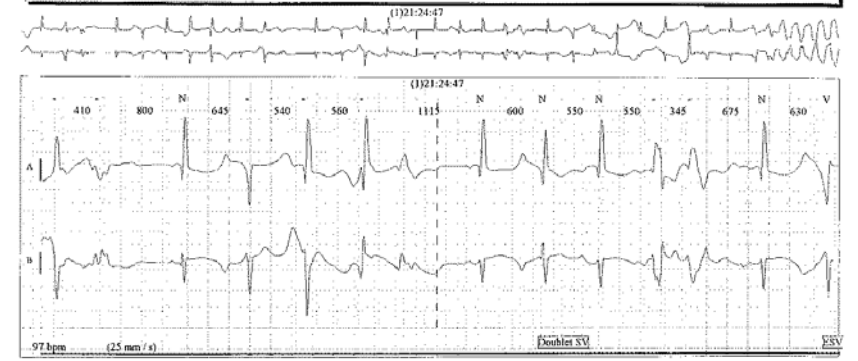
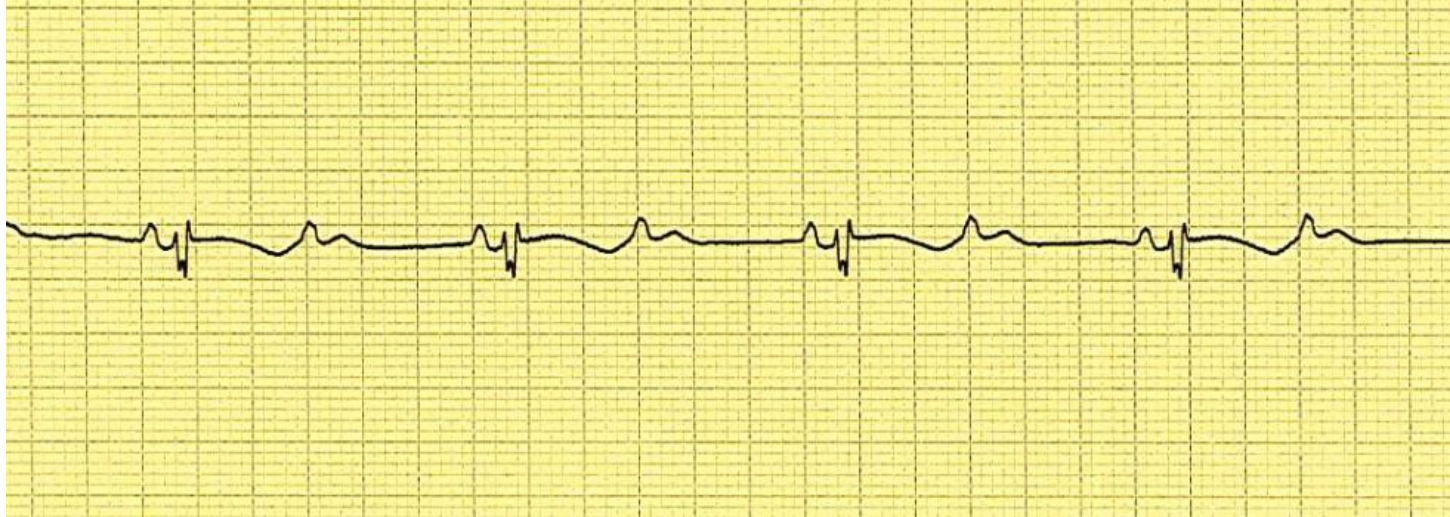


Décembre 2010

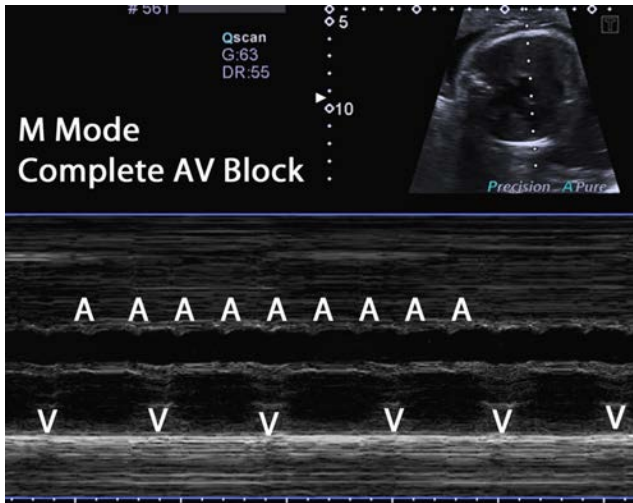


Mai 2011

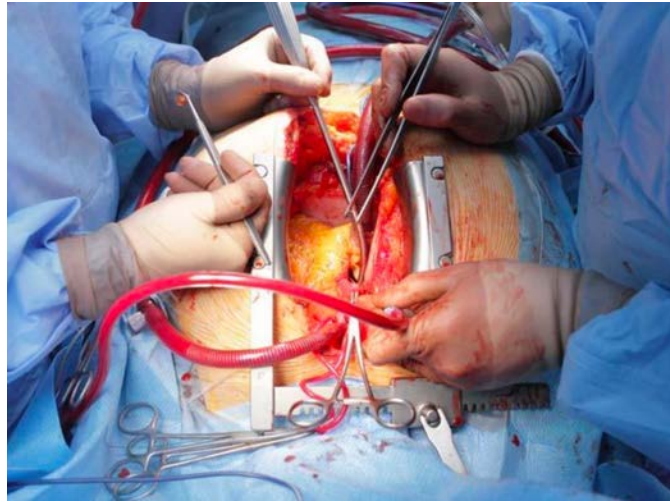
# LQTS



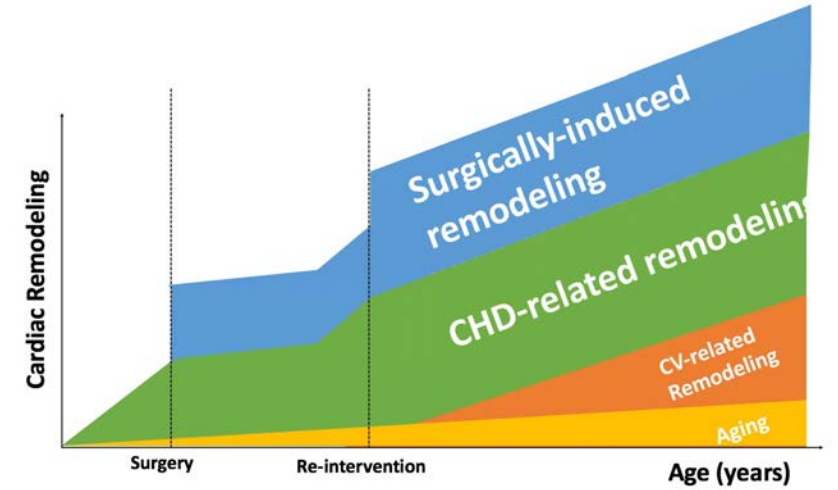




**Anatomic substrate**



**Surgical scarring**



**Progressive remodeling**

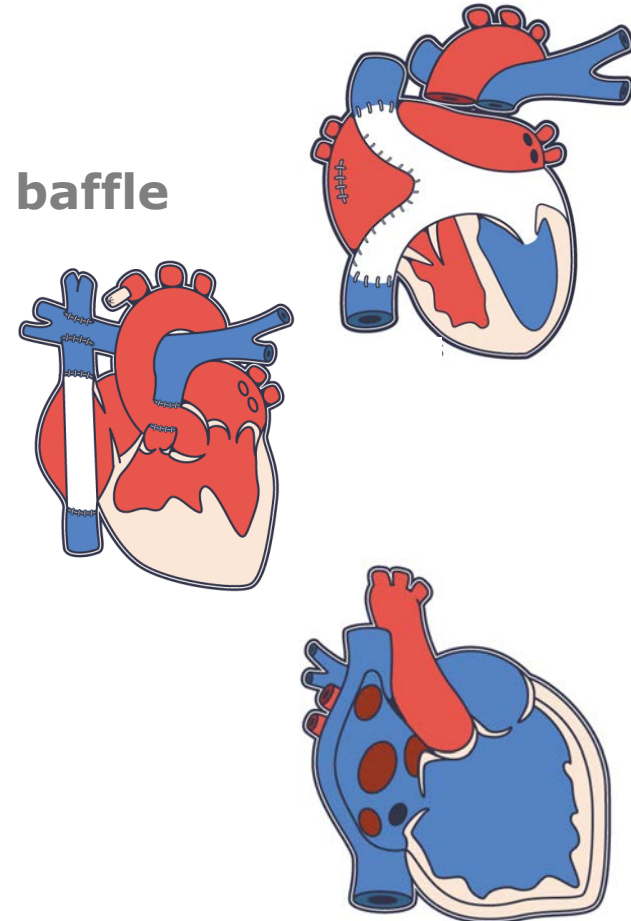
# SINUS NODE DYSFUNCTION

## Anatomic substrate

- Left-sided juxtaposition of the appendages
- Left atrial isomerism

## Surgical scarring

- Mustard/Senning baffle
- Glenn / Fontan
- Sinus venosus





# ATRIOVENTRICULAR BLOCK



## **Anatomic substrate**

- **cc-Transposition of the Great Arteries**
- **Atrioventricular septal defect**
- **Heterotaxy syndrome, single ventricles**

## **Surgical scarring**

- **Ventricular septal defect**
- **Atrioventricular septal defect**
- **cc-Transposition of the Great Arteries**
- **Tetralogy of Fallot**
- **Left ventricular outflow tract surgery**
- **Mitral or tricuspid valve replacement**

# POST SURGERY ATRIOVENTRICULAR BLOCK

**TABLE 1. Incidence of heart block with pacemaker placement by cardiac operation**

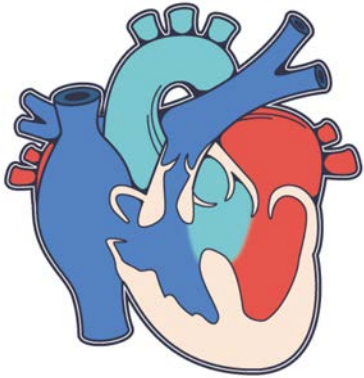
Surgery	Patients (n)	Pacemaker (n)	%	95% CI
Double switch operation	77	12	15.6	7.5-23.7
Tricuspid valve replacement	230	18	7.8	4.3-11.3
Mitral valve replacement	902	67	7.4	5.7-9.1
Atrial switch with VSD	217	14	6.5	3.1-9.7
Rastelli operation	313	15	4.8	2.4-7.1
Ebstein's repair <30 d	72	3	4.2	0-8.8
Konno operation	290	12	4.1	1.8-6.4
ASO with VSD	1517	52	3.4	2.5-4.3
Aortic valve replacement	1262	37	2.9	2.0-3.9
Sub-AS resection	1078	30	2.8	1.8-3.8
Ross operation	918	22	2.4	1.4-3.4
CAVC repair	6358	143	2.2	1.9-2.6
Mitral valvuloplasty	3802	66	1.7	1.3-2.2
Coarctation with VSD	1107	19	1.7	1.0-2.5

<p><b>3) Postoperative AV block in congenital heart disease.</b> Permanent pacing is indicated for postoperative advanced second degree or complete AV block persisting &gt;10 days.</p>	I	B	137-141
<p><b>4) Postoperative AV block in congenital heart disease.</b> Permanent pacing should be considered for persistent, asymptomatic post-surgical bifascicular block (with or without PR prolongation) associated with transient, complete AV block.</p>	IIa	C	-
			<.001
			<.001
			<.001
			<.001
			.01

**USA 2004-2013  
100,000 patients**

**~ 1% pacemaker for AV block**

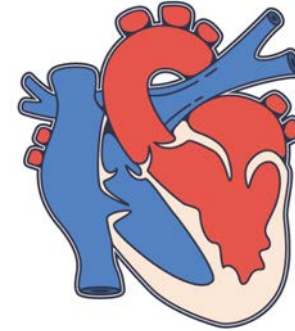
# PACEMAKER PREVALENCE IN CHD



**Fallot**

**~ 8%**

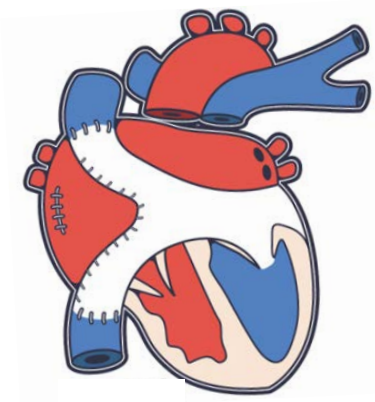
*Khairy P, et al. Circulation 2010*



**Cc-TGA**

**~ 10%**

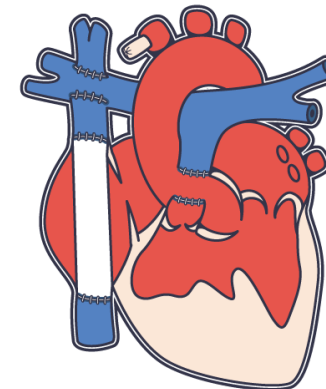
*Huhta JC M, et al. Circulation 1983*



**Mustard/Senning**

**~ 25%**

*Schwerzmann M, et al. EHJ 2009*



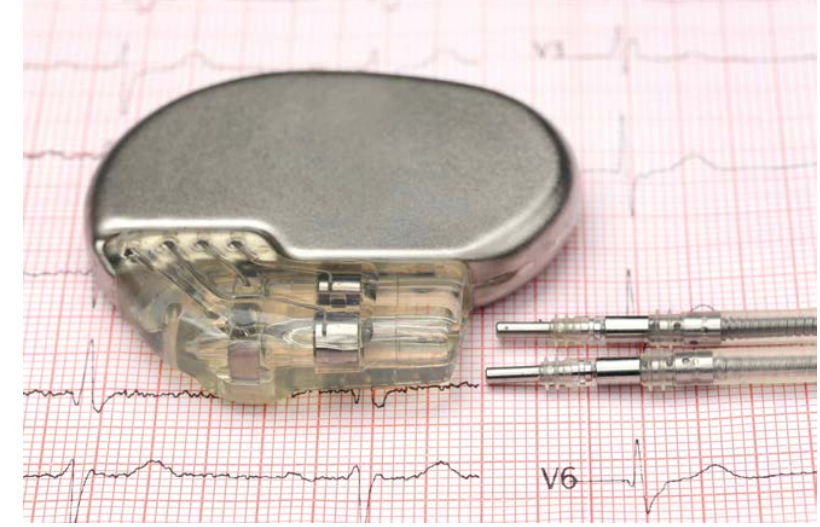
**Fontan**

**~ 20%**

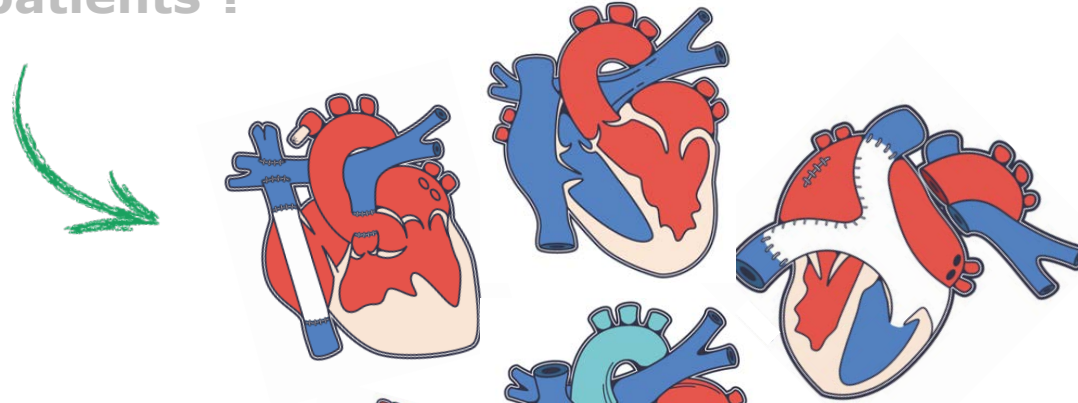
*Downing TE, et al. Circ Cardiovasc Int 2017*



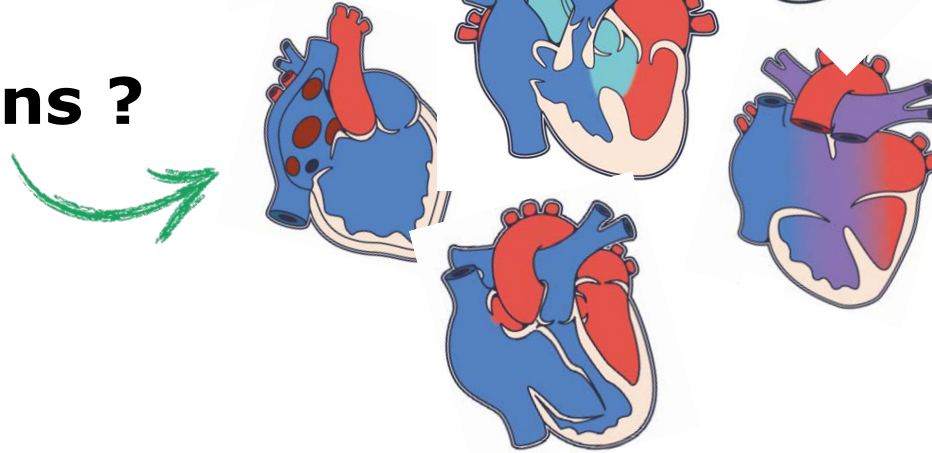
# PACEMAKER IN PEDATRIC & CHD PATIENTS



Which patients ?



**Indications ?**



Complications ?

How ?



Cardiac Resynchronization ?





# GUIDELINES



## PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease

*Developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD)*

*Khairy P, et al. Heart Rhythm 2014*



**EHRA POSITION PAPER**

**Arrhythmias in congenital heart disease: a position paper of the European Heart Rhythm Association (EHRA), Association for European Paediatric and Congenital Cardiology (AEPC), and the European Society of Cardiology (ESC) Working Group on Grown-up Congenital heart disease, endorsed by HRS, PACES, APHRS, and SOLAECE**

*Hernandez-Madrid A, et al. Europace 2018*



# GUIDELINES

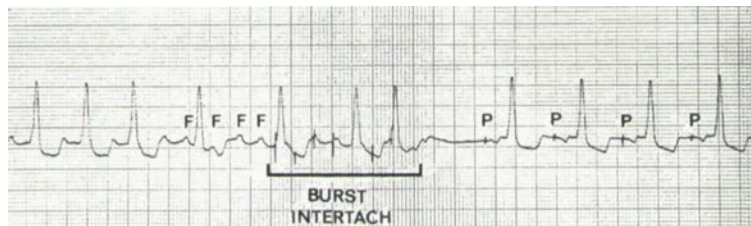
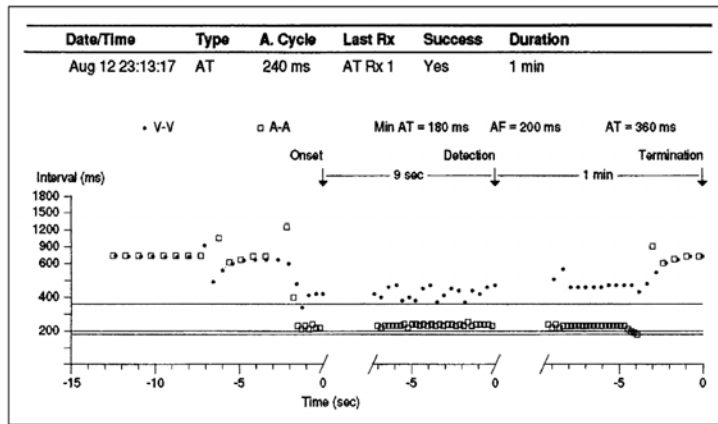


- **Symptoms ++**
- **Congenital AV block**
- **Prevention of atrial arrhythmias**
- **Hemodynamic perturbations**
- **Insufficient gain weight ?**
- **Wide QRS**
- **Ventricular arrhythmias**
- **Ventricular dysfunction**
- **Average daytime HR < 50 bpm**
- **Prolonged QT interval**

*Khairy P, et al. Heart Rhythm 2014  
Hernandez-Madrid A, et al. Europace 2018*



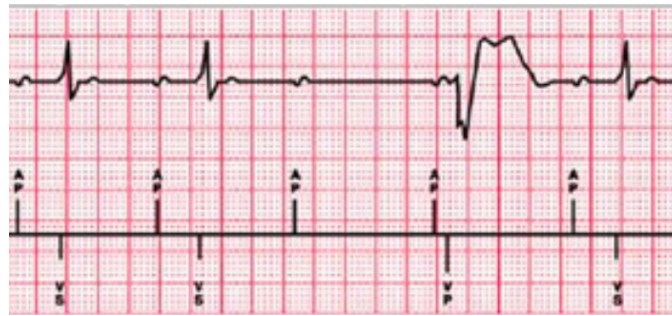
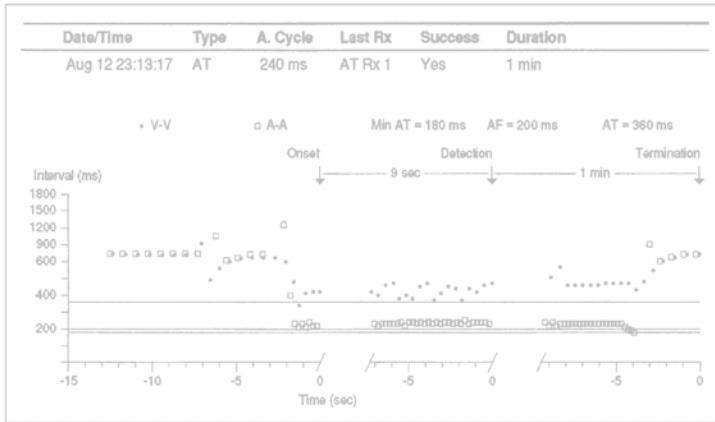
## Antitachycardia pacing



*Rhodes LA, et al. PACE 1995*  
*Stephenson EA, et al. Am J Cardiol 2003*

## Antitachycardia pacing

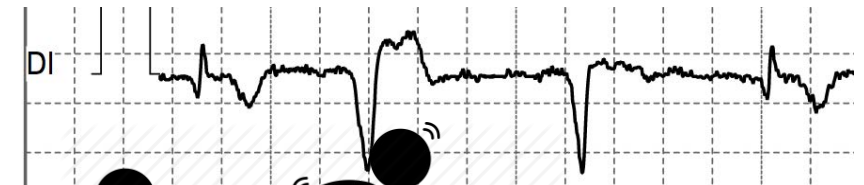
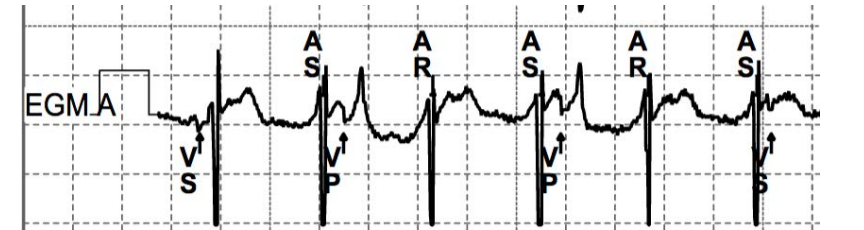
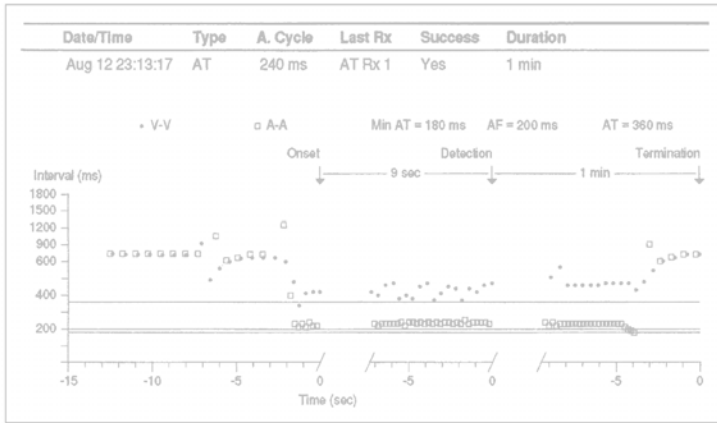
## Minimize pacing



## Antitachycardia pacing

## Minimize pacing

## Tailored settings



---

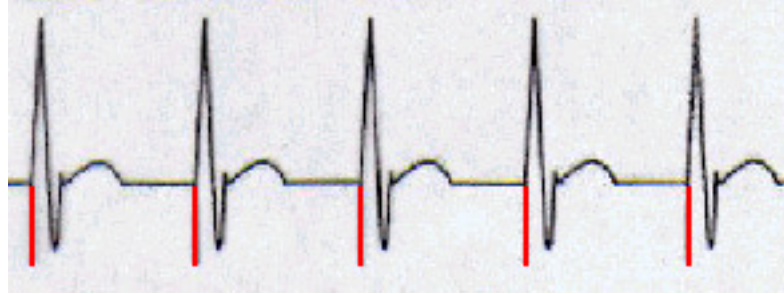
## Stimulateur cardiaque

<b>Lettre I</b>	<b>Lettre II</b>	<b>Lettre III</b>	<b>Lettre IV</b>
<b>Stimulation</b>	<b>Détection</b>	<b>Réponse à la détection</b>	<b>Fréquence</b>
O : Aucune	O : Aucune	O : Absente	O : Non Asservie
A : Auriculaire	A : Auriculaire	A : Inhibée	R : Asservie
V : Ventriculaire	V : Ventriculaire	V : Déclenchée	
D : Double (A+V)	D : Double (A+V)	D : Double (I+T)	

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# PACEMAKER — PROGRAMMATION

**VVI**

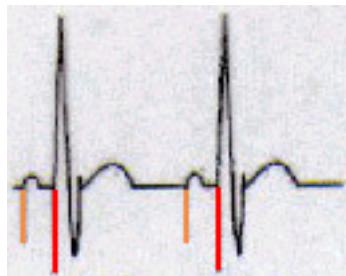


**AF**



**AV dissociation**

**DDD**



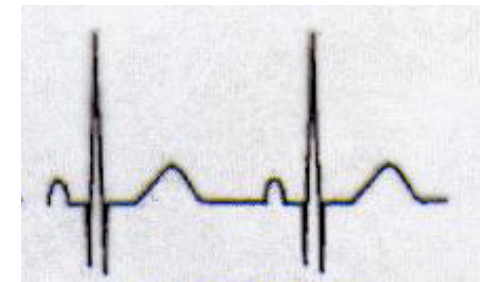
**A-V pacing**



**V pacing**



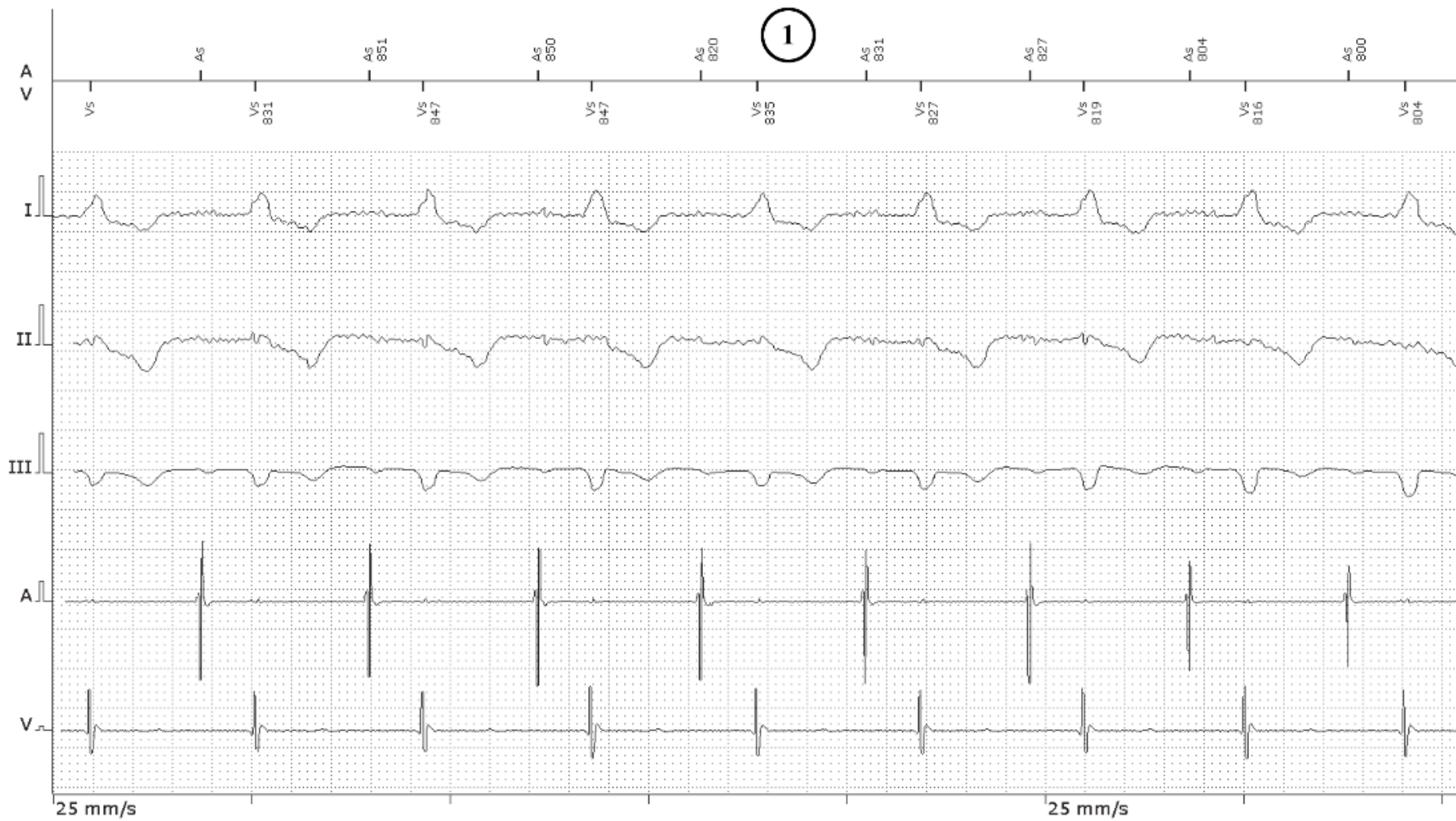
**A pacing**



**Inhibition A-V**

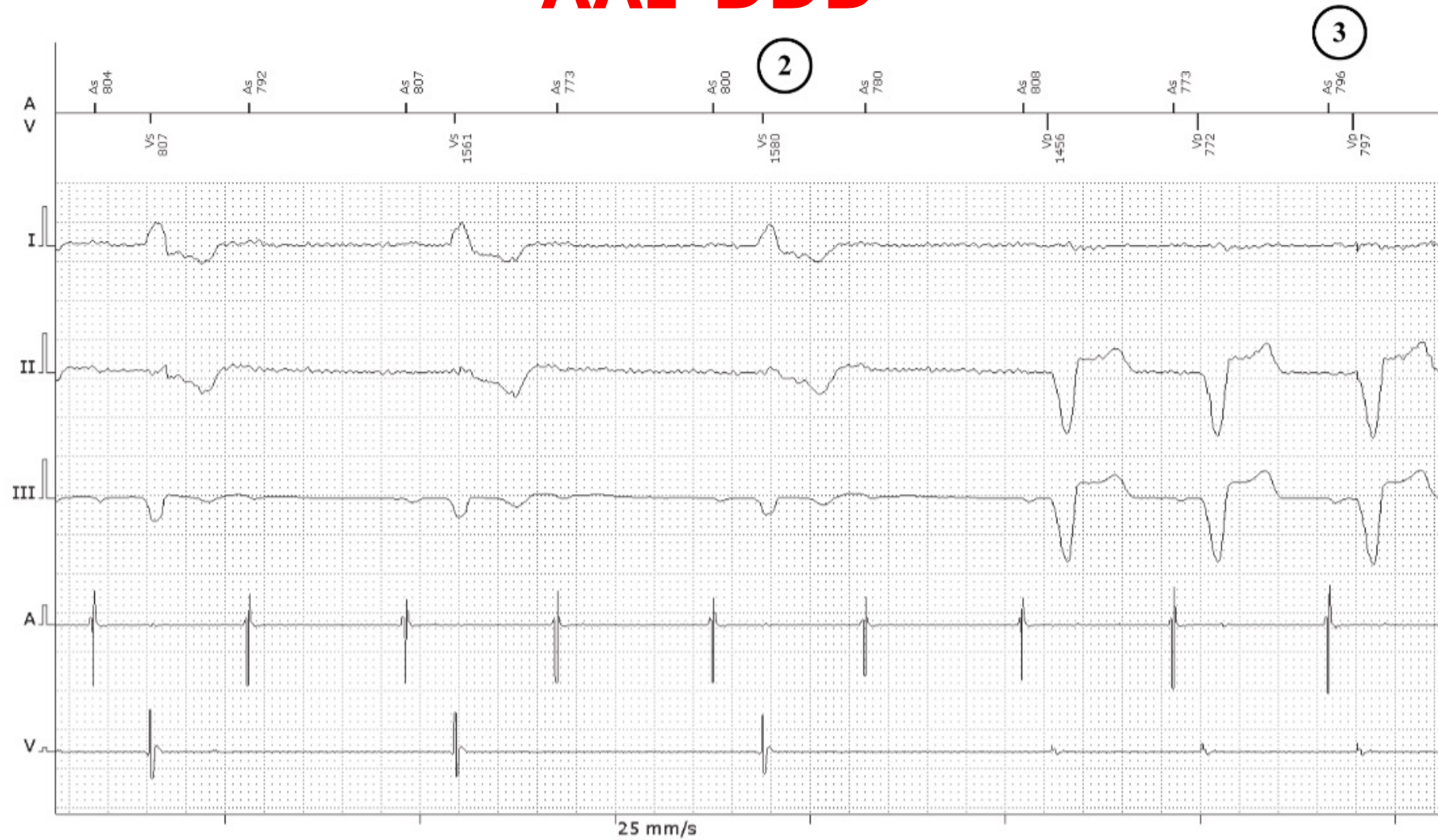
# PACEMAKER — PROGRAMMATION

## AAI-DDD



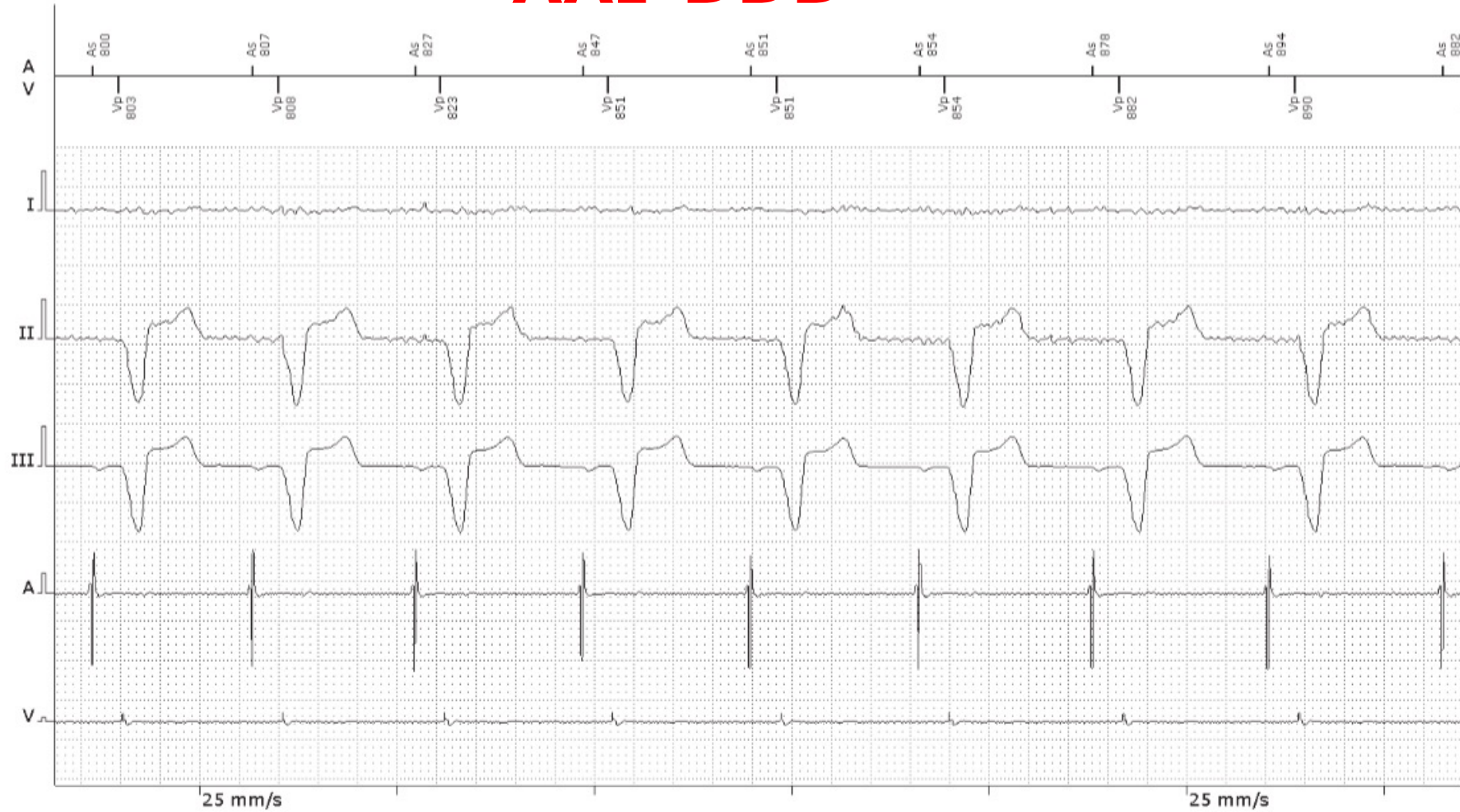
# PACEMAKER — PROGRAMMATION

## AAI-DDD



# PACEMAKER — PROGRAMMATION

## AAI-DDD





**VVI**

*1 lead*

**DDD / VDD**

*2 leads*

**AAI-DDD**

*2 leads*

**New born = VVI 120 bpm**

**Walking = VVIR**

**Back-up if paroxysmal (VVI 40...)**

**Permanent AV block**

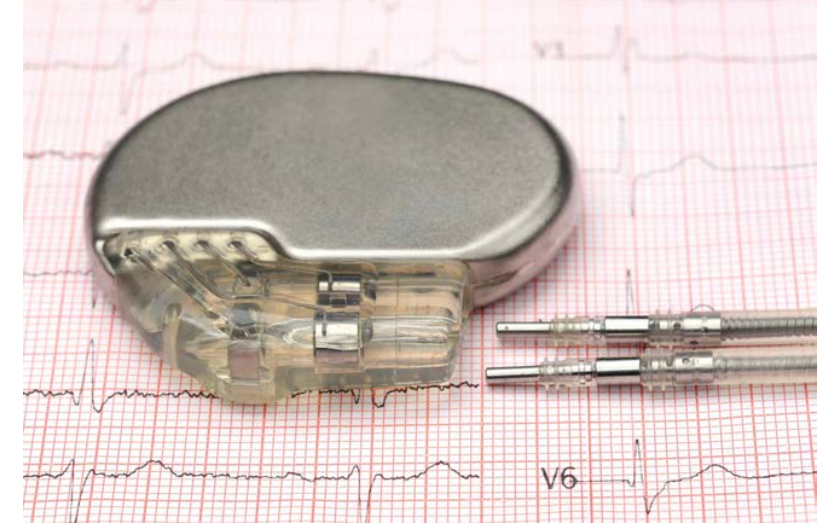
**DDDR** if sinus dysfunction associated

**Sinus node dysfunction**

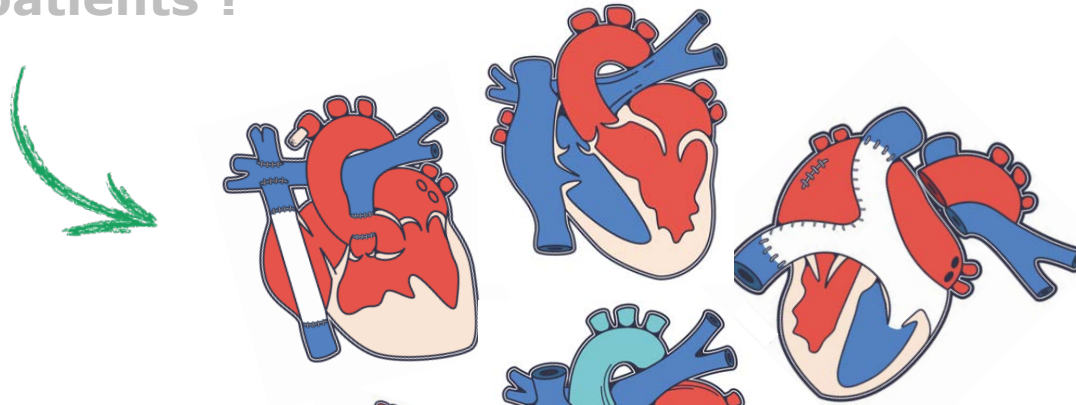
**Paroxysmal AV block**



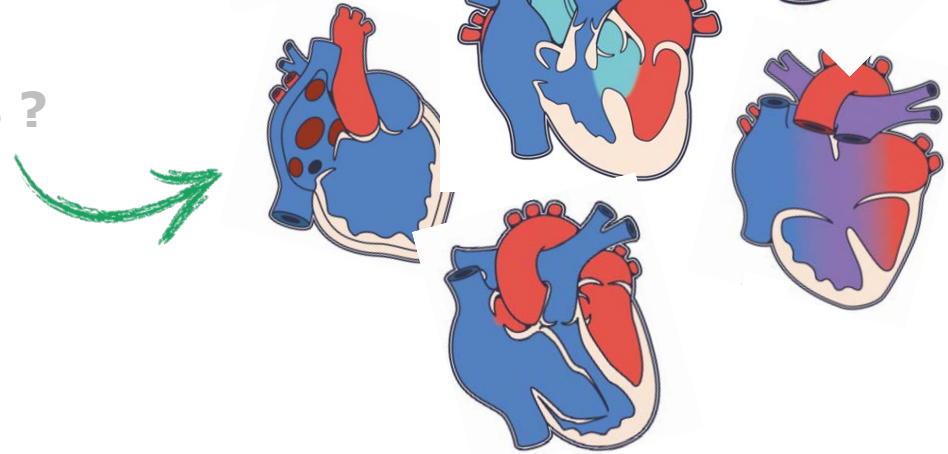
# PACEMAKER IN PEDATRIC & CHD PATIENTS



Which patients ?



Indications ?



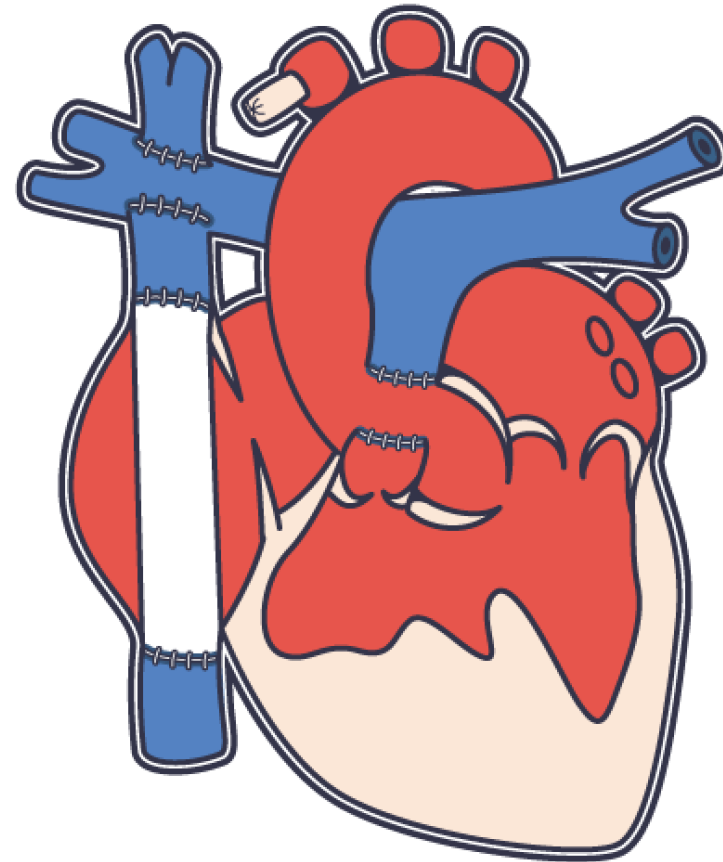
Complications ?

How ?

Cardiac Resynchronization ?

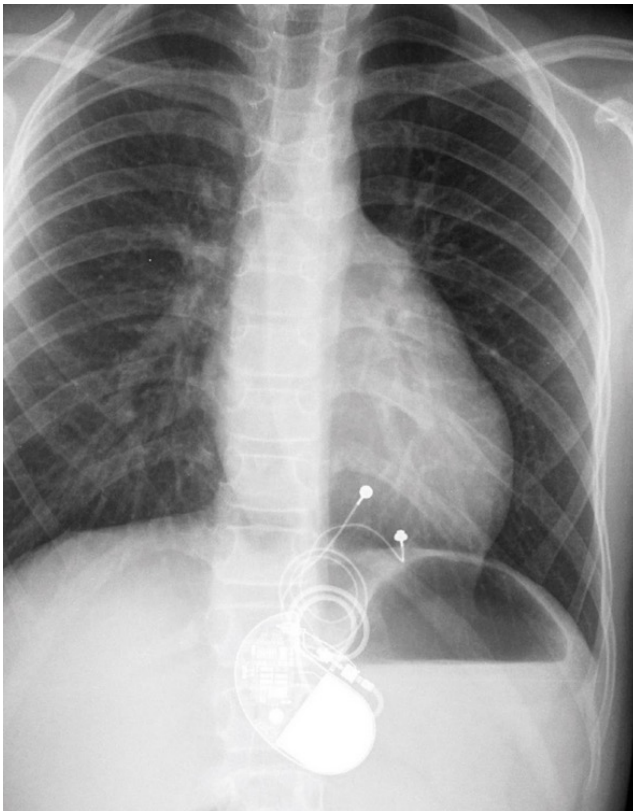
# PACE IN CHD: CHALLENGES

## Vascular access



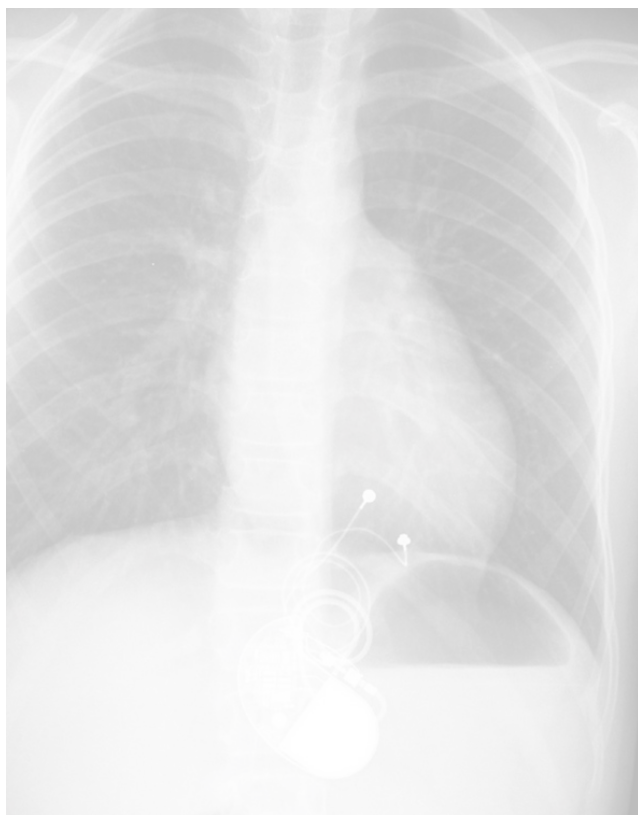
## Vascular access

### Epicardial leads

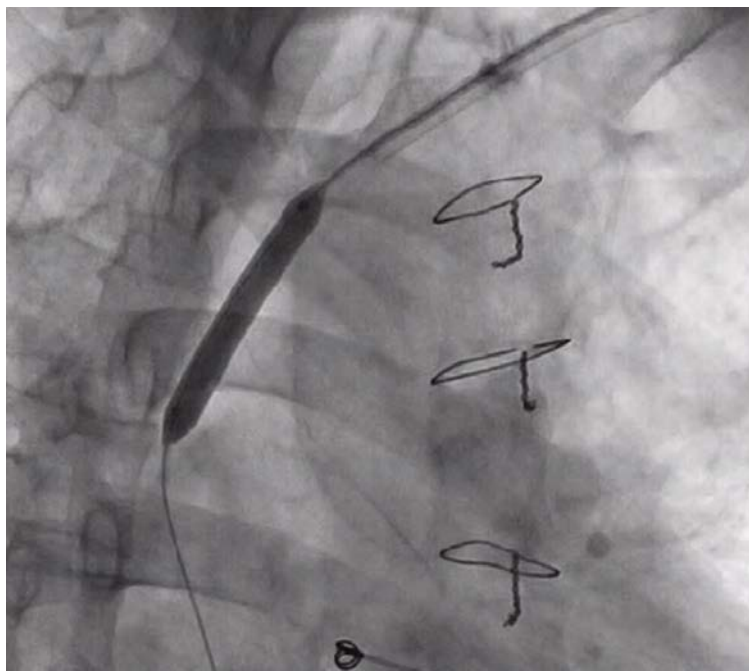


## Vascular access

### Epicardial leads

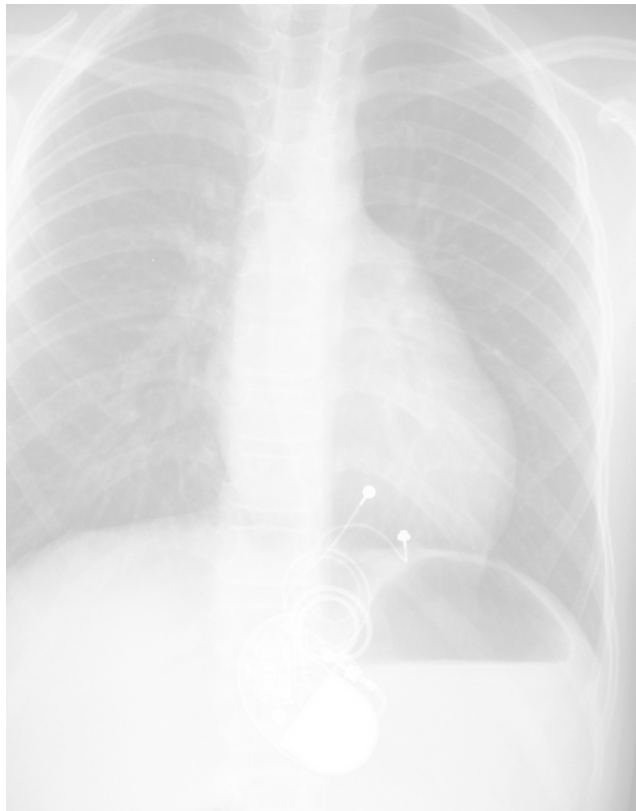


### Angioplasty

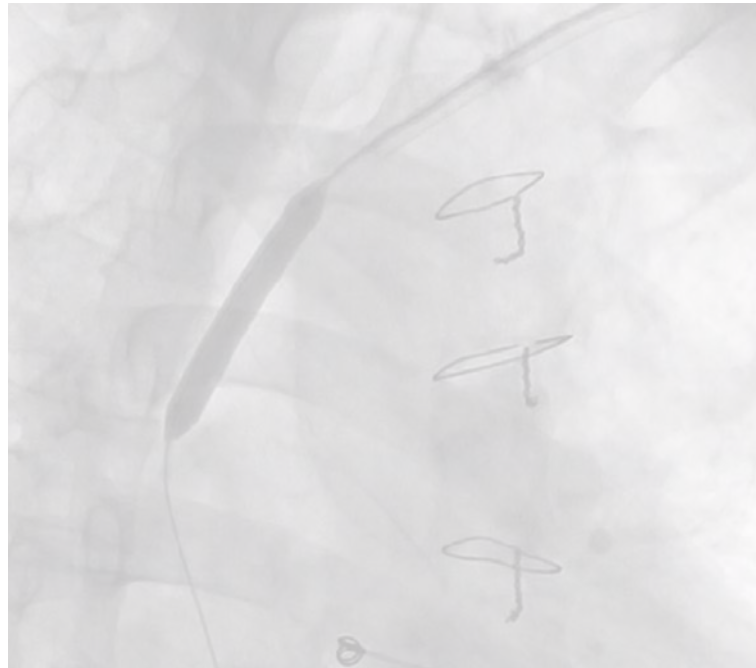


## Vascular access

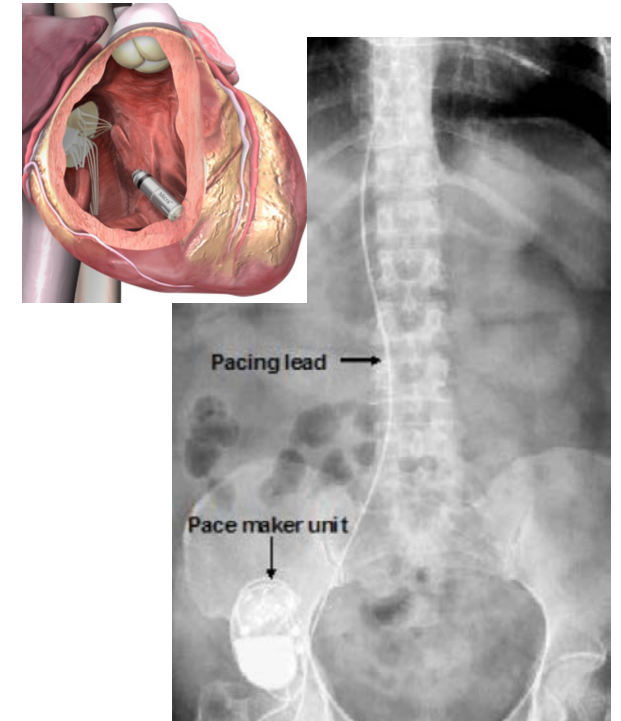
### Epicardial leads



### Angioplasty

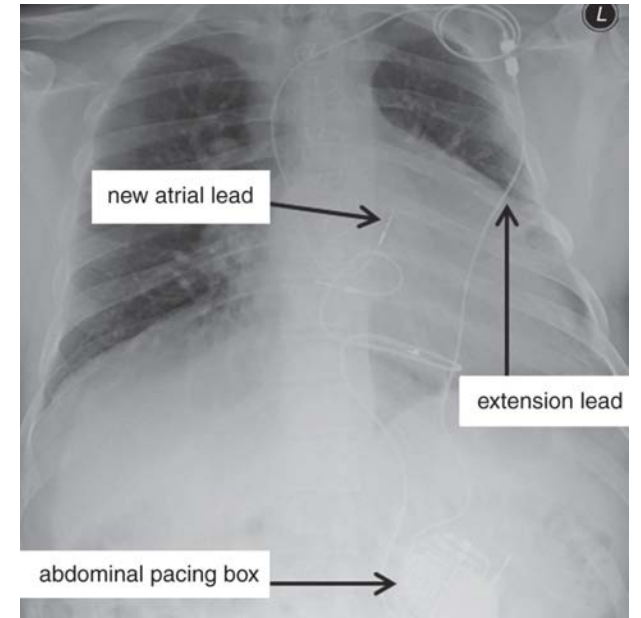
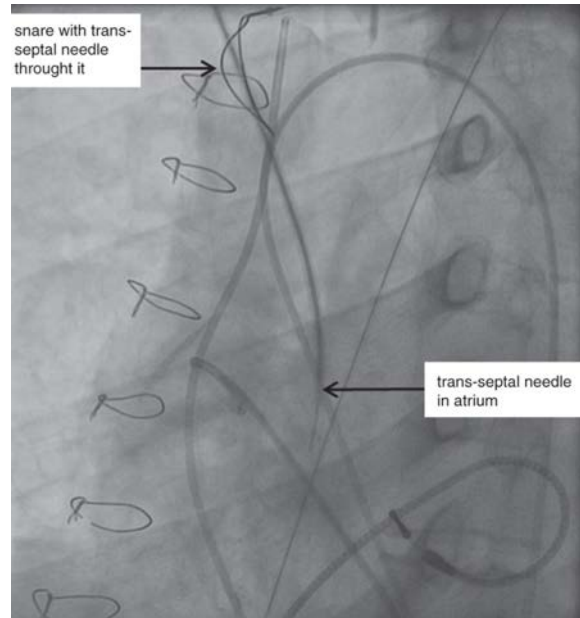
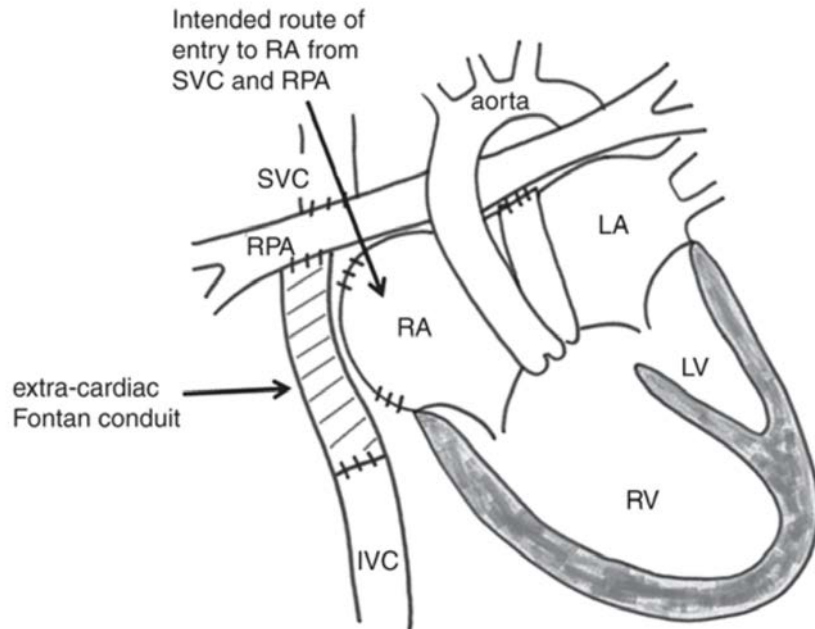


### Creative options



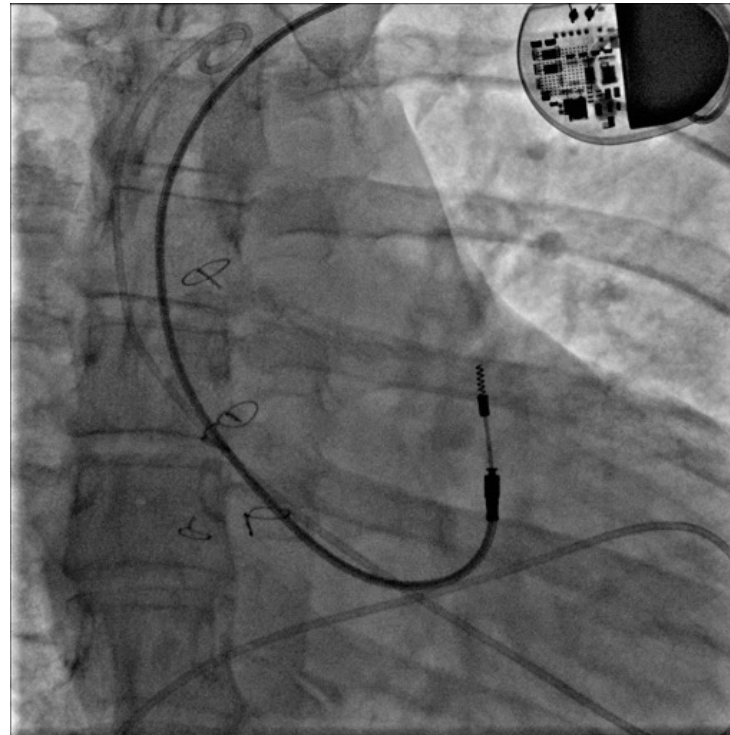
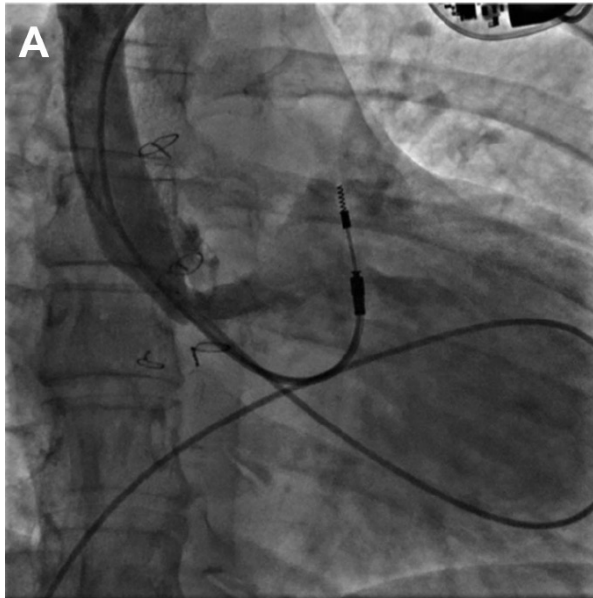
## Vascular access

### Creative options



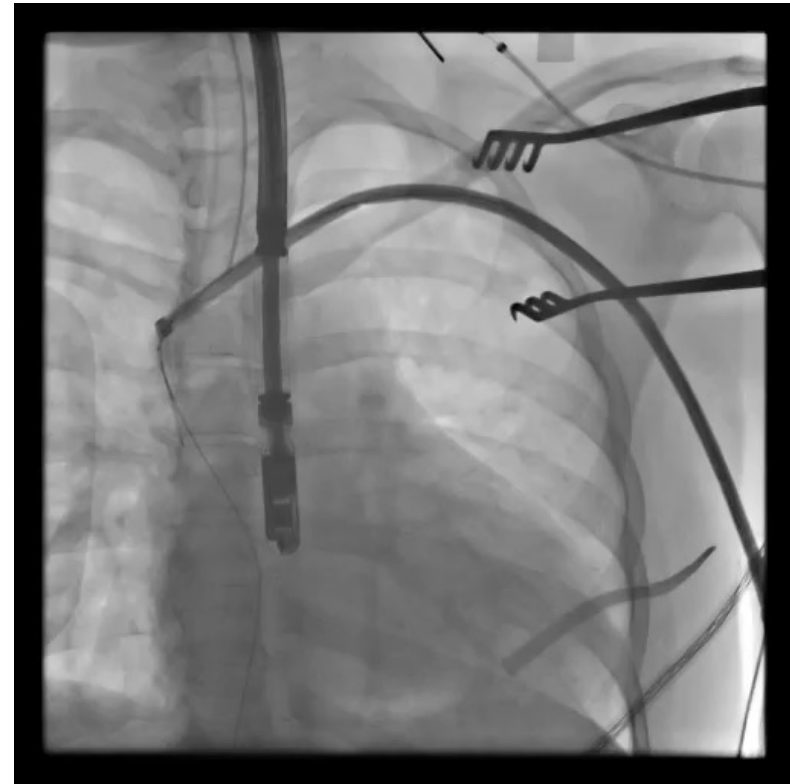
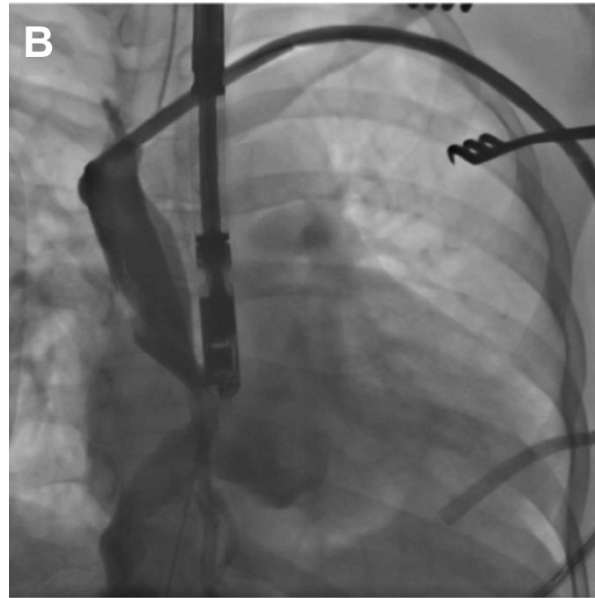
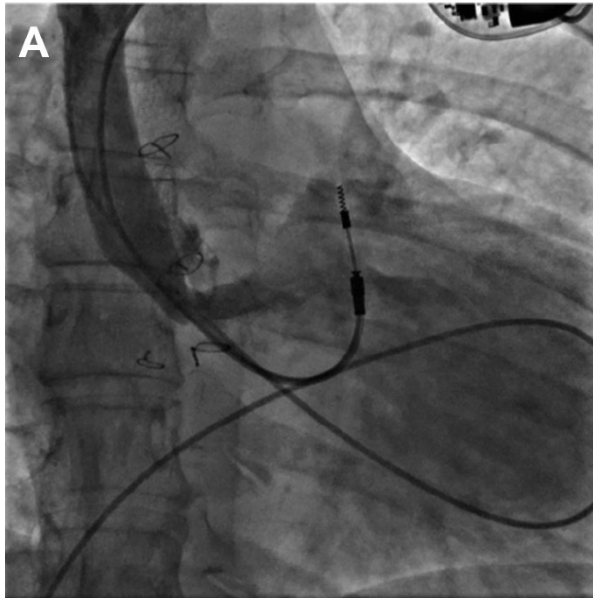
# PACE IN CHD: CHALLENGES

## Vascular access

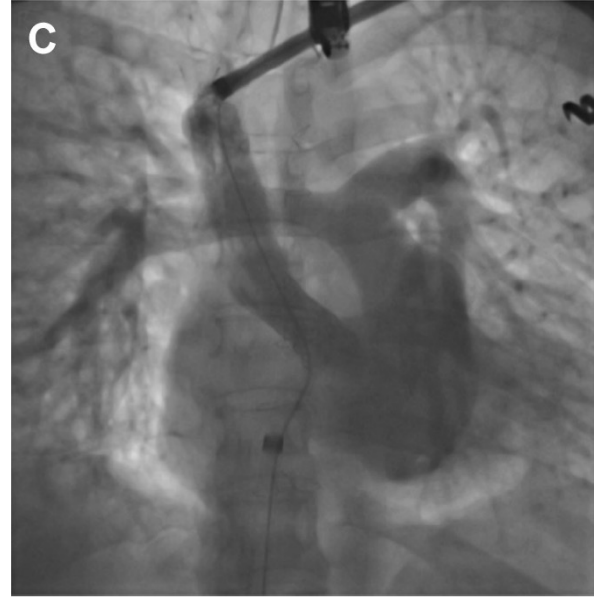
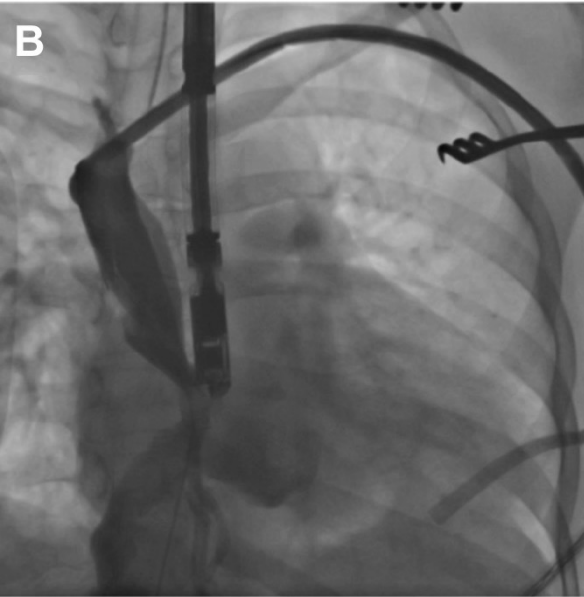
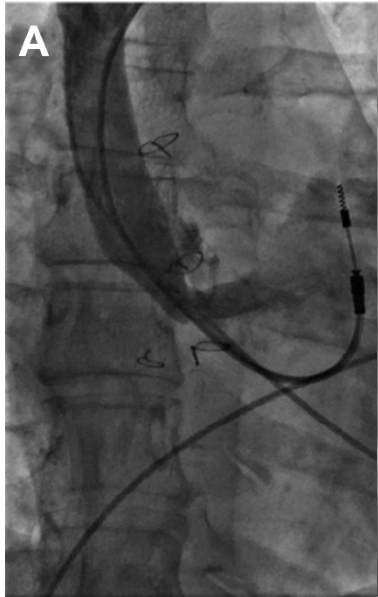




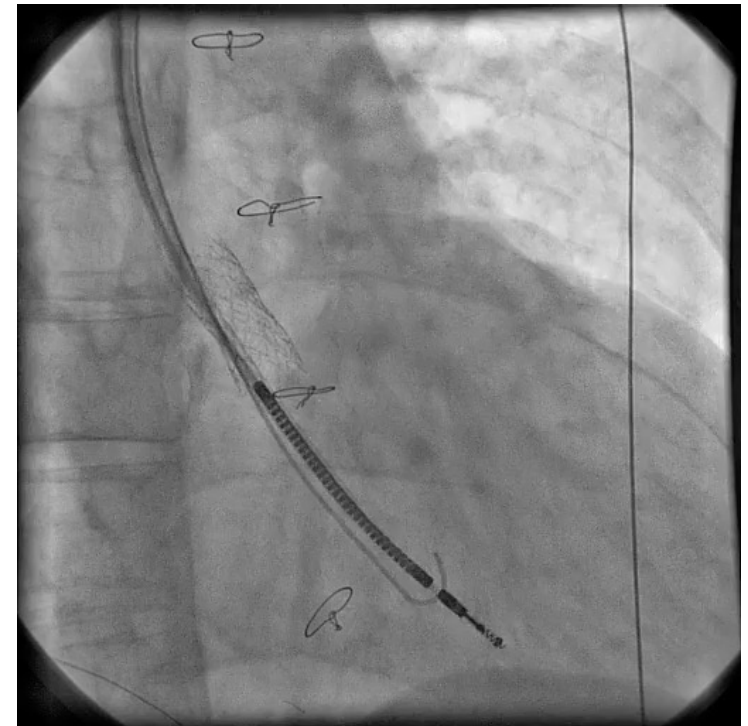
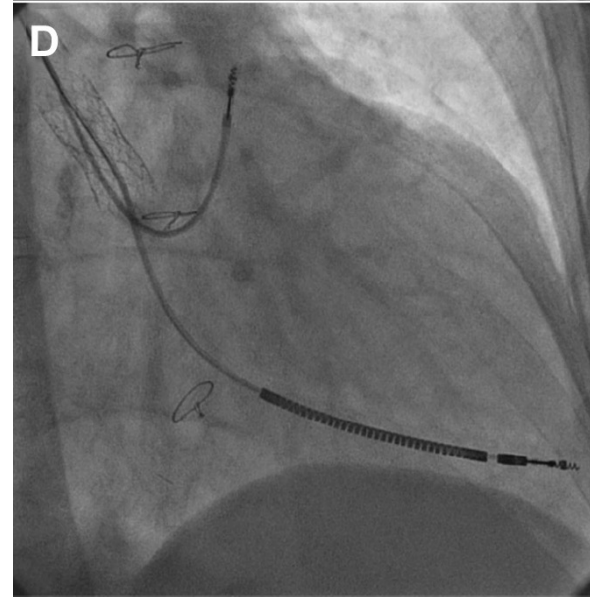
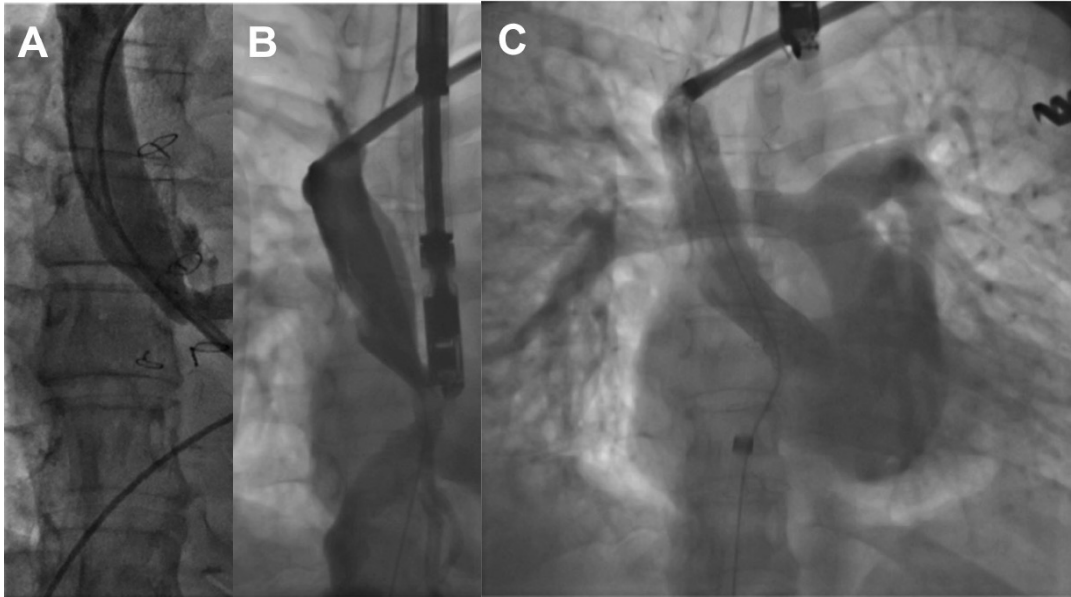
## Vascular access



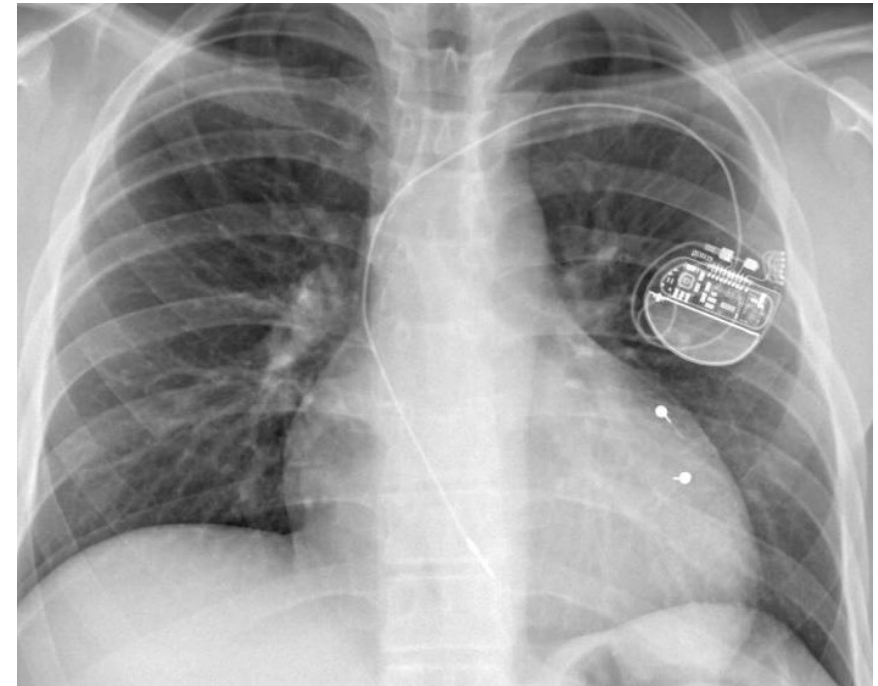
## Vascular access



## Vascular access



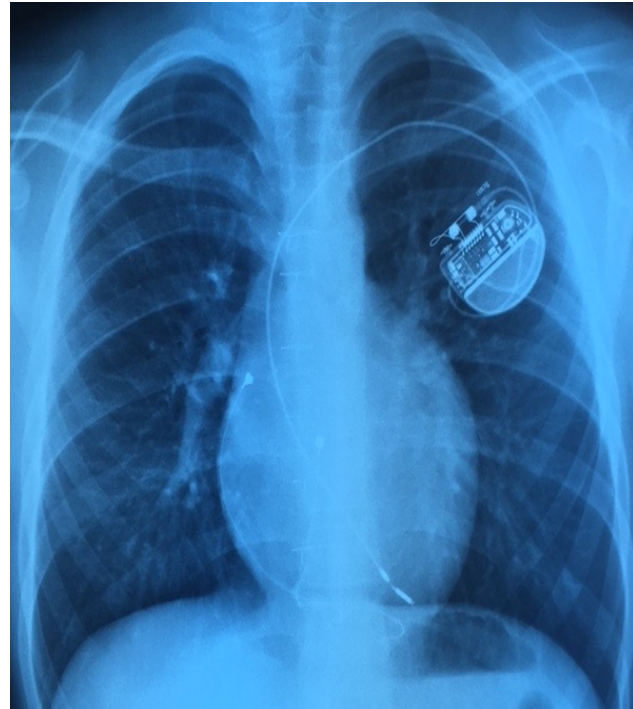
## Morphology and growth



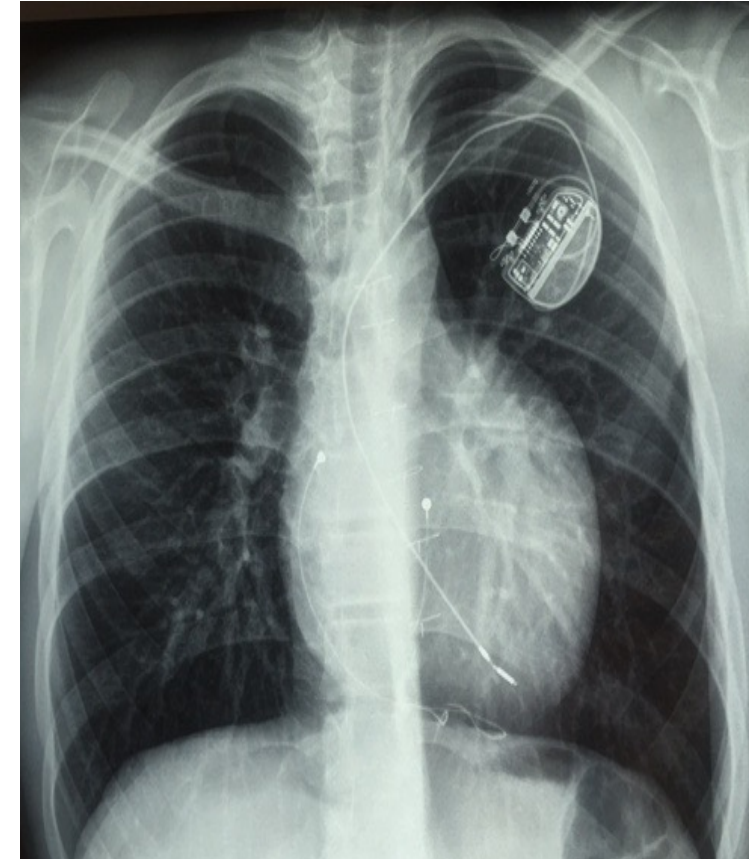
## Morphology and growth



**2003**

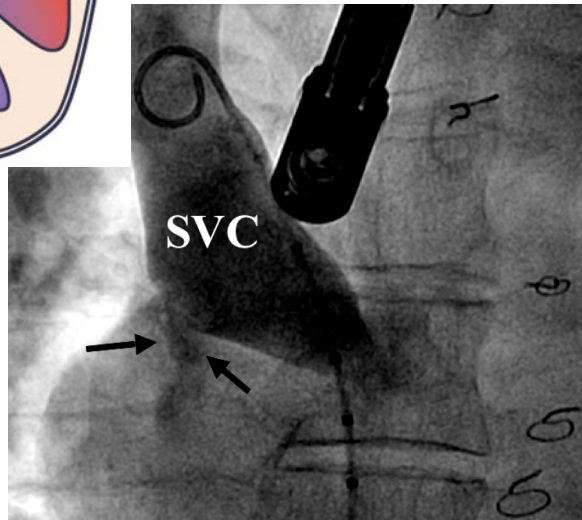
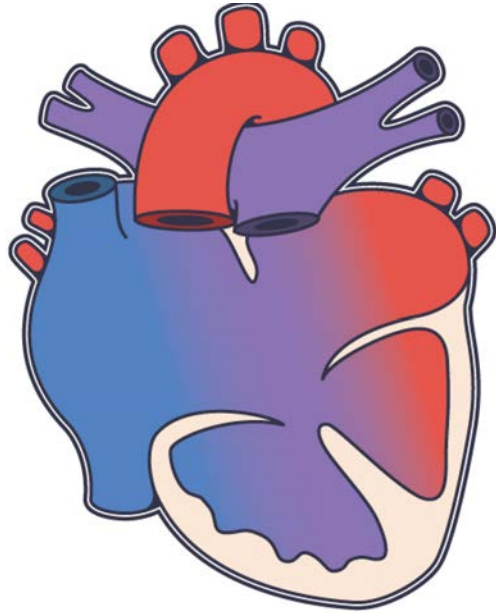


**2012**

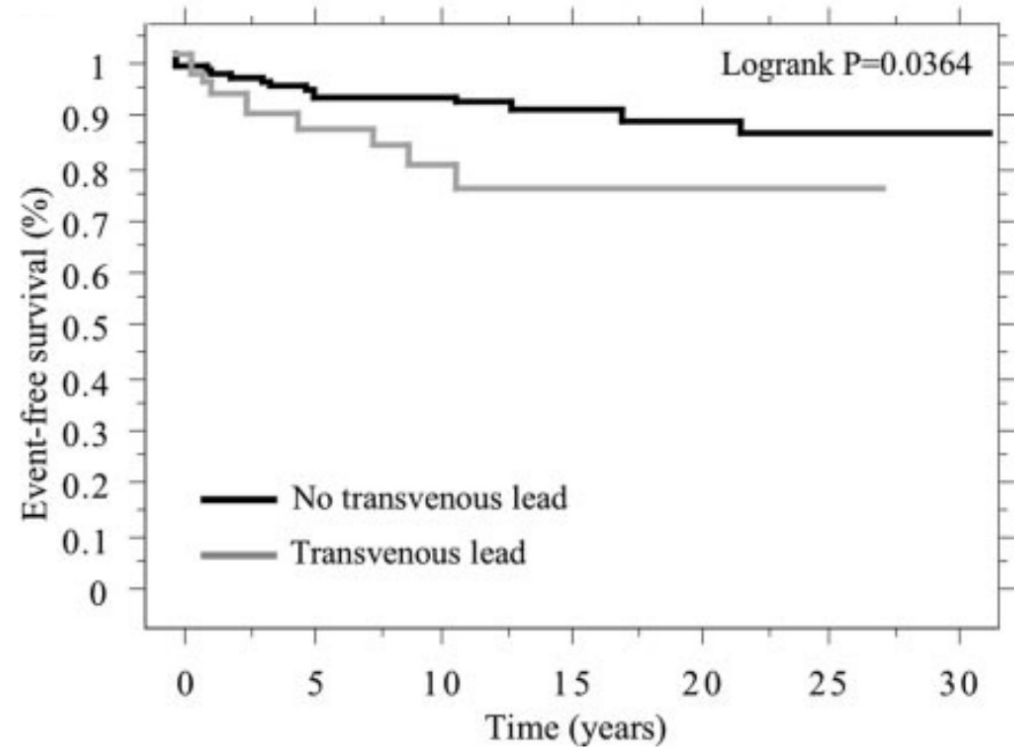


**2015 = dysfunction**

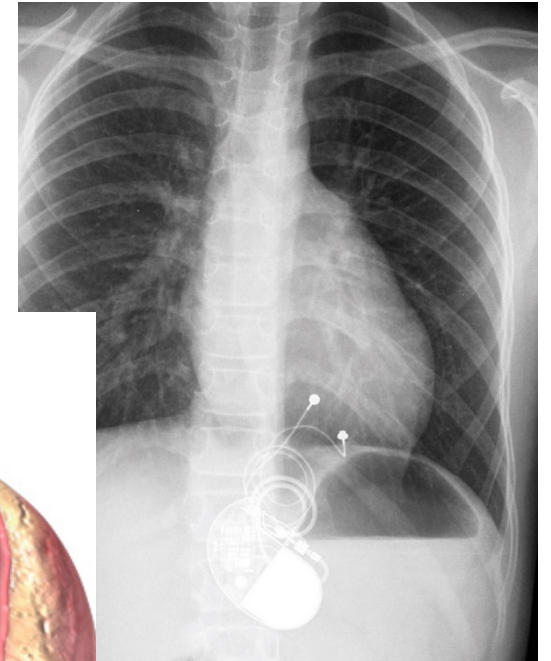
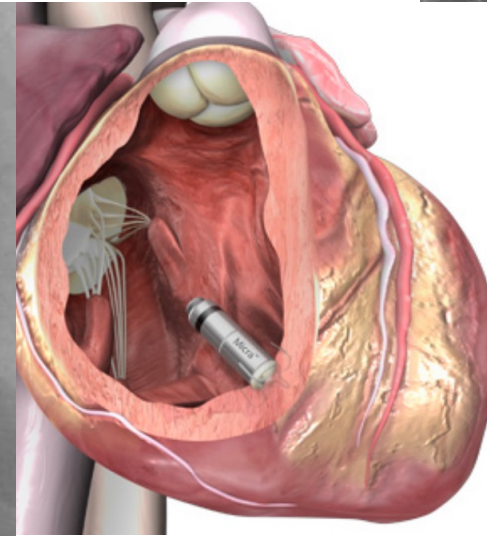
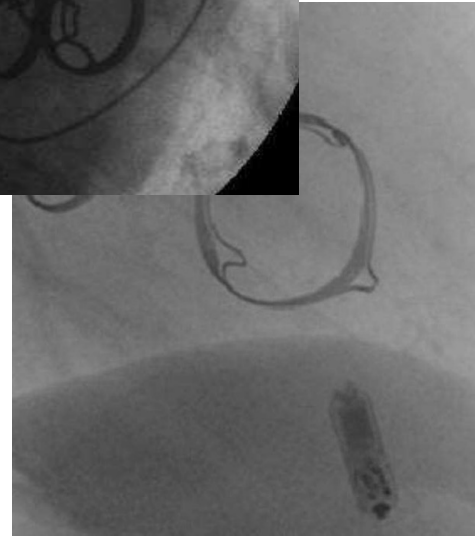
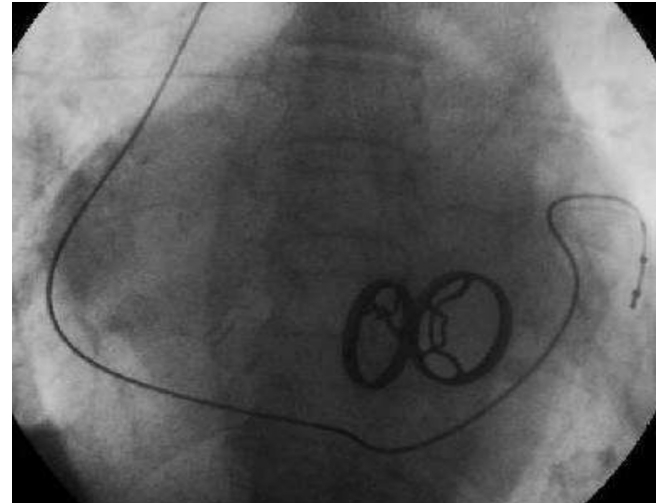
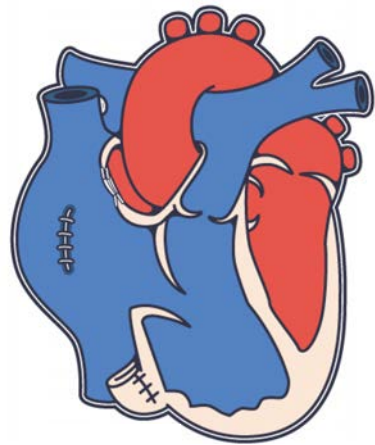
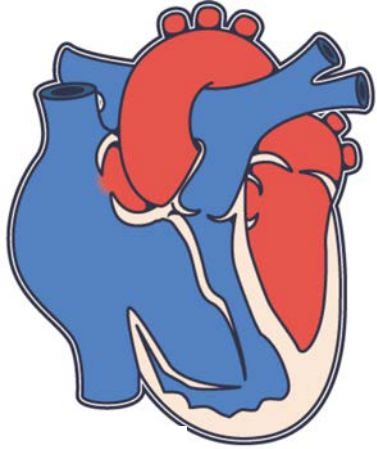
## Persistent shunts



**HR > 2 for thromboembolic events**

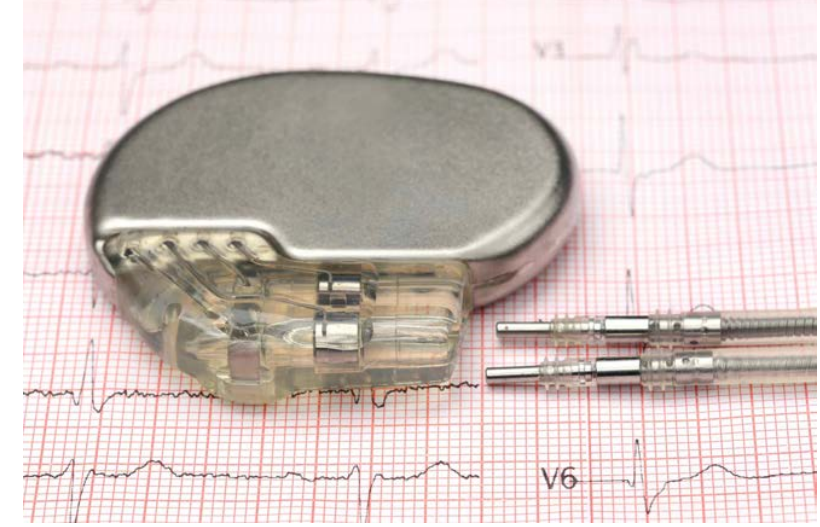


## Tricuspid valve surgery or regurgitation

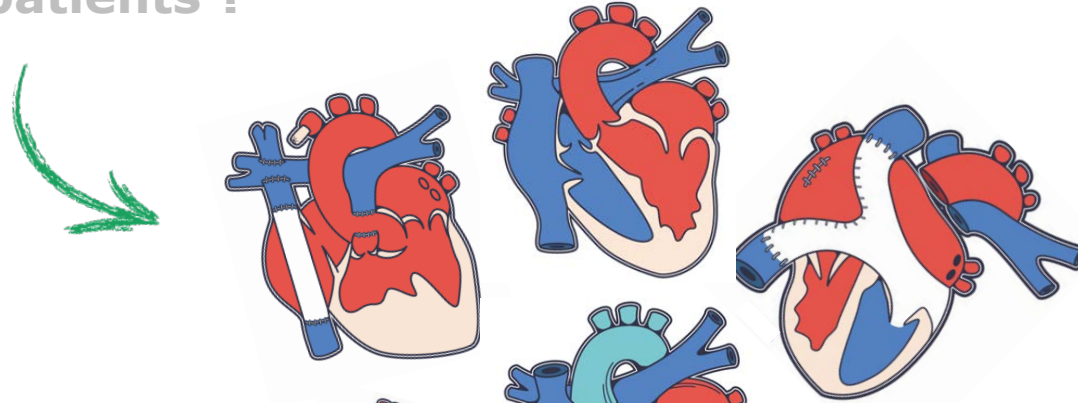




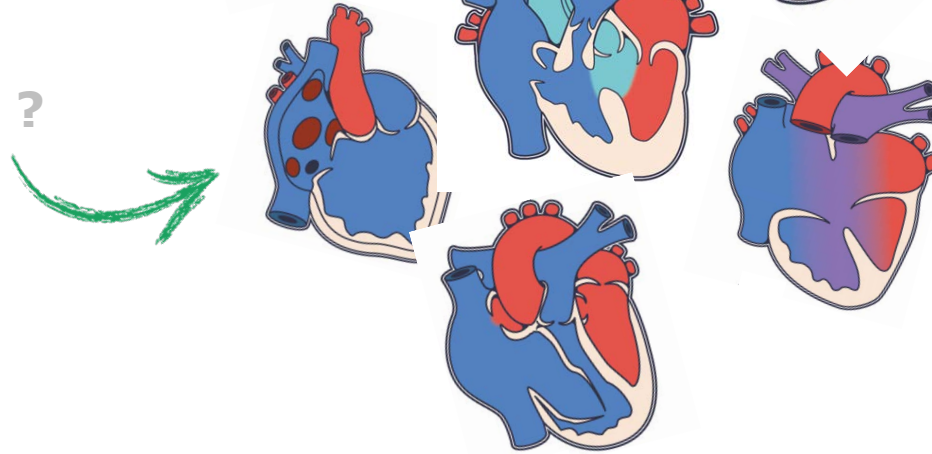
# PACEMAKER IN PEDATRIC & CHD PATIENTS



Which patients ?



Indications ?



Complications ?

How ?

**Cardiac Resynchronization ?**



## Specific features

- **RBBB > LBBB**
- **Systemic RV dysfunction**
- **Subpulmonary RV dysfunction**
- **Univentricular heart dysfunction**

Circulation: Arrhythmia and Electrophysiology

### SPECIAL REPORT

## Cardiac Resynchronization Therapy for Treatment of Chronic Subpulmonary Right Ventricular Dysfunction in Congenital Heart Disease

Jan Janoušek, MD, PhD  
 Jan Kovanda, MD  
 Miroslav Ložek, MSc  
 Viktor Tomek, MD, PhD  
 Roman Gebauer, MD  
 Peter Kubuš, MD, PhD  
 Tammo Delhaas, MD, PhD

Vol. 12, 2005  
 7/05/\$30.00  
 2005.05.096

ization

and Congenital Heart Disease Patients  
 An International Multicenter Study  
 Anne M. Dubin, MD,†  
 Margaret J. Strieper, D  
 Kevin M. Shannon, MD,†  
 Frank J. Zimmerman, MD,†  
 Amin Al Ahmad, MD,†  
 Maully Shah, MD,†  
 Mathias Emmel, MD,†  
 Anjan R...

Journal of Cardiovascular  
 Electrophysiology

Cardiac Resynchronization Therapy in Pediatric Patients

Vol. 44, No. 9, 2004  
 ISSN 0735-1097/04/\$30.00  
 doi:10.1016/j.jacc.2004.08.044

and Multisite Pacing) Case: Five Years



Europace (2017) 0, 1–8  
 doi:10.1093/europace/euw386

### CLINICAL RESEARCH

Journal of the American College of Cardiology  
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 Published by Elsevier Inc.

### EXPRESS PUBLICATION

## Cardiac Resynchronization Therapy: A Novel Adjunct to the Treatment of Systemic Right Ventricular Dysfunction in Pediatric Patients

Jan Janoušek, MD,\* Viktor Tomek, MD,\* Václav Chaloupek, MD,\* Roman A. Gebauer, MD,\* Josef Kautzner, MD, PhD,†



International Journal of  
**CARDIOLOGY**

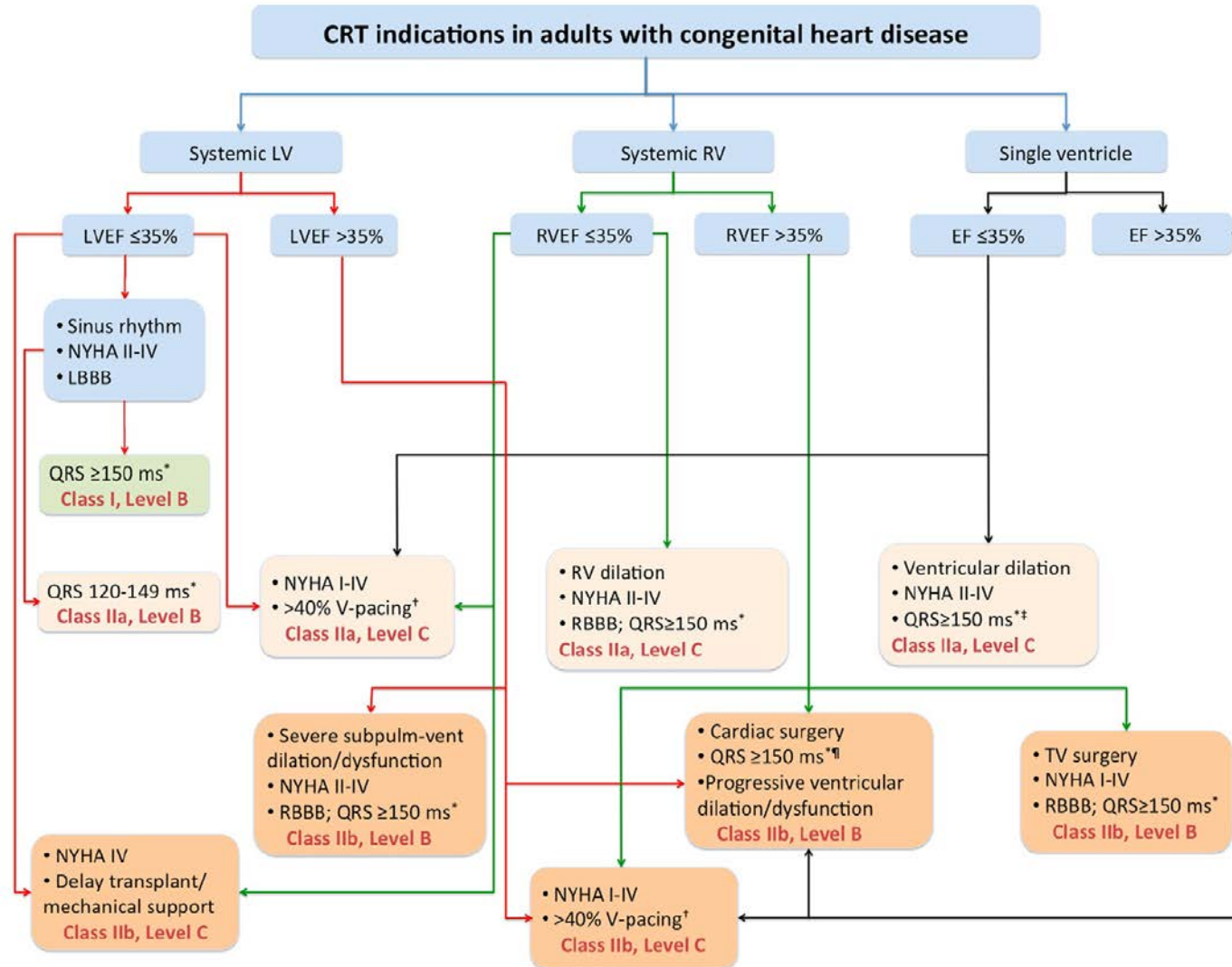
## Cardiac resynchronization therapy in congenital heart disease: Results from the German National Register for Congenital Heart Defects

Ann-Katrin Flügge<sup>a</sup>, Kristina Wasmer<sup>b</sup>, Stefan Orwat<sup>a</sup>, Hashim Abdul-Khaliq<sup>c</sup>, Paul C. Helm<sup>d</sup>, Ulrike Bauer<sup>d</sup>, Helmut Baumgartner<sup>a</sup>, Gerhard-Paul Diller<sup>a,\*</sup> for the German Competence Network for Congenital Heart Defects Investigators

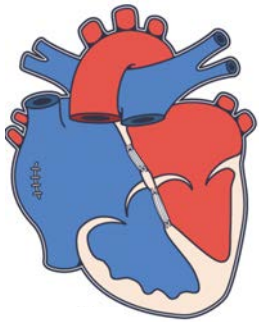
## Cardiac resynchronization therapy in adults with congenital heart disease

Zeliha Koyak<sup>1,2</sup>, Joris R. de Groot<sup>1</sup>, Ahmed Krimly<sup>3</sup>, Tara M. Mackay<sup>1</sup>, Berto J. Bouma<sup>1</sup>, Candice K. Silversides<sup>3</sup>, Erwin N. Oechslin<sup>3</sup>, Ulas Hoke<sup>4</sup>, Lieselot van Erven<sup>4</sup>, Werner Budts<sup>5</sup>, Isabelle C. Van Gelder<sup>6</sup>, Barbara J. M. Mulder<sup>1,2\*</sup>, and Louise Harris<sup>3</sup>

# RESYNCHRONIZATION IN CHD

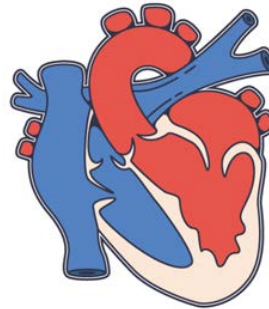


## Guidelines summary



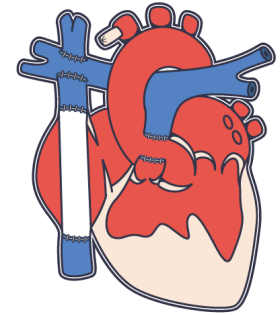
**Systemic LV  $\leq 35\%$**

LBBB  $\geq 120$  ms



**Systemic RV  $\leq 35\%$**

RBBB  $\geq 150$  ms

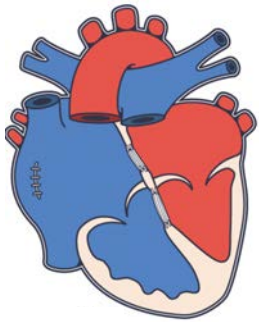


**Single V  $\leq 35\%$**

QRS  $\geq 150$  ms

**Or V-pacing  $> 40\%$**

## Guidelines summary

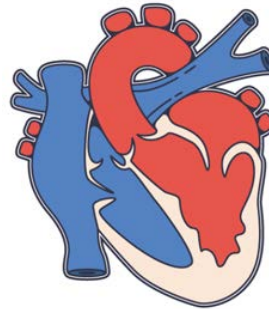


**Systemic LV  $\leq 35\%$**

**LBBB  $\geq 120$  ms**

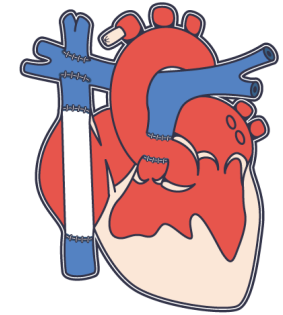
- Severe subpulm-vent dilation/dysfunction
- NYHA II-IV
- RBBB; QRS  $\geq 150$  ms\*

**Class IIb, Level B**



**Systemic RV  $\leq 35\%$**

**RBBB  $\geq 150$  ms**



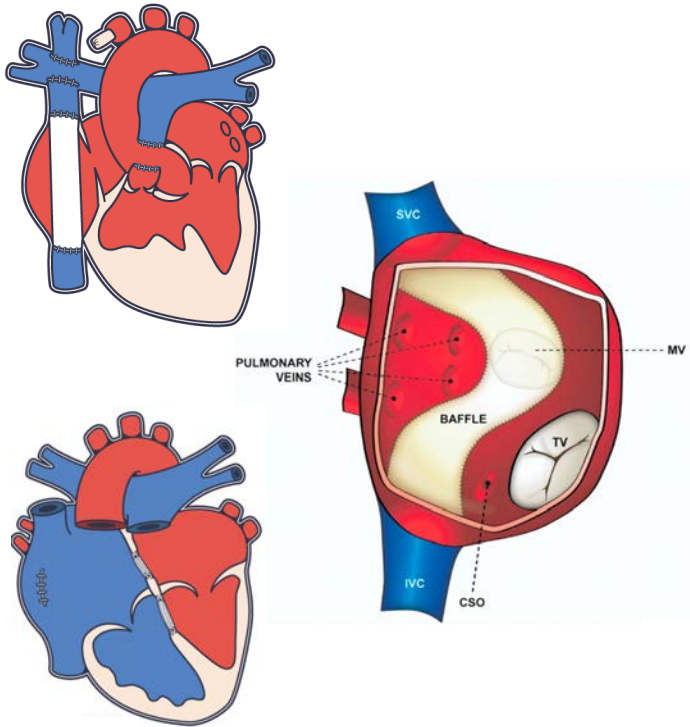
**Single V  $\leq 35\%$**

**QRS  $\geq 150$  ms**

**Or V-pacing  $> 40\%$**

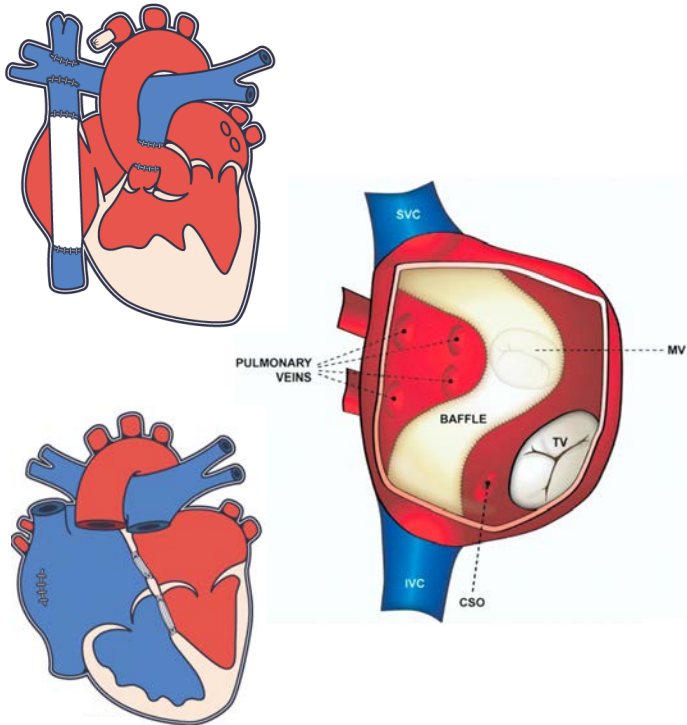
*Khairy P, et al. Heart Rhythm 2014  
Hernandez-Madrid A, et al. Europace 2018*

## Technical aspects



**Coronary sinus may be inaccessible**

## Technical aspects



Journal of the American College of Cardiology  
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 ISSN 0735-1097/05/\$30.00  
 doi:10.1016/j.jacc.2005.05.096

### FOCUS ISSUE: CARDIAC RESYNCHRONIZATION THERAPY

#### Congenital Heart Disease and Resynchronization

### Resynchronization Therapy in Pediatric and Congenital Heart Disease Patients

An International MultiCenter Study

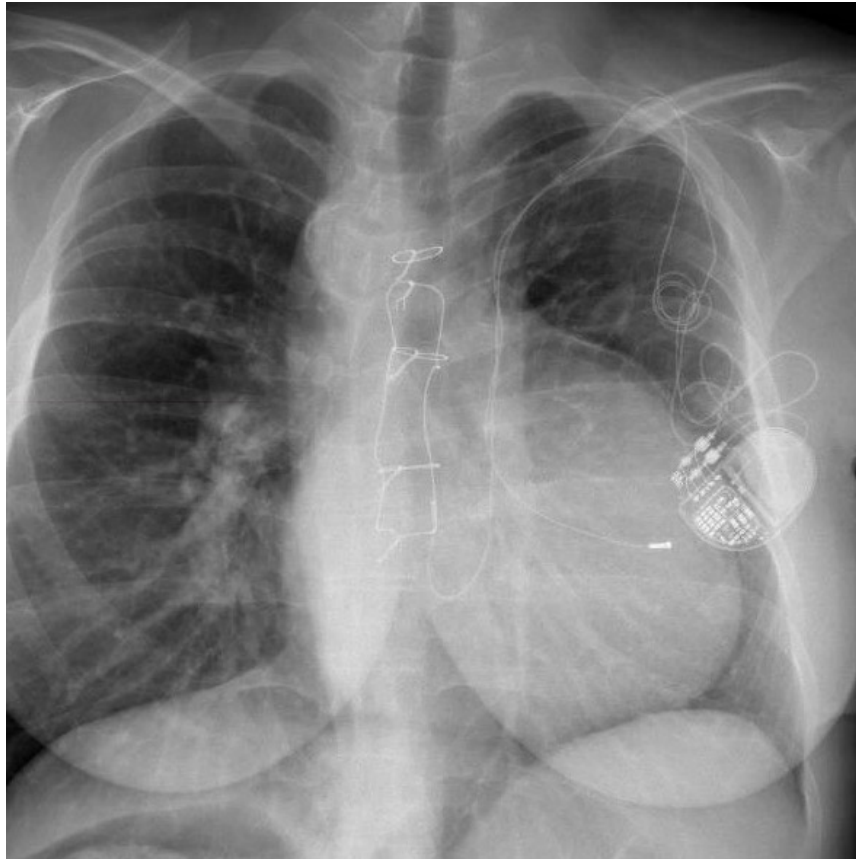
Anne M. Dubin, MD, FACC,\* Jan Janousek, MD,† Edward Rhee, MD, FACC,‡

## 73 patients with CHD

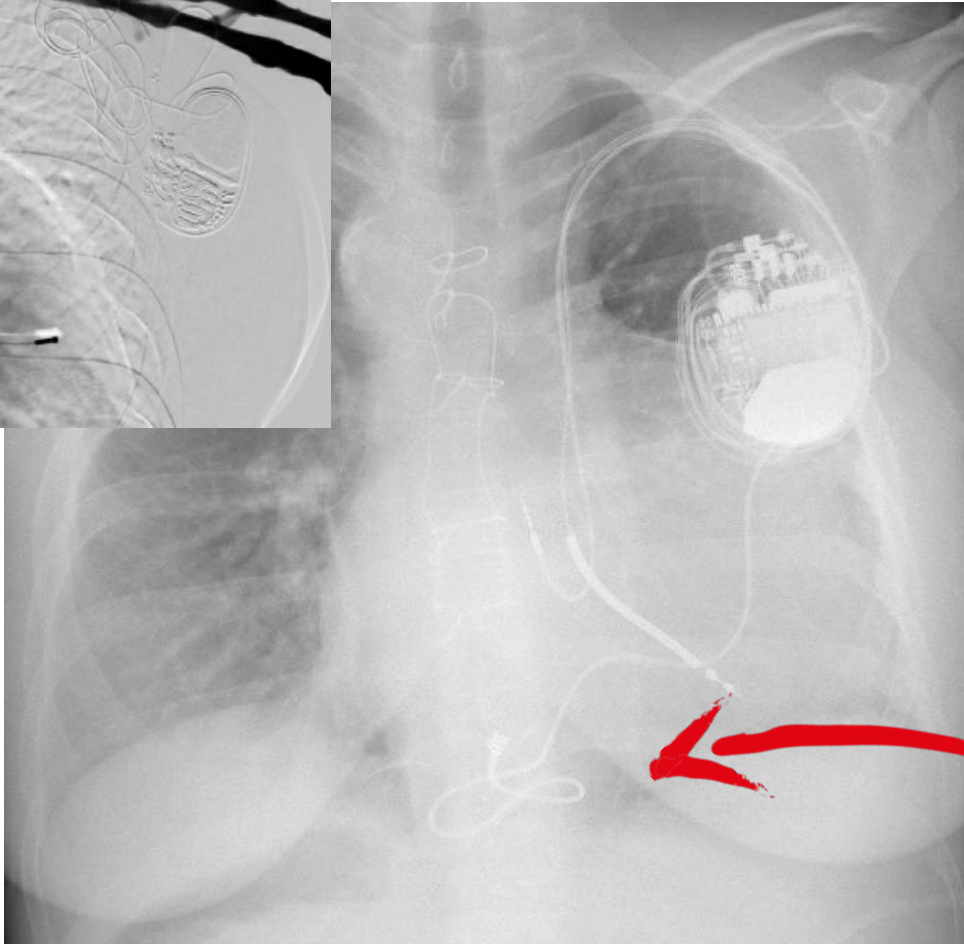
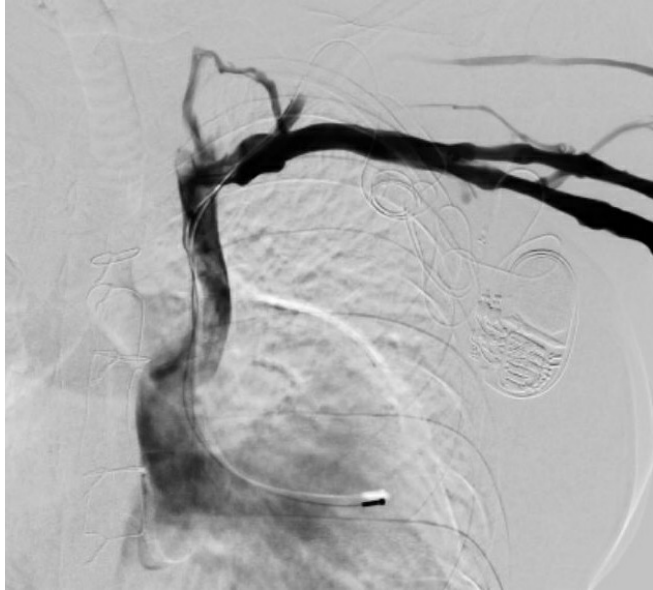
**Coronary sinus may be inaccessible**

Transvenous System (%)	Epicardial/Mixed System (%/%)
26 (36%)	37/10 (51%/14%)

## Hybrid approach



## Hybrid approach

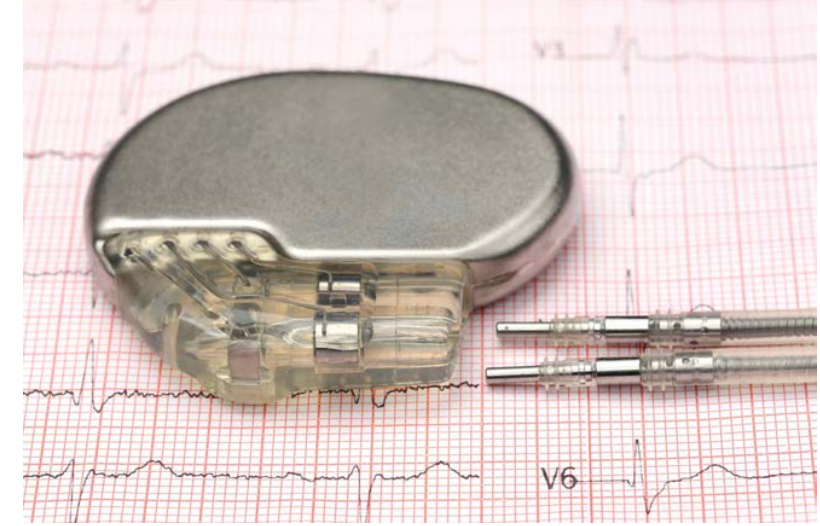


**Epicardial « left »  
tunneled lead**

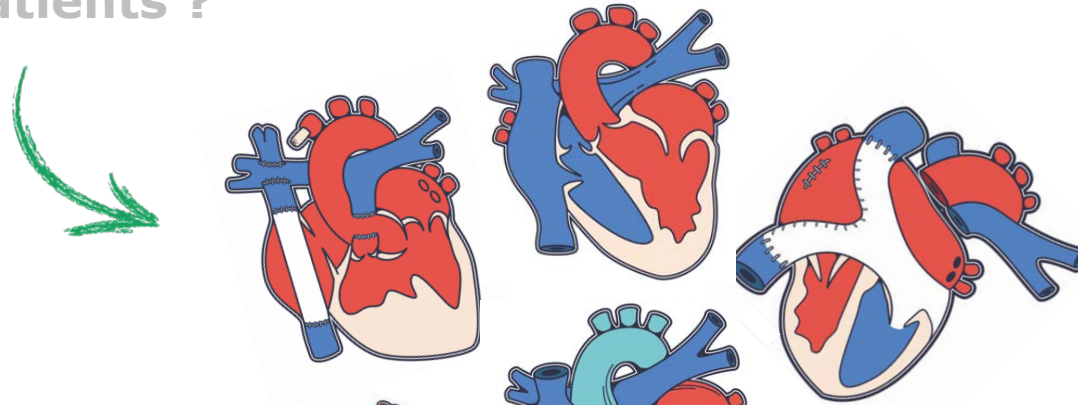




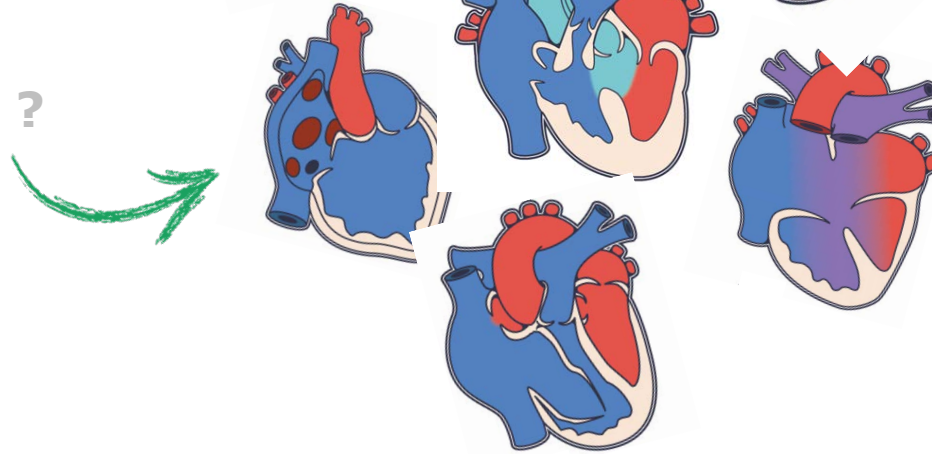
# PACEMAKER IN PEDATRIC & CHD PATIENTS



Which patients ?



Indications ?



How ?



Cardiac Resynchronization ?



Complications ?

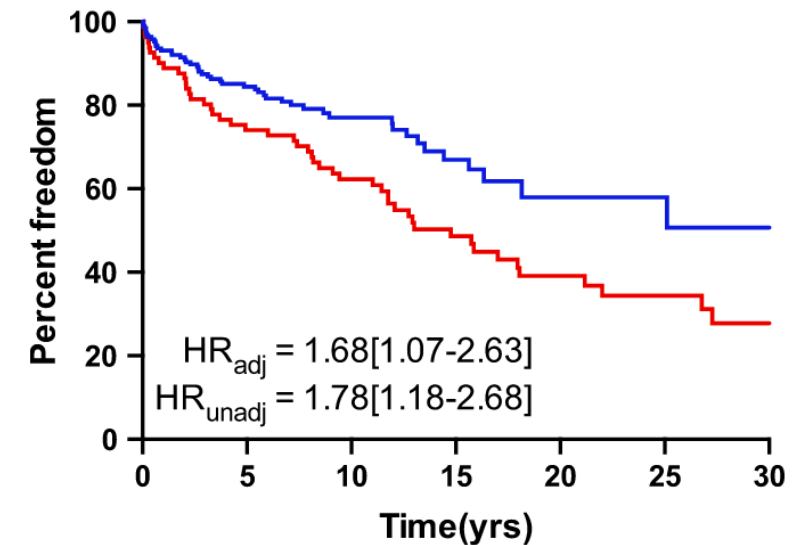
**Table 5**  
Late pacemaker-related complications.

Variable	Total population (n = 274)
Duration of follow-up (years), median [IQR]	12 [6-19]
Total number of patients	95 (34.6)
Late lead failure	68 (24.8)
Pacemaker dysfunction/early battery depletion	14 (5.1)
Pacemaker migration	13 (4.7)
Erosion	13 (4.7)
Pocket infection	12 (4.4)
Pacing system-related endocarditis	7 (2.5)
Other	9 (3.3)

Data are presented as n (%), unless stated otherwise.

**12 y of follow up**  
**35% of late complications**

**Freedom from pacemaker complications**

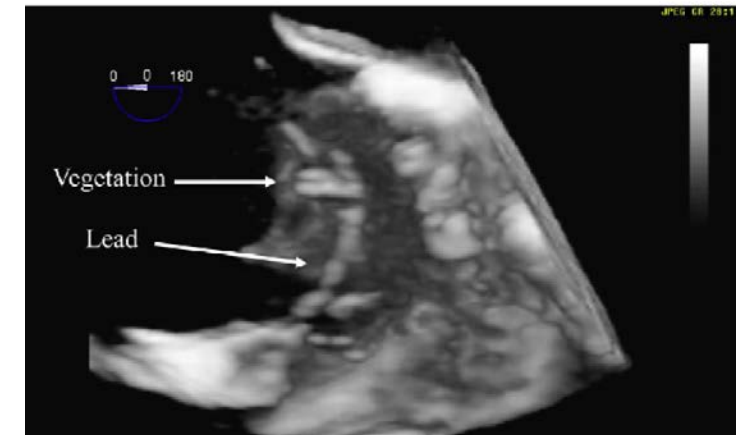
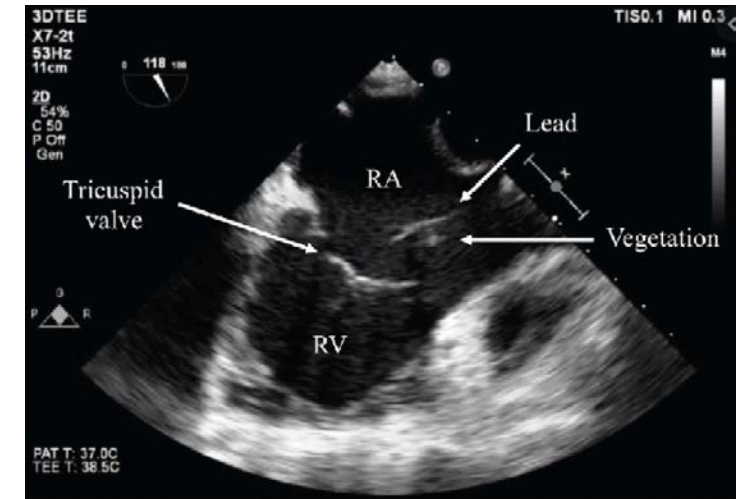
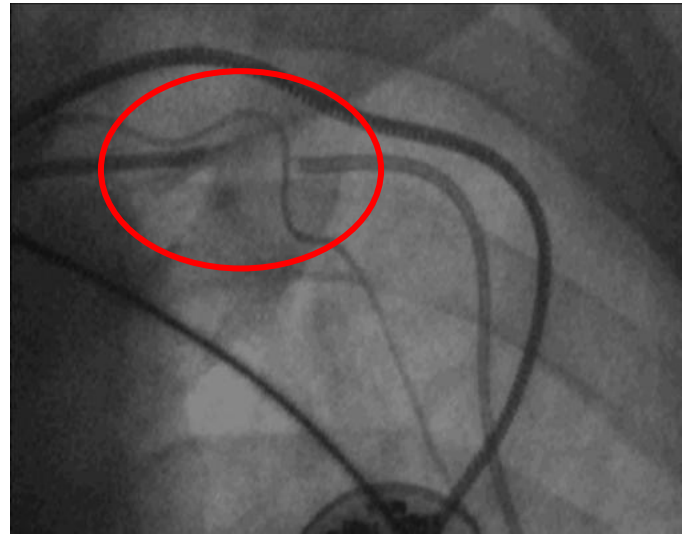


	0	5	10	15	20	25	30
<18 years	81	59	46	27	17	11	6
≥18 years	193	128	66	32	13	8	1

Numbers at risk

**Fig. 2.** Pacemaker-related complication event-free survival in two age cohorts.

# COMPLICATIONS



**Table 5**  
Late pacemaker-related complications.

Variable	Total population (n = 274)
Duration of follow-up (years), median [IQR]	12 [6-19]
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Pacing system-related endocarditis	7 (2.5)
Other	9 (3.3)

**High Burden**

- Vessels size
- Growth
- Higher activity
- Reinterventions ++

Data are presented as n (%), unless stated otherwise.

12 y of follow up  
35% of late complications

Freedom from pacemaker complications

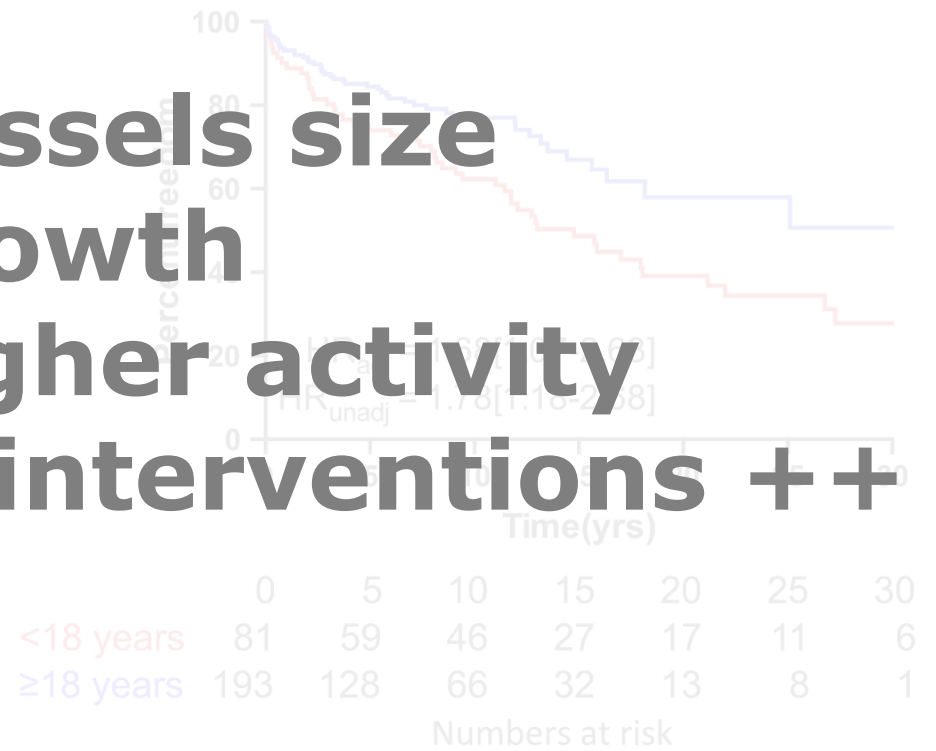


Fig. 2. Pacemaker-related complication event-free survival in two age cohorts.

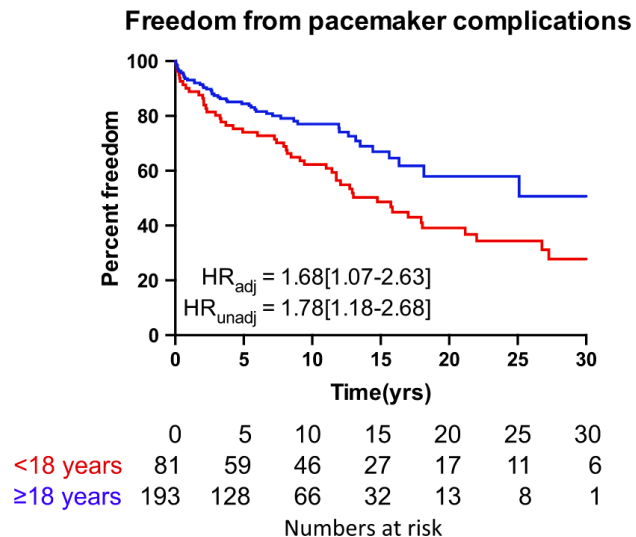
# COMPLICATIONS

## Reasons for epicardial system

**Table 5**  
Late pacemaker-related complications.

Variable	Total population (n = 274)
Duration of follow-up (years), median [IQR]	12 [6-19]
Total number of patients	95 (34.6)
Late lead failure	68 (24.8)
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Pacemaker migration	13 (4.7)
Erosion	13 (4.7)
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Pacing system-related endocarditis	7 (2.5)
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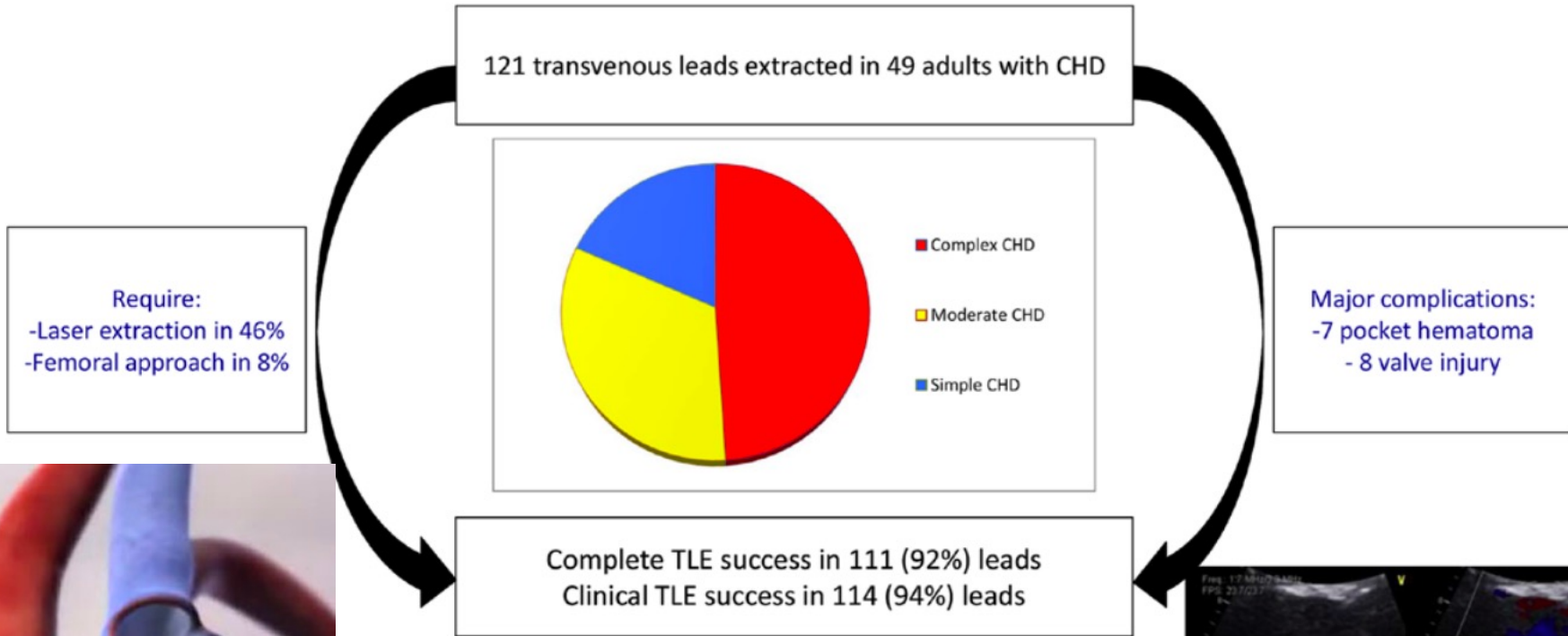
Data are presented as n (%), unless stated otherwise.



**Fig. 2.** Pacemaker-related complication event-free survival in two age cohorts.

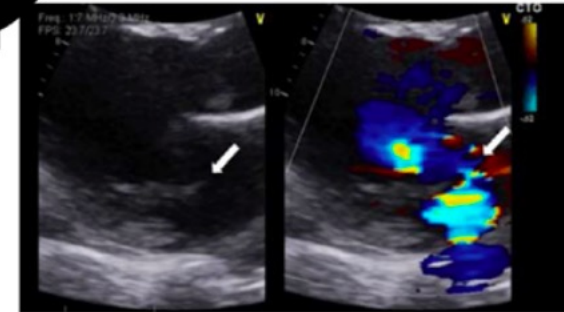
- **Body size / young age** **24%**
- **Venous access** **22%**
- **Concomitant cardiac surgery** **19%**
- **Right-sided mechanical valve** **11%**
- **Intra-cardiac shunt** **6%**
- **Endocarditis** **2%**

# COMPLICATIONS



**Laser**

Chord disruption and posterior leaflet injury (arrow) requiring sub-pulmonary atrio-ventricular valve replacement.



## Epicardial



**Preservation of venous system**  
**Left ventricular pacing**  
**Possible from birth**



**Higher stimulation/detection thresholds**  
**Lower leads longevity**  
**Coronary compression risk**

**Vs.**



**Better stimulation/detection thresholds**  
**Better leads longevity**

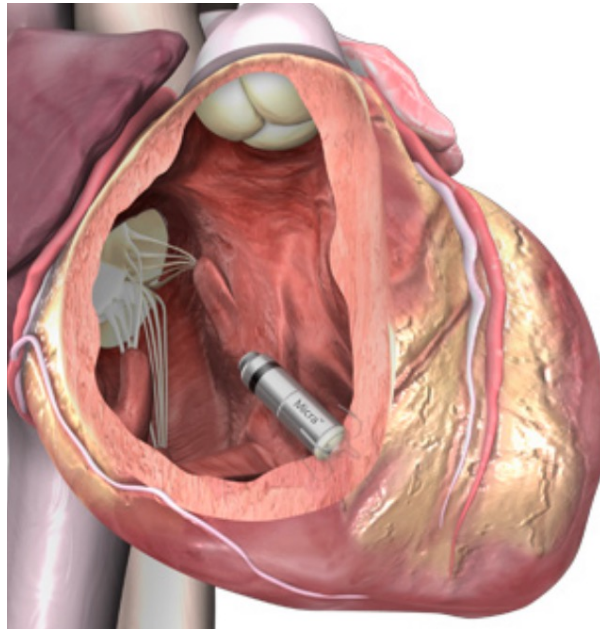
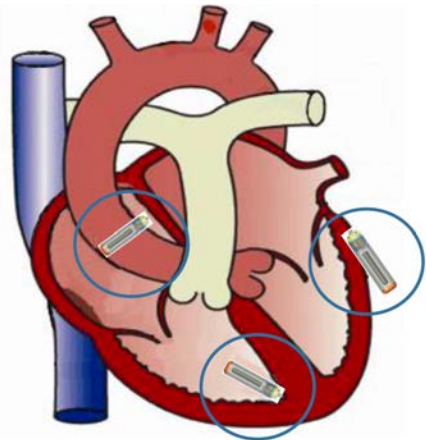
## Endovascular



**Vascular occlusion**  
**Infection/endocarditis**  
**Risks associated with extraction**  
**Right ventricular pacing**  
**Not < 10 kg**

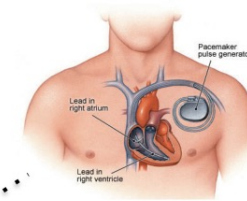
## Epicardial >> Endovascular in children (team-dependent)

### Perspectives ?



#### Conventional pacing therapy

*Implantation of a pacemaker*



#### *alternative biological approaches*

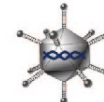
#### Cell therapy

*e.g. injection of embryonic stem cells*



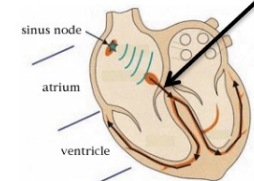
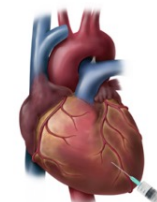
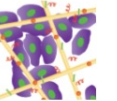
#### Gene therapy

*e.g. delivery of pacemaker genes using viral vectors*



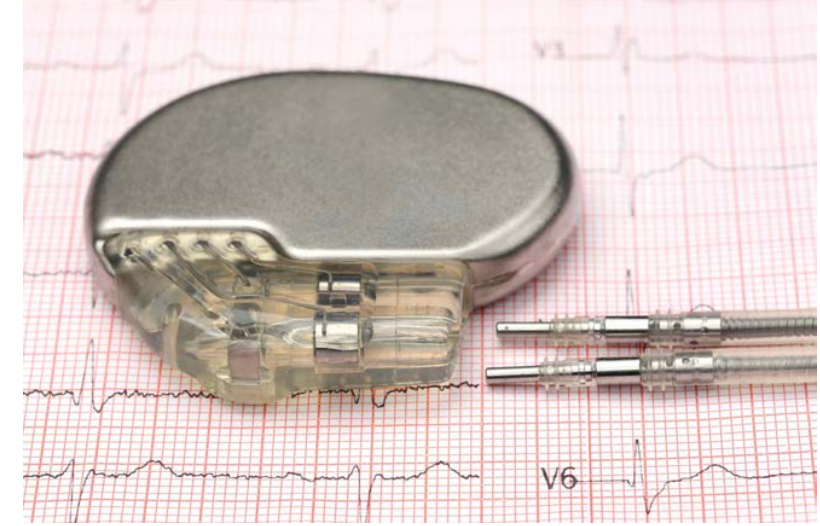
#### Tissue engineering

*autologous, conductive 3D tissue device*

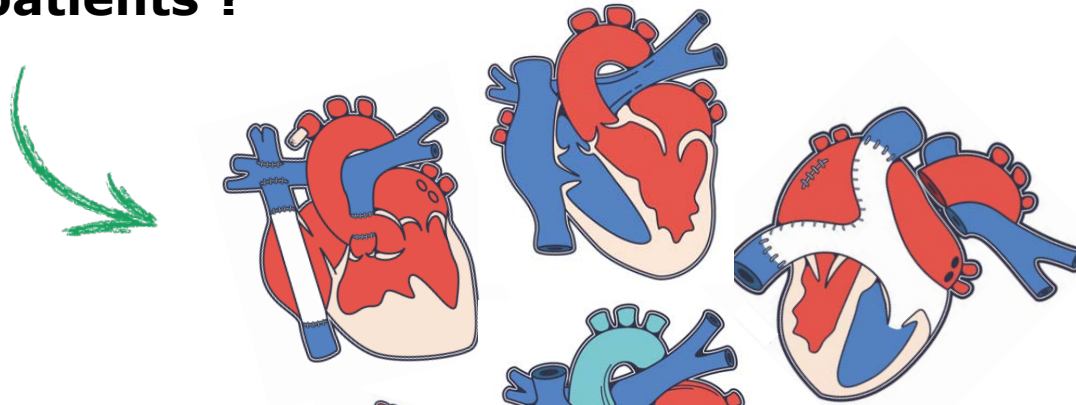




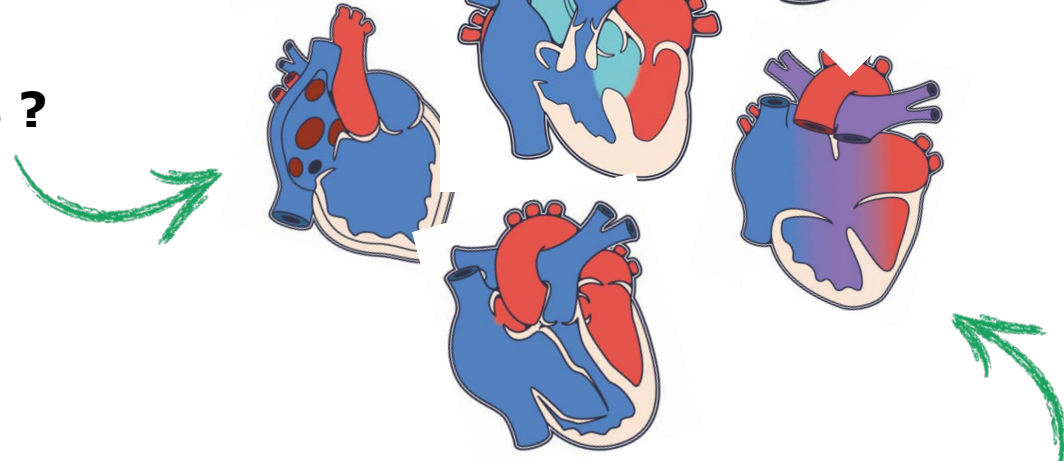
# PACEMAKER IN PEDATRIC & CHD PATIENTS



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Indications ?



Complications ?

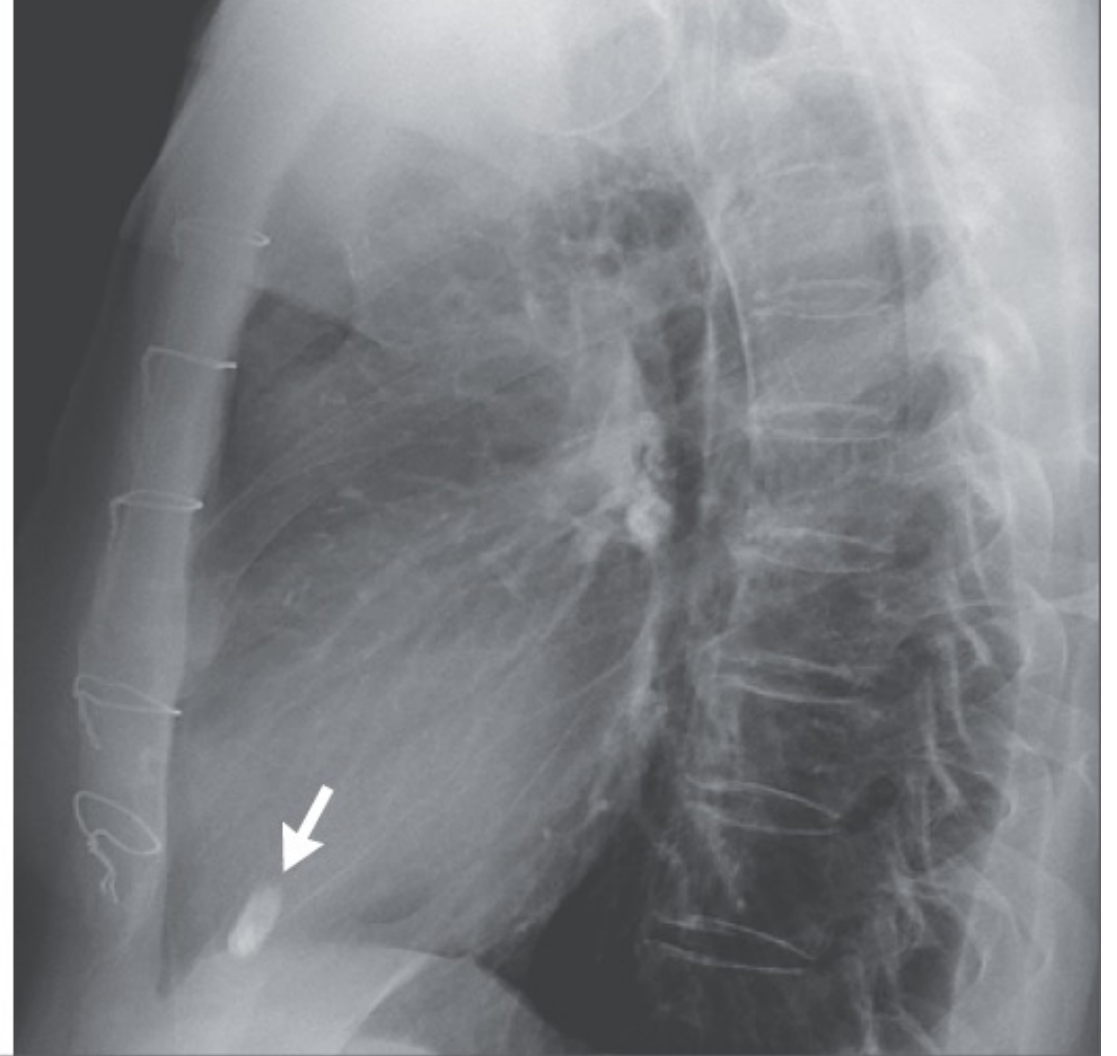
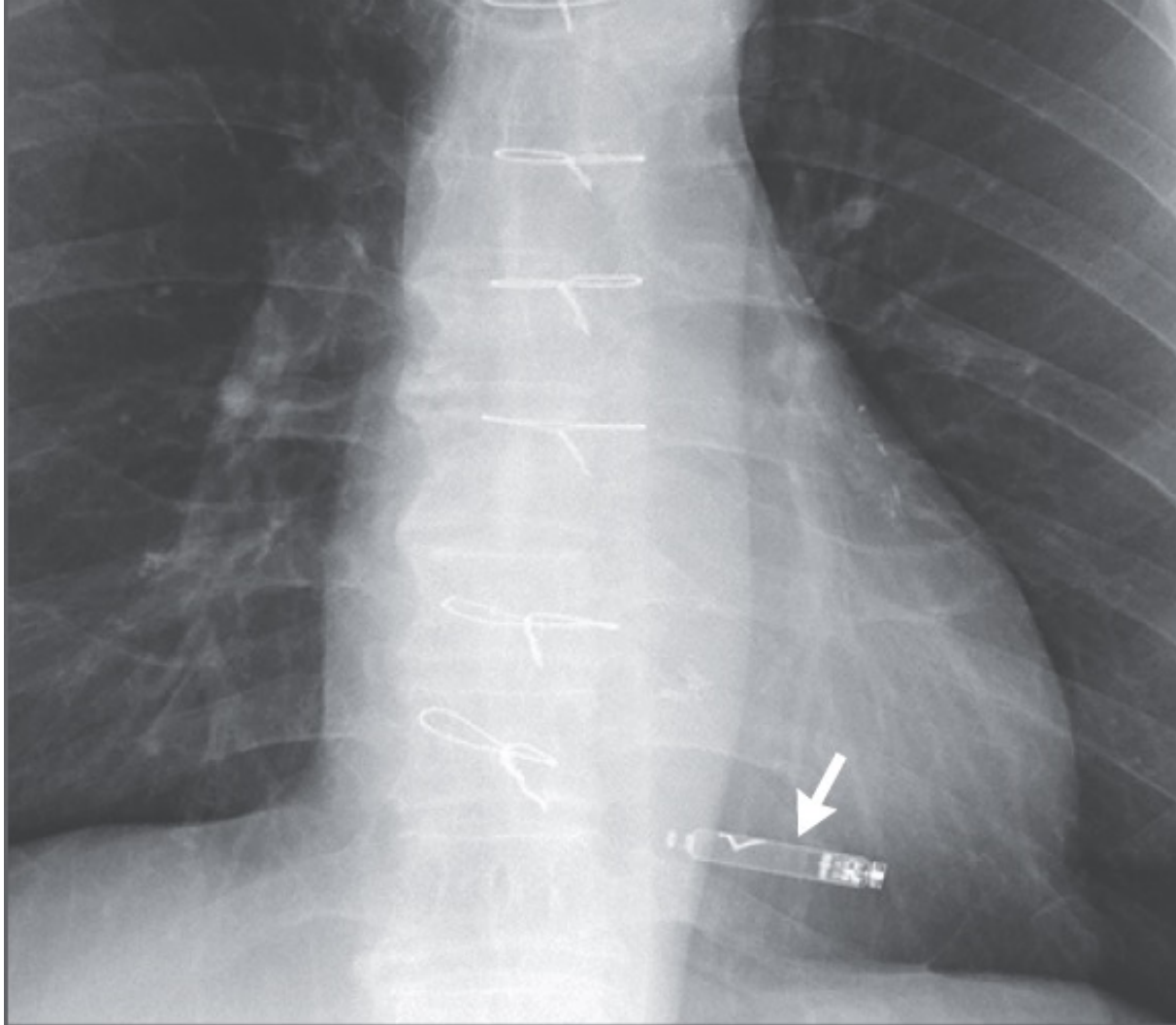
How ?

Cardiac Resynchronization ?

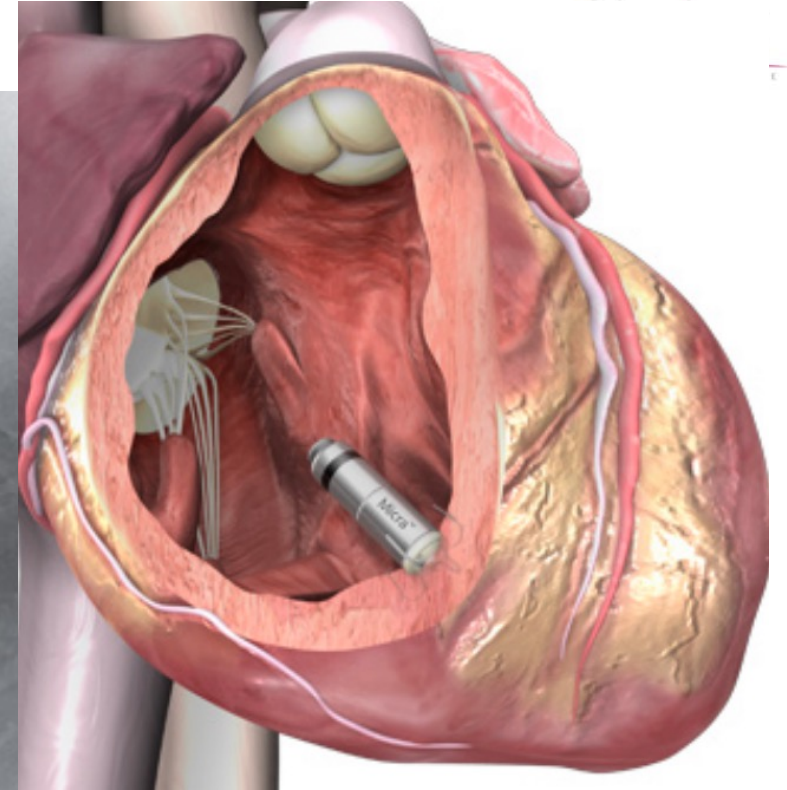
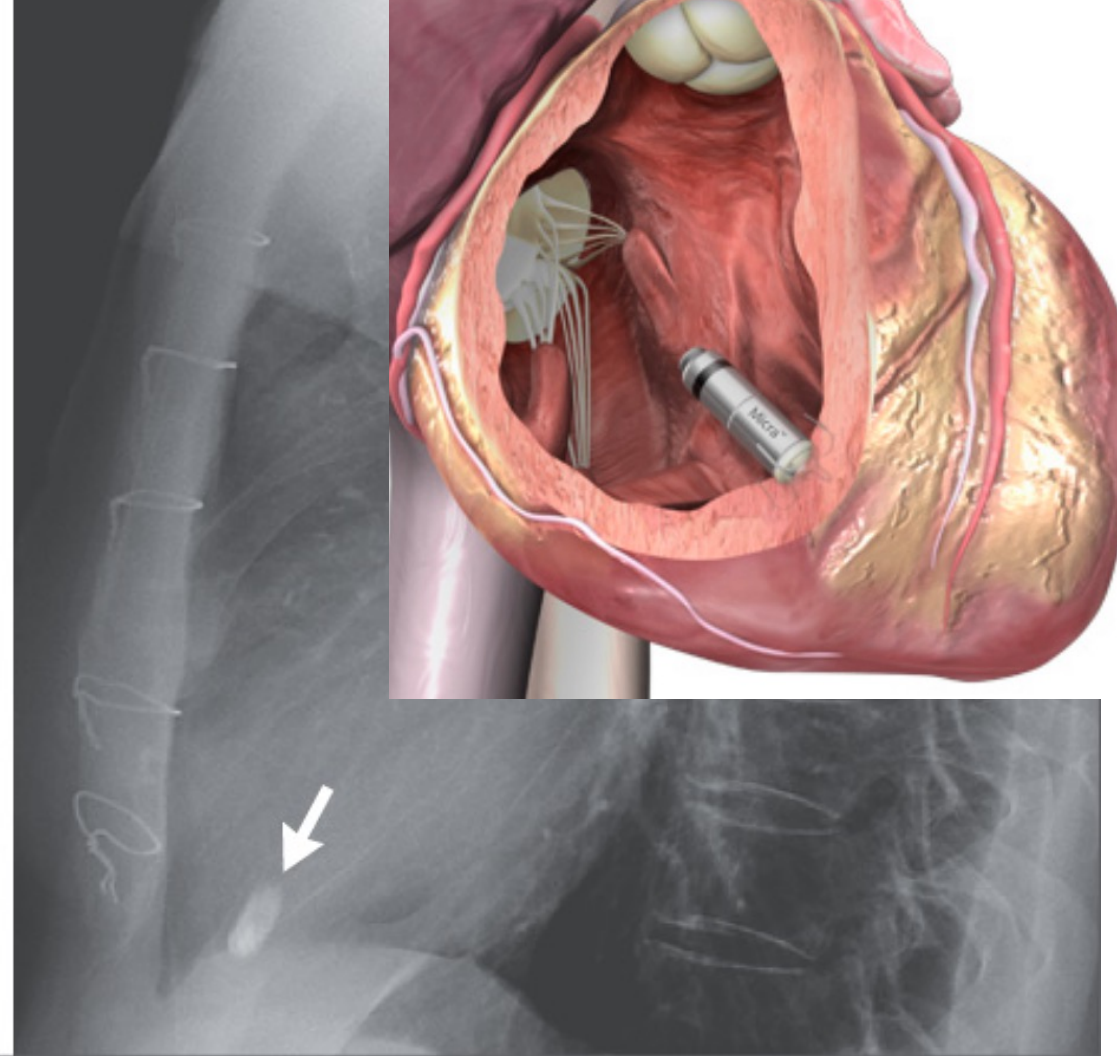
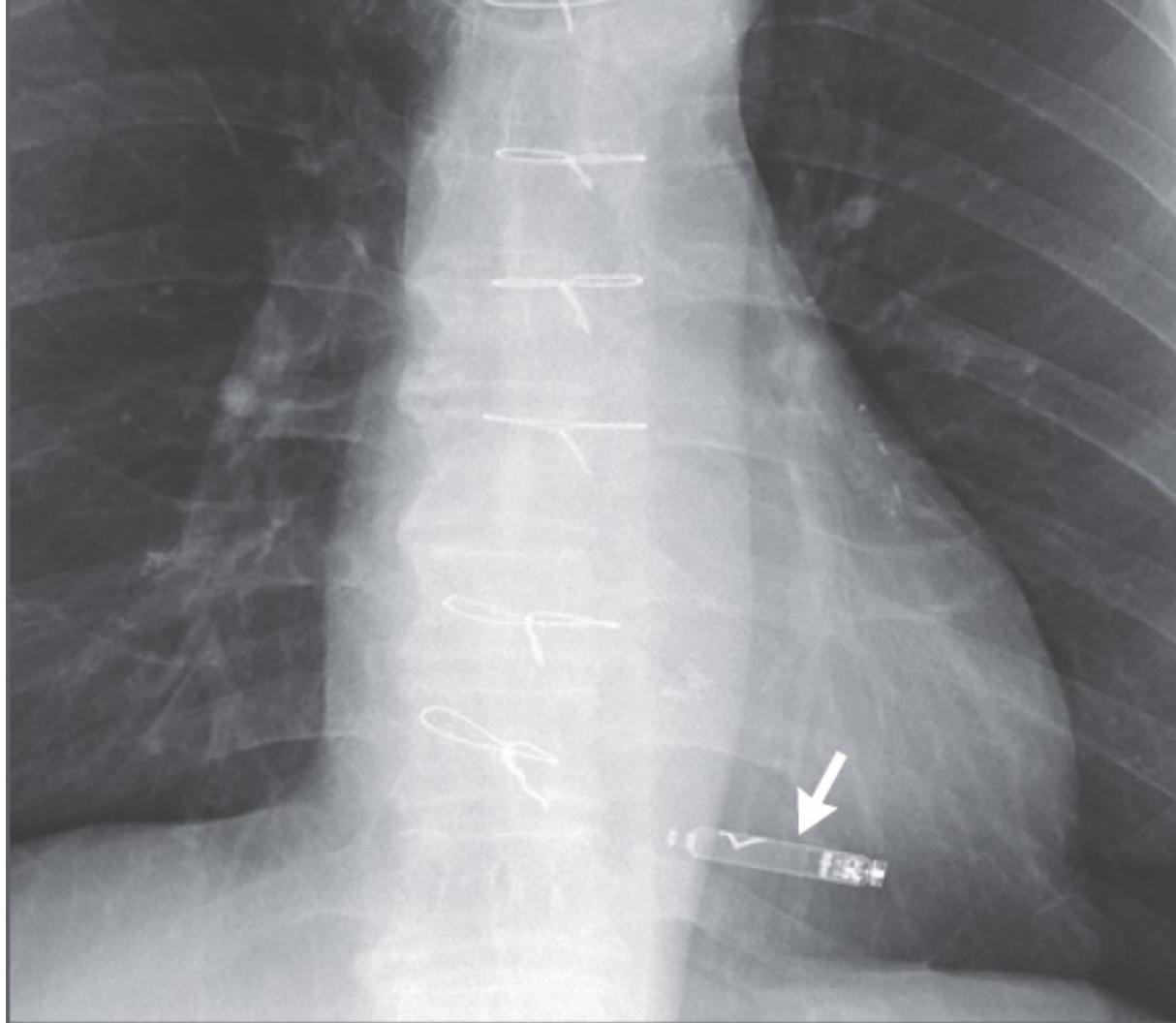
# QUIZZ – TYPE OF DEVICE ?



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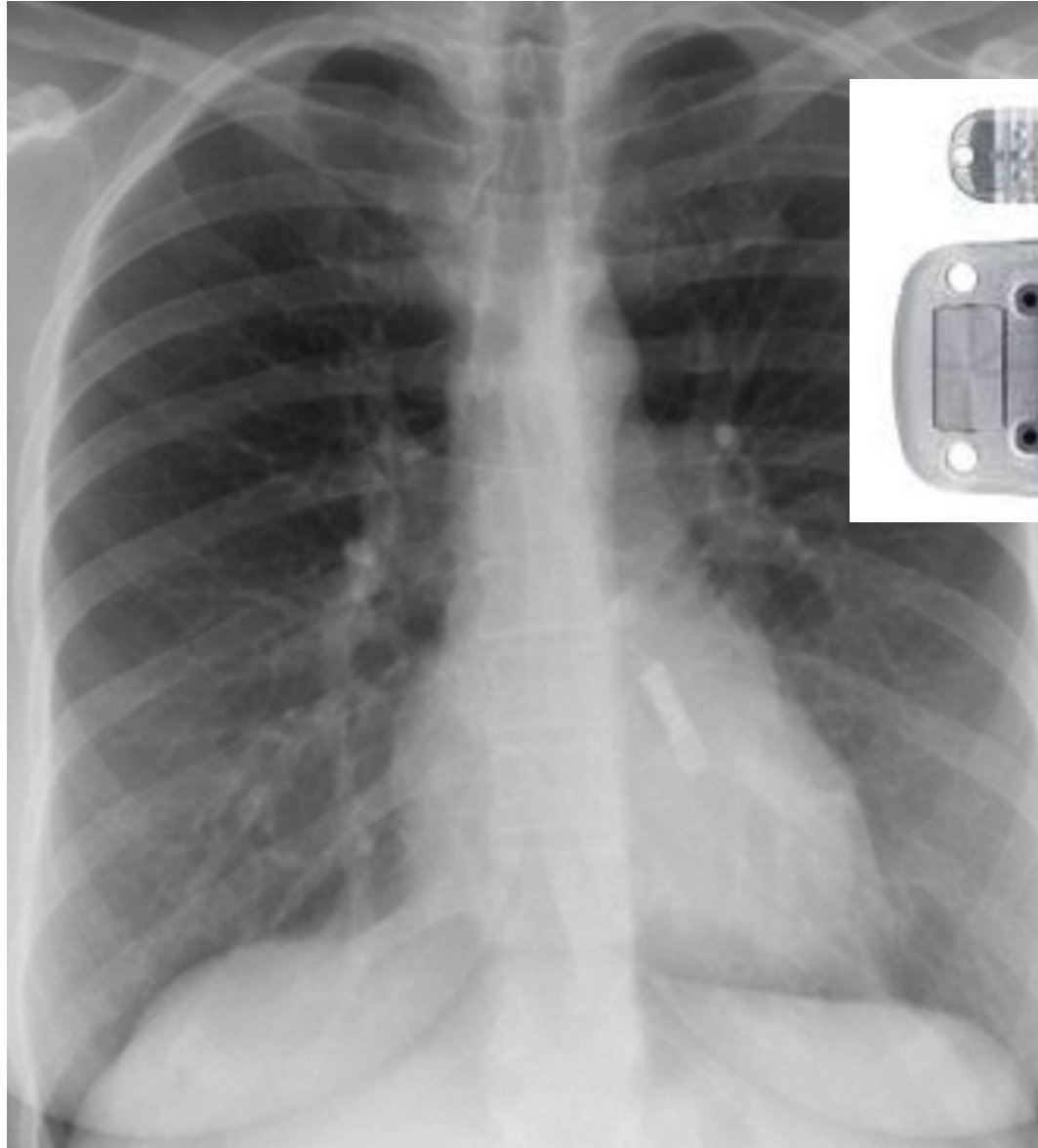
# QUIZZ – TYPE OF DEVICE ?



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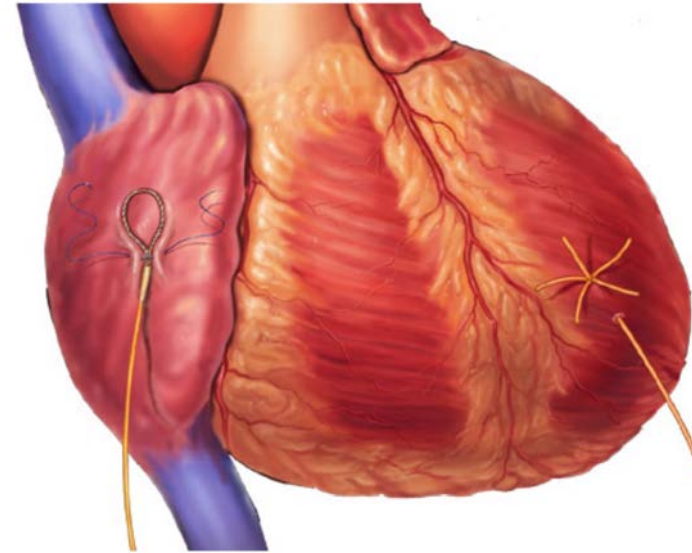
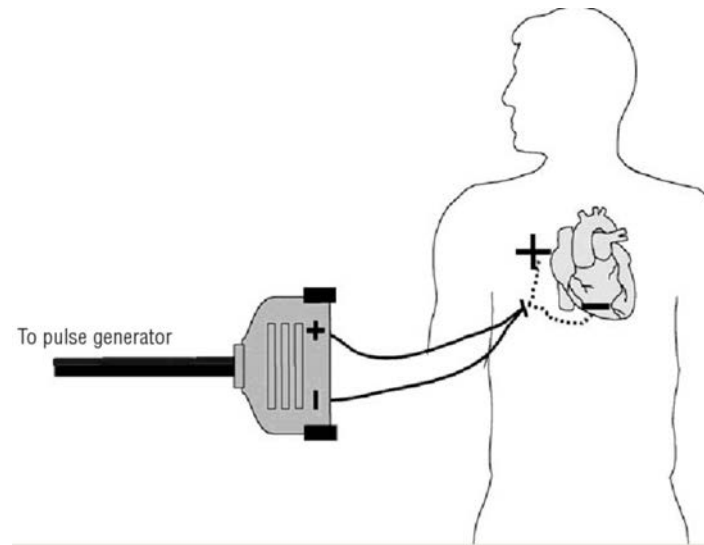
# QUIZZ – TYPE OF DEVICE ?



# BONUS — PM EXTERNES POSTOP

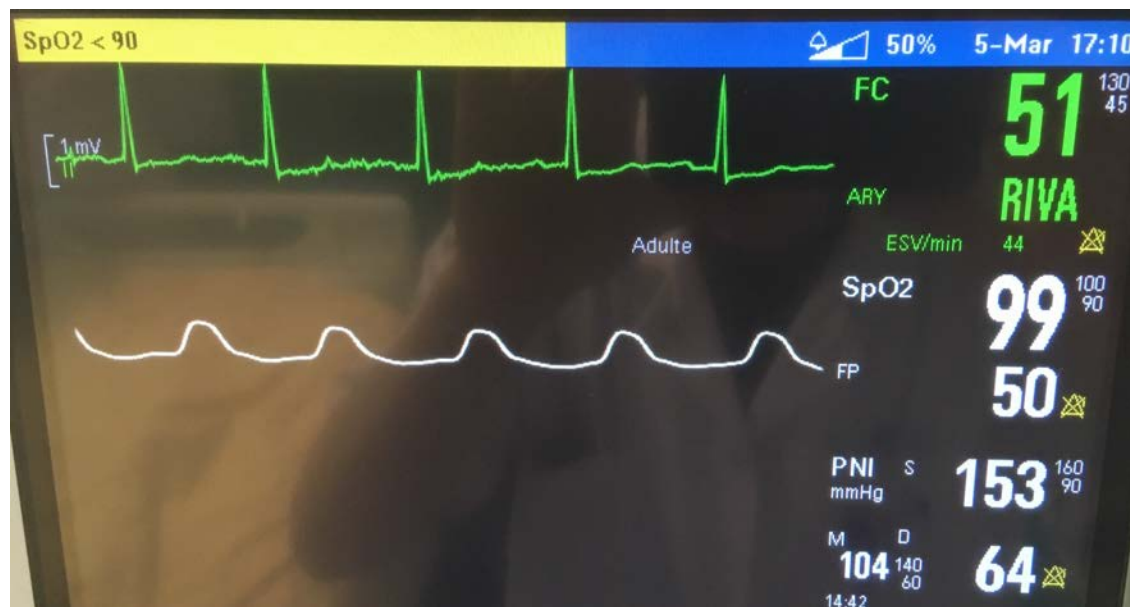


- **VVI >> DDD**



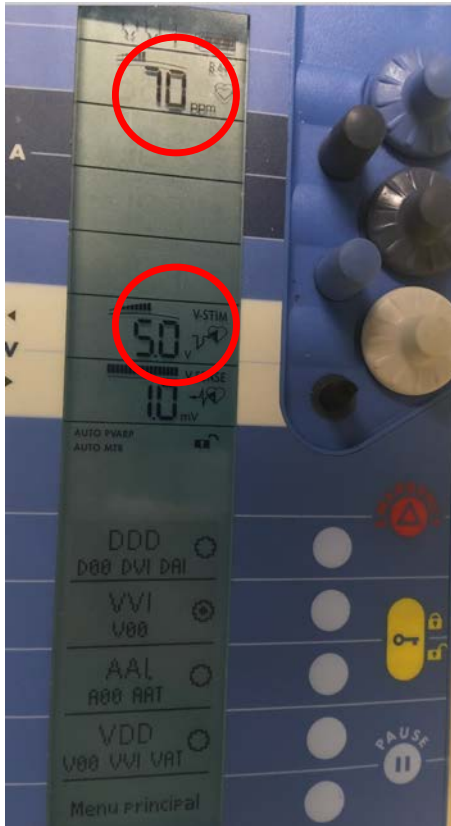


## Test du seuil



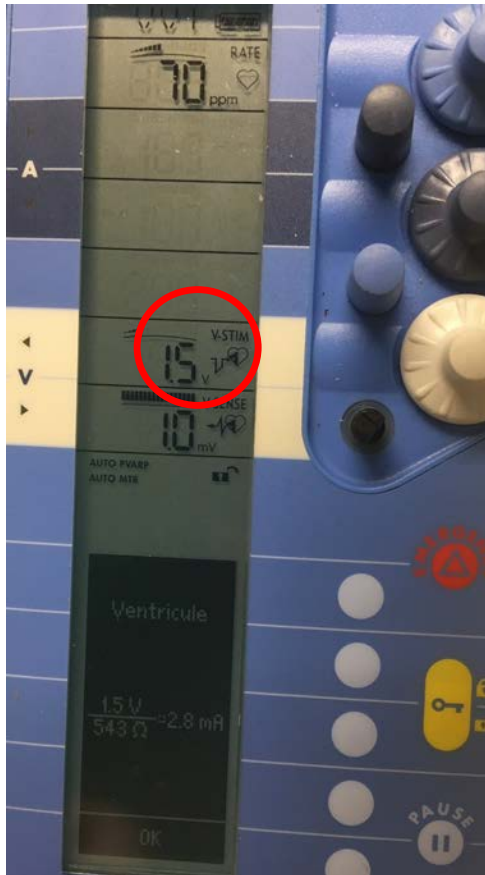
## Test du seuil

### 1/ Accélérer le PM > FC spontanée



### 2/ Baisser énergie de sortie progressivement

## Test du seuil



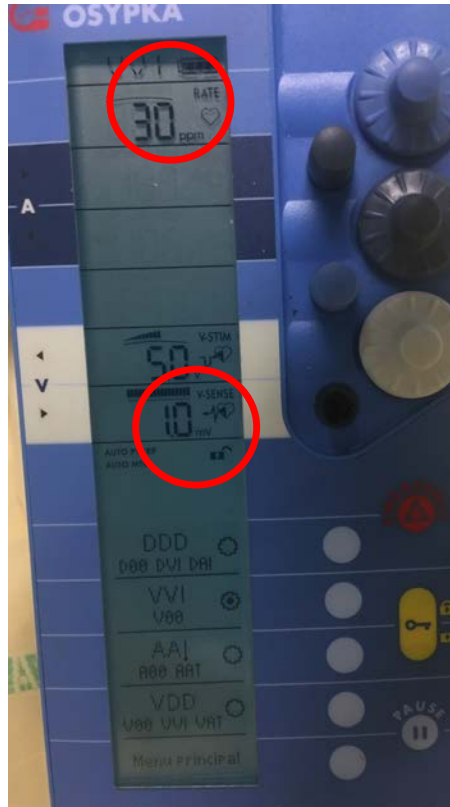
1/ Perte de capture = seuil (1,5 V)



2/ Réglage  $\geq 2x$  le seuil

## Test de sensibilité

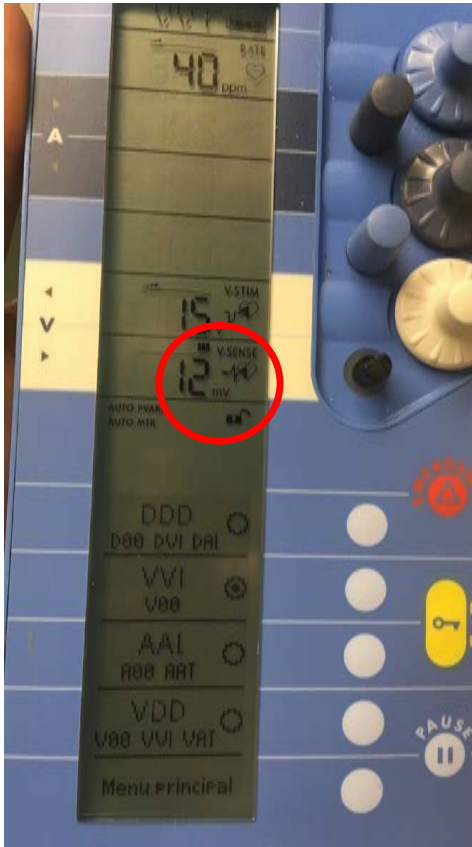
### 1/ Mettre le PM < FC spontanée



### 2/ Baisser la sensibilité progressivement

*Donc augmenter valeur, le PM n'écoute que les signaux > valeur de sensibilité réglée*

## Test de sensibilité



**1/ Spikes inappropriés = défaut d'écoute**



**2/ Régler valeur  $\leq 2$  fois la sensibilité**

## Que faire si élévation du seuil ?

**1**



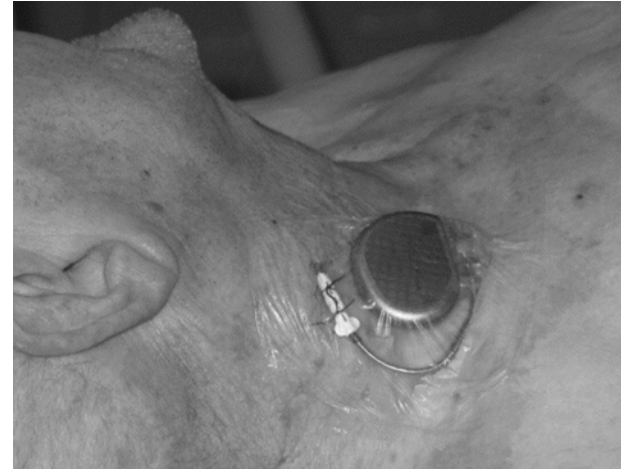
**Vérifier connectique  
Changer d'électrodes**

**2**



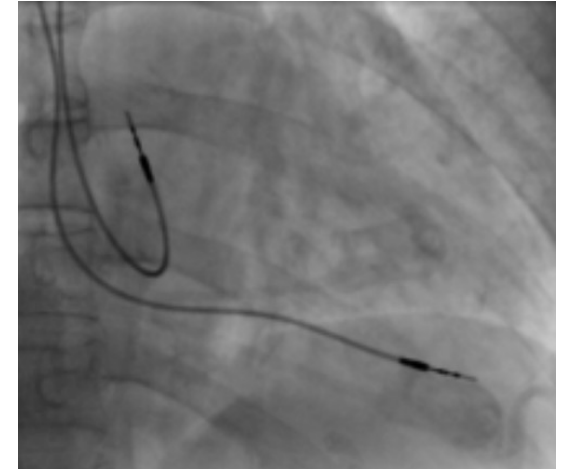
**SEES**

**3**



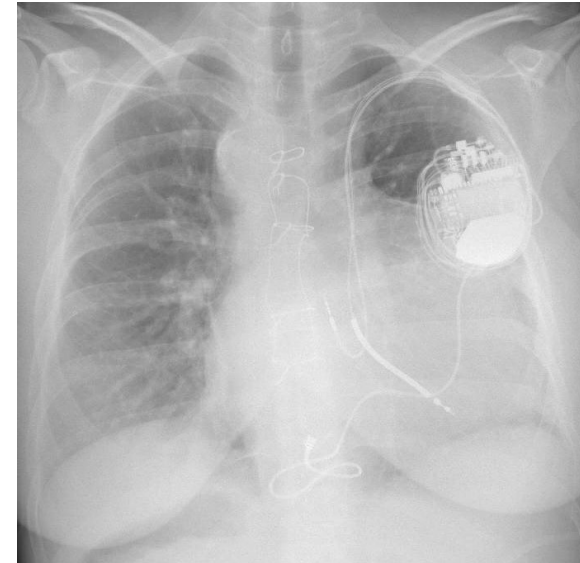
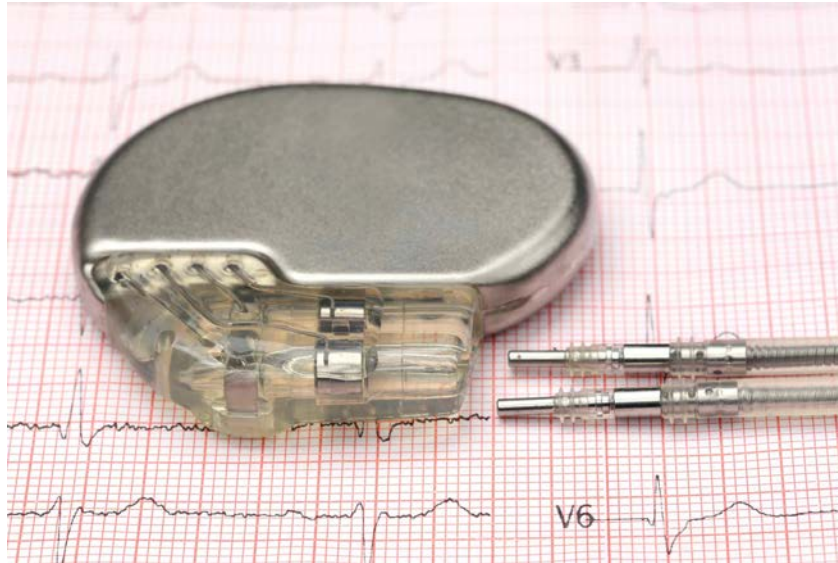
**PM vissé jugulaire**

**4**



**PM définitif**

# CARDIAC PACING IN PEDIATRIC AND CONGENITAL PATIENTS



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