15th Adult Congenital Heart Study Day

Sept 2017

Arrhythmia Management

Dr Graham Stuart

Adult Congenital Heart Study Day

Sept 2017

Arrhythmia Management

and

Sudden Cardiac Death

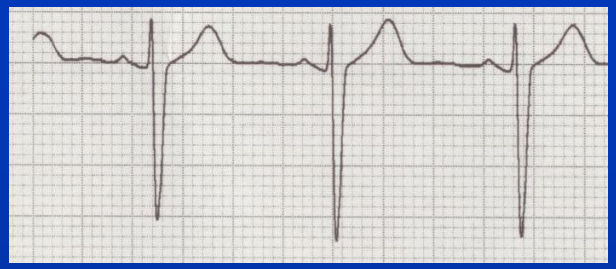
in Tetralogy of Fallot and ACHD

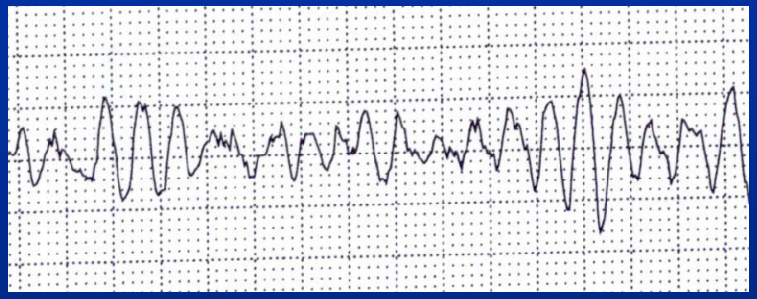
Dr Graham Stuart

Coordinated

Rhythm is fundamental ....

Meulaboh, Indonesia
Dec 26th 2004





Arrhythmias and SCD

in ToF and ACHD

- How common are arrhythmias / SCD ?
- Why do they occur?
- How do we manage them?
- The Future…….
- Thomas the Tank Engine !

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Arrhythmias in ACHD

Arrhythmias are the **main reason** for hospitalisation of GUCH patients and they are an **increasingly frequent** cause of morbidity and mortality.

ESC Guidelines for the Management of Grown Up Congenital Heart Disease
Eur Heart J 2010;31:2195-2957

Arrhythmias in ACHD

The Somerville Foundation

Help & Advice

Welcome to our Help & Advice pages where you will find lots of information for those Born with a Heart Condition.

Living with and Managing Your Heart Condition can bring up lots of questions around your Physical Health, and Emotional and Mental Health, which we hope we can help to address and answer.

www.thesf.org.uk

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Most common medical problem referred to helpline !

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

Heart Rhythm 2014;11:e102-165

Approximate Risk Estimates for arrhythmia in ACHD

Minimal

Mild

Moderate

High

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

Heart Rhythm 2014;11:e102-165

Complexity of CHD	Type of CHD	Prevalence (in CHD population)	Atrial Arrhythmia			Ventricular Arrhythmia	Other Pacing Needs		
			AT	AF	Other		SND	AV block	Dyssynchrony, heart failure
Simple	Patent ductus arteriosus	6-8%							
	Pulmonary stenosis	6-8%							
	Ventricular septal defect	30-32%							
	Secundum atrial septal defect	8-10%							
Moderate	Aortic coarctation	5-7%							
	Anomalous pulmonary venous return	0.5-2.5%							
	Atrioventricular septal defect	3-5%							
	Aortic stenosis	3-5%							
	[illegible]	[illegible]							
	Tetralogy of Fallot	8-10%							
	Primum atrial septal defect	2-3%							
Severe	Truncus arteriosus	1.5-2%							
	Pulmonary atresia	2-2.5%							
	Double outlet right ventricle	1.5-2%							
	D-transposition of the great arteries	6-7%							
	L-transposition of the great arteries	1-2%							
	Hypoplastic left heart syndrome	3-4%							
	Other (heterotaxy, other single ventricles)	7-10%							

Postoperative arrhythmias in adults with congenital heart disease: Incidence and risk factors ☆

Z. Koyak [a,1], R.C.A. Achterbergh [b,1], J.R. de Groot [a], F. Berger [c], D.R. Koolbergen [a,b], B.J. Bouma [a], W.K. Lagrand [d], M.G. Hazekamp [b], N.A. Blom [b], B.J.M. Mulder [a,*]

International Journal of Cardiology 169 (2013) 139–144

- Multicentre retrospective
  Berlin / Amsterdam
- Jan 2009 – Dec 2011
- 419 patients
  - 38 +/- 14 yrs 55% male

Excluded

- Transplantation
- Age < 18yrs

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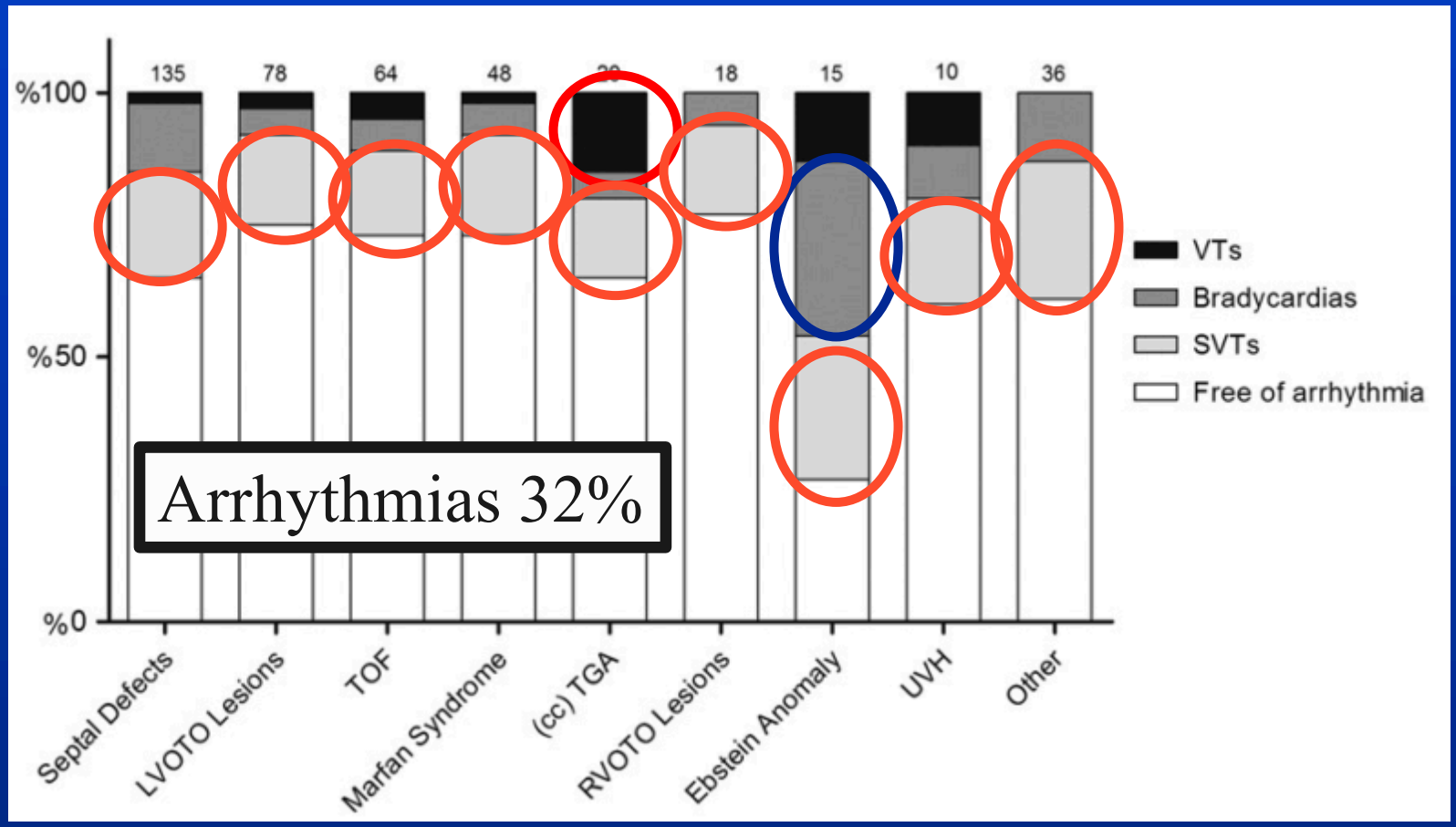

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Main Risk factors

- Age > 40yrs at surgery OR 2.5 1.4-4.6
- NYHA class>II OR 2.4 1.2-4.7
- subPulm AVVR OR 2.8 1.2-6.7
- Byass time OR 1.3 per 60 min increase

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- Post operative arrhythmias relatively common
- Older, symptomatic patients with significant valve disease
- Serious sequelae in 13%
  pacemaker implant 5%, heart failure 5%, death 3%!

Learning point !

Pay attention

!

Learning point !

Cardiac arrhythmias are ***very*** common in ACHD patients

Older, sicker, post op….

Arrhythmias and SCD

in ToF and ACHD

- How common are arrhythmias / SCD ?
- Why do they occur?
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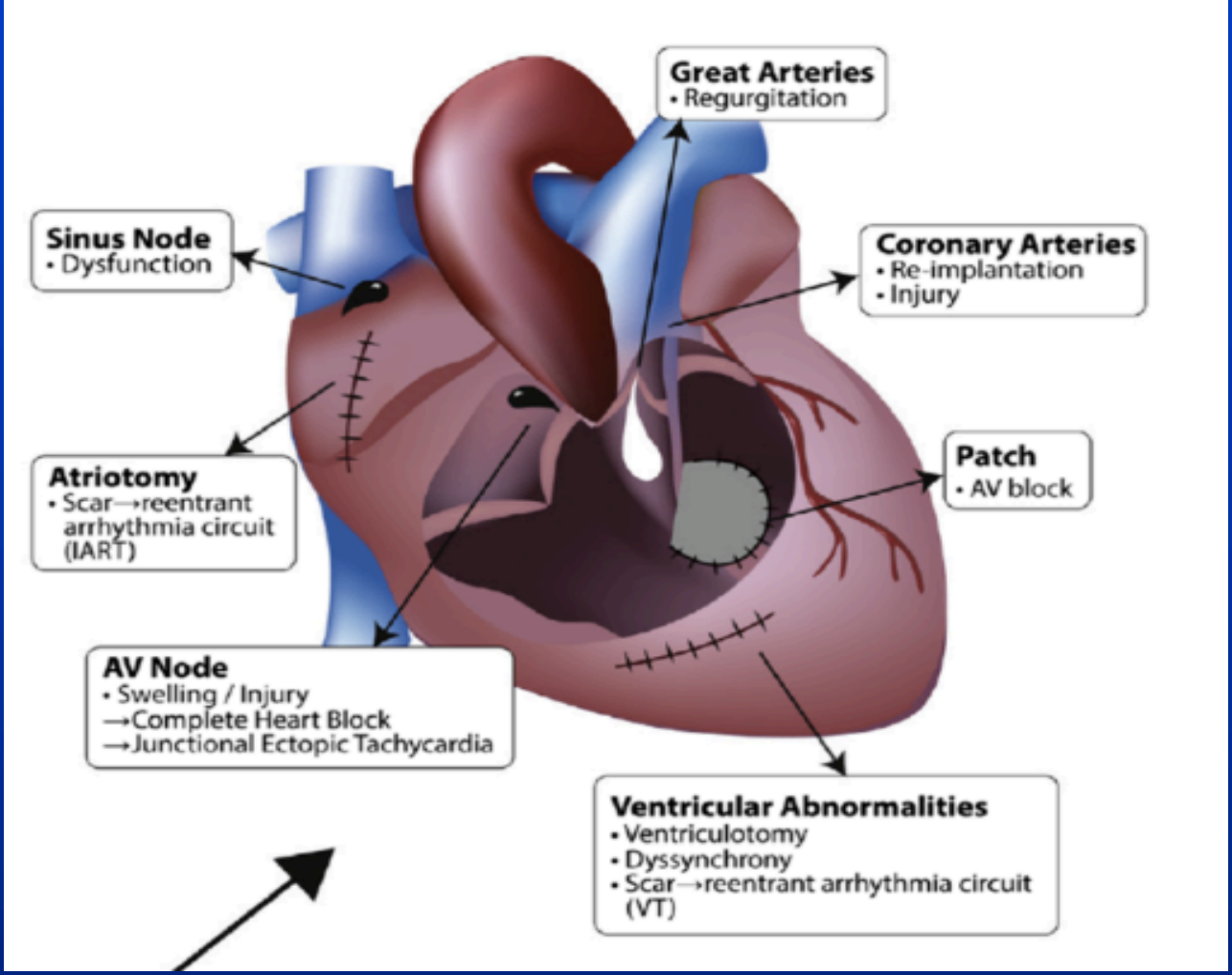
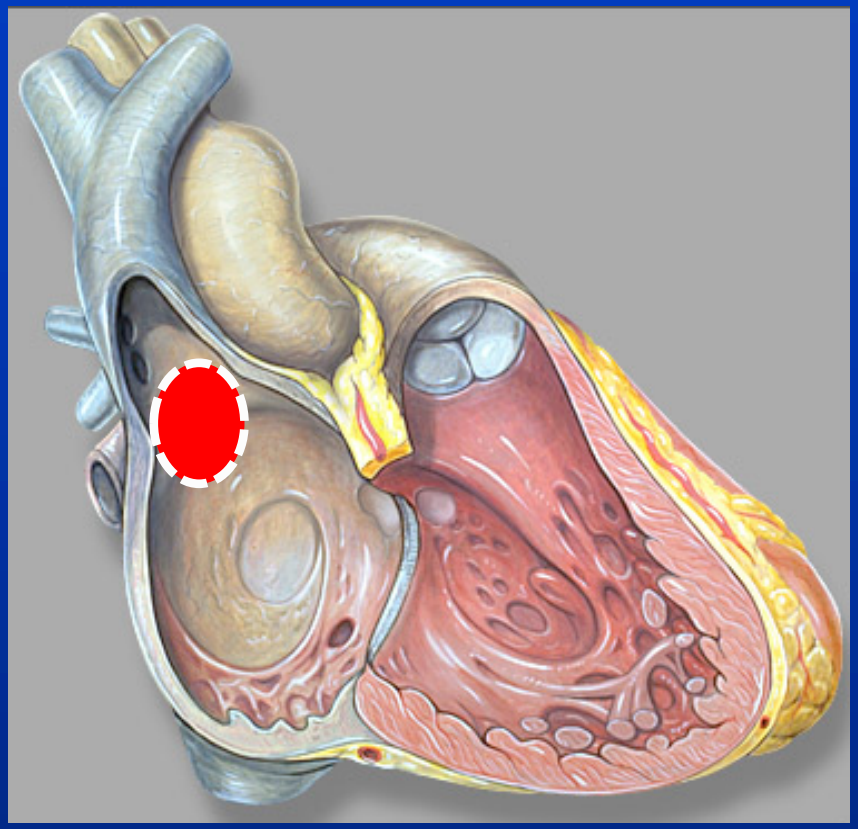
Why are arrhythmias common?

Escudero et al Can J Cardiol 2013;29(7):821-9

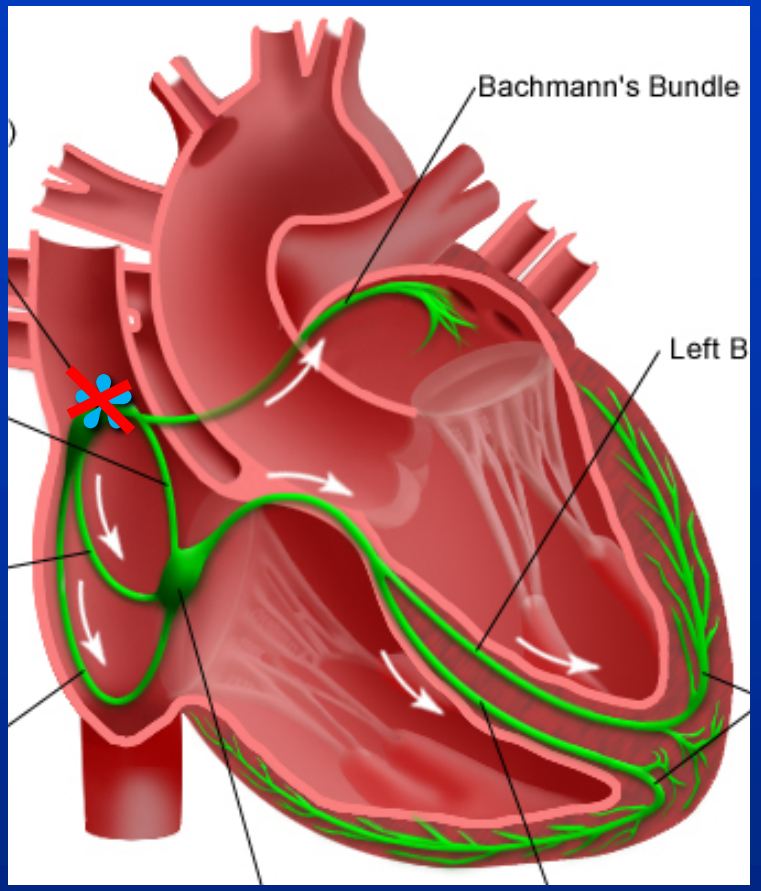


Arrhythmia mechanisms

-bradycardia



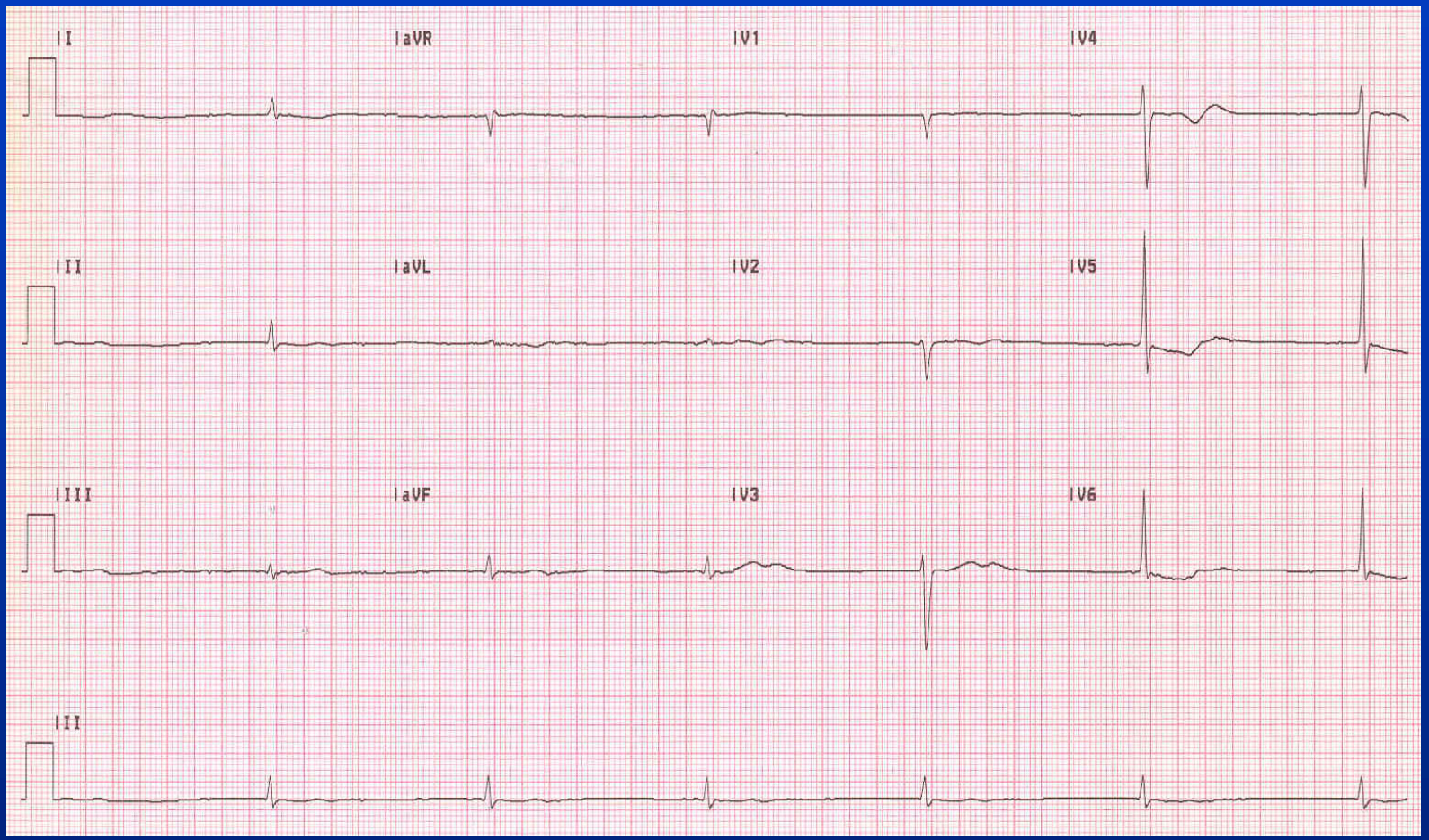
sinus venosus ASD



Arrhythmia mechanisms

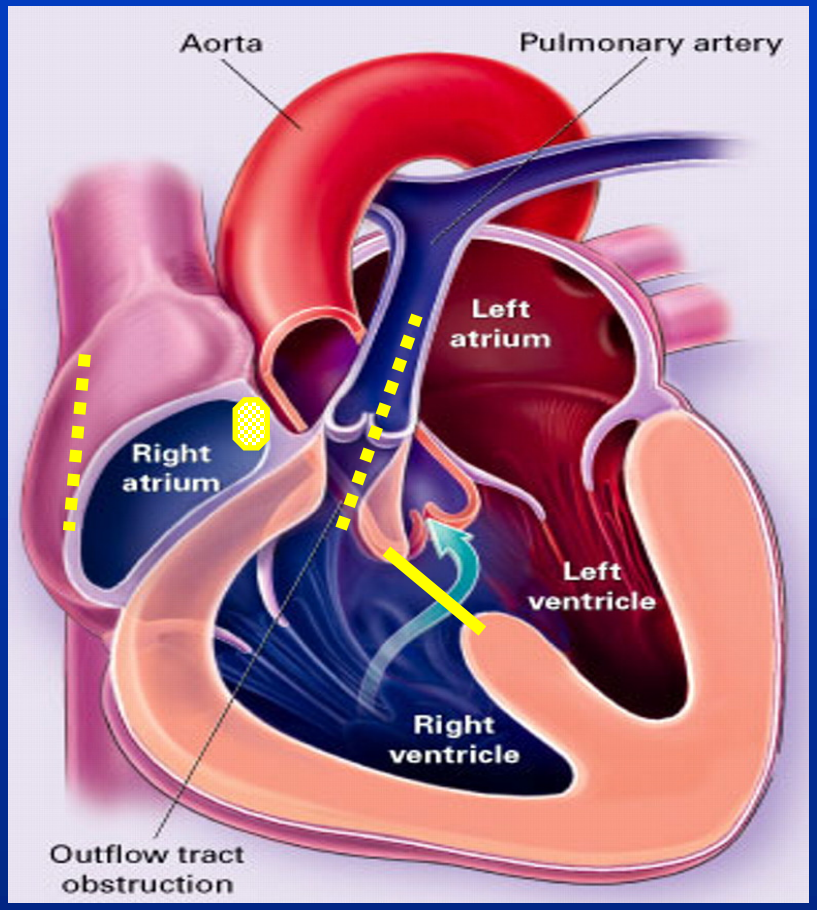
-bradycardia

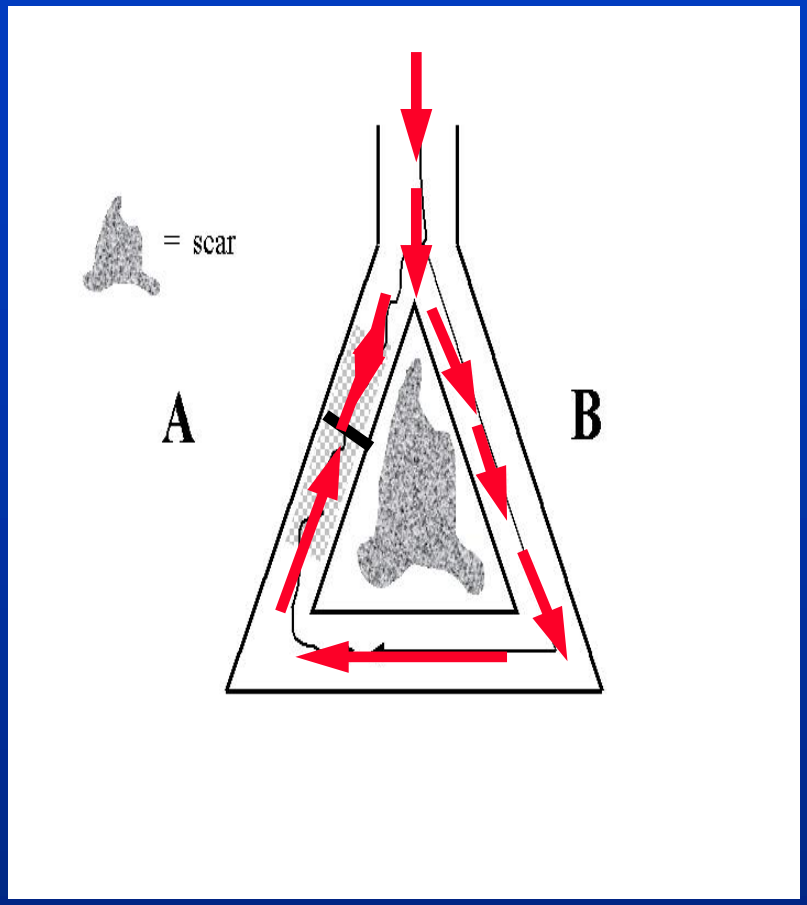
sinus venosus ASD - post operative



Scar related arrhythmias

Tetralogy of Fallot

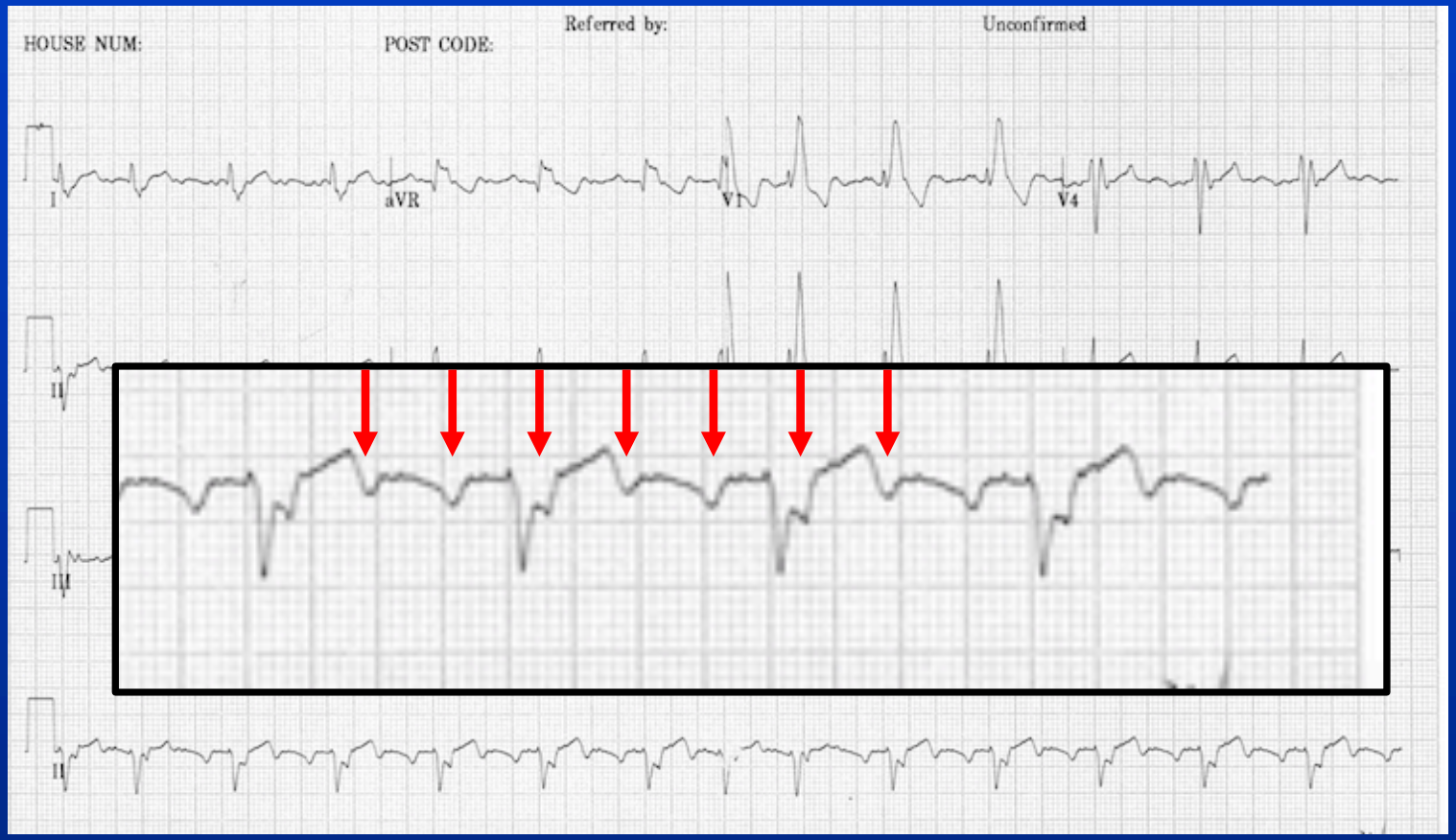




Surgical scars

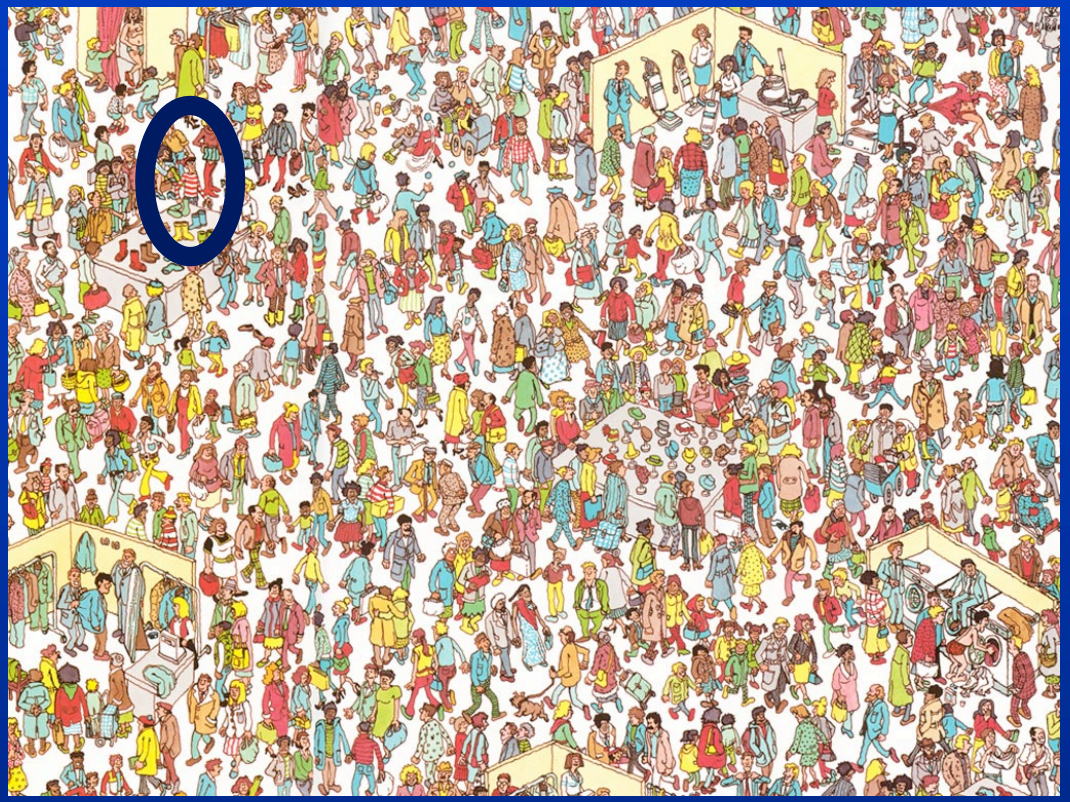
Atrial tachycardia

51yr old man with ToF



Pattern recognition

in ACHD wide complex tachycardia

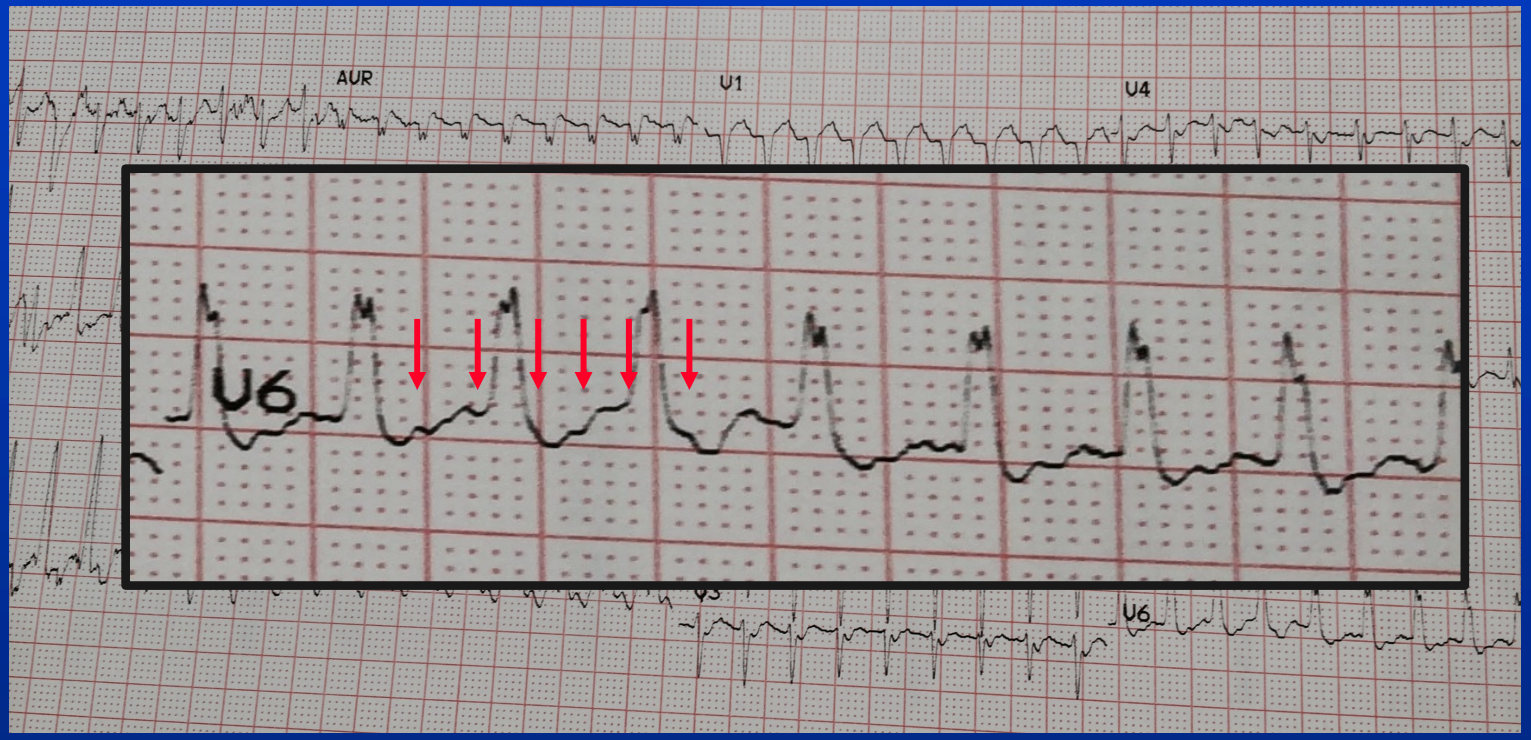


Where is the p wave?

Wide Complex Tachycardia

SVT with bundle branch block

17yr old girl. Palpitations post PAVSD/MVR/Sub AS resection



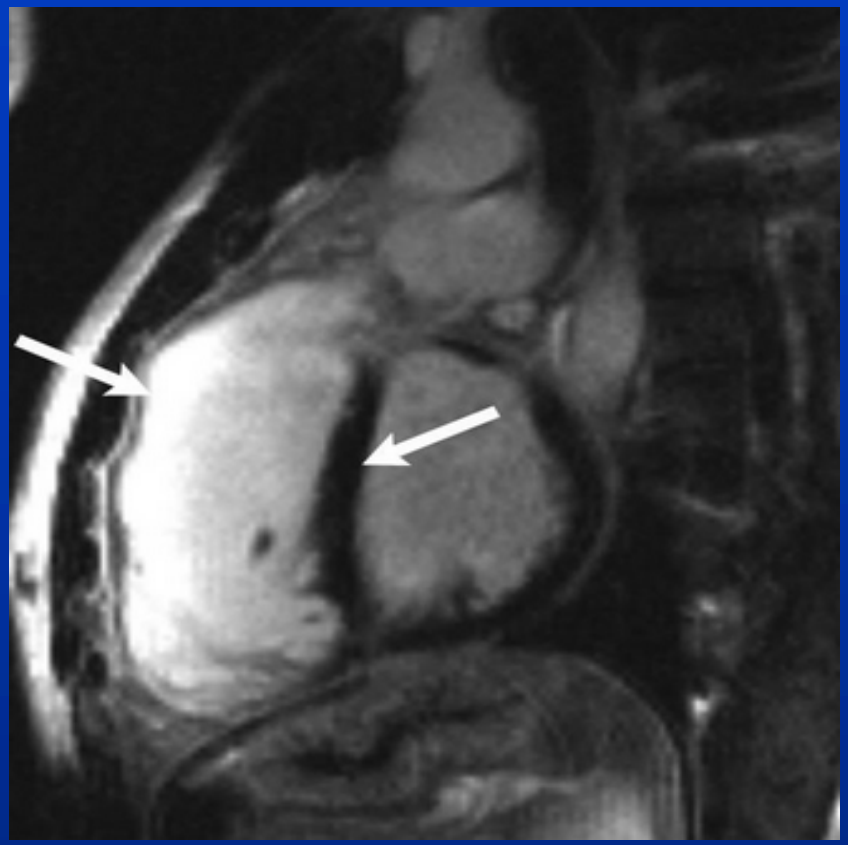
Learning point !

Myocardial scar
is a common cause of
ACHD arrhythmias

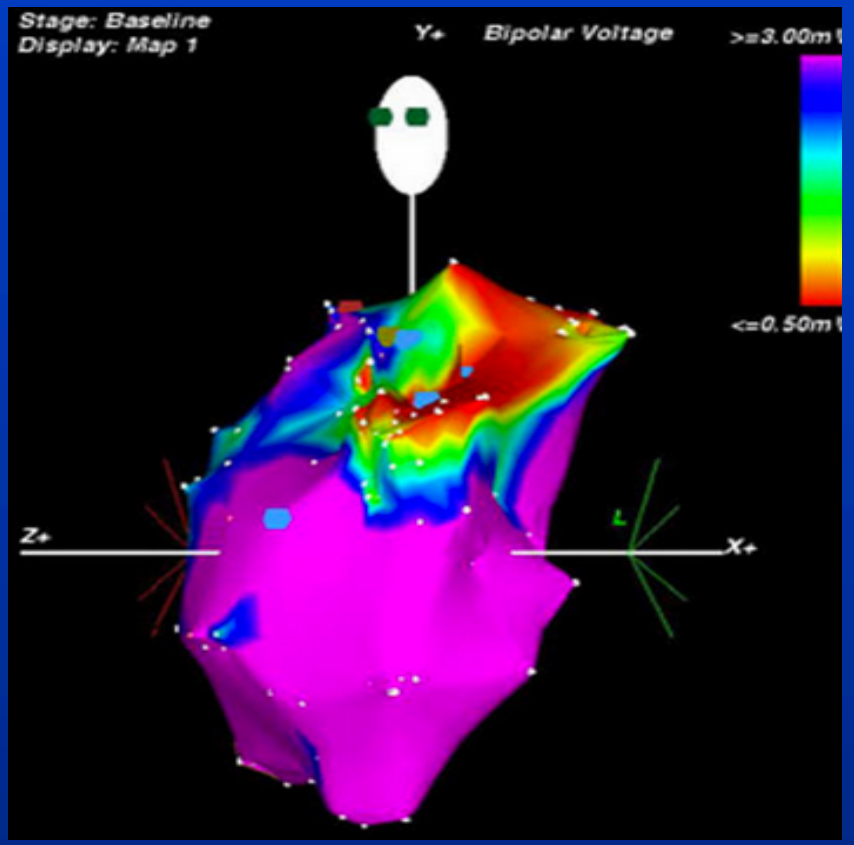
Scar ++ = arrhythmias +++

Tetralogy of Fallot

arrhythmias: scar

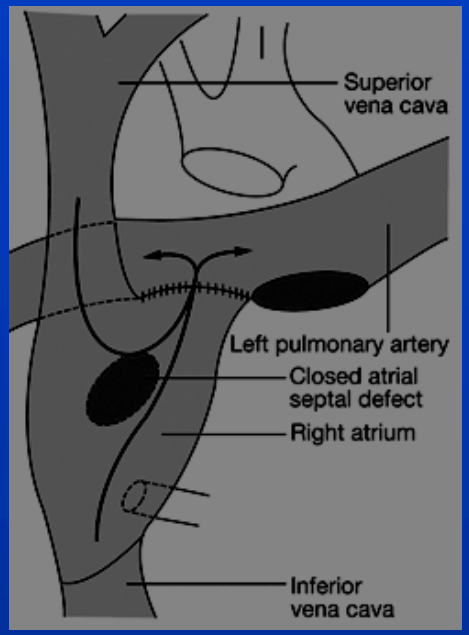


MRI : late gad + RVOT

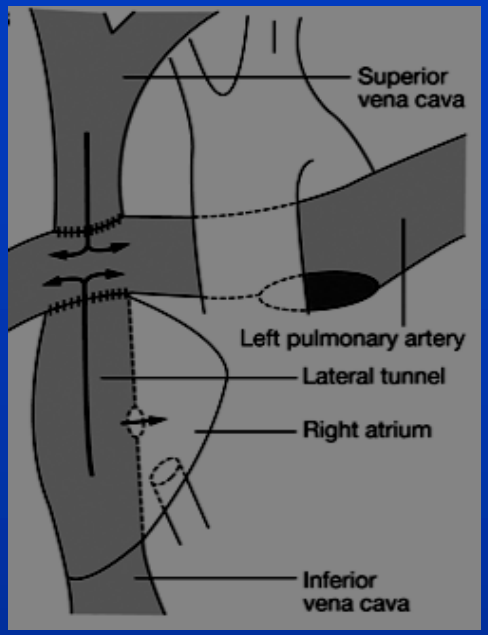


CARTO: RVOT reentry

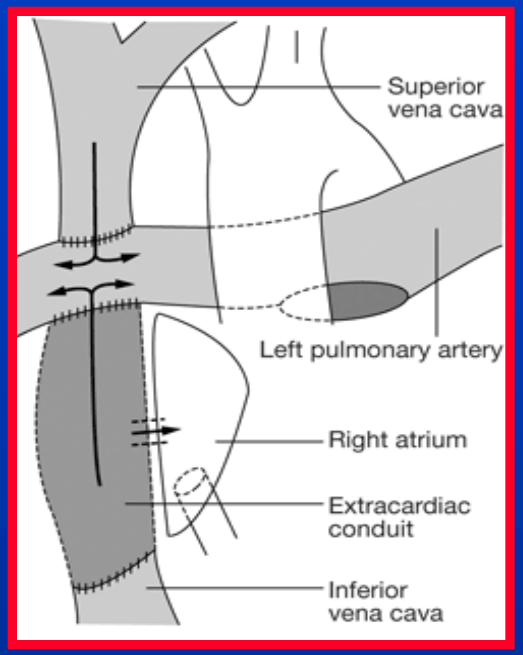
Types of Fontan Procedure



Classical atriopulmonary circuit

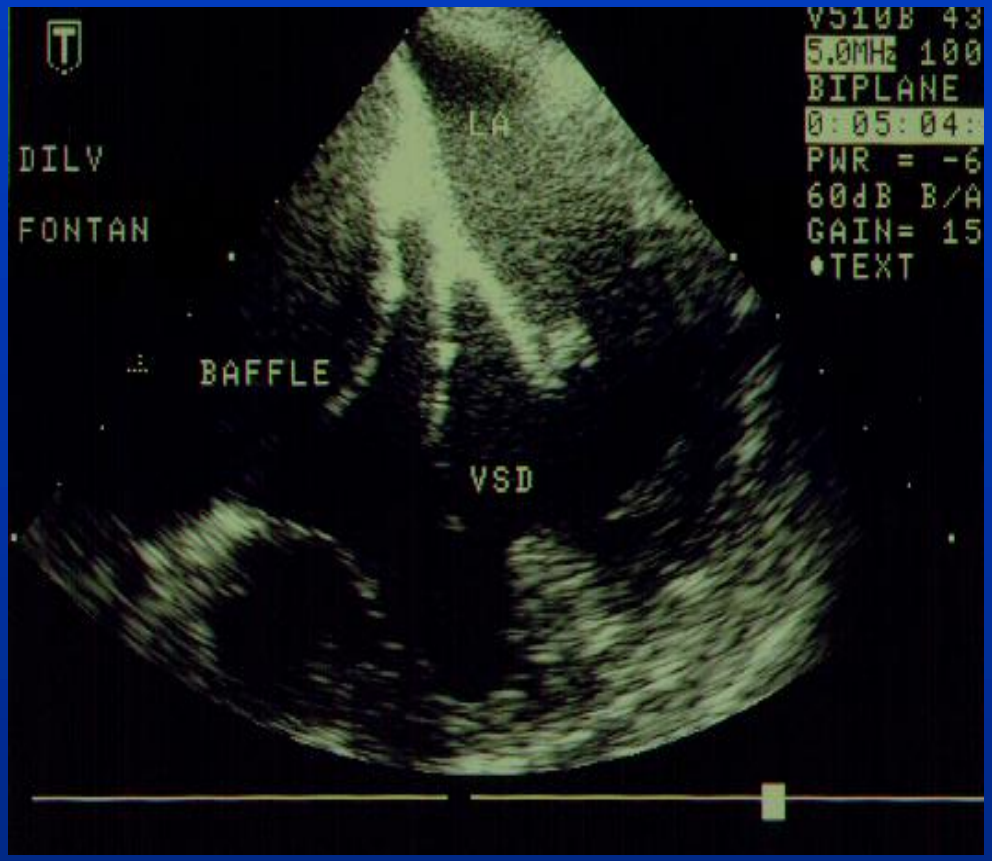


Lateral tunnel TCPC



Extracardiac conduit

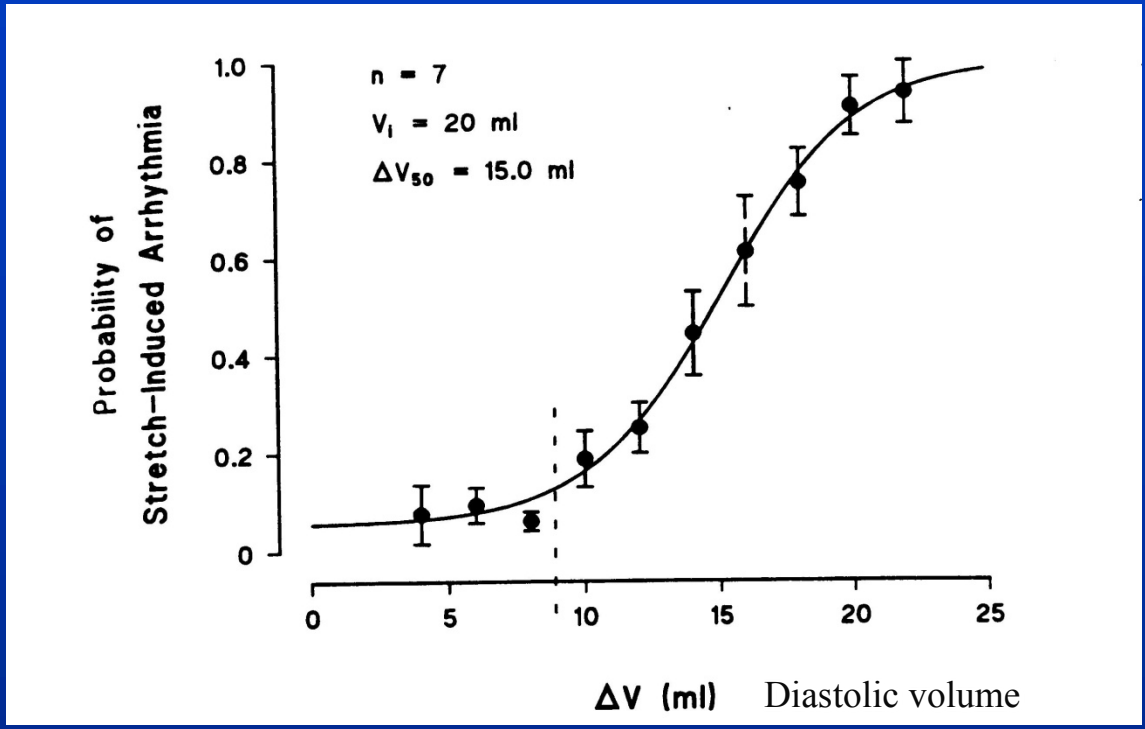
Electromechanical Failure



Dilated stretched right atrium

Stretch-induced arrhythmias

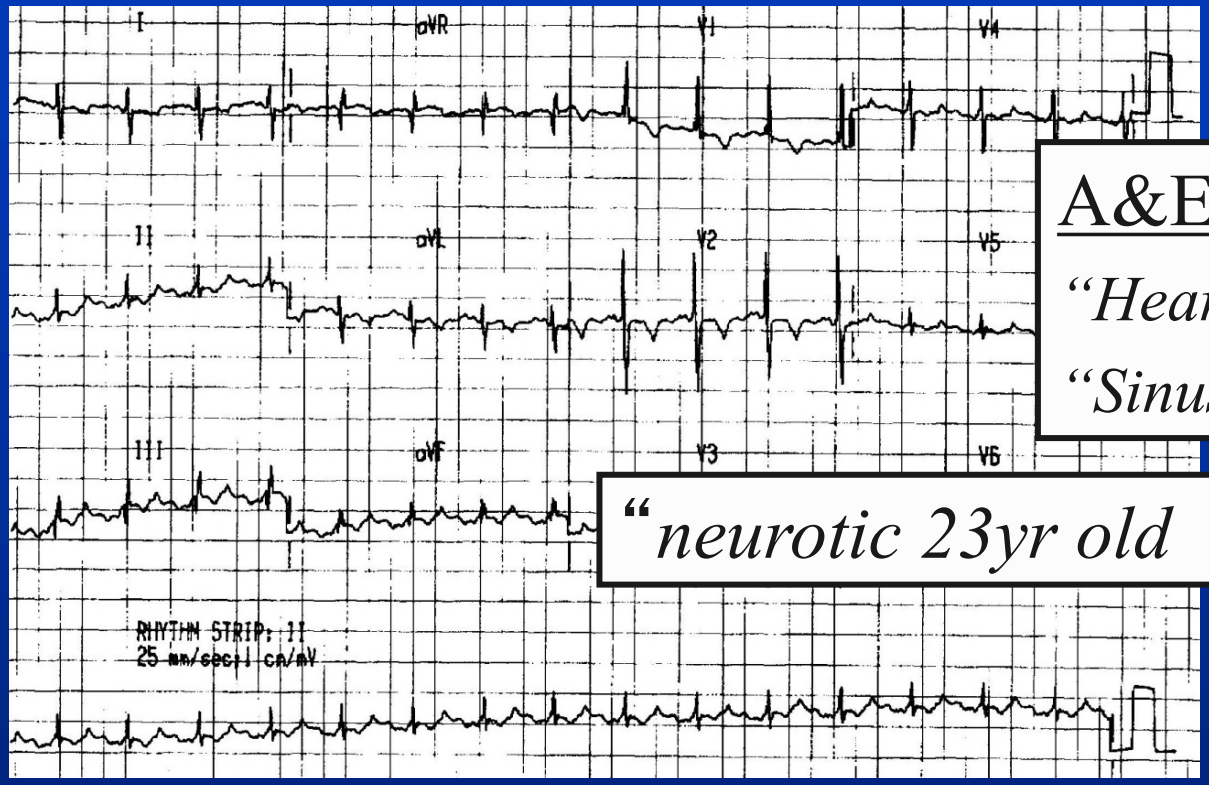
Hansen DE et al Stretch-induced arrhythmias in the isolated canine ventricle
Circulation 1990;81:1094-1105



Arrhythmias

substrates and patterns in ACHD

23yr teacher Fontan (DILV PA) = palpitations



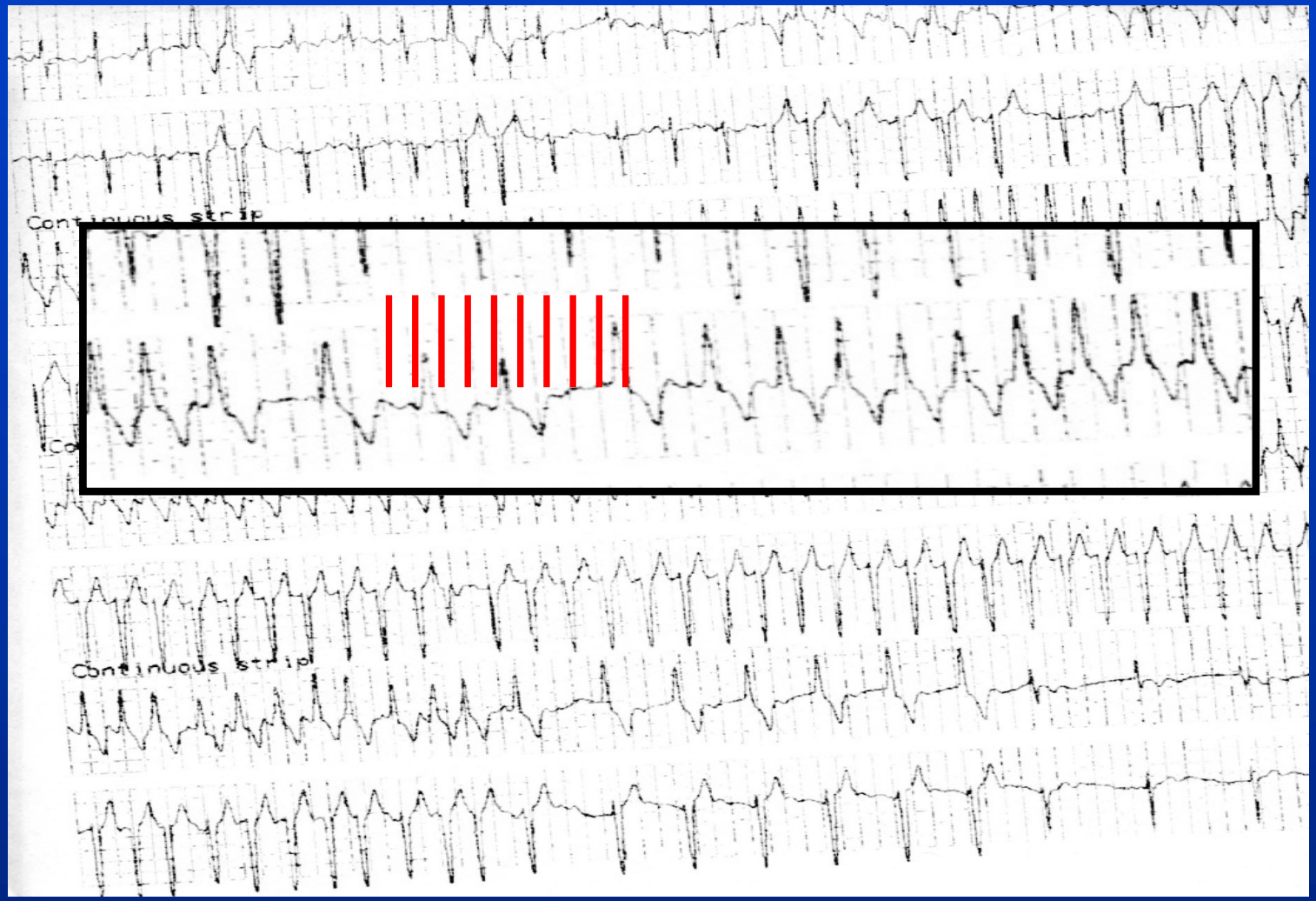
A&E Record

"Heart op as child"

"Sinus rhythm"

"neurotic 23yr old woman"

The Fontan Flutter



Learning point !

Mechanical events can have electrical consequences !

= consider haemodynamics

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in ToF and ACHD

- How common are arrhythmias / SCD ?
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- The Future……
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Arrhythmias and SCD

management in ToF and ACHD

- Lifestyle
- Drugs
- Devices
- Ablation
- The surgical role

Arrhythmias and SCD

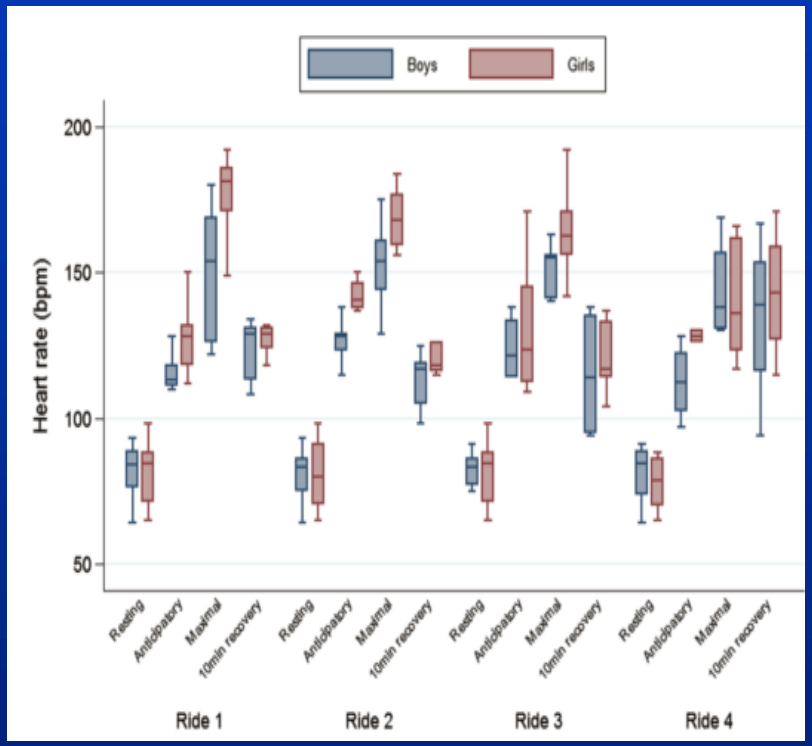
management ***lifestyle***

- Risky activities
- Drugs and alcohol…….
- Exercise

Arrhythmias and SCD

management ***lifestyle***

- Risky activities



Pieles et al Pediatr Cardiol (2017) 38:15–19

Arrhythmias and SCD

management ***lifestyle***

- Risky activities
- Drugs and alcohol…….

J Am Coll Cardiol 2016:68(23):2567-76

Alcohol and Atrial Fibrillation

A Sobering Review

Aleksandr Voskoboinik, MBBS,[a,b,c] Sandeep Prabhu, MBBS,[a,b,c] Liang-han Ling, MBBS, PhD,[a,b,c] Jonathan M. Kalman, MBBS, PhD,[c,d] Peter M. Kistler, MBBS, PhD[a,b,c]

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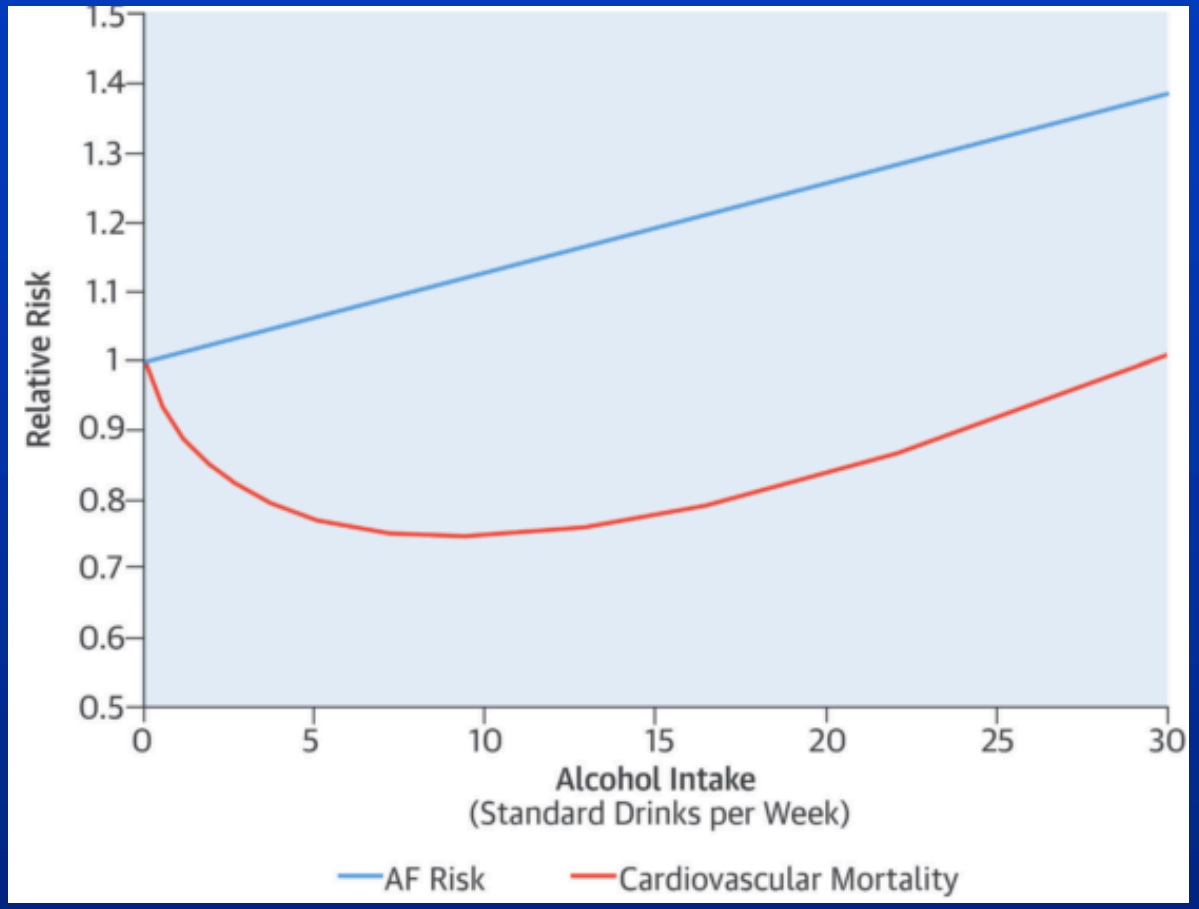


Arrhythmias and SCD

management ***lifestyle***

- Risky activities
- Drugs and alcohol……

Caffe[illegible]duced cardiac arrhyt[illegible] unrecognised danger o[illegible]od products

Marianne E Canno[illegible]ooke and James S McCarthy

Med J Aust 2001; 174 (10): 52[illegible]

Patient Died!

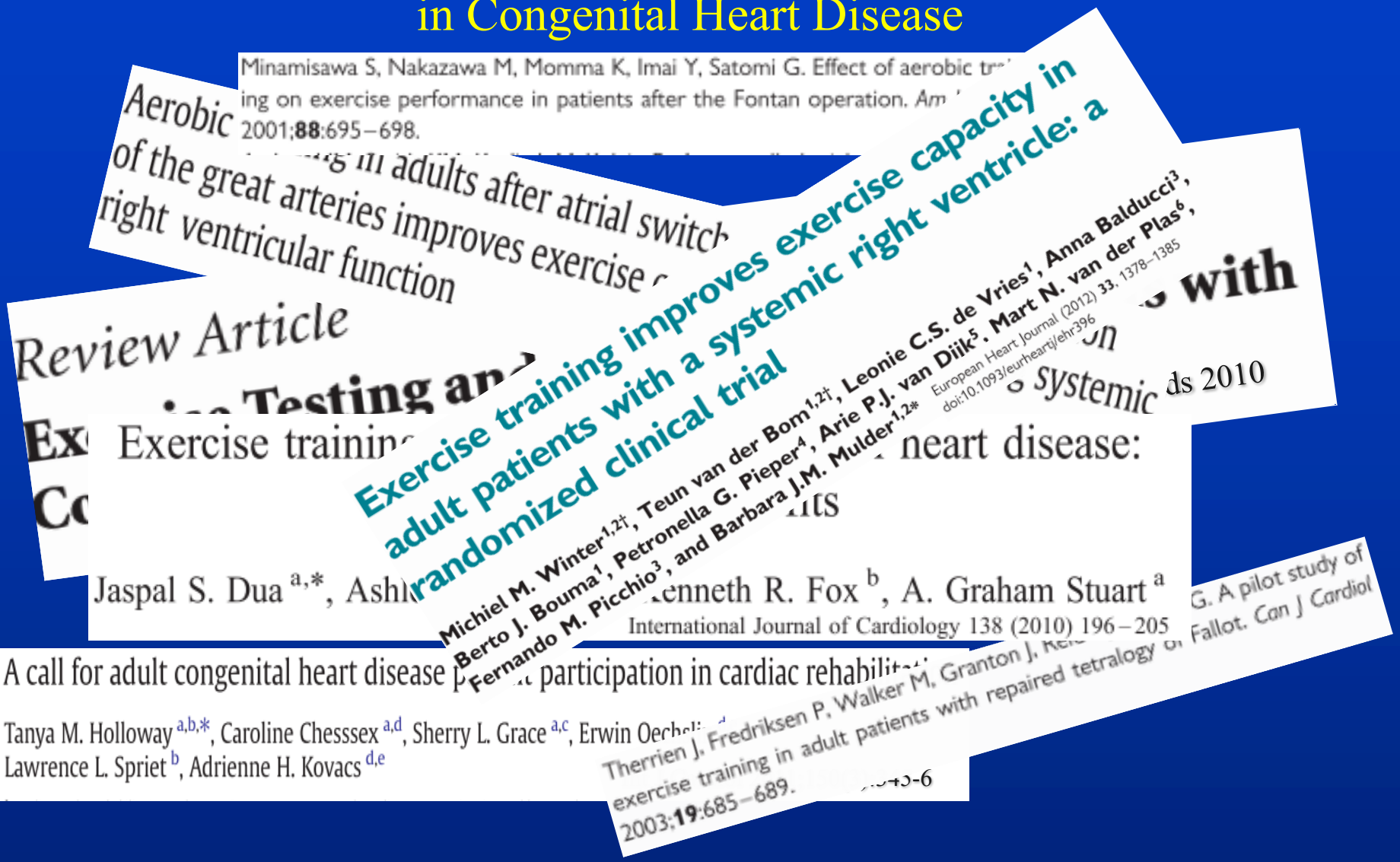
Arrhythmias and SCD

management ***lifestyle***

- Risky activities
- Drugs and alcohol…….
- Exercise

Exercise as therapy

in Congenital Heart Disease



Exercise and CVS

Congenital Heart Disease

Exercise prescription in ACHD

AHA Scientific Statement *April 2013*

Promotion of Physical Activity in Children and Adults with Congenital Heart Disease

Longmuir et al Circulation 2013 DOI:10.1161/CIR.0b013e318293688F

"counselling to encourage daily participation in appropriate physical activity should be a core component of every patient encounter…."

Physical activity in adolescents and adults with congenital heart defects; individualized exercise prescription†

Werner Budts[1,2*], Mats Börjesson[3], Massimo Chessa[4], Frank van Buuren[5], Pedro Trigo Trindade[6], Domenico Corrado[7], Hein Heidbuchel[1,2], Gary Webb[8], Johan Holm[9], and Michael Papadakis[10]

European Heart Journal Advance Access published November 7, 2013

	A	B	C	D	E
1. Ventricles	No systolic dysfunction No hypertrophy No pressure load No volume load	No systolic dysfunction No hypertrophy Mild pressure load Mild volume load	Mild systolic dysfunction Mild hypertrophy Single ventricle physiology Systemic right ventricle	Moderate systolic dysfunction Moderate hypertrophy Moderate pressure load	Severe systolic dysfunction Severe hypertrophy Severe pressure load Moderate/severe volume load
2. Pulmonary artery pressure	Low pulmonary artery pressure	Low pulmonary artery pressure	Mildly elevated pulmonary artery pressure		Moderately/severely elevated pulmonary artery pressure
3. Aorta	No/mild dilatation	Moderate dilatation	Severe dilatation	Dilatation approaching indication for repair	
4. Arrhythmia	No arrhythmia	No arrhythmia	Mild arrhythmic burden Non-malignant arrhythmia		Significant arrhythmic burden Malignant arrhythmia
5. Saturation at rest/during exercise	No central cyanosis	No central cyanosis	No central cyanosis	Central cyanosis	
Static component of sport	When all applicable Up to high static	When at least one applicable Up to moderate static			When at least one applicable Low static
Relative intensity of sport	*HIGH INTENSITY* *RPE Borg scale: 15-17* *Training HR: 75%-90% of achieved MHR during CPET*	*MODERATE INTENSITY* *RPE Borg scale: 13-14* *Training HR: 60%-75% of achieved MHR during CPET*			*LOW INTENSITY* *RPE Borg scale: 11-12* *Training HR: <60% of achieved MHR during CPET*

Learning point !

Think about and

specifically prescribe

exercise

= consider exercise stress test

Arrhythmias and SCD

management in ToF and ACHD

- Lifestyle
- Drugs
- Devices
- Ablation
- The surgical role

Arrhythmias and SCD

management in ToF and ACHD

- Lifestyle
- Drugs
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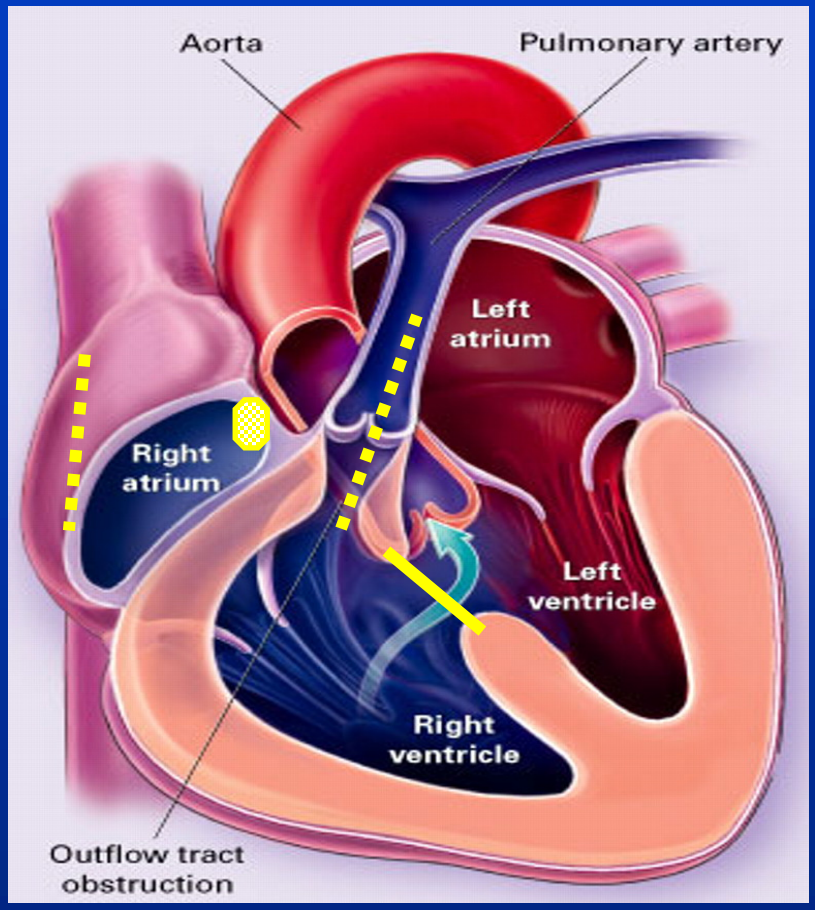
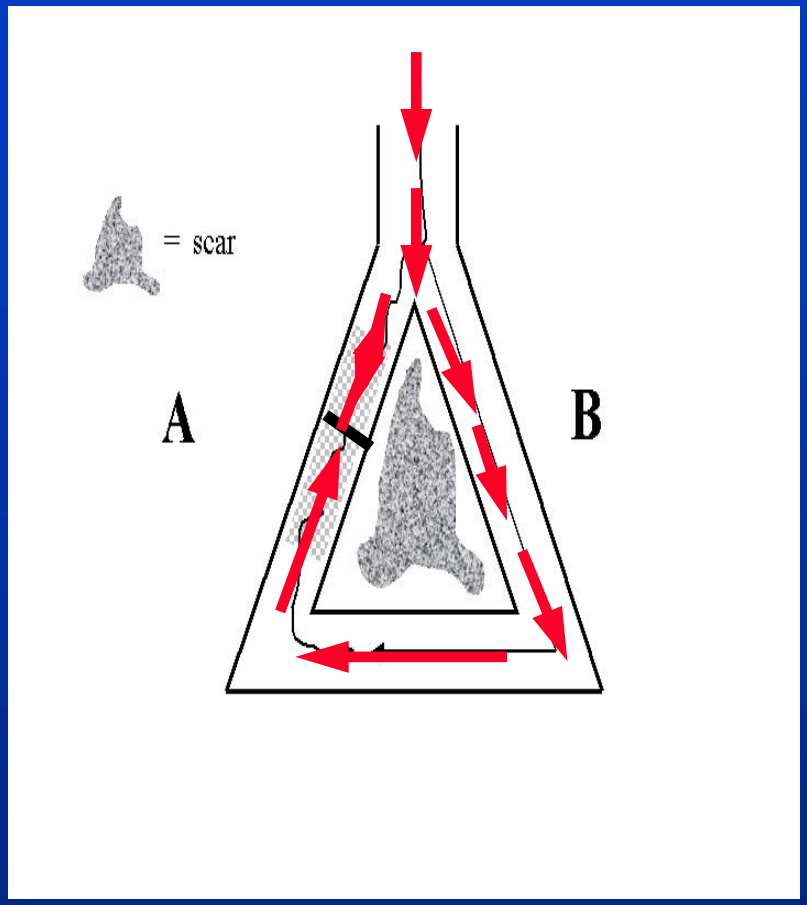
- The surgical role

Arrhythmias in ACHD

- **How might antiarrhythmic drugs work ?**
  - reducing ectopic activity ?
  - changing refractory period of A/B ?
  - changing speed of conduction of A/B ?

Scar related arrhythmias





Surgical scars

iarrhythmic Drugs

ACHD

- B
- Amiod
- Digoxin
- Others…….sotolol, ibutilide, one etc

No RCT of antiarrhythmics in ACHD!

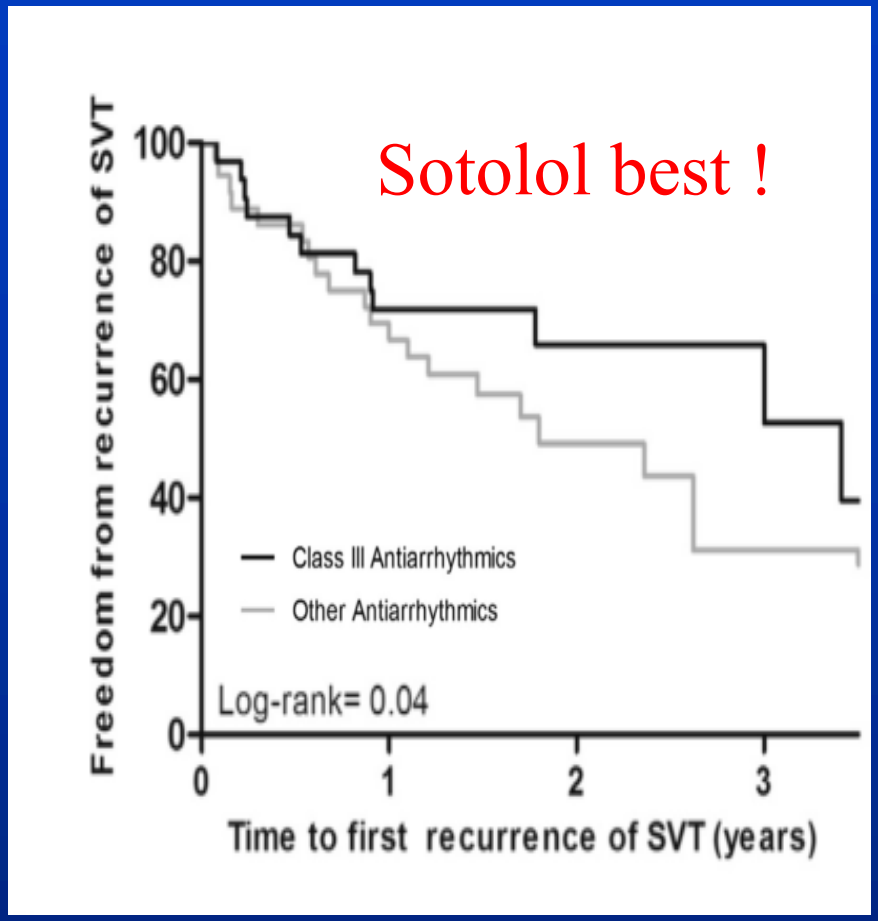
Efficacy of Antiarrhythmic Drugs in Adults With Congenital Heart Disease and Supraventricular Tachycardias

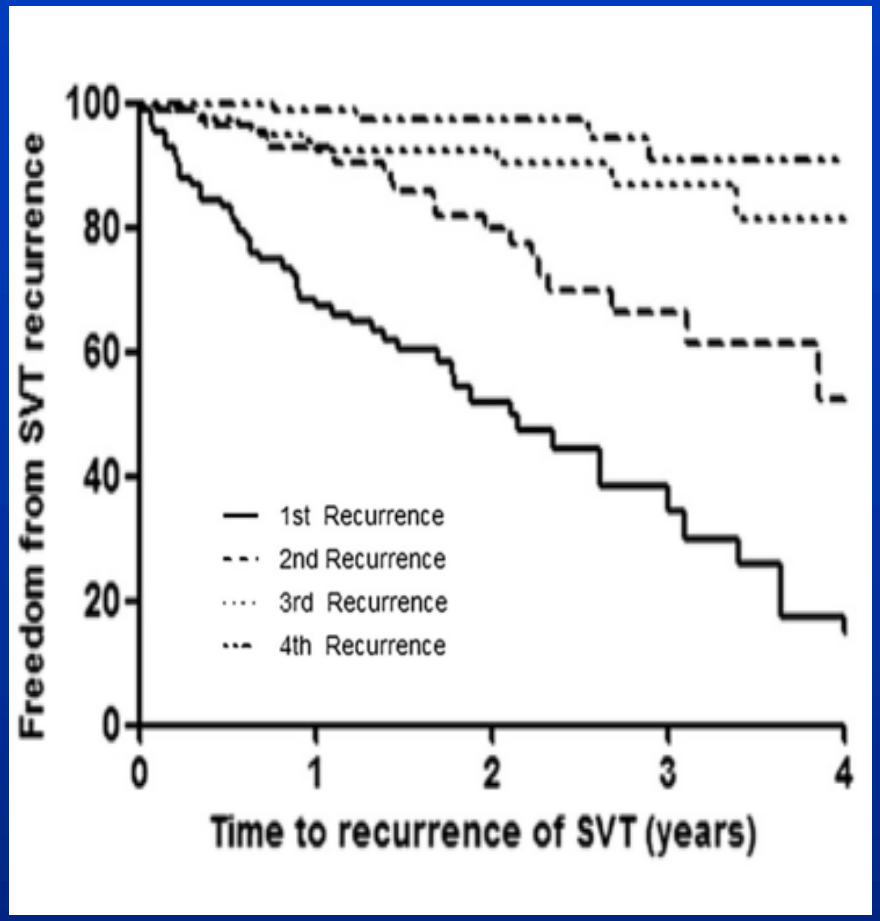
Koyak Z et al Am J Cardiol 2013;112:1461-1467

- Multicentre study/ retrospective
- Efficacy of AAD in SVT
- 2008-2011 CONCOR Database
- All new onset SVT in ACHD
  - excluded non cardiac causes of arrhythmia eg hypoT4

Efficacy of Antiarrhythmic Drugs in Adults With Congenital Heart Disease and Supraventricular Tachycardias

Koyak Z et al Am J Cardiol 2013;112:1461-1467





Efficacy of Antiarrhythmic Drugs in Adults With Congenital Heart Disease and Supraventricular Tachycardias

Koyak Z et al Am J Cardiol 2013;112:1461-1467

Conclusion

Class III most effective for SR

Sotolol should be Ist choice for SVT

What is the optimal drug Rx of ventricular arrhythmias in ACHD?

What is the optimal drug Rx of ventricular arrhythmias in ACHD?

Answer

We havent got a Scooby....

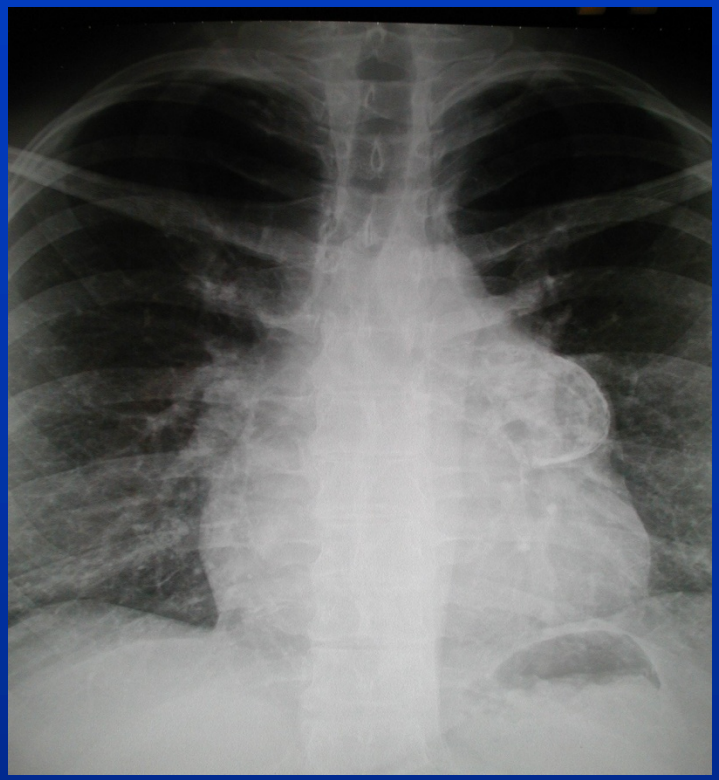
Cockney rhyming slang
Scooby's = Scooby Doo = clue Wikipedia

Are antiarrhythmic drugs helpful ?

In ACHD

- Don’t know !!
  - –Pick drugs you are familiar with
  - – Watch out for side effects
  - – Treat side effects....

Ventricular Arrhythmias in CHD

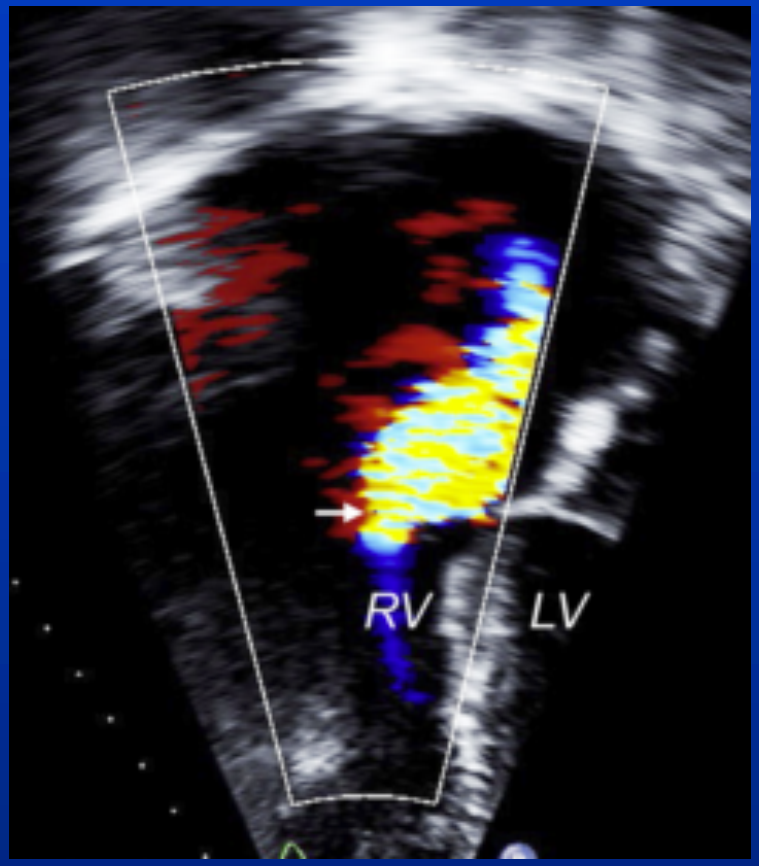


34yr old TOF - lost to follow up

Treat haemodynamic Problem……

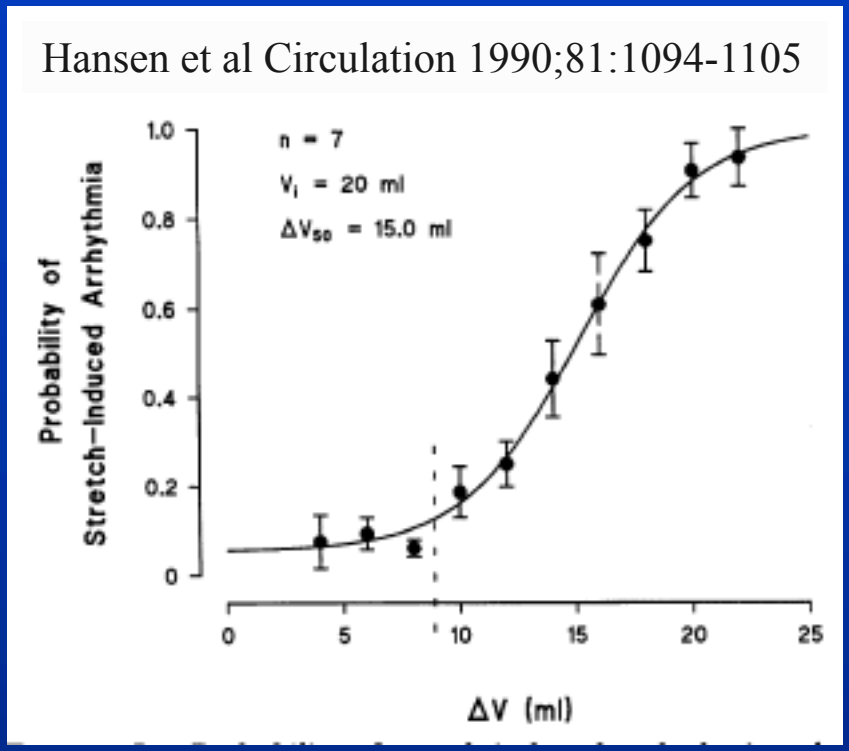
Tetralogy of Fallot

risk factors for arrhythmias



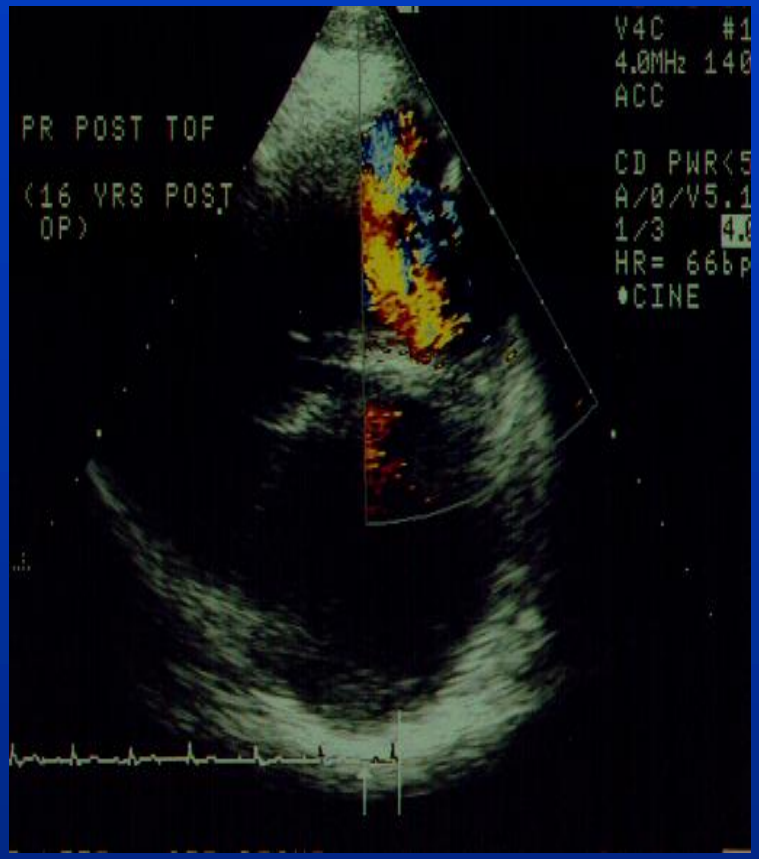
Severe TR in TOF

Hansen et al Circulation 1990;81:1094-1105



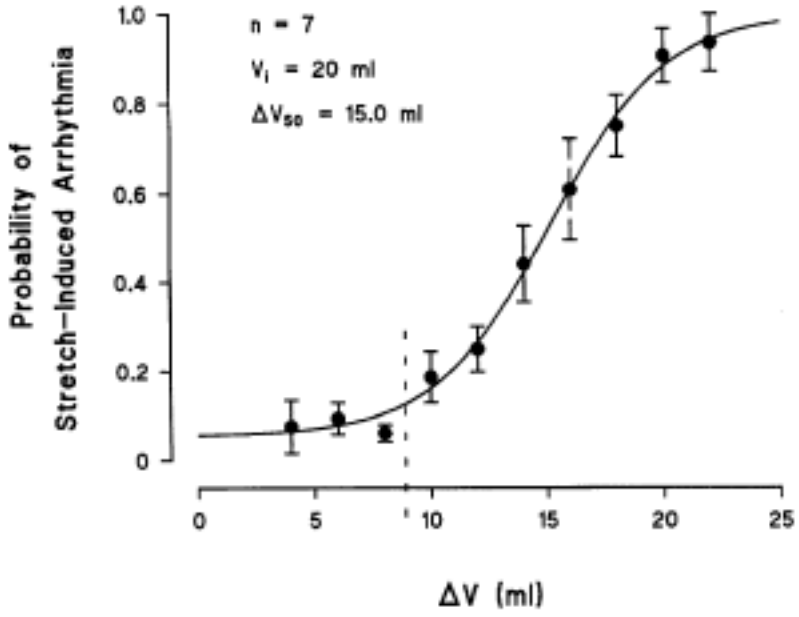
Tetralogy of Fallot

risk factors for arrhythmias

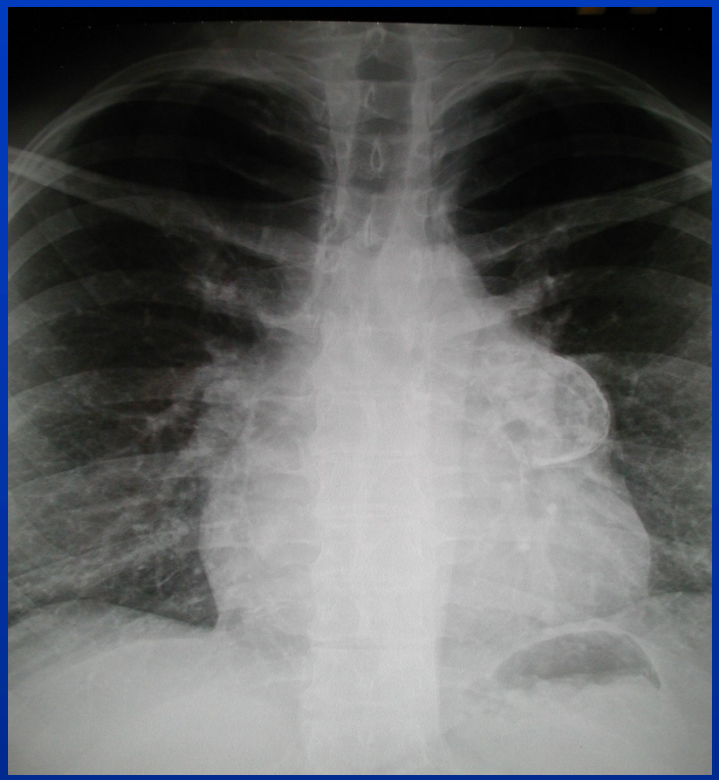


Severe PR in TOF

Hansen et al Circulation 1990;81:1094-1105



Ventricular Arrhythmias in CHD



34yr old TOF - lost to follow up

Treat haemodynamic Problem……

AA Drugs have limited role

Consider ICD +/- bivent

Arrhythmias and SCD

management in ToF and ACHD

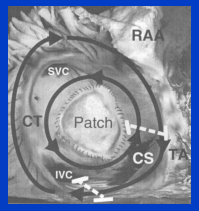
- Lifestyle
- Drugs
- Devices
- Ablation
- The surgical role

Arrhythmias and SCD

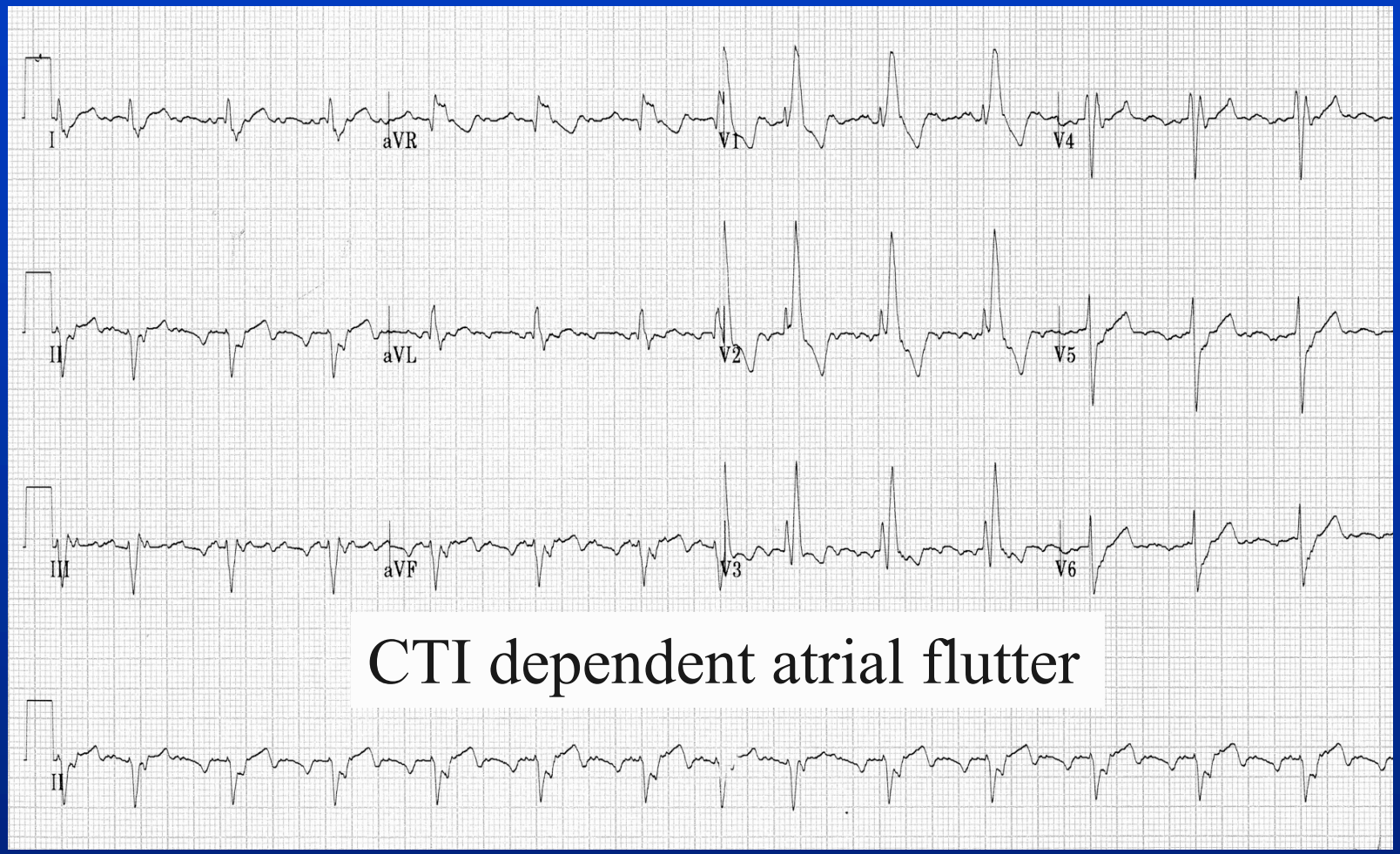
management in ToF and ACHD

- Lifestyle
- Drugs
- Devices
- Ablation
- The surgical role

Tetralogy of Fallot



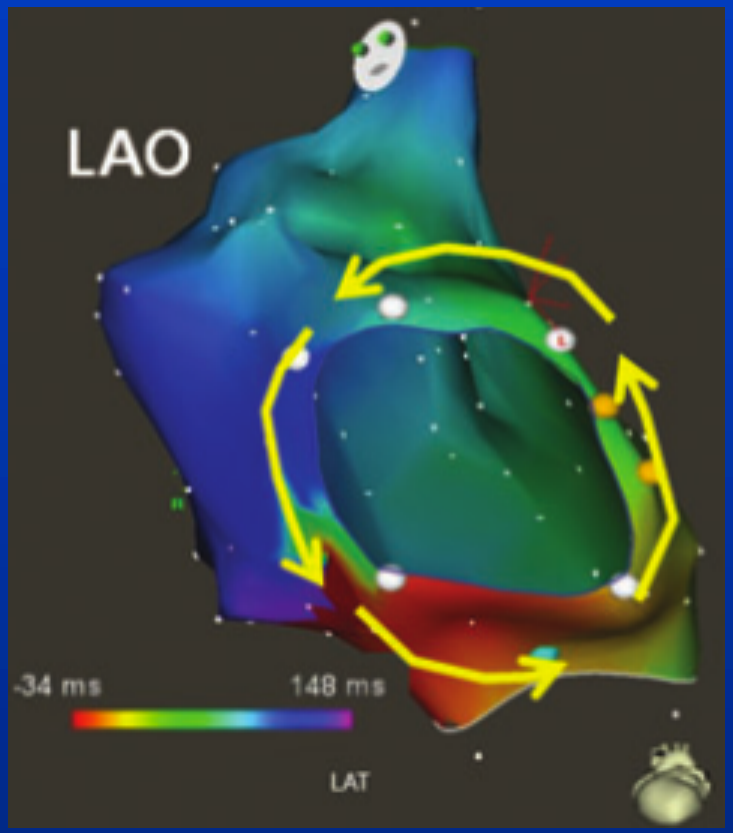
atrial arrhythmias: scar

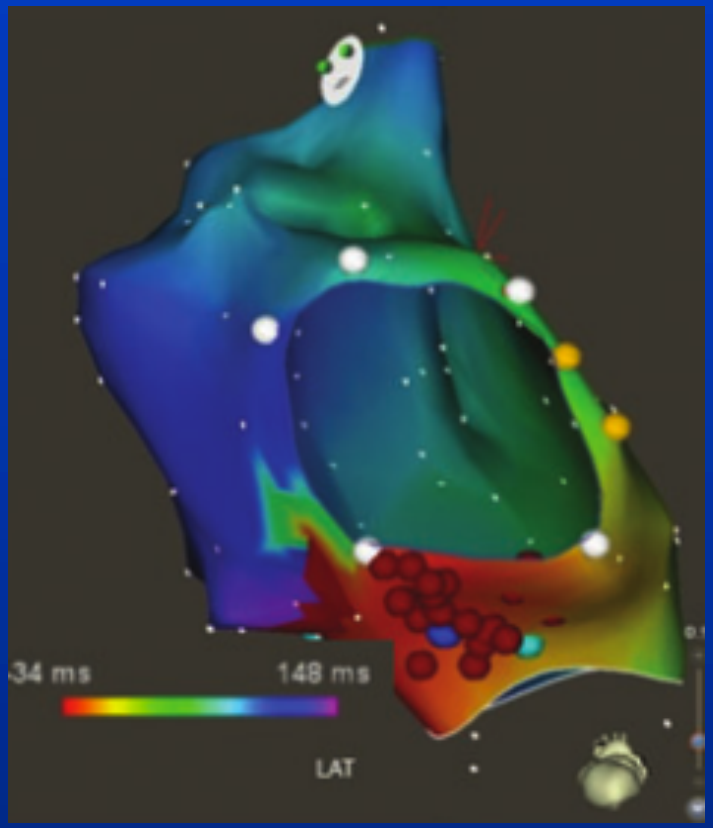


CTI dependent atrial flutter

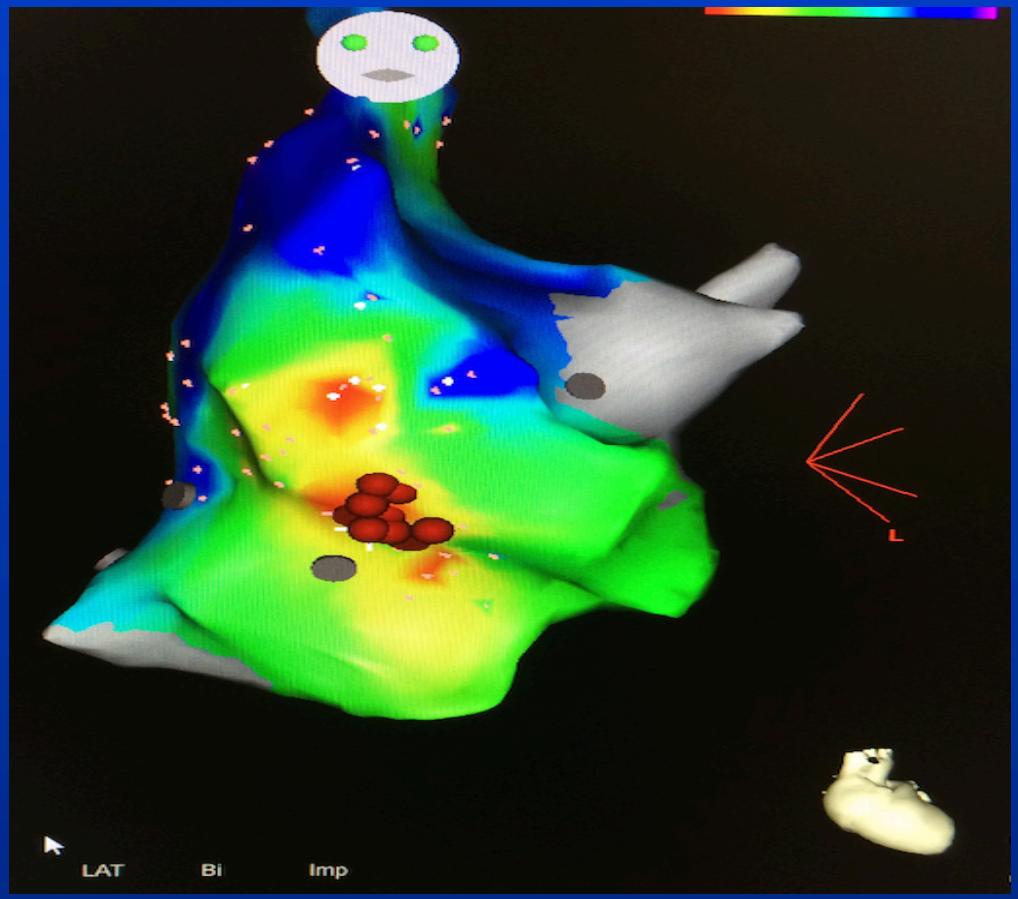
Tetralogy of Fallot

atrial arrhythmias: CTI dependent





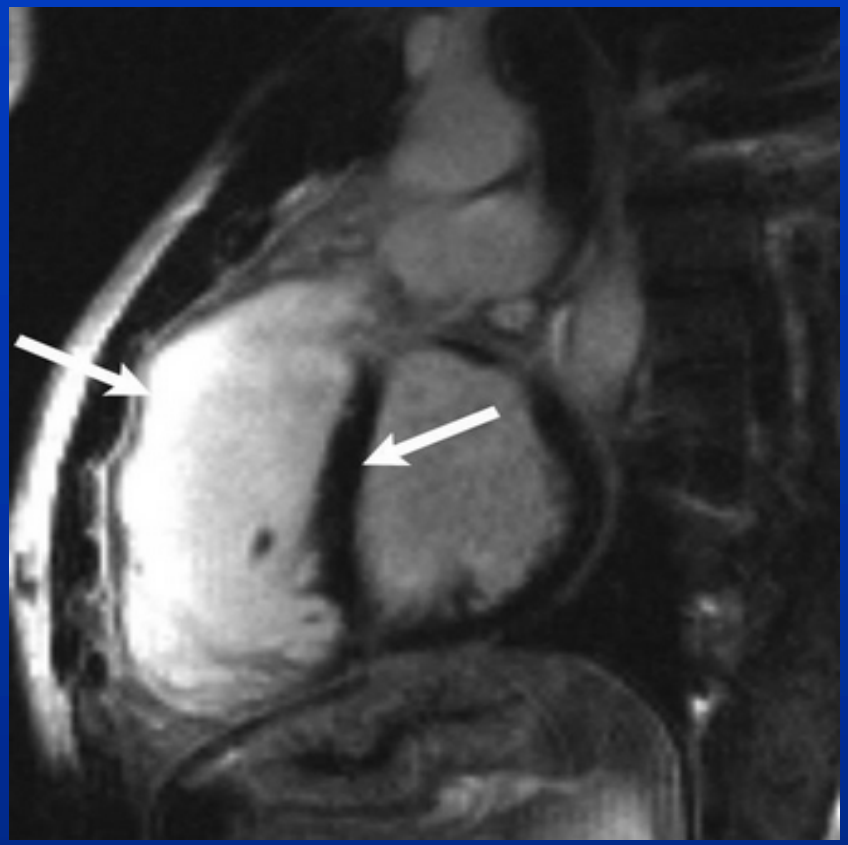
Focal Atrial tachycardia in CHD



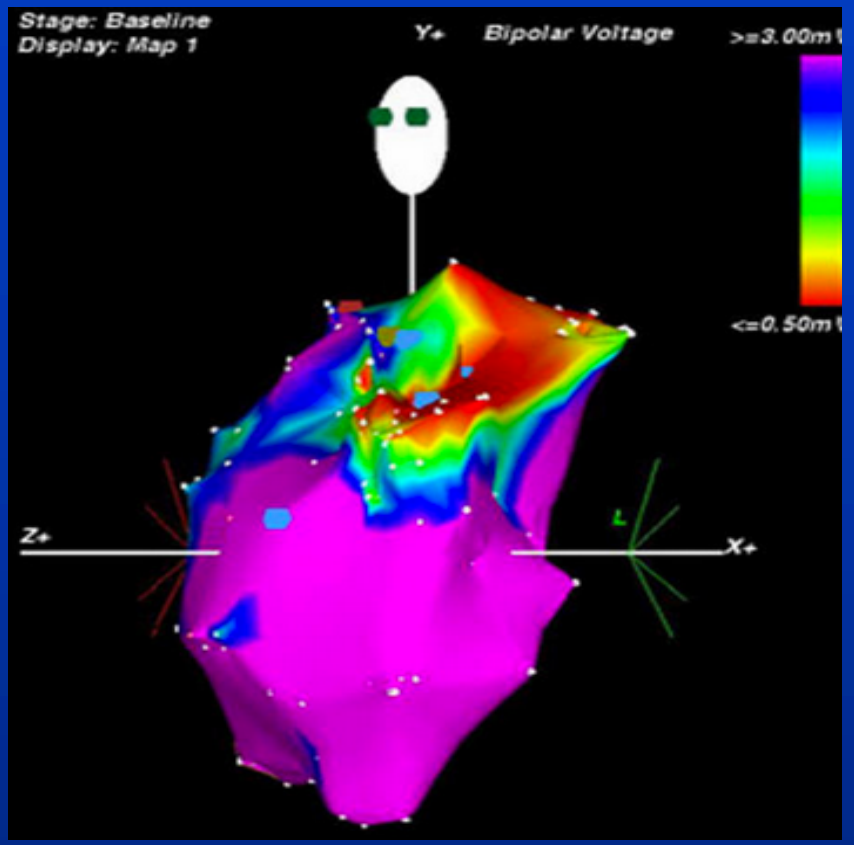
Miss HD Post MVR x3

Tetralogy of Fallot

arrhythmias: scar and VT



MRI : late gad + RVOT



CARTO: RVOT reentry

Learning point !

Essential to use up-to date technology in ablation…..

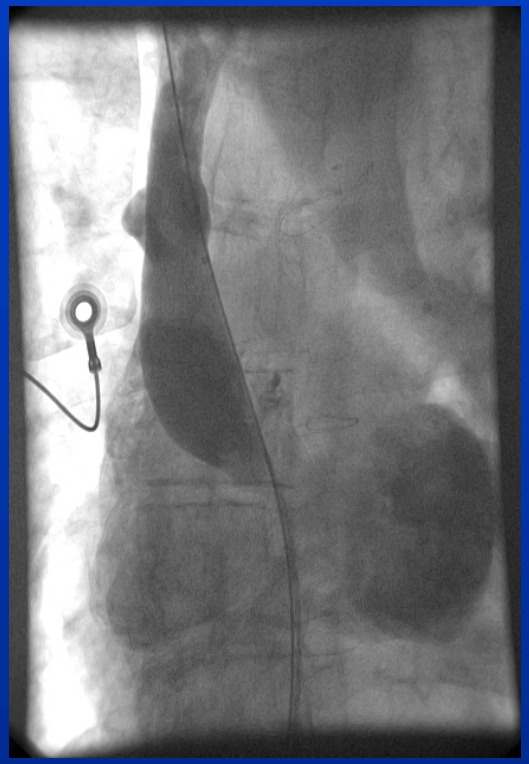
= 3D mapping, irrigated ablation, confidence software, dilithium crystals, phasers on stun etc

Arrhythmias and SCD

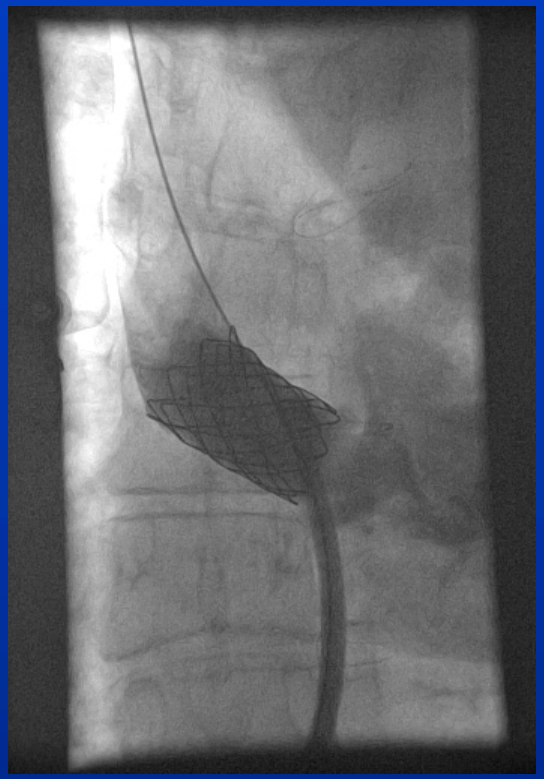
Devices in ToF and ACHD

- Pacemaker / ICD indications similar to "normal" cardiology
- Anatomy MAY be very different
- Devices have to last a long time!!

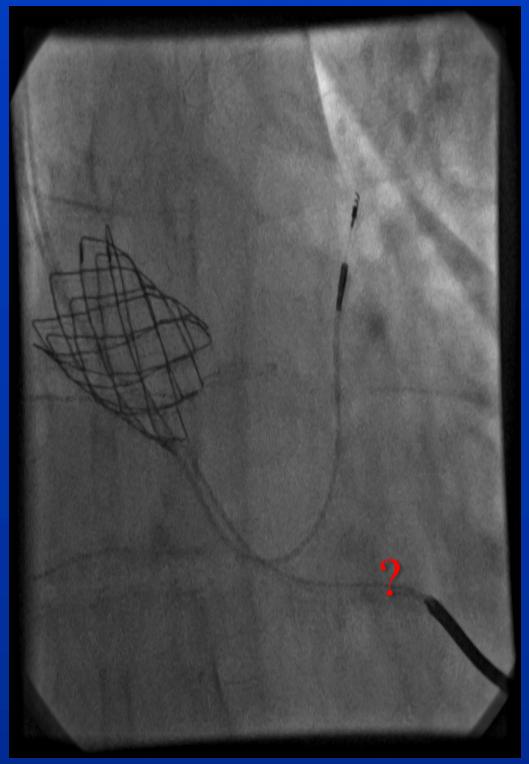
Transposition of great arteries



SVC obstruction

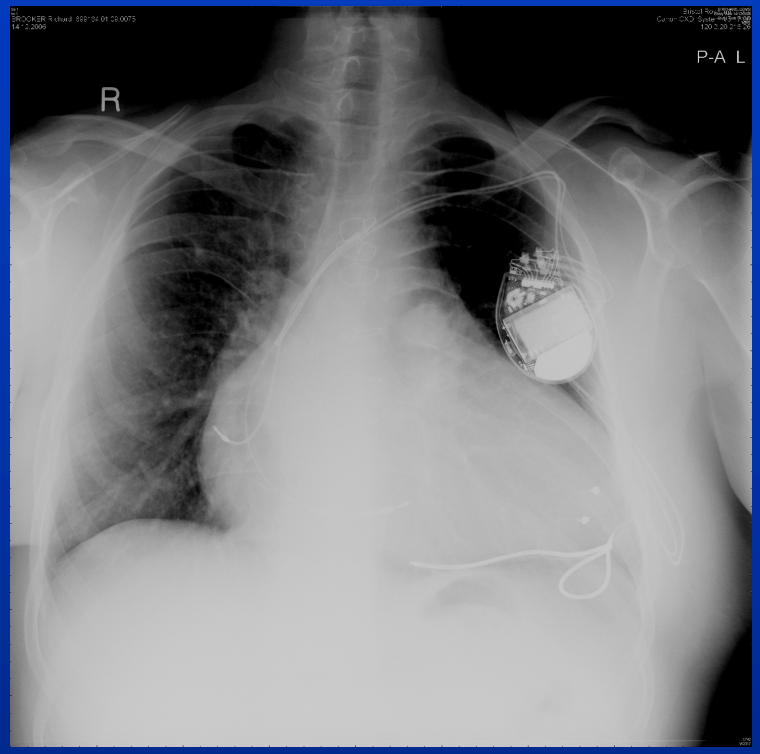


Covered stent in SVC baffle



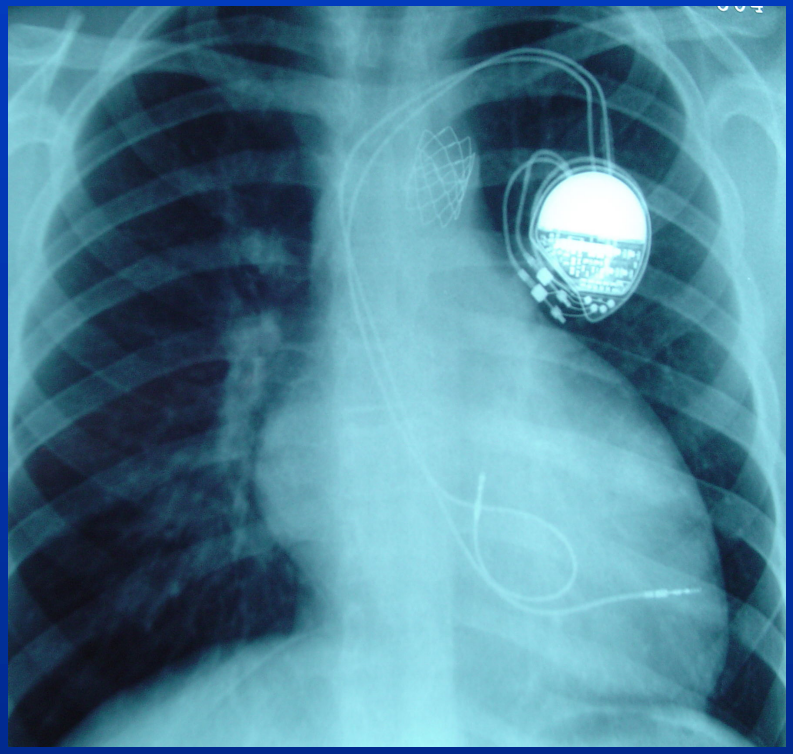
Selectsecure in “SVA”
Medtronic sprint in LV

Complex pacing in ACHD



Epicardial coils in Bivent ICD

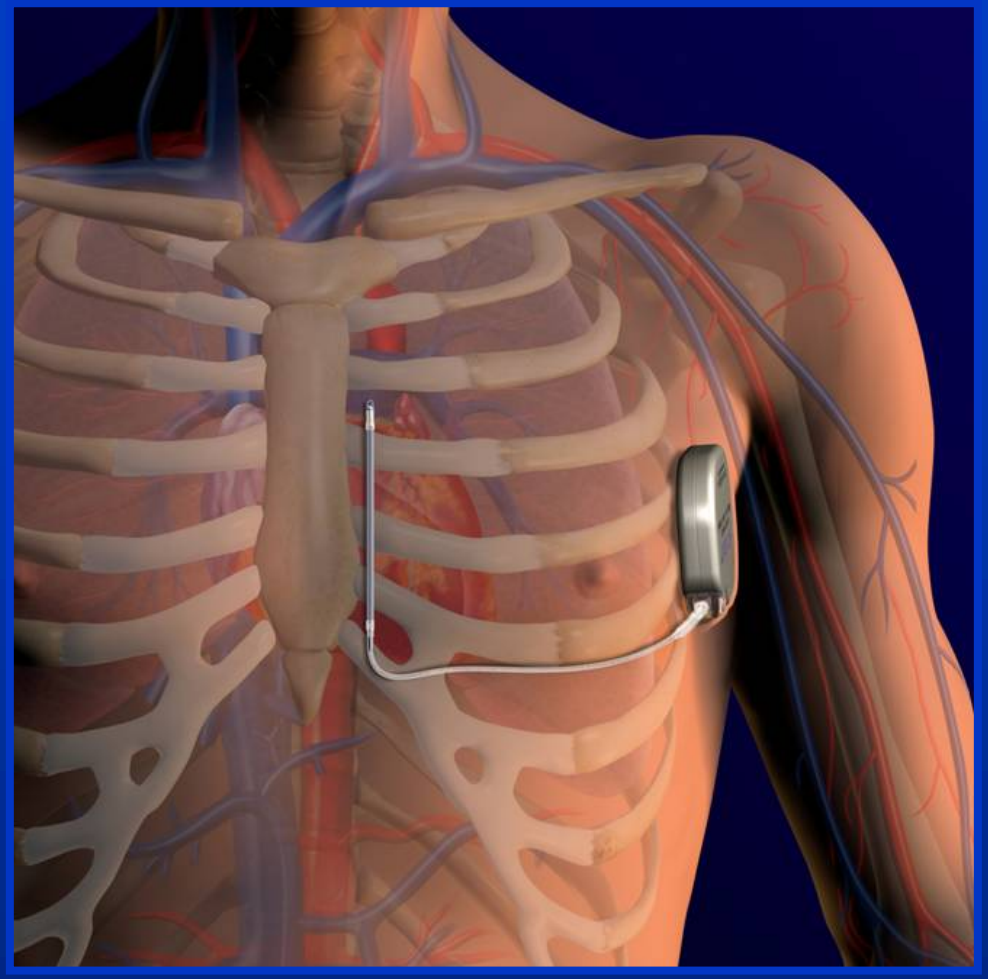
Pulmonary atresia/ VSD



Baffle puncture with transbaffle systemic ventricular lead. No coronary sinus

TGA/VSD/coarctation

Subcutaneous Defibrillator



Sometimes there is a right to left shunt !

Saturations 60%

Subcut ICD can be very useful !

Learning point !

Use patient specific device **NOT** one size fits all!!

= consider selectsecure leads / S-ICD etc

Arrhythmias and SCD

management in ToF and ACHD

- Lifestyle
- Drugs
- Devices
- Ablation
- The surgical role

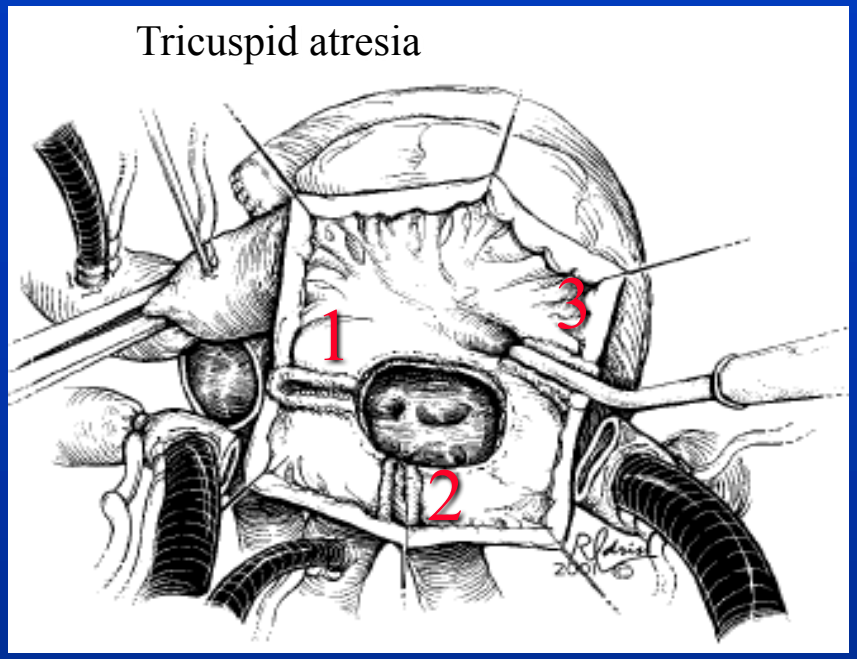
Learning point !

Surgeons need to think electrically ……

Initial surgery
complete lines of block

Redo surgery
Cox maze if known arrhythmia

Modified Right sided Maze



1. Superior Atrial septal ridge to RA appendage incised area
2. Posterior Atrial septal ridge to RA appendage incised area
3. Isthmus ablation (varies with anatomy)

IVC/ SVC transected

Atrial wall excised

ASD patch removed

Cryoablation lesions

-60 degrees for 90 secs

Arrhythmias and SCD

in ToF and ACHD

- How common are arrhythmias / SCD ?
- Why do they occur?
- How do we manage them?
- The Future…….
- Thomas the Tank Engine !

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

Heart Rhythm 2014:11(10):102-164

1. Care for ACHD arrhythmias – "coordinated by ACHD centres of excellence"

Include
- Electrophysiologist
- Interventional cardiologist
- Cardiac surgeon

} expertise in CHD

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

Heart Rhythm 2014:11(10):102-164

1. Care for ACHD arrhythmias – "coordinated by ACHD centres of excellence"

2. If symptomatic ACHD arrhythmias –

History
12 lead ECG
Ambulatory ECG

"Loop recorders"
if symptoms sporadic

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

Heart Rhythm 2014:11(10):102-164

1. Care for ACHD arrhythmias – "coordinated by ACHD centres of excellence"

2. If symptomatic ACHD arrhythmias –

Indications for haemodynamic study…..

All new onset or worsening arrhythmias or near miss – SCD **+ coronaries…**

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease Heart Rhythm 2014:11(10):102-164

1. Care for ACHD arrhythmias – "coordinated by ACHD centres of excellence"

2. If symptomatic ACHD arrhythmias –

Indications for EP study …..

All unexplained syncope +

High risk CHD substrate eg TGA/TOF/Single vent

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

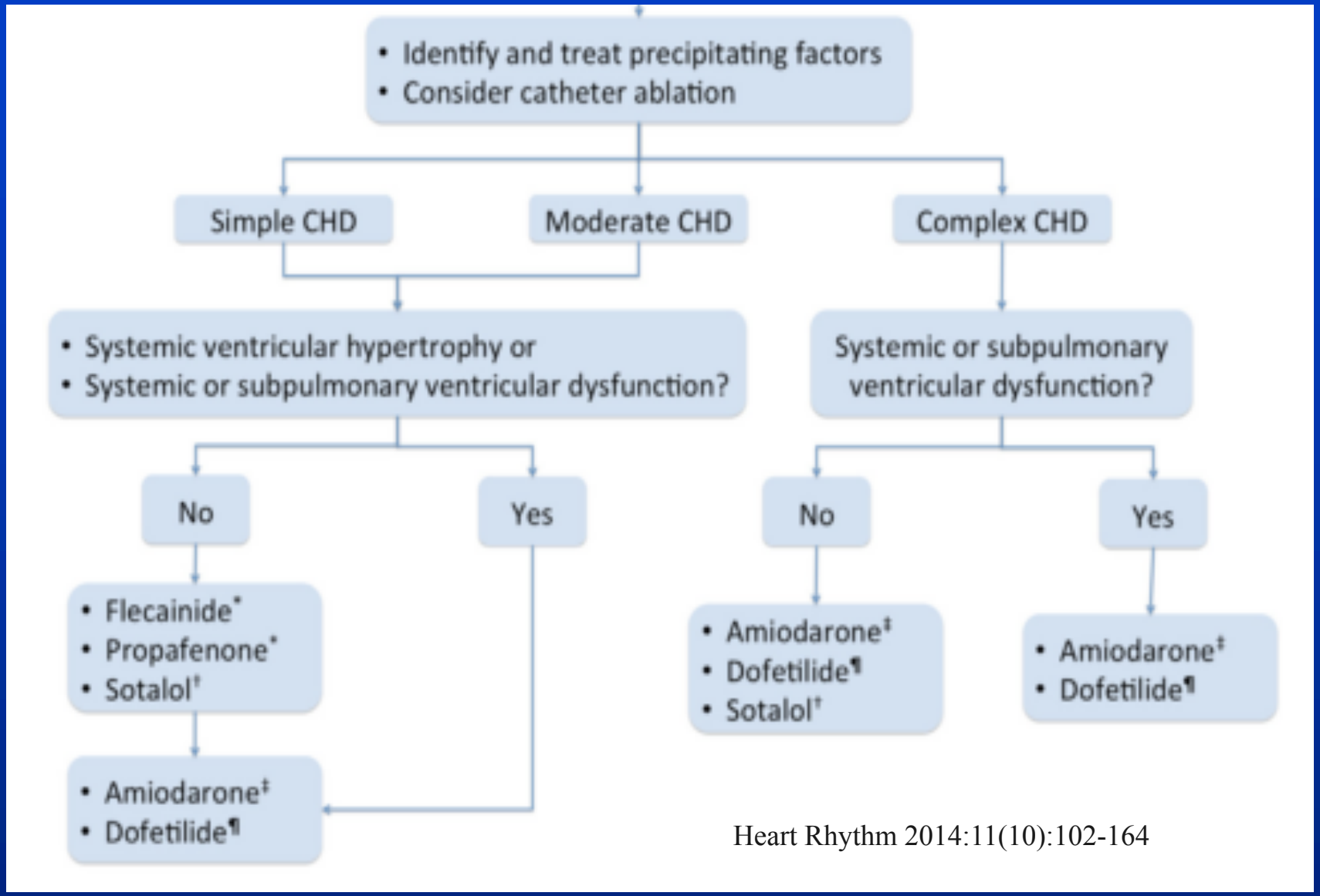
Heart Rhythm 2014:11(10):102-164

1. Care for ACHD arrhythmias – "coordinated by ACHD centres of excellence"

2. If symptomatic ACHD arrhythmias –

3. Algorithm for acute therapy

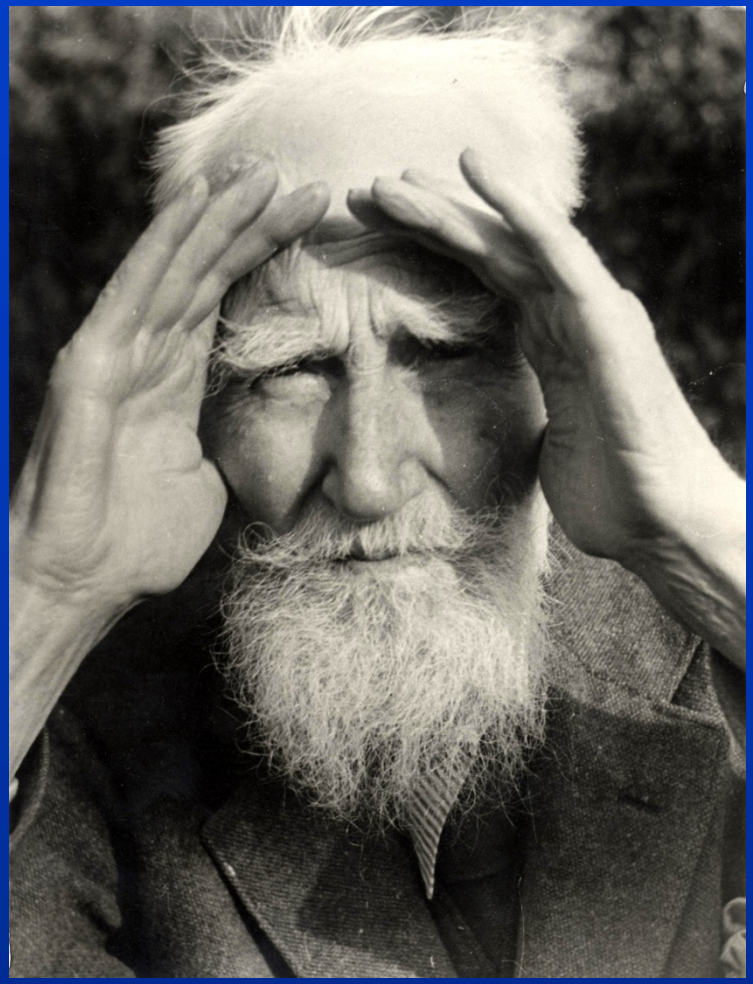
Rhythm Control in adults with CHD and IART or AFib



Heart Rhythm 2014:11(10):102-164

Interventions in CHD

success story!



George Bernard Shaw
1856 -1950

"Science is always wrong. It never solves a problem without creating ten more....."

Any Questions?

Arrhythmias and SCD

in ToF and ACHD

- How common are arrhythmias / SCD ?
- Why do they occur?
- How do we manage them?
- The Future……
- Thomas the Tank Engine !