



17th Adult Congenital Heart Study Day
Oct 2019



Life threatening arrhythmias

in Fontan patients

Graham Stuart

Hon Associate Professor, University of Bristol
Consultant Cardiologist Bristol Heart Institute

Prof Francis Fontan

1929-2018



- Father racing cyclist
“king of the mountains” Raymond



“kind to patients and kind to colleagues”

Prof Francis Fontan

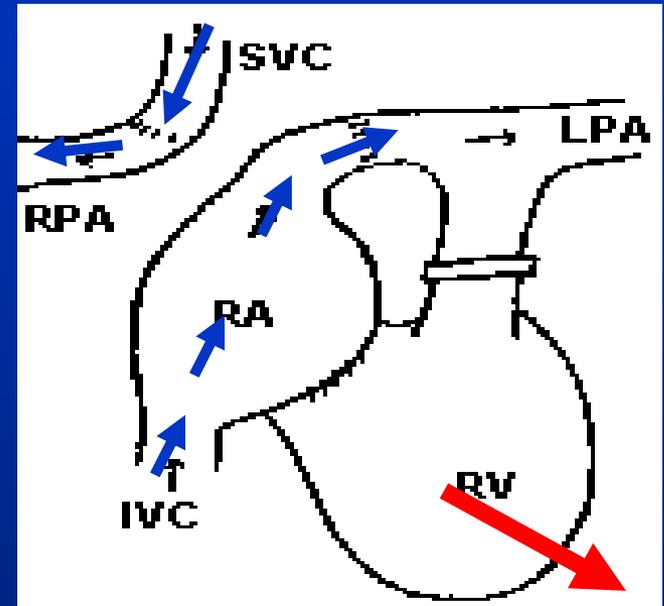
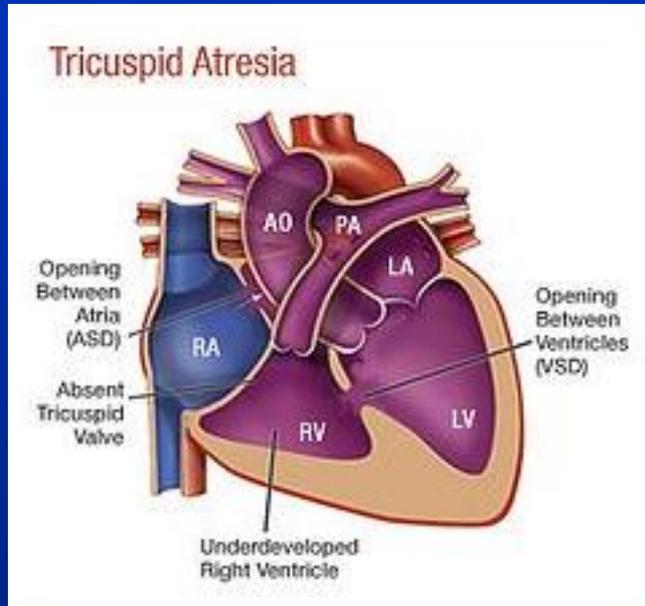
1929-2018



- Father racing cyclist
“king of the mountains”
- Cardiac Surgeon
Bordeaux 23 years
- Winner 2006
Grand Prix Scientifique de la
Fondation Lefoulon - Delalande

Prof Francis Fontan

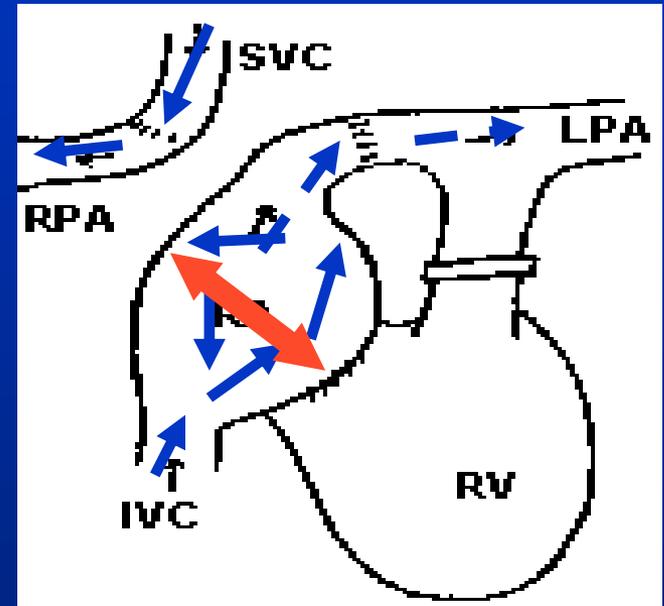
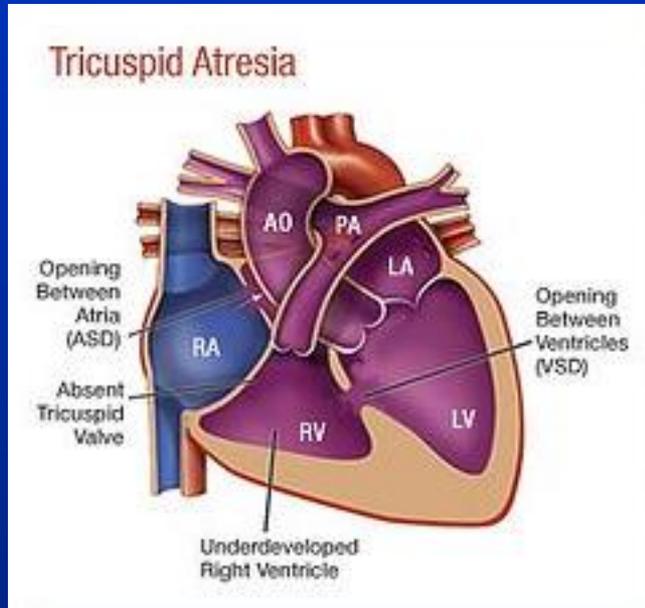
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RA = contracting pump

Prof Francis Fontan

1929-2018



RA = dilating sump

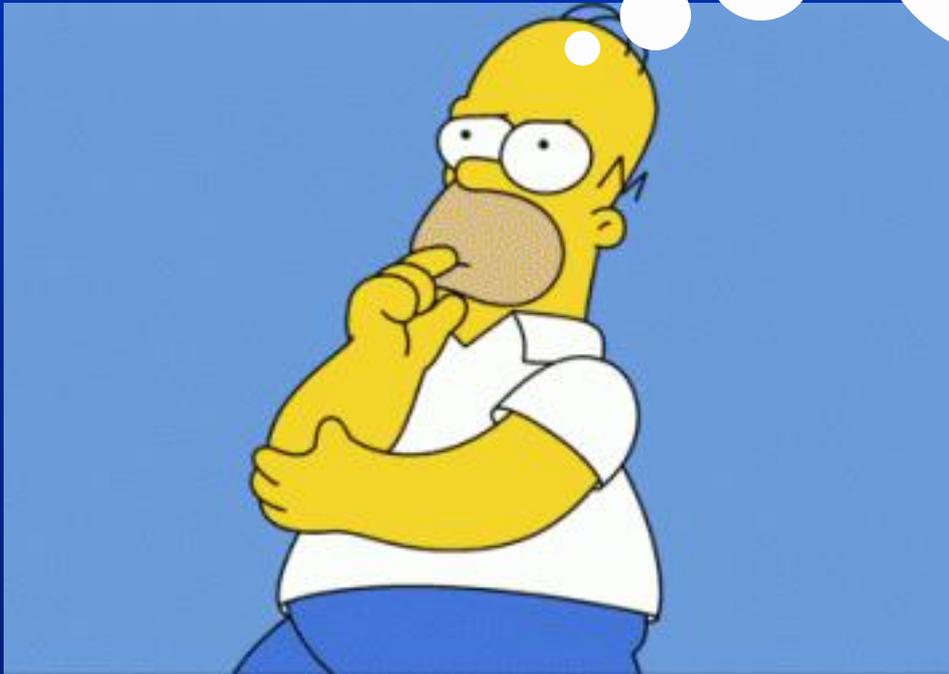
Electromechanical Failure

cruise photographer with varicose veins



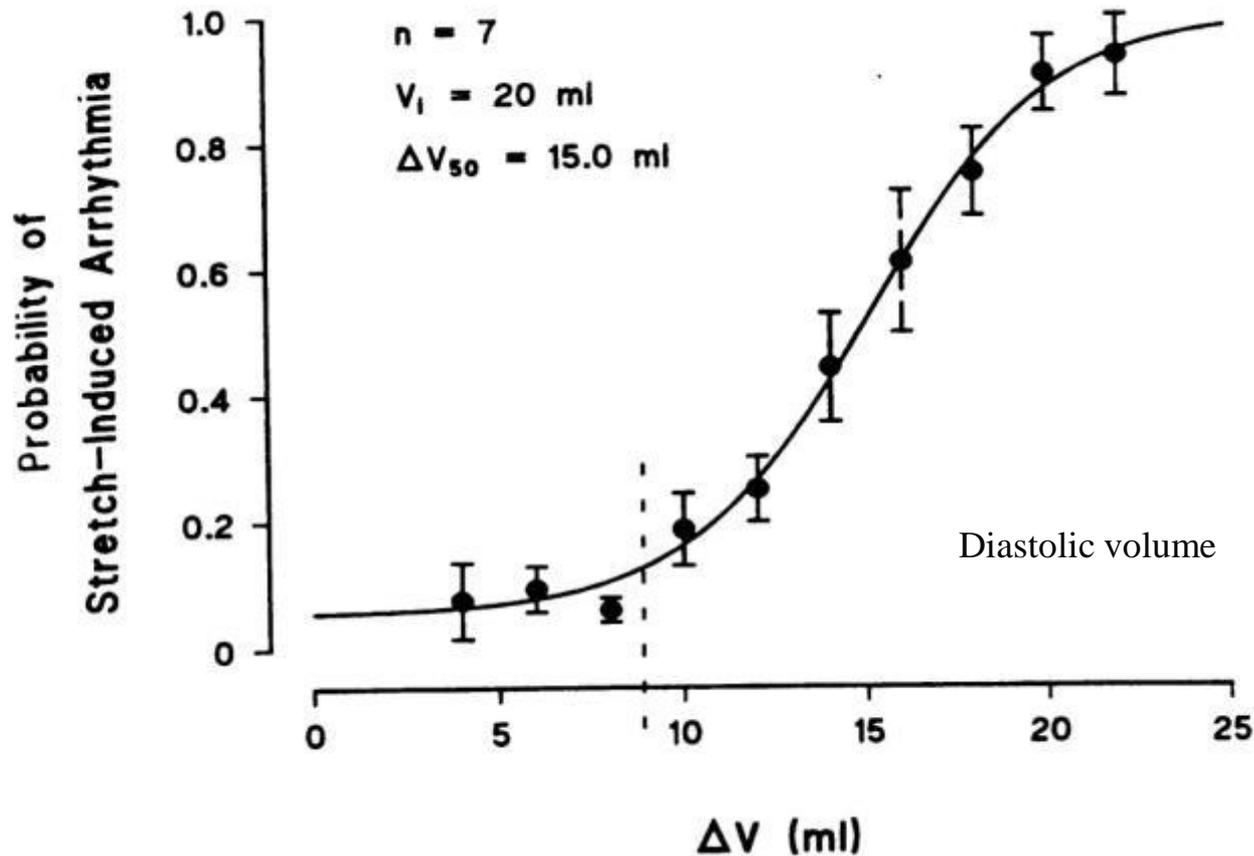
Dilated stretched
right atrium

What has this
got to do with
arrhythmias?



Stretch-induced arrhythmias

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



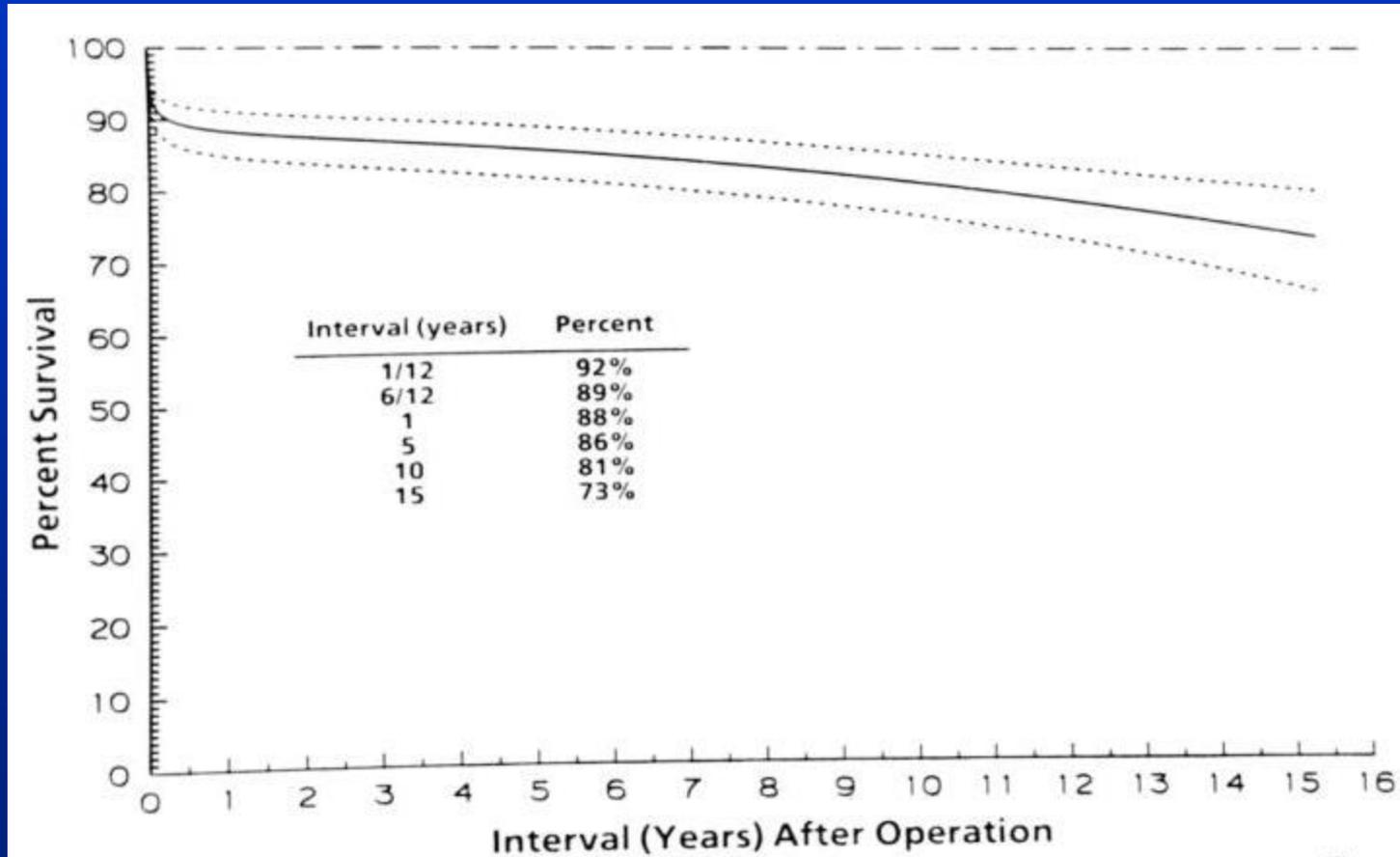
10 commandments

for the "perfect" Fontan operation

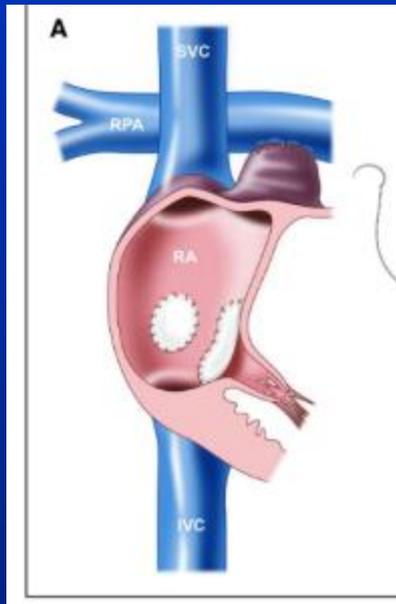
- Age > 4 years
- Sinus rhythm
- Normal systemic venous drainage
- Normal RA volume
- Mean PAP <15mmHg
- PAVR < 4 Woods units
- PA / Aorta ratio
- LVEF>60%
- Competent Mitral valve
- Well formed pulmonary arteries



“Perfect” Fontan Outcome



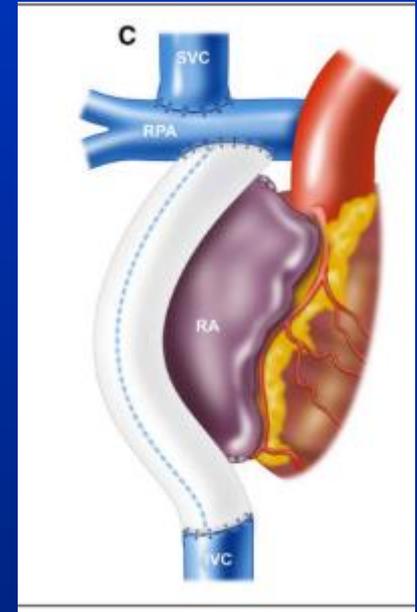
Modifications of the Fontan operation



Atriopulmonary



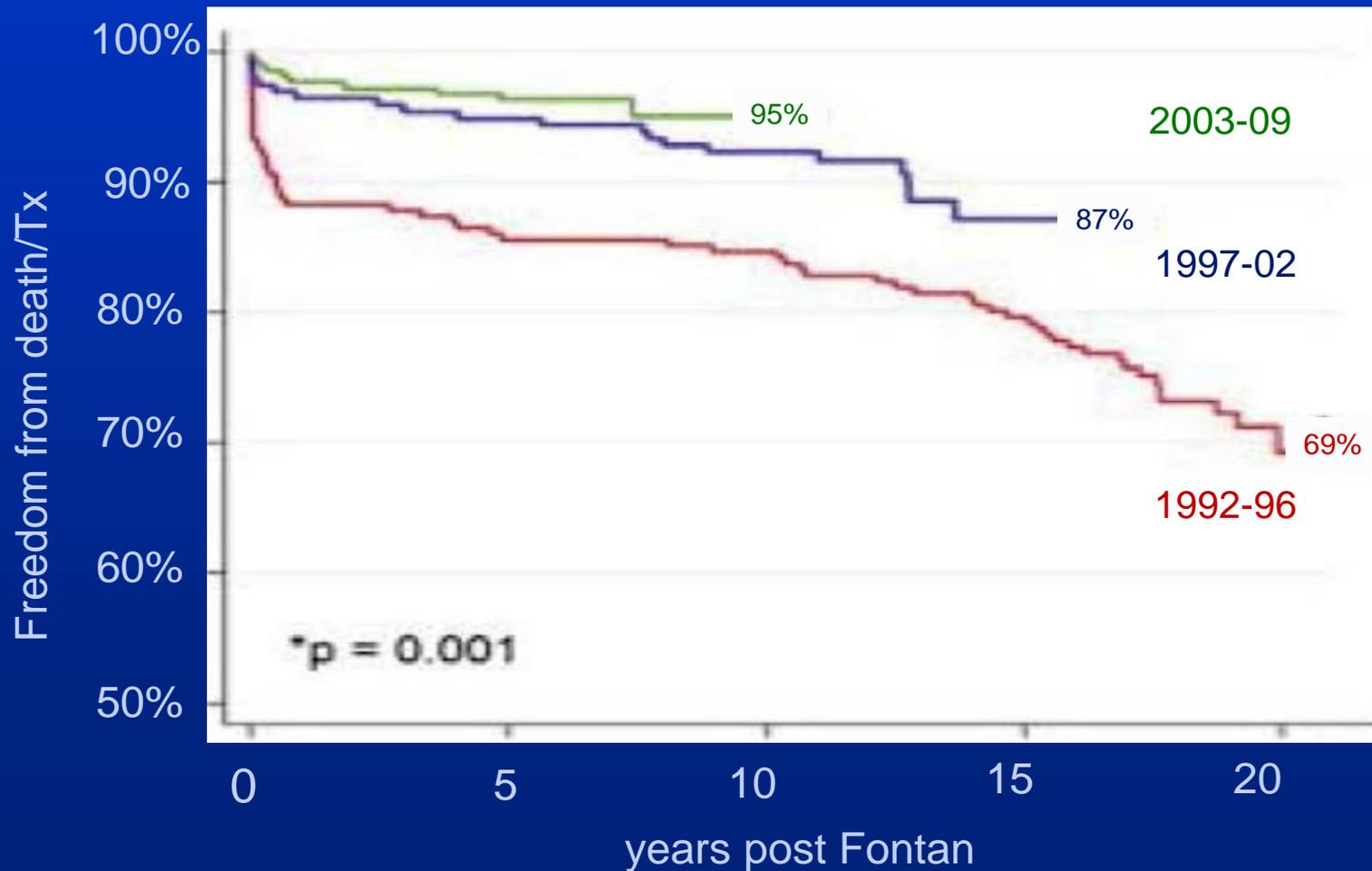
Lateral tunnel



Extracardiac tunnel

Fontan outcome

freedom from death/transplantation



Arrhythmias in ACHD

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

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.

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.

Most common
medical problem
referred to helpline !

Is this true for
Fontan
patients?



Cardiac Arrhythmias

in the Fontan operation

“ Postoperative arrhythmias, occurring in 30 - 50% of patients, are currently the most common complications following the Fontan operation. ”

Cecchin F et al Am J Cardiol 1995;76:386-391

Effect of age and surgical technique on symptomatic arrhythmias after the Fontan procedure

Cecchin F et al Am J Cardiol 1995;76:386-391

- 151 patients 1987-1993
- operation type
 - APC 46
 - TCPC 63
 - TCPC 27
plus fenestration

Effect of age and surgical technique on symptomatic arrhythmias after the Fontan procedure

Cecchin F et al Am J Cardiol 1995;76:386-391

Conclusions

- 5% preoperative arrhythmias

Effect of age and surgical technique on symptomatic arrhythmias after the Fontan procedure

Cecchin F et al Am J Cardiol 1995;76:386-391

Conclusions

- 5% preoperative arrhythmias
- 31% early post operative arrhythmias

Effect of age and surgical technique on symptomatic arrhythmias after the Fontan procedure

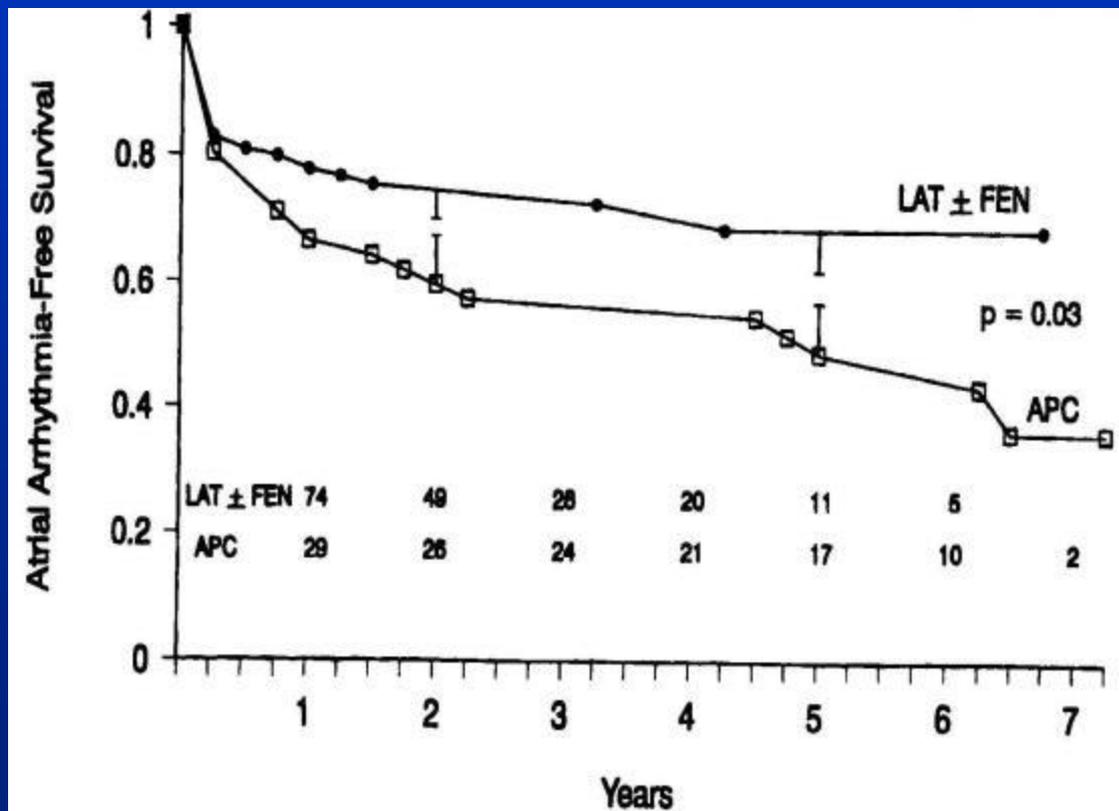
Cecchin F et al Am J Cardiol 1995;76:386-391

Conclusions

- 5% preoperative arrhythmias
- 31% early post operative arrhythmias
- 23% (30) intermediate arrhythmias

Effect of age and surgical technique on symptomatic arrhythmias after the Fontan procedure

Cecchin F et al Am J Cardiol 1995;76:386-391



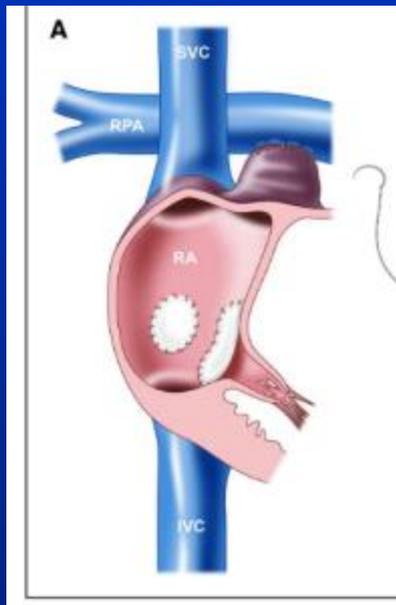
Occurrence and management of atrial arrhythmia after long-term Fontan circulation

Weipert J et al J Thor Cardiovasc Surg 2004

- 162 patients 205 patient years
- 1978- 1995
- 20yr freedom from tachycardia 46% +/- 12%

Multivariate analysis identified duration of Fontan circulation as the sole risk factor for re-entrant tachycardias.

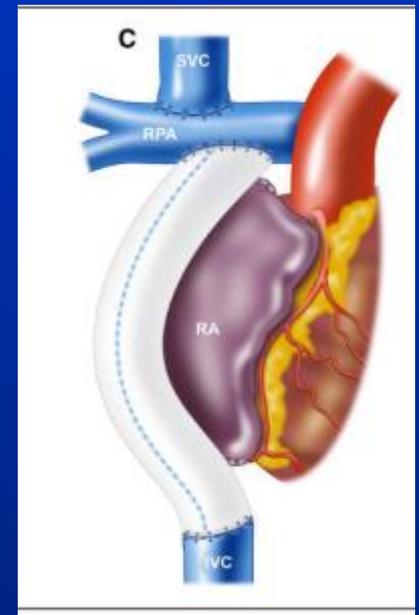
Modifications of the Fontan operation



Atriopulmonary



Lateral tunnel



Extracardiac tunnel

Is the extracardiac Fontan the answer ?

Shirai LK et al J Thorac Cardiovasc Surg 1998;115(3):499-505

- n = 16
- 1993 - 1996
- 44% had early postoperative arrhythmias
- 50% had arrhythmias at follow -up

Mostly sinus node dysfunction

“other surgical or non-surgical factors responsible”

An international multicenter study comparing arrhythmia prevalence between the intracardiac lateral tunnel and the extracardiac conduit type of Fontan operations

Balaji et al J Thorac Cardiovasc Surg 2014;148:576-81

- n=1271
 - 602 intracardiac
 - 669 extracardiac
- Median age at op
 - 2.1yrs IC; 3yrs EC
- Median follow up
 - 9.2 yrs IC; 4.7yrs EC
- Early < 30days
 - Bradyarrhythmia
 - 4% IC 11% EC
 - Tachyarrhythmia
 - 5% IC 8% EC
- Late > 30days
 - Bradyarrhythmia
 - 18% IC 9% EC
 - Tachyarrhythmia
 - 10% IC 3%EC

An international multicenter study comparing arrhythmia prevalence between the intracardiac lateral tunnel and the extracardiac conduit type of Fontan operations

Balaji et al J Thorac Cardiovasc Surg 2014;148:576-81

Conclusion after multivariate analysis

Arrhythmia burden is similar between groups

higher incidence early brady EC group

NO difference in late tachyarrhythmias

Fontan type should be based on factors unrelated to future arrhythmia risk!

What
arrhythmias
do we see?



Arrhythmias in Fontan

- Bradyarrhythmias
- Tachyarrhythmias
 - Atrial
 - Ventricular

How many are
life
threatening ?



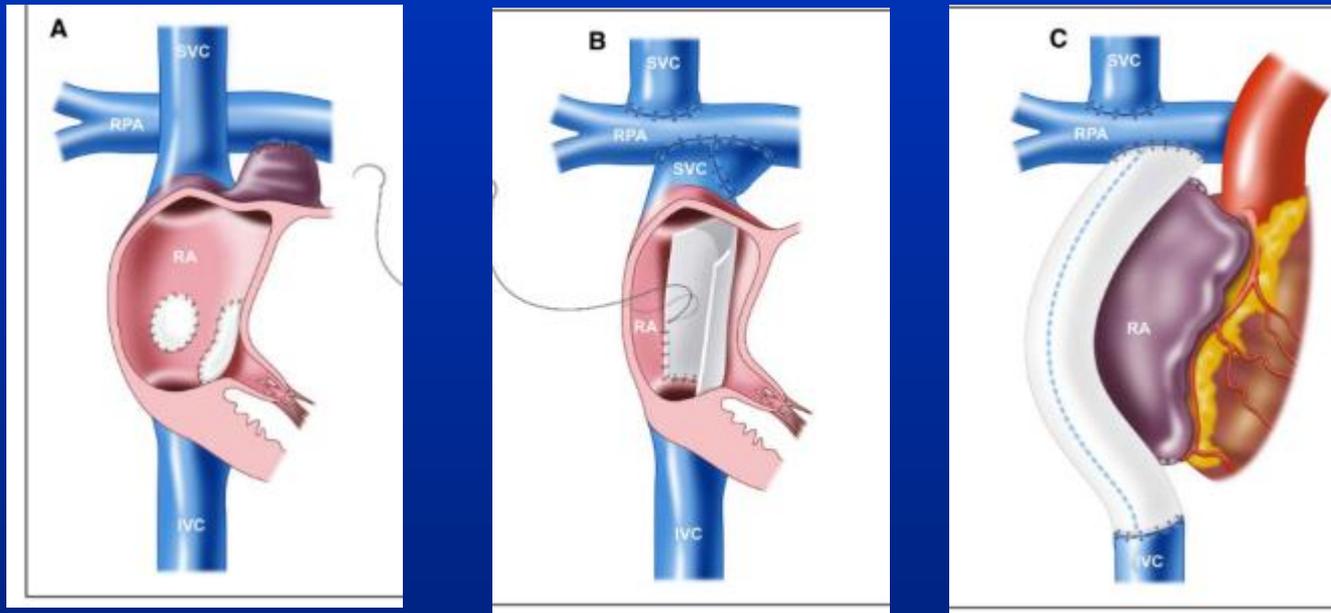
All of
them!!
!!!!



Arrhythmias in Fontan

- Bradyarrhythmia problems

Pacing in the Fontan operation



Endocardial A Epicardial v +/- Baffle puncture

Increased lead fracture/ reduced longevity/
sternotomy often needed

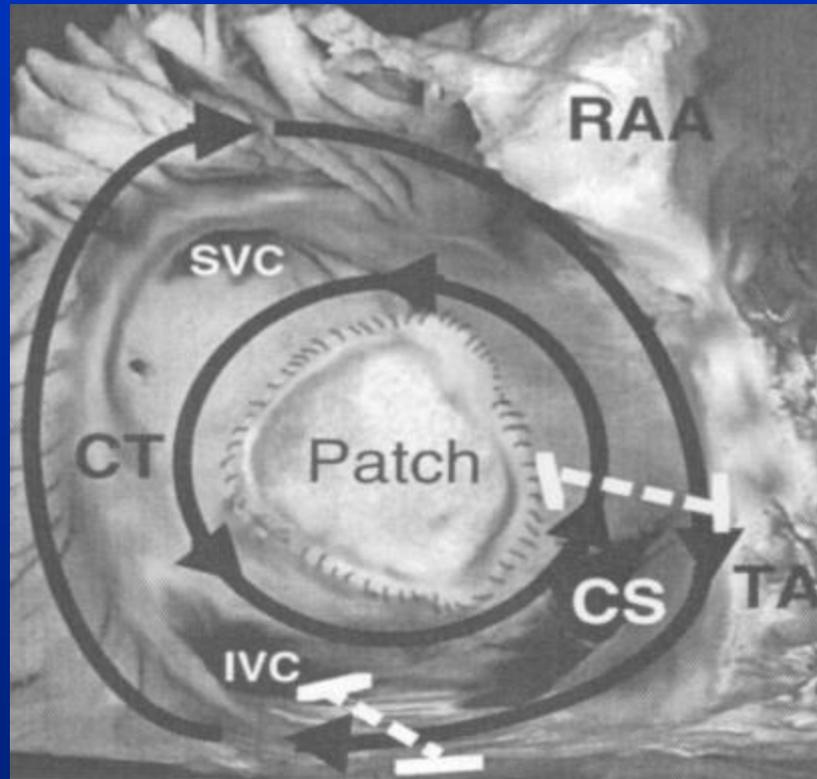
Arrhythmias in Fontan

- Bradyarrhythmias
- Tachyarrhythmias
 - Atrial
 - Ventricular

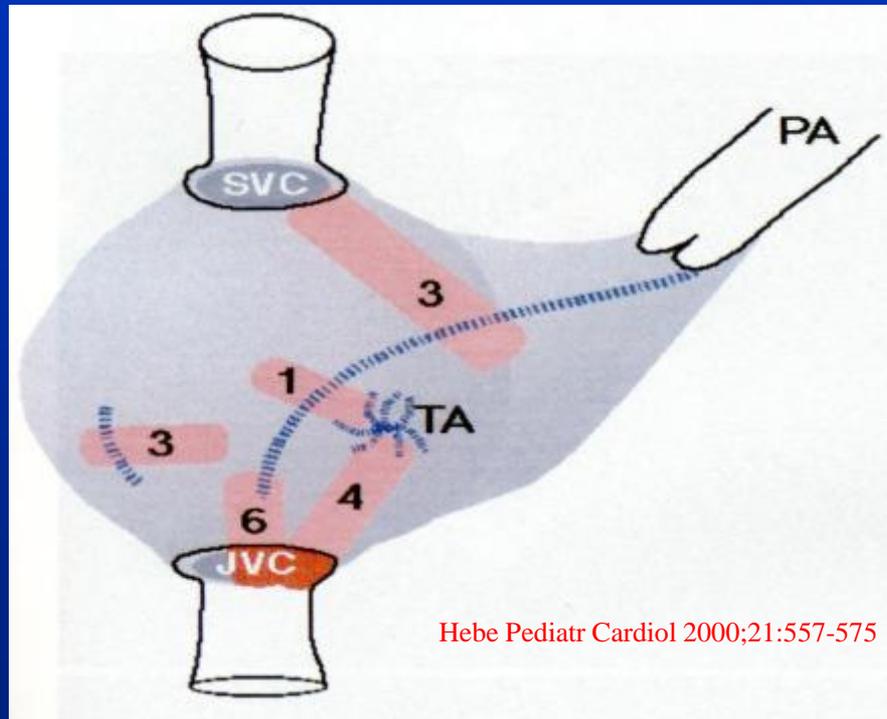
A cartoon illustration of Homer Simpson, a yellow-skinned man with a large brown beard and glasses, wearing a white t-shirt and blue pants. He is shown from the waist up, with his right hand resting on his chin in a classic 'thinking' pose. The background behind him is a light blue rectangle. A white thought bubble trail starts from his head and leads to a large white thought bubble on the right side of the image. The entire scene is set against a dark blue background.

Why do they
occur ?

Re-entry after ASD repair



“Scars” post Fontan



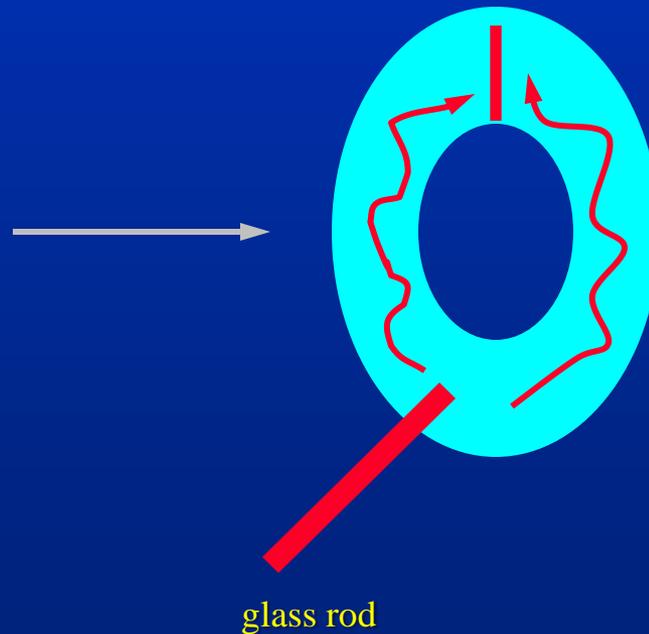
Hebe Pediatr Cardiol 2000;21:557-575

Mechano-mechanical interaction

Mayer AG. Rhythmic pulsation in a scyphomedusae
Carnegie Inst Pub 47,1906



Scyphomedusa jellyfish

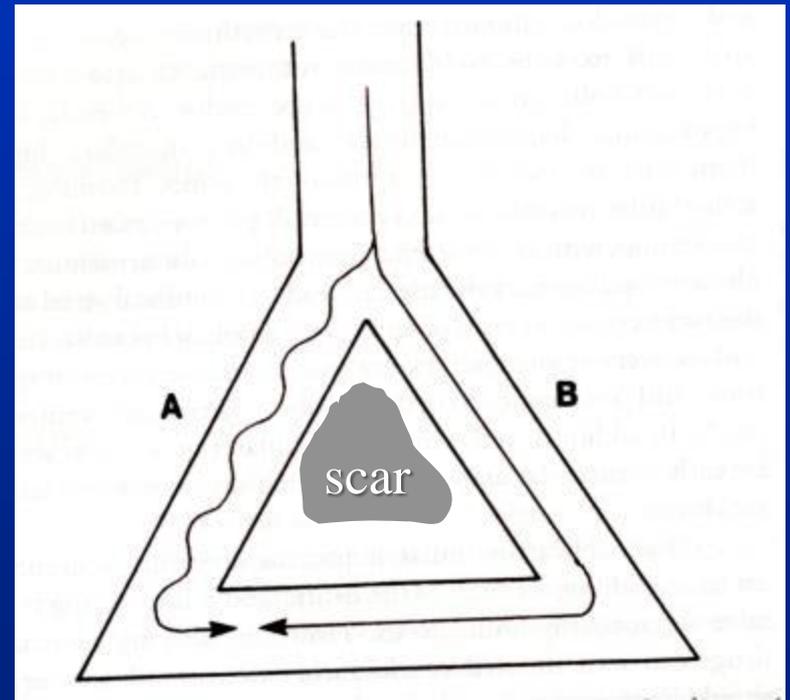


glass rod

What is re-entry?

1. must form a circuit

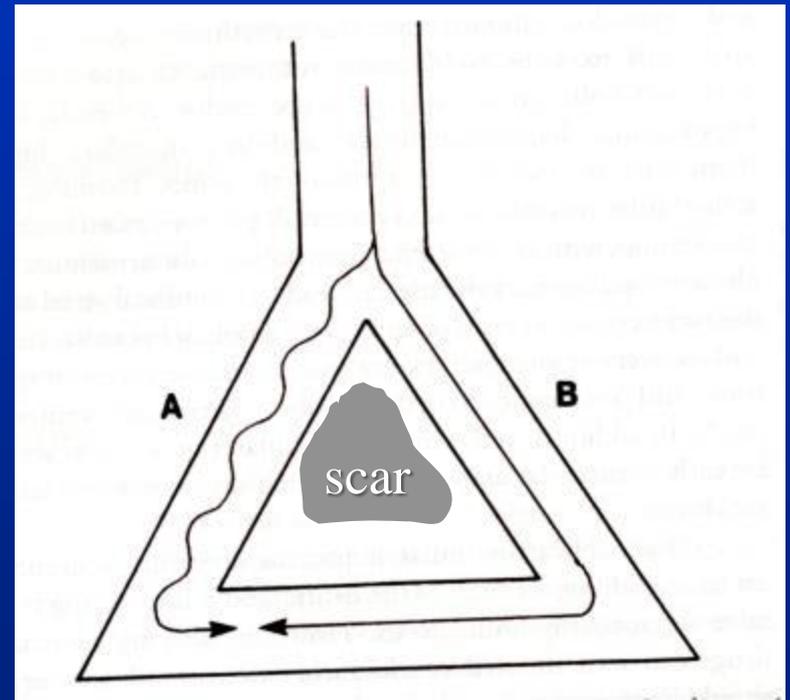
2 parallel conducting pathways



What is re-entry?

1. must form a circuit
2. refractory periods must differ ($B > A$)

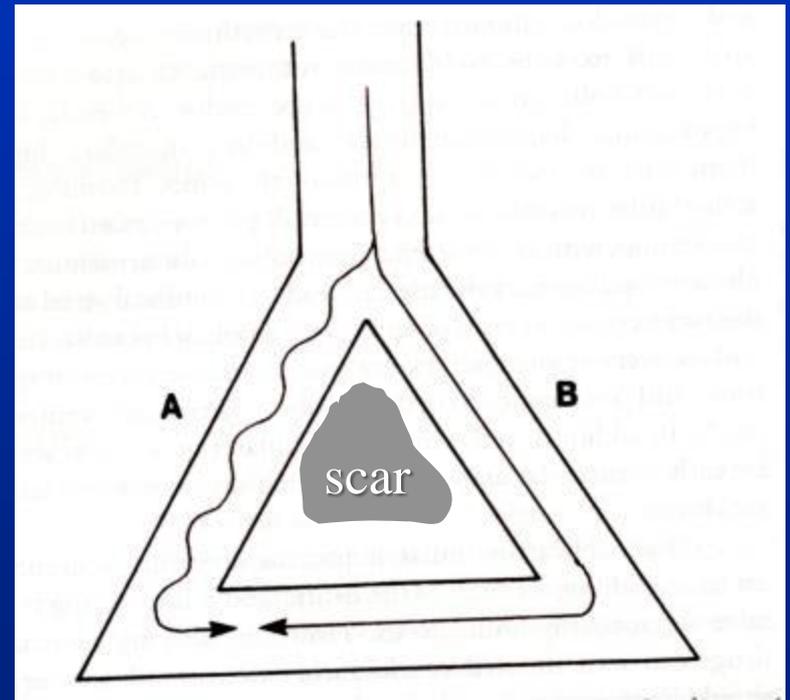
2 parallel conducting pathways



What is re-entry?

1. must form a circuit
2. refractory periods must differ ($B > A$)
3. shorter refractory period pathway (A) must conduct impulses slower than longer refractory period pathway (B)

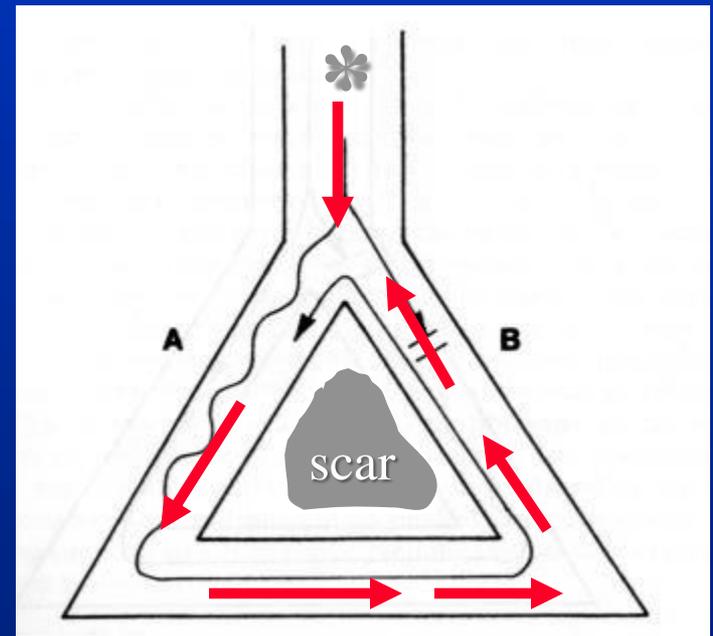
2 parallel conducting pathways



What is re-entry?

1. Impulse enters A when B is refractory
2. B recovers and impulse conducts retrograde up B.
3. Impulse then reenters A and conducts antegrade.
4. Impulses propagate to rest of myocardium.

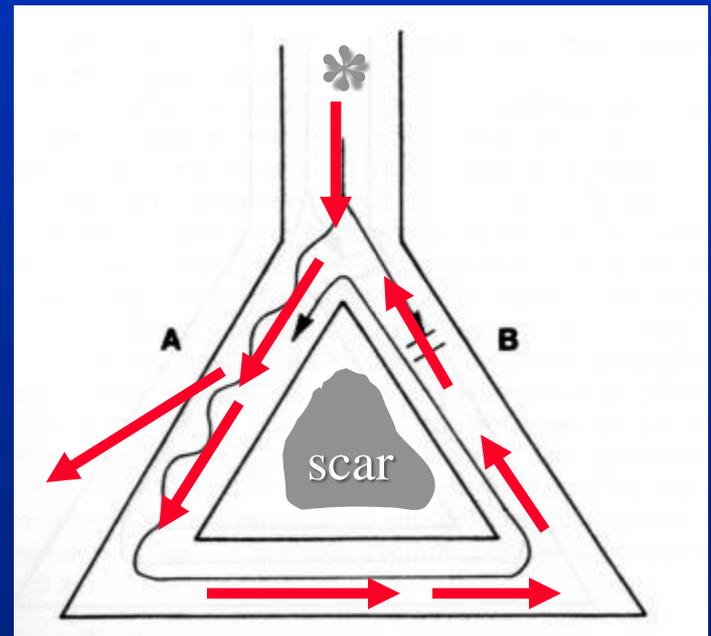
Premature impulse *



What is re-entry?

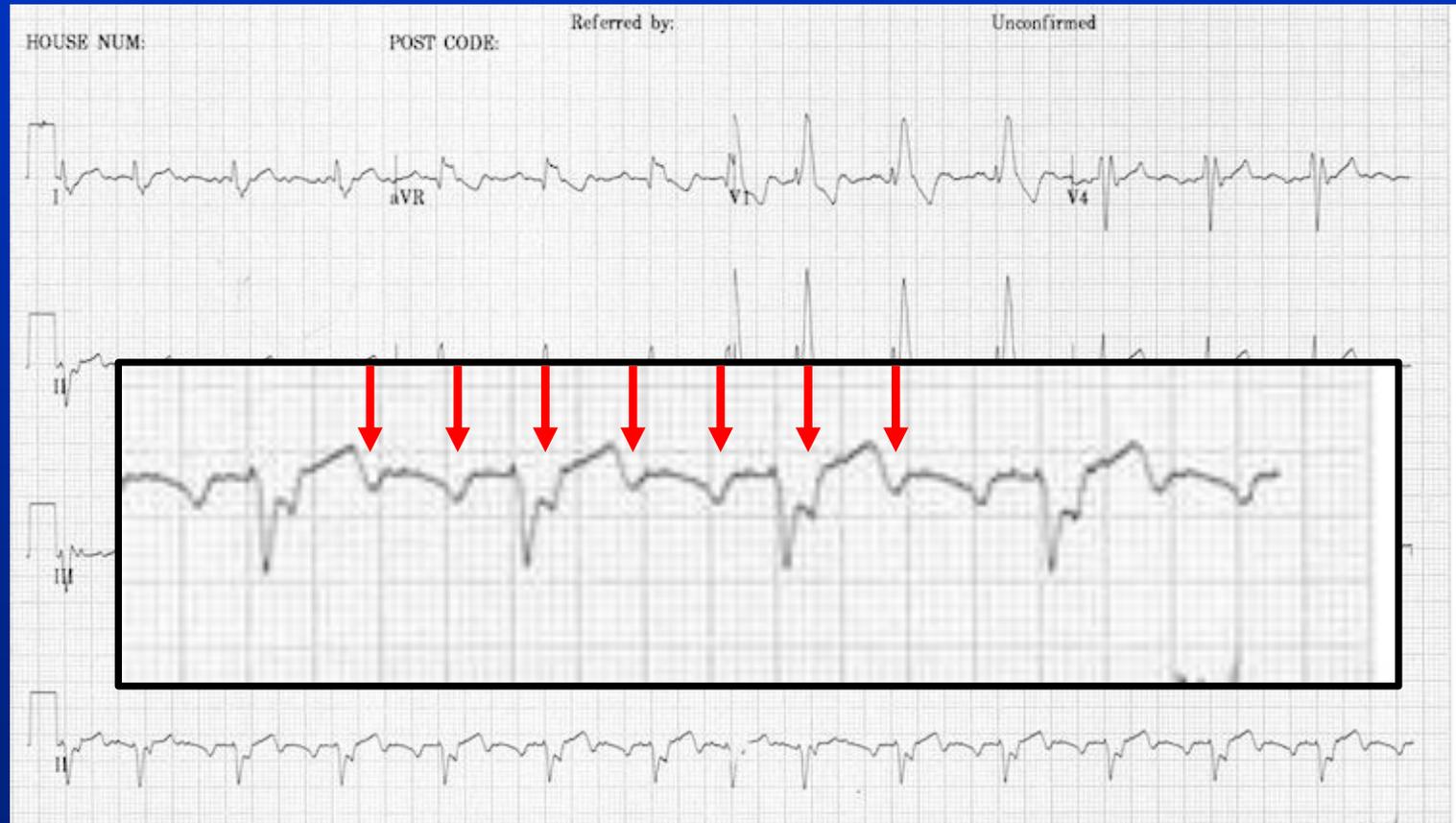
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4. Impulses propagate to rest of myocardium.

Premature impulse *



Atrial tachycardia

60yr old post-op VSD





The Fontan Flutter

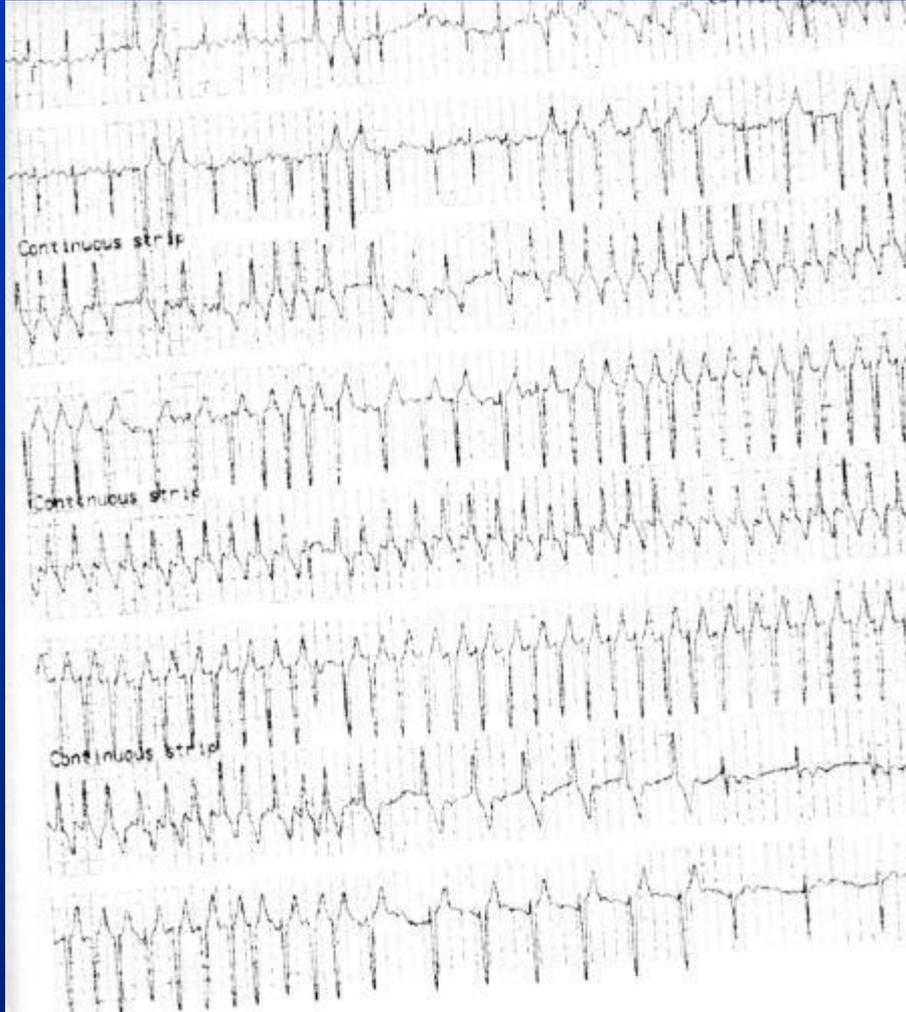
- Jan 1997 palpitations
 - seen in casualty dept.



Sinus rhythm
“neurotic”



The Fontan Flutter



Sinus rhythm
“neurotic”

24hr tape



The Fontan Flutter

- Jan 1997 palpitations
 - transferred UHW Cardiff
 - ECG classical “flutter” variable block
 - TOE: good haemodynamics, no clot, no spontaneous contrast

—————→

cardioversion 30J 60J to SR
- Warfarin added. “Normal” 48hr tape

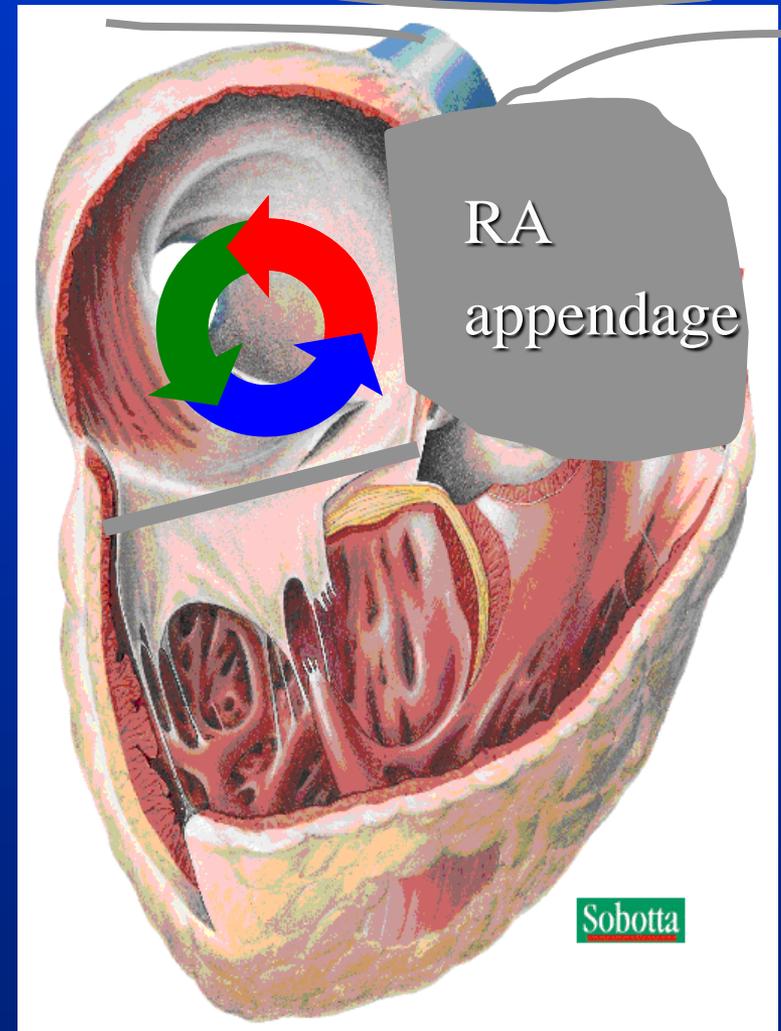


The Fontan Flutter

- Jan 1997
 - flutter Rx: cardioversion + warfarin
- Jan - Jun 1997
 - 2 episodes palpitations. Spontaneously resolved
- November 1997
 - flutter 1:1 conduction Rx amiodarone
 - 1:1 episodes frequent
 - cardiac catheter: PA pressure 12mmHg, no obstruction
- January 1998
 - EP study RF ablation- unsuccessful

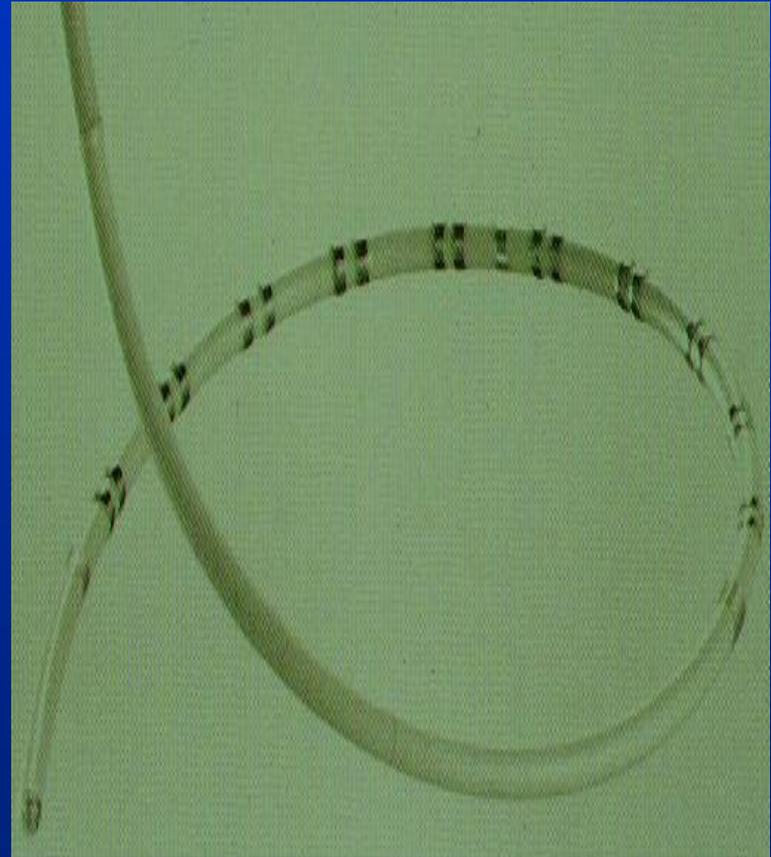
The Fontan Flutter

- Electrophysiology study
 - ? Arrhythmogenic focus
 - ? isthmus ablation
- Technique
 - Halo catheter
 - GA with TOE
 - 3 1/2 hrs 48 mins screening
 - 8 burns



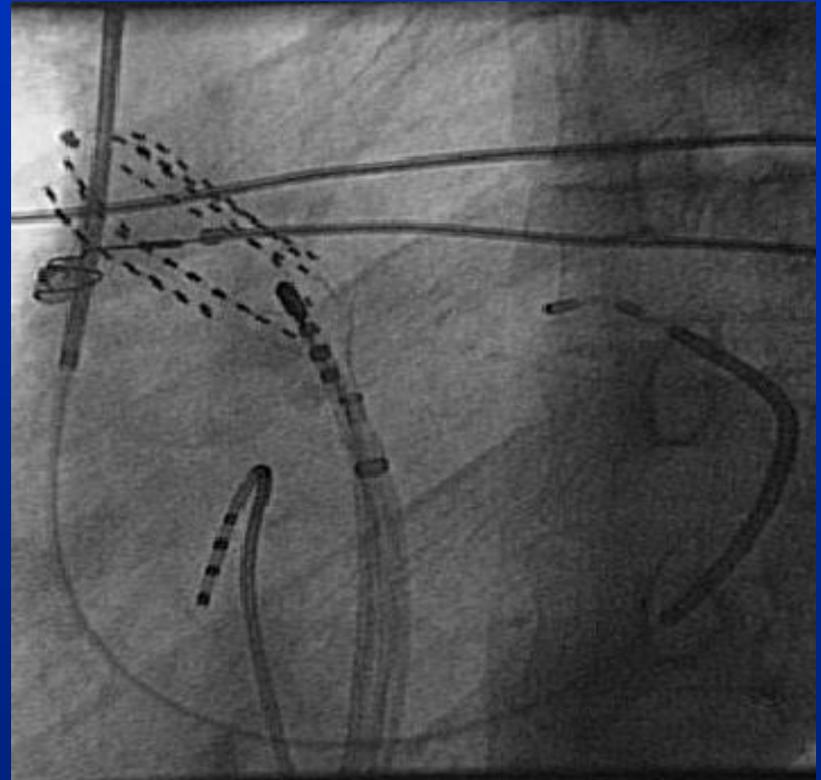
Halo Catheter

- 20 electrodes



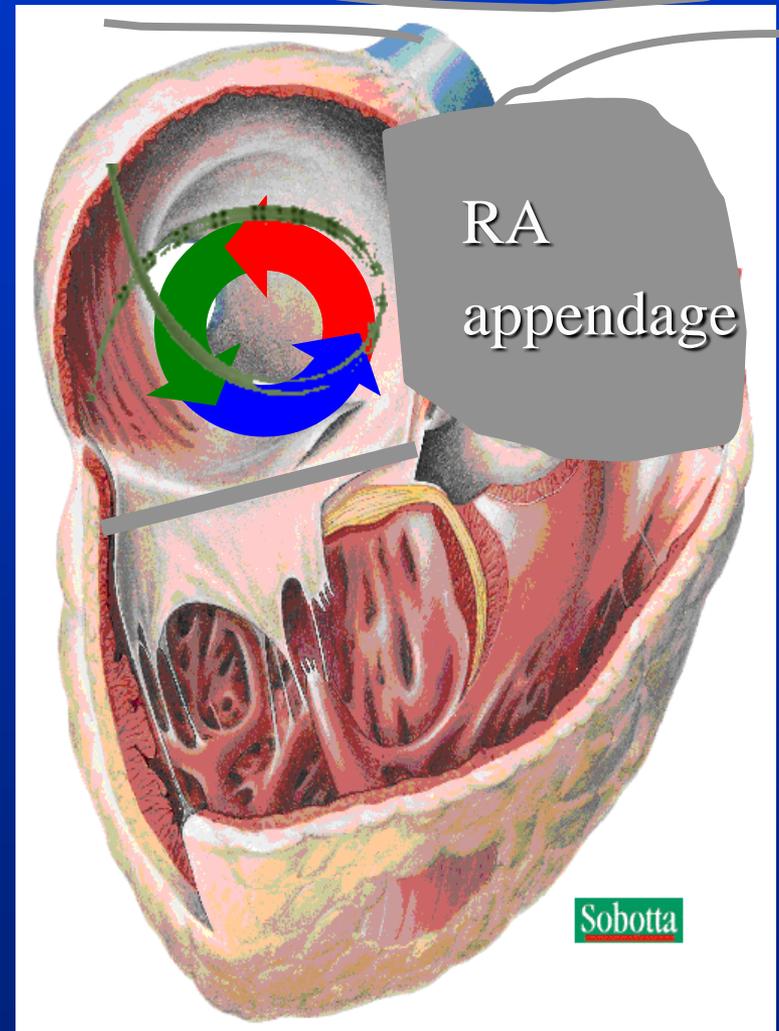
Constellation catheter

- 64 electrodes

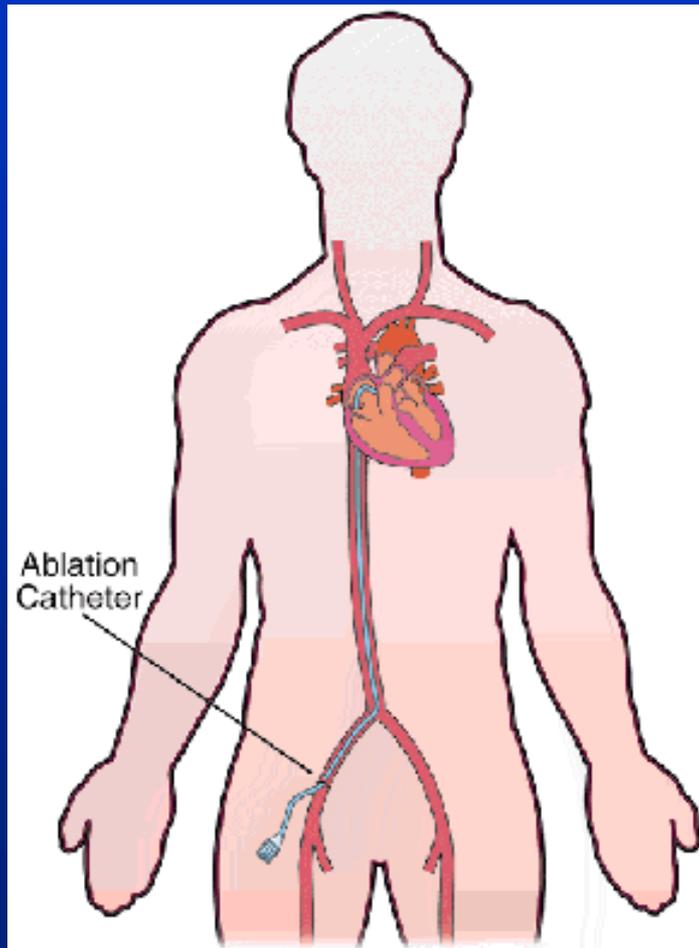


The Fontan Flutter

- Electrophysiology study
 - ? Arrhythmogenic focus
 - ? isthmus ablation
- Technique
 - Halo catheter
 - GA with TOE
 - 3 1/2 hrs 48 mins screening
 - 8 burns



Advances in RF Ablation





The Fontan Flutter

- January 98 flutter RFA ablation failed Rx verapamil, digoxin
- March 98 frequent disabling flutters Intertach II
- Aug 98 64 pace terminations; feels back to "normal"
- 1999 different atrial tachycardias



The Fontan Flutter

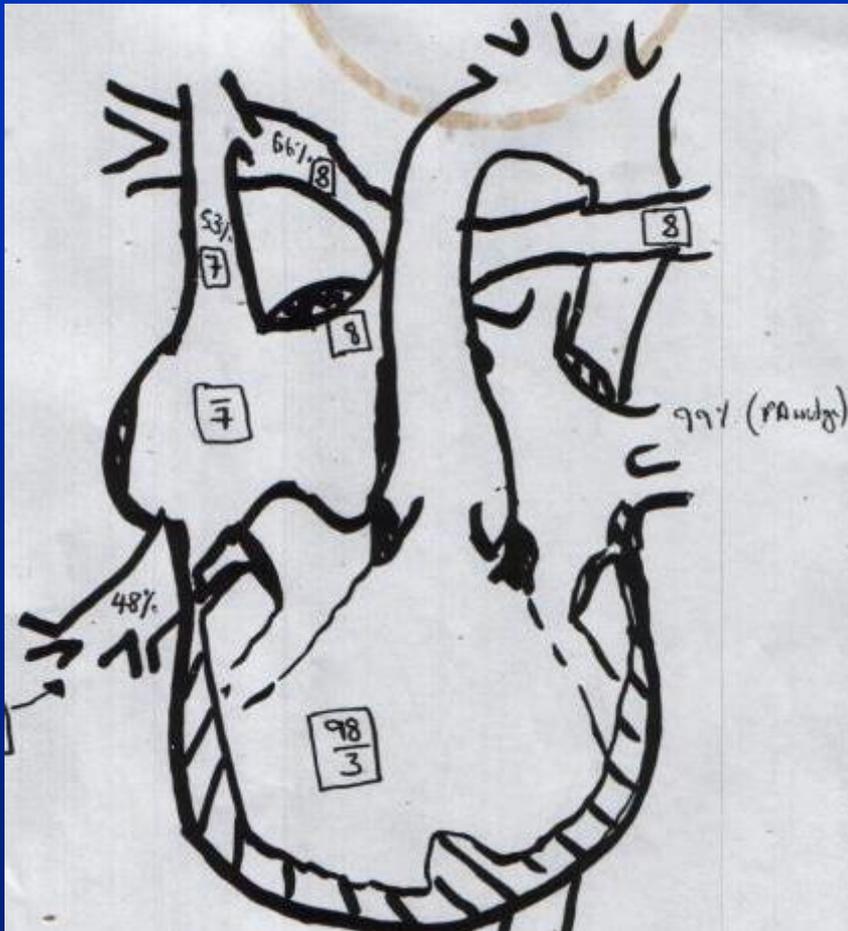
- 1999 3 admissions
- 2000 “lots of tachycardias”
- 2001 “lots more”
- 2002



The Fontan Flutter

- 1999 3 admissions
- 2000 “lots of tachycardias”
- 2001 “lots more”
- 2002 **aaaargh !!!”**

Diagram of Anatomy and Contrast Injection

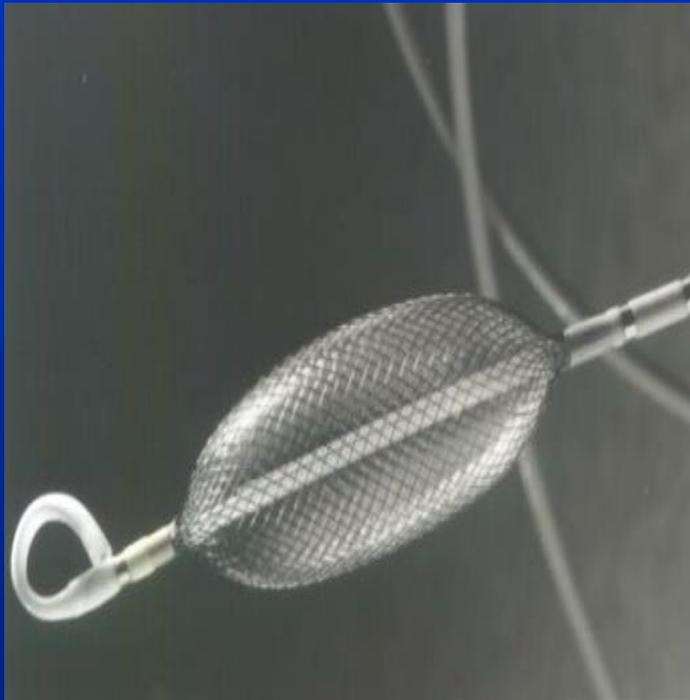


Im: 90
DFOV mm

512

kV 66.0
mA: 298
mm
Tilt: degrees
13 ms
110737
W=137,L=81

Endocardial Solutions

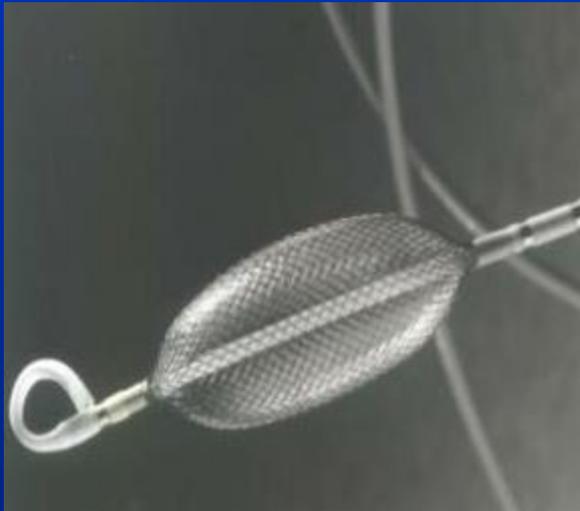


Multielectrode array catheter



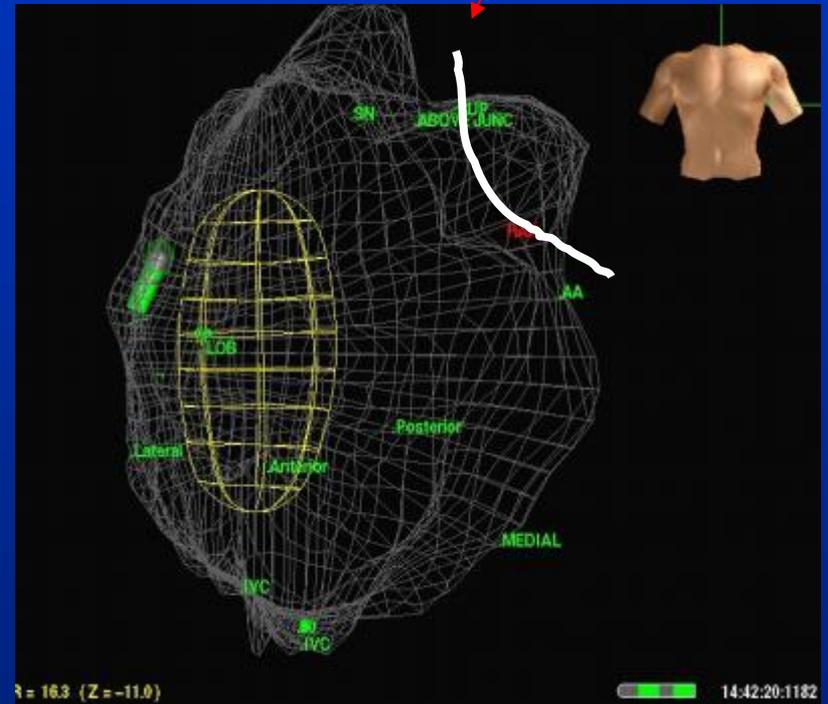
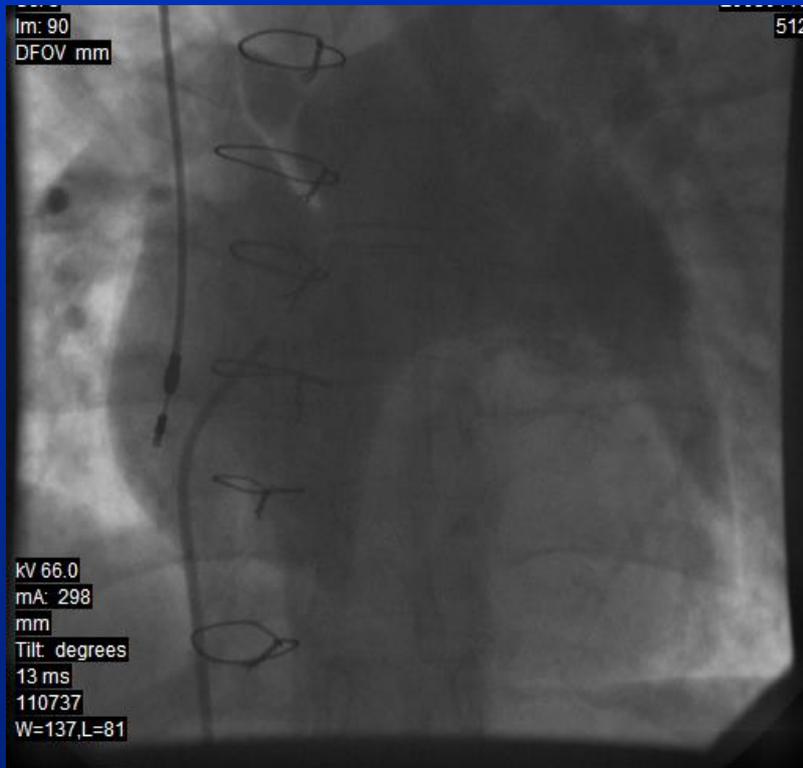
Laser etched

Endocardial Solutions

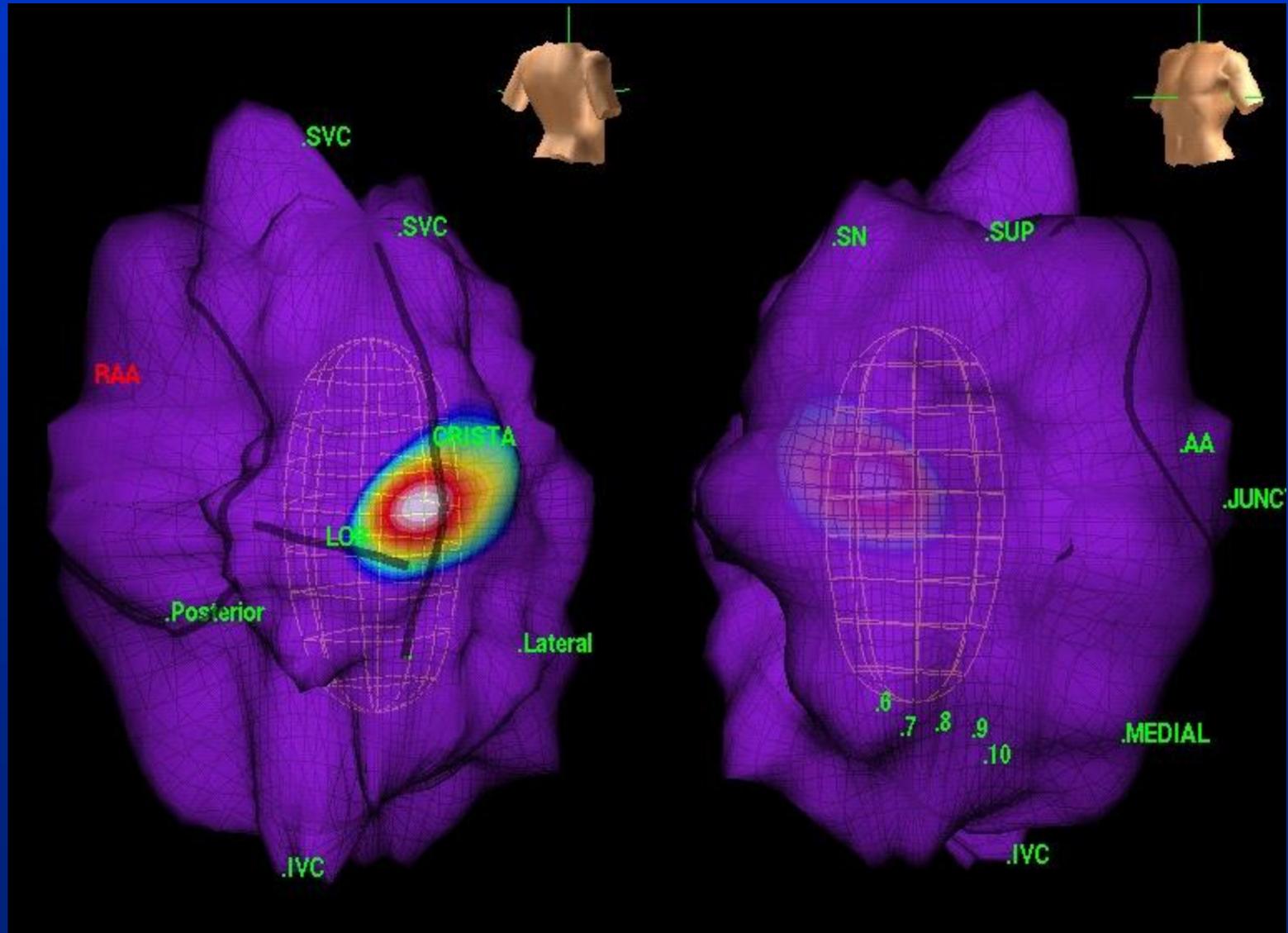


MEA / Contrast

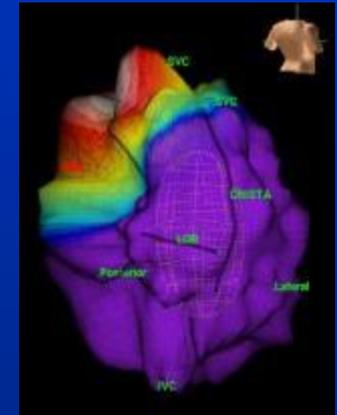
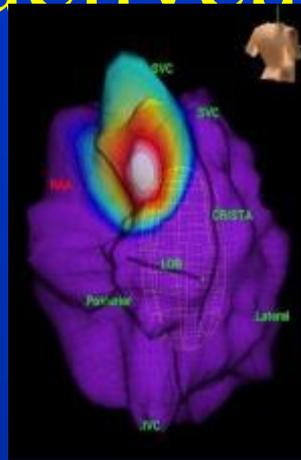
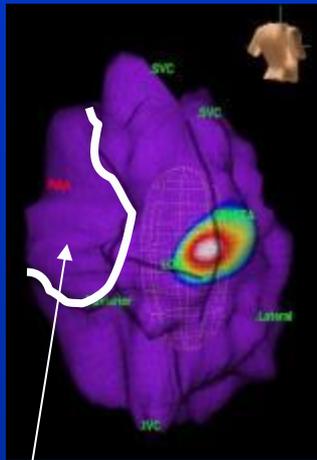
Border of
Appendage



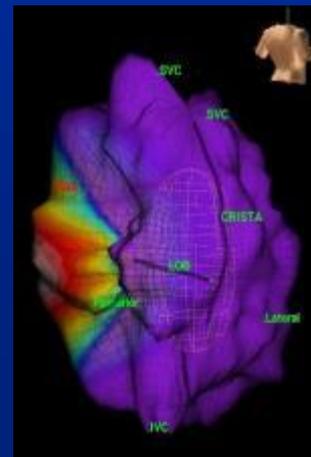
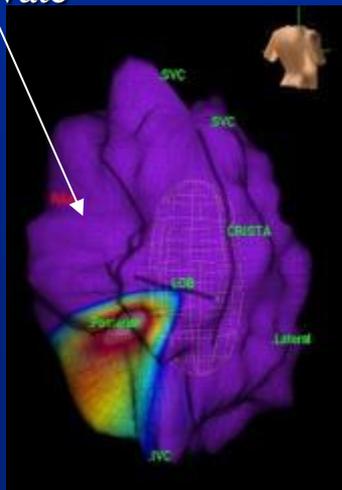
Atrial Tachycardia after Fontan



Focal Tachycardia (1)



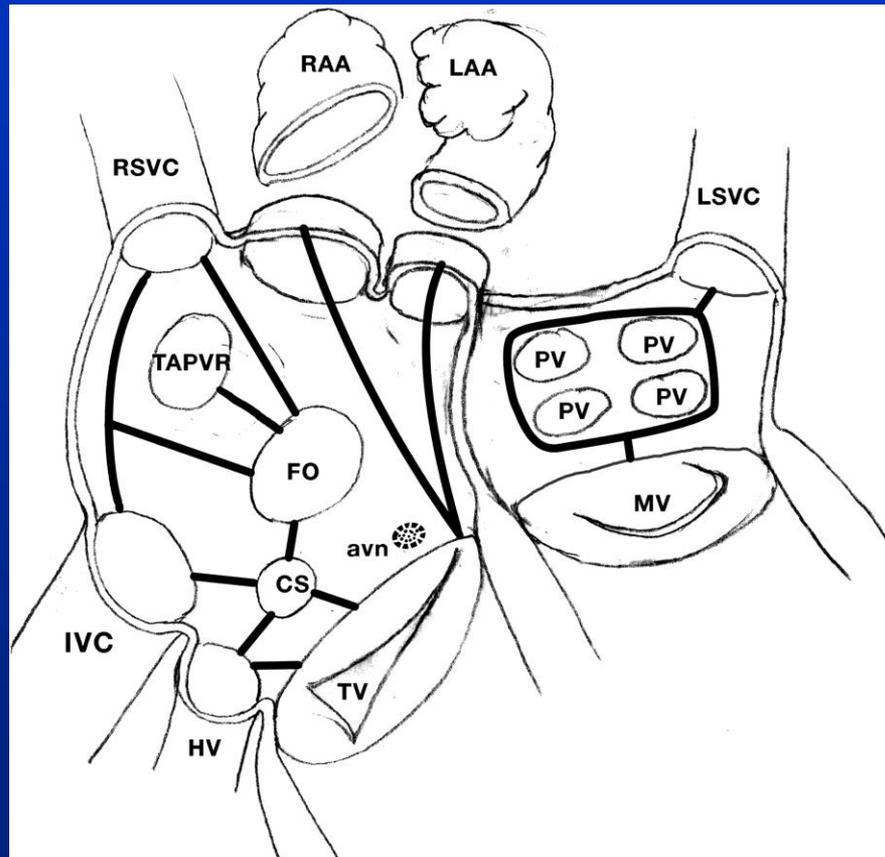
Note: Atrial Appendage does not activate



Ablation in TGA

“Catheter ablation is often considered first
?? Redo Surgery better than
Drugs or ablation in Fontan
arrhythmias

Arrhythmia Surgery in Fontan



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 +

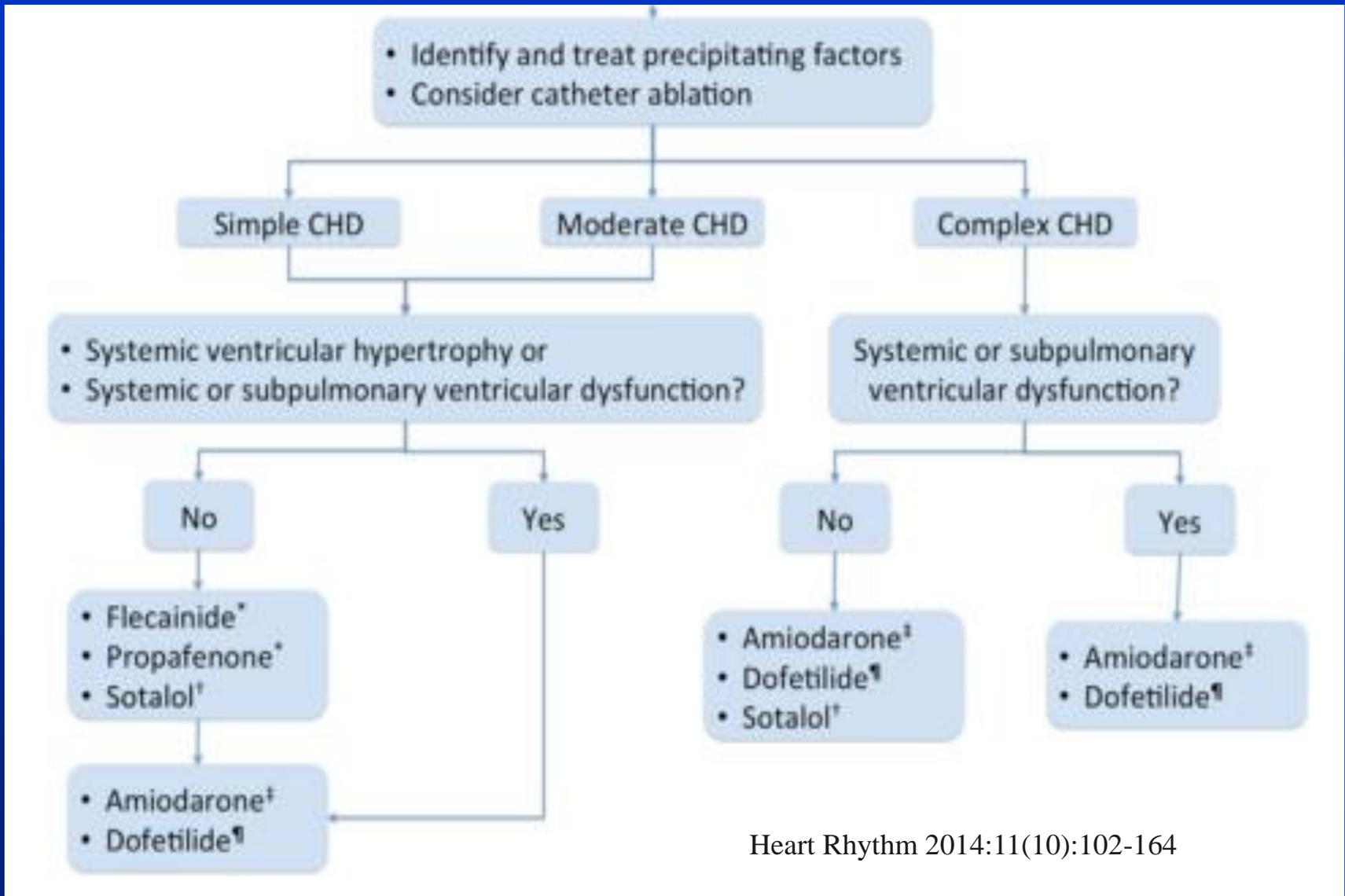
High risk CHD substrate eg TGA/TOF/Single vent

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



Learning point !

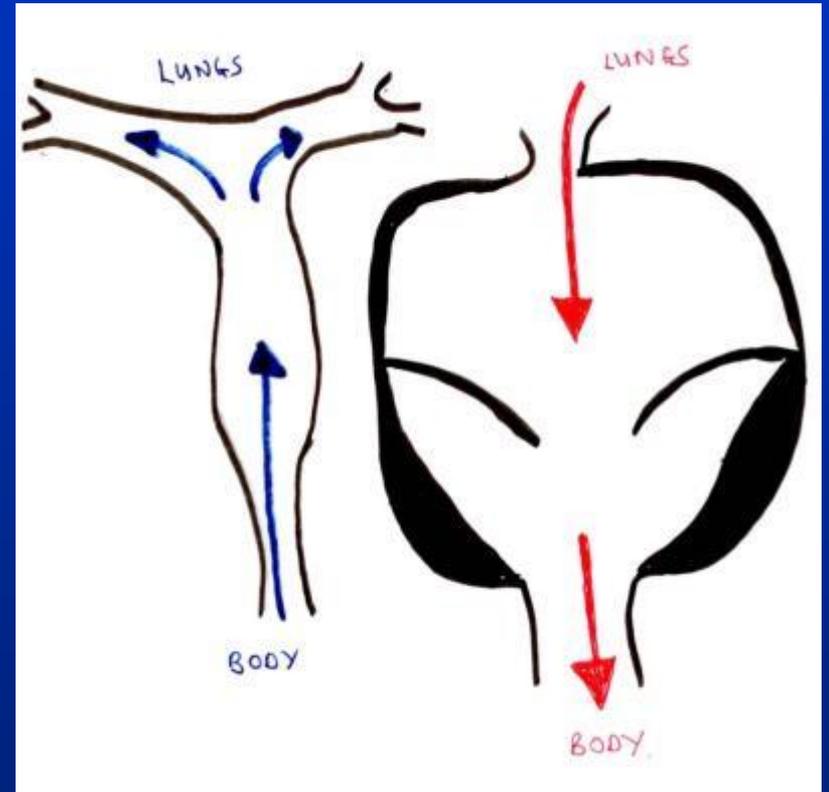


Mechanical events
can have electrical
consequences !

= consider haemodynamics

Prof Francis Fontan

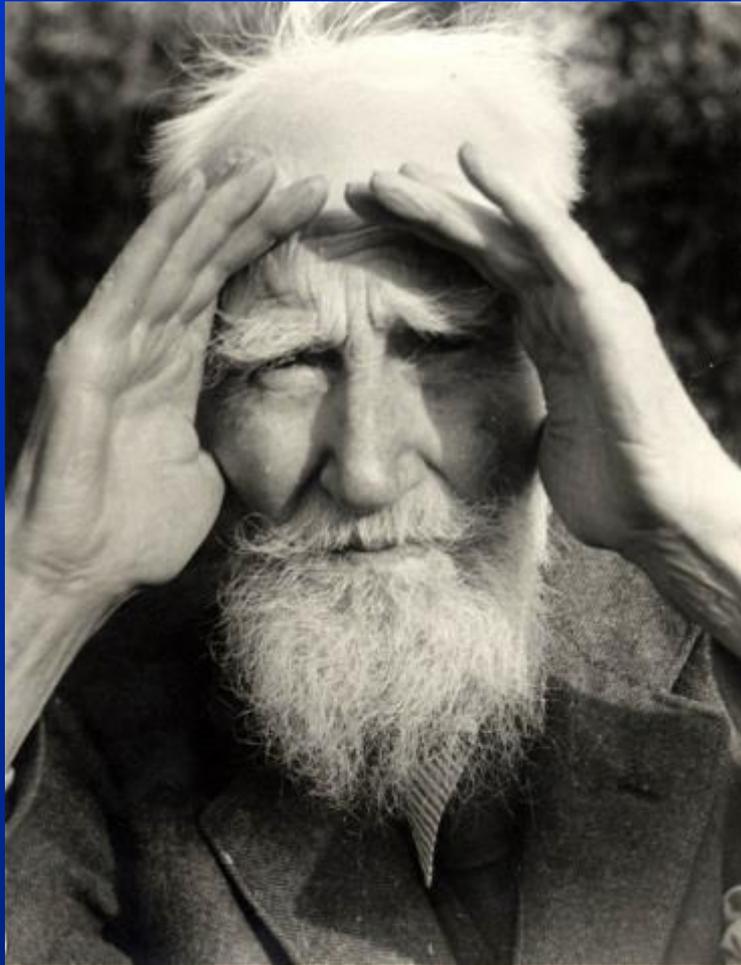
1929-2018



Fontan Surgery in Complex CHD

a success story!





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

George Bernard Shaw
1856 -1950

Any Questions?



Any Questions?

