Atrial Tachycardia in ACHD Patients Epidemiology and Evaluation

Alpay Çeliker M.D

Pediatric and Congenital Rhythm Congress VII

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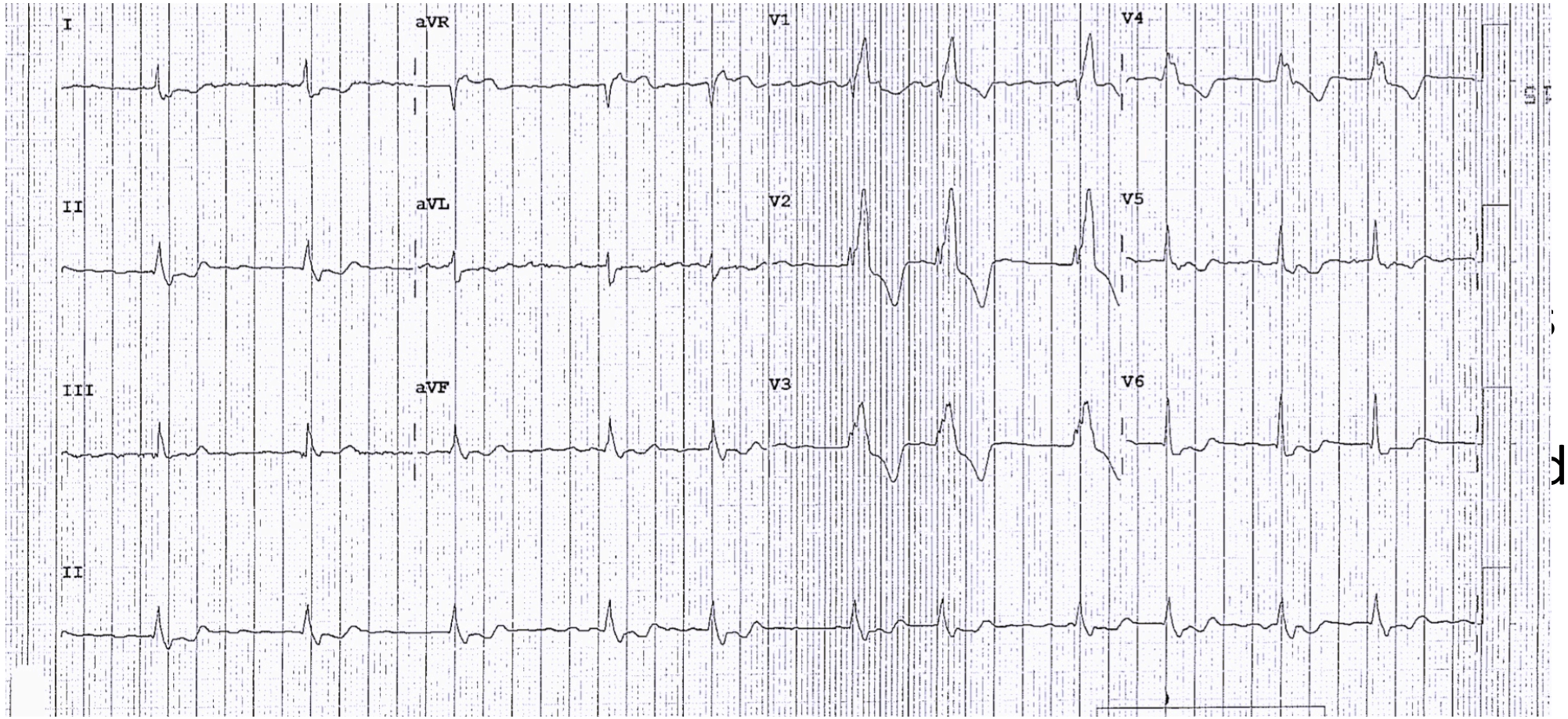
Case Presentation

- 

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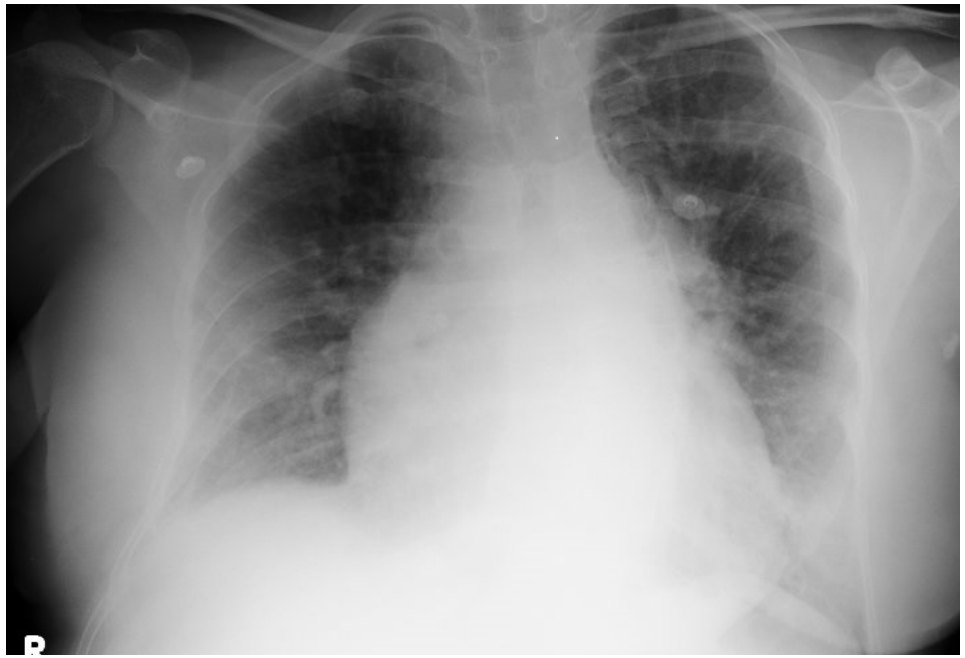
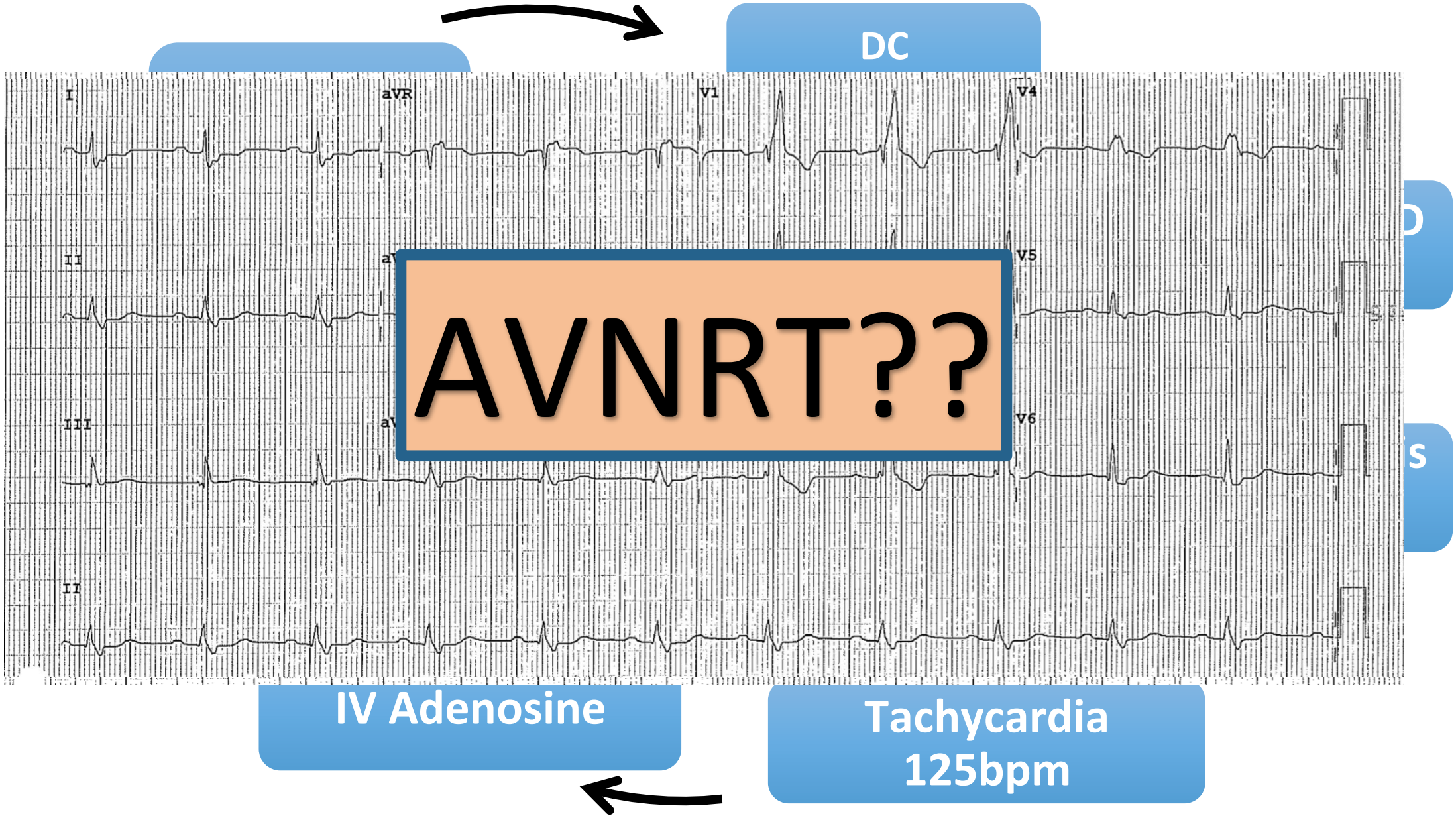
- d.

Exam

- Irregular heart beat
- Tachypneic and unable to lying down
- Distended jugular veins
- Grade 2-3/6 systolic and early diastolic murmur at the left second ICS
- Crackles at the both lung basal area
- 3cm of hepatomegaly, no ascites
- Mild edema




DC
D
S
AVNRT??
IV Adenosine
Tachycardia
125bpm

Journal of the American College of Cardiology

Published by Elsevier Inc.

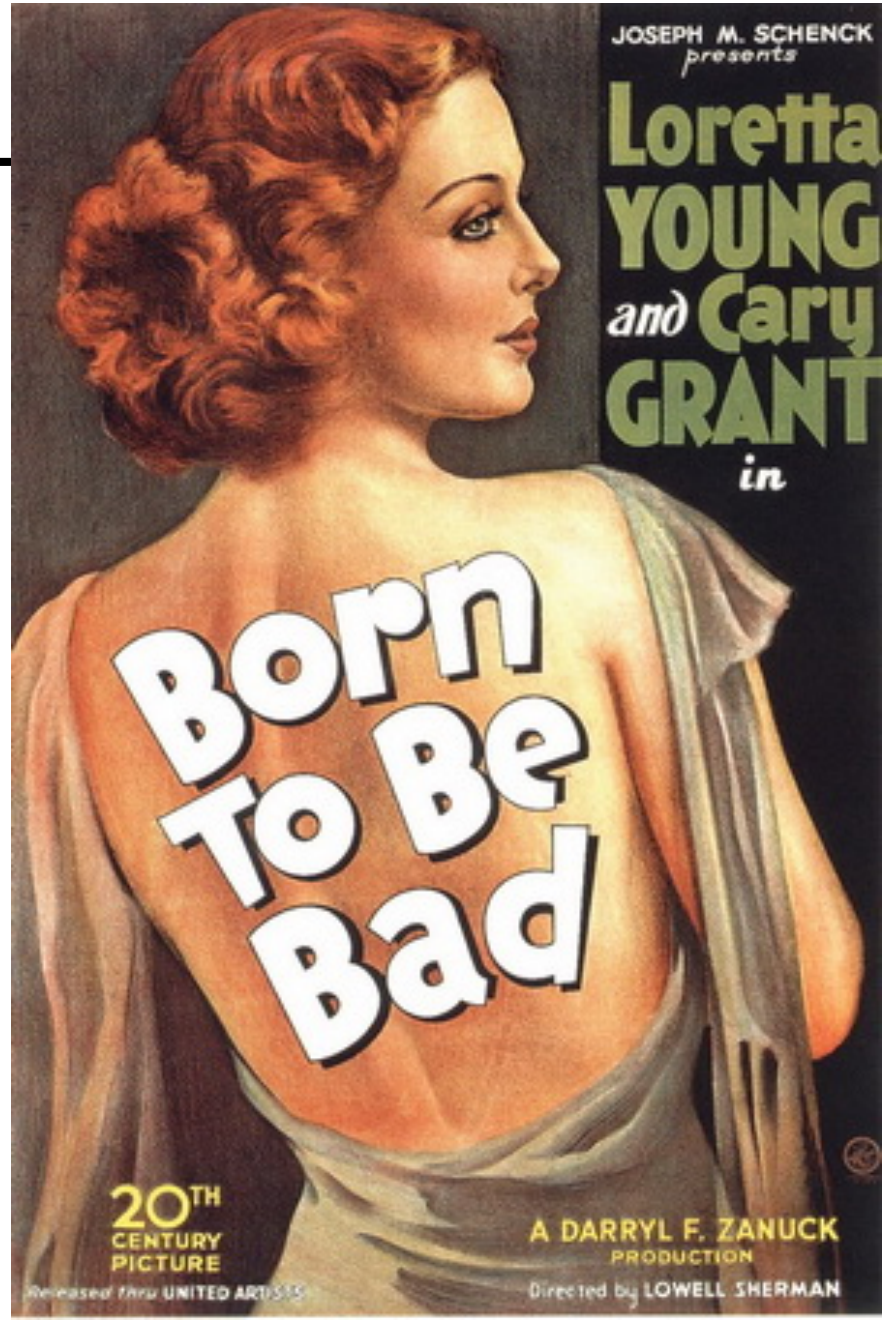
STATE-OF-THE-ART PAPER

The Adult With Congenital Heart Disease

Born to Be Bad?

Carole A. Warnes, MD, MRCP, FACC
Rochester, Minnesota

*In conclusion, are these patients "**born to be bad**"? In many ways, the answer is yes. They are seldom "cured" by surgery and continue to have cardiac problems.* The cardiology community serves them poorly, and, as we look to the future, we must make provision for lifelong care by trained physicians with expertise in their complex problems.



Arrhythmias in adults with congenital heart disease: What are risk factors for specific arrhythmias? Loomba SH et al. PACE 2016

- 27,088/ 109,168 (25%) patients with CHD, had an arrhythmia at some point.
- In those with an arrhythmia
  - **Atrial fibrillation in 85.7%.**
  - **Atrial flutter (19.5%)**
  - **Ventricular tachycardia (5.4%)**
- The largest burden of arrhythmia
  - **Tricuspid atresia with a 51% prevalence**
  - **Ebstein anomaly with a 39% prevalence.**
- Increasing age, male gender, heart failure, obstructive sleep apnea, are additional risk factors

Background

- Supraventricular tachyarrhythmia is the most common form of arrhythmia in ACHD patients, with an estimated 50 % lifetime risk for patients entering their third decade of life.
- The most common mechanism:
  - **Intraatrial reentrant tachycardia (IART)**
  - **Non- automatic focal atrial tachycardia (NAFAT)**
  - **Atrioventricular reentrant tachycardia (AVRT) mediated by twin atrioventricular (AV) nodes, typical forms of AVRT**
  - **AV node reentrant tachycardia**
  - **Atrial fibrillation (AF).**
- While almost all CHD carries a risk for SVA, there are frequent associations owing to unique embryology and subsequent surgical repair.

CHD and Arrhythmias

Table 1
Spectrum of bradyarrhythmias and tachyarrhythmias seen with selected CHD subtypes

CHD Type	IART	AF	WPW	VT/SCD	SA Node Dysfunction	Spontaneous AV Block	Acquired AV Block
VSD	+	—	—	+	—	—	+
ASD	++	+	—	—	—	—	—
TOF	+++	—	—	++	—	—	+
Aortic stenosis	—	+	—	++	—	—	+
d-TGA (Mustard or Senning)	+++	—	—	++	+++	—	—
CAVC	+	—	—	—	—	+	++
Fontan	+++	++	—	+	+++	—	—
l-TGA	+	—	+	+	—	++	+++
Ebstein's anomaly	++	—	+++	++	—	—	—

Incidence of SVT in relation to congenital defect and type

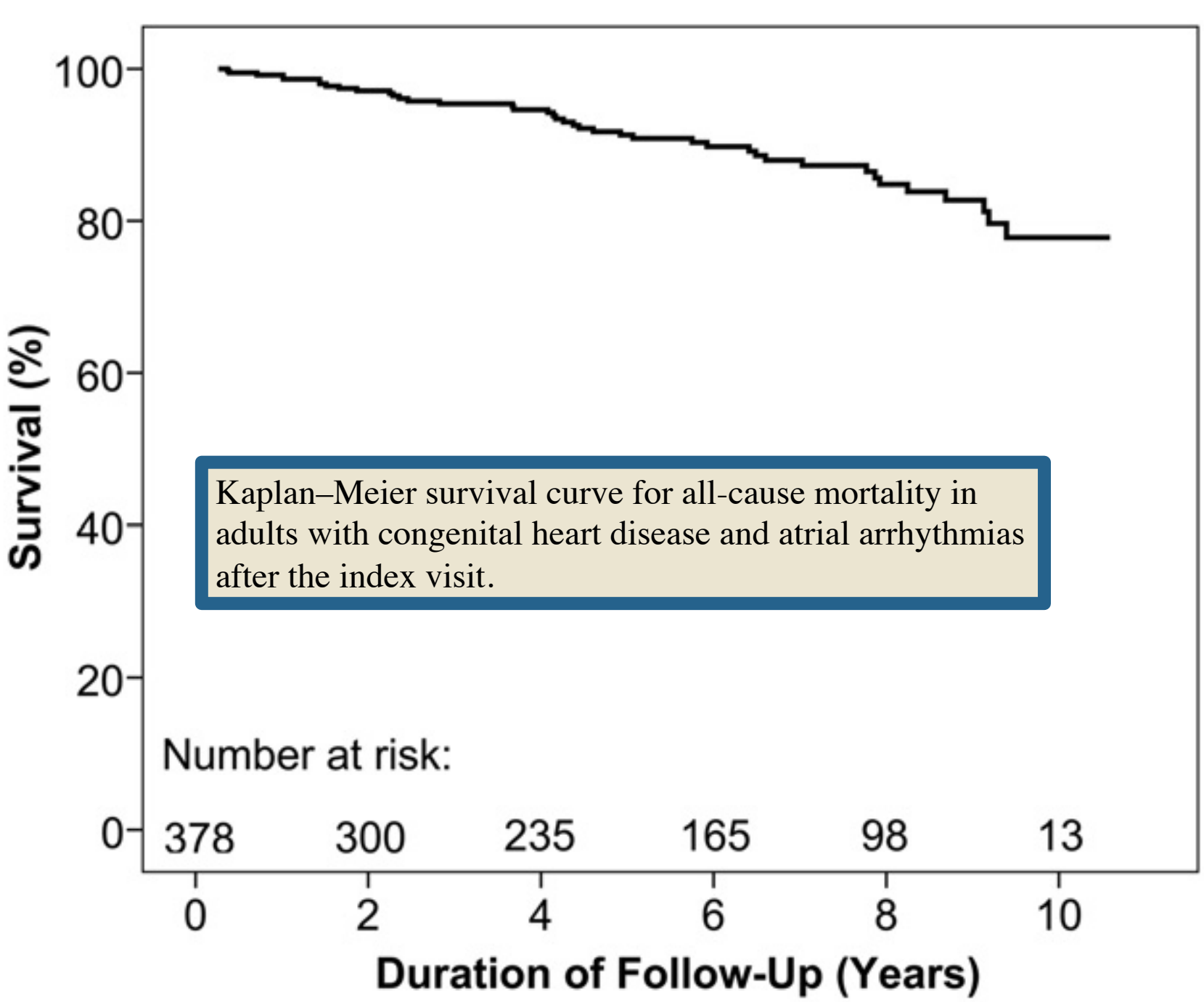
Congenital defect	Type of surgery	Incidence
ASD	Uncorrected/corrected as adult Surgical closure prior to age 15	50% 16%
TGA	Mustard/Senning Arterial switch surgery	28%–37% 5%
Single ventricle	Atriopulmonary Fontan Lateral tunnel Extracardiac tunnel	50% >10 yrs, 100% ≥26 yrs 13% after 10 yrs 0% after 10 yrs
Tetralogy of Fallot		Age-dependent up to 20%

Atrial arrhythmia & CHD type

- 378 adult patients with CHD (mean age 39 years) and atrial arrhythmias
- During a median follow-up of 5.2 years, there were 40 deaths (11%).
- Common modes of death included heart failure–related death (35%), sudden cardiac death (20%), and perioperative death (18%).
- Intra-atrial reentrant tachycardia 239 (63%) and atrial fibrillation 128 (34%)

Anomaly	N (%)
Single Ventricle	75 (20%)
Repaired TOF	58 (15%)
TGA	53 (14%)
ASD	40 (11%)
AVSD	25 (7%)
L-TGA	20 (5%)
Ebstein's	8 (5%)
VSD	15 (4%)
Others	74 (20%)

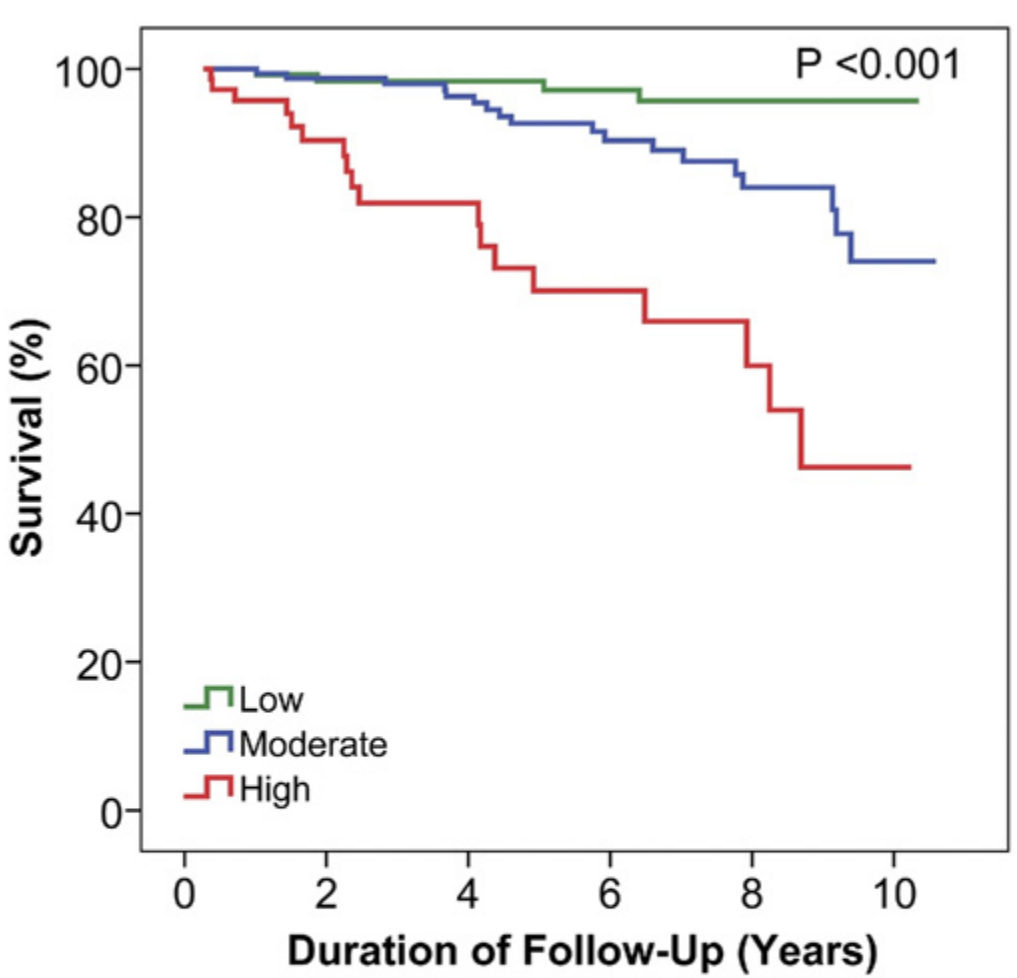
Identifying High Risk in Adults With Congenital Heart Disease and Atrial Arrhythmias. Am J Cardiol 2011



Kaplan–Meier survival curve for all-cause mortality in adults with congenital heart disease and atrial arrhythmias after the index visit.

In conclusion, in adult with CHD and atrial arrhythmias specific clinical variables identify patients at high risk for death. Importantly, the absence of any of these risk factors is associated with an excellent survival despite the presence of atrial arrhythmias.

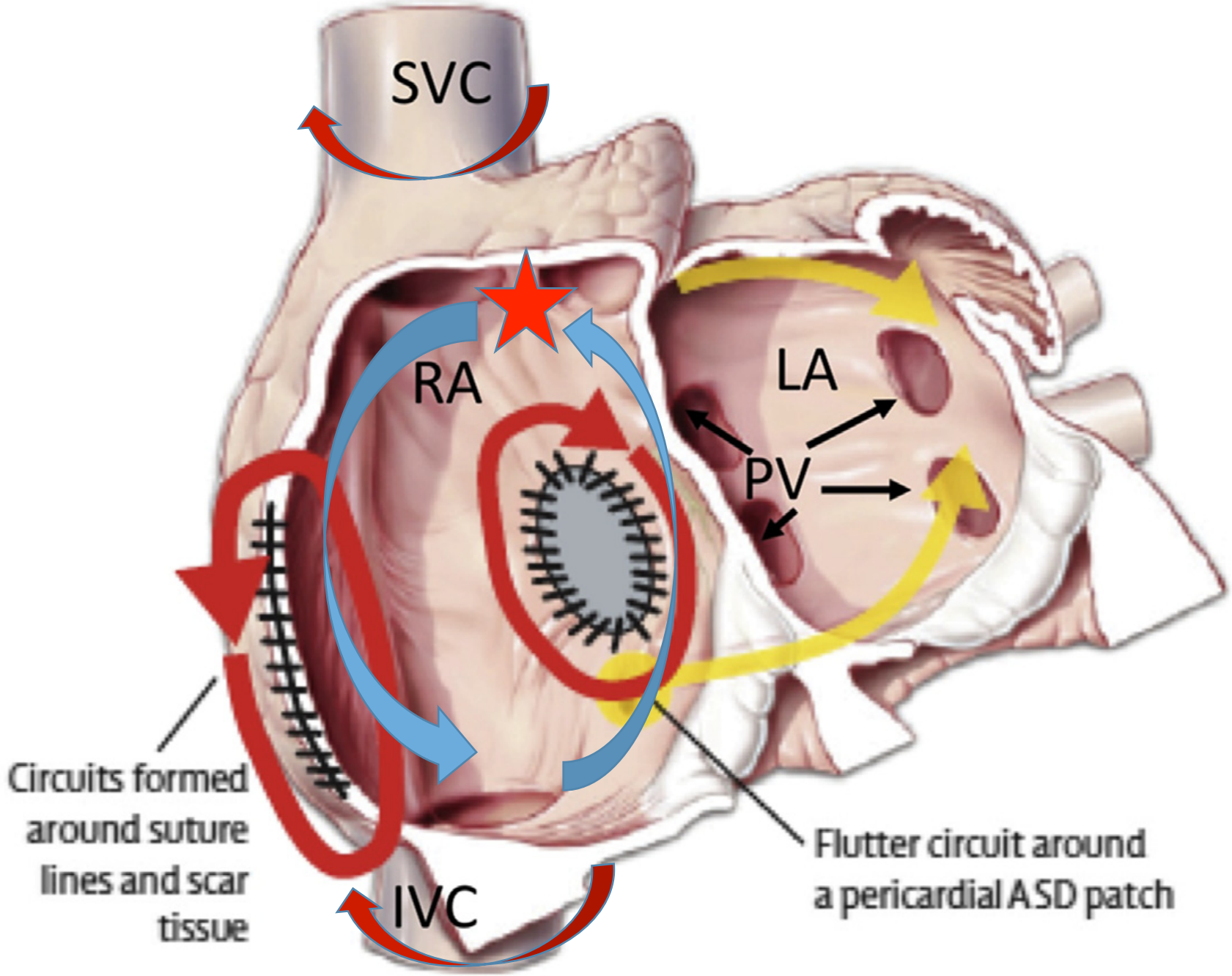
Risk Factors:
Valvular heart disease
Pulmonary hypertension
Single-ventricle physiology
New York Heart Association functional class ≥ III



Risk score	Risk category	N	Annualized Rate of Death
0	Low	133	0.5%
1	Moderate	175	1.9%
2-4	High	73	6.5%

Mechanism of Atrial Tachycardia

- Central obstacles:
  - Native structures such as the tricuspid valve annulus and vena cavae
  - Artificial structures created by prior surgical manipulation
    - Sutures
    - Scars
    - Patches
    - Incisions
- Fibrosis due to altered hemodynamics in the late postoperative setting.


SVC
RA
LA
PV
IVC
Circuits formed around suture lines and scar tissue
Flutter circuit around a pericardial ASD patch

Problems at Fontan Circulation

- Heart failure
- **Arrrhythmias**
- Thromboembolic complications
- Protein-losing enteroptahy

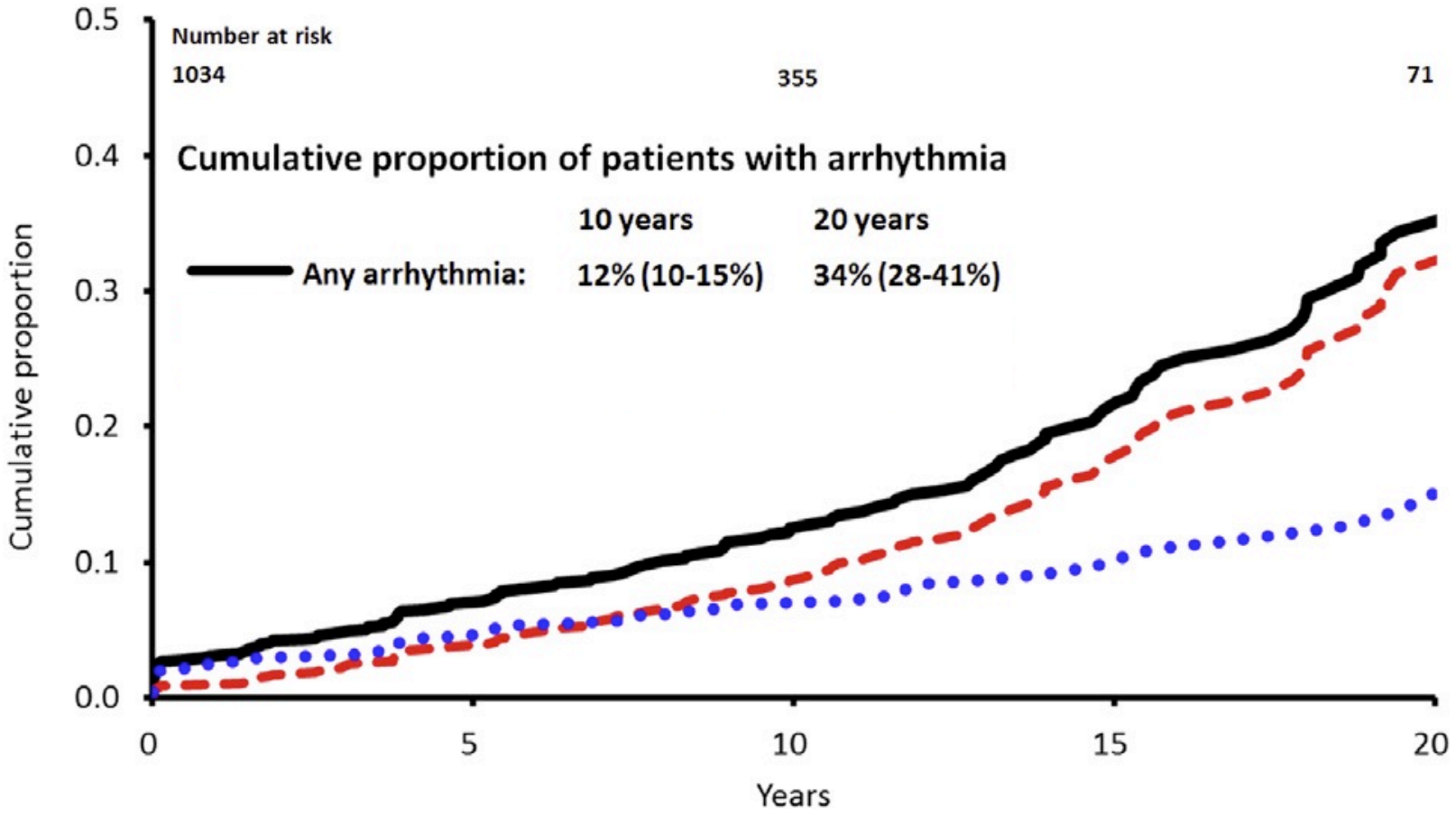
Thorax (1971), **26**, 240.

Surgical repair of tricuspid atresia

F. FONTAN and E. BAUDET

Centre de Cardiologie, Université de Bordeaux II, Hôpital du Tondu, Bordeaux, France

Surgical repair of tricuspid atresia has been carried out in three patients ; two of these operations have been successful. A new surgical procedure has been used which transmits the whole vena caval blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, 'ventriclized', to direct the inferior vena caval blood to the left lung, the right pulmonary artery receiving the superior vena caval blood through a cava-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries, which must be large enough and at sufficiently low pressure to allow a cava-pulmonary anastomosis. The indications for this procedure apply only to children sufficiently well developed. Younger children or those whose pulmonary arteries are too small should be treated by palliative surgical procedures.



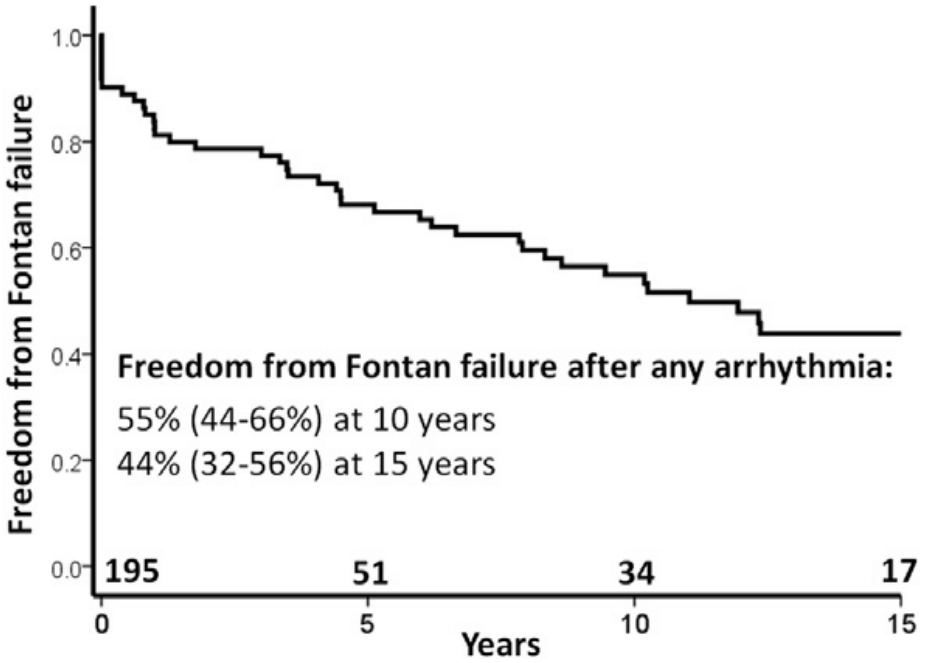
Australian and New Zealand Fontan Registry 1034 pt
Carins TA ve ark. J Thorac Cardiovasc Surg 2016;152:1355-63)

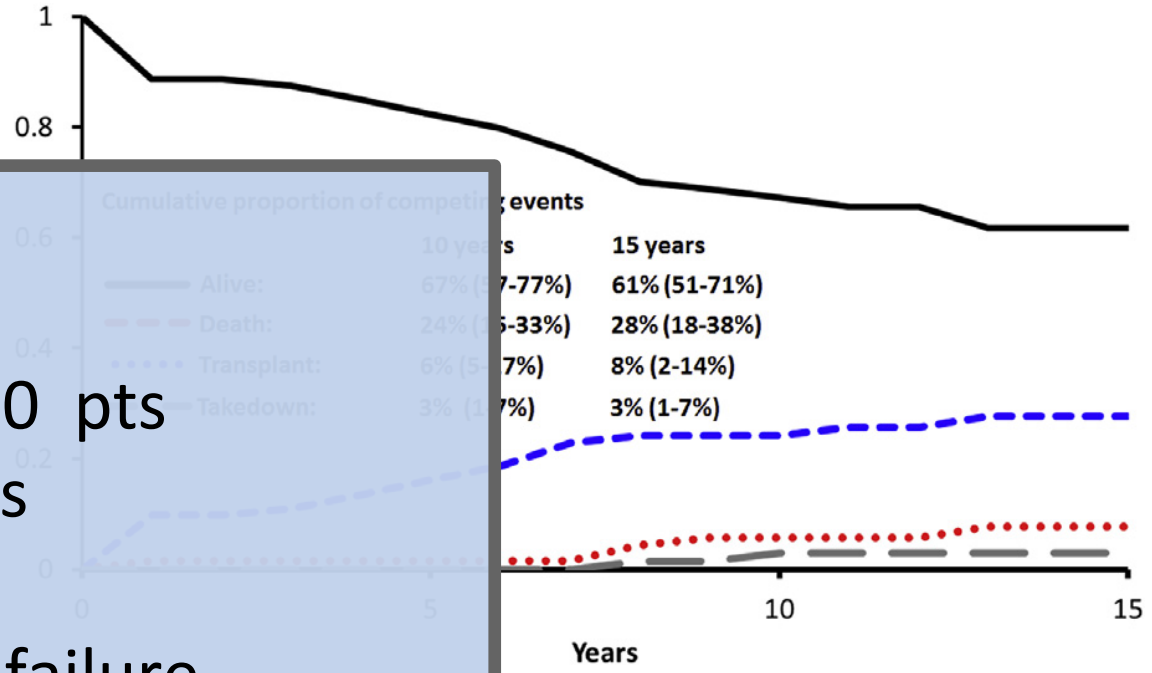
195 patient with tachycardia 162, bradyarrhythmia 74 patient, both 41 patient
Survival at 20 years without arrhythmia 66%
Extracardiac Fontan's had the lowest incidence
İzomerism leads to more arrhythmia

Predictors of developing arrhythmia post-Fontan procedure

Variable	HR	95% CI lower	95% CI upper	P value
Any arrhythmia				
ECC Fontan (vs AP)	0.23	0.10	0.51	**<.001**
LT Fontan (vs AP)	0.60	0.31	1,01	0.055
LA isomerism	3.18	.456	6.95	.004
RA isomerism	4. 00	2. 41	6. 61	**<.001**
Tachycardia				
ECC Fontan (vs AP)y.	0. 34	0.13	0. 92	033
LA isomerism (vs none)	4.17	1.77	9.79	**.001**
RA isomerism (vs none)	4.61	2.70	7.86	**<.001**
Dextrocardia	1.64	0.99	2.70	.054
Bradycardia				
ECC Fontan (vs AP)	0.06	0.02	0.20	**<.001**
LT Fontan (vs AP)	0.23	0.09	0.58	.002

Prognosis after the arrhythmia development





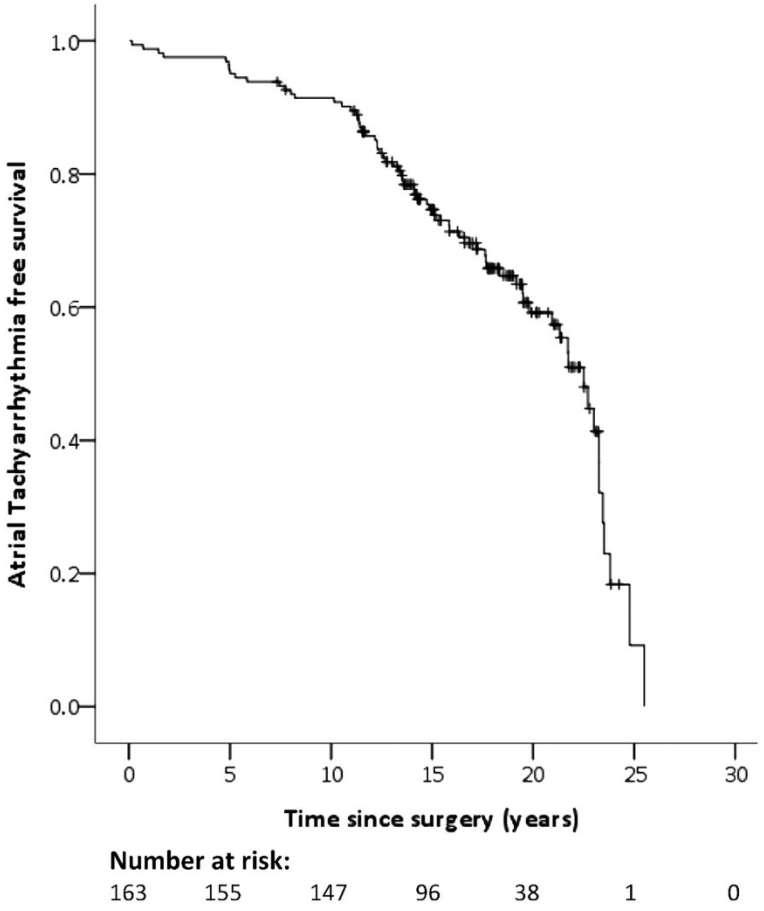
Death at 33 pts
HTX at 12 pts
Fontan conversion at 30 pts
Fontan take-down 3 pts
PLE in 12 pts
NYHA Class III-IV heart failure
Total 84 patient had Fontan problem

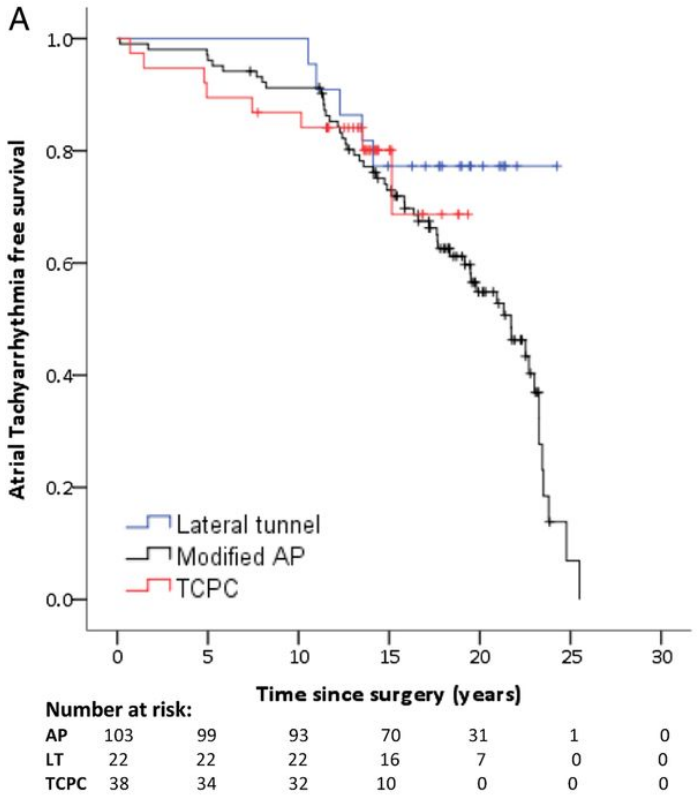
Impact of arrhythmia (as a time-varying covariate) on late outcomes: Multivariable Cox proportional hazards regression

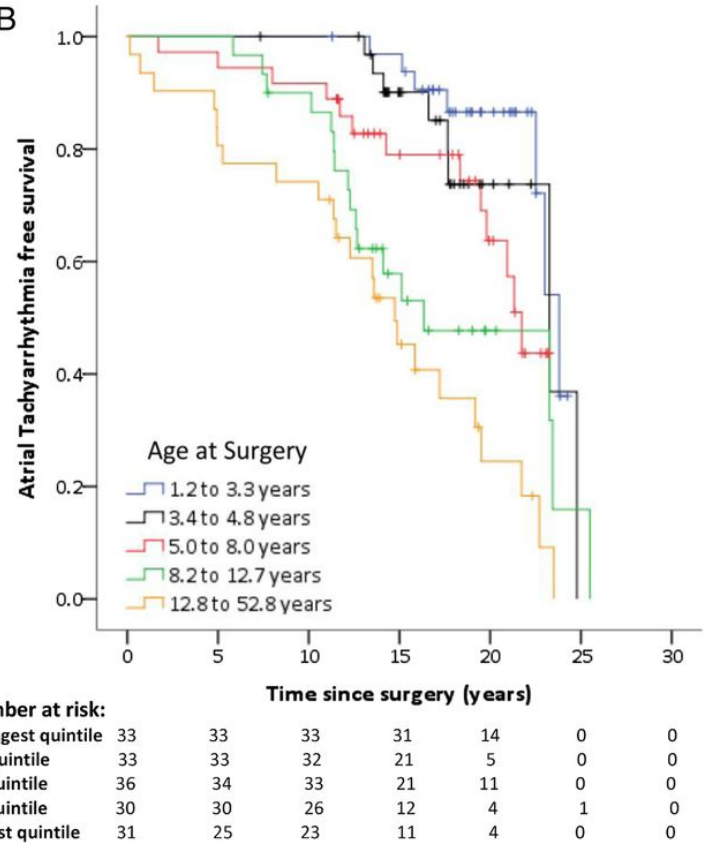
Event	HR	95% CI lower	95% CI upper	P value
Risk of event after developing any arrhythmia				
Death	1.53	0.85	2.76	.16
Death/transplantation	1.76	1.02	3.02	.042
Fontan failu				**1**
Risk of ever				
Death				
Death/transplantation	1.87	1.02	3.45	.043
Fontan failure	**2.56**	**1.60**	**4.11**	**<.001**
Risk of event after developing bradycardia				
Death	1.20	0.62	2.30	.59
Death/transplantation	1.81	1.04	3.15	.037
Fontan failure	1.85	1.16	2.95	.010

CONCLUSIONS

After the first onset of arrhythmia, patients with a Fontan circulation are at increased risk of late Fontan failure.







Quinton E et al. Prevalence of atrial tachyarrhythmia in adults after Fontan operation. Heart 2015;101:1672–1677.

Prevalence of atrial tachyarrhythmia in adults after Fontan operation

	Total	No arrhythmia	Arrhythmia	p Value
N	**166**	**96**	**70**	
Mean current age (years)	29.1 (±9.2)	25.3 (±5.6)	34.4 (±10.5)	<0.001
Mean age at Fontan (years)	9.0 (±8.5)	6.4 (±5.4)	12.6 (±10.5)	<0.001
Median age at Fontan (years)	5.6 (3.8–11.9)	4.8 (3.3–7.6)	11.0 (5.3–14.9)	<0.001
Type of Fontan				
Modified AP	104(63%)	48 (50%)	56 (80%)	
Lateral tunnel	22 (13%)	17 (18%)	5 (7%)	
TCPC	40 (24%)	31 (32%)	9 (13%)	
Mean follow-up (years)	18.6 (±4.7)	17.3 (±3.7)	20.5 (±5.2)	
Mortality	21 (13%)	9 (9%)	12 (17%)	0.21

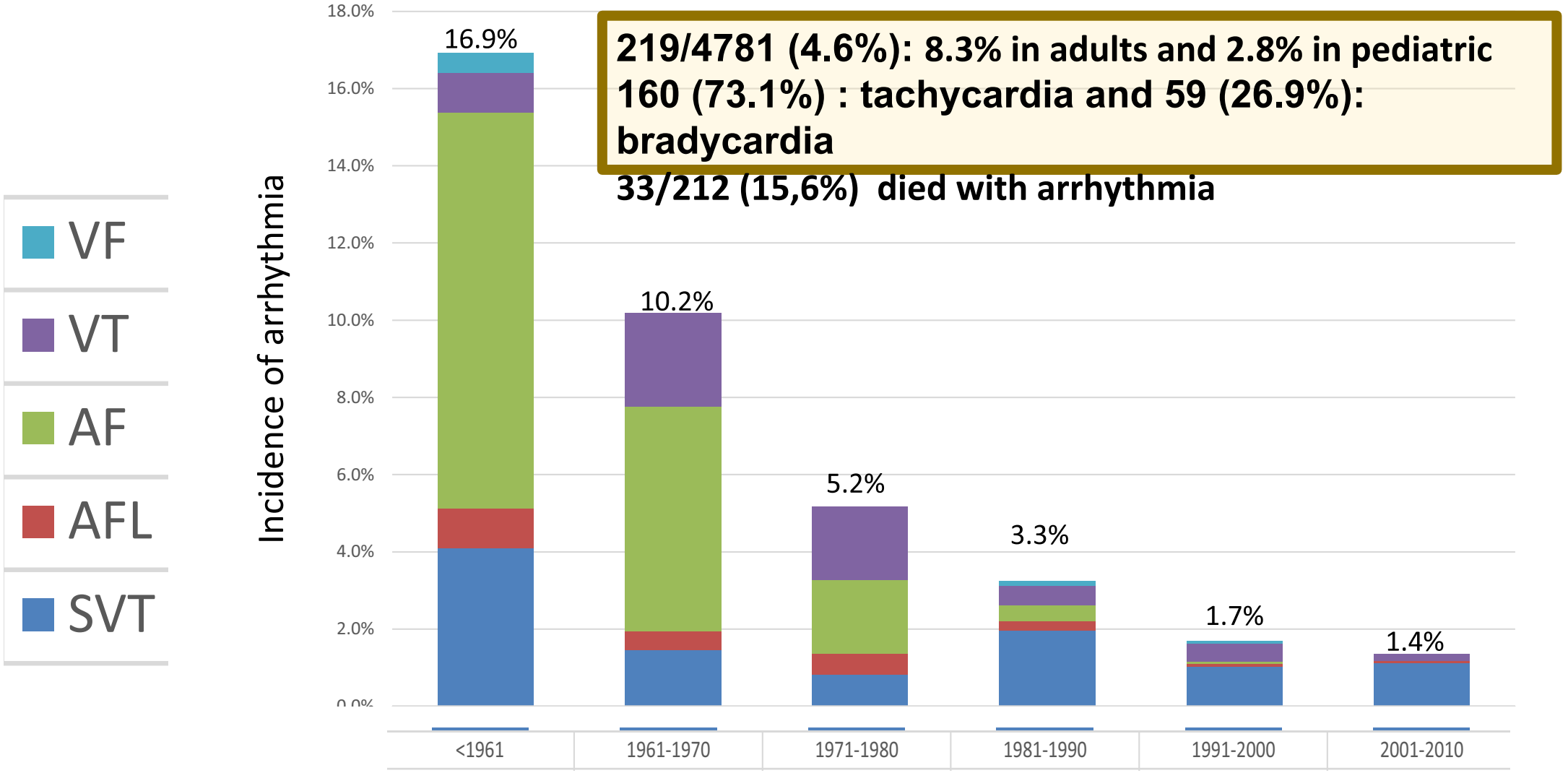
Arrhythmia Characteristics & Fontan Types

	Total	Modified AP	Lateral	TCPC
(years)				
Mean time since Fontan to arrhythmia onset (years)	14.2 (±6.4)	15.3 (±6.2)	12.3 (±1.6)	7.3 (±5.3)

Conclusion

There is a long-term risk of late arrhythmia among patients with a Fontan circulation. This study has shown that the incidence of arrhythmias increases with time since surgery. In this study arrhythmia appears almost inevitable in patients with an AP Fontan by 26 years post-surgery. It is likely that the number of patients presenting with arrhythmia will rise sharply over the next decade. This has huge implications for clinical practice and requires the development of dedicated Electrophysiologists to work with their colleagues in ACHD to manage these patients.

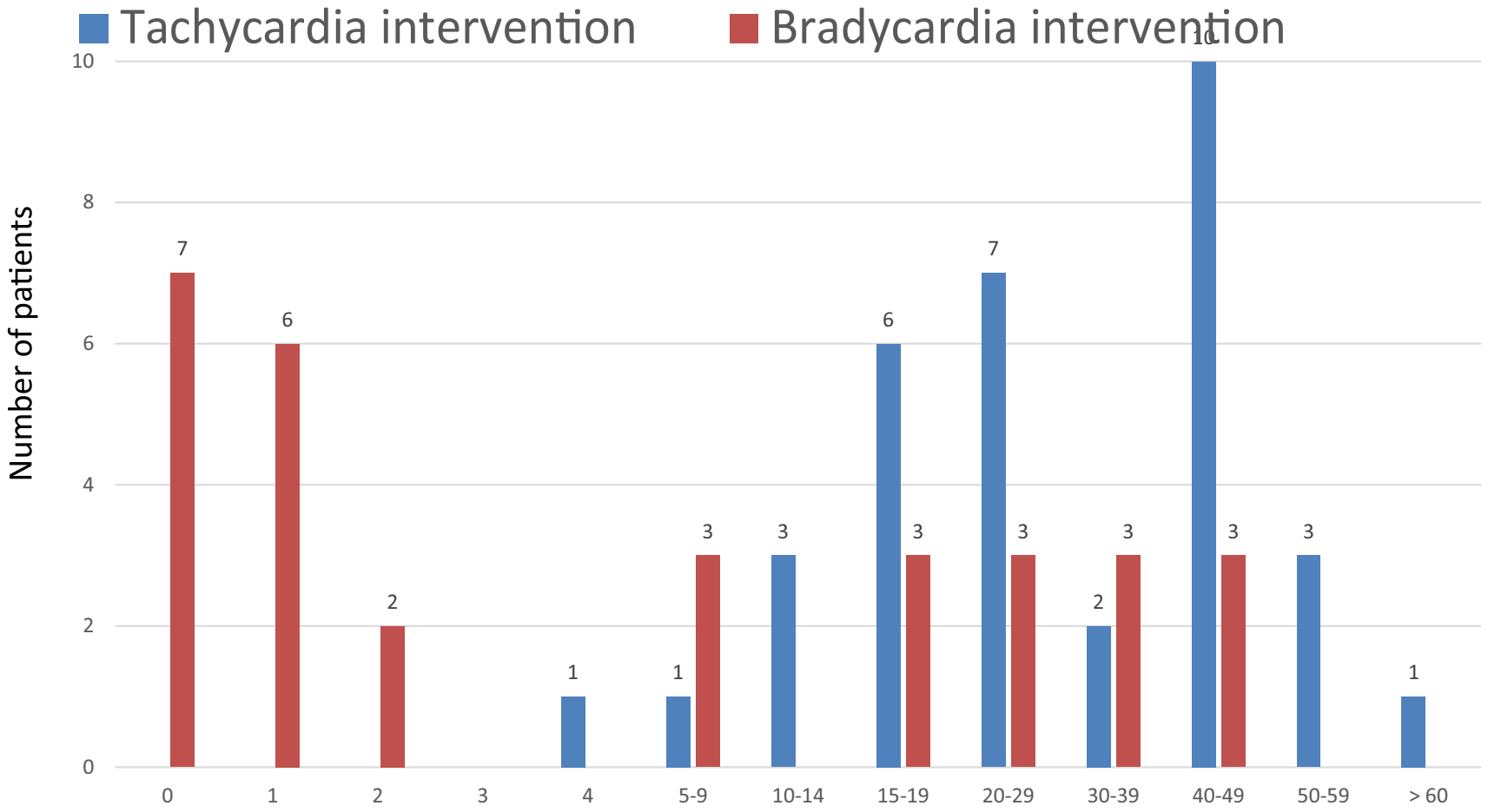
What about Tetralogy



219/4781 (4.6%): 8.3% in adults and 2.8% in pediatric
160 (73.1%) : tachycardia and 59 (26.9%): bradycardia
33/212 (15,6%) died with arrhythmia

Arrhythmic burdens in patients with tetralogy of Fallot: A national database study. Heart Rhythm, 2015

Distributions of age at transcatheter or surgical intervention for tachycardia or bradycardia among patients with tetralogy of Fallot.



Monitorization of Patients for Atrial Tachycardia

- Regular follow-up:
  - ECG, exercise test, CPET, Holter ECG
  - ECHO, MRI scanning
  - Event monitor in presumed arrhythmia symptoms
- Timely treatment of hemodynamic problems either interventional or surgically

Remote Monitoring Leads to Early Recognition and Treatment of Critical Arrhythmias in Adults After Atrial Switch Operation for Transposition of the Great Arteries

Bert Nagel, MD; Jan Janousek, PhD; Martin Koestenberger, PhD; Robert Maier, MD; Werner Sauseng, MD; Volker Strenger, PhD; Andreas Gamillscheg, PhD; Peter Zartner, MD

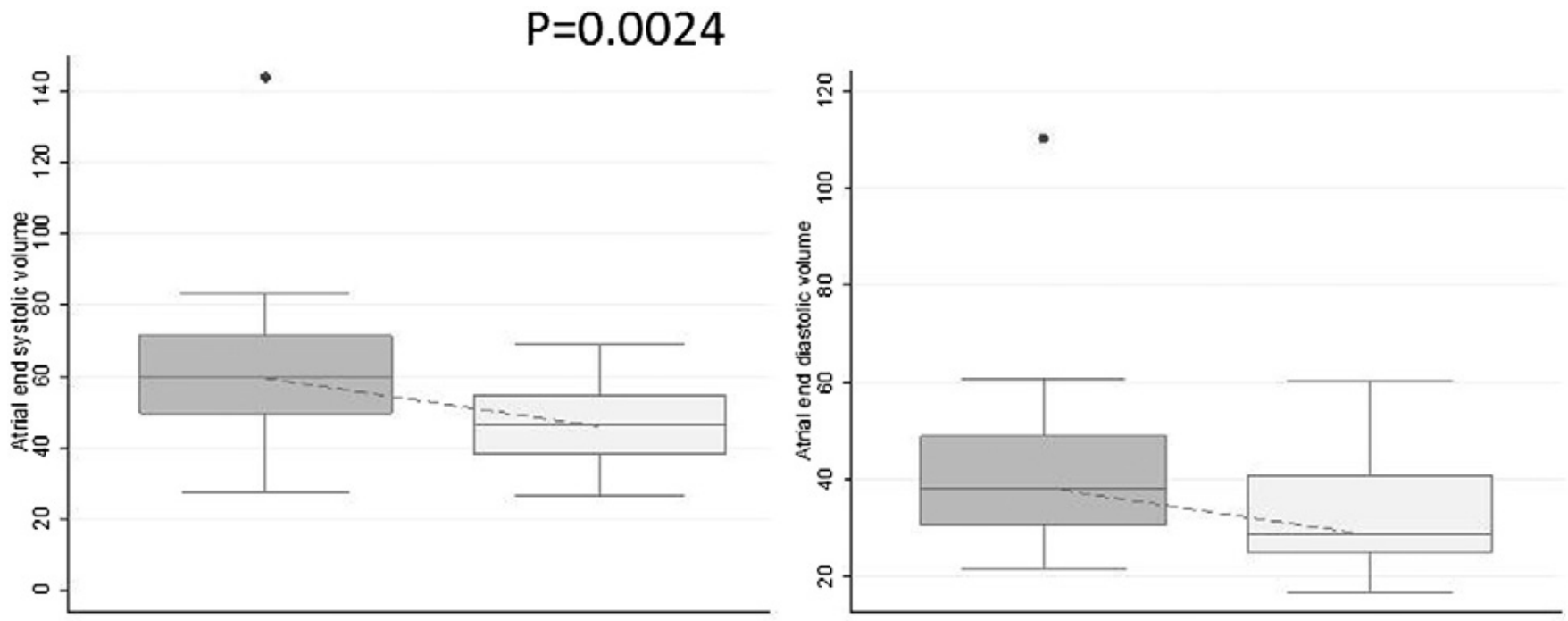
Table 4. RM Group vs. Control Group

	RM group (n=11)	Control group (n=21)
Age at enrollment (years)	29.4 (24.4–34.7)	29.4 (24.4–34.7)
Patients with detected arrhythmias	10 (91)	12 (57)
Patients with symptoms due to arrhythmias leading to advanced follow-up visit	2 (20)	3 (25)
Detected arrhythmias	16	16
SCD	0	2
Detection time (days)	3.4 (0–4.5)	108.5 (15–164)
Reaction time (days)	13.3 (1–31.8)	9.5 (1–22)‡
Time gain (days)	77.5 (10–197)	56.3 (34–72)‡
Cardiac failure due to arrhythmias	0	2
Thromboembolic event due to arrhythmia	0	1

A continuous wireless RM system allows reliable and early diagnosis of tachyarrhythmias compared to conventional follow-up in TGA patients following atrial switch repair requiring implantable cardiac devices. The majority of patients were asymptomatic at the time of adverse events.

“Pulmonary valve replacement diminishes the presence of restrictive physiology and reduces atrial volumes”: A prospective study in Tetralogy of Fallot patients

A. Pijuan-Domenech et al. / International Journal of Cardiology 177 (2014) 261–265



Change in atrial volumes , systolic and diastolic before (dark grey) and one year after PVR (light grey)

Right Atrial Volume is Increased in Corrected Tetralogy of Fallot and Correlates with the Incidence of Supraventricular Arrhythmia: A CMR Study. Pediatr Cardiol (2015)

- **The larger the RA volume was measured in our study population, the higher the incidence of SV arrhythmia.**
- **This correlation was independent of patients' age, pulmonary valve replacement, RV volume and function, and tricuspid regurgitation or pulmonary valve regurgitation.**
- **CMR-derived RA volumes may contribute valuable information to assess the risk of arrhythmia in cTOF patients.**

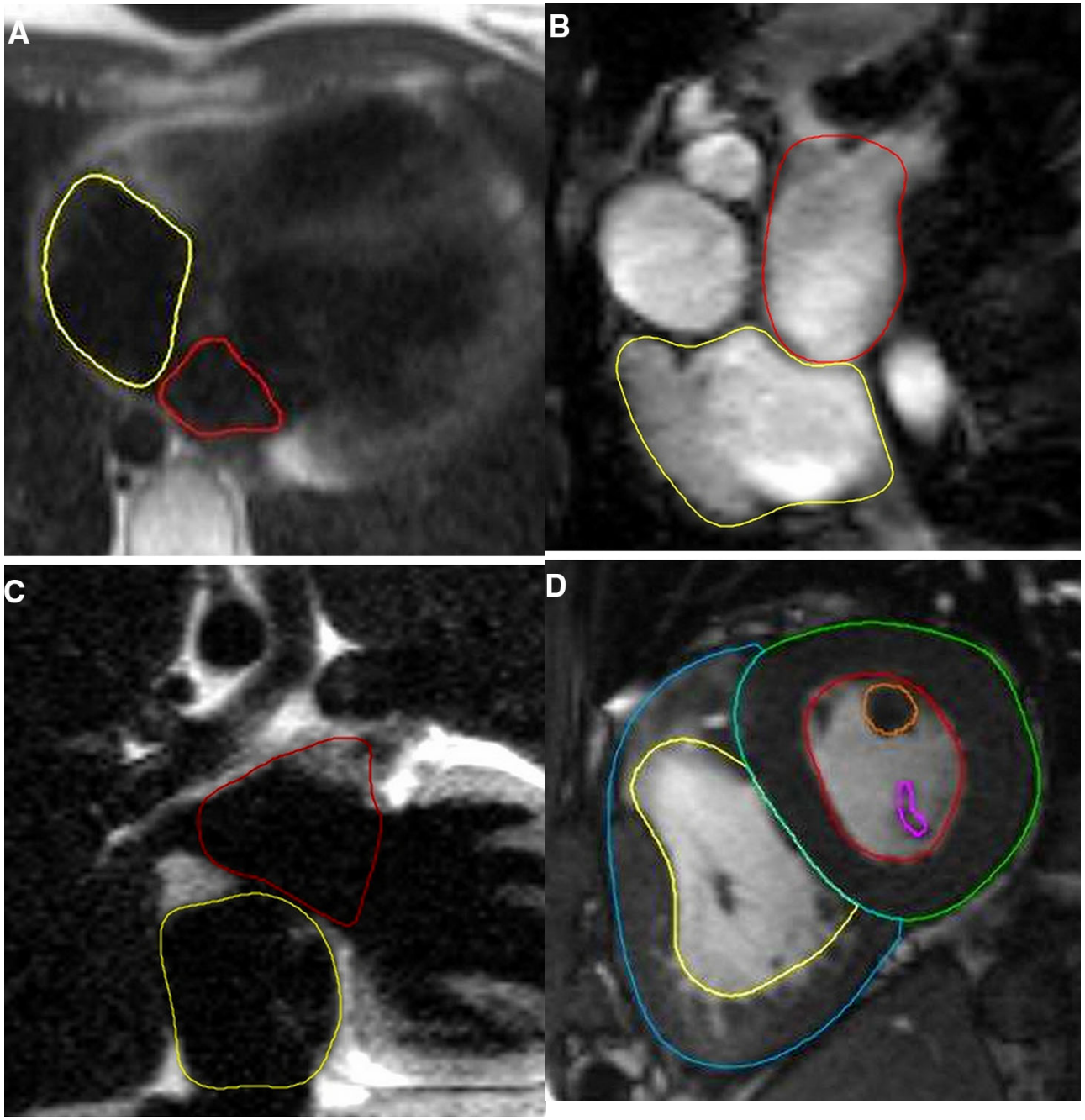


Table 1 Patients' characteristics

Patient groups	Total numbers ($n \pm$ SD)
Patients	67
Female	34
Male	33
Age at study inclusion (years)	30 ± 11.3
BMI (kg/m^2)	25 ± 4.4
Age at corrective surgery	4.2 ± 3.2
Heart beat/min	75 ± 11
Surgical correction	
Ventricular septal defect patch closure	67
Previous a–p shunt	38
RV PA conduit or homograft	40
Transannular patch	27
Myectomy	24
Pulmonary artery plasty	22
RVOT plasty	19
Commissurotomy	12
Atrial septal defect patch closure	4
Mechanical pulmonary valve	2

Conclusions

- Atrial arrhythmias are common in operated CHD patients
- Modification of surgical methods may change the incidence of atrial arrhythmias in single ventricle physiology; but the risk is not zero after this modification
- Monitoring the patients with operated Fallot tetralogy cases should be combined by ECHO and MRI studies
- Treatment of underlying hemodynamic problem in this patient group may postpone or delay the development of atrial arrhythmias
- Atrial fibrillation has been developed eventually in long-standing arrhythmia cases with tetralogy of Fallot

PEDIRHYTHM VII

Thank you

International Society for
Adult Congenital Heart Disease

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The Pediatric & Congenital Electrophysiology Society