Prevalence and Impact of Arrhythmias in Adults with Congenital Heart Disease

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Göttingen, Germany

Prague, November 5, 2022

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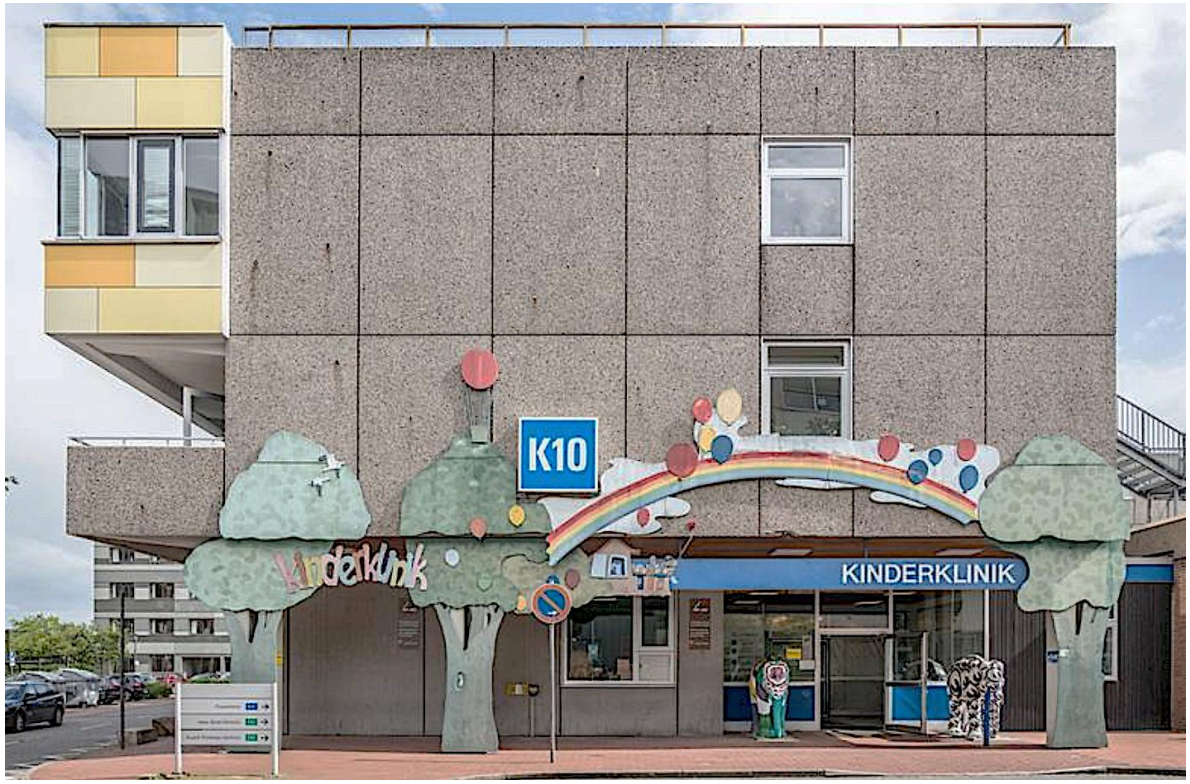
Hannover Medical School



H. C. Kallfelz, MD

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May 1991

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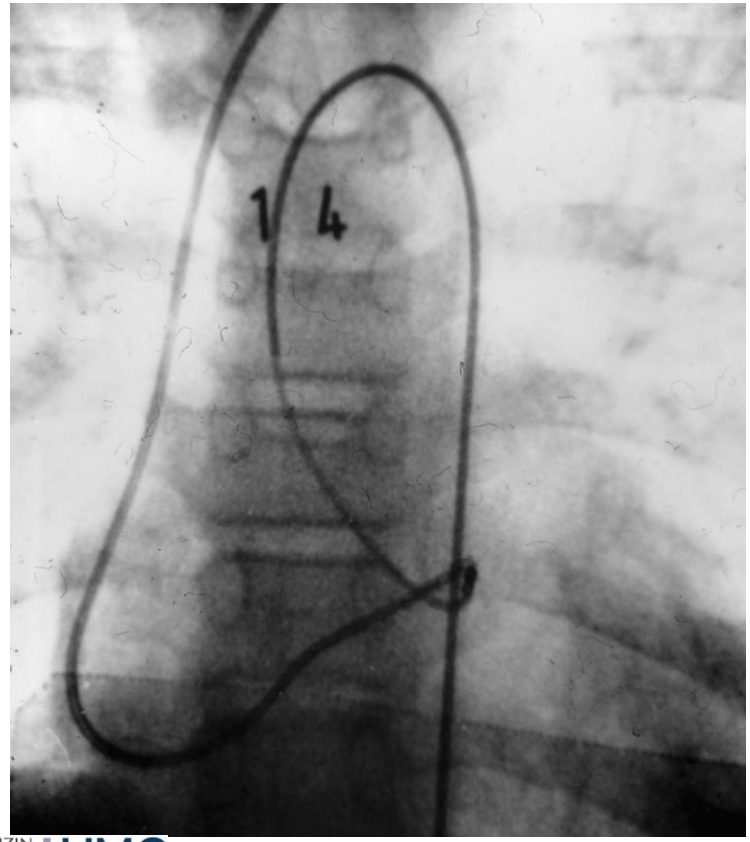

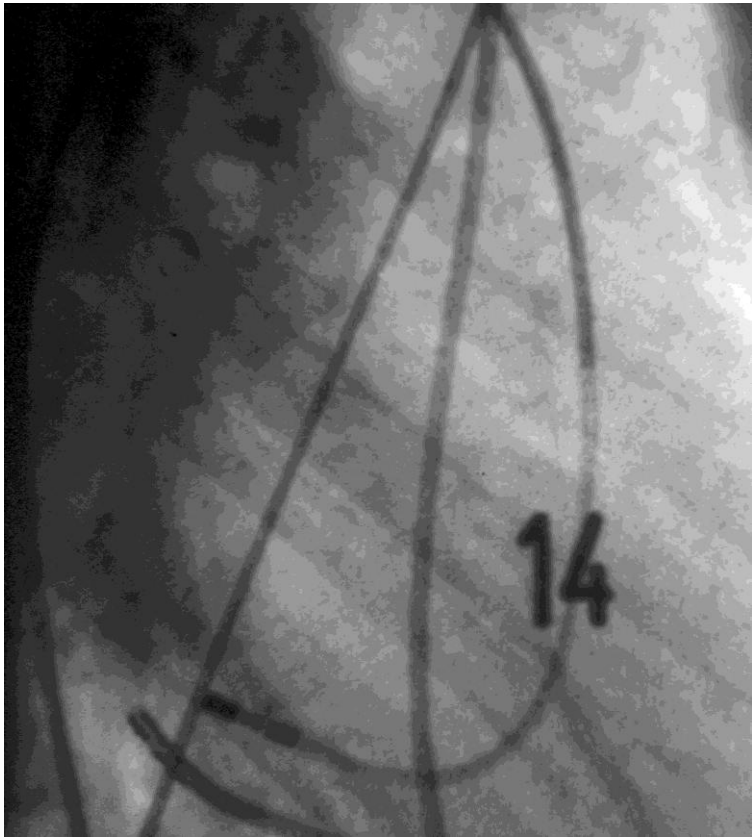

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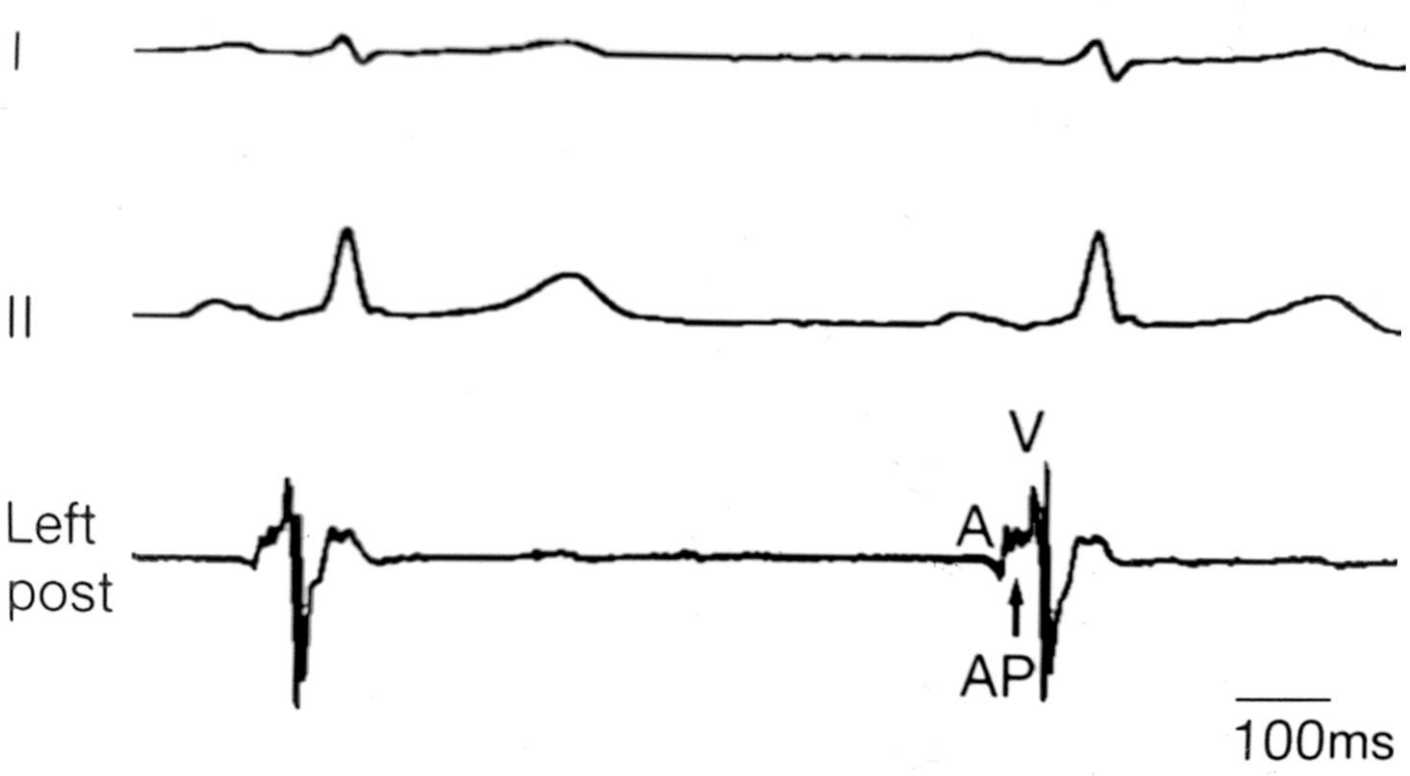

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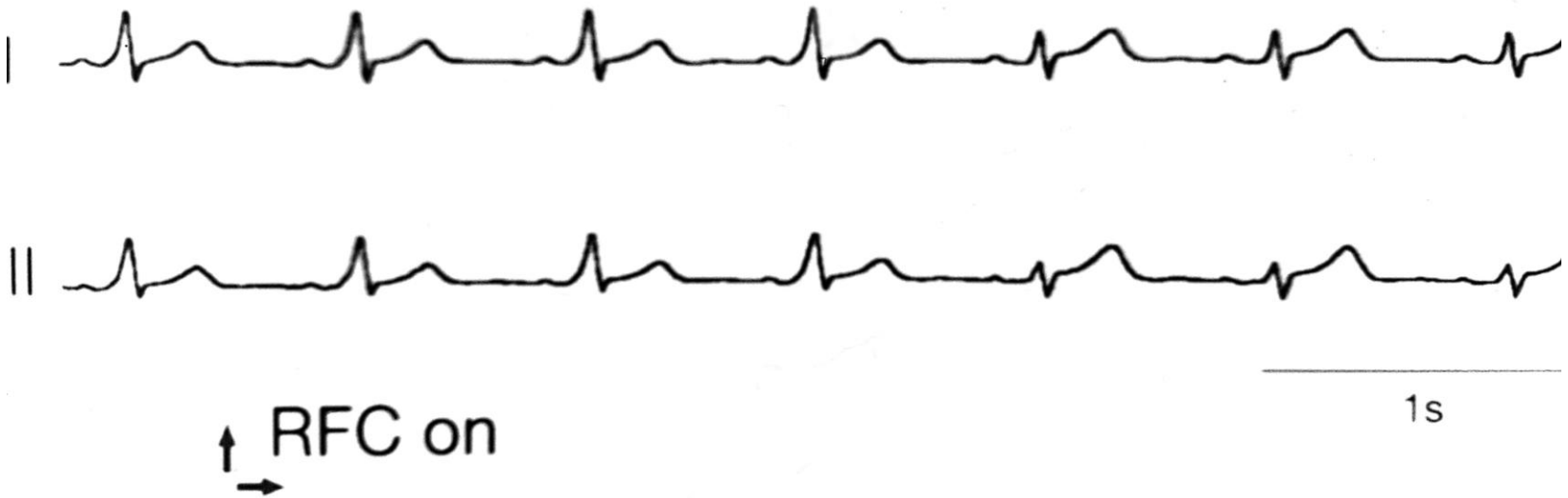
May 1991



Texas Children´s Hospital

5. listopadu 2022 Břevnovský klášter

PRAHA

PRAGUE SYMPOSIUM ON CONGENITAL HEART DISEASE 2022

FN MOTOL

ČESKÁ KARDIOLOGICKÁ SPOLEČNOST

Agátka Sovičková, 9 let

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Estimated Clinical Relevance of CHD within this Decade

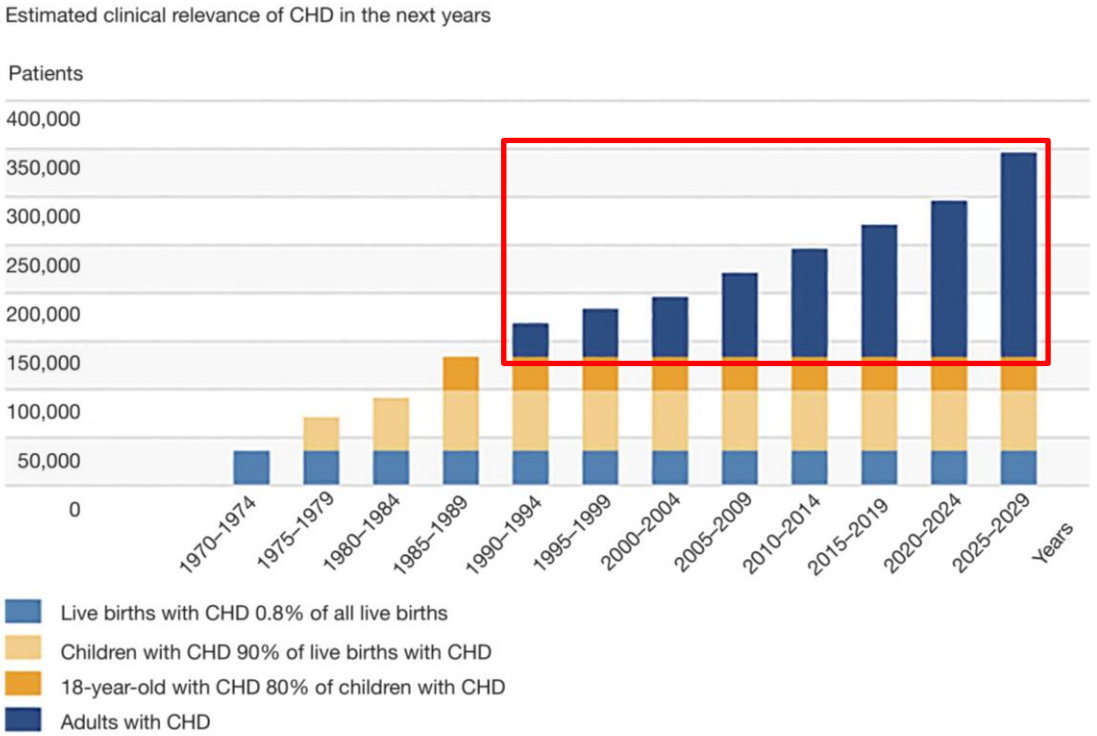
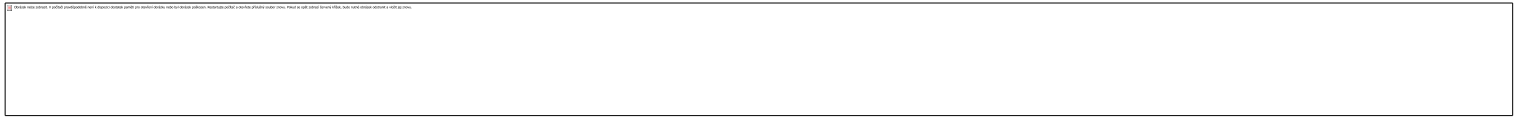
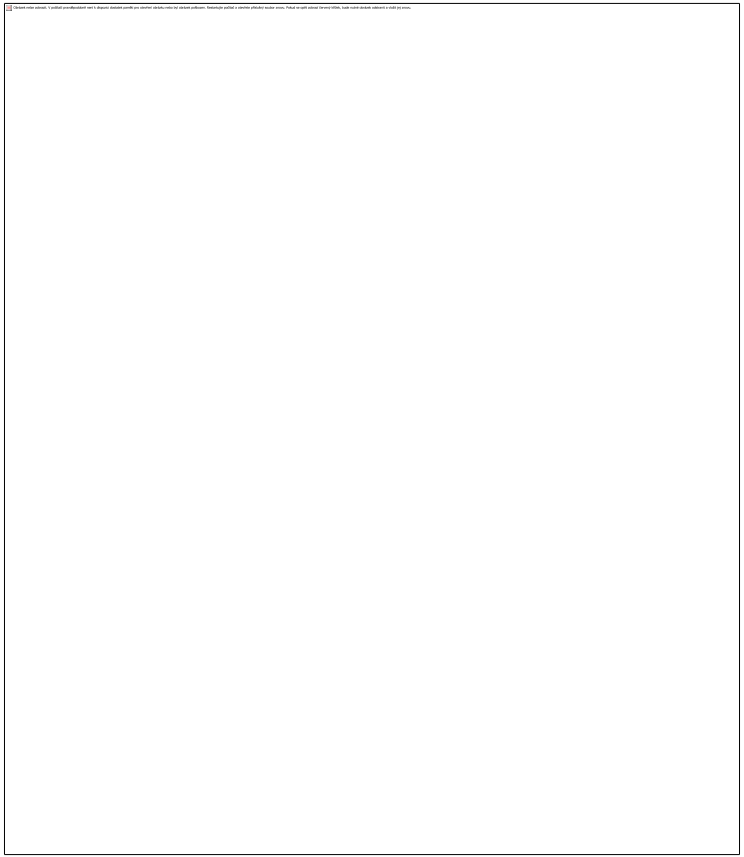


Image source: Competence Network for Congenital Heart Defects

German National Register for Congenital Heart Defects, www.kompetenznetz-ahf.de





5 ACHD centers, 201 emergency room admissions within 1 year
Kaemmerer et al., Am J Cardiol 2008

ESC European Society of Cardiology
Europace (2018) **20**, 1719–1720
doi:10.1093/europace/eux380

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

Antonio Hernández-Madrid[1]*†, Thomas Paul[2†], Dominic Abrams[3], Peter F. Aziz[4], Nico A. Blom[5,6], Jian Chen[7], Massimo Chessa[8], Nicolas Combes[9], Nikolaos Dagres[10], Gerhard Diller[11], Sabine Ernst[12], Alessandro Giamberti[13], Joachim Hebe[14], Jan Janousek[15], Thomas Kriebel[16], Jose Moltedo[17], Javier Moreno[1], Rafael Peinado[18], Laurent Pison[19], Eric Rosenthal[20], Jonathan R. Skinner[21], and Katja Zeppenfeld[22]

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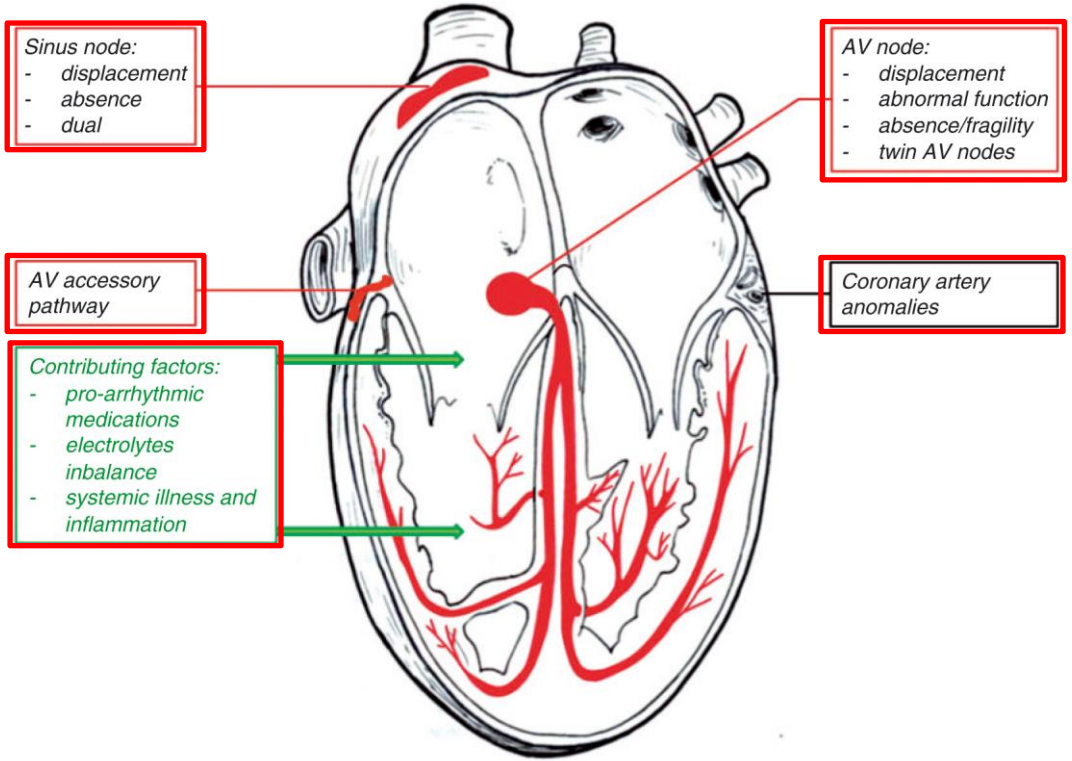
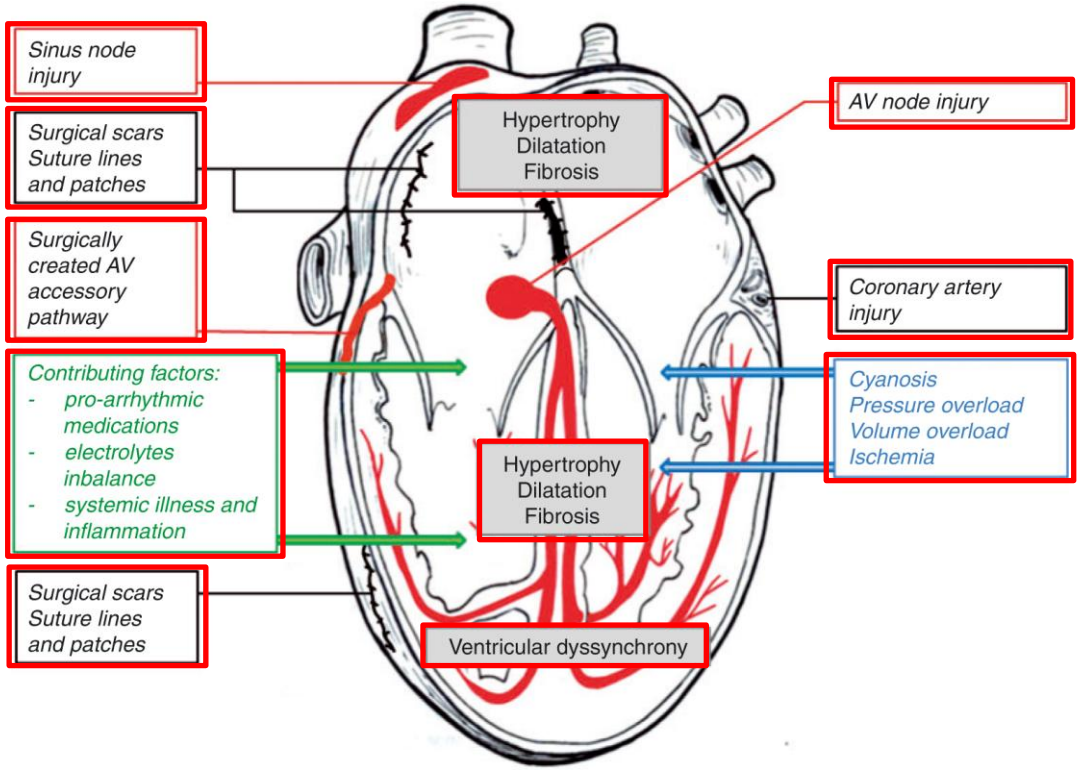
Pre-operative Factors Resulting in Supraventricular and Ventricular Arrhythmias in CHD Patients



Hernandez-Madrid A & Paul T et al., Europace 2018

Post-operative Factors Resulting in Supraventricular and Ventricular Arrhythmias in CHD Patients



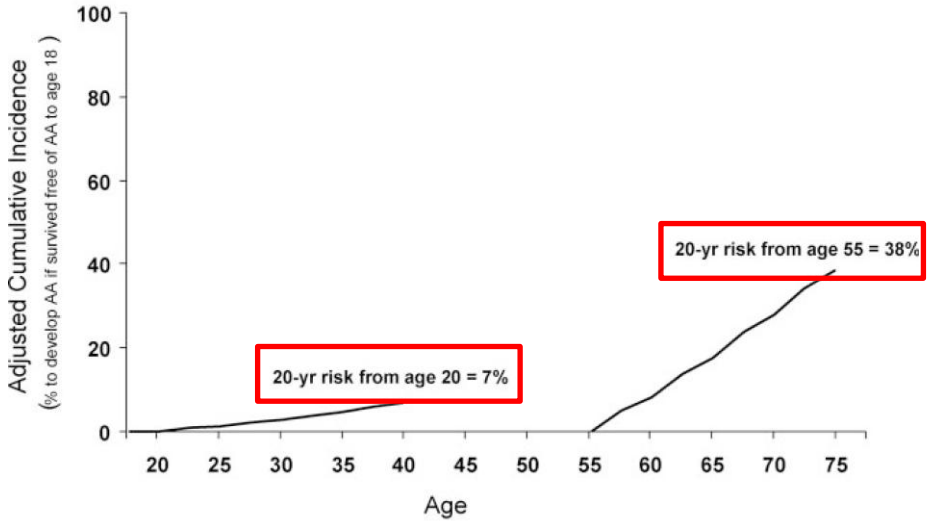
Hernandez-Madrid A & Paul T et al., Europace 2018

Atrial Arrhythmias in Adults With Congenital Heart Disease

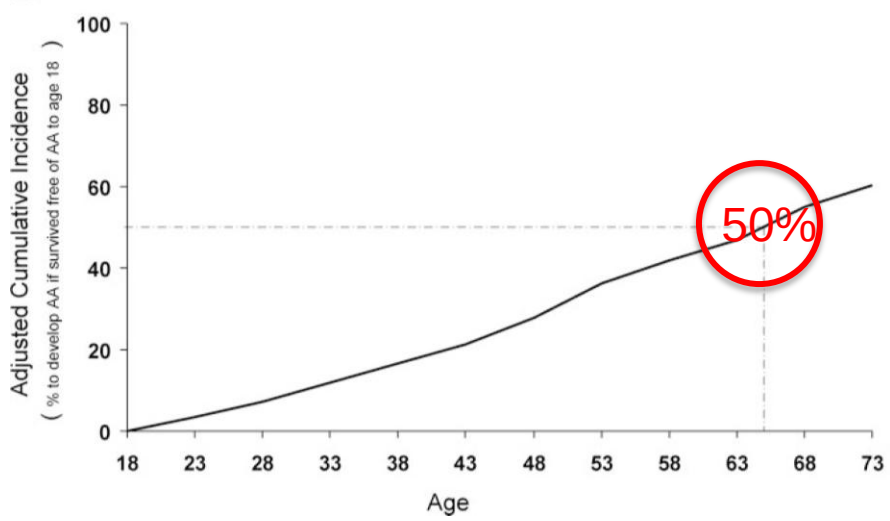
Judith Bouchardy, MD; Judith Therrien, MD; Louise Pilote, MD, MPH, PhD;
Raluca Ionescu-Ittu, MSc; Giuseppe Martucci, MD; Natalie Bottega, MD; Ariane J. Marelli, MD

Circulation 2009

Lifetime cumulative incidence of AA



Lifetime risk of AA in severe CHD

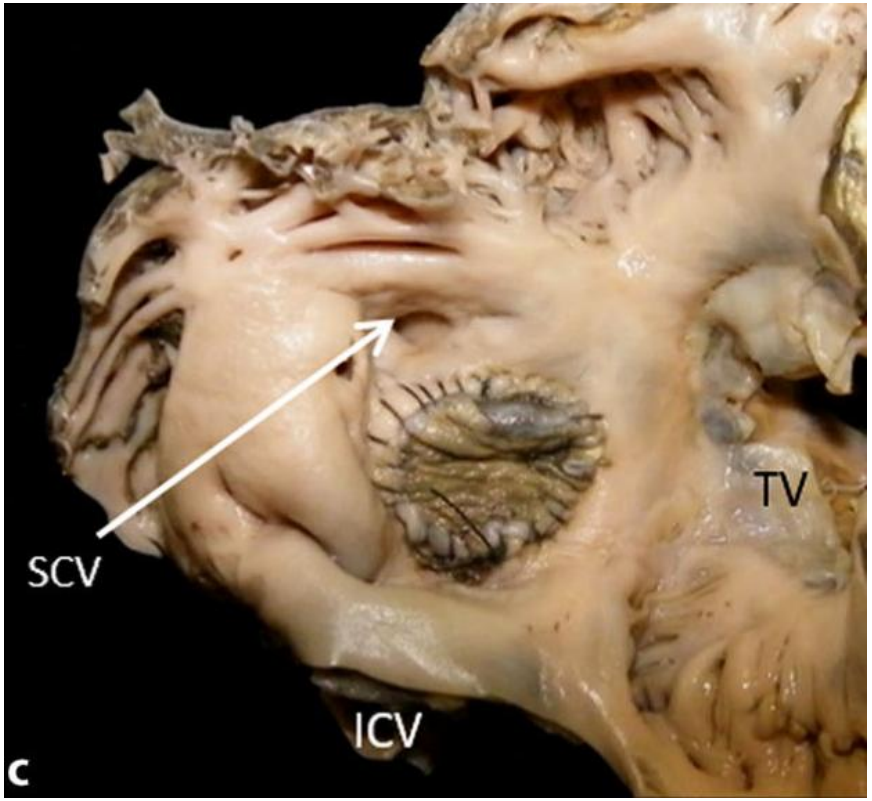


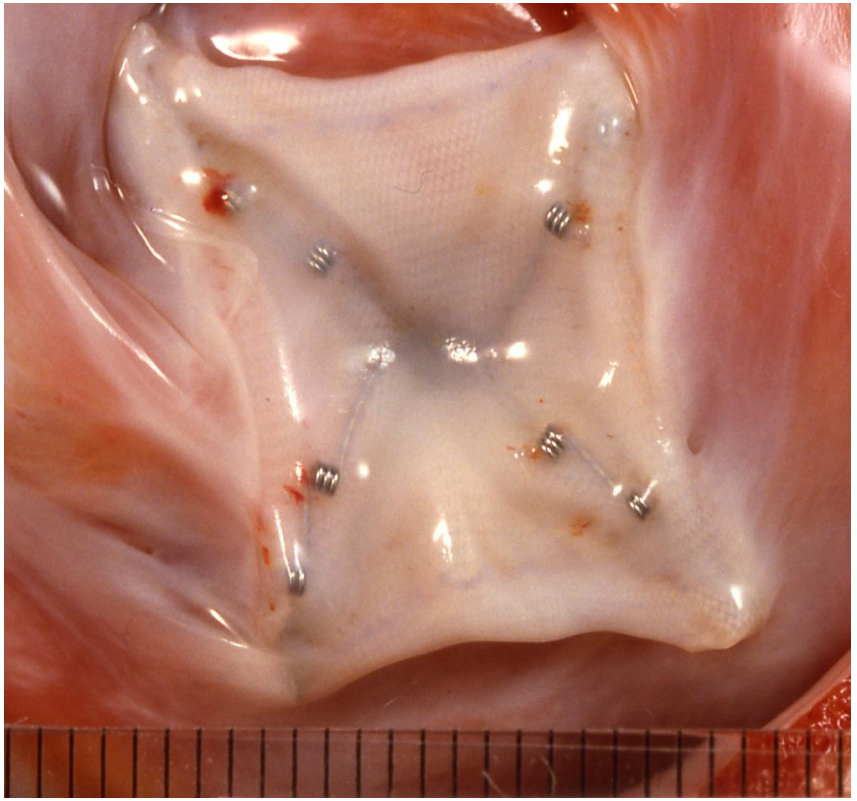
1983-2005, Quebec, n=38,428 ACHD patients, overall incidence of atrial arrhythmias 15.1%

Atrial Septal Defect

Congenital Heart Defect of *Simple Complexity*

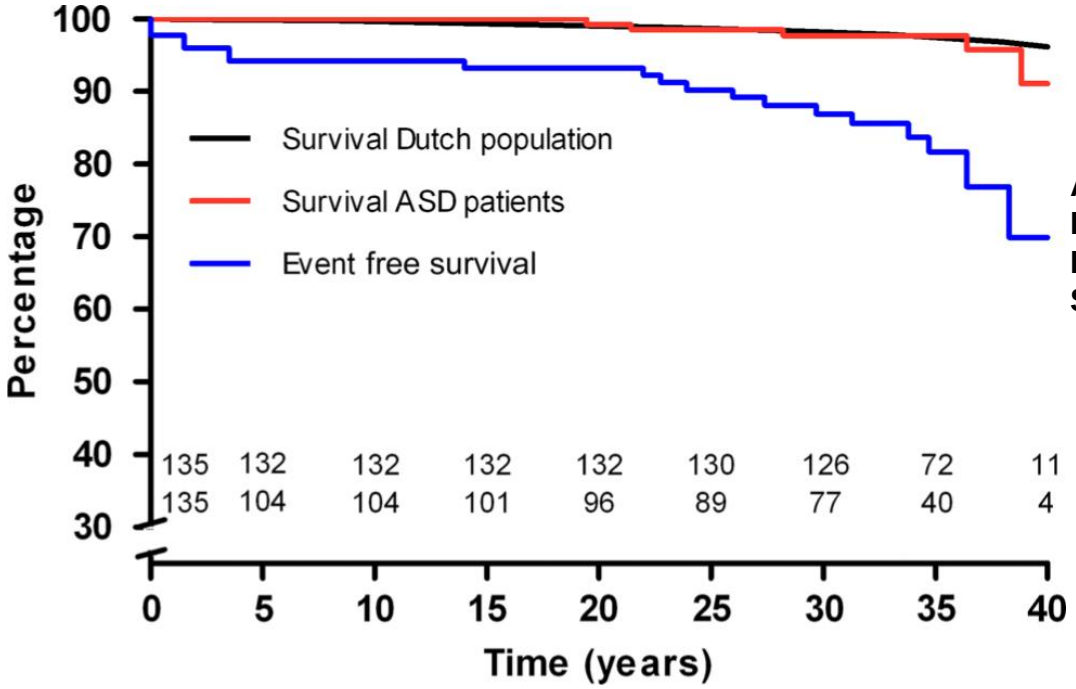
Warnes CA et al., ACC/AHA 2008 Guideline, Circulation 2008, Stout KK et al., Circulation 2018





Ernst S et al., Herzschr Elektrophys 2016

The unnatural history of an atrial septal defect: Longitudinal 35 year follow up after surgical closure at young age



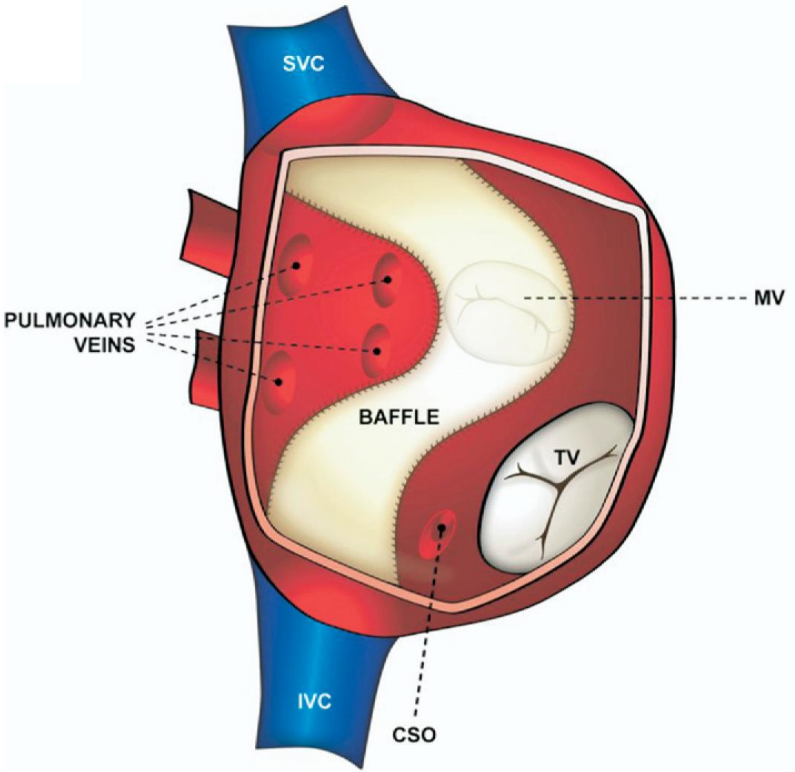
Atrial arrhythmias 16%
Pacemaker 6%
Reoperation 2%
Stroke 1%

N=135, surgical ASD closure <15 years between 1968-1980 in Rotterdam, mean f/u 35 years
Cuypers J et al., Heart 2013

d-Transposition of the Great Arteries After Atrial Switch Procedure

Congenital Heart Defect of *Severe Complexity*

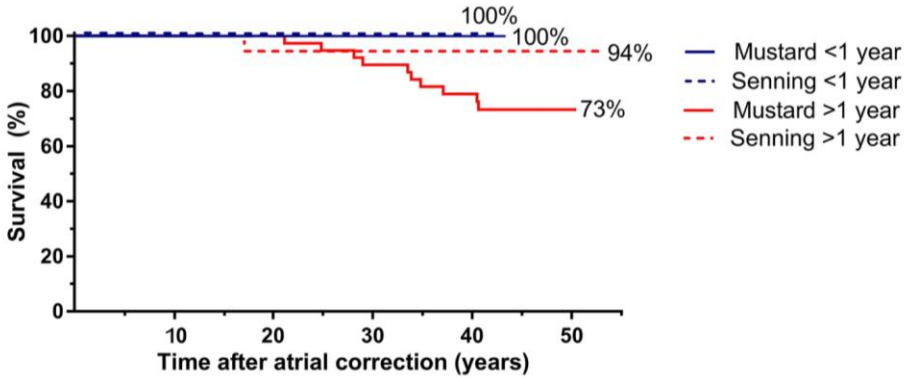
Warnes CA et al., ACC/AHA 2008 Guideline, Circulation 2008, Stout KK et al., Circulation 2018



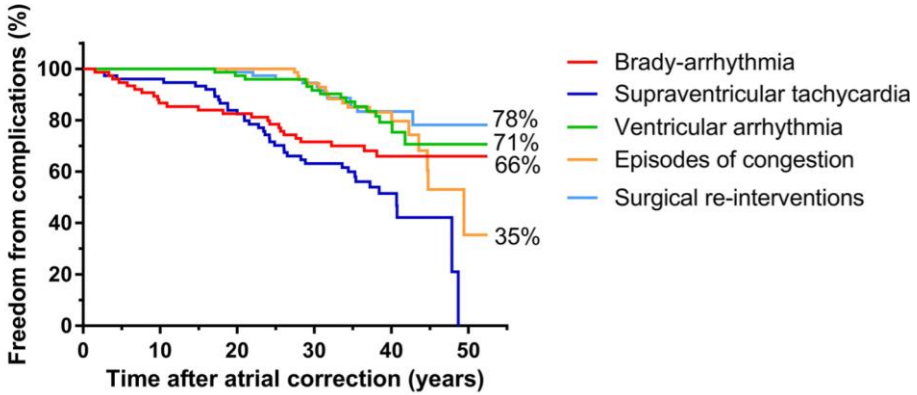
Khairy P & van Hare G, Heart Rhythm 2009

Long-term outcome after atrial correction for transposition of the great arteries

Survival curves after atrial switch



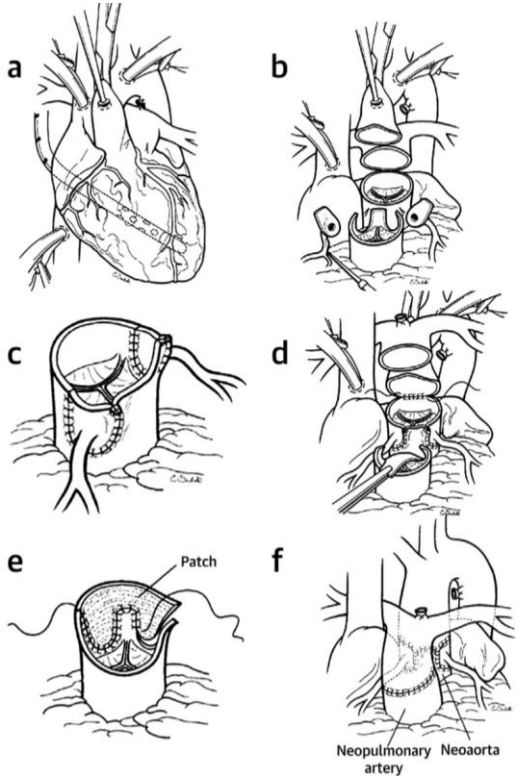
Survival curves for freedom of adverse events after atrial switch

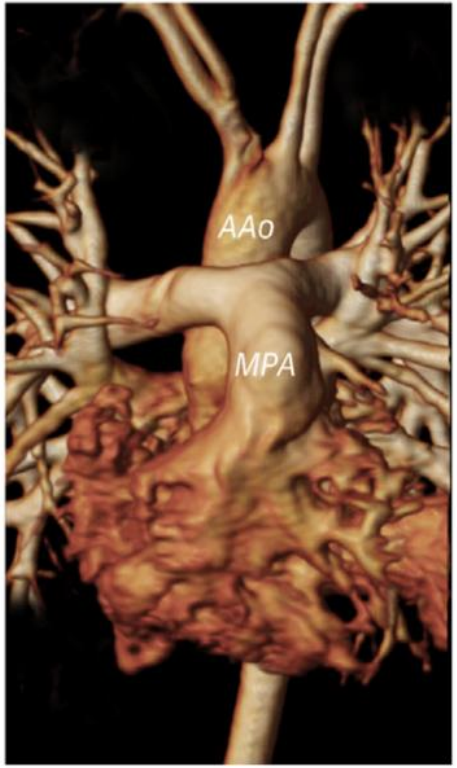


Overall survival 82% at 39.7 years after surgery

N=76 from Leiden, Couperus LE, Heart 2018

d-Transposition of the Great Arteries
- Arterial Switch Procedure -



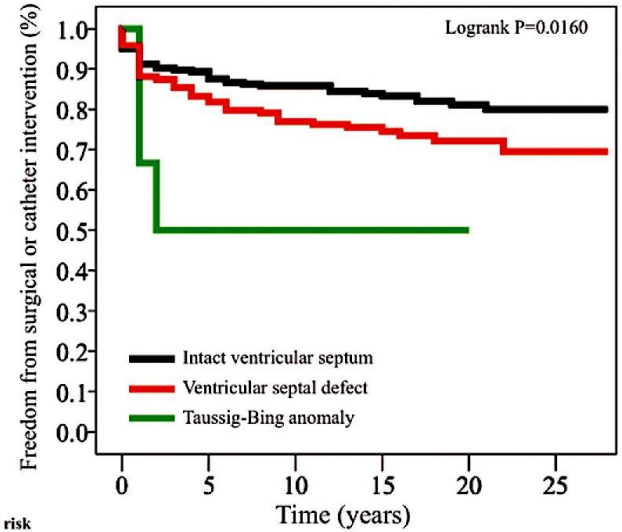


Villafane J et al., J Am Coll Cardiol 2014

Cardiovascular Outcomes After the Arterial Switch Operation for D-Transposition of the Great Arteries

Paul Khairy, MD, PhD; Mathieu Clair, MD; Susan M. Fernandes, MHP, PA-C;
Elizabeth D. Blume, MD; Andrew J. Powell, MD; Jane W. Newburger, MD, MPH;
Michael J. Landzberg, MD; John E. Mayer Jr, MD

Freedom from cardiovascular reintervention

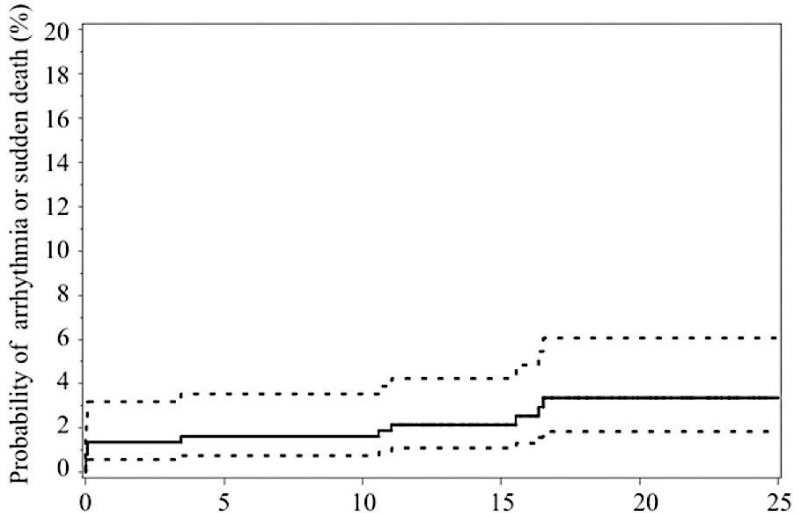


75%

Number at risk

Intact ventricular septum	225	201	193	150	82	20
Ventricular septal defect	143	119	110	78	43	4
Taussig-Bing anomaly	6	3	3	1	1	0

Late mortality due to sudden death and myocardial infarction



Number at risk

374	365	365	267	148	28

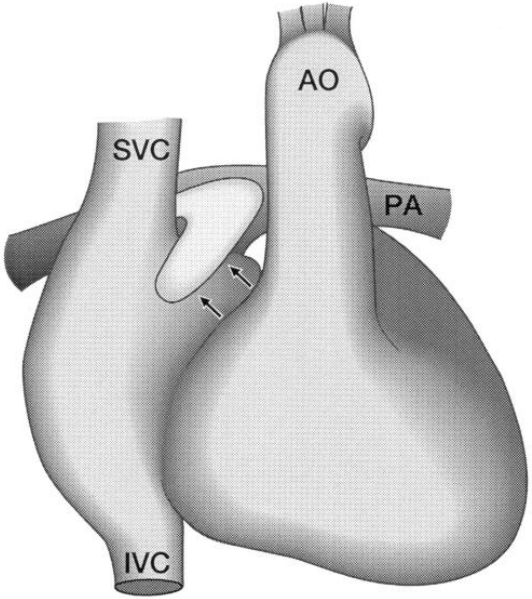
N=400, ASO between 1983 and 1999 in Boston, median f/u 18.7 years
Overall and arrhythmia-free survival at 25 years 96.6%, Circulation 2012

Fontan Operation – Modifications

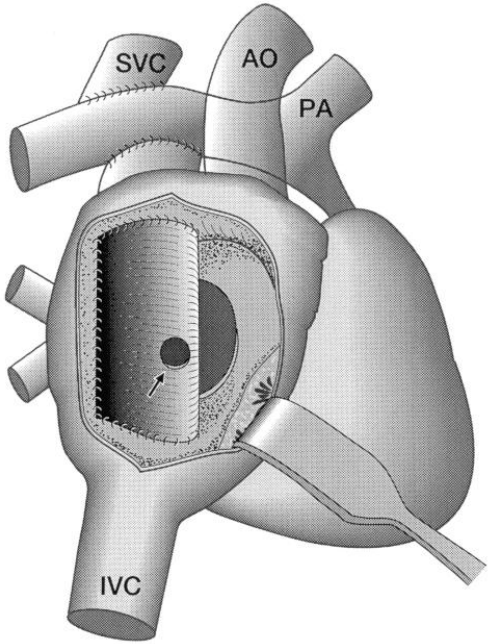
Congenital Heart Defects of *Severe Complexity*

Warnes CA et al., ACC/AHA 2008 Guideline, Circulation 2008, Stout KK et al., Circulation 2018

Atriopulmonary Connection

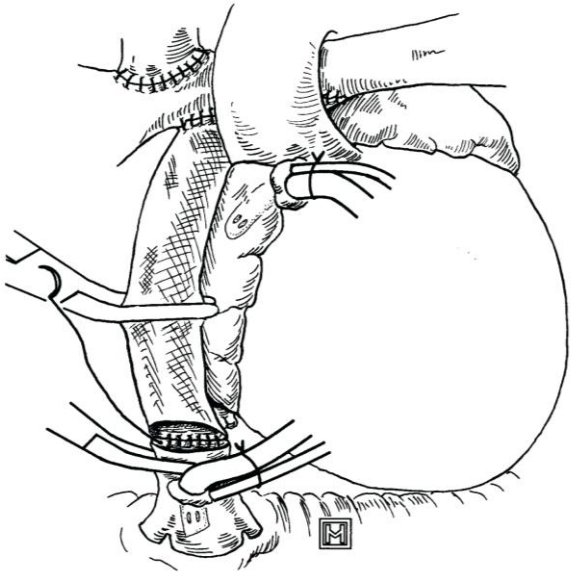


Lateral Tunnel



Courtesy R. Kaulitz

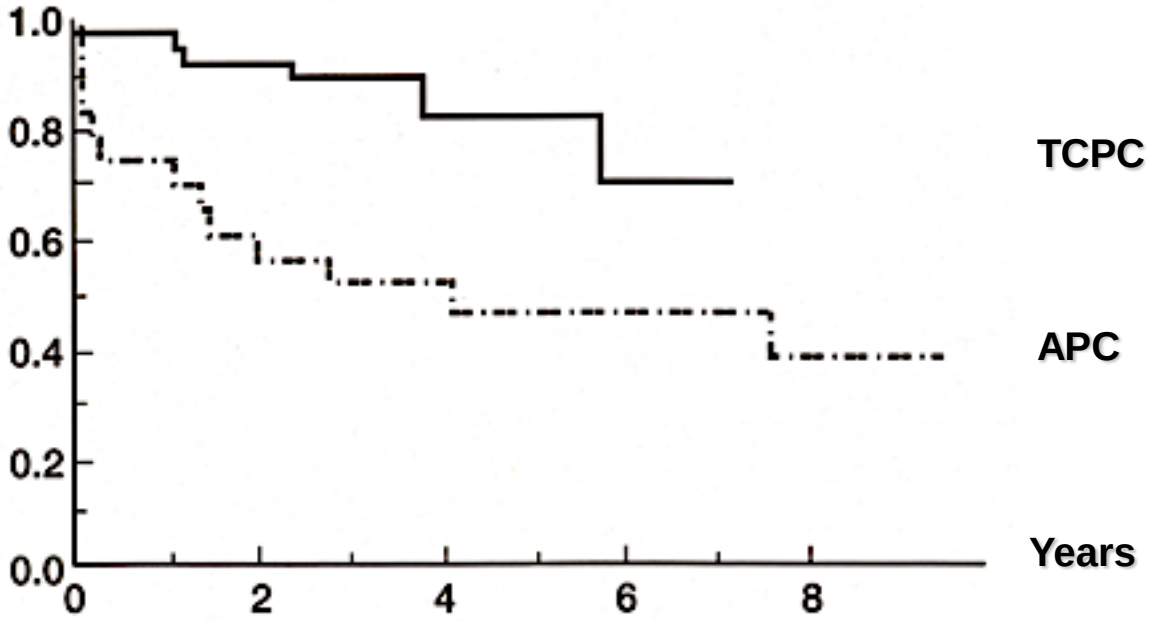
Extracardiac Conduit



McElhinney DB et al, Ann Thorac Surg 1998

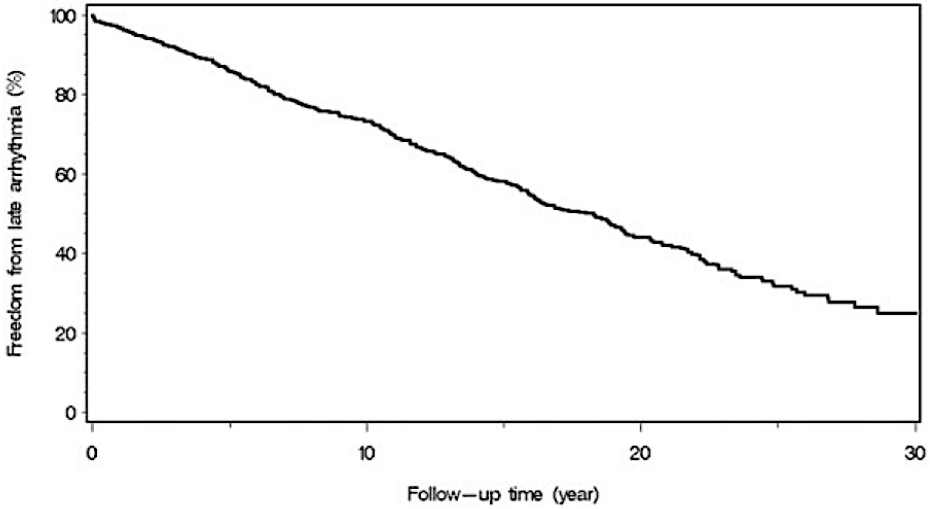
Early and late atrial dysrhythmias after modified Fontan operation: implications of preoperative hemodynamics and type of operation (atriopulmonary vs. total cavopulmonary connection) Paul T et al, Z Kardiol 1993

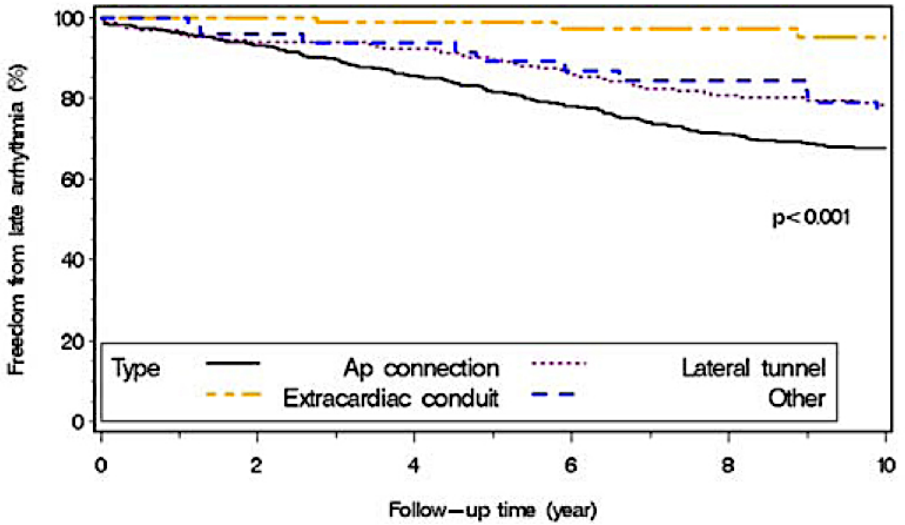
Freedom from Atrial Arrhythmias (n=95)



Sudden cardiac death and late arrhythmias after the Fontan operation

Pundi KN et al., Congenit Heart Dis 2017



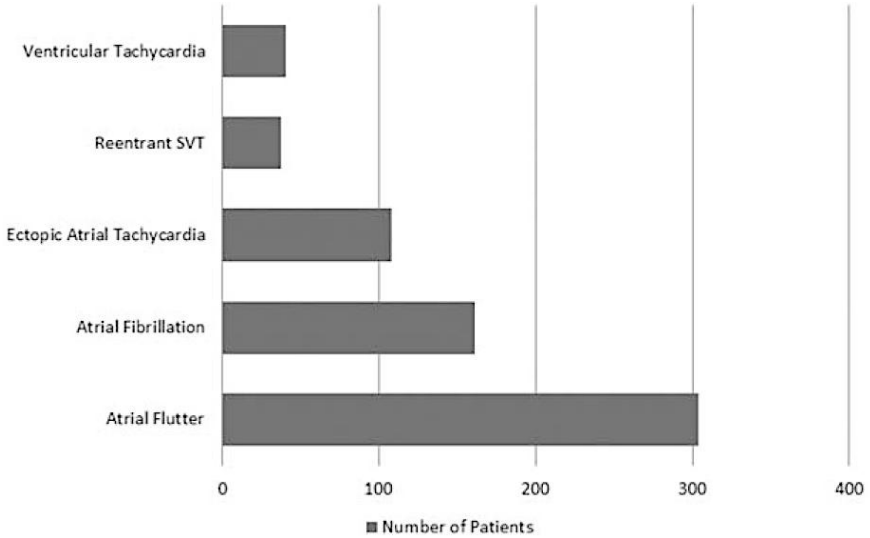


Mayo Clinic, n=864, Fontan operation 1973-2012, mean age 9.4±7.5 years, mean f/u 15.3±9.3 years
10-, 20-, and 30-year freedom from arrhythmias 71%, 42%, and 24%

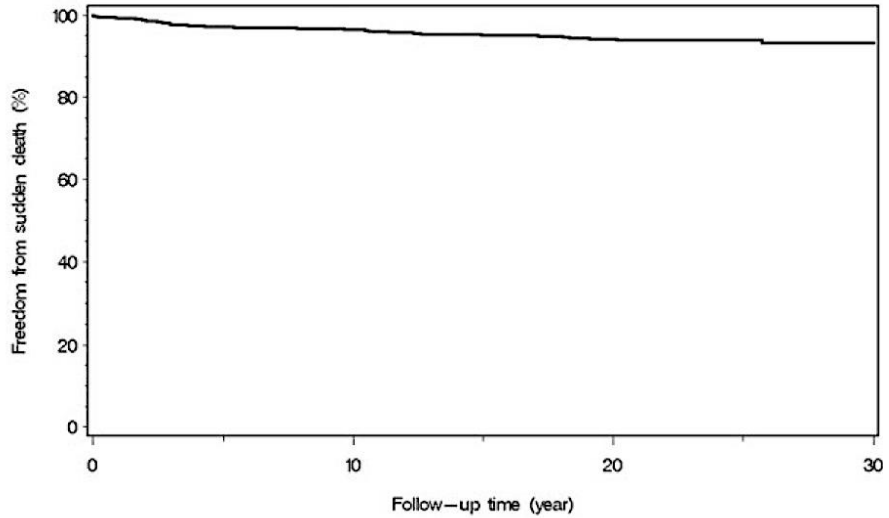
Sudden cardiac death and late arrhythmias after the Fontan operation

Pundi KN et al., Congenit Heart Dis 2017

Incidence of late arrhythmias



Freedom from sudden death



N=864, sudden cardiac death in 5%

Incidence of SCD at 10, 20 and 30 years after Fontan operation 4.6%, 6.2% and 7.1%

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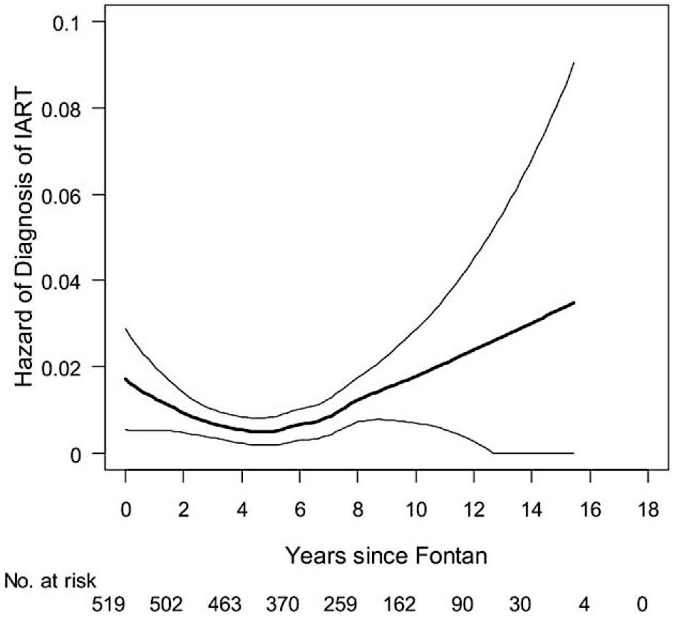
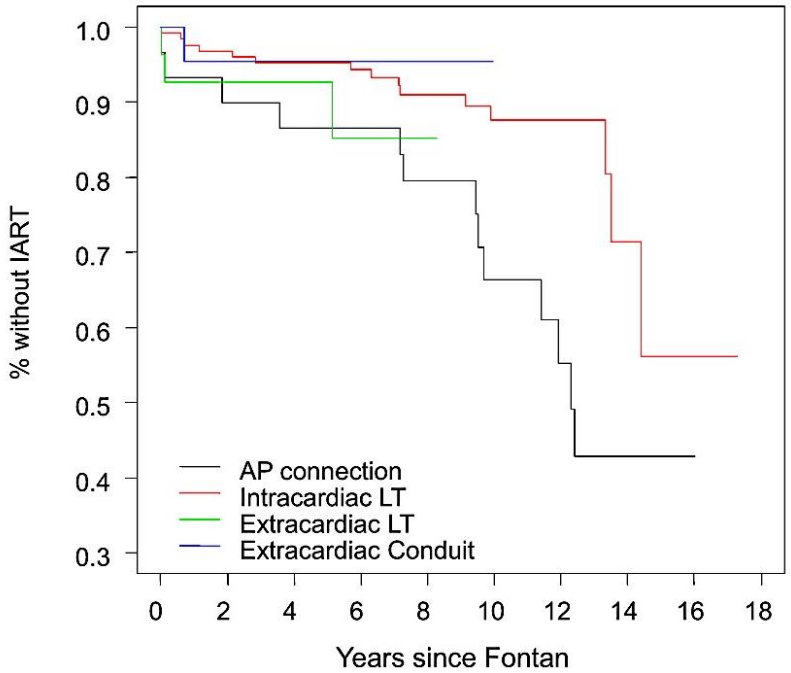
doi:10.1016/j.jacc.2010.03.079

Congenital Heart Disease

Arrhythmias in a Contemporary Fontan Cohort

Prevalence and Clinical Associations in a Multicenter Cross-Sectional Study



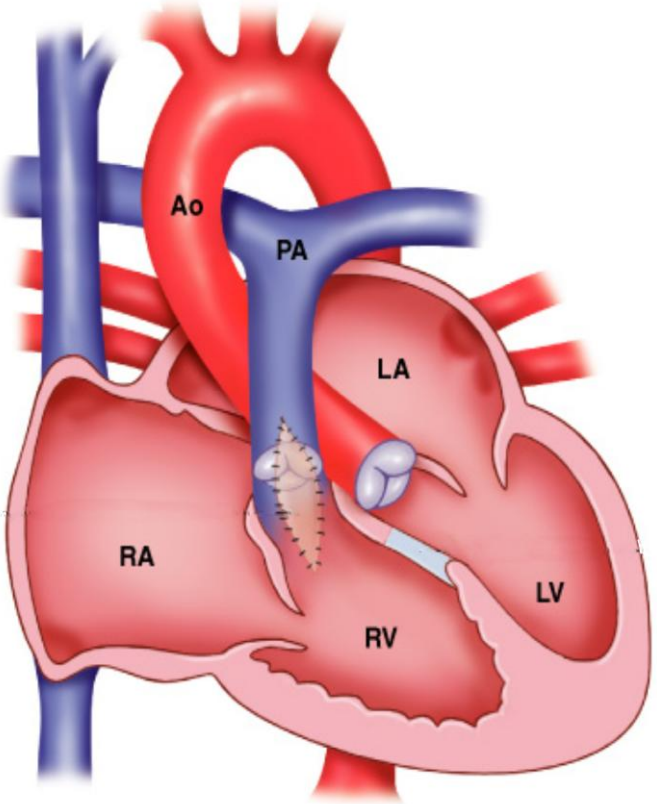


N=519 from 7 centers, mean age at Fontan operation 3.4 years
Mean follow-up 8.6 years, IART in 7%

Tetralogy of Fallot

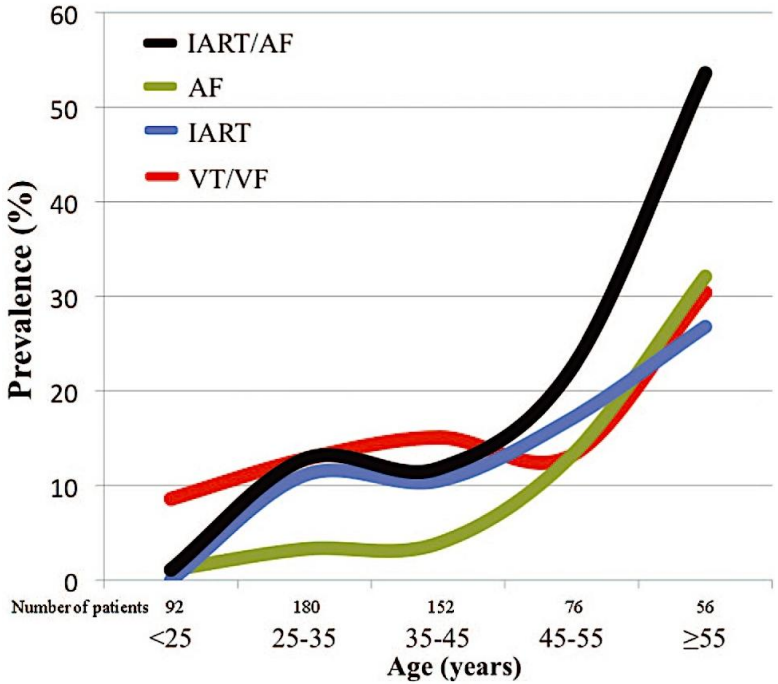
Congenital Heart Defect of *Moderate Complexity*

Warnes CA et al., ACC/AHA 2008 Guideline, Circulation 2008, Stout KK et al., Circulation 2018



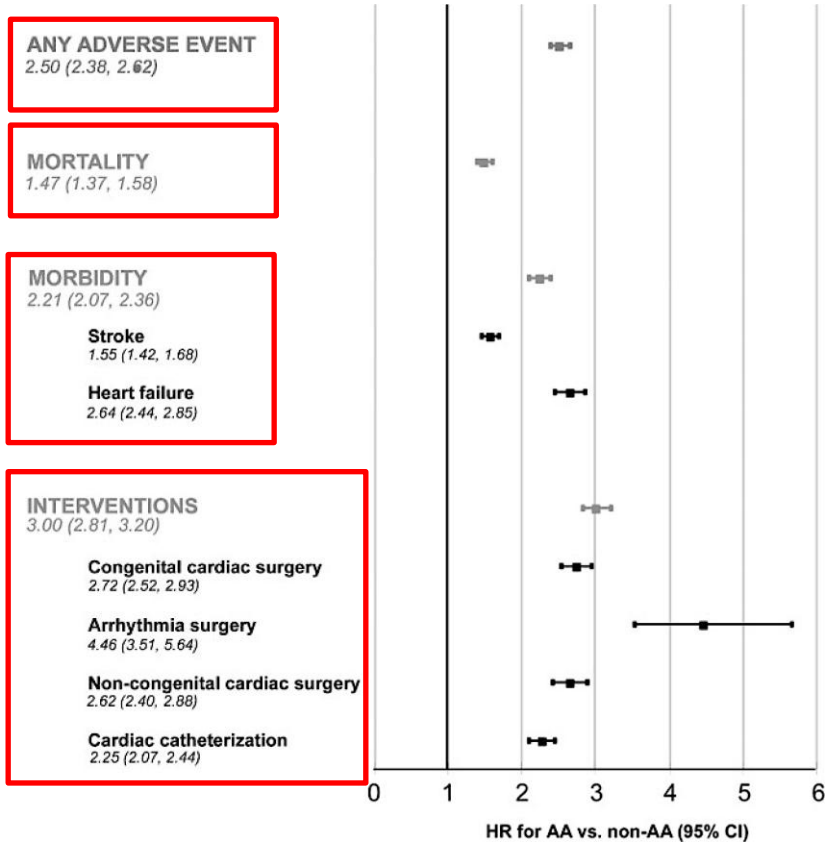
Arrhythmia Burden in Adults With Surgically Repaired Tetralogy of Fallot: A Multi-Institutional Study

Khairy P et al., Circulation 2010



N=556, 11 centers, mean age 36.8±12 years, sustained arrhythmia or arrhythmia intervention in 43.3%, sustained VT/VF in 14.6%

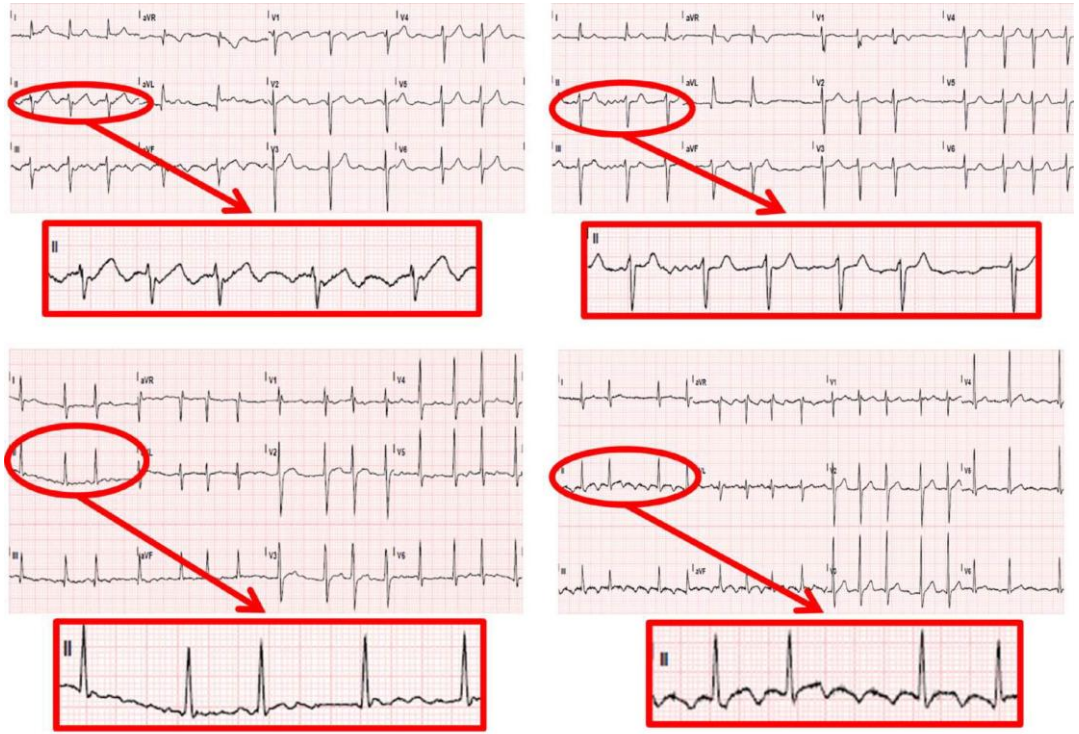
Hazard Ratio for Outcomes in ACHD with vs. without AA



Bourchardy J et al., Circulation 2009

Time Course of Atrial Fibrillation in Patients With Congenital Heart Defects

Teuwen CP et al., Circulation 2015



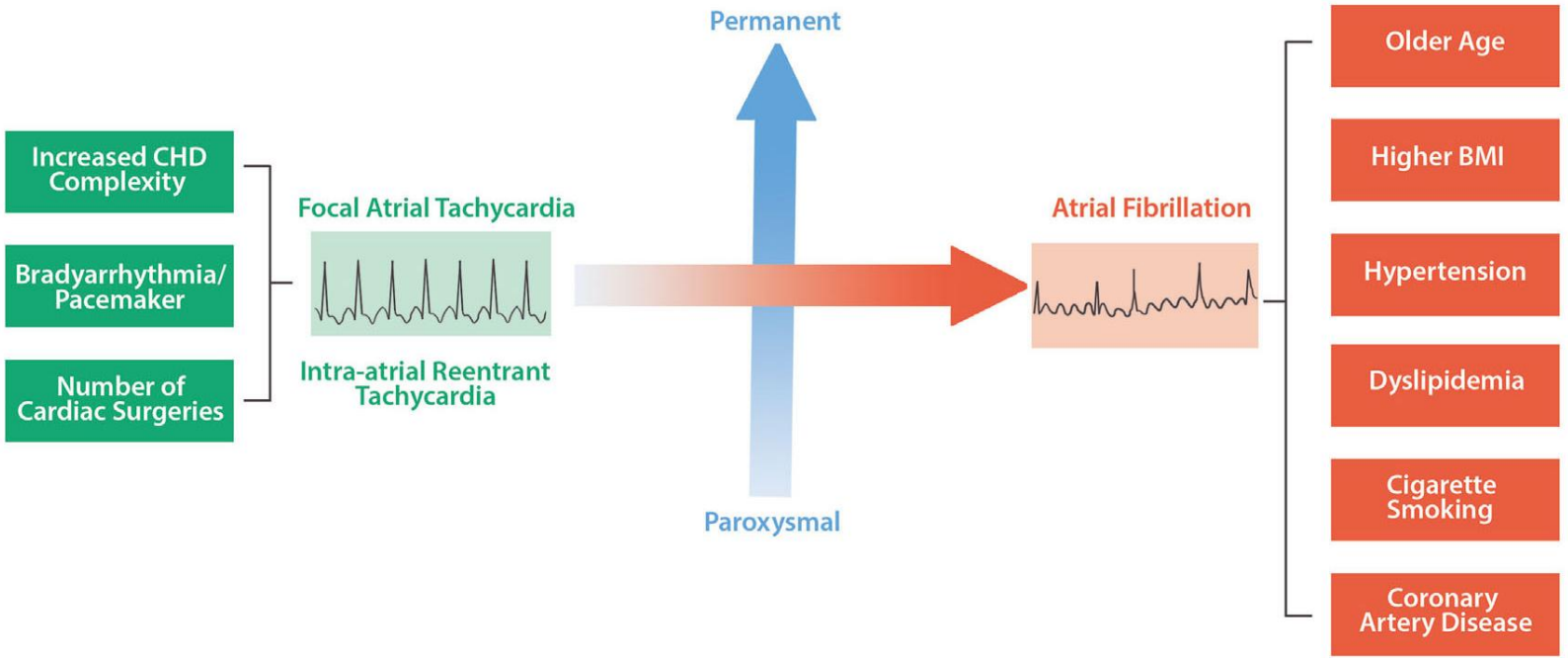
ASD Patient

PS Patient

Multicenter study, n=199 ACHD patients, mean age @AF 49±17 years, mean follow-up 5 years

Increasing Prevalence of Atrial Fibrillation and Permanent Atrial Arrhythmias in Congenital Heart Disease

Labombarda F et al., J Am Coll Cardiol 2017



12 US centers, n=482 with ACHD and atrial arrhythmias, mean age 32±18 years

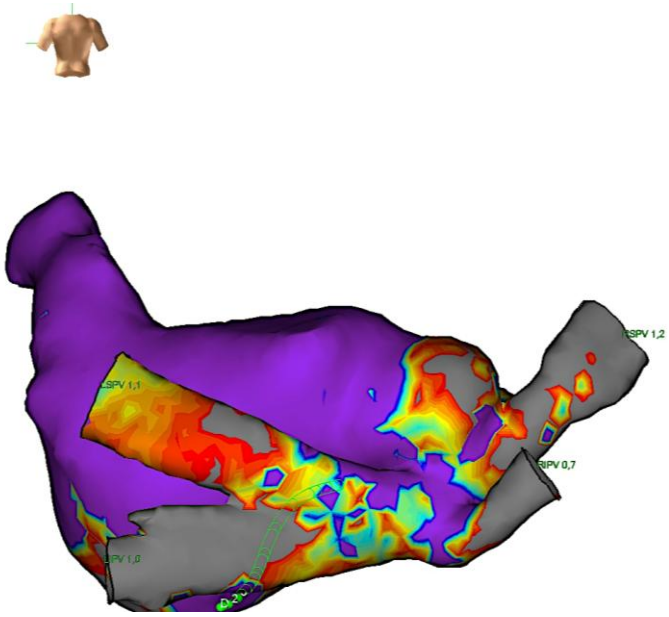
Krause U et al., J Interv Card Electrophysiol 2022

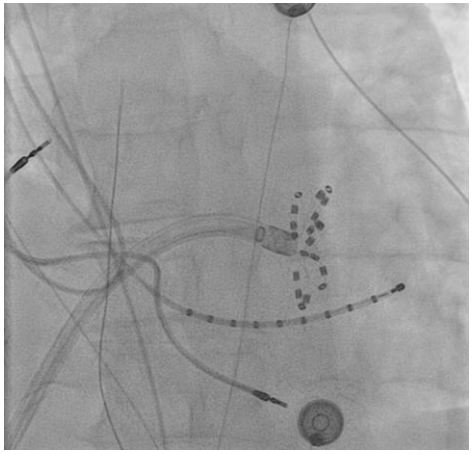
Cryoballoon + RF ablation of ART

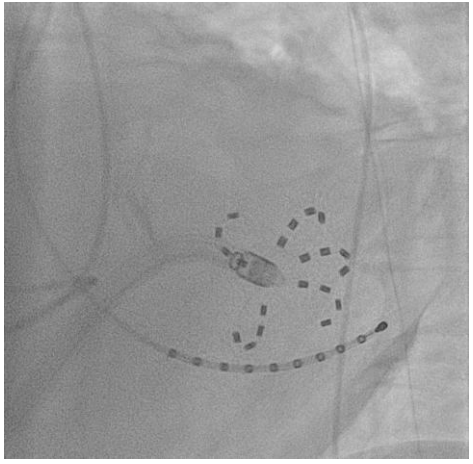
Cryoballoon only

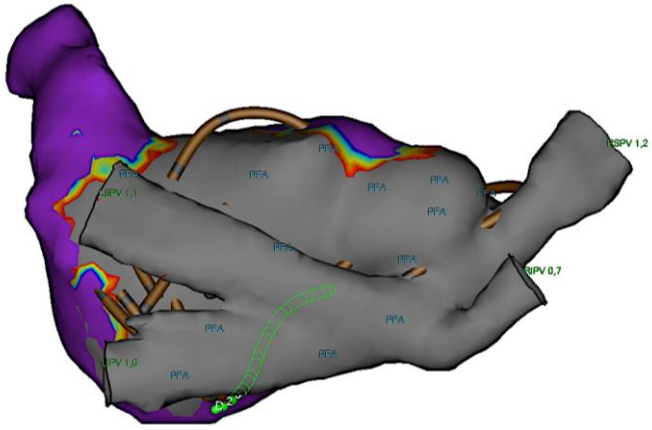
n=19 ACHD patients with AF/ART, mean age 58 years, RF ablation of ART in 9/19
Mean follow-up 26 months

Pulsed Field Ablation - Electroporation





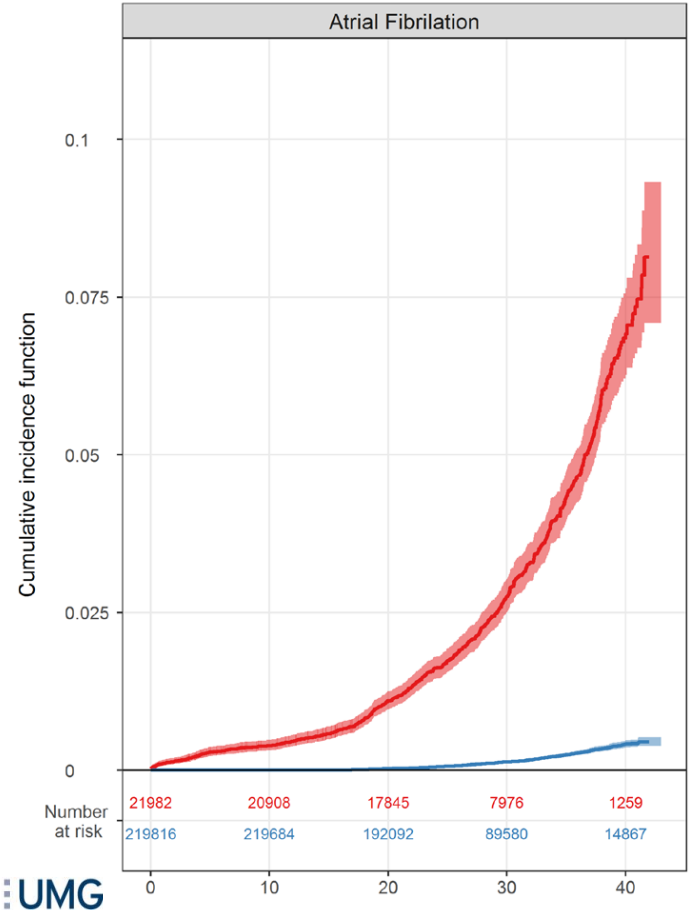





H&M

Atrial Fibrillation in ACHD Patients <40 Years

Mandalenakis et al., Circulation 2018



ACHD: n = 21,982

Controls: n = 219,816

Follow-up: 27±8.9 years