

### **Atrial arrhythmias in ACHD patients:**

## **Results of arrhythmia surgery**

7 February 2017 Jane Crosson, MD

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## History: start of major forays into A JOHNS HOPKINS arrhythmia surgery

- Akakura, Kyobu Geka, 1969: Surgical treatment for Wolff-Parkinson-White starts
- 1980s- early '90s, pinnacle for arrhythmia surgeons: WPW and VT ablations
- Catheter ablation arrives and arrhythmia surgeons lost their jobs, until....
- 1999

#### Impact of Arrhythmia Circuit Cryoablation During Fontan Conversion for Refractory Atrial Tachycardia

Barbara J. Deal, MD, Constantine Mavroudis, MD, Carl L. Backer, MD, Christopher L. Johnsrude, MD, and Albert P. Rocchini, MD

#### JOHNS HOPKINS

## **Fontan conversion and Maze**

- Designed for "failing" classic Fontan due to poor atrial mechanics &/or atrial arrhythmia
- Pioneered by Deal & Mavroudis in Chicago
- Conversion from atriopulmonary connection to either lateral tunnel or extracardiac conduit, along with Maze procedure

# Maze procedure in Fontan conversion



 Left atrial Cox maze added for for h/o atrial fibrillation



## Results of Maze with Fontan conversion



- 6% total mortality plus 5% subsequent OHT
- Late recurrence of tachycardia declined from 14% to 8% over study period (after adding leftsided Cox-maze)
- Poorer outcomes in:
  - Right or ambiguous ventricle
  - Protein-losing enteropathy
  - AV valve regurgitation, moderate to severe
  - Long cardiopulmonary bypass times

Mavroudis Ann Thorac Surg 2007



### Fontan conversion in other centers

#### Park, et al 2016:

- 21 patients with arrhythmia/failing classic Fontan had conversion with arrhythmia surgery
  - No operative deaths, 2 late deaths
  - Improved quality of life
  - 6/21 had recurrence of atrial tachycardia

## Fontan conversion in other centers: Multicenter European study



- Van Melle et al, Heart 2016: Surgical management of failing Fontan, in 22 centers
- Transplant, Fontan conversion or takedown
- 137/225 patients had conversion
- Only ½ of conversions included Maze
- Mortality 22% (mean f/u 7.7 yrs)
- In survivors, 89% in NYHA class I or II

## Surgical Maze procedure vs

- ~88% long-term control of atrial tachycardia following maze procedures in Fontan conversions
- <50% long-term control after multiple catheter ablations

PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease

Recommendations for surgical treatment:

- Class I: modified right atrial Maze is indicated during Fontan conversion, add left atrial Cox Maze if atrial fibrillation
- Class IIa: LA Cox Maze III with RA CTI in adults with CHD and atrial fibrillation

## Surgical treatment of arrhythmias JOHNS HOPKING in other CHD

#### Stulak et al, Ann Thorac Surg 2006

- 99 pts with CHD and associated atrial flutter &/or fibrillation underwent right-sided maze (no left atrial maze) during other repair
   – (ASD, TV repair or replacement, PVR, Fontan)
- Median age 43 yr, 70% had atrial fibrillation
- 6 early deaths, and mean f/u 2.7 years

   Flutter pts all free of arrhythmia (on meds)
   Atrial fib pts <sup>3</sup>/<sub>4</sub> free of arrhythmia

## Arrhythmia surgery in tetralogy of JOHNS HOPKINS Fallot



The Annals of Thoracic Surgery

Volume 81, Issue 5, May 2006, Pages 1786-1793

Cover

## 249 ToF pts reoperated

Original article

Outcomes After Late Reoperation in Patients With Repaired Tetralogy of Fallot: The Impact of Arrhythmia and Arrhythmia Surgery

Presented at the Fifty-second Annual Meeting of the Southern Thoracic Surgical Association, Orlando, FL, Nov 10-12, 2005.

Tara Karamlou, MD<sup>a</sup>, Ilana Silber, BS<sup>b</sup>, Robin Lao, BS<sup>b</sup>, Brian W. McCrindle, MD, MPH<sup>b</sup>, Louise Harris, MD<sup>b</sup>, Eugene Downar, MD<sup>b</sup>, Gary D. Webb, MD<sup>b</sup>, Jack M. Colman, MD<sup>b</sup>, Glen S. Van Arsdell, MD<sup>a</sup>, William G. Williams, MD<sup>a</sup>,  $\triangleq$ 



## Arrhythmia surgery in tetralogy of JOHNS HOPKINS Fallot



# What is the future of arrhythmia surgery?

- Changing surgical techniques with better results rendering some of this surgery obsolete, but substrate remain as long as myocardial incisions are made
- Most patients with atrio-pulmonary Fontans in their 30s+ now; conversions will be historical in another decade
- Surgery mostly now used in other diseases

## Future Directions: Prophylactic arrhythmia surgery?



- Atrial septal defect repair
- Ebstein anomaly
- Re-operation for tetralogy of Fallot
- Why not attempt amelioration of that arrhythmia risk with right-sided Maze?

## Right atrial prophylactic surge

#### Targets cavo-tricuspid isthmus



Mavroudis, Deal, Translational Pediatrics 2016

## Left atrial prophylactic surgery



## Targets atrial fibrillation substrate with pulmonary vein isolation





## Summary

- Surgical treatment of arrhythmia is constantly evolving as the substrates and alternative therapies change
- Arrhythmia surgery during Fontan conversion well-established
- Other arrhythmia surgery based on current arrhythmia is prudent
- Arrhythmia surgery based on predicted arrhythmia is an intriguing concept

## Thank you very much!







