Arrhythmia Surgery in Association With Congenital Heart Disease: Metamorphosis of a Concept in Flux

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I have no disclosures
Q49. The incidence of atrial arrhythmias in older patients with ASD is increased whether the ASD is closed or not at the time of discovery. The age that this incidence starts to increase is:

a. 25 year old  
b. 30 years old  
c. 35 years old  
d. 40 years old
Mayo Clinic: Atrial Arrhythmias in Congenital Heart Disease: Long-term Outcome After Surgical Repair of Isolated ASD Follow-Up 27 to 32 Years: 1990

- 123 patients (1956-1960) secundum or sinus venosus ASD
- Patients divided into quartiles: ≤ 11 y; 12-24 y; 25-41 y; > 41 y
- Perioperative mortality 3.3% (4 deaths)
- Patients repaired before age 25: Excellent Prognosis
- Patients repaired age 25 to 41
  - Good survival but less than age-matched controls
- Patients repaired > 41 years
  - Poor survival is out of proportion to RVH, PAP alone
- Repair in older patients-frequent complications
  - Late cardiac failure; Stroke; Atrial Fibrillation

Successful Surgical Treatment of Atrial Fibrillation: Review and Clinical Update: 1991

- 22 patients had Maze-Cox operation for AF
- No mortality; complications resolved
- Success in all patients at F/U
  - Junctional rhythm in 5/22 (23%)
  - Sinus bradycardia in 5/22 (23%)
  - Normal sinus rhythm in 17/22 (77%)
  - Dual chamber pacemakers in 7/22 (31.8%)

21 patients underwent modified Cox-maze concomitant with MV or ASD surgery

- 17 (81%) had chronic AF, 4 (19%) had paroxysmal AF; mean AF duration of 3.5 ± 3.6 yrs

Concomitant surgery on 9 (43%) mitral stenosis, 5 (24%) MR, 1 (5%) MR and AR, 3 (14%) ASD

1 death

Sinus rhythm restored before discharge in 17 (85%)

3 (15%) required antiarrhythmic therapy

3 month echo showed atrial contractility (A and E waves) in 12 (71%)

An F/U of 8 mo (range 3 to 23), 18 (90%) remained in sinus rhythm
Concomitant Mitral Valve or Atrial Septal Defect Surgery and the Modified Cox-Maze Procedure: 1996

Predictors of Sinus Rhythm Restoration After Cox-Maze Concomitant With Other Cardiac Operations: 1997

- 96 consecutive pts underwent maze III concomitant with other cardiac operations
- 10 excluded: 4 died, 6 required permanent pacemaker from sick sinus syndrome
- Mean age 59.8 yrs, 67 patients with MV disease
- Baseline characteristics of patients differ greatly from Cox patients
- Sinus rhythm was restored in 68 of 86 (79%)
- Magnitude of AF wave positively predicted postop sinus rhythm restoration
- LA diameter inversely related to postop sinus rhythm restoration
- Odds ratio of having both fine AF wave (< 1.0 mm) and enlarged LA diameter (≥ 65 mm) for patients with sinus rhythm restoration was 0.04 (95% CI, 0.01-0.28)
- AF wave and LA diameter were independent predictors of sinus rhythm restoration after maze in patients with chronic AF and organic heart disease

Maze Procedure for AF Associated With ASD: 1998

- 26 patients underwent maze concomitant with ASD closure
  - 25 were secundum, 1 was ostium primum
- With conventional right- and left-sided maze, AF disappeared in all patients
- 1 patient remained on junctional rhythm although AF ceased
  - Probably owing to sinus node dysfunction from long-standing RA pressure and volume overload
- Better results than for those with AF and MV disease
- In AF with ASD patients, LA dimension was ~1.5 times larger than normal on echos
  - Found that AF with ASD related to macroreentry circuit in enlarged LA


- 99 patients with CHD and associated atrial flutter and/or AF underwent concomitant right-sided maze at time of repair
- Median age 43 yrs (range, 9-72)
- Atrial flutter or AF was paroxysmal in 81, chronic in 18; median duration 2.9 yrs (range, < 1 mo-39.5 yrs)
- Primary diagnoses were Ebstein’s anomaly (47), other congenital tricuspid regurgitation (19), univentricular heart (11), isolated ASD (8), tetralogy of Fallot (8), other (6)
- At discharge, 83 were atrial flutter/AF free and 63 were in sinus rhythm
- Mean F/U of 2.7 yrs in 87 of the 93 early survivors extended to 8 yrs
  - 4 late deaths all from noncardiac causes
  - Of 83 known late survivors, 77 (93%) were flutter and/or AF free
  - 82 were in NYHA I or II

Incidence of Atrial Flutter/Fibrillation in Adults With ASD Before and After Surgery: 1999

- ECGs were examined before, early (days 3 and 7), and late (6 mo) after operation of 211 adults with ASD with pulmonary to systemic flow rate of 1.5:1 or greater
- Ages:
  - 18 to 40 yrs (n = 101)
  - 40 to 60 yrs (n = 83)
  - > 60 yrs (n = 27)
- Age of patients without arrhythmias before or after ASD closure was significantly lower than that of patients with flutter or AF
- Surgical correction leads to regression of atrial flutter but not AF
  - Therefore surgical repair of ASD must be combined with maze to abolish supraventricular tachyarrhythmias

Atrial Arrhythmia After Surgical Closure of ASDs in Adults: 1999

- 213 patients (82 men, 131 women) underwent surgical closure because of symptoms, substantial left-to-right shunt (ratio of pulmonary to systemic blood flow >1.5:1), or both
- 40 patients had sustained atrial flutter or AF before surgery
  - Compared with those who did not have atrial flutter or AF before surgery, those who did were older (mean age 59±11 v 37±13 yrs; P<.001)
  - They also had higher mean PA pressures (25±9.7 v 19.7±8.2 mmHg; P=.001)
- No perioperative deaths
- Late events (>1 mo postop): stroke (6, all but one with flutter or AF, 1 who died); death (2 from noncardiac causes)
- At mean F/U 3.8±2.5 yrs, 24 of 40 patients (60%) continued to have flutter or AF
  - Mean age of these patients was greater than that of the 16 who converted to sinus rhythm (P=.02)
- New onset flutter or AF was more likely to develop at F/U in older patients (> 40 yrs) than younger (5 of 67 v 0 of 106, P=.008)
- Multivariate analysis showed older age (> 40 yrs) at time of surgery (P=.001), presence of preop flutter or AF (P<.01), and presence of postop flutter/AF or junctional rhythm (P=.02) were predictive of late postop flutter or AF

Surgical Treatment of Atrial Fibrillation; Meta-analysis: 2005

- Review of AF treatment alternative sources of energy (radiofrequency-microwave and cryoablation; group I) versus classic cut and sew Cox-maze III (group II), which claims 97-99% sinus rhythm success rate
- 48 studies were reviewed with 3,832 patients (2,279 in group I and 1,553 in group II)
- Mean duration of AF, LA diameter, and LVEF were 5.4 v 5.5 yrs (P = .90); 55.5 v 57.8 mm (P = .23); 57 v 58% (P = .63)
- Postop sinus rhythm rates were 78.3 v 84.9% (P = .03)
  - Cox-maze III was used in younger patients (55 v 61.2 yrs; P = .005), more often to treat paroxysmal (22.9 v 8%; P = .05), and lone AF (19.3 v 1.6%)
  - Alternative sources of energy were used to treat permanent AF (92%), almost always as concomitant (98.4%), and increasingly with non-mitral valve surgery (18.5%)
- After correction for the variations, postop sinus rhythm rates did not differ significantly between groups (P = .260)

Surgical Ablation as Treatment for the Elimination of AF: A Meta-Analysis: 2006

- Review of 69 studies of 5,885 patients who underwent surgical ablation (67% biatrial and 33% LA) for prolonged AF (lasting > 6 or 12 mo)
- Survival rates for patients with biatrial procedures were similar to LA procedures
- Patients who underwent biatrial ablation demonstrated superior freedom from AF (return to sinus rhythm) at all time points

Barnett SD, Ad N. J Thorac Cardiovasc Surg. 2006;131:1029-1035
Surgical Treatment of Arrhythmias in Adults With CHDs: 2006, 2008

• 50 adults (mean age 39 yrs) underwent irrigated radiofrequency ablation (IRA) during cardiac surgery
  – 31 right-sided maze procedures, 13 Cox-maze III, 6 RV ablations; 14 implanted pacemakers
• 2 deaths (4%) not related to ablation; over 28 mo F/U 48 patients still alive with NYHA class I or II
• All discharged on antiarrhythmic oral tx for 3 mo; all underwent Holter at 3 and 6 mo; 5 underwent programmed VT stimulation 6 mo postop
• 43 patients still in spontaneous sinus rhythm; 2 in sinus rhythm on chronic oral antiarrhythmic tx for AF recurrence; 2 in stable AF; 1 pacemaker rhythm

Arrhythmia Surgery for AF Associated With ASD: Right-Sided Maze Versus Biatrial Maze: 2012

• 62 patients underwent surgical closure of ASD and various maze procedures for AF
  – Median age 59 yrs (range 34-79)
  – Clinical manifestations of AF were paroxysmal (8), persistent (16), longstanding persistent (38)
  – Lesions were RA (23) in group 1, RA plus PV isolation (6) in group 2, and BA (33) in group 3
• Median F/U of 54 mo (range 3-149)
  – No early and 1 late death
  – Group 1 showed significantly decreased time to recurrence of AF in comparison to group 3 (P = .0341)
  – Normal sinus rhythm without any AF recurrence at postop 2 and 5 years were 57% and 45% in group 1, 67% and 50% in group 2, and 82% and 69% in group 3

Isthmus-dependant right atrial flutter as the leading cause of atrial tachycardias after surgical ASD repair: 2013

- 54 patients were studied to evaluate AT after ASD repair
  - Median age 47.3 yrs ± 14.5 yrs
  - In 30 patients, closure by suture (55.6%); by patch (44.6%)

- Mechanisms of AT/Outcomes
  - Isthmus dependent right atrial flutter in 29 (69.0%)
  - Atriotomy dependent in 7
  - 5 patients developed AF (11%); 60% remained arrhythmia free

- Conclusions
  - Isthmus dependent AT is the most common arrhythmia
  - Mode of closure made no difference

Wasmer K et al. Munster, Germany Int J Cardiol 2013; March 26th
Therapeutic Arrhythmia Surgery

• Historical review
  – Accessory pathways (Walt Sealy)
  – Focal (automatic) tachycardia (multiple authors)
  – AVNRT (incremental success)
  – ART (Cox; Theodoro; Deal, Mavroudis, Backer)
  – AF (Cox, others)
  – VT (multiple investigators)
Transcatheter Arrhythmia RF Ablation

• Historical review
  – Accessory pathway ablation
  – AVNRT, ART, VT, Focal, AF
    • Low failure rate
    • Low complication rate
    • Benefit of multiple attempts
  – Present day practice
    • Transcatheter approach first-line therapy
When Is Surgical Ablation Performed?

Present Day Practice

• Surgery as primary therapy for arrhythmia
  – Failed catheter ablation (multiple)
  – Hemodynamic instability ↓ LV function; ECMO
  – Cavity access challenges
  • AVNRT (Senning/Mustard)

• Preoperative catheter ablation
  – Accessory pathways
  – Exceptions: Ebstein’s patients multiple
  – Cross-clamp time considered
Present Day Practice

• Arrhythmia surgery in association with repairs
  – MVR with MAZE for AF (adults, children)
  – ASD with MAZE for AF, ART (adults)
  – VSD with ablation for VT (children)
  – Redo Tetralogy with atrial MAZE and/or VT ablation (older ages)
  – AICD may be indicated after postop VT study
  – Redo Fontan atrial MAZE (L&R); pacer therapy
Prophylactic Arrhythmia Surgery

Present Day Practice

• Does it exist? YES
  – Chicago (Deal, Mavroudis, Backer) select cases
  – Mayo Clinic (Dearani)
  – Virginia (Ad)
  – Boston *(multiple authors) Only prospective randomized study for Fontan Patients; lesions based on animal study-not human
  – Others
Boston Prospective Study

Prophylactic Arrhythmia Surgery

Should it exist?

• Advantages/reasonable application
  – Treat arrhythmia which has high occurrence rate
  – Lesions have to be SAFE
  – Lesions should be studied
  – Pathology and age at therapy need to be defined
  – Control of arrhythmias, preservation of cardiac function, & prevention of dilatation?
  – Prevention precludes dilatation; therapy for arrhythmia performed after dilatation takes place
Prophylactic Arrhythmia Surgery

Should it exist?

• Disadvantages
  – Can create arrhythmias in short term (Nodal)
  – Extensive lesions have higher incidence of nodal rhythm; sinus venosus defect example
    • Cox contends that there is no difference between therapeutic and prophylactic lesions (personal)
  – Increased cross-clamp/CPB time
  – Pacemaker possibility (short- and long-term)
Prophylactic Arrhythmia Surgery
Should it exist?
What are the choices?

• Prophylactic arrhythmia surgery
  – Treat potential complications before they occur
• No prophylactic arrhythmia surgery
  – Transcatheter ablation if/when arrhythmias occur
  – “Chase the arrhythmias”
  – Left Atrial Clot/Coumadin issues
  – Antiarrhythmic agents
Prophylactic Arrhythmia Surgery

Inclusionary Criteria (Patient Selection)

- ASD > ? Age; 30; > 40
- Ebstein’s anomaly accessory connection; ART, AF
- Redo AV canal for left AV valve regurgitation without arrhythmias
- Redo Fontan without arrhythmias
- Redo TOF without arrhythmias
- Redo congenital heart surgery (in general) with
  → Right atrial dilatation; definition
  → Left atrial dilatation; definition
Review of Potential Lesions For Prophylactic Arrhythmia Surgery
ASDs in Adults / Prophylactic Maze

• For left atrial surgical Maze we know:
  – Atrial incisions can cause bleeding
  – Atrial incisions can interfere with atrial contraction

• For left atrial transcatheter Maze we know:
  – Lesions may not be transmural
  – Lesions can cause pulmonary vein stenosis/occlusion
  – Lesions can cause esophageal injury
Standard Right Atrial Maze
Redo Fontan for Single Ventricle
Therapeutic Lesions
Tricuspid Atresia
Therapeutic Lesions
Single Right Ventricle
Therapeutic Lesions
Unbalanced AV Canal
Left-Sided Maze
Potential Lesions for Anomalies of Venous Return and Atrial Appendages
Prophylactic Maze
Conclusions

- Onus for prophylactic Maze is on the clinician
  - Do no harm
  - Apply Lesions that will work
  - Weigh and balance the projected good and the projected harm
- Support prospective studies
  - Measure and record outcomes
  - Report the results; analyze the statistics
Arrhythmia Surgery in Patients with Congenital Heart Disease

May 23 - 24, 2013
Florida Hospital Nicholson Center
Orlando, Florida

Course Director:
Constantine Mavroudis, MD
- Director, Congenital Heart Center
  Florida Hospital for Children
- Site Director, Johns Hopkins
  Children’s Heart Surgery
  Florida Hospital for Children
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Objectives:
1. Explain the principles of intraoperative electrophysiologic mapping.
2. Review the clinical characteristics of prophylactic and therapeutic arrhythmia surgery.
3. Identify the various atrial and ventricular arrhythmias and up-to-date strategies for intraoperative ablation.

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Arrhythmia Surgery in Patients with Congenital Heart Disease
May 23 - 24, 2013 Orlando, Florida, USA

Barbara Deal: Atrial / Ventricular Arrhythmias in Congenital Heart Disease
Louise Harris: Electrophysiologic Mapping and Transcatheter Ablation
Niv Ad: Intraoperative Mapping Cryoablation for Atrial and Ventricular Tachycardia
Constantine Mavroudis: Anatomic Variants of Arrhythmia Surgery
John Stulak: Arrhythmia Surgery for Patients with Ebstein’s Anomaly
Sabrina Tsao: Intraoperative Electrophysiologic Analysis and Pacemaker Strategies
Carl Backer: Epicardial Lead Placement/Pacemaker Strategies in Rhythm Disturbances
George Palmer: Strategies for Difficult Transvenous Pacemaker Lead Removal
Constantine Mavroudis: Rationale of Prophylactic Arrhythmia Surgery
Niv Ad: Prophylactic Maze in Repair of Congenital Heart Defects
Duke Cameron: The Future of Cardiothoracic Surgery
Q49. The incidence of atrial arrhythmias in older patients with ASD is increased whether the ASD is closed or not at the time of discovery. The age that this incidence starts to increase is:

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