Catheter Ablation in the Fontan

Approaches in Atriopulmonary, Lateral Tunnel and Extracardiac

Mark McGuire
Royal Prince Alfred Hospital
Prince of Wales Hospital
Sydney Australia
Fontan Procedures

Atriopulmonary Connection
1971

Lateral Tunnel (TCPC)
1987

Extracardiac
1990
Types of Fontan Australia & New Zealand (n=1006)

Figure 1. Distribution of the techniques used in the growing population of Fontan patients alive in Australia and New Zealand. AP indicates atrio-pulmonary connection; ECC, extracardiac conduit; and LT, lateral tunnel.

- Extracardiac Conduit (n:532) 53%
- Lateral Tunnel (n:262) 26%
- Atrio-pulmonary (n:168) 17%

d’Udekm Y et al Circulation 2014;130:[suppl 1]S32-S38
Risk of Arrhythmia Post Fontan (n=520)

**Figure 1** Hazard of IART Following the Fontan Operation, With 95% Confidence Bands

Date of diagnosis was missing for 1 patient, thus the number at risk initially is 519. IART = intra-atrial re-entrant tachycardia.
Risk of Arrhythmia Post Fontan (n=520)
Risk Depends on Type of Procedure

Figure 2
Freedom From IART Following the Fontan Operation by Type of Fontan Procedure
ADJUSTED INCIDENCE ATRIAL ARRHYTHMIAS
No difference between LT & ECC P=0.22

Stephenson EA et al
J Am Coll Cardiol 2010;56:890–6
Modified by Khairy P
Acute Procedural Success and Freedom From Arrhythmia Recurrence After Successful Ablation

Yap S-C et al JACC 2010;56:1589
Challenges

- Multiple reentrant circuits
- Thick walled chambers
- Large chambers: catheter contact
- Low flow (coagulum formation)
- Limited access (LT, ECC, Patch over TV, ilio-femoral stenosis)
- Difficulty identifying site of conduction system
- Heart block: urgent thoracotomy
- Tachycardia poorly tolerated
- Stable timing reference difficult
- Cannulating CS
Right Atrium Post Fontan

EP Walsh
Fontan Procedures – Specific Issues

**APC**
- Chamber size +++
- Wall Thickness +++
- No. tachycardias +++
- Baffle puncture usu. not required
- Baffle puncture difficulty +

**Lateral Tunnel**
- Chamber size +
- Wall Thickness ++
- No. tachycardias ++
- Baffle puncture usu. not required
- Baffle puncture difficulty +
- Fenestrations often present

**ECC**
- Chamber Size +
- Wall Thickness +
- No. tachycardias +
- Conduit crossing always required
- Conduit puncture difficulty +++
Mr DP 22
Fontan
Tricuspid atresia
Extensive Scar Right Atrium Post Fontan

- SVC
- APC
- Bipolar
- 2-Negative > 176 Points

ML Fontan
AP View
Figure 6. Relationship between extracardiac conduit and pulmonary venous atrium. An axial view of a MRI scan is shown in a patient with an extracardiac conduit (ECC). The arrows mark the space between the ECC and neopulmonary venous atrium, composed of the right (RA) and left (LA) atria.
Transconduit Puncture: female 33, situs inversus, laevocardia, mitral atresia, ECC
Multi-centre (9)
N: 36 patients
Primary ECC: 24; Conversion 12

Procedures: 46
Access to atrium:
  Puncture: 63%, Fenestration: 26%
Acute success: 83%
No complications due to conduit puncture
Arrhythmia recurrence: 17% @ 0.4 yrs
Trapdoor Modification to Extracardiac Fontan

A

pericardial patch

B

SVC

RPA

C

TS-needle

Marker for fluoroscopy

Aboulhosn J et al, Congenit Heart Dis 2010;5:430
Robotic Magnetic Navigation in Congenital Heart Disease

N= 13
No recurrence in 10/13, mean FU 201 days

Ernst S et al, Circ Arrhythm Electrophysiol. 2012;5:131-139

• Very expensive
• Others haven’t had same results
• Several centres have abandoned
High Resolution Simultaneous Multipoint Mapping

Channel through scar
Catheter Ablation in Fontan

- 39 consecutive ablations in 28 Fontan patients, 2000-2017
- Mean age 33+/- 7 years
- Mean 2.8 tachycardias per case (range 0-10)
- Ablation strategy:
  - Targeted via activation / entrainment mapping in 15 (38%)
  - Substrate-based approach in 11 (28%)
  - Targeted + substrate in 11 (28%)
  - Slow pathway ablation for concurrent AVNRT in 2 cases
- Mean follow up 4.2 years
Ablation: Fontan Types

![Ablation: Fontan Types Diagram]
Clinical Arrhythmia Severity Score (CASS)* Following Ablation

* Triedman JK et al JACC 2002  
Moore B et al 2017
Location of the critical isthmus
Acute Procedural Success & Complications

- “Complete” success in 23/39 cases (59%)
- “Partial” success in 10/39 cases (26%)
- “Unsuccessful” in 6/39 cases (15%)
- Suspected clinical arrhythmia ablated in 79%
- Complications
  - 4 femoral vascular complications (3 required minor operative intervention)
  - 2 cases where temporary pacing was required post procedure for transient bradycardia (1 temporary pacing wire, 1 transcutaneous)
  - No strokes or pericardial effusions / tamponade
Freedom from Death, Transplant, Conversion Surgery Post-Ablation
Summary

- Difficult procedure (median proc. time 300 mins)
- Most patients benefit (acute success: 50-80%) – improvement sustained for several years
- Evolving technology likely to improve acute success rate
- Complication & mortality rate low
- Palliative rather than curative: recurrence rate significant
- Should it be always be considered before revision surgery in patients with good haemodynamics?
# Clinical Arrhythmia Severity Score*

<table>
<thead>
<tr>
<th>Category</th>
<th>Score</th>
<th>Category</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Documented Arrhythmia</td>
<td></td>
<td>Frequency of Cardioversion</td>
<td></td>
</tr>
<tr>
<td>none</td>
<td>0</td>
<td>no DCCV</td>
<td>0</td>
</tr>
<tr>
<td>non-sustained</td>
<td>1</td>
<td>one DCCV</td>
<td>1</td>
</tr>
<tr>
<td>sustained</td>
<td>2</td>
<td>Pacemaker conversion</td>
<td>2</td>
</tr>
<tr>
<td>incessant</td>
<td>3</td>
<td>2 or more DCCV</td>
<td>3</td>
</tr>
</tbody>
</table>

| Arrhythmia Severity            |       | Antiarrhythmic Drugs          |       |
| asymptomatic                   | 0     | none or digoxin only          | 0     |
| palpitations only              | 1     | class II or IV                | 1     |
| syncope/CHF/thrombosis         | 2     | class I or III                | 2     |
| cardiac arrest                  | 3     | amiodarone toxicity           | 3     |

Outcomes

• 1) Acute procedural success
  • “Complete” = all induced arrhythmias ablated (none inducible end of case)
  • “Partial” = some, but not all, induced arrhythmias ablated (≥ 1 inducible end of case)
  • “Unsuccessful” = no arrhythmias able to be ablated

• 2) Complications

• 3) Clinical arrhythmia severity score at baseline, 3-6, 12 and 24 months

• 4) Freedom from death, transplant or conversion surgery composite
Transthoracic Puncture For Access to PV Atrium Lateral Tunnel Fontan

Nehgme, RA (Heart Rhythm 2006;3:37–43)
ATRIAL FLUTTER AFTER FONTAN (N:334)
Risk Depends on Procedure Type

Fishberger  S. B. et al.; J Thorac Cardiovasc Surg 1997;113:80-86
ATRIAL FLUTTER AFTER FONTAN

Probability of Freedom from Atrial Flutter

Time Since Fontan Procedure (Years)

< 3 yrs
3-10 yrs
> 10 yrs

p < 0.001

Fishberger S. B. et al.; J Thorac Cardiovasc Surg 1997;113:80-86
REPAIRED CONGENITAL HEART DISEASE
A Perfect Storm For Arrhythmia

Substrate

- Scarring and fibrosis
- Surgical incisions and suture lines
- Atrial dilatation
- Myocyte stretch
- Hypertrophy
- Cellular hypoxia
Preparation For Ablation

- Obtain surgical report
- Imaging: CT, CMRI, TTE, TEE, venography
- Assess hemodynamic status
- Plan vascular approach: femoral or subclavian / jugular?
- Plan chamber access:
  - Retrograde aortic
  - Fenestration
  - Puncture baffle
  - Transthoracic puncture
  - Surgical assistance – hybrid access
- Plan for heart block & need for pacing in Fontan
Equipment for Ablation in Fontan and Atrial Switch

- 3D mapping
- Deflectable sheaths
- Irrigated ablation catheters
- Contact force?
- “Adequate” number catheters / electrodes
- TEE
- ICE?
- Magnetic navigation?
Site of IART in Fontan Patients
N= 19

Yap S-C et al JACC 2010;56:1589
Mr DP 22
Fontan
Tricuspid atresia
SB Fontan
Right Lateral View

Atriopulmonary Connection

SVC

TV patch

IVC

LAT

2-1-ReMap > 219 Points

1.25 cm

152ms

-133ms
Patch Over Tricuspid Valve
Hindering ablation of CTI

Atriopulmonary Connection

SVC

TV patch

IVC
Ablation Adult Congenital Heart Disease

Tips

• Use entrainment judiciously
• Check frequently that Tc has not changed
• Anaesthesiologist (anesthesiologist)
Fontan Procedures

Atriopulmonary Connection  
c.1968

Lateral Tunnel  
(TCPC)  
1987

Extracardiac  
1990
Risk of Arrhythmia Post Fontan (n=520)
Risk Depends on Type of Procedure

Figure 2
Freedom From IART Following the Fontan Operation by Type of Fontan Procedure

Stephenson EA et al
J Am Coll Cardiol
2010;56:890–6
Baffle Puncture

Baffle Materials

- Pericardium
- Cardiac flap
- Synthetic: PETE (Dacron™), PTFE (Teflon™, Gore-Tex™)

Methods

- Direct puncture with trans-septal needle / sheath
- Adjunctive radiofrequency energy
- Proprietary RF needle
- Balloon angioplasty to enlarge puncture

Tip*: Very stiff mitral valvuloplasty wire may aid access
### Adverse Events Associated with Trans Baffle Access

Boston Children’s Hospital

Puncture n=54, Baffle leak/fenestration n= 20

#### Table 6. Adverse Events

<table>
<thead>
<tr>
<th>Severity</th>
<th>Adverse Event</th>
<th>TBA</th>
<th>No TBA</th>
<th>P Value</th>
</tr>
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<tbody>
<tr>
<td>Catastrophic</td>
<td>Death</td>
<td>2</td>
<td>0</td>
<td>—</td>
</tr>
<tr>
<td>Major</td>
<td>Shunts and cyanosis</td>
<td>2</td>
<td>0</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>Permanent AV block</td>
<td>1</td>
<td>0</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>CS thrombosis</td>
<td>0</td>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>Retroperitoneal bleed</td>
<td>1</td>
<td>0</td>
<td>—</td>
</tr>
<tr>
<td>Moderate</td>
<td>Pseudoaneurysm</td>
<td>1</td>
<td>0</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td>Acute kidney injury</td>
<td>0</td>
<td>3</td>
<td>0.05</td>
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<tr>
<td></td>
<td>Hemoptysis</td>
<td>1</td>
<td>0</td>
<td>—</td>
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<tr>
<td>All</td>
<td>Total adverse events</td>
<td>8</td>
<td>4</td>
<td>1</td>
</tr>
</tbody>
</table>

AV indicates atrioventricular; CS, coronary sinus; TBA, transbaffle access.

Correa R et al: J. Am Heart Assoc 2013;2:e000325
Right Atrial Maze Procedure During Fontan Conversion

Arrhythmia After Fontan with Good Haemodynamics

Atrial Futter → Drug → Drug Effective

- yes → Continue Drug
- no
  - APC / LT
  - Extracardiac
    - ablation
      - effective
        - Conversion or Anti-tachycardia PM or Ablate & Pace
      - not effective
        - Ablate AVN? or Arrhythmia Sx
        - Transplant?
• Challenging

• Preparation & planning key to success & avoiding bad outcomes

• Modern equipment/technique facilitates ablation

• Quality of life significantly improved

• Significant risk of late arrhythmias
TITLE
Right Atrial Wall Post Fontan

(trichrome)

ENDO

EPI

8 mm
Extracardiac Fontan

Figure 1. Rendering of post-surgical anatomy
There is tricuspid valve atresia with hypoplastic right ventricle and well-developed systemic left ventricle (LV). Great arteries are D-transposed with the aorta (Ao) anteriorly dextraposed and the banded pulmonary artery (PA) posteriorly levoaposed. Superior vena cava (SVC) is connected to PA via bidirectional Glenn shunt. Extracardiac conduit connects inferior vena cava to PA. Note close proximity of extracardiac conduit to “right atrial” portion (RA) of pulmonary venous atrium. The “left atrium” (LA) and mitral valve (MV) are labeled.

Dave, A: Heart Rhythm. 2010; 7(3): 413–416
Extracardiac Fontan: Access To RA : Conduit Puncture

Dave, A: Heart Rhythm. 2010 ; 7(3): 413–416
Fontan Conversion and Arrhythmia Surgery
1994 - 2007 (n=118)

* Sedation administration, 2.5 yrs; Automobile accident, 11.3 yrs

### ATRIAL ARRHYTHMIAS AFTER SURGICAL REPAIR

<table>
<thead>
<tr>
<th>Lesion / Repair</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontan(^1,^2)</td>
<td>41-50%</td>
</tr>
<tr>
<td>Mustard / Senning(^3)</td>
<td>30%</td>
</tr>
<tr>
<td>TOF(^4)</td>
<td>34%</td>
</tr>
</tbody>
</table>

2: Ghai A. JACC 2001;37:585  
3: Flinn CJ. NEJM 1984;310:1635  
Fenestration Effect on Survival?

![Survival Probability Graph](image)

(B)
Figure 1. Two types of modified Fontan connections are shown. A, Atriopulmonary connection. B, Total cavopulmonary connection. See text for description. (Figure is used with permission of the Mayo Foundation.)
Figure 2. Surgical technique for APC-lat modification. A, Incisions in the free wall of right atrium and atrial appendage. B, Anastomosis of flap of atrial appendage to pulmonary artery. C, Intra-atrial baffle inserted to direct blood from inferior vena cava to atrial appendage–pulmonary artery connection. D, Pericardial patch to complete the connection. (Figure is used with permission of the Mayo Foundation.)
Extracardiac Fontan: Access To RA : Conduit Puncture

Figure 2.
Fluoroscopic images showing (A) still-frame from conduit angiogram, (B) transseptal needle with contrast staining of conduit prior to crossing, (C) dilator of transseptal sheath crossing conduit into pulmonary venous atrium, (D) Toray valvuloplasty wire coiled in atrium with angioplasty balloon inflated across conduit crossing, and (E) still-frame from angiography of pulmonary venous atrium.

Dave, A: Heart Rhythm. 2010 ; 7(3): 413–416
REPAIRED CONGENITAL HEART DISEASE
A “Perfect Storm” For Arrhythmia

Substrate
- Scarring and fibrosis
- Surgical incisions and suture lines
- Atrial dilatation – bigger chambers
- Myocyte stretch
- Hypertrophy & heart failure
- Cellular hypoxia
“Late” Arrhythmias

Bradycardia:
- Sinus node dysfunction
- Intra-atrial block

Tachycardia:
- Intra-atrial reentry (atrial flutter)
- Standard SVT (AVNRT & accessory pathways)
- Atrial fibrillation (following AV valve regurgitation)
- Ventricular arrhythmias with ventricular failure
ARRHYTHMIAS POST FONTAN

Bradycardia

- Sinus node dysfunction but also intra-atrial block
- Probable cause: sinus node fibrosis +/- ischaemia
- Incidence: highest LT, intermediate APC, lowest for extracardiac
Fontan Conversion – Arrhythmia Surgery – Pacemaker Implantation

Epicardial Leads

- Placement difficult because of adhesions

- Bipolar – required for anti-tachycardia pacemakers

- Both sides of ventricular mass for CRT?
Pacing Ventricle Via Coronary Sinus - APC Fontan

Figure 2  Chest radiographs in posteroanterior (A) and lateral (B) projections, after successful ventricular pacing through the coronary sinus. The redundant epicardial system is still in situ.

Blackburn M (Br HeartJ 1993;70:578-579)
(A) Angiography showing perpendicular relation of the transseptal needle (large arrows) to the Goretex baffle (small arrows) via the transhepatic route. The atrial septum has been tagged with a small amount of contrast. TE, tip of transesophageal ultrasound probe. (B) x-ray showing atrial and ventricular location of the pacing wires with a generous intracardiac loop and generator in the anterior abdominal wall.
Fenestration (lateral tunnel) No Effect on Need for PPM?


(D)

Probability of Freedom from Pacemaker Implantation

Time After Fontan Procedure (Years)

Patients at risk

f-TCPC 18 16 10 8 7 2
TCPC 50 45 45 44 40 30 11 1
APA 19 18 16 15 15 12 12 9 2 1
Atrial flutter / fibrillation / SVT

- 17 % after 5 years\(^1\)
- 16 % after 5 years\(^2\)
- 12.5% after 8.9 years\(^3\)
- 10.6 % after 3.7 years\(^4\)
- 41 % after 11 years\(^5\)
- 7.3% after 8.6 years\(^6\)

2. Fishberger SB: J Thorac Cardiovasc S 1997;113:80
5. Ghai A: J Am Coll Cardiol 2001;37:585–92
6. Stephenson EA: J Am Coll Cardiol 2010;56:890
Effect of Arrhythmia On Survival Post-Fontan?
Ghai et al (Toronto) JACC 2001:37:585

Atrial Tachyarrhythmias in Adults With Fontan

Time from Fontan Procedure (years)

| Arrhythmia Group (n) | 39 | 38 | 26 | 8 |
| Arrhythmia-Free Group (n) | 55 | 46 | 35 | 11 |

No Arrhythmia
Arrhythmia
ATRIAL FLUTTER AFTER FONTAN

Risk Factors

- Procedure type (APC vs LT vs extracardiac)
- Fenestration and or collaterals
- Age at surgery
- AV valve regurgitation
Risk of Arrhythmia Post Fontan (n=520)
Risk Depends on Type of Procedure

Figure 2
Freedom From IART Following the Fontan Operation by Type of Fontan Procedure
ATRIAL FLUTTER AFTER FONTAN
Risk Depends on Procedure Type

Probability of Freedom from Atrial Flutter

Time Since Fontan Procedure (Years)

Fishberger S. B. et al.; J Thorac Cardiovasc Surg 1997;113:80-86
# Atrial Arrhythmias After Surgical Repair

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<tr>
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</tr>
<tr>
<td>Mustard / Senning&lt;sup&gt;3&lt;/sup&gt;</td>
<td>30%</td>
</tr>
<tr>
<td>TOF&lt;sup&gt;4&lt;/sup&gt;</td>
<td>34%</td>
</tr>
</tbody>
</table>

2: Ghai A. JACC 2001;37:585  
3: Flinn CJ. NEJM 1984;310:1635  
Importance of CT/MRI scan

Dextrocardia
Interrupted IVC

RAO View

Azygos Vein

Right SVC

Left SVC

Hepatic Veins

MV

TV
Fenestration (lateral tunnel) Decreases Late Arrhythmias?

Effect of Collaterals on Tachyarrhythmias?

![Curve diagram showing the probability of freedom from tachyarrhythmia over time for patients with and without collaterals. The graph indicates that patients with collaterals are less likely to experience tachyarrhythmias, with a statistically significant difference at P=0.05.](image-url)

- **Collateral (+)**
- **Collateral (-)**

**Patients at risk**
- Collateral (+): 13, 12, 11, 10, 10, 8, 7, 5
- Collateral (-): 5, 5, 4, 4, 4, 3, 3, 3

**Time after atrio pulmonary anastomosis procedure (years):**
0, 2, 4, 6, 8, 10, 12, 14, 16, 18, 20
A, Activation map during MacroAT in ASD patient 1 shows continuous activation around smaller upper dense scar (gray area with gray tags) and line of double potentials (pink tags) and through channel (width 1.6 cm) between scars.

Mustard: trans-baffle approach to right (PV) atrium

RAO

PA

LAA
LV
Abl.

LAA
LV
Abl.
BAFFLE PUNCTURE
In Mustard/Senning & Fontan Patients

Guide wire in RSPV

Balloon

LAO view

35 atm.
Extracardiac Fontan: Access To RA : Conduit Puncture

Dave, AS: Heart Rhythm. 2010 ; 7(3): 413–416
Channel through scar
Meta-analysis Late Arrhythmias
LT vs ECC

<table>
<thead>
<tr>
<th>Study or Subgroup</th>
<th>ILT Events</th>
<th>Total</th>
<th>ECC Events</th>
<th>Total</th>
<th>Weight</th>
<th>Odds Ratio M.H, Fixed, 95% CI</th>
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</thead>
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<tr>
<td>Azakie 2001</td>
<td>1</td>
<td>43</td>
<td>2</td>
<td>54</td>
<td>5.9%</td>
<td>0.62 [0.05, 7.06]</td>
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<tr>
<td>Balaji 2014</td>
<td>9</td>
<td>602</td>
<td>17</td>
<td>669</td>
<td>54.2%</td>
<td>0.58 [0.26, 1.32]</td>
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<tr>
<td>Bossers 2015</td>
<td>4</td>
<td>46</td>
<td>1</td>
<td>69</td>
<td>2.5%</td>
<td>6.48 [0.70, 59.92]</td>
</tr>
<tr>
<td>d’ Udekerem 2007</td>
<td>4</td>
<td>105</td>
<td>1</td>
<td>48</td>
<td>4.5%</td>
<td>1.86 [0.20, 17.11]</td>
</tr>
<tr>
<td>Fiore 2007</td>
<td>11</td>
<td>113</td>
<td>4</td>
<td>48</td>
<td>17.3%</td>
<td>1.19 [0.36, 3.93]</td>
</tr>
<tr>
<td>Hakacova 2008</td>
<td>1</td>
<td>60</td>
<td>0</td>
<td>41</td>
<td>2.0%</td>
<td>2.09 [0.08, 52.64]</td>
</tr>
<tr>
<td>Lee 2007</td>
<td>2</td>
<td>67</td>
<td>3</td>
<td>98</td>
<td>8.1%</td>
<td>0.97 [0.16, 5.99]</td>
</tr>
<tr>
<td>Nakano 2004</td>
<td>2</td>
<td>88</td>
<td>0</td>
<td>79</td>
<td>1.7%</td>
<td>4.60 [0.22, 97.19]</td>
</tr>
<tr>
<td>Nürnberg 2004</td>
<td>3</td>
<td>29</td>
<td>0</td>
<td>45</td>
<td>1.2%</td>
<td>12.02 [0.60, 241.80]</td>
</tr>
<tr>
<td>Sarkis 2011</td>
<td>7</td>
<td>26</td>
<td>1</td>
<td>25</td>
<td>2.5%</td>
<td>8.84 [1.00, 78.22]</td>
</tr>
</tbody>
</table>

Total (95% CI) 1179 1176 100.0% 1.37 [0.85, 2.21]

Total events 44 29

Heterogeneity: Chi² = 12.27, df = 9 (P = 0.20); I² = 27%
Test for overall effect: Z = 1.30 (P = 0.19)

Li D et al Pediatr Cardiol 2017;38:873-880