VT Ablation in ARVD, Adult CHD

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Introduction: ARVC

• Genetically inherited disease

• Characteristics
  – Progressive replacement by fibrofatty infiltration
  – Syncope, heart failure, ventricular tachyarrhythmias and sudden death
  – Three Clinical Stage:
    • Subclinical stage, Electrical stage, Structural stage

Ellinor et al. Heart Failure Clinics 2010
Pathogenesis of ARVC VT

Transmembrane protein
Linker
Desmoplakin

DSP
DSC2
PKP2
DSG2

After increased mechanical stress

Mark S. Link et al. JACC 2014
RV dilatation

**ECG**

1) QRS in V1 ≥ 110 ms
2) epsilon wave
3) T wave inversion
4) low-voltage

**Late potentials**

**Plakoglobin**

**Control**

**ARVD/C**
Revised Task Force Criteria-ARVC
(2 major, or 1 major + 2 minor or 4 minor)

Major Criteria

• Regional RV dysfunction/structural changes (Echo/MR/Angio)-Akinesis, dyskinesis, aneurysm and RVEF < 40%

• 12 – ECG
  – Repolarization – TWI V1-V3, RBBB(-), >14 y/o
  – Depolarization – Epsilon

• Arrhythmia – VT/NSVT LBBB, Superior axis

• Biopsy – Residual myocyte < 60%, with fibrous replacement of RV+-/- fatty replacement

• Family Hx – 1° relative with Task Force Criteria or confirmed pathology

• Genetic testing – pathogenic mutation

Marcus FI et al. Eur Heart J. 2010
Catheter Ablation for ARVC

87 patients, 175 RFA procedure

49 patients, 23 endo; 26 endo+epi
Example of ARVC VT requiring epicardial ablation

Bipolar voltage map

Endocardium

0.5mV  Bipolar  1.5mV

LVZ=4.3%

Epicardium

LVZ=25%

Activation map

LAT 123  No LAVA

Lin CY et al. Europace 2017
Substrate of ARVC: Epicardial Scar Dominant
Example of ARVC VT: Endocardial Ablation Eliminate Epicardial Substrate

A Inducible Clinical VTs in the EP lab
B Epicardial VT isthmus
C Endocardial ablation eliminate epicardial LPs

D Bipolar Voltage Map
E Late Potential Map

Chung FP, Lin CY, Chen SA. KCJ 2018
Ablation Outcome - Endocardial Era in ARVC

N = 22

N = 24

Atul Verma et al. Circulation. 2005
Darshan Dalal et al. JACC 2007
Ablation Outcome—Epicardial Era

87 patients, 175 RFA procedure

49 patients, 23 endo; 26 endo+epi

Rong Bai et al. Circ AE. 2011;4:478-485
Survived SCD, Unstable VT, syncope

Stable, ns VT
Extensive disease
Early onset (<35 y/o)

Definite ARVC according to TFC
• No VT/extensive disease
• No syncope
• Irrespective of
  • Inducible VT during PVS
  • Family history

High risk
(8-10% /year)

Intermediate risk
(2% /year)

Low risk
(<1% /year)

Risk of arrhythogenic events

Risk in ARVC patients

Modified from Corrado et al. Heart 2011
Scar Distribution: Transmural Stable VT or PVC Dominant

Lin CY, Lin YJ, Chen SA. Europace 2018
Horizontal Epi Scar: VF Dominant

LVZ = 4.3%

LVZ = 65%

Epi

Endo

Bipolar Voltage Map

LAte Potential Map

HHT Map

Lin CY, Lin YJ, Chen SA. Europace 2018
Horizontal Scar: More VT/VF Recurrence

Log-Rank P value < 0.001

Lin CY, Lin YJ, Chen SA. Europace 2018
Male Gender and Late Potentials Predict Poor Outcome

Lin CY, Chung FP, Lin YJ, Chen SA. IJC 2017
Risk stratification by SAECG in ARVC patients

Patients fulfilled TF criteria ≥ 3 (N=24)  Patients fulfilled TF criteria ≤ 2 (N=40)

Liao YC, Chen SA et al. IJC 2014
SAECG in ARVC VT Recurrence

Log-rank test: P=0.019

Group 1: electrical regression
Group 2: no change
Group 3: electrical progression

VA recurrences free rate (%)

Duration (months)

Liao YC, Chen SA et al. IJC 2017
Overall Ablation Outcome in Taipei VGH

**First Procedure, N = 91**
- Recurrence (+) N = 35
- Recurrence (-) N = 56
  - Recurrence-free rate = 61.5%

**Second Procedure, N = 28**
- Recurrence (+) N = 11
- Recurrence (-) N = 17
  - Recurrence-free rate = 80.2%

**Third Procedure, N = 10**
- Recurrence (+) N = 4
- Recurrence (-) N = 6
  - Recurrence-free rate = 86.8%

**Fourth Procedure, N = 4**
- Recurrence-free rate = 90.1%

Lin CY, Chung FP et al. JCE 2019
Clinical Observation: An Endurance Athlete with ARVC VT

First Procedure

Bipolar Voltage

Unipolar Voltage

Bipolar Voltage

VF trigger origin and fast VT exit

Endocardial ablation → Inducibility (+) → Epicardial LAVA elimination → Non-inducible any VT/VF
Second Procedure

Second Procedure (3 months)

New VT isthmus and LAVA
Third Procedure (11 months)

New VT isthmus and LAVA
Fourth Procedure

Fourth ablation (23 months)

Scar Progression >> Incomplete Previous Ablation
Cause of Recurrence

Lin CY, Chung FP, Lin YJ, Chen SA et al. JCE 2019
Ablation Decrease AAD Usage in Long-Term FU

# ICD was Recommend in ARVC VT

## Recommendations for catheter ablation of VA in inherited primary arrhythmia disorders

<table>
<thead>
<tr>
<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
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<tbody>
<tr>
<td>I</td>
<td>B-NR</td>
<td>1. In patients with ARVC who experience recurrent sustained VT or frequent appropriate ICD interventions for VT in whom AAD therapy is ineffective or not tolerated, catheter ablation, at a center with specific expertise, is recommended.</td>
</tr>
<tr>
<td>I</td>
<td>B-NR</td>
<td>2. In patients with ARVC who have failed one or more attempts of endocardial VT catheter ablation, an epicardial approach for VT ablation is recommended.</td>
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<tr>
<td>IIa</td>
<td>B-NR</td>
<td>3. In patients with ARVC who experience recurrent sustained VT or frequent appropriate ICD interventions for VT in whom AAD therapy is not desired or preferred, catheter ablation, at a center with specific expertise, is reasonable.</td>
</tr>
<tr>
<td>IIa</td>
<td>B-NR</td>
<td>4. In patients with Brugada syndrome who experience recurrent sustained VAs or frequent appropriate ICD interventions, catheter ablation can be useful.</td>
</tr>
<tr>
<td>IIa</td>
<td>C-LD</td>
<td>5. In patients with ARVC, a first-line combined endocardial/epicardial approach for VT ablation is reasonable.</td>
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</tbody>
</table>
Outcomes of Catheter Ablation in Arrhythmogenic Right Ventricular Cardiomyopathy Without Background Implantable Cardioverter Defibrillator Therapy
A Multicenter International Ventricular Tachycardia Registry
Study Population

- Thirty-two patients with ARVC and VT underwent ablation without backup ICD.
  - Taipei VGH, Penn, TCAI, GGH
- Reason for absence of ICD
  - Patient refusal (63%)
  - Financial hardship (37%)

Santangeli P et al. JACC Clin Electrophysiol. 2019
ARVC without Backup ICD

Multicenter International Ventricular Tachycardia Registry: Taipei VGH, Penn, TCAI, CCH

Median follow-up of 46 months (range 26 to 65 months)
Recurrent VT episode: 19%
Mortality: none

Santangeli P et al. JACC Clin Electrophysiol. 2019
## Congenital Heart: Risk for VA/SCD

<table>
<thead>
<tr>
<th>Congenital Heart Disease</th>
<th>Incidence of VA</th>
<th>Incidence of SCD</th>
</tr>
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<tbody>
<tr>
<td>ASD</td>
<td>2-6%</td>
<td>&lt;1.5%</td>
</tr>
<tr>
<td>VSD</td>
<td>3-18%</td>
<td>&lt;3%</td>
</tr>
<tr>
<td>TOF</td>
<td>14-31%</td>
<td>1.4-8.3%</td>
</tr>
<tr>
<td>AS</td>
<td>10-34%</td>
<td>3-20%</td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>2%</td>
<td>2%</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>2%</td>
<td>3-6%</td>
</tr>
<tr>
<td>Atrial switch</td>
<td>2%</td>
<td>3-9.5%</td>
</tr>
<tr>
<td>Aterial switch</td>
<td>2%</td>
<td>1%</td>
</tr>
<tr>
<td>Cc-TGA</td>
<td>10%</td>
<td>17-25%</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>10%</td>
<td>4%</td>
</tr>
<tr>
<td>Fontan repair</td>
<td>5-17%</td>
<td>2.8-5.4%</td>
</tr>
</tbody>
</table>

2017 VA/SCD Guideline
Congenital Heart VT

CHD and Sustained Ventricular Tachycardia

Evaluate residual anatomical abnormalities, coronary arteries, ventricular function, exercise capacity

Residual abnormalities

Yes → Catheter or surgical intervention for correctable lesions; ± ablation ± ICD

No

Ablation

Yes

Recurrent ICD therapies

Yes → ICD implantation, heart failure consult

No → Polymorphic VT or single ventricle or severe CHD

ACA or severe ventricular dysfunction

Yes

High-risk CHD or recurrent VT

Yes

Simple or moderate CHD, TOF/DORV

2019 HRS/EHRA/APHRS/LAHRS Expert Consensus Statement on CA of VA
Residual VT Isthmus After OP

2019 HRS/EHRA/APHRS/LAHRS Expert Consensus Statement on CA of VA
Case Example

• 17 years-old female
• History :
  – Double outlet of right ventricle (DORV)
  – Malposition of great arteries
  – Patent ductus arteriosus (PDA)
  – Subpulmonary stenosis and subpulmonary ventricular septal defect (VSD)
• Operation History
  – Rastelli operation + PDA ligation + VSD repair on 95/7/10
  – VSD repair, RVOT reconstruction and PV replacement on 105/6/29
• Visits emergency room owing to nausea and vomiting on 105/8/5
• Syncope and several episodes of near fainting events
  ➔ ECG: hemodynamic stable and unstable VT s/p DC shock and CPR
• Frequent VT episode under the treatment of amiodarone
Operation Note
Bipolar Voltage

RAO 30    AP    LAO 60
Stump     Stump     Stump
Conduit   Conduit   Conduit
Unipolar Voltage

RAO 30
Conduit
Stump

AP
Conduit
Stump

LAO 60
Conduit
Stump
Sinus Rhythm Activation Map

RAO 90

Conduit

RAO 30

Stump

Conduit

LAO 60

Stump

Conduit
Central Isthmus: S-QRS/TCL: 33%;  
PPI-TCL < 30
VT 1

- Conduit
- Exit
- Central isthmus
- Dead end
- Proximal isthmus
- Entrance
- Tricuspid ring
VT 1 Propagation
Central Isthmus: S-QRS/TCL: 31%; PPI-TCL < 30
Take home message

• ARVC is a genetically inherited disease, characterized by progressive fibrofatty replacement
• Epicardial approach and ablation is an important strategy for ARVC VT.
• Scar distribution, baseline characteristics, SAECG were associated with outcome in ARVC
• Catheter ablation in selective ARVC patients without backup ICD was associated with a low rate of symptomatic VT recurrence without mortality during 46-month median follow-up.
• Type of CHD, previous surgical intervention, detailed pre-procedural image were important before catheter ablation