Genetic arrhythmic syndromes
mechanisms and indications for a
defibrillator and/or ablation

Venice, oct 17th 2015
NO CONFLICT OF INTEREST TO DECLARE
Amsterdam,
the Netherlands
December 4th 2015
For information and registration see www.20yrsCG.nl

www.20yrsCG.nl

Organising committee:
Karin Y. van Spaendonck
J. Peter van Tintelen
Arthur Wilde
Primary arrhythmia syndromes (2015)

- Long QT syndrome(s)
- Short QT syndrome
- Brugada syndrome
- Catecholamine-induced PMVT/VF
- Short-coupled Torsades de Pointes
- Isolated conduction disorders (AVN, BB)
- Early repolarization syndrome
- Sinus node disease, atrial standstill
- Idiopathic ventricular fibrillation
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Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes

Europace 2013, Heart Rhythm 2013, J of Arrhyth 2013
Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes

Silvia G. Priori, (HRS Chairperson)¹, Arthur A. Wilde, (EHRA Chairperson)², Minoru Horie, (APHRS Chairperson)³, Yongkeun Cho, (APHRS Chairperson)⁴, Elijah R. Behr⁵, Charles Berul⁶, Nico Blom⁷*, Josep Brugada⁸, Chern-En Chiang⁹, Heikki Huikuri¹⁰, Prince Kannankeril¹¹‡, Andrew Krahn¹², Antoine Leenhardt¹³, Arthur Moss¹⁴, Peter J. Schwartz¹⁵, Wataru Shimizu¹⁶, Gordon Tomaselli¹⁷†, Cynthia Tracy%¹⁸

Document Reviewers: Michael Ackerman (USA), Bernard Belhassen (Israel), N. A. Mark Estes III (USA), Diane Fatkin (Australia), Jonathan Kalman (Australia), Elizabeth Kaufman (USA), Paulus Kirchhof (UK and Germany), Eric Schulze-Bahr (Germany), Christian Wolpert (Germany), Jitendra Vohra (Australia), Marwan Refaat (USA), Susan P. Etheridge (USA), Robert M. Campbell (USA), Edward T. Martin (USA), Swee Chye Quek (Singapore)
Long QT Syndrome(s)

♥ Autosomal dominant/autosomal rec.
♥ genetically heterogeneous
♥ 16 genes (LQTS$_{1-16}$)
♥ $\geq 60\%$ genotyped ($\geq 90\%$ in families)
♥ gene-specific features
LQTS, risk stratification

Risk depends on:

- genotype
- phenotype
  - gender (young: male)
  - QTc ($\geq 500$ ms)
  - specific ECG features
Long QT syndrome, risk stratification

$\text{QT}_C$ Quartiles:
1: $\leq 446$ ms
2: 447 - 468 ms
3: 469 - 498 ms
4: $\geq 499$ ms

Cumulative Cardiac-Event-free Survival (%)

Age (yr)

<table>
<thead>
<tr>
<th>No. at Risk</th>
<th>1st quartile</th>
<th>2nd quartile</th>
<th>3rd quartile</th>
<th>4th quartile</th>
</tr>
</thead>
<tbody>
<tr>
<td>1st quartile</td>
<td>148</td>
<td>112</td>
<td>96</td>
<td>76</td>
</tr>
<tr>
<td>2nd quartile</td>
<td>150</td>
<td>104</td>
<td>80</td>
<td>62</td>
</tr>
<tr>
<td>3rd quartile</td>
<td>140</td>
<td>103</td>
<td>78</td>
<td>49</td>
</tr>
<tr>
<td>4th quartile</td>
<td>142</td>
<td>92</td>
<td>45</td>
<td>28</td>
</tr>
</tbody>
</table>

$P < 0.001$
Long QT syndrome

Who are the patients at risk?

♥ Aborted sudden death
♥ Syncope
♥ Patients with long QTc intervals (>500ms)
♥ Torsades de Pointes, T-wave alternans
♥ Specific mutations (compound mutations)
♥ congenital deafness (JLN)
<table>
<thead>
<tr>
<th>Class</th>
<th>ICD Recommendations</th>
</tr>
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<tr>
<td>Class I</td>
<td>ICD implantation <strong>is recommended</strong> for patients with a diagnosis of LQTS who are survivors of a cardiac arrest</td>
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<tr>
<td>Class IIa</td>
<td>ICD implantation <strong>can be useful</strong> in patients with a diagnosis of LQTS who experience recurrent syncopal events while on beta-blocker therapy.</td>
</tr>
<tr>
<td>Class III</td>
<td>Except under special circumstances, ICD implantation <strong>is not indicated</strong> in asymptomatic LQTS patients who have not been tried on beta-blocker therapy</td>
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</table>

Family history is **NOT** a risk factor
Long QT Syndrome – ICD Indications

Legend

- Class I
- Class IIa
- Class IIb
- Class III

Prior cardiac arrest?

Yes → ICD recommended

No

Recurrent syncope while on beta blocker?

Yes → ICD can be useful

Asymptomatic not treated with beta blockers

Yes → ICD is not indicated*

*Except under special circumstances, ICD implantation is not indicated in asymptomatic patients who have not been tried on beta-blocker therapy
When should an ICD be considered?

- JLNS patient with a long QTc (>500msec)
- LQT2 pt with QTc > 550
- LQT3 pt with QTc > 500
- Torsades de Pointes, T-wave alternans
- rarely LQT1!
- Family history of (a)SCD is not a riskfactor
Brugada syndrome

- Monogenetic disease? Oligogenetic!
- ≥18 genes involved
- Type 1 ECG (± drugs)
- documented VF or self terminating PMVT
- Family history of SCD < 45 y.
- 40 years of age, male
Male 39 years
Brugada Syndrome, risk stratification

Asymptomatic patients

- spontaneous variation: ++
- fragmented QRS: +
- Genotype (SCN5a or not): -
- ECG variables (HV-interval): -
- EPS inducibility: ± ?*

*: mild protocol
Brugada Syndrome, risk stratification

Symptomatic patients

❤️ documented arrhythmias/VF  ++
❤️ (presumed) arrhyth. syncope  ++
# Brugada Syndrome

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<td></td>
<td>• Are survivors of a cardiac arrest, and/or</td>
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<td>• Have documented spontaneous sustained VT with or without syncope.</td>
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<tr>
<td>Class IIa</td>
<td>ICD implantation can be useful in patients with a spontaneous diagnostic Type I ECG who have a history of syncope judged to be likely caused by ventricular arrhythmias.</td>
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<tr>
<td>Class IIb</td>
<td>ICD implantation may be considered in patients with a diagnosis of BrS who develop VF during programmed electrical stimulation (inducible patients).</td>
</tr>
<tr>
<td>Class III</td>
<td>ICD Implantation is <strong>not indicated</strong> in asymptomatic BrS patients with a drug induced type 1 ECG and on the basis of a family history of SCD alone.</td>
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# Brugada Syndrome

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Figure 2  Consensus recommendations for ICDs in patients diagnosed with Brugada syndrome.
Catecholamine-induced PMVT/VF

♥ Autosomal dominant
♥ genetic heterogeneous (5 genes, 1 locus)
♥ complaints during exercise, emotion, etc
♥ can start at young age
♥ baseline ECG = normal!
**Expert Consensus Recommendations on CPVT Therapeutic Interventions**

<table>
<thead>
<tr>
<th>Class</th>
<th>1. The following lifestyle changes are recommended in all patients with diagnosis of CPVT:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class I</td>
<td>a) Limit/ avoid competitive sports;</td>
</tr>
<tr>
<td></td>
<td>b) Limit/avoid strenuous exercise;</td>
</tr>
<tr>
<td></td>
<td>c) Limit exposure to stressful environments.</td>
</tr>
<tr>
<td></td>
<td>2. Beta-blockers are recommended in all symptomatic patients with a diagnosis of CPVT.</td>
</tr>
<tr>
<td></td>
<td>3. ICD implantation is recommended in patients with a diagnosis of CPVT who experience</td>
</tr>
<tr>
<td></td>
<td>cardiac arrest, recurrent syncope or polymorphic/ bidirectional VT despite optimal</td>
</tr>
<tr>
<td></td>
<td>medical management, and/or LCSD.</td>
</tr>
<tr>
<td>Class IIa</td>
<td>4. Flecainide can be a useful addition to beta-blockers in patients with a diagnosis of</td>
</tr>
<tr>
<td></td>
<td>CPVT who experience recurrent syncope or polymorphic/ bidirectional VT while on beta-</td>
</tr>
<tr>
<td></td>
<td>blockers.</td>
</tr>
<tr>
<td></td>
<td>5. Beta-blockers can be useful in carriers of a pathogenic CPVT mutation without clinical</td>
</tr>
<tr>
<td></td>
<td>manifestations of CPVT (concealed mutation-positive patients).</td>
</tr>
<tr>
<td>Class IIb</td>
<td>6. LCSD may be considered in patients with a diagnosis of CPVT who experience recurrent</td>
</tr>
<tr>
<td></td>
<td>syncope or polymorphic/bidirectional VT/ several appropriate ICD shocks while on beta-</td>
</tr>
<tr>
<td></td>
<td>blockers and in patients who are intolerant or with contraindication to beta-blockers.</td>
</tr>
<tr>
<td>Class III</td>
<td>7. ICD as a standalone therapy is not indicated in an asymptomatic patient with a diagnosis</td>
</tr>
<tr>
<td></td>
<td>of CPVT.</td>
</tr>
<tr>
<td></td>
<td>8. Programmed Electrical Stimulation is not indicated in CPVT patients.</td>
</tr>
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Sudden Death in a Young Man with Catecholaminergic Polymorphic Ventricular Tachycardia and Paroxysmal Atrial Fibrillation

STEPHEN PIZZALE, B.H.Sc., B.S.C.N.,* MICHAEL H. GOLLIB, M.D.,† ROBERT GOW, M.D.,† and DAVID H. BERNIE, M.B., C.H.B., M.D.,†

From the "University of Ottawa Heart Institute, Ottawa, Ontario, Canada and Children's Hospital of Eastern Ontario, Ottawa, Ontario, Canada

Sudden Death in Patient with CPVT and PAF. Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a familial condition that presents with exercise-induced syncope or sudden death in children or young adults. In most cases the disease is caused by a mutation in the cardiac ryanodine receptor (RyR2) gene. Current evidence suggests that primary therapy for CPVT is beta blockade and implantable cardioverter defibrillator (ICD) placement. There is a recent report of a patient with CPVT who died despite appropriate ICD therapies, and we report a similar case. Our patient died after probably initially receiving inappropriate ICD shocks for atrial fibrillation. We recommend that utmost efforts should be made to prevent shocks including repeated exercise testing to confirm suppression of PVT. (J Cardiovasc Electrophysiol, Vol. 19, pp. 1319-1321, December 2008)

This starts with not implanting an ICD!!
**Expert Consensus Recommendations on IVF Therapeutic Interventions**

**Class I**
1. ICD implantation *is recommended* in patients with a diagnosis of IVF.

**Class IIb**
2. Antiarrhythmic therapy with quinidine, programmed electrical stimulation guided or empirical, *may be considered* in patients with a diagnosis of IVF in conjunction with ICD implantation or when ICD implantation is contraindicated or refused.

3. Ablation of Purkinje potentials *may be considered* in patients with a diagnosis of IVF presenting with uniform morphology premature ventricular contractions in conjunction with ICD implantation or when ICD implantation is contraindicated or refused.

4. If a first-degree relative of an IVF victim presents with unexplained syncope and no identifiable phenotype following thorough investigation, then after careful counseling an ICD implant *may be considered.*
Primary arrhythmia syndromes (2015)

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- Isolated conduction disorders (AVN, BB)
- Atrial fibrillation
- Sinus node disease, atrial standstill
- Idiopathic ventricular fibrillation

Ablation?
# Primary arrhythmia syndromes (2015)

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Ablation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Long QT syndrome(s)</td>
<td>±</td>
</tr>
<tr>
<td>Short QT syndrome</td>
<td>-</td>
</tr>
<tr>
<td>Brugada syndrome</td>
<td>+</td>
</tr>
<tr>
<td>Catecholamine-induced PMVT/VF</td>
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<td>Idiopathic ventricular fibrillation</td>
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Mapping and Ablation of Ventricular Fibrillation Associated With Long-QT and Brugada Syndromes

Michel Haïssaguerre, MD; Fabrice Extramiana, MD; Mélèze Hocini, MD; Bruno Cauchemez, MD; Pierre Jaïs, MD; Jose Angel Cabrera, MD; Geronimo Farre, MD; Antoine Leenhardt, MD; Prashanthan Sanders, MBBS; Christophe Scavée, MD; Li-Fern Hsu, MBBS; Rukshen Weerasooriya, MBBS; Dipen C. Shah, MD; Robert Frank, MD; Philippe Maury, MD; Marc Delay, MD; Stéphane Garrigue, MD; Jacques Clémenty, MD

Background—The long-QT and Brugada syndromes are important substrates of malignant ventricular arrhythmia. The feasibility of mapping and ablation of ventricular arrhythmias in these conditions has not been reported.

Methods and Results—Seven patients (4 men; age, 38±7 years; 4 with long-QT and 3 with Brugada syndrome) with episodes of ventricular fibrillation or polymorphic ventricular tachycardia and frequent isolated or repetitive premature beats were studied. These premature beats were observed to trigger ventricular arrhythmias and were localized by mapping the earliest endocardial activity. In 4 patients, premature beats originated from the peripheral right (1 Brugada) or left (3 long-QT) Purkinje conducting system and were associated with variable Purkinje-to-muscle conduction times (30 to 110 ms). In the remaining 3 patients, premature beats originated from the right ventricular outflow tract, being 25 to 40 ms ahead of the QRS. The accuracy of mapping was confirmed by acute elimination of premature beats after 12±6 minutes of radiofrequency applications. During a follow-up of 17±17 months using ambulatory monitoring and defibrillator memory interrogation, no patients had recurrence of symptomatic ventricular arrhythmia but 1 had persistent premature beats.

Conclusion—Triggers from the Purkinje arborization or the right ventricular outflow tract have a crucial role in initiating ventricular fibrillation associated with the long-QT and Brugada syndromes. These can be eliminated by focal radiofrequency ablation. *(Circulation. 2003;108:925-928.)*
Long QT syndrome

Ablation experience

♥ 4 patients
♥ 1 patient RVOT ectopy,
♥ 1 patient post fascicle, 2 P-fiber activity LV
♥ ectopy as the target
♥ FU 17+7 months: arrhythmia free
Brugada Syndrome, ablation

Methods
❤ Ectopy as the target
❤ Substrate as the target
## Brugada Syndrome

<table>
<thead>
<tr>
<th>Class</th>
<th>Catheter Ablation Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class IIb</td>
<td>Catheter ablation <em>may be considered</em> in patients with a diagnosis of BrS and history of arrhythmic storms or repeated appropriate ICD shocks.</td>
</tr>
</tbody>
</table>
Prevention of Ventricular Fibrillation Episodes in Brugada Syndrome by Catheter Ablation Over the Anterior Right Ventricular Outflow Tract Epicardium

Koonlawee Nademanee, Gumpanart Veerakul, Pakorn Chandanamattha, Lertlak Chaothawee, Aekarach Ariyachaipanich, Kriengkrai Jirasirirojanakorn, Khanchit Likittanasombat, Kiertijai Bhuripanyo and Tachapong Ngarmukos

Circulation published online Mar 14, 2011;

Circulation. 2011;12:1270-9
Before ablation

1 month post ablation
Only at the epicardium of the RVOT area (anterior) one finds

- late to very late potentials
- low voltage signals
Insight into the mechanism of Brugada syndrome: Epicardial substrate and modification during ajmaline testing

Frédéric Sacher, MD, Laurence Jesel, MD, Pierre Jais, MD, Michel Haïssaguerre, MD

From the Bordeaux University Hospital and LIRYC, L’Institut de rythmologie et modélisation cardiaque, Université de Bordeaux, Bordeaux-Pessac, France.
Pre-Ajm-Area = 11.1 cm$^2$
Post Ajm-area = 27 cm²
Brugada Syndrome Phenotype Elimination by Epicardial Substrate Ablation

Running title: Brugada et al.; Brugada Syndrome Phenotype Elimination

Josep Brugada, MD\(^1\)*; Carlo Pappone, MD, PhD\(^2\)*; Antonio Berruezo, MD, PhD\(^1\);
Gabriele Vicedomini, MD\(^2\); Francesco Manguso, MD, PhD\(^2\); Giuseppe Ciconte, MD\(^2\);
Luigi Giannelli, MD\(^2\); Vincenzo Santinelli, MD\(^2\)

\(^1\)Arrhythmia Section, Cardiology Department, Thorax Institute, Hospital Clinic and IDIBAPS (Institut d’Investigació Agustí Pi i Sunyer), Barcelona, Catalonia, Spain; \(^2\)Arrhythmology Departments, Maria Cecilia Hospital, Cotignola and Policlinico San Donato, University of Milan, Milan, Italy

*contributed equally as first authors
Methods

♥ Ectopy as the target
One symptomatic RyR2+ patient, only betablocker, VES targeted approach
VPC #1: LV inferior septum origin

Pacemap:

- I
- II
- III
- aVR

VPC:

- II
- V2
- His

SR:

- Bi.

Additional notes:

- Purkinje potential
- "QS"

Time intervals:

- 300ms
- 200ms

Electrocardiograms for each lead are shown.
IVF, short coupled TdP, ablation

Methods

♥ Ectopy as the target
Mapping and Ablation of Idiopathic Ventricular Fibrillation

Michel Haïssaguerre, MD; Morio Shoda, MD; Pierre Jaïs, MD; Akihiko Nogami, MD; Dipen C. Shah, MD; Josef Kautzner, MD; Thomas Arentz, MD; Dietrich Kalushe, MD; Dominique Lamaison, MD; Mike Griffith, MD; Fernando Cruz, MD; Angelo de Paola, MD; Fiorenzo Gaïta, MD; Mélèze Hocini, MD; Stéphane Garrigue, MD; Laurent Macle, MD; Rukshen Weerasooriya, MD; Jacques Clémenty, MD

Background—Ventricular fibrillation is the main mechanism of sudden cardiac death. The feasibility of eliminating recurrent episodes by catheter ablation has not been reported.

Methods and Results—Twenty-seven patients without known heart disease (13 men, 14 women, 41±14 years of age) were studied after being resuscitated from recurrent (10±12) episodes of primary idiopathic ventricular fibrillation; 23 had received a defibrillator. The first initiating beat of ventricular fibrillation had an identical electrocardiographic morphology and coupling interval (297±41 ms) to preceding isolated premature beats typically noted in the aftermath of resuscitation. These triggers were localized by mapping the earliest electrical activity and ablated by local radiofrequency delivery. Outcome was assessed by Holter and defibrillator memory interrogation. Premature beats were elicited from the Purkinje conducting system in 23 patients: from the left ventricular septum in 10, from the anterior right ventricle in 9, and from both in 4. The interval from the Purkinje potential to the following myocardial activation varied from 10 to 150 ms during premature beat but was 11±5 ms during sinus rhythm, indicating location at peripheral Purkinje arborization. The premature beats originated from the right ventricular outflow tract muscle in 4 patients. The accuracy of mapping was confirmed by acute elimination of premature beats during local radiofrequency delivery. During a follow-up of 24±28 months, 24 patients (89%) had no recurrence of ventricular fibrillation without drug.

Conclusions—Primary idiopathic ventricular fibrillation is a syndrome characterized by dominant triggers from the distal Purkinje system. These sources can be eliminated by focal energy delivery. (Circulation. 2002;106:962-967.)

Key Words: ablation ■ death, sudden ■ heart arrest ■ fibrillation ■ mapping
Methods and Results

- 23/27 pts ectopy from LV/RV Purkinje
- 4 pts from the RVOT area
- Ectopy targeted ablation
- FU 24±28 mths, drug free
- 89% arrhythmia free survival
Electrocardiographic morphology of premature beats

Long-Term Follow-Up of Idiopathic Ventricular Fibrillation Ablation
A Multicenter Study

Sébastien Knecht, MD,*‡‡ Frédéric Sacher, MD,* Matthew Wright, MBBS, PhD,*
Mélèze Hocini, MD,* Akihiko Nogami, MD,† Thomas Arentz, MD,‡ Bertrand Petit, MD,§
Robert Franck, MD,‖ Christian De Chillou, MD,¶ Dominique Lamaison, MD,# Jéronimo Farré, MD,**
Thomas Lavernge, MD,†† Thierry Verbeet, MD,‡‡ Isabelle Nault, MD,* Seiichiro Matsuo, MD,*
Lionel Leroux, MD,* Rukshen Weerasooriya, MD,* Bruno Cauchemez, MD, §§
Nicolas Lellouche, MD,* Nicolas Derval, MD,* Sanjiv M. Narayan, MD, PhD,* Pierre Jaïs, MD,*
Jacques Clementy, MD,* Michel Haïssaguerre, MD*

Bordeaux, Saint Pierre, Paris, Nancy, and Clermond-Ferrand, France; Yokohama, Japan; Bad Krozingen,
Germany; Madrid, Spain; and Brussels, Belgium
IVF, short coupled TdP, ablation

Methods and Results

- 38 pts, multicentre study
- ectopy from LV (16) / RV (14) Purkinje
- ectopy targeted ablation
- FU median 63 months
- 7/18 (18%), recurrent VF after 4 mnths
- 5/7 arrhythmia free after re-ablation.
ICD’s and ablation options
♥ are very much disease dependent
♥ ICD’s should be carefully chosen
♥ Ablation options are available
♥ in particular in IVF, BrS
Thank you
1. BrS *is diagnosed* in patients with ST segment elevation with type 1 morphology ≥ 2 mm in ≥ 1 lead among the right precordial leads V1, V2, positioned in the 2nd, 3rd or 4th intercostal space occurring either spontaneously *or* after provocative drug test with intravenous administration of Class I antiarrhythmic drugs.

2. BrS *is diagnosed* in patients with type 2 or type 3 ST segment elevation in ≥ 1 lead among the right precordial leads V1, V2 positioned in the 2nd, 3rd or 4th intercostal space when a provocative drug test with intravenous administration of Class I antiarrhythmic drugs induces a type 1 ECG morphology.
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| Class IIb | 2. ICD implantation **may be considered** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD.  
|         | 3. Quinidine **may be considered** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD.  
|         | 4. Sotalol **may be considered** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD |
Indications for ICDs in Patients Diagnosed with Short QT

Legend
- Class I
- Class IIa
- Class IIb
- Class III

1. Prior cardiac arrest or sustained VT?
   - Yes → ICD recommended
   - No → Asymptomatic Patient with a family history of SCD?
   - Yes → ICD may be considered