Data that may change the way we practice...

- Genetic diseases
  - Sudden death in Hypertrophic Cardiomyopathy
  - Management of Brugada Syndrome

- Devices
  - New Pacing Technology
  - Wearable defibrillator
  - Lead extraction
Genetic Diseases
2011 ACCF/AHA Guidelines on Hypertrophic Cardiomyopathy

Current scheme for risk stratification for Sudden Cardiac Death in HCM

- Prior cardiac arrest or Sustained VT
  - Yes → ICD recommended Class I, LoE B
  - No → Family history-SD in first degree relative or LV wall thickness ≥ 30 mm or Recent unexplained syncope
    - Yes → ICD reasonable
    - No → Nonsustained VT or Abnormal BP response
      - Yes → Other SCD Risk Modifiers* Present
        - Yes → ICD can be usefull
        - No → ICD not recommended
      - No → ICD not recommended

Legend:
- Class I
- Class IIa
- Class IIb
- Class III
A novel clinical risk prediction model for sudden cardiac death in hypertrophic cardiomyopathy

Parameters included in the model:

- Age
- Family history of SCD
- Left atrial diameter
- Maximal left ventricular (LV) wall thickness
- Maximal LV outflow tract gradient
- Nonsustained ventricular tachycardia
- Unexplained syncope

O’ Mahony C et al. Eur Heart J 2013
HCM Risk-SCD Calculator

| Age | 54 Years | Age at evaluation |
|-----------------------------------------|------------------|
| Maximum LV wall thickness | 23 mm | Transthoracic Echocardiographic measurement |
| Left atrial size | 42 mm | Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation |
| Max LVOT gradient | 55 mmHg | The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernouilli equation: Gradient= 4V², where V is the peak aortic outflow velocity |
| Family History of SCD | No | History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis) |
| Non-sustained VT | No | 3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation |
| Unexplained syncope | No | History of unexplained syncope at or prior to evaluation |

Risk of SCD at 5 years (%): 2.34

ESC recommendation: ICD generally not indicated **

** ICD not recommended unless there other clinical features that are of potential prognostic importance and when the likely benefit is greater than the lifelong risk of complications and the impact of an ICD on lifestyle, socioeconomic status and psychological health.
New recommendations / ESC guidelines 2014

TAKE HOME MESSAGES

- **Class IIa**  ICD should be considered in patients with an estimated 5-year risk of SCD ≥ 6%

- **Class IIb**  ICD may be considered in patients with an estimated 5-year risk of SCD ≥ 4% and < 6%

- **Class IIb**  ICD is generally not advised in patients with an estimated 5-year risk of SCD < 4%
Management of Brugada Syndrome
Objectives and Study population

- Assess the efficacy of quinidine in reducing ICD shocks in Brugada Syndrome.

Number of patients → 23
Mean age (years) → 41 ± 13
Male gender → 19 (82%)
Secondary prev ICD → 20 (87%)
Mean Follow up → 47 ± 43 mo
Reduction in the median number of ICD shocks before and after Quinidine administration

Until now quinidine is only indicated for arrhythmic storms in Brugada Syndrome.

- 5 (22%) patients with at least one shock
- 18 (78%) patients without shocks

82%
Prospective data on patients with long follow up suggest efficacy of quinidine in reducing shocks in highly symptomatic patients.
New Technologies
Pacemakers rely on an internal battery which requires surgical replacement.

Batteries account for more than half of a pacemaker’s volume.

Pacemaker leads are prone to fracture and pose a risk to the patient.
A batteryless cardiac pacemaker powered by cardiac motion

- Batteryless
  - A clockwork mechanism converts the motion of the beating heart into electrical energy
  - In-vivo experiments showed that the device is able to generate 52 μW
  - As comparison, today’s modern pacemakers require about 10 μW on average

A. Zurbuchen, (CH) / 1268
The contraction of the heart may be used as an energy source.

This new device converted enough energy from the heart motion to sustain the pacemaker.

Situated directly on the heart, the system is the first batteryless and leadless pacemaker.
Can be used to bridge a decision for appropriate ICD implantation in:

- Post-MI patients
- Following coronary revascularization
- New onset non-ischaemic DCM
- Patients with high risk for SCD
- Inherited arrhythmic or congenital disorders

Provide prospective data on the safety and efficacy of a bridging strategy with the WCD in a real world setting
WEARIT-II: Study population

- N=2000 patients enrolled
- Ischaemic cardiomyopathy → 805 pts, 40.3%
- Non-ischaemic CMP → 927 pts, 46.4%
- Congenital/inherited → 268 pts, 13.4%
- Previous cardiac arrest: 8%
- Mean wearing time: 3 months (22.5 hrs/day)
WEARIT-II: Outcomes

- Low rate of inappropriate shocks (0.5%)
- Detection of VT/VF 85%
- No death

<table>
<thead>
<tr>
<th>Description</th>
<th>% Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>VTVF_trt: treated VT or VF</td>
<td>85%</td>
</tr>
<tr>
<td>SustVT_notrt: VT's that</td>
<td>65%</td>
</tr>
<tr>
<td>spontaneously terminated</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td>Other: atrial arrhythmias or SVT</td>
<td>48%</td>
</tr>
<tr>
<td>No Arrhythmia</td>
<td>39%</td>
</tr>
</tbody>
</table>

VTVF_trt: treated VT or VF, sustVT notrt: VT's that spontaneously terminated during response button use or extended detection time, other: atrial arrhythmias or SVT
WEARIT-II

TAKE HOME MESSAGE

➔ In a “real world” setting the wearable defibrillator can be safely used to bridge a decision for appropriate ICD therapy
Enrolling all consecutive patients with indications for transvenous lead extraction

Primary objective:
To evaluate the acute & long-term safety of lead extraction
**ELECTRa: Outcomes**

**Clinical Success rate (%)**
- 98.3%
- 1.7%

**Death (%)**
- All centres: 1.4%
- High volume centres: 1.2%
- Low volume centres: 2.5%

**TAKE HOME MESSAGE**

The first prospective registry on outcome of patients after lead extraction suggests that the procedure is effective and safe.
The availability of large registries for rare diseases is greatly contributing to the identification of novel risk stratification markers (HCM) and new therapies (quinidine in Brugada Syndrome).

Technology is contributing to advance arrhythmias management leading to novel devices such as leadless and batteryless pacemakers.

We are moving toward a novel vision for ICD implant using the wearable ICD as a bridge-therapy.

We can now consider transvenous lead replacement as a safe procedure.