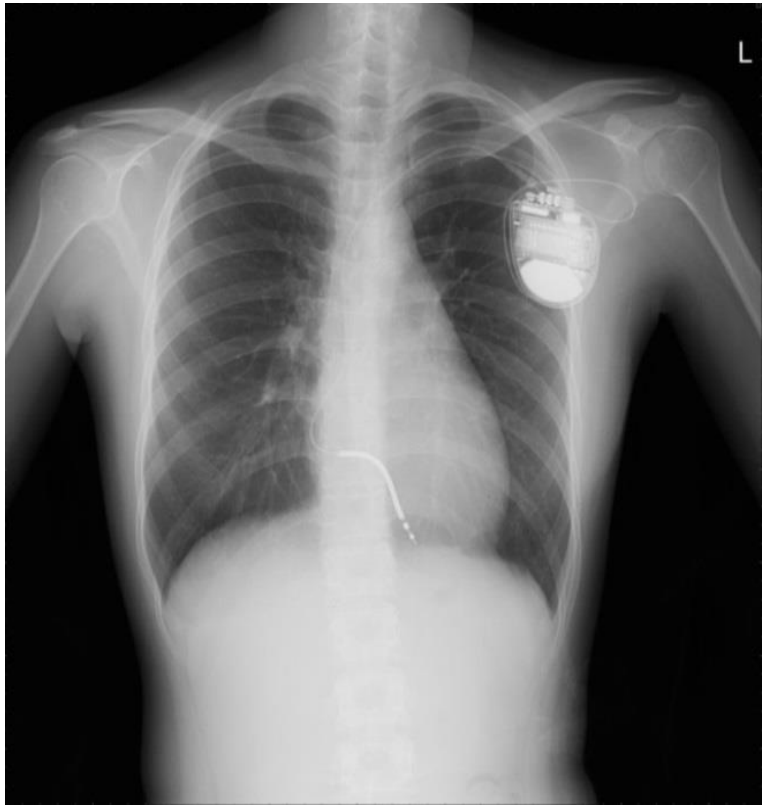
CIED in Inherited Arrhythmia

Kyungpook National University, Korea
Yongkeun Cho

Disclosure

- None.

ICD implantation in a young lady with LQTS



- Inappropriate shocks
- Lead-related complications
- Vascular occlusion or stenosis
- Device replacement
- Infection
- Psychological adjustment
- Cosmetic problem
- Social discrimination

Rush to ICD placement in patients with LQTS

- Forty-one percent of European LQTS ICD registry patients received an ICD without adequate trials of pharmacologic therapy. The majority of asymptomatic patients implanted with an ICD were those with LQT3 (Schwartz et al, Circulation 2010;122:1272-82).

- Half of LQT3 patients with ICD experienced inappropriate shocks or ICD-related complications. Two patients with frequent appropriate ICD shocks showed immediate improvement after mexiletine loading (Blaufox et al, Am J Cardiol 2012;109:1459-65).

- There is a premature rush to ICD placement. Seven percent of ICD recipients had their ICD subsequently explanted (Gaba et al, Heart Rhythm 2016;13:879-85).

European Heart Journal (2013) **34**, 3109–3116
doi:10.1093/eurheartj/eht089

REVIEW

Clinical update

The long QT syndrome: a transatlantic clinical approach to diagnosis and therapy

Peter J. Schwartz[1,2,3,4,5]* and Michael J. Ackerman[6,7,8]

Currently, there is a stunning mismatch in ICD utilization for patients with LQTS, with some programmes in the USA implanting an ICD in ~80% of their patients with LQTS, while among two of the largest LQTS Clinics in the world our ICD utilization rate is ~3% (P.J.S.) and 15% (M.J.A.).

We have many therapeutic options

Lifestyle modification

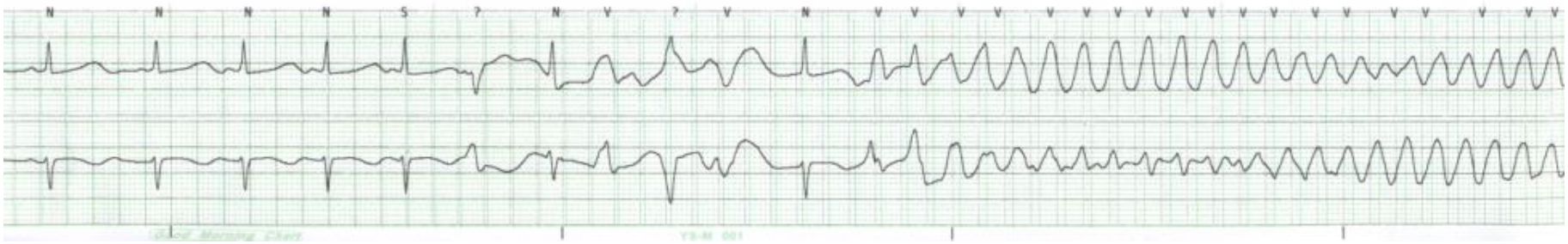
Sympathectomy

B-blocker

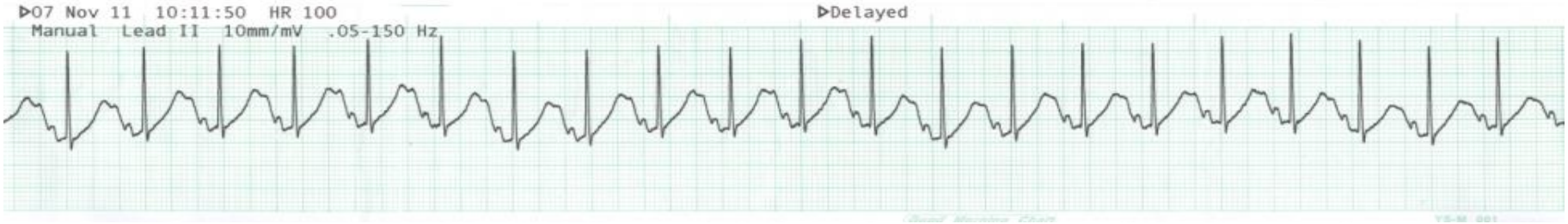
ICD

Effects of sympathectomy in another young lady with LQT2

- Before sympathectomy



- After sympathectomy



Clinical outcome after ICD implantation in patients with Brugada syndrome

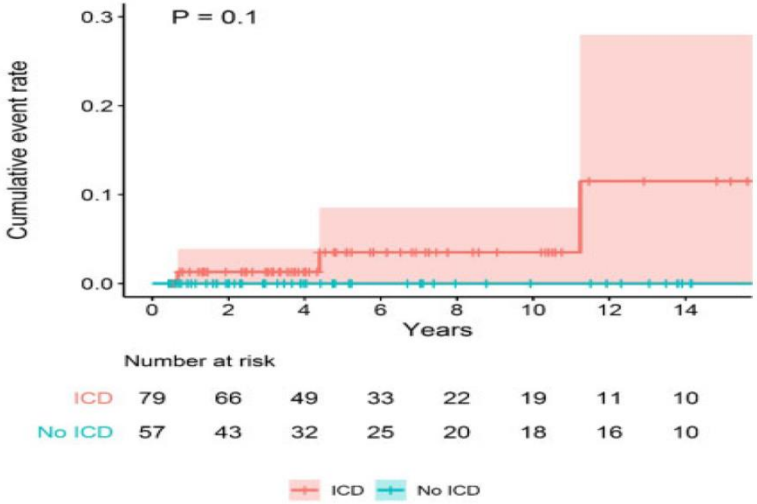
Dereci et al, JACC EP 2019;5:141–8

- Meta-analysis of 22 studies. 1,539 patients (45 years, 18% women) underwent ICD implantation for primary (79%) or secondary (21%) prevention of SCD.
- Most of the studies used a conventional TV single chamber ICD. The appropriate and inappropriate ICD intervention rates were similar as 3.1 and 3.3/100 person-years, respectively.
- ICD-related complications/100 person-years were lead malfunction (1.6), psychological complication (1.3), infection (0.6), and lead dislocation (0.4).
- Patients with Brugada syndrome at high risk for ventricular arrhythmia may significantly benefit from ICD therapy. Inappropriate ICD interventions and ICD-related complications may lead to considerable morbidity.

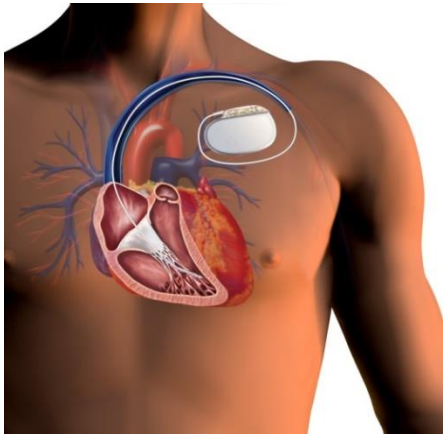
Clinical outcome after ICD implantation in patients with CPVT

- ICD use in CPVT: review (Roston et al, Heart Rhythm 2018;15:1791-9)
  - 53 studies, 503 patients with an ICD.
  - 40% had ≥1 appropriate shock and 21% had ≥1 inappropriate shock.
  - 20% had electrical storm.
  - An ICD-associated electrical storm was implicated in 4/7 deaths.
  - Additional complications such as lead failure, endocarditis, or surgical revisions in 96 of 296 patients (32%).

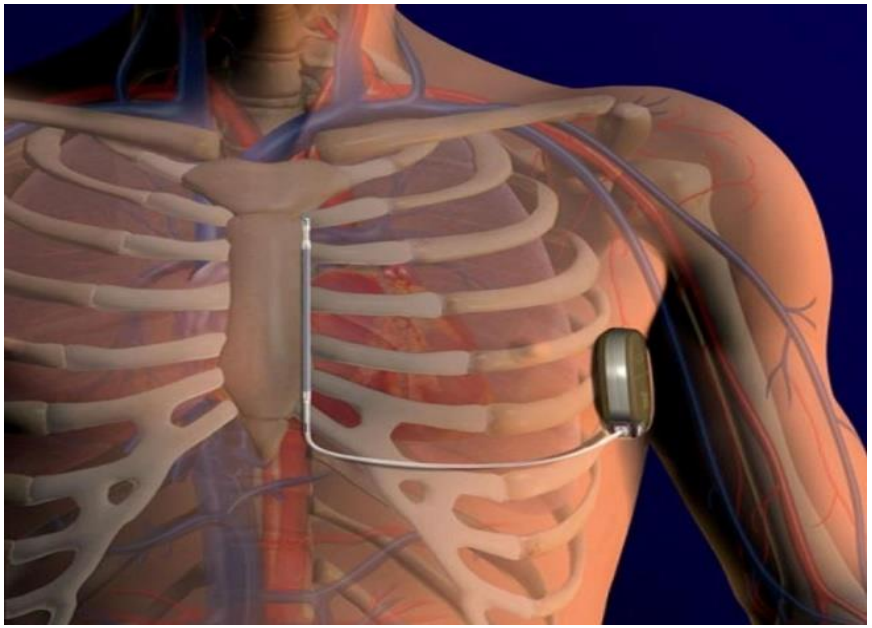
- ICDs in patients with CPVT resuscitated from SCA (van der Werf et al, Eur Heart J 2019 Epub ahead of print)
  - 136 patients with SCA, 79 patients with an ICD.
  - SCD, SCA, or appropriate ICD shocks occurred in 47% of patients with an ICD and 16% of patients without an ICD (P <0.0001).
  - The ICD did not confer a survival benefit but only ICD-associated co-morbidities including device-attributable death.



Transvenous ICD ⇒ **Subcutaneous** ICD

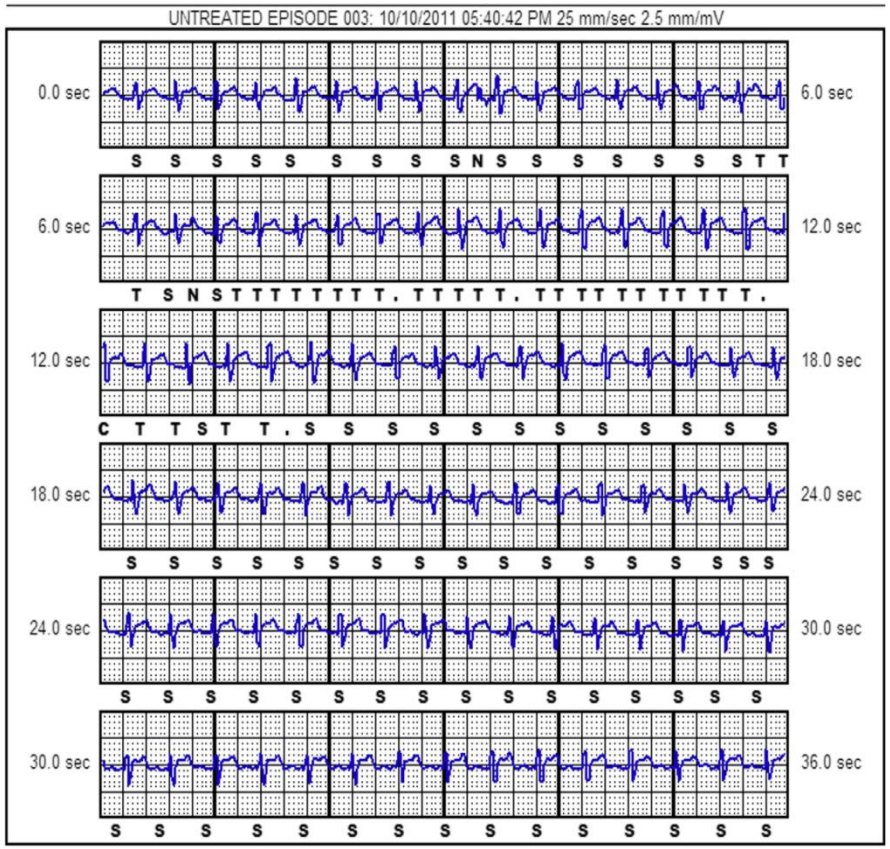


⇒

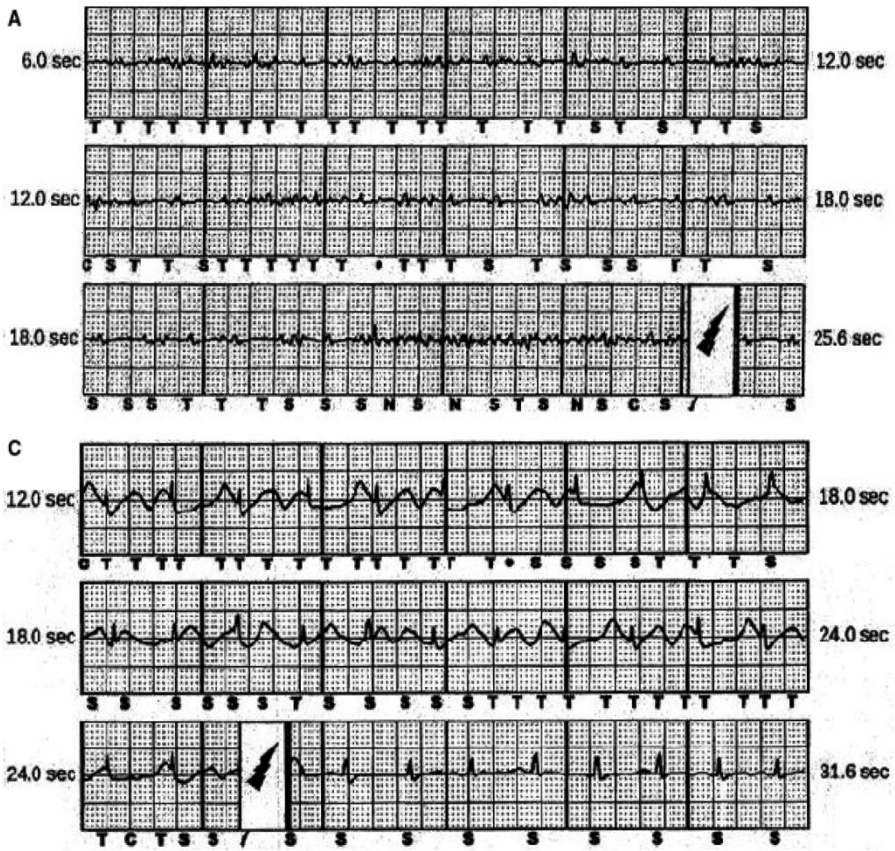


- Ideal candidates
  - Young/have a long life expectancy
  - prone to high-rate ventricular tachyarrhythmias such as VF
- Up to 30% of patients are ineligible for subcutaneous ICD (Campbell et al, Heart Rhythm 2018;15:703-7).
- Inappropriate shocks mostly due to T-wave oversensing (Kooiman et al, Heart Rhythm 2014;11:426-34).

Examples of oversensing in patients with S-ICD

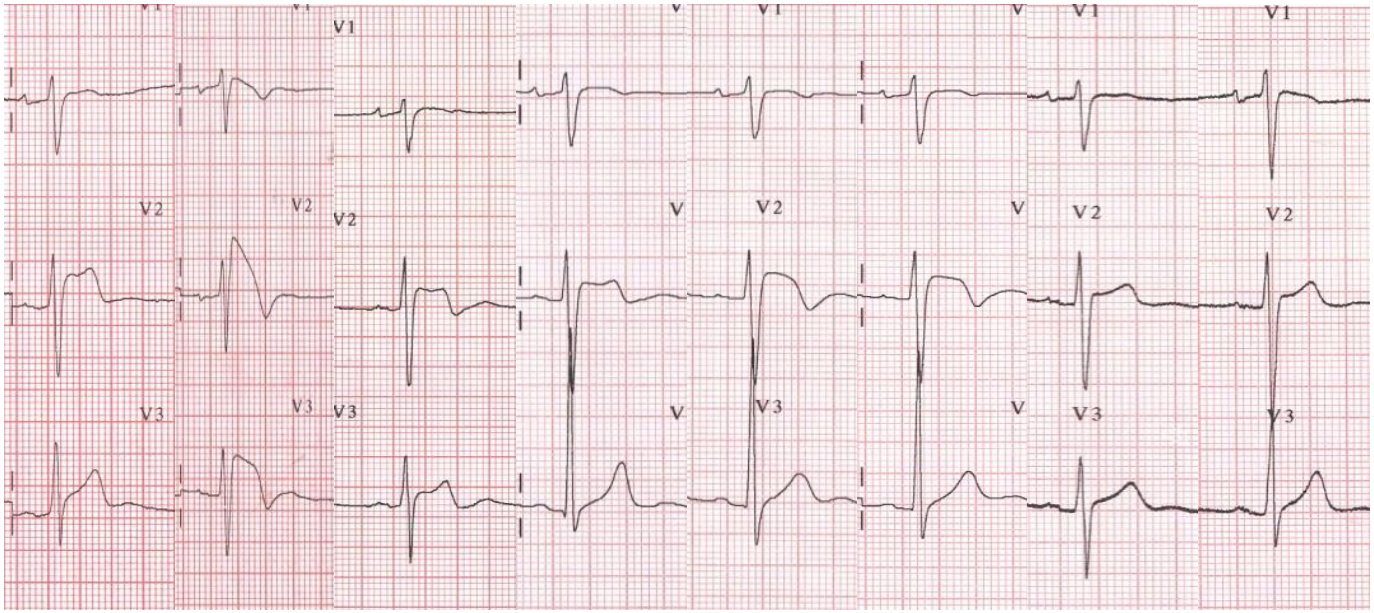


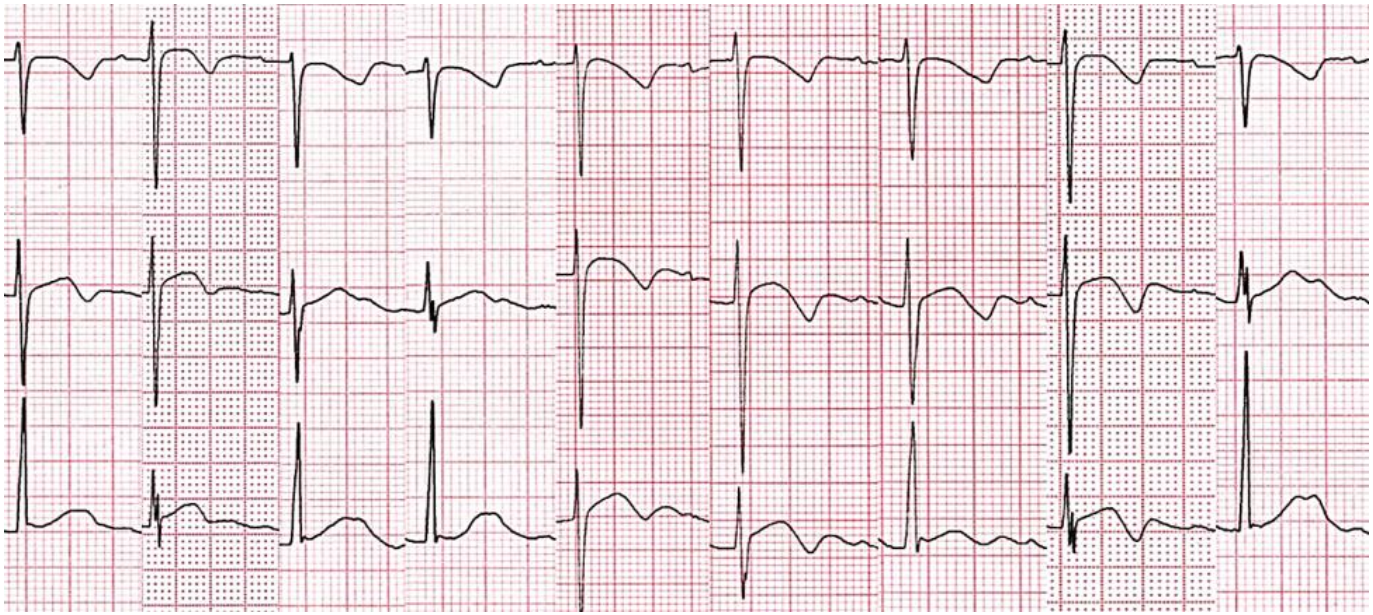
- Example of T-wave oversensing (T) (Kooiman et al, Heart Rhythm 2014;11:426–34).



- Surface ECG tracing of myopotentials (A), and movement artifact (C) (Orgeron et al, J Am Heart Assoc. 2018;7:e008782. DOI:10.1161/JAHA.118.008782.)

T-wave is changing continuously in patients with BrS & LQTS





S-ICD in patients with Brugada syndrome

- The BrS group had a significantly higher prevalence of S-ICD screening failure as compared with the non-BrS group (30% vs. 8%; P=0.003) (Kawabata, Cic J 2017;81:981-7).

- Higher rate of screening failure as compared with other channelopathies (18% vs. 5%, P = 0.07). All of patients with drug-induced BrS had appropriate morphology analysis at baseline but in 15%, S-ICD screening became inappropriate after ajmaline administration (Conte et al, Europace 2018;20:1188-93).

- Typical Brugada pattern is dynamic. Eligibility should be determined with an exercise test/drug challenge, and serial screening (Bögeholz et al, Europace 2019 in press).

ICD in patients with HCM

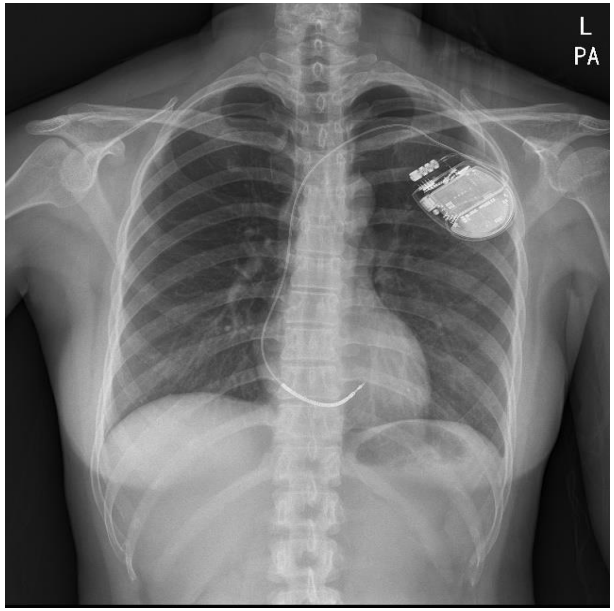
Bögeholz et al, Europace 2019 in press

- TV-ICD
  - High-risk patients may exhibit sustained monomorphic VT
  - Elderly (>65 years) patients with outflow obstruction may benefit from dual chamber pacing
  - Defibrillation threshold is increased in some patients
- S-ICD
  - Ineligibility rate from 7 - 38%
  - High R-wave/T-wave ratio and T-wave inversion is predictor of screening failure
  - Additional right parasternal screening/exercise testing should be considered

TV-ICD and S-ICD in ARVC

TV-ICD

- 4% required an additional septal lead during mean FU of 3 and 5 years, respectively (Corrado et al, Circulation 2003;108:3084-91, Bhonsale et al, JACC 2011;58:1485-96).
- ATP successfully terminated most (92%) VT episodes (Link et al, JACC 2014;64:119–25).



S-ICD

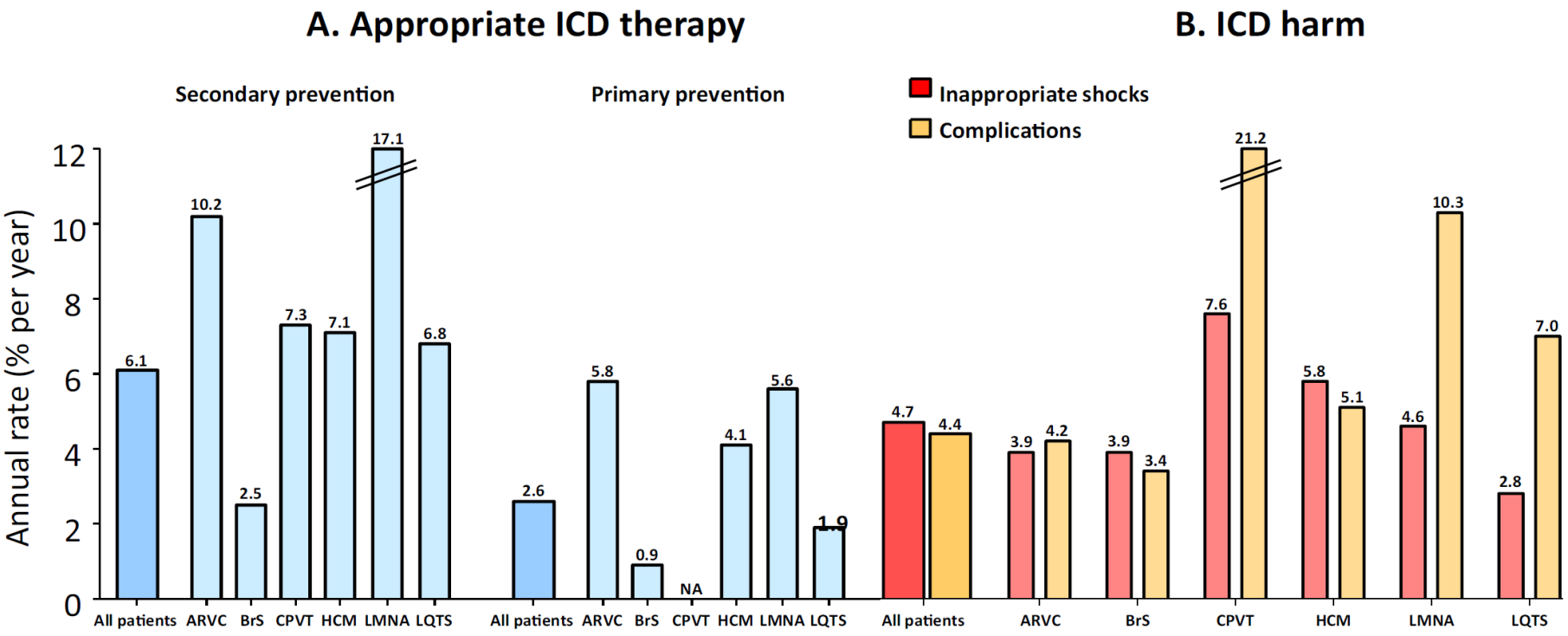
- All sustained VAs were appropriately detected /treated. 6 patients (21%) had 39 inappropriate shocks, with 3 requiring device explantation. No patient had the device removed due to the need for ATP (Orgeron et al, J Am Heart Assoc. 2018;7:e008782).
- All sustained VAs were appropriately detected/treated. 6 patients (14%) had 8 inappropriate shocks, with 1 requiring device explantation. No patients had the device explanted due to the need for ATP (Migliore et al, Int J Cardiol 2019;280:74-9).

- S-ICD for young (VF) and TV-ICD for old (VT) ?

ICD in young patients with inherited arrhythmia syndromes

Olde Nordkamp et al, Heart Rhythm 2016;13:443-54

- Mata-analysis of 63 studies, 4,916 patients (39±15 years old).
- ARVC 14%, BrS 21%, CPVT 0.6%, HCM 50%, LMNA 3.3%, LQTS 9.4%, and SQTS 1.0%.
- Inappropriate shocks occurred in 20% (4.7%/year).
- 22% experienced ICD-related complications (4.4%/year).
- 0.5% ICD-related mortality (0.08%/year).



Conclusion

- ICD does harm to patients with inherited arrhythmia syndromes.