# CIED u vrozených srdečních vad: ICD


**Jan Škoda**

**Kardiologické odd. Nemocnice Na Homolce**

jan.skoda@homolka.cz

NEMOCNICE NA HOMOLCE

# VSV-ICD evidence: sekund./prim. prevence, distribuce ICD-dg.

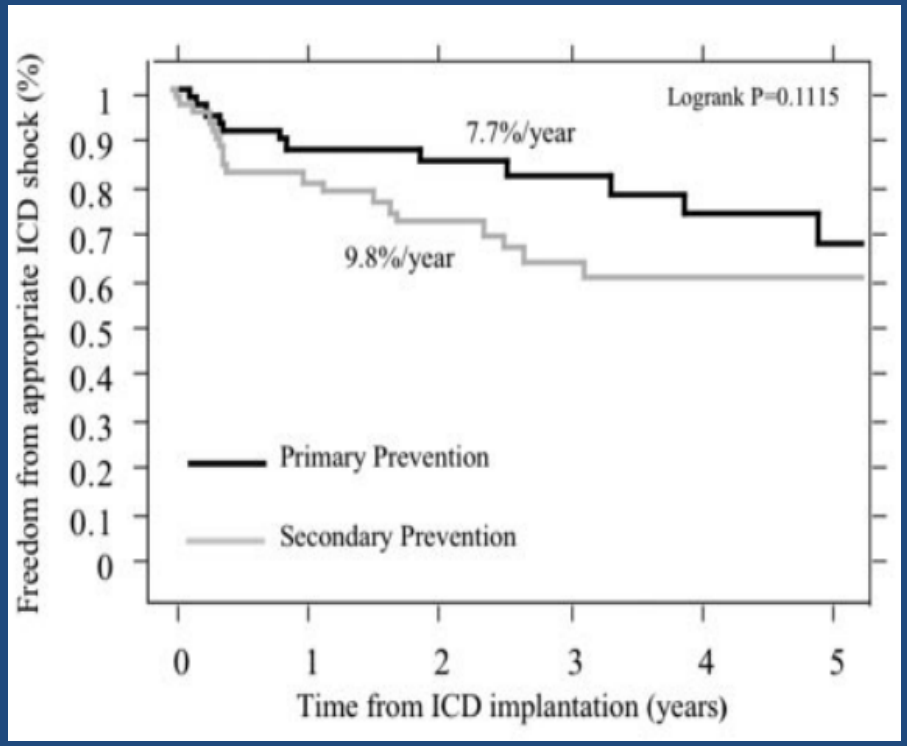
**Sekundární prevence: CA/SMVT...nepochybná implantce**

**Primární prevence, rizika ??:**

- **Nepřítomnost prospektivních či randomiz. studií**
- **Retrospektivní/Observační studie, meta, malá čísla**
- ***TOF:* nejčastější, více než polovina všech implantací, nejvíce dat, prediktory NSS definovány**
- ***dTGA-Must/Sen:* cca čtvrtina implantací, slabá data, prediktory NSS nedefinovány**
- ***ostatní dg CCTGA, UNI-V a další dg.:* <5%, data?**

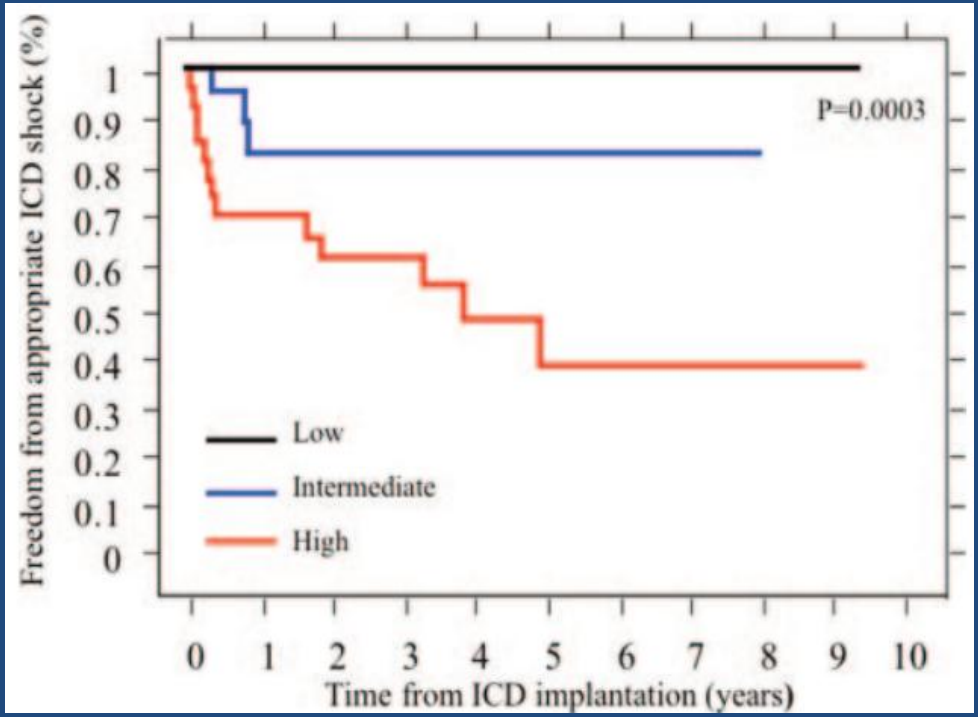
# Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

- retrosp. 11 center, 121p, věk 33let, 59%M, FUP 4R
- prim. 68/56%, sek.53/44%,
- adekv. výboje 7.7%/9.8% ročně
- prim. indep. prediktory NSVT a LVEDP
- PSK inducibilita 72%, mono/poly 7:1
- adekv.terapie FUP: PSK induc. 23/37%
  PSK noniduc. 3/12.5%
- neadekvátní výboje 6.6% ročně-mladší
- 30% komplikací (6xAcut, 25x gen-lead)
- roční mortalita 2.2%




Khairy:Circ Arrh Electrophysiology 2008; 1:250-257.

# TOF: rizikové faktory adekv výboje , skóre rizika




| Risk score | Risk category | N | Annualized rate of appropriate shocks |
|---|---|---|---|
| 0-2 | Low | 18 | 0% |
| 3-5 | Intermediate | 24 | 3.8% |
| 6-12 | High | 26 | 17.5% |

**Table 3. Risk Score for Appropriate ICD Shocks in Primary Prevention**

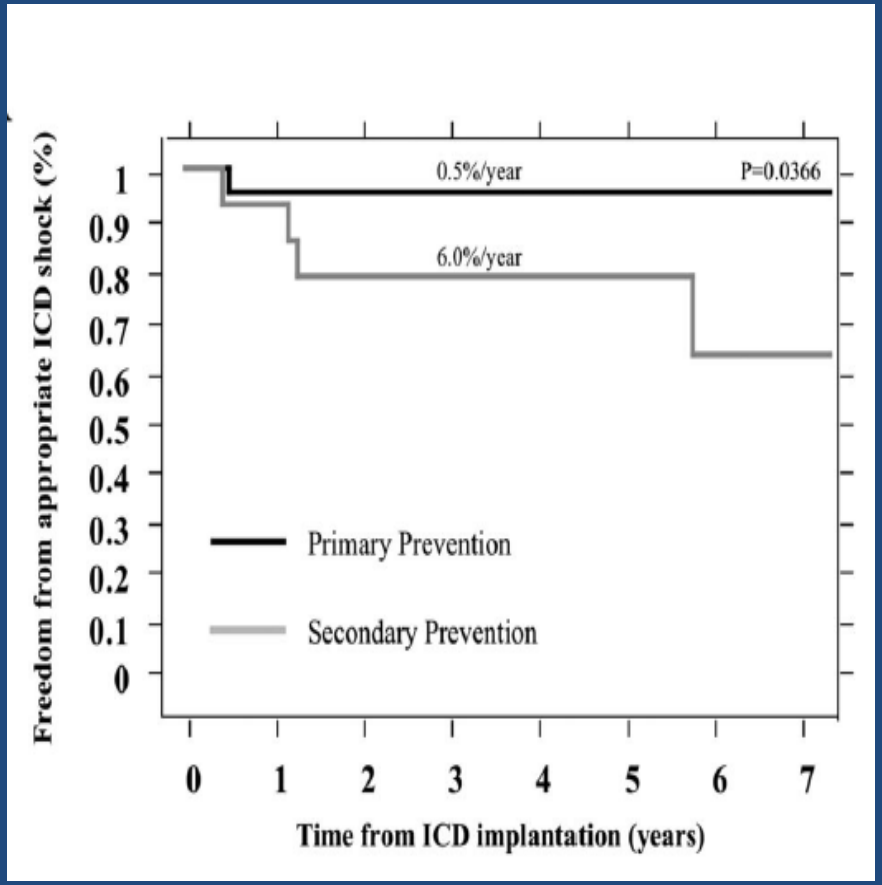
| Variable | Exp($\beta$) | Points Attributed |
|---|---|---|
| Prior palliative shunt | 3.2 | 2 |
| Inducible sustained ventricular tachycardia | 2.6 | 2 |
| QRS duration ≥180 ms | 1.4 | 1 |
| Ventriculotomy incision | 3.4 | 2 |
| Nonsustained ventricular tachycardia | 3.7 | 2 |
| LVEDP ≥12 mm Hg | 4.9 | 3 |
| Total points | . . . | 0–12 |

**Khairy:Circ Arrh Electrophysiology 2008; 1:250-257.**

# Sudden Death and Defibrillators in Transposition of the Great Arteries With Intra-atrial Baffles

## A Multicenter Study

- retrosp. 7 center, věk 28let
- 37 pac: 23prim/14sek.
- adekv. výboje jen 0.5%/6.0% ročně
- PSK nepredikuje adekv. terapii (pozit jen u 9/37, 3xmono, 6xpolyVT)
- sekund. impl. HR 18.0
- nepřítomnost BB 16.0
- 50% výskyt SVT před nebo s VT/VF
- NSVT? synkopa? RVsyst dysfunkce?
- neadekvátní výboje 6.6% ročně-mladší
- 37% komplikací (5xAcut, 12 xLate-lead)




Khairy:Circ Arrh Electrophysiology 2008; 1:250-257.

# Metaanalýza ICD u dospělých VSV: 2162 pac, FUP 3.6R, 53% prim.prev,

European Heart Journal (2016) **37**, 1439–1448
doi:10.1093/eurheartj/ehv735

META-ANALYSIS

## Implantable cardioverter-defibrillators in adults with congenital heart disease: a systematic review and meta-analysis

**Jim T. Vehmeijer[1], Tom F. Brouwer[1], Jacqueline Limpens[2], Reinoud E. Knops[1], Berto J. Bouma[1], Barbara J.M. Mulder[1,3], and Joris R. de Groot[1]***

[1]Department of Clinical and Experimental Cardiology, Heart Center, Academic Medical Center, PO Box 22700, 1100 DE, Amsterdam, The Netherlands; [2]Medical Library, Academic Medical Center, Amsterdam, The Netherlands; and [3]Interuniversity Cardiology Institute of the Netherlands, Utrecht, The Netherlands

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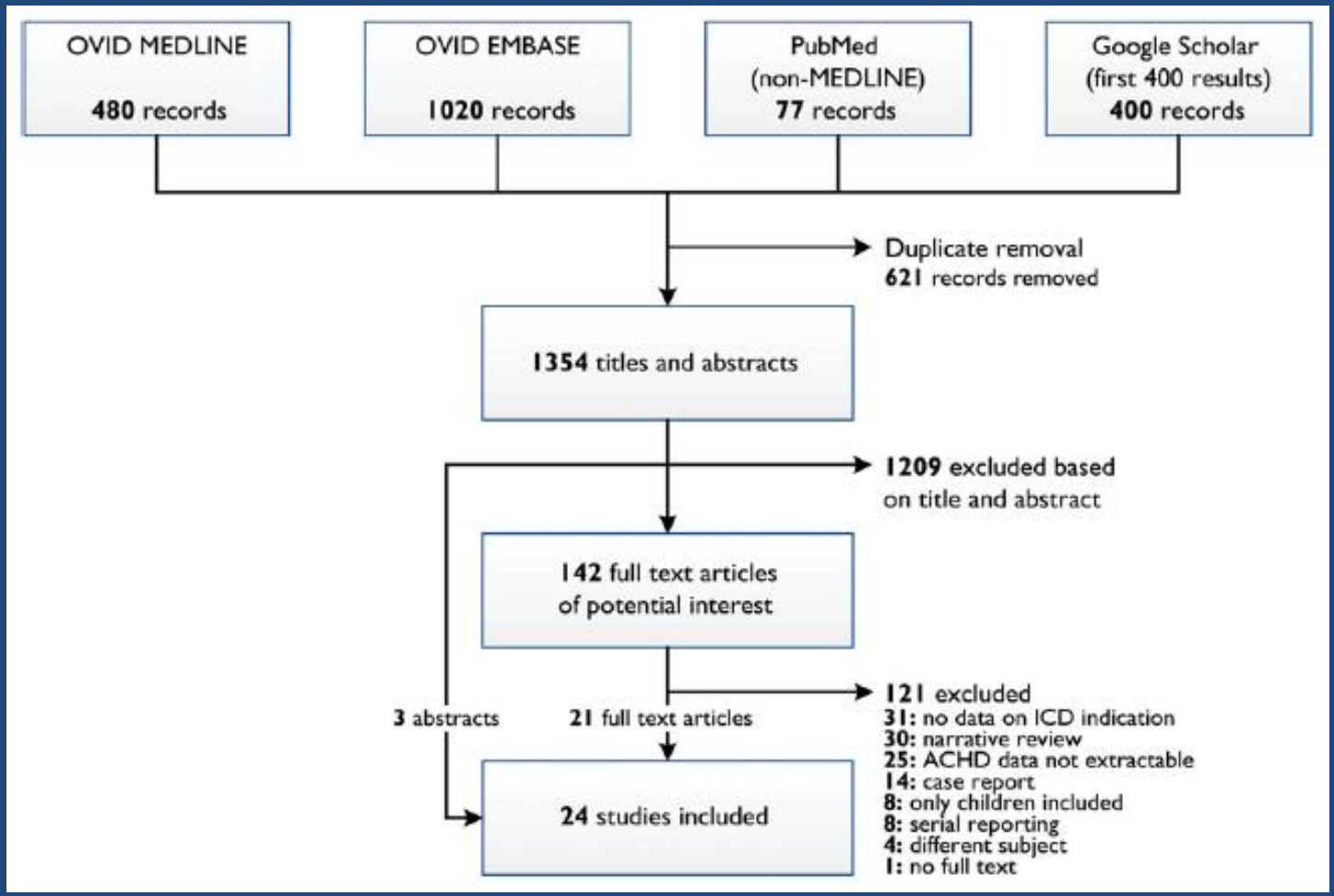
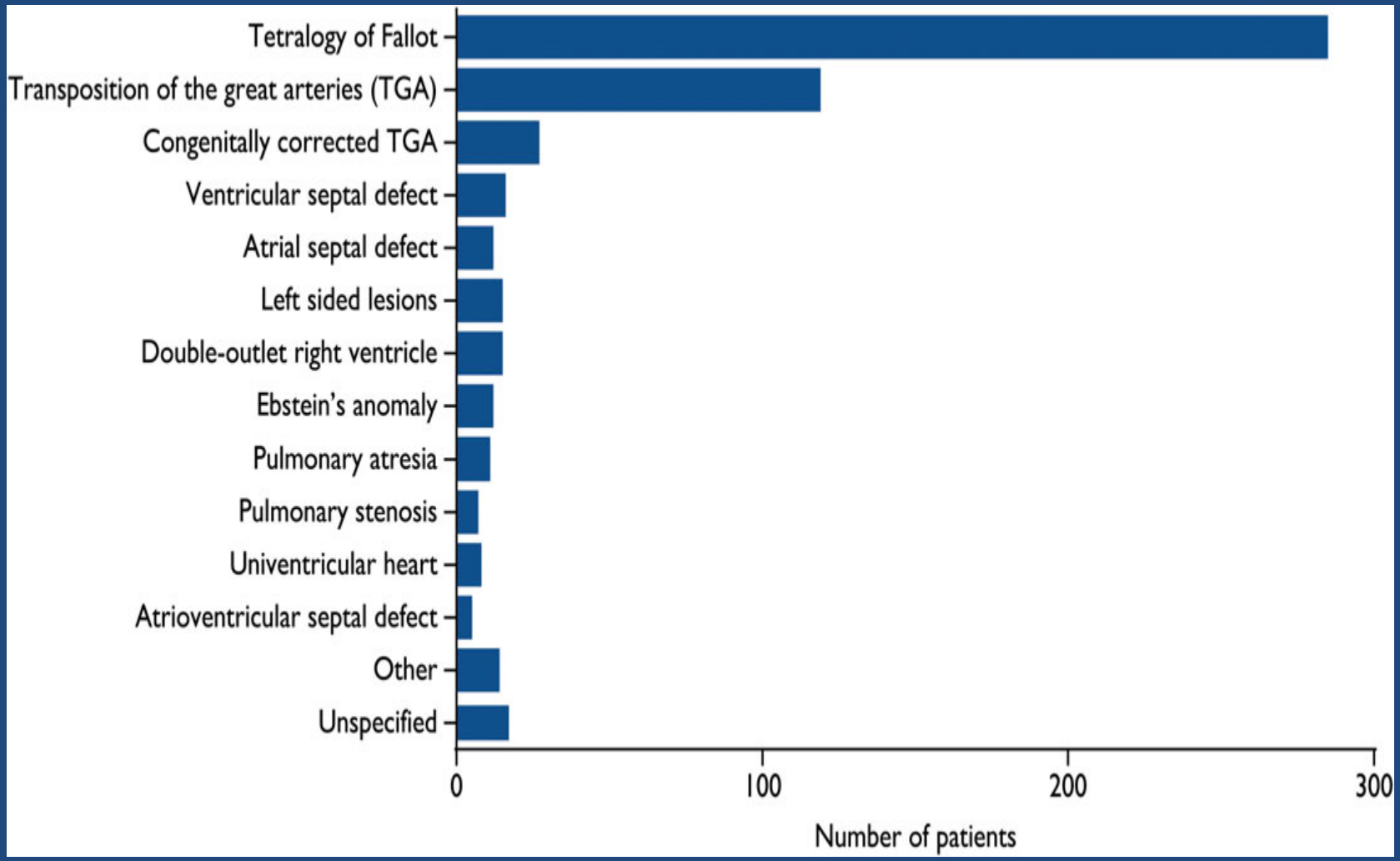
# Metaanalýza ICD u dospělých VSV: 1354 titulů-abstraktů...24 relevantních




European Heart Journal (2016) **37**, 1439–1448

# VSV-ICD: Distribuce diagnóz

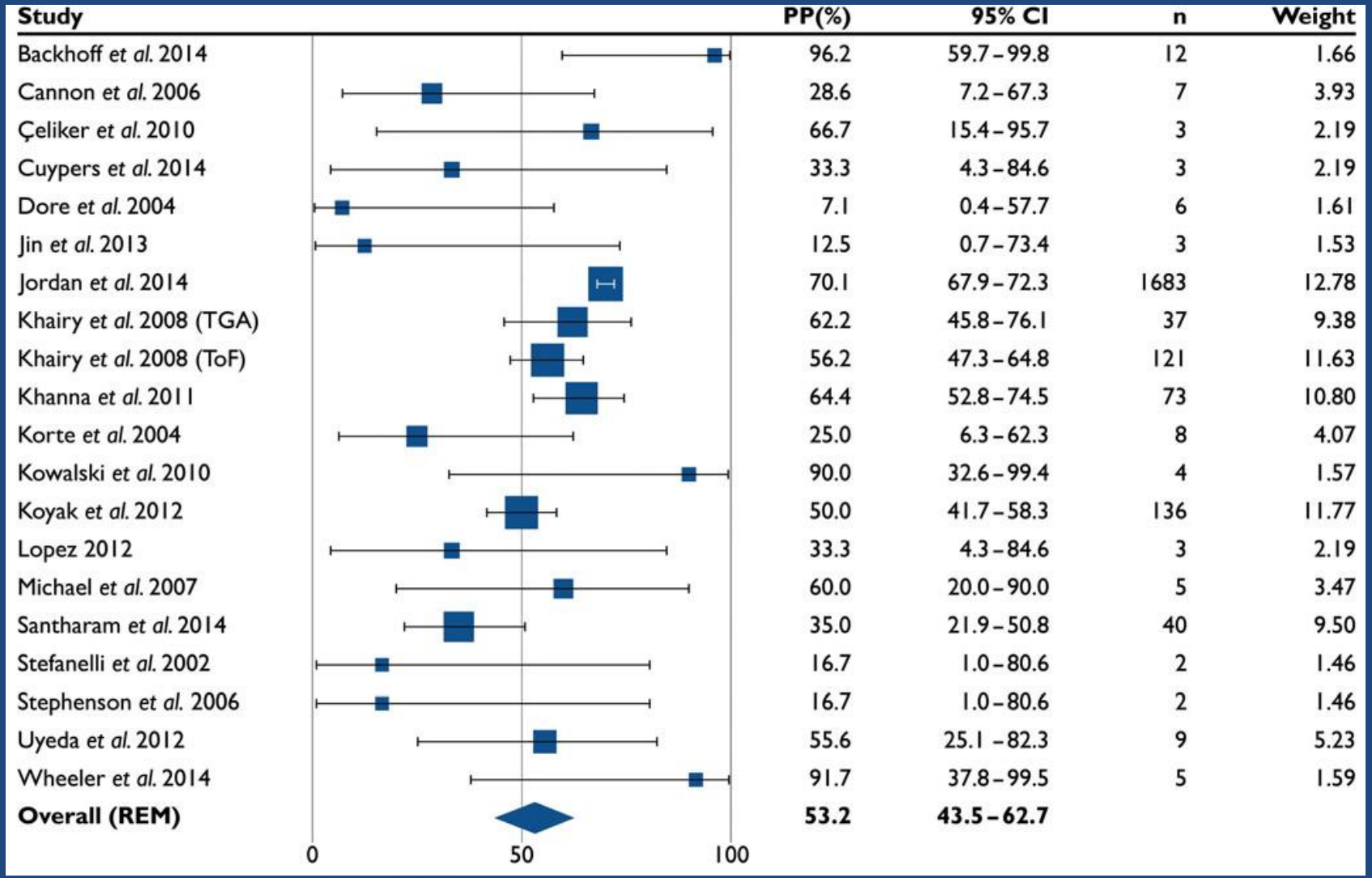
## metaanalýza 20 studií, 563 pac.




European Heart Journal (2016) **37**, 1439–1448

# VSV-ICD: primárně prev. implantací: 53%

## metaanalýza 20 studií, 2162 pac.

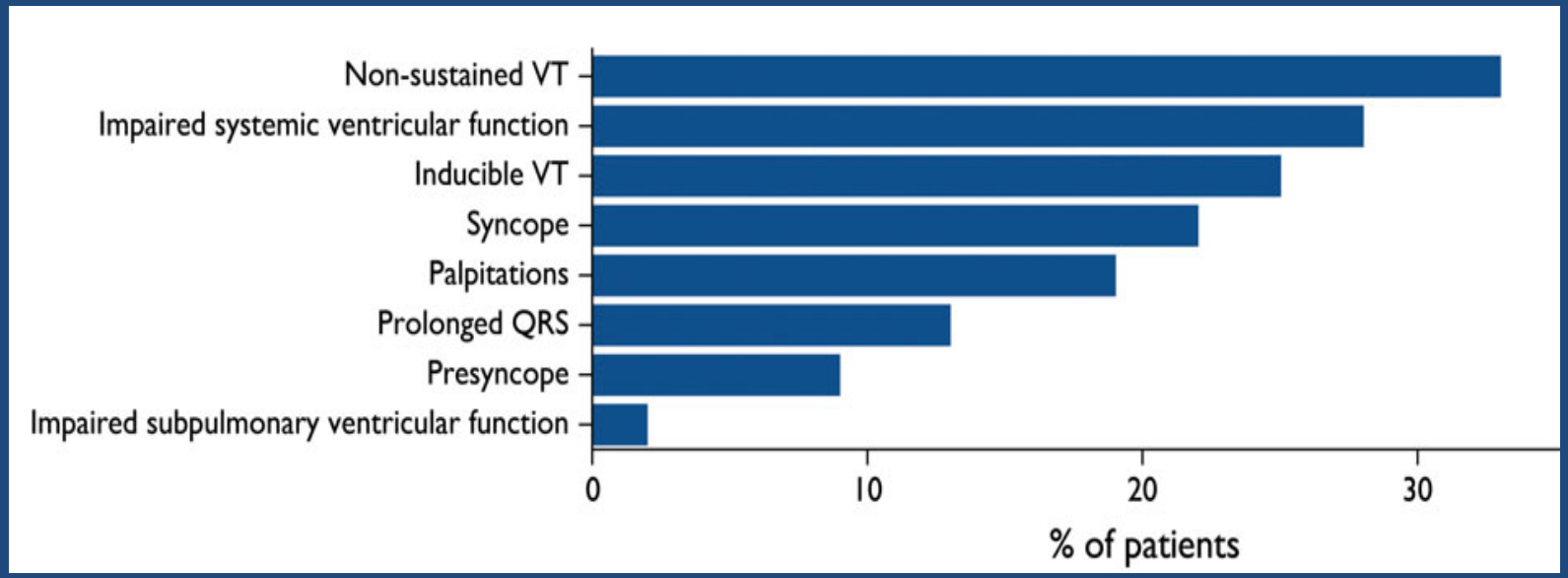


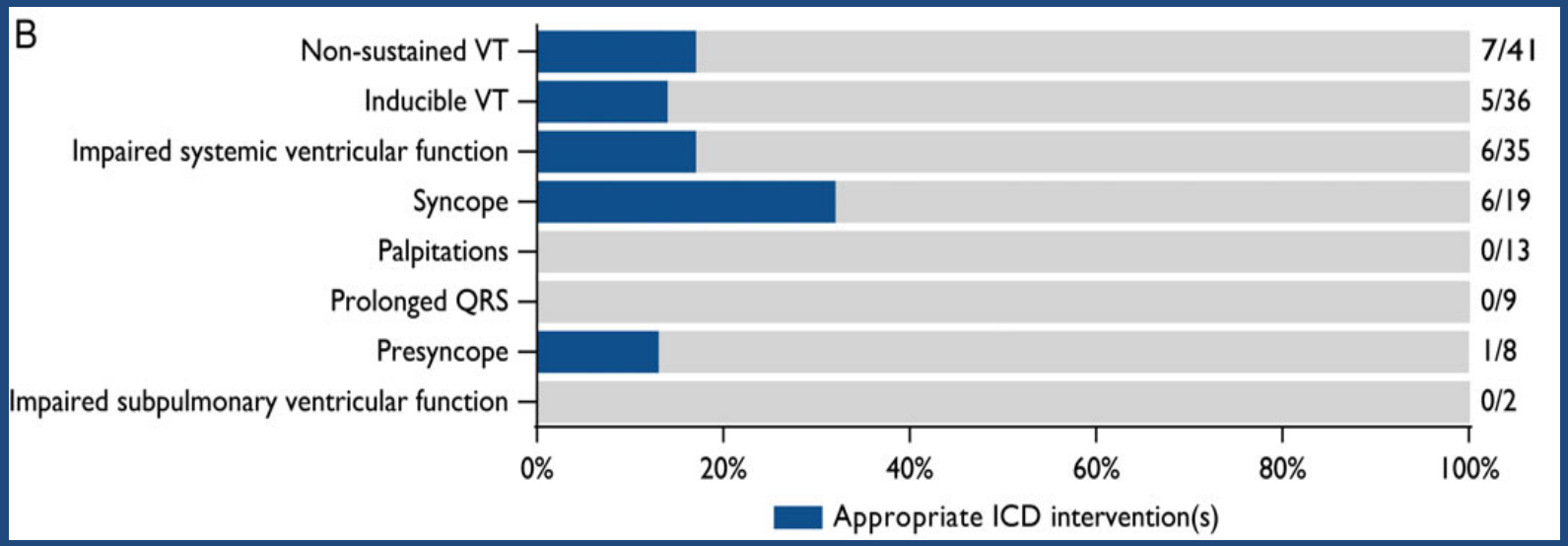
| Study | PP(%) | 95% CI | n | Weight |
|---|---|---|---|---|
| Backhoff *et al.* 2014 | 96.2 | 59.7–99.8 | 12 | 1.66 |
| Cannon *et al.* 2006 | 28.6 | 7.2–67.3 | 7 | 3.93 |
| Çeliker *et al.* 2010 | 66.7 | 15.4–95.7 | 3 | 2.19 |
| Cuypers *et al.* 2014 | 33.3 | 4.3–84.6 | 3 | 2.19 |
| Dore *et al.* 2004 | 7.1 | 0.4–57.7 | 6 | 1.61 |
| Jin *et al.* 2013 | 12.5 | 0.7–73.4 | 3 | 1.53 |
| Jordan *et al.* 2014 | 70.1 | 67.9–72.3 | 1683 | 12.78 |
| Khairy *et al.* 2008 (TGA) | 62.2 | 45.8–76.1 | 37 | 9.38 |
| Khairy *et al.* 2008 (ToF) | 56.2 | 47.3–64.8 | 121 | 11.63 |
| Khanna *et al.* 2011 | 64.4 | 52.8–74.5 | 73 | 10.80 |
| Korte *et al.* 2004 | 25.0 | 6.3–62.3 | 8 | 4.07 |
| Kowalski *et al.* 2010 | 90.0 | 32.6–99.4 | 4 | 1.57 |
| Koyak *et al.* 2012 | 50.0 | 41.7–58.3 | 136 | 11.77 |
| Lopez 2012 | 33.3 | 4.3–84.6 | 3 | 2.19 |
| Michael *et al.* 2007 | 60.0 | 20.0–90.0 | 5 | 3.47 |
| Santharam *et al.* 2014 | 35.0 | 21.9–50.8 | 40 | 9.50 |
| Stefanelli *et al.* 2002 | 16.7 | 1.0–80.6 | 2 | 1.46 |
| Stephenson *et al.* 2006 | 16.7 | 1.0–80.6 | 2 | 1.46 |
| Uyeda *et al.* 2012 | 55.6 | 25.1–82.3 | 9 | 5.23 |
| Wheeler *et al.* 2014 | 91.7 | 37.8–99.5 | 5 | 1.59 |
| **Overall (REM)** | **53.2** | **43.5–62.7** | | |



European Heart Journal (2016) **37**, 1439–1448

# VSV-ICD: distribuce prim. indikací (%) n=413
# Podíl adekvátní terapie n = 163

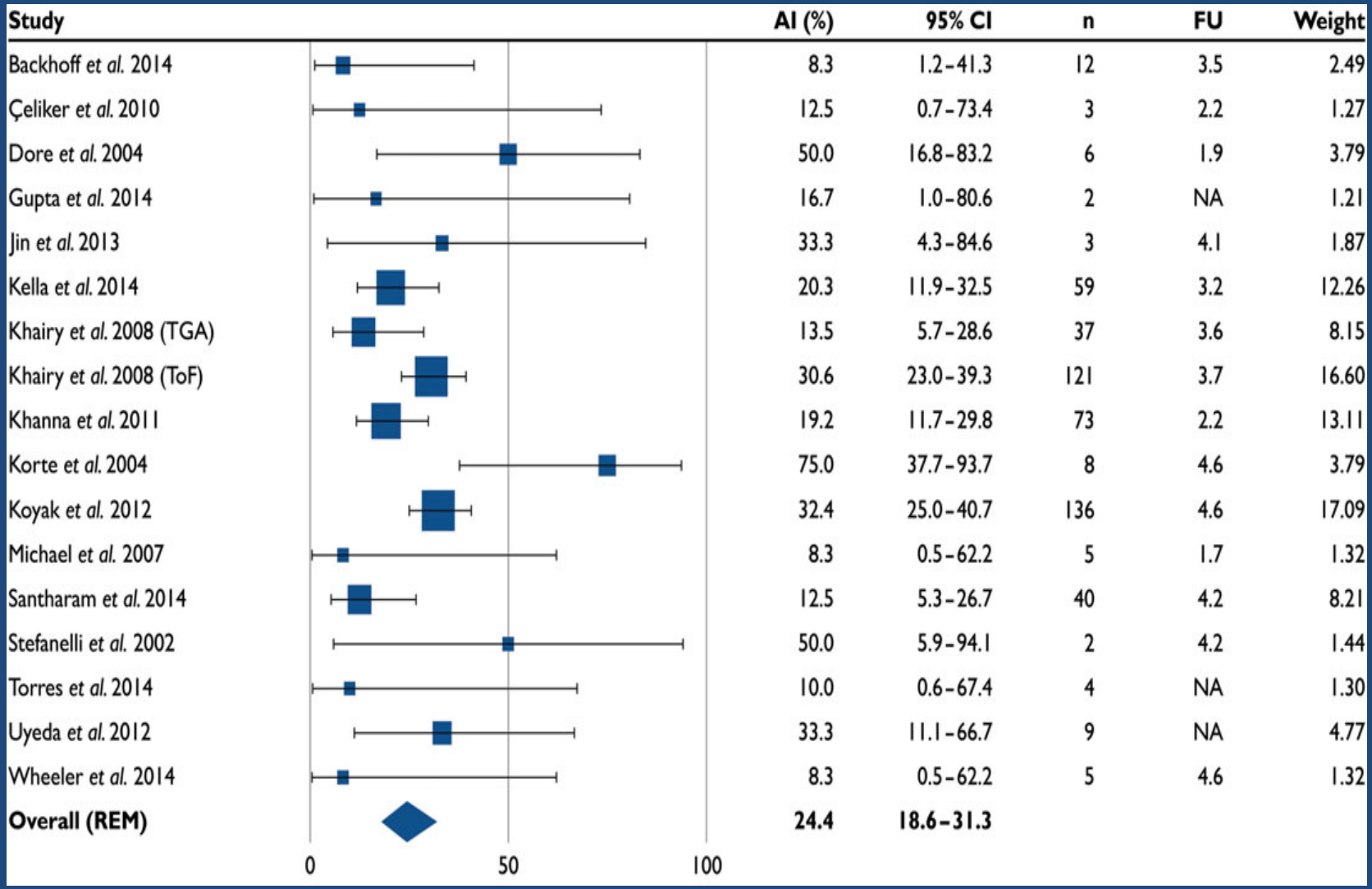



European Heart Journal (2016) **37**, 1439–1448

# VSV-ICD: adekvátní výboje - 24% pro FUP 3R

## metaanalýza 17 studií, 525 pac.



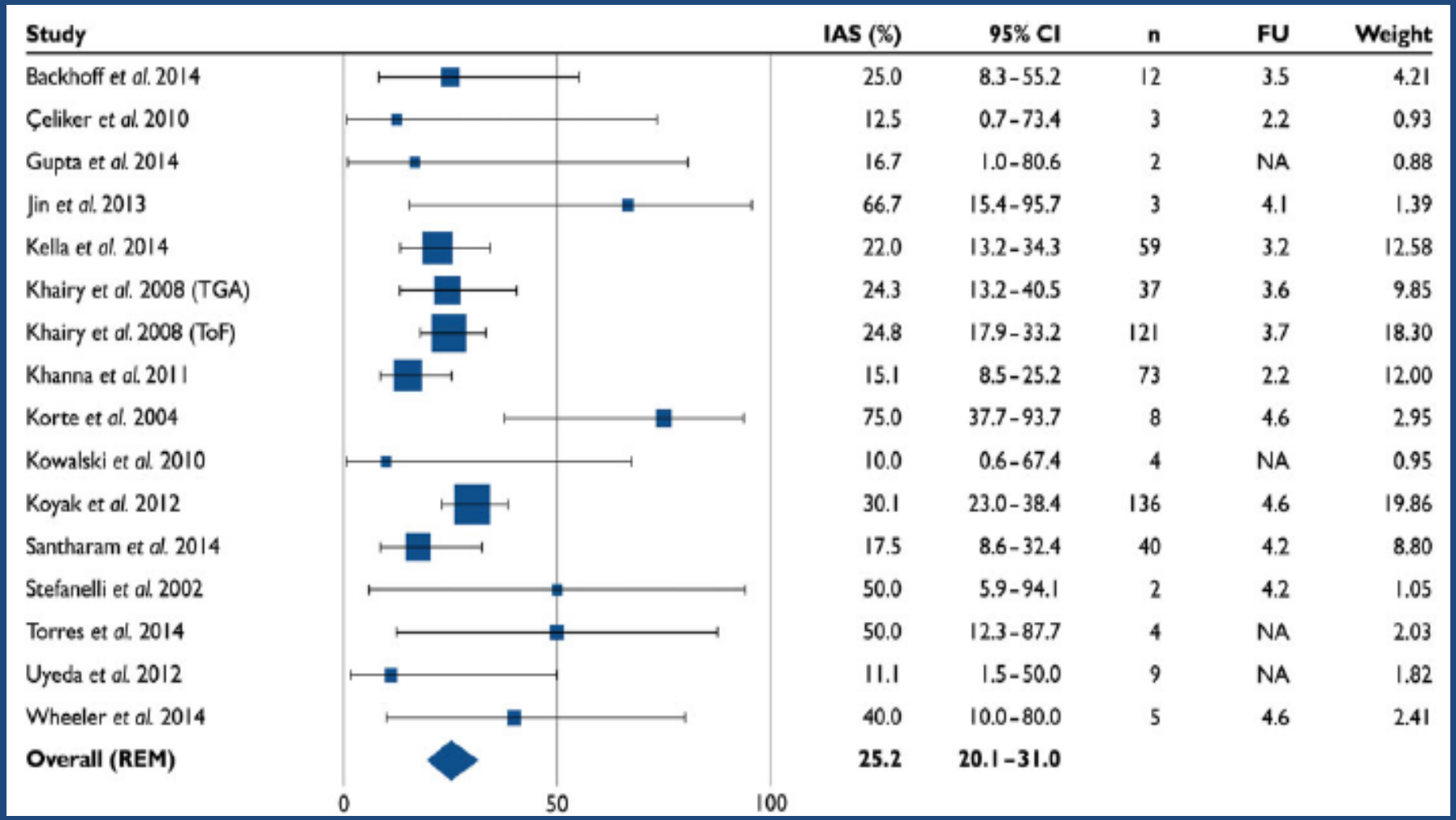
| Study | AI (%) | 95% CI | n | FU | Weight |
|---|---|---|---|---|---|
| Backhoff *et al.* 2014 | 8.3 | 1.2–41.3 | 12 | 3.5 | 2.49 |
| Çeliker *et al.* 2010 | 12.5 | 0.7–73.4 | 3 | 2.2 | 1.27 |
| Dore *et al.* 2004 | 50.0 | 16.8–83.2 | 6 | 1.9 | 3.79 |
| Gupta *et al.* 2014 | 16.7 | 1.0–80.6 | 2 | NA | 1.21 |
| Jin *et al.* 2013 | 33.3 | 4.3–84.6 | 3 | 4.1 | 1.87 |
| Kella *et al.* 2014 | 20.3 | 11.9–32.5 | 59 | 3.2 | 12.26 |
| Khairy *et al.* 2008 (TGA) | 13.5 | 5.7–28.6 | 37 | 3.6 | 8.15 |
| Khairy *et al.* 2008 (ToF) | 30.6 | 23.0–39.3 | 121 | 3.7 | 16.60 |
| Khanna *et al.* 2011 | 19.2 | 11.7–29.8 | 73 | 2.2 | 13.11 |
| Korte *et al.* 2004 | 75.0 | 37.7–93.7 | 8 | 4.6 | 3.79 |
| Koyak *et al.* 2012 | 32.4 | 25.0–40.7 | 136 | 4.6 | 17.09 |
| Michael *et al.* 2007 | 8.3 | 0.5–62.2 | 5 | 1.7 | 1.32 |
| Santharam *et al.* 2014 | 12.5 | 5.3–26.7 | 40 | 4.2 | 8.21 |
| Stefanelli *et al.* 2002 | 50.0 | 5.9–94.1 | 2 | 4.2 | 1.44 |
| Torres *et al.* 2014 | 10.0 | 0.6–67.4 | 4 | NA | 1.30 |
| Uyeda *et al.* 2012 | 33.3 | 11.1–66.7 | 9 | NA | 4.77 |
| Wheeler *et al.* 2014 | 8.3 | 0.5–62.2 | 5 | 4.6 | 1.32 |
| **Overall (REM)** | **24.4** | **18.6–31.3** | | | |



European Heart Journal (2016) **37**, 1439–1448

# Neadekvátní výboje: 25.2% pro FUP 3.3R

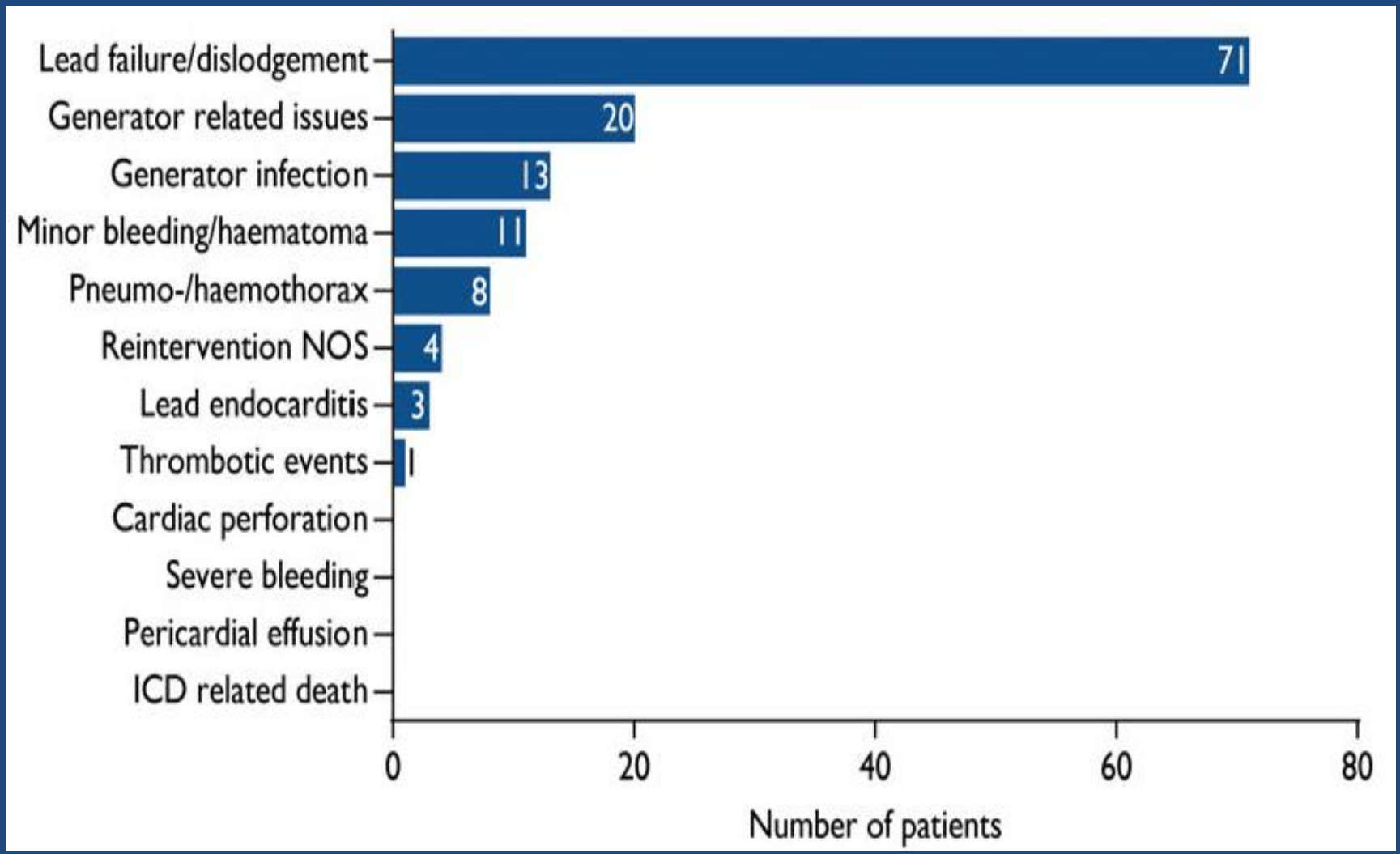
## metaanalýza 16 studií, 518 pac.




European Heart Journal (2016) **37**, 1439–1448

# VSV-ICD: Komplikace: 25% pro FUP 3.5R!!

## metaanalýza 11 studií, 433 pac.




European Heart Journal (2016) **37**, 1439–1448

ESC European Society of Cardiology
Europace (2018) 20, 1719–1720
doi:10.1093/europace/eux380

**2018** EHRA POSITION PAPER

# Arrhythmias in congenital heart disease: a position paper of the European Heart Rhythm Association (EHRA), Association for European Paediatric and Congenital Cardiology (AEPC), and the European Society of Cardiology (ESC) Working Group on Grown-up Congenital Heart Disease, endorsed by HRS, PACES, APHRS, and SOLAECE

Antonio Hernández-Madrid[1*†], Thomas Paul[2†], Dominic Abrams[3], Peter F. Aziz[4], Nico A. Blom[5,6], Jian Chen[7], Massimo Chessa[8], Nicolas Combes[9], Nikolaos Dagres[10], Gerhard Diller[11], Sabine Ernst[12], Alessandro Giamberti[13], Joachim Hebe[14], Jan Janousek[15], Thomas Kriebel[16], Jose Moltedo[17], Javier Moreno[1], Rafael Peinado[18], Laurent Pison[19], Eric Rosenthal[20], Jonathan R. Skinner[21], and Katja Zeppenfeld[22]

2013 ESC Guidelines on cardiac pac
resynchronization therapy

The Task Force on cardiac pacing and resynchroni
European Society of Cardiology (ESC). Develope
with the European Heart Rhythm Association (E

Authors/Task Force Members: Michele Brignole (Chairpers
Angelo Auricchio (Switzerland), Gonzalo Baron-Esquivias (Spain), Pierre Bordachar

PACES/HRS Expert Consensus Statement on the Recogni
Management of Arrhythmias in Adult Congenital Heart I
**2014**
*Developed in partnership between the Pediatric and Congenital Electrophysiolo*
*(PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodie*
*HRS, the American College of Cardiology (ACC), the American Heart Association*
*European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Soci*
*and the International Society for Adult Congenital Heart Disease (ISACHD).*

2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

therapy for arrhythmias in the pediatric population: EHRA and AEPC-Arrhythmia Working Group joint consensus statement

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

American Heart Association

2012 ACCF/AHA/HRS Focused Update Incorporated Into the ACCF/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities : A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society
Andrew E. Epstein, John P. DiMarco, Kenneth A. Ellenbogen, N.A. Mark Estes III, Roger A.

# VSV- ICD: Sekundární prevence NSS

- 1. Srdeční zástava při VF/nestabilní VT z nevratných příčin
- 2. Setrvalá symptomatická VT (alternativně jen katetrizační ablace či KCH u super-selektivní indikace)

| Recommendations | Consensus statement |
|---|---|
| ICD is recommended for patients with CHD who are survivors of an aborted cardiac arrest due to VF or haemodynamically unstable VT after evaluation to define the cause of the event and exclusion of any reversible causes. |  |
| ICD is recommended for patients with CHD with symptomatic sustained VT who have undergone haemodynamic and electrophysiological evaluation. |  |

Europace (2018) **20**, 1719–1720

# VSV-ICD: Primární prevence NSS

1. Syst.EFLK 35% a RV+LV a NYHA II-III
2. Synkopa: PSK+, nebo význ. V-dysfunkce
3. TOF více rizik: LV dysf, NSVT, QRS>180, PSK+
4. UNi/RVsyst. dysf + rizika: NYHA II-III, QRS>140, význ. systémová AV reg.
5. VSV + wating list Tx

Kontraindikace <1R, incess.VT/VF,term.NYHA IV, nekompatibilní s význ.psychiatr. dg.

kurativní možnost RFA

ICD is recommended in adults with CHD and a systemic LVEF ≤ 35%, biventricular physiology and NYHA functional Class II or III.

ICD implantation should be considered in patients with CHD and syncope of unknown origin in the presence of either advanced ventricular dysfunction or inducible sustained VT or VF on VPS.

ICD implantation should be considered in selected patients with TOF and multiple risk factors for SCD, including LV dysfunction, non-sustained VT, QRS duration ≥180 ms, or inducible sustained VT on VPS.

ICD therapy may be considered in patients with advanced single or systemic RV dysfunction in the presence of risk factors such as non-sustained VT, NYHA functional Class II or III, QRS duration ≥ 140 ms or severe systemic AV valve regurgitation.

ICD therapy may be considered for non-hospitalized adults with CHD awaiting heart transplantation.

ICD is contraindicated in patients with CHD with life expectancy with an acceptable functional status ≤ 1 year, incessant VT or VF, significant psychiatric illness that may be aggravated by ICD implantation or preclude systematic follow-up, and patients with drug-refractory NYHA Class IV symptoms who are not candidates for cardiac transplantation or CRT

Europace (2018) **20**, 1719–1720

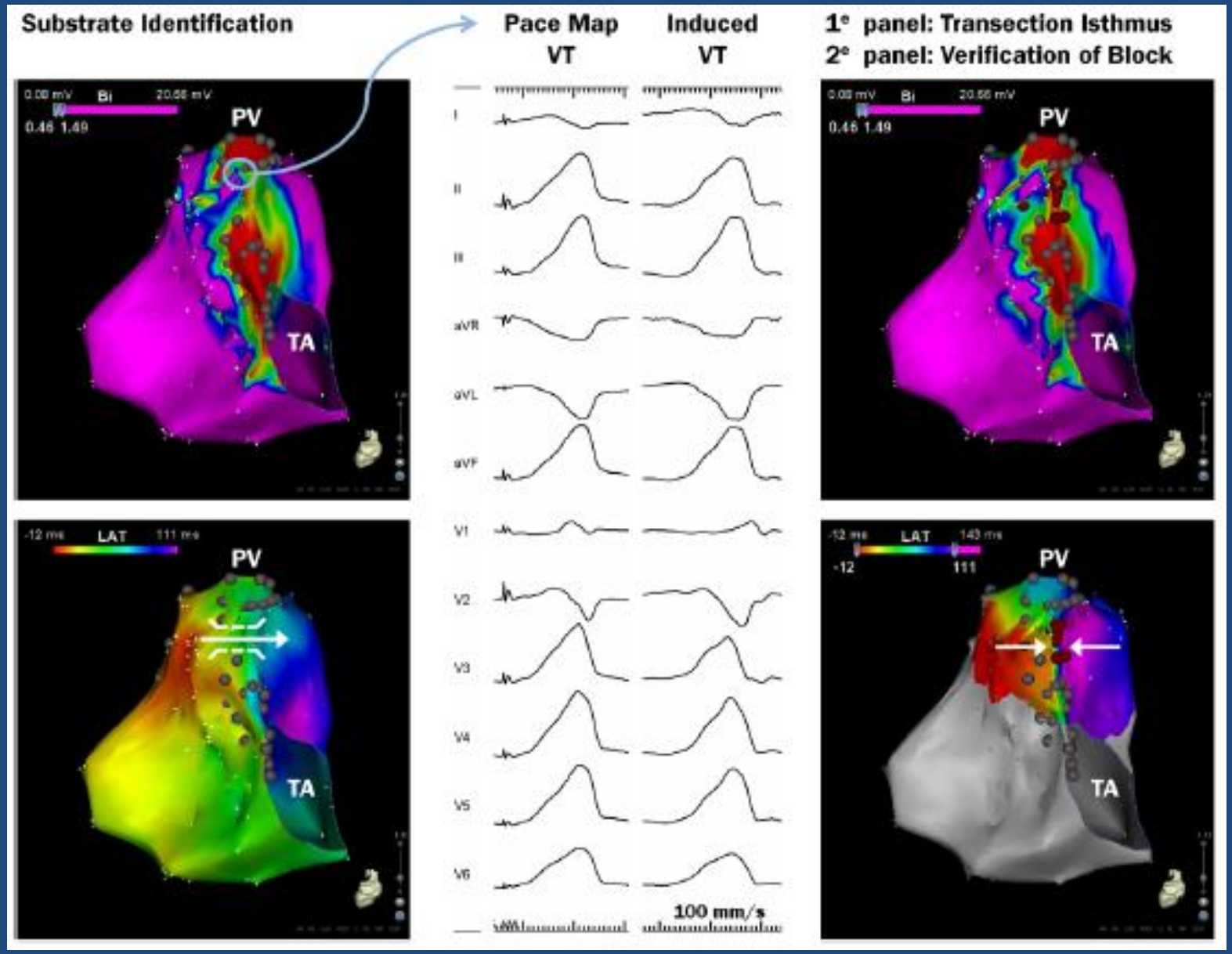
# Re-Entry Using Anatomically Determined Isthmuses
## A Curable Ventricular Tachycardia in Repaired Congenital Heart Disease

Gijsbert F.L. Kapel, MD; Tobias Reichlin, MD; Adrianus P. Wijnmaalen, MD, PhD; Sebastiaan R.D. Piers, MD; Eduard R. Holman, MD, PhD; Usha B. Tedrow, MD, MS; Martin J. Schalij, MD, PhD; William G. Stevenson, MD; Katja Zeppenfeld, MD, PhD

***Background***—Ventricular tachycardia (VT) is an important cause of late morbidity and mortality in repaired congenital heart disease. The substrate often includes anatomic isthmuses that can be transected by radiofrequency catheter ablation similar to isthmus block for atrial flutter. This study evaluates the long-term efficacy of isthmus block for treatment of re-entry VT in adults with repaired congenital heart disease.

***Methods and Results***—Thirty-four patients (49±13 years; 74% male) with repaired congenital heart disease who underwent radiofrequency catheter ablation of VT in 2 centers were included. Twenty-two (65%) had a preserved left and right ventricular function. Patients were inducible for 1 (interquartile range, 1–2) VT, median cycle length: 295 ms (interquartile range, 242–346). Ablation aimed to transect anatomic isthmuses containing VT re-entry circuit isthmuses. Procedural success was defined as noninducibility of any VT and transection of the anatomic isthmus and was achieved in 25 (74%) patients. During long-term follow-up (46±29 months), all patients with procedural success (18/25 with internal cardiac defibrillators) were free of VT recurrence but 7 of 18 experienced internal cardiac defibrillator-related complications. One patient with procedural success and depressed cardiac function received an internal cardiac defibrillator shock for ventricular fibrillation. None of the 18 patients (12/18 with internal cardiac defibrillators) with complete success and preserved cardiac function experienced any ventricular arrhythmia. In contrast, VT recurred in 4 of 9 patients without procedural success. Four patients died from nonarrhythmic causes.

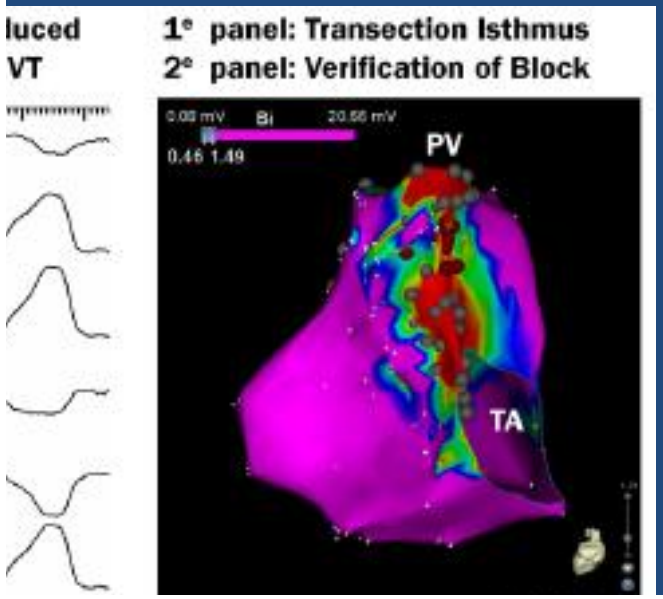
***Conclusions***—In patients with repaired congenital heart disease with preserved ventricular function and isthmus-dependent re-entry, VT isthmus ablation can be curative. (***Circ Arrhythm Electrophysiol.*** **2015;8:102-109. DOI: 10.1161/CIRCEP.114.001929.)**

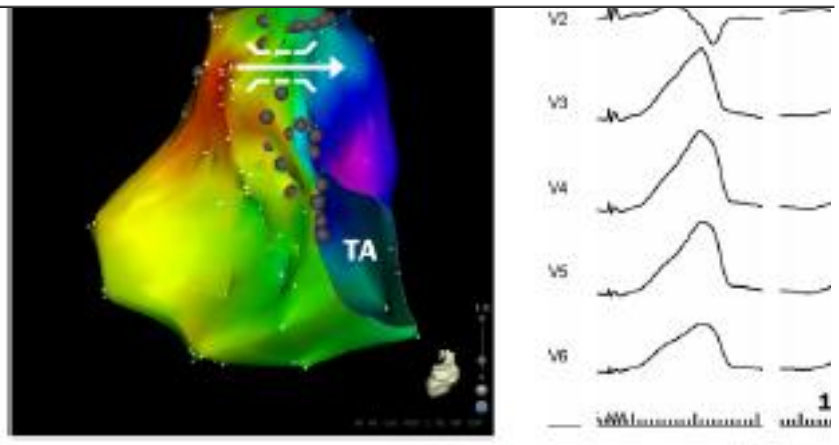



*Circ Arrhythm Electrophysiol.* 2015;8:102-109.

**Table 3. Long-Term Outcome**

| | No Complete Success (n=9) | Complete Success (n=25) | *P* Value |
|---|---|---|---|
| Follow-up, mo | 46±22 | 46±31 | 0.994 |
| SMVT | 4 (44%) | 0 (0%) | <0.001 |
| VF | 0 (0%) | 1 (4%) | 0.527 |
| ICD | 9 (100%) | 18 (72%) | 0.205 |
| Appropriate therapy | 4 (44%) | 1 (6%) | 0.042 |
| ATP | 4 (44%) | 0 (0%) | 0.007 |
| Shock | 2 (22%) | 1 (6%) | 0.350 |
| Inappropriate shock | 1 (11%) | 5 (28%) | 0.277 |
| Mortality, all-cause | 3 (33%) | 1 (4%) | 0.025 |
| Mortality, cardiac | 2 (22%) | 0 (0%) | 0.010 |




## WHAT THE STUDY ADDS

- In a large group of repaired congenital heart disease patients, transection of VT-associated anatomic isthmuses by radiofrequency catheter ablation is feasible in 74% of the patients and is highly effective to prevent VT recurrence during long-term follow-up.
- In selected patients with repaired congenital heart disease, with preserved left and right ventricular function, and anatomic isthmus dependent re-entry VT in whom other ventricular arrhythmias mechanisms are unlikely isthmus ablation may be reasonable as sole therapy for the arrhythmia.

*Circ Arrhythm Electrophysiol.* 2015;8:102-109.

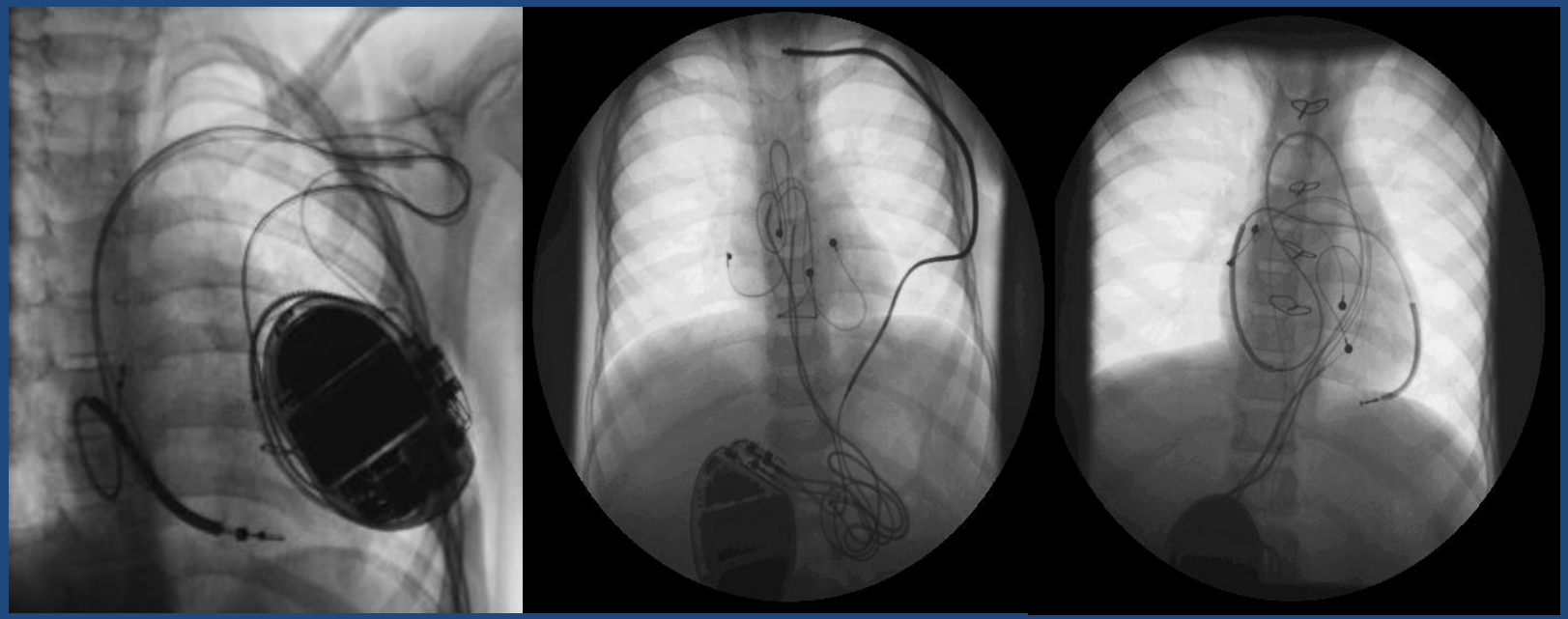
# VSV-ICD: Technika

## Neadekvátní výboje: programace a monitoring

- Vysoké detekční frekvence (sinus tachy)
- Dlouhé detekční časy (selfterminace)
- Optimalizace senzingu T vlny
- Extenzivní ATP i pro velmi vysoké detekce
- Monitorace R, stim/DFT prahů, růstové změny
- Psychologické problémy

# VSV-ICD: Technika

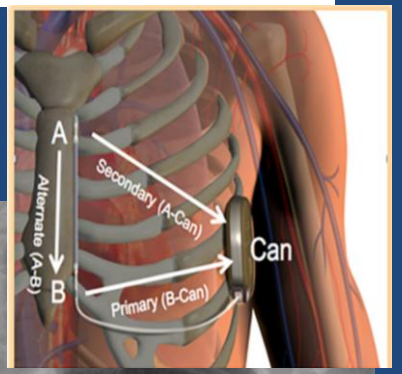
## Nepřítomnost transvenózního přístupu
## Ochrana dětského transvenóuzního přístupu do dospělosti

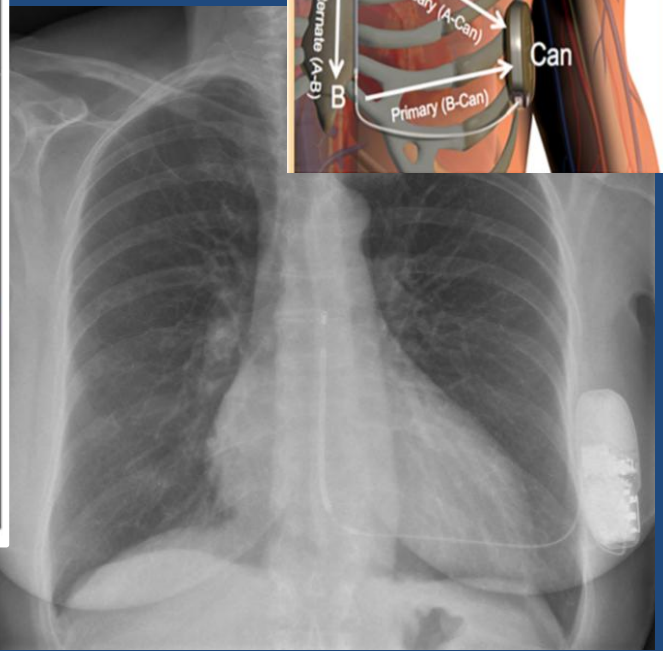


adaptováno: prof. Janoušek dětské Kardiocentrum FNM

# VSV: S-ICD

| Recommendations | Class[a] | Level[b] | Ref.[c] |
|---|---|---|---|
| Subcutaneous defibrillators should be considered as an alternative to transvenous defibrillators in patients with an indication for an ICD when pacing therapy for bradycardia support, cardiac resynchronization or antitachycardia pacing is not needed. | IIa | C | 157, 158 |
| The subcutaneous ICD may be considered as a useful alternative to the transvenous ICD system when venous access is difficult, after the removal of a transvenous ICD for infections or in young patients with a long-term need for ICD therapy. | IIb | C | This panel of experts |

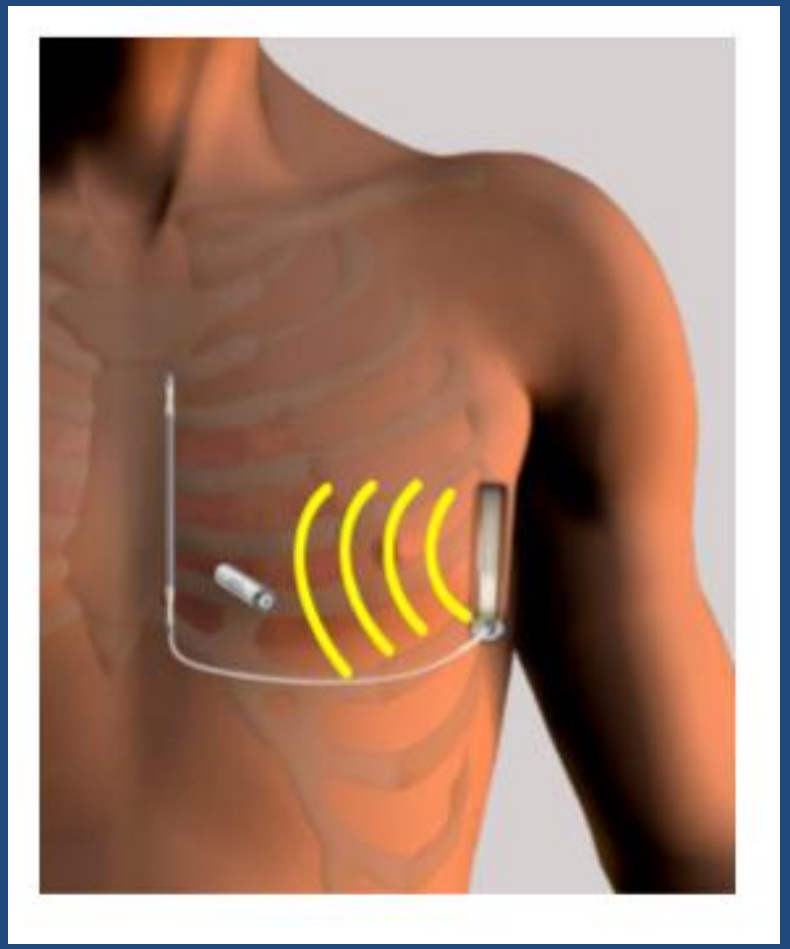



# VSV: S-ICD další vývoj

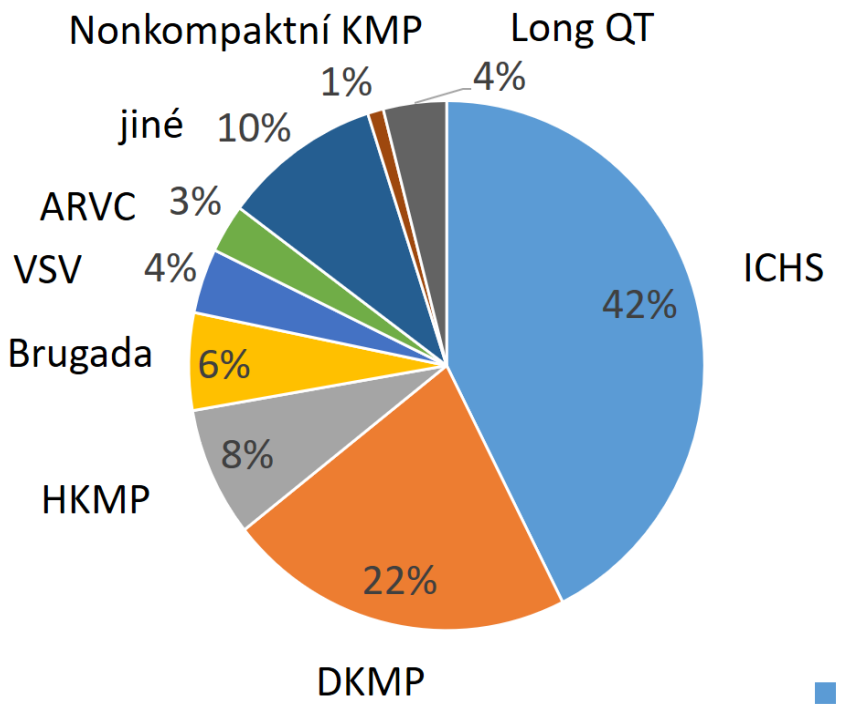
## EMPOWER™ leadless pacemaker

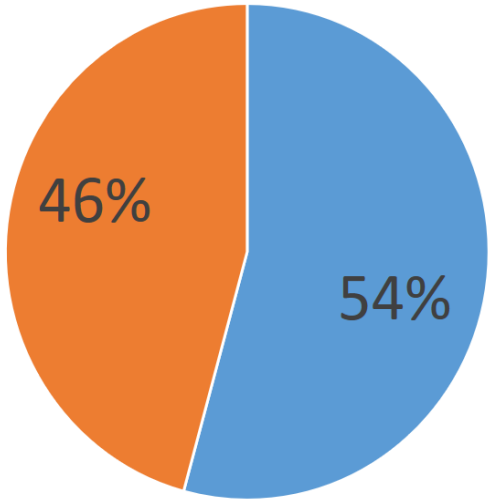
- Jednosměrná komunikace
- 99% úspěšnost wireless komunikace s S-ICD
- Správná diskriminace stimulovaného rytmu
- 99% úspěšnost vyslání ATP
- Bez ovlivnění funkce leadless KS po výboji 80J



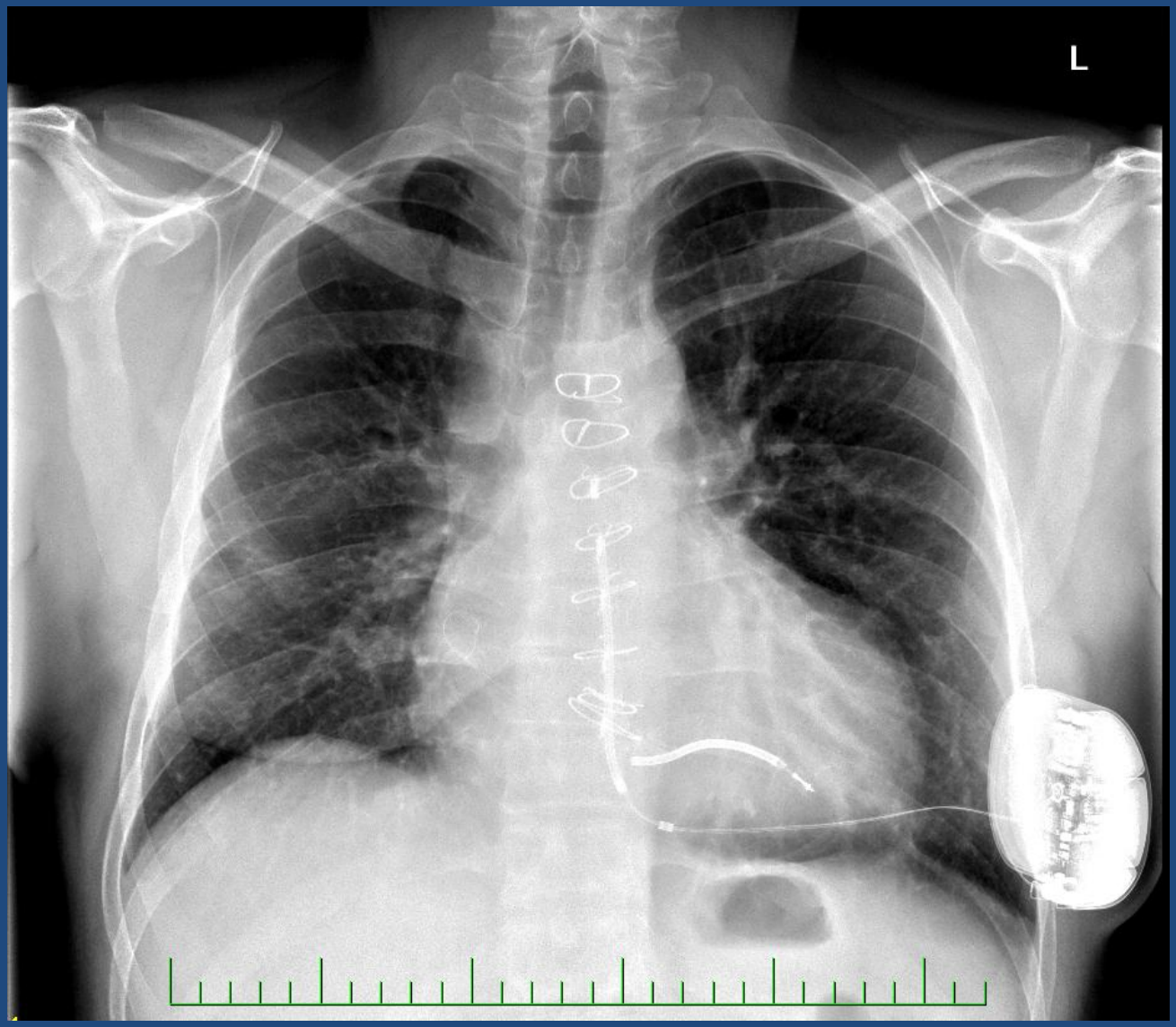
# NNH: 147 implantací S-ICD 2010-2018

## Hlavní diagnozy a indikace

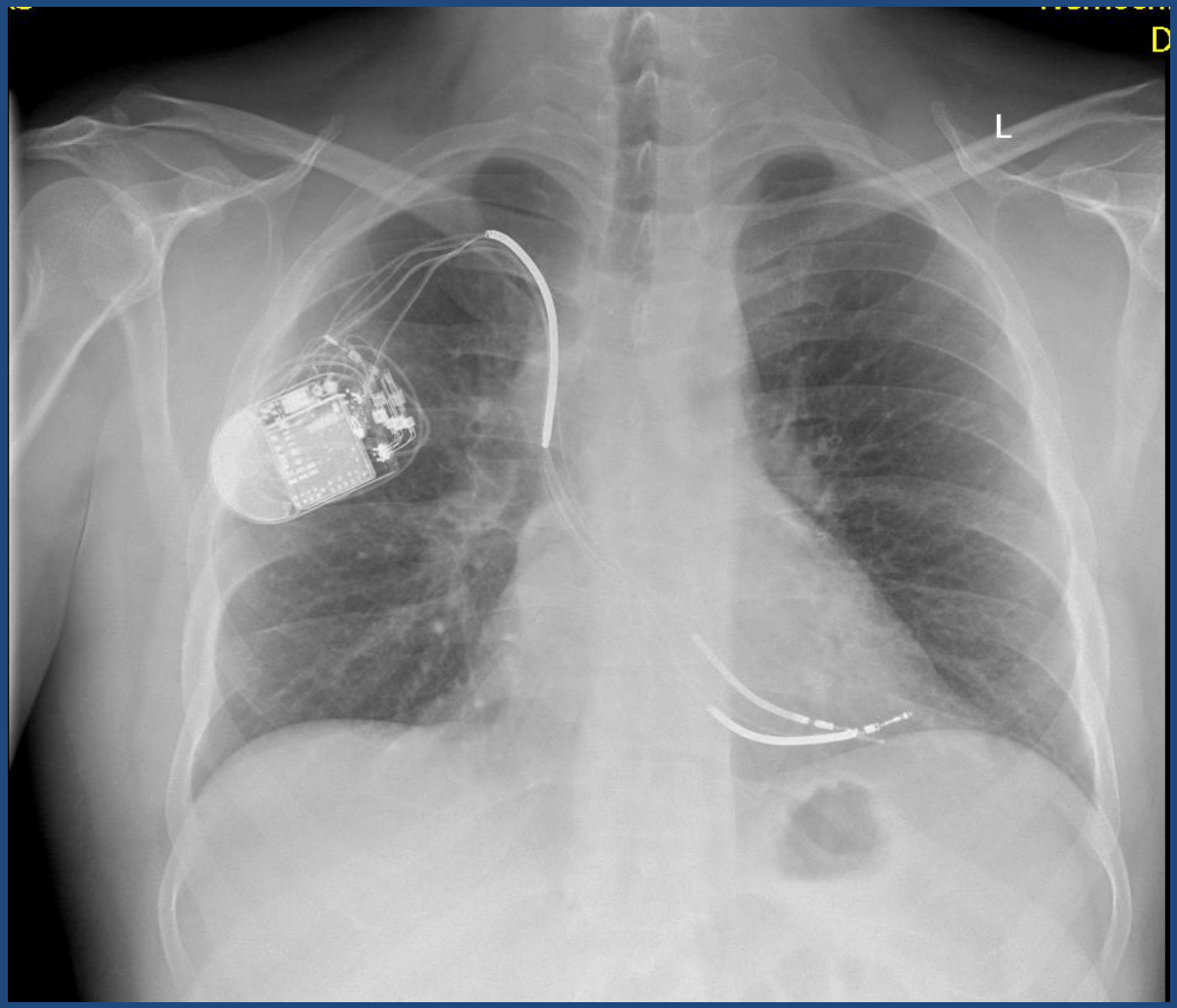


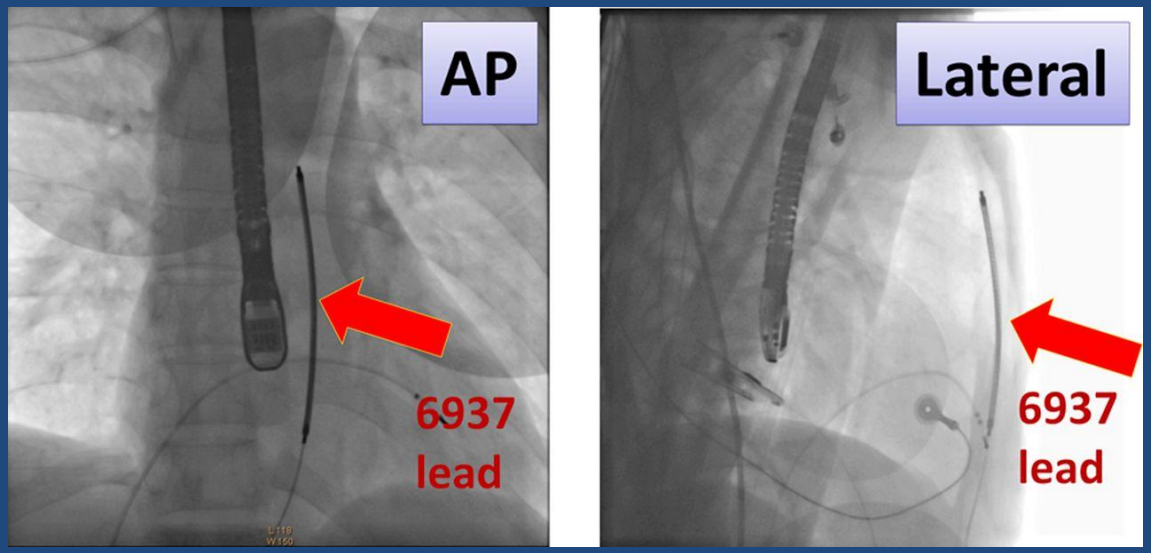
# TOF, neadekvátní výboje, neúplná extrakce



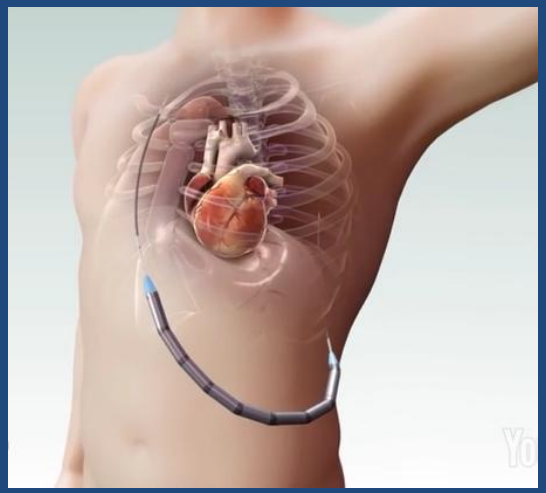
# dTGA-Mustard – opak. výměny a dysfunkce ICD elektrod



# Experimentální systémy:




EV-ICD System (Medtronic)



ISSD (Newpace)

# CIED u vrozených srdečních vad:

# ICD

Jan Škoda

Kardiologické odd. Nemocnice Na Homolce

jan.skoda@homolka.cz