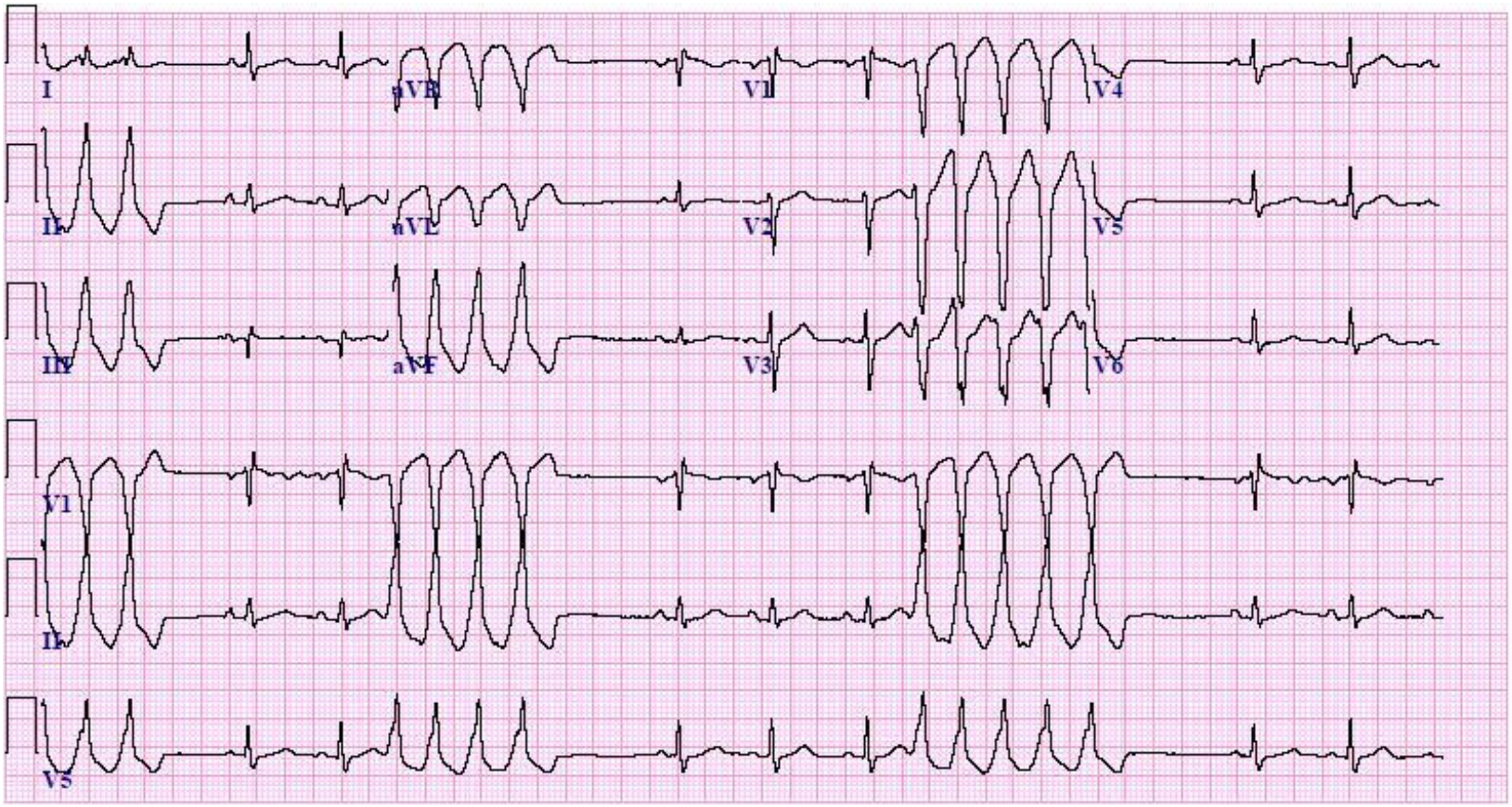
What to do for a 50-year-old woman with inoperable PA VSD and MAPCA complaining of recurrent NSVT?

이화의대
반지은

- Conflict of interest : non declared

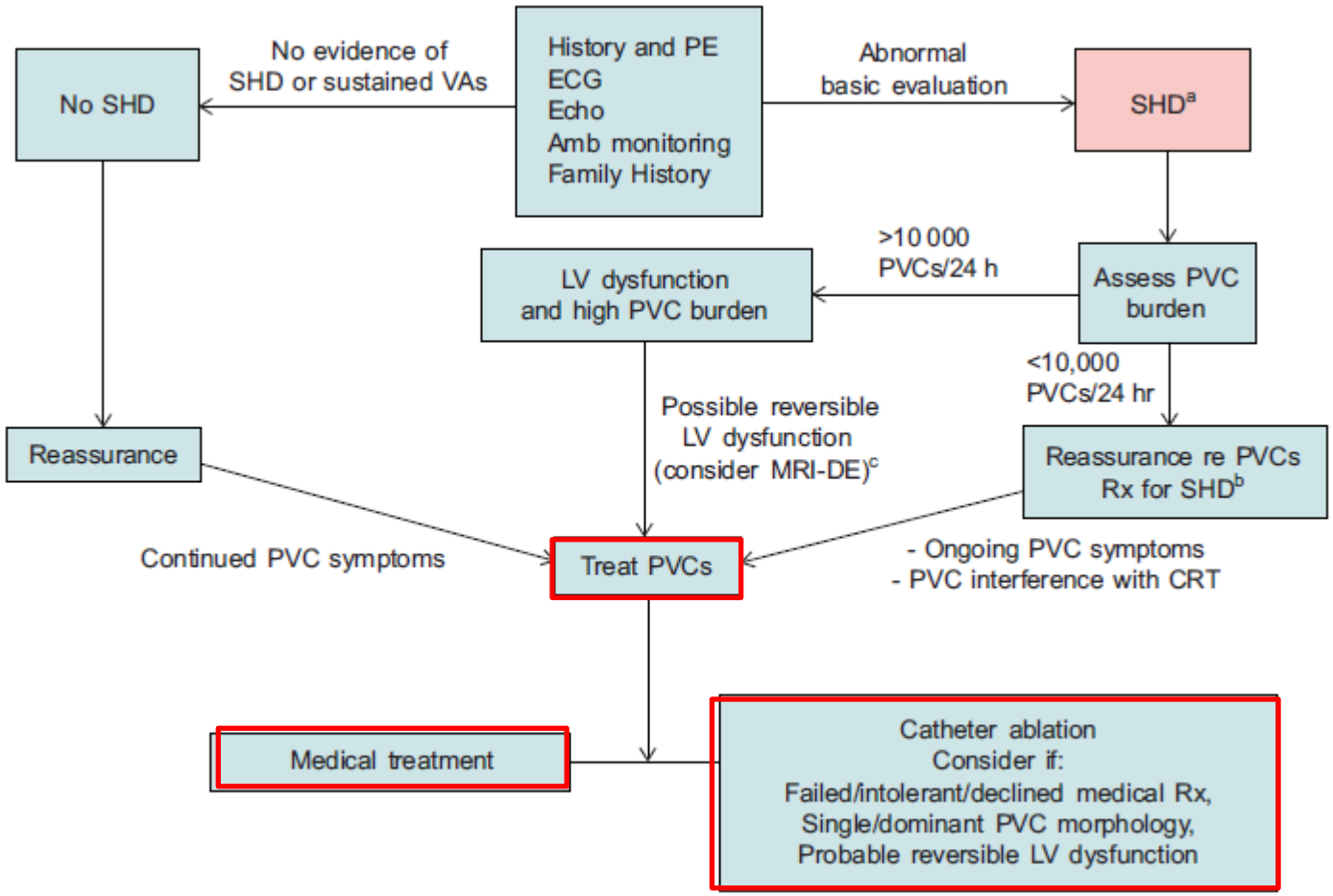
50-year-old woman, palpitation



No structural heart Ds

Management of PVCs

EHRA/HRS/APHRS Expert Consensus on Ventricular Arrhythmias, 2014



Symptomatic NSVT in normal heart

EHRA/HRS/APHRS Expert Consensus on Ventricular Arrhythmias 2014

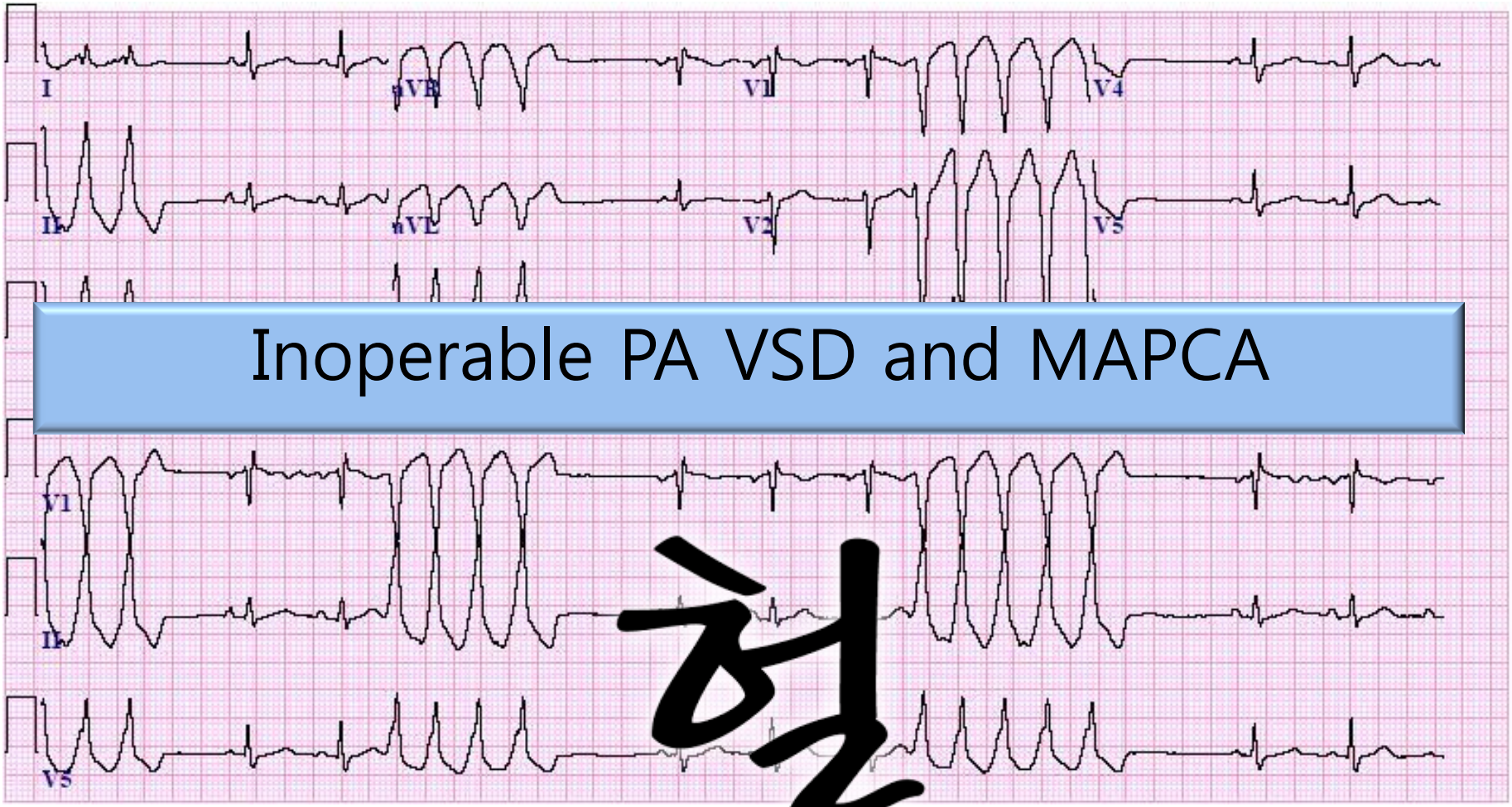
Expert consensus recommendations on non-sustained

(1) Infrequent ventricular ectopic beats, couplets, and triplets without other signs of an underlying SHD or an inherited arrhythmia syndrome should be considered as a normal variant in asymptomatic patients. IIa LOE C

(2) An invasive electrophysiological study (EPS) should be considered in patients with significant SHD and non-sustained VAs especially if accompanied by unexplained symptoms such as syncope, near-syncope, or sustained palpitations IIa LOE C

- **Beta-blockers**; considered in symptomatic patients with non-sustained VAs. (IIb, C)
- **Non-dihydropyridine calcium channel antagonist** : considered as an alternative to beta- blocker treatment. (IIb, C)
- **Antiarrhythmic drug (AAD: propafenone, flecainide, mexiletine,sotalol, amiodarone)** (IIb, C)
- **Catheter ablation** : improving symptoms or LV dysfunction (e.g. PVC >10,000/24 h) (IIa, B)

50-year-old woman, palpitation



Inoperable PA VSD and MAPCA

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Adult patient with CHD

- Asymptomatic
- Symptomatic

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease (2014)

Approach to the symptomatic adults with CHD

Sx suggestive arrhythmias (e.g., palpitation, presyncope, syncope), documented new-onset or worsening arrhythmias, or resuscitated SCD

- **Rhythm testing**

1. ECG
2. Ambulatory ECG
   * event recorder
   * implantable loop recorder
3. Cardiopulmonary exercise testing
4. Electrophysiologic study

- **Hemodynamic testing**

1. Echocardiography
2. Cardiac MRI
3. Cardiac catheterization/ angiography
   *coronary angiography

Recommendations

a. Noninvasive evaluation

- **Clinical history and physical examination**
- **Resting 12-lead ECG** (I, C)
- **Ambulatory ECG monitoring** (I, B)
- **Cardiac event loop recorders** (I, C)
- **Implantable loop recorders** ; syncope (I, B)
- **Cardiopulmonary exercise testing** : suspected exercise-induced arrhythmias (IIa, C)

b. Hemodynamic workup

- **Transthoracic or transesophageal echocardiography**
- **MRI or cardiac CT**
- **Coronary artery evaluation**: life-threatening VAs or resuscitated SCD in adults with CHD over 40 years of age, CHD associated with a higher risk of coronary ischemia

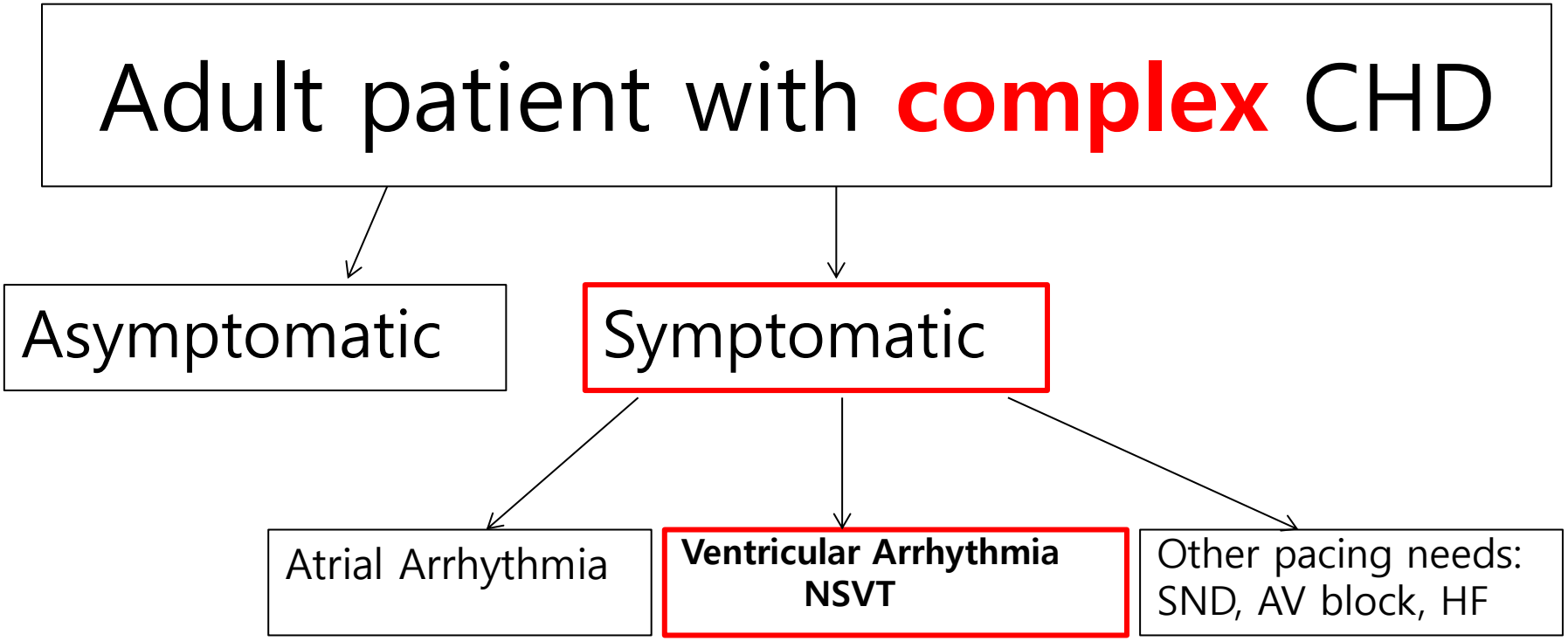
c. Electrophysiologic testing

- **EP study :** adults with unexplained syncope, 'high-risk" CHD substrates associated with primary VAs or poorly tolerated atrial tachyarrhythmias (I, C)

Class IIb	Electrophysiologic testing may be considered in adults with CHD and palpitations suggestive of sustained arrhythmia when the conventional diagnostic workup is unrevealing *(Level of evidence: C)*.[94]

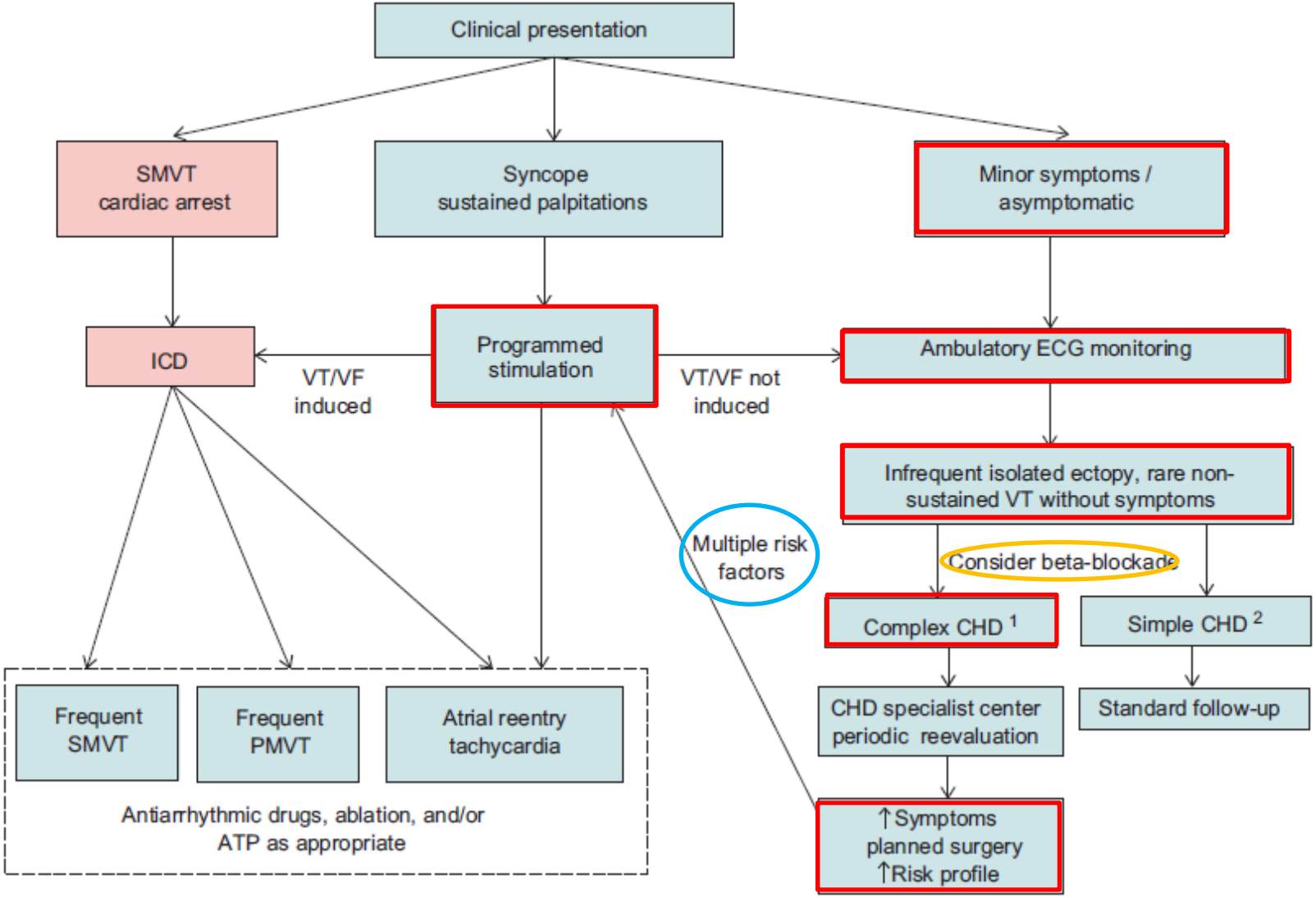
Complexity of CHD

Complexity of CHD	Type of CHD
Simple	PDA PS VSD secondum ASD
Moderate	**COA** **TAPVR** **AS** **Ebstein anomaly** **TOF** **primum ASD**
Severe	**Truncus arteriosus** **Pulmonary atresia** **DORV** **D-TGA** **L-TGA** **HLHS** **Other (heterotaxia, FSV)**


Adult patient with complex CHD
Asymptomatic
Symptomatic
Atrial Arrhythmia
Ventricular Arrhythmia
NSVT
Other pacing needs:
SND, AV block, HF

Management of VAs in adult CHD

EHRA/HRS/APHRS Expert Consensus on Ventricular Arrhythmias, 2014



Ventricular arrhythmias in Adult CHD

EHRA/HRS/APHRS Expert Consensus on Ventricular Arrhythmias, 2014

Expert consensus recommendations on VAs in CHD

(1) EP study : unexplained syncope and '**high-risk' CHD substrates associated with primary VAs** or poorly tolerated atrial tachyarrhythmias, such as **TOF, TGA with ASO, or significant systemic or single ventricular dysfunction** (I, C)

(2) In patients with CHD who have an implanted defibrillator and recurrent AHRV, VT storm, or multiple appropriate shocks, additional therapy including ATP, treatment with antiarrhythmic agents, and/or catheter ablation is indicated as adjunctive therapy to reduce the arrhythmia episodes. These therapies should be decided and initiated in an adequately trained centre. I LOE C

(3) In patients with CHD and sustained VAs who require surgical haemodynamic interventions, pre-operative electrophysiological testing and intra-operative ablation should be considered when adequate expertise is available. IIa LOE C

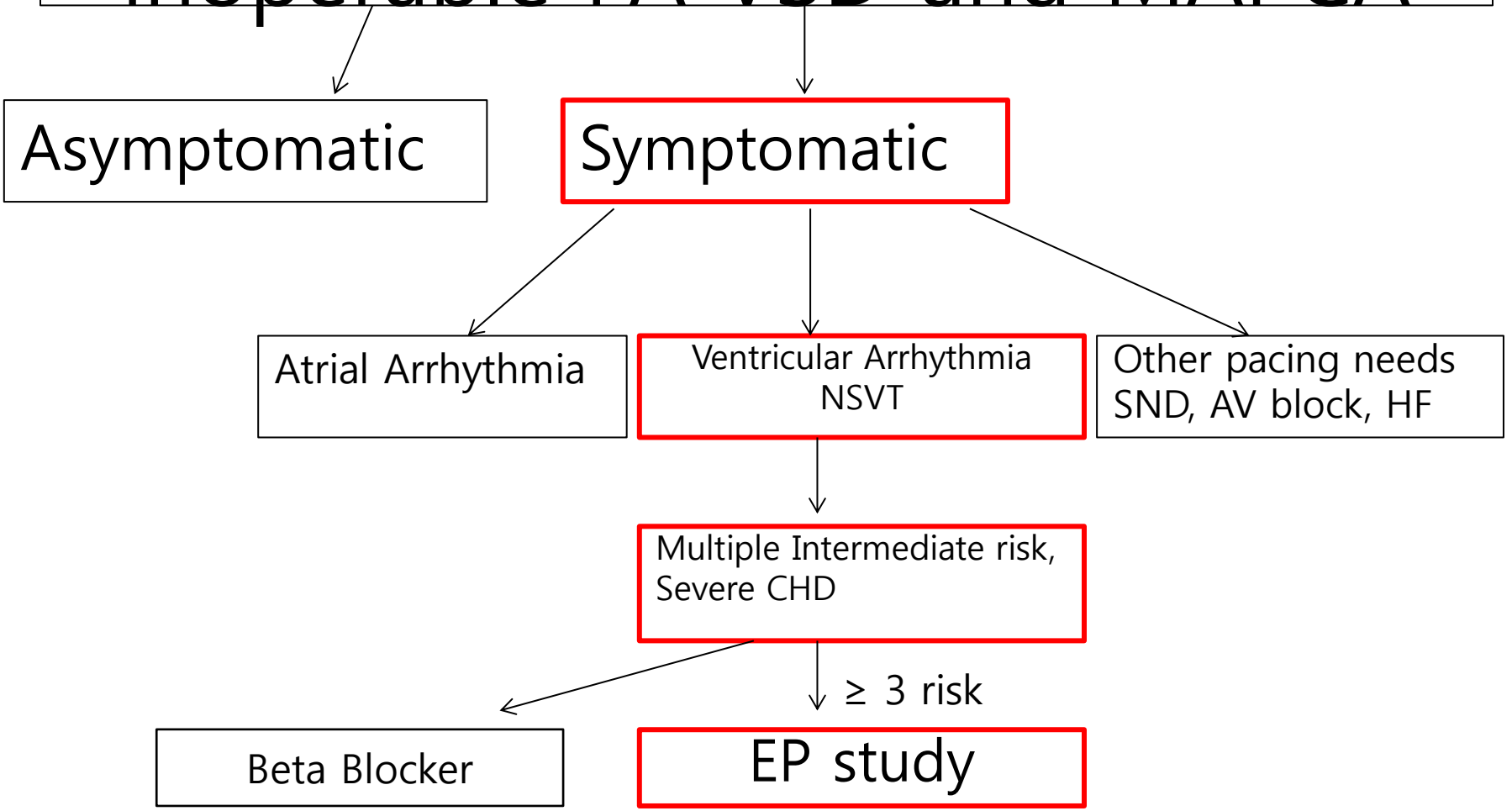
(4) Patients with good ventricular function, who are asymptomatic, have normal or near-normal ventricular haemodynamics and low-risk subtypes of CHD may reasonably be followed without advanced therapy and invasive evaluation despite the presence of moderately frequent and/or complex ventricular ectopy. IIb LOE C

(5) Catheter ablation may be appropriate for patients with CHD who have newly recognized or progressive ventricular dysfunction and a high burden of monomorphic ventricular ectopy. IIb LOE C

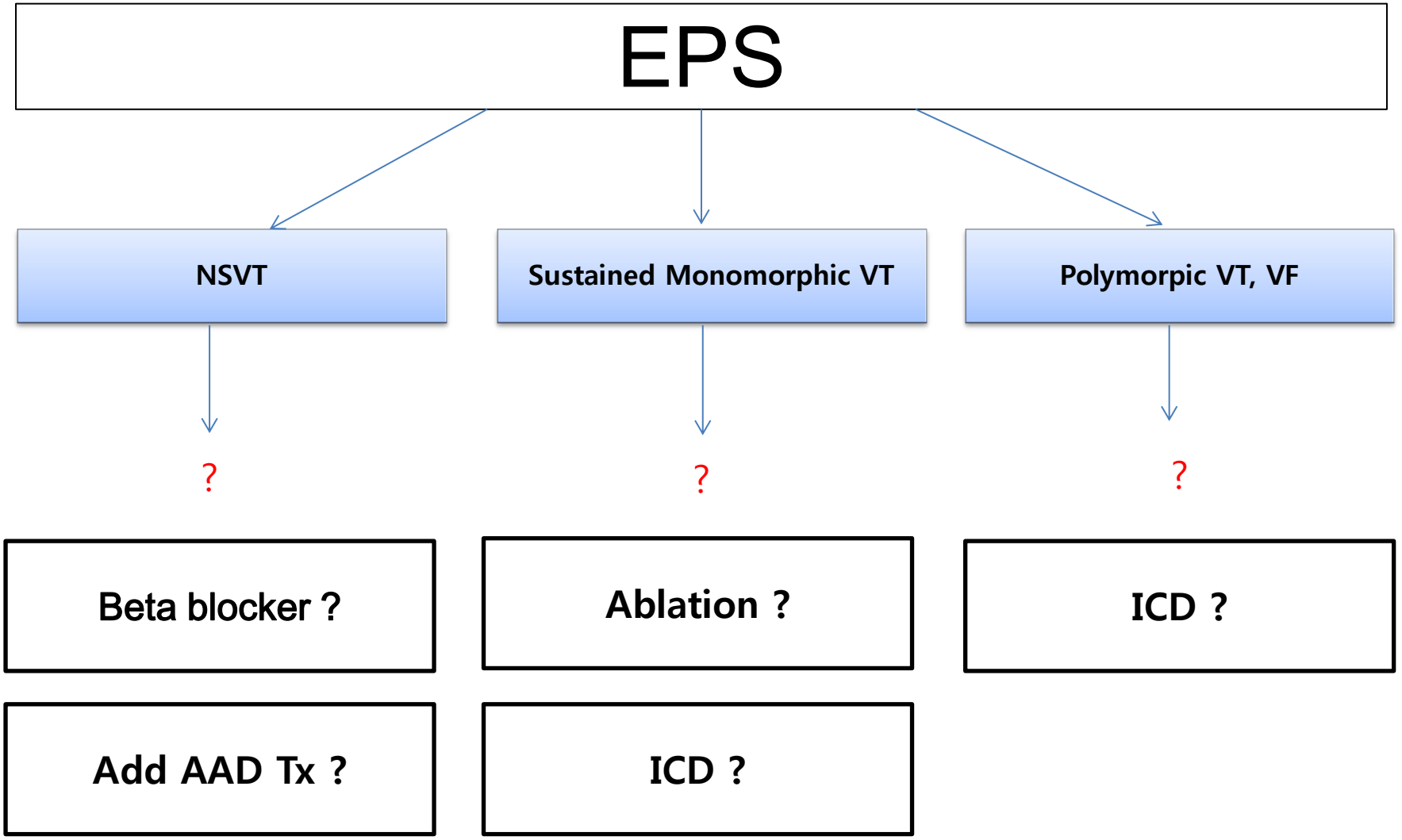
Clinical factors to consider EP study in CHD patient

	Findings	
Highest risk features	Sustained MMVT, Cardiac arrest	→ ICD Tx
High risk	Syncope, Sustained palpitations	→ Hemodynamic assess and EPS
Intermediate risk	Older age at initial repair (>1 year) Older age(>25 – 30 years) Prior palliative procedures Ventriculotomy RV haemodynamic burden : RVp >50% systemic , Moderate PR, RV EDV >150mL/m2, Increased heart size on CXR, RV function<45%, QRSd >180ms LV dysfunction(<55% for TOF) NSVT on monitoring Less clear Sx VO2 max <"20cc/kg/min T-wave alternans	→ If multiple risk, Beta blocker or EPS
Low risk	Simple 'repairs' without residual Expected or less isolated ectopy on monitoring No symptoms Good exercise capacity Good biventricular function	

Recurrent NSVT in patient with inoperable PA VSD and MAPCA



**no prospective data in CHD pts with VAs*


EPS
NSVT
Sustained Monomorphic VT
Polymorpic VT, VF
?
?
?
Beta blocker ?
Ablation ?
ICD ?
Add AAD Tx ?
ICD ?

Medical therapy

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult CHD, 2014

- Antiarrhythmic agents

Mexilletine, *JACC 1987* Phenytoin *AJC 1980, 1982*

Propafenone (?) → *CHF*

Sotalol → *QT prolongation*

Amiodarone → *lung fibrosis*

PVC suppression (Symptomatic improvement or reduction in ectopy)
≠ Mortality ↓

Catheter ablation

PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult CHD, 2014

Recommendations	
Class I	✓ Adjunctive therapy to an ICD in adults with CHD and recurrent monomorphic VT, a VT storm, or multiple appropriate shocks (I, C)
Class IIa	✓ Symptomatic sustained monomorphic VT in adults with CHD and ICDs (IIa,B)
Class IIb	✓ Adults with postop CHD and NSVT or hemodynamically poorly tolerated VT (IIb, C) ✓ Adult with CHD and frequent PVC (>15%) associated with deteriorating ventricular function (IIb, C).
Class III	1. Catheter ablation is not indicated for asymptomatic relatively infrequent ventricular ectopy in adults with CHD and stable ventricular function *(Level of evidence: C)*.[94] 2. Catheter ablation alone is not considered appropriate prophylactic therapy in adults with CHD deemed to be at increased risk for sudden cardiac death *(Level of evidence: C)*.[94]

ICD therapy in adult with CHD

Recommendations	
Class I	✓ Survivors of cardiac arrest due to VF or hemodynamically unstable VT (I,B). ✓ Spontaneous sustained VT (I,B) ✓ LV EF ≤35%, biventricular physiology, and NYHA class II or III symptoms (I,B).
Class IIa	✓ TOF and multiple risk factors for SCD, such as LV dysfunction, NSVT, QRS duration ≥180 ms, extensive RV scarring, or inducible sustained VT at EPS (IIa, B)
Class IIb	presence of additional risk factors such as complex ventricular arrhythmias, unexplained syncope, NYHA functional class II or III symptoms, QRS duration ≥140 ms, or severe systemic AV valve regurgitation *(Level of evidence: C)*.[45–48,435,467] 2. ICD therapy may be considered in adults with CHD and a *systemic ventricular ejection fraction* <35% in the absence of overt symptoms (NYHA class I) or other known risk factors *(Level of evidence of: C)*.[36,97,467] 3. ICD therapy may be considered in adults with CHD and *syncope of unknown origin* with hemodynamically significant sustained ventricular tachycardia or fibrillation inducible at electrophysiologic study *(Level of evidence: B)*.[76,97,436] 4. ICD therapy may be considered for nonhospitalized adults with CHD *awaiting heart transplantation (Level of evidence: C)*.[97,468] 5. ICD therapy may be considered for adults with syncope and moderate or complex CHD in whom there is a high clinical suspicion of ventricular arrhythmia and in whom thorough invasive and noninvasive investigations have failed to define a cause *(Level of evidence: C)*.[97,469]
Class III	1. All Class III recommendations listed in current ACC/AHA/HRS guidelines apply to adults with CHD *(Level of evidence: C)*.[97] These include: a. Life expectancy with an acceptable functional status <1 year; b. Incessant ventricular tachycardia or ventricular fibrillation; c. Significant psychiatric illness that may be aggravated by ICD implantation or preclude systematic follow-up; d. Patients with drug-refractory NYHA class IV symptoms who are not candidates for cardiac transplantation or cardiac resynchronization therapy. 2. Adults with CHD and advanced pulmonary vascular disease (Eisenmenger syndrome) are generally not considered candidates for ICD therapy *(Level of evidence: B)*.[470,471] 3. Endocardial leads are generally avoided in adults with CHD and intracardiac shunts. Risk assessment regarding hemodynamic circumstances, concomitant anticoagulation, shunt closure prior to endocardial lead placement, or alternative approaches for lead access should be individualized *(Level of Evidence: B)*.[8,54,182]

Further assessment prior to Tx

- NYHA class
- Echo : LV function, RV function
- ECG: QRS duration
- Holter : PVC burden
- Exercise test
- Cardiac MRI : extent of RV fibrosis

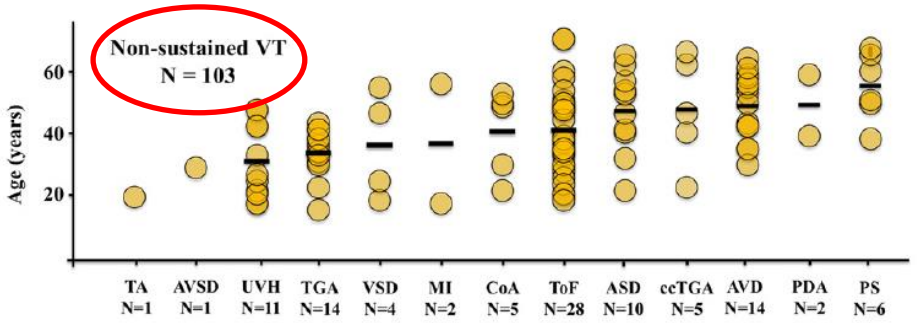
Non-sustained ventricular tachycardia in patients with congenital heart disease: An important sign?

Inter J Cardiol 2016

145 CHD Pts, 134 corrective/palliative op, 11 no op

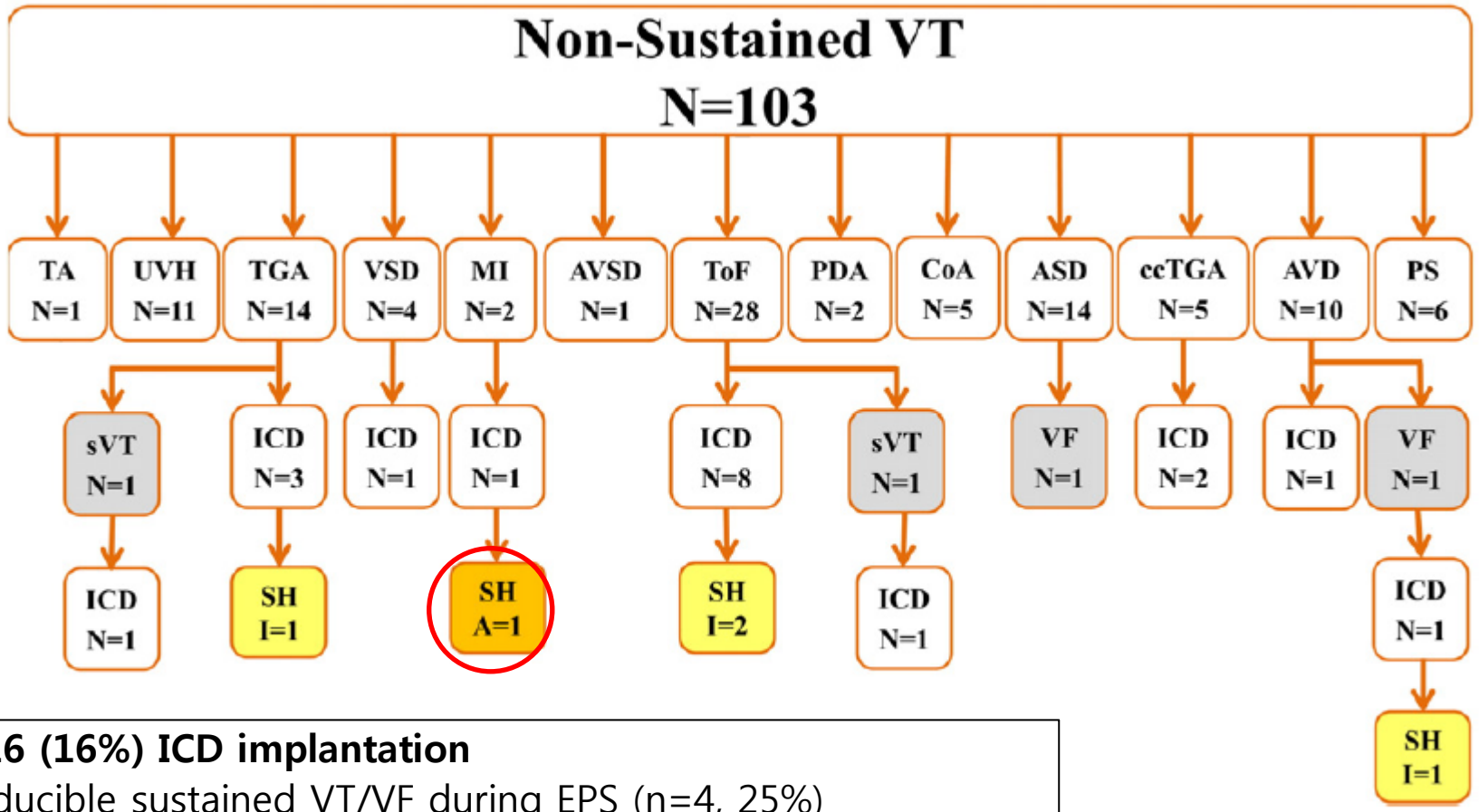
Age first procedure: 12 ± 16 (0–70) yrs

Age at VAs: 40 ± 14 (15–70) yrs



	All N = 145	NSVT N = 103	susVT N = 25	VF N = 17
Echocardiography, N (%)	122 (84)	89 (86)	22 (88)	11 (65)
Impaired ventricular function (%)	14 (11)	8 (9)	2 (9)	4 (36)
QRS, N (%)	97 (67)	80 (78)	12 (28)	5 (29)
QRS duration, mean ± SD	134 ± 32	129 ± 29*	157 ± 35*	149 ± 28
Prolonged, N (%)	58 (60)	43 (54)	10 (83)	4 (80)
ToF ≥ 180 ms, N (%)	5 (19)	2 (10)	3 (50)	0
TGA ≥ 140 ms, N (%)	5 (31)	4 (33)	1 (25)	0

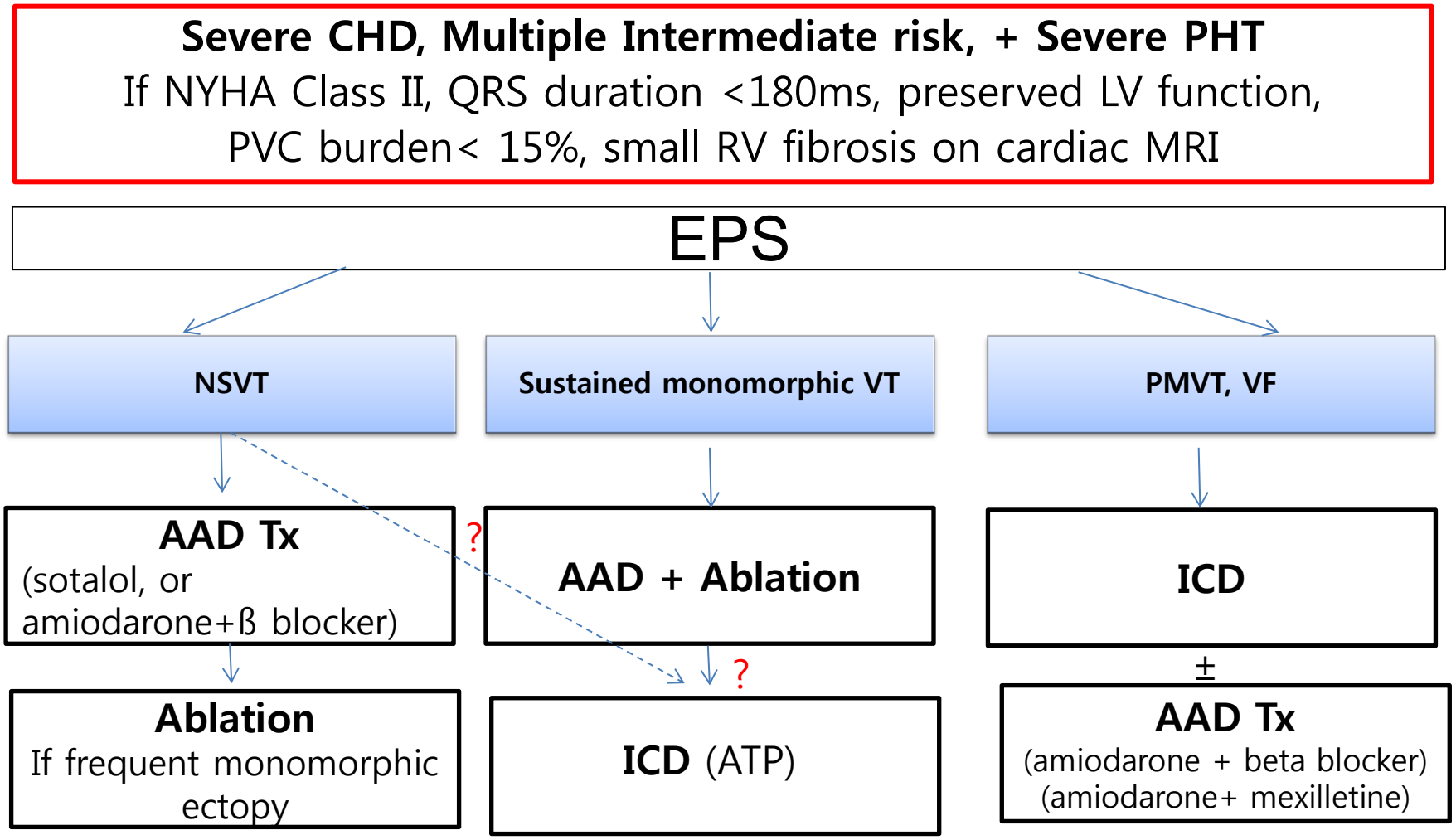
FU median period 5 yrs (range: 0-27)



- **16 (16%) ICD implantation**
  - inducible sustained VT/VF during EPS (n=4, 25%)
  - severe decreased ventricular function and NSVT (n=3, 19%)
  - symptomatic NSVT (n=2, 13%)
  - unknown (n=7)

I suggest …

Symptomatic, recurrent NSVT in adult patient with inoperable PA VSD and MAPCA



Thank you for attention !