

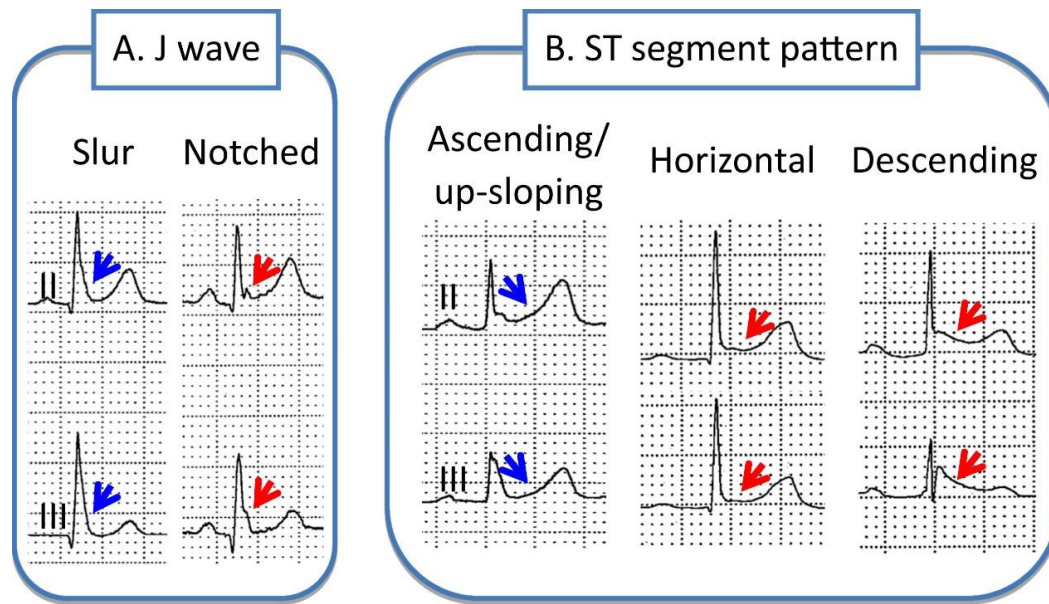


Figure 1.

Different manifestations of early repolarization. **A:** The J wave may be distinct or appear as a slur. In the latter case, part of the J wave is buried inside the QRS, resulting in an elevation of J_o . Patients with a distinct J wave have a worse prognosis than do patients with a slurred J wave. **B:** The ST segment may be upsloping, horizontal, or descending. Horizontal and descending ST segments are associated with a worse prognosis.

Antzelevitch C, et al. J-Wave syndromes expert consensus conference report: Emerging concepts and gaps in knowledge. *Europace*. 2017;19:665-694.

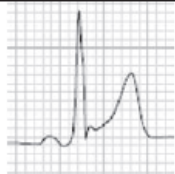
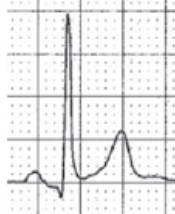
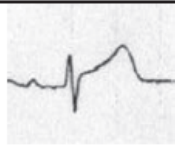
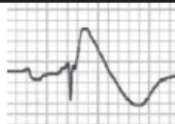
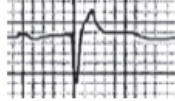
Early Repolarization Patterns	Description	Example
Terminal QRS Notching	Low frequency deflection at the end of the QRS.	
Terminal QRS Slurring	An abrupt change in the slope of the last deflection at the end of the QRS.	
ST elevation	Elevation of the ST segment above the isoelectric baseline.	
Other Repolarization Patterns	Description	Example
Brugada Type 1	Downward coved ST segment elevation, usually in V1-V3.	
Brugada Type 2	ST segment elevation with a saddleback appearance, usually	
Epsilon Wave	Low frequency, positive terminal deflection associated with ARVC, usually in V1-V3.	

Figure 2. Examples of early repolarization and other repolarization electrocardiographic patterns.

ARVC indicates arrhythmogenic right ventricular cardiomyopathy.

Electrocardiographic Early Repolarization: A Scientific Statement From the American Heart Association. *Circulation* 2016;133:1520-1529

Table 1. Diagnosis**ESC 2022 GL on VA and SCD. Recommendations for the management of patients with early repolarization pattern/syndrome****Diagnosis**

ERP is diagnosed as J-point elevation of ≥ 1 mm in two adjacent inferior and/or lateral ECG leads.	I C
ERS is diagnosed in a patient resuscitated from unexplained VF/PVT in the presence of ERP.	I C
In an SCD victim with a negative autopsy and medical chart review, and an antemortem ECG demonstrating the ERP, ERS should be considered.	IIa C
Clinical evaluation for ERP with additional high-risk features ^a for first-degree relatives of ERS patients.	IIa B
Genetic testing in ERS patients	IIb C
Clinical evaluation is not recommended routinely in asymptomatic subjects with ERP.	III C

^a ERP high-risk features: J waves > 2 mm, dynamic changes in J point and ST morphology.

Table 2. APHRS/EHRA/HRS/SOLAECE 2016 consensus report on J-wave syndromes. Differential diagnosis of early repolarization pattern**Other causes of early repolarization pattern include the following:**

- Juvenile ST pattern
- Pericardial disease (pericarditis, pericardial cyst, pericardial tumor)
- Hypothermia
- Hyperthermia
- Myocardial tumor (lipoma)
- Hypertensive heart disease
- Athlete's heart
- Myocardial ischemia
- STEMI (i.e., anteroapical myocardial infarction)
- Fragmented QRS (terminal notching)
- Hypocalcemia
- Hyperkalemia
- Thymoma
- Aortic dissection
- Arrhythmogenic right ventricular cardiomyopathy
- Takotsubo cardiomyopathy
- Neurologic causes (intracerebral bleeding, acute brain injury)
- Myocarditis
- Chagas disease
- Cocaine use

Antzelevitch C, et al. J-Wave syndromes expert consensus conference report: Emerging concepts and gaps in knowledge. *Europace*. 2017;19:665-694.

Table 3. Genetic Testing**ESC 2022 GL on VA and SCD. Recommendations for genetic testing (Class IIb)****Proband*****Initial clinical test*****Cornerstone for diagnosis**

ECG

Other tests/processes

Holter

Echocardiography

Follow-up

1-3 years dependent on level of risk

Relatives***Clinical screening***

Holter

Echocardiogram

Follow-up

Positive phenotype and/or Class IV/V variant:

1-3 years dependent on level of risk

Negative phenotype and no Class IV/V variant:

Discharge

EHRA/HRS/APHRS/LAHRs 2022 Consensus Statement on genetic testing.**Impact of genetic testing for the index case**

Disease	Diagnostic	Prognostic	Therapeutic
Early repolarization syndrome	—	—	—

Recommendations**May do**

Molecular genetic testing in unexplained cardiac arrest survivors diagnosed clinically with ERS.

Do not do

In asymptomatic individuals with only an ECG-based early repolarization pattern, genetic testing should not be performed.

Table 4. Therapy of ERS**AHA/ACC/HRS 2017 GL on VA/SCD. ERS**

Observation without treatment of asymptomatic patients with an early repolarization pattern on ECG.	I-B-NR
ICD in patients with early repolarization pattern on ECG and cardiac arrest or sustained VA.	I-B-NR
In patients with early repolarization pattern on ECG, genetic testing is not recommended.	III-B-NR No benefit

ACC/AHA/HRS 2017 GL on syncope. Early repolarization pattern

ICD in patients with early repolarization pattern and suspected arrhythmic syncope in the presence of a family history of early repolarization pattern with cardiac arrest.	IIB-C-EO
EPS should not be performed in patients with early repolarization pattern and history of syncope in the absence of other indications	III-B-NR (Harm)

ESC 2022 GL on VA and SCD. Recommendations for the management of patients with early repolarization pattern/syndrome***Risk stratification, prevention of SCD and treatment of VA***

ICD implantation in patients with a diagnosis of ERS who have survived a CA	I B
Isoproterenol infusion for ERS patients with electrical storm	IIa B
Quinidine in addition to an ICD for recurrent VF in ERS patients	IIa B
ILR in individuals with ERP and at least one risk featured or arrhythmic syncope	IIa C
PVC ablation in ERS patients with recurrent VF episodes triggered by a similar PVC non-responsive to medical treatment	IIa C
ICD implantation or quinidine in individuals with ERP and arrhythmic syncope and additional risk features. ^b	IIb C
ICD implantation or quinidine in asymptomatic individuals who demonstrate a high-risk ERP ^a in the presence of a family history of unexplained juvenile SD	IIb C
ICD implantation is not recommended in asymptomatic patients with an isolated ERP	III C

CA, cardiac arrest; ECG, electrocardiogram; ERP, early repolarization pattern; ERS, early repolarization syndrome; ICD, implantable cardioverter defibrillator; ILR, implantable loop recorder; PVC, premature ventricular complexes; PVT, ventricular tachycardia; SCD, sudden cardiac death; SD, sudden death; VA, ventricular arrhythmia; VF, ventricular fibrillation.

^a ERP high-risk features: J waves >2 mm, dynamic changes in J point and ST

morphology.

^b High-risk ERP: family history of unexplained SD <40 years, family history of ERS.

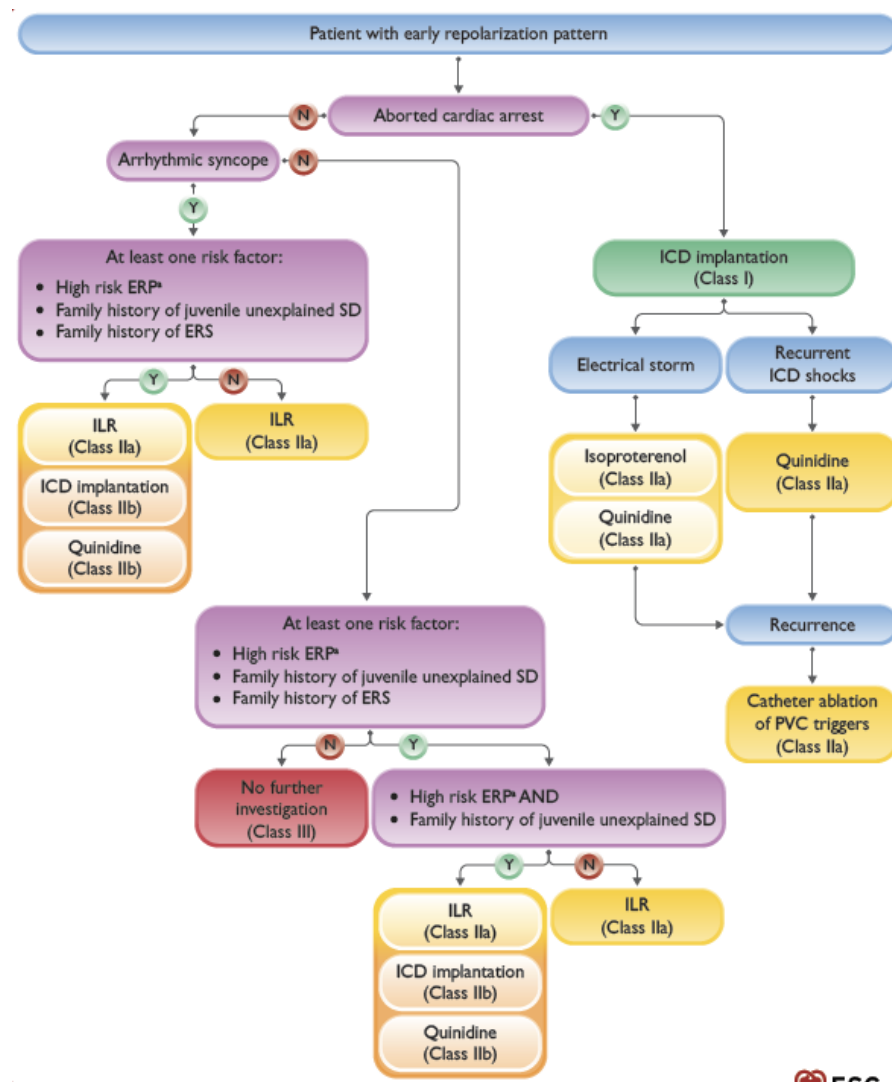


Figure 3. ESC 2022 GL on VA and SCD. Management of patients with early repolarization pattern/syndrome.

ERP, early repolarization pattern; ERS, early repolarization syndrome; ICD, implantable cardioverter defibrillator; ILR, implantable loop recorder; N, No; PVC, premature ventricular complex; SD, sudden death; Y, Yes.

^a ERP high risk features: J waves .2 mm, dynamic changes in ST morphology.