2026
해부학 · 선천성 · 유전성
부정맥 연구회
합동 심포지엄

일시 2026년 9월 5일(토) 08:50-16:00

장소 삼성서울병원 히포크라테스홀

대한부정맥학회
Korean Heart Rhythm Society

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PROGRAM

08:50~08:55	개회사	허준 (선천성-유전성부정맥연구회 회장)
08:55~09:00	개회사	박상원 (심장부정맥해부학연구회 회장)
09:00~10:20	**Session 1. Heart Anatomy; Normal and Their Congenital Defects**	
	좌장: **서정욱** (인천세종병원 병리과), **박상원** (부천세종병원 심장내과) 패널: **이주원** (서울의대 소아청소년과), **곽신형** (성균관의대 소아청소년과), **박소윤** (가톨릭의대 순환기내과), **김예지** (이화의대 심장내과)	
09:00~09:15	Anatomy of the atria and congenital heart disease (ASD, PAPVR, TAPVR)	**윤자경** (성균관의대 소아청소년과) ····· 2
09:15~09:30	Anatomy of the ventricles and congenital heart disease (VSD, TOF)	**박일근** (성균관의대 심장혈관외과) ····· 4
09:30~09:45	Anatomy of great vessels and congenital heart disease (TGA, DORV)	**곽재건** (서울의대 소아흉부외과) ····· 6
09:45~10:00	Anatomy of isomerism	**백승민** (서울의대 소아청소년과) ····· 7
10:00~10:20	Discussion	
10:20~10:40	Break	
10:40~12:00	**Session 2. Long and Short QT Syndromes; Stop the Short-Long-Short Sequence!**	
	좌장: **배은정** (서울의대 소아청소년과), **조용근** (경북의대 순환기내과) 패널: **양소영** (가천의대 심장내과), **안효정** (서울의대 순환기내과), **정주희** (고려의대 순환기내과), **차누리** (연세의대 소아청소년과)	
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11:25~11:40	Short QT syndrome; Short but not least	**김주연** (성균관의대 순환기내과) ····· 53
11:40~12:00	Discussion	
12:00~13:00	Lunch	
13:00~14:20	**Session 3. Deep Dive into Early Repolarization Syndrome**	
	좌장: **남기병** (울산의대 심장내과), **최종일** (고려의대 순환기내과) 패널: **김명중** (가톨릭의대 순환기내과), **한석문** (서울의대 순환기내과), **황태현** (연세의대 심장내과), **이경연** (고려의대 순환기내과)	
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14:20~14:40	Break	
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	좌장: **허준** (성균관의대 소아청소년과), **오세일** (서울의대 순환기내과) 패널: **이주성** (고려의대 소아청소년과), **최인수** (전남의대 소아청소년과), **이영신** (경희의대 심장내과), **이형석** (인하의대 심장내과)	
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Session 1

Heart Anatomy; Normal and Their Congenital Defects

좌장: 서정욱 (인천세종병원 병리과), 박상원 (부천세종병원 심장내과)
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Anatomy of the atria and congenital heart disease (ASD, PAPVR, TAPVR)
윤자경 (성균관의대 소아청소년과)

Anatomy of the ventricles and congenital heart disease (VSD, TOF)
박일근 (성균관의대 심장혈관외과)

Anatomy of great vessels and congenital heart disease (TGA, DORV)
곽재건 (서울의대 소아흉부외과)

Anatomy of isomerism
백승민 (서울의대 소아청소년과)

대한부정맥학회
Korean Heart Rhythm Society

Anatomy of the atria and congenital heart disease (ASD, PAPVR, TAPVR)

윤 자 경

성균관의대 소아청소년과

The interatrial septum and pulmonary venous system are among the most anatomically intricate structures of the developing heart, and a clear understanding of their embryology is fundamental to the diagnosis and management of the most common congenital cardiac anomalies. This presentation provides an integrated review of atrial and pulmonary venous anatomy, tracing normal development from the primitive heart tube through to the clinically relevant anatomy encountered in modern imaging and intervention.

Atrial septation begins with looping of the primitive cardiac tube and expansion of its venous inlet pole to form the atria, into which the horns of the sinus venosus drain asymmetrically before being incorporated into the developing right atrium. Around the fourth week of gestation, the septum primum arises as a crescentic fold that grows toward the fusing endocardial cushions, transiently leaving the ostium primum before this closes and the septum primum fenestrates to form the ostium secundum. The septum secundum subsequently develops as an infolding of the atrial roof, overlapping the septum primum to create the floor of the oval fossa — the substrate for the foramen ovale, which functionally closes after birth as left atrial pressure rises. Failure of adequate valve-rim overlap results in a patent foramen ovale, found in up to a third of the population, while true deficiency of septal tissue produces the secundum atrial septal defect (ASD), the most common form of interatrial communication. We review the clinical anatomy of the true atrial septum — emphasizing that much of the so-called “septal” rim seen on imaging is in fact infolded atrial wall (Waterston's/Sondergaard's groove) rather than true septal tissue — along with its proximity to critical structures including the aortic root, atrioventricular node, coronary sinus, and mitral and tricuspid valves, which has direct implications for the safety of transseptal puncture and device or surgical closure. Beyond the true septum, we discuss superior and inferior sinus venosus defects, coronary sinus

(unroofed coronary sinus) defects, and ostium primum defects as interatrial communications lying outside the confines of the oval fossa, each with distinct morphology, embryologic origin, and surgical considerations.

The second half of the talk turns to the pulmonary veins, whose embryologic origin has historically been debated. Contrary to older teaching that the pulmonary veins arise from the systemic venous sinus, current molecular and morphologic evidence indicates that the pulmonary vein is an entirely new structure, canalizing independently from mediastinal (second heart field) myocardium and opening initially as a solitary orifice adjacent to the atrioventricular junction before remodeling — uniquely in the human heart — to form four separate venous connections at the corners of the left atrial roof. This distinct origin explains why the myocardial sleeves extending onto the pulmonary veins are electrically distinct from adjacent atrial tissue and why they serve as the dominant source of ectopic triggers for atrial fibrillation, underpinning the rationale for pulmonary vein isolation as first-line ablation therapy for Atrial fibrillation ablation. We review the normal pulmonary venous anatomy (right and left superior and inferior veins found in 60–70% of individuals) and its common variants, before addressing anomalous pulmonary venous drainage (APVD). Partial anomalous pulmonary venous return (PAPVR) — most commonly the right superior pulmonary vein draining into the superior vena cava — is frequently associated with a sinus venosus ASD and produces a left-to-right shunt with right heart volume loading. Total anomalous pulmonary venous return (TAPVR) is classified into supracardiac, cardiac, infracardiac, and mixed types based on the level of anomalous connection, and typically presents in the neonatal period with cyanosis and heart failure requiring urgent surgical correction. Related entities, including scimitar syndrome and cor triatriatum, are briefly discussed. Throughout, we highlight the essential role of cross-sectional imaging (echocardiography, cardiac CT, and cardiac MRI) in delineating this anatomy — for pre-procedural planning of ASD closure and transseptal access, for surgical planning in PAPVR/TAPVR, and for mapping pulmonary venous anatomy prior to catheter ablation of atrial fibrillation.

By synthesizing developmental anatomy with its clinical and imaging correlates, this presentation aims to provide cardiologists, imaging specialists, electrophysiologists, and surgeons with a practical and unified framework for understanding and managing this spectrum of congenital atrial and pulmonary venous anomalies.

Anatomy of the ventricles and congenital heart disease (VSD, TOF)

박 일 근

성균관의대 심장혈관외과

Background: Successful repair of congenital heart defects requires an understanding of ventricular anatomy that remains valid despite abnormal cardiac position, connections, and chamber morphology. A systematic morphologic approach can clarify the relationships among the ventricular components, ventricular septal defects (VSDs), the cardiac conduction axis, and the right ventricular outflow tract.

Methods: This educational review integrates established surgical-anatomic concepts and morphologic specimens to describe the ventricles, classify VSDs, and interpret the anatomy of tetralogy of Fallot (TOF). Particular emphasis is placed on structures that determine operative exposure, patch placement, conduction-system risk, and the safe extent of muscular resection.

Results: Both ventricles can be analyzed in terms of inlet, apical trabecular, and outlet components. When conventional positional features are unreliable, the coarse apical trabeculations of the morphologically right ventricle and the fine trabeculations of the morphologically left ventricle provide the most dependable distinction. Within the right ventricle, the septomarginal trabeculation and its limbs form the principal framework for identifying the outlet septum, medial papillary muscle, moderator band, and conduction axis.

VSDs should be described using three complementary features: the phenotype of their borders, the ventricular component into which they open, and the presence or absence of septal malalignment. The border phenotype - muscular, perimembranous, or doubly committed and juxta-arterial - is the most important predictor of the conduction axis. In perimembranous defects, the conduction axis typically courses along the posteroinferior margin, whereas in muscular inlet defects it lies anterosuperiorly. Straddling of the tricuspid valve remains an important exception in which the conduction pathway cannot be predicted by standard landmarks.

In TOF, anterocephalad deviation of the outlet septum provides a unifying explanation for the VSD,

overriding aorta, and subpulmonary obstruction. Operative repair requires superficial suturing on the right ventricular aspect of the septal crest because an unseen atrioventricular bundle may branch astride the septum. Relief of obstruction may include resection of the septal insertion of the outlet septum and septoparietal trabeculations, while protecting the right coronary artery, aortic valve leaflets, and tricuspid papillary muscle.

Conclusion: A morphology-based framework links normal ventricular anatomy directly to VSD and TOF repair. Reading the defect border first, identifying reliable landmarks, and distinguishing septal from parietal structures can improve operative planning and reduce injury to the conduction system, coronary arteries, and arterial valves.

Keywords: ventricular anatomy; ventricular septal defect; tetralogy of Fallot; conduction axis; right ventricular outflow tract; surgical anatomy

Anatomy of great vessels and congenital heart disease (TGA, DORV)

곽 재 건

서울의대 소아흉부외과

Double outlet right ventricle (DORV) and transposition of the great arteries (TGA) represent two of the most anatomically and physiologically complex conotruncal malformations encountered in congenital cardiac surgery, and together they illustrate the spectrum of ventriculo-arterial connection abnormalities that determine surgical strategy in the modern era.

DORV is characterized by both great arteries arising entirely or predominantly (>50%) from the right ventricle, with the VSD serving as the effectively obligatory outlet for the left ventricle. The position of the VSD, VSD-great vessel relationship (subaortic, subpulmonary/Taussig-Bing, doubly committed, or non-committed), together with the presence or absence of pulmonary stenosis, dictate whether the lesion behaves physiologically like a VSD, tetralogy of Fallot, or TGA, and correspondingly determines the surgical approach: intraventricular baffling of the left ventricle to the aorta, baffling with right ventricular outflow tract reconstruction or RV-to-PA conduit, arterial switch operation with VSD closure (Taussig-Bing type), Rastelli-type repair, or aortic translocation (Nikaidoh procedure).

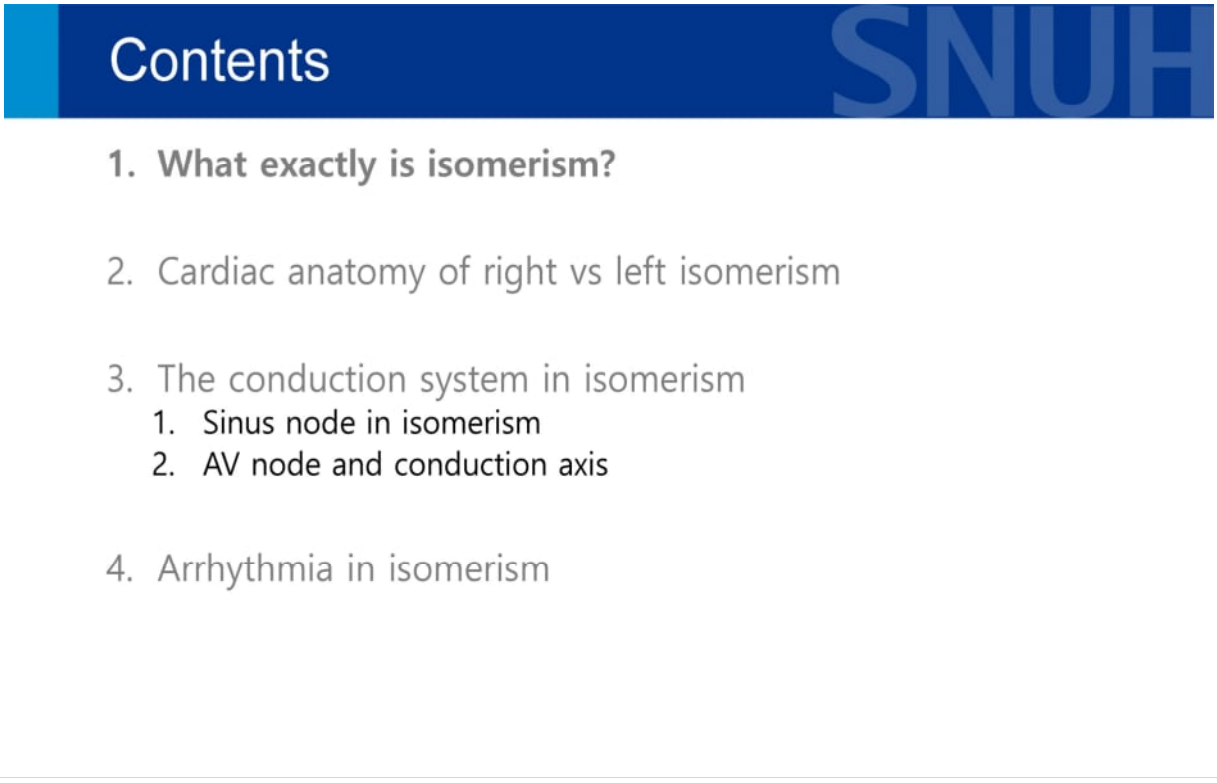
d-TGA is defined by the combination of atrio-ventricular (AV) concordance and ventriculo-arterial (VA) discordance—the aorta arising from the morphologic right ventricle and the pulmonary artery from the morphologic left ventricle—resulting in a parallel rather than the normal spiral arrangement of the great vessels. Whereas, l-TGA (congenitally corrected TGA, ccTGA) represents a double discordance—both AV and VA discordance—resulting in a distinct anatomic arrangement in which the morphologic right ventricle supports the systemic circulation and the morphologic left ventricle supports the pulmonary circulation.

In this presentation, we will review the key anatomic features of these two conditions using schematic drawings.

Anatomy of isomerism

백 승 민

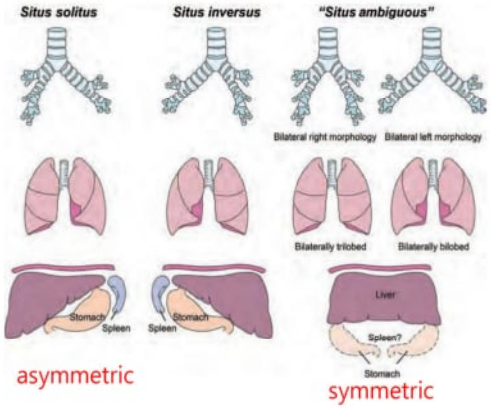
서울의대 소아청소년과



Isomerism

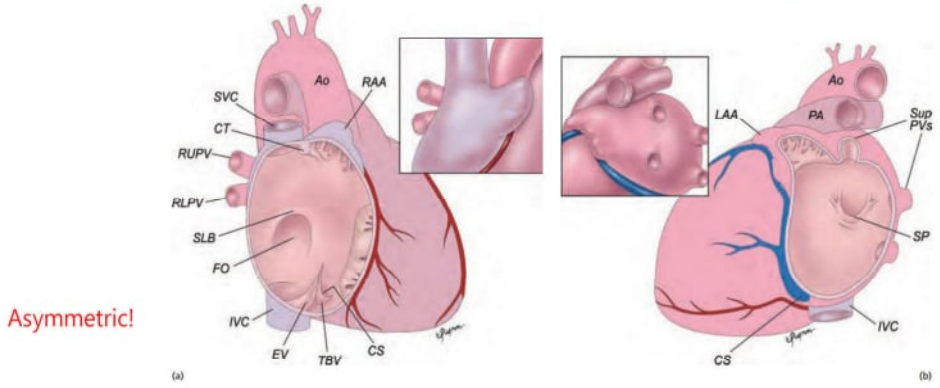
SNUH

- **Symmetrical development** of anatomical structures that are usually **asymmetric.**
  - liver, lungs, bronchi, and atria.

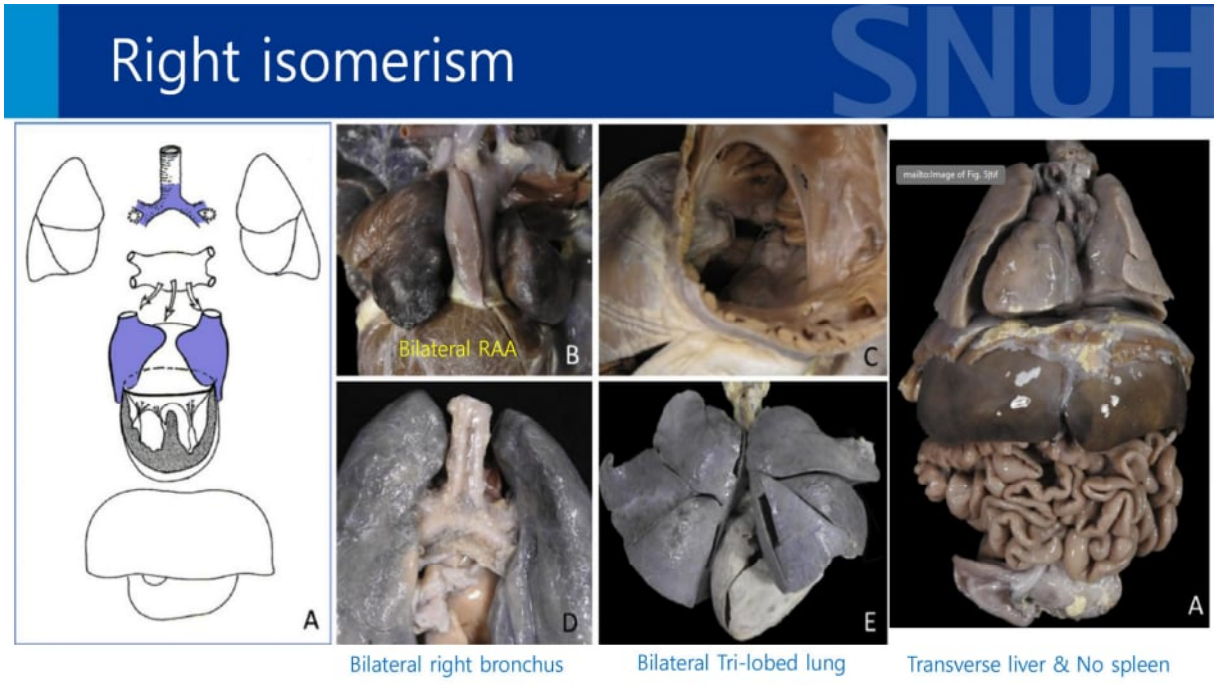


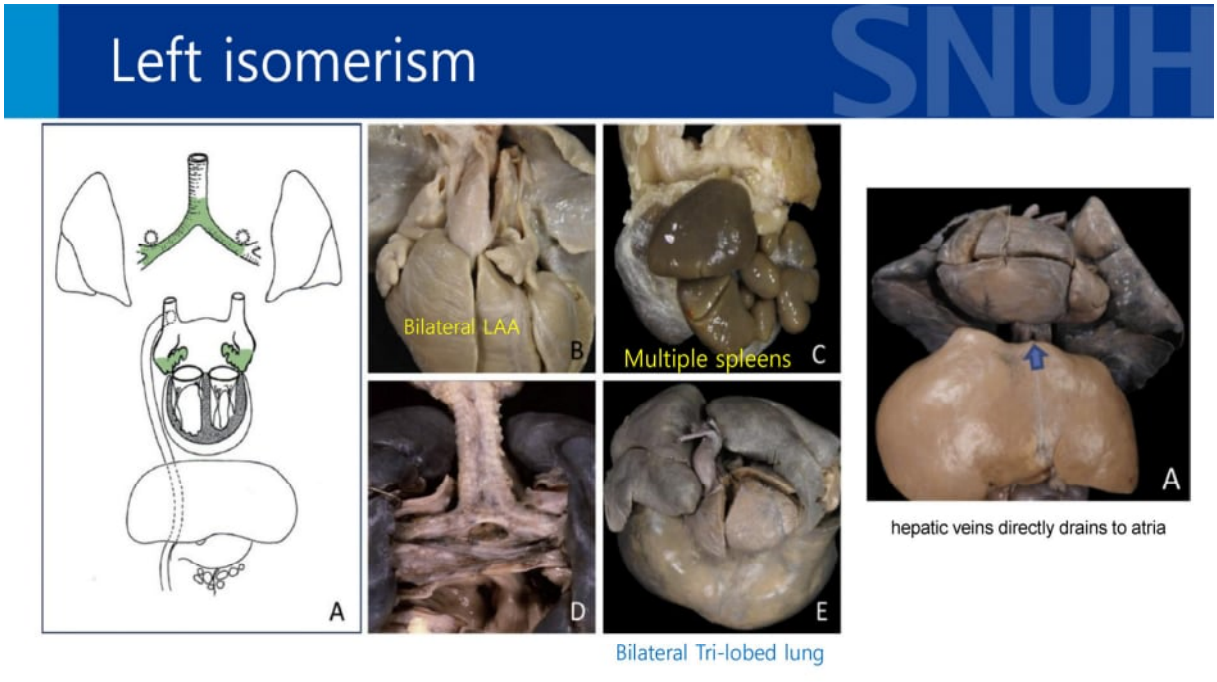
Normal atrial anatomy

SNUH



	Right	Left
Myocardial features	Crista terminalis, tinea sagittalis, extension of pectinate muscles toward AV valve	Pectinate muscles confined to appendage No Crista terminalis
Appendage	Broad-based, triangular	Long and narrow (finger-like)
Septum	Septum secundum (limbus of the fossa ovale)	Septum primum (valve of the foramen ovale)


Right isomerism
SNUH
A
Bilateral RAA
B
C
D
E
A
Bilateral right bronchus
Bilateral Tri-lobed lung
Transverse liver & No spleen


Left isomerism
SNUH
A
Bilateral LAA
B
Multiple spleens
C
D
E
A
Bilateral Tri-lobed lung
hepatic veins directly drains to atria

Terminology

SNUH

- Right isomerism, Left isomerism??

  Right isomerism = isomerism of RAA
  Left isomerism = isomerism of LAA

- Asplenia, Polysplenia??

  Right isomerism: frequently, ***but not always*** associated with asplenia
  Left isomerism: mostly, ***but not always*** associated with polysplenia

- Situs ambiguous??

  Not ambiguous but "recognizable" as bilaterally left or right.

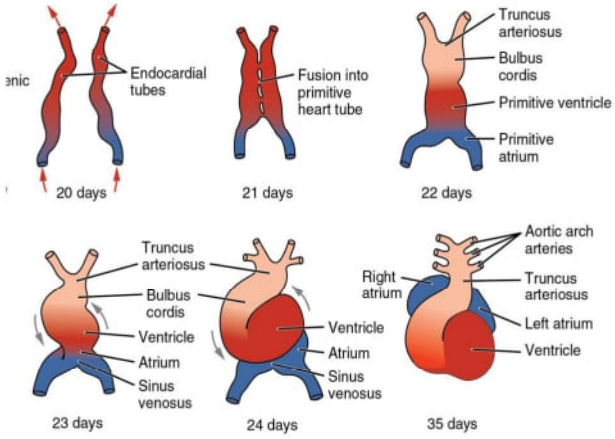
- Heterotaxy syndrome??

  failure in development of 'normal asymmetry'
  Heterotaxy syndrome ∋ right/left isomerism, polysplenia/asplenia, mixed form

Why atria?

SNUH

- No evidence of ventricular/arterial isomerism.
  - LV/RV is 'proximal → distal' structure, not left vs right
  - Truncal septation occurs in different stage (not right & left endocardial tuve)



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SNUH

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2. **Cardiac anatomy of right vs left isomerism**
3. The conduction system in isomerism
   1. Sinus node in isomerism
   2. AV node and conduction axis
4. Arrhythmia in isomerism

1. Venous drainage

SNUH

Anomalies	Details	Isomerism of RAA	Isomerism of LAA
Systemic venous drainage	Bilateral SVC	**24 (53%)**	**14 (70%)**
	Single right SVC	16 (35%)	3 (15%)
	Single left SVC	5 (11%)	3 (15%)
	Absent CS	45 (100%)	–
	Interrupted IVC	2 (4%)	**15 (75%)**
Pulmonary venous drainage	Symmetric PV drainage	–	13 (65%)
	PVs to right-sided atrium	10 (22%)	3 (15%)
	PVs to left-sided atrium	6 (13%)	4 (20%)
	PVs to left SVC	7 (15%)	–
	PVs to right SVC	15 (33%)	–
	PVs to portal system	7 (15%)	–

Absent CS is pathognomonic

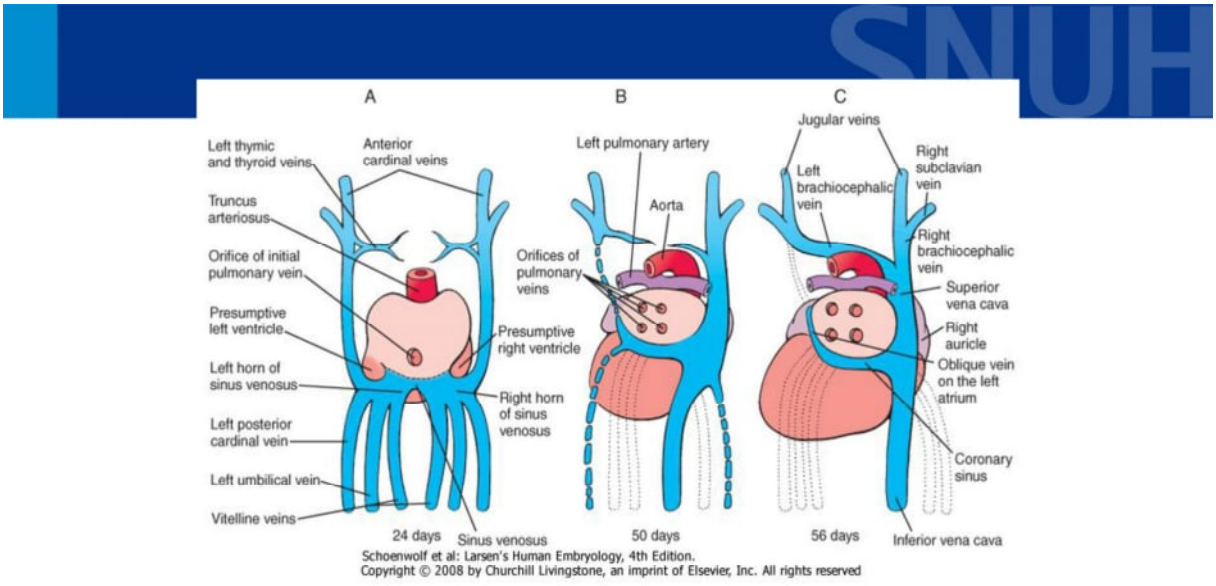
2PVs to left sided atrium
2PVs to right sided atrium

Cardiac

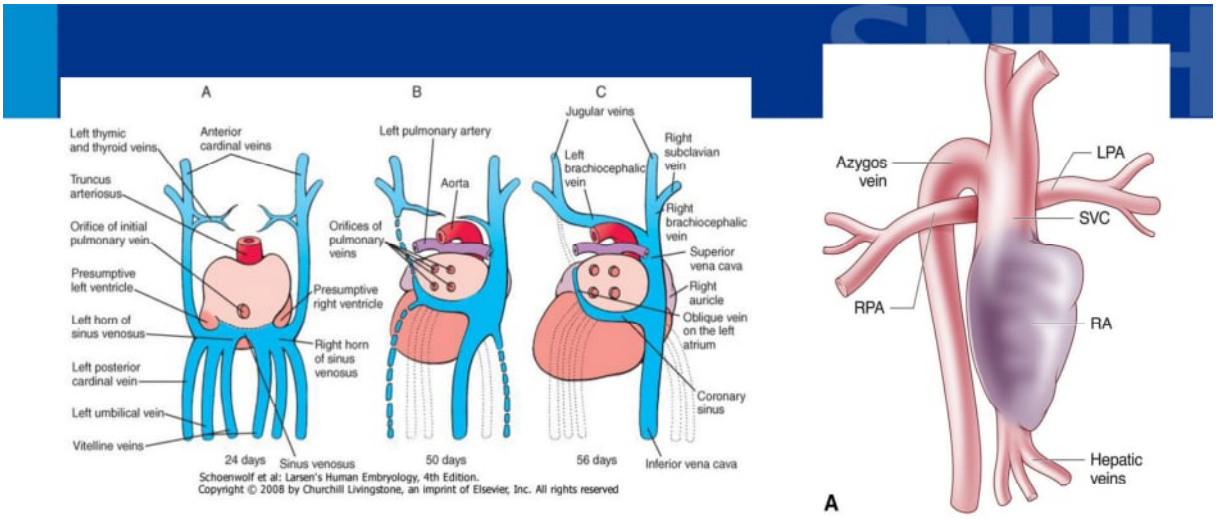
Supracardiac

Infracardiac

PVs **always** anomalous in Right isomerism



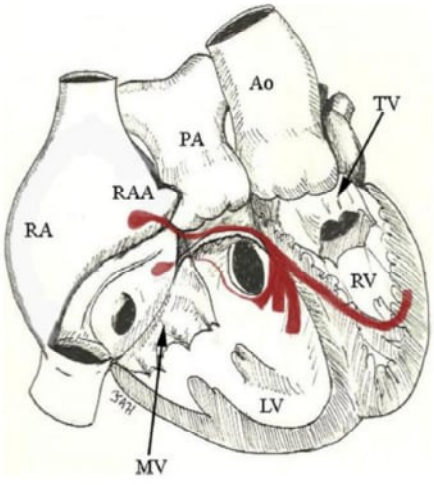
- **CS & PVs are left side structure** → always anomalous in right isomerism
  - Absent CS; difficult to set a reference catheter



- **IVC is right side structure** → frequently interrupted in left isomerism
  - Femoral vein → azygos → atrium
  - Hepatic vein cannulation may be needed in **Septal puncture**

2. Cardiac position, AV connection, Ventricle

Anomalies		Isomerism of RAA	Isomerism of LAA
Dextrocardia		14 (31%)	6 (30%)
Mesocardia		–	1 (5%)
Juxtaposition of the atrial appendages		–	1 (5%)
Ventricular loop	D-loop	24 (53%)	15 (75%)
	L-loop	21 (47%)	5 (25%)
Atrioventricular connection	Biventricular	23 (51%)	13 (65%)
	Univentricular-DIRV	16 (36%)	
	Univentricular-DILV	6 (13%)	3 (6%)
	Absent Rt AV connection	-	1 (2%)
	Absent Lt AV connection	-	3 (6%)



- **L- loop is associated Duplicated AV nodes**

3. Septal defects

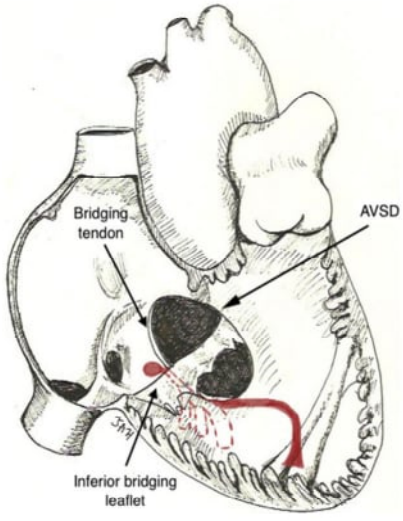
SNUH

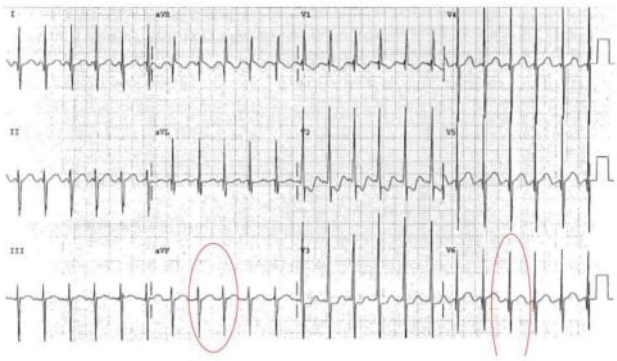
Anomalies	Isomerism of RAA	Isomerism of LAA
Atrial septal defect (fossa ovalis type)	17 (38%)	5 (25%)
Common atrium	25 (55%)	8 (40%)
Partial AVSD	**4 (9%)**	3 (15%)
Complete AVSD	**41 (91%)**	6 (30%)
Perimembranous VSD	1 (2%)	8 (40%)
Muscular VSD	4 (9%)	–
Multiple muscular VSD	–	1 (5%)
Perimembranous + muscular VSD	–	2 (10%)

AVSD in 100% of right isomerism, 45% of left isomerism
→ displaced AV node

AVSD and AV node, His bundle

SNUH





Superior QRS axis

4. VA connection and semilunar valves

Anomalies		Isomerism of RAA	Isomerism of LAA
VA connection	2 to 2 Concordant	–	9 (45%)
	2 to 2 Discordant	4 (9%)	1 (5%)
	1 to 2 Double outlet RV	21 (47%)	4 (20%)
	Single outlet – pulmonary atresia	18 (40%)	3 (15%)
	Single outlet – aortic atresia	2 (4%)	3 (15%)
Bicuspid/unicuspid PV		5/1 (13%)	4 (20%)
Valvular/subvalvular VS		11/16 (58%)	7/4 (55%)

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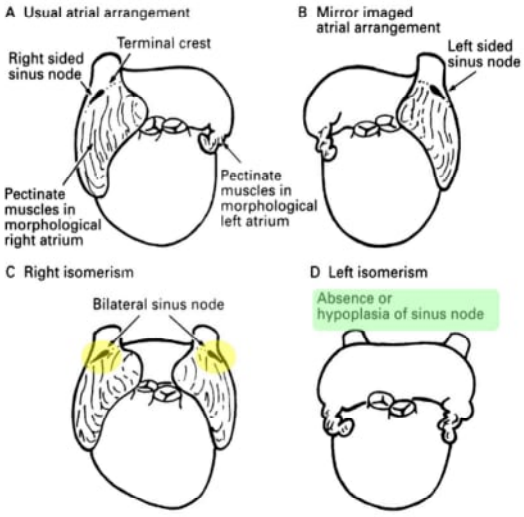
   1. Sinus node in isomerism
   2. AV node and Conduction axis
4. Arrhythmia in isomerism

SA node

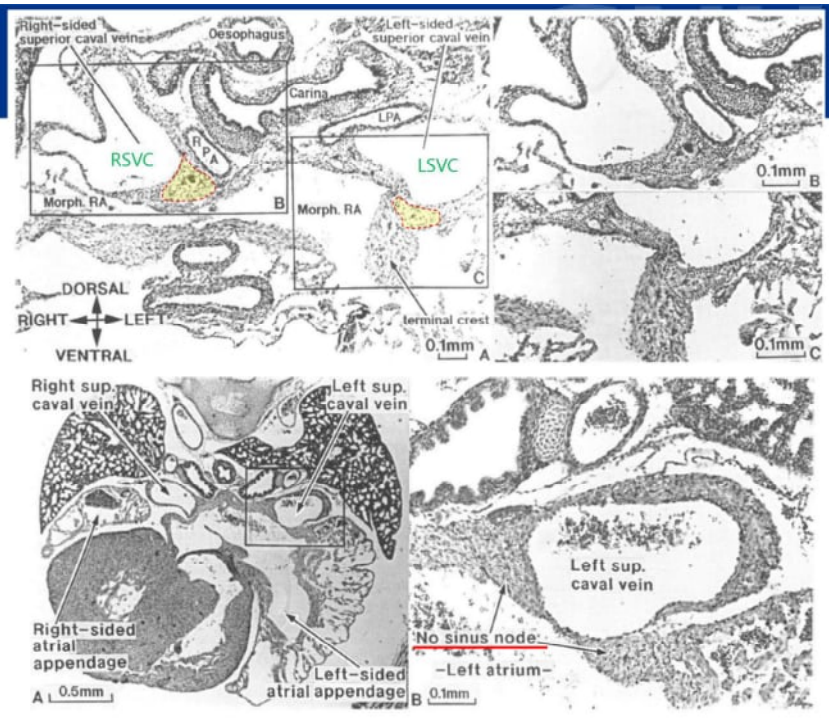


- SA node is '**unique**' right atrial structure
- Human fetus
  - 2 right isomerism
  - 12 left isomerism
- Mouse

Ho SY, Seo JW, et al. Morphology of the sinus node in human and mouse hearts with isomerism of the atrial appendages. Br Heart J. **1995**

Right isomerism

Left isomerism



SA node

SNUH

Original Article

Characterization of atrial morphology and sinus node morphology in heterotaxy syndrome: an autopsy-based study of 41 cases (1950–2008)

Adam L. Ware, MD[a,1], Dylan V. Miller, MD[b,2],
Co-burn J. Porter, MD[c], William D. Edwards, MD[b,*]

[a]Mayo Medical School, Mayo Clinic, Rochester, MN, USA
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Received 29 June 2011; received in revised form 19 December 2011; accepted 28 December 2011

- SAN is 'not necessarily' associated with a morphologic RA

2012 Ware AL, et al. Cardiovac Pathol PMID: 22285192.

SA node

SNUH

Right isomerism (N=28)

- Bilateral: (54%)
  - 29%: 2 normal size
  - 18%: 1 normal & 1 hypoplastic
  - 7%: 2 hypoplastic
- Single: (43%)
  - 28%: right/left, normal sized
  - 14%: left, hypoplastic
- Absent: (4%)

25% had hypoplastic or absent SAN

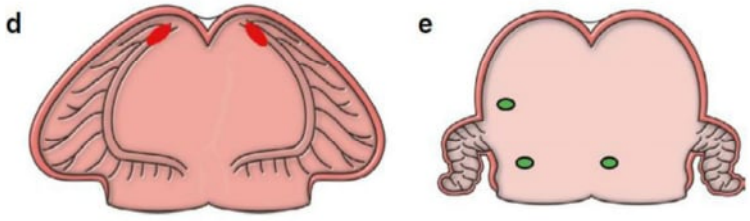
Left isomerism (N=8)

- SAN 'always' absent in right-sided atrium
  - 63%: normal sized
  - 25%: hypoplastic
  - 13%: Bilaterally absent

38% had hypoplastic or absent SAN

2012 Ware AL, et al. Cardiovac Pathol PMID: 22285192.

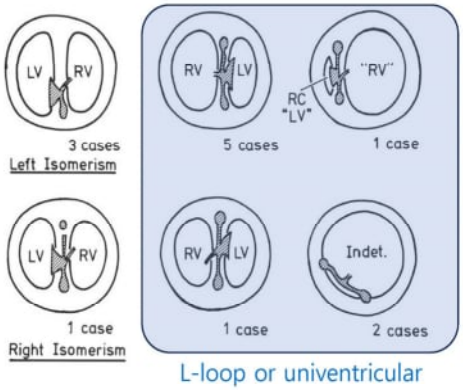
SA node



- **SAN location in right isomerism:**
  - Normal size: normal location
  - Hypoplastic: more inferior & posterior than usual along CT
- **SAN location in left isomerism:**
  - SVC/atrial junction or ridge of appendage

AV node and Conduction axis

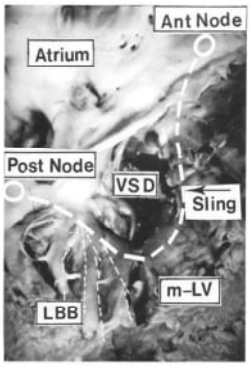
- Right & Left isomerism
  - Regular posterior position (triangle of Koch)
- In case of L-loop
  - Presence of "duplicated AV nodes"



Dickinson, D. F., et al. "The Cardiac Conduction System in Situs Ambiguus." *Circulation*

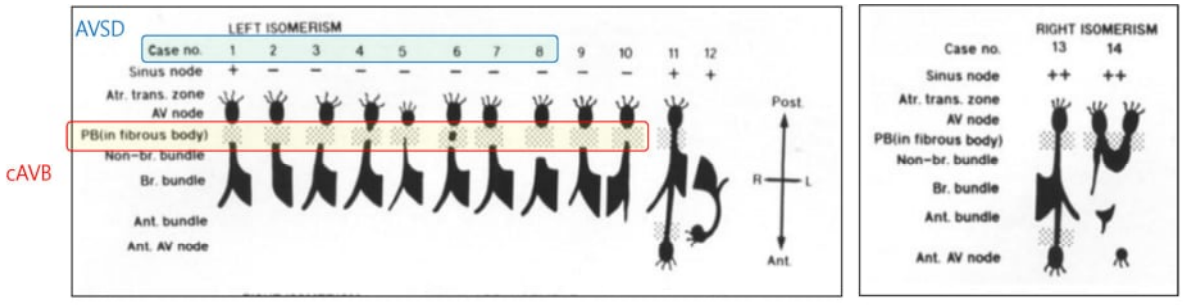
AV node and Conduction axis

- Duplicated AV nodes
  - Posterior AVN: constant position, related to tendon of Todaro
  - Anterior AVN (or anterolateral): behind aorta/pulmonary trunk
  - Connected to a sling of two AV bundles
    - On the inferior rim of VSD



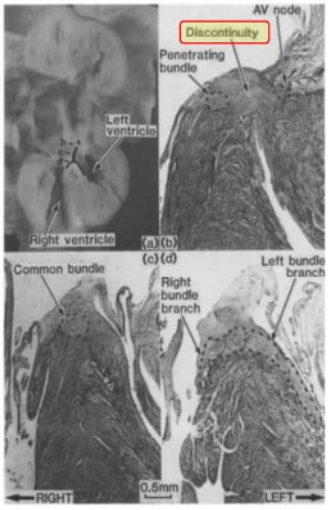
AV node and Conduction axis

- AV block
  - Frequent in left isomerism, especially with AVSD
  - Rare in right isomerism (although AVSD is also common)



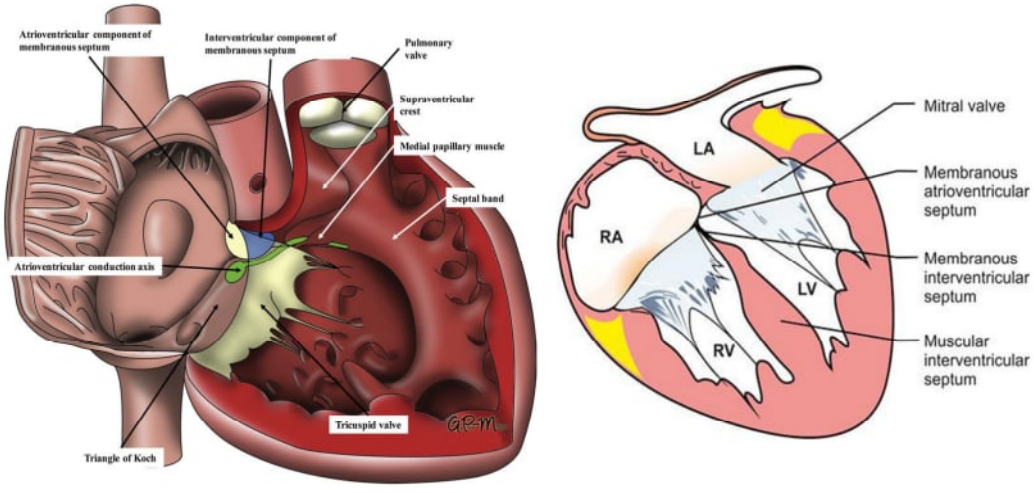
Ho SY, et al. Disposition of the atrioventricular conduction tissues in the heart with isomerism of the atrial appendages. JACC. 1992 PMID: 1527300.

AV node and Conduction axis SNUH



Discontinuity between a **large regularly situated AVN** and the **nonbranching bundle.**
→ **nodoventricular discontinuity**.

AV node and Conduction axis SNUH



ToK, Atrioventricular MS: RA structure

Contents

1. What exactly is isomerism?
2. Cardiac anatomy of right vs left isomerism
3. The conduction system in isomerism
   1. Sinus node in isomerism
   2. AV node and conduction axis
4. **Arrhythmia in isomerism**

Twin atrioventricular nodes, arrhythmias, and survival in pediatric and adult patients with heterotaxy syndrome

Mei-Hwan Wu, MD, PhD,* Jou-Kou Wang, MD, PhD,* Sheunn-Nan Chiu, MD, PhD,* Wei-Chieh Tseng, MD,† Chun-Wei Lu, MD, PhD,* Hsin-Chia Lin, MD,† Ming-Tai Lin, MD, PhD,* Chun-An Chen, MD, PhD*

Table 2 Distribution of arrhythmia types in different subtypes of isomerism

Variable	Isomerism subtype			
	RAI (n = 326)	LAI (n = 35)	Indeterminate isomerism (n = 5)	Total (N = 366)
Arrhythmia	115 (35.3)	18 (51.4)	3 (60.0)	136 (37.2)
Tachycardia	115* (35.3)	9† (25.7)	3 (60.0)	127 (34.7)
SVT	105* (32.2)	1 (2.9)	3 (60.0)	109 (29.8)
Atrial flutter/IART	1 (0.3)	1 (2.9)	1‡ (20.0)	3 (0.8)
Atrial fibrillation	2‖ (0.6)	3 (8.6)	0	5 (1.4)
Ventricular tachyarrhythmias	8 (2.5)	4† (11.4)	0	12 (3.8)
Bradycardia paced	3* (0.9)	11† (31.4)	0	14 (3.8)
SSS	2 (0.6)	10† (28.6)	0	20 (5.5)
AVB	1 (0.3)	3 (8.6)	0	4 (1.1)
Unspecified bradycardia	1 (0.3)	0 (0)	0	1 (0.3)
None	211 (64.7)	17 (48.6)	2 (40.0)	230 (62.8)

Wu MH, et al. Twin atrioventricular nodes, arrhythmias, and survival in pediatric and adult patients with heterotaxy syndrome. Heart Rhythm. 2021 Apr;18(4):605-612.

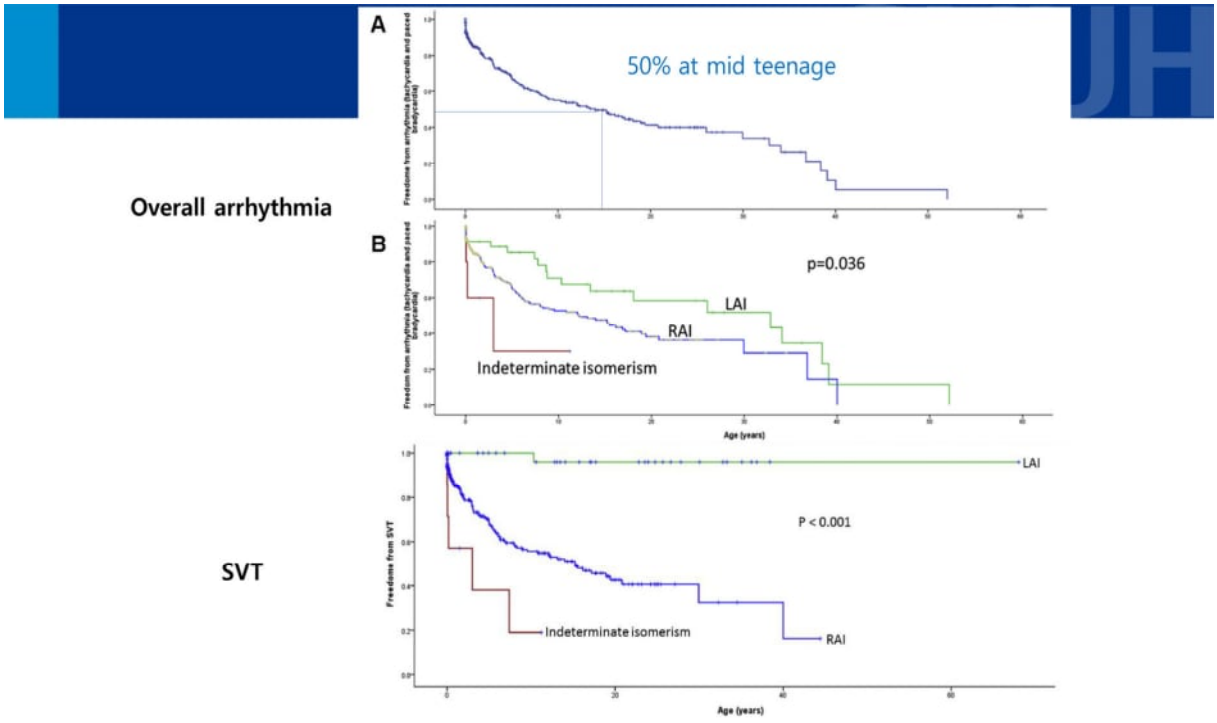


Table 1 Associated cardiac structural and conduction abnormalities in 366 heterotaxy syndrome

Variable	Right isomerism (n = 326)	Left isomerism (n = 35)	Indeterminate isomerism (n = 5)	*P*
Morphological characteristics				
IVC interruption	0 (0)	32 (94)	1 (20)	<.001
Dextrocardia	84 (25.8)	9 (27.3)	1 (20.0)	.419
TAPVR	320 (98.2)	1 (2.9)	4 (80)	<.001
TAPVR to the systemic vein	135 (41.4)	1 (2.9)	0 (0)	<.001
Pulmonary venous stenosis	128 (39.3)	1 (2.9)	0 (0)	<.001
Common atrium	301 (92.3)	18 (51.4)	3 (60.0)	<.001
Common AV valve	321 (97.9)	20 (57.1)	3 (60.0)	<.001
Balanced ventricle	80 (24.5)	24 (68.6)	1 (20.0)	<.001
DORV or DOV	288 (88.3)	25 (71.4)	3 (60.0)	.005
PS/PA	321 (98.5)	26 (74.3)	5 (100)	<.001
MAPCA	23 (7.1)	0 (0)	1 (20.0)	.131
Coarctation/IAA	6 (1.8)	2 (5.7)	0 (0)	.312
Conduction characteristics				
Twin AV nodes	50 (51.5)	2 (8.7)	2 (40.0)	.001
Accessory pathway	3 (0.9)	0 (0)	1 (20.0)	<.001
Nonsurgical sinus bradycardia	0 (0)	17 (48.7)	1 (20.0)	<.001
Nonsurgical AV block	0 (0)	3 (8.6%)	0 (0)	<.001

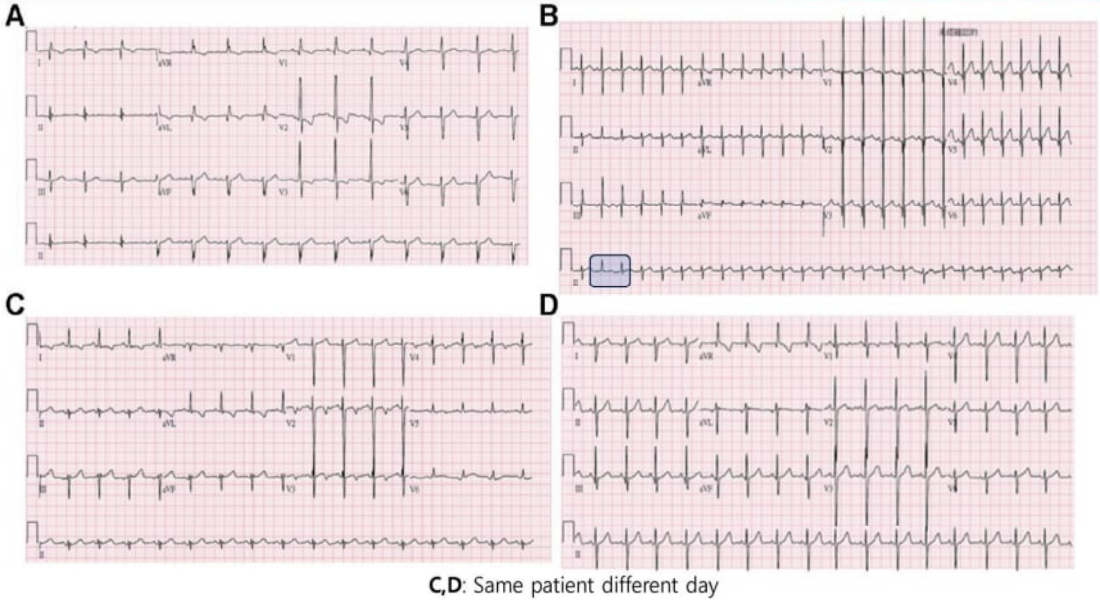
Wu MH, et al. Twin atrioventricular nodes, arrhythmias, and survival in pediatric and adult patients with heterotaxy syndrome. Heart Rhythm. 2021 Apr;18(4):605-612.

Table 3 Risk analysis of time-related supraventricular tachycardia (n = 109)

Variable	Univariate analysis	Multivariate analysis		
	P (log-rank test)	Odd ratio	95% CI	*P*
Male sex	.907			
Isomerism type	<.001			
RAI		Reference		
LAI		0.098	0.008–1.180	.067
Indeterminate isomerism		4.470	1.185–16.859	.027
Morphological characteristics				
Dextrocardia	.990			
Pulmonary vein stenosis	.004	1.278	0.648–2.518	.479
TAPVR	<.001	0.583	0.111–3.063	.524
Common atrium	<.001	2.967	0.799–11.02	.104
Common AV valve	.002	0.793	0.049–12.85	.871
Single ventricle	.355			
Moderate or more than moderate AVR	.992			
Ventricular dysfunction	.060			
Conduction characteristics				
Twin AV node	<.001	4.814	2.330–9.948	<.001
Sinus bradycardia	.005	0.718	0.050–10.299	.807
Preexcitation	.237			
Open heart surgery	.220			

Wu MH, et al. Twin atrioventricular nodes, arrhythmias, and survival in pediatric and adult patients with heterotaxy syndrome. Heart Rhythm. 2021 Apr;18(4):605-612.

Twin AV node ECG



C,D: Same patient different day

Twin AV Node and Induced Supraventricular Tachycardia in Fontan Palliation Patients

EUN-JUNG BAE,* CHUNG-IL NOH,* JUNG-YUN CHOI,* YONG-SOO YUN,* WOONG-HAN KIM,† JEONG-RYUL LEE,† and YONG-JIN KIM†

Table I.

Morphological Characteristics of the 52 Patients

Visceroatrial situs	
Ambiguus	20 (38%)
Right isomerism	18
Left isomerism	2
Solitus	22 (43%)
Inversus	10 (19%)
Atrioventricular connections	
Concordant	39 (75%)
Discordant	8 (15%)
Ambiguous	5 (10%)
Atrioventricular junctional anomaly	
Tricuspid atresia or mitral atresia	5 (10%)
Single indeterminate AV valve*	7 (12%)
Double inlet right ventricle	19 (37%)
Double inlet left ventricle	4 (8%)
Criss cross AV connections	2 (4%)
Biventricular 2 valves	5 (10%)
Biventricular common valve	10 (19%)
Complete AVSD†	18 (35%)
Ventriculoarterial connections	
DORV	35 (67%)
Concordant	6 (12%)
Discordant	11 (21%)

Table II.

Twin AV Node and Induced Supraventricular Tachycardia Involving Two AV Nodes

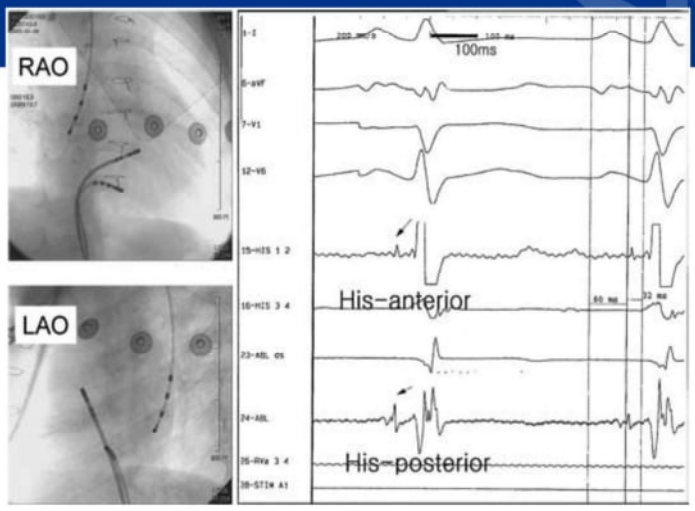
Morphology	2 QRS (ECG/Paced)	Retrograde VA Conduction	Two HBE Recorded	AVRT (Induced/ Clinical)	Mode of Induction	TCL (ms)	VA (ms)	Antegrade Limb
{A.D.D}, RI, AVSD, BV	+/+	Decremental	+	+/+	V single	272	98	Post.
{A.D.D}, RI, AVSD, BV	+/+	Decremental	–	+/+	A & V single	330	188	Ant.
{A.D.D}, RI, AVSD, RV	+/+	Decremental	+	+/+	V single	330	113	Ant.
{A.D.D}, RI, AVSD, BV	+/+	Decremental	+	+/+	A burst	332	164	Post.
{A.D.D}, RI, TVst.	+/+	Decremental	+	+/–	A burst	280	98	Post.
{A.D.D}, RI, AVSD, BV	+/+	Decremental	–	+/–	A burst	296	128	Post.
{A.D.D}, RI, AVSD, RV	+/+	Dissociation	ne	–/–				
{A.D.D}, RI, AVSD, RV	+/+	Dissociation	ne	–/–				
{A.D.D}, RI, SIRV	+/+	Dissociation	ne	–/–				
{I.D.D}, TA	+/+	Dissociation	ne	–/–				
{A.D.D}, RI, AVSD, RV	±	Decremental	ne	–/–				
{S.D.D}, AVSD, RV	±	Dissociation	ne	–/–				
{A.L.L}, RI, AVSD, BV	±	Decremental	ne	–/–				

6/10 had twin node AVRT

Either Ant/Post can be antegrade limb

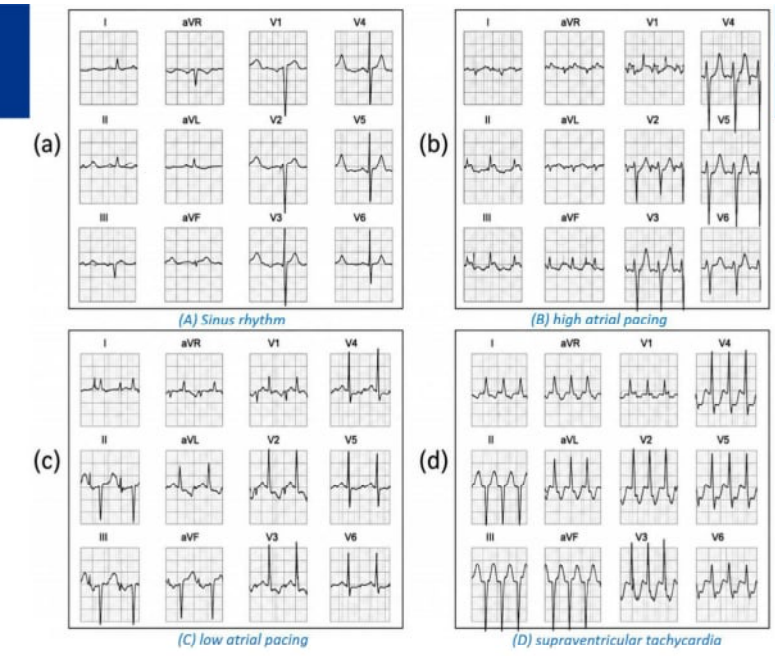
Not exclusive, but predominant in isomerism (9/10 in twin AVN)
9/20 (45%) in isomerism

Bae EJ, et al. Twin AV node and induced supraventricular tachycardia in Fontan palliation patients. PACE. 2005 Feb;28(2):



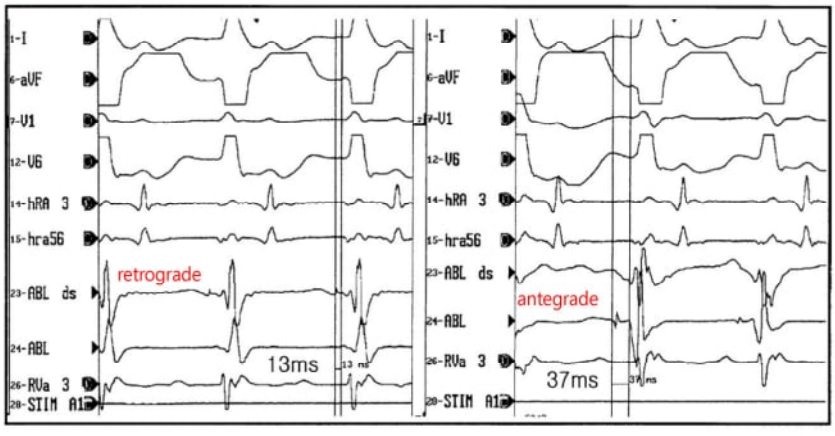
Separate His bundle EGM during sinus rhythm

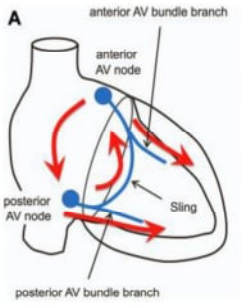
Bae EJ, et al. Twin AV node and induced supraventricular tachycardia in Fontan palliation patients. PACE. 2005 Feb;28(2):



Different non-preexcited QRS complex
→ not an accessory pathway

SVT QRS ≒ low atrial paced QRS
→ post AVN as antegrade limb





• Antegrade limb may be determined by HV intervals

Bae EJ, et al. Twin AV node and induced supraventricular tachycardia in Fontan palliation patients. PACE. 2005 Feb;28(2):

Table IV.
Categorization of AV Node Characteristics in Complex Cardiac Anomaly

	2QRS ECG	2QRS Paced	Two HBE	Slurred QRS	HBE-retro Preceding V During AVRT	AVRT	JT with AV Dissociation
	–	–	–	–	–	–	Possible
	+	+	+	–	+	Possible	Possible
	+	+	+	–	–	Possible	Possible
	–	–	+	–	+	Possible	Possible
	+	±	±	+	+	Possible	Possible

- 1 AVN & 1 conduction bundle.
- 2 AVNs & 2 conduction bundles w sling.
- 2 AVNs & 2 conduction bundles w/o sling
- 2 AVNs & 1 common conduction bundle.
- 1 dominant AVN with a nodofascicular, nodoventricular, or fasciculoventriclar pathway

Summary

	Right isomerism	Left isomerism
Key feature	Bilateral RAA	Bilateral LAA
Venous drainage	TAPVC, absent CS	IVC interruption
SA node	**Often bilateral** may be hypoplastic	**Often hypoplastic/absent**
AV node	**Normal posterior AVN** (duplicated in L-loop)	
AV conduction axis	Normal	Discontinuous penetrating bundle
Rhythm problem	**SVT** (twin node reentry)	**SND/ AVB**

2026
해부학·선천성·유전성
부정맥 연구회 합동 심포지엄

Session 2

Long and Short QT Syndromes; Stop the Short-Long-Short Sequence!

좌장: 배은정 (서울의대 소아청소년과), 조용근 (경북의대 순환기내과)
패널: 양소영 (가천의대 심장내과), 안효정 (서울의대 순환기내과), 정주희 (고려의대 순환기내과), 차누리 (연세의대 소아청소년과)

Diagnosis and genetics of long QT syndrome; How long is long?
백재숙 (울산의대 소아청소년과)

How to manage each type of long QT syndrome; Long life with long QT
송미경 (서울의대 소아청소년과)

Sympathectomy for long QT syndrome; Surgical β-blocker
김하은 (연세의대 흉부외과)

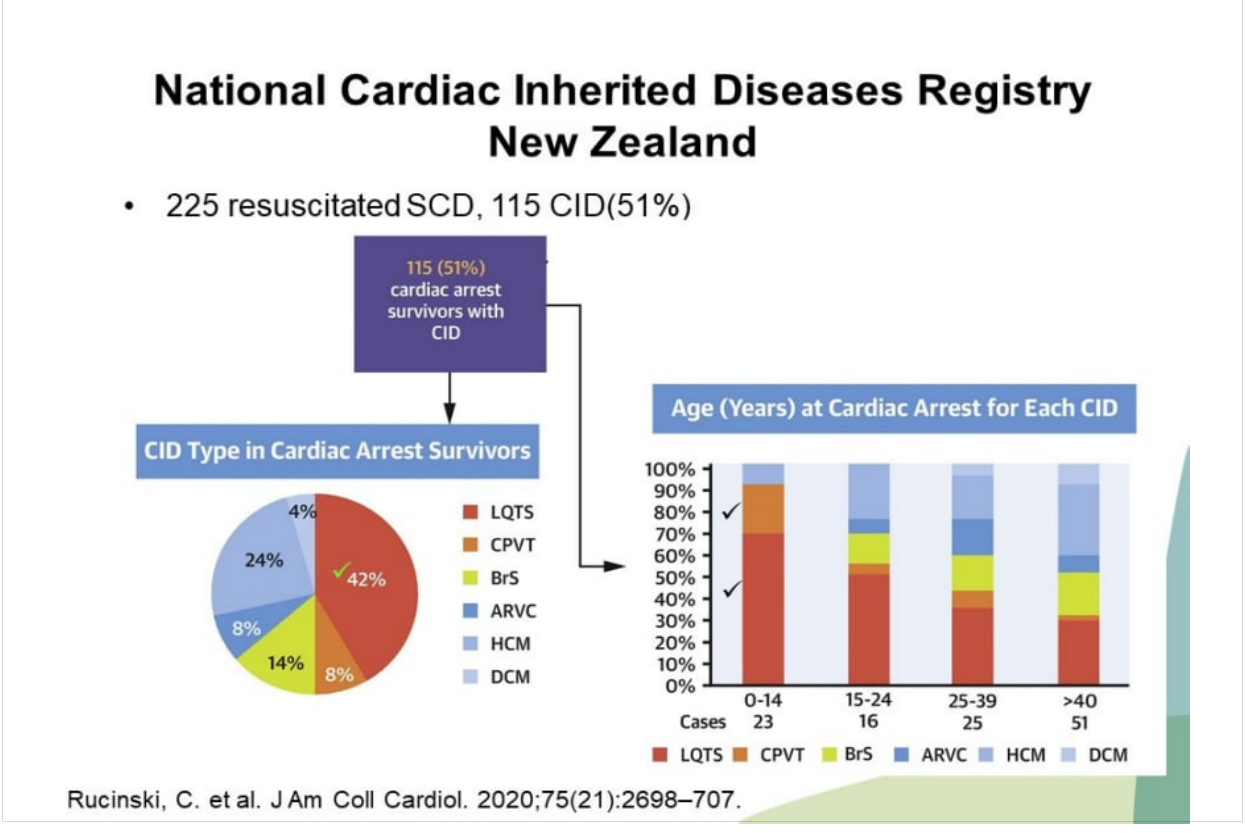
Short QT syndrome; Short but not least
김주연 (성균관의대 순환기내과)

대한부정맥학회
Korean Heart Rhythm Society

Diagnosis and genetics of long QT syndrome; How long is long?

백 재 숙

울산의대 소아청소년과



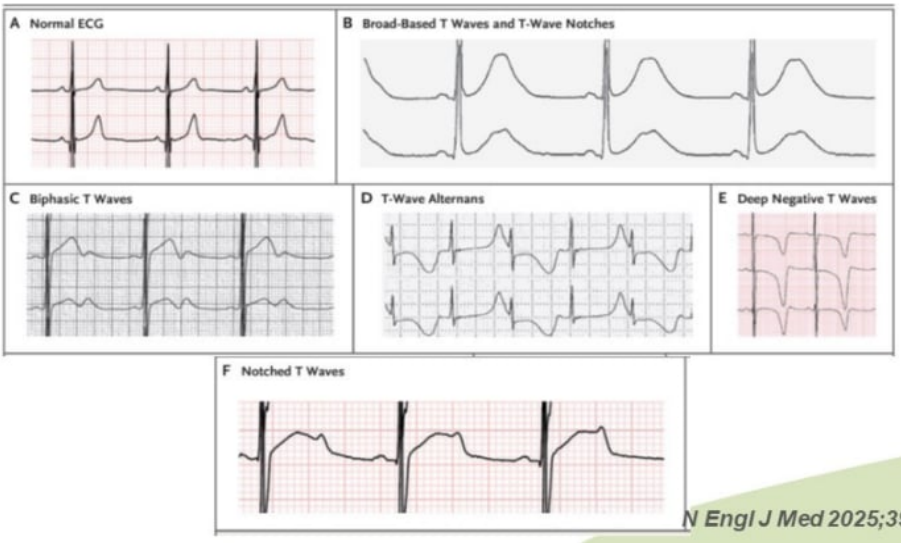
Long QT syndrome

- A genetic disorder disrupting electrical activity in the heart
- Life-threatening arrhythmias and sudden cardiac death
- Initial patient descriptions
  - *Jervell and Lange-Nielsen syndrome*
    - Described in 1957
    - Prolonged QT on ECG , congenital deafness
    - Autosomal recessive
  - *Romano-Ward syndrome*
    - More common
    - Describe in 1963 & 1964
    - with normal hearing, AD

Schwartz PJ, Crotti L. N Engl J Med 2025;393:2023-2034

Diagnosis; ECG

- Markedly prolonged QT + bizarre morphologic changes with regard to ventricular repolarization (biphasic and notched T waves)



N Engl J Med 2025;393:2023-34.

Measurement pitfalls

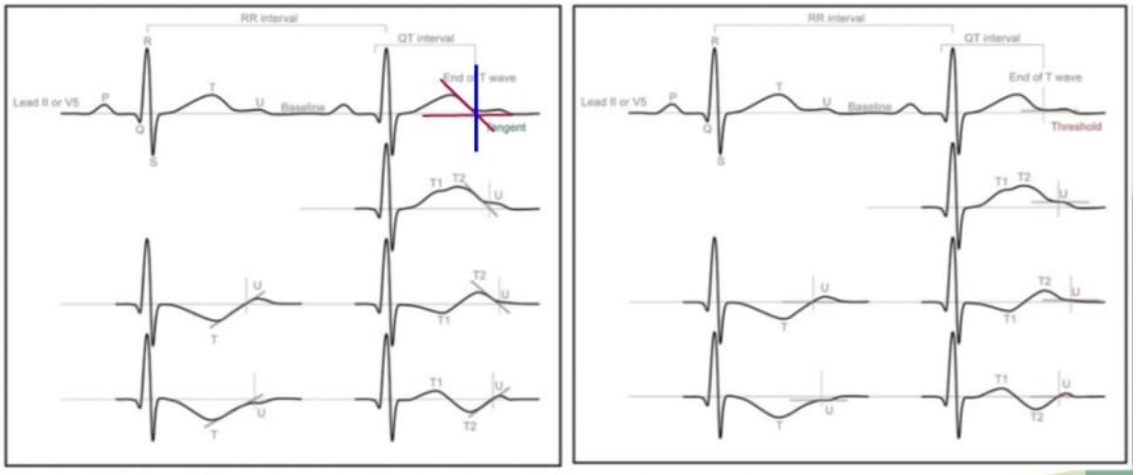
Where dose measurement goes wrong?

- T-wave offset
  - Wide inter-reader variability when it merges with a U wave
- Lead / beat choice
  - Recommended: lead II or V5, average of 3 beats
- Max QT vs. mean QT — taking only the longest inflates false positives
- Correction formula
  - Bazett over-corrects at fast heart rates

Schwartz PJ, Crotti L. N Engl J Med 2025;393:2023-2034

QT measurement

- Tangent technique
- Threshold technique



Tangent reads 10–20 ms shorter — borderline by tangent can be diagnostic by threshold

J Am Coll Cardiol EP 2022;8:687–706

Comparing correction formulas

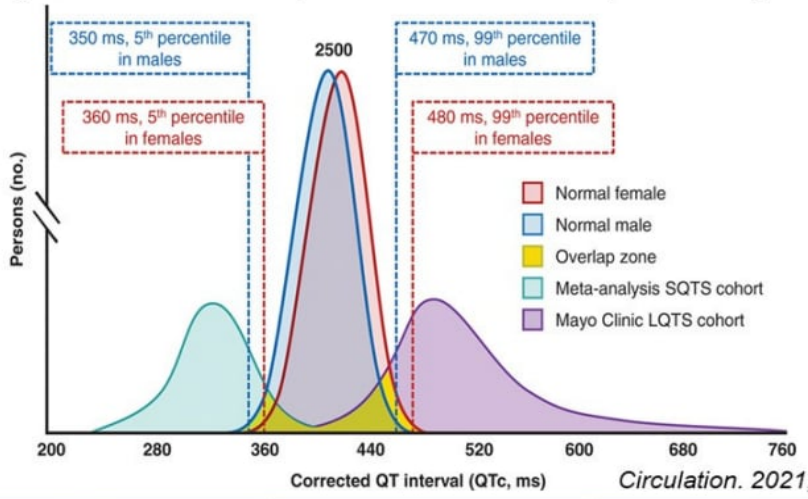
The four formulas agree in the middle and disagree at the extremes of heart rate

Formula	QTc =	Behaviour
Bazett	QT / √RR	Over-corrects when fast, under-corrects when slow
Fridericia	QT / $\sqrt[3]{RR}$	Stable across a wider heart-rate range
Framingham	QT + 0.154 (1 – RR)	Linear, population-derived
Hodges	QT + 1.75 (HR – 60)	Rate-based, rarely used at the bedside

Bazett remains the basis of the diagnostic cutoffs and the Schwartz score — NEJM 2025

How long is long?

- Diagnostic threshold: repeated QTc ≥ 480 ms (ESC 2022)

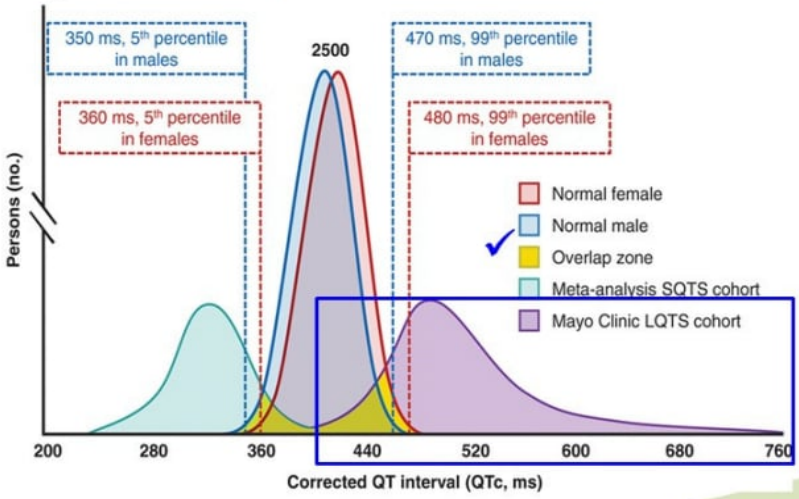


Circulation. 2021;143:739–752

QTc (Bazett)	Men	Women
Normal	< 440 ms	< 460 ms
Prolonged	≥ 460 ms	≥ 480 ms

Borderline QTc

- 25% of patients with LQTS may have a ECG showing borderline prolonged QT interval✓



Circulation. 2021;143:739–752

Borderline QTc

QTc (Bazett)	Men	Women
Borderline	440–459 ms	460–479 ms

- ESC 2022

Recommendation Table 41 — Recommendations for the management of patients with long QT syndrome

Recommendations	Class[a]	Level[b]
Diagnosis		
The LQTS diagnosis should be considered in the presence of a QTc ≥460 ms and <480 ms in repeated 12-lead ECGs in patients with an arrhythmic syncope in the absence of secondary causes for QT prolongation.[952,962,963]	IIa	C

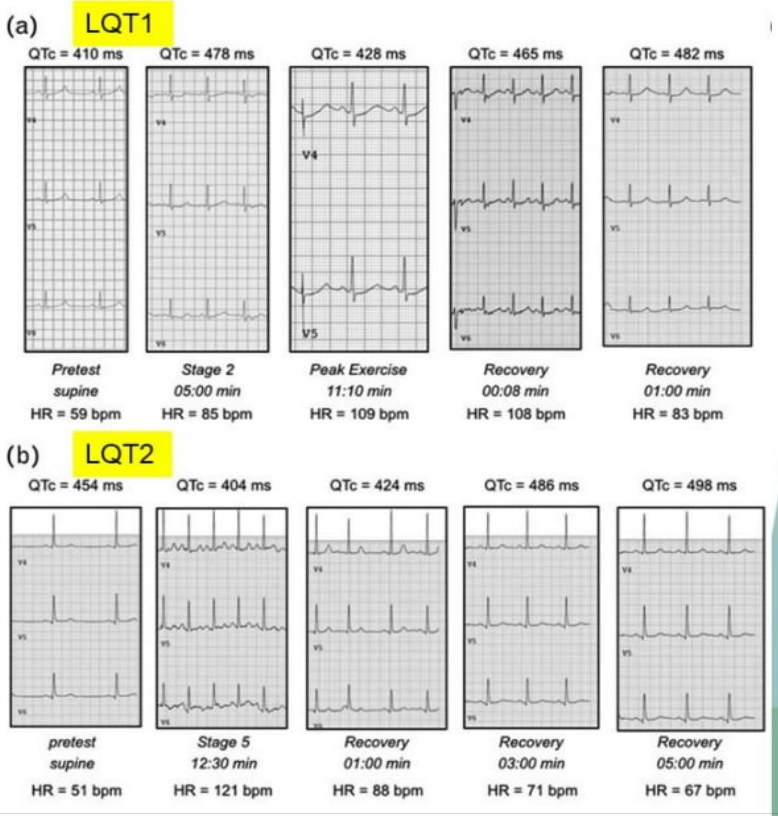
Provocation Testing

- Overlap zone
- Assessment of repolarization in response to autonomic changes
  - During acceleration & deceleration of HR
  - Maladaptive repolarization response is frequently seen in LQTS

- **Exercise stress test — the only truly useful test**
  - Marked QT prolongation at 4 min of recovery is highly specific
  - T–P fusion at peak exercise also supports it
- 24-hour 12-lead Holter often unmasks changes at night
- Stand-up test — of limited value
- Epinephrine challenge — should not be used
  - Arrhythmogenic, and alters repolarization in normal hearts

Schwartz PJ, Crotti L. N Engl J Med 2025;393:2023-2034

Exercise stress test



Modified diagnostic score

- **ESC 2022**
- Score-based
  - LQTS diagnostic score > 3 (ESC 2022)
- ECG alone
  - Repeated QTc ≥ 480 ms, whatever the symptoms (ESC 2022)
  - ≥ 460 ms is enough with unexplained arrhythmic syncope
- Genotype alone
  - A pathogenic variant diagnoses LQTS even if the QTc is normal

Findings			Points
ECG	QTc	≥480 ms	3.5
		=460–479 ms	2
		=450–459 ms (in males)	1
		≥480 ms during 4th minute of recovery from exercise stress test	1
	Torsade de pointes		2
	T wave alternans		1
	Notched T wave in 3 leads		1
	Low heart rate for age		0.5
Clinical history	Syncope	With stress	2
		Without stress	1
Family history	Family member(s) with definite LQTS		1
	Unexplained SCD at age <30 years in first-degree family		0.5
Genetic finding	Pathogenic mutation		3.5

ECG, electrocardiogram; LQTS, long QT syndrome; SCD, sudden cardiac death.
Diagnosis of LQTS with a score >3.

Genetics and diagnosis

A pathogenic variant diagnoses LQTS by itself

- **What the genotype adds beyond the number**
  - It explains the trigger — exercise, startle, or sleep
  - It guides gene-specific management
  - It finds relatives with one targeted test
- **What it does not do**
  - T-wave shape hints at genotype but cannot replace it
  - A variant of uncertain significance is not a diagnosis

Schwartz PJ, Crotti L. N Engl J Med 2025;393:2023-2034

Genetic testing

- ESC 2022

Recommendation Table 41 — Recommendations for the management of patients with long QT syndrome

Recommendations	Class[a]	Level[b]
Diagnosis		
In patients with clinically diagnosed LQTS, genetic testing and genetic counselling are recommended.	I	C
It is recommended that LQTS is diagnosed in the presence of a pathogenic mutation, irrespective of the QT duration.	I	C

Europace 2022;24:1307-1367

Genetic testing

Table 1 Classification of genetic evidence by the Clinical Genome Resource (ClinGen) for genes previously associated with LQTS

Gene	Protein	Level of evidence
AKAP9	A kinase anchor protein 9	Disputed
ANK2	Ankyrin-2	Disputed
CACNA1C	Calcium voltage-gated channel α1c subunit	Moderate
CALM1	Calmodulin-1	Definitive
CALM2	Calmodulin-2	Definitive
CALM3	Calmodulin-3	Definitive
CAV3	Caveolin-3	Limited
KCNE1	Potassium voltage-gated channel subfamily E regulatory subunit 1	Disputed
KCNE2	Potassium voltage-gated channel subfamily E regulatory subunit 1	Disputed
KCNH2	Potassium voltage-gated channel subfamily H member 2	Definitive
KCNJ2	Potassium voltage-gated channel subfamily J member 2	Limited
KCNJ5	Potassium voltage-gated channel subfamily J member 5	Disputed
KCNQ1	Potassium voltage-gated channel subfamily Q member 2	Definitive
SCN4B	Sodium voltage-gated channel β subunit 4	Disputed
SCN5A	Sodium channel voltage-gated α subunit 5	Definitive
SNTA1	Syntrophin α1	Disputed
TRDN	Triadin	Strong

- Definitive genes: KCNQ1, KCNH2, SCN5A, CALM1–3
- Indication
  - Offer it to every index patient with a high probability of LQTS
  - Probability from history, family history, baseline ECG, Holter and exercise test (Schwartz score ≥ 3.5)
  - Relatives: cascade testing for the family variant only

Wilde AAM, et al. Heart 2022;**108**:332–338

Concealed LQTS

- 3,386 genotyped subjects from 7 multinational LQTS registries

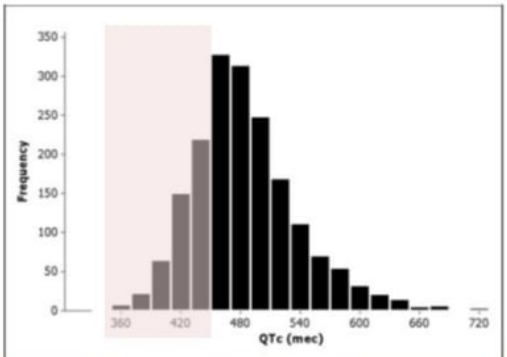


Figure 1 Distribution of QTc Interval Duration in Genotype-Positive Patients With LQTS

Distribution of corrected QT (QTc) interval durations in genotype-positive study patients. LQTS = long-QT syndrome.

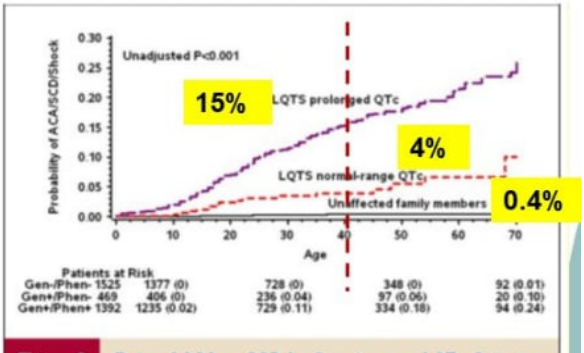


Figure 2 Rate of ACA or SCD by Genotype and QTc Category

Kaplan-Meier cumulative probabilities of aborted cardiac arrest (ACA) and sudden cardiac death (SCD) by genotype and corrected QT (QTc) subgroup. LQTS = long-QT syndrome.

- Normal-range QTc ≤440ms
  ➔ n = 469 (13.8%)

Goldenberg, JACC 2011;57:51-59

Concealed LQTS

- **Test even when the resting QTc is normal**
  - A family variant is already known — cascade testing
  - Unexplained arrest, or autopsy-negative sudden death
  - Unexplained drowning, or syncope triggered by startle
  - QTc prolongation at 4 min of recovery, or abnormal T waves

Major genotypes: LQT1–3

- KCNQ1, KCNH2 and SCN5A, identified in 1995–1996
- **LQT1 · KCNQ1 (~50%) — loss of I_{Ks}**
  - I_{Ks} is the prevalent current under adrenergic drive
  - If QT does not shorten as rate rises, VF may ensue
- **LQT2 · KCNH2 (~40%) — loss of I_{Kr}**
- **LQT3 · SCN5A (~10%) — gain of function, I_{Na}**
  - Late inward sodium current prolongs repolarization

Schwartz PJ, Crotti L. N Engl J Med 2025;393:2023-34

Less common genotypes

- **Jervell–Lange-Nielsen syndrome**
  - Homozygous or compound heterozygous KCNQ1 or KCNE1
  - Recessive, with congenital deafness
- **Timothy syndrome · CACNA1C**
  - Long QT with skeletal and neurodevelopmental features
- **Calmodulinopathies · CALM1, CALM2, CALM3**
  - Three genes on different chromosomes, one protein
  - Impaired calcium-channel inactivation prolongs QT
- **Andersen–Tawil syndrome · KCNJ2**
  - Periodic paralysis, dysmorphism, prominent U waves

Schwartz PJ, Crotti L. N Engl J Med 2025;393:2023-2034

Genotype–phenotype relationships

- The genotype also gives the trigger and guides therapy
- Specific genotype–phenotype relationships have been described for the three most common subtypes: LQTS types 1, 2 and 3

	Type 1	Type 2	Type 3
Gene	*KCNQ1*	*KCNH2*	*SCN5a*
Protein	$K_v7.1$	$K_v11.1$	$Na_v1.5$
Effect on current	$I_{Ks}\downarrow$	$I_{Kr}\downarrow$	$I_{Na1.5}\uparrow$
Effect on action potential			
Frequency among LQTS	± 35%	± 30%	≤ 10%
Penetrance	± 65%	± 80%	± 90%
Typical resting ECG (V_5)			
QT change with exercise	Failure to shorten	Normal	Supranormal
Main trigger of events	Exercise (swimming)	Arousal	Rest
Age of onset arrhythmias	Childhood	Puberty	Puberty
Gender most at risk	♂	♀	♀
Onset of arrhythmias	*Not pause-dependent*	*Pause-dependent*	At lower heart rates
Therapy	Beta-blockers (+++) Left stellectomy	Beta-blockers (++) Left stellectomy Potassium suppletion	Beta-blockers (++) Sodium channel blocker Pacemaker

Wilde AAM, et al. Heart 2022;**108**:332–338

Same QTc, different risk

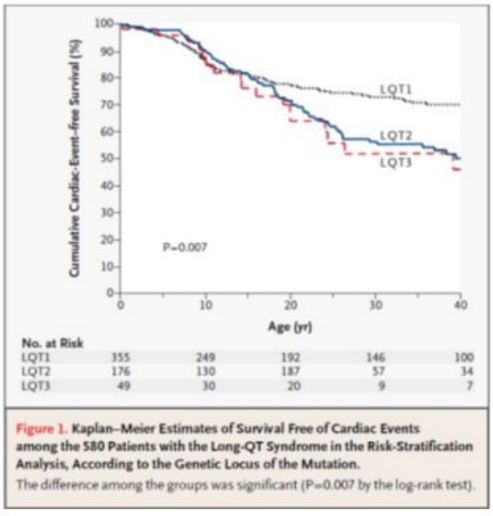


Figure 1. Kaplan–Meier Estimates of Survival Free of Cardiac Events among the 580 Patients with the Long-QT Syndrome in the Risk-Stratification Analysis, According to the Genetic Locus of the Mutation.
The difference among the groups was significant (P=0.007 by the log-rank test).

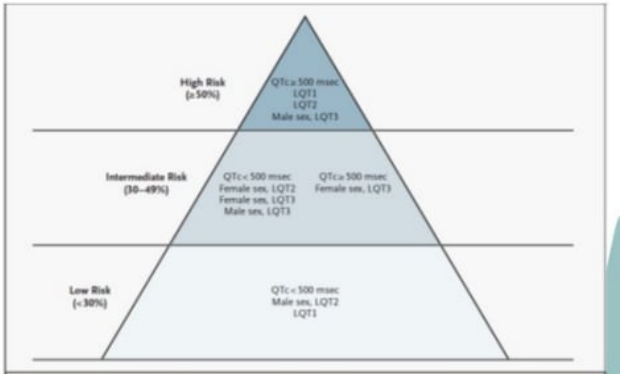
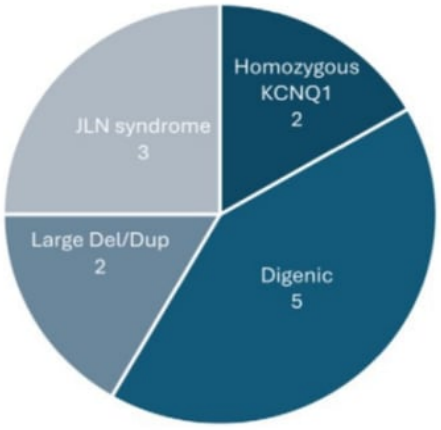


Figure 4. Proposed Scheme for Risk Stratification among Patients with the Long-QT Syndrome According to Genotype and Sex.
The risk groups have been defined on the basis of the probability of a first cardiac event (syncope, cardiac arrest, or sudden death) before the age of 40 years and before therapy. A probability of 50 percent or higher defines the high-risk group, a risk of 30 to 49 percent the intermediate-risk group, and a risk below 30 percent the low-risk group.

N Engl J Med 2003;348:1866-74

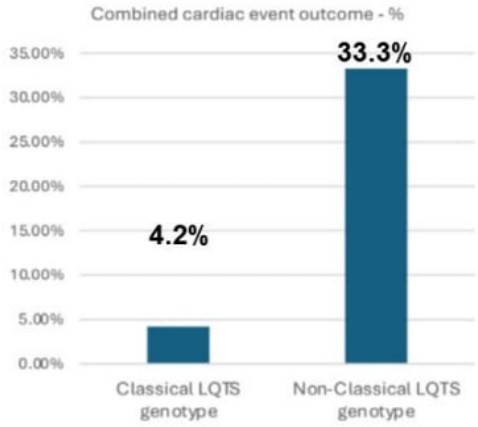
High-Risk Nonclassical Genotypes

Panel B : Genotype of cases with Non classical LQTS



- Classical n= 713
- Nonclassical, n= 12

Panel C: Event rate in classical and Non-classical LQTS cases



Circ Genom Precis Med. 2024;17:e004554

Variant interpretation

- **Variants of uncertain significance are common**
  - Reclassified from phenotype strength, cosegregation and functional testing

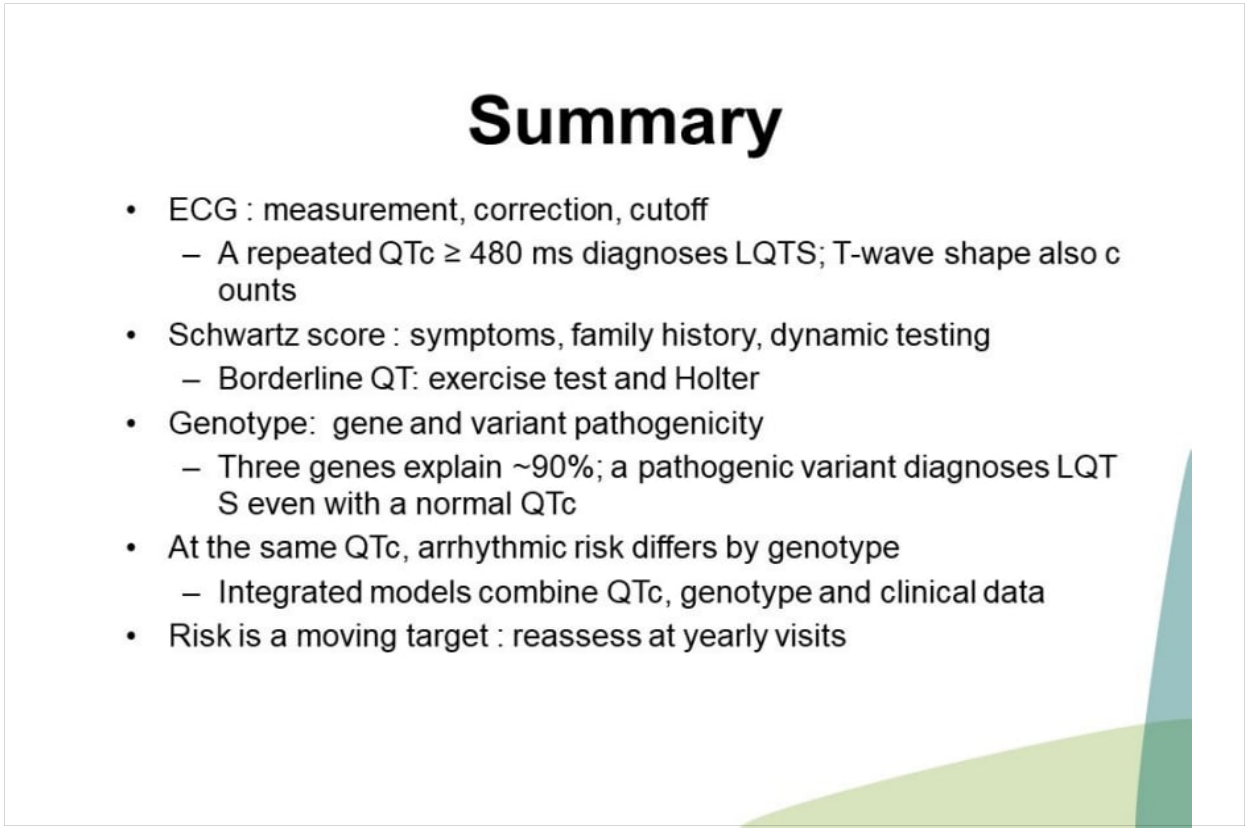
Schwartz PJ, Crotti L. N Engl J Med 2025;393:2023-34

Table 4 Comparing subgroup analysis comparing reclassification rate of variants according to initial variant classification

Initial variant classification	Upgrade, n/N (%)	Downgrade, n/N (%)	Unchanged, n/N (%)	P-value[a]
VUS				
Definite/moderate gene group	5/18(27.8)	4/18 (22.2)	9/18 (50)	0.0403[a]
Limited/disputed gene group	0 (0)	6/8 (50)	2/8 (25)	
(Likely) pathogenic				
Definite/moderate gene group	N/A	1/31 (3.2)	30/31 (96.8)	0.0057[b]
Limited/disputed gene group	N/A	2/2 (100)	0	

- Among 59 variants, 18 variants (30.5%) were reclassified.
- Hong Kong Children's Hospital

Pediatric Cardiology (2024) 45:1023–1035


Summary
• ECG : measurement, correction, cutoff
– A repeated QTc ≥ 480 ms diagnoses LQTS; T-wave shape also c ounts
• Schwartz score : symptoms, family history, dynamic testing
– Borderline QT: exercise test and Holter
• Genotype: gene and variant pathogenicity
– Three genes explain ~90%; a pathogenic variant diagnoses LQT S even with a normal QTc
• At the same QTc, arrhythmic risk differs by genotype
– Integrated models combine QTc, genotype and clinical data
• Risk is a moving target : reassess at yearly visits

How to manage each type of long QT syndrome; Long life with long QT

송 미 경

서울의대 소아청소년과

Congenital long QT syndrome (LQTS) is potentially lethal but highly treatable. In contemporary cohorts, the majority of patients are asymptomatic at first evaluation, and the risk of a sentinel cardiac event is very low once genotype- and phenotype-tailored therapy is in place. Breakthrough events today are driven less by the disease itself than by non-adherence and by treatment that is mismatched to the patient's arrhythmia phenotype. The clinical goal has therefore shifted from preventing death to preventing both under-treatment and over-treatment, so that patients can live full and unrestricted lives.

Non-selective beta-blockade with nadolol or propranolol remains the foundation for every genotype and carries a Class I recommendation, together with avoidance of QT-prolonging drugs, electrolyte and fever management, availability of an automated external defibrillator, and cascade family screening. Genotype then determines the second therapeutic layer. In LQT1, beta-blockers are most effective and adherence is the principal determinant of outcome, with supervised rather than prohibited swimming. In LQT2, environmental control of auditory and emotional triggers, maintenance of serum potassium in the upper normal range, and heightened vigilance during the postpartum window are essential; mexiletine also shortens the QT interval in this subtype. In LQT3, late sodium current blockade with mexiletine is a Class I recommendation for patients with a prolonged QT interval, ideally preceded by an acute mexiletine challenge because the response is variant-specific, and the threshold for pacing or an implantable cardioverter-defibrillator is lowest in this group. Rare and malignant forms, including Jervell and Lange-Nielsen syndrome, calmodulinopathy, Timothy syndrome, and Andersen-Tawil syndrome, require distinct strategies.

Whether left cardiac sympathetic denervation (LCSD) works differently across genotypes has been debated. Long-term single-center experience suggests that protection is governed by pre-operative

clinical severity and by the degree of postoperative QTc shortening rather than by genotype, whereas other cohorts report markedly higher breakthrough rates in LQT3. Recent data reconcile these observations by showing that, independent of the underlying genotype, patients whose pre-LCSD events were adrenergically triggered derive substantially greater benefit. The practical question is therefore not which LQTS subtype the patient has, but whether the events are sympathetically mediated. A post-LCSD QTc below 500 ms serves as a single, genotype-independent marker of therapeutic response.

This lecture reviews subtype-specific management, and discusses shared decision-making for exercise and competitive sports participation.

Sympathectomy for long QT syndrome; Surgical β-blocker

김 하 은

연세의대 흉부외과

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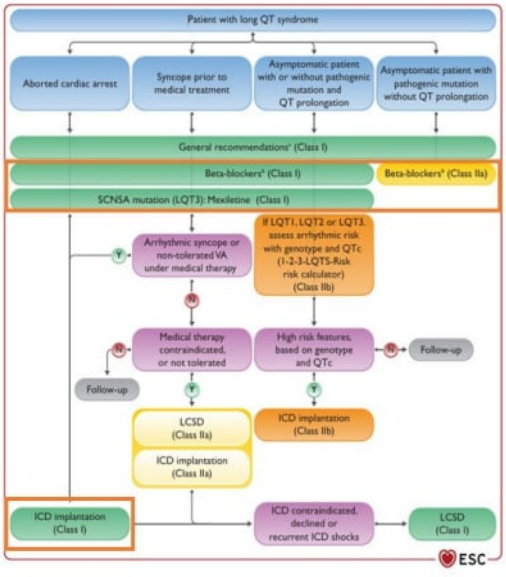
Medical β-blocker for Long QT syndrome



- β-blockers as 1st line treatment (IB)
  - ideally non-selective β-blockers
- Mexiletine for SCN5A mutation (IC)
- ICD implantation (IC)
  - symptomatic patients while receiving β-blockers

→ Contraindications for ICD therapy
→ Medical intolerance or low compliance
→ Need medication with an ICD
→ Multiple shocks, VT/VF, or syncope despite medication

European Heart Journal (2022) 43, 3997–4126

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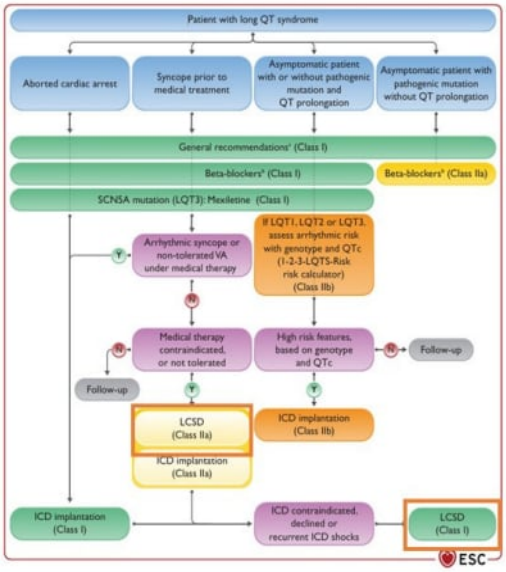
Medical β-blocker for Long QT syndrome



→ Contraindications for ICD therapy
→ Medical intolerance or low compliance
→ Need medication with an ICD
→ Multiple shocks, VT/VF, or syncope despite medication

➔ Considering **surgical β-blocker**
- class I, if ICD is contraindicated/declined, or recurrent ICD shocks are identified
- class IIa, if medical treatment is contraindicated or non tolerated

European Heart Journal (2022) 43, 3997–4126

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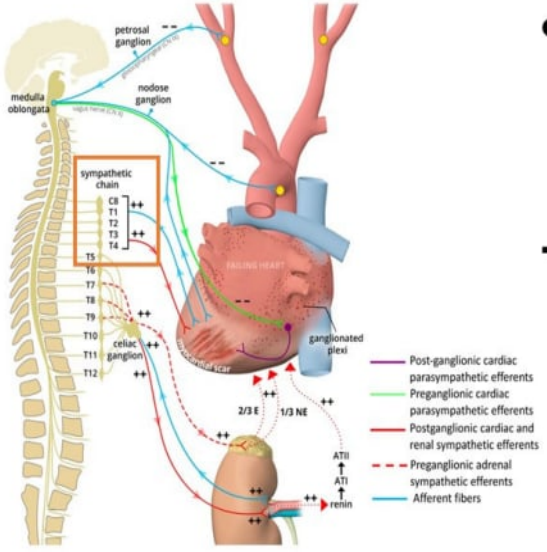
Left Cardiac Sympathetic Denervation (LCSD)



● Extracardiac intrathoracic ganglia
- sympathetic ganglia of sympathetic chain
- sympathtetic stimulation increases in positive chronotropy, inotropy, dromotropy via catecholamines

➔ Removal sympathetic ganglia for preventing surge of catecholamines
= surgical β-blockers

Circulation (2004) 109, 1826-1833
Pacing Clin Electrophysiol (2020) 43(2) 172-180

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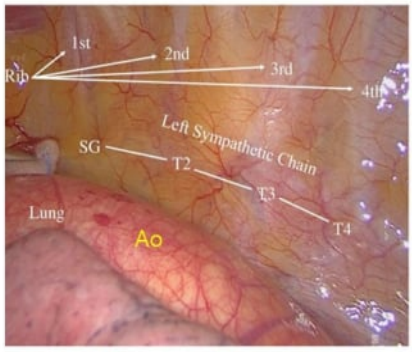
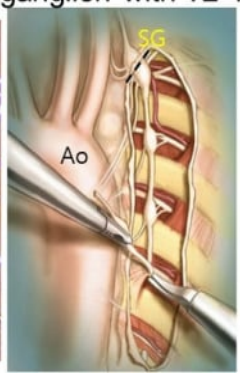
Left Cardiac Sympathetic Denervation (LCSD)

➔ Removal sympathetic ganglia for preventing surge of catecholamines
- first introduced in late 1960s (left stellectomy)
- first intervention in 1970 by J Moss
- removal of lower half of stellate ganglion with T2-T4





Circulartion (1985) 71(1), 17-21
Circulation (2004) 109, 1826-1833
Pacing Clin Electrophysiol (2020) 43(2), 172-180
Heart Rhythm O 2 (2025) 6, 1652-1658

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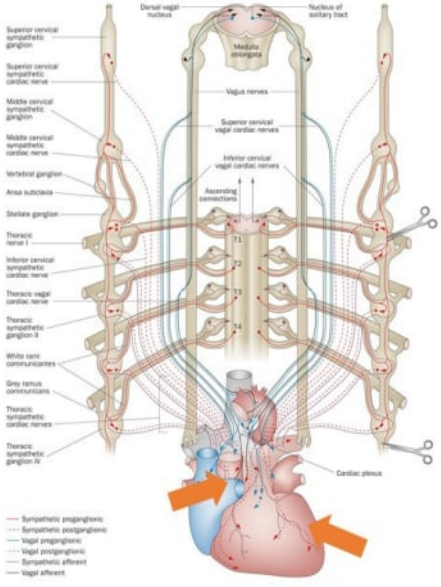
Left Cardiac Sympathetic Denervation (LCSD)



- Right sympathetic chain
  - primary innervation target: SA node
  - dominantly chronotropic effect
  - less directly responsible for ventricular triggers
- Left sympathetic chain
  - primary innervation target: AV node and ventricular myocardium
  - more likely related to ventricular arrhythmias

➔ For LQTS, LSCD is primarily indicated

Nature review of cardiology (2014) 11, 346-353

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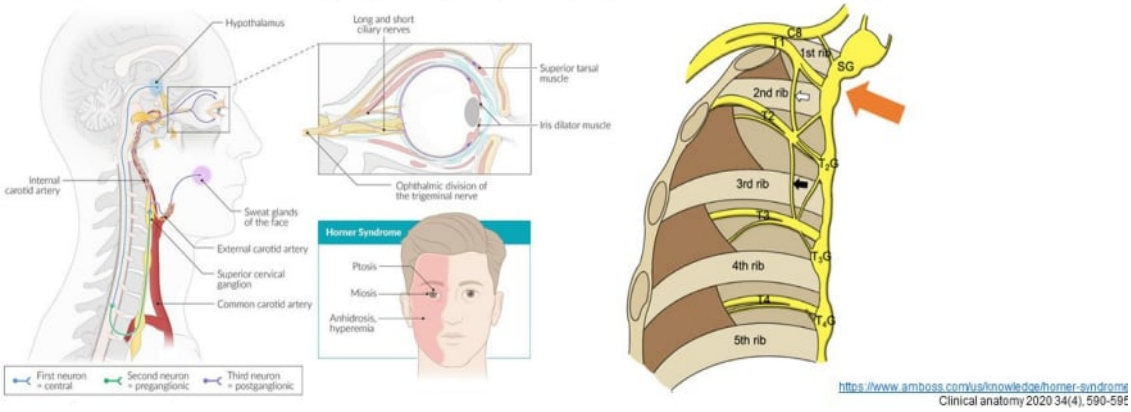
Left Cardiac Sympathetic Denervation (LCSD)

- Anatomical details: lower half of left stellate ganglion & T2-T4 ganglion
  - for preventing Horner's syndrome
    - → upper half of stellate ganglion (C7) for sympathetic nerve on eye & face



https://www.amboss.com/us/knowledge/horner-syndrome
Clinical anatomy 2020 34(4), 590-595

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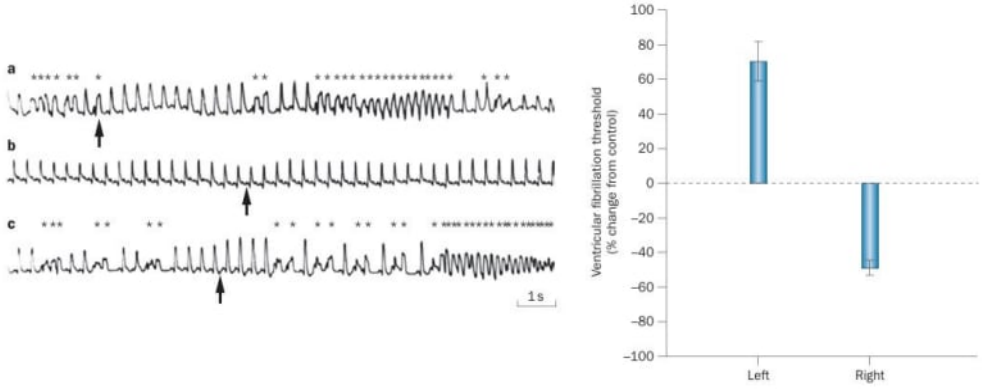
Left Cardiac Sympathetic Denervation (LCSD)

- Expected outcomes after LCSD
  - increasing threshold for life-threatening ventricular arrhythmia



Nature review of cardiology (2014) 11, 346-353

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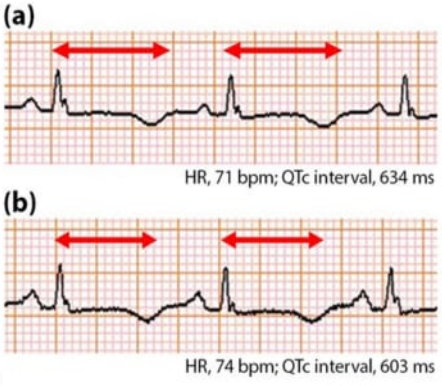
Left Cardiac Sympathetic Denervation (LCSD)

- Expected outcomes after LCSD
  - decrease in QTc interval: about 30-40ms

Table 1 Patient characteristics, LCSD secondary prevention: Summary of patients who received an LCSD at our institution as secondary preventative therapy

Case no.	Sex	Age at LCSD (years)	Genotype	Prior failed therapies	QTc Pre/Post (ms)	No. events/ACA/ ICD shocks before LCSD	No. events/ACA/ ICD shocks since LCSD
1	M	2 mo	LQT−	Beta-blockers, lidocaine, mexiletine	651/645	1	0
2	F	42	LQT2	Beta-blockers	584/560	15	1
3	F	1	LQT−	Beta-blockers, lidocaine, flecainide, mexiletine	451/494	1*	0
4	M	7 mo	LQT2	Beta-blockers	477/464	2	0
5	M	6	JLNS1	Beta-blockers	507/440	1	2
6	M	3 mo	LQT3	Beta-blockers, lidocaine	687/575	1.5 TdP/day	1.4 TdP/day
7	F	16	CPVT	Beta-blockers, mexiletine	N/A	10	0
8	M	1	LQT−	Beta-blockers	529/444	1	0
9	M	9	LQT1	Beta-blockers	500/498	2	0
10	F	4	LQT−	Beta-blockers, mexiletine	546/598	16	0
11	M	15	JLNS1	Beta-blockers	562/512	40	0



Hear Rhythm (2009) 6, 752-759
Clinical Reports (2025) 11, 60

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Left Cardiac Sympathetic Denervation (LCSD)

- Expected outcomes after LCSD

TABLE 2. Effect of LCSD on the Rate of Events During Follow-Up

	Patients, n	Median Event Count per Patient (IQR)	Mean Annual Event Rate (95% CI)*	IRR (95% CI)	Change in Expected Count, %	P
Any event						
Before LCSD	139/141	6 (3–11)	1.32 (1.25–1.40)	1		
After LCSD	77/141	1 (0–2)	0.19 (0.17–0.22)	0.09 (0.06–0.14)	91	<0.001
ACA/SD						
Before LCSD	65/141	0 (0–1)	0.13 (0.11–0.15)	1		
After LCSD	32/141	0 (0–0)	0.06 (0.04–0.07)	0.44 (0.19–0.98)	56	0.046
Any event (patients with syncope only)						
Before LCSD	74/74	5 (3–10)	1.23 (1.13–1.33)	1		
After LCSD	41/74	1 (0–2)	0.24 (0.20–0.28)	0.09 (0.05–0.17)	91	<0.001

Circulation (2004) 109, 1826-1833
Heart Rhythm O 2 (2025) 6, 1652-1658

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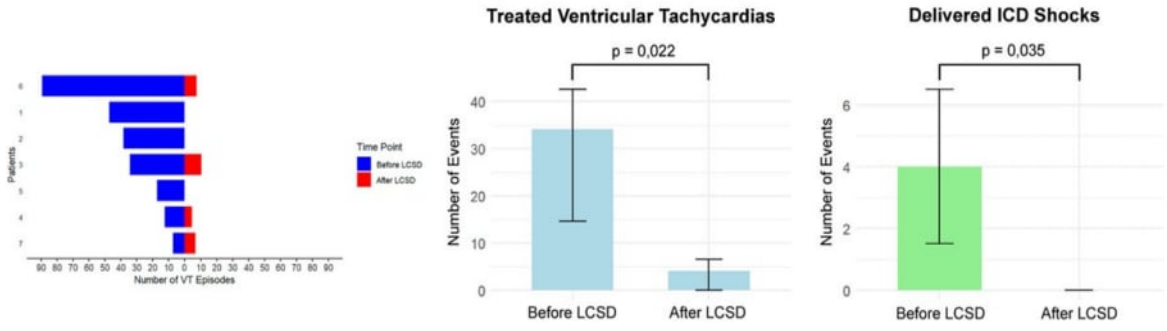
Left Cardiac Sympathetic Denervation (LCSD)

- Expected outcomes after LCSD



Circulation (2004) 109, 1826-1833
Heart Rhythm O 2 (2025) 6, 1652-1658

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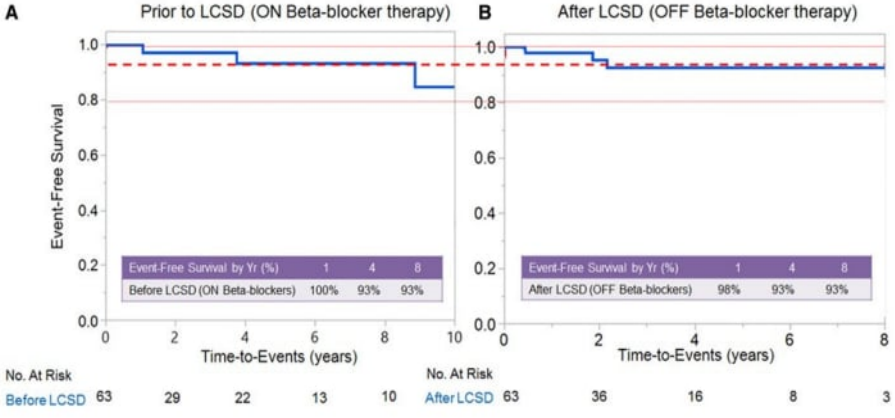
Left Cardiac Sympathetic Denervation (LCSD)

- Expected outcomes after LCSD



Circ Arrhythm Electrophysiol 2020 (13), 1443-1450

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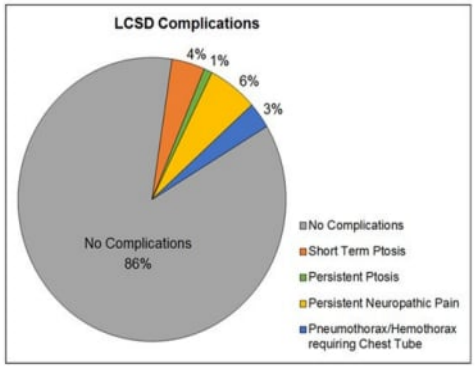
Left Cardiac Sympathetic Denervation (LCSD)

- Expected complications
  - ptosis/Horner syndrome: 5~20%
  - pneumothorax / hemothorax: 10~20%
  - neurogenic pain

Table. Comparison of Left Cardiac Sympathetic Denervation in Arrhythmia and Hyperhidrosis Literature

First Author	Yr of Publication	n	Any Complication, %	Compensatory Hyperhidrosis, %	Ptosis/Horner Syndrome, %	Pneumothorax, %
Arrhythmia literature						
Bos et al[11]	2013	52	13	-	8	-
Hofferberth et al[13]	2014	24	13	-	-	8
Roston et al[15]	2015	18	16	-	11	3
Olde Nordekamp et al[14]	2014	17	23	-	6	18
Coleman et al[12]	2012	27	26	-	11	15
Vaseghi et al[16]	2014	41	37	10	7	10
Schneider et al[18]	2013	10	70	-	70	6
Waddell-Smith et al[10]	2015	47	95	56	21	11



Circ Arrhythm Electrophysiol 2015 (8), 1007-1009
Circ Arrhythm Electrophysiol 2020 (13), 1443-1450

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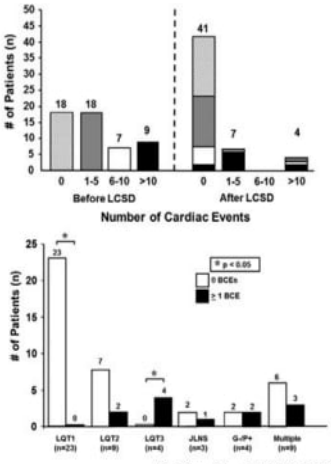
Left Cardiac Sympathetic Denervation (LCSD)

- For special populations: pediatric patients
  - incidence of major cardiac events decreases more than 85~90%
  - free from ICD and bridges to ICD
  - especially effective for LQT1 patients

	Study Cohort	Primary Prevention	Secondary Prevention	*P* Value	0 BCEs	≥1 BCE	*P* Value
n	52	33	19		40	12	
Males/females	24/28	12/21	12/7	0.06	17/23	7/5	0.5
Age at diagnosis, y	10.0±10	12.0±8	6.4±11	0.05	11.2±8	5.7±12	0.1
Baseline QTc, ms	528±74	505±44	568±97	0.01	507±45	597±106	0.01
β-Blocker, n (%)	51 (98)	32 (97)	19 (100)	1.0	39 (98)	12 (100)	1.0
Na-blocker, n (%)	45 (87)	1 (3)	6 (32)	0.007	2 (5)	5 (42)	0.005
ICD before LCSD, n (%)	16 (31)	3 (9)	13 (68)	<0.001	6 (15)	10 (83)	<0.001
Indication for LCSD							
β-Blocker intolerance, n (%)	17 (33)	17 (52)	0 (0)		17 (43)	0 (0)	
Severe LQTS, n (%)	25 (48)	16 (48)	9 (47)	<0.001	18 (45)	7 (58)	0.005
Breakthrough event pre LCSD, n (%)	10 (19)	0 (0)	10 (53)		5 (13)	5 (42)	
Age at surgery, y	14.1±10	15.6±9	11.3±11	0.1	15.4±9	9.5±12	0.1
Follow-up time, y	3.6±1.3	3.3±1	3.9±1	0.1	3.3±1	4.2±1	0.06
BCE post LCSD, n (%)	12 (23)	0 (0)	12 (63)	<0.001	…	…	



Ann Thorac Surg (2008) 86, 1620-1625
Clin Res Cardiol (2013) 102, 33–42
Circ Arrhythm Electrophysiol (2020) 13. 1443-1450

YONSEI UNIVERSITY

Left Cardiac Sympathetic Denervation: real world cases

- 38/F

- VF arrest d/t long QT syndrome 2012-10-17
  - ✓ QTc 493ms, Torsade de Pointes, syncope without stress, sudden death in family members <30years without defined cause → total 6.5point, LQTS diagnosed
  - ✓ 2012-10-17 ~ 2012-10-19 refractory VF, about 45 defibrillation needed
  - ✓ 2012-11-02 ICD insertion
- Refractory & symptomatic event since 2020
  - ✓ 2019년 이전 1년에 2~3차례 / 2020년 이후 2-3개월에 2-3차례 event 빈도 증가함
  - ✓ 2023-08-18 ICD generator change
- 2025-08-21 ER 내원 ICD shock *5

  → decide LCSD 2025-09-05

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Left Cardiac Sympathetic Denervation: real world cases

- 38/F LCSD 2025-09-05

YONSEI UNIVERSITY

Left Cardiac Sympathetic Denervation: real world cases

- 2/M
- VF arrest d/t long QT syndrome 2025-03-08
- 2025-05-28 ICD insertion
  - ✓ QTc 543ms, syncope with stress
  - ✓ SCN5A mutation
  - ✓ 2025-05-28 ICD insertion
- 2026-05-04 ~ 2026-05-12 recurrent ICD shock, LOC
  → decide LCSD 2026-05-19

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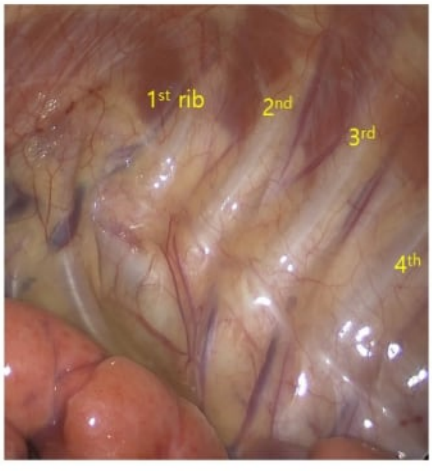
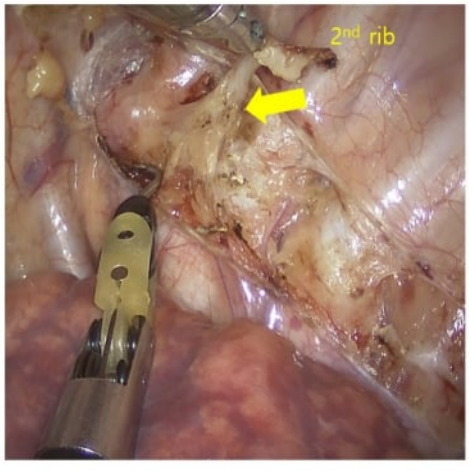
Left Cardiac Sympathetic Denervation: real world cases

- 2/M LCSD 2026-05-19





YONSEI UNIVERSITY

Left Cardiac Sympathetic Denervation: real world cases

- 2/M Follow up
  - no more shock event (VF self termination)
  - Horner's syndrome
    - ✓ Thermal injury
    - ✓ Traction injury
    - ✓ Ischemic damage
    - ✓ Anatomical variation

    → Need to find out temporary vs permanent symptoms

Short QT syndrome; Short but not least

김 주 연

성균관의대 순환기내과

10:40~12:00	Session 2. Long and Short QT Syndromes; Stop the Short-Long-Short Sequence!	
	좌장: **배은정** (서울의대 소아청소년과), **조용근** (경북의대 순환기내과) 패널: **양소영** (가천의대 심장내과), **안효정** (서울의대 순환기내과), **정주희** (고려의대 순환기내과), **차누리** (연세의대 소아청소년과)	
10:40~10:55	Diagnosis and genetics of long QT syndrome; How long is long?	백재숙 (울산의대 소아청소년과)
10:55~11:10	How to manage each type of long QT syndrome; Long life with long QT	송미경 (서울의대 소아청소년과)
11:10~11:25	Sympathectomy for long QT syndrome; Surgical β-blocker	김하은 (연세의대 흉부외과)
11:25~11:40	Short QT syndrome; Short but not least	김주연 (성균관의대 순환기내과)
11:40~12:00	Discussion	
12:00~13:00	Lunch	

Contents

- Definition
- Epidemiology
- Clinical presentation
- Risk stratification
- Treatment
- Future direction

Short QT syndrome

- Inherited arrhythmia syndrome characterized by a markedly shortened QT interval on the surface electrocardiogram (ECG) and a high risk of sudden cardiac death

- Short QT syndrome (SQTS) is a rare genetic disorder characterised by a short QT interval, premature AF and VF in the context of a structurally normal heart

Pivotal observation was made in 1998

- Family with a dramatic history of sudden cardiac death
- Extremely short QT, with an almost absent ST segment and tall, narrow, peaked T waves
- 6 family members experienced sudden cardiac death or resuscitated cardiac arrest, 1 family member also had early-onset atrial fibrillation
- EPS : markedly shortened atrial and ventricular effective refractory periods, and programmed electrical stimulation demonstrated easy inducibility of ventricular fibrillation

Heart Rhythm 2026;23:1233–1235

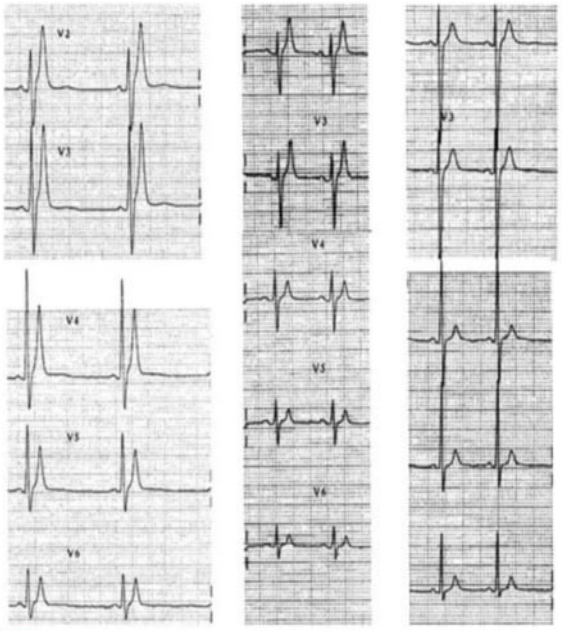


Figure 1
12-lead electrocardiograms (paper speed 25 mm/s) of family 1 patients. *Left panel*: Patient 1: heart rate 52 beats/min and QT interval 280 ms. *Middle panel*: Patient 2: heart rate 96 beats/min and QT interval 220 ms. *Right panel*: Patient 3: heart rate 92 beats/min and QT interval 260 ms. Adapted with permission from Wolters Kluwer Health, Inc., Gaita et al.[7]

CARDIOLOGY

Arrhythmias, Electrophysiology and Electrocardiography

Cardiology 2000;94:99–102
Received: May 22, 2000
Accepted after revision: August 3, 2000

Idiopathic Short QT Interval: A New Clinical Syndrome?

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Stephen L. Kopecky[a] Bernard R. Chaitman[d] Preben Bjerregaard[d]

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2000, Gussak

Case report

2003, Gaita, Borggrefe

The short QT syndrome is characterized by familial sudden death, short refractory periods, and inducible ventricular fibrillation

Circulation
Volume 108, Issue 8, 26 August 2003; Pages 965-970
https://doi-org.libproxy.samsunghospital.com/10.1161/01.CIR.0000085071.28695.C4

CLINICAL INVESTIGATION AND REPORTS

Short QT Syndrome

A Familial Cause of Sudden Death

Fiorenzo Gaita, MD, Carla Giustetto, MD, Francesca Bianchi, MD, Christian Wolpert, MD, Rainer Schimpf, MD, Riccardo Riccardi, MD, Stefano Grossi, MD, Elena Richiardi, MD, and Martin Borggrefe, MD

Definition

5. Short QT Syndrome (SQTS) *Expert Consensus Recommendations on* ***Short QT Syndrome Diagnosis***

1. SQTS ***is diagnosed*** in the presence of a QTc ≤330 ms.
2. SQTS ***can be diagnosed*** in the presence of a QTc <360 ms and one or more of the following: a pathogenic mutation, family history of SQTS, family history of sudden death at age ≤40, survival of a VT/VF episode in the absence of heart disease.

Expert Consensus Recommendations on ***Short QT Syndrome Therapeutic Interventions***

Class I	1. ICD implantation ***is recommended*** in symptomatic patients with a diagnosis of SQTS who a. Are survivors of a cardiac arrest *and/or* b. Have documented spontaneous sustained VT with or without syncope.
Class IIb	2. ICD implantation ***may be considered*** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD. 3. Quinidine ***may be considered*** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD. 4. Sotalol ***may be considered*** in asymptomatic patients with a diagnosis of SQTS and a family history of SCD.

Expert Consensus Statement on Inherited Primary Arrhythmia Syndromes

Recommendation Table 46 — Recommendations for the management of patients with short QT syndrome

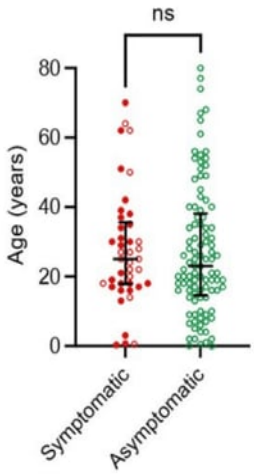
Recommendations	Class[a]	Level[b]
Diagnosis		
It is recommended that SQTS is diagnosed in the presence of a QTc ≤360 ms and one or more of the following: (a) a pathogenic mutation, (b) a family history of SQTS, (c) survival from a VT/VF episode in the absence of heart disease.[1061,1068]	I	C
Genetic testing is indicated in patients diagnosed with SQTS.[1063]	I	C
SQTS should be considered in the presence of a QTc ≤320 ms.[1064–1067,1073,1074]	IIa	C
SQTS should be considered in the presence of a QTc ≥320 ms and ≤360 ms and arrhythmic syncope.	IIa	C
SQTS may be considered in the presence of a QTc ≥320 ms and ≤360 ms and a family history of SD at age <40 years.	IIb	C

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Clinical presentation

- Of the 162 patients with SQTS, 57 (35.2%) had arrhythmic symptoms related to ventricular arrhythmias, including sudden death in 9 patients, cardiac arrest with resuscitation in 23, recorded malignant ventricular arrhythmias in 5, and malignant syncope in 39 patients, for a total of 76 events.
- The median age at the time of arrhythmic symptoms was *25.0* years
- The disease has high lethality in all age groups, including the first months of life. The probability of a first CA by the age of 40 years is >40%

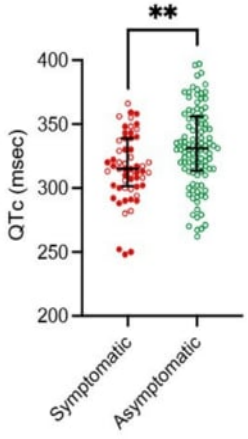
Age distribution of symptomatic and asymptomatic patients.



Heart Rhythm 2026;■:1–5

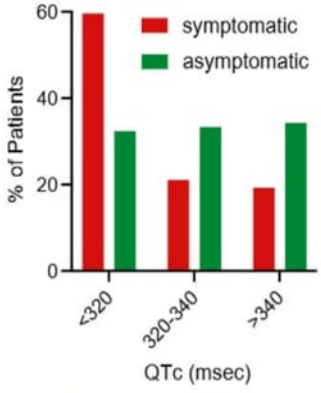
QTc and risk

QTc distribution of symptomatic and asymptomatic patients



Heart Rhythm 2026;■:1–5

Distribution of patients by QTc and the risk of symptoms.



The QTc shortens below 320 ms, the risk of malignant arrhythmic events increases significantly.

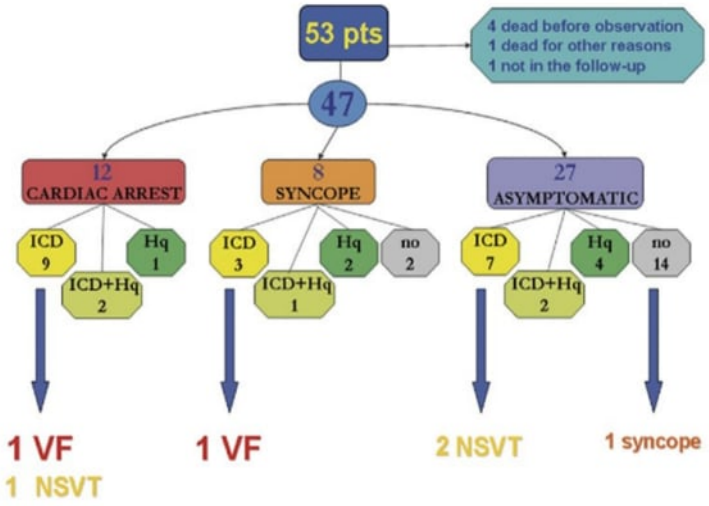
Male and risk

- The prevalence of arrhythmic symptoms was 42.5% for males and 21.4% for females
- Males were overrepresented in the entire cohort, accounting for 106 patients with SQTS (65.4%).
- Males demonstrated markedly higher odds of being symptomatic, with an OR of *3.26* (95% CI 1.75–6.09), than females.

Risk stratification, SCD prevention and treatment of VA		
ICD implantation is recommended in patients with a diagnosis of SQTS who: (a) are survivors of an aborted CA and/or (b) have documented spontaneous sustained VT.[1063]	I	C
ILR should be considered in young SQTS patients.	IIa	C
ICD implantation should be considered in SQTS patients with arrhythmic syncope.	IIa	C
Quinidine may be considered in (a) SQTS patients who qualify for an ICD but present a contraindication to the ICD or refuse it, and (b) asymptomatic SQTS patients and a family history of SCD.[1069–1071]	IIb	C
Isoproterenol may be considered in SQTS patients with an electrical storm.[1075]	IIb	C
PES is not recommended for SCD risk stratification in SQTS patients.	III	C

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Long-Term Follow-Up of Patients With Short QT Syndrome



JACC Vol. 58, No. 6, 2011:587–95

Treatment

- ICD
- Quinidine (KCNH2 mutation)

- The optimal strategy for primary prevention of cardiac arrest in SQTS is not clear

Clinical and electrocardiographic characteristics of patients with short QT interval in a large hospital-based population

Akashi Miyamoto, MD, PhD,* Hideki Hayashi, MD, PhD,* Tomohide Yoshino, MD,*
Tamiro Kawaguchi, MD,* Atsushi Taniguchi, MD,* Hideki Itoh, MD, PhD,*
Yoshihisa Sugimoto, MD, PhD,* Makoto Itoh, MD, PhD,* Takeru Makiyama, MD, PhD,†
Joel Q. Xue, PhD,‡ Yoshitaka Murakami, PhD,§ Minoru Horie, MD, PhD*

BACKGROUND Short QT syndrome is one of the underlying disorders associated with ventricular fibrillation. However, the precise prognostic implication of a short QT interval remains unclear.

OBJECTIVE The purpose of this study was to investigate the prevalence and long-term prognosis in patients with a shorter-than-normal QT interval in a large hospital-based population.

METHODS We chose patients with a short Bazett QTc interval from a database consisting of 114,334 patients to determine the clinical characteristics and prognostic value of a short QT interval.

RESULTS A total of 427 patients (mean age 43.4 ± 22.4 years) had a short QT interval with about a 1.2 times higher male predominance (234 men). The QTc interval was significantly longer in female than in male patients (363.8 ± 6.1 ms vs 357.1 ± 5.8 ms, $P < .0001$). The age-specific prevalence of patients with short QT interval was biphasic, peaking at young and old age. Atrial fibrillation and early repolarization were complicated with short QT interval in 39 (9.1%) and 26 (6.1%) patients, respectively. The prognosis of 327 patients (182 men; mean age, 46.4 ± 27.3 years) with a short QT interval could be assessed (mean follow-up period, 54.0 ± 62.0 months). During the follow-up, 2 patients, 1 of whom had early repolarization, developed life-threatening events, in contrast to 6 patients who died of noncardiac causes and did not have early repolarization.

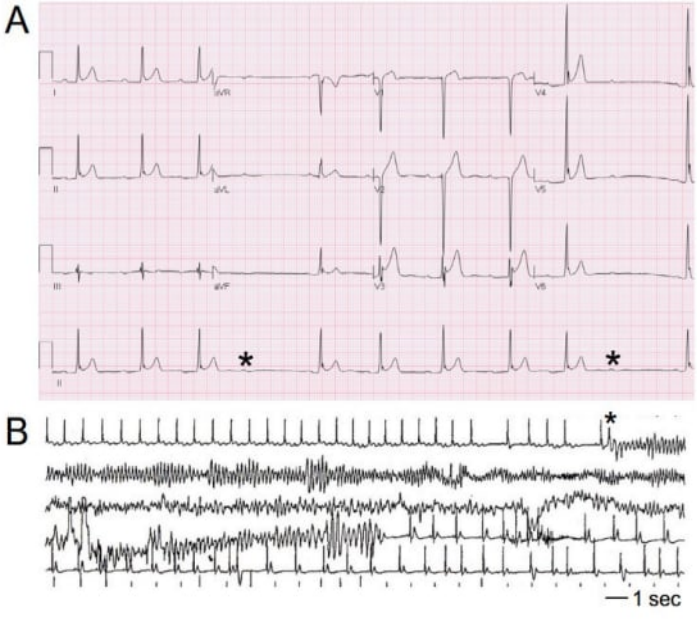
CONCLUSION The prevalence of a short QT interval showed a slight male preponderance and biphasic age-dependent distribution in both genders. The complication rate of atrial fibrillation was higher in those with a short QT interval than in general populations. The long-term outcome suggested that early repolarization in a short QT interval might be associated with potential risk of lethal arrhythmia.

KEYWORDS Electrocardiography; QT interval; Prevalence; Prognosis; Repolarization

ABBREVIATIONS **AF** = atrial fibrillation; **CI** = confidence interval; **ECG** = electrocardiogram

Heart Rhythm 2012;9:66–74

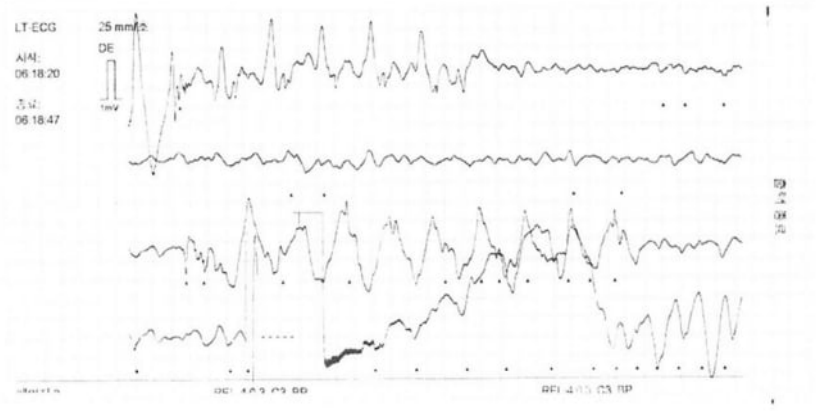
22-year-old man who exhibited early repolarization



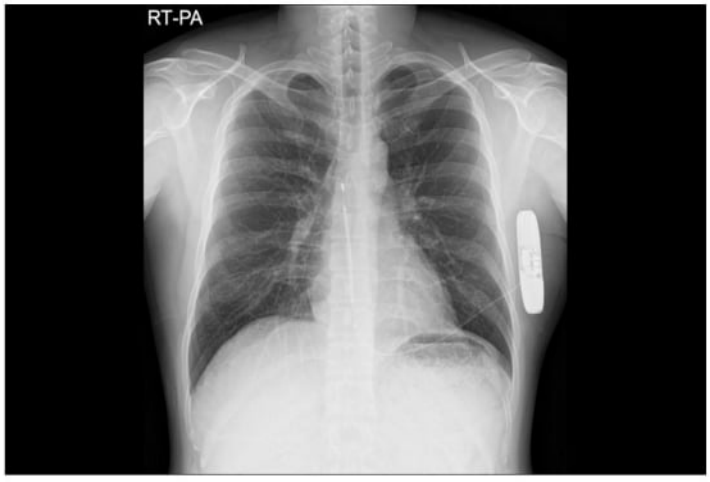
Heart Rhythm 2012;9:66–74

M/31

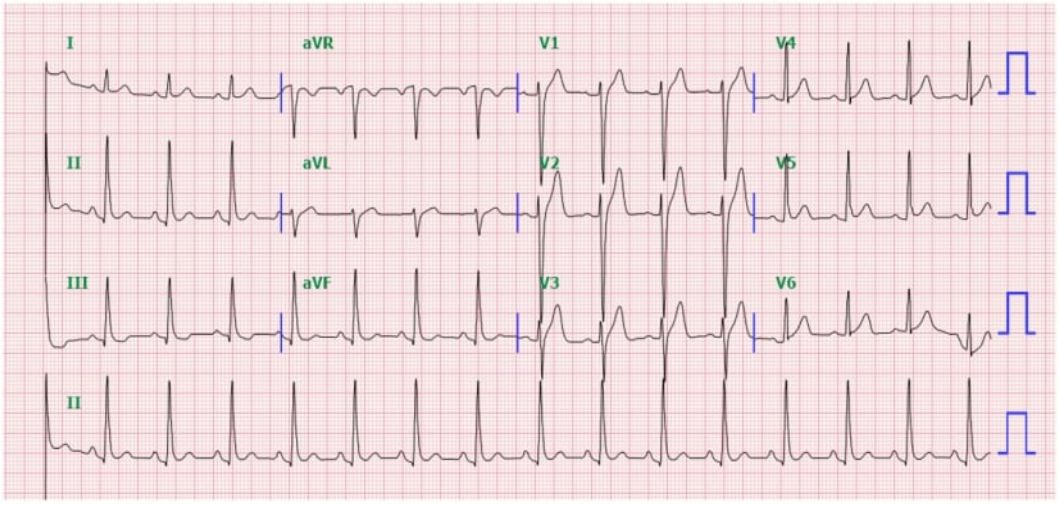
- 수면 중 자다가 손위로 드는 자세 취하며 눈돌아간 모습 보호자에 의해 확인 후 119 연결, 119에서 VF 2회 제세동 후 ROSC



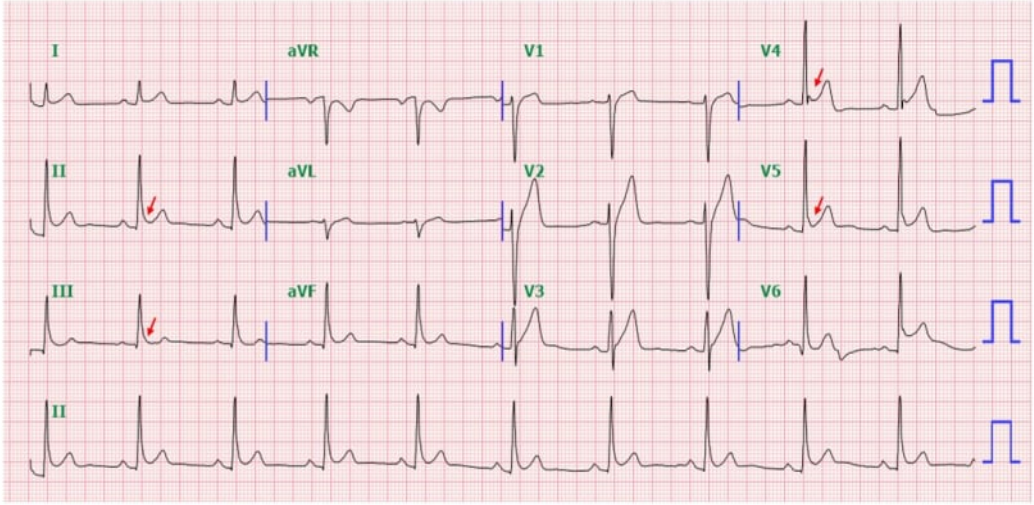
Echocardiogram, CAG, CMR : normal



QTc 377ms



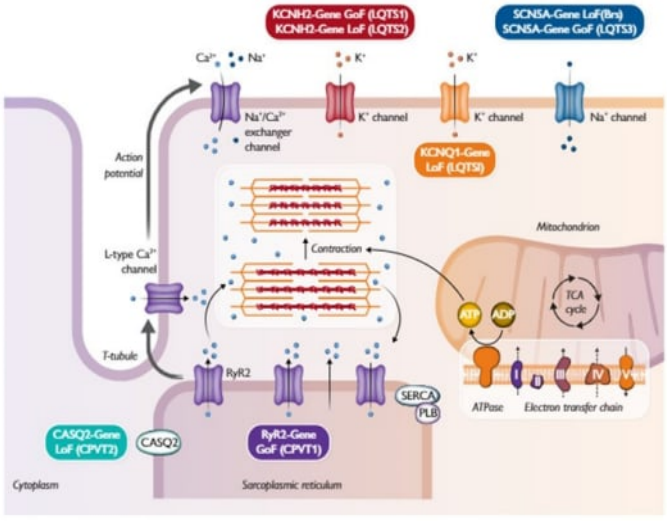
QTc 383ms



Genetic basis

Genetic basis

- DNA variants in 3 potassium channel genes : KCNH2,KCNQ1,KCNJ2
- Gain of function mutations in KCNH2, KCNQ1 and loss of function in SLC4A



European Heart Journal (2026) 47, 2982–2998

Table 1 Genetic results of genotype-positive patients

Culprit gene	Reported mutations	Accession	Number of cases	ACMG classification	Allele frequency
KCNH2 ↑	E50D	NM_000238.4:c.150G>T	1	LP	6.85e-7
	I560T	NM_000238.4:c.1679T>C	1	LP	0
	N588K	NM_000238.4:c.1764C>A	11	P	0
	T618I	NM_000238.4:c.1853C>T	1	P	0
	R1135H	NM_000238.4:c.3404G>A	3	VUS	2.1e-6
KCNQ1	V307L	NM_000218.3:c.919G>T	1	LP	0
	V141M	NM_000218.3:c.421G>A	13	P	0
KCNJ2	D172N	NM_000891.3:c.514G>A	2	P	0
	M301K	NM_000891.3:c.902T>A	1	P	0
	K346T	NM_000891.3:c.1037A>C	2	VUS	0
SLC4A3 ↓	R343H*	NM_005070.4:c.1028G>A	21	P	6.8e-7
	F347S	NM_005070.4:c.1040T>C	4	LP	0
	G359V*	NM_005070.4:c.1076G>T	2	LP	0
	P428R†	NM_005070.4:c.1283C>G	1	VUS	0
	R573C	NM_005070.4:c.1717C>T	1	LP	0
	R925H	NM_005070.4:c.2774G>A	2	VUS	2.1e-6
	R1016G	NM_005070.4: c.3046C>G	11	LP	0
	A1068V‡	NM_005070.4:c.3203C>T	1	VUS	3.4e-6
	P1069H	NM_005070.4:c.3206C>A	3	LP	0

Heart Rhythm 2026;■:1–5

(1) Variant specific characteristics

Circulation Journal
Circ J 2026; **90**: 767–772
doi:10.1253/circj.CJ-24-0927

REVIEW

Congenital Short QT Syndrome
— Review Focused on *KCNQ1* p.Val141Met Variant —

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Short QT syndrome (SQTS) is a very rare inherited arrhythmia characterized by extremely short QT intervals on electrocardiograms and sudden cardiac death in young patients. Among the genotypes of SQTS, gain-of-function variants in the potassium voltage-gated channel subfamily Q member 1 (*KCNQ1*) gene are accountable for SQTS type 2 (SQT2). Pathogenic variants for SQT2 are rare and, among them, the p.Val141Met is relatively prevalent. This review summarizes findings for 5 SQTS patients harboring p.Val141Met we recently encountered and compares them to another 14 patients reported in the literature.

KCNQ1 p.Val141Met Variant

- While most SQT variants primarily cause isolated ventricular arrhythmias, KCNQ1 p.Val141Met exhibits a uniquely complex phenotype.

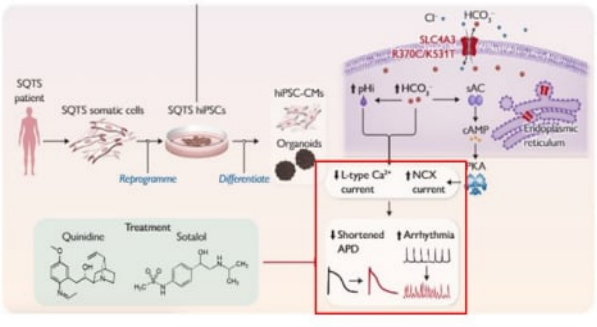
- Distinct clinical feature
  - Perinatal/fetal bradycardia and SSS
  - AF SVR and atrial standstill with junctional rhythm
  - Bradycardia induced early onset LV dilatation

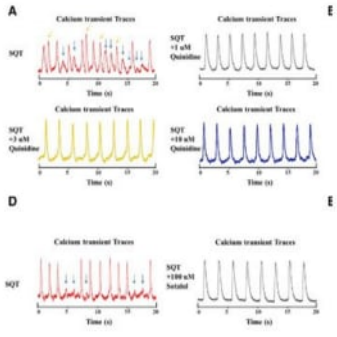
Table. (A) Demographics of *KCNQ1* Val141Met-Positive Probands, (B) Demographics of *KCNQ1* Val141Met-Positive Family Members of the Probands

(A) Proband	Age at genetic testing (years)	Sex	Pattern of inheritance	Rhythm	RR (ms)	QT (ms)	QTc (ms)
1	2	M	Paternal	SR	1,162	326	302
2	1	F	Paternal	SR	1,304	358	315
3	4	M	De novo	SR	650	252	313
4	0	F	De novo	SR	1,000	280	280
5	3	F	ND	ND	ND	ND	<330
6	2	F	ND	SR	870	260	279
7	6	F	ND	AF	1,000	200	292
8	9	F	ND	AF	675	260	317
9	10	F	Paternal	JR	938	280	293
10	1	F	De novo	AF	ND	ND	270
11	1	M	De novo	AF	1,160	312	290
12	1	F	De novo	AF	1,280	328	290
13	19	F	ND	SR	840	260	283
14	26	F	ND	SR	ND	ND	330
15	0	F	ND	ND	ND	ND	277
16	1	F	De novo	SR	ND	ND	330
(B) Family member							
17 (father of Proband 1)	37	M	ND	AF	1,065	349	339

(A) Proband	Perinatal bradycardia	AF (age at onset; years)	LV dilatation	VA (age at onset; years)	Quinidine	Device therapy (age at implantation; years)	Reference
1	Yes	Yes (2)	Yes	No	No	PM (2)	14
2	Yes	Yes (0)	Yes	No	No	PM (3)	This study
3	Yes	No	Yes	No	No	PM (4)	This study
4	Yes	Yes (fetus)	No	No	No	No	6
5	Yes	Yes (3)	ND	No	No	No	3
6	Yes	No	ND	No	Yes	No	9
7	Yes	Yes (fetus)	ND	No	Yes	PM (0)	8
8	Yes	Yes (fetus)	ND	No	No	No	8
9	Yes	Yes	No	No	No	PM (0)	11
10	Yes	Yes	Yes	No	No	PM (2)	10
11	Yes	Yes	Yes	No	Yes	PM (0)	10
12	Yes	Yes	Yes	No	No	No	10
13	Yes	Yes	No	No	Yes	PM (2)	12
14	ND	Yes	No	No	No	PM (8) → ICD (26)	13
15	ND	Yes	ND	No	No	No	16
16	Yes	Yes (0)	Yes	No	No	PM (1)	15
(B) Family member							
17 (father of Proband 1)	Yes	Yes (8)	Yes	VF (42)	No	PM (20) → ICD (42)	14

(2) SLC4A3-related short QT syndrome

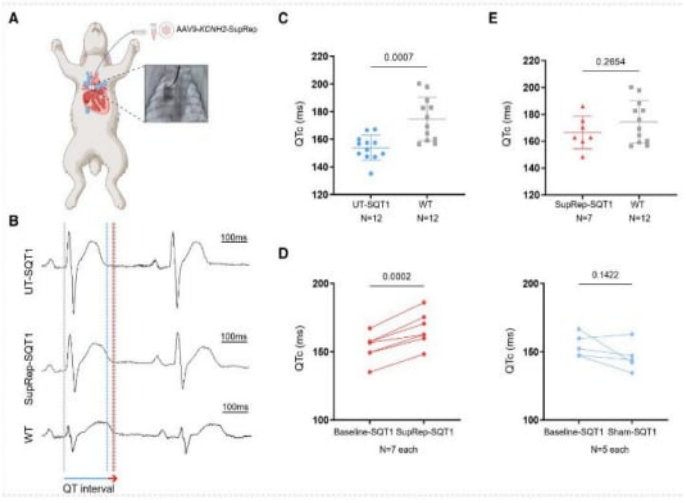




- a loss of activity of the chloride/bicarbonate exchanger
- increase in intracellular pH and a shortened QT interval
- reduced L-type calcium channel current, leading to a shortened action potential duration
- enhanced Na/Ca exchange current promoting delayed afterdepolarization-mediated arrhythmias.

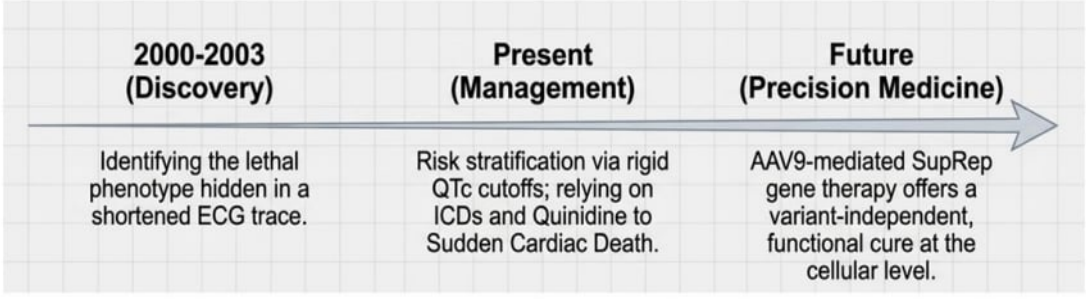
European Heart Journal (2026) 47, 2982–2998

(3) Gene therapy



- AAV9-mediated KCNH2suppression-replacement (KCNH2-SupRep) effectively prolonged the shortened QTc in transgenic SQT1 rabbit model without inducing undesirable regional repolarization heterogeneity in vivo.

European Heart Journal (2026) 47, 199–213



2026
해부학·선천성·유전성
부정맥 연구회 합동 심포지엄

Session 3

Deep Dive into Early Repolarization Syndrome

좌장: 남기병 (울산의대 심장내과), 최종일 (고려의대 순환기내과)
패널: 김명중 (가톨릭의대 순환기내과), 한석문 (서울의대 순환기내과), 황태현 (연세의대 심장내과), 이경연 (고려의대 순환기내과)

Diagnosis and genetics of early repolarization syndrome
심재민 (고려의대 순환기내과)

Risk stratefication of early repolarization syndrome; Which early repolarization is malignant?
윤남식 (전남의대 순환기내과)

How to prevent VF and sudden death in early repolarization syndrome
안민수 (연세원주의대 심장내과)

Catheter ablation for early repolarization syndrome; Is it really effective?
엄재선 (연세의대 심장내과)

대한부정맥학회
Korean Heart Rhythm Society

Diagnosis and genetics of early repolarization syndrome

심 재 민

고려의대 순환기내과

Early repolarization (ER) was traditionally considered a benign electrocardiographic variant. However, its association with idiopathic ventricular fibrillation (VF) established the concept of early repolarization syndrome (ERS). ERS and Brugada syndrome (BrS) are now recognized as two distinct but related entities within the spectrum of J-wave syndromes (JWS), sharing several electrophysiological, genetic, autonomic, and pharmacological characteristics.

A fundamental distinction should be made between the early repolarization pattern (ERP) and ERS. According to the 2026 JWS expert consensus, ERP is characterized by an end-of-QRS notch or J-wave augmentation >0.1 mV on the downslope of a prominent R wave, with or without ST-segment elevation, in at least two inferolateral leads. In contrast, ERS represents a clinical syndrome in which a characteristic ER pattern occurs in an appropriate clinical context, particularly otherwise unexplained VF or polymorphic ventricular tachycardia (VT). Thus, ERP is an ECG phenotype, whereas ERS is a clinical diagnosis.

The 2026 modified Shanghai score integrates clinical history, 12-lead and ambulatory ECG findings, and family history. Unexplained cardiac arrest or documented VF/polymorphic VT carries the greatest diagnostic weight, while the magnitude and morphology of ER and other high-risk ECG features provide additional information. Importantly, at least one ECG criterion is required, and genetic test results are not included in the ERS diagnostic score.

ERS and BrS may share an electrophysiological framework involving an outward shift of currents during the early phases of the ventricular action potential. Reduced inward sodium or calcium currents or increased outward currents can accentuate regional repolarization differences, enhance the J-wave phenotype, and promote ventricular arrhythmogenesis. However, particularly in BrS, depolarization abnormalities and an abnormal right ventricular outflow tract substrate may also contribute

to the phenotype.

Familial aggregation of ERP supports a genetic contribution, but heritability does not necessarily imply monogenic inheritance. Earlier candidate-gene studies implicated several cardiac ion-channel genes, including KCNJ8, SCN5A, CACNA1C, CACNB2, CACNA2D1, and KCND3. Subsequent reassessment has weakened the evidence for many of these genes as highly penetrant causes of ERS. For example, the relatively high population frequency of the historically reported KCNJ8 p.S422L variant argues against a highly penetrant monogenic role. Rare SCN5A loss-of-function and KCND3 gain-of-function variants have also been reported in selected ERS cases or cohorts, but neither gene can currently be regarded as a definitive monogenic cause of ERS.

Genome-wide association studies of ERP have identified common susceptibility loci, including KCND3, supporting a complex genetic contribution to the ER phenotype. Importantly, genetic associations identified for the relatively common ERP phenotype should not be assumed to represent causal genes for the much rarer ERS phenotype. Overall, current evidence suggests that the genetic architecture of ERS is heterogeneous and likely more complex than initially proposed by candidate-gene studies.

Consequently, the clinical utility of genetic testing in ERS remains limited. Routine genetic testing is not recommended in asymptomatic individuals with isolated ERP. In survivors of unexplained cardiac arrest with a convincing ERS phenotype, particularly in familial cases, selective genetic testing may be considered after comprehensive phenotyping and genetic counseling. A positive genetic finding does not by itself establish ERS, while a negative result does not exclude the diagnosis. Current expert guidance therefore supports a phenotype-first rather than genotype-first approach to ERS.

In summary, ERS remains a phenotype-driven rather than genotype-driven diagnosis. Although familial and genomic studies support a genetic contribution, no single gene currently explains a substantial proportion of ERS. The available evidence favors genetic heterogeneity and a potentially complex or polygenic architecture, emphasizing the importance of careful phenotyping and cautious interpretation of genetic findings.

Risk stratefication of early repolarization syndrome; Which early repolarization is malignant?

윤 남 식

전남의대 순환기내과

Early repolarization (ER) was traditionally considered a benign electrocardiographic finding, particularly in young individuals and athletes. However, accumulating evidence has demonstrated an association between specific ER phenotypes and idiopathic ventricular fibrillation (VF) or sudden cardiac death (SCD), establishing early repolarization syndrome (ERS) as a distinct arrhythmogenic entity. Nevertheless, because the ER pattern is relatively common in the general population, identification of the small subset of individuals at clinically significant risk remains challenging.

Several clinical and electrocardiographic features have been associated with an increased arrhythmic risk. High-amplitude J waves, particularly >2 mm, horizontal or descending ST segments, inferior or widespread J-wave distribution, and dynamic augmentation of J-point elevation are considered potentially malignant features. Clinical markers, including arrhythmic syncope, aborted cardiac arrest, a family history of unexplained SCD at a young age, and a family history of ERS, further increase concern. Importantly, J-wave and ST-segment abnormalities may be highly dynamic and can become most prominent immediately before or after ventricular arrhythmias, limiting the predictive value of a single resting electrocardiogram. Increased dispersion of repolarization may also contribute to the underlying arrhythmogenic substrate.

Current guidelines define ERS primarily in patients with an ER pattern who have survived cardiac arrest and emphasize careful evaluation of high-risk clinical and electrocardiographic features. An implantable cardioverter-defibrillator remains the cornerstone of secondary prevention. In patients with recurrent VF or ICD shocks, catheter ablation targeting triggering premature ventricular complexes, frequently originating from the Purkinje system, or localized arrhythmogenic substrates may reduce recurrent events. Experimental and clinical studies further suggest that abnormalities of repolarization and subtle structural substrates may contribute to J-wave syndromes, providing poten-

tial targets for substrate ablation and novel pharmacological therapies.

Thus, most individuals with an ER pattern have a benign prognosis, whereas a small subgroup carries substantial arrhythmic risk. Risk assessment should therefore integrate the clinical presentation, family history, J-wave amplitude and distribution, ST-segment morphology, and dynamic electrocardiographic changes rather than relying solely on the presence of ER.

How to prevent VF and sudden death in early repolarization syndrome

안 민 수

연세원주의대 심장내과

2022 ESC Recommendations for the management of patients with early repolarization pattern/syndrome

Recommendations	Class	Level
ICD implantation is recommended in patients with a diagnosis of ERS who have survived a CA	I	B
Isoproterenol infusion should be considered for ERS patients with electrical storm	IIa	B
Quinidine in addition to an ICD should be considered for recurrent VF in ERS patients	IIa	B
ILR should be considered in individuals with ERP and at least one risk featured or arrhythmic syncope	IIa	C
PVC ablation should be considered in ERS patients with recurrent VF episodes triggered by a similar PVC non-responsive to medical treatment	IIa	C
ICD implantation or quinidine may be considered in individuals with ERP and arrhythmic syncope and additional risk features	IIb	C
ICD implantation or quinidine may be considered in asymptomatic individuals who demonstrate a high-risk ERPc 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

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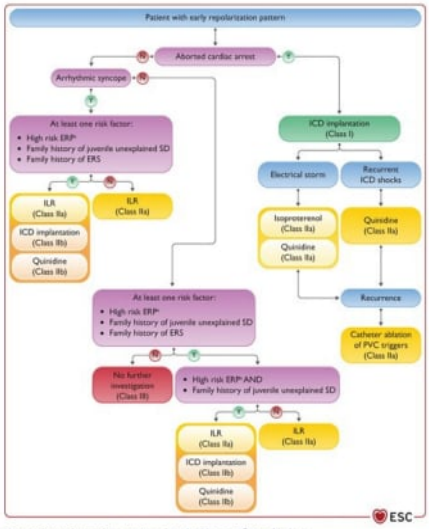
2022 ESC Recommendations for the management of patients with early repolarization pattern/syndrome



Isoproterenol infusion is effective in acute suppression of recurrent ICD discharges and electrical storm.

AADs that block the transient outward potassium current can prevent VF.

A retrospective multicentre study showed a reduction of recurrent VF after initiation of quinidine.

Phosphodiesterase-3 inhibitors such as cilostazol and milrinone also reduced the recurrence of VF.

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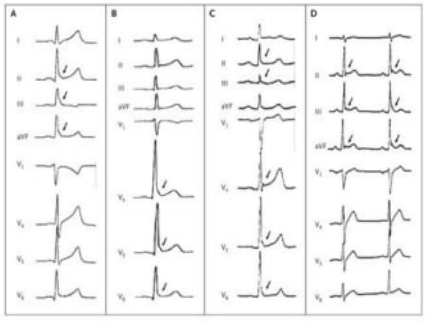
SCD associated with Early Repolarization

The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Sudden Cardiac Arrest Associated with Early Repolarization



22 international tertiary care arrhythmia centers

All subjects with the diagnosis of idiopathic VF

Baseliein ECG with early repolarization,

an elevation of the QRS–ST junction (J point) in at least two leads, (II, III, and aVF), lateral lead (I, aVL, and V4 to V6)

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N Engl J Med 2008;358:2016-2023

SCD associated with Early Repolarization

Actuarial Curves for Case Subjects, According to the Presence or Absence of Early Repolarization.

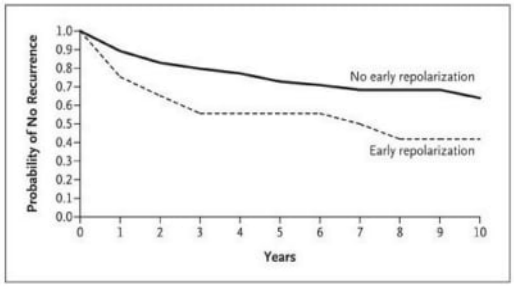


Table 2. Outcome after Initial Aborted Sudden Cardiac Arrest.*

Variable	Early Repolarization (N=64)	No Early Repolarization (N=142)	P Value
Duration of follow-up (mo)			0.81
Mean	60±45	62±52	
Median	49	54	
Interquartile range	24–90	17–92	
No. of recurrent episodes of ventricular fibrillation per patient			0.001
Median	8	2	
Interquartile range	2–35	1–6	
Successful treatment (no./total no.)†			
Beta-blockers	2/13	9/17	
Amiodarone	0/7	3/7	
Flecainide, cifenline, or pilsicainide	0/10	2/4	
Quinidine or disopyramide	4/4	1/3	
Verapamil	0/5	3/8	
Mexiletine	0/5	0/2	
Catheter ablation	5/8	6/7	
Current outcome			
No. of subjects alive	63	142	
No. of subjects with recurrence in the past 12 mo	5‡	5	

* Plus–minus values are means ±SD.
† Successful treatment was defined as no ventricular fibrillation for at least 12 months, as documented by an implantable defibrillator.
‡ Quinidine was recently prescribed for three subjects.

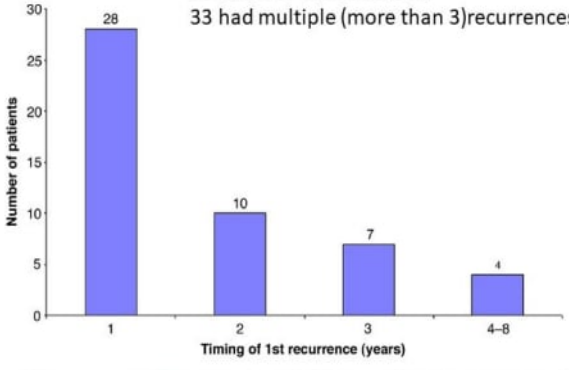
Characteristics of Recurrent Ventricular Fibrillation Associated With Inferolateral Early Repolarization

Thirty tertiary care arrhythmia centers
122 patients with the diagnosis of idiopathic VF and early repolarization on the ECG

Table 1 Characteristics of the Population

Variable	n (Available Data)	Mean ± SD or Percentage
Age (yrs)	122	36 ± 12.4
Sex		
Women	32	26%
Men	90	74%
Familial history of sudden death		
Yes	21	18%
No	98	82%
Syncope		
Yes	39	33%
No	78	67%
PR >200		
Yes	5	4%
No	110	96%
Amplitude of J-wave (mV)	112	0.21 ± 0.09
QRS duration (ms)	119	91.8 ± 10.4
QTc (ms)	117	394.7 ± 26.0
EP study: VF induced		
Yes	22	28%
No	57	72%

73 had no recurrence of VF,
16 had 1 to 3 recurrences,
33 had multiple (more than 3)recurrences.



Timing of Recurrence of Ventricular Fibrillation After the Initial Cardiac Arrest

Haïssaguerre M, et al. J Am Coll Cardiol. 2009;53(7):612–619.

Characteristics of Recurrent Ventricular Fibrillation Associated With Inferolateral Early Repolarization

Table 2 Characteristics of 33 Patients With Multiple VF Versus Other 89 Patients

Variables	n = 33	n = 89	p Value
Age (yrs)	38 ± 13.5	36 ± 11.9	0.34
Amplitude of J-wave (mm × 10)	24.2 ± 11.5	19.5 ± 7.8	**0.04**
QRS duration	93.9 ± 13.5	91.1 ± 9.0	0.29
QTc	393.7 ± 31.9	395.1 ± 23.8	0.82
Men (vs. women)	21 ± 63.6%	69 ± 77.5%	0.12
Family history of sudden death	5 ± 15.6%	16 ± 18.4%	0.73
Syncope	19 ± 57.6%	20 ± 23.8%	**5.10^{-4}**
EP study: VF induced	11 ± 47.8%	11 ± 19.6%	**0.01**

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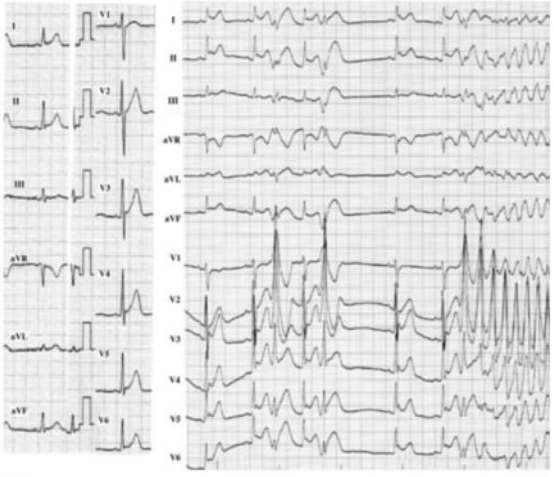
Haïssaguerre M, et al. J Am Coll Cardiol. 2009;53(7):612–619.

Characteristics of Recurrent Ventricular Fibrillation Associated With Inferolateral Early Repolarization

Amplification of Early Repolarization Pattern During the Arrhythmic Period



Twelve lead ECGs recorded during the arrhythmic periods showed a consistent increase in the amplitude of early repolarization compared with that seen in baseline

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Haïssaguerre M, et al. J Am Coll Cardiol. 2009;53(7):612–619.

Characteristics of Electrical Storms Associated With Inferolateral Early Repolarization

Table 3 Clinical and Therapeutic Data in 16 Patients With Electrical Storm

Sex, Age (yrs)	VF During Storm (n)	Origin of Ectopy	Timing After ICD Implantation (Months)	Ineffective AAD	Effective Therapy	Long-Term Therapy
M, 13	>50	LV + RV	0	Lido Procai BB Verap	Isoproterenol	No AAD
F, 52	>200	LV	0	BB Amio		*
M, 37	18	RV	84	Lido BB Amio Bretyl	Sedation	Quinidine
M, 59	52	LV	1	Lido BB Amio	Isoproterenol/quinidine	*
F, 45	50	LV + RV	0	Amio	Isoproterenol	No AAD
F, 16	38	LV + RV	15	Lido Fleca BB Amio Verap	Isoproterenol/quinidine	Quinidine
F, 37	8	LV	96	Lido Mexi BB	Quinidine	Quinidine
M, 24	28	RV	0	Lido	Isoproterenol	Cath ablation
F, 26	6	RV	0	Lido BB Amio	Isoproterenol	Cath ablation
M, 34	8	LV	0		Amio	No AAD
M, 29	5	LV	0	BB	Amio	No AAD
F, 48	17	LV	0	Mexi BB Verap	Sedation	Cath ablation
M, 38	14	LV	3	BB	Amio	No AAD
M, 50	20	RV	0	Lido BB Amio	Isoprenaline	Cath Ablation
F, 59	4	LV	20	Lido	Spontaneous resolution	Quinidine
F, 50	20	RV	0	Lido BB Amio	High rate pacing	Pacing + BB

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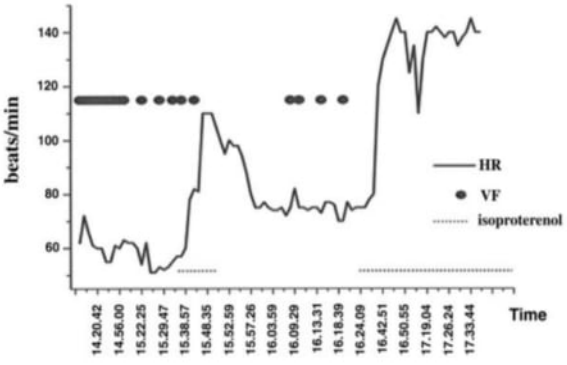
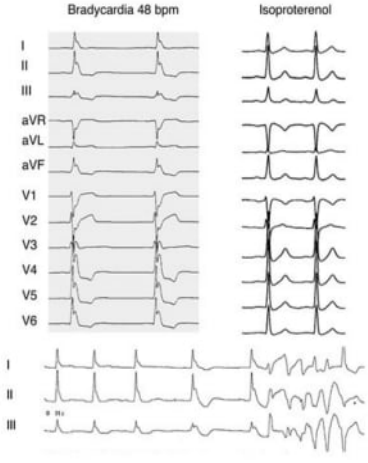
Haïssaguerre M, et al. J Am Coll Cardiol. 2009;53(7):612–619.

Isoproterenol in Electrical Storm



Immediate Correction by Isoproterenol Infusion
Sinus rate ≥120 bpm



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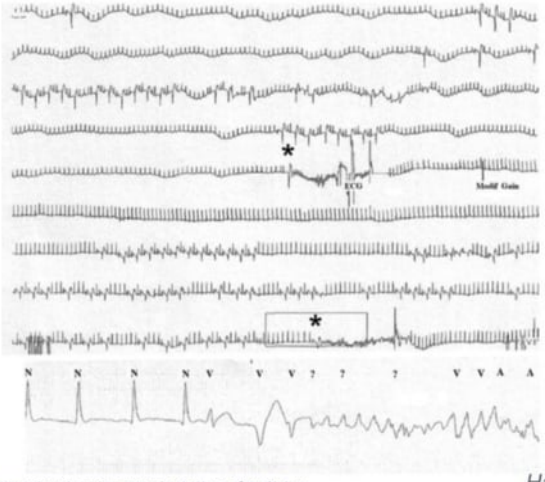
Haïssaguerre M, et al. J Am Coll Cardiol. 2009;53(7):612–619.

Isoproterenol in Electrical Storm

Recurrence of VF After Discontinuation of Isoproterenol Infusion



Any attempt to reduce isoproterenol infusion and heart rate was associated with recurrence of VF in 3 of the patients. Isoproterenol was infused for a period ranging from 6 h to 5 days.

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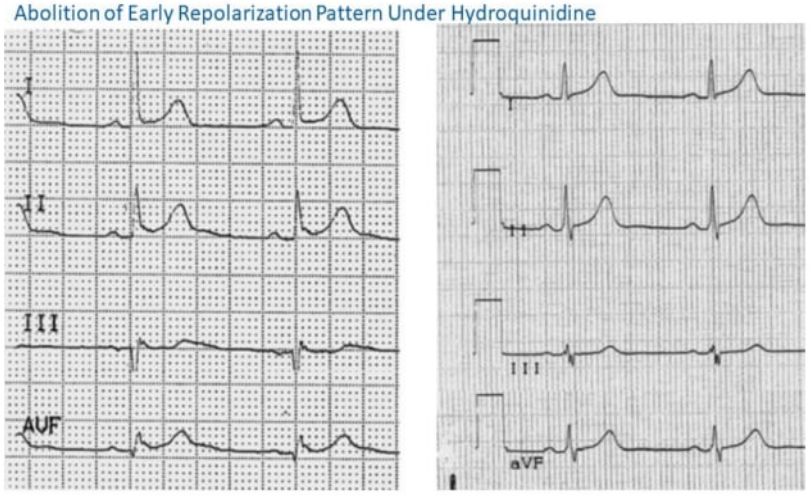
Haïssaguerre M, et al. J Am Coll Cardiol. 2009;53(7):612–619.

Quinidine in Early repolarization

Abolition of Early Repolarization Pattern Under Hydroquinidine



Quinidine (in 3) or hydroquinidine (in 6) was totally successful in 9 of 9 patients decreasing the number of recurrent VF from a mean of 33 35/median of 25 (IQR 8 to 62) episodes to nil with a current follow-up of 25 18 months on therapy.

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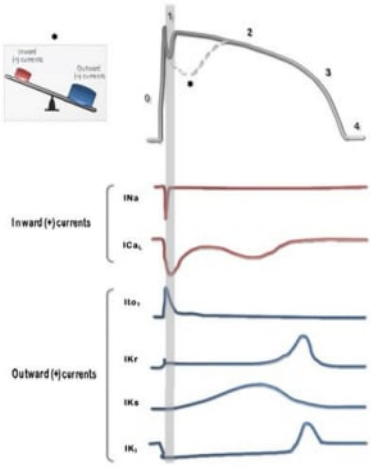
Haïssaguerre M, et al. J Am Coll Cardiol. 2009;53(7):612–619.

Cellular and Ionic Mechanisms

Normal ventricular myocyte action potential and the major ionic currents involved in each one of the phases



imbalance between outward and inward positive currents during phase 1, favoring repolarization and the appearance of a particular notch in the action potential mediated by the outward transient potassium currents (Ito)

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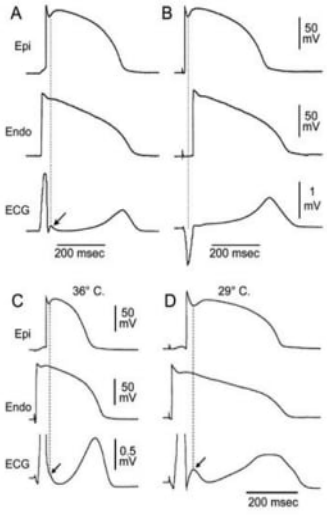
Prog Cardiovasc Dis. 2008 Jul-Aug;51(1):1-22

Ionic and cellular mechanisms for the J wave



The ventricular epicardium commonly displays action potentials with a prominent Ito-mediated notch or spike and dome.

A prominent Ito-mediated action potential notch in ventricular epicardium but not endocardium produces a transmural voltage gradient during early ventricular repolarization that registers as a J wave or J-point elevation on the ECG.

The goal of a pharmacological approach

to produce an inward shift in the balance of current flowing during the early phases of the LV epicardial AP.

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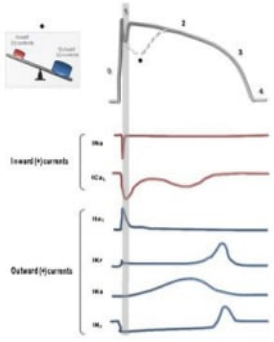
Antzelevitch C Heart Rhythm 2010;7:549 –558)

Genetic mechanism

- Gain of function
  - Ito (KCNE5 mutation and rare polymorphismin DPP10)
  - ATP-sensitive potassium current(IK-ATP) (KCNJ8 and ABCC9)
- Loss of function
  - L-type calcium current(ICa) (CACNA1C, CACNB2, and CACNA2D1)
  - Cardiac voltage-gated fast sodium current (INa) (SCN5A and SCN10A)



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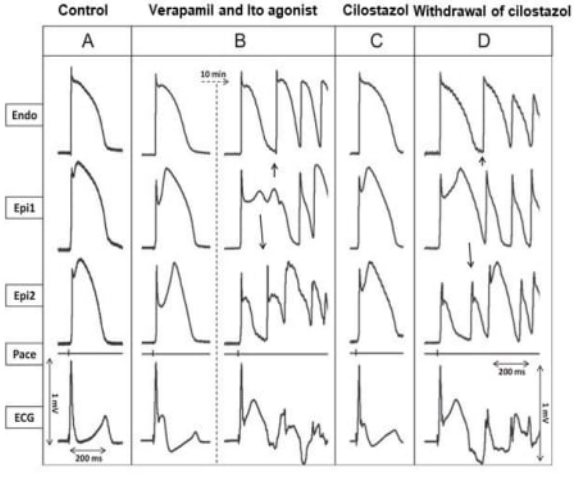
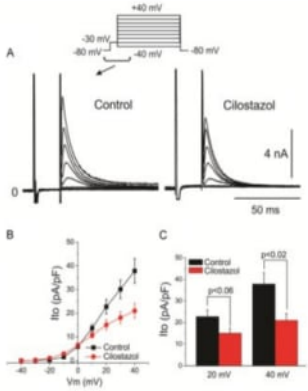
Antzelevitch C Heart Rhythm2016;13:1326–1334

Ameliorative effect of cilostazol



PDE-3 inhibitors reduce Ito, significantly contributing to their ameliorative effect in J-wave syndromes.



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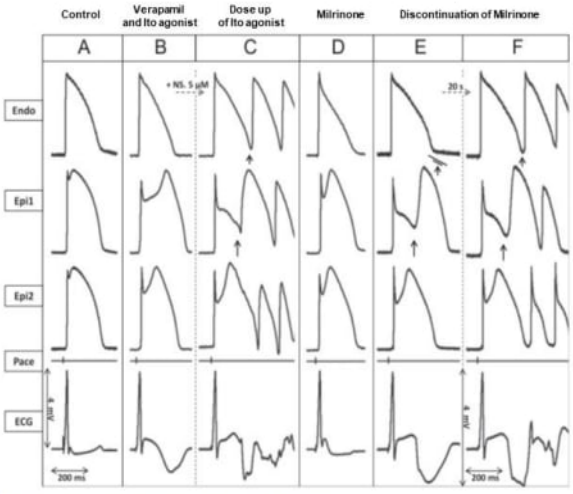
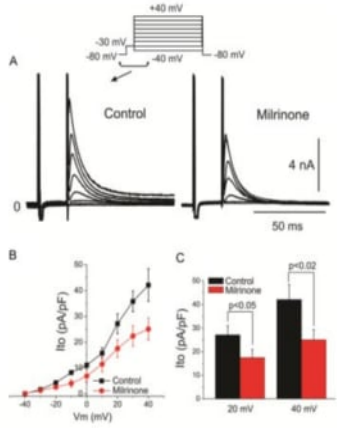
Cilostazol inhibition of the transient outward potassium current

Ameliorative effect of milrinone





Milrinone inhibition of the Ito

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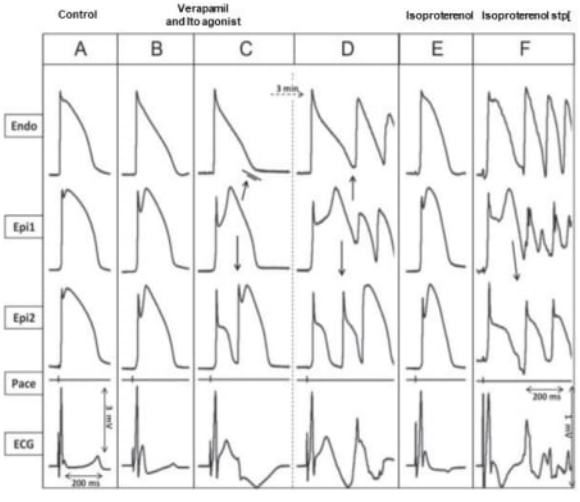
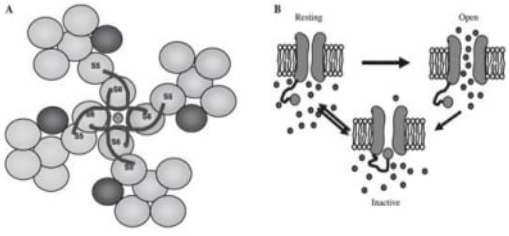
Antzelevitch C Heart Rhythm2016;13:1326–1334

Ameliorative effect of Isoproterenol



Ito : slow recovery from inactivation,
Ito is reduced following an acceleration of heart rate, resulting in a decrease in the magnitude of the J wave.



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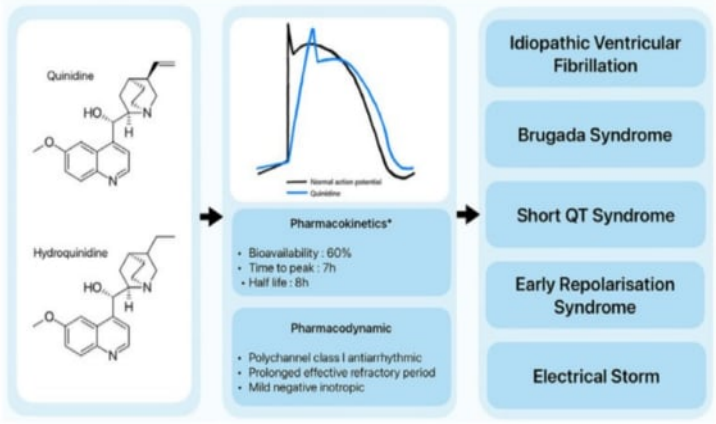
Antzelevitch C Heart Rhythm2016;13:1326–1334

Quinidine to prevent VF



quinidine binds and blocks Kv4.3 and Kv1.4, responsible for the *I* to current

anticholinergic effect that enhances its antiarrhythmic action

Yonsei university, Wonju College of Medicine

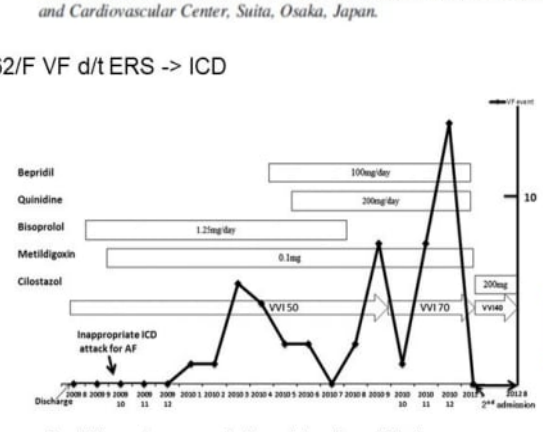
Trends in Cardiovascular Medicine 35 (2025) 73–81

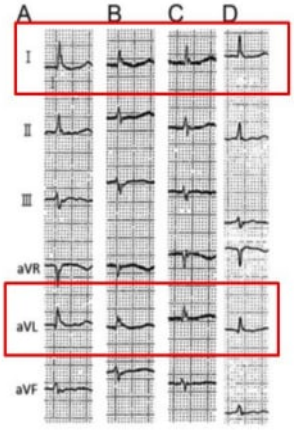
Beneficial effects of cilostazol in a patient with recurrent ventricular fibrillation associated with early repolarization syndrome

Kohei Iguchi, MD, Takashi Noda, MD, PhD, Shiro Kamakura, MD, PhD, Wataru Shimizu, MD, PhD

From the Division of Arrhythmia and Electrophysiology, Department of Cardiovascular Medicine, National Cerebral and Cardiovascular Center, Suita, Osaka, Japan.

62/F VF d/t ERS -> ICD





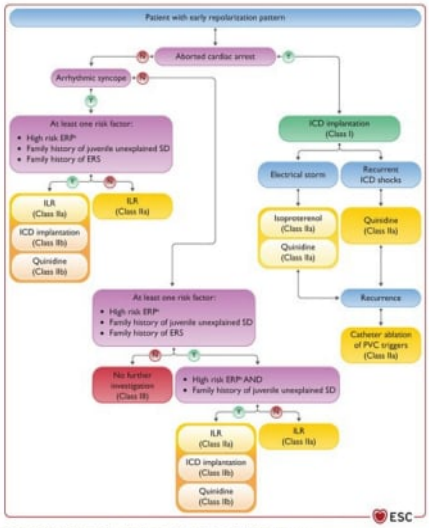
A : baseline
B : after pilsicainide
C : isoproterenol
D : Cilostazol

Bepridil is a unique, non-selective calcium channel blocker

Yonsei university, Wonju College of Medicine

Heart Rhythm 2013;10:604– 606

2022 ESC Recommendations for the management of patients with early repolarization pattern/syndrome



Isoproterenol infusion is effective in acute suppression of recurrent ICD discharges and electrical storm.

AADs that block the transient outward potassium current can prevent VF.

A retrospective multicentre study showed a reduction of recurrent VF after initiation of quinidine.

Phosphodiesterase-3 inhibitors such as cilostazol and milrinone also reduced the recurrence of VF.

Yonsei University, Wonju College of Medicine

Catheter ablation for early repolarization syndrome; Is it really effective?

엄 재 선

연세의대 심장내과

2026
해부학·선천성·유전성
부정맥 연구회 합동 심포지엄

Session 4

Finally, Let's Open the Ebstein Files!

좌장: 허준 (성균관의대 소아청소년과), 오세일 (서울의대 순환기내과)
패널: 이주성 (고려의대 소아청소년과), 최인수 (전남의대 소아청소년과), 이영신 (경희의대 심장내과), 이형석 (인하의대 심장내과)

Anatomy & embryology of Ebstein anomaly
반지은 (부천세종병원 소아청소년과)

Tricuspid valve surgery for Ebstein anomaly
신유림 (연세의대 심장혈관외과)

Accessory pathways in Ebstein anomaly
이의재 (부천세종병원 심장내과)

Ventricular arrhythmia and sudden cardiac death in Ebstein anomaly
김준 (울산의대 심장내과)

대한부정맥학회
Korean Heart Rhythm Society

Anatomy & embryology of Ebstein anomaly

반 지 은

부천세종병원 소아청소년과

Ebstein anomaly is a rare congenital malformation characterized by a broad spectrum of abnormalities involving the tricuspid valve, right ventricle, and right atrioventricular junction. Although traditionally described as an apical displacement of the tricuspid valve, it is more accurately understood as a complex developmental disorder of both the valve and ventricular myocardium. The marked anatomical variability explains its heterogeneous clinical presentation, ranging from severe heart failure and cyanosis in the neonatal period to an incidental diagnosis in asymptomatic adults.

During normal cardiac development, the tricuspid valve leaflets are formed through remodeling of the endocardial cushions and delamination of valvar tissue from the underlying right ventricular myocardium. The mural and septal components of the valve progressively separate from the ventricular wall, while the subvalvar apparatus, including the chordae tendineae and papillary muscles, develops. In Ebstein anomaly, this delamination process is incomplete, predominantly affecting the septal and inferior leaflets. Consequently, these leaflets remain variably adherent to the ventricular septum or inferior wall, and their hinge points are displaced apically from the true atrioventricular junction. However, the precise molecular and developmental mechanisms responsible for this abnormality remain incompletely understood.

The functional tricuspid valve orifice is displaced not only apically but also rotationally toward the right ventricular outflow tract. The septal and inferior leaflets are frequently hypoplastic, dysplastic, or muscularized. In contrast, the anterior leaflet is often large and sail-like but may exhibit fenestrations, restricted mobility, and abnormal muscular or chordal attachments. The true tricuspid annulus is usually dilated, and the malformed leaflets fail to achieve adequate coaptation, producing tricuspid regurgitation of variable severity.

Apical displacement of the functional valve divides the morphologic right ventricle into two

components. The proximal portion, located between the true atrioventricular junction and the displaced functional valve, becomes the atrialized right ventricle. This segment is typically thin-walled, dilated, and mechanically inefficient. The remaining distal portion constitutes the functional right ventricle and includes variable proportions of the apical trabecular and outlet components. In severe forms, the functional right ventricle may be markedly reduced in size and unable to provide adequate pulmonary blood flow.

Associated abnormalities include right atrial enlargement, atrial septal defect or patent foramen ovale, pulmonary stenosis or atresia, and abnormalities of the left ventricular myocardium. The malformed right atrioventricular junction also provides an anatomical substrate for accessory atrioventricular pathways, which are commonly located along the posterior or posteroseptal tricuspid annulus and may be multiple. These pathways, together with right atrial enlargement and surgical scars, contribute to the high burden of supraventricular and ventricular arrhythmias.

A comprehensive understanding of the embryology and three-dimensional anatomy of Ebstein anomaly is essential for accurate imaging, electrophysiological assessment, and surgical planning. Contemporary cone reconstruction reflects this anatomical concept by mobilizing available leaflet tissue, recreating a circumferential valve at the true atrioventricular junction, and restoring a more physiological relationship between the tricuspid valve and right ventricle.

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3. da Silva JP, Baumgratz JF, da Fonseca L, et al. The cone reconstruction of the tricuspid valve in Ebstein's anomaly: the operation, early and midterm results. *J Thorac Cardiovasc Surg*. 2007;133:215–223. PubMed
4. Walsh EP. Ebstein's anomaly of the tricuspid valve: a natural laboratory for re-entrant tachycardias. *JACC Clin Electrophysiol*. 2018;4:1271–1288. PubMed
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Tricuspid valve surgery for Ebstein anomaly

신 유 림

연세의대 심장혈관외과

Valve repair for Ebstein anomaly

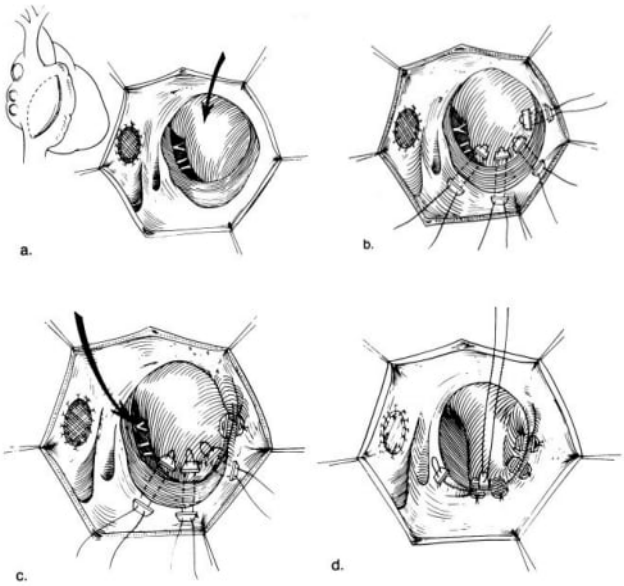
- Feasibility of valve repair in Ebstein anomaly depends on the presence of an anterior leaflet of adequate size.
- Septal and posterior leaflets have no importance for repair.

- Anterior leaflet
- Atrialized ventricle
- Feasibility for biventricular repair

Severance

Danielson repair

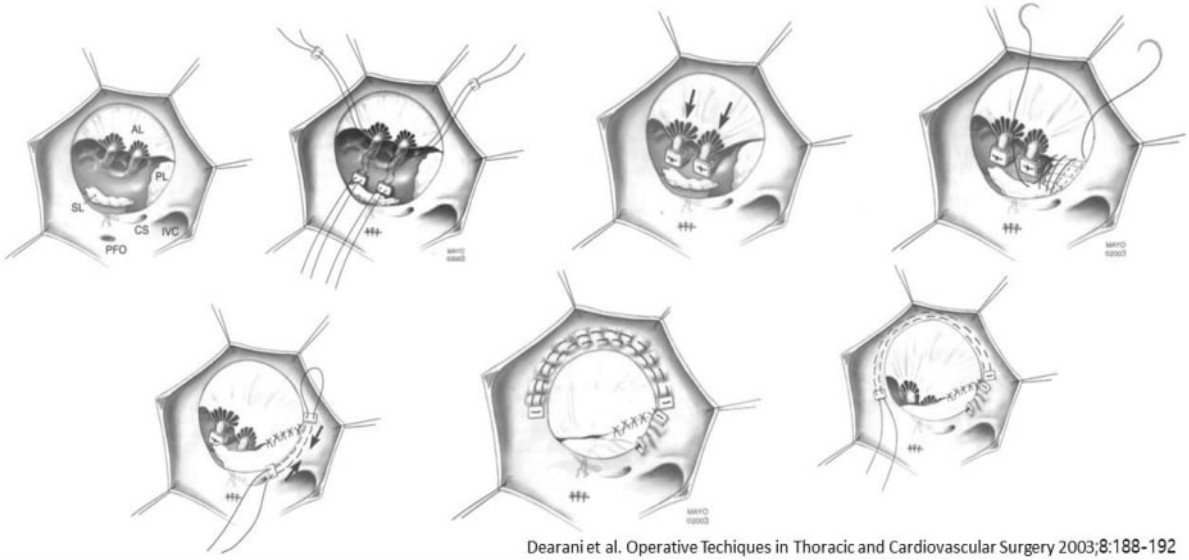
- Plication of the free wall of the atrialized RV
- Posterior tricuspid annuloplasty
- RA reduction



Daneilson et al. Ann Surg 196(4): 1982; 499-504

Severance

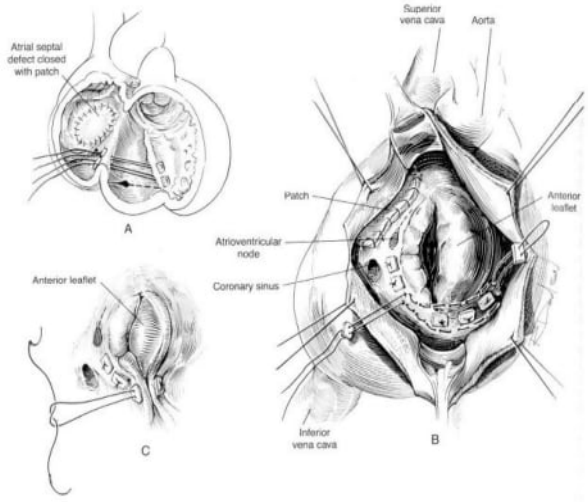
Danielson repair



Dearani et al. Operative Techiques in Thoracic and Cardiovascular Surgery 2003;8:188-192

Severance

Danielson repair



Dearani et al. Operative Techiques in Thoracic and Cardiovascular Surgery 2003;8:188-192

Severance

Danielson repair

- 1972-1991, 189 pts, Mayo Clinic

Age	No. of patients
11-23 mo	9
2-9 yr	44
10-19 yr	54
20-29 yr	46
30-64 yr	36
Total	189

Table V. *Early deaths*

Procedure	No. of patients	Early death	
		n	%
Plication and valvuloplasty	110	8	7.3
Tricuspid valve replacement	69	4	5.8
Plication and Fontan procedure	2	0	0
Other	8	0	0
Total	189	12	6.3

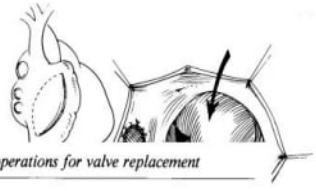
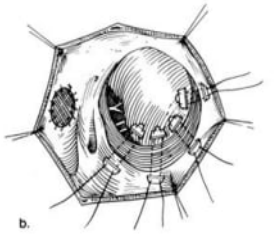
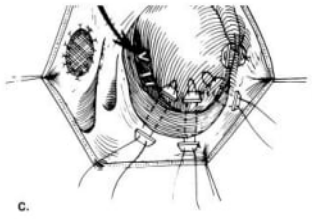
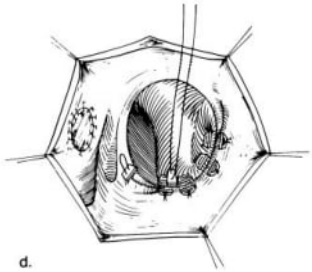


Table VI. *Reoperations for valve replacement*

Age at repair (yr)	Interval (yr)	Anatomy
1	6.5	Residual valvular incompetence
21	1.4	Distortion by inferior infarction
34	14.1	Dilatation of anulus and ventricle
39	6.0	Dilatation of anulus and ventricle







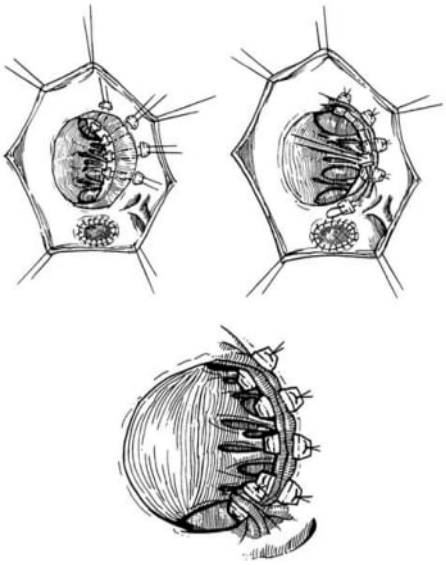
Danielson et al. J Thorac Cardiovasc Surg 1992;104:1195-1202

Severance

Danielson repair

- EP mapping for localization of accessory pathways
- Closure of ASD
- Horizontal plication of the atrialized RV
- Repair of replacement of TV
- Correction of associated anomalies

- Creation of a competent monocuspid valve from the large anterior leaflet + obliteration of the atrialized RV

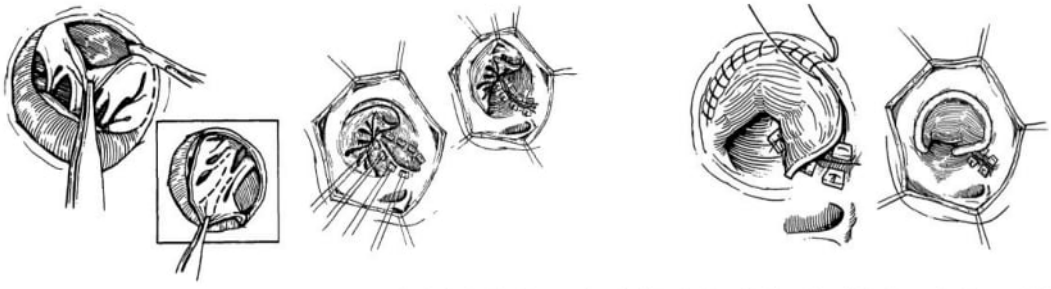


Pediatric Cardiac Surgery Annual of the Seminars in Thoracic and Cardiovascular Surgery 1999:35-50

Severance

Carpentier repair

- Exclusion of atrialized RV by longitudinal plication (preservation of the height of the ventricle) + mobilization of the tricuspid leaflet tissue to create a bileaflet valve

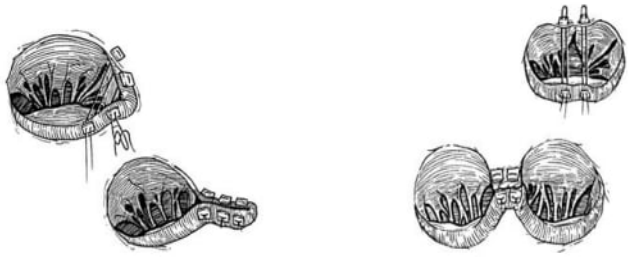


Pediatric Cardiac Surgery Annual of the Seminars in Thoracic and Cardiovascular Surgery 1999:35-50

Severance

Hetzer repair

- Reduction of the tricuspid valve to cover the orifice with the most mobile leaflet
- Plication of the atrialized RV is not used.



Pediatric Cardiac Surgery Annual of the Seminars in Thoracic and Cardiovascular Surgery 1999:35-50

Severance

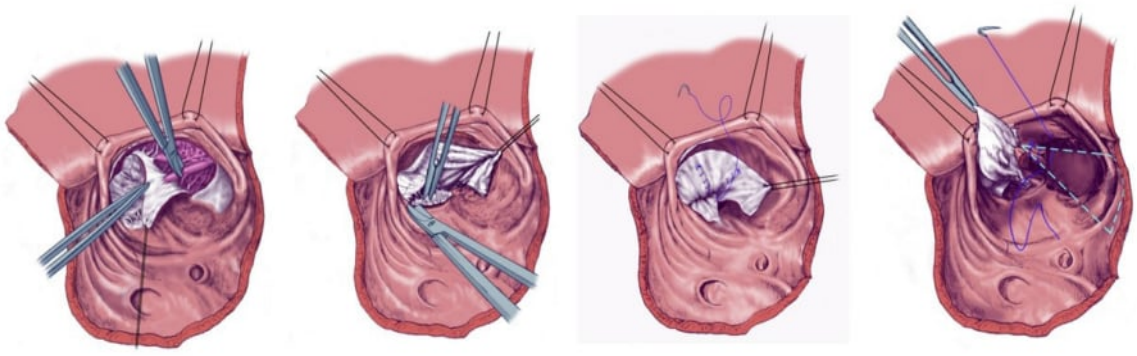
Cone reconstruction

- In contrast to previous techniques that focused on an anterior leaflet monocusp method, the cone repair results in 360' of tricuspid leaflet tissue surrounding the right atrioventricular junction
- Leaflet to leaflet tissue coaptation
- Reconstructed tricuspid valve is reattached at the true annulus
- Atrialized ventricle is plicated.

Dearani et al. JTCVS Techniques 2020;3:269-76

Severance

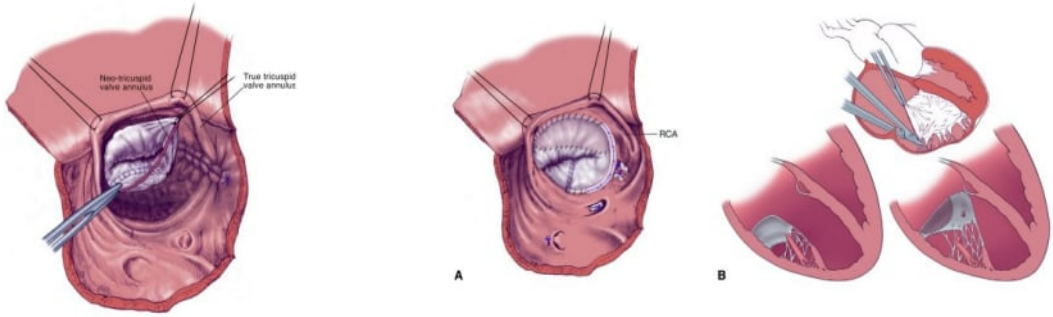
Cone reconstruction



Dearani et al. JTCVS Techniques 2020;3:269-76

Severance

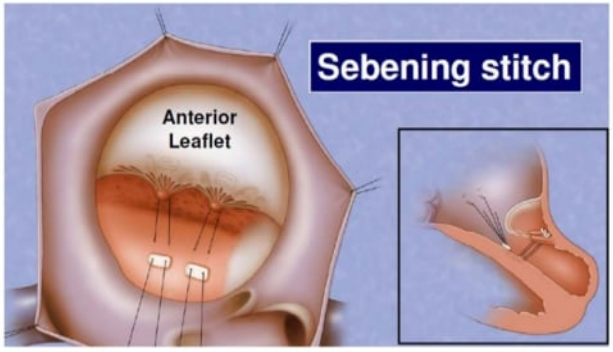
Cone reconstruction

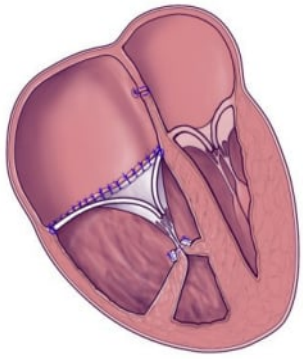


Dearani et al. JTCVS Techniques 2020;3:269-76

Severance

Sebening stitch



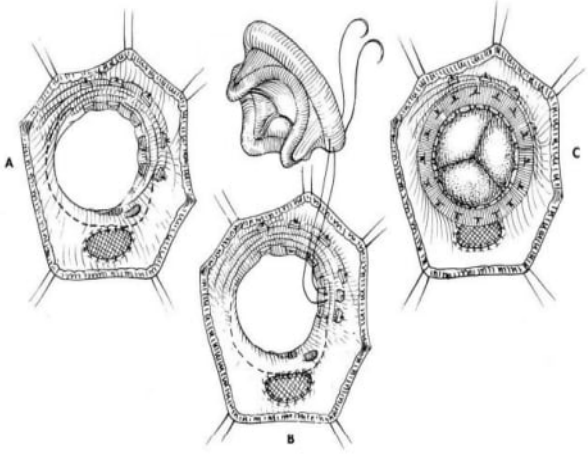


Modified Sebening stitch

Dearani et al. JTCVS Techniques 2020;3:269-76

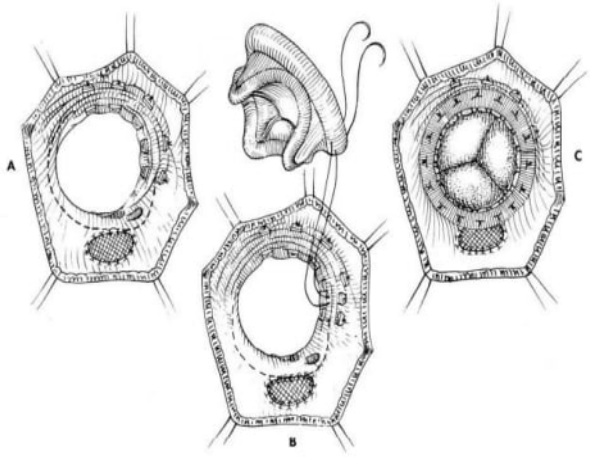
Severance

Tricuspid Valve Replacement



Severance

Tricuspid Valve Replacement



Severance

Summary

- VAD therapy provides an effective means to manage HF as a bridge to HT or as destination therapy in single ventricle patients
- Because of the unique anatomical and physiologic features of single ventricle physiology, special consideration should be given to the implantation techniques.

Severance

Accessory pathways in Ebstein anomaly

이 의 재

부천세종병원 심장내과

Ebstein anomaly (EA)는 tricuspid valve (TV)의 septal leaflet과 posterior leaflet이 하방으로 전위되어 부착하면서 RV가 atrioventricular annulus를 기준으로 “atrialized RV”와 functional RV로 분리되는 선천성 심질환으로, 선천성 심질환 중 1% 미만을 차지하는 드문 질환이지만 부정맥 발생 빈도가 유난히 높아 전기생리학적으로 특히 중요한 질환이다.

Ebstein anomaly에서는 fetus 시기에 TV tissue와 그 아래 RV muscle의 delamination 실패로 서로 유착되어 있으며, 이로 인해 triangle of Koch가 정상 심장보다 작고 AV node의 위치와 형태가 왜곡된다. 또한 CS ostium이 확장되어 있고, 이 부위를 따라 fibromuscular ridge가 형성되는데, 이 능선은 true AV annulus의 위치를 나타내는 중요한 해부학적 지표이자 동시에 다수의 accessory pathway (AP)가 위치하는 부위이다. Septal TA와 중심섬유체(central fibrous body) 사이의 불연속이 AP 형성의 해부학적 기질로 알려져 있으며, 최근 연구에서는 BMP/Alk3 및 Notch 신호전달 경로 이상에 의한 valve-annulus fibrosus 형성 장애가 EA와 WPW syndrome이 공존하는 발생학적 기전으로 제시되고 있다.

EA는 선천성 심질환 중 AP 동반 빈도가 가장 높은 질환으로, 연구에 따라 10~38%에서 AP가 확인되며, 이 중 98~100%가 right side에 있고 특히 posteroseptal 및 posterior 부위에 호발한다. 정상 심장에 비해 multiple APs의 빈도가 현저히 높아 15~34%에서 2개 이상의 AP가 발견된다. ECG는 일반적으로 RBBB를 동반하는데, 만약 RBBB 패턴이 없는 경우 right-side AP 동반을 91~98%의 sensitivity, 92%의 specificity로 예측할 수 있어 진단적 가치가 크다. 그러나 typical RBBB가 관찰된다고 해서 AP가 없는 것이라 할 수 없다. 또한 atrium 내 conduction delay로 인해 delta wave가 있음에도 PR interval이 normal ~ prolongation 될 수 있다.

EA는 부정맥 스펙트럼이 넓어(AVRT, AVNRT, atriofascicular fiber tachycardia, AT, AF, 드물게 atrialized RV에서 기원하는 VT까지) symptomatic tachycardia 환자뿐 아니라, 무증상이더라도 surgical repair (특히 Cone operation)을 계획하는 모든 환자에서 preoperative EPS가 권고된다(class IIa). 이는 repair된 annulus 또는 tissue에 catheter 접근이 어려워지고 특히 annular ring이 mechanical barrier로 작용할 수

있어, 가능하면 수술 전에 AP를 확인하고 제거하는 것이 유리하기 때문이다.

EA에서의 catheter ablation은 구조적으로 정상인 심장보다 성공률이 낮고 재발률이 높다. 그 이유는 (1) atrialized RV에서 기록되는 fractionated EGM이나 atrial muscular fiber가 annulus와는 다른 방향으로 주행해 EGM pattern interpretation이 어렵고, (2) RA/RV dilatation, true tricuspid annulus 식별의 어려움, unusual anatomy에 의한 poor catheter manipulation, thin atrialized RV tissue 등의 해부학적 문제, (3) 일부 환자에서 hemodynamic unstability 등이 있다. Acute success rate은 80~97%로 비교적 양호하나 1 year recurrence rate이 20~40%로 높게 보고되며, 최대 50%의 환자에서 multiple APs가 확인되어 redo procedure가 필요할 수 있다.

Ventricular arrhythmia and sudden cardiac death in Ebstein anomaly

김 준

울산의대 심장내과

** 52 patients of Ebstein's anomaly; surgery

Patient	Age/ Sex	Arrhythmia			Operation	Follow-up		
		Preop	Periop	Postop in hospital		Status	Duration(Mo)	Medications
1	12/M	PSVT	VT/VF	VPC	Plastic repair; ASD closure; multiple DC shocks	SD	1	Digitalis
3	18/M	PSVT	VT/VF	VT/VF	Plastic repair; ASD closure; EPS	SD	1 day	None
24	34/M	PAF	*	-	Plastic repair; ASD closure; multipleDC shocks	SD	6	Procainamide, digitalis
32	12/M	VPC	VT/VF	VT/VF(POD#2)	TV replacement; PV dilation; ASD closure; multiple DC shocks	SD	27	Digitalis
35	14/M	PSVT	PSVT	-	Plastic repair	SD	9days	Lidocaine, procainamide
13	35/M	PSVT	PSVT/VT	-	TVR; ASD closure; repair of PAPVR; bypass traction ablation	Asympto matic	12	None
17	10/M	PSVT/VT with EPS	VT	VT	Plastic repair; ASD closure; EPS	BA	51	Quinidine fo 3 mo
21	34/M	PAF	PAF/VF	PVC	Plastic repair; ASD closure; DC shocks	PAF	14	None
49	15/F	-	VF	PVC	Modified Fontan; ASD closure; closure of left subclavian artery	No F/U		None

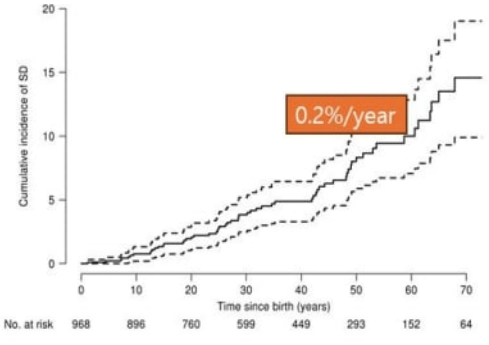
Oh JK. J Am Coll Cardiol 1985;6:1351-7

Sudden cardiac death

- N=968
- mean F/U 13.2 yrs (SD 10.5 yrs)
- mean age 24.3 (SD 18.8 yrs)
- 41.5% male
- cardiac surgery (27.4%)
- syncope (17.1%)
- Heart failure (22.3%)
- accessory pathway (18.6%)
- atrial arrhythmia (22.3%)
- ventricular arrhythmia (6.1%)
- sudden cardiac arrest (0.74%)
- ICD (0.74%)
- PPM (3.0%)
- severe EA (79.85)
- severe TR (82.1%)
- RVE (69.4%)
- RVD (74.7%)
- ASD (59.9%)
- PFO(18.6%)
- LVEF 58.3%

Attenhofer Jost. Eur Heart J 2018;39:1970-1977a

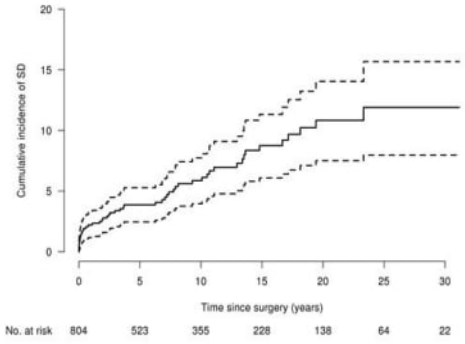
The Kaplan–Meier curve demonstrating the incidence of sudden death from birth (dotted lines represent 95% confidence intervals). SD, sudden death



Clinical variable	Adjusted HR (95% CI)	*P*-value
Heart failure	5.64 (3.20, 9.94)	<0.001
Ventricular tachycardia	6.02 (3.17, 11.44)	<0.001
Pulmonic stenosis	3.42 (1.62, 7.25)	0.001
Haemoglobin > 15 g/dL	2.05 (1.09, 3.87)	0.026
Syncope	2.03 (1.13, 3.65)	0.019
Tricuspid valve surgery at institution	5.94 (2.31, 15.25)	<0.001

Attenhofer Jost. Eur Heart J 2018;39:1970-1977a

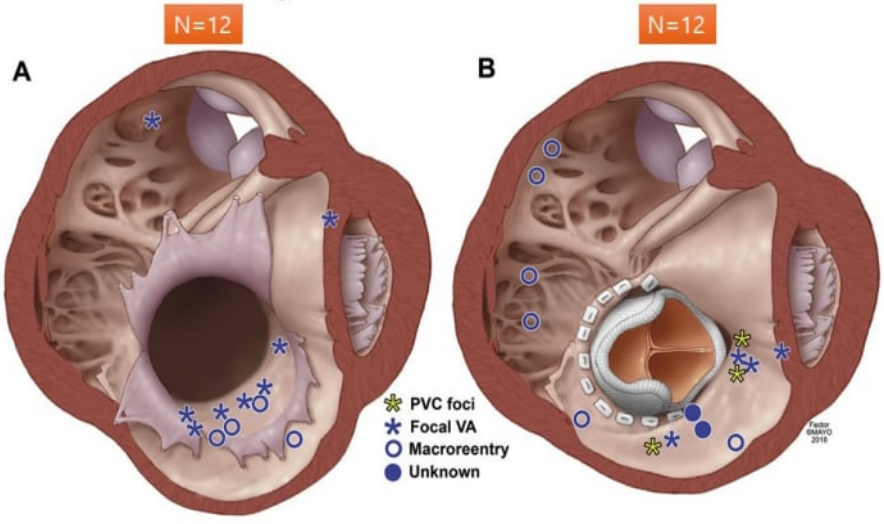
The Kaplan–Meier curve demonstrating the incidence of sudden death following tricuspid valve surgery (dotted lines represent 95% confidence intervals). SD, sudden death.



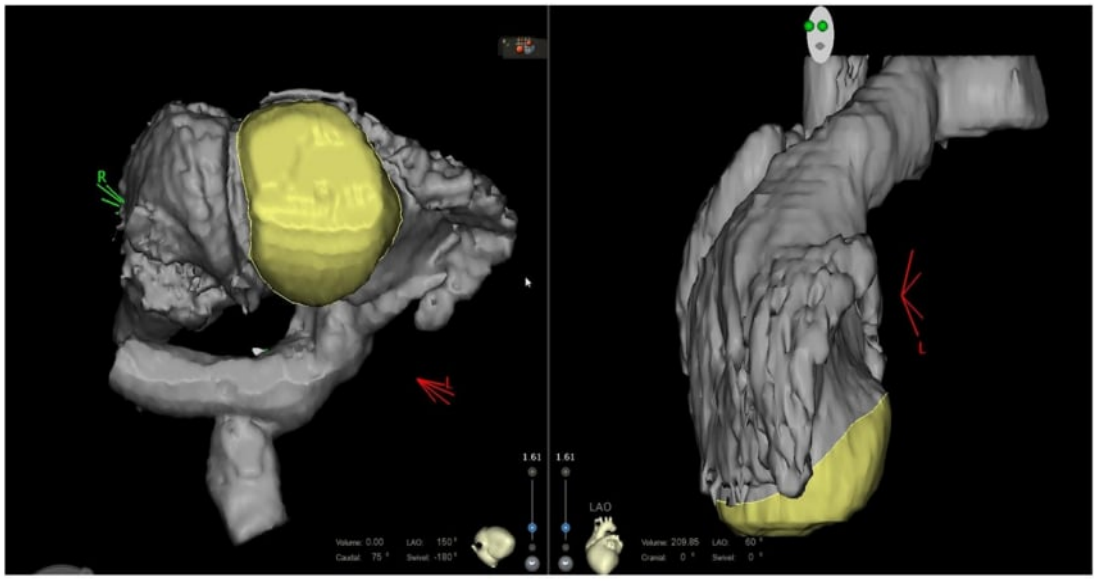
Clinical variable	Adjusted HR (95% CI)	*P*-value
Heart failure	6.33 (3.48, 11.53)	<0.001
Ventricular tachycardia	6.37 (3.30, 12.30)	<0.001
Pulmonic stenosis	3.41 (1.54, 7.57)	0.003
Haemoglobin > 15 g/dL	2.61 (1.30, 5.27)	0.007
Syncope	1.76 (0.94, 3.27)	0.077

Attenhofer Jost. Eur Heart J 2018;39:1970-1977a

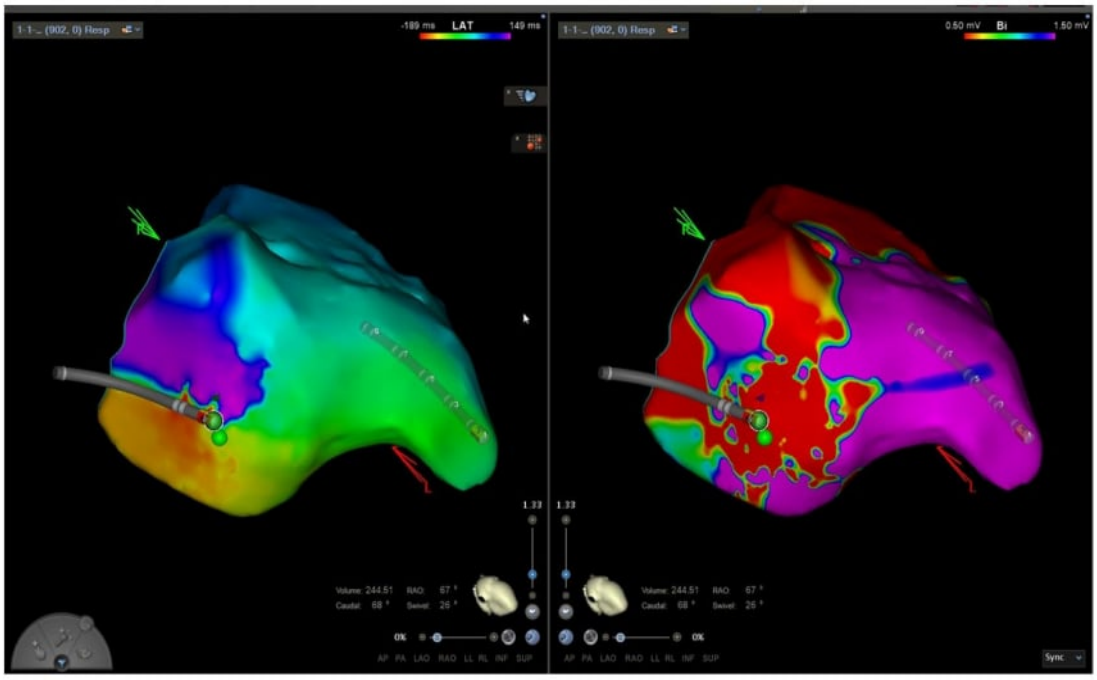
Catheter Ablation of Ventricular Arrhythmia for Ebstein's Anomaly in Unoperated and Post-Surgical Patients

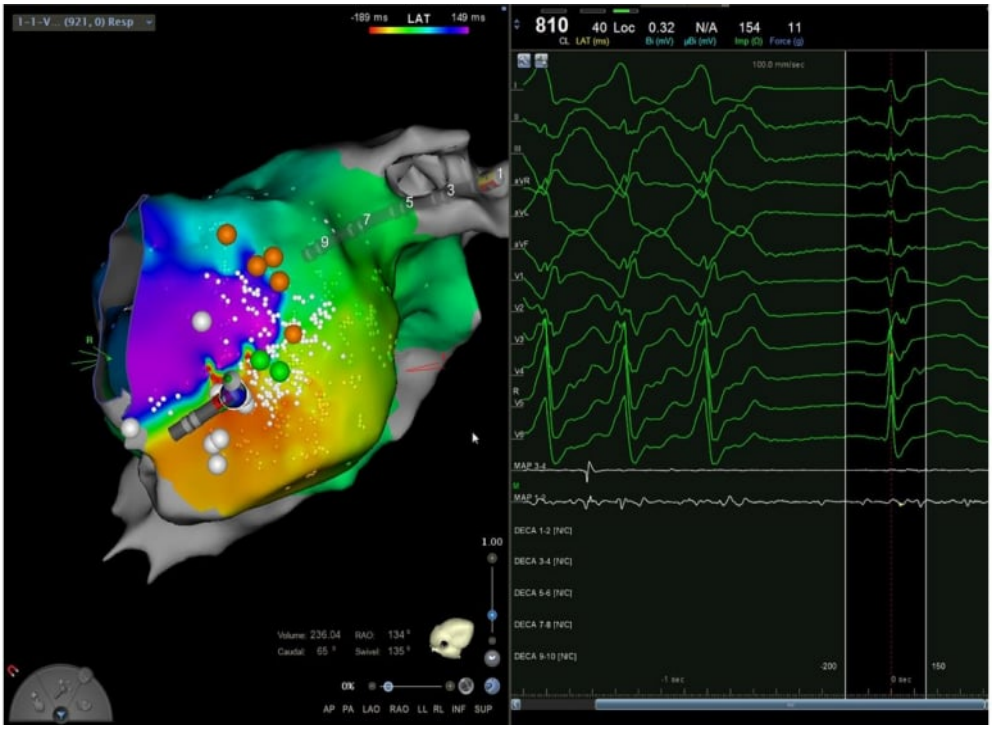


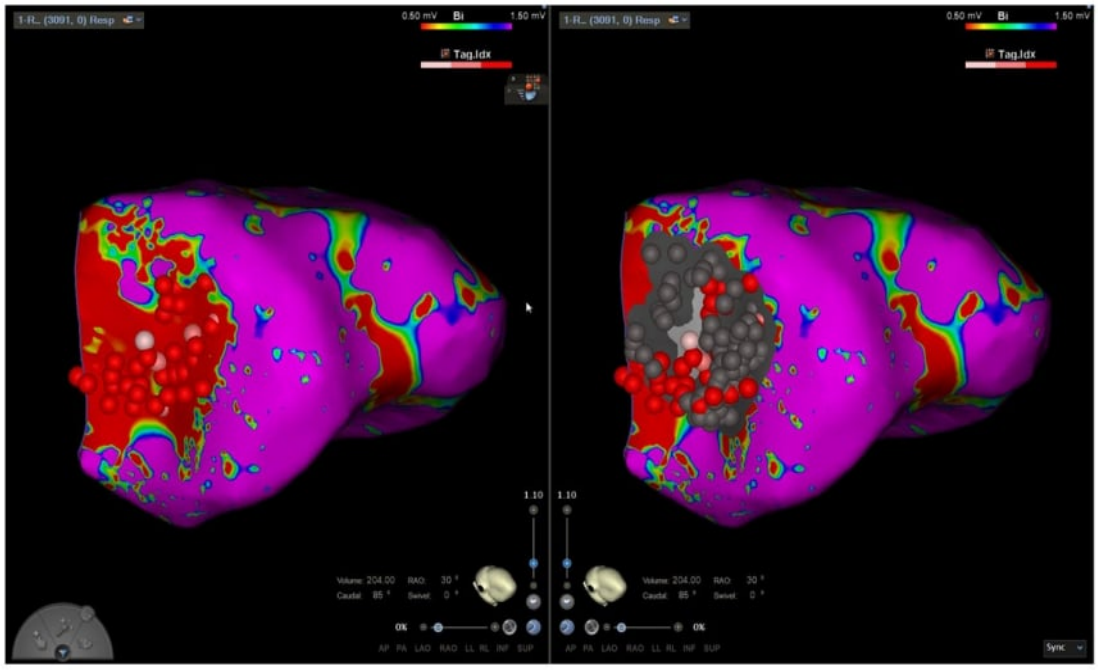
Moore JP. JACC Clin Electrophysiol 2018;4:1300-1307


R
1.61
1.61
LAO


LAT
341


LAT
-189 ms
149 ms
Bi
0.50 mV
1.50 mV
Sync


LAT
-189 ms
149 ms
810
40 Loc
0.32
N/A
154
11
CL
LAT (ms)
Bi (mV)
pBi (mV)
Imp (Ω)
Force (g)
100.0 mm/sec
MAP 3-4
MAP 1-2
DECA 1-2 [NC]
DECA 3-4 [NC]
DECA 5-6 [NC]
DECA 7-8 [NC]
DECA 9-10 [NC]
AP PA LAO RAO LL RL INF SUP



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