

## Brugada 증후군

가  
이 만 영

## Brugada Syndrome

Man Young Lee, MD

Department of Internal Medicine, College of Medicine, The Catholic University of Korea, Seoul, Korea

## ABSTRACT

The occurrence of ventricular fibrillation in the absence of any structural heart disease is classified as "primary electrical disorder". The paradigm of primary electrical disease is the long-QT syndrome. In 1992, Brugada and Brugada first reported a unique electrocardiographic syndrome in which ventricular fibrillation could occur without obvious structural heart disease. Their report drew attention to this condition as another form of primary electrical disorder and Brugada syndrome has subsequently been recognized in virtually all parts of the world. Brugada syndrome is electrocardiographically characterized by ST-segment elevation in the right precordial leads, either with or without right bundle branch block. Although its incidence and distribution have not been confirmed, it may be considerably more common in Southeast Asia. Sudden death is common, may be the first manifestation of disease during its clinical course, and is not effectively prevented by anti-arrhythmic drug therapy. This has led to the recommendation for the placement of an implantable cardioverter-defibrillator in symptomatic patients. The purpose of this paper is to describe the current understanding of Brugada syndrome. (**Korean Circulation J 2002;32(6):461-466**)

**KEY WORDS** : Death, sudden, cardiac ; Ventricular fibrillation ; Bundle-branch block.

## 서 론

(V<sub>1-3</sub>) ST

5%

Brugada

2)

(idiopathic ventricular fibrillation)

(sodium channel) alpha subunit

(enco-

1)

ding) SCN5A, 3p21

가

3)

Brugada

: , 403 - 720

6

long - QT

665 가

: (032) 510 - 5500 · : (032) 520 - 5683

(primary electr-

E - mail : mylee@olmh.cuk.ac.kr

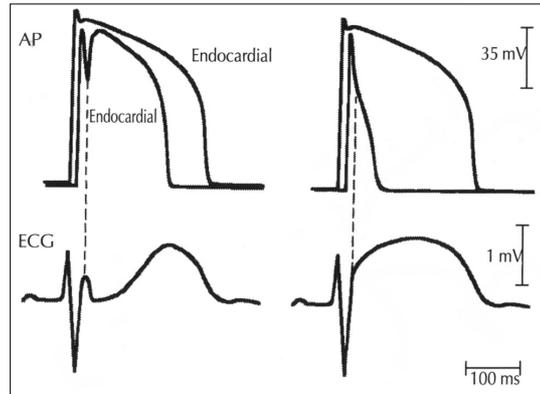
ical disorder)

4)5)

**병태생리**

**유전학적 측면**

가 . 25%  
 , 15%  
 가 Brugada 25%  
 가 50%  
 Brugada  
 Brugada 20 25%  
 SCN5A 가  
 5)6) chromosome 3 - linked long QT Bruga-  
 ada  
 (allelic disorder) 7) Brugada



**Fig. 1.** Difference between epicardial and endocardial membrane action potential (AP) morphology. Top tracings show epicardial and endocardial APs ; bottom, surface ECG. Epicardial AP is characterized by pronounced phase 1 ;“ spike and dome ” (top left). Loss of epicardial AP dome (top right) shortens AP duration. This causes transmural heterogeneity and ST-segment elevation in surface (ECG bottom right).<sup>10)</sup>

5)8)  
 “ dome ”  
 4 - 6) (dispersion) 11)

**심근세포 단위에서의 전기적 변화**

outward current( $I_{to}$ ) transient movement reentry가 circus  
 2 (phase 2) “ notch ” Phase 2 - reentry  
 “ spike and dome ” (Fig. 1). 5)10)12) Brugada

Itage gradient) (vo- 가 Brugada  
 J  
 9) Yan Antzelevitch<sup>10)</sup> 13)

pinacidil, K<sup>+</sup> opener  
 flecainide “ dome ”  
 항부정맥제와 심근세포 단위에서의 전기적 변화  
 (sodium channel blocker) Br-  
 ugada

ST (Fig. 1), ST  $I_{to}$   
 4 - aminopyridine  
 ( $I_{Na}$  current) 가  
 가 1

2 “dome” ST

ST<sup>10)</sup>

**임상양상**

1992 Josep Pedro Brugada가  
(V<sub>1-3</sub>) ST  
aborted sudden cardiac death  
Brugada<sup>2)</sup>

8

ST  
QT

8

4

가

가

Brugada

ST

10)

Brugada

I<sub>to</sub> current  
I<sub>to</sub> current  
inward current  
I<sub>to</sub> current가

가

가

10)

Brugada

I<sub>to</sub> current  
I<sub>to</sub> current  
quinidine I<sub>to</sub> cu-  
“dome”

가

Sudden and Unexpected Death Syndrome(SUDS)  
Sudden Unexpected Nocturnal Death Syndrome(SUNDS)

Brugada

quinidine

Brug-

Brugada

quinidine

10)15)

Brugada

Brugada

disopyramide I<sub>to</sub> I<sub>Na</sub> current

가

Alings Wilde<sup>17)</sup>

가 , 가 58%

가 가

. 22%

가

10)

8)16) class I fl-  
ecainide, ajmaline procainamide I<sub>to</sub>  
I<sub>Na</sub> current Brugada

가

Brugada

28 , 76

14) lidocaine class IA IC  
가 Bru-

가

(stress testing, isoproterenol)  
Ca<sup>++</sup>

가 가 21

ST , 17 가

진 단

가  
 가 (V<sub>1-3</sub>) RSR ST  
 50%  
 가  
 50%  
 가

심전도 소견

Brugada

(V<sub>1-3</sub>) ST  
 ST downsloping  
 T  
 ST  
 S 가

ST (early high takeoff of (V<sub>1-3</sub>))  
 the J wave)  
 (Fig. 2).<sup>18)</sup>

0.05  
 0.16%

Brugada 가  
 가 .<sup>6)</sup>  
 Brugada , 40%

ajmaline, procainamide, flecainide propafenone  
 Brugada

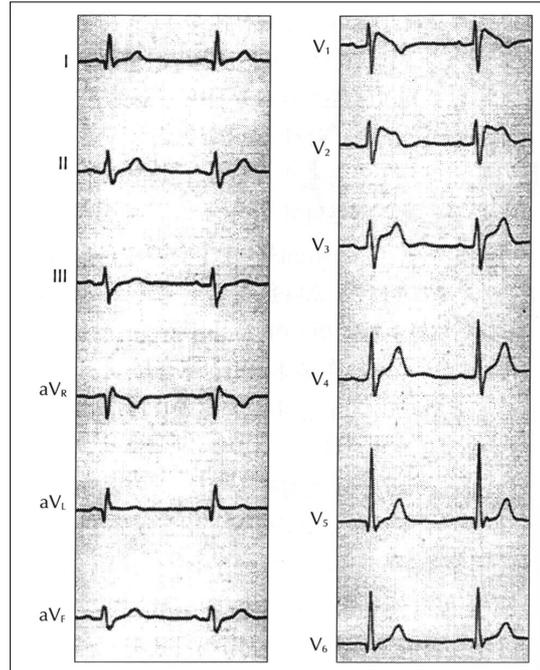


Fig. 2. ECG characteristics with Brugada syndrome displaying RBBB pattern with right precordial ST-segment elevation.

가  
 , 1)  
 ST , 2) class  
 ST , 3)  
 , 4) , 5) 6) 3  
 SCN5A  
 ,  
 Brugada  
 ,  
 Brugada  
 ,  
 , long QT , RSR  
 , ST

, Holter 가  
 torsade de pointes Brugada  
 Brugada 가  
 pause - dependent  
 (coupling interval) 4)6)  
 6) long QT 4)12)17)24)  
 syndrome HV 가 , amiodarone  
 signal averaged ECG 25) ICD 19 34.7 ± 19.4  
 27 22 46  
 eraged ECG가 가 signal av-  
 20) 가  
 16)17) Brugada 22)

### 예후 및 치료

Brugada 21) Brugada 16)24)  
 63 34 27% ST 가  
 가 가  
 가 가  
 가

### 결론

Brugada 가  
 16) Atarashi 23) ST  
 (coved - type) 3 Brugada  
 67.6%, 93.4%  
 Brugada 22) Brugada  
 334 Brugada 가  
 71 , 73 ,  
 190 ,  
 62%,  
 19% 8% (V<sub>1-3</sub>) ST

Brugada

중심 단어 :

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