

Brugada 증후군

가
이 만 영

Brugada Syndrome

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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

(V₁₋₃) 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

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ical disorder)

(primary electr-

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 Brugada
 ada
 (allelic disorder) 7) Brugada

5)8)

4 - 6)

심근세포 단위에서의 전기적 변화

transient
 outward current(I_{to}) 1 (phase 1)
 2 (phase 2)
 "spike and dome"
 "notch"
 (Fig. 1).

(vo-
 ltag gradient)
 J

9) Yan Antzelevitch¹⁰⁾
 K^+ opener
 pinacidil, flecainide
 "dome"

ST
 (Fig. 1), ST I_{to}
 4 - aminopyridine

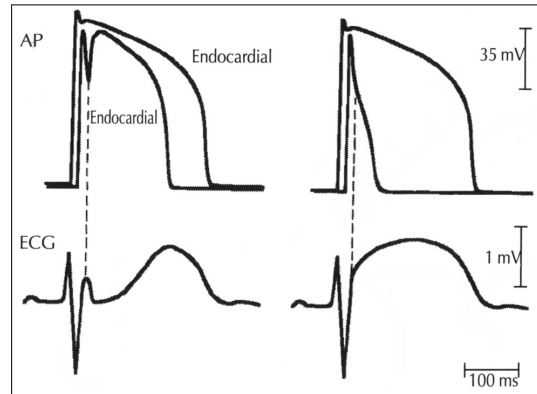


Fig. 1. Difference between epicardial and endocardial membrane action potential (AP) morphology. Top tracings show epicardial and endocardial APs ; bottom, surface ECG. Epicardial AP is characterized by pronounced phase 1 ; "spike and dome" (top left). Loss of epicardial AP dome (top right) shortens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

"dome"

(dispersion)

11)

Phase 2 - reentry
 movement reentry가

circus

5)10)12)

Brugada

가 Brugada

항부정맥제와 심근세포 단위에서의 전기적 변화

(sodium channel blocker) Br-
 ugada

(I_{Na} current)

가
 가

가
 1

2 “dome” ST

ST¹⁰⁾

임상양상

1992 Josep Pedro Brugada가 (V₁₋₃) ST aborted sudden cardiac death Brugada²⁾

8

ST QT

8

4

가

가

ST

Brugada

I_{to} current

I_{to} current, inward current I_{to} current가

I_{to} current quinidine I_{to} cu- “dome”

Brugada

quinidine

quinidine Brugada

quinidine

disopyramide I_{to} I_{Na} current

class I fl-⁸⁾¹⁶⁾

ecainide, ajmaline procainamide I_{to} Brugada

I_{Na} current

Brugada

lidocaine class IA IC 가 Bru-

gada

(stress testing, isoproterenol) Ca⁺⁺

ST

Alings Wilde¹⁷⁾

가 가 , 가 58%

가 가 . 22%

가

104

28 , 76

가

가 가 21

17 가

진 단

가
가 50%
가 (V₁₋₃) RSR ST
가
50%
가

심전도 소견

Brugada

(V₁₋₃) ST
ST downsloping
T
ST
S 가
ST (early high takeoff of (V₁₋₃)
the J wave)
(Fig. 2).¹⁸⁾
0.05
0.16%
Brugada 가
가⁶⁾
Brugada , 40%
가 가
ajmaline, procainamide, flecainide propafenone Brugada
Brugada , 1)
ST , 2) class
가 , 3)
ST , 5) 6) 3
SCN5A
Brugada
Brugada
Brugada
19)
long QT , RSR
ST

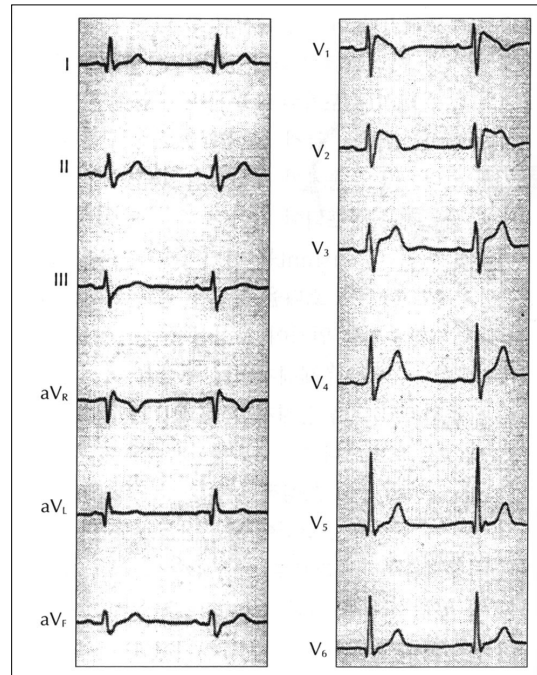


Fig. 2. ECG characteristics with Brugada syndrome displaying RBBB pattern with right precordial ST-segment elevation.

가, Holter 가 Brugada

torsade de pointes

Brugada 가

pause - dependent (coupling interval) 4)6)

가 (ICD)

6) long QT 4)12)17)24)

syndrome HV 가 , amiodarone

가

signal averaged ECG 25) ICD 19 34.7 ± 19.4

27 22 46

signal av-

eraged ECG가 가 가

20)

16)17) Brugada 22)

예후 및 치료

Brugada 21) Brugada 16)24)

63 34 27% Priori ST 가

가 가

가

가

가

Brugada 가

16) Atarashi 23) ST

(coved - type)

3

67.6%, 93.4% Brugada

Brugada 22)

334 Brugada

71 , 73 , 가

190 ,

62%,

19% 8% (V₁₋₃) ST

결론

Brugada

중심 단어 :

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