Electrocardiographic parameters to Detect Cardiac Involvement in Fabry Disease

Original Articles

Yu Wang, MD[1#], Jie Kong, Mphil[1#], Jun Wang, MD[2#], Ru Zhao, MD[1], Heng Wang, MD[1], Jing-Fen Zhu, MD[2], Bing-Yuan Zhou MD, PhD[1], Cao Zou MD, PhD[1], Ting-Bo Jiang MD, PhD[1], Jia-Li Fan, MD[1]*, Si-Jia Sun MD, PhD[1]*

[1]Division of Cardiology, Department of Medicine, The First Affiliated Hospital of Soochow University, Suzhou, China.

[2]Department of Radiology, The First Affiliated Hospital of Soochow University, Suzhou, China.

#These authors have contributed equally to this work.

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*Address for Correspondence:
Si-Jia Sun, MD, PhD or Jia-Li Fan, MD
Division of Cardiology, Department of Medicine, The First Affiliated Hospital of Soochow University, No. 188 Shizi Street, Suzhou, China.
E-mail: u3005150@connect.hku.hk or Fanjiali3@163.com
Tel: +86-0512-67781727. Fax: +86-0512-67781727.

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Supplemental Table 1. Reproducibility of ECG analysis.

Variable	ICC (95% CI)
P wave width	0.985 (0.978-0.990)
PR interval	0.974 (0.962-0.983)
Tp-e/QT	0.919 (0.838-0.963)
QTc	0.988 (0.976-0.994)
QRS width	0.957 (0.919-0.980)
II ST depression	0.985 (0.972-0.993)
II negative or biphasic T	0.991 (0.987-0.994)
fQRS	0.996 (0.992-0.998)
Sokolow-Lyon index	0.941 (0.899-0.974)
V5 ST depression	0.993 (0.986-0.997)
V5 negative or biphasic T	0.995 (0.992-0.998)
U wave	0.901 (0.788-0.959)

Supplemental Table 2. Mutations and type of variant in Fabry patients.

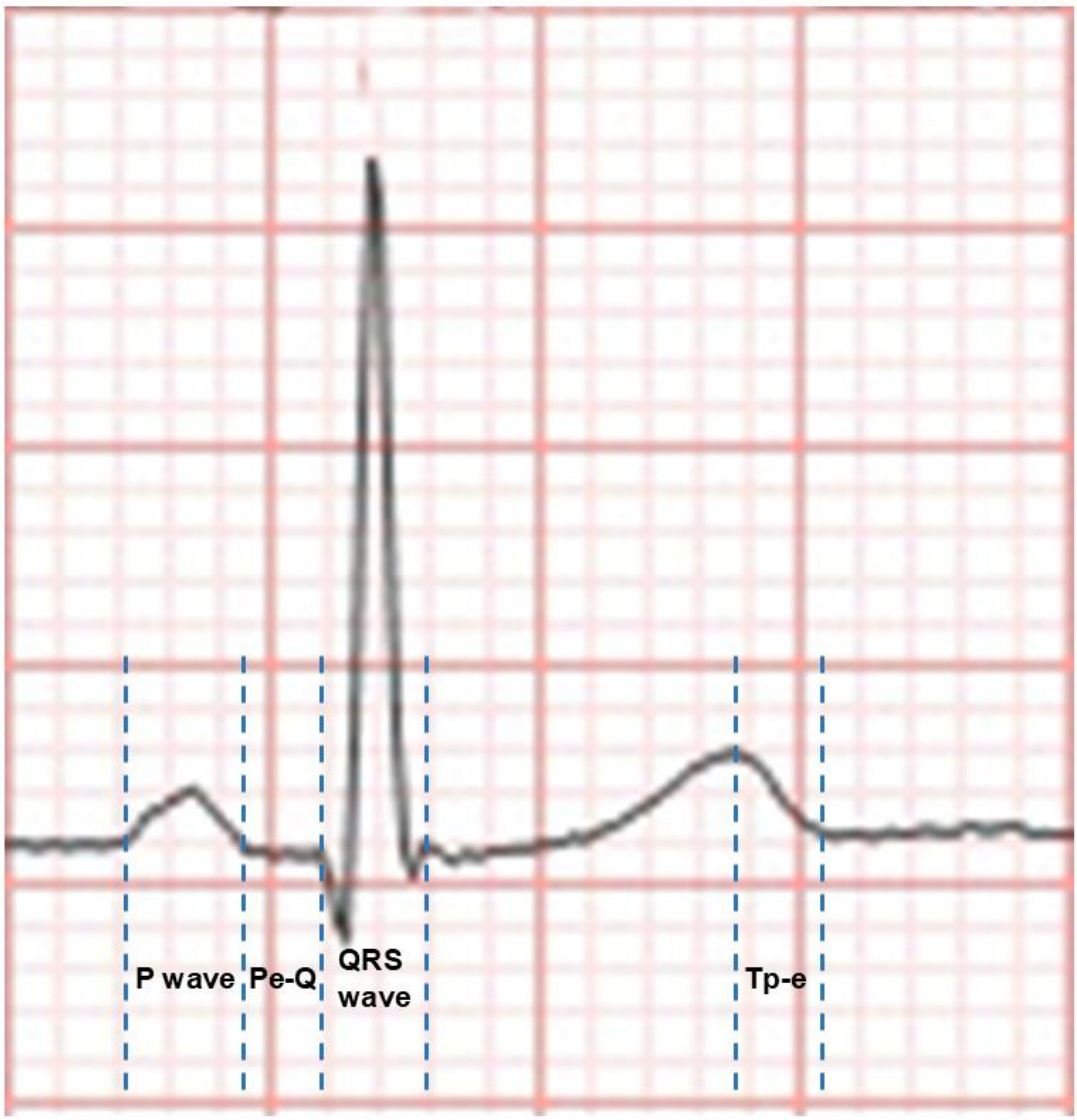
Type of Variable	Mutations (variant)
Classical	c.395G>A(p.G132E), c.827G>T(p.S276I), c.298A>T(p.R100X), c.605G>A(p.C202Y), c.708G>A(p.Trp236Ter), c.709A>T(p.Lys237Ter), c.463G>A(p.Asp155Asn), c.695T>G(p.Ile232Ser), c.680G>A(p.Arg227Gln), c.1080-1082delTGG(p.Gly361del), c.748C>A(p.Gln250Lys), c.834T>A(p.Asn278Lys), c.388-390dupAAG(p.lys130dup), c.334C>T(p.Arg112Cys), c.1072-1074delGAG(p.Glu358del), c.1151T>A(p.1384N), c.1046G>A(p.Trp349*), c.898C>T(p.Leu300Phe)
Late onset	c.486G>C(p.Trp162Cys), c.1080-1082delTGG(p.Gly361del), c.658C>T(p.Arg220Ter), c.1087C>T(p.Arg363Cys), c.1066C>T(p.Arg356Trp), c.1013A>G(Glu338Gly), c.1020G>A(p.Trp340*), c.605G>T(p.Cvs202Phechrx), c.335G>A(p.Arg122His), c.272T>C(p.191T), c.902G>A(p.Arg301Gln), c.463G>A(p.Asp155Asn), c.644A>G(p.Asn215Ser), c.962A>G(p.Gln321Arg), c.334C>T(p.Arg112Cys), c.898C>T(p.Leu300Phe)

Supplemental Table 3. Correlation between α-galactosidase A and globotriaosylsphingosine with GLS, LVMI, Low T1 and LGE.

Variable	α-galactosidase A		globotriaosylsphingosine	
	R	P	R	P
GLS	0.203	0.166	-0.280	0.069
LVMI	-0.009	0.951	0.254	0.100
Low T1	-0.435	**0.004**	0.491	**0.001**
LGE	-0.187	0.236	0.273	0.073

Supplemental Table 4. Linger regression.

	R^2	Durbin-Watson test	B	t	P
Model for GLS	0.671	2.036			
Constant			-26.490	-5.889	**<0.001**
Gander (Female)			-2.609	-3.093	**0.003**
Age			0.107	2.523	**0.014**
Tp-e/QT			-35.818	-2.623	**0.011**
QRS width			-0.109	4.017	**<0.001**
Sokolow-Lyon index			0.710	2.845	**0.006**
Model for LVMI	0.611	1.797			
Constant			27.935	0.499	**0.620**
Gander (Female)			-36.644	-3.242	**0.002**
Tp-e/QT			-559.648	-3.225	**0.002**
QRS width			2.343	6.616	**<0.001**



Supplemental Figure 1. Schemic illustration of electrocardiographic parameters of interest.