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Abstract

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Key words: β -glucuronidase; β -glucuronidase deficiency; β -glucuronidase gene; β -glucuronidase gene mutation; β -glucuronidase gene mutation analysis

1. Introduction

K, **a**, , **r** f
a, **r**, **a** **y y**
y **r**. I **y** **r** **a** **a** **r**-
a **r** (**r**, 1978; **r**, **r**, **r**,
, & **r**, 2001; . 1). **r**-
r **r** **a** **r** **f**
r, **a** **r** **r**
a **a** (**a**) **r** **a** **r**
a **m**- **m**- **r** (**)** **r** **r**. I
a **r** **f** **a** **y** **y** **a** **y** **m**, **a**
r **a** **a** **r** **r** **a**

r a ~ ~ ~ a ~ ~ ~ f -
 ~ y r r ~ a f a ~ ~ a r r ~ a
 a ~ r ~ f ~ ~ ~ ~ ~ I ~
 ~ a r ~ ~ ~ r r a ~ a ~ ~ ~
 ~ ~ r ~ r ~ ~ a ~ ~ a
 r r ~ a .
 ~ a a r y f ~ r a ~ r f r ~ ~ r ~
 f r r ~ . ~ ~ a ~ r a r ~ r a r ~ r ~
 a ~ r a a ~ ~ ~ ~ ~ ~ ~
 ~ ~ ~ fa ~ a r ~ . y ~ y ~
 ~ r y ~ a ~ r fa ~ a ~ ~
 r ~ a ~ y ~ ~ r ~ r ~ y ~ a ~
 a r ~ r a r r a ~ r ~ r ~ r a ~
 a r r a ~ y r ~ r a r ~ ~ r ~
 r (~ . , r ~ a / /) , ~ r a ~ ~
 ~ a r ~ r ~ y ~ ~ , ~ y ~ a ~
 r ~ ~ ~ a ~ r r ~ ~ r ~
 ~ ~ y ~ ~ ~ ~ r ~ a ~ r ~ r ~

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E mail address: zz a. @ . (. a.).

, r 3/1) a a m 3ar 3
r3a 3 3a y r m 3y/r 3a y.
A, fr 3 y r3 ar 3a 3r 3a 3r 3a
r3a 3r 3a 3r (3, 1992; 3 3 3r, 1985; 3 3
& a, 1987)a 3 3a 3r 3a 3r 3a
3a 3r (3, 1992; 3 3a, & 3, 1994). m 3
3a 3a 3a f r3a 3a 3a
fa 3 3 3a m 3. 3r 3 f 3a r-
a 3r 3 (f 3a 3r, / 1/), a a m a-
r3a 3a 3r, 3a r 3a fa m-
3m 3r3r3 3a yfr m 3 3r
3r 3a 3a 3a y r 3a f 3r
m 3a 3r 3a 3a 3a 3a 3a (3.,
33, a, , z3, & , 2005). r 3r m r3a 3
r3a y 3a 3r3a 3a r3a 3a 3r 3a
3r 3a r3a 3a 3a m 3r 3a 3
f 3 3a 3a r3a 3r3a 3a 3a, 3.,
r3a 猜(3, /a 1/), / 1/, 3r 3a f
3a 3a (青, r 3, / 1/). 3a r3a f r3a
a 3a 3a A 3r 3a 3a m 3r A
3r r y 3a 3a (1999) 3a 3a & 3r r,
1992).

a k3 3r, 3a 3a 3a 3a 3a
3a 3r 3a r3a 3a m a r3a 3a
3a 3r, a 3a 3a 3r 3a 3a
m 3 f 3 3a 3a 3a 3a 3a 3a
3a 3a (3., , & , 1999) a 3a 3a
3a r 3a m 3a 3a 3a 3a 3a
r 3a f r3a fa 3a 3a 3a 3a
y a 3a) a r3a r a 3a, 3a m a 3a
r 3a 3a 3a 3a 3a 3a 3a 3a
m m y 3a 3a y r 3a f l e x i c a l r 3a f 3a r-
a 3r 3a 3a 3a. A 3a 3r3a 3a 3a
r 3r 3a 3a. 1, 3r3a 3a r 3f r
r3a 3a 3a r a 3a 3a 3a 3a
r 3a 3a r3a 3a (3a) a 3a
3a 3a 3a 3a y 3a 3a 3a 3a
3a 3a 3a y 3a 3a y m y a m
r3a 3a r3a 3a r 3a a 3a 3a
f 3a 3a m 3a 3a r 3a 3a. A
3a 3a 3a r 3r, 3a 3a r 3a 3a m
a r3a 3a k 3a 3a r 3a 3a r3a 3a
a a 3a 3a r3a 3a 3a f 3a
3a 3a r3a 3a m r3a 3a m r3a
m a f r 3a m fr m a 3a k3 (3a 3a
3a, 1997)a Y (a, , 3a, 2005), a r3a
a 3a 3a m 3a 3a 3a 3a 3a 3a
3a 3a r3a 3a 3a 3a 3a f
3a k a 3a r3a 3a 3a 3a 3a
3a 3a y m y 3a 3a 3a r 3a. D a
fr m 3a k3r 3a 3a 3a 3a y 3a m a
y 3a fa 3a a m 3a. 3a 3a m a
y 3a y 3a f 3a r3a 3a r a fa 3a
r 3a m a 3a 3a 3a 3a 3a r 3a
a a r3a 3a r 3a. 3a 3a m r3a y a 3a f r3a
a m a 3a r3a 3a r3a 3a r3a 3a
a m a 3a m 3a f 3a r3a, a 3a

3a 3a f 3a 3a r 3a (r3a r 3a
a) . 1 f 3a r3a 3a 3a 3a 3a f
3a 3a 3a f r 3a 3a 3a 3a 3a
3a 3a 3a 3a 3a r3a 3a 3a f 3a
3a r3a 3a m 3a 3a r3a a 3a
3a r3a (3., , 3a r 3a - , 1997, 1999)a
3f r3a 3a a 3a r3a 3a y m 3a m
(3a r 3a) 3a 5.

A 3r 3a f3a r3a f 3a 3a 3a 3a
3a f 3a 3a 3a, 3a r3a m 3a m a 3a
3a m r 3a (3., m 3a) 3a 3a 3a
3a r3a 3a 3a 3a f r3a f 3a 3a
a 3a m a 3a r 3a. k3 3a 3a, 3a
3a r3a 3a 3a y r3a 3a. A r 3a y, 88%
f 3a 3a 3a r3a 3a transparent r semi transparent
3a 3a, 3a, 河(r 3r), 溪(3a k3), 海(3a),
湿(3a), 汗(3a), 洗(3a), a 3a 3a 3a 3a
3a, 3a y 3a, m 3a 3a r3a 3a. A 12% f
3a 3a 3a 3a 3a opaque 3a 3a 3a
y a m 3a f 3a m 3a 3a r3a 3a
3a 3a 3a 3a 3a, 3a, 消, a 3a, / a 1/
(3a, 3a, A 3r , , & a, 2003). I
a 3a r3a 3a 3a r 3a 3a r3a f
3a 3a 3a 3a 3a 3a a
f 3a 3a r3a y. 3a 3a f 3a 3a
3a r3a 3a 3a 3a 3a 3a
3a f 3a 3a f r m 3a 3a y 3a m
3a r3a.

I 3f 3a, 3a r3a 3a f 3a
3a y a r 3a 3a 3a r3a 3a
a 3a k. I 3r m 3a 1, 3a m a r3a
3a r3a a k 3a k 3a f r3a 3a
3a 3a m 3a j m 3a, 3a m f 3a
3a 3a f 3a 3a. I 3r m 3a 2, 3a
f 3a 3a 3a 3a 3a 3a 3a
m a 3a r3a 3a r3a 3a r3a
3a 3a m 3a y a 3a 3a a
3a 3a r3a y f 3a 3a.

2. Case background

a 75-year-old male patient
- r a r y 3a, a f r m r y r k3 a a
a 3a 3a r3a f 3a j y. m y
r3a r3a 4 y a r f 3a r3a m m r y a r 3a
3a 16 3a 3a 3a f 3a 3a r a-
3a 3a 3a r3a 3a 3a 3a y f 3a
3a r m f 3a f 3a a 3a (. 2). A
3a 3a D 3a 3a 2001 3a 3a 3a
3a 3a, 3a 3a f 3a m 3a 3a 3a f
3a 3a 3a f r 3a r3a y 3a 3a
3a 3a 3a 1999a, 3a y 2004.

I 3a r m a r y r3a 3a 3a 3a 3a 1999,
3a r 3a m a 3a 3a 3a k, 3r3a
3a r3a 3a 3a r m a 3a 15a
3a 3a (15/15). 3a 3a r3a 3a r a

Table 1

Frequency of correct responses for each condition (N = 162 for each condition)

	N	Correct	Incorrect	Correct	Incorrect	Correct	Incorrect
Condition 1: r / r	162	56 (91)	22 (35)			3 (5)	
Condition 2: r / r	162	41 (67)	20 (32)			1 (2)	
Condition 3: r / r	226	45 (101)	26 (59)	3 (7)	0 (0)	18 (40)	8 (19)
Condition 4: r / r	226	94 (213)	0	1 (3)	1 (2)	3 (8)	0

Note: The data in Table 1 are the frequency of correct responses for each condition. The data in parentheses are the percentage of correct responses. The data in the last two columns are the frequency of correct responses for the two conditions that were not included in the main analysis. The data in the last two columns are the frequency of correct responses for the two conditions that were not included in the main analysis.

If the data in Table 1 are the frequency of correct responses for each condition, the data in parentheses are the percentage of correct responses. The data in the last two columns are the frequency of correct responses for the two conditions that were not included in the main analysis. The data in the last two columns are the frequency of correct responses for the two conditions that were not included in the main analysis.

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A (Continued)

1	2	3	4	5	1	2	3	4	5
捷	/jie2/	an	/jie2/	a	阔	/fa2/	V a	/fa2/	a a y
绅	/shen1/	a a r	/shen1/	r	伸	/shen1/	V	/shen1/	an
键	/jian4/	y	/jian4/	r	伦	/lun2/		/lun2/	an
距	/ju4/	D a	/ju4/		腐	/fu3/	r	/fu3/	
密	/mi4/		/mi4/		滚	/gun3/	r	/gun3/	a r
伟	/wei3/	r	/wei3/	an	仗	/hang4/	a	/hang4/	an
慢	/man4/		/man4/	r	奖	/jiang3/	A a r	/jiang3/	
蚊	/wen2/		/wen2/	I	泥	/ni2/		/ni2/	a r
碑	/bei1/	an	/bei1/	k	沾	/han1/		/han1/	a r
材	/cai2/	a a	/cai2/		葱	/cong1/	a	/cong1/	
拌	/ban4/		/ban4/	a	揍	/ou4/		/ou4/	a
奶	/nai3/	an	/nai3/	an	返	/fan3/	r	/fan3/	k
舱	/cang1/		/cang1/	a	洋	/ang2/		/ang2/	a r
盯	/ding1/	a r	/ding1/	y	抬	/tai2/	a r y	/tai2/	a
秧	/ang1/		/ang1/		苞	/bao1/		/bao1/	
护	/hu4/	r	/hu4/	a	渔	/u2/		/u2/	a r
惊	/jing1/	r r	/jing1/	r	躬	/gong1/		/gong1/	y
捶	/chui2/		/chui2/	a	枝	/hi1/		/hi1/	
评	/ping2/	A	/ping2/		绒	/rong2/		/rong2/	r
译	/i4/	I r	/i4/		粮	/liang2/		/liang2/	
瞄	/miao2/	A	/miao2/	y	叮	/ding1/		/ding1/	
爸	/ba4/	D	/ba4/	a r	惜	/xi1/		/xi1/	r
窃	/qie4/		/qie4/	A r	褐	/he4/	r	/he4/	
逗	/dou4/	A	/dou4/	k	供	/gong4/	r	/gong4/	an
邮	/ou2/		/ou2/		犹	/ou2/	A k	/ou2/	A
陷	/xian4/	k	/xian4/		寞	/mo4/		/mo4/	an
漠	/mo4/	D r	/mo4/	a r	型	/xing2/		/xing2/	r
径	/jing4/	a a y	/jing4/	an	低	/di3/		/di3/	an
球	/qiu2/	a	/qiu2/	a	稀	/xi1/	r	/xi1/	
依	/i1/	D	/i1/	an	丛	/cong2/		/cong2/	
消	/xiao1/	D a r	/xiao1/	a r	苍	/cang1/	r y	/cang1/	
郊	/jiao1/	k r	/jiao1/		颊	/jia2/	a	/jia2/	f
狐	/hu2/		/gua1/	A	瞎	/xia1/		/hai4/	y
扭	/niu3/	a r	/chou3/	a	污	/wu1/		/kui1/	a r
悦	/ue4/	a y	/dui4/	r	扯	/che3/			

