

IV

• •

2015 .

ОГЛАВЛЕНИЕ

| | |
|----------|----|
| | 3 |
| | 3 |
| | 3 |
| | 3 |
| | 4 |
| | 4 |
| | 5 |
| | 5 |
| | 5 |
| | 5 |
| | 7 |
| | 7 |
| IV | 7 |
| | 8 |
| | 10 |

/

:

1.

()

| | | | | | | |
|-----|----------|---|---------------|-----------|---------|--------|
| | | | | | - | OMIM |
| | | | | | - | |
| I | I H | | <i>IDUA</i> | -L- | 4p16.3 | 607014 |
| | I H/S | - | | | | 607015 |
| | I S | | | | | 607016 |
| II | II | | <i>IDS</i> | -2- | Xq28 | 309900 |
| III | III | | <i>SGSH</i> | -N- | 17q25.3 | 252900 |
| | III | | <i>NAGLU</i> | N- -D- | 17q21.2 | 252920 |
| | III | | <i>HGSNAT</i> | - - N- | 8p11.21 | 252930 |
| | IIID | | <i>GNS</i> | N- - | 12q14 | 252940 |
| IV | IV | | <i>GALNS</i> | -6- | 16q24/3 | 253000 |
| | IV | | <i>GLBI</i> | - | 3p21.33 | 253010 |
| VI | VI | - | <i>ARSB</i> | N- -4- | 5q14.1 | 253200 |
| VII | VII | | <i>GUSB</i> | - | 7q21.11 | 253220 |
| IX | IX | | <i>HYALI</i> | | 3p21.31 | 601492 |

IV

:

,

-

,

,

,

-

,

-

,

-

,

,

-

-

.

-6- - (IV) b- (IV),

IV 1:250 000 , IV

IV - , GALNS 16q24.3.
 I. , GBS 3q21.33.
 b-

IV

(.2).

2.
 IV.

| | | | | | |
|---|---|---|-----|-----|-----|
| - | | - | - | + | + |
| | | - | - | + | ++ |
| - | | - | - | + | + |
| | | - | - | + | + |
| | | - | - | + | ++ |
| - | - | - | +++ | +++ | +++ |
| | | - | - | ± | ± |
| | | - | - | ++ | ++ |
| | | - | - | + | + |
| | | - | - | +++ | +++ |

| | | | | | |
|---|-----------|-----|-----|-----|-----|
| | | | | | |
| | () | - | - | + | + |
| | | - | ++ | ++ | ++ |
| | | - | - | ++ | ++ |
| | | - | + | +++ | +++ |
| | | - | + | ++ | ++ |
| | | - | - | + | + |
| - | | n - | | | n - |
| - | | n - | n - | n - | n |
| | N- -6- | | | | |
| | | - | n - | n - | n |

1-3 ; 7-8

80-115)

IV

()

IV

- 1.
- 2.
- 3.
- 4.
- 5.

(
 -6- (IVA), b-
 (IVB)
 : GALNS (
 IVA) GBS (IVB).

IV.

IV

IV

6-12

21-28

.2.

3

IV

| | | | | |
|--|--|---|----|--|
| | | 6 | 12 | |
|--|--|---|----|--|

4

3-