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(Good Practice Points – GPPs):

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· [10,12]

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Q80

Q80.0

Q80.1

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[« »]

Q80.8

Q80.9

1. _____ (_____)

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✓ (∴) - ,
) (Q80.0);

- ✓ **MEDNIK** (, ,) , , ,
- ✓ , ,) -
- ✓ / (, ,)
- ✓ , **IgE**, « ») (.:)
- ✓ (, , ,)
- ✓ (.:)(**10** , , ,)
- ✓ (.:)(, ,)
- ✓ - (, - , ,) , « »- ,)
- ✓ (**BIDS-** (, ; **IBIDS-** : **BIDS-** + , **(Icthyosis)**; **PIBIDS-** : **IBIDS+** **(Q80.9)**)
- **(L85.0)**

Вульгарный ихтиоз

1q22. (R501 2282del4).

) 1:5300 (

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1:250 (

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2.

(3-7

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(5).

3.

4.

40-50%.

5. (, , - :
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 - 2.
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4.

(. 1).

8-12

(D) 3500-6000

7-8

1-4

[1-5].

2

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1. (35-38°) (D) [5].
 2. (38°), 15-20 (D) [5].
 3. - (0,1-0,4 /), 36-37° , 8-12 (12-14); 2,6 , (36°), 10-15-20 (D) [5].
 4. (D) [1,4,5].

X-сцепленный ихтиоз

22.32. 1:2000-1:9500.

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(20%) .

(50%),

6.

(KALI, Xp22.3) ,

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(D) 6000-8000

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(D) 0,3-0,5

6-8

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80-90%

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10%

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4-6

- ... 90%
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Аутосомно-рецессивные врожденные ихтиозы

«...».

14q11)

1:100 000.

Небуллезная врожденная ихтиозиформная эритродермия

17p13.1(

14q11.2 b
-12-

3).
1:100 000 1:20 000.

80-90%

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[36,37]

1. :

2. -

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2-3-

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0,3-0,7 /

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- 1.
- 2.

1 2

6-7

- 3.
- 4.

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- 1.

5

- 2.
- 3.

(1).

(D) 0,3-0,5

3

– 2-3
6

[1-8].

2-

[2,5,46,56].

Буллезные (эпидермолитические; кератинопатические) ихтиозы

[41].

(,)

10).

12q11-13 17q12-q21 (

1

1:300 000.

DiGiovanna (Bale,1994).
(1-3)

(4-6)

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(

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0,3 / / .

1:6000-1:10 000.

(, -).

(D) 0,3-0,5
[1,6].

Ихтиоз иглистый Курта-Маклина

« » 1.

- - .
, « ».

Ихтиоз буллезный типа Сименса (ихтиоз эксфолиативный)

, - , 2 .

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Особые формы ихтиозов

() - -

ABCA12,

2q34.

1:300 000.

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80%

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[29,34].

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[47].

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2.

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3.

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2-5

[1,9].

1-1,5

Синдром отслаивающейся кожи (пилинг-синдром)

-	-	-
-	-5 (TGM-5	15q15.2),
-	1:100 000	(CD5N 1: 500 000.
-		6p21.3).

3-6

IgE

1.

(_____);

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20-50%

(_____).

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40%,
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- 0,1%
5%
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(5%)
(Davila-Seijo et al 2014; Bassolti et al 2011) [21] .
(Blanchet-Bardon et. al 2012)
15% 10% [15] .
10%
(Tadini et al.,2011) [54].

0,1%

2

Craiglow BG et al,2013. Hofmann B, Stege H, Ruzicka T, Lehmann P 1999; Barbarot S, Acher-Chenebaux A, Stalder JF 2006 .; Marulli GC, Campione E, Chimenti MS, Terrinoni A, Melino G, Bianchi L.2003; Nguyen et al.,2007) [13,20,32,40] .

(Litwin et al. 2015;) [25,38,48,53]

(Sigurdsson

et al.,2015) [51] ;

(Nayak et al.2011;) [42]

CHILD-

10%

(Paller et al. 2011)

[45],

2%

2%

2011

Bodemer et al., 44

3

«

»,

20

40

50% [16].

30%

3-4

70-80%

○ , , , , .
○ . [47, 20].

○ : «
» (D) 2-3 (D), 0,5% (D), [5,6].
(D) : 10% (D), 2-5% (D), 8%
(D) 2 [1,4,5]. _____

— : 1%
— ()
• :
— , 5% . 5-10
— 5-10%; (5%;
1-2%,)

• , « »
5% , 5%

: ,
« »

2011) [27] (45 1 / 0,5 /) (Ganemo et al 2012)/. Rajpopat et al 83-86% 7 , 76% 3 . 92% (Shibata et al.,2014) [49] .

[26]. [35]. (Fontao et al.,2011) Maatouk et al.,2012; Kaminska et al.2012) [33,39] .

[50,55] .

[18,19,23,36].

10-14 .

1.

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				80-90%	()	
				80-90%	()	

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4. : . - , 1996. – 441 .
5. : . / : , 2005. – 882 .
6. : . - : , 2008. – 736 .
7. : . - -2013.
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