

E
Job

*

Job syndrome		IgE		IgE
1999/1/1				
6		18	2003/12/31	
	.1/2		.2002	
	%83.3			
		%39		% 55.6
		%72.2		%72.2
		%38.9		%33.3
.%71.4	14/10			
		E		

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Hyperimmunoglobulinemia E (Hyper IgE) Syndrome with Recurrent Infections or Job Syndrome

Isam Anjak *

Summary

The hyper immuno-globuline E (IgE) syndrome with recurrent infections or Job syndrome is a rare immunodeficiency characterized by recurrent skin and pulmonary abscesses and extremely elevated levels of IgE in serum with eczematous dermatitis. Associated facial and skeletal features have been recognized but their frequency is unknown. The genetic basis of this syndrome is poorly understood.

A retrospective clinical study was conducted in the Children's Hospital of Damascus University between 1/1/1999 and 31/12/2003 and 18 patients were hospitalized during period with the diagnosis of Job syndrome, six of them were in the year 2002. The sex ratio was 2/1. Some cases were hospitalized more than once because of recurrent infections.

The symptoms appeared in the first year of life in 83.3 % of the cases. The family history was positive in 39% of the cases.

The most common symptom was fever which was seen in 72.2% of cases. The skin was affected in 72.2% and cough was present at the same percentage.

Laboratory studies revealed increase in white blood cells count in 38.9% and in neutrophils in 33.3% . The percentage of eosinophils was elevated in 10/14 patients (71.4%). IgE was high in all cases.

Most cases were treated by antibiotics according to the site of infection and the expected organisms. Treatment was changed according to the cultures and the sensitivity to antibiotics or to the clinical responses. One patient died during the study.

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IgE

12

(IgE)E

.(3) IgE

IgE

.(1)

:

.(4,1)

1966 Job

1972

4q

Buckley

.(5,4,3,2)

.(2)IgE

:

:

60

%80

.(2) 4

8

.(2)

60

.(6)

IgE

.(2)

:

Atopy

.(2)

.(6)

.(2)

:

.(1)

IgE

/

2000

(2,1)

(2)

.(1) D

.B .(2,1)

: CD8- ,CD4- ,CD3-
. (1) ,CD2-
:

.Craniosynostosis

.(2) Aspergillus

:

(9,8,7)

alpha

.(10) and gamma interferon

:

5

1999/1/1

E

2003/12/31

:

:

(18)

(1)

1999	3	16.7
2000	4	22.2
2001	3	16.7
2002	6	33.3
2003	2	11.1
	18	

(1)

(2)

	11	61.1
	4	22.2
	3	16.7
	18	

(2)

(3)

	12	66.7
	6	33.3
	18	

(3)

(2)

(4)

	15	83.3
3-	1	5.6
6-4	1	5.6
10-7	1	5.6
	18	

(4)

(5)

	3	16.7
3-	9	50
6-4	3	16.7
10-7	3	16.7
	18	

(5)

(6)

	3	16.7
	1	5.6
	1	5.6
	2	11.1
	7	39

(6)

(7)

	10	55.6
	2	11.1
	2	11.1
	1	5.6
	1	5.6
IgE	1	5.6
IgE	1	5.6

(7)

(8)

	9	50
	6	33.3

	7	38.9
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(8)

(9)

	13	72.2
-	13	72.2
-	12	66.7
-	13	72.2
-	9	50
	2	11.1
	1	5.6

(9)

(10)

-	10	55.6
-	13	72.2
-	5	27.8
-	1	5.6

-	2	11.1
-	1	5.6

(10)

(11)

	7	38.9
	6	33.3
	14/10	71.4
C	17/16	94.1
	14/11	78.6

(11)

(12)

IgG	10	-	8
IgM	12	5	1
IgA	14	-	4
IgE	-	-	18

(12)

Nitro-blue tetrazolium dye test

7

(NBT)

:

(13)

9

	5	27.8
	1	5.6
+	1	5.6

+ +	1	5.6
+	1	5.6
	9	50

(13)

: :
4 -
(-)

(14)		
	13	72.2
	1	5.6
	1	5.6
	3	16.7
	18	

(14)

(15)

	3	16.7
2-1	5	27.8
4-3	6	33.3
4	4	22.2
	18	

(15)

:

E

(11)

8

(5)

30

5

%38.9

14

18

2002

1/2

10

(5,2)

%55.6

(5,4,2,1)

IgE

(%83.3)

%72.2

13

%16.7

/

15

IgE

IgE

14

%39

IgE

2000

8/7

%87.5

C

%78.6, %94.1

(5)Renner et al

IgG, IgM,

IgA, IgE

IgG

8

IgM

5

7

IgA

%38.9

IgE

4

6

14120 345

%33.3

2000

10

2000

%71.4

(11)

IgE

7 NBT .(2)

9

4

(11) Chamlin et al

8 6

5

.(5)

4

13

4-2

4

4

8

6

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.2004/3/15:

.2004/7/13: