

Scoring System with Clinical and Laboratory Tests for Individuals in Kindreds with HIES

CLINICAL FINDINGS	POINTS <sup>a</sup>									
	0	1	2	3	4	5	6	7	8	10
Highest serum-IgE level (IU/ml) <sup>b</sup>	<200	200-500			501-1,000				1,001-2,000	>2,000
Skin abscesses	None		1-2		3-4				>4	
Pneumonia (episodes over lifetime)	None		1		2		3		>3	
Parenchymal lung anomalies	Absent						Bronchiectasis		Pneumatocele	
Retained primary teeth	None	1	2		3				>3	
Scoliosis, maximum curvature	<10°		10-14°		15°-20°				>20°	
Fractures with minor trauma	None				1-2				>2	
Highest eosinophil count (cells/ $\mu$ l) <sup>c</sup>	<700			700-800			>800			
Characteristic face	Absent		Mildly present			Present				
Midline anomaly <sup>d</sup>	Absent					Present				
Newborn rash	Absent				Present					
Eczema (worst stage)	Absent	Mild	Moderate		Severe					
Upper respiratory infections per year	1-2	3	4-6		>6					
Candidiasis	None	Oral	Fingernails		Systemic					
Other serious infections	None				Severe					
Fatal infection	Absent				Present					
Hyperextensibility	Absent				Present					
Lymphoma	Absent				Present					
Increased nasal width <sup>e</sup>	<1 SD	1-2 SD		>2 SD						
High palate	Absent		Present							
Young-age correction	>5 years			2-5 years		1-2 years		$\leq$ 1 year		

<sup>a</sup> The entry in the furthest-right column is assigned the maximum points allowed for each finding.

<sup>b</sup> Normal <130 IU/ml.

<sup>c</sup> 700/ $\mu$ l = 1SD, 800/ $\mu$ l = 2 SD above the mean value for normal individuals.

<sup>d</sup> For example, cleft palate, cleft tongue, hemivertebrae, other vertebral anomaly, etc. (see Grimbacher et al. 1999a).

<sup>e</sup> Compared with age- and sex-matched controls (see Farkas et al. 1994).