

Table 1**Scoring System with Clinical and Laboratory Tests for Individuals in Kindreds with HIES**

CLINICAL FINDINGS	POINTS ^a									
	0	1	2	3	4	5	6	7	8	10
Highest serum-IgE level (IU/ml) ^b	<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/ μ l) ^c	<700			700–800			>800			
Characteristic face	Absent		Mildly present			Present				
Midline anomaly ^d	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 ^e	<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		

^a The entry in the furthest-right column is assigned the maximum points allowed for each finding.

^b Normal <130 IU/ml.

^c 700/ μ l = 1SD, 800/ μ l = 2 SD above the mean value for normal individuals.

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

^e Compared with age- and sex-matched controls (see Farkas et al. 1994).