**Table 1** supplementary materials: General characteristics of the patients with HIES. The calculated NIH HIES scores are presented.

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| Genetic | Patients | Sex | Consanguinity | Family historyof child death or miscarriage | HIES score | Status |
| *DOCK8+* | P1 | Male | No | Yes (death in sibling) | 31 | Alive |
| P2 | Male | Yes (first cousin) | Yes (death in sibling) | 49 | Alive |
| P3 | Female | Yes (first cousin) | Yes (death in sibling) | 38 | Alive |
| P4 | Female | Yes (first cousin) | No | 68 | Alive |
| P5 | Male | Yes (first cousin) | No (only child) | 37 | Dead(Sepsis) |
| P6 | Female | Yes (second cousin) | Yes (HIES in sibling) | 44 | Dead(Lymphoma) |
| P7 | Male | Yes (second cousin) | Yes (HIES in sibling) | 38 | Alive |
| P8 | Male | No | Yes (HIES in siblings) | 40 | Alive |
| P9 | Female | No | Yes (HIES in siblings) | 26 | Alive |
| P10 | Female | No | Yes (HIES in siblings) | 58 | Alive |
| *STAT3* | P11 | Male | No | No | 34 | Alive |
| P12 | Male | Yes (first cousin) | Yes (death in siblings) | 37 | Dead(Sepsis) |
| P13 | Male | No | No | 58 | Alive |
| P14 | Male | Yes (first cousin) | No | 36 | Alive |
| Undefined genetic | P15 | Male | Yes (beyond second cousin) | Yes (miscarriage) | 80 | Alive |
| P16 | Male | No | No | 18 | Alive |
| P17 | Female | Yes (first cousin) | Yes (death in siblings) | 49 | Alive |
| P18 | Male | No | No | 40 | Dead (Sepsis) |

**Table 2** Supplementary materials: Immunological characteristics of the studied HIES patients.

|  |  |  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Genetic | Patients | Highest IgE level (IU/mL) | Highest eosinophil count (cells/mL) | IgMg/L | IgGg/L | IgAg/L | CD3+cells | CD4+cells | CD8+cells | CD4+/CD8+cells | CD16+cells | CD19+cells |
| *DOCK8+* | P1 | >2000 | 2800 | 2.4 | 18.8 | 0.9 | 85 | 30 | 50 | 0.6 | 12 | 3 |
| P2 | 5360 | 3553 | 7.3 | 20 | 2.1 | 44.9 | 31.1 | 14.2 | 2.2 | 22.6 | 12.8 |
| P3 | >2000 | 18696 | 0.7 | 12.6 | >5.9 | 45 | 10 | 25 | 0.4 | 32 | 24 |
| P4 | 2744 | 21889 | 0.3 | 21.4 | 1.9 | 31 | 8 | 22 | 0.4 | 23 | 41 |
| P5 | 1203 | 18600 | 3.08 | 14.76 | 1.03 | 66 | 45 | 18 | 2.5 | 5 | 29 |
| P6 | 7000 | 4768 | <0.2 | 8.5 | 2.0 | 51 | 15 | 29 | 0.5 | 26 | 22 |
| P7 | 1447 | 1288 | 1.7 | 9.4 | 1.9 | 74 | 12 | 62 | 0.2 | 25 | 1 |
| P8 | 114 | 2400 | 0.3 | 14.5 | 3.5 | 68 | 42 | 27 | 1.5 | 10 | 25 |
| P9 | 650 | 572 | 0.8 | 7.2 | 1 | 59 | 40 | 28 | 1.4 | 8 | 38 |
| P10 | 2171 | 14535 | 0.3 | 3.5 | 5.6 | 71 | 26 | 41 | 0.6 | 11 | 18 |
| *STAT3* | P11 | 1305 | 19941 | 3.2 | 6.3 | 0.7 | 64 | 45 | 23 | 1.9 | 10 | 29 |
| P12 | 1281 | 1560 | 0.2 | 4.6 | 2.1 | 63 | 17 | 36 | 0.5 | 13 | 24 |
| P13 | >2000 | 285 | 1.2 | 8.7 | 1.3 | 83 | 45 | 33 | 1.4 | 2 | 15 |
| P14 | 1417 | 1680 | 0.6 | 16.9 | 2.8 | 78 | 45 | 33 | 1.4 | 6 | 16 |
| Undefined genetic | P15 | 2643 | 2136 | 1.6 | 15.1 | 1.8 | 67 | 37 | 32 | 1.2 | 9 | 23 |
| P16 | 6860 | 300 | 0.9 | 10.8 | 2.0 | 75 | 34 | 38 | 0.9 | 11 | 17 |
| P17 | 1459 | 198 | 2.6 | 8.9 | 1.7 | 50 | 27 | 20 | 1.3 | 7 | 43 |
| P18 | 4490 | 1008 | 0.9 | 7.8 | 1.1 | - | - | - | - | - | - |

**Table 3** Supplementary materials: Clinical features of four HIES patientswith undefined genetic defect

|  |  |
| --- | --- |
| Clinical Manifestations | Number of affected patients |
| Pneumonia | 4 |
| Bronchiectasis | - |
| Pneumatocele | 2 |
| Eczema | 3 |
| Newborn Rash | 1 |
| Skin Infection | 3 |
| Candidiasis | 1 |
| Sepsis | 2 |
| Meningitis | - |
| Chronic Diarrhea | 1 |
| Otitis | 1 |
| Sinusitis | 1 |
| Hepatomegaly | 1 |
| Cancers | - |
| Vascular abnormality | 1 (JRA) |
| Cardiovascular diseases | - |
| Viral infections | 3 |
| Failure to Thrive | - |
| Neurologic syndrome | 1 |
| Food Allergy | - |
| Characteristic Face | 1 |
| Bone fragility | 2 |
| Connective tissue problems | 1 |
| Scoliosis | 1 |
| Osteopenia | - |
| Osteomyelitis and Septic Arthritis | 1 |
| Retained Primary Teeth | 1 |
| Cellulitis | 1 |
| Brain Abscess | 1 |
| Seizure | 1 |
| Keratoacanthoma | 1 |
| Death | 1 (Sepsis) |