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|  |  | **Punwani D, et al.** | **Jabara H, et al.** | **McKinnon M, et al.** |
| **P1** | **P2** | **1 patient** | **2 siblings** | **1 patient** |
| **Consanguinity** | Yes | Yes | No | Yes | Yes |
|  | **Pre-HSCT** | **Post-HSCT** | **Pre-HSCT** | **Post-HSCT** | **Pre-HSCT** | **Post-HSCT** | **HSCT not done** |
| **Infections** |
| Skin/mucosa | Candida, HSV-1 Pseudomonas | HSV1 | Candida, HSV-1 | HSV1 | Candida, S.aureus | No |  | S. aureusHSV-1, VZV |
| Pulmonary | S.pneumoniaePneumocistis jirovecii, EBV, CMV, adenovirus | No | Adenovirus | AdenovirusClinical bacterial pneumonia | CMV, RSV | No | S.pneumoniae, S.aureusH. influenzae, K. pneumoniaePseudomonasCandida | S. aureusS.pneumoniaeCMV |
| Gastrointestinal tract | Salmonella, Campilobacter jejuniirotavirus, adenovirus, EBV, CMV (chronic) | No | Salmonella | No | Clostridium difficile | No | Candida |  |
| Blood | S. aureus, S.pneumoniae | Transient CMV reactivation |  | Transient CMV reactivation | CMV | No |  |  |
| Others | HSV-1 keratitis | HSV-1 keratitis |  | No |  | No | S.pneumoniae meningitisH.influenzae meningitis CMV (urine) |  |
| **Clinical manifestations** |
|  | Poor or delayed growth | Yes | Resolved | No | No | Yes | Resolved | Yes | Yes |
|  | Oral lesions\* | Yes | Resolved | Yes | Partly resolved | Yes | Resolved | Yes | Yes |
|  | Eczema | Yes | Resolved | Yes | Partly resolved | Yes | Resolved | Not reported | Yes |
|  | Enteropathy | Severe diarrhea, malabsorbtion, massive lymphocyte infiltration, subtotal villous atrophy  | Resolved  | No (no histology) | No | Bloody diarrhea (no histology) | Resolved | Intraepithelial lymphocytosis villous atrophy | Severe intestinal inflammation |
|  | Bronchiectasis | Yes | NA | CT-scan not done | NA | No | No | Yes, respiratory failure | Yes |
|  | Other findings | Dysmorphic facies, severe shortsightedness | NA | Dysmorphic faciesPeanut allergy | NA |  |  | Mastoiditis | Dysmorphic facies, bone fractures, granulationtissue on vocal cord, larynx, ear canal |
| **Treatments** |
|  | Ig IV | Yes | No | No | No | Yes | Yes | Yes | Yes |
|  | Anti-microbial prophylaxis | TMP-SMXAcyclovir | TMP-SMXAcyclovir | TMP-SMX | TMP-SMXAcyclovir | Yes | No | Yes | Yes |
|  | Immunosupresive therapy | MethylpredisoloneTacrolimus | No | No | No | Not reported | Not reported | Not reported | Not reported |
| **Outcome** |  | Alive, well at D400(8 years)  | Alive, well at D330( 5years) | Alive, well at 7 years | Deceased, respiratory failure,13 years, 7 years | Alive |

 **Supplementary Table 2**: P1's and P2's summaries, compared to previously reported patients, before and after haematopoietic stem cell transplantation (HSCT).

\*: aphthous ulcers, cheilitis, gingivitis, thrush

HSV-1: Herpes virus type 1; VZV: varicella virus; S. aureus: Staphylococcus aureus ; S. pneumoniae : Streptococcus pneumoniae ; EBV : Eptein-Barr virus ; CMV : cytomegalovirus ; RSV: Respiratory syncytial virus; H. influenzae: Haemophilus influenzae; K. pneumonia: Klebsiella pneumonia; CT: computerized tomography; Ig IV: immunoglobulin intravenous; TMP-SMX: Trimethoprim-sulfamethoxazole; GVHD: graft versus host disease; D330: day 330 post HSCT; NA: Not available