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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. aureus  HSV-1, VZV |
| Pulmonary | | S.pneumoniae  Pneumocistis jirovecii, EBV, CMV, adenovirus | | No | Adenovirus | Adenovirus  Clinical bacterial pneumonia | | CMV, RSV | No | | S.pneumoniae, S.aureus  H. influenzae, K. pneumoniae  Pseudomonas  Candida | S. aureus  S.pneumoniae  CMV |
| Gastrointestinal tract | | Salmonella, Campilobacter jejunii  rotavirus, 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 meningitis  H.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 facies  Peanut allergy | NA | |  |  | | Mastoiditis | Dysmorphic facies, bone fractures, granulation  tissue on vocal cord, larynx, ear canal |
| **Treatments** | | | | | | | | | | | | |
|  | Ig IV | | Yes | No | No | | No | Yes | | Yes | Yes | Yes |
|  | Anti-microbial prophylaxis | | TMP-SMX  Acyclovir | TMP-SMX  Acyclovir | TMP-SMX | | TMP-SMX  Acyclovir | Yes | | No | Yes | Yes |
|  | Immunosupresive therapy | | Methylpredisolone  Tacrolimus | 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