

Supplemental information

Mucus sialylation determines intestinal host-commensal homeostasis

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Table S1. Clinical Features of ST6GALNAC1-Deficient Patients, related to Figure 4.

Patient	Pt 1	Pt 2	Pt 3
Age of onset (months)	18	18	3
Current Age (years)	10	9	14
Failure to thrive	Y	Y	Y
Presentation	Recurrent Diarrhea	Recurrent Diarrhea	Oral and perianal inflammation/ulcerations, blood in stool, fever
Endoscopic findings	Aphthous ulcers at the terminal ileum and nodularity in the caecum	Ulcers, edema, fibrin in ileo-cecal region	Superficial ulcer in the esophagus, perianal skin tag and fissures
Imaging findings			Terminal ileitis
Histopathology			
Eosinophil infiltration	Ileum	Esophagus, Ileum, Colon	Stomach
Granulomas	ND	Stomach, Duodenum, Ileum	Mouth
Lymphocytic Infiltration	Colon, Rectum	Duodenum	Esophagus
Goblet cells	Decrease	Decrease	
Cryptitis	Rectum	Ileum	

Table S2. Clinical hematological and immune parameters, related to Figure 4.

	Pt. 1	Normal ranges	Pt. 2	Normal ranges	Pt. 3	Normal ranges
Lymphocyte populations						
% (absolute number)						
CD3	62	60-83 (714-2266)	81 (2859)	60-83 (714-2266)	N.D.	60-83 (714-2266)
CD4/CD3%	24	31-62 (359-1565)	43 (1518)	31-62 (359-1565)	N.D.	31-62 (359-1565)
CD4 naïve (CD62L+/CD45RA+)	49	7.6-37 (102-1041)	N.D.	7.6-37 (102-1041)	N.D.	7.6-37 (102-1041)
CD4 Effector memory (CD62L-/CD45RA-)	26	2.3-15.6 (42-225)	N.D.	2.3-15.6 (42-225)	N.D.	2.3-15.6 (42-225)
CD4 Central Memory (CD62L+/CD45RA-)	21	10.4-30.7 (162-614)	N.D.	10.4-30.7 (162-614)	N.D.	10.4-30.7 (162-614)
CD8/CD3%	28	11-35 (178-853)	33 (1165)	11-35 (178-853)	N.D.	11-35 (178-853)
CD8 naïve (CD62L+/CD45RA+)	29	5.7-19.7 (85-568)	N.D.	5.7-19.7 (85-568)	N.D.	5.7-19.7 (85-568)
CD8 Effector memory (CD62L-/CD45RA-)	40	1.1-9.2 (24-175)	N.D.	1.1-9.2 (24-175)	N.D.	1.1-9.2 (24-175)
CD8 Central Memory (CD62L+/CD45RA-)	2	1.5-10.3 (25-180)	N.D.	1.5-10.3 (25-180)	N.D.	1.5-10.3 (25-180)
CD8 TEMRA (CD62L-/CD45RA+)	N.D.	0.7-7.8 (11-172)	N.D.	0.7-7.8 (11-172)	N.D.	0.7-7.8 (11-172)
CD4/CD8 ratio	0.85	1.1-5.17	1.3	1.1-5.17	N.D.	1.1-5.17
αβDNT Cells	N.D.	1.3-9.2(18-185)	N.D.	1.3-9.2(18-185)	N.D.	1.3-9.2(18-185)
CD20	18	3-19% (59-329)	14 (494)	3-19% (59-329)	N.D.	3-19% (59-329)
Memory B cells (CD20 ⁺ /CD27 ⁺)	N.D.	0.8-3.6 (12-68)	N.D.	0.8-3.6 (12-68)	N.D.	0.8-3.6 (12-68)
Plasma Blasts (CD20 ⁺ /CD38 ⁺)	N.D.	1.2-17.6	N.D.	1.2-17.6	N.D.	1.2-17.6
NK	N.D.	6-34% (126-729)	N.D.	6-34% (126-729)	N.D.	6-34% (126-729)
Immunoglobulin levels						
IgG mg/dL	1140	572-1474	1070	670-1530	910	572-1474
IgM mg/dL	114	34-305	158	52-274	160	34-305
IgA mg/dL	69	20-263	79.8	48-179	150	20-263
IgE IU/mL	16	0-90	289	<52	41	0-90
WBC *10 ⁹ /L	6.6	4.5-11	8.61	5-14.5	19	5.0-12.0
ANC *10 ⁹ /L	2.9	1.5-8	2.95	1.8-8	N.D.	1.8-8
ALC *10 ⁹ /L	2.8	1.5-6.5	1.27	1.5-6.5	N.D.	1.5-6.5
eo *10 ⁹ /L	0.2	(0-0.6)	0.77	(0-0.6)	1	.02-5
Hb g/L	127	121-172	135.5	115-135	118	110-140
Platelets *10 ⁹ /L	403	130-440	437	130-440	322	150-400
Albumin g/L	47	33.0-46.0	29	33.0-46.0	38.7	33.0-46.0
CRP mg/L	N.D.	<1.0	230	0-8	0.4	<1.0
ESR mm/h	5	<10	65	<10	8	<10
Neutrophil count *10 ⁹ /L	2.9	1.5-8.5	N.D.	1.5-8.5	12.4	1.5-8.5
Lymphocyte count *10 ⁹ /L	2.8	2.0-8.0	N.D.	2.0-8.0	4.3	2.0-8.0
ANA	N.D.	negative	1:160	negative	negative	negative
ASCA IgA KEU/L	negative	<20	N.D.	<20	1	<20
ASCA IgG KEU/L	positive	<20	43.5	<20	10	<20
ANCA	negative	negative	negative	negative	negative	negative
Neutrophil oxidative burst index	N.D.	32-300	normal	32-300	303	32-300

Abbreviations

TEMRA	Terminally differentiated effector Memory T cells
αβDNT	CD4 and CD8 Double Negative alpha/beta T-cell receptor T cells
NK	Natural Killer cells
WBC	White Blood Cells
ANC	Absolute Neutrophil Count
ALC	Absolute Lymphocyte Count
eo	eosinophil count
Hb	Hemoglobin
CRP	C-Reactive Protein
ESR	Erythrocyte Sedimentation Rate
ANA	Antinuclear Antibodies
ASCA	Anti-Saccharomyces Cerevisiae Antibodies
ANCA	Antineutrophil Cytoplasmic Antibodies
N.D.	Not determined