

Supplementary Tables

Supplementary Table 1. Parotid gland biopsy results in a subpopulation of patients with major salivary gland enlargement

	pedSjD n=18	diagnostic adSjD n=12	pedSjD vs diagnos- tic adSjD p-value	trial adSjD n=33	pedSjD vs. trial adSjD p-value
Focus score*	2.7 (1.75-3.55)	1.1 (1.0-1.78) [^]	0.010	1.84 (1.32-4.72)	0.72
CD45+ cells * (% of parenchyma)	20.65 (9.82-36.33)	15.22 (3.76-25.41)	0.19	19.84 (6.97-32.94)	0.622
LELs/mm ² *	0.05 (0.00-0.31)	0.16 (0.0125-0.26)	0.73	0.14 (0.00-0.49)	0.52
Presence of LELs, n(%)	9 (50)	9 (75)	0.26	21 (66)	0.28
LEL severity (1-3)*	2 (1-3)	2 (1-2.5)	0.78	1 (1-2.5)	0.38
CD21+ FDC- networks/mm ² *	0.21 (0.09-0.86) [^]	0.00 (0.10-0.38)	0.14	0.26 (0.00-0.49)	0.41
Bcl6+ Ectopic GC/mm ² *	0.15 (0.08-0.34)	0.00 (0.00-0.13)	0.024	0.00 (0.00-0.32)	0.098
CD3+ cells/mm ² *	1444 (977-2058)	470 (177-1406)	0.090	718 (258-1960)	0.168
CD20+ cells/mm ² *	1735(592-2818)	488 (98-877)	0.014	651 (136-1510)	0.020
CD20/(CD20+CD3)ratio*	0.52 (0.36-0.59)	0.39 (0.27-0.50)	0.042	0.40 (0.24-0.48)	0.011
CD3/CD20 segregation, n(%)	14 (79)	9 (75)	0.60	19 (58)	0.22
IgA/IgG plasma cell shift, n(%)	9 (50)	7 (58)	0.47	17 (52)	0.92
IgM+ plasma cells/mm ² *	70 (32.5-168)	24.1 (12.5-51.7-93)	0.018	18.4(4.3-80.4)	0.002

Data is reported as median (IQR)* or n(%).

Abbreviations: LEL: lymphoepithelial lesion, mm²: square millimeter, IQR: interquartile range, FDC: follicular dendritic cell, GC: germinal center

Significant p-values are italicized and bold

Missing data: ^0-5, \$6-10, *11-15, #16-20

Supplementary Tabel 2. Parotid gland biopsy results in a subpopulation of anti-SSA positive patients

	pedSjD n=13	diagnostic adSjD n=31	pedSjD vs diagnostic adSjD p-value	trial adSjD n=36	pedSjD vs. trial adSjD p-value
Focus score*	2.8 (1.95-3.7)	1.1 (0.3-1.8) [^]	0.002	2.42 (1.29-5.22)	0.89
CD45+ cells * (% of parenchyma)	28.5 (16.4-40.4-)	10.80 (3.36-24.44)	0.006	20.05 (8.57-34.82)	0.28
LELs/mm2*	0.25 (0.00-0.47)	0.14 (0.00-0.26)	0.31	0.14 (0.00-0.46)	0.70 [^]
Presence of LELs, n(%)	8 (62)	18 (58)	1.00	23 (64)	0.88
LEL severity (1-3)*	2 [^] (1-3)	2 (1-3)	0.89	1 * (1-2)	0.32
CD21+ FDC-networks/mm2 *	0.43 (0.09-1.1) [^]	0.00 (0.00-0.36)	0.009	0.27 (0.00-0.49)	0.16
Bcl6+ Ectopic GC/mm2*	0.2 (0.09-0.37)	0.00 (0.00-0.13)	<0.001	0.00 (0.00-0.30)	0.032
CD3+ cells/mm2*	1738 (1031-2648)	474 (170-1450)	0.015	962 (438-1962)	0.21
CD20+ cells/mm2*	2793(1423-3156)	381 (66.5-1039)	<0.001	700 (292-1591)	0.007
CD20/(CD20+CD3)ratio*	0.58 (0.51-0.60)	0.41 (0.26-0.48)	0.013	0.41 (0.237-0.52)	0.004
CD3/CD20 segregation, n(%)	10 (77)	17 (55)	0.20	24 (67)	0.73
IgA/IgG plasma cell shift, n(%)	8 (62)	18 (58)	0.83	24 (67)	0.75
IgM+ plasma cells/mm2*	71(30-180)	44 (10-94)	0.083	33 [^] (3.7-94)	0.021

Data is reported as median (IQR)* or n(%).

Abbreviations: LEL: lymphoepithelial lesion., Mm2: square millimeter, IQR: interquartile range, FDC: follicular dendritic cell, GC: germinal center

Significant p-values are italicized and bold

Missing data: ^0-5, \$6-10, *11-15, #16-20