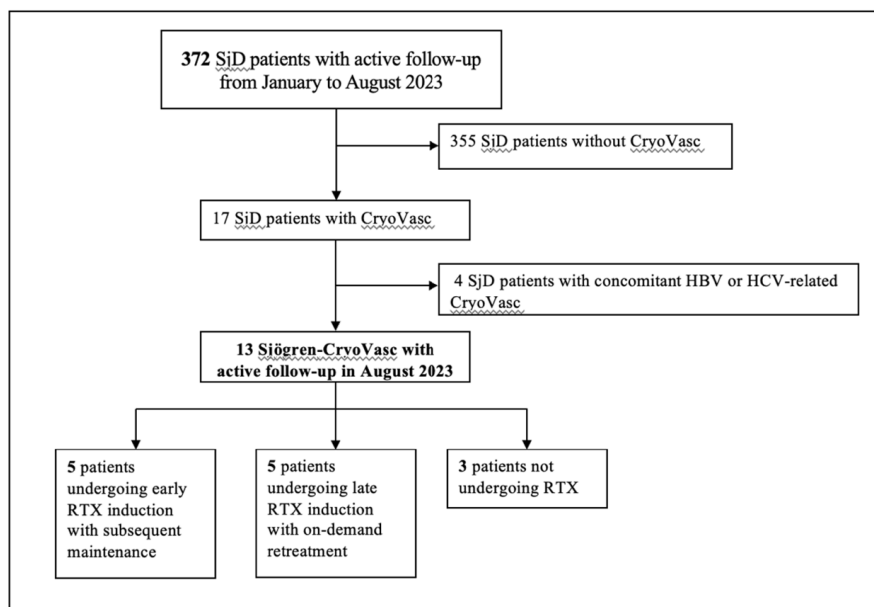




Supplementary Fig. S1. Flow chart of Sjögren-CryoVasc patient selection process.

SjD: Sjögren's disease; Sjögren-CryoVasc: cryoglobulinaemic vasculitis secondary to Sjögren's disease; RTX: rituximab.

Supplementary Table S1. Demographic, clinical and laboratory data at Sjögren-CryoVasc diagnosis.

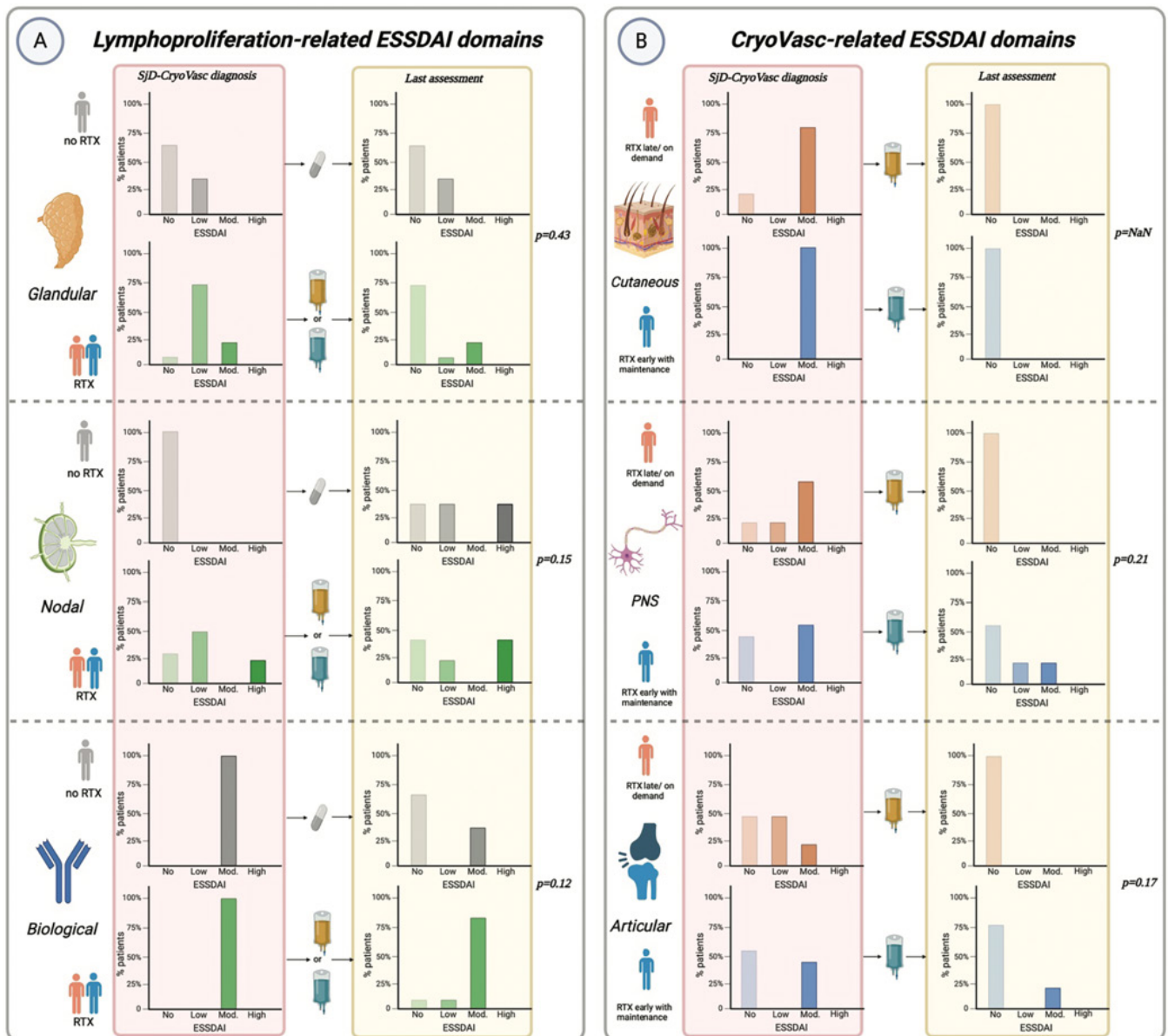
	Group 1 (n=5)	Group 2 (n=5)	Group 3 (n=5)	p-value
Age at Sjögren-CryoVasc diagnosis median years (IQR)	65 (55-67)	62 (51.5-73)	64 (N/A)	0.84
Female, n (%)	3 (60%)	5 (100%)	3 (100%)	29
SjD duration, median years (IQR)	11 (5-17.5)	15 (14.5-21)	9 (N/A)	30
Sjögren-CryoVasc duration, median years (IQR)	4 (4-8)	7 (4.5-11)	5 (N/A)	0.57
Autoantibodies (anti-SSA/SSB +), n (%)	4 (80%)	5 (100%)	3 (100%)	0.9
Cryoglobulins, n (%)	5 (100%)	5 (100%)	3 (100%)	1
Hyper/hipogammaglobulinaemia n (%)	2 (40%)	2 (40%)	1 (33%)	1
Low C4, n (%)	5 (100%)	5 (100%)	1 (33%)	0.03
RF positivity, n(%)	5 (100%)	5 (100%)	3 (100%)	1
Serum monoclonal component,	1 (20%)	3 (60%)	1 (33%)	76
Persistent or recurrent glandular swelling, n (%)	5 (100%)	4 (80%)	1 (33%)	0.18
Previous NHL, n (%)	0 (0%)	2 (40%)	0 (0%)	0.29

IQR: interquartile range; Sjögren-CryoVasc: cryoglobulinaemic vasculitis secondary to Sjögren's disease; SjD: Sjögren's disease; RF: rheumatoid factor; NHL: non-Hodgkin lymphoma.

Supplementary Table S2. Overall ESSDAI score and single ESSDAI domains at Sjögren-CryoVasc diagnosis.

ESSDAI		Group 1 (n=5)	Group 2 (n=5)	Group 3 (n=3)	p-value
Constitutional ESSDAI n, (%)	No	4 (80%)	5 (100%)	3 (100%)	0.99
	Low	1 (20%)	0 (0%)	0 (0%)	
	Moderate	0 (0%)	0 (0%)	0 (0%)	
	High	0 (0%)	0 (0%)	0 (0%)	
Glandular ESSDAI n, (%)	No	0 (0%)	1 (20%)	2 (66%)	0.13
	Low	4 (80%)	3 (60%)	1 (34%)	
	Moderate	1 (20%)	1 (20%)	0 (0%)	
	High	0 (0%)	0 (0%)	0 (0%)	
Cutaneous ESSDAI n, (%)	No	1 (20%)	0 (0%)	0 (0%)	0.99
	Low	0 (0%)	0 (0%)	0 (0%)	
	Moderate	4 (80%)	5 (100%)	3 (100%)	
	High	0 (0%)	0 (0%)	0 (0%)	
Nodal ESSDAI n, (%)	No	0 (0%)	3 (60%)	3 (100%)	<0.001
	Low	5 (100%)	0 (0%)	0 (0%)	
	Moderate	0 (0%)	0 (0%)	0 (0%)	
	High	0 (0%)	2 (40%)	0 (0%)	
Articular ESSDAI n, (%)	No	2 (40%)	2 (40%)	2 (66%)	0.5
	Low	2 (40%)	0 (0%)	1 (34%)	
	Moderate	1 (20%)	3 (60%)	0 (0%)	
	High	0 (0%)	0 (0%)	0 (0%)	
Peripheral nervous system ESSDAI n, (%)	No	1 (20%)	2 (40%)	2 (66%)	0.82
	Low	1 (20%)	0 (0%)	0 (0%)	
	Moderate	3 (60%)	3 (60%)	1 (34%)	
	High	0 (0%)	0 (0%)	0 (0%)	
Renal ESSDAI, n (%)	No	5 (100%)	5 (100%)	3 (100%)	1
	Low	0 (0%)	0 (0%)	0 (0%)	
	Moderate	0 (0%)	0 (0%)	0 (0%)	
	High	0 (0%)	0 (0%)	0 (0%)	
Pulmonary ESSDAI, n (%)	No	5 (100%)	5 (100%)	3 (100%)	1
	Low	0 (0%)	0 (0%)	0 (0%)	
	Moderate	0 (0%)	0 (0%)	0 (0%)	
	High	0 (0%)	0 (0%)	0 (0%)	
Biological ESSDAI n, (%)	No	0 (0%)	0 (0%)	0 (0%)	1
	Low	0 (0%)	0 (0%)	0 (0%)	
	Moderate	5 (100%)	5 (100%)	3 (100%)	
	High	0 (0%)	0 (0%)	0 (0%)	
Overall ESSDAI, median (IQR)		21 (18.5-27)	22 (15-28)	10 (N/A)	0.10

IQR: interquartile range.



Supplementary Fig. S2. Results. (A) Change in glandular, nodal and biological ESSDAI domains in patients treated RTX (groups 1+2) vs. no RTX (group 3). (B) Change in cutaneous, PNS and articular ESSDAI domains in patients treated with early RTX with maintenance (group 1) vs. late RTX/on-demand retreatment (group 2).

CryoVasc: cryoglobulinaemic vasculitis; RTX: rituximab; PNS: peripheral nervous system.