Neurologische Manifestationen des Sjögren Syndroms

Neuroimmunologische Ambulanz

Schwerpunktambulanz für neuroimmunologische Erkrankungen

- Betreuung und Behandlung von Patienten
- Konsultation zur Zweitmeinung
- Interdisziplinärer Austausch
- Beobachtungs- und Therapiestudien
- Enge Anbindung an Grundlagenwissenschaft

Was hat das mit dem Sjögren Syndrom zu tun?

Kompetenznetz Multiple Sklerose

Klinische Manifestation

Sjogren syndrome

Etiology
Secondary form associated with RA, PBC

Epidemiology
♀ >> ♂

Antibody serology
Anti-Ro/SSA antibody and anti-La/SSB antibody, Rheumatoid factors

Complications
Lymphadenopathy, increased risk of B-cell lymphoma

Cardinal symptoms
Sicca symptoms: dry mouth, dry eyes

Note
transplacental transmission possible!

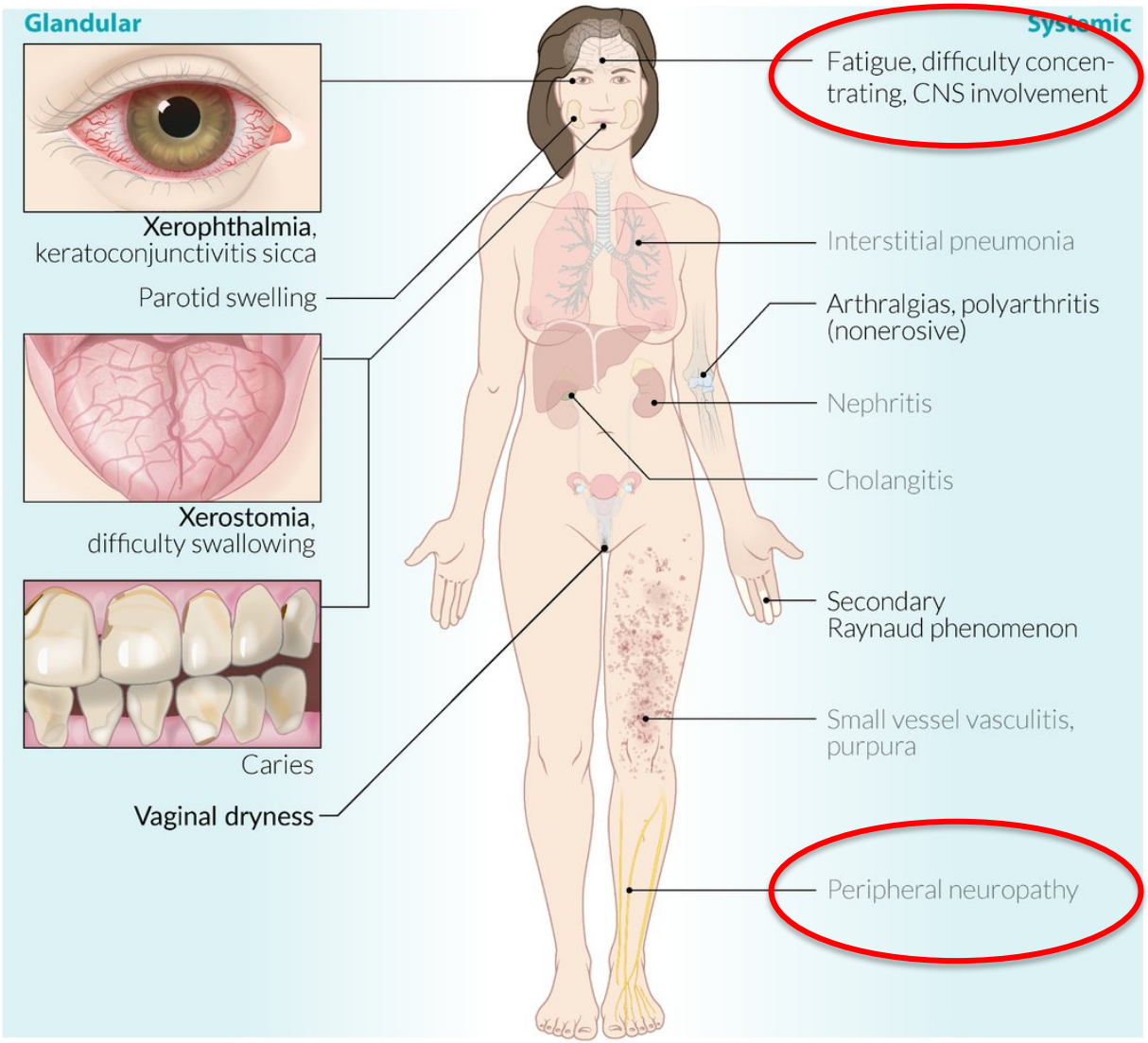

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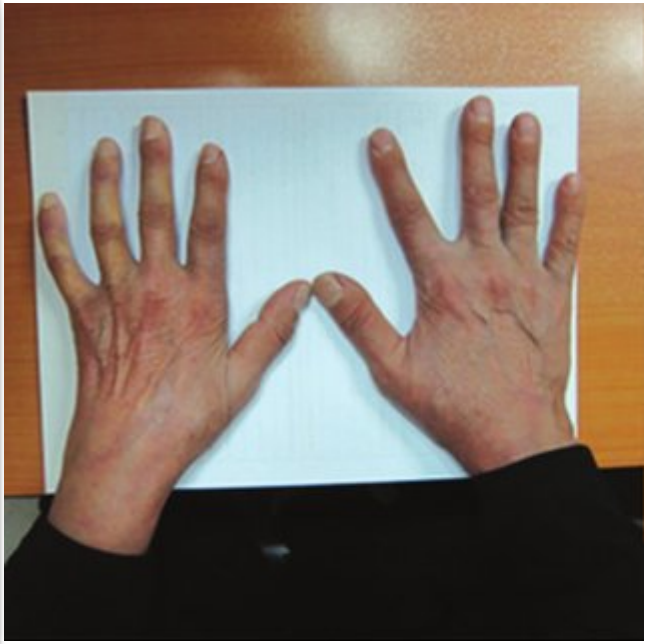
Table II. Epidemiological features, glandular involvement, systemic involvement and immunological profile in patients with primary Sjögren's syndrome with and without features out of the ESSDAI.

Variable	Patients with features out of ESSDAI (n=1641)	Patients without features out of ESSDAI (n=4690)	*p*-value	Adjusted *p*-value	OR [CI95%]
Gender (female)	1534/1641 (93.5)	4383/4690 (93.5)	1	1	1 [0.8-1.27]
Age at diagnosis	51.7 ± 13.3	52.6 ± 14.1	0.018	0.023	
Ethnicity			<0.001	<0.001	
White	1410/1634 (86.3)	3543/4668 (75.9)			
Asian	55/1634 (3.4)	445/4668 (9.5)			
Hispanic	126/1634 (7.7)	455/4668 (9.7)			
Black/African American	20/1634 (1.2)	70/4668 (1.5)			
Others	23/1634 (1.4)	155/4668 (3.3)			
Dry eye	1559/1641 (95)	4404/4690 (93.9)	0.114	0.132	1.23 [0.96-1.61]
Dry mouth	1489/1641 (90.7)	4413/4690 (94.1)	<0.001	<0.001	0.61 [0.5-0.76]
Abnormal ocular tests	1336/1530 (87.3)	3650/4251 (85.9)	0.169	0.187	1.13 [0.95-1.36]
Positive minor salivary gland biopsy	995/1148 (86.7)	3139/3525 (89)	0.033	0.039	0.8 [0.65-0.98]
Abnormal oral diagnostic tests	1034/1325 (78)	2808/3607 (77.8)	0.918	0.949	1.01 [0.87-1.18]
Anti-Ro antibodies	1212/1623 (74.7)	3175/4624 (68.7)	<0.001	<0.001	1.35 [1.18-1.53]
Anti-La antibodies	718/1613 (44.5)	1858/4602 (40.4)	0.004	0.006	1.18 [1.05-1.33]
ANA positive	1338/1618 (82.7)	3643/4582 (79.5)	0.006	0.008	1.23 [1.06-1.43]
RF positive	650/1484 (43.8)	1883/4160 (45.3)	0.346	0.37	0.94 [0.83-1.06]
C3 low	252/1448 (17.4)	383/3962 (9.7)	<0.001	<0.001	1.97 [1.65-2.35]
C4 low	209/1452 (14.4)	381/3984 (9.6)	<0.001	<0.001	1.59 [1.32-1.91]
Positive cryoglobulins	91/1053 (8.6)	122/2222 (5.5)	0.001	0.001	1.63 [1.21-2.18]
ESSDAI	10.3 ± 11.9	5.5 ± 6.3	<0.001	<0.001	
ClinESSDAI	10.8 ± 12.9	5.7 ± 6.9	<0.001	<0.001	
DAS			<0.001	<0.001	
Low	662/1559 (42.5)	2555/4359 (58.6)			
Moderate	476/1559 (30.5)	1311/4359 (30.1)			
High	421/1559 (27)	493/4359 (11.3)			
ESSDAI domains					
Constitutional	262/1637 (16)	348/4565 (7.6)	<0.001	<0.001	2.31 [1.94-2.75]
Lymphadenopathy	235/1641 (14.3)	436/4585 (9.5)	<0.001	<0.001	1.59 [1.34-1.89]
Glandular	520/1635 (31.8)	1029/4562 (22.6)	<0.001	<0.001	1.6 [1.41-1.82]
Articular	782/1638 (47.7)	1787/4564 (39.2)	<0.001	<0.001	1.42 [1.26-1.59]
Cutaneous	265/1641 (16.1)	379/4585 (8.3)	<0.001	<0.001	2.14 [1.8-2.54]
Pulmonary	322/1641 (19.6)	410/4585 (8.9)	<0.001	<0.001	2.49 [2.11-2.92]
Renal	147/1638 (9)	169/4564 (3.7)	<0.001	<0.001	2.56 [2.02-3.24]
Muscular	120/1638 (7.3)	67/4560 (1.5)	<0.001	<0.001	5.3 [3.87-7.3]
PNS	190/1635 (11.6)	200/4558 (4.4)	<0.001	<0.001	2.86 [2.32-3.54]
CNS	53/1635 (3.2)	63/4560 (1.4)	<0.001	<0.001	2.39 [1.62-3.52]
Haematological	438/1634 (26.8)	943/4562 (20.7)	<0.001	<0.001	1.41 [1.23-1.6]
Biological	904/1570 (57.6)	2059/4408 (46.7)	<0.001	<0.001	1.55 [1.38-1.74]

Retamozo et al (2019), Clin Exp Rheumatol

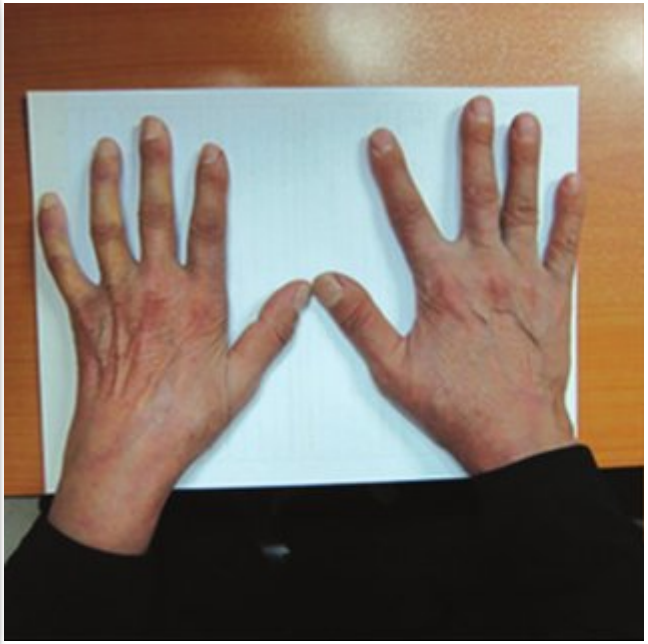
Fallvignette I

- An.: 23j Patientin mit schmerzhafter Schwäche beider Hände seit 9 Monaten
- KU: Hochgradige sensomot. Parese im Medianus-, Ulnaris- sowie Radialisversorgungsgebiet bds.
- Keine eindeutige Keratoconjunctivitis sicca bei kongenitalem Glaukom bds.

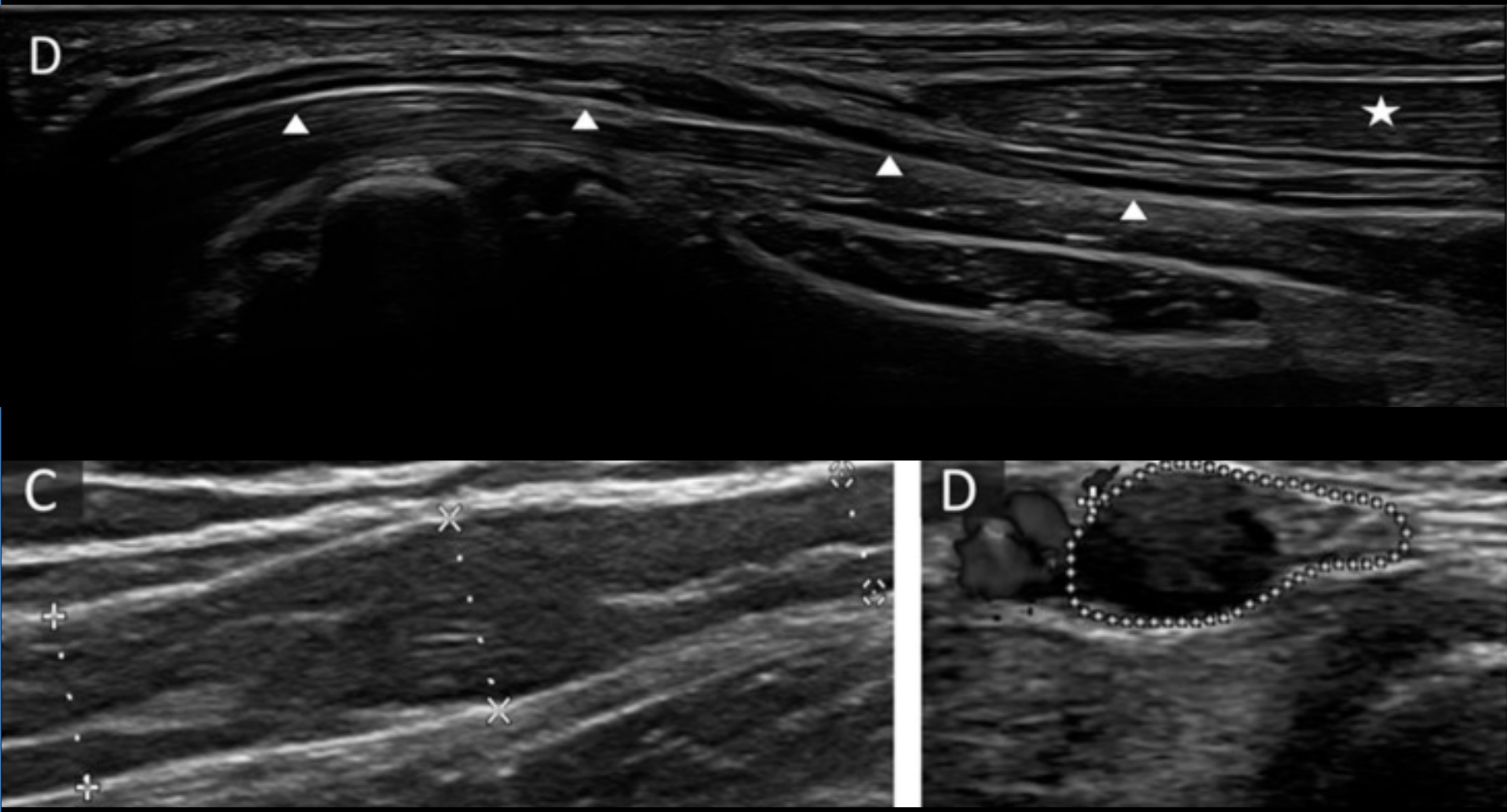


Fallvignette I

- ENG/EMG: Polyneuropathie (genauer: Mononeuritis multiplex)



Nervenultraschall



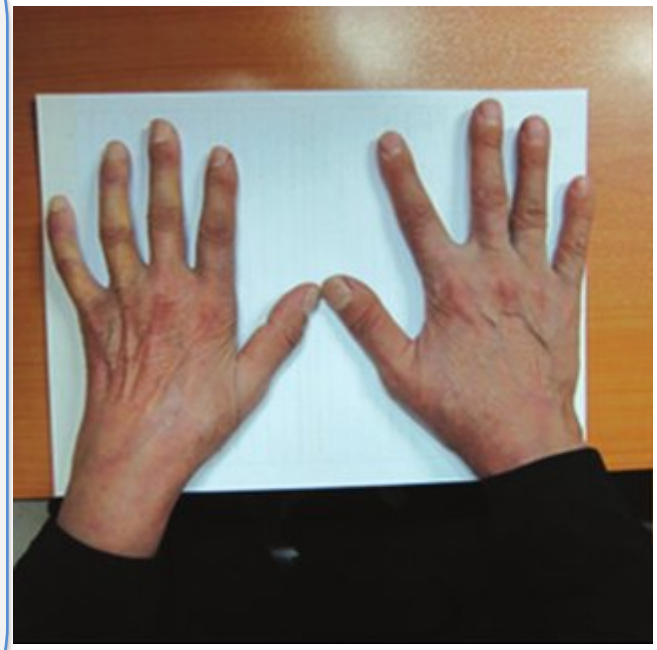
Kramer et al. (2021), Diagnostics (Basel)

Fallvignette I

- Di: ENG/EMG, MR-Extremitäten, Nervenultraschall + Labor
- Labor: ANA 1:800, ANA-Diff. unauffällig
- Lippenbiopsie: C&H 4°

⇒ 7x Plasmapherese + Rituximab

⇒ Nach 3,5a Behandlung beschwerdefrei



Fallvignette II

Wilhelm H and Schabet M, 2015, Dtsch Arztebl

cMRT

Afzali et al, in preparation

Fallvignette II

- 2009 Kribbeln der Zehen seit 2 Jahren
- 2010 neue Sehstörung rechts

⇒ Diagnose: chron.-entz. ZNS-Erkrankung

⇒ Kriterien für MS und NMOSD nicht erfüllt

- Im weiteren Verlauf bildgebende Krankheitsaktivität
- 2012 Arthralgien der Finger und des linken Handgelenks, trockene Augen und Mund

⇒ 2013 Diagnose pSS

⇒ Auf Patientenwunsch keine Behandlung, seit 2014 stabiler Krankheitsverlauf

Kohorte MRI 2017 - 2019

- Bei 14 Patientinnen führte ein neurologisches Symptom zur Erstvorstellung und zur Diagnose
- 5 Patientinnen mit Sicca bei Erstvorstellung
- 4 Patientinnen mit Sicca im Verlauf
- 5 Patientinnen ohne Sicca

Afzali et al, in preparation

Behandlung

Table 2 | **Treatment of primary Sjögren syndrome according to the ESSDAI score by domain**

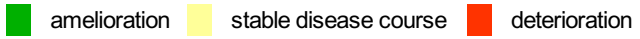
Domain	ESSDAI score*		
	Low	**Moderate**	**High**
Constitutional	• Advise patients with fatigue to exercise (B)	• Hydroxychloroquine (C) • Short-term oral glucocorticoids (C)	• NA
Lymphadenopathy	• Abstention (D)	• Abstention (D)	• Treatment as for lymphoma (D)
Glandular	• Abstention (D)	• Abstention (D)	• Short-term oral glucocorticoids (D) • Sialendoscopy (D) • Intraductal glucocorticoids (D)
Arthralgia or arthritis	• Treatment as for chronic pain, with NSAIDs (C)	• Hydroxychloroquine (C) • Methotrexate (D) • Short-term oral or intra-articular glucocorticoids if arthritis (C)	• Hydroxychloroquine (C) • Methotrexate (D) • Second-line DMARDs as for rheumatoid arthritis if arthritis (C) • Oral glucocorticoids but as briefly as possible (D)
Cutaneous	• Abstention (D) • Cutaneous topical agents (C)	• Abstention (D) • Cutaneous topical agents (C) • Hydroxychloroquine (C)	• Hydroxychloroquine (C) • Oral glucocorticoids (C)
Respiratory	• Treatment of sicca, inhaled glucocorticoids or β2 adrenergic agonists (D)	• Careful monitoring or oral glucocorticoids (D)	• Oral or IV glucocorticoids, immunosuppressants, pirfenidone or nintedanib (C)
Renal	• Abstention and careful monitoring (D)	• Glomerular disease: glucocorticoids (D) • Tubulopathy: K^+ and HCO_3 if necessary (D)	• Glomerular disease: glucocorticoids (C) • Tubulopathy: K^+ and HCO_3 if necessary (D) • Rituximab if cryoglobulinaemia (D)
Muscular	• Abstention (D)	• Glucocorticoids (D)	• Methotrexate plus glucocorticoids (D)
Peripheral nervous system	• Treatment as for chronic pain (D)	• Oral or IV glucocorticoids or IVIg or both (D)	• IV glucocorticoids or IVIg or immunosuppressants (D)
Central nervous system	• NA (D)	• Oral or IV glucocorticoids (D)	• Glucocorticoids or immunosuppressants (D)

Saraux et al. (2016), Nat Rev Rheumatol

Fig. 1

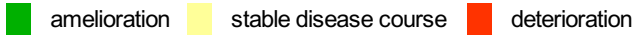
CNS treatment response

A



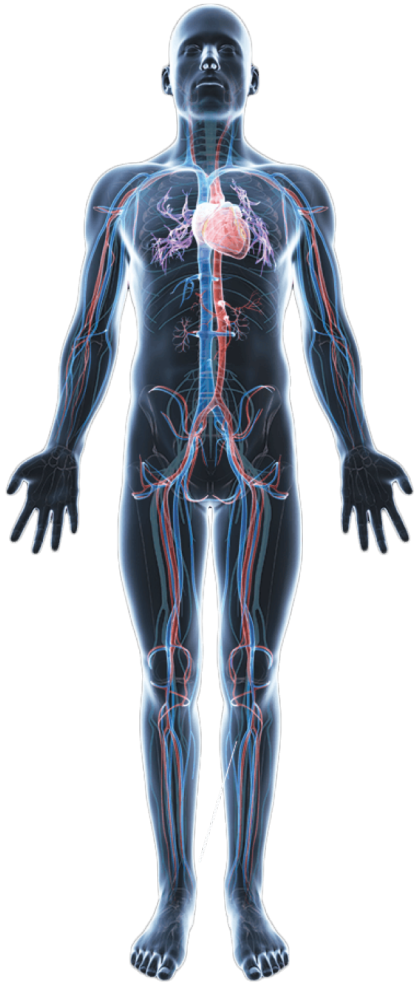
PNS treatment response

B



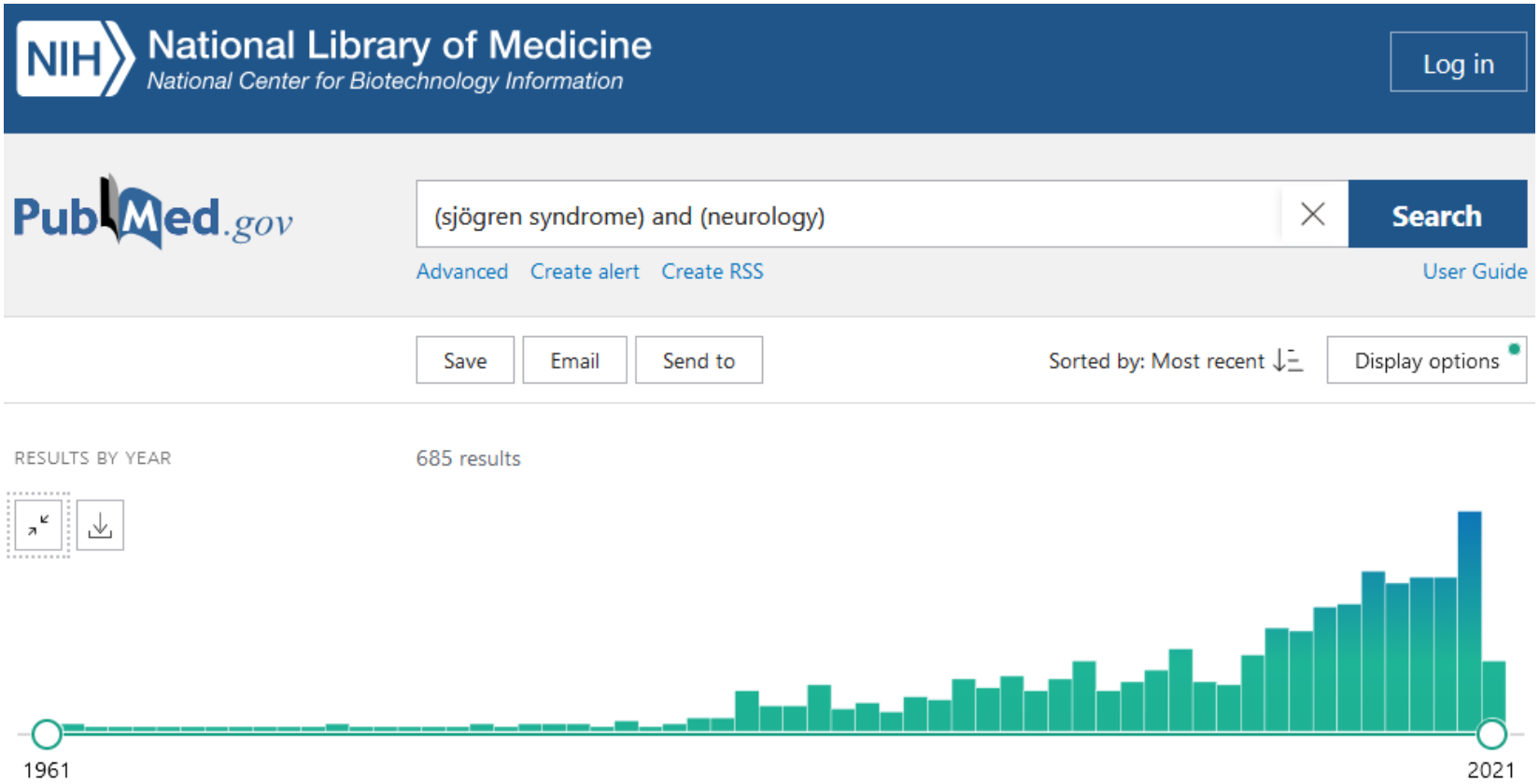
Afzali et al, in preparation

Neurolog. Symptome



- Kopfschmerzen/Migräne
- Kognitive Dysfunktion
- Angst-, affektive Störungen
- Sehstörungen, Doppelbilder
- Motorisch Schwäche
- Taubheit, Kribbeln, neuropathischer Schmerz
- Koordinationsstörungen
- Blasen-/Mastdarmfunktionsstörung
- Gang-/Standstörung

Ausblick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