

ABS0652

### ACCURACY OF ULTRASONOGRAPHY AND INTRANEURAL DOPPLER IN DETECTING CARPAL TUNNEL SYNDROME

**Keywords:** Diagnostic test, Ultrasound

V. Chiarvetto<sup>1</sup>, E. Deltin<sup>1</sup>, N. Marin Zucaro<sup>1</sup>, J. Zacarias<sup>1</sup>, M. Scolnik<sup>1</sup>, E. R. Soriano<sup>1</sup>, J. Rosa<sup>1</sup>. <sup>1</sup>Hospital Italiano de Buenos Aires, Rheumatology Section, Medical Clinic Service, Ciudad Autonoma de Buenos Aires, Argentina

**Background:** Carpal Tunnel Syndrome (CTS) is the most common entrapment neuropathy [1]. The diagnosis of CTS relies on a combination of clinical findings and abnormalities in electrodiagnostic studies (EDX). Although EDXs report high specificity and moderate sensitivity [2], they are invasive and have limited availability. In this context, we aimed to evaluate the diagnostic capacity of combined clinical tests and Doppler ultrasonography using EDXs as a reference standard. Intraneural Doppler has also been proposed as a complementary technique to evaluate vascular changes associated with nerve compression [3].

**Objectives:** To determine the diagnostic utility of cross-sectional area of the median nerve (CSAMN) and intraneural Doppler using ultrasonography (US) for classifying patients with suspected CTS, using EDX as the gold standard.

**Methods:** Cross-sectional, analytical, observational study. Patients over 18 years of age with clinical suspicion of CTS were included, starting from May 2022 to November 2024. Clinical tests, EDX, and US were performed. Patients were classified into the CTS group (positive EDX) or the suspected CTS group (normal EDX). Sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) of intraneural Doppler and CSAMN were calculated. The ultrasonographic technique will be carried out according to the recommendations of EULAR [4].

**Results:** A total of 112 patients were included, 72.3% (95% CI: 63.2–79.9) female, with a mean age at diagnosis of 65.9 years (SD 13.8). Eighty-eight patients were classified as CTS group (80%, 95%CI: 71.4–86.5). The clinical and ultrasound variables are shown in Table 1. The CTS group was significantly older compared to the suspected CTS group (P = 0.016). Only 9 patients presented with positive intraneural Doppler signals, of which 5 were part of the CTS group. The diagnostic performance metrics are summarized in Table 2. Regarding the Likelihood Ratios (LR), the positive LR and negative LR for a CSAMN >10 mm<sup>2</sup> were 0.9 and 1.1, respectively. ROC curves were generated for the CSAMN (AUC: 0.54, 95% CI: 0.44–0.64). A CSAMN ≥18 mm<sup>2</sup> demonstrated a specificity of 100%.

**Table 1.** Baseline characteristics and CTS classification according to EDX.

Characteristics by groups	Total (n= 112)	CTS (n= 88)	Suspected CTS (n= 24 )	Valor P*
Female sex; n; % (CI 95%)	81: 72.3% (63.2-79.9)	64: 72.7% (62.3-81.1)	17:70.8 % (48.9-86.1)	ns
Age at diagnosis; years, median; (SD)	65.9 (13.8)	67.8 (13.1)	57.9 (14.1)	0,0016
Right Index Hand; n; % (CI 95%)	67: 59.8 % (50.4-68.5)	51: 57.9 % (47.3-67.9)	16: 66.6% (44.9-83.1)	ns
Positive Durkan test; n; % (CI 95%)	68: 62.3% (52.8-71.1)	53: 61.6% (50.8-71.4)	15: 65.2% (43.1-82.2)	ns
Positive Tinel test; n; % (CI 95%)	84: 75% (66.1-82.2)	67: 76.1% (65.9-83.9)	17: 70.8% (48.9-86.0)	ns
Positive Phalen test; n; % (CI 95%)	71: 63.6% (54.1-72.1)	56: 65.1% (54.3-74.5)	14: 58.3% (37.3-76.7)	ns
CSAMN; median (SD)	15,1 (4,3)	15,3 (4,2)	14,2 (4,7)	p=0,13
CSAMN ≥ 10 mm <sup>2</sup> ; n; % (CI 95%)	96: 85.7% (77.8-91.1)	78: 88.6% (80.0-93.8)	18: 75%(53.1-88.8)	ns
Positive Doppler signal; n; % (CI 95%)	9: 8.1% (4.2-14.9)	5: 5.6% (2.3-13)	4**: 17.4% (6.3-39.7)	ns
EDX Mild;n; % (CI 95%)	37: 33.1% (24.9-42.3)	36: 40.9% (31.1-51.6)	-	-
EDX Moderate;n; % (CI 95%)	28: 25% (17.7-33.9)	28: 31.8% (22.8-42.3)	-	-
EDX Severe; n; % (CI 95%)	26: 23.2% (16.2-32.1)	24: 27.2% (18.9-37.6)	-	-

CSAMN (cross-sectional area of the median nerve), CTS (Carpal Tunnel Syndrome), MN (Median Nerve), and EDX (Electrodiagnostic Studies), ns (not statistically significant).

\*\*They were classified as suspected CTS due to the absence of EDX at the time of analysis.

**Table 2.** Diagnostic Performance Metrics.

Metrics	Sensitivity	Specificity	PPV	NPV
Positive Doppler signal	5.6 %	82%	55%	18.6 %
CSAMN ≥ 10mm	93%	35%	88%	75%
CSAMN ≥ 10mm or Positive Doppler signal	81%	29%	82%	30%

CSAMN (cross-sectional area of the median nerve)

**Conclusion:** The CSAMN showed moderate performance as a diagnostic test (high S y low ES). While the number of patients with positive intraneural Doppler signals was low, it demonstrated a specificity of 82% for diagnosing CTS. Notably, a CSAMN ≥18 mm<sup>2</sup> achieved a specificity of 100%.

#### REFERENCES:

- [1] Atroshi I, Gummesson C, Johnsson R, Ornstein E, Ranstam J, Rosén I. Prevalence of carpal tunnel syndrome in a general population. *JAMA*. 1999;282(2):153.
- [2] Jablecki CK, Andary MT, Floeter MK, Miller RG, Quarty CA, Vennix MJ, Wilson JR; American Association of Electrodiagnostic Medicine; American Academy of Neurology; American Academy of Physical Medicine and Rehabilitation. Practice parameter: Electrodiagnostic studies in carpal tunnel syndrome. Report of the American Association of Electrodiagnostic Medicine, American Academy of Neurology, and the American Academy of Physical Medicine and Rehabilitation. *Neurology*. 2002 Jun 11;58(11):1589-92.
- [3] DeJaco C, Stradner M, Zauner D, Seel W, Simmet NE, Klammer A, Heitzer P, Brickmann K, Gretler J, Fürst-Moazedzi FC, Thonhofer R, Husic R, Hermann J, Graninger WB, Quasthoff S. Ultrasound for diagnosis of carpal tunnel syndrome: comparison of different methods to determine median nerve volume and value of power Doppler sonography. *Ann Rheum Dis*. 2013 Dec;72(12):1934-9.
- [4] Costantino F, Carmona L, et al. EULAR recommendations for the reporting of ultrasound studies in rheumatic and musculoskeletal diseases (RMDs). *Ann Rheum Dis*. 2021 Jul;80(7):840-847. doi: 10.1136/annrheumdis-2020-219816. Epub 2021 Jan 22.

**Acknowledgements:** NIL.

**Disclosure of Interests:** None declared.

**DOI:** 10.1136/annrheumdis-2025-eular.B2458

© The Authors 2025. This abstract is an open access article published in *Annals of Rheumatic Diseases* under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>). Neither EULAR nor the publisher make any representation as to the accuracy of the content. The authors are solely responsible for the content in their abstract including accuracy of the facts, statements, results, conclusion, citing resources etc.

ABS0671

### IGG4-RELATED DISEASE IN OVERLAP WITH IDIOPATHIC MULTICENTRIC CASTLEMAN DISEASE: CLINICAL-PATHOLOGICAL CHARACTERISTICS AND THERAPEUTIC IMPLICATIONS OF AN ORPHAN DISEASE

**Keywords:** Observational studies/ registry, Rare/orphan diseases

J. Mahajne<sup>1</sup>, L. Bongiovanni<sup>1</sup>, T. Calimeri<sup>1</sup>, F. Erbella<sup>1</sup>, M. Lanzillotta<sup>1</sup>, A. Ferreri<sup>1</sup>, L. Dagna<sup>1</sup>, M. Ponzoni<sup>1</sup>, E. Della Torre<sup>1</sup>. <sup>1</sup>IRCCS San Raffaele Hospital, Milan, Italy

**Background:** Historically, IgG4-Related Disease (IgG4-RD) and idiopathic Multicentric Castleman Disease (iMCD) have been considered to be mutually exclusive; however, rising evidence of significant overlap between the two exists. Recently, a new clinical entity sharing common features of both IgG4-RD and iMCD, named IgG4-CD, has been described but its features have yet to be characterized.

**Objectives:** To describe the demographic, clinical and histological characteristics of IgG4-CD, its prognosis and response to immunosuppressive treatment.

**Methods:** This retrospective observational study included patients with IgG4-RD referred to the San Raffaele Hospital IgG4-RD Clinic. Patients were evaluated looking for features of iMCD. IgG4-CD was defined histologically based on the concomitant presence on lymph node biopsy of more than 10 IgG4+ plasma cells per HPF, an IgG4/IgG plasma cell ratio >40%, and by common features of CD, such as atrophic germinal centers, enlarged interfollicular areas, and mature plasma cell sheet-like proliferation. Patients with pathology samples positive for human herpes virus 8 were excluded. Patients with concomitant infectious, autoimmune, allergic, and neoplastic conditions were excluded.

**Results:** Six patients (5 males) with a median age of 64 years (range 49-71) at the time of IgG4-CD diagnosis were included. Patients fulfilled the available diagnostic criteria for both IgG4-RD and iMCD. All patients presented with constitutional symptoms (arthralgia, malaise, fatigue) but body temperature was never >38°C. Two patients presented with splenomegaly, detected either on abdominal ultrasound or Computed Tomography (CT) scan. Fluid accumulation, eruptive cherry hemangiomas, and lymphocytic interstitial pneumonitis were not detected in any of the patients. All patients had lymph node (LN) biopsies showing features consistent with IgG4-CD, with prominent sheet-like plasmacytosis. Thoracic and abdominal LNs were involved in 5 cases each, pelvic LNs and cervical LNs in 4 cases each, and axillary LNs in 3 cases. IgG4-RD involved the salivary glands in 3 cases, the pancreas and the lacrimal glands in 2 cases, the bile ducts and mesentery in 1 case each (Table 1). All patients had elevated serum IgG4 (median 5732 mg/L, normal <864 mg/L). The median serum IgG4/IgG ratio was 24% (range 7%-56%). ESR was elevated in all patients (range 44-120 mm/h, normal <15 mm/h). CRP was elevated in 4 patients (range 1.5-11.3 mg/L, normal <6 mg/L). IL-6 was elevated in 4 patients (range 8.7-21.5 pg/mL, normal <7 pg/mL). Serum B2-microglobulin was elevated in 3 patients (range 3-10.9 mg/L, normal <3 mg/L). Soluble IL2R assessed in only one patient was 4420 pg/mL (normal <2000 pg/mL). Serum IgA concentration was within normal range in all patients. Serum IgE concentration was elevated in 4 patients (range 2-468 IU/mL, normal <100 IU/mL). Polyclonal hypergammaglobulinaemia was observed in 4 patients. Three patients were anemic and one patient was thrombocytopenic. Four patients were treated with glucocorticoid monotherapy, 1 patient was treated with combined glucocorticoids and cyclophosphamide; 1 patient was kept on watchful waiting. Second line therapies included azathioprine 1 case, and rituximab in 2 cases. Third line rituximab was used in the 2 patients treated with azathioprine and cyclophosphamide due to persistent disease activity (Table 2). Rituximab led to IgG4-CD remission in all 4 cases but disease relapsed in 3 patients up to 3 times. The 5-year mortality rate from the onset of IgG4-CD symptoms was 0%.

**Table 1.** IgG4-CD patients' demographics and organ involvement.

Patient	Age	Gender	IgG4-RD organ involvement	IgG4-CD LN involvement
1	73	M	Salivary glands, bile ducts, pancreas	Cervical, thoracic, abdominal, pelvic, axillary
2	72	F	Salivary glands, lacrimal glands	Cervical, thoracic, pelvic
3	70	M	Lacrimal glands	Thoracic, abdominal
4	56	M	Salivary glands	Cervical, thoracic, abdominal, axillary
5	61	M	Mesentery	Abdominal, pelvic
6	68	M	Pancreas	Cervical, abdominal, pelvic, axillary, thoracic

**Table 2.** Laboratory values and treatment status in IgG4-CD patients. Normal (N); low (L); not assessed (NA); Yes (Y); prednisone (PDN); rituximab (RTX); cyclophosphamide (Cyc); azathioprine (AZA)

Pt	IgG4 (mg/L)	IgG4/IgG (%)	IgA	IgE (IU/mL)	ESR (mm/h)	CRP (mg/L)	IL6 (pg/mL)	B2-MG (mg/L)	sIL2R (ng/mL)	Hb	PLT	Polyclonal hyper-Ig	Treatment
1	1400	8.55	N	2	75	11.3	8.7	10.9	NA	N	N	Y	PDN/RTX
2	8234	30.68	N	468	91	1.5	21.5	0	NA	L	N	Y	PDN/RTX
3	11200	56.00	N	NA	0	0	14.6	3	NA	L	N	No	PDN/RTX/RTX
4	6400	9.28	N	255	120	2	0	6	NA	L	L	Y	PDN/Cyc/RTX
5	1183	7.51	N	282	44	7	9	0	4420	N	N	No	WW
6	5974	29.50	N	200	120	5.4	0	NA	NA	N	N	Y	PDN/AZA/RTX

**Conclusion:** We here describe six patients presenting with IgG4-CD. Our findings suggest that IgG4-CD is a relapsing-remitting condition characterized by elevated ESR, CRP, IL-6, and IgG4 levels, tissue plasmacytosis, good response to anti-CD20 therapy, and a favorable prognosis. Larger cohort studies will be needed to further characterize this novel entity.

**REFERENCES:**

[1] Zhang X, Zhang P, Peng L, Fei Y, Zhang W, Feng R, Zhang W. Clinical characteristics of a concurrent condition of IgG4-RD and Castleman's disease. *Clin Rheumatol.* 2018 Dec;37(12):3387-3395. doi: 10.1007/s10067-018-4165-4. Epub 2018 Jun 14. PMID: 29948354.

[2] Fajgenbaum DC, Uldrick TS, Bagg A, Frank D, Wu D, Srkalovic G, Simpson D, Liu AY, Menke D, Chandrakasan S, Lechowicz MJ, Wong RS, Pierson S, Paessler M, Rossi JF, Ide M, Ruth J, Croglio M, Suarez A, Krymskaya V, Chadburn A, Colleoni G, Nasta S, Jayanthan R, Nabel CS, Casper C, Dispenzieri A, Fossà A, Kelleher D, Kurzrock R, Voorhees P, Dogan A, Yoshizaki K, van Rhee F, Oksenhendler E, Jaffe ES, Elenitoba-Johnson KS, Lim MS. International, evidence-based consensus diagnostic criteria for HHV-8-negative/idiopathic multicentric Castleman disease. *Blood.* 2017 Mar 23;129(12):1646-1657. doi: 10.1182/blood-2016-10-746933. Epub 2017 Jan 13. PMID: 28087540; PMCID: PMC5364342.

[3] Umehara H, Okazaki K, Masaki Y, Kawano M, Yamamoto M, Saeki T, Matsui S, Yoshino T, Nakamura S, Kawa S, Hamano H, Kamisawa T, Shimosegawa T, Shimatsu A, Nakamura S, Ito T, Notohara K, Sumida T, Tanaka Y, Mimori T, Chiba T, Mishima M, Hibi T, Tsubouchi H, Inui K, Ohara H. Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011. *Mod Rheumatol.* 2012 Feb;22(1):21-30. doi: 10.1007/s10165-011-0571-z. Epub 2012 Jan 5. PMID: 22218969.

**Acknowledgements:** NIL.

**Disclosure of Interests:** None declared.

**DOI:** 10.1136/annrheumdis-2025-eular.B2526

© The Authors 2025. This abstract is an open access article published in *Annals of Rheumatic Diseases* under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>). Neither EULAR nor the publisher make any representation as to the accuracy of the content. The authors are solely responsible for the content in their abstract including accuracy of the facts, statements, results, conclusion, citing resources etc.

**ABS0709 CLINICAL AND IMAGING FEATURES OF HAEMOCHROMATOSIS ARTHROPATHY - A SYSTEMATIC LITERATURE REVIEW AND META-ANALYSIS TO INFORM THE DEVELOPMENT OF EULAR CLASSIFICATION CRITERIA**

**Keywords:** Imaging, Epidemiology, Systematic review

S. Engelhardt<sup>1,2</sup>, S. Buroh<sup>3</sup>, G. Schwarzer<sup>4</sup>, A. Kerschbaumer<sup>5</sup>, A. Braner<sup>6</sup>, M. Herber<sup>7</sup>, P. Machado<sup>8,9,10</sup>, P. Kiely<sup>11,12</sup>, S. Finzel<sup>1</sup>. <sup>1</sup>University of Freiburg, Faculty of Medicine, University of Freiburg, Department of Rheumatology and Clinical Immunology, Medical Center, Freiburg, Germany; <sup>2</sup>Klinikum Freising, Freising, Department of Internal Medicine, Cardiology, Klinikum Freising, Freising, Germany; <sup>3</sup>Library of the Medical Center - University of Freiburg, Faculty of Medicine, Library of the Medical Center - University of Freiburg, Faculty of Medicine, University of Freiburg, Freiburg, Germany; <sup>4</sup>University of Freiburg, Freiburg, Germany, Institute of Medical Biometry and Statistics, Faculty of Medicine and Medical Center - University of Freiburg, Freiburg, Germany, Freiburg, Germany; <sup>5</sup>Medical University of Vienna, Vienna, Austria, Department of Medicine 3, Division of Rheumatology, Vienna, Austria; <sup>6</sup>MVZ Aschaffenburg, Aschaffenburg, Germany; <sup>7</sup>Strasbourg Centre University, Strasbourg, France, Department of Internal Medicine, Strasbourg, France; <sup>8</sup>University College London, London, UK, Department of Neuromuscular Diseases, London, United Kingdom; <sup>9</sup>University College London Hospitals NHS Foundation Trust, London, UK, National Institute for Health Research, University College London Hospitals Biomedical Research Centre, London, United Kingdom; <sup>10</sup>London North West University Healthcare NHS Trust, London, UK, Department of Rheumatology, Northwick Park Hospital, London, United Kingdom; <sup>11</sup>St George's University Hospitals NHS Foundation Trust, London, UK, Department of Rheumatology, London, United Kingdom; <sup>12</sup>City St George's, University of City St George's, University of London, UK, UK, Institute of Medical and Biomedical Education, City St George's, University of London, UK, United Kingdom

**Background:** Haemochromatosis arthropathy (HA) has been little researched to date. Arthropathy frequently is amongst the first clinical manifestations of haemochromatosis, but might be overlooked or misdiagnosed as being different origin, which impacts significantly on the patient's joint function and health due to delayed diagnose and therapy initiation.

**Objectives:** To summarize the imaging and clinical features of haemochromatosis arthropathy to inform the development of EULAR classification criteria for HA.

**Methods:** We performed a systematic literature review by searching Medline (PubMed, Ovid), Cochrane Library and Embase, and conference abstracts. We