events, 30%, predominantly in group E (75%), and infection, 30%, predominantly in group A. The median from diagnosis to death was 3 years (range 1 - 13).

Figure 1. Group A (red line) according to ACG 1990 criteria and Group B (green line) according to ACG criteria and implementing TAUS for rapid diagnosis

Conclusion: The implementation of temporal artery ultrasound (TAUS) is associated with a significant improvement in the survival rate of patients with GCA and a reduction in treatment-related complications in patients who were diagnosed with ultrasound in less than 7 days compared to those diagnosed by the conventional healthcare attention routes.

REFERENCES:

- [1] Gonzalez-Gay MA, et al. Giant cell arteritis: epidemiology, diagnosis, and management. DOI: 10.1007/s11926-010-0135-9
- [2] Patil P, et al. Fast track pathway reduces sight loss in giant cell arteritis: results of a longitudinal observational cohort study. PMID: 26016758
- [3] Breuer GS, et al. Survival of patients with giant cell arteritis: a controversial issue. PMID: 31969222
- [4] Diamantopoulos AP, et al. The fast-track ultrasound clinic for early diagnosis of giant cell arteritis significantly reduces permanent visual impairment: towards a more effective strategy to improve clinical outcome in giant cell arteritis? 10.1093/rheumatology/kev289

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Vasculitis – small vessel vasculitis

AB0385

THE EFFECTIVENESS OF THE ANTI-TUMOR NECROSIS FACTOR THERAPY INFliximab IN NEURO-BEHÇET'S DISEASE: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: The use and benefit of biologic targeted therapy in the treatment of refractory manifestations of behcet's syndrome remain unclearly defined due to the relative paucity of published research. Neuro-behçet's disease represents a challenging clinical presentation. The lack of identified effective strategies in the management of refractory neuro-behçet's adds to the disease burden and contributes to lack of tight control. [1-3]

Objectives: The aim of this systematic review and meta-analysis is to examine the effect of anti-TNF therapy on refractory NBD by performing a systematic review and meta-analysis of the previously published studies assessing the effectiveness of anti-TNF therapy in patients with BD having refractory or recurrent neurological manifestations.

Methods: Authors used the PICO Model (P= population, I= intervention, C= comparator, O= outcome) used for designing the research question. The PRISMA statement was used for developing the study protocol search methodology. Literature searches were done for articles in published in English language only from January 2000 till January 2020 (considering the first till the last available publication that addresses the research question and satisfied eligibility criteria).

The study was registered on the Prospero PROSPERO website for systematic reviews and meta-analysis. Random-effects meta-analysis was performed. Inter-study heterogeneity was explored using I² statistics. Cumulative meta-analysis was conducted to assess temporal trend for accumulating evidence from accruing published studies. Meta-regression was used to analyze possible confounders.

Authors of the current systematic review and meta-analysis worked on a simple score "Hamdy and Woldeamanuel simple response score, 2020", the aim behind the score was to incorporate standard evaluation parameters for assessment of responses driven by the measures used in each study. The authors used numerical values to grade the pattern of response in each parameter considered attempting to standardize the quantification of the responses and minimize tendency towards overestimation. The score included clinical and radiographic points rated on a scale of 5.

Results: Twenty-one studies involving 64 patients were included, 52 male patient and 12 females at a ratio of 4.3:1, with a mean age of 38 .21 years, mean disease duration of 84.76 5 months. Effect size analysis showed that 59 out of 64 patients i.e. 93.7% of the treated patients with neuro-behçet's disease in the analysis were responders to infliximab therapy (CI 88% - 99.3%). There was no significant inter-study heterogeneity (I² = 0%, p = 0.744). Cumulative analysis showed accumulating evidence favoring increasing effectiveness over the last 20 years. There was no statistically significant confounding of infliximab effect size by age (p = 0.89), sex ratio (p = 0.29), and disease duration (p = 0.67).

Conclusion: In this systematic review and meta-analysis Infliximab showed a strong therapeutic effectiveness in the treatment of refractory neuro-behçet's disease. There is an increasing trend of effectiveness observed in the last two decades which may be due to improved neuro-behçet's diagnostic accuracy.

REFERENCES:

- [1] Uygunoğlu U, Siva A. Behçet's Syndrome and Nervous System Involvement. *Curr Neurol Neurosci Rep.* 2018;18(7):35. Published 2018 May 23. doi:10.1007/s11910-018-0843-5.
- [2] Ohno S, Ohguchi M, Hirose S, Matsuda H, Wakisaka A, Aizawa M. Close Association of HLA-Bw51 With Behçet's Disease. *Arch Ophthalmol.* 1982; 100(9):1455–1458. doi:10.1001/archophth.1982.01030040433013.
- [3] Kalra, S., Silman, A., Akman-Demir, G., et al. Diagnosis and management of Neuro-Behçet's disease: international consensus recommendations. *Journal of neurology*, 2014; 261(9): 1662–1676. <https://doi.org/10.1007/s00415-013-7209-3>.

Figure 1 a. Forest plot summarizing the meta-analysis results on the effectiveness of infliximab to treat Neurobehçet disease. Overall, 94% of patients responded to infliximab.

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AB0386

CORONAVIRUS INFECTION AND VASCULITIS: IDENTIFYING ASSOCIATIONS MINING THE BIOMEDICAL LITERATURE

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Background: Based on recent publications suggesting an association between COVID-19 and vascular Inflammation.

Objectives: Our aim was to explore new associations between coronavirus infections and vasculitis utilizing semantic mining of PubMed results.

Methods: The following literature search string: "(vasculitis OR vascular inflammation OR vascular damage) AND (coronavirus OR SARS virus OR

MERS-CoV OR Covid-19)" was used to retrieve abstracts from the whole PubMed database, using Semantic MEDLINE 2. on 6/7/2020. This application represents a network of semantic predications (triples of the form subject-predicate-object, e.g. COVID-19 causes Disease) on a knowledge graph. The system allows for choosing the maximum number of nodes represented, the central topic, and the length of the network. For our network we chose to display all relations, COVID-19 (31 edges) as the central term, 3 lengths, and selecting the most informative nodes. Automatic summarization eliminated the less informative predications.

Results: The search string retrieved 152 citations from PubMed and identified 1,028 predications. The

network (Figure 1), displayed using COVID-19 as the central term, consisted of 72 nodes and 140 edges. The 5 most connected nodes were 'Patients: 19 nodes', 'COVID-19: 13', 'Inflammation: 13', 'Lung: 11', and 'Disease: 11'. Multiple links have been found between coronavirus and vasculitis. Animal coronaviruses, including the one causing feline infectious peritonitis (FIP), the murine coronavirus mouse hepatitis virus (MHV), the SARS-CoV in transgenic mice and coronavirus in ferrets, are known to cause vasculitis in animals. It is known that coronaviruses that infect animals can evolve and become new human coronaviruses. SARS produces inflammation in blood vessels. In 2005, a link between the coronavirus HCoV-NL63 or New Haven Coronavirus (HCoV-NH) and KD was reported, although later studies concluded that HCoV-NH did not play a dominant role in the etiology or

pathogenesis of KD. In 2014, serological testing suggested the possible involvement of CoV-229E in the development of KD. There has also been a report of KD patients being infected by coronavirus OC43/HKU1. COVID-19 may infect the vessels and trigger inflammatory reactions like those of vasculitis, including vasculitis-like cutaneous lesions. COVID-19 patients develop thrombosis, and increased risk of thrombosis is also present in primary vasculitic syndromes. Children, many of whom tested positive for COVID-19 antibodies, developed Multisystem Inflammatory Syndrome in Children (MIS-C), an inflammatory condition similar to Kawasaki Disease (KD).

Conclusion: Knowledge integration and discovery methods are an efficient and powerful way of retrieving and analyzing relevant information from multiple papers. Their main advantages are finding relations among biomedical concepts, generating new hypotheses, and opening them to literature-based discovery.

SARS-CoV-2 may cause vasculitis or vasculitis-like syndromes. The KD-like syndrome reported mainly in children with COVID-19 revives the previous suspicion of coronavirus as a possible triggering agent of KD and the decades-old hypothesis of infection involvement in the pathogenesis of vasculitis.

REFERENCES:

- [1] Rindfleisch TC, et al. Semantic MEDLINE: An advanced information management application for biomedicine. *Information Services & Use* 2011;31:15-21.

Figure 1. Semantic Networks Resulting from PubMed. All relations COVID-19 (edges:140) 3 lengths Max nodes: F (all nodes considered relevant).

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AB0387

SYSTEMIC AND LOCAL THERAPIES FOR EAR, NOSE AND THROAT MANIFESTATIONS IN ANCA-ASSOCIATED VASCULITIS: A SYSTEMATIC REVIEW

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Background: Ear, nose and throat (ENT)-involvement is common in antineutrophil cytoplasmic autoantibody- (ANCA)-associated vasculitis (AAV), can lead to permanent damage and is correlated with reduced quality of life. Yet, guidelines on management of ENT-manifestations are lacking.

Objectives: The aim of this study was to evaluate the efficacy of systemic and local therapies for ENT- involvement in AAV.

Methods: A systematic review was conducted. Clinical studies between January 2005 – April 2020, in adults with AAV and ENT-involvement using outcomes reflecting disease activity, relapse and damage were included. Case reports, congress abstracts and non-English articles were excluded.

Results: The search yielded 4,586 results. For full-text assessment, 109 articles were selected. Finally, 25 articles were included (Figure 1). Two placebo-controlled trials were available. The other observational studies had a limited level of evidence (LoE) of 2b-4 according to the Oxford Centre for Evidence-based Medicine.

Ten studies (n=127) with subglottic stenosis evaluated dilatation (n=94), intralelational steroids (n=63), surgery (n=51) or rituximab (n=8), which mostly resulted in temporary response (12-34 months) (Table 1). Otologic manifestations were treated with various disease modifying anti-rheumatic drugs (DMARDs) in 251 patients in six studies. All studies demonstrated improvement in hearing thresholds, but relapse rates were high (47%). Five studies assessed DMARDs or surgery for sinonasal symptoms showing remission in 21-82%. Six studies, which did not specify ENT-symptoms of the included patients, reported remission in 86-96%. Due to heterogeneity across studies no meta-analysis could be performed.

Figure 1. Flowchart of article selection