2018 update of the EULAR recommendations for the management of large vessel vasculitis

12 March, 2020, Kilchberg, Switzerland – The European League Against Rheumatism, EULAR, has announced the publication of new recommendations for the management of large vessel vasculitis, modifying its previous recommendations for the condition, published in 2009.

A EULAR task force conducted a systematic literature review of recently published clinical trials and cohort analyses and sought opinion from 20 experts from 13 countries. Their new report, published in the January 2020 issue of *The Annals of the Rheumatic Diseases*, aims to facilitate the translation of current scientific evidence and expert opinion into better management and improved outcome of large vessel vasculitis (LVV) patients. To date, the recommendation has already been cited many times.

The task force formulated three overarching principles and 10 recommendations that address diagnosis and therapy for patients with LVV.

The three overarching principles are:

A. Patients with LVV should be offered best care which must be based on a shared decision between the patient and the rheumatologist, considering efficacy, safety and costs.
B. Patients should have access to education focusing on the impact of LVV, its key warning symptoms and its treatment (including treatment-related complications).
C. Patients with LVV should be screened for treatment-related and cardiovascular comorbidities. We recommend prophylaxis and life-style advice to reduce cardiovascular risk and treatment-related complications.

The 10 recommendations are:

1. All patients presenting with signs and symptoms suggestive of giant cell arteritis (GCA) should be urgently referred to a specialist team for further multidisciplinary diagnostic work-up and management.
2. All patients presenting with signs and symptoms suggestive of Takayasu arteritis (TAK) should be referred to a specialist team for multidisciplinary diagnostic work-up and management.
3. A suspected diagnosis of LVV should be confirmed by imaging (ultrasound or MRI for temporal or other cranial arteries, ultrasound, CT, positron-emission-tomgraphy (PET)T-CT or MRI for the aorta/extracranial arteries) or histology (TAB).
4. High dose glucocorticoid (GC) therapy (40–60 mg/day prednisone-equivalent) should be initiated immediately for induction of remission in active GCA or TAK. Once disease is controlled, we recommend tapering the GC dose to a target dose of 15–20 mg/day within 2–3 months and after 1 year to ≤5 mg/day (for GCA) and to ≤10 mg/day (for TAK).
5. Adjunctive therapy should be used in selected patients with GCA (refractory or relapsing disease, the presence or an increased risk of GC related adverse effects or complications) using Tocilizumab (TCZ). Methotrexate may be used as an alternative.
6. Non-biological disease modifying agents should be given in combination with GC in all patients with TAK. TCZ or TNF-inhibitors can be considered in case of relapsing or refractory disease despite conventional disease modifying anti-rheumatic drug (DMARD) therapy.
7. In case of major relapse (either with signs or symptoms of ischaemia or progressive vascular inflammation) we recommend reinstitution or dose escalation of GC therapy as recommended for new onset disease. For minor relapses we recommend an increase in GC dose at least to the last effective dose. Initiation or modification of adjunctive therapy should be considered particularly after recurrent disease relapses.
8. Antiplatelet or anticoagulant therapy should not be routinely used for treatment of LVV unless it is indicated for other reasons (eg, coronary heart disease or cerebrovascular disease, etc). In special situations such as vascular ischaemic complications or high risk of cardiovascular disease, these might be considered on an individual basis.
9. In LVV, elective endovascular interventions or reconstructive surgery should be performed in phases of stable remission. However, arterial vessel dissection or critical vascular ischaemia requires urgent referral to a vascular team.

EULAR Secretariat | Seestrasse 240 | 8802 Kilchberg | Switzerland
T: +41 44 716 30 30 | F: +41 44 716 30 39
eular@eular.org | www.eular.org

10. Regular follow-up and monitoring of disease activity in patients with LVV is recommended, primarily based on symptoms, clinical findings and ESR/CRP levels.

Rapid diagnosis and effective treatment are required in LVV in order to treat symptoms, but more importantly, to reduce the risk of complications such as blindness in GCA and aortic aneurysm or vascular stenosis in GCA and TAK.

About EULAR

The European League against Rheumatism (EULAR) is the European umbrella organisation representing scientific societies, health professional associations and organisations for people with RMDs. EULAR aims to reduce the burden of RMDs on individuals and society and to improve the treatment, prevention and rehabilitation of RMDs. To this end, EULAR fosters excellence in education and research in the field of rheumatology. It promotes the translation of research advances into daily care and fights for the recognition of the needs of people with RMDs by the EU institutions through advocacy action.

About The Annals of the Rheumatic Diseases (ARD)

ARD is the EULAR Journal and the highest ranking rheumatology journal for original research, publishing research from clinical, translational and basic sciences related to rheumatological diseases as well as recommendations and points to consider for various diseases.

Contact

Ursula Aring, EULAR Public Affairs Manager, ursula.aring@eular.org Tel. +41 44 716 30 38

Notes to Editors

EULAR Strategy: https://www.eular.org/eular_strategy_2018.cfm
EULAR School: https://esor.eular.org/
EULAR Campaign: https://www.eular.org/eular_campaign.cfm
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EULAR Secretariat | Seestrasse 240 | 8802 Kilchberg | Switzerland
T: +41 44 716 30 30 | F: +41 44 716 30 39
eular@eular.org | www.eular.org

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eular@eular.org | www.eular.org