2.6) had significant effect on SDI. HA at the time of diagnosis or occurring at any time during disease course did not contribute to mortality.

Conclusion: Lymphopenia contributes independently damage accrual in European background. Lymphopenia and thrombocytopenia at early disease appear to be independent risk factors for subsequent damage.

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THU0285

UMBILICAL ARTERIAL DOPPLER ULTRASONOGRAPHY PREDICTS LATE PREGNANCY OUTCOMES IN PATIENTS WITH LUPUS NEPHRITIS: A MULTICENTER STUDY FROM SOUTHERN CHINA

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Background: Compared with the general population, patients with lupus nephritis (LN) are still at high risk of adverse pregnancy outcomes (APOs). Umbilical artery is the last vessel upstream of the placenta and is particularly important for placental perfusion and foetal development. Increased umbilical artery resistance could be traced by Doppler velocimetry, which is frequently related to pre-eclampsia, intrauterine growth restriction (IUGR), and foetal distress. Umbilical artery Doppler is used as a screening tool for placenta-related diseases in the general population. However, the predictive value in complications of LN pregnancies has not been widely assessed.

Objectives: To investigate the fetal adverse pregnancy outcomes (APOs) and the predictive value of umbilical arterial Doppler ultrasonography in the third trimester in pregnant women with LN.

Methods: A retrospective cohort study enrolling 203 LN patients with pregnancies from 2007 to 2017 from three centers was performed. These patients received umbilical arterial Doppler ultrasound examination during 28-34 gestational weeks. Doppler velocity waveforms of the umbilical arteries were assessed with resistance index (RI), systolic/diastolic ratio (S/D) and pulsatility index (PI).

Results: Fetal APOs occurred in over half of the patients (103/203, 50.7%). Sixty-six pregnancies (66/203, 32.5%) ended with preterm births. The incidence rate of IUGR was 18.2% (37/203). Fetal distress was noted in 23 pregnancies (23/203, 11.3%). All the Doppler parameters were higher in patients with IUGR, fetal distress and composite APOs than in patients without any APOs. Patients with IUGR and fetal distress had significantly higher values of PI, RI, and S/D. It was noted that PI, RI and S/D was higher in patients with premature delivery than those with full-term delivery. However, only the difference of RI and S/D reached statistical significance. RI indicated the highest risk of IUGR and composite APOs. The cut-off values were 0.66 and 0.67, respectively. Sensitivities were 51.4% and 33.7%, and specificities were 87.4% and 92.1%. S/D was also a best predictor for IUGR, with the optimal cut-off value of 2.88. Sensitivity and specificity were comparable with RI. PI over 0.84 was an ideal indicator for fetal distress with an optimal combination of sensitivity (89.5%) and specificity (51.6%).

Conclusion: Fetal complications were frequent in patients with LN. Umbilical arterial Doppler ultrasonography was a useful measure to predict late IUGR, fetal distress, and the composite APOs.

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17. Vasculitis

THU0286

MANAGEMENT OF TAKAYASU ARTERITIS: A SYSTEMATIC LITERATURE REVIEW INFORMING THE 2018 UPDATE OF THE EULAR RECOMMENDATIONS FOR THE MANAGEMENT OF LARGE VESSEL VASCULITIS

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Background: European League Against Rheumatism (EULAR) recommendations on large vessel vasculitis (LVV) management were published in 2009.¹ However, given recent advances, especially in therapeutics, an update was needed.

Objectives: To collect available evidence on management of LVV to inform the 2018 update of the EULAR management recommendations.

Methods: Two independent systematic literature reviews (SLRs) were performed, one focused on diagnosis and monitoring and the other on drugs and surgical treatments. Using a predefined PICO strategy, Medline, Embase and Cochrane databases were accessed, and eligible papers reviewed. This abstract reports the main results for Takayasu arteritis (TAK). Results on giant cell arteritis (GCA) are presented elsewhere.

Results: A total of 283 articles were selected from the SLRs. Relevant heterogeneity precluded a meta-analysis.

The clinical pattern of TAK disease may differ according to age and gender. Males appear to have more complications than females. The presence of major complications (e.g. aneurysm formation), a progressive disease course and a weaker inflammatory response appear to relate to a more unfavorable prognosis, with survival rates as low as 43% at 15 years after diagnosis. Thus, follow up is essential, but there is no good quality evidence regarding the best timing and methods for disease monitoring.

High-quality evidence to guide the treatment of TAK was not found. Glucocorticoids (GC) are widely accepted as first line treatment. However, many patients require doses above 10 mg prednisolone per day for disease control. Conventional immunosuppressive drugs and TNF-inhibitors appeared to be beneficial in case series and open-label studies, but high-quality evidence for their use is lacking. Tocilizumab showed modest signs of efficacy in a randomized controlled clinical trial, although the primary endpoint was not achieved. Revascularization procedures (aneurysm/stenosis) may be required and should be performed during stages of inactive disease.

Conclusion: Evidence to guide monitoring and treatment of TAK patients is predominantly derived from observational studies with low level of evidence. Therefore, higher-quality studies are needed in the future.

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THU0287 ANEURYSM IN BEHÇET'S DISEASE: MANAGEMENT, PROGNOSIS AND MORTALITY

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Background: Arterial aneurysms are one of the unique features of Behçet's disease (BD). Although arterial aneurysms are relatively rare, they carry risk of rupture and have poor prognosis.

Objectives: To evaluate the medical and interventional therapies and long-term outcomes of BD patients with extracranial arterial aneurysms.

Methods: We retrospectively reviewed the medical records of 441 BD patients according to ISG criteria between 2013 and 2018 at the Hacettepe University Vasculitis Center (HUVAC). We determined 45 BD patients with an arterial aneurysm. Six patients with isolated carotid and/or cranial arterial aneurysms who were followed by other clinics excluded from the analysis. Overall, 39 BD patients with an extracranial and extra carotid aneurysm included in the study. Data regarding demographic characteristics, clinical, laboratory and vascular imaging findings, history of medical treatments and outcomes were collected. Vascular intervention of patients were grouped as surgery or endovascular intervention. Radiological response after vascular intervention divided into 3 groups as regression, stable and progression according to radiological evaluation. Regression was defined as a decrease or disappearance of the aneurysm. Stable was defined as no change in the aneurysm diameter. Progression was defined as an increase in diameter of a previous aneurysm or occurrence of a new vascular event. We colligated the stable and regression groups and compared with the progression group. Relapse was defined the progression of the aneurysm which underwent an intervention.

Results: A total of 39 BD patients (Male, 80%) with an arterial aneurysm were analyzed in this study. Mean age and mean age of diagnosis patients were 40.9±11.0 and 29.7±7.7 years, respectively. The clinical features of BD was as follows; oral ulcer 100%, genital ulcer 80.6%, acneiform lesion 47.2%, erythema nodosum 33.3%, pathergy positivity 34.5%, arthritis 27.8%, fever 24.3%, uveitis 34.5%, neurological involvement 30.6% and, gastrointestinal involvement 7.7%. Distribution of arterial aneurysms were 5.1% subclavian, 5.1% coronary artery & cardiac, 38.8% pulmonary, 10.3% thoracic aorta, 33.3% abdominal aorta, 5.1% renal, 25.6% iliac and 12.8% femoral. Induction and maintenance treatments are shown in figure 1. In our cohort, 25 (64%) patients needed a vascular intervention, including 9 (23%) open operations and 16 (41%) endovascular interventions. Of 7 (18%) patients had to perform an urgent open operation or endovascular intervention but we didn't find any relation between the progression of an aneurysm and urgent intervention. First induction treatment options and prognosis of arterial aneurysms shown in figure 2. At median 71.8 months follow up, 7 (18%) patient died in the BD aneurysm group. The rate of progression of the aneurysm was 20% in BD patients with pulmonary artery involvement and 64% in BD patients with aortic and its branch involvement (p=0.038). In the absence of medical induction treatment, 5 patients had progression, 1 had regression and 1 had no follow-up.

Abstract THU0287 – Table 1.

	Induction treatment, n=39 (%)	Maintenance treatment*, n=39 (%)
No treatment, n (%)	7** (17.9)	3 (7.6)
Colchicine	32 (82)	15 (38.4)
Medium-high dose steroid, n (%)	31 (79.4)	11 (28.2)
Cyclophosphamide, n (%)	18 (46.1)	9 (23.0)
Interferon-α, n (%)	21 (53.8)	18 (46.1)
Azathioprine, n (%)	5 (12.8)	11 (28.2)
TNFi, n (%)	1 (2.5)	5 (12.8)
Anticoagulant n (%)	12*** (30.7)	7 (17.9)

*Some patients have received more than one type of immunosuppressive in maintenance therapy
** These patients were followed by rheumatology department after the surgical or endovascular intervention.
*** Patients already using anticoagulants due to venous thrombosis

Abstract THU0287 – Table 2.

Figure 2. Evaluation of response according to vascular intervention and induction treatments *

	Stabile/Regression, n=18	Progression, n=18	p
Vascular intervention			
Done, n=23 (%)	10 (43.5)	13 (56.5)	0.298
Not done, n=13 (%)	8 (61.5)	5 (38.5)	
Induction treatment			
Cyc	2	1	0.315
Cyc+IFN-α	10	5	
IFN	3	2	
Anti-TNF	0	1	
Azathioprine	2	3	
No treatment	1	5	
Only steroid	0	1	

*3 patients have no follow-up data
Cyc: Cyclophosphamide, IFN: Interferon-α

Conclusion: Arterial aneurysms in BD have poor prognosis and high mortality. Glucocorticoids, cyclophosphamide and interferon-α are the most preferred treatments for induction and maintenance therapy for aneurysm of the BD. Approximately 2/3 of the patients required a vascular intervention. Clinical and radiological progression was more prominent in the aorta and its branches. As expected, there is more progression in interventional procedures without medical treatment.

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THU0288 LONG-TERM FOLLOW-UP OF ANTI-IL6-RECEPTOR TOLICIZUMAB IN REFRACTORY UVEITIS IN PATIENTS WITH BEHÇET'S DISEASE. MULTICENTER STUDY OF 14 PATIENTS IN CLINICAL PRACTICE

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Background: Ocular involvement in Behçet's disease (BD) is a potential severe and disabling complication. Anti-TNF-α agents have shown an improvement of visual outcome in BD-related uveitis refractory to conventional immunosuppressive (IS) drugs. However, these drugs do not achieve control of intraocular inflammation in all patients or are not well tolerated. Tocilizumab (TCZ) has shown efficacy in different refractory ocular inflammatory diseases.

Objectives: To assess the efficacy of long-term therapy with TCZ in refractory uveitis associated to extraocular manifestations (EOM) due to BD.

Methods: Multicenter study of patients with BD refractory to standard systemic treatment.

Results: We followed up 14 patients (9 men/5 women) (26 affected eyes); mean age 40.8±19.5 years. Pattern of ocular involvement: panuveitis (10; 4 with retinal vasculitis), anterior (3) and posterior (1) uveitis; 8 recurrent and 6 chronic; 9 with cystoid macular edema. At TCZ onset the following EOM were present: oral and/or genital ulcers (10), arthritis (6), folliculitis/pseudofolliculitis (6), erythema nodosum (3), livedo reticularis (1), intestinal affection (1) and neurological involvement (3). Before TCZ, they had received corticosteroids (13 intraocular, 12 oral and 12 iv), conventional IS drugs and biologic agents: methotrexate (11), cyclosporine (8), azathioprine (10), colchicine (1), cyclophosphamide (2), mycophenolate mofetil (1), adalimumab (10), infliximab (6), golimumab (3), canakinumab (1), or etanercept (1). TCZ was used in monotherapy (7) or