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THU0325 RELATIONSHIP OF THE INITIAL SYMPTOMS TO THE DIAGNOSIS DELAY AND POOR PROGNOSIS IN PATIENTS WITH TAKAYASU ARTERITIS

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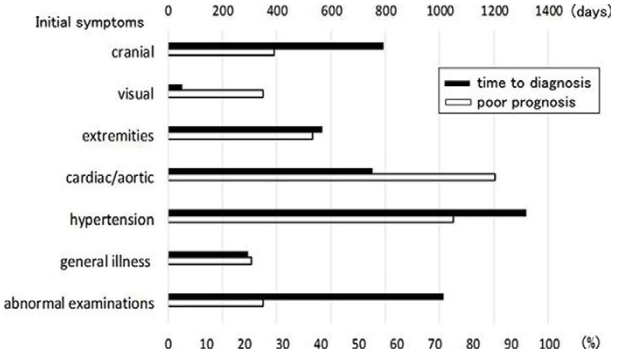
Background: Clinical courses of Takayasu arteritis are of great variety. Its non-specific initial symptoms can cause the delay in diagnosis and lead to poor prognosis. However, the symptoms of very early phase of Takayasu arteritis and their effects on diagnosis delay and prognosis are unclear.
Objectives: To clarify the characteristics of initial symptoms of Takayasu arteritis, the delay in diagnosis, and its relationship with prognosis.
Methods: The consecutive patients with Takayasu arteritis with analysable information in our hospital were enrolled. Initial symptoms, laboratory findings before diagnosis, the duration from symptom onset to diagnosis, and prognosis were investigated. Initial symptoms were divided into 7 groups; cranial symptoms (dizziness, syncope, headache, neck pain, hemi-paralysis, and jaw claudication), visual symptoms (vision loss and visual field loss), extremities symptoms (claudication of extremities, coldness of limbs, bilateral difference in blood pressure, and limb numbness), cardiac/aortic symptoms (dyspnea on exertion, palpitation, and chest compression), hypertension, general illness (fever, fatigue, body weight loss, and arthralgia), and abnormal medical examinations (heart murmur, bruit on any extremities, and abnormal chest X-ray). Poor outcome was defined as a cardiovascular surgery or death.
Results: A total of 98 patients were enrolled with the median observation period of 12.1 years (range, 1 month to 59 years). Eighty-seven (88.7%) were female and the mean age at diagnosis was 37 years. The mean duration from the initial symptoms to diagnosis was 600 days. Thirty-four (34.7%) patients had poor outcomes. The initial symptoms before diagnosis were cranial symptoms in 25%, visual symptoms in 5%, extremities symptoms in 20%, cardiac/aortic symptoms in 7%, hypertension in 9%, general illness in 26%, and abnormal medical examinations in 8%. The duration from symptom onset to diagnosis was 792, 52, 567, 752, 1318, 293 and 1014 days (p=0.10), respectively; the rate of poor outcome was 28, 25, 38, 86, 75, 22, and 25% (p=0.010), respectively. The duration from symptom onset to diagnosis was longer in the patients with poor prognosis than those without (837 vs 493 days, p=0.06). The patients with extremities symptoms were younger (29.6 vs 38.9 years, p=0.026) than those with the other symptoms, and patients with cardiac/aortic symptoms were older (49.7 vs 36.3 years, p=0.044). The patients without general illness showed lower levels of C-reactive protein (3.26 vs 5.86 mg/dl, p=0.048), erythrocyte sedimentation rate (38.3 vs 66.6 mm/h, p=0.01), and platelet counts ($29.4*10^4$ vs $34.0*10^4/\mu l$, p=0.09), and resulted in poor outcome more frequently than those with (42.6 vs 22.6%, p=0.058).



Conclusions: The initial symptoms of Takayasu arteritis before diagnosis varied widely, and majority of them were non-specific. Lacking inflammatory signs were related with delayed diagnosis and poor prognosis.
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THU0326 SHORT AND LONG-TERM FOLLOW-UP WITH ADALIMUMAD IN REFRACTORY UVEITIS ASSOCIATED TO BEHÇET'S DISEASE. MULTICENTER STUDY OF 74 PATIENTS

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Objectives: To evaluate the efficacy of adalimumab (ADA) in short and long term follow-up in refractory uveitis of Behçet's disease (BD)
Methods: Multicenter study. Ocular inflammation was evaluated according to "SUN working Group" (Am J Ophthalmol 2005;140:509–516), and the macular thickening with OCT. A comparison was carried out between baseline, and follow-up visits. Results are expressed as mean±SD or median [IQR]. Continuous variables were compared with Wilcoxon test.
Results: We studied 74 patients/132 affected eyes (39M/35W); mean age 38.7±11.3. The ocular pattern was panuveitis (n=45), posterior uveitis (n=14), anterior uveitis (n=14) and intermediate uveitis (n=1). Before ADA, systemic treatment with corticosteroids, iv metilprednisolone (n=23), Cyclosporin A (58), azathioprine (33), metotrexate (31) and other drugs (28) was used. The dose of ADA was 40 mg/2 weeks/ sc in monotherapy (n=22) or combined (n=52). Most patients showed a rapid and progressive improvement (TABLE). The 24 patients (37 affected eyes) with CME showed a significant improvement.
ADA was optimized in 23 (31.1%) that were in remission for 15.3±9 months. Interval of administration was increased to 3 (n=6), 4 (13), 5 (1), 6 (1) and 8 weeks. After a mean follow-up of 13.0±9.7 months after optimization, 21 patients were stable and 2 had a severe flare. In 4 patients ADA was stopped after 35.2±9.3 months in remission. The main adverse effects observed were lymphoma (n=1), pneumonia (1), and 2° bacteriemia by E. Coli (1)

TABLE. EVOLUTION OF OCULAR PARAMETERS

	Basal	2nd week	1st month	6th month	1st year	2nd year	3rd year
VA (mean±SD)	0.5±03 n=146	0.6±0.3* n=144	0.6±0.2* n=143	0.7±0.2* n=120	0.8±0.2* n=112	0.8±0.2* n=89	0.8±0.2* n=41
Cells in the anterior chamber (median [IQR])	1 [0-2] n = 144	0 [0-2]* n= 142	0 [0-1]* n=142	0 [0-0]* n= 120	0 [0-0]* n = 116	0 [0-0]* n = 92	0 [0-0]* n = 42
Vitritis (median [IQR])	1 [0-2] n= 145	1 [0-1]* n= 143	0 [0-1]* n= 143	0 [0-0]* n= 124	0 [0-0]* n= 116	0 [0-0]* n= 92	0 [0-0]* n= 42
Retinal vasculitis (% affected eyes)	53.3% n= 144	43.2%** n= 141	36.4%* n= 142	11.4%* n= 123	4.1%* n= 116	0.6%* n= 92	0.6%* n= 42
OCT (μ) (mean±SD)	345.5±134.5 n= 82	341.1±108.6* n= 65	309.8±8* n= 64	268.1±38.4* n= 52	254.4±38.5* n= 55	259.5±41.2* n= 42	254.5±48.9* n= 20

*p<0.05
Abbrev: VA= visual acuity

Conclusions: ADA was effective in short and long-term follow-up in refractory uveitis associated to BD. Optimization or even suspension of ADA is possible.
Disclosure of Interest: None declared
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THU0327 ASSOCIATION BETWEEN THE TC/HDL RATIO AND DISEASE ACTIVITY IN PATIENTS WITH TAKAYASU ARTERITIS

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Background: Accelerated atherosclerosis has become the main cause of morbidity in patients with autoimmune diseases such as RA and SLE [1]. The Cholesterol/High-density Lipoprotein Cholesterol (TC/HDL-C) ratio is a high discriminatory power index for coronary heart disease. A high TC/HDL-C ratio has been intensively used as a predictor of CVDs [2]. EULAR Task Force recommended that the TC/HDL-C ratio should be regarded as an important prognostic indicator for future cardiovascular disease (CVD) in patients with rheumatoid arthritis (RA) [3]. However, the relationship between the TC/HDL-C ratio and disease activity of Takayasu arteritis (TAK) is unclear.
Objectives: To investigate changes in the TC/HDL-C ratio and to evaluate the relationship between the TC/HDL-C ratio and disease activity of TAK.
Methods: A retrospective study of 103 patients with TAK and 73 healthy controls was performed. We compared the triglyceride (TG), TC, low-density lipoprotein cholesterol (LDL-C), HDL-C and TC/HDL-C ratio between patients and healthy controls, and we analyzed correlations between the lipid parameters and indexes

of TAK activity. A ROC curve was used to determine the predictive value of TC/HDL-C ratio in patients group.

Results: The TG level was higher in patients with TAK than in the controls (p=0.000). The TC, LDL-C, and HDL-C levels were significantly lower in patients with TAK than in the controls (p=0.000, p=0.000, and p=0.000, respectively). The HDL-C level was significantly lower in the active TAK group than in the inactive TAK group (p=0.005). The TC/HDL-C ratio was significantly increased in patients with disease activity (p=0.001), and it exhibited a positive relationship with the high-sensitivity C-reactive protein level (r=0.234, p=0.003) and Kerr score (r=0.219, p=0.031). In addition, the TC/HDL-C ratio of 3.698 was determined as a predictive cut-off value of TAK (sensitivity 61.0%, specificity 78.6%, area under the curve=0.743) (Figure 1).

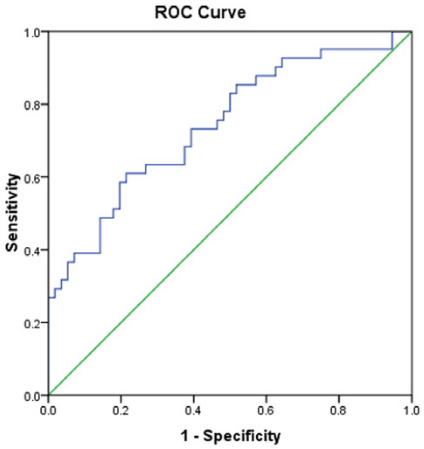

Figure 1.Receiver-operating characteristic curve for the TC/HDL-C ratio.
The area under the curve was 0.743, with sensitivity and specificity of 61.0%and 78.6%

Conclusions: For the first time, we showed that serum levels of TC, LDL-C, and HDL-C were significantly lower in patients with TAK, and the TC/HDL-C ratio has a positive relationship with the disease activity of TAK, suggesting that the TC/HDL-C ratio may be a potential indicator for monitoring the disease activity of patients with TAK.

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THU0328 PREDICTORS OF SEVERE DAMAGE IN ANCA-ASSOCIATED VASCULITIS: DATA FROM A MONOCENTRIC INCEPTION COHORT

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Background: Granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA) are multi-systemic diseases associated with anti-neutrophil cytoplasmic antibodies (ANCA). Patients develop severe and irreversible damage, even in early stages of the disease, but data about prognostic factors are limited.

Objectives: To assess items and predictors of severe damage in a monocentric cohort of ANCA-associated vasculitis (AAV) patients, classified as GPA or MPA.

Methods: Clinical and serological data of patients, followed-up in a daily practice setting, were retrospectively revised. Severe damage, defined as a Vasculitis Damage Index (VDI) ^{3} 5, was assessed after 12 months and at last examination (LE). The five most important items, predictors of severe damage and their increase rate were analysed at the beginning and at the last examination.

Results: 69 patients were included, 53 (76.8%) classified as GPA and 16 (23.2%) as MPA. According to EUVAS classification 7 (10.1%) were affected by limited disease, 14 (20.3%) by early systemic disease, 35 (50.7%) by generalized disease and 13 (18.9%) by severe disease. Patients were middle aged at diagnosis (54.6±15 years) with a diagnostic latency of 9.6±16.4 months, mostly caucasian (98.6%) and equally represented in sex (female 53.6%). Mean follow-up was 54.5±41.9 months. The number of flare/year at LE was significantly higher in anti-PR3 patients if compared to anti-MPO (1.0±1.1 vs 0.3±0.5, p=0.028).

The VDI items most frequently observed are listed in Figure 1.

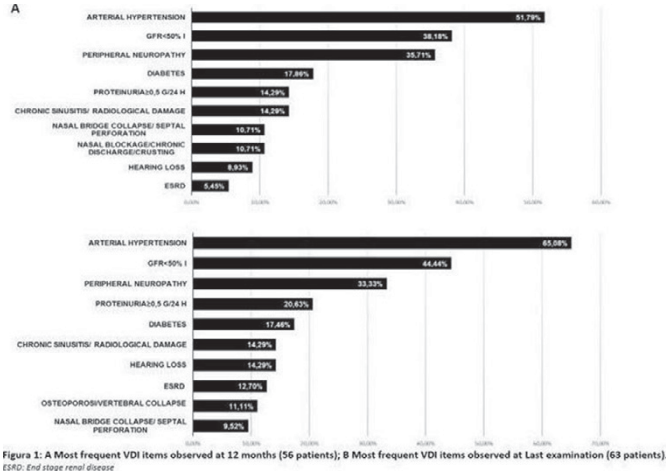

Figura 1: A Most frequent VDI items observed at 12 months (56 patients); B Most frequent VDI items observed at Last examination (63 patients).
ESRD: End stage renal disease

We measured their frequencies and increase rate from diagnosis to the LE: arterial hypertension increased from 3 (4.7%) to 41 (65.1%) (+60.3% (IC 95% 48–71%), p<0.0001), GFR<50% from 1 patient (1.6%) to 28 (44.4%) (+42.8 (IC 95: 31–55), p<0.0001), neuropathy from 4 (6.3%) to 22 (34.9%) (+28.6% (IC 18–41), p<0.0001), diabetes from 0 (0%) to 11 (17.5%) (+17.5% (IC 10–29); p=0.0011) and proteinuria>0.5 g 24 h from 1 (1.6%) to 13 (20.6%) (+19% (IC 11–31); p=0.0006).

Severe damage, defined as VDI≥5, increased over time, being present in none patient at diagnosis and in 14 (25.9%) at last examination.

At univariate analysis, number of flares/year (p=0.012), BVASv3 at diagnosis (p=0.03), steroid pulses therapy (p=0.04) and steroid cumulative dose (p=0.03) were identified as predictive factors of severe damage in the first year. Conversely, creatinine at LE (p=0.04), numbers of hospitalizations (p=0.009) and steroid cumulative dose at LE (p=0.47) were found to be predictors of long-term severe damage. Interestingly, ANCA pattern did not influence damage severity and only slightly differences in VDI items was noted between anti-PR3 and anti-MPO positive patients.

Conclusions: Our results highlights that knowing severe damage predictors is key in short and long term treat to target patient's management.

Disclosure of Interest: None declared
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THU0329 BUDD-CHIARI SYNDROME IN BEHÇET'S DISEASE: A RETROSPECTIVE MULTICENTER STUDY

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Background: The aim of this study was to determine the demographic, clinical,