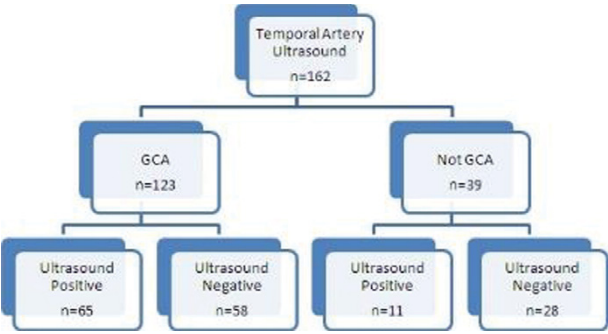


had a sensitivity of 48.8% (95% CI 39.7, 57.9) and specificity of 97.4% (95% CI 84.9, 99.9). A hypothetical sequential strategy of US followed by biopsy in the case of negative US had a sensitivity of 78.9% (95% CI 70.1, 85.5) and specificity of 71.8% (95% CI 54.9, 84.5), equivalent to a simultaneous testing strategy. Time on glucocorticoids did not significantly impact the results of US or biopsy. The only factor independently predictive of a positive US was male sex (OR 5.53, 95% CI 2.72 to 11.22, $p<0.001$). The only factor independently predictive of a positive biopsy was jaw claudication (OR 2.40, 95% CI 1.11, 5.21, $p=0.027$).

Table 1. Performance Characteristics of Ultrasound, Biopsy, and Combination Strategies

Strategy	Sensitivity	Specificity	PPV	NPV	PLR	NLR
Ultrasound alone	52.8	71.8	85.6	32.6	1.87	0.66
Biopsy alone	48.8	97.4	98.0	37.6	19.02	0.53
Sequential (Biopsy only if US positive)	22.8	97.4	96.6	28.6	8.88	0.79
Sequential (Biopsy only if ultrasound negative)	78.9	71.8	89.8	51.9	2.80	0.29
Simultaneous	78.9	71.8	89.8	51.9	2.80	0.29
Procedure, glucocorticoid duration						
Ultrasound, all, n=162	52.8	71.8	85.6	32.6	1.87	0.66
Ultrasound, 0 days, n=80	51.9	76.9	82.4	43.5	2.25	0.63
Ultrasound, >0 ≤3 days, n=27	46.2	100	100	6.0	Infinity	0.54
Ultrasound, >3 ≤7 days, n=25	60.0	40.0	80.0	20.0	1.00	1.00
Ultrasound, >7 ≤14 days, n=18	64.3	50.0	81.8	28.6	1.29	0.71
Ultrasound, >14 days, n=12	44.4	100	100	37.5	Infinity	0.56
Biopsy, all, n=162	48.8	97.4	98.0	37.6	19.02	0.53
Biopsy, 0 days, n=53	46.9	100	100	55.3	Infinity	0.53
Biopsy, >0 ≤3 days, n=24	66.7	83.3	92.3	45.5	4.00	0.40
Biopsy, >3 ≤7 days, n=38	47.1	100	100	18.2	Infinity	0.53
Biopsy, >7 ≤14 days, n=29	47.8	100	100	33.3	Infinity	0.52
Biopsy, >14 days, n=18	37.5	100	100	17.0	Infinity	0.63



Conclusions: TA US is a useful tool in the diagnosis of GCA; however false positive tests occur in 7% and thorough clinical assessment remains crucial. Prior glucocorticoid treatment has no clear impact on results.

Disclosure of Interest: None declared
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THU0313 INFERIOR AND SUPERIOR VENA CAVA THROMBOSIS IN BEHÇET'S DISEASE. MOROCCAN EXPERIENCE

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Background: Behçet's disease (BD) is a systemic disorder with a vascular tropism where the vessels might be affected. Venous thrombosis is the most common vascular complication. Among its locations, vena cava thrombosis (VCT) are rare but can be life-threatening.

The purpose of this work is to specify the frequency of VCT in the course of Behçet's disease in morocco and to analyse epidemiological, clinical and therapeutic modalities.

Objectives: This retrospective study was conducted in the internal medicine department of the University Hospital IbnRochd of Casablanca, over a period of thirty-five years between 1980 and 2016.

Methods: Where included 1618 cases of Behçet's disease, all diagnosed in our service and meeting the diagnosis criteria as defined by the international study group (ISG) for Behçet's disease.

Results: 52 patients with VCT – 47 men and 4 women - were gathered during this period, representing a rate of 3.30% of 1572 cases, with a mean age of 35 years (ranges 17–54).

The diagnosis of VCT and BD was concomitant in 10 cases and occurred during the course of the disease in 42 cases after an average of 6 years of evolution.

Among the localization. Inferior VCT is the most frequent localization (32 patients, including 15 cases of Budd- Chiari syndrome). The superior VCT was reported in 24 patients, whereas 5 patients had both a superior and inferior VCT.Vena cava thrombosis was associated with deep venous thrombosis (DVT) of the lower limbs in 24 cases and with an aneurysm in 7 cases (pulmonary artery: 4 cases, femoral artery: 2 cases, abdominal aorta; 1 case).

Regarding the therapeutic modalities, 41 patients were under anti-coagulant treatment, 45 patients received corticosteroids and 51 of the patients were under immunosuppressive agents (cyclophosphamide,azathioprine).

The evolution was significantly improved after the introduction of the immunosuppressive therapy, which were firstly initiated in 2006.

Conclusions: Vena cava thrombosis in the context of Behçet's disease is a very serious pathology threatening the patient's vital and functional prognosis. Preventive measures, early diagnosis and effective treatment are the keys to a successful management of such complications' risks.

Disclosure of Interest: None declared
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THU0314 RADIOLOGIC ACTIVITY IS THE MAJOR DETERMINANT FOR PHYSICIANS WHILE DECIDING ACTIVE DISEASE IN TAKAYASU ARTERITIS

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Background: There are no valid follow-up parameters in the assessment of disease activity in Takayasu arteritis (TA).

Objectives: We investigated the impact of incorporation of vascular imaging into ITAS in the assessment of disease activity in TA.

Methods: 52 patients who fulfilled the ACR criteria were included in the study. PGA, Kerr et al.'s criteria and ITAS2010/ITAS-A scores were evaluated in all patients in serial visits. All the patients were followed using 3–6 monthly B-mode/Doppler ultrasonography (USG) and 6–12 monthly magnetic resonance angiography (MRA).

Radiological activity (Rad) was defined based on the presence of any of the 3 parameters including new vessel involvement by any technique (5 points),increase in vessel wall thickness on USG (3 points) and vessel wall edema on MRA (3 points).Then we incorporated these scores with ITAS-A to obtain a composite disease activity index (ITAS2010-A-Rad) (Table 1). Active disease was defined as ITAS-A-Rad >4 points.

Results: Total 410 visits of 52 TA patients (mean age 50.7 yrs, F: 92.3%, mean follow-up duration:6.4±2.9 yrs) were evaluated. Radiological assessment was done in 359 visits (by USG in 271 and by MRA in 190). Patients were categorized as having active disease in 194 visits (47.4%) according to PGA and 72 visits (17.5%) according to Kerr et al. criteria.The agreement between them was fair (66%, κ : 0.29). Radiological activity was determined in 105 out of 359 visits (29.2%). The total agreement between radiological activity and Kerr et al. criteria was 83% (κ : 0.58). It was found to be 76% (κ : 0.52) between radiological activity and PGA. Mean ITAS-A-Rad scores were found to be significantly higher in visits with active disease compared to visits with inactive disease according to both PGA and Kerr et al. criteria (Table 2). The ITAS-A-Rad was significantly correlated with all the other activity parameters including ITAS2010, ITAS-A, and APRs.

There were 43 visits with new vessel involvement by any radiologic technique; all visits included patients with active disease based on both PGA and Kerr et al. criteria. Whereas in 50% of these visits, patients had normal CRP, and %49 had normal ESR.

The agreement between ITAS2010 and PGA was fair (69%, κ : 0.38).When APR was added (ITAS-A), it did not improve (68%, κ : 0.34). But the agreement between

Table 1. The definition of ITAS-A-Rad Score

Clinical	ITAS2010		0–
Laboratory	APR	ESR	0 for ESR<20 1 for 21–39 2 for 40–59 3 for >60 mm/h
		CRP	0 for CRP≤5 1 for 6–10 2 for 11–20 3 for >20mg/l
Radiology	Radiological activity	New vessel involvement with any radiological method	5
		B-mode Doppler USG	Progression on vessel wall thickness 3
		MRA	Presence of vessel wall edema 3
Total	ITAS-A-RAD Score		

ITAS-A-Rad Score >4 → Active.

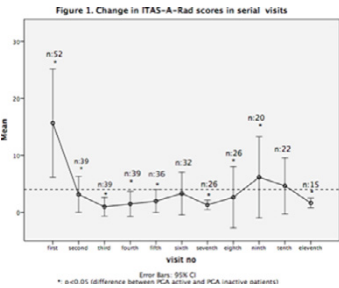


Table 2. Response to change: The mean ITAS-A-Rad scores in serial visits

Visit no	ITAS-A-Rad score (mean±SD)		P
	Active	Inactive	
1	16.72±6.3	5.25±4.9	<0.001*
2	6.23±4.6	1.92±3.8	<0.001*
3	5.12±3.3	1.40±1.88	<0.001*
4	4.11±2.47	1.60±1.72	<0.001*
5	6.36±5.6	1.36±1.7	<0.001*
6	3.20±2	2.95±39	0.29
7	4±2.3	1.76±2.7	<0.001*
8	5.3±3.8	1.3±1.77	<0.001*
9	4.33±6.02	1.21±2.3	0.02*
10	6.67±4.5	1.51±2.3	0.06
11	4±2.7	1.10±0.99	0.028*

ITAS-A-Rad and PGA (72%, κ : 0.50) and also Kerr et al. criteria (82%, κ : 0.56) was found to be moderate. Interestingly, when only USG (ITAS-A-USG) or only MRA (ITAS-A-MRA) was used, the agreement with PGA was remained unchanged (73%, κ : 0.45 and 76%, κ : 0.52, respectively).

When responsiveness to change of ITAS-A-Rad score was evaluated by serial visits of patients, it was found that the mean value of the score was discriminative for activity according to PGA in 9 of 11 visits (Figure 1).

Conclusions: The results of this study suggest that ITAS-A-Rad may be used to be a valuable follow-up parameter in the assessment of disease activity.

Disclosure of Interest: None declared

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THU0315 CENTRAL NERVOUS SYSTEM INVOLVEMENT IN PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS: A SINGLE CENTER EXPERIENCE

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Background: Peripheral nerve involvement is relatively frequently encountered in patients with granulomatosis with polyangiitis (GPA). Central nervous system (CNS) manifestations are reported to occur in about 10% of GPA patients.

Objectives: We aimed to estimate the prevalence of CNS involvement in Greek patients with GPA, describe the related clinical characteristics, and identify possible predicting factors for its occurrence. We also compared the clinical picture and long term outcomes of GPA patients with and without CNS involvement.

Methods: The medical charts of all patients with ANCA-associated and biopsy proven small vessel vasculitis (AAV), diagnosed in our hospital between 1995–2015, were retrospectively reviewed and GPA patients with CNS involvement were identified. Demographics, serological and clinical features, at the time of AAV diagnosis, of CNS involvement and during the follow-up time, were recorded. Comparisons of disease characteristics and outcomes, including patient survival, relapse rate and treatment-related adverse events, were performed between GPA patients with and without CNS involvement.

Results: 77 patients with GPA were identified in our AAV registry. Of these, nine (11.7%) developed CNS manifestations, either at clinical presentation (33.3%) or during the follow-up (66.7%). At the time of CNS involvement, all patients were characterized by increased acute phase reactants and all but one patients had vasculitic manifestations in several other organs/systems and increased titers of ANCA. CNS symptomatology included: sensor and/or sensorimotor symptomatology (33.3%), severe headache and hearing loss (33.3%), delirium and seizures (22.2%), diplopia (11.1%) and cerebellar symptoms (11.1%). Findings from MRI were: cerebral ischemic lesions (55.6%), focal dural thickening with enhancement (22.2%), orbital mass formation (11.1%) and mastoiditis causing facial nerve palsy bilaterally (11.1%). Patients with CNS involvement, compared to those without, at initial AAV diagnosis, experienced vasculitic manifestations of the ENT system more frequently (77.8% versus 25.4%, $p=0.004$) and they had a lower disease activity, as assessed by the BVAS score, while during the overall course of the disease experienced lung vasculitis less frequently (44.4% vs. 79.4%, $p=0.02$). Comparisons between GPA patients with and without CNS manifestations did not reveal any differences in long-term outcomes including relapse rate/100 person-months, (95% CI) [1.812 (0.920–3.229) Vs 1.033, (0.757–1.378), respectively] ($p=0.171$), survival (Mantel-Cox test, $p=0.244$) and treatment-related adverse events

Conclusions: CNS involvement was recorded in 11.7% of our GPA patients, either during the initial phase or as a late disease sequela. ENT involvement and low BVAS score at disease onset were more common in GPA patients with CNS manifestations. Based on our results, CNS involvement did not affect the long term outcomes of GPA patients.

Disclosure of Interest: None declared

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THU0316 EPIDEMIOLOGY OF TAKAYASU ARTERITIS IN NORTHERN ITALY

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Background: Takayasu arteritis (TAK) is a large vessel vasculitis of unknown etiology, predominantly affecting the aorta and/or its major branches and occurring before the age of 40 years. TAK has been described worldwide with an annual incidence in Europe ranging from 0.4 to 1.3 /1.000.000. TAK is more common in female. In the largest study from Japan, the female to male ratio was eight to one. In a recent Swedish epidemiologic study no male were identified. There are no epidemiological studies regarding Takayasu arteritis in Italian population.

Objectives: To investigate the epidemiology of Takayasu arteritis in a Northern Italy area.

Methods: All patients with incident TAK diagnosed between 1997 and 2015 living in the Reggio Emilia area were identified by capture and re-capture checking at

computerized discharge diagnosis codes (ICD10) and at outpatients databases from rheumatology, internal medicine, surgery, pathology, imaging departments of Reggio Emilia Hospital and by examining the Reggio Emilia district database for rare diseases. The Reggio Emilia population is predominantly of Caucasian origin (92.5%) with a yearly increase in general population of 0.5% (from 438.588 inhabitants in 1997 to 533.827 in 2015).

Results: There were 5 women satisfying ACR 1990 criteria for TAK diagnosis during the 18 years study period. The overall age and sex-adjusted 18 years incidence per 100.000 persons aged <40 years was 2.2. (95% CI:0.71 to 5.09). The overall age- and sex-adjusted 20 years incidence per 100,000 women aged <40 years was 4.5. (95% CI:1.46 to 10.48). Median age at disease onset was 36y and at diagnosis was 39 years. The prevalence of TAK in the general population on December 31 2015 was 0.9 (95% CI: 0.31 to 2.19) while in female population aged <40 years was 4.4 (95% CI 1.47 to 10.54). All patients are still alive on December 31 2015.

Conclusions: As observed in other epidemiological studies, TAK is a rare disease also in Northern Italy with a large prevalence of female.

Disclosure of Interest: None declared

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THU0317 CARDIOVASCULAR RISK FACTORS AND COMORBID DISEASES IN TAKAYASU ARTERITIS

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Objectives: Cardiovascular complications are the main causes of morbidity and mortality in the course of Takayasu arteritis (TA). In addition to the occlusive vasculitis, hypertension and accelerated atherosclerosis are possibly other factors playing role in the etiopathogenesis of these complications. Although, strict management of traditional cardiovascular risk factors is advocated to diminish the effect of cardiovascular complications, we still do not know whether traditional risk factors and other comorbid conditions are increased or operative. In this study we looked at the frequency of traditional atherosclerotic risk factors and comorbid conditions among patients with TA and suitable diseased and healthy controls.

Methods: We studied 88 (77F, 11M) consecutive TA patients and 71 (66F, 5M) SLE patients, between May and November 2016. In addition, age and gender matched, 96 (80F, 16M) healthy controls were included. Study participants were interviewed with the help of a standardized questionnaire that assess the presence or absence of traditional atherosclerotic risk factors and several comorbid conditions according to the Charlson comorbidity index. Additionally, Framingham coronary heart disease risk score was calculated.

Results: Smoking was more frequent among the healthy controls, whereas hypertension and family history of cardiovascular diseases were more common among TA patients. Patients with SLE were found to have less hyperlipidemia. The Framingham risk scores did not differ among the groups. Pericardial/pleural and renal diseases were more frequently observed in SLE patients, whereas cardiovascular diseases and chronic lung diseases were more common in TA patients. Inflammatory bowel diseases were only observed in TA patients.

	Takayasu arteritis (n=88; 77F/11 M)	SLE (n=71; 66 F/5 M)	Healthy controls (n=96; 80 F/16 M)	P value
Age, mean \pm SD	44 \pm 12	42 \pm 12	45 \pm 11	NS
Age at disease onset, mean \pm SD	31 \pm 12	29 \pm 11	–	NS
Smoking (current and past), n (%)	27 (31)	30 (42)	47 (49)	<0.05
Hyperlipidemia, n (%)	32 (44)	15 (25)	41 (48)	0.018
Hypertension, n (%)	47 (53)	23 (32)	22 (23)	<0.01
Diabetes mellitus, n (%)	8 (9)	6 (7)	15 (16)	NS
Median [IQR] Framingham risk score	3 [1–9]	3 [1–5]	3 [2–7]	NS
Familial history of cardiovascular diseases, n (%)	41 (47)	15 (21)	22 (23)	<0.01
Cardiovascular diseases, n (%)	47 (53)	13 (18)	1 (1)	<0.01
Valvular heart disease, n (%)	34 (39)	12 (17)	7 (7)	<0.01
Chronic pulmonary diseases, n (%)	11 (13)	1 (2)	5 (6)	<0.05
Pleural/pericardial diseases, n (%)	7 (8)	14 (20)	0	<0.01
Renal diseases, n (%)	10 (11)	19 (27)	8 (8)	<0.05
Other accompanying rheumatologic diseases and amyloidosis, n (%)	10 (11)	15 (21)	1 (1)	<0.01
Inflammatory bowel diseases, n (%)	9 (11)	0	0	<0.01
Inflammatory upper/lower back pain, n (%)	21 (24)	23 (32)	6 (6)	<0.01
Family history of rheumatologic diseases, n (%)	26 (30)	25 (35)	4 (4)	<0.01

Conclusions: Traditional atherosclerotic risk factors are not increased in TA. Comorbidities in these patients are mainly due to the complications of vascular involvement. The frequency of inflammatory bowel diseases and inflammatory upper/lower back pain is substantially high and deserves further research. Moreover, the increased incidence of cardiovascular and rheumatologic diseases among the first-degree relatives of TA patients suggest that genetic mechanisms may play role in TA

Disclosure of Interest: None declared

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