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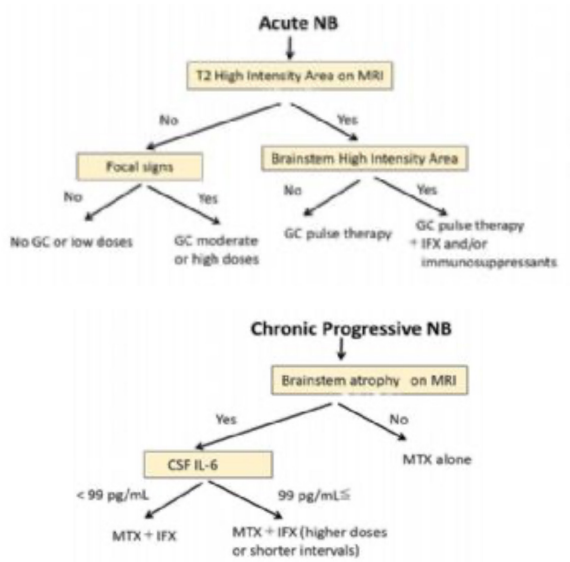
Abstracts
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O-01

DEVELOPMENT OF ALGORITHMS FOR TREATMENT OF NEURO-BEHÇET'S DISEASE

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Background/Objective. The Japanese Behçet's disease clinical guideline 2020 describes algorithm for diagnosis and treatment of neuro-Behçet's disease (NB). However, it does not provide specific policies on which patients should be treated and how. We tried to develop algorithms that would be useful in determining treatment policies for acute NB (ANB) and chronic progressive NB (CPNB) in real-life clinical practice.
Methods. Sixty-one patients with ANB and 26 patients with CPNB who met the ISG criteria for Behçet's disease were included. For ANB, treatment intensity scores (TIS) were set based on the treatment details (*e.g.* glucocorticoid [GC] dosage). A retrospective analysis was conducted to determine the relationship of various endpoints with TIS. For CPNB, provisional severity levels were set according to treatment and outcome, as Group 1 (Gr 1): no progression with MTX (methotrexate) alone, Gr 2: no progression with MTX and IFX (infliximab), Gr 3: MTX and IFX with progression. Retrospective analysis was conducted to determine the relationship of various endpoints with the provisional severity levels.
Results. In ANB, higher TIS were significantly associated with worse recovery from acute attacks, indicating the TIS reflected the severity. Moreover, the TIS were significantly correlated with the presence of focal brain symptoms and MRI T2 high-intensity areas (any brain region or brainstem). In CPNB, CSF IL-6 was significantly lower in Gr 1 than in Gr 2+ Gr 3. ROC analysis showed that a cut-off of 98.5 pg/ml of CSF IL-6 discriminated Gr 1 from Gr 2+ Gr 3 with 80% sensitivity and 70% specificity. Notably, no progression of symptoms was observed in the 3 patients without brainstem atrophy on MRI. Based on these results, new algorithms for treatment of ANB and CPNB were established (Fig. 1).
Conclusion. The new algorithms for treatment of ANB and CPNB might be useful in determining treatment strategies.
Key words: neurological involvement, severity, treatment, algorithm, magnetic resonance image



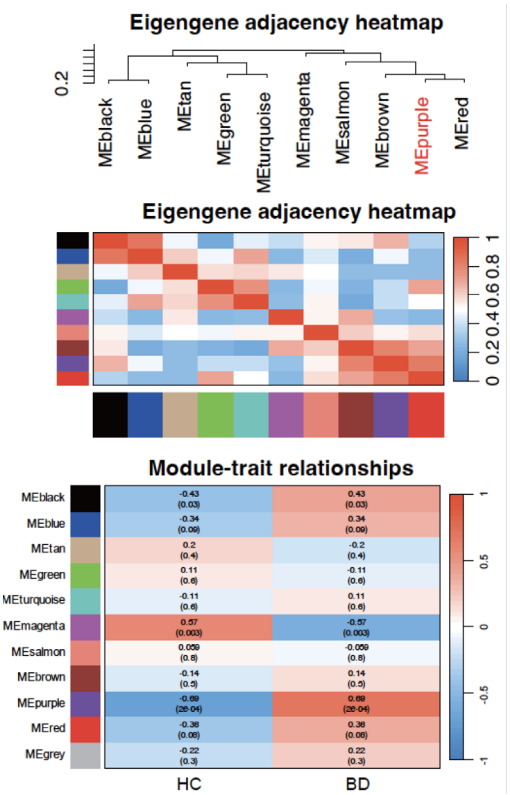
O-01 Fig. 1. Proposed algorithms for treatment of acute neuro-Behçet's disease (NB) and chronic progressive NB.

O-02

EXPLORATION OF BIOMARKERS AND IMMUNE MECHANISMS IN INTESTINAL BEHÇET'S SYNDROME BASED ON WHOLE BLOOD TRANSCRIPTOMIC FEATURES

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Background. Intestinal Behçet's Syndrome (BS) is frequently complicated by severe, refractory gastrointestinal ulcers and life-threatening events. Early identification remains a major clinical challenge due to the lack of specific symptoms and reliable molecular biomarkers. Whole-blood transcriptomics serves as a cost-effective, non-invasive 'liquid biopsy' that captures broad circulating transcriptomic signals – including those from immune cells and extracellular vesicles – offering a highly promising strategy for biomarker discovery.
Objective. To elucidate the whole-blood transcriptomic characteristics of intestinal BS, uncover underlying systemic immune regulatory mechanisms, and screen specific candidate genes for early diagnosis.
Methods. Whole-blood transcriptomic profiling was conducted comparing treatment-naive intestinal BS patients (n=15) with healthy controls (n=10). Analysis included Principal Component Analysis (PCA) and immune-related pathway enrichment. A transcription factor co-regulation network was constructed using differentially expressed transcription factors (DETFs) and the ChEA3 database. Additionally, Weighted Gene Co-expression Network Analysis (WGCNA) identified disease-associated hub genes, which were then intersected with core transcription factor pathways to isolate key candidate genes.
Results. Patients with intestinal BS exhibited distinct transcriptional profile including increased secretion of inflammatory cytokines, enhanced response of innate immune and enhanced activation of inflammation pathways. Network analyses revealed that the transcription factor PU.1 (SPI1) together with



O-02 Fig. 1. WGCNA Identifying Candidate Key Regulatory Genes in Intestinal BS
WGCNA screening of whole blood transcriptome illustrated gene set modules significantly related to intestinal BS. The intersection of PU.1 transcription factor regulatory pathway and gene set module identified Candidate Gene Regulatory Genes in intestinal BS.

downstream regulated genes plays a central regulatory role in the immune pathogenesis of the disease. Intersection of WGCNA analysis and transcription factor co-regulation network identify candidate key regulatory genes.
Conclusion. Patients with intestinal BS exhibit a distinct hyper-inflammatory whole-blood transcriptomic profile. The transcription factor PU.1 (SPI1) plays a central regulatory role in its immune pathogenesis and demonstrates high potential as specific biomarkers for intestinal BS.
Key words: Behçet's syndrome, whole-blood transcriptomics, PU.1 (SPI1)

O-03

CLINICAL, RADIOLOGICAL, AND PROGNOSTIC CHARACTERISTICS OF NEURO-BEHÇET'S SYNDROME: ANALYSIS OF 507 PATIENTS

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Objective. To determine the diagnostic value of imaging; to assess the relationship between clinical and laboratory features and disease progression; and to evaluate how these parameters influence prognosis in Neuro-Behçet's syndrome (NBS).
Background. Neurological involvement of Behçet's syndrome (BS), termed NBS, primarily presents as parenchymal NBS (p-NBS) or vascular NBS, mainly cerebral venous sinus thrombosis (CVST). Defining the clinical spectrum and prognostic determinants is essential for optimized management and outcome prediction.
Methods. Medical records of 2,640 patients followed at the Neuro-Behçet Outpatient Clinic, Istanbul University–Cerrahpaşa, between 1983 and 2024 were reviewed. Among them, 507 patients who fulfilled BS criteria and were definitively diagnosed with NBS were included. Clinical, radiological, and laboratory data were analyzed to identify prognostic factors and treatment-related outcomes.
Results. The study included 507 patients (365 males, 142 females; M/F 2.57; median age 33.2 ± 24.8 years). At presentation, 154 patients had isolated neurological symptoms, while 351 reported multiple complaints. Motor weakness and dysarthria at onset were associated with poorer outcomes, whereas isolated headache correlated with lower disability at last follow-up (last EDSS). CVST was associated with a more favourable course, while myelitis predicted higher disability and increased relapse risk. Patients with lower disability at last follow-up more often had CVST and fewer relapses. Infliximab use was associated with fewer relapses and lower last EDSS at follow-up.
Conclusion. In this large single-center cohort, initial disability, relapse pattern, and infliximab therapy emerged as key predictors of long-term outcomes. These results support the importance of early recognition and aggressive treatment in high-risk subgroups to prevent irreversible neurological disability.
Key words: Behcet, Neurobehçet, NBS

O-04

MULTICENTER VALIDATION OF THE 23-VALENT POLYSACCHARIDE PNEUMOCOCCAL VACCINE-INDUCED SKIN PATHERGY TEST IN BEHÇET'S DISEASE

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Background/Objective. The skin pathergy test (SPT) remains a useful diagnostic tool in Behçet's disease (BD); however, its declining sensitivity has limited its clinical utility. We previously demonstrated that stimulation with the 23-valent pneumococcal polysaccharide vaccine (PS-23) markedly improved sensitivity up to 80.3% in active BD (aBD), further increasing to 91.3% when the number of prick sites was doubled in replication cohort, while maintaining 100% specificity. This study aimed to validate these findings in a multicenter cohort.
Methods. This multicenter study (December 2023–March 2026) included patients evaluated for suspected BD. The cohort comprised BD, recurrent aphthous stomatitis (RAS), and other disease controls. Standard SPT (20G-SPT) was performed using a 20G needle with 3–5 mm oblique insertion. For PS23-SPT, 10 μL of PS-23(Pneumovax) was applied prior to pricking. Four pricks were performed on each forearm (two standard, two PS23-SPT). Test sites were evaluated at 48 hours; induration ³2 mm with erythema was considered positive. Patients with ³1 active manifestation were classified as aBD.
Results. A total of 894 patients were included: 259 BD (200 active, 59 remission), 269 RAS, and 366 controls. PS23-SPT showed a sensitivity of 69.5% and specificity of 100% in all BD, and 85.0% sensitivity with 100% specificity in aBD, compared to 4.2% and 5.5% sensitivity for conventional SPT. At least one pustule was observed in 132/259 (51%) of all BD and 127/200 (69.5%) of aBD; none occurred with conventional SPT. In remission, SPT positivity was 10/59 (16.9%), with pustules in 5/59 (8.5%). Strong correlations were observed between prick sites and arms (p<0.0001; r=0.70–0.85). Sensitivity was higher in treatment-naive aBD (90.5%, 154/170) *vs*. treated patients (53.3%, 16/30); pustules were also more frequent (70.6% *vs* 23.3%).
Conclusion. This multicenter study robustly validates the enhanced sensitivity of PS23-SPT, particularly in active and untreated BD, while maintaining specificity. Increasing prick sites improves diagnostic yield and reduces operator dependency. PS23-SPT is a promising adjunctive diagnostic tool, with international validation ongoing.
Key words: Behçet' disease, skin pathergy reaction, polysaccharide pneumococcal vaccine, 23-valent, multicenter study

O-05

BEHÇET DISEASE IN PEDIATRIC VERSUS ADULT PATIENTS (UPDATES FROM IRAN BD REGISTRY)

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Objective. We report here the clinical spectrum of pediatric Behçet's disease (PED-BD) and highlight the differences with adults from Iran's BD registry.
Methods. In a retrospective cohort study on 7856 patients with BD, those with both disease onset and diagnosis before 18 years (PED-BD) were com-

pared with those with disease onset after 18 years (adult-BD). Proper statistical comparisons were done, and *p*-value <0.05 considered significant.
Results. We identified 308 patients with PED-BD (3.9%). The male/female ratio was 0.96/1, and the mean age at onset was 11.2±3.7. Positive familial history for BD was present in 8.6%. As the first manifestation, oral aphthosis (OA) was the most frequent (79.2%) followed by ocular lesions in 14.3%. The prevalence rates of various manifestations were as follows: OA: 95.5%; genital ulcer: 48.7%; skin: 56.2% (pseudofolliculitis 44.5%, erythema nodosum 14.3%); ocular: 62% (anterior uveitis 51.3%, posterior uveitis 56.5%, retinal vasculitis 37.3%); articular: 34.7%; neurological: 5.8%; large vessel involvement: 6.2% (venous 5.2%, arterial 1.9%); gastrointestinal: 6.8%; epididymo-orchitis: 7.3% (boys); high ESR (^{3}20): 51.4%; abnormal urine: 14.1%; positive pathergy test: 51.2%; positive HLA-B5/51: 50.7%. ICBD criteria have the highest sensitivity for the classification of PED-BD patients in Iran.
In comparison with 6529 adult-BD patients, PED-BD patients showed lower mean disease duration (p:0.0006) and lower mean diagnosis delay (*p*:0.000007), lower male/female ratio (*p*<0.006), and higher positive familial history (*p*:018). At disease onset, lower OA (*p*<0.05) and skin lesions (*p*<0.03), and higher ocular lesions (*p*<0.004) were present in PEDBD. Among the major manifestations, PEDBD showed lower mucocutaneous lesions (*p*<0.03), but higher and more severe ocular lesions (*p*<0.004). Lower venous thrombosis (*p*<0.02) was the only minor manifestation that differed with adults.
Conclusion. The clinical spectrum of PED-BD in the Iranian cohort was somehow different with adult-onset patients with higher and more severe ocular lesions.
Key words: Behcet disease, pediatric, juvenile, clinical, Iran

O-06

CD177+ NEUTROPHILS PROMOTE ENDOTHELIAL SENESCENCE VIA HISTONE 3.1-ENRICHED NETS IN BEHÇET'S DISEASE

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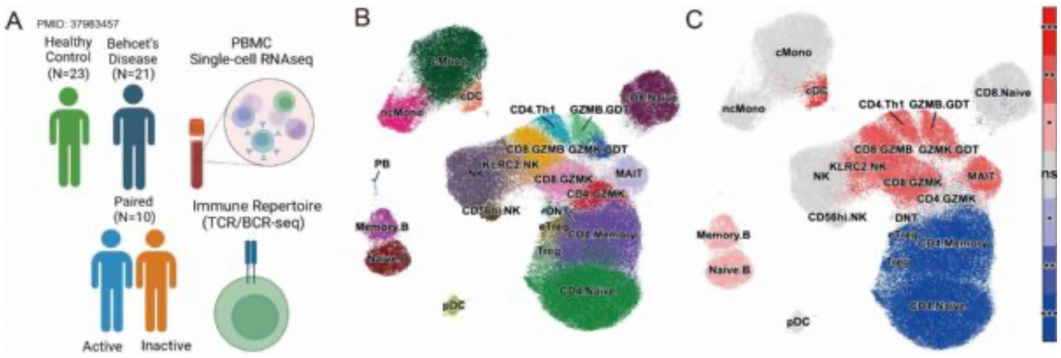
Background/Objective. To investigate CD177+PMN dysregulation on endothelial cells in Behçet's disease (BD), a chronic systemic vasculitis characterized by polymorphonuclear neutrophil (PMN) activation and endothelial dysfunction.
Methods. We reanalyzed BD-PMN RNA sequencing data and quantified CD177+PMN in BD peripheral and vascular tissue. We then analyzed BD and HC CD177+PMN using RNA sequencing, measured neutrophil extracellular traps (NETs) production, which were used to stimulate vascular endothelial cells (VECs) for RNA sequencing analysis. Differentially expressed proteins in NETs were determined by Mass spectrometry. Senescence, senescence-associated secretory phenotype (SASP) and PMN chemotaxis of VECs treated with BD CD177+PMN-derived NETs were assessed.
Results. Transcriptome analysis revealed BD-PMN overexpressed CD177 (Fig. A). CD177+PMN subset was expanded in active and severe systemic BD and was accumulated in BD aortic walls (Fig. B-C). Enhanced NETosis in BD CD177+PMN was noted in RNA sequencing, which was confirmed in both resting and activated BD CD177+PMN (Fig. D). BD CD177+PMN-derived NETs induced greater VECs activation than HC CD177+PMN-derived NETs, which were sustained after DNase I digestion (Fig. E). BD CD177+PMN-derived NETs promoted senescence, SASP and PMN chemotaxis in VECs using RNA sequencing analysis (Fig. F), which were validated by upregulated IL-6, CXCL8, ICAM-1, SA-β-gal (Fig. G-H). The level of endothelial cell senescence in the aortic wall tissue of BD patients was significantly elevated (Fig. I). Mass spectrometry identified BD CD177+PMN-derived NETs contained more Histone 3.1 (H3.1) (Fig. J). H3.1 induced senescence, SASP and PMN chemotaxis in VECs via Toll like receptor (TLR) 4 signaling, which was attenuated by senolytics (Fig. K-O).
Conclusion. Increased CD177+PMN in BD patients promoted senescence, SASP in VECs through secreting H3.1-enriched NETs, which recruit PMN and collectively contribute to vasculitis in BD.
Key words: Behçet's disease, CD177+PMN, endothelial, senescence

O-07

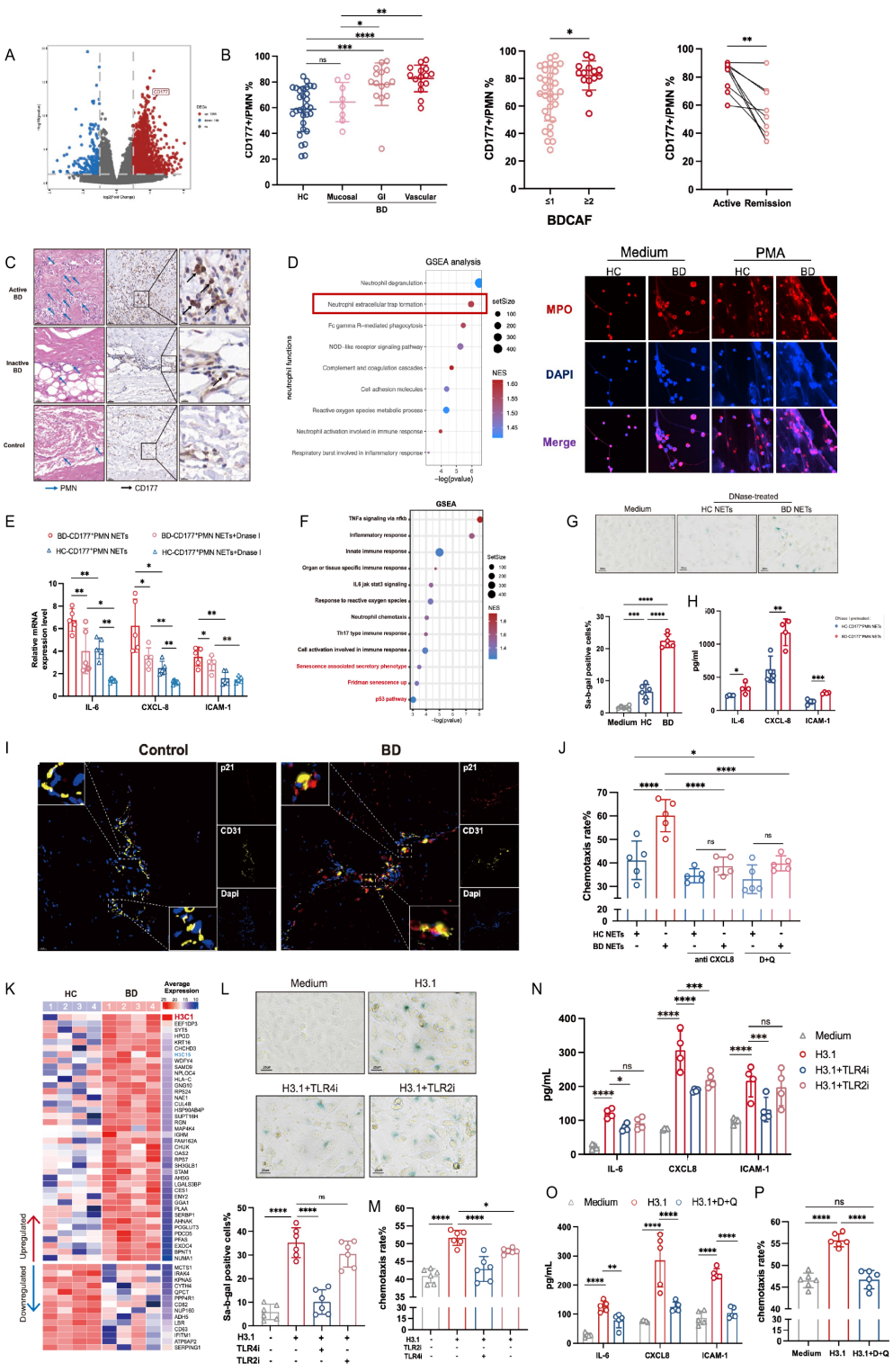
SUSTAINED CYTOTOXIC T CELL-DENDRITIC CELL CROSSTALK UNDERLIES CHRONIC INFLAMMATION IN BEHÇET'S DISEASE

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Background/Objective. Behçet's disease (BD) is a rare multisystem inflammatory disorder characterized by heterogeneous clinical manifestations, with its underlying immune cell-mediated pathogenic mechanisms remaining incompletely understood. This study aimed to elucidate the immune landscape and pathogenic cell populations driving BD using longitudinal paired samples from active and inactive disease states.
Methods. We analyzed paired peripheral blood samples from Korean BD patients during active and inactive phases. Comprehensive immune profiling was performed to assess cellular composition, cytokine expression, signaling pathway activation, and T cell receptor (TCR) clonality. Comparative analyses were conducted against healthy controls and external BD cohorts, including a Japanese dataset, to validate findings.
Results. BD patients exhibited a significant expansion of cytotoxic CD8 T cells and unconventional T cell populations, accompanied by elevated secretion of TNF and IFN-γ. Key inflammatory pathways, including IL-12, TNF,



O-07 Fig. 1. A. Schematic overview of the study design. Peripheral blood (PB) samples were collected from healthy controls (HC) and patients with Behçet's disease (BD) during active and inactive disease states. Single-cell RNA sequencing (scRNA-seq) was performed along with immune repertoire profiling (TCR/BCR sequencing).
B. Uniform Manifold Approximation and Projection (UMAP) visualization of major immune cell populations identified from PB samples, including CD4 and CD8 T cell subsets, natural killer (NK) cells, B cells, dendritic cells (DCs), and monocyte populations. Distinct clusters such as cytotoxic CD8 T cells (GZMB+, GZMK+), MAIT cells, γδ T cells (GDT), regulatory T cells (Treg), and memory/naïve subsets are annotated.
C. Differential cell-type distribution between BD and HC. Red and blue colours indicate relative enrichment or depletion of specific cell populations in BD compared to HC, respectively, with statistical significance denoted (*p*<0.05, *p*<0.01, *p*<0.001; ns, not significant). Notably, cytotoxic CD8 T cells, unconventional T cells, and dendritic cells are expanded, whereas CD4 T cell populations are reduced in BD.



O-06 Fig. 1. A. Differentially expressed genes in BD-PMN (GSE205867).
B. CD177+PMN levels in HC, active BD, BD subtypes, and BD remission.
C. Representative IHC of CD177 in aortic wall lesions (BD *vs*. control).
D. GSEA pathway analysis of sorted CD177+PMN in BD and Representative IF of NETosis in HC and BD CD177+PMN with or without PMA.
E. VECs stimulated with BD or HC CD177+ PMN-derived NETs, with or without DNase I pretreatment.
F. GSEA of VECs stimulated with BD CD177+ PMN-derived NETs (DNase I pretreated).
G-H. SA-ß-gal staining and IL-6, CXCL8, ICAM-1 levels in NET-stimulated VECs.
I. Representative IF of p21 (red) and CD31 (yellow) in aortic wall lesions (BD *vs*. control).
J. Mass spectrometry analysis of NETs from CD177-PMN.
K-M. SA-ß-gal staining, neutrophil chemotaxis, and IL-6, CXCL8, ICAM-1 in H3.1-stimulated VECs, with or without TLR2/4 inhibitor.
N-O. IL-6, CXCL8, ICAM-1 in VECs treated with H3.1 and D+Q, and chemotaxis of neutrophils toward the supernatant.
**p*<0.05, * *p*<0.01, ****p*<0.001

VEGF, and TGF-β signaling, were upregulated, while CD4 T cell proportions were reduced. Clonal expansion of CD8 effector T cells was markedly increased compared to healthy controls, with persistent clones maintained longitudinally for periods ranging from 4 months to 5 years, displaying sustained cytotoxic profiles. Dendritic cells were also expanded and exhibited enhanced secretion of VEGFA and IL-18, along with increased engagement in TNF, TGF-β, and IFN-γ-mediated interaction networks. These immunological features were distinct compared to related diseases and were consistently replicated in an independent Japanese BD cohort.

Conclusion. These findings demonstrate that BD pathogenesis is driven by persistent cytotoxic immune activation, characterized by long-lived clonally expanded CD8 T cells and activated dendritic cells that sustain inflammatory signaling over time. Cross-cohort validation highlights the robustness and translational relevance of these immune signatures, providing potential targets for therapeutic intervention.

Key words: Behcet's disease, cytotoxic CD8 T cells, clonal expansion, dendritic cells, persistent immune activation

O-08

VΔ2 T-CELL-DRIVEN B-CELL DYSREGULATION AND AUTOANTIBODY PRODUCTION IN BEHÇET'S SYNDROME

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Background/Objective. The pathogenesis of Behçet's syndrome (BS) is complex and associated with dysregulated immune responses. B-cell dysregulation contributes significantly to BS, yet the mechanisms behind this remain incompletely defined. We previously showed that Vδ2-T cells from BS patients adopt a Tfh-like functional profile and enhance B-cell differentiation into plasma cells. To further our understanding, we present detailed B-cell immunophenotyping and functional assays in in patients with BS presenting with either mucocutaneous or ocular manifestations.
Methods. PBMCs from 61 patients and 42 healthy controls were analysed *ex vivo* and following 5-day HMBPP-stimulation. A 22-colour spectral flow cytometry panel was used for high-dimensional B-cell immunophenotyping. For Vδ2 cells, cytokine production assay was performed in both *ex vivo* and after HMBPP stimulation (6, 12, 24 hours). Anti-HSP60 autoantibodies were quantified in serum and in long-term PBMC co-culture supernatants.
Results. Vδ2 T cells from BS patients showed expanded CXCR5+PD-1+ Tfh-like subsets, accompanied by higher early cytokine production (IFN-γ, TNF-α, IL-10, IL-21) compared with healthy controls. *Ex vivo*, BS B cells displayed reduced total B-cell and plasma-cell numbers but heightened activation signatures (IRF4hiPAX5lo, elevated pre-switched and IgM memory cells). After HMBPP coculture, BS samples showed amplified plasma-cell differentiation, increased double negative B cells, and stronger induction of BAFF-R, BCMA, IL-21R, IL-4Rα, CXCR4, BLIMP-1, and HLA-DR, revealing a dysregulated and more rapidly differentiating B-cell program. Finally, BS patients exhibited higher anti-HSP60 autoantibody levels in serum and markedly increased autoantibody production in HMBPP-stimulated cocultures, particularly in ocular disease.
Conclusion. Together, these findings indicate that both Vδ2 T cells and B cells in BS carry intrinsic phenotypic abnormalities, and their interaction diverges fundamentally from healthy controls, promoting pathogenic B-cell activation and autoantibody production.
Key words: Behçet's syndrome, Vδ2 T cells and B cell interaction, plasma cells, autoantibody production, heat shock protein 60

O-09

CLINICAL EFFICACY OF WEEKLY ADALIMUMAB IN BEHÇET UVEITIS: A RETROSPECTIVE ANALYSIS

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Background/Objective. Uveitis is an ocular manifestation of Behçet's syndrome and may remain active despite standard immunosuppressive therapies and, in some patients, despite every-other-week adalimumab. Evidence for weekly adalimumab escalation in this setting is limited. This retrospective study aimed to evaluate the efficacy of weekly adalimumab in patients with Behçet's uveitis.
Methods. Patients with Behçet's uveitis followed at the Department of Ophthalmology, IUC-Cerrahpaşa Medical Faculty, and initiated on weekly adalimumab between January 2022 and December 2025 were included. Medical records were retrospectively reviewed for sociodemographic characteristics, medical treatments used during follow-up, reasons for switching to weekly adalimumab therapy, subsequent dose reduction and treatment modification status, and visual outcomes.
Results. Of the 44 patients, 23 (52.3%) were male and 21 (47.7%) were female. The mean age was 39.0 ± 10.4 years (range, 22–63 years). The mean follow-up duration from uveitis diagnosis to the last visit was 6.5 ± 3.3 years, and the mean duration from escalation to weekly adalimumab to last follow-up was 2.44 ± 1.49 years. Prior to adalimumab, azathioprine was the most commonly used non-biologic agent (34/44, 77.3%). The most frequent reason for escalation to weekly adalimumab was diffuse vascular leakage and optic disc staining on fundus fluorescein angiography (34/44, 77.3%). Fourteen patients (31.8%) remained on weekly adalimumab, 17 (38.6%) were de-escalated due to clinical remission, and 13 (29.5%) required switching to another biologic agent due to refractory disease; infliximab was the most common alternative treatment (9/13, 69.2%). Among eyes with available paired measurements, visual acuity was generally stable or improved at both 6 and 12 months after escalation to weekly adalimumab. No adalimumab-related adverse events occurred.
Conclusion. Weekly adalimumab may represent a treatment option for selected patients with Behçet's uveitis; however, treatment switching may still be required, and larger real-world studies are needed to better define its role.
Key words: Behçet's disease, ocular involvement, adalimumab.

O-10

FIRST-LINE ANTI-TNF TREATMENT IN BEHÇET'S UVEITIS

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Background/Objective. To evaluate the long-term functional and anatomical outcomes of anti-tumour necrosis factor (anti-TNF) therapy initiated as first-line treatment in patients presenting with an acute attack of Behçet's uveitis.
Methods. In this retrospective study, medical records of patients presenting with an acute Behçet's uveitis attack over the past 10 years and initiated on anti-TNF therapy in combination with azathioprine as first-line treatment were reviewed. Best-corrected visual acuity (BCVA), central macular thickness (CMT) measured by optical coherence tomography (OCT), presence of macular oedema, and systemic corticosteroid use were evaluated at baseline and during follow-up visits at months 3, 6, 12, and 24, as well as at the final examination. In addition, fluorescein angiography (FA) scores were calculated to quantify retinal vascular leakage.
Results. A total of 50 patients (92 eyes) were included. The mean age was 28.7±8.7 years and 36 patients (72%) were male. Mean BCVA improved significantly from 0.51±0.65 logMAR at baseline to 0.16±0.41 logMAR at the final evaluation (p<0.001). Mean CMT decreased from 350.9±119.3 μm at baseline to 260.7±42.8 μm at the final visit (p<0.001). The prevalence of macularoedema declined markedly from 66/92 eyes (71.7%) at baseline to 7/92 eyes (7.6%) at the final evaluation (p<0.001). Similarly, the mean FA score decreased significantly from 14.79±5.61 to 5.79±5.43 (p<0.001). At baseline, 35 of 50 patients (70%) required systemic corticosteroids. This rate decreased to 4% at the final evaluation (p<0.001).
Conclusion. Initiation of anti-TNF therapy as first-line treatment in patients with Behçet's uveitis provides rapid and sustained control of ocular inflammation, resulting in significant improvements in visual acuity and macular structure. Early biologic therapy was associated with a marked reduction in angiographic inflammatory activity, substantial resolution of macular edema, and a pronounced steroid-sparing effect, supporting the early use of anti-TNF agents in the management of Behçet's uveitis.
Key words: anti-TNF therapy, Behçet's disease, uveitis

O-10 Table I. Changes in clinical and anatomical parameters during follow-up

Baseline	Month 3	Month 6	Month 12	Month 24	Final	*p*
0.51 ± 0.65	0.25 ± 0.43	0.20 ± 0.40	0.18 ± 0.42	0.13 ± 0.34	0.16 ± 0.40	<0.001
350.8 ± 119.3	302.6 ± 84.4	280.5 ± 55.9	269.2 ± 38.5	261.6 ± 35.0	260.6 ± 42.8	<0.001
1.04 ± 1.18	0.07 ± 0.24	0.03 ± 0.16	0.08 ± 0.34	0.05 ± 0.30	0.00	<0.001
66 (71.7)	38 (41.3)	22 (23.9)	7 (7.6)	5 (5.4)	7 (7.6)	<0.001
35 (70)	8 (16)	6 (12)	2 (4)	2 (4)	2 (4)	<0.001

logMAR: logarithm of the minimum angle of resolution

O-11

SEVERE VISUAL IMPAIRMENT IN BEHÇET SYNDROME IN TIME: A COMPARISON OF FOUR TIME PERIODS

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Background/Objective. The prognosis of Behçet syndrome (BS) uveitis seems to have improved considerably over time. We wanted to better quantitate this impression and tabulate the prognosis of Behçet syndrome uveitis in the previous four decades in a dedicated BS outpatient clinic by utilizing a severe and irreversible visual acuity impairment definition as the main outcome measure.
Methods. Medical records of 9289 patients with BS registered between 1976 and 2019 were reviewed for uveitis and severe visual impairment (SVI). SVI was defined as irreversible visual acuity ≤20/200 for ≥3 months. After excluding 527 patients with inadequate data, 8762 were included. Patients were grouped by registration period (<1990, 1990–1999, 2000–2009, 2010–2019). Data were collected using a standardized form. Men and women were analyzed separately.
Results. Overall, 4409/8762(50.3%) patients had uveitis, and 776/4409(17.6%) developed SVI. The frequency of SVI decreased over time. In males, the rate of bilateral SVI at presentation declined from 9.5% among patients registered before 1990 to 3.8% among those registered between 2010-2019.Through the same time periods, progression from unilateral to bilateral SVI decreased from 6.3% to 0.8%, while new-onset bilateral SVI in those without baseline SVI declined from 5.1% to 0.2%. The reduction was more pronounced in patients without baseline SVI. Progression was higher in the pre-2000 than in the post-2000 period (32% *vs*. 16%, *p*=0.002). In females, bilateral SVI decreased from 3.8% to 0.5%, with lower rates than in males. Treatment patterns evolved, with increased use of advanced therapies, including interferons and anti-TNF agents, in recent decades.
Conclusions. These data emphasize the strikingly different disease course between males and females and confirm that BS uveitis can now be managed more effectively. The SVI definition likely reflects improved medical management rather than a milder natural disease course, supported by the greater reduction in bilateral SVI among patients without baseline SVI. Formal drug trials should be sex-stratified or adequately powered for subgroup analyses.
Key words: Behçet syndrome, uveitis, prognosis

O-11 Table I. Visual outcomes in male and female patients with Behçet uveitis.

Male patients

	GROUP 1 (1972-1989)	GROUP 2 (1990- 1999)	GROUP 3 (2000-2009)	GROUP 4 (2010-2019)
Number of BS patients	778	1518	1830	1152
Patients with uveitis, n/N (%)	524/778 (67.4)	819/1518 (54)	970/1830 (53)	630/1152 (54.7)
Number of patients without uveitis at registration	3/524	3/819	1/970	0/630
Patients with bilateral SVI at registration n/N (%)	50/524 (9.5)	59/819 (7.2)	43/970 (4.4)	24/630 (3.8)
Patients with initial unilateral SVI who developed bilateral SVI during follow up, n/N (%)	30/474 (6.3)	34/760 (4.5)	14/927 (1.5)	5/606 (0.8)
*at first year, n/N (%)**	*17/467 (3.6)*	*20/752 (2.7)*	*9/921 (1)*	*3/603(0.5)*
*at third year, n/N (%)***	*25/450 (5.6)*	*26/739 (3.5)*	*13/907 (1.4)*	*3/595 (0.5)*
Patients without SVI in either eye who developed bilateral SVI during follow-up, n/N (%)	24/474 (5.1)	18/760 (2.4)	6/927 (0.6)	1/606 (0.2)
*at first year, n/N (%)**	*10/467 (2.1)*	*8/752 (1.1)*	*2/921 (0.2)*	*1/603 (0.2)*
*at third year, n/N (%)***	*15/450 (3.3)*	*13/739 (1.8)*	*2/907 (0.2)*	*1/595 (0.2)*

Female patients

	GROUP 1 (1972-1989)	GROUP 2 (1990- 1999)	GROUP 3 (2000-2009)	GROUP 4 (2010-2019)
Number of BS patients	416	899	1244	925
Patients with uveitis, n/N (%)	238/416 (57)	327/899 (36)	537/1244 (43)	364/925 (39)
Number of patients without uveitis at registration	3/238	3/327	1/537	0/364
Patients with bilateral SVI at registration n/N (%)	9/238 (3.8)	11/327 (3.4)	10/537 (1.9)	2/364 (0.5)
Patients with initial unilateral SVI who developed bilateral SVI during follow up, n/N (%)	1/229 (0.4)	4/316 (1.3)	8/527 (1.5)	3/362 (0.8)
*at first year, n/N (%)**	*0*	*2/315 (0.6)*	*6/526 (1.1)*	*2/361 (0.6)*
*at third year, n/N (%)***	*1/228 (0.4)*	*4/311 (1.3)*	*7/524 (1.3)*	*2/361 (0.6)*
Patients without SVI in either eye who developed bilateral SVI during follow-up, n/N (%)	2/229 (0.9)	2/316 (0.6)	1/527 (0.2)	1/362 (0.3)
*at first year, n/N (%)**	*1*	*0*	*0*	*0*
*at third year, n/N (%)***	*1*	*0*	*0*	*0*

All percentages were calculated among the patients without initial bilateral SVI.
*Patients who were followed for less than a year were excluded from the analysis
** Patients who were followed for less than 3 years were excluded from the analysis.

O-12

CLINICAL COURSE AND RADIOLOGICAL OUTCOMES OF PULMONARY ARTERY THROMBOSIS IN BEHÇET'S DISEASE: A COMPARATIVE STUDY WITH NON-BEHÇET PULMONARY THROMBOSIS

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Background/Objective. Pulmonary artery involvement is one of the most severe and life-threatening manifestations of Behçet's Disease (BD). Whether BD-related PAT represents a distinct clinical and radiological course compared with non-Behçet patients remains unclear. To compare the anatomical distribution, radiological evolution, and clinical outcomes of PAT in patients with BD and non-Behçet patients.
Methods. We conducted a retrospective cohort study including patients with radiologically confirmed PAT followed at our tertiary rheumatology center.
Results. A total of 169 patients were included: 137 with BD and 32 non-Behçet patients. Age at diagnosis of vascular involvement was similar between groups (36.0 ± 11.7 *vs* .36.0 ± 10.8 years, *p*=0.852). However, BD patients were predominantly male (69.1%), whereas non-Behçet patients were predominantly female (68.8%, *p*<0.001). During a median follow-up of 102 months, relapse occurred in 26.3% of patients, with a median time to first relapse of 24.5 months. Relapse occured in 6.3 % of non-BD pa-

O-12 Table I. Comparison of pulmonary artery involvement in Behçet's disease and non-Behçet patients.

Feature	Behçet disease (n=137)	Non-Behçet disease (n=32)	*p*-value
Age at vascular diagnosis (years)	36.0 ± 11.7	36.0 ± 10.8	0.852
Male sex, n (%)	95 (69.1)	10 (31.3)	<0.001
Pulmonary artery thrombosis distribution			
Main pulmonary artery	17 (12.4)	2 (6.3)	0.321
Lobar arteries	17 (12.4)	7 (21.5)	0.167
Segmental arteries	54 (39.4)	14 (43.8)	0.653
Subsegmental arteries	37 (27.0)	9 (28.1)	0.898
Small-caliber distal arteries	12 (8.8)	0 (0)	0.080
Radiological outcome (last imaging)			<0.001
Regression	65 (56.5)	23 (71.9)	
Stable	15 (13.0)	9 (28.1)	
Progression	35 (30.4)	0 (0)	
Relapse	36 (26.3)	2 (6.3)	0.017
Death	7 (5.1)	0 (0)	0.192

PAT was classified according to the most proximal anatomical level affected (main, lobar, segmental, subsegmental, and small-caliber distal arteries). Radiological evolution on follow-up imaging was categorized as regression, stability, or progression

tients during median 53 months follow-up. Immunosuppressive therapy was used in 137/137 (100%) BS patients (azathioprine 70.8%, cyclophosphamide 14.6%, anti-TNF 14.6%). Anticoagulant therapy had been administered to 85 patients (62.0%). All non-BD patients received anticoagulation. The anatomical distribution of PAT at the main, lobar, segmental, and subsegmental levels was comparable between the two groups (Table I). Radiological outcomes differed markedly between groups. On last imaging, progression was observed in 30.4% of BD patients but in none of the non-Behçet patients, whereas regression was more frequent in the non-Behçet group (71.9% *vs.* 56.5%) ($p<0.001$). Relapse occurred significantly more often in BD patients (26.3 % *vs.* 6.3%, $p=0.017$).

Conclusions. PAT in BD represents a more aggressive vascular phenotype than non-Behçet pulmonary artery thrombosis. Although proximal distribution is similar, BD shows exclusive distal small-vessel involvement, more radiological progression, and higher relapse rates, suggesting persistent inflammatory vasculopathy despite treatment. These findings support close radiological follow-up and stronger immunosuppressive management in BD-related PAT.

Key words: Behçet's disease, pulmonary artery thrombosis, radiology

O-13

PREDICTION FOR THE DEVELOPMENT OF INTESTINAL BEHÇET'S DISEASE BASED ON EXTRAINTESTINAL FACTORS

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Background/Objective. Intestinal involvement is one of the severe manifestations of Behçet's disease (BD), forming a distinct subgroup as the intestinal-type cluster in East Asia. Besides intestinal lesions, this cluster is characterized by the presence of arthritis, the absence of ocular lesions, and a low frequency of HLA-B51. This study aimed to determine whether these extraintestinal characteristics serve as risk factors for the subsequent development of intestinal lesions

Methods. We retrospectively analyzed data from 657 patients in the Yokohama City University (YCU) registry (1991–2018) and 6,754 patients in the Ministry of Health, Labour and Welfare (MHLW) national registry including newly diagnosed BD cases in Japan (2003-14). Patients without intestinal lesions at the baseline were enrolled. A score from 0 to 3 was assigned based on the presence of arthritis, absence of ocular lesions, and HLA-B51 negativity. We then evaluated the incidence of newly developed intestinal lesions according to this scoring system.

Results. Among eligible cases, 23 of 397 patients (5.8%) in the YCU registry and 90 of 1,455 patients (6.2%) in the MHLW registry developed new intestinal lesions. Using cases with a score of 0 or 1 as the reference, the odds ratios for developing intestinal lesions were 10.89 [2.96–40.13] for score 2 and 15.99 [3.98–65.76] for score 3 in the YCU registry. In the MHLW registry, the corresponding odds ratios were 2.25 [1.33–3.81] for score 2 and 2.87 [1.59–5.20] for score 3. No significant associations were found with vascular and neurological involvement.

Conclusion. This study identified arthritis, absence of ocular lesions, and HLA-B51 negativity as risk factors for the development of intestinal lesions in BD. Considering racial and genetic differences, further identification of additional associated genetic factors may improve predictive accuracy for intestinal involvement.

Key words: intestinal Behçet's disease, cluster, risk factor, HLA-B51

O-14

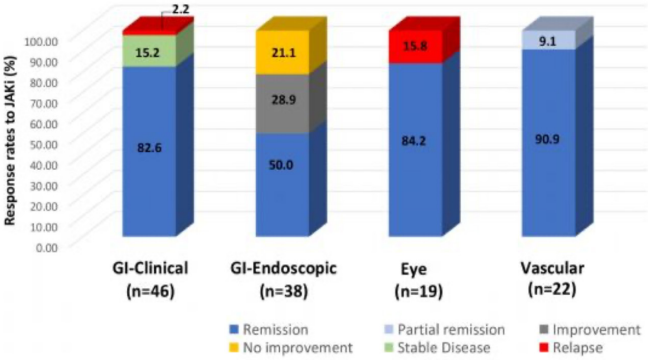
EFFICACY AND SAFETY OF JAK INHIBITORS IN BEHÇET SYNDROME: SYSTEMATIC LITERATURE REVIEW

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Objective. Given the active role of the JAK signaling pathway and the multifactorial immune mediators observed in Behçet syndrome (BS), JAK inhibitors (JAKi) may be promising agents. In this systematic literature review (SLR), we aim to evaluate the efficacy and safety of JAKi in BS.

Methods. A systematic literature search was conducted in Embase, PubMed, and the Cochrane Library to identify all reports on the efficacy and safety of JAKi in patients with BS. The SLR protocol was registered with the International Prospective Registry of Systematic Reviews (CRD420251000172). We additionally presented our patient with gastrointestinal involvement treated with upadacitinib.

Results. Among the 92 patients (48 men, mean (SD) age 38.5 (13.5) years), 46 used tofacitinib, 31 used baricitinib, and 15 used upadacitinib. All but 1 patient had used at least one previous conventional immunosuppressive, and 57.6% were refractory to at least one prior tumour necrosis factor inhibitor. JAKi were used in combination with conventional immunosuppressive and/or glucocorticoids in 90.6% of the patients. The primary indications for JAKi initiation were gastrointestinal involvement in 46 (50%) patients including our patient, vascular involvement in 22 (23.9%), uveitis in 19 (20.6%), and mucocutaneous and/or articular involvement in 5 (5.6%) patients. The remission rates for overall, gastrointestinal, vascular, and eye involvement were 85.9%, 82.6%, 90.9%, and 84.2% (Fig. 1). Serious adverse events were observed in 5.4% of the patients. No thrombotic events were reported.



O-14 Fig. 1. Response rates to JAKi

Conclusion. This SLR showed that JAKi may be effective in refractory BS patients with a favorable safety profile.

Key words: Behçet syndrome, upadacitinib, tofacitinib, baricitinib, JAK inhibitors.

O-15

CONCORDANCE OF CLINICAL PHENOTYPES, HLA-B51, AND ERAP-1 VARIATIONS AMONG FIRST-DEGREE RELATIVES WITH BEHÇET SYNDROME

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Objective. We aimed to evaluate a sizeable cohort of familial BS patients for the concordance of sex, clinical phenotypes, HLA-B51 positivity, ERAP-1 variations, and their associations among first-degree relatives both of whom have BS.
Methods. 713 patients with at least one first-degree relative diagnosed with BS (familial BS) were included. Twenty-four HLA-B51 positive family pairs were selected for ERAP-1 sequencing based on discordance for uveitis or vascular involvement. Additionally, 1,744 non-familial BS patients were identified for comparison.
Results. Familial BS patients had a younger age at first symptom and at disease onset, higher rates of pathergy and HLA B51 positivity, and lower rates of genital ulcer, ocular, and vascular involvements (Table I). Concordance rate among familial pairs were 73/231 (31.6%) for uveitis, 12/121 (9.9%) for vascular involvement, 5/40 (8%) for nervous system involvement, and 2/13 (15.3%) for gastrointestinal involvement. Among the 210 family pairs discordant for sex, major organ involvement was more common in men than in women (47.5% *vs*. 26.4% for uveitis, 25.0% *vs*. 6.2% for vascular involvement, 8.0% *vs*. 4.1% for nervous system involvement, 2.5% *vs*. 0.5% for gastrointestinal involvement). However, discordance rates for organ involvement were similar among family pairs discordant for sex and those concordant for sex (106/210, 50.5% *vs*. 89/191, 46.6%, *p*=0.50). Among the 143 family pairs with HLA-B51 data, 71 pairs were concordant for organ involvement. Among these, only 36 (50.7%) were concordant for HLA-B51 positivity (Table II). The sequencing of exons of the ERAP1 gene detected variants that are common in the general population.
Conclusion. Discordance for sex does not seem to be the main driver of discordance for organ involvement within family pairs. HLA-B51 positivity does not seem to be associated with concordance of clinical phenotypes within families. ERAP1 variants also did not contribute to clinical concordance, among HLA-B51 positive family pairs.
Key words: Behçet syndrome, familial BS, HLA-B51, ERAP-1, genetic epistasis

O-15 Table I. Comparison of clinical and demographic characteristics and HLA-B51 positivity between familial and sporadic Behçet syndrome patients HLA

	Familial Behçet syndrome (n=713)	Non-familial Behçet syndrome (n=1,744)	*p* value
Age, mean ± SD (years)	50.01 ± 13.6	46.40 ± 12.07	<0.001
Male, n (%)	382 (53.6)	1007 (57.7)	0.060
Type of familial relationship, n (%)*			
Mother-child	88 (21)	NA	
Father-child	76 (19)	NA	
Siblings	237 (60)	NA	
Age at first symptom onset, n (%)			
<16 years	179 (25.1)	249 (14.3)	<0.001
>16 years	534 (74.9)	1495 (85.7)	
Age at diagnosis, n (%)			
<16 years	65 (9.1)	41 (2.4)	<0.001
>16 years	648 (90.9)	1703 (97.6)	
Clinical features, n (%)			
Oral ulcers	707 (99.2)	1714 (98.3)	0.137
Genital ulcers	434 (60.9)	1284 (73.6)	<0.001
Papulopustular lesions	485 (68.0)	1186 (68.0)	1.000
Erythema nodosum	322 (45.2)	769 (44.1)	0.655
Positive pathergy test	349 /713(48.9)	567/1300 (40.9)	<0.001
Joint involvement	176 (24.7)	476 (27.3)	0.191
Eye involvement	263 (36.9)	828 (47.5)	<0.001
Vascular involvement	125 (17.5)	438 (25.1)	<0.001
Neuro-Behçet	33 (4.6)	78 (4.5)	0.915
Gastrointestinal involvement	16 (2.2)	47 (2.7)	0.576
Fulfilment ISG criteria, n (%)	652 (91.4)	1270 (72.8)	<0.001
Index age, mean ± SD (years)	30.48 ± 10.11	NA	
HLA-B51, n/N (%)	240/339 (70.8)	246/400 (61.3)	0.008
*Although 401 family pairs were identified, the total number of patients was 713 because families with three members were analyzed as two independent pairs.			

O-15 Table II. Concordance of organ involvement and HLA-B51 status among family pairs with known HLA-B51 status

Clinical features	Clinical features	Concordant for organ involvement (n=143)	HLA B51 +/+ (n=80)	HLA B51-/- (n=16)	HLAB51 +/- (n=47)
No major organ involvement	No major organ involvement	40	18 (45)	6 (15)	16 (40)
Uveitis	Uveitis	23	13 (56.5)	4 (17.3)	6 (26.2)
Vascular	Vascular	6	3 (50)	1 (16.6)	2 (33.4)
Neuro-Behçet	Neuro-Behçet	1	1	0	0
Gastrointestinal	Gastrointestinal	1	1	0	0

O-16

POST-THROMBOTIC VASCULAR REMODELLING IN BEHÇET SYNDROME

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Background/Objective. Vascular involvement, particularly deep vein thrombosis (DVT), affects nearly half of Behçet syndrome (BS) patients. Severe post-thrombotic syndrome (PTS) develops in up to 20%, yet its predictors remain poorly understood. This study aimed to identify clinical and ultrasonographic parameters associated with venous remodeling and severe PTS risk factors.
Methods. This study included BS patients with DVT (n=91), BS patients without vascular involvement (n=52), and healthy controls (HC, n=49). Clinical severity was assessed using CEAP and Villalta scores. Two blinded radiologists performed bilateral ultrasound and shear wave elastography (SWE) on the common femoral (CFV) and great saphenous veins (GSV). Predictors for severe PTS and stasis ulcers were identified via logistic regression.
Results. BS patients with DVT showed significantly higher CFV and GSV wall thickness compared to other groups ($p<0.001$). Importantly, BS patients without vascular involvement also had thicker vein walls than HCs ($p<0.001$). A mean CFV wall thickness cut-off of 0.687 mm predicted vascular involvement (%69.2 sensitivity and %73.5 specificity, AUC: 0.776, $p<0.001$). CFV wall SWE was significantly higher in DVT patients than HCs ($p=0.001$). While thrombus stiffness did not significantly correlate with thrombus age, higher values were observed within the first six months. Multivariate analysis identified venous reflux time [OR: 3.359; 95% CI (1.247–9.053), $p=0.017$] and fibrotic bands [OR: 4.927; 95% CI (1.112–21.837), $p=0.036$] as independent sonographic predictors for severe PTS (CEAP>3) and stasis ulcers. The number of relapses was the strongest clinical predictor for severe PTS per Villalta category [OR: 3.060; 95% CI (1.285–7.286), $p=0.011$].
Conclusion. Venous remodelling in BS is characterized by wall thickening and fibrosis. Prolonged reflux and fibrotic bands are critical sonographic markers for severe PTS, while clinical relapses amplify this damage. Early identification of these parameters is essential for managing PTS in BS.
Key words: thrombosis, remodelling, elastography

O-17

PHENOTYPE-SPECIFIC IMMUNE SIGNATURES IN BEHÇET'S DISEASE: A TRANSCRIPTOMIC AND CYTOKINE-BASED APPROACH

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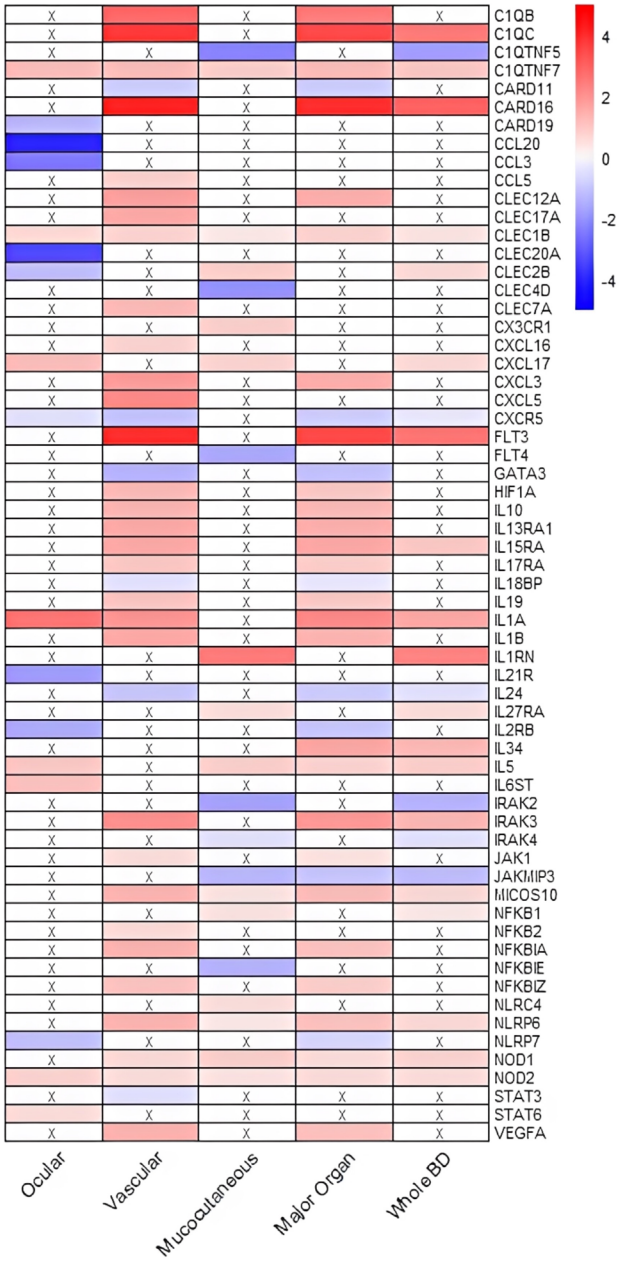
Background/Objective. Behçet's Disease (BD) is a chronic multisystem inflammatory disorder with heterogeneous phenotypes, most commonly mucocutaneous, ocular, and vascular. Clarifying the differential contributions of innate and adaptive immune mechanisms across these subgroups may advance personalized therapeutic strategies.

Methods. Serum samples were collected from BD patients during active disease (aBD) and remission (rBD), classified by phenotype. Patients with overlapping or gastrointestinal/neurological involvement were excluded. Pro- and anti-inflammatory cytokine concentrations were quantified using sandwich ELISA. Transcriptomic profiling of peripheral blood mononuclear cells (PBMCs) was performed with Illumina NovaSeq6000 sequencing. Quality control included FastQC, Trimmomatic, HISAT2 alignment to hg38, and HTSeq read quantification. Differentially expressed genes (DEGs) were identified using DESeq2 and NOISeq. Comparative analyses focused on innate immunity, inflammasome activity, adaptive pathways, and angiogenesis-related gene networks.

Results. Forty-one aBD patients (median age 29 years; M/F 9/32), 35 rBD patients (M/F 9/26), and nine healthy controls (HC) (M/F 3/6) were analyzed. Phenotypic distribution among aBD was: 19 mucocutaneous (m-aBD), 11 ocular (o-aBD), and 11 vascular (v-aBD). IFN-γ levels were elevated in aBD versus rBD (116 *vs*. 92 pg/ml, p=0.022). IL-35 was higher in HC compared with both aBD and rBD (p=0.05). IL-17-related cytokines were reduced in o-aBD but increased post-treatment, whereas they decreased in m-aBD and v-aBD. Transcriptome analysis (Fig. 1) confirmed lower Th17-related gene expression in o-aBD. Vascular BD displayed marked upregulation (4–13 fold) of angiogenesis and hematopoiesis-related genes (VEGF, HIF1A, FLT3). Distinct innate immune activation and inflammasome-related signatures further differentiated v-aBD from other groups.

Conclusion. Innate immunity and Th1-driven adaptive responses play central roles in BD pathogenesis, with phenotype-specific patterns. Suppressed Th17 responses in o-aBD may underlie the limited efficacy of IL-17 blockade in uveitis. Vascular BD is characterized by enhanced angiogenesis and inflammasome activation, offering insights for phenotype-tailored therapeutic strategies. These findings refine the immunopathogenic map of BD and may inform precision medicine approaches.

Key words: Behçet's disease, transcriptomics, innate immunity, IL-1 pathway, vascular inflammation



O-17 Fig. 1. Heat-map analysis of phenotypes comparing to healthy controls.

O-18

CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION SECONDARY TO PULMONARY ARTERY INVOLVEMENT OF BEHÇET'S SYNDROME

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Background. Pulmonary artery involvement in Behcet's syndrome (BS) remains a major cause of morbidity and mortality despite immunosuppressive therapies. Massive hemoptysis from pulmonary artery aneurysm (PAA) is the main cause of death, while elevated pulmonary artery pressure may also contribute to adverse outcomes. Data on the frequency and determinants of chronic thromboembolic pulmonary hypertension (CTEPH) in BS are limited.
Objective. This study aimed to assess the frequency of chronic thromboembolic pulmonary hypertension and its associated clinical, radiological, and hemodynamic features in patients with Behçet's syndrome and pulmonary artery involvement.
Methods. BS patients with pulmonary artery involvement followed at our center between June 2023 and December 2024 were included. Demographic and clinical features, functional class (NYHA), six-minute walk distance, pulmonary function tests, and pro-BNP levels were recorded. Pulmonary hypertension (PH) probability was assessed by transthoracic echocardiography (TTE) per 2022 ESC/ERS guidelines, and high-risk cases underwent right heart catheterization (RHC). Computed tomography (CT) pulmonary angiography and/or single-photon emission computed tomography (SPECT)-CT were reviewed for chronic thromboembolic disease (CTED).
Results. A total of 108 BS patients (88M/20F; median age 46 years, disease duration 9 years) were studied. Forty-seven (43.5%) had PAA, and the remaining had isolated pulmonary artery thrombosis (PAT) which was more frequent in women (75%). Echocardiography showed intermediate/high PH probability in 22.2%. PH was confirmed in 12 cases by RHC. CTED and CTEPH were identified in 36.1% and 5.5% of patients. Female gender increased CTED risk 3.4-fold. NYHA ≥2 (OR:18.7, p=0.001) and perfusion defect (OR:13.2, p=0.025) independently predicted PH.
Conclusion. CTEPH and CTED occur more often than expected in Behçet's patients with pulmonary artery involvement. Functional class and perfusion defects predict PH. Our results suggest the need for systematic assessment to improve treatment and clinical outcome.
Key words: pulmonary artery involvement, pulmonary hypertension, chronic thromboembolic pulmonary hypertension

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