14th International Conference on Behçet's Disease

8th - 10th July 2010

Queen Mary University Of London - Mile End Campus
London, United Kingdom

ABSTRACT BOOK

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ISBD
INTERNATIONAL SOCIETY FOR BEHÇET'S DISEASE

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WELCOME LETTER

On behalf of the United Kingdom Behçet's Disease Forum and the United Kingdom Behçet's Syndrome Society, we have great pleasure in organising the 14th International Conference on Behçet's Disease, in London from 8th-10th July 2010 under the auspices of the International Society for Behçet's Disease.

The venue of the Conference is the Queen Mary University Conference Centre at Mile End, London E1. The venue is within five minutes walk from the Mile End Underground Station, and a short taxi ride from hotels situated at St Catherine's dock near Tower Bridge.

The Conference Steering Committee has planned to provide an excellent scientific programme, aiming at bringing new developments in basic and clinical science to bear on the specific issues of Behçet's Disease (BD). Topics include Immunology of BD, Vasculitis in BD, Genetic basis of BD, Regional Inflammation and Age-related BD. The programme also includes debates on topics of current interest or controversy.

Besides these main activities, you will be able to enjoy the rich cultural life, and tourist attractions of London and the rest of the UK.

We welcome you to London .

Professor Dorian Haskard, United Kingdom Behçet's Disease Forum

Honorary President of the 14th International Conference

Dr Graham Wallace, United Kingdom Behçet's Disease Forum

Honorary General Secretary of the 14th International Conference

Dr Colin Barnes, United Kingdom Behçet's Disease Forum

International Affairs Secretary of the 14th International Conference

COMMITTEES

Executive Committee - International Society for Behçet's Disease

President	Sungnack LEE	(Korea) - Dermatology
Past-President	Hasan YAZICI	(Turkey) - Rheumatology
Vice-President	Kenneth CALAMIA	(USA) - Rheumatology
Secretary	Dongsik BANG	(Korea) - Dermatology
Treasurer	Samir ASSAAD KHALIL	(Egypt) - Internal Medicine
President of 14th International Congress	Dorian HASKARD	(UK) - Rheumatology
President Past International Congress	Michael SCHIRMER	(Austria) - Rheumatology
Member - Dermatology	Eun-So LEE	(Korea) - Dermatology
Member - Internal Medicine	Petros SFIKAKIS	(Greece) - Internal Medicine
Member - Scientific Affairs	Haner DIRESKENELI	(Turkey) - Rheumatology
Member - Publication Affairs	Graham WALLACE	(UK) - Immunology

Hon. Life Presidents - International Society for Behçet's Disease

Colin G BARNES	(UK) - Rheumatology
Nihat DILSEN	(Turkey) - Rheumatology
George EHRLICH	(USA) - Rheumatology
Thomas LEHNER	(UK) - Immunology
Desmond O'DUFFY	(USA) - Rheumatology

Local Organising Committee

Honorary President	Dorian HASKARD	(UK) - Rheumatology
International Affairs Secretary	Colin G BARNES	(UK) - Rheumatology
Honorary General Secretary	Graham WALLACE	(UK) - Immunology
	Farida FORTUNE	(UK) - Oral Medicine
	Robert MOOTS	(UK) - Rheumatology
	Miles STANFORD	(UK) - Ophthalmology
Representatives - Behçet's Syndrome Society	Jan MATHER	(UK) - BSS
	Chris PHILLIPS	(UK) - BSS

International Scientific Committee

Adnan AL-ARAJI	(UK) - Neurology
Clive ARCHER	(UK) - Dermatology
Samir ASSAAD KHALIL	(Egypt) - Internal Medicine
Dongsik BANG	(Korea) - Dermatology
Paul BROGAN	(UK) - Paediatrics
Kenneth CALAMIA	(USA) - Rheumatology
Anne CHAMBERLAIN	(UK) - Rheumatology
Hormoz CHAMS	(Iran) - Ophthalmology
Jorge CRESPO	(Portugal) - Rheumatology
Fereydoun DAVATCHI	(Iran) – Rheumatology
Andrew DE BURGH-THOMAS	(UK) – Genito-Urinary Medicine
Haner DIRESKENELI	(Turkey) - Rheumatology
Ahmet GUL	(Turkey) - Rheumatology
Martin van HAGEN	(Netherlands) - Medicine
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Habib HOUMAN	(Tunisia)- Internal Medicine
Fumio KANEKO	(Japan) - Dermatology
Ina KÖTTER	(Germany) - Ophthalmology
Isabelle KONE-PAUT	(France) - Paediatrics
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Desmond KIDD	(UK) - Neurology
Eun-So LEE	(Korea) - Dermatology
Sungnack LEE	(Korea) - Dermatology
Susan LIGHTMAN	(UK) - Ophthalmology
Wafa MADANAT	(Jordan) - Rheumatology
Ravinder MAINI	(UK) - Rheumatology
Shigeaki OHNO	(Japan) - Ophthalmology
Sofia OLIVIERA	(Portugal) - Research Biologist
Yang PEIZENG	(China) - Ophthalmology
Richard POWELL	(UK) - Clinical Immunology
Jeremy SANDERSON	(UK) - Gastroenterology
Michael SCHIRMER	(Austria) - Rheumatology
Petros SFIKAKIS	(Greece) - Internal Medicine
Alan SILMAN	(UK) - Epidemiology
Hasan YAZICI	(Turkey) - Rheumatology
Yusuf YAZICI	(USA) - Rheumatology
Manfred ZIERHUT	(Germany) - Ophthalmology
Christos ZOUBOULIS	(Germany) - Dermatology

SCIENTIFIC PROGRAMME

8th July 2010, Thursday

8:00 – 9:00 Registration

9:00 – 9:15 President's Welcome Speech

9:15 – 11:30 **Scientific Session 1 – Immunology - Incorporating therapy**
Co-chairs Dr. Ina Kotter (Germany) and Dr Y. Kaneko (Japan)

9:15 – 9:45 Outside speaker - Professor Adrian Hayday (UK)
The evidence for lymphoid stress-surveillance of tissues

9:45 – 10:15 ISBD invited speaker – Dr Bahram Bodaghi (France)
Regulatory T-cells in non infectious uveitis : a new therapeutic strategy

10:15 – 11:30 Selected speakers from abstracts

O-001 Altered Expression of Costimulatory Molecules in Behçet's Disease According to Clinical Activity (Ref: 074)
[1]Ji Hyun Sim, M.d., [1]Mi Jin Park, B.s., [2]Sun Park, M.d., Ph.d., [1]Eun-so Lee, M.d., Ph.d.
[1]Department of Dermatology, Ajou University School of Medicine, [2]Department of Microbiology, Ajou University School of Medicine .

O-002 Expression of pro-inflammatory protein S100A12 (EN-RAGE) in Behçet's disease and its association with disease activity (Ref:111)
[1]Eun Chun Han, [1]Sung Bin Cho, [1]Keun-jae Ahb, [1]Sang Ho Oh, [1]Jihyun Kim, [2]Dong Soo Kim, [1]Kwang Hoon Lee, [1]Dongsik Bang .
[1]Department of Dermatology and Cutaneous Biology Research Institute, Yonsei University College of Medicine, Seoul, Korea, [2]2department of Pediatrics, Yonsei University College of Medicine, Seoul, Korea

O-003 Identification of JAK1 as a candidate inflammatory signalling pathway by genome-wide expression profiling in monocytes from patients with Behcet's Disease (Ref:159)
[1]H. Direskeneli, [4]Jj. Boyle, [2]Ft. Ozdemir, [3]V. Yılmaz, [2]E. Ekşioğlu-demiralp, [4]D. Haskard, [3]G. Saruhan-direskeneli .
[1]Department of Rheumatology, Marmara University, Faculty of Medicine, Istanbul, Turkey, [2]Department of Immunology, Marmara University, Faculty of Medicine, Istanbul, Turkey, [3]Department of Physiology, Istanbul University, Istanbul Faculty of Medicine, Istanbul, Turkey, [4]Cardiovascular Sciences Centre, Imperial College, London, Uk

O-004 A novel murine model of Behcet's-like eye disease associated with proteoglycan induced arthritis in the absence of gamma interferon. (Ref:263)
[1]Jelena Kezic, [1]Michael Davey, [1]Tibor Glant, [1]Katie Dahlhausen, [1]Stephen Planck, [1]Tammy Martin, [1]James Rosenbaum, [1]Holly Rosenzweig .
[1]Casey Eye Institute, Oregon Health & Science University, Portland, Or., [2]Veterans Affairs Medical Center, Portland, Or., [3]Rush University Medical Center, Chicago, Il.

O-005 The functional analysis of MICA polymorphism with an emphasis on Behcet's disease (Ref:314)
[1]Seema Shafi, [1]Miles R Stanford, [1]Graham R Wallace, [1]Adrian C Hayday .
[1]Dept Immunobiology Guy's Hospital, [2]Dept Ophthalmology St Thomas' Hospital, [3]Academic Dept Ophthalmology, Birmingham

11:30 – 13:00 **Coffee Break and Posters**

13:00 – 14:00 **Lunch**

14:00 – 16:15 **Scientific session 2 – Vasculitis**
Co-chairs – Dr Michael Schirmer (Austria) and Professor Samir Assad-Khalili (Egypt)

14:00 – 14:30 Outside speaker – Dr Justin Mason (UK)
Imaging studies in large vessel vasculitis

14:30 – 15:00 ISBD invited speaker – Professor Hasan Yazici
The vasculitis of Behçet's syndrome: What do we know and not know?

15:00 – 16:15 Selected speakers from abstracts

O-006 Management of Thrombosis In 62 Patients With Behçet Syndrome Over 15 Years (Ref: 009)
[1]Puja Mehta, [2]Mike Laffan, [1]Dorian Haskard .
[1]Department of Rheumatology, Imperial College Healthcare NHS Trust, [2]Department Of Haematology, Imperial College Healthcare NHS Trust

O-007 Pulmonary Artery Hypertension in Behçet's Syndrome (Ref: 194)
[1]Emire Seyahi, [2]Murat Baskurt, [1]Melike Melikoglu, [3]Canan Akman, [3]Deniz Cebi Olgun, [4]Eda Simsek, [1]Vedat Hamuryudan, [1]Serdar Kucukoglu, [1]Hasan Yazici .
[1]Cerrahpasa Medical Faculty, Rheumatology Department, [2]University of Istanbul, Institute of Cardiology, Department of Cardiology, [3]Cerrahpasa Medical Faculty, Radiology Department, [4]Cerrahpasa Medical Faculty, Internal Medicine Department

O-008 Cutaneous Vasculitis in Behçet's Disease (Ref: 219)
[1]Ko-ron Chen .
[1]Saiseikai Central Hospital

O-009 Pulmonary Perfusion Scintigraphy Findings in Behçet's Disease (Ref: 309)
[1]Fulya Cosan, [2]Isik Adalet, [1]Bahar Artim-esen, [1]Orhan Aral, [1]Ahmet Gul .
[1]Istanbul Faculty of Medicine, Department of Internal Medicine, Division of Rheumatology, Istanbul University, [2]Istanbul Faculty of Medicine, Department Nuclear Medicine, Istanbul University

O-010 Efficacy of Adalimumab in patients with Behçet's Disease unsuccessfully treated with Infliximab (Ref: 316)
[1]Salvatore d'Angelo, [1]Ignazio Olivieri, [1]Pietro Leccese, [1]Angela Padula, [1]Carlo Palazzi, [2]Loredana Latanza.
[1]Rheumatology Department of Lucania, San Carlo Hospital of Potenza and Madonna Delle Grazie Hospital of Matera, Potenza and Matera; Italy, [2]Section of Ocular Immunopathology, Department of Ophthalmology, "a.cardarelli" Hospital, Naples, Italy

16:15 – 16:30 **Coffee Break**

16:30 – 17:00 Debate – Autoimmunity vs autoinflammation
Professor Haner Direkeneli and Dr Graham Wallace

17.15 -18.00 **Special lecture -** Tom Greeves MA PhD (Archaeologist and Historian) & Chris Chapman (Photographer and Film Maker) to speak on *'Joined by the Ears - the Remarkable Journey of the Three Hares along the Silk Route from Ancient China to Devon...and the Bones!'.*

18.00-20.00 **Opening Cocktail in the Octagon, Queen Mary University of London**

9th July 2010, Friday

09:15 – 11:30 Scientific session 3 - Regional inflammation
Co-chairs Dr Dongsik Bang (Korea) and Dr Wafa Madanat (Jordan)

09:15 – 09:45 Outside speaker Dr Dennis McGonigle (UK)
Behcets Disease classification as an intermediate between adaptive and innate mediated immune disease

09:45 – 10:15 ISBD invited speaker Dr Ahmet Gul (Turkey)
Multiple faces of inflammation in Behcet's disease

10:15 – 11:30 Selected speakers from abstracts

O-011 Effects of infliximab in the treatment of refractory posterior uveitis of Behçet´s disease after withdrawal of infusions (Ref: 016)
[1]Alfredo Adan, [1]Victoria Hernandez, [1]Laura Pelegrin, [1]Gerard Espinosa, [1]Marina Mesquida, [1]Raimon Sanmarti .
[1]Hospital Clinic Barcelona

O-012 AIN457, a fully human monoclonal anti-interleukin-17A monoclonal antibody, for the adjunctive treatment of posterior segment uveitis secondary to Behçet's Disease: The SHIELD study (Ref: 062)
[1]Ilknur Tugal-tutkun, [1]On Behalf of The Shield Study Group .
[1]Department of Ophthalmology, Istanbul University, Istanbul Faculty of Medicine, Istanbul, Turkey

O-013 Methotrexate in Ocular lesions of Behcet's Disease; a Longitudinal Study (Ref: 097)
[1]Fereydoun Davatchi, [1]Bahar Sadeghi Abdollahi, [1]Hormoz Shams, [1]Farhad Shahram, [1]Abdolhadi Nadji, [1]Cheyda Chams-davatchi, [1]Tahereh Faezi, [1]Massoomeh Akhlaghi, [1]Farima Ashofteh .
[1]Rheumatology Research Center, Tehran University of Medical Sciences

O-014 Neuro-Behcet's disease in Japan: a multicenter retrospective survey (Ref: 104)
[1]Shunsei Hirohata, [2]Hirotoshi Kikuchi, [3]Tetsuji Sawada, [4]Hiroko Nagafuchi, [5]Masataka Kuwana, [6]Mitsuhiro Takeno, [6]Yoshiaki Ishigatsubo .
[1]Kitasato University School of Medicine, [2]Teikyo University School of Medicine, [3]Tokyo Medical University, [4]St. Marianna University School of Medicine, [5]Keio University School of Medicine, [6]Yokohama City University Graduate School of Medicine

O-015 A Comparative Study of Clinical, Endoscopic and Histologic Findings in Patients with Gastrointestinal Behçet's Disease and Crohn's Disease (Ref: 298)
[1]Aykut Ferhat Celik, [1]Ibrahim Hatemi, [2]Gulen Hatemi, [1]Emine Satir, [1]Yusuf Erzin, [1]Gürhan Şişman, [3]Suha Goksel, [4]Orhan Sami Gültekin, [2]Hasan Yazici .
[1]Istanbul University, Cerrahpasa Medical School, Gastroenterology, [2]Istanbul University, Cerrahpasa Medical School, Rheumatology, [3]Istanbul University, Cerrahpasa Medical School, Pathology, [4]Endotip Diagnosis Center

11:30 – 13:00 **Coffee Break and Posters**

13:00 – 14:00 **Lunch**

14:00 – 16:15 **Scientific session 4 – Age-related BD**
Co-chairs – Dr Paul Brogan (UK) and Dr Martin van Hagen (Netherlands)

14:00 – 14:30 Outside speaker - Professor Michael Beresford (UK)
UK Paediatric Rheumatology Clinical Studies Group: Driving Forward Care & Understanding

14:30 – 15:00 ISBD invited speaker Dr Isabelle Kone-Paut (France)
Pediatric Behcet's Disease: diagnostic problems and treatment peculiarities

15:00 – 16:15 Selected speakers from abstracts

O-016 Ethnic and gender patterns of age distribution and duration of Adamantiades-Behçet's disease (ABD) course in Germany (Ref: 098)
[1]Nestor G. Papoutsis, [1]Nikolaos G. Bonitsis, [1]Andreas Altenburg, [1]Helmut Orawa, [1]Ina Kötter, [1]Lothar Krause, [1]Uwe Pleyer, [1]Djalil Djawari, [1]Rudolf Stadler, [1]Uwe Wollina, [1]Peter Kohl, [1]Wilhelm Kirch, [1]Falk Ochsendorf, [1]Peter Martus, [1]Christos C. Zouboulis .

[1]*German Registryof Adamantiades- Behçet's Disease, Dessau, Germany,* [2]*Institute of Medical Informatics, Biometry and Epidemiology, Charité Universitaetsmedizin Berlin, Berlin, Germany*

O-017 Systemic Involvements and Currently Preferred Immunosuppressive Agents In A Large Population Composed of Relatively Young Behcet's Patients From Countrywide (Ref: 166)
[1]Omer Karadag, [1]Veli Yazisiz, [1]Sedat Yilmaz, [1]Battal Altun, [1]Mustafa Gezer, [1]Murat Karaman, [1]Muhammet Cinar, [1]Hakan Erdem, [1]Salih Pay, [1]Ayhan Dinc .
[1]*Gulhane Schoolof Medicine, Rheumatology Division Ankara, Turkey*

O-018 Cross-sectional survey of neurologic and psychiatric symptoms in North American patients with Behcet's syndrome. (Ref: 250)
[1]Ilya Kister, Md, [2]Maria Filopoulos, [2]Monalyn De Los Reyes Labitigan, Md, [3]Sara Crystal, Md, [4]Matthew Robbins, Md, [1]Joseph Herbert, Md, [2]Yusuf Yazici, Md .
[1]*1. ms Care Center, Department of Neurology, NYU School of Medicine, Ny,* [2]*2. Behcet's Syndrome Evaluation, Treatment, Research Center, Department of Medicine, NYU School of Medicine, NY,* [3]*3. Department of Neurology, NYU School of Medicine, Ny,* [4]*4. Department of Neurology, Albert Einstein College of Medicine, Bronx, NY*

O-019 Eye disease in juvenile patients with Behçet's syndrome (Ref: 264)
[1]Yilmaz Ozyazgan, [2]Emire Seyahi, [1]Ahmet Sarica, [2]Serdal Ugurlu, [2]Gulen Hatemi, [2]Sebahattin Yurdakul, [2]Hasan Yazici .
[1]*University of Istanbul, Cerrahpasa Medical Faculty, Department of Ophthalmology,* [2]*University of Istanbul, Cerrahpasa Medical Faculty, Department of Internal Medicine, Division of Rheumatology .*

O-020 Work Disability In Behcet's Syndrome (Ref: 267)
[1]Gulen Hatemi, [1]Vedat Hamuryudan, [1]Koray Tascilar, [1]Serdal Ugurlu, [1]Hasan Yazici .
[1]*Istanbul University, Cerrahpasa Medical School, Rheumatology*

16:15 – 16:30 **Coffee Break**

16:30 – 17:00 Debate – Geographical differences in BD
Professor Shigeaki Ohno (Japan) and Professor Miles Stanford (UK)

20:00 **Gala Dinner in The Great Hall, St. Bartholomew's Hospital**

10th July 2010, Saturday

09:00 – 11:15 Scientific session 5 – Genetics
Co-chairs – Dr Sofia Oliveira (Portugal) and Dr Jan van Laar (Netherlands)

09:00 – 09:30 Outside speaker – Dr Oliver Brand (UK)
Genetic of complex diseases:lessons from Graves' disease

09:30 – 10:00 ISBD invited speaker Dr Eun-Bong Lee (Korea)
Perspective of candidate gene analysis in Behcet's disease

10:00 – 11:15 Selected speakers from abstracts

O-021 Methylenetetrahydrofolate reductase (MTHFR) genetic polymorphism in patients with Behcet disease (Ref: 060)
[1]Farhad Shahram, [2]Niloofar Mojarad Shafiee, [3]Mehrdad Behmanesh, [1]Arash Tehrani Banihashemi, [1]Alireza Faridar, [1]Fereydoun Davatchi .
[1]*Rheumatology Research Center, Tehran University of Medical Sciences, Tehran, Iran,* [2]*Department of Biology, Science and Research Branch, Islamic Azad University, Tehran, Iran,* [3]*Department of Genetic, Faculty of Biological Sciences, Tarbiat Modares University, Tehran, Iran*

O-022 Impact of HLA-B51(5) on Behçet's Disease Clinical Phenotype: Systematic Review and Meta-Analyses on Phenotype-Genotype Correlates (Ref: 129)

[1]Mahr Alfred, [1]Maldini Carla, [1]Cheminant Morgane, [2]Lavalley Michael P., [1]Guillevin Loïc .
[1]Hospital Cochin, Université de Paris 5, Paris, France, [2]Boston University School of Public Health, Boston, Massachusetts, United States .

O-023 Genome-wide association studies define two susceptibility loci for Behçet's disease (Ref: 185)

[1]Nobuhisa Mizuki, [1]Akira Meguro, [2]Masao Ota, [3]Yeong Wook Song, [3]Eun Bong Lee, [4]Nobuyoshi Kitaichi, [5]Kenichi Namba, [5]Yukihiro Horie, [6]Mitsuhiro Takeno, [7]Sunao Sugita, [7]Manabu Mochizuki, [8]Seiamak Bahram, [6]Yoshiaki Ishigatsubo, [9]Hidetoshi Inoko, [10]Shigeaki Ohno .
[1]Department of Ophthalmology and Visual Science, Yokohama City University Graduate School of Medicine, [2]Department of Legal Medicine, Shinshu University School of Medicine, [3]Department of Internal Medicine, Seoul National University College of Medicine, [4]Department of Ophthalmology, Health Sciences University of Hokkaido, [5]Department of Ophthalmology and Visual Sciences, Hokkaido University Graduate School of Medicine, [6]Department of Internal Medicine and Clinical Immunology, Yokohama City University Graduate School of Medicine, [7]Department of Ophthalmology and Visual Science, Tokyo Medical and Dental University Graduate School of Medicine, [8]Laboratoire D'immunogénétique Moléculaire Humaine, Centre de Recherche d'Immunologie et d'Hématologie, Faculté de Médecine, Université de Strasbourg, [9]Department of Molecular Life Science, Division of Molecular Medical Science and Molecular Medicine, Tokai University School of Medicine, [10]Department of Ocular Inflammation and Immunology, Hokkaido University Graduate School of Medicine .

O-024 A Twin Study In Behçet's Syndrome (Ref: 191)

[1]Seval Masatlioglu, [2]Emire Seyahi, [3]Eda Tahir Turanli, [2]Izzet Fresko, [4]Feride Gogus, [5]Ebubekir Senates, [6]Fatma Oguz Savran, [2]Hasan Yazici .
[1]Division of Rheumatology, Haydarpaşa Numune Research and Education Hospital, [2]Division of Rheumatology, Department of Medicine, Cerrahpaşa Medical Faculty, [3]Molecular Biology and Genetics Department, Dr. Orhan Öcalgiray Molecular Biology and Genetic Research Center (mobgam), Istanbul Technical University, [4]Department of Physical Medicine and Rehabilitation, Division of Rheumatology, University of Gazi, [5]Division of Gastroenterology, Haydarpaşa Numune Research and, Education Hospital, [6]Department of Medical Biology, Istanbul Medical Faculty, University of Istanbul

O-025 A Genome-wide Association Study Identifies Common Variants of the IL10 and IL23R Genes that Contribute to Behçet's Disease Susceptibility (Ref: 224)

[1]Elaine F. Remmers, [2]Fulya Cosan, [1]Yohei Kirino, [1]Michael J. Ombrello, [3]Barbara Yang, [4]Virginia G. Kaklamani, [5]William E. R. Ollier, [6]Dongsik Bang, [7]Graham R. Wallace, [3]Massimo Gadina, [1]Daniel L. Kastner, [2]Ahmet Gül .
[1]Laboratory of Clinical Investigation, National Institute of Arthritis and Musculoskeletal and Skin Diseases, [2]Department of Internal Medicine, Istanbul Faculty of Medicine, [3]Office of Science and Technology, National Institute of Arthritis and Musculoskeletal and Skin Diseases, [4]Division of Hematology/oncology, Northwestern University, [5]School of Immunity and Infection, University of Manchester, [6]Dept of Dermatology, Yonsei University College of Medicine, [7]Academic Unit of Ophthamology, Universtiy of Birmingham .

11:15 – 11:45 **Coffee Break**

11:45 – 12:00 Presentation of Awards

12:00 – 12:30 Closing Remarks Professor Songnack Lee (President)

General Notes

Scientific sessions

The aim of the conference organisers is to have the sessions based on concepts rather than BD in specific tissues. It is hoped that this will lead to the identification of common ground in the pathogenesis of the disease in different sites, and to highlight differences that may be important. The invitation of outside speakers should provide current knowledge in the particular fields that will inform and encourage participants.

Scientific session 1

The aim of this session is to address the wider issues of the immune response in BD. Several publications report a Th1 polarised response yet in several tissues i.e. anterior chamber, joint and CNS it is reported as a predominantly a neutrophilic infiltrate. Moreover, roles for NKT, NK and γδ T cells have all been implicated in the pathogenesis of BD. The session will seek, along with the related debate, to identify a common process and the kinetics of such a response.

Scientific session 2

Vasculitis is a common feature of many of the clinical manifestations of BD. However, it is still not clear which cells and processes are involved. For example, what are the differences between parenchymal and non-parenchymal CNS disease. What are the vascular responses in the skin in conditions such as erythema nodosum. Is the retinal vascular occlusion seen in BD due to neutrophil activation? Most importantly, is the endothelium of these sites activated and why. It is envisaged that these and other concepts will be addressed.

Scientific session 3

The current concept of regional inflammation is particularly relevant to BD. Why do certain patients get one manifestation and others a different form. Importantly, the potential difference between the inflammatory response at the mucosal surfaces compared to other immune privileged sites will be addressed. The effect of treatment on inflammation at different sites will also be discussed.

Scientific session 4

BD is acomplex disease that can occur in children as well as adults. Paediatric BD is a difficult diagnostic problem as well as being a complicated treatment issue. It is not clear whether paediatric BD is the same condition as adult BD or what the outcome for juvenile patients will be. Similarly, should the same treatment be given to children as to adults? These points will be addressd in this session.

Scientific session 5

BD has long been considered as having a genetic component. In this session current studies will be discussed. Recent data from genome-wide analysis studies, from different ethnic groups will be presented. Similarly case controls studies of candidate genes in different ethnic groups will be discussed.

Debates

The plan is to hold two debates on current topics of general interest in BD. These debates will feature two selected individuals one who will speak for the proposal and one to speak against. Speakers will have 10 minutes to present their case, followed by 5 minutes each for a rebuttal. The questioning will then be opened up to the floor, before a vote (for fun) will be taken.

Debate 1 Autoimmunity versus auto inflammation

A case for BD being an auto inflammatory response has been presented in several recent publications. Is it therefore time to change the textbook definition of BD, or is autoimmunity still a better term to describe the responses seen in BD?

Debate 2 Geographical differences in BD

It has long been suggested that BD has a different clinical picture in different parts of the world. Can this be supported in the light of current data. If so what may be the causes of such differences. Data from Japan suggests that BD is decreasing, but is this seen in other countries?

ORAL PRESENTATION INDEX

Board Number	Ref. No	Topic	Title	Date	Time
O-001	74	Immunology	Altered Expression of Costimulatory Molecules in Behçet's Disease According to Clinical Activity	08.07.2010	10:15-10:30
O-002	111	Immunology	Expression of pro-inflammatory protein S100A12 (EN-RAGE) in Behçet's disease and its association with disease activity	08.07.2010	10:30-10:45
O-003	159	Immunology	Identification of JAK1 as a candidate inflammatory signalling pathway by genome-wide expression profiling in monocytes from patients with Behcet's Disease	08.07.2010	10:45-11:00
O-004	263	Immunology	A novel murine model of Behcet's-like eye disease associated with proteoglycan induced arthritis in the absence of gamma interferon.	08.07.2010	11:00-11:15
O-005	314	Immunology	The functional analysis of MICA polymorphism with an emphasis on Behcet's disease	08.07.2010	11:15-11:30
O-006	9	Vasculitis	Management of thrombosis in 62 patients with Behçet Syndrome over 15 years	08.07.2010	15:00-15:15
O-007	194	Vasculitis	Pulmonary Artery Hypertension in Behçet's Syndrome	08.07.2010	15:15-15:30
O-008	219	Vasculitis	Cutaneous Vasculitis in Behçet's Disease	08.07.2010	15:30-15:45
O-009	309	Vasculitis	Pulmonary Perfusion Scintigraphy Findings in Behçet's Disease	08.07.2010	15:45-16:00
O-010	316	Vasculitis	Efficacy of Adalimumab in patients with Behçet's Disease unsuccessfully treated with Infliximab	08.07.2010	16:00-16:15
O-011	16	Regional	Effects of infliximab in the treatment of refractory posterior uveitis of Behçet ´s disease after withdrawal of infusions	09.07.2010	10:15-10:30
O-012	62	Regional	AIN457, a fully human monoclonal anti-interleukin-17A monoclonal antibody, for the adjunctive treatment of posterior segment uveitis secondary to Behçet's Disease: The SHIELD study	09.07.2010	10:30-10:45
O-013	97	Regional	Methotrexate in Ocular lesions of Behcet's Disease; a Longitudinal Study	09.07.2010	10:45-11:00
O-014	104	Regional	Neuro-Behcet's disease in Japan: a multicenter retrospective survey	09.07.2010	11:00-11:15
O-015	298	Regional	A Comparative Study of Clinical, Endoscopic and Histologic Findings in Patients with Gastrointestinal Behçet's Disease and Crohn's Disease	09.07.2010	11:15-11:30
O-016	98	Paediatric	Ethnic and gender patterns of age distribution and duration of Adamantiades-Behçet's disease (ABD) course in Germany	09.07.2010	15:00-15:15
O-017	166	Paediatric	Systemic involvements and currently preferred immunosuppressive agents in a large population composed of relatively young Behcet's patients from countrywide	09.07.2010	15:15-15:30
O-018	250	Paediatric	Cross-sectional survey of neurologic and psychiatric symptoms in North American patients with Behcet's syndrome.	09.07.2010	15:30-15:45
O-019	264	Paediatric	Eye disease in juvenile patients with Behçet's syndrome	09.07.2010	15:45-16:00
O-020	267	Paediatric	Work disability in Behcet's syndrome	09.07.2010	16:00-16:15
O-021	60	Genetics	Methylenetetrahydrofolate reductase (MTHFR) genetic polymorphism in patients with Behcet disease	10.07.2010	10:00-10:15
O-022	129	Genetics	Impact of HLA-B51(5) on Behçet's Disease Clinical Phenotype: Systematic Review and Meta-Analyses on Phenotype-Genotype Correlates	10.07.2010	10:15-10:30
O-023	185	Genetics	Genome-wide association studies define two susceptibility loci for Behçet's disease	10.07.2010	10:30-10:45
O-024	191	Genetics	A twin study in Behçet's syndrome	10.07.2010	10:45-11:00
O-025	224	Genetics	A Genome-wide Association Study Identifies Common Variants of the IL10 and IL23R Genes that Contribute to Behçet's Disease Susceptibility	10.07.2010	11:00-11:15

ORAL PRESENTATIONS

Book No: O-001
Ref. No: 74
Topic: Immunology

Altered Expression of Costimulatory Molecules in Behçet's Disease According to Clinical Activity

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Objectives: For a balanced immune response, key inhibitory costimulartory molecules that critically affect peripheral T cell tolerance are important. To understand the implication of costimulartory molecules in Behçet's disease (BD), we investigated the expression of CTLA-4 and PD-1 on T cell subsets and of their ligands, CD80, CD86 and PD-L1 on antigen presenting cells (APCs).
Methods: PBMCs of subjects (11 patients with active BD, 8 patients with inactive BD, 8 patients with recurrent aphthous ulcer as disease control and 10 healthy volunteers) were cultured and stained for analysis by flow cytometry. To measure the sCTLA-4 concentrations in culture supernatants, ELISA was performed. To investigate the mRNA expression level of PD-L1, real time PCR was performed.
Results: Reduced expression of CTLA-4 on CD4+ T cells after stimulation and its ligand, CD86 on APCs on the resting state was shown in active BD group compared with the HC group. There is significantly decreased frequency of PD-L1 expressing APCs in the BD group compared with the other group on the resting state and this was sustained after stimulation. There were no differences in concentration of sCTLA4 among each group in culture supernatants at 24 hours and 48 hours after stimulation. Decreased expression of PD-L1 mRNA in the active BD group was demonstrated compared with the HC group.
Conclusion: This study suggests the possibility of impaired function of CTLA-4/B7 pathway and PD-1/PD-L1 pathway as a part of pathogenesis in BD.

Book No: O-002
Ref. No: 111
Topic: Immunology

Expression of pro-inflammatory protein S100A12 (EN-RAGE) in Behçet's disease and its association with disease activity

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Objective. S100A12 is a member of the S100 family of calcium-binding proteins and is secreted in inflamed tissues or in the bloodstream by activated neutrophils. Expression of S100A12 has been reported in various diseases, especially non-infectious inflammatory diseases such as Kawasaki disease, giant cell arteritis and inflammatory bowel disease. This study was conducted to determine tissue expression and serum levels of S100A12 in Behçet's disease (BD) patients and the correlation of S100A12 serum level with disease activity of BD.
Methods. We included ten BD patients who fulfilled the criteria for diagnosis according to the International Study Group for BD. The activity of BD was calculated using the BD Current Activity Form. Serum concentrations of S100A12 and interleukin-8 were measured by an enzyme-linked immunosorbent assay before and after treatment. Immunohistochemical studies were also performed to detect skin S100A12 expression.
Results. Serum S100A12 level was significantly increased in the active BD period ($P < 0.001$), in the inactive BD period ($P = 0.041$) and in patients with active Kawasaki disease ($P = 0.028$) compared with that of the healthy controls. Serum S100A12 level decreased significantly from baseline compared to post-treatment ($P = 0.017$). The activity score of BD was significantly correlated with serum S100A12 level (Spearman's coefficient = 0.464, $P = 0.039$). Immunohistochemical studies showed that S100A12 was strongly expressed in erythema nodosum-like skin lesions of BD patients.
Conclusions. S100A12 may contribute to BD pathogenesis related to neutrophil hyperactivity and reflects disease activity in some patients with BD.

Book No: O-003
Ref. No: 159
Topic: Immunology

Identification of JAK1 as a candidate inflammatory signalling pathway by genome-wide expression profiling in monocytes from patients with Behcet's Disease

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Aim: To obtain a global view of the immune responses in Behcet's Disease (BD) compared to Familial Mediterranean Fever (FMF), a systemic, auto-inflammatory disorder.
Method: Twenty-eight BD (F/M: 9/19, mean age: 33.4), 13 FMF (F/M: 9/4, mean age: 30.4) patients and 8 controls (F/M: 4/4, mean age: 30.6) were enrolled. Whole-genome microarray profiling was performed with human U133 (Plus 2.0) microarrays on an Affymetrix platform using isolated CD14+monocyte and CD4+T-lymphocyte subsets. Data was analysed with Genespring (Version 10.0). RT-PCR was used for the validation of JAK1 expression.
Results: Among 28792 transcripts analysed, in CD14+ monocytes, 1188 transcripts reached a significant difference level with a minimum 2 fold difference observed in 279 genes. In CD4+T-lymphocytes, 2880 transcripts showed significant difference with at least 2 fold difference in 109 genes. Among over-expressed genes in BD CD14+ monocytes, oxysterol binding protein-like 8 (OSBPL8)(3.8 fold), cell-division-cycle-27 homolog (S. cerevisiae)(CDC27)(3.1 fold) and myeloid/lymphoid or mixed-lineage leukemia 3 (MLL3)(3.1 fold) were among the highest. However, in principal component analysis, Januse-kinase-1 (JAK1)(2.6 fold) and metallothionein 1X, (MT1X)(2.1 fold) are shown to be the major regulatory molecules of associated-signalling pathways. Validation by RT-PCR also showed an increased JAK1 expression compared to FMF (BD: 9.5 vs. FMF: 5.1 fold, p=0.045).
Discussion: Whole-genome microarray analysis demonstrated a selective activation of BD monocytes compared to FMF, suggesting their significant role between innate and adaptive immune responses. Activation of JAK1 through various cytokines such as IL-2, IL-6, IL-15 and interferon-g may be the dominant signaling pathway driving inflammation in BD.

Book No: O-004
Ref. No: 263
Topic: Immunology

A novel murine model of Behcet's-like eye disease associated with proteoglycan induced arthritis in the absence of gamma interferon.

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Uveitis, including retinal vasculitis and retinitis, is often the dominant clinical manifestation of Behcet's disease. We recently described a model characterized by uveitis and arthritis in mice whose T cell receptors are transgenic to recognize the proteoglycan (PG) aggrecan (Rosenzweig et al., 2008). These TCR-Tg mice develop mild anterior uveitis at 3 weeks post-immunization with PG. As PG-induced arthritis is known to be IFN-g dependant, we investigated the effects of IFN-g deficiency on ocular inflammation. TCR-Tg and IFN-g knockout (KO)/TCR-Tg mice were immunized with PG and ocular inflammation analyzed over 6 weeks. Intravital microscopy and histology revealed a progressive mild anterior uveitis in TCR-Tg mice. By comparison, the number of rolling, adherent and infiltrating cells in the iris was higher in PG-treated IFN-g KO/TCR-Tg mice than TCR-Tg mice. Histological analysis of IFN-g deficient mice at 2 to 4 weeks post-immunization revealed a heavy infiltrate in both anterior and posterior segments, with vitritis, retinal vasculitis and retinal folding. Immunofluorescence staining of uveitic eyes from IFN-g KO/TCR-Tg mice demonstrated a predominance of neutrophils and mononuclear MHC Class II-positive cells. In contrast to the eye, joint inflammation was reduced in IFN-g deficient mice. This study describes and characterizes a new model of ocular inflammation that accompanies joint and spine disease. Whilst IFN-g deficiency is protective in the joints, severe anterior uveitis with marked posterior involvement akin to the ocular disease in Behcet's is evident in the eye. Our findings have potential implications for the treatment of systemic diseases with accompanying uveitis.

Book No: O-005
Ref. No: 314
Topic: Immunology

The functional analysis of MICA polymorphism with an emphasis on Behcet's disease
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Behcet's disease (BD) is a multi-organ inflammatory disease. A main factor of genetic predisposition locates to. HLA-B51, but MICA*009 is also reported to be strongly associated with BD. The aim is to undertake a functional analysis of MICA polymorphism, and determine the functional effects of the HLA-B51-MICA*009 combination.

Isogenic stable cell lines expressing MICA*009,*008,*004 and ULBP2 were separately created. These are used in killing assays to compare the cell biology of the different MICA alleles and to assess whether MICA*009 varies from other alleles in its capacity to promote killing by NKG2D positive cells from healthy controls and BD patients. Similarly, double transfectants: HLA-B51-MICA*009 and HLA-B52-MICA*009 (control) were created and used in similar assays to determine the inhibitory effect, if any, of HLA-B51 in patients.

Data from killing assays show a most unanticipated, donor-to-donor variation in the hierarchy with which different transfectants are targeted by healthy controls and patients. These hierarchies seem stable longitudinally and these data are validated by the CD107a assay. Differential killing cannot easily be explained by an affinity hierarchy because people kill different targets with different hierarchies. Variation among only patients is even greater: 25% show MICA*009 as the primary target which may be a clue to a functional significance of MICA*009 in BD. HLA-B51 shows comparable inhibition of MICA*009 targeting as does HLA-B52 in controls. HLA-B51 shows a distinctly stronger level of inhibition of MICA*009 targeting than does HLA-B52 in 20% of patients. This may be a clue to a functional significance HLA-B51 in BD

Book No: O-006
Ref. No: 9
Topic: Vasculitis

MANAGEMENT OF THROMBOSIS IN 62 PATIENTS WITH BEHÇET'S SYNDROME OVER 15 YEARS
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Vascular involvement in Behçet's syndrome (BS) is predominantly due to inflammation, and immunosuppression is generally regarded as appropriate.The role of anti-coagulation is controversial, due to the risk of bleeding from aneurysms and the perceived rarity of pulmonary embolism.We reviewed vascular involvment in the 657 patients referred for assessment to our tertiary Behçet's clinic between 1994-2009. 62 patients (9%) had a history of thrombosis, excluding isolated thrombophlebitis. Thrombosis was the event that eventually led to the diagnosis of BS in 39. 17 were considered by the referring physician to have had a pulmonary embolus. The large majority (55 patients; 89%) were treated with warfarin at the time of thrombosis and before referral to our centre. Warfarin was discontinued because of complications in only two patients, one due to haemoptysis secondary to pulmonary aneurysms and the other due to an upper gastro-intestinal bleed. Thus, patients in the UK tend to be anti-coagulated at the time of thrombosis prior to referral to specialist centres. Arguements in defence of this include:(i) the rarity and frequent ambiguity of the diagnosis of BS in the UK,(ii) lack of familiarity of most UK acute physicians with the condition, (iii) the chances of thrombosis in an undiagnosed patient being due to more common causes, and (v) the risk of pulmonary embolism in our series being as great, or greater, than that of bleeding.Our future focus will be encouraging early specialist referral for assessment of the risk of anti-coagulation and review of treatment.

Book No: O-007
Ref. No: 194
Topic: Vasculitis

Pulmonary Artery Hypertension in Behçet's Syndrome

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Objectives: We prospectively surveyed the frequency of pulmonary hypertension (PH) by echocardiography among Behcet's syndrome (BS) patients with pulmonary artery involvement (PAI) along with diseased and healthy controls.
Methods: Pulmonary symptoms were evaluated by a standardized questionnaire. Right and left ventricular functions and pulmonary artery pressure were studied using transthoracic echocardiography. The cut-off value for PH was determined as ≥ 35 mmHg. Six-minute walking distance and CO diffusing capacity (DLCO) were also measured. Serum levels of endothelin-1 (ET-1), vascular endothelial growth factor (VEGF) and pro-brain natriuretic peptide (pro-BNP) were assessed.
Results: The frequency of individuals having PH was significantly increased only among patients with systemic sclerosis (SS) and BS with PAI. Six-minute walking distance was significantly diminished among patients with SS and BS patients with vascular disease not having PAI. ET-1 levels were found to be above normal limits only among patients with SS. Pro-BNP levels were significantly increased among SS patients and BS patients with PAI while serum levels of VEGF did not differ among the study groups.
Conclusion: BS may cause mild PH especially when it involves pulmonary arteries. This correlates with a diminished DLCO and increased pro-BNP levels. This novel finding indicates that BS may involve small/micro pulmonary vessels as well as large pulmonary arteries.

Table: Demographic and clinical characteristics of the study groups

	BS withPAI(n = 30)	BS vascular disease (without PAI)(n= 26)	BS Mucocutaneous involvement only(n=21)	Healthy controls(n=21)	Systemic sclerosis(n=23)	P
M/F	27/3	23/3	19/2	19/2	2/21	<0.001
Mean age, years	35 ± 8	39 ± 8	36 ± 7	36 ± 7	48 ± 10	<0.001
Disease duration, years	8 ± 5	13 ± 7	9 ± 4	-	11 ± 7	0.009
Exercise dyspnea, n (%)	15 (50)	10 (38)	1 (4)	0	14 (61)	<0.001
PAH (cut-off 35 mmHg), n (%)	5 (17)	2 (8)	0	0	6 (26)	0.016
6 minute walking distance, m	489±79	448±107	508±88	532±71	392±96	<0.001
Pro-BNP ng/ml	680±383	552±175	566±185	453±113	831±311	<0.001
DLCO, %	72 ± 15	92 ±4	95 ± 14	95 ±18	52 ±22	<0.001

Book No: O-008
Ref. No: 219
Topic: Vasculitis

Cutaneous Vasculitis in Behçet's Disease
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Cutaneous Vasculitis in Behçet's disease
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Ko-Ron Chen, M.D., Ph.D.
Background: We have previously demonstrated that near half of the Behçet's disease (BD) (19/40) with either erythema nodosum-like lesions or infiltrated erythema had cutaneous vasculitis characterized by superficial thrombophlebitis or dermal venulitis. (Chen KR, et al. J Am Acad Dermatol 1997;36:689-696.)
Objective: Whether and how often the cutaneous vasculitis could be identified in the recent BD cases.
Methods: Four cases met the criteria of the International Study Group for BD since 2007 were investigated histopathologically.
Results: All the 4 cases were confirmed to have dermal venulitis with underlying lobular neutrophilic panniculitis. The cutaneous manifestations reveal erythema nodosum-like lesions in 3 cases and infiltrated erythema in 1 case with coexistent papulopustular lesions.
Conclusions: Erythema nodosum-like lesions and infiltrated erythema in BD are vasculitis-associated skin lesions. Not only the superficial thrombophlebitis but the features of dermal venulitis with underlying lobular neutrophilic panniculitis are also the diagnostic clues to BD.

Book No: O-009
Ref. No: 309
Topic: Vasculitis

Pulmonary Perfusion Scintigraphy Findings in Behçet's Disease
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Behçet's disease (BD) patients may develop various pulmonary manifestations such as pulmonary artery aneurysms, organizing pneumonia, or pulmonary thrombosis. Despite high frequency of venous thrombosis, pulmonary thromboembolism (PTE) is considered as an unexpected finding in BD. In this retrospective study, we analyzed the data from 105 patients (74 male, 31 female) with BD, who were investigated with pulmonary scintigraphy (PS) between 2008 and 2009. PS was requested for the screening of vascular involvement (n=46), or further investigation of patients with vascular involvement and dyspnea (n=49), or abnormal chest X-ray/thorax CT findings (n=10). Half of the patients (n=53) had known vascular involvement. PS findings were found to be normal in 55 patients (%52). In the remaining, 29 patients had bilateral (13 widespread, 9 local, 7 heterogeneous) and 21 patients had unilateral (7 widespread, 12 local, 2 heterogeneous) abnormal perfusion findings. There was a significant association between the abnormal scintigraphy findings and arterial aneurysms (P=0.035, OR=3.2) and any vascular involvement (P=0.007, OR=2.8). All patients with abnormal PS findings were investigated further with thorax CT, and no association was documented with the scintigraphy findings and PTE. PS findings suggest unrecognized small vessel and/or parenchymal lesions especially in those patients with vascular involvement. These perfusion defects may indicate a tendency to thrombosis in small vessels along with large ones, and they should not be interpreted as findings of PTE.

Book No: O-010
Ref. No: 316
Topic: Vasculitis

Efficacy of Adalimumab in patients with Behçet's Disease unsuccessfully treated with Infliximab
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[1]Rheumatology Department of Lucania, San Carlo Hospital of Potenza and Madonna Delle Grazie Hospital of Matera, Potenza and Matera; Italy, [2]Section of Ocular Immunopathology, Department of Ophthalmology, "a.cardarelli" Hospital, Naples, Italy. .

We analyzed the effects of adalimumab in 17 patients with BD (12 M, 5 F, median age 37 yrs, mean disease duration 11.3 yrs) in whom immunosuppressive therapy had failed. All patients had received infliximab which was discontinued in 2 patients for infusion reaction, in 2 patients for no response and in the remaining 13 patients there was a lack of efficacy. Indication for adalimumab treatment were uveitis in 4 patients, severe mucocutaneous manifestations in 9, CNS vasculitis in 1, leg ulcers in 1 and refractory seizure in the remaining 2.
In patients with ocular involvement we observed complete remission in 3 and a stable disease in 1. No results have been observed in the 2 patients with seizure. Rapid improvement was obtained in patient with severe CNS vasculitis, occurred during treatment with infliximab. In the patient with leg ulcers adalimumab resulted in a partial remission.
In patients with severe mucocutaneous involvement we observed complete remission in 4, partial remission in 3, stable disease in 1 and progressive disease in 1.
Until now, 12 patients are still receiving adalimumab (median duration treatment 20 months), in 5 patients adalimumab is administrated in monotherapy and in the other 7 patients concomitant immunosuppressive therapy has been tapered. No serious adverse events were recorded.
In our experience adalimumab has been showed effective and safe in patients with refractory BD.

Book No: O-011
Ref. No: 16
Topic: Regional

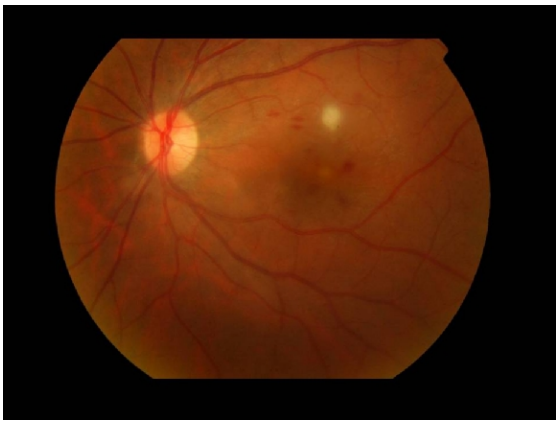
Effects of infliximab in the treatment of refractory posterior uveitis of Behçet ´s disease after withdrawal of infusions
[1]Alfredo Adan, [1]Victoria Hernandez, [1]Laura Pelegrin, [1]Gerard Espinosa, [1]Marina Mesquida, [1]Raimon Sanmarti .
[1]Hospital Clinic Barcelona .

Purpose : To determine the efficacy of infliximab treatment in refractory posterior uveitis in Behçet´s disease (BD) after withdrawal of infusions
Methods: Four patients with posterior uveitis secondary to Behçet´s disease were treated with infliximab until complete remission and were followed after withdrawal of infusions. Intraocular inflammation was assessed using the binocular indirect ophthalmoscopy score, best corrected visual acuity (BCVA) and foveal thickness measured by optic coherence tomography (OCT).
Results: All of the patients included received treatment with infliximab during a minimum of 12 months and were in complete remission. All these patients were off steroids and immunosupressants. Main follow-up after withdrawal of infusions was 7.5 months.Two out of 4 patients (50%) maintain complete remission of posterior uveitis. BCVA was stable in 7 eyes . OCT showed worsening in macular edema in the two eyes of the patients with reactivation.
Conclusions: Infliximab is efficient long-term treatment of refractory posterior uveitis in BD. Repeated infusions are required to maintain long-term remission which may be sustained on discontinuation of the drug .

Patient	Infusionsnumber	Follow-upIFX	Follow-upafter WD-IFX	BCVA afterWD-IFX	FinalBCVA	Foveal thickness (µm)WD-IFX	Final Foveal thickness(µm)	Relapse	TreatmentAfter relapse
(1)	14	20 months	10 months	R 20/80L 20/30	R 20/80L 20/30	R 187L 201	R 194L 312	YES	Adalimumab

(2)	12	16 months	8 months	R 20/30L 20/30	R 20/20L 20/20	R 210L 225	R 205L 214	NO	None
(3)	11	16 months	6 months	R 20/20L 20/40	R 20/20L 20/20	R 203L 219	R 214L 340	YES	Adalimumab
(4)	14	17 months	6 months	R NLPL 20/20	R NLP20/20	L 212	L 220	NO	None

Case 3



Fundus photograph of the left eye of a 26 year-old female patient with Behçet disease showing vitritis with retinal exudate,retinal haemorrhages and macular edema

Book No: O-012
Ref. No: 62
Topic: Regional

AIN457, a fully human monoclonal anti-interleukin-17A monoclonal antibody, for the adjunctive treatment of posterior segment uveitis secondary to Behçet's Disease: The SHIELD study

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Background and purpose
Uveitis occurs in 60-80% of patients with Behçet's disease (BD) and recurrent uveitis exacerbations cause blindness in 25-50% of cases despite systemic immunosuppressive therapy (IST). Elevated interleukin-17A (IL-17A) in patients with BD may contribute to the ocular and systemic manifestations. Systemic IL-17A blockade with AIN457, a human monoclonal antibody, is a novel therapeutic approach for patients with Behçet's uveitis.

Methods
The SHIELD study is an ongoing phase III, 24-week, multicenter, randomized, double-masked, placebo-controlled trial in adults with BD 2 exacerbations of intermediate, posterior or panuveitis≥experiencing requiring IST within 6 months of enrollment. Patients with active or quiescent uveitis were randomized to subcutaneous AIN457 300 mg (every 2 weeks or monthly after loading doses) or placebo injections given adjunctive to prescribed IST which is gradually tapered. Uveitis exacerbations, defined as either a >/= 2+ increase in vitreous haze, new retinal infiltrates/vasculitis/hemorrhages, or >/= 10 ETDRS letter decrease in visual acuity (VA), are treated with rescue therapy. The primary efficacy endpoint is the rate of recurrent ocular exacerbations during 24 weeks of treatment. Secondary endpoints are change from baseline at week 24 in immunosuppressive medication score, visual acuity, foveal thickness, and vitreous haze grades, and safety. Non-ocular outcomes are explored.

Results
A total of 118 patients were enrolled globally. Rationale for AIN457 in posterior segment uveitis, study design, and baseline demographic data will be presented.

Conclusions
The SHIELD study seeks to demonstrate the efficacy and safety of AIN457 as an adjunct to systemic immunosuppressants for therapy of posterior segment Behçet's uveitis.

Book No: O-013
Ref. No: 97
Topic: Regional

Methotrexate in Ocular lesions of Behcet's Disease; a Longitudinal Study
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Purpose: Cytotoxic drugs are the gold standard and the first line treatment for ophthalmological manifestations of Behcet's Disease (BD). The purpose of this study was to evaluate methotrexate on long-term follow-up, in a longitudinal study on 15 years.
Materials and Methods: methotrexate was given as 7.5 to 15 mg/weekly. Prednisolone was associated as 0.5 mg/kilogram/daily. Prednisolone was tapered later, according to the improvement of inflammation.
Visual acuity (VA) was calculated on Snellen chart (10/10). Disease Activity Index (DAI) was calculated for anterior uveitis (AU), posterior uveitis (PU), and retinal vasculitis (RV) according to Ben-Ezra. Total Inflammatory Activity Index (TIAI) and Total Adjusted Disease Activity Index (TADAI) were also calculated. Results, before the treatment and after different intervals (from 3 months to 15 years), were compared together by Student paired t test. Confidence interval (CI) at 95% was calculated for percentages.
Results: Five-hundred-ninety-seven (597) patients received methotrexate. patients-year-follow-up was 2231. Good results (stabilized or improved eyes from the baseline to last evaluation) were: VA 62.4%, PU 88.7%, RV 68.1%, TIAI 81.7%, TADAI 76.4%. The mean VA improved from 5.1 to 5.5, mean PU improved from 1.8 to 0.6, mean RV improved from 1.9 to 1.2, mean TIAI improved from 13.3 to 6.1, and mean TADAI improved from 25.7 to 19. The difference was statistically significant for all parameters $p<0.001$). Longitudinal study showed maintenance of good results all over the study time.
Conclusion: methotrexate with prednisolone was effective in ocular lesions of Behcet's disease. Good results were maintained on long-term study.

Book No: O-014
Ref. No: 104
Topic: Regional

Neuro-Behcet disease in Japan: a multicenter retrospective survey
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The present study was designed to delineate the clinical characteristics of neuro-Behcet's disease (NBD) and to determine the reliable diagnostic parameters. A multicenter retrospective survey was performed on BD patients who fulfilled the diagnostic criteria of the international study group and presented neurological manifestations between 1987 and 2008. The diagnosis of either NBD or neurological manifestations due to causes other than BD (non-NBD) was confirmed by retrospective review of the clinical records. NBD was further classified into acute and chronic progressive NBD according to the clinical courses. A total of 142 BD patients were studied, including 73 with acute NBD, 36 with chronic progressive NBD, and 33 with non-NBD. Smoking and HLA-B51 were correlated with chronic progressive NBD ($p=0.0018$ and $p=0.0037$, respectively), whereas significantly higher numbers of acute NBD patients had been taking cyclosporine A. Cerebrospinal fluid (CSF) cell counts were most prominently elevated in acute NBD, but within normal limit in approximately 15% of chronic progressive NBD. Whereas CSF IL-6 was elevated in both acute and chronic progressive NBD, it decreased only in acute NBD after treatment. High intensity lesions on MRI T2-weightened images were found in 63.2% of acute NBD, 57.1% of chronic progressive NBD, and 43.8% of non-NBD, whereas brainstem atrophy was observed in 4.4% of acute NBD, 71.4% of chronic progressive NBD, and 6.3% of non-NBD. The results disclosed characteristics of acute NBD and chronic progressive NBD. Moreover, the data demonstrated the efficacy of CSF cell counts and IL-6 in the

Book No: O-015
Ref. No: 298
Topic: Regional

A Comparative Study of Clinical, Endoscopic and Histologic Findings in Patients with Gastrointestinal Behçet's Disease and Crohn's Disease

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Background:
Gastrointestinal Behçet's Disease (GIBD) and Crohn's disease (CD) have similar clinical and endoscopic findings which are difficult to distinguish from one another. The aim of this survey is to analyze the symptoms, clinical, endoscopic and histopathological findings of our GIBD patients and compare them to our CD patients followed in the same setting.

Methods:
All of our inflammatory bowel disease patients including GIBD patients are recorded under the same outpatient registry. GIBD is defined as any patient who fulfill ISG criteria and have colonic ulcers in the abscence of NSAID use. CD was diagnosed with clinical, endoscopic, radiologic and histologic findings. None of our CD patients fulfilled ISG criteria. Between 2000 and 2010, 36 patients have been diagnosed as GIBD. For the purposes of this study, we selected 2 CD patients for each GIBD patient. These were selected as the CD patient before and the one after each GIBD patient in our records.

Results:
We evaluated 36 GIBD (20 men, 16 women, mean age 37.8 ± 7.8 SD), and 72 CD patients (41men, 31 women, mean age 43.5 ± 13.2). Clinical and endoscopic findings are summarised in the table.

Conclusion:
The location of intestinal involvement is similar in the two diseases. Focal single ulceration and complications like perforation and gross rectal bleeding are more suggestive of GIBD. GIBD patients have more frequent and earlier surgical interventions than CD patients. Absence of granuloma in GIBD is not a distinctive feature for differentiating the two diseases.

	GIBD (N=36)	CD (N=72)	p
Age at diagnosis	31.1±6.9	36.8±13.2	0.02
Abdominal pain	75 %	69.4 %	NS
Diarrhea	36 %	51.3 %	NS
Bloody diarrhea	8.3 %	20.8 %	NS
Gross rectal bleeding	19.4 %	1.3 %	0.00
Perforation	19.4 %	1.3 %	0.00
Weight loss	2.2 %	19.4 %	0.00
Being Operated	52.7 %	30.5 %	0.03
Age at the operation	30.8±8.89	38.7±13.7	0.04
Gastrointestinal disease duration (years) at time of the operation	0.8±1.4	3.5±5.7	NS
Area of involvement			
Colonic	47.2 %	26.3 %	NS
Ileocolonic	19.4 %	34.7 %	NS
Ileocecal	11.1 %	8.3 %	NS
Ileal	19.4 %	27.7 %	NS
Other	2.7 %	2.7 %	NS
Ulcer Location			
Focal single	27.7 %	5.5 %	0.002

Focal multiple	22.2 %	19.4 %	NS
Segmental	44.4 %	63.8 %	NS
Diffuse	5.5 %	11.1 %	NS
Number of ulcers			
Single	33.3 %	5.5 %	0.000
Multiple	50 %	73.6 %	0.019
Aphtous lesions	16.6 %	11.1 %	NS
Cobblestone appereance	0 %	9.7%	NS
Granuloma in mucosal biopsies (%)	0 / 26 (0 %)	7/56 (12.5 %)	0.09

Book No: O-016
Ref. No: 98
Topic: Paediatric

Ethnic and gender patterns of age distribution and duration of Adamantiades-Behçet's disease (ABD) course in Germany

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Objective: To determinate the differences of age and duration on the course of ABD among ethnic and gender populations.
Patients and methods: 590 patients [344 male (m) and 246 female (f) - 227 German (G), 267 Turkish (T) and 96 Patients of other ethnic origin) residing in Germany reported to the Adamantiades-Behçet's registry.
Results: The overall mean age(MA) on onset of ABD was 26 years (yr) (m:25.4 vs f:27.6, ns, G:28.0 vs T:25.7, ns). The frequency of onset-manifestation in the third life-decade (20-29yr) was 40.7% (m:48.1% vs f:30.8%, ns, G:33.3% vs T:44.2%, ns). Age at onset ranged between 1-72yr (13.2% <16yr, 86.8% ≥16yr). MA at second-manifestation was 28.0yr (m:26.6 vs f:28.5, ns, G:28.9 vs T:27.8, ns). MA at full-development of ABD was 29.5yr (m:28.3 vs f:30.7, p<.005, G:31.0 vs T:28.5, ns), full ABD was observed at females 2.5yr later compared to males. MA at diagnosis was 31.0yr (m:29.6 vs f:33.3, p<.005, G:34.0 vs T:33.0, p<.005), f and G were diagnosed 4yr later. Mean duration(MD) from disease-onset to full-development of ABD lasted 3.0 months (m:3.0 vs f:1.9, ns, G:5.9 vs T:1.0, ns). MD from disease-onset to diagnosis counted 36.0 months (m:28.5 vs f:48.0, p<.005, G:53.0 vs T:27.0, p<.005), f were diagnosed 1.5 and G 2yr later. MD from diagnosis to full-development of ABD counted 0.4yr (m:0.2 vs f:0.4, p<.005, G:0.8 vs T:0.3, p<.005). T after being diagnosed developed full-picture of ABD a half yr faster.
Conclusion: The presented data indicate distinct ethnic and gender patterns regarding the course of ABD.

Book No: O-017
Ref. No: 166
Topic: Paediatric

SYSTEMIC INVOLVEMENTS AND CURRENTLY PREFERRED IMMUNOSUPPRESSIVE AGENTS IN A LARGE POPULATION COMPOSED OF RELATIVELY YOUNG BEHCET'S PATIENTS FROM COUNTRYWIDE

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Background: Prognosis and treatments in Behcet's disease (BD) depend on the site and the degree of clinical involvement. The immunosuppressive drugs are widely added to the treatment when a systemic involvement takes place besides mucocutaneous lesions. This study is aimed to investigate the extent of clinical involvement and preferred treatment approaches in a single center which admits relatively young male Behcet's patients from the whole country.
Patients and Methods: The files of all Behcet's patients admitted from 2007 to 2009 were screened for the disease characteristics and their treatments. All patients were diagnosed as for being fulfilled International Society BD criteria.

Results: A total of 863 patient files (Female/Male: 160/703) were evaluated. The mean age and disease duration were 33.3 ± 10.1 and 7.2 ±5.1 years, respectively. Besides mucocutaneous findings 37.7% of patients have ophthalmologic and 20.2% of patients have venous involvement. Frequencies of patients with neurological, arterial and gastrointestinal involvements were 2.9%, 2.3% and 1.2%, respectively. The mostly ever used drugs were colchicine in (88.5%) and corticosteroids (54.5%). Countrywide, immunosuppressive agents preferred for organ involvements were shown (Table).
Conclusion: In this relatively young population composed from all over the country; the frequency of ophthalmologic, venous and neurological involvement is somewhat lower than previous reported cohorts. Most patients have been commenced yet on azathioprine, cyclosporine A as a chronic immunosuppressive agent. However, interferon alpha seems to be emerged as an alternative in resistant cases; other choices such that infliximab or thalidomide is rarely needed.

Drugs	Number of patients (%)
Colchicine	763 (88.5%)
Corticosteroids	470 (54.5%).
Azathioprine	332 (38.5%)
Cyclosporin A	95 (11.0%)
Interferon alpha	84 (9.7%)
Sulphasalazine	51 (5.9%)
Cyclophoshamide	48 (5.6%)
Methotrexate	15 (1.7%)
Benzatin penicillin	14 (1.6%)
Mycophenolate mofetil	8 (0.9%)
Infliximab	4 (0.5%)
Thalidomide	3 (0.4%)

Book No: O-018
Ref. No: 250
Topic: Paediatric

Cross-sectional survey of neurologic and psychiatric symptoms in North American patients with Behcet's syndrome.
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Objectives: To assess frequency of neurologic and psychiatric symptoms in patients attending a Behcet's syndrome (BS) clinic in the US.
Background: There is paucity of data on neurological and psychiatric symptoms in BS patients in North America.
Design/Methods: A questionnaire designed to elicit detailed history of neurologic and psychiatric symptoms and of neurologic work-up (brain MRI and lumbar puncture) was administered to consecutive BS patients attending Behcet's Clinic at the New York University, NY.
Results: Forty six (46) consecutive BS patients completed the questionnaire. Average age was 35.4 +/-12.7 years, disease duration was 7.6 +/-6.7 ; 93% were women. Thirty eight patients (83%) reported history of one or more non-painful neurological symptom; the most common was 'progressive cognitive difficulties' (30 patients, 65%). Headache was reported by 87% (40 patients), of whom 50% met criteria for migraine and 33% - chronic daily headaches. Thirty one patients (67%) underwent brain MRI and 18 (39%) both MRI and lumbar puncture, mostly for the purpose of ruling out meningitis. Moderate-severe depression was diagnosed in 28% of patients (PHQ-9>14) and moderate-severe anxiety (GAD-7>10) in 37%.
Conclusions: In the cross-sectional survey of neurologic and psychiatric symptoms in BS patients attending specialized clinic in North America there was a remarkably high prevalence of neurologic and psychiatric symptoms. Headaches were seen in almost 90% of patients, and one third of them were chronic daily headache with a marked female predominance.

Book No: O-019
Ref. No: 264
Topic: Paediatric

Eye disease in juvenile patients with Behçet's syndrome

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Objective: To describe clinical features of eye disease in patients with juvenile onset Behçet's syndrome (BS).
Patients and Methods: There were 136 (71 M/ 65 F) patients with juvenile onset BS who were registered between 1980 and 2006 in our multidisciplinary outpatient clinic at Cerrahpasa Medical Faculty. We identified 76 (49 M/ 27 F) (56 %) patients with eye disease. We retrospectively surveyed clinical features, damage score and complications.
Results: The mean age of the patients at initial presentation was 14.3 ± 1.7 years. The median follow-up time was 10 years [IQR: 2-13] in the remaining. Eye disease was present at the initial visit in 70 (46 M/ 24 F) patients and developed during the follow-up in 6 (3 M/ 3F). Ocular involvement was bilateral in 66 patients and unilateral in the remaining 10. Anterior uveitis was observed in 7 eyes (5 %; right: 5, left: 2), posterior uveitis in 49 eyes (35 %; right: 25, left: 24) and panuveitis in the 86 eyes (60 %; right: 42, left: 44). The damage scores, number of activations and complications are shown in Table.
Conclusions: As seen in adults, eye disease was more common among boys (69 %) than girls (42 %). It was mostly bilateral and in the form of posteror or panuveitis. However, eye disease in juvenile BS patients seems to have more favorable outcome compared to adults.

	Right eyes n= 72	Left eyes n = 70
Damage score, mean ±SD	2.0± 1.43	2.0±1.5
Activations, mean ±SD	2.8± 4.8	2.6± 3.8
Retinal vasculitic infiltrations, mean ±SD	0.8± 1.8	0.8±1.7
Hypopyon, n (%)	3 (4)	5 (7)
Papillary stasis, n (%)	10 (13)	10 (13)
Cataract operation, n (%)	1 (1.3)	2 (3)
Phtysis bulbi, n (%)	4 (5)	3 (4)

Book No: O-020
Ref. No: 267
Topic: Paediatric

WORK DISABILITY IN BEHCET'S SYNDROME

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BACKGROUND: Behcet's syndrome (BS) is most active during working years thus affecting productivity. We aimed to survey the frequency and associated reasons of work disability among BS patients.
METHODS: Consecutive patients who attend our BS outpatient clinic for their routine visits were evaluated using a standard questionnaire about their work and education status, modified MDHAQ and their RAPID3 scores (1) were calculated. Unemployment rate was compared with the general population over the age of 25 in Turkey (2).
RESULTS: 216 patients (127 women, mean age 38.7±10.9) were surveyed. 105/216 were not eligible for employment (85 homemakers, 13 retired and 7 students). Among the remaining 111 (78 men, mean age 36.6±8.6) who were eligible for work, 23 (21%; 15 men, mean age 37.7±7.7) were unemployed. Unemployment rate in Turkey as officially announced in March 2009 was 13.6%. All except 1of these patients had previously been employed. 18 (78%) considered their disease as the cause of unemployment (eye involvement in 6, vascular in 5, joint in 3, neurologic in 2, pleural effusion and severe mucocutaneous involvement in 1 patient, each). RAPID3 scores of unemployed patients were significantly higher than employed ones (12.1±5.1 vs 7.9±6.5, p=0.009) while the disease duration was similar between the same 2 groups (10.5±7.5 SD years vs 9.6±7.3 SD years; p=ns)
CONCLUSION: Work disability rate is high (21%) among BS patients. It is associated with major organ involvement and more active disease.
1) Yazici, Y. J Rheumatol. 2008 Apr;35(4):603-9
2) Turkish State Statistical Institute.

Book No: O-021
Ref. No: 60
Topic: Genetics

Methylenetetrahydrofolate reductase (MTHFR) genetic polymorphism in patients with Behcet disease

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Introduction: Increased plasma homocysteine level has been associated with a tendency to induce thrombosis in patients with Behcet disease (BD). The aim of this study was to evaluate four genetic polymorphisms in methylenetetrahydrofolate reductase (MTHFR) gene in BD patients with thrombosis.
Methods: In a case control study 88 BD patients (39 with thrombosis and 49 without vascular involvement) and 49 healthy individuals were included. All patients were fulfilled the ISG criteria for BD. Genomic DNA was extracted by salting out method, and four genetic polymorphisms in MTHFR gene were evaluated (C1727T by PCR-RFLP; C677T, SNPrs45438591 and SNPrs45589033 by sequencing method). Allelic and genotype frequency were compared by chi square ($\chi 2$) test. Plasma homocysteine level was measured by ELISA in all and was compared between the different groups using ANOVA (F-test).
Results: The frequency of TT genotype for C677T polymorphism was 15.4% in BD patients with thrombosis, 4.1% in BD patients without vascular involvement and 8.2% in controls ($\chi 2$=5.66, p=0.22). Homocysteine levels showed no significant relation with CC, CT or TT genotypes both in BD patients with thrombosis (F=0.949, p=0.40) and in those without vascular involvement (F=0.692, p=0.51), as well as in healthy subjects (F=0.509, p=0.61). The frequency of TT genotype for C1727T and AA genotype for SNPrs45438591 and SNPrs45589033 were zero in all three groups.
Conclusion: We have found no significant relationship between above mentioned polymorphisms in MTHFR gene and increased plasma homocysteine level or thrombosis in patients with BD.

Book No: O-022
Ref. No: 129
Topic: Genetics

Impact of HLA-B51(5) on Behçet's Disease Clinical Phenotype: Systematic Review and Meta-Analyses on Phenotype-Genotype Correlates

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Background: HLA-B51(5) is closely linked to Behçet's disease (BD) susceptibility but it remains unclear to what extent this allele also influences the BD clinical phenotype.

Objective: To clarify the relationship between HLA-B51(5) carriage and BD phenotype using meta-analyses.

Methods: Relevant publications were identified by a systematic literature research. Eligible studies had to provide comparative frequencies for at least 1 BD characteristic for HLA-B51(5) carriers and non-carriers. Pooled odds ratios (OR) were calculated for those variables for which at least 10 relevant studies were identified. Computations used random-effects models.

Results: Among the 861 publications evaluated, 73 studies and 7594 BD subjects (including 4321 carriers and 3273 non-carriers of HLA-B51(5)) met eligibility criteria. Pooled OR [95% confidence intervals] for the 10 analyzed variables were: male sex (38 comparisons), 1.44 [1.20–1.73] (P<0.001); genital ulcers (29 comparisons), 1.82 [1.35–2.46] (P<0.001); ocular involvement (47 comparisons), 1.55 [1.23–1.96] (P<0.001); skin involvement (25 comparisons), 1.62 [1.17–2.23] (P=0.004); erythema nodosum (13 comparisons), 1.10 [0.64–1.88] (P=0.74); positive pathergy test (15 comparisons), 1.44 [0.98–2.12] (P=0.06); articular involvement (30 comparisons), 0.92 [0.71–1.19] (P=0.52); central nervous system disease (29 comparisons), 0.91 [0.64–1.30] (P=0.61); gastrointestinal disease (16 comparisons), 0.59 [0.35–1.01] (P=0.05); and thrombophlebitis (18 comparisons), 1.47 [0.88–2.45] (P=0.14).

Conclusion: The results of these meta-analyses indicate that, for BD patients, HLA-B51(5) carriage predominates in men and determines increased propensity for developing genital ulcers, ocular disease and skin involvement. However, the relatively small magnitude of these HLA-B51(5) effects on clinical characteristics suggests that this allele is not a major determinant in the BD clinical phenotype.

Book No: O-023
Ref. No: 185
Topic: Genetics

Genome-wide association studies define two susceptibility loci for Behçet's disease

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Objective: The etiology of Behçet's disease (BD) is currently believed that certain environmental factors are able to trigger the symptomatology in individuals with a particular genetic background. The molecular nature of this "genetic background" remains mostly unknown with the exception of the association with the HLA class I region. To identify susceptibility genes for BD, especially outside the HLA region, we conducted a genome-wide association study (GWAS).
Methods: We conducted a GWAS in the Japanese population (612 BD patients and 740 controls) using 500,568 SNPs from the Affymetrix GeneChip Human Mapping 500K Array Set. After the GWAS, to validate our results, we exchanged data with colleagues performing a GWAS in the Turkish population. Finally we conducted a meta-analysis of loci, which showed strong association in both Japanese and Turkish GWAS, in the Japanese, Turkish and Korean populations.
Results: A total of 35 distinct loci outside the HLA region showed association with the disease at $P < 0.0001$ in Japanese GWAS. By comparing results of Turkish GWAS, we identified two susceptibility loci for BD. A meta-analysis of these two loci in the three populations showed genome-wide significant associations ($P < 5.0\times10^{-8}$).
Conclusions: In this study, we were able to identify several new susceptibility loci for BD. Two loci with the strongest association signals are located within genes involved in the immune response, suggesting that these two immune-related genes are involved in the largely unknown pathophysiology of BD in which no major locus outside the HLA region has been reliably identified.

Book No: O-024
Ref. No: 191
Topic: Genetics

A TWIN STUDY IN BEHÇET'S SYNDROME

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Objectives: We are not aware of formal twin studies in Behçet's syndrome (BS). We sought the frequency of MZ and dizygotic (DZ) twin births in BS and compared it to a healthy population sample. We also looked for the concordance rate among the MZ and DZ twins.
Methods: 1705 (1039 M/ 666 F) patients attending a dedicated BS outpatient clinic and 7761 medical school students were asked about having a MZ or DZ twin sibling. MZ and DZ twins identified among both patients and controls were individually seen at the clinic. All twins were contacted 8 years later for new emergence of disease.
Results: There were 14 (0.82 %) patients with BS and 120 (1.55 %) controls who had a twin sibling ($P = 0.022$). MZ twin frequency was similar between BS patients (6/ 1705; 0.35 %) and control population (28/ 7761; 0.36 %). The pairwise

concordance rate for BS was 2/6 for MZ and 1/8 for DZ twins (P =0.538). After 8 years of follow-up, 4 of 6 MZ and 6 of 7 DZ twin pairs were still discordant.
Conclusions:
The frequency of MZ twin births in BS is not different than that in the general population while the DZ twins were seen less frequently among the BS patients. The concordances for BS were higher in MZ compared with DZ twins, suggesting genetic predisposition. The persistence of discordance after 8 years of follow up among the remaining MZ twins demands further research to understand non- genetic factors.

Book No: O-025
Ref. No: 224
Topic: Genetics

A Genome-wide Association Study Identifies Common Variants of the IL10 and IL23R Genes that Contribute to Behçet's Disease Susceptibility

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Although a genetic contribution to Behçet's Disease (BD) has been well-established, with the exception of HLA-B51, which explains less than 20% of the genetic liability, the identities of specific alleles that are responsible for the complex inheritance of this disease have remained enigmatic. We performed a genome-wide association study (GWAS) using 311,459 informative SNPs in a collection of 1215 BD patients and 1278 healthy controls from Turkey. We also genotyped these cases and controls for HLA-B51. This study confirmed the known strong disease association with HLA-B51 and provided evidence for a second, independent susceptibility locus in the Class I region of the major histocompatibility complex. In addition, we found one SNP with genome-wide evidence for disease association ($P < 5.0 \times 10\text{-}8$) within the gene encoding the immunoregulatory cytokine, interleukin-10 (IL10). A meta-analysis of ethnically matched case/control collections from diverse genetic backgrounds, including a total of 2430 cases and 2660 controls, established associations with the IL10 variant (rs1518111, P = 3.54E-18, odds ratio 1.45 with 95% confidence interval 1.34 to 1.58) and with a variant located between the interleukin-23 receptor (IL23R) and interleukin 12 receptor β2 (IL12RB2) genes (rs924080, P = 6.69E-09, odds ratio 1.28 with 95% confidence interval 1.18 to 1.39). The disease-associated IL10 variant was associated with diminished mRNA expression and low protein production by cells obtained from healthy blood donors, suggesting novel therapeutic targets for BD.

POSTER PRESENTATIONS INDEX

Board Number	Date of Display	Ref. No	Topic	Title
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POSTER PRESENTATIONS

Immunology & Vasculitis will be hang on 08.07.2010
Regional & Paediatric & Genetics will be hang on 09.07.2010

Board No: P-001
Ref. No: 43
Topic: Immunology

Treatment of Neuro-Behcet's Disease with Infliximab. An International Multicentre Case-Series of 18 patients

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Background:
TNF blockers are increasingly reported to be effective and relatively safe for the serious non-neurological complications of BD. Few single case reports and a single centre case series of 8 patients reported favourable outcomes for the use of infliximab (IFX) for neuro-BD (NBD).

Objectives:
To evaluate the use, efficacy and safety of IFX in treating NBD in a multicentre case-series

Methods:
We preformed a retrospective analysis of BD patients treated with IFX for their neurological complications from 15 centres in 10 countries. Information was collected through structured data collection forms. Inclusion criteria required cases to satisfy the ISG criteria for BD, have clear neurological presentations compatible with parenchymal NBD, and have clear data for follow-up and outcome.

Results:
Eighteen (12M, 6F) cases were included. The mean age at NBD presentation was 29.4 yr (17-49yr). The mean duration of IFX use was 20.9 months (range: 2-53). The mean duration of follow-up after starting IFX was 32.8 months (range: 5-76); 12/18 were followed for 2 or more years. Seventeen patients used IFX because of unresponsiveness to other immunotherapies. The outcome was favourable in 17/18; one patient with predominant cognitive impairment did not show improvement. Two patients switched to alternative TNF blockers. No serious side effects were reported except for a patient who was switched to another TNF blocker and developed later radiological signs of CNS demyelination.

Conclusions:
Infliximab seems to be effective when the first line immunotherapies fail to prevent or control parenchymal NBD. CNS demyelination is a potential risk.

Board No: P-002
Ref. No: 58
Topic: Immunology

Therapeutic possibility for patients with Behcet's disease by the peptides of heat shock protein-65/60 derived from oral streptococci

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Behcet's disease (BD) generally starts with oral aphthous ulceration and develops to other organ involvements. BD patients have hypersensitivity against oral streptococci increased in their oral cavity. Heat shock proteins-65/60 (HSP-65/60) derived from S. sanguinis and the damaged mammalian tissues, respectively, are supposed to play a role in the lesions. The antigen presenting cells taken HSP-65 via Toll-like receptors are known to lead T cells undergo apoptosis. The human HSP60 peptide (336-351 aa) combined with recombinant cholera toxin B subunit was reported to be a therapeutic agent for BD patients with advanced uveitis. In our experiments, the peptide (249-264 aa; designated Lo1) of HSP60, that shows highly homologous to the T cell-epitope, induced CD4+, CD8+ T cells apoptosis in peripheral blood mononuclear cells (PBMCs) from BD patients.

To know the effects of the HSP60-peptides, the five peptides (Lo1:249-264, IIIa:365-384, IIIb:395-413, Lo2:480-499, and UK:311-326) were applied, respectively, on BD patients' PBMCs. Also, to understand the reduction of cytokines from the patients' PBMCs, the effects of Lo1 on NOMO-1 cells (human macrophage cell line) stimulated with S. sanguinis antigen were analyzed by the DNA-chip procedure. Although proinflammatory cytokines were naturally produced from the patients' PBMCs, IL-8 production was significantly inhibited by Lo1, IIIa and IIIb, and also IL-12 production was reduced by Lo1, Lo2, Lo3, IIIa and UK. In the NOMO-1 cells, mRNA expressions of IL-16, IL13, IL-28, and IL-17-receptor were reduced, but CD58 and/or FK506 binding protein were enhanced by Lo1, indicating the HSP60-peptides influence the cytokine production.

Board No: P-003
Ref. No: 61
Topic: Immunology

New HLA B allele association to Behçet disease in Moroccan patients

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Human leukocyte antigen HLA-B51 is the most strongly associated gene with Behçet disease (BD) in different ethnic populations. To analyze the influence of HLA-B allele in BD predisposition in Moroccan population and its association with clinical manifestations; the HLA-B phenotype frequencies were analyzed by Serologic HLA class I typing and by polymerase chain reaction sequence-specific oligonucleotide (PCR-SSO) reverse dot blot hybridization in 120 unrelated Moroccan patients; all of whom fulfilled the international study group criteria for Behçet's disease, and in 111 ethnically matched healthy controls.
Besides HLA-B*51(16.66%) and HLA-B*15 (15%) alleles, a significant increased frequency of the HLA-B*27 allele was found in Moroccans patients with Behçet's disease when compared to Controls (13.33% of patients versus 2.7% of controls, chi square = 8.63, OR = 5.54, 95% IC [1.57-19.57] and particularly in the patients who presented a uveitis (23.52 %).

Key words: Behçet's Disease, HLA-B 27, association, Moroccan population

Board No: P-004
Ref. No: 63
Topic: Immunology

Serum leptin level in patients with Behcet disease

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Introduction: Leptin, as a peptide hormone with main effect on modulation of body weight, has also proinflammatory functions and may influence different aspects of immune system. This study was to compare serum leptin level in patients with Behcet disease (BD) with control group, and its potential relation to ocular involvement, disease activity or disease duration.
Methods: In a case control study 105 consecutive BD patients (according to ICBD criteria) and 105 controls were included. Controls were either healthy subjects or those with recurrent oral aphthosis or ocular lesions in whom BD was ruled out, all matched regarding age, sex and body mass index. Serum leptin levels have been measured in all by enzymatic assay (ELISA method) using DBC leptin kits (normal range: 4-19 ng/dL). Disease activity was determined according to physician global assessment (PGA) in BD patients. Comparisons were done by independent t-test between means and by ANOVA (F-test) between groups.

Results: There was no significant difference in the mean serum leptin level between the BD patients and controls (14.5±12.2 vs. 12.9±10.8, p=0.31). It was not related neither to disease duration (F=1.955, p=0.12) nor to disease activity (12.6±11.4 in active vs. 16.8±12.8 in non-active disease, p=0.08) in BD patients. There was also no significant difference between those with and without ocular involvement (13.7±12.8 vs. 15.4±11.6, p=0.46).
Conclusion: Serum leptin level was not higher in our patients with BD, and had no correlation with disease duration, disease activity or ocular involvement in them.

Board No: P-005
Ref. No: 72
Topic: Immunology

Immune and inflammatory gene expressions are different in Behcet's disease compared to Familial Mediterranean Fever

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Aim: Immune classification of Behcet's Disease (BD) is still controversial. In this study, we aimed to compare immune/inflammatory gene expressions of BD with Familial Mediterranean Fever (FMF), an autoinflammatory disorder with innate-immune activation.
Method: Ten BD (F/M: 6/4, age: 36.7), 6 FMF (F/M: 3/3, age: 29.2) patients and 4 controls (F/M: 2/2, age: 32.4) were enrolled. mRNA's were extracted from CD14+monocyte and CD4+T-lymphocyte subsets. Expressions of 440 immune/inflammatory genes were analyzed with a DNA microarray system (OligoGEArray, SABiosciences). Up-(>1,5 fold) and down-regulated (<0,8 fold) gene expressions were presented.
Results: Expressions of CXCR-3 (1.7), IL-7 (1.7) and prokineticin-2 (1.7) were higher in BD compared to FMF group in CD4+T-lymphocytes. In CD14+monocytes, CCL3 (2.7), CCL5 (1.5), IL-8 (2.3) and TNF-α (1.7) were up-regulated, whereas CX3C-R 1 (0.6), TLR-2 (0.7) and TNF ligand (0.6) were down-regulated.
When compared to controls, CCR1 (2.3) expression in BD and CCR1 (1.6), CCR7 (1.7), TGF-β-RII (1.5) and TLR2 (1.5) expressions in FMF group were higher in CD4+ T-lymphocytes. On the other hand, allograft inflammatory factor 1 (0.6), FOS (0.3) and lactoferrine (0.3) levels of BD and CCL3 (0.6), C/EBPb (0.6) CSF-3 receptor (0.6) and FOS (0.4) levels of FMF group were lower. CCL3 (2.7), CCr1 (1.7), FOS (1.5), IL-8 (1.8) and TNF-α (1.8) were also upregulated in BD compared to controls in CD14+monocytes.
Conclusion: Immune/inflammatory gene expressions were differently present in BD compared to FMF, suggesting that "auto-inflammation" is possibly not the sole mechanism driving the immune activation in BD.
Key words: Behcet's Disease, Familial Mediterranean Fever, inflammation, gene expression.

Board No: P-006
Ref. No: 78
Topic: Immunology

Effect of intracameral triamcinolone acetonide irrigation on postoperative inflammation and intraocular pressure after cataract surgery in patients with Behcet Disease

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Purpose: To evaluate the effect of intracameral triamcinolone acetonide (TA) irrigation on postoperative intraocular pressure (IOP) and controlling ocular inflammation in patients with Behcet Disease undergoing cataract surgery.

Methods: Twenty eyes of 20 patients with Behcet Disease undergoing cataract extraction with phacoemulsification at the Department of Ophthalmology, Eskisehir Osmangazi University Medical Faculty. The patients were randomized into two groups. Eyes in group 1 (10 eyes) irrigated of TA into the anterior chamber, but eyes in group 2 (10 eyes) did not. TA irrigation was done before intraocular lens implantation. 4 mg/0.1 ml TA was injected into the anterior chamber via sideport. TA was stayed twenty seconds in the anterior chamber. and then the anterior chamber was cleaned with balanced salt solution. Postoperatively; in two groups, topical dexamethasone 1% eye drops were administered six times per day for 7 days, then four times per day for 30 days, to control postoperative inflammation. Anterior chamber cells, anterior chamber flare, fibrinoid reaction, visual acuity (VA), and IOP measurements were evaluated preoperatively and at 24 h, 1 week, and monthly until 6 months postoperatively.

Results: There were no statistically significant differences in mean VA and mean IOPs between two groups. Anterior chamber cells, flare and fibrinoid reaction were seen less in group 1 than 2. ($p<0.05$)

Conclusion: Intracameral irrigation of TA after phacoemulsification surgery in patients with Behcet Disease was found to be an effective procedure to control postoperative inflammation. This procedure had no significant negative effects on postoperative VA and IOP.

Board No: P-007
Ref. No: 79
Topic: Immunology

PATHERGY REACTION IN BEHCET DISEASE:Past and Present (An Overview)

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Pathergy reaction(PR)is an intersting and enigmatic phenomenon observed in Behcet disease(BD). The aim of this presentation is to summarize our experience and literature.Pathergy(P) term was coined by Rössle in 1933 and Blobner observed PR in an patient with recurrent hypopion iritis in 1937. In 1941 Jensen applied the skin pathergy test(SPT) in an patient with BD and named it as "needle-prick test". Berlin,and Nazzaro in 1960 mentioned its absolute specificity in 1960.SPT was included into the diagnostic criteria of Japan in 1972 and later on into the others and ours too. In 1985 London Conference we proposed the followings:high specificity for BD;the advantages of using multiple pricks and thicker needles;no good correlation between SPT and clinical features;reading time of SPT must be 48 hr;its heterogeneity and convertibility;it is a genetic marker.PR was elicited and observed in different organs too,by various authors. Despite to the excellent investigations the exact etiopathogenesis of PR and BD have not been elucidated yet.Conclusions: we think that PR is an exagerated model of the reaction induced in the healthy subjects. We should try to increase its sensitivity by augmenting the traumatic character of the needles and method further. We believe that at present SPT remains one of the most practical and reliable method in the diagnosis of BD.

Board No: P-008
Ref. No: 80
Topic: Immunology

The Immunoregulatory Effects of Interferon-α therapy on T cell responsiveness in Ocular Behçet's disease
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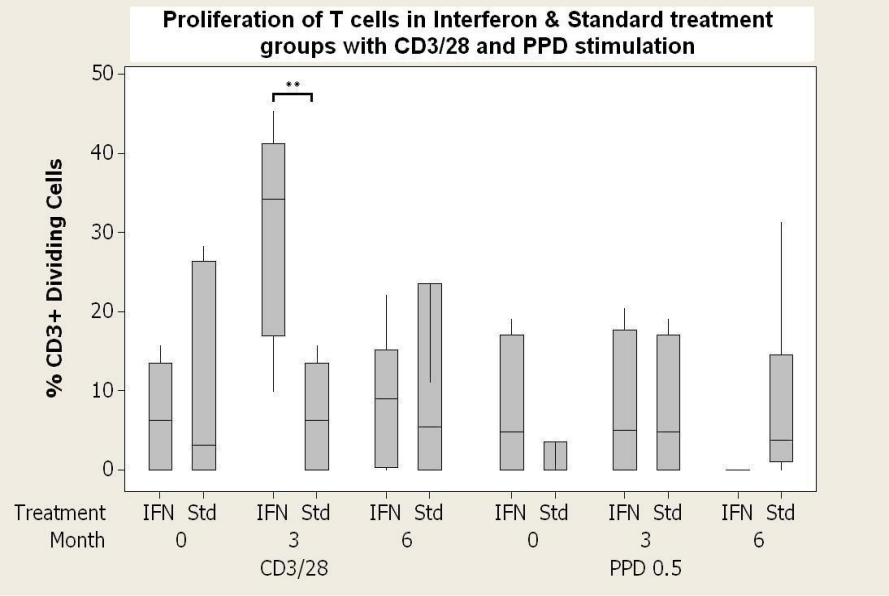
Aims: As part of an ongoing randomized clinical trial of Interferon-α2b therapy in patients with ocular Behçet's disease (BD), we aimed to investigate the effects of Interferon therapy on T cell responsiveness ex vivo.

Methods: Blood samples were collected at 0, 3, 6 months from BD patients during which patients received either standard therapy (prednisolone + cyclosporin; n=6, mean age=33.67, 5 male), or standard therapy with pegylated Interferon-α2b (IFN α2b; once weekly for 3 months; n=6, mean age=40.17, 4 male). To determine the antigen responsiveness, whole blood was exposed to different stimuli (anti-CD3/28, purified protein derivative (PPD), α-tropomyosin, retinal soluble antigen (sAg) peptide, heat shock protein 65 and, at 5 days, dividing cells were identified by coexpression of propidium iodide and PCNA, using flow cytometry.

Results: Increased levels of T cell proliferation were detected in response to anti-CD3/28 stimulation in those receiving IFN α2b at 3 months ($P<0.05$) in comparison to those receiving standard therapy alone. This increase was not observed for any other stimuli and was not detected at 6 months post therapy. Intracellular cytokine staining showed increased expression of IFN gamma in CD3+ T cells in the IFN α2b treated group at 3 months when compared to the standard group ($p<0.01$).

Conclusions: IFN α2b is a recognised therapy for Behçet's disease. An increase in T cell proliferation in response to anti-CD3/28 stimulation combined with an increase in IFN gamma expression was observed in the IFN α2b-treated group at 3 months', suggesting an upregulation of Th1 cells is occurring in response to treatment.

Proliferation of T cells in Interferon and Standard treatment groups with CD3/28 and PPD stimulation



This graph clearly demonstrates the increased proliferation of T cells with CD3 stimulation seen in the Interferon treated group at 3 months. This effect was temporary and was not seen with any other form of stimulation

Board No: P-009
Ref. No: 99
Topic: Immunology

Complement pathways involved in the inflammatory central nervous system disorders

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The complement system is known to play an important role in the pathogenesis of inflammatory central nervous system disorders. To investigate the involvement of different complement pathways in these disorders, levels of breakdown products for classical (C4d), alternative (FBb) and common (sC5b-9) pathways were measured in the sera of neuro-Behçet's disease (NBD) (n=30), neuromyelitis optica (NMO) (n=28), multiple sclerosis (MS) (n=29) patients and healthy controls (HC) (n=36) by ELISA. Both the classical and alternative pathways were activated in NBD, only the classical pathway was activated in NMO and none of these pathways were activated in MS patients and HC. Complement breakdown product levels were comparable among aquaporin-4 antibody positive versus negative NMO and parenchymal versus vascular NBD patients. While MS patients with lower EDSS scores had significantly higher complement breakdown product levels, a similar association could not be shown for NBD and NMO patients. Our results suggest that NBD and NMO differ from MS by the predominance of complement system involvement as a pathogenic mechanism. Since antibodies are the major activators of the complement system in autoimmune disorders, antibody-mediated tissue destruction appears to play a significant role in NBD pathogenesis. Increased classical pathway activation in aquaporin-4 negative NMO patients suggest that non-aquaporin-4 specific antibodies could be found in these patients' sera. The complement activation appears to be somewhat involved in MS pathogenesis in the earlier inflammatory phase of the disease, whereas it regresses as the degenerative phase ensues.

Board No: P-010
Ref. No: 105
Topic: Immunology

TIM3-TIM3 Ligand interaction ameliorate Herpes Simplex Virus-induced Behcet's Disease-like symptoms
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Tim3-Tim3L interaction serves as a specific down-regulator of the Th1 immune response. Galectin-9 (Gal-9) was identified as a Tim3 ligand. We found that Tim3-Tim3L interaction would affect symptoms in Herpes Simplex Virus induced Behcet's Disease (BD)-like mice. The expression of Gal-9 in macrophages of BD-like mice was lower than control mice. So we injected 100 μg of Gal-9 to BD-like mice for 5 times with 3 days interval and subsequently observed the change of symptoms for 15 days. The effect of Gal-9 was the improvement of symptoms, the decrease of severity score, and the increase of regulatory T cell expression. Pro-inflammatory cytokine levels were decreased in Gal-9 treated group than control group. Gal-9 appeared to play an important role of recovery in induced BD symptoms through up-regulation of regulatory T cell and down-regulation of pro-inflammatory cytokines. Therefore administration of Gal-9 was effective in recovery of symptoms in BD-like mice.

Board No: P-011
Ref. No: 106
Topic: Immunology

Vitamin D3 ameliorated the inflammation in herpes simplex virus induced Behcet's disease-like mouse model through down-regulation of Toll-like receptors
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The purpose of this study is to understand the role of vitamin D3 through regulation of TLRs in herpes simplex virus induced Behcet's Disease (BD)-like mice. Serum 25-hydroxyvitamin D levels were lower in BD mice (12.4±5.4 ng/ml) than in asymptomatic BD normal mice (BDN, 17.5±7.2 ng/ml). TLR2 and TLR4 was expressed higher level in macrophages of BD mice. To see the effect of vitamin D3 in vitro, peritoneal macrophages were isolated from normal and BD mice and then incubated with 10-9 M and 10-8 M of 1,25-Dihydroxyvitamin D3 (1,25(OH)2D3) for 24 to 72 h. The frequencies of TLR2 and TLR4 were down-regulated in 1,25(OH)2D3 treated macrophages. To demonstrate the effect of 1,25(OH)2D3 in improvement of BD symptoms, 1,25(OH)2D3 was orally once daily administered for 10 days. 1,25(OH)2D3 improved the mucocutaneous symptoms in 6 out of 11 BD mice, decreased the BD severity score from 2.36±0.4 to 2.09±0.8, and down-regulated the frequencies of TLR2 and TLR4. 1,25(OH)2D3 also downregulated the cytokine levels in sera of BD mice. Interleukin-6 was changed from 434.3±94.1 ng/ml to 94.1±45.8 ng/ml and TNF alpha was from 100.1±5.1 pg/ml to 50.5±2.5 pg/ml. Vitamin D affected the improvement of HSV induced BD-like symptoms by down-regulating the expression of TLRs and pro-inflammatory cytokines in vivo mouse models.

Board No: P-012
Ref. No: 110
Topic: Immunology

Joint involvement and anti-cyclic citrullinated peptide antibodies in Behçet's disease
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Background: We aimed to determine the prevalence of anti-CCP antibodies in a large group of Korean patients with Behçet's disease (BD), with and without joint involvement, and to compare these findings to the prevalences of anti-CCP antibodies in patients with rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE).

Methods: We tested 189 patients with BD, 105 with RA, and 36 with SLE for anti-CCP antibodies and IgM rheumatoid factor (RF) in serum. We reviewed the medical records of patients with BD to investigate their personal and clinical characteristics and laboratory test results.
Results: Anti-CCP antibodies were detected in seven of the 189 BD patients (3.7%), at a mean titer of 30.6 ± 44.4 U/ml, in 86 of the 105 RA patients (81.9%) with a mean titer of 198.8 ± 205.7 U/ml, and in nine of the 36 SLE patients (25%) with a mean titer of 180.4 ± 113.9 U/ml. Five of the seven anti-CCP-positive BD patients (71.4%) had polyarticular joint involvement, and two patients (28.6%) had oligoarticular involvement.
Conclusion: We determined the prevalence of anti-CCP antibodies in a large group of Korean BD patients with and without joint involvement and compared these findings with the prevalences of anti-CCP antibodies in RA and SLE patients. The typically negative anti-CCP test in patients with BD may help to differentiate BD from RA and SLE that present with similar clinical features.

Board No: P-013
Ref. No: 112
Topic: Immunology

HnRNP-A2/B1 as a Target Antigen of Anti-Endothelial Cell IgA Antibody in Behçet's Disease
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BACKGROUND Streptococcus sanguis has been proved its major role in immunopathogenesis of Behçet's disease (BD) which mainly inhabit the mucous membrane of the mouth, throat, colon, and female genital tract. Because IgA is responsible for mucosal immunity, the aim of our study was to identify the anti-endothelial cell IgA antibody-binding human dermal microvascular endothelial cell (HDMEC) antigen.
METHODS We detected a target protein by using Western blotting and immunoprecipitation, and then searched for a similar protein after the amino acids were sequenced by nanoflow liquid chromatography/electrospray ionization/tandem mass spectrometry (LS-EOI-MS-MS) analysis. We next searched for the DNA sequence of the target protein at the National Center for Biotechnology Information and purified the recombinant target protein by gene cloning. We then investigated the reactivities of the recombinant target protein in BD.
RESULTS A 36-kD protein band was detected in all seven patients' serum samples by Western blotting of extracts of HDMECs. The 36-kD protein band obtained from immunoprecipitation was excised and analyzed by nanoflow LS-EOI-MS-MS and a peptide fingerprint was obtained. We found that the protein band showed the amino acid sequences of hnRNP-A2/B1. Reactivity to hnRNP-A2/B1 was detected in 35 of 44 BD patients (79.5%) on Western blots, whereas none of 20 healthy controls and 13 IgA nephropathy patients presented reactivity against hnRNP-A2/B1.
CONCLUSIONS The hnRNP-A2/B1 protein is the target protein of serum anti-endothelial cell IgA antibody in BD patients. This is the first report of the presence of IgA antibodies to hnRNP-A2/B1 in endothelial cells from the serum of BD patients.

Board No: P-014
Ref. No: 115
Topic: Immunology

Effect of interferon-alfa 2a therapy on peripheral blood CD4+CD25+ T regulatory cells in patients with Behçet's uveitis
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Purpose: This study was designed to evaluate the phenotypical and functional effect of interferon alfa-2a (IFN α-2a) therapy on peripheral blood CD4+CD25+ T regulatory (Treg) cells in patients with Behçet's uveitis.
Methods: Blood samples were taken from 5 patients with Behçet's panuveitis thrice: (1) before initiation of any systemic therapy while the patients had active panuveitis, (2) at the termination of conventional immunosuppressive agent due to unresponsiveness of uveitis/before initiation of IFN α-2a, and (3) while panuveitis was inactive under IFN α-2a. Five age-matched healthy subjects were also studied. For functional analysis Treg cells were separated by means of magnetic-assisted cell sorting and subsequently co-cultured for 3 days. The levels of IFN-γ, TNF-α, IL-4, IL-17, IL-18 and IL-10 in the

supernatants were determined by ELISA.

Results: Our data showed a slightly elevated percentage of Treg cells in patients with active Behçet's uveitis when compared with healthy controls (median±SEM; 5.30±4.09% vs. 5.10±1.21%). The intensity of Foxp3 expression of Treg cells indicated by mean fluorescence intensity was also slightly elevated in patients with active uveitis (median±SEM; 543.0±67.79 vs. 473.0±32.85). IFN α-2a led to a borderline significant decline of Treg cells [median±SEM; (1): 5.30±4.06% vs. (3): 0.80±0.87%, p=0.06] and elevation of IL-10 [median±SEM; (1): 0.12±0.45 pg/ml vs. (3): 2.39±0.51 pg/ml, p=0.06].

Conclusions: IFN α-2a therapy led to a decline in the dysfunctional CD4+CD25+Foxp3+Treg cell population. IL-10 may play a major role in IFN α-2a mediated control of Behçet's uveitis.

Board No: P-015
Ref. No: 119
Topic: Immunology

Differential Expression of T cell Immunoglobulin- and Mucin- Domain-Containing Molecule-3 (TIM-3) According to Activity of Behçet's disease

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Objective: T cell immunoglobulin mucin-3 (TIM-3) is recently described as a TH1-associated cell surface molecule that regulates TH1 responses. Increased TH1 immune response is one possible pathogenesis of a chronic inflammatory disorder, Behçet's disease (BD). This study aimed to examine the expression of TIM-3 in BD patients and to determine if TIM-3 expression is correlated with disease activity.
Methods: Behçet's patients (n=67), healthy control (13) and psoriasis patients (14) were involved. The frequency of TIM-3 in PBMCs was analyzed by flow cytometry. The expression of TIM-3 protein was evaluated by Western blotting. TIM-3 expression in cutaneous lesion was revealed by immunohistochemistry
Results: TIM-3 expression in BD patients was significantly up-regulated compared with those in controls presented by flow cytometry and Western blot. The frequency of TIM-3 in CD8+ and CD56+ cells was significantly increased in BD and immunohistochemistry showed positivity for TIM-3 was co-localized with CD4+ cells. Active BD group showed decreased TIM-3 expression compared to stable group. In the same patient, TIM-3 expression was increased after turning into stable state compared to those in active state. After disease stabilization in the same patient, TIM-3 frequency on each leukocyte subpopulations was increased, especially on CD4+ and CD56+ cells.
Conclusion: This study may imply the differential expression of human TIM-3 molecules in the PBMCs of TH1-driven Behçet's disease according to disease activity and suggest that there were altered kinetics in the expression of TIM-3 molecule that might modulate immunologic response in Behçet's disease.

Board No: P-016
Ref. No: 137
Topic: Immunology

Infliximab therapy may greatly change ocular inflammation and visual prognosis in Behcet's disease

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Objectives: Infliximab has been used in Behcet's disease to try to control refractory ocular inflammation. In this study, we systematically analyzed the clinical effects of infliximab in a large number of Behcet's disease patients with recurrent ocular inflammation.
Methods: Subjects included a total of 50 Behcet's patients with refractory ocular inflammation being followed at 8 uveitis specialty clinics in Japan. Infliximab was administered intravenously at 0, 2 and 6 weeks, and every 8 weeks thereafter. Clinical findings of the patients during the 6 months just prior to starting infliximab therapy were compared to that during the

first 6 months on infliximab.
Results: Over the first 6 months on infliximab therapy, 66% of patients had marked efficacy, 20% moderate efficacy, 12% no efficacy, and 2% worsening. The average number of ocular attacks per 6 month-period decreased from 2.6 before starting infliximab to only 0.44 while on infliximab therapy. of the 50 patients, 38 (76%) had zero recurrences while on infliximab therapy. At least one adverse effect was observed in 19% of patients, including tonsillitis, upper respiratory symptoms and infusion reactions, however none were serious.
Conclusions: Similar to previous reports, we found infliximab therapy to be safe and effective for refractory ocular inflammation in Behcet's disease. Since the typical clinical features of Behcet's disease such as hypopyon iridocyclitis and recurrent uveoretinitis appeared to be strongly suppressed, we believe that infliximab therapy may greatly change the course of ocular disease and the visual prognosis of patients with Behcet's disease.

Board No: P-017
Ref. No: 138
Topic: Immunology

Brain MRI imaging findings in Behcet's disease patients with neurological involvement
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Objects : We assessed the MRI imaging and clinical manifestations in BD patients with central nervous involvement.
Methods: Brain MRI imaging were conducted in 27 BD patients (16 male, 11 female) who received care in Yokohama City University Hospital from 1991 to 2007. The imaging conditions included T1 WI, T2WI, and FLAIR images. We analyzed the abnormal imaging findings and neurological manifestations.
Results: A total of 77 abnormal signals were identified by any imaging conditions in the 27 patients. The lesions were distributed in the cerebrum (77.8%), the brain stem (59.3%), basal ganglia (37.0%) and cerebellum (22.2%).
Sporadic T2 high intensities were most commonly found in the cerebral subcortex. Atrophic changes were found in the cerebellum and cerebrum in 8 (38.1%) and 3 (14.2%) of 27 patients, respectively. of neurological clinical features, cranial nerve involvement was more common in patients with the brainstem lesions than the others (p=0.027). Cerebellar and/or cerebral brain atrophy was associated with personality changes (p=0.005) and speech disorder (p=0.018). The other symptoms such as headache and meningeal signs were not related with any particular abnormal MRI findings. There was no significant association of abnormal MRI findings with gender, age and HLA-B phenotype.
Conclusions: MRI is useful to detect the brainstem lesions, which are responsible for cranial nerve injury, and parenchymal brain atrophy, leading to personality changes and speech disorder in patients with BD.

Board No: P-018
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Topic: Immunology

INFLIXIMAB TREATMENT FOR UVEITIS IN PATIENTS WITH BEHCET'S DISEASE
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Objectives: We evaluated clinical efficacy and safety issues in infliximab (IFX) for Behcet's disease (BD) patients.
Methods: We studied 13 BD patients receiving IFX for refractory uveitis to therapies with colchicines, cyclosporine A, and prednisolone. IFX was given at 0, 2, and 6 weeks and thereafter every 8 weeks. Frequency of ocular attacks and visual acuity were assessed.
Results: IFX therapy commenced 7.4+6.0 years after disease onset. Post-therapy observation period was 1.5+0.8 years. Attack frequency was reduced from 2.4 + 0.6/6 months before therapy to 0.9 + 0.9/6 months after the therapy. Since most ocular attacks were found 6 weeks post-infusion, intervals were shortened from 8 to 6 weeks in 5 patients. Visual acuity was improved in 14 eyes, unchanged in 5 eyes, and deteriorated in 6 eyes. Early introduction of IFX therapy led to good visual

prognosis. Surgery was performed in 3 eyes for cataract and glaucoma, respectively, without operation-related ocular attacks. Serious adverse events including tuberculosis were not observed except for cytomegalovirus infection in a patient.
Conclusions: IFX therapy significantly suppresses ocular attacks, promoting improved visual acuity in BD patients with uveitis. Particularly, early introduction of IFX therapy is encouraged in BD patients with serious uveitis.

Board No: P-019
Ref. No: 145
Topic: Immunology

Unbalanced T cell response in Behçet's disease
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Purpose: The interleukin (IL)-23/IL-17 pathway has been demonstrating to be upregulated in certain inflammatory diseases. This study was designed to contribute to understanding the pathogenesis of Behcet's disease (BD) from a view of the pathway.
Methods: mRNA of helper T cell related cytokines (IL-10, IL-17, IL-17F, IL-21, IL-22, IL-23, IFN-gamma), TGF-beta/Smad and IL-6/STAT signaling proteins in peripheral blood mononuclear cells (PBMCs) of four active BD patients were examined using quantitative RT-PCR, compared with those of healthy volunteers. We studied the expression of INF-gamma, IL-17 and TGF-beta on infiltrating T cells into BD patients' skin lesions.
Results: We didn't detect mRNA expressions of almost all Th17 cytokines in BD's PBMCs before lectin stimulation. After stimulation, mRNA expression of Th1 and Th17 cytokines were increased to some degree in BD. TGF-beta receptor 1, 2 and Smad2 ($p<0.03$) mRNA expressions were accelerated clearly in BD's PBMCs, while both ROR-C and Foxp3 had decreasing trends. Infiltrating CD4 T cells on BD skin lesion showed the co-existence of INF-gamma, IL-17 and TGF-beta.
Conclusion: We revealed the impairment of cell signaling and the disorder in immuno-reacting cytokine production by BD T cells. We speculate that imbalanced T cell response may play a critical role in the development and maintenance of recurrent acute inflammation in BD.

Board No: P-020
Ref. No: 150
Topic: Immunology

Inducible human heat shock protein (HSP70) is elevated in the saliva of Behçet's patients.
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Objectives: Inducible human HSP70 was measured in serum and saliva of Behcet's patients and healthy volunteers (HV). Human antibodies specific for HSP70 were also investigated.
Methods: Saliva samples were collected from BD patients (n=57) and HV (n=19). Saliva was centrifuged and stored at -200C. Plasma samples (BD, n=25; HV, n=12) were separated from whole EDTA blood and stored at -200C. Inducible heat shock protein was assayed in saliva and plasma using a high sensitivity EIA kit. IgG antibodies were analysed by indirect ELISA using recombinant human HSP70. The results are expressed as end point titre and statistical analysis was carried out using one way ANOVA, Student's t test and Mann-Whitney-U.
Results: The level of HSP70 in BD saliva was significantly higher than in HV ($p<0.001$) when measured by ELISA or by qualitative western blot analysis. There was no significant difference between the levels of HSP in the serum of BD vs. HV ($p=0.9015$). Similarly, the levels of HSP 70 in HV saliva and serum were not significantly different. There was a marked elevation in the BD saliva compared with serum ($p<0.0001$). IgG antibody responses to recombinant Human HSP 70 were also significantly elevated in BD vs. HV ($p<0.001$) and showed an interesting seasonal variation with peak levels in spring.
Conclusion: This is the first evidence that inducible HSP70 is elevated in the saliva of BD patients compared with HV and may contribute to a pro-inflammatory milieu driving the oral manifestations of the disease.

Board No: P-021
Ref. No: 152
Topic: Immunology

Infliximab therapy for intestinal Behcet's disease in Japan
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Objects : We examined clinical efficacy and safety issues in infliximab therapy for patients with intestinal Behcet's disease in Japan.
Methods: We retrospectively collected clinical data of 121 patients who had received infliximab therapy more than once for intestinal Behcet's disease from 38 institutes in Japan by using questionnaires. of them, 89 patients (male 45, female 42, 43.3 + 14.3 y.o) who met the Japanese criteria revised in 1987 were included in the study.
Results: Patients presented with abdominal pain (88%), diarrhea (55%), and bleeding (51%). The lesions were mainly distributed in ileum (84%), cecum (45%), and ascending colon (34%). Previous therapies included corticosteroids (83%), mesalazine (69%), colchicine (55%), and any immuosuppressants (70%). Surgical operation was conducted in 31 patients (35%). IFX (3 to 5 mg/kg) was give one to 48 times. Nineteen patients received IFX more than 20 times over three years. Subjective improvement was noted in 81% of patients. Endoscopic improvement and/or steroid sparing effect were confirmed in 52%. Favorable responses were found in patients having ocular lesions and arthritis, latter of which was proven as an independant factor by multivariate logistic regression analysis, whereas esophageal lesions gave a negative impact on the outcome. Eleven adverse events were noted, including 8 infections. The therapy was discontinued in 24 patients (27%) because of remission (5 patients), adverse events (10 patients) and exacerbation or insufficient efficacy (9 patients).
Conclusions: IFX therapy showed favorable clinical outcomes even in Behcet's disease patients who had refractory intestinal lesions to conventional therapies.

Board No: P-022
Ref. No: 169
Topic: Immunology

The Oral Health and Microbiota of UK Behcet's Disease patients
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Objectives: Investigating the oral health and microbiota in Behcet's disease (BD).

Methods: The oral health of 22 BD and 17 age and sex matched healthy controls (HC) was assessed by using decayed, missing and filled teeth index (DMFT), plaque index (PI), gingival index (GI), sulcus bleeding index (SBI), periodontal pocket depth (PPD) and attachment loss (AL).

Saliva and oral swabs (Copan, UK) collected from the same cohort were cultured on blood agar, chocolate agar, colistin nalidixic acid agar, MacConkey agar, gonococcus (GC) agar, sabouraud agar and fastidious anaerobic agar. Plates were incubated at 37°C for 48 hours in CO2, 37°C for 48 hours in O2 and 37°C for 7 days anaerobically. Isolated colonies were purified and extracted peptides were subjected to MALDI-TOF analysis for bacterial identification (Bruker Daltonics, Germany).

Results: 22 BD (F/M: 12/10, mean age: 44.5±13.6) and 17 HC (F/M: 7/10, mean age: 41.9±13.1) were included in the oral health assessment. The BD cohort had statistically higher DMFT ($p=0.039$), SBI ($p<0.0001$), GI ($p=0.004$) and AL ($p=0.004$). From 36 samples collected (9BD, 8HC) 48 different oral microbial species were isolated giving a mean of 6.75±2.156 different species from each sample. There was higher colonization of the oral cavity of BD with candida albican ($p<0.05$).

Conclusion: The BD cohort showed less favorable oral health status in comparison to HC. However, their oral microbiota was comparable to the HC except they exhibited higher candidal species colonization. This might indicate a defect in the microbial defense mechanisms in BD.

Board No: P-023
Ref. No: 178
Topic: Immunology

Why does Behcet's disease decline in Japan? -Possible association between economic development and decreased risk of Behchet's disease

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Objectives: Behcet's Disease (BD) is mainly present along the ancient Silk Road. It has been reported that the number of new patients is gradually declining and the disease is becoming milder only in Japan. In this study, we examined why BD is in decline in Japan.
Methods: To explore a possible link between economic change and decreased risk of BD, we examined the relationship between the number of new patients reported in Inaba's report, 2005 (BD and other 46 intractable diseases) and GDP in Japan. Subsequently, we examined oral health behavior and eating habit. Results: Decline in new cases of BD was still seen in Hokkaido University, Japan in 2005-2008. GDP was significantly associated with the number of new patients with BD from 1955 to 2004 ($P<0.01$, $r=0.9503$) and from 1970 to 2004 ($P<0.01$, $r=0.9170$). Increase in various intractable diseases was also associated with GDP. Recently, tooth brushing custom changed in patients with BD in Japan. New market of drinkables, especially green tea, is associated with GDP. Human behavior could bring the changes of oral microbial flora, which is suggested as an initiating factor of BD. Conclusions: There was a clear link between the incidence of BD and GDP in Japan. GDP brings better oral health behavior and eating habit. GDP estimates the basis of systems of national account and it is possible to compare this among various countries. We can speculate that increase in GDP and better oral health behavior may decrease the incidence and severity of BD.

Board No: P-024
Ref. No: 179
Topic: Immunology

Expression of Th17 and related cytokines according to clinical activity of Behcet's disease

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Background: Recent studies have shown that IL-17 is an important proinflammatory cytokine and is upregulated in certain autoimmune and inflammatory disease. Previous studies on Behcet's disease (BD) showed elevate production of IL-23, IL-17, and IFN-g by PBMCs and increased frequencies of IL-17-producing and IFN-g producing T cells in BD patients with active uveitis.
Purpose: We investigated Th17 and related cytokine expression pattern according to clinical disease activity.
Material and Methods: Flow cytometry was used to evaluate the frequencies of IL-17-producing and IFN-g-producing T cells and the expression of CD45RO. The levels of IL-17, IL-23 and IFN-g in sera or peripheral blood mononuclear cells (PBMCs) were detected by ELISA. IL-17, IL-23p19 and IL-12/23p40 mRNA in PBMCs was examined using RT-PCR.
Result: The frequencies of IL-17-producing and IFN-g-producing T cells from PBMCs were significantly upregulated in active BD patients. IL-17-producing CD4+CD45RO+ memory T cells were higher in active BD patients.
Conclusion: We confirmed previous findings that Th17 is associated in the pathogenesis of BD. Further study to investigate variations in Th17 and related cytokines during deterioration or improvement in the patient's clinical condition is ongoing.

Board No: P-025
Ref. No: 180
Topic: Immunology

Expression of the NALP3 inflammasome in skin lesions of Behçet's disease patients
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Introduction: Necrotic cells release endogenous molecules, such as heat shock proteins that alert the innate immune system of the danger associated with tissue damage and infection. NALP3, a pattern recognition receptor, recognizes messenger molecules generated inside cells in response to these insults. After activation of the NALP3 inflammasome, subsequent release of pro-inflammatory cytokines such as interleukin-1β (IL-1β)occur.
Purpose: As there is strong evidence for pro-inflammatory role of IL-1β in Behçet's disesase (BD), we explored the expression of the different components of the NALP3 inflammasome in tissue specimens from patients with BD.
Method: The expression of NALP3, apoptosis-associated speck-like protein containing CARD domain (ASC) in erythema nodosum lesions of 40 BD patients and 10 non-BD patients were examined by immunohistochemistry.
Result: The expression of NALP3 and apoptosis associated speck-like protein containing a CARD domain (ASC) showed higher tendencies in BD patients. NALP3 was expressed in CD68+, MPO+, CD31+ cells. These results may indicate that macrophages and neutrophils are the principle sources of inflammasome-mediated IL-1β production in BD patients.
Conclusion: This study reveals a role for the NALP3 inflammasome complex in BD. Inhibition of the NALP3 inflammasome activity may delay the inflammatory process and chronic morbidities noticed in patients with BD. Further study on the mechanistic insight to the NALP3 mediated modulation of host immune responses is underway.

Board No: P-026
Ref. No: 186
Topic: Immunology

IL-17A Plays An Important Role In The Acute Attacks of Behçet's Disease
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Th17 cells and IL-17 has recently been reported to be responsible for the enhanced inflammatory reaction in Behçet's disease (BD) patients with active uveitis.
We particularly aimed to investigate the role of IL-17A in the activity and different organ involvements of BD in 45 patients (24 women, 21 men; mean age, 40 years) and 33 age- and sex-matched healthy controls (HC) (17 women, 16 men; mean age, 36). Serum IL-17A levels were examined by ELISA. In vitro IL-17A response of PBMC of BD patients and HC after stimulation with S. sangius, E. coli and PHA were evaulated by ELISPOT. The proportion of IL-17A secreting cells was detected by flow cytometry.
Active BD patients showed significantly higher levels of IL-17A compared with inactive patients and HC ($p<0,001$). IL-17A levels of BD patients with active stages of uveitis ($p=0,003$), oral ulcers ($p<0,001$), genital ulcers ($p<0,001$) and articular symptoms ($p=0,002$) were significantly higher than patients with inactive stages of these symptoms. A significant increase was observed in the number of IL-17A producing cells obtained from BD patients after stimulation with S. sangius ($p=0,003$), E. coli ($p<0,001$) and PHA ($p=0,008$). There was a significant increase in the percentage of IL-17A, CD4(+) IL-17A(+) and CD4(-) IL-17A(+) T cells after E. coli ($p=0,004$) and PHA ($p=0,006$) stimulation.
Taken together, our results indicate that Th17 ve IL-17 pathway are active in BD patients, and play an important role particularly in acute attacks of the disease.

Board No: P-027
Ref. No: 198
Topic: Immunology

Title: Psychopathology in Behcet's disease
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Objective: To characterize depression and anxiety in Behcet's disease (BD) and to investigate possible associations with demographic, clinical and quality of life variables.
Patients and methods: The Hospital Anxiety and Depression Scale (HADS) and the SF-36 were administered to 52 BD patients in the nonactive phase of their illness (73.1% women; mean age=43.25, s.d.=11.72; mean education=9.48, s.d.=4.34; mean disease duration=13.92 years, s.d.=8.27; 28.8% with CNS involvement) selected from the CHP's Clinical Immunology Unit. Both HADS sub-scales were calculated and a cut-off score of 11 was used to classify pathology. The eight scales that comprise SF-36 were calculated. Non-parametric tests (Mann-Whitney, Chi-square or Fisher's Exact) were applied for group comparisons.
Results: The frequency of pathological anxiety was 40.4% and pathological depression was 19.2%. The classification of pathological anxiety and pathological depression was not associated with gender, age, education, CNS involvement, disease duration, or current prednisolone intake. Patients with higher levels of anxiety had poorer quality of life on the following SF-36 indexes: bodily pain ($p=0.031$), general health ($p=0.003$), vitality ($p=0.006$), social functioning ($p<0.001$), role emotional ($p<0.001$), and mental health ($p<0.001$). Pathological depression was associated with poorer physical functioning ($p=0.003$), general health ($p=0.03$), vitality ($p<0.001$), social functioning ($p=0.001$), role emotional ($p=0.001$), and mental health ($p=0.001$).
Conclusions: Elevated levels of anxiety and depression were found to be associated with the patient's quality of life reports but not with the analyzed demographic (i.e., gender, age, and education) and clinical variables (i.e., CNS involvement, disease duration, or current prednisolone intake).

Board No: P-028
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Topic: Immunology

Biomarkers for Adamantiades-Behçet's disease
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Objective: To detect serological predictive course-parameters in Adamantiades-Behçet's disease (ABD).
Methods: Serum/blood of 122 ABD patients in active/inactive stages and of 75 controls was screened for IL1alpha, IL1beta, IL8, TNFalpha, sICAM1, bFGF, CRP, ESR. Serum IL18, IL8, MIP1alpha and CRP and ESR were investigated prospectively in another 18 ABD patients (11 with disease exacerbations in two years) and 16 controls.
Results: Increased IL8 serum levels, but not IL1alpha, IL1beta, IL6, TNFalpha, sICAM1, bFGF or ESR were found in active compared with inactive ABD patients and controls. Patients with oral aphthae and neurological features presented higher IL8 levels. An association of IL8 levels with the number of active clinical signs and the presence of severe oral aphthae was detected. Patients having systemic symptoms along with oral aphthae had significantly higher CRP and VEGF levels during the symptomatic period. In the longitudinal prospective evaluation, IL18 was increased in active and decreased in inactive periods in the individual patient's course. IL18 was elevated even in the presence of few oral aphthae or mild uveitis. Comparably, CRP was a useful course-indicator, while MIP1alpha and ESR showed no reliable correlation with disease activity.
Conclusion: IL18 is the most sensitive, single sign-parameter correlating with disease activity in ABD, followed by IL8, which increases with the number of clinical signs. CRP and VEGF can also be used as biomarkers to identify systemic but not mucocutaneous disease. The combination of these four serological parameters can be useful in the follow-up of ABD patients, especially in therapeutic clinical trials.

Board No: P-029
Ref. No: 206
Topic: Immunology

Clinical use of small bowel evaluation by double balloon enteroscopy in Behçet's patients with abdominal complaints

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Introduction: Gastrointestinal symptoms are often present in patients with Behçet's disease (BD), but objective gastrointestinal inflammation occurs infrequently. Double balloon enteroscopy (DBE) enables visualization of the entire small bowel (SB) together with tissue sampling and thus enhances the yield of gastroentestinal investigations of BD patients with abdominal complaints. Evaluation of Th1 and Th17 skewed key BD-cytokines might improve the understanding of the pathophysiological processes in this systemic vascuitis.
Patients and methods: 10 BD patients with therapy resistant abdominal complaints were evaluated with DBE. Serum cytokine profiles and lymphocyte subsets were analyzed.
Results: SB lesions were identified in four (40 %) patients. The SB lesions were located in the distal ileum in three, and distal jejunum and terminal ileum in one patient. In all patients the SB lesions were out of reach for conventional gastroduodeno- or ileocolonoscopy. No complications occurred during or after the DBE procedure.
In two of the patients with SB lesion (50%) the serum C-reactive protein (CRP) was elevated, while one patient without SB lesions (17%) had an elevated CRP level. Serum IFN-γ levels were elevated (> 10 pg/ml) in three patients (75%) with SB lesions and in one (17%) without SB lesions. Serum levels of TNF-α, IL1β and IL-10 were also elevated in patient 2.
Conclusion: DBE is a valuable and safe tool to evaluate the involvement of the entire gastrointestinal tract in BD patients possibly leading to a change in therapy. Inflammatory cytokines are involved, however no clear Th1 or Th17 profile was observed.

Table1

		Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8	Patient 9	Patient 10
A) Patient data	Age (yrs)	42	55	61	38	40	45	43	45	33	43
	Sex	M	F	M	M	F	M	M	M	M	M
	BDCAF	18	31	11	23	28	11	38	5	30	11
	Abdominal pain (# weeks/month)	1	4	1	3	2	2	1	1	2	1
	Diarrhoea (# weeks/month)	0	2	1	3	1	2	0	1	2	0
	Other symptoms	Oral ulcers, pustules	Oral ulcers, pustules, athralgia	Oral ulcers	Oral ulcers	Oral ulcers, pustules, athralgia, eye activity	Athralgia	Oral ulcers, genital ulcers, athralgia	Oral ulcers	Oral ulcers, genital ulcers, pustules, athralgia	Athralgia
	Medication	anti-TNF-α	Etoricoxib, Colchicine, Hydroxy-chloroquine	Mycofenolic acid, Azathioprine, Prednisolon	Infliximab, Cyclosporine	Prednisolon, Metotrexate, Adalimumab	None	Celecoxib, Cidesporine	Cidesporine, Sulfasalazine	Celecoxib, Ciclosporine, Colchicine	Prednisolon, Azathioprine
B) DBE Results	SB visualization	50%	Complete	Complete	Complete	Complete	Complete	50%	Complete	Complete	Complete
	Inflammation	Grade 2; Moderate active small ulcerations distal ileum	Grade 2; two aphtous lesions distal ileum	Grade 2; Moderate activity distal ileum, coecum and colon ascendens	Grade 1; Superficial inflammation distal jejunum and terminal ileum. Grade 3; Multiple severe ulcers in distal colon	None	None	None	None	None	None
C) Lab results	CRP (<10 mg/l)	1	8	29	24	<1	3	8	17	1	3
	Cytokine (pg/ml)										
	IFN-γ (<10)	22	14	5	30	<5	<5	<5	<5	18	<5
	TNF-α (<10)	<5	32	<5	<5	<5	<5	<5	<5	<5	<5
	IL-10 (<10)	8	10	<5	5	<5	<5	<5	<5	<5	<5
	IL-5 (<10)	<5	<5	<5	1	<5	<5	<5	<5	<5	<5
	IL-4 (<10)	5	<5	<5	<5	<5	<5	<5	<5	<5	<5
	IL-2 (<10)	7	<5	<5	<5	<5	<5	<5	<5	<5	<5
	IL-12 p70 (<10)	<5	5	<5	<5	<5	<5	<5	<5	<5	<5
	IL-8 (<10)	7	<5	5	9	<5	<5	<5	<5	<5	<5
	IL-1beta (<10)	<5	33	<5	3	<5	<5	<5	<5	<5	<5

Table 1. Table 1a) Patient characteristics (Behçet's Disease Current Activity Form; BDCAF); b) DBE results; c) laboratory results (normal range) (a) Age in years, b) SB= small bowel, C) CRP = C-reactive protein, IFN-γ = inter feron gamma,

Board No: P-030
Ref. No: 207
Topic: Immunology

Long term results of Adalimumab in Behçet's disease

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Objective: anti-TNF-α therapy has proven to be effective in patients with autoimmune diseases such as Behçet's disease (BD). In 2006 we presented a case series of 8 BD patients successfully treated with the anti-TNF-α agent Adalimumab. We kept monitoring these patients in order to analyze the long term efficacy and side effects.
Patients and Methods: a case series of 8 BD patients with active, mostly severe systemic refractory disease started treatment of 40 mg Adalimumab in 2006. Initial symptoms included uveitis, colitis, oesophageal, oral and genital ulcers, central nervous system involvement, erythema nodosum and arthritis and were refractory to most non-anti-TNFα immunosuppressive drugs. Patients were followed during their treatment by BDCAF as well as subjective disease response. Furthermore data on side effects was collected.
Results: Initially Adalimumab yielded swift objective responses and tapering of earlier used immunosuppressive therapy in all patients. The mean BDCAF decreased from 35.8 to 9.2. of the 8 patients 2 stopped the anti-TNF treatment after 190 and 608 days respectively and stayed in remission with conventional treatment. One moved abroad and was excluded after treatment 1119 days disease free period. The remaining 5 patients are treated from 1455 to 2000 days (4 to 5.5 years). BDCAF is still in the range of the initial respons, average of 8.8. No significant side effects occured.
Conclusions: Adalimumab constitutes a well-tolerated and effective modality in long term therapy for at least 5.5 years in BD patients.

Board No: P-031
Ref. No: 214
Topic: Immunology

Role of CD4+CD25hiCD127lo/-FoxP3+ regulatory T lymphocytes in the pathogenesis of Behçet disease in children

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Introduction : Behçet disease (BD) is an idiopathic and recurrent inflammatory disease. Physiopathology of BD shows a role of neutrophils and cytotoxic T lymphocytes.
Our aim is to assess the role of regulatory T lymphocytes (Tregs) in the pathogenesis of BD in children in comparison to adults.
Patients&Methods: We first assessed blood Tregs levels in 22 healthy children. We then studied 18 patients with active BD (10 children and 8 adults) and 18 age and sex-matched healthy controls. Percentages of blood CD4+CD127-CD25hiFoxP3+ Tregs and other T/B and NK cells subpopulations were performed by flow cytometry. We measured serum cytokines by Luminex and ELISA. We compared groups by using the Wilcoxon-Rank-signed test. Values were expressed as median.
Results: In children, Tregs population is high, especially in males and decreases with age. In BD children and adults, there is no difference in Tregs compared to controls (3 vs 4%). In BD children, Tregs are significantly less activated (CD4+CD25hiCD38+: 49 vs 63%). In BD adults, naïve Tregs (CD4+CD25hiCD127-CD45RA+) are low (1.6 vs 2.2, $p<0.05$). Naive CD8 (CD8+CD45RA+CD62L+) T cells are high in BD children but low in adults. In BD adults, the effector memory CD8 T cells subset is high (CD8+CD45RA-CD62L-: 24.5 vs 18%, $p<0.05$). There is no difference concerning B and NK cell populations. There is a trend for high IFN-γ, IL-1β, TNF-α, IL-15, IL-2, IL-17 levels in BD populations but not significant.
Conclusion: there is no deficit in Tregs number during BD acute phases. The Tregs ability to regulate CD8 cytotoxic T cells needs to be assessed.

Board No: P-032
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Topic: Immunology

EFFECTS OF AZITHROMYCIN ON IN VITRO INTRACELLULAR CYTOKINE RESPONSES IN BEHCET'S DISEASE

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Aim: The aim of this study was to investigate the effects of azithromycin on in vitro intracellular cytokine responses in patients with Behcet's disease (BD).
Materials and Methods: Ten BD patients with active mucocutaneous findings and impaired oral health who were not under any treatment modalities (F/M: 8/2, age: 38.6±10.9 years) and 9 healthy controls (F/M: 6/3, age: 33.2± 7.1 years) were included in the study. Patients were treated with azithromycin (1500 mg/week) for 4 weeks. Clinical and immune responses were evaluated in pre-treatment and post-azithromycin treatment periods. The peripheric blood mononuclear cells (PBMCs) of patients and controls were stimulated by lipopolysaccharide (LPS), lipoteichoic acid (LTA), heat shock proteins (HSP-60) and Streptococcus sanguis (S.sanguis) for 3 hours. In vitro intracellular interferon-gamma (IFN-gamma) and tumor necrosis factor-alpha (TNF-alpha) levels were measured in culture supernatants by ELISA. Flow cytometry was used to analyse neutrophil expressions of CD11b and CD16.
Results: Percentages of stimulated intracellular IFN-gamma responses with S. sanguis were higher in BD (7.5±6.2) in pre-treatment than HC (3.9±2.7) and post-treatment period (3.3±4.3)(p=0.05 and p=0.052, respectively). Percentages of stimulated intracellular IFN-gamma responses with LTA were also higher in BD (4.9±2.4) in pre-treatment than post-treatment period (3.2±1.9), however without reaching significance (p=0.07). In post-treatment, the percentage of CD16 expression was decreased in BD (91.6±5.4) compared to HC (97.4±0.79) and pre-treatment period (95.17±4.23)(p=0.04 and p=0.02, respectively).
Conclusion: Azithromycin treatment decreased both intracellular IFN-gamma responses to S. Sanguis and neutrophil expressions of CD16 in short-term PBMC cultures, suggesting an immunomodulatory effect in BD.

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Topic: Immunology

Symptoms of Behcet's Syndrome in a representative group of adults in the UK

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Objective: To assess the type and number of symptoms reported by adults with BD
Methods: A questionnaire was mailed 650 members of the Behçet's Syndrome Society (UK). Participants provided information on their socio-demographic characteristics, use of health care services, current symptoms, and whether symptoms were under control.
Results: 447 adults returned the questionnaires (68.8% response rate), of whom 410 (63%) had information on the variables selected for this analysis. The mean age of participants was 49.9 years (Standard Deviation: 12.7, range: 14 to 81), 76% were females; 94% were White British. All of them had a confirmed diagnosis of Behçet's Syndrome. Only 2% of participants were free from symptoms. The mean number of symptoms experienced was 5.3 (SD: 2.3, range: 0 to 10). The most commonly reported symptoms were Fatigue (83.7%), Joint problems (79.0%) Mouth ulcers (76.1%), headaches (55.1%), stomach/bowel problems (52.4%), skin lesions (45.8%), genital ulcers (43.8%), eye problems (42.9%), neurological problems (28.4%), pathergy reaction (16.5%). Only 17.8% of participants considered that their symptoms were controlled. Several types of health professionals contributed to the treatment of BD; GP (72.6%), Rheumatologist (63.7%), Dentist/Oral Consultant (42.0%), Ophthalmologist (32.3%), Neurologist (17.0%), Dermatologist (12.8%), Gastroenterologist (12.1%), Immunologist (11.9%), and Obstetrician/Gyneacologist/Sexual Health Professionals (7.2%). The mean number of doctors treating a participant was 2.72 (SD: 1.6, range: 0 to 10).
Conclusion: The number of symptoms reported by patients with BD was relatively high. A collaborative multidisciplinary health care team and the administration of multiple medications are necessary to take care of patients with BD.

Age group	Male		Female		TOTAL	
	n	%	n	%	n	%
18-29	6	1.4	21	4.9	27	6.3
30-39	19	4.4	49	11.3	68	15.7
40-49	30	6.9	84	19.4	114	26.4
50-59	22	5.1	97	22.5	119	27.5
60-69	25	5.8	53	12.3	78	18.1
70-79	3	0.7	21	4.9	24	5.6
80+	0	0.0	2	0.5	2	0.5
Total	105	24.3	327	75.8	432	100.1

Board No: P-034
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Topic: Immunology

Anti –TNF Therapy in severe uveitis of Behçet's Syndrome: Report of 27 patients

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Objective: To present the experience of a dedicated center with anti-TNF agents in the treatment of severe uveitis of Behçet's syndrome (BS).
Methods: A retrospective survey of the medical charts revealed 50 BS patients treated with anti TNF agents between October 2001 and January 2010. The indication was severe uveitis refractory to immunosuppressive treatment in 27 patients (21 M, 6 F; mean age 34 ±8 SD years; mean duration of eye disease: 9 ±5 SD years). 24 (89%) patients were treated with infliximab, 2 (7%) with etanercept and 1 (4%) with adalimumab. The changes in the visual acuities (VA) and in the numbers of ocular attacks before (in the preceding year) and during treatment were the main outcome measures.
Results: The mean duration of treatment was 25±18 SD months. Azathioprine was the most frequently used concomitant agent (19 patients, 70 %). All patients used prednisolone in varying dosages. Initial VA's were 0.31 ± 0.4 SD in the right and 0.27 ± 0.3 SD in the left eye. VA remained stable in the right (0.29 ± 0.38 SD) and increased significantly in the left eye (0.35±0.37 SD; p = 0.04) under anti TNF treatment. The frequency of ocular attacks decreased in both eyes but this was significant only for the left eye (before treatment: 1.8 ± 1.9 SD, under treatment: 1.0 ± 1.1 SD; p = 0.016).
Conclusion: Our results support the efficacy of anti TNF agents in severe and refractory uveitis of BS.

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Topic: Immunology

Age, gender and disease related platelet and neutrophil activation in Behcet's

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Activated platelets are important in leucocyte recruitment and are linked to pathological disorders associated with vascular inflammation
Objectives: To investigate the effect of disease activity, gender and age (ranges 20-30, 31-50, 50>)on platelet and neutrophil

activation in Behcet's disease (BD).
Methods: Blood was collected from 81 healthy controls (HC) (24-71 years;47 female) and 43 patients with BD (23-72 years;26 female). Whole blood was collected first into K3EDTA and then immediately transferred to a vacutainer containing CTAD. Anticoagulated blood was held at 4°C until flow cytometric analysis 6 hours after venesection.
Results:Inactive (2.78 ± 0.56%;p=0.0009;3.11±0.78%;p<0.0001) and active (2.28 ±0.84%, p<0.0001;3.071±0.67%,p=0.0031) BD patients had a significantly higher percentage of CD62P expressing platelets and % CD62P+ platelet microparticles as compared to HCs (0.84± 0.1%,1.23±0.14%) respectively. Neutrophil platelet aggregates were significantly higher in the inactive (2.89± 0.54%;p=0.0169) BD patients only
The percentage of CD62P+ platelets and CD62P+ platelet microparticles in female BD (2.1±0.46%;p=0.0029, 2.57±0.46%,p=0.0009) and male BD (3.43±0.95%,p=0.0038, 3.91±1.44%,p=0.048) patients respectively were significantly higher than that expressed by female HCs (0.87±0.15%, 1.22±0.15%)) and male HCs (0.79±0.16%,1.26±0.26%). Neutrophil platelet aggregates were significantly higher in male BD only (3.77±0.99%,p=0.0113). The %CD62P+ microparticles were significantly increased in the 20-30 (p=0.0301) and 31-50 (p<0.0162) age ranges but not in the older over 50 age group of BD patients.
Conclusion: Microparticles have an essential role in inflammation. Data suggests that there is a decrease in inflammation with increasing BD patient age since the percentage of CD62P+ microparticles were not significantly increased in the over 50 BD age group.

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Topic: Immunology

Secretory leukocyte protease inhibitor (SLPI) in Behcet's patients saliva.
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Secretory leukocyte protease inhibitor (SLPI) is a member of the innate immunity-associated proteins. SLPI is found in human saliva and it has been shown to have antimicrobial activity and thought to participate in mucosal defence.

Objectives: 1) To measure SLPI in the saliva of Behcet's disease (BD) patients and healthy controls (HC). 2) To measure mRNA and protein expression in buccal epithelial cells of BD and HC.

Methods: Saliva and buccal swabs were collected from 34 BD, 4 recurrent aphthous stomatitis (RAS) patients and 9 healthy controls (HC). SLPI was measured in whole unstimulated saliva by ELISA and RNA was isolated from buccal scrapes and reverse transcribed to cDNA. Gene expression of SLPI and the control gene 18SrRNA were measured by real time PCR and protein expression on buccal epithelial cells was measured by Western blotting.

Results: SLPI levels in BD (0.197±0.027µg/ml,p=0.0003) and RAS (p=0.0028) patients were significantly lower than in HC (0.655±0.124µg/ml). SLPI levels were lower in male (p=0.018) and female BD (p=0.008) versus HC as were inactive (p=0.0004) and active (p=0.0028) BD levels. QPCR data showed that SLPI mRNA was significantly higher in the active BD patients (p=0.011) but not inactive BD patients (p=0.055). Western blotting showed the presence of SLPI with positive bands corresponding to higher molecular weight proteins than that expected for SLPI.

Conclusions: SLPI inhibits elastases which can cause cellular damage at sites of inflammation. Lower levels of SLPI in BD could be a clue to the instability of the oral mucosa in BD.

Board No: P-037
Ref. No: 262
Topic: Immunology

Expression of NK Receptors on gd T Cells In Behcet's Disease
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Considering the increased expression of CD94 in Behcet's Disease (BD) and the regulatory functions of NKG2 molecules in heterodimer with CD94, the presence of NK receptors on T cell subsets are screened in this study.
NKG2A/C/D molecules on gd T cells, CD56+ NK cells or CD8+ T cells were analyzed in 38 active (ABD) and 32 inactive (IABD) patients with BD, 27 tuberculosis patients (TB) and 26 healthy controls (HC) by using flow-cytometry. CD16 and CCR7 molecules were evaluated on Vg9d2 T cells.
gd T cells were increased in patients with IABD compared to HC and TB (4.4 vs. 3 and 2.9%, p=0.029 and 0.026), whereas Vd2 T cells were lower among gd T cells (50 vs. 70.6%,p=0.014). Vg9Vd2 T cells of the BD patients had decreased CCR7 (68.4 vs. 83.8%, p=0.0001), but increased CD16 (21.8 vs. 15.6, p=0.02). Increased expression of activating NKG2C molecules was observed on gd T cells of BD and ABD (0.4, 0.5 vs. 0.2, p= 0.008), but NKG2A expression on the T cells was not statistically different in both groups. Activating NKG2D receptors on the gd T cells were found to be decreased in patients with BD and TB compared to HC (78.5 and 79.8 vs. 92.5%, p=0.001 and 0.004). NKG2C was also higher on CD8+ T cells of BD patients (7.2 vs. 3.3, p=0,024).
The lower CCR7 and NKG2D and the higher NKG2C and CD16 on gd T cells of BD patients implicate that these receptors are involved in the regulation of immune response in this disease.

Board No: P-038
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Topic: Immunology

Immunomodulatory mechanisms induced by IFN alpha 2b
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Interferon alpha has been recently used successfully in the management of patients with Behcet's disease and, in contrast to conventional immunosuppressive agents in this disease, seems to be able to induce disease remission in some patients which is maintained after the IFN-α has been discontinued. The mechanisms underlying this are uncertain but regulatory T cells are thought to be involved. In this study we investigated the effect of IFN-α2b on healthy donor cells in vitro and PBMCs derived from BD patients either receiving IFN alpha2b or conventional immunosuppression. We report that IFN-α treatment produces a significant increase in FoxP3-expressing CD4+ and CD4+CD25hi T cells as well as an upregulation of IL-10 and TGFβ production by T cells in vitro. Similar findings are seen in patients with BD who have received IFN alpha 2b and, crucially, are maintained after the discontinuation of IFN-α. We conclude that upregulation of Tregs may underlie the response of some BD patients to IFN-α.

Board No: P-039
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Topic: Immunology

Clinical Characteristics and Serum Cytokines in a US Cohort of Patients with Behcet's Disease at the NIH
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Objective: To describe the clinical characteristics and serum cytokine profiles of patients with Behcet's Disease (BD) seen at the NIH

Design, Setting, and Patients: Prospective cohort of patients evaluated at the NIH beginning May 2009 fulfilling the International Criteria for Behcet's Disease. Cytokine levels were measured using the Luminex technology in 18 patients and 25 healthy donors.

Results: Mean age was 31.25 (+/-13.48). 23% of patients were male and 27% were HLA B51 positive. Oral ulcers were present in 100% of patients, genital ulcers in 91%, arthritis in 86%, pustular skin disease in 86%, gastrointestinal ulceration in 55%, ocular disease in 45%, psychiatric disorders in 36%, neurologic disease in 27%, and vascular thrombosis in 23%.

Median serum levels in pg/mL of IL-17, MIP-1ß, and IL-9 were significantly lower in BD patients than in controls: 100.8 vs. 205.3 (p=0.01), 1280.1 vs. 2130.1 (p=0.03), and 253.8 vs. 426.4 (p=0.02), respectively. There was a non-significant trend towards lower IL-2: 58.9 vs. 185.8, MIP-1a: 43.7 vs. 74.9, and G-CSF: 56.3 vs. 89.7 and higher TNF-a: 517.8 vs. 304.8. IL-6, IL-10, IL-1-b, IL-1 Ra, IL-8, and IFN-g were similar.

Conclusions: A cohort of patients with BD is building at the NIH. Lower serum levels of some cytokines in the TH-17 pathway are demonstrated implying that IL-17 is important in BD pathogenesis. The benefit of targeting IL-17, or other regulatory cytokines such as IL-1 or IL-23, can be tested clinically as effective therapeutics become increasingly available.

Board No: P-040
Ref. No: 280
Topic: Immunology

Plasma and saliva cytokine profiles in Behcet's Disease: Do cytokine levels in plasma correlate with levels in saliva?
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Objectives: 1) To compare Th1 and Th2 cytokines in plasma and saliva of Behcet's patients using the FlowCytomix™ Multiplex array (Bender MedSystems). 2) Compare the cytokine/chemokine results from the FlowCytomix™ array to a standard ELISA method completed on a larger cohort of patients.

Methods: In a pilot study IL-12p70, IFN-gamma, IL-2, IL-10, IL-8, IL-6, IL-4, IL-5, IL-1ß, TNF-alpha and TNF-ß concentrations were measured in matching plasma and saliva from active Behcet's disease (BD) (n=2), inactive BD saliva (n=2) and healthy control saliva (HC) (n=4) using a bead based assay to detect multiple analytes by flow cytometry. ELISA was used to measure IL-8 levels in plasma (BD n=50, HC n=11) and saliva (BD n=35, HC n=9).

Results: Analyte concentrations were calculated using the FlowCytomix Pro software and from the 11 cytokines investigated in plasma and saliva, IL-8 and IL-1ß were found to be the most abundant. Levels of IL-8 in BD saliva (778.7±237.5pg/ml) were notably higher than in BD plasma (23.0 ±11.0 pg/ml) but comparable to HC saliva (874.8 ±437.4 pg/ml). Using ELISA with a greater number of BD and HC plasma and saliva samples, IL-8 was found at low levels in plasma (BD 4.0 ±1.4 pg/ml, HC 2.1 ±0.7 pg/ml) and high levels in saliva (BD 720.6 ±42.36 pg/ml, HC 705.7 ±68.4 pg/ml).

Conclusions: Raised IL-8 levels have been reported as a possible marker of BD however this data suggests that there is no significant difference between BD and HC IL-8 levels in plasma and saliva.

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Topic: Immunology

Alemtuzumab (CAMPATH-1H) as remission induction therapy in Behcet's Disease
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Behcet's Disease is a chronic, relapsing inflammatory disorder, characterised by recurrent oral and genital ulceration, but the more severe, life threatening manifestations are neurological, gastrointestinal or vascular in nature. The precise aetiology and pathogenesis are unclear, but there is increasing evidence that T cells play a key role in the development of disease, and thus T cell depleting agents, such as the anti-CD52 humanised monoclonal antibody, alemtuzumab (CAMPATH-1H) are a potential therapeutic strategy. We present our experience of 20 patients treated with alemtuzumab since 1998 in Addenbrooke's Hospital, Cambridge, UK. Patient demographics and outcomes after alemtuzumab therapy as presented in table 1. Six months after treatment, 74% of patients were in complete remission. Many of these remissions have been sustained over years, but in those that did relapse, re-treatment with alemtuzumab was effective. There were significant reductions in prednisolone dose and BVAS (a measure of disease activity) after treatment. Alemtuzumab was well tolerated. 25% of patients experienced infusion reactions, but in only one did the reaction necessitate termination of the treatment. There were no infectious complications directly attributable to alemtuzumab, but six patients developed new autoimmune thyroid dysfunction. Autoimmune diorders following alemtuzumab, including thyroid disease and ITP, have also been

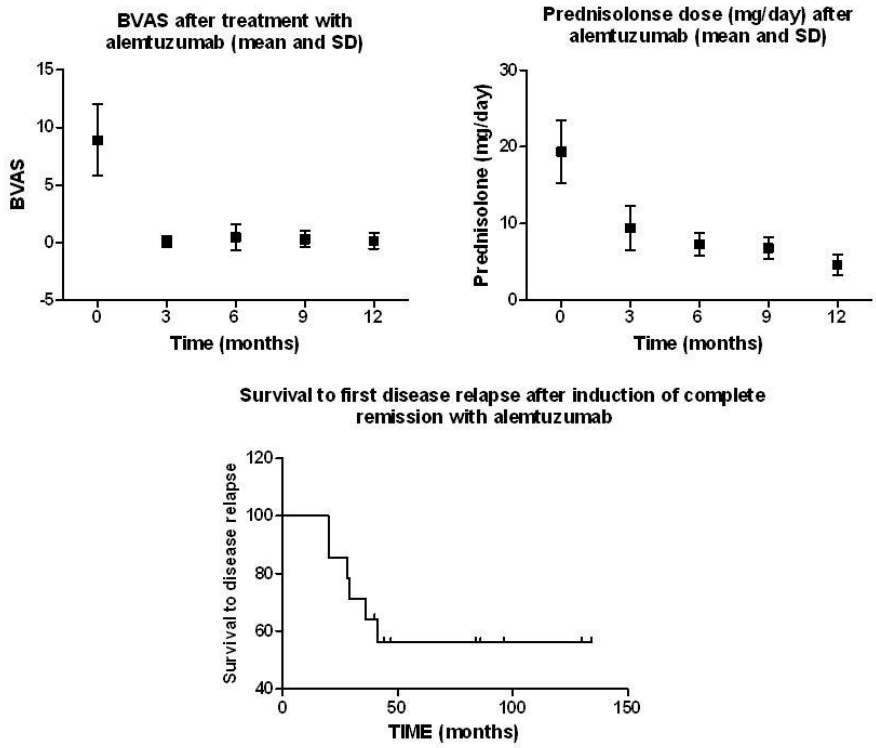
reported in multiple sclerosis. Alemtuzumab is a safe and effective therapy for the treatment of Behcet's Disease, particularly in those cases which prove to be refractory or life threatening.

No	Age, Sex	Organs Involved	Disease Duration (months)	Previous Therapies	ALM date	Response at 6 months	Follow Up (months)	Time to relapse (months)	Further treatment	Current Status
1	34, F	OGU, Eye, GIT, J, S	64	P, AZA, CsA, CYC, IVIG	2/98	CR	84			CR
2	42, F	OGU, GIT, J, S	47	P, HC, TL MTX, CsA, AZA, IVIG	1/98 9/02 4/03	CR	80	28 2* PR	P 2nd ALM 3rd ALM IFX, ETN	CR
3	18, F	OGU, S, J	93	P	12/98	CR	86			CR
4	28, F	OGU, Eye, J, CNS	1	P	2/99	CR	40			CR
5	39, F	OGU, Eye, GIT, J, S	12	P, AZA, CsA	1/99	PR	120	12 41 51	P, MMF P, HC, MMF P, TL, MMF	CR
6	49, F	OGU, J, S, CNS	12	P, PE	1/99	CR	130			CR
7	36, M	OGU, Eye, J, S, V, GIT, CNS	203	P, AZA, CYC, TL, PE	9/98	CR	96	9	P, AZA, CsA	CR
8	59, M	OGU, J, S, V	107	P, CsA, CYC	10/98	CR	134			CR
9	21, F	OGU, J, S, V	84	P, AZA	12/98 4/08	CR CR	134	29 87 112	CsA IFX 2nd ALM ADM	PR
10	41, F	OGU, Eye, GIT, J, S	108	P, AZA, MMF, CsA, MTX, TL, IVIG	2/99 8/04	PR CR	133	25 38 66 11* 35	P CsA 2nd ALM CsA IFX, ETN	CR
11	29, M	OGU, Eye, J, CNS, S	12	P	12/98 4/03 10/05	CR CR CR	108	49 28*	TL, 2nd ALM HC, 3rd ALM	CR
12	23	OGU, Eye, GIT, J, S, CNS	72	P, CYC	1/99 5/02 8/03	CR PR PR	108	36 14* 11*	AZA, FK 2nd ALM 3rd ALM	

					7/08 2/10	CR		22 33 40 57 19*	RTX RTX RTX RTX 4th ALM 5th ALM	CR
13	20, F	OGU, J, S	21	P, HC, AZA, FK, TL, MTX	1/03	CR	86	41	P	CR
14	45, M	OGU, Eye, GIT, S, V	161	P, HC, FK, MTX, IVIG, AZA	7/06	CR	44			CR
15	45, M	OGU, Eye, GIT, J, S, Lung, CNS, PNS	26	P, HC, AZA, IFX	3/08 8/08	PR PR	20		RTX	RIP
16	53, F	OGU, GIT, J, S, CNS, PNS	53	P, AZA, MMF, FK, CYC, TL, IFX, HC, ETN	10/06	PR			CsA, MMF, TL	CR
17	35, F	OGU, GIT, J, S	23	P, AZA, TL, IVIG, IFX, ETN, ADM, RTX	6/07 12/08 12/09	CR	33	18 11*	2nd ALM 3rd ALM	CR at 4/12
18	35, F	OGU, Eye, GIT, J, S, CNS, V	13	P, MMF, CsA, FK	2/03 10/04	CR PR	85	20 2*	2nd ALM RTX, IFX	CR
19	23, F	OGU, GIT, J, CNS	46	P, HC, FK, IFX, ADM	4/06	PR	47		IVIG	PR
20	49, F	OGU, Eye, GIT, J, S	38	P, MMF, FK, TL, IFX, ETN, ADM, HC, MTX	7/05 8/06 11/09	PR CR	56	10 33*	P 2nd ALM P, 3rd ALM	CR - 4/12

CNS, central nervous system; GIT, gastrointestinal tract; J, joint; OGU, oral genital ulceration; PNS, peripheral nervous system; S, skin; V, vascular; ADM, adalimumab; ALM, alemtuzumab; AZA, azathioprine; CsA, ciclosporin; CYC, cyclophosphamide; ETN, etanercept; FK, tacrolimus; HC, hydroxychloroquine; IFX, infliximab; IVIG, intravenous immunoglobulin; MMF, mycophenolate mofetil; MTX, methotrexate; P, prednisolone; PE, plasma exchange; RTX, rituximab; TL, thalidomide; CR, complete remission; PR, partial remission

Prednisolone doses, BVAS, and relapse free survival post alemtuzumab



Board No: P-042
Ref. No: 297
Topic: Immunology

Safe, Rapid-Onset, and Sustained Biological Activity of IL-1beta Regulating Antibody XOMA 052 in Resistant Uveitis of Behçet's Disease: Preliminary Results of a Pilot Trial

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Overexpression of proinflammatory cytokines, including IL-1β has been considered to play a critical role in Behçet's disease (BD). XOMA 052, a recombinant humanized monoclonal anti-IL-1β antibody, may produce rapid and sustained efficacy in IL-1β-mediated diseases.
This study evaluated the safety and PK of XOMA 052 in BD uveitis. We enrolled seven patients with a posterior/panuveitis or retinal vasculitis attack despite cyclosporine and/or azathioprine treatment. We administered XOMA 052 as a single 0.3 mg/kg intravenous infusion on Day 0, discontinued immunosuppressive agents, and maintained prednisolone at ≤10mg/day (6 patients) or 20mg/day.
Four patients completed the study period (98 days). No adverse events related to XOMA 052 were observed so far. Intraocular inflammation started to resolve in all patients on Day 1, and complete resolution of retinal findings and vitreous haze was achieved in 4-21 days. All but one patient were in remission on Day 28, and the efficacy was sustained up to Day 98 in one. Five patients received a second infusion for new retinal infiltrates between Day 49 and 95, and one patient for macular edema in the other eye on Day 29. Five patients had recurrences of oral ulcers and folliculitis despite resolution of intraocular inflammation.
Administration of XOMA 052 appears safe, and IL-1β regulation using XOMA 052 shows a rapid-onset effect for treatment of uveitis attacks in BD. This favorable effect of XOMA 052 was observed despite discontinuation of immunosuppressives as of infusion day and without an increase in corticosteroids.

Board No: P-043
Ref. No: 301
Topic: Immunology

Expression of Transcription Factors of CD4+ T Helper Cells in Behcet's Disease

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Objective: According to the recent studies; spontaneous and/or induced overexpression of pro-inflammatory cytokines (mainly Th1 type) from various cellular sources seems responsible for the enhanced inflammatory reaction in Behcet's disease (BD). IL-17 is the recently discovered pro inflammatory cytokine and is up-regulated in BD with uveitis. In this study, we have screened the expression levels of Th17 transcription factor (retinoic-acid-related orphan receptor (ROR)-gamma t) of the CD4+ T cells.

Methods: CD4+ T cells were purified from the peripheral blood mononuclear cells of 14 active BD, 6 tuberculosis (TB) patients, and 11 healthy controls (HC) by using the magnetic beads (MACS, positive selection). RNAs of these donors were purified and transcribed into cDNA. Expression of ROR-gamma t in CD4+ T cells were determined by using semi-quantitative RT-PCR (Sybr Green). Expression levels were evaluated with GAPDH gene by comparing 2^{-ddCT} and analyzed by non-parametric tests.

Results: The expression levels of Th17 transcription factor were higher among BD patients compared to HC (1.42 vs. 1.0), however the difference was not statistically significant. The ROR-gamma t transcription factor was down-regulated in TB patients compared to BD (0.55 vs.1.42, p=0.009) and HC (1.00, p=0.024).

Conclusion: The relatively higher expression of ROR-gamma t transcription factor in BD in contrast to lower levels in TB implicates differential regulation Th17 cells in these diseases.

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Board No: P-044
Ref. No: 302
Topic: Immunology

Genetic Predisposition for Low IL-10 Expression is Associated with Behçet's Disease

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Genetic predisposition to Behçet's disease (BD) plays an important role in its pathogenesis. Our recent genome-wide association study (GWAS) performed in 1215 BD patients and 1279 controls from Turkey identified the interleukin-10 (IL10) gene as a new non-HLA risk factor for BD. Five SNPs in the IL10 gene, all in strong linkage disequilibrium with one another, were found strongly associated with BD, one of them (rs1518111 G>A) with genome-wide significance. A meta-analysis of ethnically matched case/control collections from diverse populations (2430 cases, 2660 controls) established associations with the IL10 variant. We tested the functional significance of rs1518111 polymorphism using different methods. Allelic imbalance measurement in monocyte pre-mRNA from 8 heterozygous healthy individuals showed that the level of transcripts with rs1518111 A allele was found to be reduced to 35% of the level of the transcript with G allele. Similarly, LPS-stimulated mononuclear cells from healthy Turkish donors homozygous for A allele produced significantly lower amounts of IL-10 compared with individuals with one or two G alleles. Also, stimulation of cultured monocytes from

ethnically diverse healthy donors from the United States with MDP and Pam_3Cys resulted in production of significantly less IL-10 in individuals homozygous for the disease-associated A allele than individuals with one or two G alleles, whereas no statistically significant variation was found in TNFα production.
These findings suggest that a genetic predisposition for low IL-10 expression at both mRNA and protein level is a risk factor for BD, which may provide new targets for the treatment of BD.

Board No: P-045
Ref. No: 303
Topic: Immunology

Role of Gamma Delta+ T Cells in Behcet's Disease
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Behcet's Disease (BD) is a multisystemic disorder with muco-cutaneous, ocular, arthritic, vascular or central nervous system involvement. The role of gd T cells is implicated in BD and the regulatory role of these cells by cytokine and chemokine secretion is evaluated in this study.

Peripheral blood mononuclear cells (10^5 cells per well) were stimulated with a phosphoantigen (BrHPP) and responsive gd T cells expanded with IL-2. Cultures of donors with at least 70% gd^+ T cells (23 active BD, 20 inactive BD, 20 controls) were stimulated with the PolyI:C (TLR-3 agonist), BrHPP or PolyI:C+BrHPP. The concentrations of IL-17, IL-13, IFN-g, TNF-a, GM-CSF, MIP-1ß, RANTES were measured in the supernatants after 24 hours of the stimulation by multiplex ELISA.

gd T cells of BD patients expanded with BrHPP and stimulated by TLR-3 and BrHPP produced less IL-13 (p=0.029), IFN-g(p=0.008), GM-CSF (p=0.02), MIP-1ß (p=0.012), TNF-a (p=0.021) and RANTES (p=0.04) compared to controls. All the reduced secretion levels were more pronounced in the inactive BD patients group and BrHPP alone also induced lower levels of GM-CSF, MIP-1ß and TNF-a in this group. IL-17 did not show any difference among study groups.

Lower cytokine response of gd^+ T cells, particularly in inactive phase of the disease implicate down modulation of these cells in BD.

This study is supported by Istanbul University Research Fund (#501).

Board No: P-046
Ref. No: 307
Topic: Immunology

The medium-term efficacy of recurrent uveitis in Behcet disease with etanercept
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OBJECTIVE: To investigate the medium-term efficacy of recurrent uveitis in Behcet disease (BD).
METHODS: Retrospective review of 5 recurrent BD uveitis patients (10 eyes) who were followed up in the Department of internal medicine of the military hospital of tunis among 160 case of BD during the period from April 2004 to April 2010 . All patients were men with a mean age of 31 years old with a refractory posterior uveitis who had failed at least one immunosuppressive drug. Patients received etanercept for treatment of uveitis on the basis of conventional corticosteroid and immunosuppressive therapy. Two of them had severe BD lesions: one a neurological and the other intestinal lesions. Subcutaneous injection of 3 – 7,5 mg/kg etanercept twice per week was instituted initially, followed by an injection in day 45 than each two months. Complete ocular examinations including visual acuity, intraocular pressure, anterior segment and fundus conditions were well documented each visit. RESULTS: of 5 patients, 4 had shown improvement on visual acuity at least in one eye and one patient was not improved. No serious adverse reaction was observed during follow up.
CONCLUSION: On the basis of corticosteroid and immunosuppressive therapy, etanercept has good medium term efficacy and safety for recurrent BD uveitis.

Board No: P-047
Ref. No: 310
Topic: Immunology
EFFICACY AND SAFETY OF TNF-ALPHA ANTAGONISTS IN THE MANAGEMENT OF BEHCET'S SYNDROME: A SYSTEMATIC REVIEW
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BACKGROUND: There is growing evidence on the efficacy of TNF-alpha in BS, mainly based on open studies or case series. This systematic review aims to analyze the current literature on the efficacy and safety of TNF-alpha antagonists in BS patients.
METHODS: We performed a systematic literature search covering articles in PubMed until April 2010, abstracts from the annual Meetings of EULAR, ACR and ICBD.
RESULTS: 102/216 articles and 17/32 abstracts were included in the analyses. Open studies and case series of eye involvement showed a rapid onset of response, improvement in visual acuity, suppression of retinal vasculitis, decrease in uveitis attacks and corticosteroid use. However relapses were observed when the drug was stopped. A good clinical response and regression in MRI findings was observed in neurologic involvement. Use in gastrointestinal involvement resulted in rapid healing of ulcers and resolution of bloody diarrhea. The response in gastrointestinal involvement seems to be more sustained compared to other organs. The only RCT with etanercept showed a medium effect size for oral ulcers (0.59) and papulopustular lesions (0.51). Tuberculosis, and other infections were the most common adverse events.
CONCLUSION: TNF-alpha antagonists seem to be promising agents especially for resistant eye, GI and CNS involvement of BS. Controlled studies are needed since current evidence which relies mostly on open studies and case reports might have caused a false impression of efficacy due to publication bias. Side effects are similar to those reported for TNF-alpha antagonist use in other diseases.

Board No: P-048
Ref. No: 400
Topic: Immunology

Anti-TNF-α (adalimumab) in Ocular Behηet's disease

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Introduction: Behçet's disease (BD) is a systemic vasculitis of unknown origin. Amongst several systemic inflammatory complaints, uveitis is one of the most urgent immunosuppressive requiring conditions. Azathioprine and cyclosporine are the only drugs for uveitis in BD that are proven effective in randomized trials up till now. The introduction of TNF-α blocking agents, such as the chimeric monoclonal antibody (MoAb) infliximab, heralded a new era in the treatment of (auto)inflammatory diseases. Also BD and uveitis patients respond remarkably well to TNF-α blocking therapy. Hence, EULAR recommendations include the initiation of infliximab as a second step in BD patients with a drop in visual acuity of more than 2 lines and/or retinal disease (vasculitis or macular involvement). Adalimumab is a fully humanized MoAb with TNF-α blocking characteristics of which therapeutic efficacy in BD and uveitis patients has been described in several case series. We have retrospectively investigated the long term results of adalimumab in BD patients with therapy refractory uveitis.
Patients and methods: From 2005 till onward, out of of 56 uveitis patients with a follow up of at least 3 years, 10 BD patients were selected and evaluated. Clinical parameters included Visual Acuity (VA), Uveal Disease Activity (UDA), number of ophthalmologic outpatient clinic visits.
Results: VA improved form 0.3 to 0.5 after 2 years (p=0.015) and remained at this level for at least 3 years. UDA was high (93%) at the time of initiation adalimumab. Swift significant improvement was seen, leveling to about 12% within 9 months. Both observations decreased the need of ophthalmologic controls form 6 every year before adalimumab, to 1.8 afterwards.
Conclusion: In this retrospective case series we demonstrate objective therapeutic efficacy of adalimumab in patients with severe therapy refractory BD associated uveitis. These results need randomized conformation in larger studies.

Board No: P-049
Ref. No: 55
Topic: Vasculitis

Fever in Behçet's disease. 107 cases
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Background:Fever is unusual in Behcet's disease(BD).Nevertheless,it can be observed in relapses of oral and genital ulcers,joint,vascular and neurological involvements.Rarely long standing fever can be inaugural.
Purpose:to evaluate clinical manifestations with fever and the diagnostic problems of long standing fever in BD.Results:of 1294 cases of BD registered between 1981 and 2009, in one department of Medicine,107 had fever(8.25%).Subject included 73 males and 34 females:31.78% for a sex ratio of 2.14.Average age was 26.08.Youngest was 13 and oldest was 54.Fever revealed BD in 9 patients:isolated in 2 cases.Fever was present in 48 patients(45%)with attacks of oral and genital ulcers especially in giant aphtosis;in joint involvement (n=4):2 of them had erythema nodosum; 20 patients presented vascular involvement:Vena cava thrombosis(VCT)(n=6) :Superior VCT(n=4),Inferior VCT (n=2); Budd-Chiari syndrome in 2 cases, one of them was associated with superior VCT;arterial aneurysms(n=6). Neurological involvement was observed : parenchymal (n=10), non parenchymal (n=3);ulcerative colitis (n=3) .One patient had audiovestibular disturbance and another had orchiepididimytis .6 cases of long standing fever revealed neurological involvement (n=3),arterial aneurysm(n=2)and pneumopathy(n=1).Bacterial infections were observed in 14 patients:one of them had disseminated tuberculosis(liver, adenopathy, lung)with long standing fever .One patient had malignant tumor.Treatment was colchicine and penicillin in patients with fever and oral,genital ulcers;corticosteroids in vascular, digestive and neurological involvements.Antibiotics were used in infections.One patient died from bronchial carcinoma.Conclusion:Fever is rare and underrecognized in BD.It can be associated with attacks of BD.Long standing fever can also inaugurate BD raising diagnostic problems.Infections and malignant tumor are not the first cause of fever.

Board No: P-050
Ref. No: 73
Topic: Vasculitis

Infliximab in the Treatment of Behçet´s disease
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Objectives:Behçet's disease is a chronic-relapsing, multisystemic disease of unknown etiology, classified among the vasculitides[1]. It has been reported in the literature cases with severe refractory mucocutaneous manifestations, with a good response to anti TNF alpha agents[2].
Case:C.I. 41 years old, female, with a 15 years history of recurrent oral ulcers, symmetrical and non-erosive polyarthritis, recurrent cutaneous ulcers with residual scarring involving the entire body. She had no history of genital ulcers or ocular involvement.
HLA B51 and pathergy reaction were positive.
She was treated with hydroxychloroquine, azathioprine, dapsone, thalidomide, methotrexate, prednisolone (up to 40mg / d) and NSAIDs without response.
The authors decided to start infliximab (0.3 mg / kg 6 / 6 weeks), with complete regression of skin lesions and improvement of polyarthritis after 2 weeks. She is in the 7th week after initiation of infliximab maintaining clinical stability.

Conclusions:This case supports the results observed in other cases of Behcet's disease with severe manifestations resistant to conventional therapies, successfully treated with anti TNF alpha[3].
The use of infliximab showed a rapid onset of action, with remission of the mucocutaneous manifestations and improvement of polyarthritis, however it will be need a longer follow-up to confirm the effectiveness.

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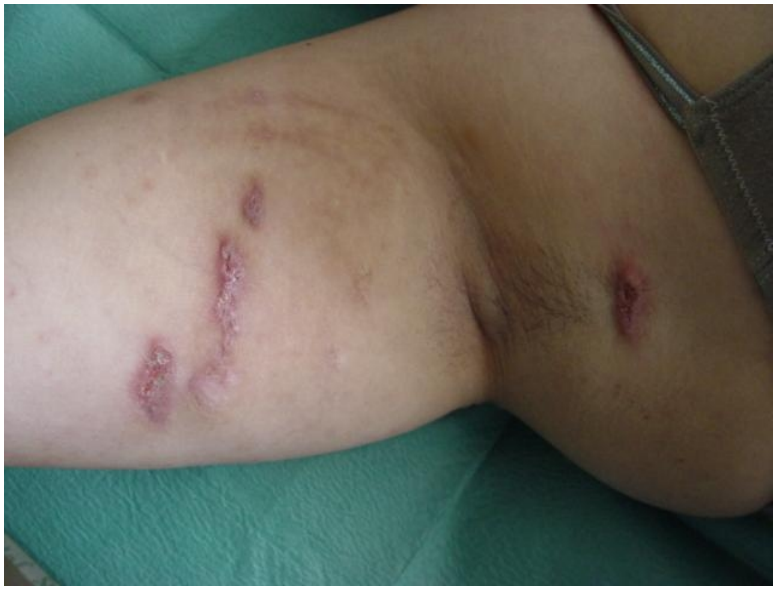

Board No: P-051
Ref. No: 85
Topic: Vasculitis

SERUM ADIPONECTIN AND VASPIN LEVELS IN BEHÇET'S DISEASE

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Background: Behçet's disease (BD) is a chronic systemic inflammatory disorder characterized by vasculitis that can affect all types and sizes of vessels in any localization. It is known that cardiovascular disease occurs at a significantly higher rate in patients with chronic inflammatory diseases. Adipocytokines exert insulin-sensitizing and anti-atherogenic effects. Adiponectin inhibits the expression of adhesion molecules and attenuates neointimal thickening in mechanically injured arteries. Moreover, vaspin a novel adipocytokine has been reported to be inversely associated with carotid artery stenosis. Objectives: To determine serum adiponectin and vaspin levels and their associations with predictors of atherosclerosis in BD. Methods: The study involved 37 BD patients and 29 healthy controls (HC). Serum adiponectin and vaspin levels, homeostasis model assessment (HOMA-IR) index and common carotid intima-media thickness (IMT) were determined. Results: The adiponectin level and IMT were higher and vaspin level was lower in the BD group compared to the HC group (Table), but neither adiponectin nor vaspin levels was associated with the IMT. In the BD group, 17 patients were active and 20 patients were inactive. In the active BD subgroup, the adiponectin level was higher, while the vaspin level was lower than in the inactive BD subgroup (p=0.004 and p=0.020, respectively) and the HC group (p<0.001and p=0.003, respectively). Conclusion: Despite of increased adiponectin and decreased vaspin levels in BD, these adipocytokines are associated with neither HOMA-IR index nor IMT. Further studies are needed to understand the regulation of these adipocytokine and determination of their relations with predictors of atherosclerosis in BD.

Table: Demographics and laboratory data

	BD (n=37)	HC (n=29)
Age (years)	35.3±10.0	38.0±10.3
Female/Male	19/18[a]	23/6
BMI (kg/m^2)	23.3±3.9[a]	25.9±4.7
SBP (mm/Hg)	109±7[a]	102±16
DBP (mm/Hg)	68±6[b]	62±10
TC (mg/dl)	161±31[c]	206±36

LDL-C (mg/dl)	108±23[c]	134±28
HDL-C (mg/dl)	37±11[c]	52±12
TG (mg/dl)	108±42	128±47
FBG (mg/dl)	83±9	89±13
Insulin (IU/ml)[†]	6.77±5.13	6.77±3.31
C-peptide (ng/ml)	2.95±0.94[c]	1.94±0.58
HOMA-IR	1.43±1.15	1.49±0.75
ESR (mm/h)	39.8±31.6[c]	18.4±9.8
CRP (mg/l)	28.9±36.5[c]	6.5±7.9
IL-6 (pg/ml)[†]	6.78±14.53	3.64±4.92
TNF-α (pg/ml)[†]	17.9±17.5	11.4±9.5
Adiponectin (µg/ml)	45±28[c]	9.2±3.8
Vaspin (pg/ml)[†]	128±325[c]	434±343
IMT (mm)	0.675±0.073[c]	0.547±0.035

[†]Logarithmic transformations were applied to data with skewed distribution before entering statistical analysis.

vs. the HC group: [a]p<0.05, [b]p<0.01, [c]p<0.001.

Board No: P-052
Ref. No: 86
Topic: Vasculitis

SERUM SALUSIN-α LEVEL IN BEHÇET'S DISEASE

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[1]Firat University, Faculty of Medicine, Department of Rheumatology, [2]Firat University, Faculty of Medicine, Department of Cardiology, [3]Firat University, Faculty of Medicine, Department of Biochemistry .

Background: Endothelial cell activation and/or injury are the characteristic features of Behçet's disease (BD), a chronic systemic inflammatory disorder. Endothelial dysfunction is widely regarded as being the initial lesion in the development of atherosclerosis. Moreover, accelerated atherogenesis has previously been shown in inflammatory rheumatic diseases. Salusin-α and β are novel bioactive peptides. Salusin-α suppresses macrophage foam cell formation, and decreased salusin-α level has been reported in coronary artery disease.
Objectives: To assess serum salusin-α level and its association with predictors of atherosclerosis in BD.
Methods: The study included 37 BD patients and 29 healthy controls (HC). TNF-α, IL-6 and salusin-α levels, homeostasis model assessment of insulin resistance (HOMA-IR) index and common carotid intima-media thickness (IMT) were determined.
Results: The salusin-α level and IMT were higher in the BD group compared to the HC group (Table). But the salusin-α level was not directly associated with the IMT.
Conclusion: Serum salusin-α level is increased in patients with BD, although they have increased IMT. Salusin-α has been reported to have anti-atherogenic effects. However, it seems that salusin-α does not directly affect the atherogenesis in BD in our study. Further studies are needed in order to evaluate whether salusin-α might be a biomarker for subclinical atherosclerosis in BD.

Table: Demographics and laboratory data		
	BD (n=37)	HC (n=29)
Age (years)	35.3±10.0	38.0±10.3

Female/Male	19/18[a]	23/6
BMI (kg/m^2)	23.3±3.9[a]	25.9±4.7
SBP (mm/Hg)	109±7[a]	102±16
DBP (mm/Hg)	68±6[b]	62±10
TC (mg/dl)	161±31[c]	206±36
LDL-C (mg/dl)	108±23[c]	134±28
HDL-C (mg/dl)	37±11[c]	52±12
TG (mg/dl)	108±42	128±47
FBG (mg/dl)	83±9	89±13
Insulin (IU/ml)[†]	6.77±5.13	6.77±3.31
C-peptide (ng/ml)	2.95±0.94[c]	1.94±0.58
HOMA-IR	1.43±1.15	1.49±0.75
ESR (mm/h)	39.8±31.6[c]	18.4±9.8
CRP (mg/l)	28.9±36.5[c]	6.5±7.9
IL-6 (pg/ml)[†]	6.78±14.53	3.64±4.92
TNF-α (pg/ml)[†]	17.9±17.5	11.4±9.5
Salusin-α (pg/ml)	97.0±3.2[b]	94.2±2.5
IMT (mm)	0.675±0.073[c]	0.547±0.035

[†]Logarithmic transformations were applied to data with skewed distribution before entering statistical analysis.

vs. the HC group: [a]p<0.05, [b] p<0.01, [c]p<0.001.

Board No: P-053
Ref. No: 103
Topic: Vasculitis

Evaluation of Some Rheological Parameters in Patients with Behçet's Disease; Impact of Disease Activity on Blood Viscosity & Yield Stress

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BACKGROUND: BD is a multisystem disease characterized by a pro-thrombotic status which is not fully understood. Moreover, the contribution of rheological alterations has been seldom investigated in this disease. The present study aimed at studying some rheological markers namely blood viscosity and yield stress in these patients.

SUBJECTS & METHODS: The patients group comprised 25 BD patients (15 males & 10 females). The control group included 15 healthy volunteers matched for age and sex. Viscosity measurements were obtained at shear rate ranging from 38.4 sec^{-1} to 576 sec^{-1}; using a DV-II +Pro Brookfield Cone & Plate Viscometers Spindle CPE-42. As blood is a non-Newtonian fluid whose viscosity decreases as shear rate increases, we chose a single reading point representing viscosity at shear rate 115 sec^{-1} and temperature 37°C. Analysis of the flow curve was performed by applying Bingham plastic model.

RESULTS: In BD patients with activity, the mean blood viscosity (5.610±0.965 cp) was significantly higher than those without disease activity (3.647±0.573 cp) (p<0.001) and higher than the control subjects (4.113±0.379 cp) (p=0.003). Similarly, the Bingham Model based plastic viscosity was significantly higher in BD patients with activity, compared with those without activity (p<0.001) and the control group (p=0.031). The yield stress was significantly higher in BD patients

with activity (1.660±0.526), compared with patients with inactive disease (0.972±0.321) (p=0.007), and normal subjects (1.026±0.101) (p=0.011).

CONCLUSION: These abnormal rheological findings denote reduced RBCs deformability and can explain, at least in part, the pro-thrombotic status that can occur in some BD patients.

Board No: P-054
Ref. No: 108
Topic: Vasculitis

Detection of Cardiovascular System Involvement in Behçet's Disease using Fluorodeoxyglucose Positron Emission Tomography

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Background: Behçet's disease (BD) theoretically affects all sizes and types of vessels, however, there have been few reports describing the clinical efficacy of using 18F-fluorodeoxyglucose positron emission tomography (FDG PET) in patients with BD.
Methods: Eight patients who were registered at the BD Specialty Clinic of Severance Hospital between 2004 and 2008 underwent FDG PET scans to evaluate the cardiovascular presentations associated with BD. Medical records and FDG PET images of the patients were retrospectively reviewed to determine the clinical significance of the cardiovascular findings.
Results: The median quantitative FDG uptake index was 1.46 (range 0.58-2.61). FDG uptake was detected in multiple pseudoaneurysms, aortitis and arteritis associated with aortic regurgitation and aneurysmatic dilatation of the sinus of Valsalva, atherosclerotic change of the proximal ascending aorta associated with aortic regurgitation, and multiple pulmonary artery aneurysms. The quantitative FDG uptake intensity was significantly associated with the ESR level.
Conclusion: Because FDG PET scans the entire body, it may have clinical value as a baseline workup study for patients with BD who have cardiovascular presentations.

Board No: P-055
Ref. No: 113
Topic: Vasculitis

Major arterial aneurysms and pseudoaneurysms in Behçet's disease: results from a single center

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Objective. Behçet's disease (BD) with arterial involvement is closely correlated with mortality and morbidity due to life-threatening complications such as arterial occlusion and aneurysm rupture. We aimed to demonstrate clinical characteristics of BD patients with aneurysms and pseudoaneurysms in the major arterial systems.
Methods. Medical records of 30 BD patients diagnosed with aneurysms or pseudoaneurysms in major arterial systems were reviewed in order to investigate the clinical characteristics of BD, the sites and types of arterial aneurysms or pseudoaneurysms, laboratory test results, and responses to treatments.
Results. A total of 47 aneurysms and pseudoaneurysms (32 saccular aneurysms, eight fusiform aneurysms, and seven pseudoaneurysms) were detected in 30 patients. Most aneurysms and pseudoaneurysms (27 patients, 90%) had not ruptured. Symptomatic lesions presented in 21 patients (70%), whereas asymptomatic lesions were incidentally detected in nine patients (30%). Ten of the 30 patients (33.3%) presented two or more aneurysmal lesions. Recurrence was observed in five patients (16.7%) after treatment with stent graft (n=3), graft interposition (n=1), or graft embolization (n=1).
Conclusions. We suggest that BD patients diagnosed with major arterial aneurysms be further evaluated to detect possible associated venous or arterial thrombosis formations or aneurysmal lesions at other sites.

Board No: P-056
Ref. No: 114
Topic: Vasculitis

Vascular manifestations of Behçet's disease in Japan: a survey of two university hospitals
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Objectives: Differences of vascular features in Behçet's disease (BD) have not yet been compared among ethnic groups. Here, we analyzed vascular manifestations of BD in Japan.
Methods: We retrospectively reviewed clinical charts of 412 patients who fulfilled the 1987 Japanese criteria for BD and received care in two Yokohama City University hospitals from July 1991 to December 2007.
Results: We found that 26 patients (6%) presented with vascular lesions. The mean age of BD diagnosis and onset of vascular episode were 39.7 and 42.4 years, respectively. HLA-B51 was positive in 6 patients of 15 patients (40%). Arterial and venous lesions were found in 8 (31%) and 21 patients (81%), respectively, and 3 patients (12%) had both. Pulmonary artery occlusion was the most common type of arterial lesion (n=5, 19%), followed by aneurysm (n=2, 8%), and thrombosis (n=1, 4%). Deep vein thrombosis in the limbs was the leading phenotype of venous lesions (n=19, 73%), followed by central retinal vein thrombosis (n=1, 4%) and transverse sinus thromboses (n=1, 4%), respectively. In extravascular manifestations, ocular involvement was significantly infrequent in patients with vascular lesions when compared with the other BD patients (42% versus 66%, $p<0.05$), whereas gastrointestinal involvement was more commonly found (31% versus 9%, $p<0.001$). Three patients received surgical operation (12%). Prednisone and immunosuppressants (cyclophosphamide, azathioprine) were given to 13 (50%) and 7 patients (27%) respectively. Two patients (8%) received infliximab for concurrent gastrointestinal involvement.
Conclusions: Our study illustrates characteristics of vascular involvement in BD patients in Japan.

Board No: P-057
Ref. No: 120
Topic: Vasculitis

Comparison of clinical features between retinal vasculitis in Behçet patients and idiopathic retinal vasculitis
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Purpose: Retinal vasculitis is one of the common manifestations in Behçet disease. This study was aimed to compare clinical features between retinal vasculitis in Behçet patients and idiopathic retinal vasculitis (IRV).
Subjects & Methods: Medical records of patients with retinal vasculitis in Behçet patients and IRV with follow-up duration of 12 months or more were reviewed retrospectively. Clinical features including age, gender, HLA-B typing, fundoscopic and angiographic findings, treatment methods, and visual outcome were compared. Visual prognosis was evaluated by Kaplan-Meier survival analysis.
Results: 47 patients with IRV and 153 Behçet patients with retinal vasculitis were included. Behçet patients have the more bilaterality and HLA-B51 positivity than IRV patients. Fluorescein angiography found that diffuse macular leak and disc leak were more common in Behçet patients than IRV. While cystoid macular edema (CME) was common in both groups, the edema tended to be persistent in Behcet's disease even after anti-inflammatory treatment. Visual prognosis including cumulative risk of visual acuity 20/200 or worse at 2 years was significantly worse in Behçet patients.
Conclusions: Retinal vasculitis associated with Behçet's disease has different clinical and angiographic manifestations from IRV.
* This study was supported by a grant from the Ministry of Health and Welfare, Republic of Korea (A080588).

Board No: P-058
Ref. No: 122
Topic: Vasculitis

Mortality in Behçet's disease

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Objective. To report the long-term mortality in Behçet's disease (BD).
Methods. We analyzed the causes of death, the standardized mortality ratio (SMR), and the factors associated with mortality in a cohort of 817 patients, fulfilling the international criteria of BD, from a single center.
Results. Among 817 BD patients, 41 (5%) died after a median follow-up of 7.7 years. The mean (± SD) age at death was 34.6 ± 11.5 years with 95.1% of male. Main causes of death included major vessel disease (mainly arterial aneurysm and Budd-Chiari syndrome) (43.9%), cancer and malignant hemopathy (14.6%), and central nervous system involvement and sepsis (12.2%). The mortality rate at 1 and 5 years was of 1.2% and 3.3%, respectively. There was an increased mortality among the 15-24 years [SMR with 95% confidence interval, 2.99 (1.54-5.39)], and the 25-34 years, [SMR 2.90 (1.80-4.49)] as compared to age and sex matched healthy controls. The mortality decreased in patients older than 35 years [SMR, 1.23 (0.75-1.92)]. In multivariate analysis, male gender (HR: 4.94, CI: 1.53-16.43), arterial involvement (HR: 2.51, CI: 1.07-5.90), and a high number of BD flare (HR: 2.37, CI: 1.09-5.14) were independently associated with mortality.
Conclusions. The overall mortality in our Behçet cohort was 5% after a median follow-up of 7.7 years. Male gender, arterial involvement and the number of flare were associated with mortality in BD.

Board No: P-059
Ref. No: 135
Topic: Vasculitis

Vascular manifestations of Behçet's disease in Japan: a survey of 98 patients

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Objectives: Differences of vascular features in Behçet's disease (BD) have not yet been compared among ethnic groups. Here, we analyzed vascular manifestations of BD in Japan.
Methods: We collected data of 98 BD patients having vascular symptoms from 37 hospitals from wide range of Japan. Clinical features and backgrounds including vascular features were retrospectively analyzed by questionnaires.
Results: of 98 patients, 69 (70%) were male and 29 (30%) were female. The mean age of BD diagnosis and onset of vascular episode were 40.5 and 45.2 years, respectively. HLA-B51 was positive in 19 patients of 51 patients (37%). Arterial and venous lesions were found in 64 (65%) and 51 patients (52%), respectively, and 17 patients (17%) had both. Arterial aneurysm was the most common type of arterial lesion (n=41, 42%), followed by arterial thrombosis (n=20, 20%), and pulmonary artery occlusion (n=11, 11%). The most frequent type of venous involvement was deep vein thrombosis in the limbs (n=40, 78%), followed by inferior caval thrombosis (n=6, 6%), and superior caval thrombosis (n=4, 4%), respectively. Fifteen patients received surgical operation (51%). Prednisone and immunosuppressants (cyclophosphamide, azathioprine) were given to 69 (70%) and 23 patients (23%) respectively. Five patients (5%) received infliximab, by which one patient was treated for vascular involvement, and the other 4 patients were for concurrent gastrointestinal involvement.
Conclusions: Our study illustrates the features of vascular involvement of BD in Japan in part, though selection bias should be considered.

Board No: P-060
Ref. No: 144
Topic: Vasculitis

Interferon Treatment in Behçet's Disease and Change in Carotid Atherosclerosis:

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Aim

We are studying the effect of IFN-α on progression of carotid atherosclerotic plaque burden and cardiovascular function in Behçet's disease.

Background

Vascular involvement in Behçet's Disease is the leading cause of death with a prevalence of 25% (1). The pathological hallmark is enhanced inflammation causing endothelial dysfunction and vascular changes (2). Venous complications are more common than coronary artery disease and cerebrovascular events (1,3,4).

Materials and Methods

We prospectively recruited 30 patients who were referred from rheumatology and ophthalmology clinics. All patients who consented for the cardiovascular magnetic resonance (CMR) sub-study attended for 3 CMR scans; baseline (prior to randomisation), 6 months and 12 months. Each scan consisted of a carotid artery study, brachial artery reactivity assessment (BAR) and evaluation of cardiac function. All images were acquired on a 1.5 Tesla magnetic resonance scanner (Siemens, Erlangen, Germany).

Results

Baseline characteristic data from the patients who completed the study are presented below (table 1). There was no significant difference at baseline between the mean carotid wall volumes in the interferon versus the standard treatment arm, (701.0 ±133.8mm3 vs 738.1 ±187.1mm3; p=0.32). Similarly, there was no significant difference in the BAR between the groups (14.6 ±7.7% vs 10.8 ±4.6%; p=0.13). Cardiac parameters were also non-significantly different.

Future directions

The full set of data analyses from all the patients recruited are in progress to determine whether IFN-α treatment affects progression of atherosclerosis in Behçet's disease.

References

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	Standard N=16	Interferon N=14	p value
Age/ years, mean (SD)	41.2 (10.3)	37.4 (7.3)	0.266
Male	7 (43.8)	8 (57.1)	0.464
Disease duration/ years, mean (SD)	9.6 (9.2)	6.4 (5.4)	0.26
Smoker	6 (37.5)	6 (42.9)	0.765
Hypertension	7 (43.8)	4 (28.6)	0.389
Systolic BP/ mmHg, mean (SD)	133.0 (15.6)	127.0 (15.9)	0.307
Diastolic BP/ mmHg, mean (SD)	81.4 (10.7)	75.9 (11.6)	0.187
Diabetic	1 (6.3)	0 (0)	0.341
Creatinine/ mmol/l, mean (SD)	78.3 (14.9)	74.7 (12.3)	0.487
Framingham score, mean (SD)	6.9 (4.3)	5.1 (4.4)	0.346
Total cholesterol/ mmol/l, mean (SD)	5.6 (1.0)	5.0 (0.8)	0.105
HDL cholesterol/ mmol/l, mean (SD)	1.6 (0.4)	1.6 (0.6)	0.771
Azathioprine	5 (31.3)	3 (21.4)	0.544
Cyclosporin A	4 (25.0)	6 (42.9)	0.301

Prednisolone	12 (75.0)	11 (78.6)	0.818
Tacrolimus	1 (6.3)	0 (0)	0.341
Methotrexate	4 (25.0)	1 (7.1)	0.19
Mycophenylate	3 (18.8)	6 (42.9)	0.151
Oral ulcers	15 (93.8)	13 (92.9)	0.922
Genital ulcers	12 (75.0)	10 (71.4)	0.825
Ocular involvement	10 (62.5)	10 (71.4)	0.605
Musculo-skeletal involvement	14 (87.5)	13 (92.9)	0.626
Skin lesions	13 (81.3)	11 (78.6)	0.855
Headaches	8 (50.0)	4 (28.6)	0.232
CNS	0 (0)	0 (0)	NA
Vascular	1 (6.3)	1 (7.1)	0.922
Renal	0 (0)	0 (0)	NA

All values N (%) unless otherwise stated

Board No: P-061
Ref. No: 161
Topic: Vasculitis

Venous thrombosis in Behçet's disease: Study of 182 patients

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Introduction: The vascular injury in Behçet's disease (BD) affects mainly the veins (10 to 40% of cases), all the veins can be achieved.
Materials and methods: It is a retrospective study including patients treated for BD at the Internal Medicine Department of Monastir for a period ranging from 1990 to 2008.
Results: There were 111 men and 71 women with mean age at diagnosis of BD 29.15 years. Vascular disease was noted in 37.6% and was significantly more frequent in men (p = 0.003).Venous thrombosis was reported in 50 patients (27.47%). Their distribution was as follows: lower limbs (32 cases), Superior cava vein (7 cases), upper limbs (5 cases), inferior cava vein (3 cases), supra hepatic vein (1 case), ovarian vein (1 case) and spleen vein (1 case).VT were statistically more frequent in men (p = 0, 015) and were multiple in 15 patients. We noted in our series, 2 types of vascular complications: Pulmonary embolism (4 cases) and recurrent VT (6 cases). All patients were placed on anticoagulants with good evolution in 86% cases.
Conclusion:
The vascular tropism of the MB is obvious and is a situation with high mortality risk. A TV in a young, male and Mediterranean moreover, should suggest priority diagnosis of BD and lead to appropriate therapy.

Board No: P-062
Ref. No: 165
Topic: Vasculitis

LARGE VESSELS INVOLVEMENT IN BEHCET'S SYNDROME: A STUDY ON 182 TUNISIAN PATIENTS

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Background:Although vascular lesions are not listed among the International criteria for diagnosis of Behçet syndrome (BS), up to 25-35% of patients develop venous or arterial large vessel complications throughout their disease course. Management of vascular complications is still controversial.
Objectives: Report the demographic, clinical and therapeutical aspects of large vessels involvement in BS
Patients and methods: 182 patients (mean age 39 years, 111 males and 71 females) recruited from the Internal Medicine Department of Fattouma Bourguiba Hospital, all fulfilling the International Study Group Criteria for diagnosis of Behçet's Disease, were studied. Demographic, clinical, and therapeutic aspects were determined for patients with large vessels involvement.
Results: Fourth (46/182) of patients with BS had large vessels involvement. They were 37 males and 19 females. Vascular events occurred at a mean age of 32.46 (16-51 years). In 10 cases the event happened 3.9 years (1-12 years) before the

diagnosis of BS. Vascular manifestations were deep vein thromboses in 39 cases, arterial thromboses in 2 cases and arterial aneurysms in 7 cases. Arterial aneurysms affected pulmonary arteries in 5 cases, the mesenteric artery and the coronary arteries in the other two cases. Pulmonary embolism occurred in 7 cases. Thromboembolic disease was more frequent in men($p < 0.005$). Patients with peripheral deep vein thromboses were treated by anticoagulation. Steroids and immunosuppressive therapy were used in arterial aneurysms and in addition to anticoagulation in case of large vessel thromboses.
Comments: Mortality can be affected by large vessels involvement in BS. However its treatments still remains controversi

Board No: P-063
Ref. No: 173
Topic: Vasculitis

CEREBRAL BLOOD FLOW VELOCITY IN BEHCET DISEASE

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Background and aims: The aim of this study was to determine the possible blood flow velocity (BFV) changes in patients with Behçet Disease (BD), Neuro-Behçet (NBD), and control subjects, using Transcranial Doppler sonography (TCD).
Methods: Using a 2MHz probe, Mean BFV (mBFV) of intracranial arteries was measured in 18 patients with BD, 22 patients with NBD, and 35 normal subjects.
Results: Study included 75 subjects (38 male), with no significant age and sex difference between 3 groups (P=0.09 & P=0.811, respectively). There was no significant difference between mBFV of MCA, ACA, PCA, Terminal extracranial ICA, and vertebral arteries. Also, there was no significant difference in mBFV of Ophthalmic arteries and carotid siphons between BD and NBD patients; but there was significant difference in mBFV of Ophthalmic arteries (24.1±3.9 and 19.8±5.2, P=0.001) and Carotid Siphons (55.7±9.7 and 45.5±8.6, P=0.000) between patient group (BD & NBD patients) and control subjects, with higher flow velocity in the patient group. Also, there was a marginal difference in mBFV of Basilar artery between 3 groups (P=0.050), but the difference between NBD group and control subjects (49.3±10.3 and 42.5±8.9, P=0.035) was more significant than the difference between BD group and control subjects (47.6±7.2 and 42.5±8.9, P=0.067).
Conclusion: Difference in mBFV of Ophthalmic arteries and Siphons between patient and control group may be related to eye involvement in BD; but difference in mBFV of Basilar artery between NBD and control groups may be suggestive of more vulnerability of posterior circulation of the brain in BD.

Group	Mean	SD	
MCA	control	60.94	9.34
	BD	65.76	12.75
	NBD	63.90	13.98
ACA	control	50.87	7.04
	BD	52.64	8.57
	NBD	55.04	12.95
PCA	control	32.73	4.38
	BD	35.22	7.65
	NBD	32.62	4.13
Opht.	control	19.83	5.16
	BD	24.86	4.43
	NBD	23.57	3.48
Siph.	control	45.46	8.57
	BD	53.72	9.95
	NBD	57.38	9.40

TEICA	control	36.14	5.66
	BD	39.15	6.19
	NBD	38.26	7.65
VA	control	35.41	8.18
	BD	39.04	7.98
	NBD	37.66	9.26
BA	control	42.54	8.89
	BD	47.64	7.16
	NBD	49.25	10.33

Board No: P-064
Ref. No: 188
Topic: Vasculitis

Vitamin D status and Endothelial dysfunction in Behcet's disease
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Aim: Endothelial dysfunction (ED) is present in Behcet's disease (BD) and is recently shown to be associated with Vitamin D deficiency in healthy controls (HC). We aimed to evaluate the status of serum 25(OH)Vit D3 levels and its association with disease activity, endothelial function and carotis intima media thickness (CIMT) in patients with BD.
Methods: Forty-two BD (F/M: 29/13, mean age: 39.9 years) patients and 85 healthy controls (F/M: 50/35, mean age: 33.7 years) were studied. Endothelial function was evaluated by brachial artery flow mediated dilatation (FMD) and CIMT was measured with B-Mode ultrasound.
Results: Less than 50 nmol/L levels of 25(OH)Vit D3 were present in 66% (n=27) of BD and 36% (n=28) of HC and a significant difference was observed between the levels of 25(OH)Vit D3 among the groups (BD: 42 (9-112) vs HC: 56 (14-149), p=0,02). 25(OH)VitD3 levels were observed to be less than 50 mmol/L in 33% (10/13) of active patients (p=0,3). When active manifestations were separately analyzed, only joint involvement was associated with lower levels of 25(OH)VitD3 (p=0,04). CIMT and FMD were also significantly different between BD and HC [0.57 (0.35-9.26) mm vs 0.28 (0-0.52) mm and 5.2 (0.62-30.58)% vs 9.04 (-0.69-34.17)%, p:0,00 and p:0,02, respectively]. However, no correlation was observed between 25(OH)VitD3 and CIMT or FMD (r:0,1, p=0,3 and r:-0,09, p=0,6, respectively).
Conclusion: A high presence of Vitamin D insufficiency was observed in BD patients from Turkey. However, impaired vascular endothelial function did not correlate with vitamin D levels.

Board No: P-065
Ref. No: 189
Topic: Vasculitis

A one year prospective cost-of-illness-study among patients with Behçet's syndrome
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Objective: Our previous and retrospective cost analysis of Behçet's syndrome (BS) had shown a considerable economic burden for the health care system in Turkey (1). This time, we investigated the economic impact of newly registered patients followed prospectively for 1 year.
Methods: A total of 326 (175 M / 151 F) consecutive patients with BS had registered for the first time in a multidisciplinary outpatient clinic for BS between January and December 2007. Among these 326 patients, 208 (64 %) (102 M/ 106 F) who were living in or near to the center were studied prospectively for 1 year assess direct and indirect costs. Clinical characteristics of the 208 patients studied were similar to 118 patients in whom cost data were not available. Patients were interviewed with a standardized questionnaire concerning a) direct costs such as medication, laboratory or hospitalization fees and b) indirect costs such as lost workdays, wages and lodging or transportation expenditures at each visit within 1 year of follow-up.

Results: The mean age was 32 ± 10 years and median disease duration was 2 years [IQR: 6 months- 3 years]. The mean number of visits was 3.3 (min:2, max: 8). Direct and indirect medical costs are shown in Table 1. The mean direct cost of those with any major organ involvement was significantly higher than that of those with mucocutaneous and or joint disease (P <0.001). The direct cost accounted for the 84 % of the total cost. Medication expenses made up 90 % of the total direct cost.
Conclusions: Economic burden of BS is still high. Drug costs were still the major cost driver and their share (from 79 % to 90 %) had increased within 5 years. Our inclusion of patients with less severe disease (earlier disease, more female patients and patients with less vascular / neurological disease) may be responsible from our finding a lower cost. Prominent changes in the health care policies for the last 5 years might have also played role.
Reference:
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Table 1: Direct and indirect costs according to clinical subgroups

	Costs, mean ± SD per capita per year (in Euro)		
	Direct	Indirect	Total
Eye involvement (n =92)	1635 ± 2392	221 ± 482	1855 ± 2639
Neurological disease (n =7)	1334 ± 2131	58 ± 43	1392 ±2124
Vascular disease (n =14)	1244 ± 1788	97 ± 118	1342 ±1793
Mucocutaneous and/or joint disease (n =108)	154 ± 130	116 ± 192	270 ± 260
Overall (n = 208)	813 ± 1750	161 ± 352	974 ± 1928

Board No: P-066
Ref. No: 202
Topic: Vasculitis

FIRST REPORT OF THE COEXISTENCE OF BEHCET'S SYNDROME AND BUERGER'S DISEASE IN A SAME PATIENT

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Introduction:
Behçet syndrome (BS) is a chronic multisystem inflammatory disease. It is a vasculitis that can affect blood vessels of all sizes. Buerger disease (BuD) is also an inflammatory disease that can concern arteries and veins. However, BuD affects only small and medium size vessels and is usually confined to the distal circulation. The vessel wall is relatively spared in contrary to BS. BS and Bu can sometimes be confounded. Here is a first report of the coexistence of these two diseases in a same patient.
Observation:
A 47 male patient, with 17 years history of tobacco use, was diagnosed as having BS because of a bipolar aphtosis, necrotizing pseudofolliculitis and a deep venous thrombosis. 5 years after, he developed recurrent leg ulcers. The blood count was normal, antiphospholipid antibodies were negative and there was no deficiency in protein C, S and antithrombine III. There was also no resistance to activated protein c. Cardiac ultrasound was normal. Arterial doppler ultrasounography showed a distal arteriopathy confirmed by the angiography that revealed distal and segmental occlusive lesions of legs and arms. Our patient met the 5 diagnostic criteria of Shionoya. He was diagnosed as having also BuD. Smoking cessation was indicated. The evolution was marked by stabilization of ulcers.
Comments:
Because leg ulcers are not common in BS, other causes must be ruled out. Some vasculitis are classically considered as exclusion criteria for BuD but BS can't explain here this distal and segmental occlusive arteriopathy responsible of recurrent leg ulcers.

Board No: P-067
Ref. No: 203
Topic: Vasculitis

Diversity of skin symptoms in Behçet's disease

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Behçet's disease (BD) is a chronic, relapsing, multi-system vasculitis of unknown aetiology with a wide spectrum of clinical presentations. Skin lesions are a common symptom of the disease occurring in the majority of patients. Several sets of diagnostic criteria have been defined and all of these criteria have included the presence of "typical skin lesions" and a positive pathergy test. Typical skin lesions include erythema nodosum-like lesions, papulopustular or acneiform lesions and pseudofolliculitis. Other recognized skin manifestations, which have been reported in numerous case reports, are Sweet's syndrome-like lesions, pyoderma gangrenosum-like lesions, cutaneous vasculitis lesions which may present as palpable purpura, bullous or necrotizing lesions and acral purpuric papulonodular lesions, extragenital ulcers and superficial thrombophlebitis.
The two main nodular skin lesions – erythema nodosum and superficial thrombophlebitis – are equally frequent and may not be clinically differentiated.
Lesions offten occur in combination with other skin or organ symptoms (eg, erythema nodosum–like lesions and papulopustular eruptions, acneiform lesions and arthritis, superficial thrombophlebitis and thrombosis in large veins).
It is often difficult to recognize and diagnose the disease, because of the possibility that lesions mimick similar lesions occurring in other conditions.
The diagnosis therefore relies not only on the constellation of symptoms and signs, but also on histological assessment. Namely, it has been postulated that only those lesions documented by neutrophilic vascular reactions or leucocytoclastic vasculitis should be included as cutaneous lesions of BD.

Board No: P-068
Ref. No: 204
Topic: Vasculitis

Investigation of the integrity of venous vessels in Behcet's patients with no known vascular event by using Doppler ultrasonography

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Background&Aims: In Behcet's disease (BD), deep venous thrombosis (DVT) occurs primarily in the lower extremities. Total recanalization rate is low, so thrombotic segment could be detected by imaging afterward. For disclosing vein involvement, leg swelling is commonly queried in the history taking. However, there is no data about presence of "silent" thrombosis in BD. We aimed to investigate the integrity of venous vessels in BD, without known vascular event by using Doppler ultrasonography (DU).
Materials&Methods: The study population comprised of BD patients fulfilling ISG criteria and classified as lacking vascular involvement. Patients having past events revealed in the vascular questionnaire or physical findings attributable to vascular disease were excluded. Demographic features of study groups are presented in the Table.
Results: Various degree of venous insufficiency was detected in 74 patients in BD (74%), 24 out of 33 patients (72%) in AS and in 8 out of 34 (25%) in HC group. All were at the lower extremities and the frequency did not differ between BD and AS, while both were significantly higher than HC (p=0.001, and 0.004, respectively). Six patients with BD (6%) have chronic venous thrombi at the lower extremities and none in AS and HC. All thrombotic segments were at the popliteal vein.
Discussion: Vascular involvement is a poor prognostic factor in BD and warrants systemic immunosuppressive treatment. As a noninvasive method, DU of lower extremities may disclose "silent" thrombosis. Venous insufficiency in those patients should be considered cautiously as an indicator of vein involvement.

	Behcet Disease (BD)	Ankylosing Spondylitis (AS)	Healthy Control(HC)
Number of cases	100	33	34
Sex (male/female)	85/15	29/4	28/6
Age (mean±SD)	28.6±7.1	26.4±5.5	26.9±6.9

Board No: P-069
Ref. No: 232
Topic: Vasculitis

Acute myocardial infarction in a patient with Behçet's disease
[1]Ben Abdelghani Khaoula, [1]Yahyaoui Safia, [1] Jerbi Mouna, [1] Hajri Saida, [1] Hriz Anis, [1]Mahfoudhi Mediha, [1]Turki Sami, [1]Kheder Adel .
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A 25-year-old man, a known case of Behcet's disease with vascular complications: recurrent thrombophlebitis, was admitted with the diagnosis of acute anterior myocardial infarction which was complicated by a ventricular fibrillation. The patient did not suffer from any symptoms, myocardial infarction, or readmission in the nine-month follow-up. About 25 cases of myocardial infarction associated with Behcet's disease have been reported previously. Although coronary involvement is rare in Behcet's disease, it is especially important because it affects young individuals and often presents as acute coronary syndromes. The etiopathogeny, the causal relationship and the treatment are yet unknown

Board No: P-070
Ref. No: 233
Topic: Vasculitis

Pseudotumoral presentation of neurobehçet
[1]Ben Abdelghani Khaoula, [1]Jerbi Mouna, [1]Yahyaoui Safia, [1]Hriz Anis, [1]Hajri Saida, [1]Mahfoudhi Mediha, [1]Ben Taarit Chokri, [1]Turki Sami, [1]Kheder Adel .
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Neurological involvement occurs in 5.3% to 30% of patients with Behçet's disease. Neurological manifestations can be secondary to direct central nervous system involvement or vascular angeitis. Neurological pseudotumoral presentation is rarely reported. We report a case of 26-year-old woman suffering from left hemiplegia. MRI was performed and showed pseudotumoral lesion in the protuberance and the right cerebral pedicule. Oral and genital aphtous ulcers were found. HLA B 51 was positive. The patient was improved with steroid therapy.

Board No: P-071
Ref. No: 234
Topic: Vasculitis

Arterial aneuvrysm in Behçet's disease: 6 cases
[1]Ben Abdelghani Khaoula, [1]Hajri Saida, [1]Hriz Anis, [1]Yahyaoui Safia, [1]Jerbi Mouna, [1]Mahfoudhi Mediha, [1]Ben Taarit Chokri, [1]Turki Sami, [1]Kheder Adel .
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Behçet's disease is a systemic vasculitis generally involving in the venous system. Arterial manifestations, usually aneurysm or more rarely occlusion, are less common. We analysed 6 cases of Behçet's disease with arterial aneurysm complications. There were 5 men and 1 woman, mean age 35 years. Mean delay to arterial complications was 8 years after the first sign of the disease. Five patients showed evidence aneurysm of the pulmonary arteries. One patient developed an aneurysm of the aortic and iliac artery.
Phlebitis was associated with arterial involvement in 4 patients. Combined corticosteroid and cyclophosphamid therapy enabled regression of pulmonary aneurysm in 2 patients. One was operated with a favorable outcome. One patient was overlooked and 2 patients died of massive hemoptysis

Board No: P-072
Ref. No: 244
Topic: Vasculitis

The Investigation of Varicocele and Epididymitis in Behcet Disease
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Introduction: Varicocele, the abnormal dilation of the spermatic veins, is associated with an increased risk in male infertility. Behcet disease (BD) is a systemic vasculitis, therefore can involve epididymis and scrotal veins. We aimed to investigate frequency of varicocele and epididymitis in patients with BD.

Methods: The study groups comprised of 47 BD, 31 ankylosing spondylitis (AS) patients and 31 healthy controls (HC). Scrotal physical exam and Doppler ultrasonographic exam were performed by a blind clinician and a radiologist, respectively. Measurements of thickness of epididymis and its echogenicity were used for diagnosis of epididymitis.

Results: Varicocele was detected in 57.4% (n=27) in BD, 29.0% (n=9) in AS and 41.9% (n=13) in HC with physical examination and in 55.3% (n=26) in BD, 35.5% (n=11) in AS and 29% (n=9) in HC with Doppler ultrasonographic exam ($p<0.05$). Comparing to HC, Odds ratio for developing varicocele in BD was 3.02. Physical examination showed moderately agreement with the results of Doppler ultrasonographic exam (kappa value=0,609). Eight patients (17%) with BD had epididymitis, whereas no cases with epididymitis was detected in AS and HC groups.

Conclusion: It seems that the frequency of varicocele and epididymitis were increased among patients with BD compared to AS and HC. Although clinical significance of this finding may need to be investigated further, given the known negative effects of these disorders with respect to male infertility, it should be recommended that BD patients should be investigated for varicocele and epididymitis at least by a careful physical examination.

Board No: P-073
Ref. No: 249
Topic: Vasculitis

A Case Report and Review of Behcet's Disease in the African American Population: Is There a Higher Prevalence of Vascular Manifestations Among African Americans?
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Behcet's Disease (BD) is a multi-systemic syndrome most commonly afflicting populations along the "Silk Road," and rarely reported in the African American (AA) population. In this abstract, we present a case of BD with pseudoaneurysm in an AA male.
A 28 year-old AA male presented to an urban emergency room (ER) with 1 week of fevers and severely painful oral and genital ulcers. His past medical history was significant only for a post-traumatic femoral pseudoaneurysm that was repaired by coiled embolectomy. The physical exam recorded from this previous admission revealed chancroid lesions on his penis. When he returned to the ER 1 year later, he was found to have severe mucosal ulcerations with areas of hemorrhage and necrosis in his mouth, oropharynx, and penis, and a papulopustular rash in his upper extremities. Pathergy test was positive and the diagnosis of BD was made. The patient improved with colchicine and prednisone.
Previous case series have described BD in West African and Afro-Caribbean patients. There is a paucity of BD reports in the AA population, however, and a review of these cases is summarized below. Although this may represent reporting bias, a high prevalence of vascular manifestations is apparent among AA patients. Because of possible genetic drift, larger epidemiologic studies are necessary to determine the prevalence and natural history of BD in the U.S. If there is a higher prevalence of vascular disease among AA patients, then physicians must be educated for early diagnosis.

Authors	Patients	Sex	Age	Vascular Involvement	Other Manifestiations	Treatment	Location	HLA-B51
Pandrea, et al. 2007	1	M	26	Axillary/common carotid aneurysm Subclavian DVT	Hypopyon, retinal vasculitis Pathergy positive	Cyclophosphamide Infliximab Prednisone	CT	Negative
Endo, et al. 2007	1	F	13	Bilateral pulmonary artery aneurysm Popliteal artery occlusion	Cardiogenic thrombi	Infliximab	AL	Unknown
Gallardo, et al. 2004	1	F	28	None	Hypopyon, panuveitis, necrotizing retinintis Post-steroid-injection hypopigmentation	Injected traimcinolone IV steroids Etanercept Methotrexate Cyclophosphamide Cyclosporine	TX	Unknown
Cohle, et al. 2002	1	M	10	Bilateral pulmonary artery aneurysm	Endocardial fibrosis	Died prior to treatment	MI	Unknown
Ecker, et al. 2000	1	M	20	None	Vitritis, peripheral retinitis, hypopyon Oral-genital ulcers Pathergy negative	IV Solumedrol Optical corticosteroids Oral prednisone	VA	Positive
Winer-Muram, et al. 1994	2	M	20s	Bilateral pulmonary artery aneurysm Pulmonary artery infarction	Pathergy negative	Unknown	Unknown	Unknown
Puckette, et al. 1994	1	F	35	Bilateral pulmonary aneurysms	Oral-genital ulcers	Oral prednisone Colchicine	VA	Unknown

Board No: P-074
Ref. No: 265
Topic: Vasculitis

Some Manifestations Disappear Earlier than Others in Behçet's Syndrome

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We had observed that the frequency of all mucocutaneous lesions decreased in frequency with time (1). In this study we investigated whether some manifestations disappeared before others.
Patients and Methods:
We studied consecutive BS patients who were seen in the rheumatology outpatient clinic between February 2009 and April

2010. Only BS patients whose disease duration and follow-up were 10 years or longer were included in the study. Patients were asked whether skin mucosa lesions, arthritis and uveitis attacks occurred for the preceding one year. If not, they were asked about the date when the manifestation occurred for the last time. Also a pathergy test was done to those who volunteered.

Results:

We studied 115 (60 M, 55 F) patients. The mean age of the patients was 48 ± 8 and the mean disease duration 20 ± 6 years. 63 patients had eye, 20 vascular and 6 neurological disease. Vascular and neurological attacks were not evaluated due to rarity. The frequency of those with any BS manifestation during the preceding one year including pathergy positivity was decreased significantly at the final visit compared to that found at the beginning (Table). As seen in the Kaplan-Meier curve (Figure), eye attacks were the first to cease followed by arthritis, genital ulcers and skin lesions. Oral ulcer was the most frequent lesion after 30 years of disease course.

Conclusions:

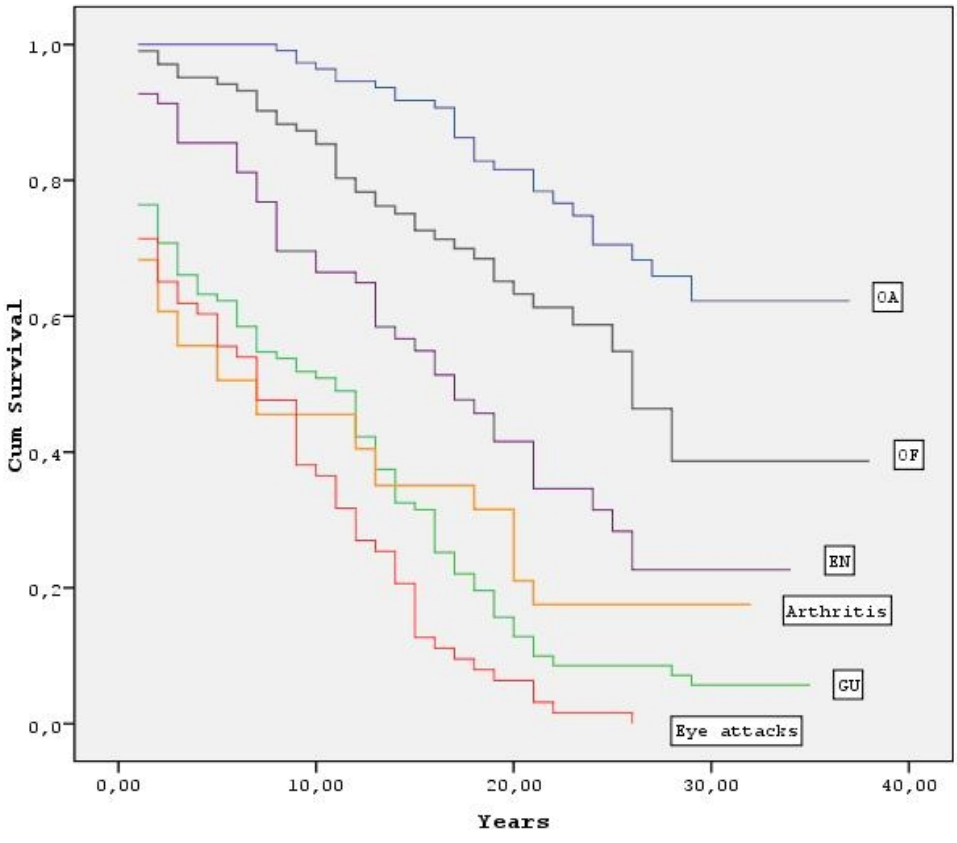
In BS some disease manifestations disappear earlier than others. These findings may have important pathogenetic and clinical implications.

Reference:

1) Kural-Seyahi et al. Medicine 2003.82:60-76.

Frequency n (%)	Initial visit	Final visit	P
Oral ulcers	115 (100)	84 (73)	< 0.001
Genital ulcers	105 (115)	13 (11)	< 0.001
Papulopustular lesions	104 (90)	61 (53)	< 0.001
Erythema nodosum	72 (63)	23 (20)	< 0.001
Arthritis	39 (34)	8 (7)	< 0.001
Pathergy positivity	85/109 (78)	6/39 (15)	< 0.001

Kaplan Meier survival plot for duration of BS manifestations



OA: oral ulcer, OF: papulopustular lesions, EN: erythema nodosum, GU: genital ulcer,

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Ref. No: 266
Topic: Vasculitis

Venous Severity Assessment in Behçet's syndrome

[1]Osman Serdal Cakmak, [2]Emire Seyahi, [3]Fatih Kantarci, [1]Ayla Yesilova, [3]Ayse Seda Ergin, [3]Atilla Suleyman Dikici, [4]Hasan Tuzun, [2]Hasan Yazici .
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Lower extremity venous thrombosis (LEVT) is the most common type of vascular involvement in Behçet's syndrome (BS). Little data is available on its clinical and radiological evaluation. We assessed clinical characteristics and severity scores among BS patients with LEVT and suitable controls.
Patients and Methods:
We studied 66 (63 M/3 F) BS patients with LEVT and 39 (36 M/ 3 F) BS patients without LEVT. Also 37 (20 M/ 17F) non–BS patients with LEVT who were followed by the vascular surgery outpatient clinic were included. Venous severity was assessed using venous clinical severity scoring (VCSS) used frequently by vascular surgeons to determine the severity of chronic venous insufficiency (1). Intermittent claudication was assessed (2). Venous Doppler USG of lower extremities was done in all patients with LEVT.
Results:
As shown in the Table, BS patients with LEVT had significantly more severe lower extremity disease in every aspect compared to non-BS patients with LEVT. Popliteal and femoral veins were the most commonly involved veins among both BS and non-BS patients with LEVT. The mean number of thrombosed veins was significantly higher among BS patients (5.2 ± 2.9) than non-BS patients (3.8 ± 2.3) ($P < 0.01$).
Conclusions:
Venous thrombosis due to BS run a more severe disease course compared to those due to non-BS causes. VCSS may be a useful disease assessment tool in BS.
Reference:
1) Rutherford et al. J Vasc Surg 2000.
2) Rose GA, et al. Br J Prev Soc Med 1977.

	BS patients with LEVT, (n = 66)	BS patients without LEVT, (n =39)	Non-BS patients with LEVT, (n =37)	P
Mean age ± SD, years	37 ± 10	36 ± 8	42 ± 13	< 0.001
Intermittent claudication, n (%)	24 (36)	1 (3)	5 (14)	< 0.001
Pain, n (%)	49 (74)	13 (33)	22 (60)	< 0.001
Varicose veins, n (%)	45 (68)	0	14 (38)	< 0.001
Venous edema, n (%)	40 (61)	1 (3)	14 (38)	< 0.001
Skin pigmentation, n (%)	38 (58)	0	5 (14)	< 0.001

Inflammation, n (%)	51 (77)	0	3 (8)	< 0.001
Active ulcers, n (%)	11 (17)	0	1 (3)	< 0.001
Venous clinical severity score, mean ± SD	6.8 ± 5.0	0.4 ± 0.7	4.3 ± 3.6	< 0.001

Board No: P-076
Ref. No: 274
Topic: Vasculitis

Vena cava thrombosis in Behçet's disease. About 6 cases
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AIM: To study the clinical characteristics and the evolution of vena cava thrombosis (VCT) in Behçet's disease (BD), as well as their association with other severe symptoms.

PATIENTS AND METHODS: Among 152 BD, we selected those with VCT. All patients fulfilled the diagnostic criteria of the international study group of Behcet's disease. Different clinical and paraclinical parameters were determined and evaluated the severity of BD in these patients. Protein C, protein S, antithrombin, homocyctein and anticardiolipin antibody (aCL) levels were measured .

RESULTS: Six patients had a vena cava thrombosis (4%). They were all male with an average age of 43 years (range: 28-57). We had 4 cases of superior vena cava thrombosis, 2 cases of inferior VCT one associated to Budd Chiari syndrom and the second to deep venous thrombosis. The average delay to diagnosis of the VCT from the date of the BD diagnosis was 12 years (range: 9 months-14 years). All patients were clinically symptomatic and the installation of the symptoms were progressive and insidious in all cases. Protein C, protein S and antithrombin and homocyctein levels were normal in all cases. No patient was positive for IgG aCL or for a beta 2GPI.

All our patients were treated by anticoagulation therapy and high-dose prednisone combined to colchicine. One patient died due to pulmonary embolism. The 5 others are clinically improved (83%) after an average 8 year course.

Board No: P-077
Ref. No: 276
Topic: Vasculitis

Behçet's disease and hypereosinophilic syndrome:a case report
[1]Mabrouk Khalifa, [1]Chiraz Atig, [1]Neirouz Ghannouchi, [1]Amor Alaoua, [1]Elhem Benjazia, [1]Ahlem Braham, [1]Amel Letaief, [1]Fethi Bahri .
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Introduction: The hypereosinophilic syndrome is a multisystem disease characterized by infiltration of eosinophils in bone marrow, heart, and other organs. It has occurred in association with several specific disorders, including parasitose, some malignancies and vasculitis. We report the case of a patient with co-existing Behçet's disease (BD) and hypereosinophilic syndrome.
Observation: A 32-year- old woman was followed for BD since 5 years. BD diagnosis was based on recurrent oro- genital ulceration and an episode of venous thrombosis treated by Acenocoumarol for three months. She was admitted to our hospital in March 2007 for abdominal pain and diarrhoea. Physical examination has found a lungale aphtose and an ascites of medium abundance. A numeration of the bloody formula revealed a hypereosinophilia at 10100 elements/mm3; as well 97% of eosinophiles were objectived in the liquid ascites. A myelogram objectived a hyperplasy of the eosinophile line with an absence of blasts. The digestive endoscopy and the parasitological examination of excrements were normal. The diagnosis of idiopathic hypereosinophilic syndrome was retained, and the patient was treated by corticotherapy at the dose of 1mg/kg/day.

The evolution was marked by the disappearance of ascites and the normalisation of the bloody numeration, without relapse.
Conclusion: Behçet's disease and hypereosinophilic syndrome could have common symptoms such as mucosal ulcers involving the mouth and genitalia. The co-existing of these two deseases and their physiopathologic links remained unclear.

Board No: P-078
Ref. No: 284
Topic: Vasculitis

Assymmetric dimethylarginine serum levels in patients with Behçet's disease
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Introduction:
It is now widely accepted that endothelial dysfunction as a result of reduced bioavailability of nitric oxide (NO) plays a central role in the pathogenesis of vascular disease. The assymmetric dimethylarginine (ADMA) is an endogenous inhibitor of NO synthase (NOS), involved in endothelial dysfunction. One of the prominent features of Behçet's disease (BD) is vasculitis and thrombosis as a result of endothelial dysfunction.

Objective:
To define level of ADMA in plasma of BD patients and its relation with the main manifestations of the disease.

Patients and methods:
A case–control study was carried out using 78 unrelated Tunisian BD patients who were classified according to The International Study Group criteria for the disease. Seventy nine healthy control subjects from a similar ethnic background were age and sex matched. Exclusion criteria were hepatic and renal diseases (chronic renal failure), thyroid disease, diabetes, essential hypertension, congestive heart disease.
ADMA measurement was carried out by employing an ELISA technique. The normal values are ranged between 0.4 and 0.75 µmol/l. Statistical analysis was performed with Statistical Package for the Social Sciences for Windows (SPSS Inc, version 11, Chicago, IL, USA).

Results:
Fifty three male and 25 female represent the group of patients with BD. The mean age was 39.3 ± 1.2 years.
Seventy six patients had oral aphtosis and 63 had genital aphtosis (80.7%). Cutaneous lesions were present in 77 patients (98.7%). Thirty six of the 78 patients (62.7%) had ocular manifestations. Twenty three patients (29.4%) complained of neurological symptoms.
Plasma ADMA levels were significantly elevated (3.28 versus 0.6 µmol/l; $p < 0.05$) in BD subjects compared with controls. Plasma ADMA level were higher in patients with neurological manifestations (5.73 versus 2.25 µmol/l, $p=0.09$). Regarding other BD manifestations, plasma ADMA level did not differ significantly.

Conclusion:
Elevated ADMA level in serum may be responsible for the endothelial damage in BD and therefore will be correlated to the disease.
In this study plasma ADMA concentration was significantly higher in BD patients and associated with neurological manifestations. In our best knowledge, such results were not reported in other ethnic grou

Board No: P-079
Ref. No: 291
Topic: Vasculitis

Clinical features at diagnosis in 98 Behçet patients
[1]Bel Feki Nabil, [1]Alaoua Amor, [1]Khalifa Mabrouk, [1]Ghannouchi Neirouz, [1]Hasni Yosra, [1]Letaief Amel, [1]Bahri Fethi .
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Background: Behçet disease is a multisystem inflammatory disorder characterized by recurrent oral ulcers, genital ulcers and ocular inflammation, and which frequently involves the joints, skin, central nervous system (CNS) and gastro intestinal tract.
Objective: the aim of our study was to review the clinical features at diagnosis of 98 patients with Behçet's disease.

Patients and Methods: We report 98 cases with Behçet's disease diagnosed at the internal medicine department between 1985 and 2010; the diagnose was made using the international criteria for classification of Behçet's disease.
Results: The male to female ratio was 1.88. the mean age at onset of disease was 35.3 years (range 12 to 61 years).Oral lesions were present in 52%, genital aphtosis in 39.8%,bipolar ulcers in 34 %, Joint involvement in 31.6% ocular lesions in 20%,neurological manifestations in 18% and vascular in 8.16%
Conclusion: Our data show that the major symptoms at diagnosis in our patients were benign

Board No: P-080
Ref. No: 292
Topic: Vasculitis

Venous involvement in Behçet's disease
[1]Bel Feki Nabil, [1]Alaoua Amor, [1]Khalifa Mabrouk, [1]Ghannouchi Neirouz, [1]Bouker Ahmed, [1]Letaief Amel, [1]Bahri Fethi .
[1]Department of Internal Medicine, Farhat Hached University .

Background: Behçet's disease is a chronic relapsing systemic vasculitis, involving both arteries and veins of various sizes.
Objectives: The aim of this study was to determine the prevalence and characteristics of venous involvement in Behçet's disease. Among 98 patients seen at the internal medicine department between 1985 and 2010, 15 (17, 2 %) had venous involvement and were included in our retrospective study. All these patients fulfilled International Study Group criteria for BD.
Results: Men with BD were more likely to have venous involvement (10/64) than women (5/34) and were younger at diagnosis of vascular disease (24, 6 vs. 44 years). Many patients had venous manifestations at more than one site : 11 had deep venous thrombosis, 2 had superficial venous thrombosis, 5 had cerebral thrombophlebitis, 1 had diffuse thrombosis(mesenteric vein, portal vein,caval thrombosis).Combined treatment with glucocorticoids and anticoagulants was effective in the most venous manifestations.
Conclusion: Venous manifestations of BD are common in our patients, particularly deep venous thrombosis in lower extremities. Combined glucocorticoids and anticoagulants is the major treatment of thrombosis in Behçet's disease.

Board No: P-081
Ref. No: 293
Topic: Vasculitis

Arterial involvement in Tunisian patients with Behçet's disease: About 8 cases
[1]Ghannouchi Jaafoura Neirouz, [1]Turki Emna, [1]Khalifa Mabrouk, [1]Alaoua Amor, [1]Ben Jazia Elhem, [1]Krifa Ahlem, [1]Letaief Amel, [1]Bahri Fethi .
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Introduction: Venous thrombosis is the most common manifestation (30 % of cases) for vascular involvement in Behçet's disease (BD). Arterial involvement (AI) is rare (2.7 to 7 %) and it's associated with poor prognosis and a high mortality.
Purpose: describe clinical features, treatment and prognosis of AI in Tunisian patients with BD.
Patients: Among 114 patients fulfilled International Study Group criteria for BD, seen between 1985 and 2009 in the department of internal medicine in Sousse, we studied retrospectively patients with AI (aneurysm and/or arterial occlusion).
Results: Eight patients (6 men and 2women, mean age 29.6 years) had AI (7%). We found 5 aneurysms formation (pulmonary arteries in 4 cases, abdominal aorta in one case) and 3 thrombosis (Brain infraction) with a median delay respectively of 2.2 and 6.5 after BD diagnosis. Combination of venous manifestations occurred in 4 patients. Neurological symptoms were frequently noted in patients with AI (6/8). Five patients underwent corticosteroids and immunosuppressive therapy and two received only corticosteroids. Two patients were treated with pulmonary embolism and one was successfully operated for aneurysms. One patient died of massive hemoptysis. Seven patients were alive after a mean of 2 years follow-up
Conclusion: Aneurysm formation is more common than arterial occlusion. Men with BD were more likely to have AI than women. AI in Behçet's disease raises treatment challenges because the lesions tend to recur and can cause complications.

Board No: P-082
Ref. No: 308
Topic: Vasculitis

Mean Platelet Volume in Patients with Behçet's Disease

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[1]Süleyman Demirel University Department of Internal Medicine Division of Rheumatology, [2]Süleyman Demirel University Department of Internal Medicine, [3]Süleyman Demirel University Department of Internal Medicinedivision of Endocrinology, [4]Süleyman Demirel University Department of Cardiology .

Venous as well as arterial thrombosis is a common complication of BD but exact pathogenetic mechanism of the thrombotic tendency is not well known. Although the coagulation system, fibrinolytic activity, and thrombophilic factors were studied extensively in patients with BD, platelet function which is a major component of hemostasis and thrombosis, was evaluated only in a few reports. Mean platelet volume (MPV) is an indicator of platelet activation and that larger platelets have higher thrombotic potential. The aim of this study was to assess the MPV in patients with BD.

We studied 39 patients (18 M/ 21F; mean age 38.7±10.4 years) and 30 control subjects (10M/ 20F; mean age 34.6±7.6 years). In all study group body mass index (BMI) calculated, systolic and diastolic blood pressure (SBP, DBP) and smoking habits were all recorded. Fasting plasma glucose, serum lipids, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), platelet counts and mean platelet volumes were detected.

There were no differences between two groups according to BMI, SBP, DBP, serum lipids and platelet counts. However, the MPV values of patients were found significantly higher than those of the controls (8.6±0.8 fl versus 8.0±0.8 fl; p=0.004, respectively). The platelet count detected was not different significantly between patient and control groups. However, ESR and CRP levels were found significantly high in patients with BD (p=0.01, p<0.001, respectively).

In conclusion, our results suggest that patients with BD tend to have an increased platelet activation. MPV may be considered as a useful hematological parameter for early and easy identification of patients at higher risk of thrombosis. Further data are needed to clarify the role of MPV in patients with BD.

	BD(n=39)	Control(n=30)	P value
Age (years)	38.7±10.4	34.6± 7.6	0.01
Gender (M/F)	18/21	10/20	0.28
BMI ($kg/m^{2.}$)	26.7±3.0	25.7±3.9	0.21
SBP (mm Hg)	125.1±15.1	120.6±8.6	0.15
DBP (mm Hg)	77.9±8.3	75.2±9.5	0.21
Smoking (%)	2 (5 %)	3 (10 %)	0.43
Glucose (mg/dl)	92.7± 12.9	88.8±9.5	0.16
Triglycerides (mg/dl)	113.6± 42.5	108.6± 59.9	0.69
LDL-cholesterol (mg/dl)	110.2±29.7	110.7±34.3	0.94
ESR (mm/h)	16.0±12.2	9.7±3.7	0.01
CRP (mg/dl)	12.3±18.3	2.2±0.8	< 0.001
Platelet count ($\times 10^9$)	270.7±76.1	243.9±60.6	0.11
MPV (fl)	8.6±0.8	8.0±0.8	0.004

Board No: P-083
Ref. No: 311
Topic: Vasculitis

Progesterone as a possible factor in Behcet Disease attacks
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Abstracts:
The factors that aggravate the activation in BD are still unclear. In this report, we aimed to draw attention to progesterone as a possible symptom activating factor in BD by a Behcet case with clinical activation after progesterone administration.
Case:
A 27 year-old female patient with BD presented to our clinic because of a new developed erytema nodosum, many oral ulcerations, thrombophlebitis and pustular lesions. She was diagnosed as BD for 7 years. She was symptom free except from minor oral ulcers with colchicine and benzathine penicillin for two years. Many oral ulcerations, pustular lesions, thrombophlebitis and erythema nodosum were developed after approximately one week receiving Progesterone capsule 200 mg due to menstrual irregularities. She was advised to stop progesterone and her symptoms disappeared approximately within three weeks. Her gynecologist administered progesterone again for the next period. Same symptoms were seen in our patient again. Similarly, after stopped progesterone again approximately three weeks later symptoms disappeared.
Discussion:
Some medications have been considered as related factors in the activations of BD. The progestins could aggravate acneiform lesions. In several studies, phelotrombosis has been seen to be caused by medroxyprogesterone acetate. It has been observed that in veins, progestins may lead thrombosis. In arteries, progestins may act as vasoconstrictors at sites of injured endothelium. Also hormones have been considered as affecting factors in BD and progesterone has been considered as a major hormone influencing the course of BD. Our results may contribute the studies showing possible oral progesterone effect in BD.

Board No: P-084
Ref. No: 312
Topic: Vasculitis

BEHCET DISEASE PRECEDED BY FEVER OF UNKNOWN ORIGIN
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Introduction: We report a case of Behcet's disease (BD) that present as fever of unknown origin.
Case report: A 30 year-old-man was with six weeks history of fever and pain in the left legs was admitted. He had suffered from both genital and oral ulcers over five months A color Doppler sonography showed thrombosis of left common femoral veins. The fever resolved when glucocorticoid and anticoagulant therapy was given.
Conclusion: BD is an extremely rare cause of protracted fever of unknown origin. However, BD should be considered in patients from the Mediterranean rim, and a fever in a patient with suspected or documented BD should prompt a search for thromboembolism

Board No: P-085
Ref. No: 31
Topic: Regional

Prick Test with Self-saliva as an Auxiliary Diagnostic Measure in Behcet's Disease
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Behcet's disease (BD) generally starts with oral aphthous ulceration followed by the systemic disorder. Although it is not so difficult to make a diagnosis for typical cases, some difficulties occur in cases without apparent aphthous ulceration. We have

previously demonstrated a trial of the prick test with oral streptococcal antigens as one of diagnostic ways, because BD patients show hypersensitivity against streptococci.
It is well known that saliva of BD patients contains many oral streptococci including uncommon serotype of S. sanguinis. We performed the prick tests with self-saliva and saline as a control on the forearm of 7 BD patients, 3 recurrent aphthosis patients with rheumatoid arthritis (RA), a herpetic oral ulcer patient, 2 non-BD patients with erythema nodosum, and 5 healthy controls. Five of 7 BD patients and a RA patient showed erythematous reaction (more than 10mm in diameter) with a small pustule 48 hours after the prick test with self-saliva. The trial of the self-saliva prick tests seemed to be more helpful for the diagnosis of BD compared with the prick test with streptococcal antigens. This method may also suggest that the pathergy reaction seen in BD patients is due to the reaction toward the bacteria on the skin surface, although this hypersensitivity has been believed as one of clinically mysterious characters when making a diagnosis for this disorder.

Board No: P-086
Ref. No: 32
Topic: Regional

Clinical, Cytological, Cytogenetic and Biochemical Analysis of Behcet's disease and recurrent aphthous ulceration in Iraqi patients

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Background: Behcet's disease (BD) is a chronic multisystemic vasculitic inflammatory disorder of undetermined etiology
Objective: To determine and compare the clinical pattern, biochemical markers, cytological presentation of exfoliating epithelial cells and the chromosomal status of Iraqi BD patients with that of recurrent aphthous ulcerations.
Methods:Seventy two patients who were suffering from BD,forty one patients from RAU and Thirty five individuals were considered clinically as healthy control .Saliva was collected from patients and healthy control for the biochemical analysis of enzymes such as ALP, CK, LDH and TSP and essential trace elements (Zn, Ca, Mg and Fe). Furthermore, cytocentrifuging was used to study the exfoliating epithelial cells.
Heprenized blood samples of BD patients were cytogenetically investigated to study the chromosomal status.
Results: Statistically significant increased salivary ALP was seen in both BD&RAU groups as compared to healthy group, Furthermore, Statistically significant increase salivary CK was observed when compared BD group with that of RAU, while highly significant decrease was noticed when comparing RAU &BD with HC group. There was statistically significant decrease in Saliva zinc in BD as compared with RAU and HC.Statistically significant higher level of salivary iron was seen in BD group when compared with RAU and HC.saliva of BD patients showed an increased number of superficial epithelial cells with pyknotic nuclei while patients with RAU showed an increased number of intermediate epithelial cells

Board No: P-087
Ref. No: 34
Topic: Regional

Re-evaluation of pathergy test in Iraqi patients with behcet disease

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Background : pathergy test was introduced as an important criteria in diagnosis of Behcet disease . The aim of the present work is to reevaluate this important test and especially to perform this reaction on oral mucosa and legs .
Individuals and methods : Thirty five Arab patients with untreated Behcet disease, their ages ranged between 21-51 years with male to female ratio of 3/2 .
For 15 patients, pathergy tests were performed on left and right forearms using G27, 23,20 needle from above down ward and for remaining 20 patients on both right and left legs and forearms .Using G 20 needle lower lip mucosa pathergy test were performed for all patients .
35 individuals with other dermatological disease were taken as control group by doing pathergy tests for 15 of them on both right and left forearms and for remains 20 patients, bilateral legs and forearms pathergy were done using G 20 needle and for all control group lower lip mucosa pathergy was done using Gauge 20 .
Results :The ordinary pathergy test was positive in 65.7% of all patients & on oral mucosa in 45.7% . There was no

difference in correlated gauge of needle.
The sensitivity and degree of positivty of the test on legs was more than that on forearms although statistically not significant (T=0.083).
Conclusion : pathergy test is better to be carried out on legs using G20 and on any side with no difference .

Board No: P-088
Ref. No: 35
Topic: Regional

Re-evaluation of pathergy test in Iraqi patients with behcet disease
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Background : pathergy test was introduced as an important criteria in diagnosis of Behcet disease . The aim of the present work is to reevaluate this important test and especially to perform this reaction on oral mucosa and legs .
Individuals and methods : Thirty five Arab patients with untreated Behcet disease, their ages ranged between 21-51 years with male to female ratio of 3/2 .
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Results :The ordinary pathergy was positive in 65.7% of all patients & on oral mucosa in 45.7% . There was no difference in correlated gauge of needle.
The sensitivity and degree of positivty of the test on legs was more than that on forearms although statistically not significant (T=0.083).
Conclusion : pathergy test is better to be carried out on legs using G20 and on any side with no difference .

Board No: P-089
Ref. No: 38
Topic: Regional

GENDER AND CLINICAL MANIFESTATIONS OF BEHCET`S DISEASE: A REVIEW OF 95 CASES IN RUSSIA
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Background: Behcet's disease (BD) is well-known and more severe course of this disease observed among young males. The gender difference in Behcet's disease has never been studied before in Russia.
Objectives: To determine the effect of gender on the severity and clinical features of BD patients.
Methods: 95 BD pts were evaluated in the Institute of Rheumatology within the period from 01.2006 to 10.2009. BD was diagnosed according to criteria of International Study Group for Behcet's Disease (ISGBD). The mean age (years) of males 28,5 [23-34] and 30,9 [25-35] - females, the mean disease duration - 8,8 [3-14] and 11,7 [6-15] years, the mean age at the disease onset was 20,3 [15-26] and 18,8 [13-23] respectively.
Results: We observed the difference in respect to a gender: males had of more eye lesions (68,6% vs 32,0%, p=0,003), retinal vasculitis (62,8% vs 28,0%, p=0,005) and skin lesions (91,4% vs 64,0%, p<0,003) compared with female pts. Severe vascular involvement (Badd-Chiari syndrome, vena cava and ileac thrombosis, arterial thrombosis and arterial aneurisms) was found only in males - 10%. The female pts had more frequent joint involvement if compared with males (88,0% vs 58,6%, p<0,01). Score of the BD severity was calculated according to I. Krause [1]: mild in 15,7% of male pts and 44% of females (p=0,009), moderate 7,1% vs 16%, (p>0,05) and severe 77 % vs 40 %, (p=0,001) respectively. Two male pts (18 and 20 yrs) died and no deaths were observed among the female patients.
Conclusion: We observed the differences in clinical pictures of BD in respect to gender. Male pts in Russia have severe BD more freqently than female pts.

1. Krause I., Uziel Y., Guedj D., et al. Childhood Behcet's disease: clinical features and comparison with adult-onset disease. Rheumatology (Oxford). 199

Board No: P-090
Ref. No: 39
Topic: Regional

CLINICAL ASPECTS OF BEHCET`S DISEASE: A REVIEW OF 95 CASES IN RUSSIA
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[1]State Institute of Rheumatology Russian Academy of Medical Science Moscow, Russia. .

Objectives: We described the clinical features of the Behcet's disease (BD) in Russia.
Methods: 95 BD pts were evaluated in the Institute of Rheumatology within the period from 01.2006 to 10.2009. The group consisted of representatives of 25 ethnic subpopulations, of Caucasoid 92 pts (mainly inhabitants of Nord Caucasus and Transcaucasia, n=72) and Asians 3 pts.
Results: The male-to-female ratio was 3,7:1, mean age 29.7 [23-35] years, the mean disease duration - 9,6 [4-15] years. The mean age at the disease onset was 19,9 [14-25] yrs.
The clinical features in pts with BD were (%): oral ulcers in 98,9, genital ulcers - 81,0, eye lessions - 58,9, skin lesions - 84,2, patergy reaction 33,7 pts, articular - 52,6, neurological - 14,7, gastrointestinal -22,1, vascular - 26,3 pts, (thrombophlebitis 13,7, deep venous thrombosis 16,8 and arterial lesions 3,2 pts), 12,8 pts had epididimitis. 2,2% pts (18 and 20 yrs old male) died and the cause of death was vessel involvement.
The clinical features of BD in Russia are similar with those reported by ISGBD, but russian pts had higher frequences of deep venous thrombosis (16,8% vs 5%, p=0,01) and neurological manifestations (14,7 vs 5%, p=0,03).
Conclusion: The clinical features of BD in Russia are similar with those reported by ISGBD, but BD pts in Russian population showed higher frequency of deep venous thrombosis and neurological manifestations.

Board No: P-091
Ref. No: 40
Topic: Regional

Ethnic distributions of Behcet's disease in Russia
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Background: It has long been discussed that Behcet's disease (BD) has a different clinical features in various ethnic groups.
Aim: To evaluate the clinical features of Behcet's disease (BD) patients in different ethnic groups.
Methods: 82 patients with BD (M:F - 2,5:1) were included in our study. BD was diagnosed according to criteria of International Study Group for Behcet's Disease (ISGBD). The test for HLA B5 was performed using microlymphocytotoxity method. The mean age was 29.3 [23-34] years, the mean age at the disease onset - 20,3 [15-26] years, the mean disease duration - 8,8 [3-14] years. Patients have been divided into the 5 ethnic groups: Russians, Azerbaijanians, Armenians, Chechens and inhabitants of Dagestan, which are include 10 small ethnic groups.
Results: Ophthalmologic manifestations were found more in Azerbaijanian population and inhabitants of Dagestan in comparison with Russians pts. Vascular involvement was not observed in Azerbaijanian population. Neurological manifestations were seen more often Russian pts. Frequency of HLA-B5 was rare in Russian pts compared to pts of other groups. HLA-B5 correlated with ophthalmologic manifestations (retinal vasculitis) mainly in male patients.
Conclusion: Clinical picture of BD in Russia varied in ethnic subpopulation that allows predicting severe manifestations of diseases.

Groups	n=82	M:F	HLA B5(+)	Manifestation, (%)			
				uveitis	vascular	gastrointestinal	neurological
Inhabitants of Dagestan	31	3,5:1	83,9	66,7	35,5	22,6	9,7
Armenians	15	4:1	80	53,3	33,3	13,3	-

Azerbaijanians	11	1,7:1	81,8	72,7	-	18,2	9,1
Chechens	13	1,6:1	83,3	53,8	30,8	23,0	15,4
Russian	12	2:1	25	33,3	25,0	25,0	33,3

Board No: P-092
Ref. No: 46
Topic: Regional

Epidemiological and Clinical Characteristics of Behchet Disease in Japan- using a clinical database for patients receiving financial aid for treatment

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Objective: The aim of this study was to explore the clinico-epidemiological features of Behchet disease using a clinical database for patients receiving financial aid for treatment.
Methods: In the fiscal year of 2005, 16627 patients with Behchet disease were registered to receive public financial aid from the Ministry of Health, Labour and Welfare (MHLW) of Japan. From 2003, the MHLW of Japan started an on-line registration system of 45 intractable diseases including Behchet disease. We obtained the 2005 clinical database of Behchet disease from the on-line registration system of the MHLW. We analyzed clinical data from 9416 patients (input rate was 56.5%) including sex, age, onset age, activity in daily life, major symptoms, prevalence of vascular, neuro-, or intestinal Behchet, severity, and laboratory data.
Results: The sex ratio (male/female) of the patients was 0.76. This disease most frequently afflicted the 50-59 years age group in both males and females. The onset age of Behchet disease peaked at 30-39 years in both males and females. The prevalence of vascular Behchet was 2.4%, neuro-Behchet was 4.7%, and intestinal Behchet was 9.0%, respectively. The proportion of patients with severity levels (stages 1-5) of 1 and 2 (mild) was 66.9%, 3 (medium) was 9.3%, and 4 and 5 (severe) was 27.7%.
Conclusion: Using a clinical database of 9416 Behchet disease patients, we clarified the clinico-epidemiological features of Behchet disease in Japan.
Acknowledgements: This study was supported by a Grant-in-Aid for Research Committee of Intractable disease, provided by the MHLW, Japan.

Board No: P-093
Ref. No: 48
Topic: Regional

COMPARISON of INTERFERON ALPHA VERSUS CYCLOSPORINE-A FOR BEHÇET UVEITIS

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Purpose: The aim of this prospectively randomized study was to compare the efficacy of interferon alpha treatment and cyclosporine-A therapy in recurrent uveitis associated with Behçet's disease, which is refractory to conventional steroid therapy

Patients and Methods: Fifty two Behçet patients with ocular inflammatory reactions that could not be suppressed, or had frequent uveitis attacks with classical uveitis treatment were randomly divided into 2 groups of 26 each with patients of similar ages, sex distribution and severity of disease. Both groups received corticosteroids according to standard protocol. At the beginning of the steroid therapy, interferon alpha was added 6 million IU per day subcutaneously with a total of 120 million IU to the patients in group 1; and cyclosporine-A (5 mg/kg/day) was added to the patients in group 2.

Results: The results were compared between two groups, according to remission period and attack number during follow up. Cyclosporine-A was efficient in all cases with a mean follow-up of 35 months (26-55 months). The mean remission periods are 5.2 (±1.8) and 9.6 (±3.2) months in group 1 and 2, respectively. The numbers of acute episodes of uveitis during the follow up were 6.7 ± 2.3 and 3.4 ± 1.1 in group 1 and 2, respectively. The difference is clinically and statistically significant (p=0.001).

Conclusion: Cyclosporine-A proved to elongate the remission period and reduce the acute episodes, promising in the management of refractory forms of uveitis in Behçet's disease.

Board No: P-094
Ref. No: 57
Topic: Regional

Intraocular surgery under systemic infliximab therapy in patients with Behçet's disease

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Purpose
Infliximab is a new drug treatment for Behçet's disease with strong therapeutic efficacy. The action of infliximab is to suppress immunity that may increase the risk of systemic or local infections after surgery in patients who are on infliximab. To see if this is a possible undesirable consequence of infliximab treatment, we reviewed the outcomes of patients treated with infliximab for Behçet's disease who underwent intraocular surgery.
Methods
We conducted a retrospective study of 5 patients with Behçet's disease who underwent intraocular surgery while receiving systemic infliximab therapy. From 2007 to 2009, we treated seven eyes of five patients. All patients were male, and the mean age at the time of surgery was 44.2 years (30-70 years old). Four eyes had surgery for phacoemulsification, and three eyes had surgery for trabecletomy. The mean duration since the onset was 110 months (34-180 months). The mean dose frequency of infliximab at the time of surgery was 4th (2th-20th).
Results
Only 1 patient presented with uveitis after surgery, once in the right eye and twice in the left eye. Systemic short-term administration of steroid and steroid subtenon ocular injection could suppress the ocular inflammation. All patients receiving phacoemulsification recovered visual acuity. Moreover, ocular tension was well controlled in all patients that had a trabeclectomy. No infections were observed in any of the patients after surgery.
Conclusions
Based on our limited sampling of infliximab treated patients with Behçet's disease, infliximab treatment didn't complicate any subsequent intraocular surgery.

Board No: P-095
Ref. No: 65
Topic: Regional

Interferon-alfa therapy in Turkish patients with Behçet uveitis

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Purpose: To report on the results of interferon-α 2a (IFNα) treatment in Turkish patients with Behçet uveitis
Methods: We retrospectively analyzed the medical records of 121 patients treated with IFNα between April 2001 and May 2009. IFNα was administered as monotherapy or with low-dose (<10mg/day) steroids. The initial dose of IFNα was 6miU/day in 102 patients, 3miU/day in 15 patients and 3miU every other day in 4 patients. Main outcome measures were recurrence of uveitis attacks, changes in visual acuity, and side effects of treatment.
Results: Median duration of IFNα treatment was 13.5 months (range, 1-60 months). The recurrence rate decreased from 2.38 attacks/patient/year to 1.15 attacks/patient/year during IFNα therapy. Nine patients had recurrent attacks at an IFNα dose of

6miU/day, 22 patients at 3miU/day, and 42 patients at lower doses. There was no attack in 48 patients (39.7%). Remission was sustained after discontinuation of treatment in 33 of 44 patients. Mean logMAR visual acuity improved from 0.59±+0.55 and 0.66±0.66 to 0.24±0.39 and 0.37±0.56 in the right and left eyes, respectively, during treatment. The most common side effects, apart from flu-like symptoms, included elevation of liver enzymes (20%), leucopenia (17%), anemia (9.9%), weight loss (8.3%), and depression (6.6%). Treatment was discontinued in 46 patients (38%) because of unresponsiveness (13.2%)), side effects (6.6%), or noncompliance (18.2%).
Conclusions: IFNα therapy reduces the frequency of uveitis attacks in patients with Behçet uveitis. However, side effects and noncompliance with treatment limit the use of this therapeutic modality in one forth of our patients.

Board No: P-096
Ref. No: 69
Topic: Regional

Musculoskeletal manifestations in Behcet disease: an 18-month prospective study in 1495 patients
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Aim: To evaluate prospectively the different aspects of musculoskeletal manifestations in patients with Behcet disease (BD).
Methods: All patients with BD referred to our weekly outpatient clinic were evaluated for musculoskeletal symptoms. The type of manifestations and their relation to age, sex, HLA-B5 and HLA-B27 were noticed. All patients were followed until the end of study. A confidence interval at 95% (CI) was calculated for each item. The comparisons were done by chi square test.
Results: In an 18-month period, 4501 visits were done for 1495 patients (2.01/patient/year). Musculoskeletal manifestations were seen in 181 patients, with no relation to previous involvement (p:0.18). It was developed for the first time in 97, showing an annual incidence rate of 7.3% (CI:5.6-9.1) in those without these involvements (890). Peripheral arthritis was seen in 52%, arthralgia in 24%, periarthritis in 20%, ankylosing spondylitis in 13% and fibromyalgia in 4%. Monoarthritis was the most frequent pattern of peripheral joint involvement (60%), and knees were the most frequent involved joints (46%). We have found no age or sex difference in the prevalence or type of articular involvement except for peripheral arthritis seen more in males (p=0.01). Apart from expected higher prevalence of HLA-B27 in those with inflammatory back pain (p=0.002), there was no correlation with HLA-B5 or HLA-B27.
Discussion: In comparison with previous retrospective studies done in Iranian BD patients, this study showed a different pattern of joint involvement (monoarthritis vs. oligoarthritis) and no correlation with HLA-B5. This may be due to the shorter follow-up in this study.

Board No: P-097
Ref. No: 77
Topic: Regional

THE EFFECTS OF DENTAL AND PERIODONTAL TREATMENTS ON ORAL HEALTH RELATED QUALITY OF LIFE IN BEHCET'S DISEASE
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Aim: The aim of this study was to examine the effects of dental and periodontal treatments on oral health related quality of life in Behcet's disease (BD).
Materials and Method: Twenty-nine BD patients (F/M:15/14, mean age:39.6±6.9 years) were in the intervention group with dental and periodontal treatments performed. Control group including 29 BD patients (F/M: 15/14, 39.4±10.6 years) was only given oral hygiene education. Patients were examined in the pre-treatment period and 6 months after treatment. The number and healing time of oral ulcers were noted in both periods. Oral health related quality of life was examined by oral health impact profile-14 (OHIP-14) in the prospective study.
Results: The number of oral ulcers was similar in intervention (4.8±3.2) and control groups (3.7±2.3) in the pre-treatment period (p=0.13). However, healing time of oral ulcers (9.7±3.8 days) was higher in intervention group than controls (7.2±2.5

days) (p=0.008). Six month after treatment, the number of oral ulcers (1.9±1.5) and score OHIP-14 (34.2±11.6) were significantly decreased compared to those (4.8±3.2 and 40.2±8.5, respectively) in pre-treatment period of the intervention group (p=0.000 and p=0.02, respectively). In contrast, OHIP-14 score and the number of oral ulcers were found to be similar in pre-treatment (21.2±15.9 and 3.7±2.3, respectively) and after treatment periods in control group (21.5±17.4 and 2.8±2.4, respectively) (p=0.91, and p=0.15, respectively).
Conclusion: Oral health related quality of life status was improved and the number of oral ulcers decreased more prominently after dental and periodontal treatments compared to basic hygiene education in BD.

Board No: P-098
Ref. No: 81
Topic: Regional

ORAL HEALTH RELATED QUALITY OF LIFE IS RELATED TO ORAL ULCER ACTIVITY INDEX IN BEHCET'S DISEASE

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Aim: The aim of this study was to evaluate the relationship between oral ulcer activity and oral health related quality of life status in patients with Behcet's disease (BD) and recurrent aphthous stomatitis (RAS).
Materials and Methods: In this cross-sectional study, 79 BD patients with active oral ulcers (F/M: 43/36, mean age: 37.4±11.9 years) who were under immunosupressives or colchicine treatment and 31 RAS patients (F/M: 21/10, mean age: 33.53±10.51 years) treated with topical medications were included. The number and healing time of oral ulcers were noted during the last 3 months. Oral ulcer activity was examined by Composite index (CI). Pain (CI-pain) and functional subscales (CI-function) of CI were calculated in both patient groups.
Oral health related quality of life was examined by oral health impact profile-14 (OHIP-14). Scores of OHIP-14 subscales - physical, psycho-social and psychological - demonstrated by factor analysis in active oral ulcers were calculated in BD and RAS.
Results: Scores of CI, OHIP-14 and the number of oral ulcers were significantly higher in RAS (6.94±2.19, 35.67±9.98 and 7.58±5.27) compared to BD (6.02±2.05, 19.37±15.20 and 5.07± 3.11)(p=0.040, p=0.00 and p=0.018, respectively). CI score correlated with OHIP-14 subscales scores regarding physical, psycho-social and psychological both in BD and RAS ($p<0.05$).
Conclusion: Oral health related quality of life status was related to ulcer activity index and its subscales in both groups. A lower oral health related quality of life was observed in patients with RAS, treated only with topical medications, compared to BD patients.

Board No: P-099
Ref. No: 82
Topic: Regional

COULD MINIMAL CLINICALLY IMPORTANT IMPROVEMENT FOR OHIP-14 REFLECT CHANGES IN ORAL ULCER ACTIVITY IN BEHCET'S DISEASE ?

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Aim: The aim of this prospective study was to evaluate the relationship between minimal clinically important improvement (MCII) for oral health impact profile-14 (OHIP-14) and oral ulcer activity examined by Composite index (CI) in Behcet's disease (BD).
Materials and Method: In this prospective study, 91 BD patients (F/M:54/37, mean age: 40.18±10.19 years) were enrolled. Oral ulcer activity was evaluated by CI. Scores of CI and subscales of CI-Pain and CI-functional status were calculated. The follow-up period was 4.01±2.4 months between the visits. The threshold level of percent change (-38.1 %) in OHIP-14 score generated by ROC analysis was used for the analysis for MCII (sensitivity: 88.6 % and specificity: 97.1 %). Patients were

categorised as improved or non-improved according to the cut-off value for MCII.
Results: 23.1% of BD patients (n=21) were in the improved group. Although the ratio of patients who were under immunosuppressive medications (25%) were higher compared to those treated with colchicine (21.9 %) in the improved group, no significant difference was found (p=0.07). CI scores in baseline and follow-up periods were significantly lower in the improved group (7.50±2.11 and 1.94±2.04) than those in non-improved group (9.33±1.18 and 9.84±4.53)(p=0.042 and p=0.000).
Conclusion: A close relationship was observed between oral ulcer activity and quality of life status. A core assessment of mucosal disease in BD may include both composite index as an objective and oral health related quality of life as a subjective outcome parameter and both seem to be useful for clinical studies.

Board No: 100
Ref. No: 83
Topic: Regional

BEHÇET'S DISEASE IN SOUTHERN TURKEY: CLINICAL AND DEMOGRAPHIC CHARACTERISTICS OF 406 PATIENTS

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Purpose: To describe and investigate the demographic and clinical features, ocular and systemic manifestations of the patients with Behçet's Disease living in Cukurova region.
Design: Nonrandomized, retrospective clinical study.
Materials-methods: The study was conducted from 406 patients who met the classification criteria of the International Study Group for Behçet's Disease who have been followed at Cukurova University Uvea-Behçet Clinic, which is a tertiary referral center. Clinical and demographic characteristics of the patients including sex, age, anterior and/or posterior segment involvement, visual acuity, systemic manifestations and treatment modalities were reviewed retrospectively.
Results: of the 406 patients; 306 (75.4%) were male and 100 (24.6%) were female. The mean age at onset was 27.6±7.2 years in male patients and 29.0±9,3 years in females. The frequency of ocular involvement (p=0.163) and bilaterality (p=0.67) were similar at both genders. The most common initial presenting manifestation of the disease was oral aphthous ulcer which was seen in 71.9% of the patients followed by ocular involvement in 23.4%. The leading clinical features were oral aphthous ulcers (92.6%), genital ulcers (82.8%) and ocular manifestations (80.3%) respectively. Males had more frequent and more serious ocular involvement than females. Cyclosporine was more commonly preferred for patients with frequent inflamatory episodes. There was no significant difference in final visual acuity between genders.
Conclusions: Ocular involvement in Behçet's Disease is significantly more severe in men. The main prognostic criteria affecting the final visual acuity are the severity and frequency of ocular involvement.

Board No:101
Ref. No: 89
Topic: Regional

An audit of Behçet's Syndrome research: A 10-year survey

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Objective: We had the impression and some preliminary data suggesting that the use of diseased control groups and proper use of power calculations were neglected in manuscripts published even in widely read rheumatology and medicine journals. We aimed to formally survey these and other methodological shortcomings in published manuscripts on one specific topic, Behçet's syndrome (BS). We reason that recognizing such methodological shortcomings will eventually lead to better quality clinical and basic science manuscripts.

Methods: The articles published in the 15 highest-impact-factor rheumatology, ophthalmology, dermatology and general medicine journals between January 1999 and January 2009 were searched for original papers on BS. The study designs (study types and time element), control groups, gender ratio and mean age of patients, the use of power calculations and reporting of negative results were specifically tabulated.

Results: 280 articles qualified for the survey. Prospective longitudinal studies were few (7%). In a considerable proportion of papers, some basic demographic data was missing (21%). Power calculations were rare (3%) and were not even considered in clinical hypothesis testing. Diseased control groups were present in slightly over one half of clinical and laboratory original research while, as we had noted earlier, only 9% of genetic association studies included diseased controls. Only 12% of all manuscripts concerned mainly negative outcomes.

Conclusion: A considerable number of the research articles on BS with methodological weaknesses in study design appeared in our better journals. The generalizibitiy of what we observed in BS to other research topics needs to be formally studied.

Table: Some of the methodological issued addressed in the survey.

	% (n)
Mean age of the patients not mentioned	19.9% (55)
Gender of the patients not mentioned	8.3% (23)
Prospective longitudinal study designs	7.1% (13)
Retrospective longitudinal study designs	9.9% (18)
Cross-sectional study designs	83.0% (151)
Diseased control groups present	43.8 (71)
Power calculations mentioned	2.8% (6)
Negative results mentioned	12.1% (23)

The survey of the time element, control groups, power calculations and presence of negative results were considered only in the article types suitable for the inclusion of such elements in the study design.

Board No:102
Ref. No: 91
Topic: Regional

Gender Influence on Ocular Manifestations and its Outcome in Behcet's Disease

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Introduction: It is of general belief that male gender is prone to more severe manifestations, and less favorable outcome. The aim of this study was to evaluate this hypothesis in ophthalmological manifestations (OM) of Behcet's Disease (BD).
Materials and Methods: Visual acuity (VA), anterior uveitis (AU), posterior uveitis (PU), and retinal vasculitis (RV) disease activity indexes (DAI) were checked according to Ben Ezra. The data at baseline and last visit were compared. Total Inflammatory Activity Index (TIAI) and the Total Adjusted Disease Activity Index (TADAI) were calculated too.
Results: Male/female ratio was 1.2 in the BD registry (6568 patients) and 1.51 for OM patients (Chi2=27.732, p<0.0001). 1514 patients were treated for OM. The patients-year-follow-up was 4895.
All parameters improved significantly from baseline to the last visit.
Mean VA improved from 3.9 to 4.4 (males) and 4.5 to 5.1 (females). Difference between males/females improvement was significant (p<0.0001). Percent Improvement males/females: 47.1%/48.8% (p=0.4).
Mean PU (male/female) improved from 2.1/2.0 (difference male/female NS) to 0.8/0.6 (difference male/female improvement: NS). Percent Improvement males/females: 75.4%/81% (p=0.002).
Mean RV (male/female) improved from 2.4/2.4 (no difference) to 1.4/1.2 (difference male/female p<0.001). Percent Improvement males/females: 62%/64.4% (p=0.3).
Mean TIAI (male/female) improved from 17.3/16 (difference male/female: p=0.06) to 7.8/6.3 (difference male/female p<0.27). Percent Improvement males/females: 77.1%/80.6% (p=0.09).
Mean TADAI (male/female) improved from 27.4/26.5 (difference male/female: p=0.22) to 18.7/18.0 (difference male/female p<0.39). Percent Improvement males/females: 72%/73.6% (p=0.49).
Discussion and Conclusion: Male gender is more prone to ocular manifestations, but the severity and therapeutic outcome was the same for the majority of parameters.

DAI	Subjects	Mean Baseline	Mean Improv	*P*	% Eyes improv
Visual Acuity	Male	3.9	0.5	*<0.001*	47.1
	Female	4.5	0.6	*<0.001*	48.8
	P Value	*<0.001*	*<0.001*	-	*0.4*
Posterior Uveitis	Male	2.1	1.3	*<0.001*	75.4
	Female	2.0	1.4	*<0.001*	81
	P Value	*0.11*	*0.11*	-	*0.002*
Retinal Vasculitis	Male	2.4	1.0	*<0.001*	62.0
	Female	2.4	1.2	*<0.001*	64.4
	P Value	*No diff*	*<0.001*	-	*0.3*
TIAI	Male	17.3	9.5	*<0.001*	77.1
	Female	16.0	9.7	*<0.001*	80.6
	P Value	*0.06*	*0.27*	-	*0.09*
TADAI	Male	27.4	8.7	*<0.001*	72.0
	Female	26.5	8.5	*<0.001*	73.6
	P Value	*0.22*	*0.39*	-	*0.49*

Board No:103
Ref. No: 95
Topic: Regional

Incidence of uveitis due to Behcet disease and complications.

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Incidence of uveitis due to Behçet disease and complications.
Kabasele Paul Muambi Buana MD Mphil, Taylor Simon Richard Phd, Lightman Susan Louise Phd.

Introduction
To report the incidence of uveitis due to Behçet disease and complications at a tertiary referral centre.
Patients and methods.
A cross sectional prospective observational study. Case notes review of patients attending with at least six months follow up from June 2008 to January 2010.
Results.
Out of a population of 1444 patients with uveitis, 63 (4.3%), 105 eyes had uveitis due to Behçet disease. of these, 37 males and 26 females (ratio 1.4), mean age 44.9 (limits 23-66 years). Mean follow up 7.3 years (limits 6months- 38 years). Twelve eyes (11.4%) developed anterior uveitis, 15 (14.2%) intermediate uveitis, 36(34%) panuveitis. Posterior uveitis accounted for 3.8% while vasculitis was found in 31.4% of eyes.
Macular ischemia developed in 10 eyes, accounting for 18.1%. Epiretinal membranes (ERM) were found in 16.3% of eyes, cataract and cystoids macular edema (CME) developed equally in 12.7% of eyes each. Optic atrophy and glaucoma were found in 10.9% and 9% respectively.
Fifty eight eyes (55.2%) had their best corrected visual acuity (BCVA) of 6/12 or worse at first visit. At last follow up visit, 56 eyes (53.3%) had BCVA≤ 6/12. Four patients (6.3%) were legally blind.
Conclusion.
Patients with Behçet disease can develop all types of uveitis. However, panuveitis and vasculitis are the most common. Most complications occur in the posterior segment and are sight threatening.

No grant received for this study.

Board No:104
Ref. No: 101
Topic: Regional

Papillitis presenting as the initial ocular sign of Behçet Disease
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Purpose: To report a patient with Behçet's disease presenting with unilateral papillitis as the initial ocular sign.
Methods:A 36 year-old male patient with Behçet's disease and without previous ocular involvement was admitted to the author's clinic by the complaint of a central scotoma in his right eye.Ophthalmologic examination, fundus florescein angiography (FFA), visual field test,macular optical coherence tomography (OCT), pattern visual evoked potential test(VEP), cranial Magnetic Resonance Imaging (MRI) were performed.
Results:Visual acuity was 1.0 in the right eye and 1.0 in the left eye. No inflammation sign and sequaele was seen in slit lamp examination. Hyperemic swelling of the right optic disc with blurred margins were observed in fundus examination. The left optic disc examined as normal. Fundus florescein angiography revealed late hyperfluorescence and leakage at the right optic nerve head. No abnormality was found in macular OCT, visual field test, pattern VEP and cranial MRI.Neurologic examination was normal.With the diagnosis of papillitis in the right eye, medical therapy was initiated as intravenous methylprednisolone 1000 mg /dayx3 and followed by an oral regimen 1 mg/kg/day tapered in 8 weeks combined with oral cyclosporin A 5mg/kg/day.At the end of the 8 weeks follow-up period, the patient determined as almost in a remission, since he has had a very few leakage in upper quadrant of the right optic nerve head in his last FFA.
Conclusion: Papillitis is a rare finding as an initial sign in ocular Behçet's disease.

Board No:105
Ref. No: 107
Topic: Regional

Infliximab therapy for chronic progressive neuro-Behçet's disease: A four-year follow-up study
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We previously reported that infliximab (IFX) prevented the progression of chronic progressive neuro-Behçet's disease (CPNB) by markedly reducing cerebrospinal fluid (CSF) IL-6 in a 14-week open trial. In this study, we further performed a follow-up survey for 5 patients who had completed the 14-week trial, for as long as 4 years. In one patient, although IFX was withdrawn without any increase in the dose of MTX, CSF IL-6 remained below 20pg/ml throughout the course. In another patient, who was complicated by refractory uveitis, IFX was continued. His CSF IL-6 remained below 20pg/ml throughout the course. In another 3 patients, IFX was stopped after 3 additional infusions. However, since the CSF IL-6 became elevated over 20pg/ml, IFX was resumed along with increasing the dose of MTX in all 3 of these patients. In 1 of the 3 patients, IFX could be stopped by increasing the dose of MTX up to 25mg/week, which caused the CSF IL-6 levels to decrease below 20pg/ml. In another of the 3 patients, although IFX was continued every 6 weeks along with the increased dose of MTX up to 17.5mg/week, CSF IL-6 were sustained above 20pg/ml with progression of urinary incontinence and myoclonus. In the final of the 3 patients, his CSF IL-6 were sustained around 20pg/ml after IFX was resumed, spasticity was aggravated. These results indicate that IFX has a beneficial effect in the treatment of CPNB by reducing CSF IL-6 levels. Further study is needed in order to establish the effective treatment regimen for CPNB.

Board No:106
Ref. No: 109
Topic: Regional

Association of HLA-A26 with Behçet's disease

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Association of HLA-B51 with Behçet's disease (BD) is well-known. Recent reports have suggested that HLA-A26 might also be associated with BD. We therefore investigated the frequency of HLA-A26 in BD and its relevance with clinical features. HLA phenotyping data were obtained from 161 patients (76 males and 85 females), who had been diagnosed as BD at Teikyo University Hospital from 1989 until 2009 according to the 1987 criteria of BD Research Committee of Japan. The frequencies of B51 and A26 were 48.4% and 29.2%, respectively, which were significantly higher than those of healthy Japanese population ($p<0.0001$ and $p=0.0117$, respectively). The incidence of mucocutaneous lesions, ocular manifestation, arthritis and epididymitis were not significantly correlated with A26 or B51. of note, the incidence of vascular involvement (VBD) were significantly lower in A26-positive patients compared with A26-negative patients (4.3% vs. 21.1%, $p=0.0165$ [Chi-square test]), whereas there was no significant correlation of A26 with neurological involvement (NBD) and intestinal involvement (IBD). By contrast, in B51-positive patients, the incidence of NBD was significantly higher (33.3% vs. 10.8%, $p=0.0011$) and that of IBD was significantly lower compared with B51-negative patients (12.8% vs. 30.1 %, $p=0.0136$). The results confirm the significant association of HLA-A26 with BD. Moreover, the data suggest that HLA-A26 and HLA-B51 might have significant influences on the expression of various organ involvements. Further studies to explore how HLA-A26 contributes to the pathogenesis would be important.

Board No:107
Ref. No: 118
Topic: Regional

Long-term efficacy and safety of low dose and dose escalating interferon alfa-2a therapy in refractory Behçet's uveitis

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Purpose: To investigate long-term efficacy and safety of low dose and dose escalating therapy of interferon alfa-2a (IFN α-2a) in the treatment of Behçet's uveitis.
Methods: For this prospective study, 37 patients with refractory panuveitis due to Behçet's disease were included. All were unresponsive to previous therapy with conventional immunosuppressive(s). For induction IFN α-2a was given 3 million international-units (IU) subcutaneously (sc) daily for 14 days. Maintenance was achieved with IFN α-2a given as 3 million IU 3x/week sc. The dose was increased sequentially to 4.5, 6 and 9 million IU 3x/week, if uveitis relapses occurred. Total therapy duration was 24 months. Primary outcome measure was control uveitis with quiescence while on maintenance therapy. Ocular relapses per patient-year (PY) before and after initiation of IFN α-2a and steroid-sparing effect were secondary outcomes. We also estimated the rate of remission after discontinuing IFN α-2a.
Results: During maintenance IFN α-2a was able to control uveitis in 94.5% of patients. In 51.4% of patients a maintenance dose of 3 million IU 3x/week was able to control uveitis without any relapse. The rate of uveitis relapses decreased from 3.52/PY before initiation to 0.70/PY after initiating IFN α-2a. Survival analysis estimated the rate of remission after discontinuation of IFN α-2a as 75% by 3 months. The rate of remission stayed stable thereafter.
Conclusions: A treatment protocol utilizing a low dose and dose escalating therapy of INF α-2a was able to control and achieve remission of uveitis in the majority of patients.

Board No:108
Ref. No: 125
Topic: Regional

Behcet's disease in Germany: differences and similarities in patients of German and Turkish origin – a single center experience

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Objective. To evaluate the relationship between ethnic origin, disease manifestations and the association of HLA-B 51 to BD in German and Turkish patients living in Germany.
Methods. 238 patients with BD according to ISBD criteria from a an interdisciplinary reference centre were retrospectively analysed (112 German, 126 Turkish). All patients were seen by a rheumatologist and ophthalmologist and received a laboratory workup including HLA-B51 genotyping.
Results. In both patient groups oral aphthae were most common (100%), followed by genital aphthae and ocular involvement . Disease manifestations and severity were comparable in both groups. HLA B51 was positive in 36 % of the German and 77 % of Turkish patients ($p<0,001$). Ocular involvement (posterior uveitis and panuveitis, retinal vasculitis) was significantly associated with HLA-B 51 in both patient groups, in German patients ($p<0,007$) even more than in Turkish ($p<0,05$). Moreover, in Turkish patients, a significant association between papulopustules and HLA-B 51was found.
Conclusion. In 2004 we published our data on 60 patients and could not find any clear cut differences between German or Turkish patients. Meanwhile, with a much larger cohort, we – in agreement with others (Krause et al. 2009) can support the association of HLA-B51 with ocular disease in both patient groups. Furthermore, HLA B51 was significantly more common in Turkish patients and associated with papulopustules. The expression of the disease manifestations does not differ significantly between both groups, hence BD in Turkish patients in Germany is not more severe than that of German patients.

Board No: P-109
Ref. No: 127
Topic: Regional

RENAL INSUFFICENCY DUE TO AA-AMYLOIDOSIS IN A PATIENT WITH BEHÇET'S DISEASE

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A 44 year old Moroccan female, was admitted with fatique, dyspnea, diarrhea, vomiting and fever for 2 weeks. She had a history of Behçet's disease (BD) with oral and genital ulcera, arthritis and erythema nodosum successfully treated with NSAID's, steroids cyclosporine, and mycophenolates. Her disease showed periods of relapses, however recently there was no disease activity and she was only treated with prednisone 5 mg once two daily. Her complaints started both after endoscopic appendectomy and vaccination against the Mexican flu. During the surgical procedure she reacted with anaphylactic shock to antibiotics (clindamycin). In the course of a few weeks laboratory evaluation showed a sudden decrease in renal function. Despite rehydration the renal function only partially improved. A renal biopsy revealed tubulointerstitial disorder and glomerulosclerosis with amyloidosis. Staining with potassium permanganate indicated AA-amyloidosis (see figure 1). Viral infections (Mexican flu, influenza, noro, adeno, entero, rota) and other systemic triggers such as hematological malignancies with AL amyloidosis and periodic fever syndromes were excluded.
Our patient presented with a combination of acute on chronic renal failure. Acute tubulus necrosis during surgery and hypotensive episodes after the anaphylaxis combined with AA-amyloidosis might have caused the renal deterioration. Serum sickness after vaccination can be excluded because of the absence of immunecomplexe deposition in the renal biopsy.
In summary, our patient presented with a combination of acute on chronic renal failure. Amyloidosis is considered a common cause if BD patients develop renal failure with proteinuria or nephrotic syndrome.

Renal biopsy

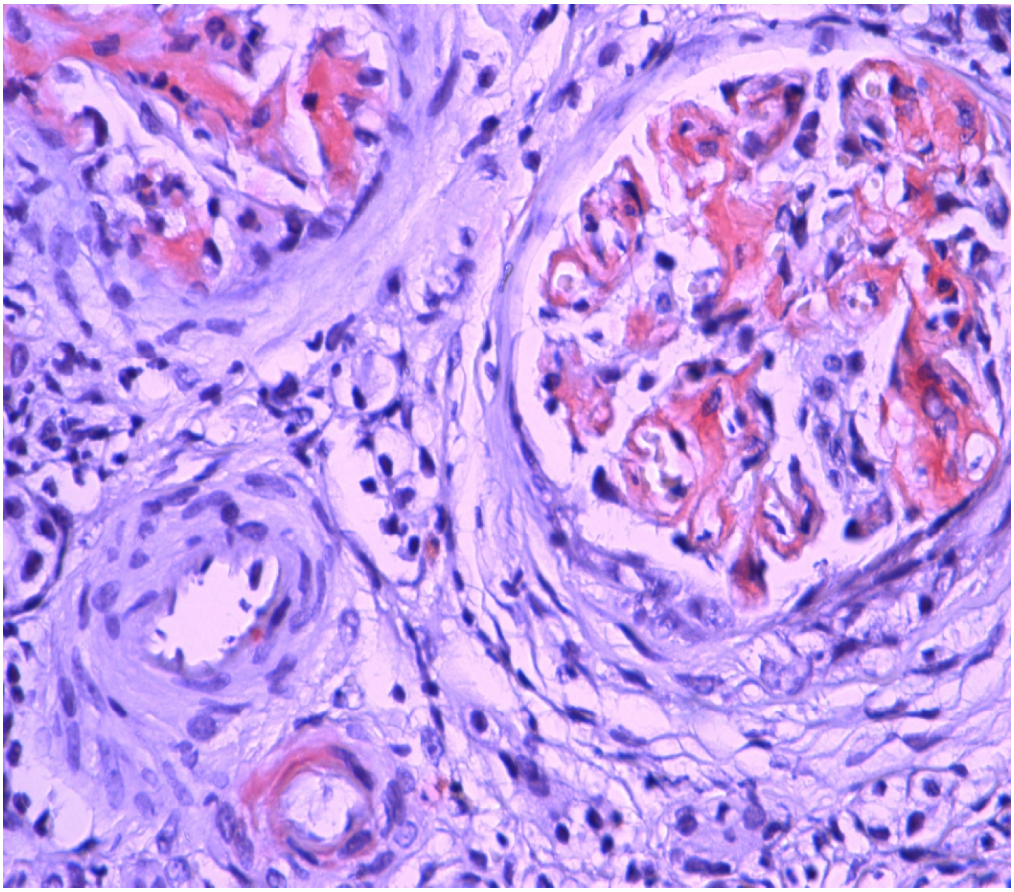

Fig 1: amyloidosis in glomeruli stained with congo red

Board No:110
Ref. No: 128
Topic: Regional

Particularities of Behçet's disease in Tunisia through a study of a homogenous group of 430 patients.

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Objectives:
To determine demographic, clinical and genetic particularities of BD in a homogenous group of Tunisian patients

Patients and methods:
Files of 430 BD patients (ISG criteria),seen during the last 20y, were analyzed for demographic, clinical presentation, investigations, complications and treatment. Results were compared with those in other ethnic groups. Data were recorded and analyzed using SPSS 11.0.
Results:
Records of 430 patients were reviewed. They were 295 males and 135 females (sex-ratio= 2.2). The average age at BD onset and diagnosis was respectively 29 and 34 years. The average delay to BD diagnosis was 5 years.
Frequencies of clinical and genetic features in our patients are shown in Table1.
Frequency of genital ulcers (p= 0.003), positive pathergy test (p= 0.001) and deep venous thrombosis DVT (p< 0.001) were significantly less frequent in patients with ocular involvement; whereas HLA B51+ was significantly more frequent in these patients (p= 0.049).
Frequency of arterial aneurism was significantly more frequent in neuroBD patients (p<0.001) but HLAB51+ was

significantly less frequent in these patients (p=0.04).
DVT were significantly more frequent in males (p<0.001). Genital ulcers (p=0.014) and arterial aneurisms (p<0.001) were significantly more frequent in patients with DVT while ocular involvement (p<0.001) were significantly less frequent in this group of patients.

Conclusion,
Our series shows many similarities with the majority of others reported in different countries all around the word. Yet, it shares with similar ethnic and geographic groups many particularities. These findings confirm once more the ethnic and geographic BD presentation's variability.

	n = 430
Oral ulcers n (%)	430 (100)
Genital ulcers n (%)	341 (85)
Pseudofolliculitis n (%)	320 (74.4)
Positive pathergy test n (%)	176/305 (57.7)
Arthritis/Arthralgia n (%)	195 (45.7)
Ocular involvement n (%)	200 (46.5)
Neurological involvement n (%)	121 (28.1)
Vascular involvement n (%)	150 (34.9)
Deep vein thrombosis n (%)	136 (31.6)
Arterial aneurysms n (%)	23 (5.3)
Arterial thrombosis n (%)	6 (1.4)
Intestinal involvement n (%)	7 (1.6)
HLA B51 +	84/177 (47.5)

Board No:111
Ref. No: 147
Topic: Regional

Quantitative analysis of brainstem areas on magnetic resonance imaging in neuro-Behçet's disease
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Quantitative evaluation of brainstem on magnetic resonance imaging (MRI) was conducted in patients with neuro-Behçet's disease (NB) in order to delineate the differences in the pathogenesis of acute NB and chronic progressive NB. Areas of the mesencephalic tegmentum and pons were measured on sagittal sections of T1-weightened images of MRI using Image J in 12 patients with acute NB and 18 patients with chronic progressive NB. Areas of the mesencephalic tegmentum (acute NB: 136.08 ± 24.37 mm^2 [mean ± SD], chronic progressive NB: 95.82 ± 20.37 mm^2) as well as those of the pons (acute NB: 532.77 ± 83.62 mm^2, chronic progressive NB: 399.63 ± 78.93 mm^2) were decreased significantly in chronic progressive NB compared with acute NB. Three patients with chronic progressive NB with persistent elevation of CSF IL-6 in spite of treatment with MTX and infliximab showed progression of brainstem atrophy. By contrast, in three patients with chronic progressive NB, whose CSF IL-6 levels were controlled by treatment, brainstem atrophy did not progress. These results confirm that brainstem atrophy is more prominent in chronic progressive NB than in acute NB. Moreover, the data suggest that quantitative analysis of brainstem areas might be useful for early diagnosis as well as for evaluation of the disease activity in chronic progressive NB.

Board No:112
Ref. No: 148
Topic: Regional

GOOD RESPONSE TO STEROID THERPAY IN TWO CASES OF INTESTINAL BEHCET'S DISEASE

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INTRODUCTION
The gastrointestinal involvement of BD is characterised by single or multiple deep penetrating ulcers, mostly in the terminal ileum, the ileoceacal region and the colon. There is yet no evidence-based treatment that can be recommended for the management of intestinal BD. We report here two cases of patients with intestinal BD who responded well to steroid therapy.
OBSERVATIONS
Case 1: A 42 years old male, having a recurrent oral aphtosis, necrotizinig pseudofolliculitis lesions and bilateral posterior uveitis, was diagnosed as BD. Six years after diagnosis, he developed a chronic diarrhea and an important weight loss. Endoscopic exam showed a normal colic mucosa but revealed deep ulcers on ileal mucosa. Pathological exam of these ulcers showed vasculitis lesions without granulomas. The patient was treated with 1mg/kg/day of prednisone and aspirine. He responded very well to therapy without any relapse after seven 7 years of follow-up.
Case 2: A 24 years old female with a 2 years history of oral, genital aphtae and necrotizing pseudofolliculitis presented with abdominal pain and haemorrhagic diarrhea. Endoscopic exam showed inflammation and multiples erosions within the colic mucosa. There were no granulomas at the pathological exam but vasculitis signs. The patient was treated by colchicines, aspirine and 1 mg/kg/day of prednisone. The improvement was total with a complete disappearance of abdominal pain and diarrhea.
CONCLUSION
We think that steroids can be considered as a first line treatment in intestinal BD. Agents such as azathioprine, TNFα antagonists and thalidomide can be used in refractory – non complicated of perforation- cases

Board No:113
Ref. No: 155
Topic: Regional

The influence of behcet disease on pregnancy

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Behçet's disease (BD) is a multisystem disorder affecting mainly young adults. Conflicting reports of the effects of pregnancy on the course of BD have been reported.

Patients and methods:
We retrospectively analyzed the relationship between BD and pregnancy, in order to detect any possible interaction between these two multisystemic processes. We studied 147 pregnancies in 46 women with BD.

Results:
The diagnoses were made according to the criteria of the ISGBD. There was remission of BD during 24 % of pregnancies. The disease became exacerbated during 36% of pregnancies. There were no changes in the clinical course of BD in 40% of pregnancies.
The most frequent manifestations of the clinical exacerbation were increases in the intensity and severity of outbreaks of oral ulcers during pregnancy (76%), venous thromboses were observed in 8 % of outbreaks of genital ulcers; eye inflammation and arthritis were other signs of exacerbation.

Conclusion:
The influence of pregnancy on the clinical course of BD is quite variable between patients and even during different pregnancies in the same patient. We speculate that pregnancy in general, doesn't seem to markedly affect the natural course of BD.

Board No:114
Ref. No: 156
Topic: Regional

coexisting behcet and crohn's disease
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The Association of Behcet's disease (BD) and Crohn's disease (CD) in the same patient or in the same family was rarely described, and raises a nosologic problem.

Case report:
A 47 year old man developed since 2006, recurrent oral and genital ulcers, polyarthralgia. pseudofolliculitis, and positive pathergy test were noted. The HLA B51 was positive.
His brother developed at the age of 40 years, diarrhea and abdominal pain. Endoscopic colonoscopy examination showed a diffuse colitis. Histological examination revealed epitheloid granuloma confirming the CD. The HLA B27 was positive. He was treated by oral prednisone (1mg/kg/day) with a good remission after 5 months.

CONCLUSION:

While being of different pathogenic origin, BD and CD may coexist in the same family or in the same patient. It's sometimes difficult to distinguish the gastrointestinal involvement of BD from that of CD causing therapeutic problems.

Board No:115
Ref. No: 160
Topic: Regional

Brhcet's disease after the age of 50 years
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Introduction :

Behçet's disease (BD) is a multisystem disorder affecting young adults with a typical onset of the disease between the age of 20 and 40 years, varying from the first few months of life to the age of 72 years. It's exceptionally revealed after the age of 60 years.

Patients and methods:

We analyzed retrospectively the course of BD in 182 patients in a period of 17 years. The diagnosis was made according to the criteria of the ISGBD.
We include in this study patients after the age of 50 years, and discuss the particularities of this onset.

Results:

Seventy patients were diagnosis having BD after the age of 50 years (14 males and 3 females). The mean age was 57 years.
The most frequent signs were cutaneous (11 cases), eye inflammation (n=4), genital ulcers (n=1) and thrombosis (n=1)
The disease becomes exacerbed in 10 patients: Neurologic complications (6), ocular complications (8), vascular complications (6) and arthritis (3).
The treatment was based on immunosuppressive therapy (8 patients) and colchicine® (all patients).

Conclusion:

BD affects the young adult but has to be evoqued even at an advanced age, in order to treat adequately and prevent complications. The clinical signs should be actively looked for, even in on elderly patient. Only an adequate treatment may prevent severe complications in these patients.

Board No:116
Ref. No: 162
Topic: Regional

Association of psoriasis and Behçet's disease: Report of three cases

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Introduction

Psoriasis is a chronic inflammatory skin disorder, which is characterized by erythematous, sharply demarcated papules, and rounded plaques covered by silvery micaceous scale.
The inclusion of Behçet's disease (BD) among seronegative spondyloarthropathies is still being debated. We described three cases of psoriasis associated with (BD).
Cases:
Case 1
A 66-year-old man had a 20-year history of BD with recurrent aphthosis, genital ulcerations, necrotizing pseudofolliculitis and posterior uveitis complicated with blindness. He complained of sharply demarcated, erythematous, scaling plaques in the elbows, knees and scalp. The diagnosis was a psoriasis associated to BD.
Case 2
A 35-year-old man was treated in dermatology department for psoriasis. After five year, he developed bipolar aphtosis and necrotizing pseudofolliculitis, so we considered as psoriasis accompanied with BD.
Case 3
A 30-year-old man had BD according to the criteria of International Study Group of Behcet's disease (recurrent aphthosis, genital ulcerations, and necrotizing pseudofolliculitis).This patient was diagnosed one year after, as psoriasis by dermatologist, based on micaceous skin lesion in back and elbow.

Conclusion
The association between BD and psoriasis is rarely reported in literature. Physiopathologic links may be exist and should be searched. Many authors suggested that BD could be included in spondyloarthropathy, but sacroiliitis in BD was quite lower than that in psoriasis. In addition, the of eye involvement of BD were different from that of spondylarthropathy, so it is still being debated

Board No:117
Ref. No: 168
Topic: Regional

Prevalence of Behçet's disease in south-west Germany among the adult Turkish and German population

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Objectives: To estimate the prevalence of Behçet's disease (BD) among the adult Turkish and German population in South Wuerttemberg and Baden-Wuerttemberg.

Methods and Materials: The study was conducted using two sources: A questionnaire was sent to 8000 general practitioners, rheumatologists, internal specialists, orthopaedic specialists, dermatologists, ENT-specialists and ophthalmologists in Southern Wuerttemberg. In addition, a random sample of 96 physicians of the same specialities was taken. Furthermore, all university hospitals, hospitals of maximum medical care and rheumatologists in Baden-Wuerttemberg were contacted. Then the results were correlated to the population of Southern Wuerttemberg and Baden-Wuerttemberg, based on data of the State Statistics Office.

Results: 31 patients were reported in Southern Wuerttemberg and 200 patients in Baden-Wuerttemberg. The prevalence among the Turkish population is 36/100.000 in Baden-Wuerttemberg and 44/100.000 in Southern Wuerttemberg. The prevalence among the German population does not differ between the two regions and amounts to 0,8/100.000 inhabitants.

When correlatedto inhabitants without migration background, the prevalence is much higher (1,3/100.000).

Discussion: The prevalence of BD among the Turkish population in Germany is much lower than in Turkey, where it is in the range between 80-420/100.000. This supports other studies which proposed strong environmental influences on the expression of BD. Furthermore a south-north-division becomes apparent. Similar results have also been described for Japan. The prevalence in the German population is comparable to the prevalence in other north-western European populations. The difference to the prevalence in Berlin, the highest nationwide with 1,47/100.000 inhabitants, is probably caused by the multi-ethnic character of the urban Berlin population.

Board No:118
Ref. No: 175
Topic: Regional

Differences in the distribution of clinical signs between Adamantiades-Behçet's disease patients of Turkish and German origin in Germany

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Among the 725 patients with Adamantiades-Behçet's diseas reported to the German Registry of Adamantiades-Behçet's Disease until February 2010, 317 were of Turkish (43.7%) and 287 of German origin (39.6%). Androtopism was found among patients of Turkish origin (men:women=1.84:1, p<0.001), while no gender predilection was recorded in German patients (men:women=0.93:1, ns). Altogether 9.3% of patients showed a positive family history with significant difference among Turkish and German patients (14.3 vs 3.7%, respectively, p<0.001). In general, there was a trend of more frequent ocular involvement in Turkish than in German patients (59.6 vs 49.7%, p=0.044), but there was no statistical difference in the risk of blindness (8.7 vs 4.5%; n=18 vs. 8). A similar trend could be observed for folliculitis beeing more frequent in Turks than in Germans (67.9 vs 58.7%, p=0.05), whereas sterile prostatitis/epidimytitis was more frequent in German patients (17.4 vs 7.6%, p=0.02). 48.3% of the patients had HLA-B5 antigen; of these 64.5% were of Turkish and 35.5% of German origin (p<0.001). Frequencies of other clinical signs showed no significant differences between Turks and Germans: oral aphthae (98.9 vs 98.0%), genital ulcers (63.2 vs 67.1%), arthralgia (50.2 vs 51.3%), erythema nodosum (42.5 vs 35.1%), thrombophlebitis (13.1 vs 13.4%), gastrointestinal (10.1 vs 15.2%) and CNS involvement (9.7 vs 12.0%), pos. pathergy test (31.5 vs 30.5%). In the Turkish as well as in the German patients group, oral aphthae were the most common first (85.6 vs 84.7%, ns), genital ulcers the most common second symptom (52.7 vs 47.3%, ns).

Board No:119
Ref. No: 176
Topic: Regional

CHALLENGE OF DIAGNOSIS OF NEURO-BEHÇET: A PROSPECTIVE STUDY

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Background and aims: Neurological involvement in Behçet Disease (BD) or Neuro-Behçet (NBD), although potentially a serious complication, seems to be overdiagnosed. The aim of this study was to determine potential clues to the correct diagnosis.
Methods: During September 2008 to February 2010, 918 BD patients were referred for evaluation of possible neurological involvement. of these, 114 had one or more neurological symptoms/signs, which were examined by a neurologist, experienced in NBD, and detailed relevant paraclinical investigations including blood and CSF studies, imaging methods (MRI- MRV-MRA, cerebral angiography), TCD, EEG, and EPs were performed. Results were correlated with all other data (such as retinal vasculitis, HLA typing, etc).
Results: of 114 BD patients with one or more neurological symptoms/signs, 44 were diagnosed as NBD, and 70 patients were

diagnosed as having neurological syndromes not related to BD. The most characteristic NBD syndromes were cerebral sinus thrombosis, meningoencephalitis, brainstem stroke, and movement disorder. Headache, seizures, and hemispheric stroke, although occurring with considerable frequency in both groups, do not help in diagnosing NBD. Complaints about depression and memory problem were considerably more frequent in patients with neurological syndromes not related to BD. Conclusion: No diagnostic clinical entity or paraclinical test for NBD was found. Clues to the correct diagnosis of NBD still appear to be definitive diagnosis of BD by international criteria, reasonable consistency of neurological condition with pathophysiology of BD, and absence of better explanation based on obtaining precise past history, family history, and performing complete investigations relevant to each patient.

Neurological Diagnosis	Group	
	NBD(%)	BD & unrelated neurologic symptom (%)
Headache	31.8	42.9
Sinus Thrombosis	15.9	0.0
seizure	13.6	7.1
Stroke, Hemispheric	13.6	4.3
Menningoencephalitis	13.6	0.0
Stroke, Brain Stem Syndrome	9.1	0.0
Paresthesia of Limbs	9.1	0.0
Movement Disorder	4.5	0.0
Paraplegia	4.5	0.0
Memory Problem	4.5	11.4
Limb Aneurysm	2.3	0.0
Brain Aneurysm	2.3	0.0
Dementia	2.3	2.9
Glosopharyngeal Neuralgia	2.3	0.0
Depression	2.3	30.0
Mononeuritis Multiplex	2.3	0.0
Thyrotoxic Myopathy	0.0	1.4
Essential Tremur	0.0	7.1
Neuropathy	0.0	4.3
ADEM	0.0	1.4
Benign Positional Vertigo	0.0	1.4
Vestibular Neuronitis	0.0	5.7
Dysphagia	0.0	1.4
Parkinson's Disease	0.0	1.4
Carpal Tunnel Syndrome	0.0	4.3
Non-Specific Complaints	0.0	20.0

Board No:120
Ref. No: 183
Topic: Regional

Interim analysis of the clinical trial INCYTOB (Interferon alpha-2a versus Cyclosporin A for the treatment of severe ocular Behçet´s disease)

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Purpose:
To provide an interim analysis of the clinical trial INCYTOB (Interferon alpha-2a versus Cyclosporin A for the treatment of severe ocular Behçet´s disease) and to evaluate problems that may arise during conducting an investigator initiated trial for a rare disease in Germany.

Methods:
INCYTOB is a national, prospective, controlled, randomized multicenter trial performed by an interdisciplinary cooperation between rheumatologists and ophthalmologists to assess the efficacy and safety of interferon (IFN) alpha-2a compared to cyclosporine A (CSA) in patients with severe eye involvement due to Behçet´s disease (BD). To be eligible for inclusion, patients have to suffer from complete BD and have to present with active ocular disease affecting the posterior eye segment. Randomization is done in a 1:1 ratio either to IFN alpha-2a or to CSA.
In case of inefficacy or side effects a crossover is planned.

Results and Conclusion:
Since 2007 thirty patients (of 100 patients who will be necessary for statistical analysis) in currently 10 sites have been included. An interim safety analysis of 27 patients revealed a higher crossover rate from CSA to IFN than vice versa (6 vs. 1). The number of adverse events was not different between both treatment arms. Reasons for crossover were mainly inefficacy or adverse effects (eg. CNS- manifestations under CSA). Recruitment rate is insufficient, and more centres are presently recruited to overcome this problem.

Board No:121
Ref. No: 195
Topic: Regional

IS BEHCET'S DISEASE A RISK FACTOR FOR COLON PERFORATION DURING THE COLONOSCOPIC EXAMINATION?

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Introduction/aim: The intestinal lesions of Behcet's disease most commonly found in the ileocecal area, and colonic involvement is less frequent. In this study patients, who had perforation as a complication of colonoscopic perforation during or after colonoscopic procedure, were analyzed retrospectively.
Materials and methods: May 2002 to December 2007; for symptoms or findings of constipation, diarrhea, rectal bleeding and anemia. 2615 colonoscopic procedure performed, of which 135 of the cases had Behcet's disease. All the procedures were performed by one experienced endoscopists. Iatrogenic perforation occurred in 3 patients [(median age 42, (range, 21-62), (2 men, 1 woman)] (incidence, 0.11%). All these 3 cases had Behcet's disease (incidence, 2.2%). In 2 of these patients colon explored through caecum . Proximal colonic and ileal deep ulcers with the diameter up to 2 cm were found in these cases during the procedure. In the 3rd case colon explored through midtransvers colon. Ulcers were observed in sigmoid, decendens and transverse colon. Perforations were diagnosed shortly after the procedure with the symptoms and signs of perforation. All these three patients underwent surgery and ileal resection and right hemicolectomy were performed.
Discussion: Colonoscopies are performed not only for diagnosis but also periodically in the follow-up to evaluate the effectiveness of the therapy in Behcet's disease. However, colonic perforation can occur during the colonoscopic procedures. Especially, discrete ulcers with typical round or oval ''punched-out'' appearance have a tendency to bleed or perforate.

Board No:122
Ref. No: 200
Topic: Regional

Risc factors for the clinical severity of Behçet's disease
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Behçet's disease is a chronic, relapsing, systemic vasculitis of unknown aetiology. The disease runs a chronic course with unpredictable exacerbations and remissions. In the present study, we aimed to determine the effects of possible risc factors on the clinical severity of Behçet's disease.
A total of 224 patients with Behçet's disease (96 female, 128 male; mean±SD age, 36.4±10.4 years), diagnosed according to the criteria of the International Study Group for Behçet's Disease were included in the study. The demographic findings and clinical features were recorded. Patients were also assessed for clinical severity score as previously described.
In logistic regression analysis, male gender ($p=0.05$) and increased numbers of symptoms at the diagnosis ($p<0.001$) were found to be a significant risk factors for the severity of Behçet's disease. Clinical severity score was higher in those patients carrying HLA-B51 antigen. However, the difference was not statistically significant ($p=0.097$).
Our results indicate that male sex and patients with increased numbers of organ involvement at the diagnosis are associated with more severe disease. Therefore, close monitoring and appropriate treatments are mandatory to decrease the morbidity and mortality of the disease in these patients.

Board No:123
Ref. No: 208
Topic: Regional

GLOBAL PATTERN OF NEUROLOGICAL INVOLVEMENT IN TUNISIAN PATIENTS: A STUDY ON 182 PATIENTS
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Introduction:
Prevalence of Behçet's syndrome (BS) clinical manifestations is characterized by a great geographic and ethnic variety. Neurological involvement (NI) is seen in 3-44% of patients with BS. There are two major subtypes of NI in BS: parenchymal CNS involvement and cerebral vascular lesions involvement.
Objective:
Report the epidemiological and clinical characteristics of NI in Tunisian patients with BS.
Patients and methods:
182 patients (mean age : 39 years, sex ratio: 1.56) recruited in the Internal Medicine Department of Fattouma Bourguiba Hospital and fulfilling the international study group criteria for diagnosis of Behçet's disease, were retrospectively enrolled in the study. Demographic and clinical aspects were determined for patients with NI.
Results:
Fourth of the patients had NI (43/182). They were 26 males and 17 females (SR: 1.53). Their mean age was 41 years. The CNS was involved in 35/43 cases. There were isolated headaches in 65% of cases. An isolated intracranial hypertension was observed in 14% of patients. Fifth of the patients had a parenchymal CNS involvement and a cerebral vein thrombosis was shown by a magnetic resonance angiography in 15/43 cases. Cranial nerves were affected in only 3 cases. Psychiatric involvement was observed in 11 patients.
Comments:
In Tunisia, prevalence of NI in BS is quite the same as other Arabic countries (Egypt, Lebanon and Jordan). All over the world, the clinical pattern of NI has globally the same characteristics: a male predominance, frequency of CNS parenchymal involvement and thrombosis of dural sinuses. Peripheral NI remains rare.

Board No:124
Ref. No: 210
Topic: Regional

Investigation of Bactericidal Effect of Uroepithelial Cells Against Streptococcus pyogenes and Escherichia coli in Behcet's Disease Patients

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Although etiology of Behçet's disease is unknown, viral and bacteriologic agents, genetic, environmental and immunologic factors are thought to be responsible. Epithelial cell besides being a barrier, has critical role in immun regulation and tolerance. The aim of the present study is to investigate the bactericidal effect of epithelial cells against S. pyogenes and E. coli in Behcet's disease patients (BDP). Urine epithelial cells as effector cells and S. pyogenes ATCC 49619 and E. coli O75 strains as target cells were prepared and stimulated with bacteria in plates with an effector/target ratio of 1/1. After 1 hour incubation, supernatants were collected for growth inhibition assay and were inoculated onto blood agar plates for S. pyogenes and agar plates for E. coli. Following overnight incubation at 37ºC, colony forming units were enumarated and bactericidal effect were calculated. According to the results, the bactericidal effect against S. pyogenes and E. coli in BDP were 31,6% and 23,3%, respectively. These percentages were 46.8% and 60% for healty controls. In terms of bactericidal effect against S. pyogenes, there was no statistically significant difference between BDP and healty controls ($p>0.001$). On the other hand, the bactericidal effect of BDP against E. coli was significantly lower than that of healty controls ($p<0.001$). To our knowledge, this is the first study explored the bactericidal effect of urine epithelial cells against S. pyogenes and E. coli. Our findings might depend upon the lack of productive immun mechanisms in host defence against E. coli, the most frequent agent of urinary tract infections, in BDP.

Board No:125
Ref. No: 211
Topic: Regional

Endoscopic Findings, and Gastrointestinal Involvement of Behçet Patients; Do They Mean The Same?

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Introduction: Frequency of Gastrointestinal System(GIS) involvement of Behcet's disease(BD) varies between 3–60%. However, there are endoscopic abnormalities in up to 93% of these patients. The aim of this study is to describe significance of endoscopic findings, their pathologic, and clinical correlation.
Material and Methods: The study was conducted at Ankara University Multidisciplinary Behçet Clinics, and Gastroenterology Endoscopy Departments between 2005–2010 with 115 consecutive cases (40,9% male) who have undergone, upper or lower GIS endoscopy. Mean age was 39,6±10,2. 151 endoscopies were performed in 108 cases[Median=1,(Min-Max=1-6)], whereas 48 colonoscopies were done in 37 cases[Median=1,(Min-Max=1–3)]. Pathology, hemoglobin, sedimentation rate(ESR), and C–reactive protein(CRP) related with inflammatory activity were collected.
Results: There were 9(7,8%) patients having GIS involvement clinical or pathologically, However, many endoscopic, and colonoscopic abnormalities were detected(Table 1). At only 4,9% of endoscopies, vasculitis was documented pathologically. Median ESR was 16(Min-Max=1–103), CRP 3,0(Min-Max=0–162), and Hb level was 12,6±1,9. In the correlation analysis, vasculitis is inversely proportional with Hb($r=-0,391$, $p=0,0001$), whereas it has no affect on ESR, and CRP.
At 7(18,9%) colonoscopies, vasculitis was detected pathologically. ESR level was 39,4±31,8, median CRP was 3(Min-Max=0-126), and Hb was 11,4±2,1. In the correlation analysis, only CRP was found related to vasculitis($r=0,372$, $p=0,04$). Although elevated ESR seemed to be related to activity, it didn't reach significance.
Conclusion: Despite typical endoscopic appearance, pathology doesn't always confirm vasculitis, so clinical judgement is important. In our country, prevalance of GIS involvement is lower than many others, possibly due to unknown environmental, and genetic factors.

Endoscopic Finding	Percentage (%)

Relaxed LES	47,6
Antral Gastritis	33,1
Reflux Esophagitis	29,0
Atrophy	20,0
Pangastritis	13,8
Sliding Hiatal Hernia	10,3
Gastric Ulcer	10,3
Bulbitis	9,0
Duodenal Ulcer	6,9
Colonoscopic Finding	
Normal	32,4
Edema – erythema	17,6
Apthous ulcer	35,3 (Ileocecal20,6 %, Rest of colon 17,6 %)
Ulcer >1cm	14,7 (Ileocecal5,8 %, Rest of colon 8,9 %)

,**Board No:126**
Ref. No: 212
Topic: Regional

Urethral meatus ulcer with dysuria sypmtom in Behcet's disease
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Genital ulcers are the second main symptom encountered in Behcet's disease. They are similar in appearance and course to oral ulcer, but may not recur as often and can have a scarring tendency. The scrotum is the most frequently involved site in males. Ulcers can also be observed on the shaft and glans penis. In females, the ulcers most commonly occur on the labia but the vaginal mucosa and rarely the cervix can also be affected. Vaginal ulcers often cause a discharge. Genital ulcers may occur in both sexes in the groin, perineal and perianal area. We present a 35 year old male patient had oral ulcer, painful genital ulcer with dysuria sypmtom and papulopustuler lesions. In dermatologic examination, we observed genital ulcer on the scrotum and inside of the urethral meatus. Therefore, we recommend a careful examination urethral meatus in patients with dysuria sypmtom and Behcet's disease should assest in conditions presenting with meatal/urethral ulcer.

Board No:127
Ref. No: 215
Topic: Regional

A MUCOCUTANEOUS ACTIVITY INDEX FOR BEHCET'S DISEASE
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Aim: The aim of this study was to develop a disease specific mucocutaneous activity index in Behcet's disease (BD).
Materials and Method: Forty-five BD patients (F/M:29/16, mean age:36.6±11.1 years) were included in the study. Pain and functional disability were subscales of both Genital ulcer activity index (GI) and Erythema nodosum activity index (EI). Mucocutaneous index (MI) was composed of GI, EI and Composite index, previously validated, for oral ulcer (Mumcu 2009). Score of MI could be between 0 and 30 (0-10 points for each involvement). The score was evaluated in patients with both active and inactive disease for content validity.
Results: MI score (7.2±6.5) was higher in active patients (n=30, 10.7±4.9) than inactives (n=15,0±0)(p=0.000). Scores of CI, GI and EI were also lower in inactives (0±0) compared to active ones (7.9±1.7, 7.7±1.3 and 6.8±2.8, respectively) for each involvement. MI score was lower in patients treated with immunosuppressives (1.7±4.2) than colchicine (11.1±4.8) (p=0.000). In addition, MI was correlated with disease severity score (7.2±2.1)(r=0.85 p=0.000).
Conclusions: We developed a mucocutaneous activity index which helps to evaluate mucocutaneous-specific clinical symptoms and the effects of treatment modalities in BD.

Board No:128
Ref. No: 218
Topic: Regional

CHANGES IN ORAL HEALTH IN PATIENTS WITH BEHCET'S DISEASE: 10-YEAR FOLLOW UP.

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Aim: The aim of this study was to evaluate changes in oral health parameters in patients with Behcet's disease (BD) in a 10-year follow-up.
Method: Non-selected consecutive 18 BD patients (F/M: 12/6, mean 9.9 years) followed by clinical and laboratory examinations±age:36.4 for 10 years regularly were included in the study. Oral health was evaluated by dental and periodontal indices. Patients were given oral hygiene education in each visit regularly.
Results: Disease severity score (4.4±1.8) was lower at baseline than 10-year follow up (5.6±1.6)(p=0.000). Although scores of periodontal indices were lower and frequency of tooth brushing was higher in the 10-year follow up than those of baseline, no significant difference was observed (p>0.05). The number of extracted teeth was found to be similar at baseline and follow-up.
Conclusion: Data from this study indicate that, in BD patients, dental and periodontal health remained stable by motivation and education for oral hygiene in a 10-year follow-up. However, whether better oral hygine is reflected in improving the disease course requires further studies.

	Baseline	10-year follow-up	
	Mean SD	Mean SD	p
Plaque index	2.1± 0.9	1.8± 0.9	0.32
Gingival index	2.3± 0.9	2.1± 1.05	0.55
Sulcus bleeding index	2.3± 0.7	2.1± 0.9	0.64
Periodontal pocket depth	3.1± 0.7	3.02± 0.6	0.87
DMFT	9.5± 5.9	11.0± 6.5	0.36
The number of extracted teeth	6.8± 5.9	7.4± 7.5	0.70
The number of carious teeth	3.4± 2.4	2.6± 2.2	0.32
Frequency of tooth brushing	0.9± 0.6	1.7± 1.2	0.06

Board No:129
Ref. No: 222
Topic: Regional

A Tunisian version of the Behçet's Disease Clinical Activity Form

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INTRODUCTION
Behçet's syndrome (BS) is characterized by a heterogeneous vessel involvement, a fluctuating natural history and by the absence of biological markers correlated to disease activity that's why objective clinical scores are needed for the assessment of its activity. Many activity scores were proposed. The Behçet's Disease Clinical Activity Form (BDCAF) is the most recent and used one.
Aim:
do a cross-cultural adaptation of the BDCAF to the Tunisian dialect (Arabic language) and test its reliability among Tunisian patients.
Patients and methods:
Two Tunisian Internal Medicine physicians translated the BDCAF from English to Tunisian dialect. The two versions were then back translated. Translations and back translations were all reviewed by clinicians and English Teachers and finalized into one Tunisian BDCAF form called Tu-BDCAF. To test its reliability, the activity of BS was assessed in 40 BS Tunisian. Patients were questioned by two BS specialists the same day (Tu-BDCAF1 and Tu-BDCAF2) to test inter-observer reproducibility and twice by the same physician at 24 hours interval to assess the intra-observer reproducibility (Tu-BDCAF1

and Tu-BDCAF3). K coefficient was used to test the concordance between qualitative variables and correlation between quantitative variables was evaluated used Pearson coefficient.
Results:
Patients were 23 males and 17 females with a mean age of 38 years. Results of Inter and intra-observers agreement concerning all clinical manifestations involved in the BDCAF score are listed in table 1
Comments:
there was a good correlation between global scores calculated by the two physicians the same day (Tu-BDCAF1–Tu-BDCAF2) (r = 0.94, p<0.0001) and also between the scores calculated by the same clinician at different times Tu-BDCAF1-Tu-BDCAF3 (r =0.98, p <0.

Clinical manifestation	Interobserveragreement ( ) (n=40)	Intraobserveragreement ( ) (n=30)
Headache	0,59	0,89
Buccal aphtosis	0,92	1
Genital aphtosis	0,68	0,85
Erythema nodosum	0,56	0,77
Necrotizing pseudofolliculitis	0,73	0,81
Arthralgias	0,78	0,84
Arthritis	0,48	0,34
Nausea/Itching	0,56	0,51
Diarrheas	0,25	0,71
Ocular involvement	0,79	0,93
Neurological involvement	0,81	1
Large vessels involvement	0,75	1

Board No:130
Ref. No: 229
Topic: Regional

The condition of "Behçet syndrome" in some of major cities in Iran that no referred to Rheumatology Research Centre

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Background: "Behçet syndrome" is a disorder which involves several organs. Several diagnostic criteria have been developed based on clinical findings. The purpose of this study was to express the so-called outcomes.
Materials and Methods:The current research was performed on 202 patients with the syndrome, of whom no one had referred to Rheumatology Research Centre in" Tehran University", before. All diagnoses were done according to International criteria. Referees' data were derived from their medical files and then analyzed.
Results:70% of studied population was males and 30% were females. Average age was 31.6±6.9. Average duration of involvement was 4.8±2.7. All patients; due to International criteria; had suffered from oral aphthous ulcer. Other clinical findings were as follows: Anterior Uveitis 30.2%, Posterior uveitis 12.4%, Pan uveitis 6.9%,Retin vasculitis 4.5%, Arthralgia 71.3%,Arthritis 29.2%,Genital aphthous ulcer 37.6%, Pseudofolliculitis 57.4%, Erythema nodosum 47.5%, Phlebitis 11.4%, Thrombophlebitis 8.4%,Arterial aneurism 0.5%,Brain parenchyma involvement 0.5%.No sign of brain vessels involvement was observed. laboratorial results were as follows: high ESR 55.9%, positive CRP 52.5%, HLA-B5 30.7%, HLA-B51 30.2%, HLA-B27 11.4%, RPR 5.9%. Pathergy test in 69.8% of cases was positive.
Conclusion:
After oral aphthous ulcer, the most visible clinical demonstration in patients involved with "Behçet syndrome" are Arthralgia and Pseudofolliculitis which the longer the duration of involvement, the more pervasive they are. The most prevalent ocular involvement is Anterior Uveitis. The most prevalent laboratorial findings are high ESR and positive CRP. The most seen HLA in these patients is HLA-B5. Pathergy test approximately will be positive in 2/3 of cases.

Board No:131
Ref. No: 235
Topic: Regional

Rheumatologic manifestations of Behçet's disease
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The purpose of our work was to define the epidemiology and the clinical features of Behçet's disease giving special attention to unusual forms.
We retrospectively reviewed the medical records of 250 cases with joint manifestations among 330 of Behçet's disease seen over 20-year period who met the international study group of Behçet's disease criteria.
Joint manifestations were present in 75% and were inaugural in 30% of cases. Inflammatory arthralgias were the most common manifestation and observed in 81%, interesting mainly the large lower limb joints. Arthritis was less common: oligoarthritis (35%), monoarthritis (25%) and polyarthritis (15%). Sacroiliitis was observed in 7% of patients. Unusual forms like destruction polyarthritis, popliteal cyst, myositis were rare.
Joint involvement is common in BD and could be the first manifestation of the disease. Most of the patients present with inflammatory arthralgias of the large lower limb joints. Disease course is usually favorable, spontaneously or with treatment. However, in our study population, skeletal manifestations were responsible for significant disability

Board No:132
Ref. No: 236
Topic: Regional

Behçet's disease and renal involvement
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The renal involvement in Behcet's disease is rare. The clinical features vary from urinary sediment's abnormalities to end stage renal failure.
We propose to study the clinical, biological and histological data, the therapeutic management and the prognosis of patients.
We report a retrospective study including 8 patients representing 1.23 % of all cases of Behçet's disease.
The average age of the patient was of 37 ± 12.35 years with a clear male prevalence.
The urinary signs were discovered fortuitously by the strips in the majority of the cases after an average of 18 months. It was essentially proteinuria and hematuria. Renal insufficiency and hypertension were rare. Pathological study highlighted 3 cases of amyloidosis, 2 cases of IgA nephropathy, 1 case of minimal change disease, 1 case of endo and extracapillary glomerulonephritis and at least 1 case of interstitial nephropathy.
The patients having GN were treated by corticoids and immunosuppressive agents and whose having an interstitial nephropathy was treated symptomatically with good evolution in the majority of the cases.
Only one patient was dead, he had amyloidosis. The prognostic depended on the precocity of diagnosis, the histological type and the treatment.
The renal involvement during Behçet's disease is rare. Amyloidosis and Ig A nephropathy are the most frequent. The treatment is much discussed.

Board No:133
Ref. No: 237
Topic: Regional

Multiple Aseptic Osteonecrosis in Behcet's disease: report of one case
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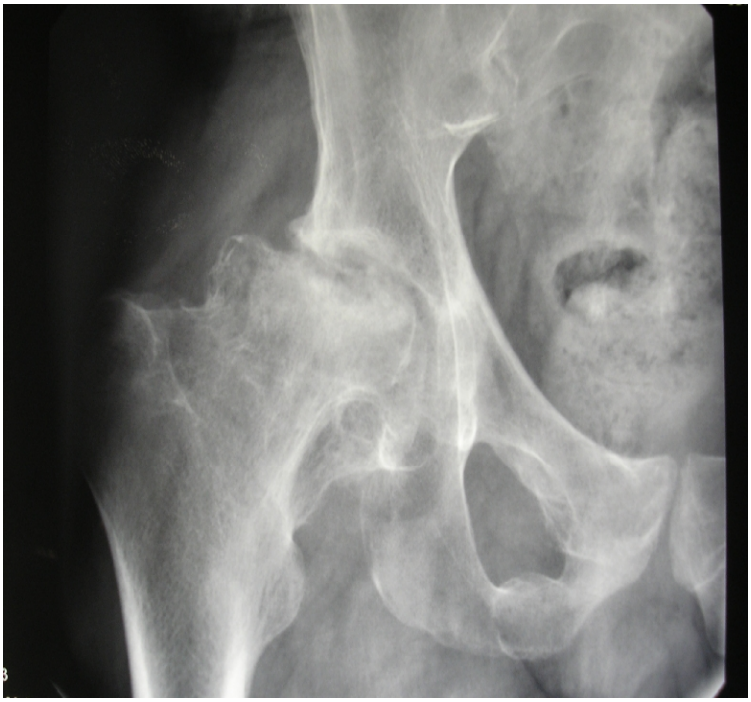
Osteonecrosis in association with Behcet's disease is rarely reported.
We describe one case with Behcet's disease (BD) developing osteonecrosis the 2 humeral heads and the 2 femoral heads, at coxite stage in the right femoral head. He had had a past history of significant corticosteroid administration to treat ocular complications of BD such as uveitis. Anticardiolipin (aCL) antibodies were negative.
Corticosteroids are useful in Behçet's disease, but they probably act also on the underlying immune vascularitis. The occurrence of pain in weight-bearing joints during treatment should suggest osteonecrosis and in such cases, beside radiography, CT scan and MRI might provide an early diagnosis. The cases described in literature of osteonecrosis are often in relation with the presence of aCL antibodies, we didn't find these antibodies in our case.

Aseptic Osteonecrosis of the right femoral head



Board No:134
Ref. No: 238
Topic: Regional

Late onset of Behçet's disease
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Behçet's disease (BD) affects mostly the young adult, usually in the third and fourth decades, with an exceptional onset after the age of 60 years. We report one case of 66-year-old woman with a history of thrombophlebitis and pulmonary embolism who presented with oral ulcerations, pseudofolliculitis and pustular rash. Ophthalmologic examination showed bilateral uveitis. All stabilized by colchicine and immunosuppressive therapy. HLA B 51 was positive. The clinical course of BD is not indolent in the patients with late-onset BD. Physicians should be aware that BD can occur in older patients, and close attention regarding their disease activities is warranted as their clinical courses may not be as benign as previously believed.

Board No:135
Ref. No: 239
Topic: Regional

COLCHICINE DOES NOT DECREASE THE NEED FOR IMMUNOSUPPRESSIVE USE AT LONG TERM IN BEHÇET'S SYNDROME (BS)

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BACKGROUND: A recent survey looking at the long term outcome of patients who had entered a controlled trial of thalidomide showed a trend for less immunosupressive use among those who had previously used colchicine (1). This time we looked at the long term outcome of patients who took part in a 2 year, randomized, placebo controlled trial of colchicine.
METHODS: 116 BS patients (60 men, 56 women) with mucocutaneous involvement were re-evaluated 16.6±1.1 years after the trial ended. The main outcome measure was the need for immunosuppressive use during follow-up.
RESULTS: Outcome information was achieved in 90/116(78%) patients. of these, 51 were men and 39 were women (mean age 27.1±5.3, disease duration 8.4±8.1 months at randomization). The group distribution of patients who could not be reached was similar. During follow-up, 18/51(36%) men and 8/39(20%) women had received immunosuppresives (Table). There was no difference in immunosuppresive use between patients who had been randomized to colchicine or placebo arms (men: 11/25 colchicine vs 7/26 placebo and women 3/18 colchicine vs 5/21 placebo).
CONCLUSION: Initial use of colchicine does not decrease the need for immunosuppressive use, hence the development of major organ involvement of BS patients at long term.

1. Hamuryudan V et al. Rheumatology (Oxford). 2010 Jan;49(1):173-7

	Men (n=51)		Women (n=39)	
	Colchicine (n=25)	Placebo (n=26)	Colchicine (n=18)	Placebo (n=21)
Eye involvement	1	-	1	1
DVT	2	2	1	-
Pulmonary artery aneurysm	-	1	-	-
Neurologic involvement	2	1	-	-
Gastrointestinal	1	-	-	1
Arthritis	2	3	1	3
Mucocutaneous involvement	3	-	-	-

Board No:136
Ref. No: 242
Topic: Regional

TNF-ALPHA AND OSTEOPROTEGERIN IN ASSESSMENT OF BONE METABOLISM IN PATIENTS WITH BEHCET'S DISEASE

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Behcet's disease (BD) is a systemic vasculitis with recurrent attacks that TNF-alpha is reported to be increased. TNF-alpha both stimulates osteoclastogenesis and inhibits osteoblast function. Osteoprotegerin (OPG), a member of the TNF receptor family, is reported to inhibit the formation and activity of osteoclasts. OPG prevents the bone resorption caused by TNF-alpha. We aimed to assess bone metabolism of Behcet patients.
41 Behcet patients (F/M 21/20, mean age 42.3±11.65) and 36 healthy controls (F/M 21/15, mean age 41.7±10.99) were

enrolled. Serum TNF-alpha, OPG, osteocalcin, thyroid function tests, total testosteron, calcium, phosphorus, ALP, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) levels and urinary creatinine and deoxypyridinolin (DPD) from second morning void were tested. Bone mineral densitometry (BMD) of both lumbar spine and femoral neck was calculated by DEXA method at the same day of serum sampling.
In patient group, CRP, DPD, urinary DPD/creatinin (DPD/cre), and TNF-alpha levels were significantly high ($p<0.05$, $p<0.001$, $p<0.05$, $p<0.001$, respectively). Despite, the osteocalcin levels were high in patient group this was not significant statistically. No significant difference was found in OPG levels. BMD levels of femoral T and Z score, vertebral T and Z scores, femoral neck, trochanter and wards triangle were all significantly lower than the controls. Nobody had osteoporotic levels in patient group. In patient group no correlation was found between the parameters studied. OPG levels were lower but no significant difference was found between the controls. However, there were positive correlations between OPG and both ESR and CRP.
All these results suggested a minimal decrease in BMD that probably be the result of the inflammatory activity and intermittent character of the BD.

Board No:137
Ref. No: 246
Topic: Regional

Infliximab long-term treatment for uveitis in Behçet's disease
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Objective: to evaluate the safety and efficacy of long-term administration of infliximab in Behçet's Disease patients (BD) with severe uveitis refractory to conventional treatment.
Patients and methods: Patients with BD were treated with infliximab (5 mg/kg on day 1 and 15, every months for 4-6 months and every 2 months thereafter) if they had failed to respond to conventional therapy with corticosteroids plus at least two different courses of immunosoppressive drugs (azathioprine, cyclosporine or methotrexate). The patients were monthly submitted to complete ophthalmological evaluation and laboratory tests. Changes in visual acuity and laboratory values, presence of uveitis relapses, and steroids and immunosuppressives dose changes were recorded throughout the follow-up.
Results: 10 BD patients with a mean disease duration of 79.1±52.5 months (range 22-203 months) were enrolled. The mean duration of infliximab treatment was 44.7±28.2 months (range: 12 – 88 months). Twenty uveitis relapses were recorded during follow-up (mean: 0.57 relapse/patient/year) in 7 patients, 3 patients (30%) being relapse-free since their first infusion. Eight relapses (40%) were related to delayed scheduled infusion (3 because of concomitant infections). The mean daily prednisone dose of the patients dropped from 13.9 mg/day in the pre-treatment period to 5 mg/day in the infliximab period ($p<0.0001$). Eight patients (80%) stopped completely steroids; six (60%) stopped immunosuppressive drugs also. Only 2 patients (20%) presented a transitory transaminase elevation. One patient (10%) had to stop infliximab because of headache and vomiting (brain nuclear magnetic resonance normal). Visual acuity remained unchanged from baseline in 13 eyes (65%), and improved in 7 eyes (35%)
Conclusion: infliximab therapy is safe and effective in preventing uveitis relapses being able to maintain baseline visual acuity in all the patients after a mean follow-up of 3.5 years

Board No:138
Ref. No: 247
Topic: Regional

Multi-drug resistance and side-effects in a patient with Behçet's disease
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OBJECTIVE: to report the uveitis course in a multidrug-resistance patient with Behçet's disease. CASE REPORT: a 22 year old male with BD (bilateral panuveitis, retinal vasculitis, oral ulcers, erythema nodosum, arthralgia, epididymitis) was treated from 1999 to 2007 with steroids (mean daily dose >20 mg/prednisone/day plus peribulbar injections as needed) and immunosuppressive drugs sequentially with the following results and drug-related side-effects: Cyclosporine A (5 mg/kg/day), unsuccessful, decrease of renal function and nephrolithiasis; Methotrexate (20 mg/week) alone or in combination with CyA or Azathioprine, unsuccessful, transaminases elevation; Interferon alpha (3-4.5 MU three times/week), severe

depression with a suicide attempt; Chlorambucil, severe leucopenia requiring withdrawal; two brief courses of Infliximab (5 mg/kg/infusion) which were complicated by the onset of pulmonary tuberculosis (PPD and Chest X ray negative prior therapy), and by a pulmonary legionellosis, respectively one year apart; Azathioprine (1,5 mg/kg/day) and Mycofenolate mofetil (2 gr/day), unsuccessful. Despite these therapies during follow-up the patient showed multiple relapses of uveitis and of systemic BD related lesions, and he developed severe ostheoporosis with multiple vertebral fractures, bilateral cataract and steroid-associated glaucoma. Since 2007 he has been taking only prednisone and he is finally free from ocular uveitis and systemic symptoms. His final visual acuity is 9/10 in the right eye and counting fingers in the left one.
CONCLUSION: Usually BD patients are responsive to immunosuppressive drugs, especially the newest ones (anti-TNF alpha). Nevertheless there is also a possibility of a multi-drug resistance as well as of a multiple drug-related side effects.

Board No:139
Ref. No: 248
Topic: Regional

Dexamethasone exerts dual actions in reducing monocyte thrombogenic profile by enhancing fibrinolytic activity and reducing procoagulant activity

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BACKGROUND
Patients with Behcet's syndrome (BS) have an increased predisposition to venous thrombosis due to vascular inflammation. Although corticosteroids are often used for treating thrombosis in BS, it is not known whether they have a direct beneficial effect on thrombogenicity or whether they just act indirectly by suppressing the inflammatory response.

OBJECTIVES
We sought to determine the effect of dexamethasone on monocyte thrombogenicity.

METHODS
Monocytes were isolated from citrated venous blood drawn from 10 healthy donors. Monocyte fibrinolytic and procoagulant activities were assessed using in vitro assays, (a) colorimetric Coomassie clot lysis assay, and (b) turbimetric clotting assay. Monocytes were either untreated or treated with dexamethasone 10nM. In the clot lysis assay, fibrinolysis was determined from the amount of Coomassie released by the clot (absorption at 540nm). In the clotting assay, clot turbidity was measured using absorption (405nm), and turbidity curves were analyzed to determine (i) lag time (T_{lag}); (ii) maximum rate of turbidity change (V_{max}) and (iii) maximum turbidity (ABS_{max}).

RESULTS
Monocytes enhanced clot lysis by 33% at 24h ($p<0.001$) and 45% at 96h ($p<0.001$). The addition of dexamethasone 10nM enhanced clot lysis by 97% at 24h ($p<0.001$) and 108% at 96h ($p<0.001$). Dexamethasone also reduced monocyte procoagulant activity at 24h with 68% increase in T_{lag} ($p=0.019$), 54% decrease in V_{max} ($p=0.049$) and 13% decrease in ABS_{max} ($p=0.004$).

CONCLUSIONS
Dexamethasone has beneficial effects on monocyte thrombogenic profile by (a) enhancing fibrinolytic activity; and (b) reducing procoagulant activity. These in vitro data suggest that corticosteroid therapy may have a direct effect in reducing thrombotic complications of BS.

Figure 1
Effect of monocytes ± dexamethasone on clot lysis as determined using a colorimetric Coomassie clot lysis assay (* denotes statistical significance compared to monocytes, p<0.001)

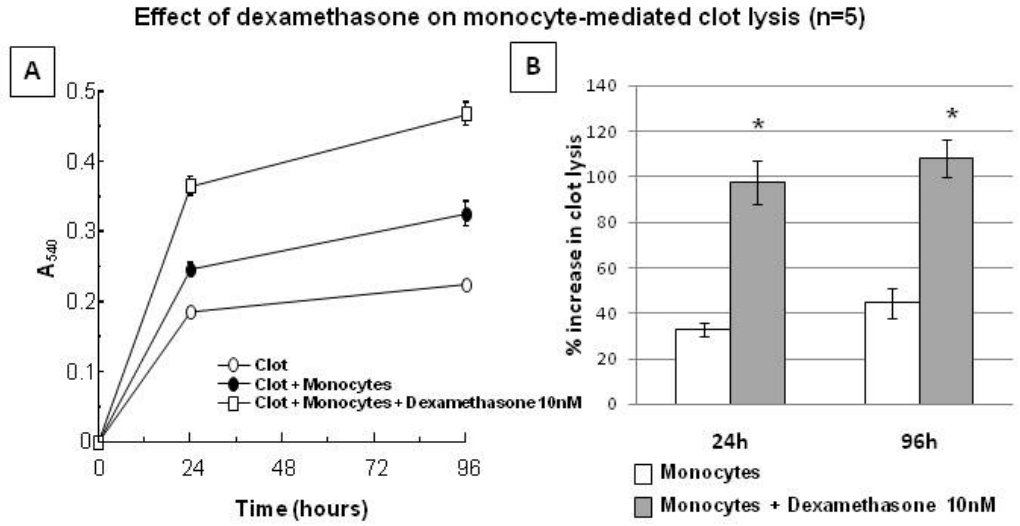
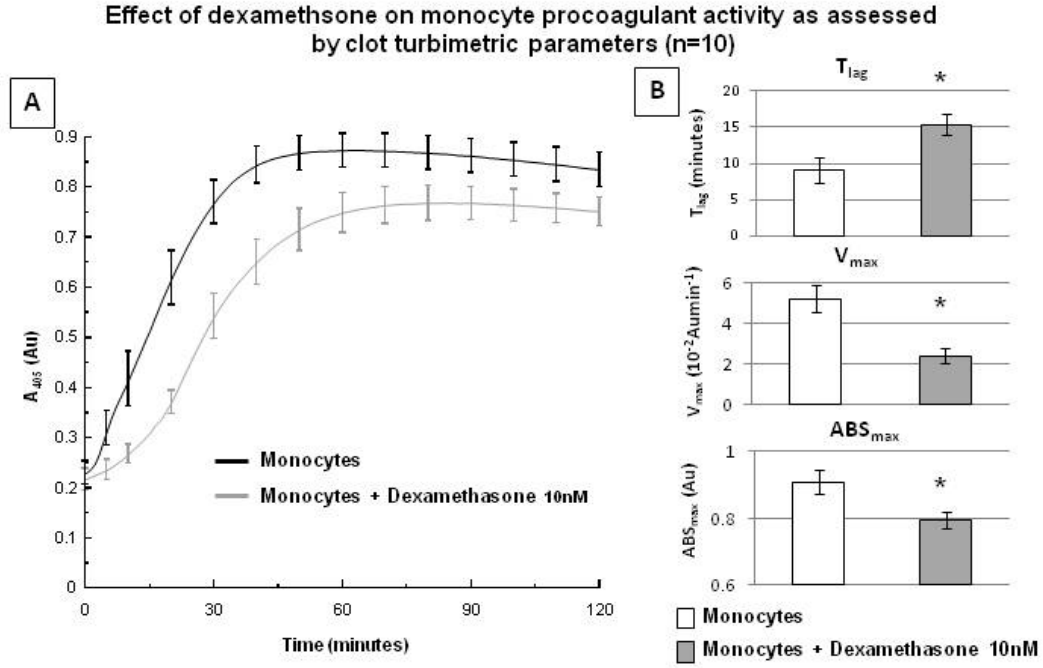


Figure 2
Effect of dexamethasone on monocyte procoagulant activity as assessed using a turbimetric clot assay at 24 hours (* denotes statistical significance compared to monocytes , p<0.05)



Board No:140
Ref. No: 251
Topic: Regional

Behcet's syndrome in the United States: Clinical characteristics, treatment and ethnic/racial differences in manifestations of 518 patients

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Background: Behcet's syndrome (BS) is a systemic vasculitis is rare in northern Europe and the US. Previous reports have suggested that there may be ethnic and racial differences in disease presentation and possible clustering of manifestations. We started a dedicated Behcet's clinic in 2004 and now report on the disease characteristics of the first 518 patients.

Methods: All patients seen at the center have completed a MDHAQ, and a questionnaire about past medical history, medication use, Behcet's specific history, ethnic and demographic information. These data are prospectively collected and updated each visit. About 2/3 of patients live within driving distance of New York City while patients from over 45 states have been seen. Patients were analyzed as the whole cohort and then also separated into to 2 groups: Group A= with ethnic background in northern Europe and North America and/or self declared Caucasians without background around the Mediterranean and/or the Far East; Group B= Patients with an ethnic background in the Mediterranean, Middle East, North Africa, and Far East. These groups were compared for disease manifestations, demographic information and medication use.

Results: 518 patients (398 (77%) female, mean (SD) disease duration 4(6.5) years, age 35 (13.6), 324 (60%) fulfilled International Behcet's classification criteria) were analyzed. Disease characteristics of the whole cohort and those fulfilling criteria are given in Table 1. When divided by ethnic background, Group A had statistically more significant GI disease (47% vs. 28%, $p<0.001$) and more females compared to Group B (83% vs. 71%, $p=0.005$) for those meeting criteria. For the whole cohort, less eye disease and vascular involvement than other centers was reported. There was only one blind eye.
Most commonly used medication at baseline was low dose prednisone (62%),, followed by colchicine (46%), TNF inhibitors (21%) azathioprine (19%) and methotrexate (10%).

Conclusions: In this cohort of 518 Behcet's patients, largest cohort to be reported in the US. demographic and clinical differences were noted between patients with different ethnic backgrounds. There were significantly more female patients in the non-ethnic groups. The frequency of GI disease was also significantly more in this group. Eye disease prevalence for the whole cohort was far less than reported from other centers. These finding point to considerable heterogeneity in disease expression in BS at different geographic areas.

	Total cohort (%)	Meeting criteria (%)	Not meeting criteria (%)
N	518	313	205
Oral ulcers	481 (90%)	313 (100%)	151 (74%)
Genital ulcers	376 (73%)	285 (91%)	91 (44%)
Skin	342 (66%)	287 (92%)	55 (27%)
Arthritis	268 (52%)	199 (64%)	69 (34%)
GI	179 (35%)	121 (39%)	58 (29%)
Eye	144 (28%)	127 (41%)	17 (8%)
CNS	81 (16%)	59 (19%)	22 (11%)
DVT	20 (4%)	17 (5%)	3 (1%)
Pathergy	41 (8%)	37 (12%)	4 (2%)
HLA B51	58 (11%)	37 (12%)	21 (10%)

Board No:141
Ref. No: 259
Topic: Regional

Diagnosis and Management of Neuro-Behçet's Disease: International Consensus Recommendations
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Background:
The neurological complications of BD (neuro- Behçet's disease, NBD) are amongst the most serious and disabling manifestations. Evidence based research on NBD is scarce and difficult to conduct for various reasons.

Objectives:
To develop internationally agreed consensus recommendations on the main diagnostic and management issues in NBD.

Methods:
The project committee (SK, AAA, AS) invited specialists with interest in BD to form an international advisory group (52 members from 20 countries, including 23 neurologists).

SK completed literature search through Cochrane, Medline and Embase databases till January 2010. Drafts of recommendations are being drawn up by the project committee and will be sent to the advisory group members for comments.

A consensus group of 12 members, with neurologists in majority, will be chosen for voting on amended recommendations using the 9-point Likert scale and scores ≥7 will be considered as indicative of agreement.

Results:
The advisory group agreed on the scope of the project to include: criteria for NBD diagnosis, classification of patterns of presentations, differentiating NBD from mimics, role of certain investigations in diagnosis, and headache in BD. Management issues include: management of parenchymal and cerebral venous presentations, headache, role of disease markers, identification of prognostic factors, and outcome measures.

Two thirds of the advisory group members preferred to use the term neuro-Behcet's disease to describe the neurological manifestations of BD.

The agreed recommendations will be presented and published later in a peered review journal.

Board No:142
Ref. No: 260
Topic: Regional

Sclerosing cholangitis in Behcet's disease: a rare condition
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Introduction: The hepatobiliary conditions reported to occur in patients with Behcet's disease (BD) are cirrhosis, acute and chronic hepatitis, primary biliary cirrhosis and hepatocellular carcinoma. Sclerosing cholangitis (SC) is a very rare condition in BD.

Case report: A 43-year-old North African man, with known BD (recurrent oral and genital ulcers, pseudofolliculitis and thrombophlebitis), was admitted in our institution with jaundice and fatigue. Biological investigations showed raised alkaline phosphatase, transaminases and gamma-glutamyl transpeptidase. Viral hepatitis was excluded. Cholangio-MRI showed distal stricturing of the common bile duct; there was no evidence of gallstone, tumor or other pathologic process. Histological examination of liver biopsy showed non-specific changes.
Discussion: Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease of intra and/or extrahepatic bile ducts. A concentric obliterative fibrosis occurs which leads to bile duct strictures. The etiology and pathogenesis of PSC are still unknown. Secondary sclerosing cholangitis (SSC) is a sclerosing cholangitis that occurs secondary to a known pathogenic process or injury such as obstruction of the bile ducts due to tumor or gallstones, bacterial infections, surgical, chemical or ischemic injury. We think that involvement of the parabiliary arteries by BD vasculitis has probably caused ischemic damage to the common bile duct and led to sclerosing cholangitis in this patient.
Conclusion: SC should be considered in patients with BD presenting hepatobiliary abnormalities.

Board No:143
Ref. No: 269
Topic: Regional

Current Trends in the Management of Behcet's Syndrome
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OBJECTIVE: The aim of this study is to determine the current trends in the management of various aspects of BS in our clinic.

METHODS: This study was conducted in a dedicated multidisciplinary Behcet's syndrome clinic where there are over 7,000 registered patients. We recorded all current and past medications, indications for treatment and for discontinuation of treatment of all BS patients who visited our clinic during one month.

RESULTS: A total of 327 BS patients (175 men, 152 women; mean age 37.6±10.6; mean disease duration 9.9±10.3 months) were evaluated in our clinic in one month. 21 of these were new patients. Current drugs according to each system predominantly involved is given in the table. The usual trend for managing eye involvement was to start with azathioprine solo or together with cyclosprine-A and go on to interferon and later to TNF antagonists in resistant cases or when adverse events occur. Corticosteroids were started during flares, but in some patients it was hard to taper and stop corticosteroids due to recurrences. Among patients with deep vein thrombosis, only 2 were receiving warfarin. One had Factor V Leiden mutation and the other had antiphosholipid antibodies.

CONCLUSION: Patients with only mucocutaneous and joint involvement were mostly using colchicine. Among patients with predominant eye involvement, similar number of patients were using interferon and TNF-alpha antagonists. All patients with DVT were using or had used azathioprine. The 3 patients with pulmonary artery aneurysms had used cyclophosphamide and high dose corticosteroids and 2 are now using azathioprine.

Board No:144
Ref. No: 283
Topic: Regional

Neuro Behçet's Disease in Tunisia: Evaluation of 29 patients
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Background: Behçet's disease (BD) is a multisystem relapsing inflammatory disorder of unknown aetiology. Neurological involvement has been reported in the range of 5 to 40%. Neurological involvement should be carefully investigated because of worse prognosis.
Objectives: To determine the prevalence, clinical features and neuroimaging data of Neuro Behçet's disease (NBD) in our patients.
Patients and Methods: From a database of 98 patients with BD, all fulfilling the International Study Group criteria for the diagnosis of BD. Patients were divided into two groups according to the presence or not of NBD.

Results: the prevalence of NBD was 29.6%. There were 19 males and 10 females (M/F= 1.88). The mean age at neurological onset was 36 years. NBD was concomitant or preceded the diagnosis of BD in 10 cases. Headache occurred in 18 cases, 10 patients had pyramidal signs, 6 had motor defect and 6 patients had cerebellar syndrome. Psychiatric manifestations were noted in 6 patients. CSF was abnormal in 5 cases: lymphocytic meningitis (n=4), sub-arachnoid haemorrhage (n=1). In 9 patients MRI showed parenchymal lesions. One patient died and motor and cognitive sequels were seen in another patient.
Conclusion: prevalence of NBD in our population, with clinical and neuroimaging features, is similar to those of other populations.

Board No:145
Ref. No: 304
Topic: Regional

NEW INTERNATIONAL CRITERIA FOR BEHÇET'S DISEASE (ICBD)

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Background: Concerns over low sensitivity of the International Study Group (ISG) criteria for Behçet's Disease (BD) led to the need to reassess the criteria.
Methods: Physicians from 27 countries [International Team for the Revision of the International Criteria for Behcet's Disease] submitted data from 2556 patients with a clinical diagnosis of BD, as well as 1163 controls having at least one major sign of BD. These were randomly divided into training and validation sets. We used logistic regression, 'leave-one-country-out' cross-validation, and clinical judgment to develop a new International Criteria for Behcet's disease (ICBD) with the training data.
Results: In the training set, the ISG criteria had estimated 81.2% sensitivity considered too low, and 95.9% specificity. The new ICBD criterion assigns 2 points to each of ocular lesions, oral aphthosis and genital aphthosis, and assigns 1 point to each of skin lesions, central nervous system involvement, and vascular manifestations; ≥ 4 points indicates BD. In the training set the ISCB had 93.9% sensitivity and 92.1% specificity. In the validation set, the ISG had 85.0% sensitivity and 96.0% specificity. The new criteria had 94.8% sensitivity (95% confidence interval, CI: 93.4% to 95.9%), and 90.5% specificity (95% CI: 87.9% to 92.8%). Where pathergy skin testing is performed, slightly higher sensitivity can be achieved by assigning an additional point for a positive pathergy result.
Conclusions: The ICBD criteria has improved sensitivity yet slightly lower specificity compared to the ISG criteria. It is proposed that the ICBD be adopted to guide the diagnosis of BD.

Board No:146
Ref. No: 306
Topic: Regional

FUNCTIONAL RESULTS FOLLOWING CATARACT SURGERY IN OCULAR ADAMANTIADES-BEHÇET'S DISEASE

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Background: Adamantiades-Behçet´s disease (ABD) is a multi-system disorder with recurrent oral and/or genital ulcerations, skin lesions and ocular involvement. Left untreated, it leads to blindness and often to loss of the eye through secondary complications like phthisis or painful glaucoma. Secondary cataract formation often occurs due to recurrent inflammation and due steroid treatment and requires intraocular surgery.
Methods: Retrospective study of patients with ocular ABD who underwent cataract surgery. From 1982 to 2009 we treated 28 eyes from 17 patients with Phacoemulsification and intraocular lens implantation (n=16). Nine eyes remained aphacic. The mean Follow up was 124,5 months (9,1-267,7 months).
Results: 20 out of 25 eyes had better visual acuity following surgery. Eight eyes did not benefit from cataract surgery because of optic nerve atrophy or maculopathy. We observed a better mean visual acuity of +1.2 lines. The main postoperative complication was recurrent ueitis in 4 eyes.
Conclusion: In most of the cases a better visual acuity could be achieved with cataract surgery in patients with ocular ABD. Optic atrophy and maculopathy limit the functional results.

Board No: P-147
Ref. No: 096
Topic: Regional

Prevalence of atopic disease in the normal population and in Behçet's disease (BD) in Iran

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Background: Atopy is a hereditary polygenic disease, regrouping Atopic Dermatitis (AD), asthma, and allergic rhino-conjunctivitis. Epidemiologic studies show an increase of atopic diseases in Europe. The hypothesis of this increase is the urbanization and the life style of population. Objective: To study the prevalence of Atopy in BD and in normal population in Iran. Material & Methods: 348 subjects were enrolled; 108 BD (female 42.6%, male 57.4%) and 240 controls (female 41.3%, male 58.8%), p=0.814. The mean age in BD was 38.05, standard deviation (SD)=11.415, and 30.36 in controls (SD=11.421). Past or present history of atopy were checked in patients and their family. Result: In BD, 20.4% of the patients had either an atopy themselves or in their familly versus 51.3% in controls (Chi2=29.221, p<0.0001). In BD, 10.2% had an atopy themselves, versus 30% in controls (Chi2=16.102, p<0.001). In BD, familial history of atopy was found 15.7% versus 38.8% in controls (Chi2=18.241, p<0.001). In BD, the presence of atopy, both in patients and their familly together, was seen in 5.6% versus 17.5% in controls (Chi2=7.961, p=0.005). Individual atopic manifestations were as follow: asthma in BD versus control (0.9%/2.5%, p=0.333). Atopic dermatitis (7.4%/10.4%, p=0.375). Rhino-conjunctivitis (5.6%/20.8% p<0.001). Discussion: Both BD and atopic diseases are developed by a combination of genetic and environmental factors, but of different kind. The decrease of one is reported by the increase of the other. Conclusion: Atopic diseases were seen more frequently in the normal population of Iran than in BD. These results warrants a larger study.

Board No: P-148
Ref. No: 19
Topic: Paediatric

The prevalence of Behcet disease in a Druze community in Israel

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Objectives- To evaluate the prevalence of Behçet's disease (BD) in a Druze community in Israel.
Methods- We conducted a two-stage clinic-based survey in an Israeli Druze town. The first stage aimed to identify patients with recurrent aphthous stomatitis (RAS) in all patients who visited 3 of the largest clinics in the town during a period of 6 months. The second stage aimed to identify those patients with RAS who fulfilled the diagnostic criteria for BD according to the International Study Group (ISG) criteria.
Results- One thousand and eighty three subjects were interviewed, 63 of whom had RAS (5.8%). Two patients fulfilled the ISG criteria for BD, resulting in a calculated prevalence of 185:100,000. Another two patients with oral and genital aphthosis but without eye or skin lesions were diagnosed as suspected BD.
Conclusions- The very high prevalence of BD, as found in our study, places the Druze among the populations with the highest prevalence of the disease all over the world, though selection bias could account for overestimation of the actual BD prevalence. Our findings call for genetic studies to explore whether there is a genetic predisposition to BD in this population.

Board No: P-149
Ref. No: 20
Topic: Paediatric

Prevalence of Behcet disease in Israeli Yemenite Jews
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Objective: To assess the prevalence of Behcet's disease (BD) in the Yemenite Jewish community in Israel, in whom the frequency of HLA-B5 has been found to be very high (39%).
Methods: Patients attending a community health center in an Israeli town with a predominant Yemenite Jewish population were asked whether they had recurrent aphthous stomatitis (RAS). Those who had had 3 or more aphthous episodes during the previous year underwent an extensive interview and examination for the presence of other manifestations of BD.
Results: A total of 1500 subjects were surveyed, of whom 41 had RAS. All patients with RAS were interviewed and examined, yet none was found to have BD according to the ISG criteria.
Conclusions: Despite the high frequency of HLA-B5 among the Yemenite Jewish population in Israel, no cases of BD were found among 1500 individuals of this population. Our results point to a very low prevalence of BD among Israeli Jews of Yemenite origin. Furthermore, our findings imply that the presence of HLA-B5 may not be a risk factor for future development of BD in this population.

Board No: P-150
Ref. No: 59
Topic: Paediatric

Cognitive impairment in patients with Behcet disease, a case-control study
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Aim: To study cognitive impairment in patients with Behcet disease (BD), and the impact of different disease characteristics on it.
Methods: In a cross sectional study 65 BD patients without overt neurological symptoms and 43 healthy controls were enrolled. Three main aspects of cognitive functions including memory, visuospatial ability and frontal lobe executive functions were measured by Wechsler Memory Scale test, Wisconsin card sorting test and WAIS block design test respectively in all. We have also measured level of anxiety (Beck test) and depression (Hamilton test). Demographic features in all, and different disease characteristics such as disease activity, disease duration, prednisolone use were determined in BD patients. Comparison between means was done by t test, and logistic regression analysis was used for associations of different factors.
Results: Memory deficit was not statistically different between the two groups, both in prevalence (30.8% in BD vs. 18.6% in controls, p=0.18) and the mean scale (93.4±20 vs. 96.9±17.8, p=0.35). This was also true for mean visuospatial ability scale (24±9.9 vs. 25.2±9.5, p=0.55), and the prevalence of executive dysfunction (44.7% vs. 38.9%, p=0.64). The difference was still non-significant after adjustment for anxiety, depression, education level, disease duration, and prednisolone use. They were not related to disease activity (p>0.1). Disease duration was the only influencing factor on memory deficit in BD patients (p=0.02).
Conclusion: Cognitive impairment was not higher in BD patients without neurological involvement than healthy controls. Disease activity had no influence, while longer disease duration was the only influencing factor on memory deficit.

Board No: P-151
Ref. No: 84
Topic: Paediatric
Chronological manifestations in Japanese patients with Behçet's disease: Retrospective cohort study in two university hospitals
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Clinical phenotypes of Behçet's disease (BD) vary among ethnics. We chronologically analyzed clinical manifestations in 412 patients meeting the Japanese criteria for BD who visited two Yokohama City University hospitals from July 1991 to December 2007. Onset of individual symptoms in each patient was examined.
A single initial symptom appeared earlier than any other manifestations in 78% of patients. Duration from the initial symptom to diagnosis was 8.6 ± 10.1 years. Oral ulcer, the most common initial manifestation, preceded the diagnosis by 7.5 ± 10.2 years. Genital ulcer, eye and skin involvements appeared one or two years prior to diagnosis, whereas gastrointestinal, central nervous or vascular involvement developed lately. Frequency of eye involvement was significantly higher in patients with neurological lesions, but significantly lower in those with gastrointestinal or vascular involvement. However, there was no particular combination of major symptoms which predicted development of the organ involvement. The ratio of complete type patients having all of oral ulcer, genital ulcer, eye and skin lesions has been recently decreasing, whereas frequencies of arthritis, gastrointestinal, and vascular involvement have been increasing.
Further assessment may allow the detection of early predictors of more aggressive disease, requiring more intensive treatment.

Board No: P-152
Ref. No: 100
Topic: Paediatric

Quality of Life in Behcet's Disease
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Introduction
Behçet's disease (BD) is a multisystem inflammatory disorder which affects the mucocutaneous, ocular, cardiovascular, pulmonary, neurological, articular and gastrointestinal systems. Symptoms usually relapse and remit and may lead to temporary or permanent functional disability, which in turn may restrict an individual's lifestyle and mental well-being. Quality of life (QoL) is therefore an important outcome factor in BD.
Purpose
The authors set out to evaluate patients' perspectives on QoL.
Methods
Behçet's patients attending uveitis and/or immunology clinics were questioned. Each patient was given two questionnaires to complete; the BD-QoL (devised by Tennant et al) and a fatigue questionnaire. The BD-QoL assessed physical limitations in daily activities, dependence on others and the effect on relationships, self-esteem and self-confidence. The fatigue questionnaire centred on three areas: physical strength, lethargy and energy levels, concentration and memory, and mood and lack of interest in things.
Results
A total of 65 questionnaires were completed. Thirty patients suffered with systemic disease not affecting the eyes, and thirty-five patients had both systemic and ocular disease. The BD-QoL questionnaire was scored out of 30 and the fatigue questionnaire scored out of 46. The higher the score, the greater the negative impact of BD on QoL. The average score for all patients for QoL was 14/30 and fatigue was 24/46. Non-ocular patients scored higher on average with QoL 17/30 and fatigue 26/46 whilst ocular patients had lower scores of QoL 12/30 and fatigue 21/46.
Conclusions

Our survey demonstrates that BD has a significant impact on QoL. Perhaps surprisingly from an ophthalmic point of view, non-ocular manifestations are the predominant impactors on QoL.

Board No: P-153
Ref. No: 121
Topic: Paediatric

EVALUATION OF GALLBLADDER VOLUME AND EJECTION FRACTION IN PATIENTS WITH BEHÇET'S DISEASE

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Bacground&Aims: Gastrointestinal involvement in Behcet's disease mainly appears in mucosa, however, all patients do not have endoscopic findings although they have upper gastrointestinal sypmtoms. It has been shown that some of these upper gastrointestinal symptoms may be the result of esophageal motor dysfunction. In addition to the abnormal motor activity of the esophagus, Behçet's disease may be associated with extraesophageal dysmotility involving gallbladder. The aim of this study was to investigate possible dysfunction of the gallbladder in patients with Behçet's disease.
Methods: Thirty patients with Behçet's disease (22 women, mean age: 41.9 ± 9.6 years) and 30 healthy volunteers (22 women, mean age: 42.4±10.7 years) were included into the study. All subjects were investigated after 12 hours of fasting and 30 minutes after a standard test meal. Fasting and postprandial gallbladder volumes (PPGV) were studied ultrasonographically and ejection fraction of the gallbladder (GEF) was calculated.
Results: The mean fasting gallbladder volume (FGV) in Behçet's disease was lower than the control group (22.1±9.91 vs. 30.31±11.29 cm3, p=0.033). The mean PPGV in Behçet's disease group was lower than the control group (12.12±5.42 vs. 14.02±8.20 cm3, p=0.295). The GEF (%) of Behçet's disease group was lower than that of the control group (45.86±17.28 vs. 54.84±14.47, p=0.04).
Conclusion: The results of this study indicated that Behçet's disease patients have smaller fasting gallbladder volumes and ejection fraction compared to healthy controls. Altered motility of gallbladder may help to explain a part of unexplained upper gastrointestinal symptoms in patients with Behçet's disease.

Board No: P-154
Ref. No: 153
Topic: Paediatric

PED-BD: An international cohort study on pediatric Behçet's disease.One year data of 110 patients

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Aim: is to set-up an international cohort of patients suspected with BD and selected on homogenous criteria. The cohort is aimed at defining an algorithm for definition of the disease in children, reflecting the natural history.
Methods: Centers specializing in pediatric BD have been called to collaborate documenting their patients into a database. An international expert committee has defined inclusion criteria. Data are updated every year and patient's files are classified by the expert committee into 3 groups: definite, probable and not BD. Statistical analysis are performed at the end to compare the 3 groups.
Results: In January 2010, 110 patients (56M/54F) from 16 centres of 11 countries have been included. Mean age at first symptom: 8.1y (median 8.2). Mean age at BD suspicion: 11.8y, (median 14.4y). 38 % of them had only 1 symptom associated with OA, 31% had 2 and 31% had at least 3. 106 first visits have been done. 93% were receiving treatment: colchicine 59%, steroids 58%, and azathioprine 14%. 57 patients underwent first year visit, 36 had no new symptom, 12 had

one, and 9 had 2.
The expert committee has examined 48 files and classified 30 as definite and 18 as probable. Among our patients classified as definite, 27 (90%) fulfilled the ISG criteria,17/18 (94%) classified as probable did not meet the international criteria.
Conclusion: The expert committee has classified the majority of patients in the BD group although they presented few symptoms and this at inclusion independently from BD classification criteria.

Table1: Distribution of 110 Ped-BD patients by country and gender

Country	Female	Male	Total
France	20	17	37
Italy	8	11	19
Iran	11	7	18
Turkey	3	9	12
Morocco	3	4	7
Egypt	1	3	4
Germany	3	1	4
Saudi Arabia	1	2	3
Switzerland	2	1	3
Spain	1	1	2
Denmark	1	0	1
Total	54	56	110

Board No: P-155
Ref. No: 172
Topic: Paediatric

Personal hygiene habits of Behçet's disease patients: Can bad hygiene cause Behçet's disease?
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Several epidemiological studies have shown that autoimmune and allergic disorders are more prevalent in developed countries with good personal hygiene habits and sanitation conditions. Our aim was to test the influence of personal hygiene habits and environmental conditions on the development of Behçet's disease (BD). Consecutive Behçet's disease (n=50), multiple sclerosis (MS, n=50) and headache (HA, n=50) patients followed in our outpatient clinic were subjected to a 21 question, multiple-choice questionnaire to assess the personal hygiene habits and social/educational and living conditions. BD patients had lower educational levels and monthly income rates than MS and HA patients and were living in smaller houses with more residents. Significantly more BD patients were living in soil-based houses near the animal shelters and eating from the same plate and less BD patients had the habits of taking bath, brushing the teeth and washing the hands. As compared to the MS group, BD patients had higher rates of dealing with agriculture/cattle breeding, living in houses with poor sanitation, giving birth at home, using dried cow dung as fuel and developing parasite infections. MS patients had better personal hygiene and living conditions as compared to both BD and HA patients. Our results indicate that BD patients live in poor hygiene conditions and have low educational and income levels. These living conditions might be increasing the susceptibility of exposure to infectious or non-infections agents that might trigger the immune responses ultimately causing BD.

Board No: P-156
Ref. No: 174
Topic: Paediatric

Update of epidemiologic and clinical data of Adamantiades-Behçet's Disease in Germany (2010)
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The German Registry of Adamantiades-Behçet's Disease (ABD) is a registered charity, founded 1990 and providing epidemiologic data for ABD in permanent residents of Germany. The current prevalence of ABD based on this data is 0.88:100,000. of the 421 male and 304 female ABD patients, 287 are of German origin (39.6%), 317 of Turkish origin (43.7%), and 12 of Italian (1.7%), 10 of Greek (1.4%), and 9 of Lebanese origin (1.2%). Another 83 patients originate from 27 other countries. The frequencies of major clinical manifestations are: oral ulcers 98.5%, skin lesions 74.4%, genital ulcers 63.8%, arthritis 52.1%, ocular manifestations 51.6%, and pos. pathergy test 30.5%. Severe ocular involvement is significantly associated with HLA-B5 ($p<0.001$) and male gender ($p=0.001$). Oral ulcers were with 83.8% the most common onset, genital ulcers with 40.9% the most frequent second sign. Among skin lesions, papulopustules could be detected in 58.5%, erythema nodosum-type lesions in 37.5%, pyoderma in 11.8%, thrombophlebitis in 11.3%, and skin ulcers in 10.8%. Verified CNS involvement was diagnosed in 11.8%, gastrointestinal involvement in 11.1%, prostatitis/ epididymitis in 10.5%, lung and cardiac manifestation in 3.5% and 2.7%, resp., renal vasculitis in 1.8%. Severe courses occurred in 10.9%, fatal outcome in 1.2%, n=7. Median age of onset is 28 years (range 0 to 72 years). The complete clinical picture developed in 5 months (median). Interval between onset and diagnosis is 50 months (median) being significantly longer than the duration of development of the complete clinical picture ($p<0.001$).

Board No: P-157
Ref. No: 261
Topic: Paediatric

Pseudotumoral Neurobehçet disease mimicking brain tumour
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Introduction: Neurological involvement occurs in 10-20% of patients with Behçet's disease (BD). Neurological pseudotumoral presentation is rarely reported and brain biopsies are few described. We report two cases of neurological pseudotumoral presentation of BD in patients not previously diagnosed with BD. Patients: First: A 63 years old man was evaluated for hemiparesis, gait disturbance and change of mood during two weeks. He had genital and oral ulcerations. Brain MRI showed a thalamic mass. CSF and thoracoabdominal scan were normal.. He was diagnosed with BD and after being treated with corticosteroids and azathioprine symptoms disappeared. Three months later brain MRI was normal. After 10 years of follow up and treatment with AZA the patient remained asymptomatic. Second: A 35 years old woman was admitted because a seizure. MRI revealed a mass lesion at the left frontoparietal area. After 20 days of treatment with dexametasone MRI showed a dramatically reduction of the lesion. CSF and thoracoabdominal scan were normal. Three months later, patient developed mouth and genital ulcers. Neuro-BD was diagnosed and cyclophosphamide was started. One month after finishing treatment she had a new seizure. MRI showed an increase of the lesion. A brain biopsy revealed a mild granulomatous vasculitis. Due to the poor response to cyclophosphamide and azathioprine, infliximab was started. There has been a decrease of lesion size in MRI and no other epileptic seizures have appeared. Conclusion: Brain involvement in BD can mimic a brain tumour and must be considered in the differential diagnosis of brain mass lesions.

Board No: P-158
Ref. No: 64
Topic: Genetics

Familial aggregation of Behcet disease in a group of Iranian patients
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Introduction: Familial occurrence of Behcet disease (BD), although well known, is not common. The aim of this study was to determine the prevalence of BD in first-degree relatives of patients with BD.
Methods: All available first-degree relatives (parents, children and siblings) of 100 consecutive BD patients attending Behcet's disease Clinic were evaluated for the presence of BD. First, the major signs of BD were detected by a screening questionnaire, and if suspected they were visited by rheumatologist and pathergy test performed. The diagnosis of BD was accepted according to ICBD criteria. The prevalence of the disease in this group was compared to the BD prevalence in Iran by one-sample t-test, and the recurrence risk ratio was calculated.
Results: The 100 probands had 901 relatives. Seventy of them were deceased and 12 were not available, thus the remaining 819 (90.9%) participated in this study. There were 485 siblings, 139 parents and 195 children. We have found 9 cases of BD (1.1%). Regarding the prevalence of 0.08% of BD in Iranian normal population, the recurrence risk ratio calculated was 13.74 in the first-degree relatives of index patients. These cases were 3 in parents (2.2%), 6 in siblings (1.2%) and none in children. There were no significant differences in disease prevalence comparing parents with siblings (p=0.42) or children (p=0.07), and comparing siblings with children (p= 0.13).
Conclusion: The recurrence risk ratio for BD is high in first-degree relatives of patients with BD. This may be in favor of strong genetic predisposition in the pathogenesis of BD.

Board No: P-159
Ref. No: 66
Topic: Genetics

TNF-α gene polymorphisms in Iranian Azeri Turkish patients
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Genetic factors that predispose individuals to Behcet's disease (BD) are considered to play an important role in the development of the disease. The serum level of tumor necrosis factor (TNF) is elevated in patients with BD, and a dramatic response to anti- TNF-α antibody treatment further supports the role of TNF in BD. We investigated the distribution of TNF-α promoter -1031T/C and -308G/A polymorphisms in 53 BD patients of Iranian Azeri Turks and 79 matched healthy controls, via the PCR–RFLP technique. The frequency of the TNF-α -1031C allele was significantly higher in Behcet's patients than in healthy controls ($p < 0.0001$, OR = 3.08; 95% CI = 1.73–5.47), whereas the frequency of the TNF-α -308A allele was similar in the two compared groups. The frequency of CG haplotype was significantly higher ($p < 0.0001$, OR = 3.42; 95% CI = 1.89–6.18), and that of the TA haplotype was significantly lower in BD patients than in healthy controls. These results suggest that TNF-α is a susceptibility gene for BD in patients from Iranian Azeri Turk ethnic group.

Board No: P-160
Ref. No: 68
Topic: Genetics

Association of TLR4 polymorphisms with Behcet's disease in Japanese and Korean populations
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Objectives: We recently reported that the single nucleotide polymorphisms (SNPs) of the Toll-like receptor 4 gene (TLR4) on chromosome 9 are associated with Behcet's disease (BD) among Japanese and Korean populations. One of the SNPs, rs7037117, is significantly associated with Japanese, and a haplotype of TLR4 is increased in Korean patients. In the present study, we analyzed TLR4 polymorphisms in Japanese and Korean Behcet's patients regardless of their nationality.
Methods: We enrolled 319 patients (200 Japanese and 119 Korean) and 243 healthy controls (102 Japanese and 141 Korean) at the Hospitals of Yokohama City University, Hokkaido University, and Seoul National University. 49.2% of the patients were HLA-B*51 positive. Nine SNPs in TLR4 were analyzed by TaqMan 5' exonuclease assay or direct sequencing method.
Results: Any SNPs showed no significance between whole cohort of Behcet's and healthy subjects. However, there were significant associations between HLA-B*51 positive patients and healthy people. The homozygous for major allele, rs11536889 was significantly increased in HLA-B*51 positive BD patients (64.3%) compared with controls (53.5%) [odds ratio (OR)=1.57, 95%confidence interval (CI) =1.04-7.83, p=0.032]. Also, the allele frequency of rs7045953 was significantly increased in HLA-B*51 positive patients (12.7%) compared with healthy controls (7.6%) [OR=1.77, 95%CI=1.11-8.29, p=0.016].
Conclusions: There were significant differences of TLR4 polymorphisms between HLA-B*51 positive BD and healthy people in East Asian populations. It suggests that signal pathway through Toll-like receptor 4 may be an important role in the development of BD. Further studies are required to find the cause of the dissociation between HLA-B*51 positive and negative patients.

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Ref. No: 93
Topic: Genetics

FAMILIAL BEHÇET'S DISEASE

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Introduction :
Behçet's disease (BD) is an inflammatory disease of unknown origin presenting with recurrent oral and genital ulcers and ocular involvement. Familial disease is uncommon. A positive family history is observed in up to 12 % and a sibling risk ratio of 11.4-52.5 has been reported.
Objectives :
To investigate the characteristics of familial BD and review literature.
Patients and methods:
Out of 193 patients we report 22 patients from 15 families (13 men, 9 women, mean aged 28.2 years).
Results :
All patients presented with recurrent oral and genital aphthous. They also have pseudofolliculitis (n=21), positive pathergy test (n=19), arthritis or arthralgia (n=16), uveitis and or vasculitis (n=6), vascular involvement (n=8), neurological manifestation (n=4).
A history of familial BD concern brothers (n=8), sisters (n=5), cousin (n=3), uncle (n=1), aunt (n=1). Six patients have more than two patients from their family with BD. When we compared patients with or without family history, there is a difference in only cutaneous manifestations which were more frequent in the group with family history.
Conclusion :
The aetiology of BD remains unknown. Family history suggests a genetic component to the disease similar to that observed in other complex genetic disorders such as inflammatory bowel disease.

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Ref. No: 94
Topic: Genetics

CONJUGAL BEHCET'S DISEASE
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Introduction :
The occurrence of conjugal Behçet's disease (BD) is exceptional as in other inflammatory diseases. We report the case of a BD patient's wife who developed the onset of the disease within a 10 year period of marriage.
Observations:
A 37 year-old tunisian man, in whom the diagnosis of BD was confirmed at the age of 20 years, complained of dyspnoea, oral and genital persistent ulcers which resist to symptomatic treatment. Vasculitis and bilateral uveitis are confirmed and the patient was treated by pulse corticosteroid (CS) and cyclophosphamide (CP). Although immunosuppressive treatment, he was complicated with blindness. Furthermore, computed chest tomography showed a pulmonary embolism. Heparinotherapy with the immunosuppressive treatment was started with favourable evolution. In the same period, the diagnosis of BD was confirmed in his wife with retinal vasculitis, recurrent oral and genital ulcers, pseudofolliculitis and cerebral thromphlebitis. There were no consanguinity between them. Immunosuppressive agents (BCS and CP) allows favourable evolution.
Conclusion:
The aetiology of BD remains unknown. Environnemental factors may play a role in its pathogenesis. To our knowledge, this is the first reported case of conjugal BD.

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Ref. No: 124
Topic: Genetics

HLA-antigens and their importance as prognostic-marker in Adamantiades-Behçet's disease (ABD) - Is HLA-Bw4 a new prognostic marker?
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Objective: To explore correlations between genetic predisposition vs clinical manifestations and course of ABD.
Patients and methods: 590 patients (t) [344 male (m) and 246 female (f) - 227 German (G), 267 Turkish (T) and 96 patients of other ethnic origin] from the German Registry of Adamantiades-Behçet's disease. Retrospective statistic analysis of the presence of HLA-A, -B, -C-antigen vs clinical manifestations (mf). Chi-square testing and at multiple testing Bonferroni- and Benjamini-Hochberg-correction (BH) was conducted. Controls (c) were ethnically and residentialy matched.
Results: The prevalence (Pr) of HLA-B5 was t=58.8% (G=44.1%, T=73.2%). The Odds ratio (OR) was G=4.8, T=6.0 (both $p<0.005/c$). Pr of HLA-A2 was t=66.5% (G=68.9%, T=68.1%), odds ratio G:OR=2.2, T:OR=2.9 (both $p<0.005/c$). $P<0.005$ was observed among t between HLA-B5-positivity(+) vs cutaneous and ocular mf (erythema nodosum, hypopyon-iritis, retinitis and blindness). Among HLA-Bw4+ patients (ft=67.9%, G=59.3%, T=76.1%), which contains HLA-B5, $p<0.005$ independently from B5+ vs mf was observed in ocular (conjunctivitis, retinitis, blindness), arthritis, pulmonary, neurologic mf and +pathergy test. Coexisting HLA-B5 and HLA-Bw4 in Turkish males was associated with severe course of ABD (BH). Other correlations after BH: (T f Bw4+B5 vs arthritis), (G m Bw4 vs pulmonary mf), (T m Bw4+B5 vs headache and vs vascular mf). $P<0.005$ at mean age onset of disease vs HLA-Bw4 (without B5+) at T (Bw4+=24 yr vs −=32 yr), and among duration in months (ms) from onset mf to full clinical ABD: (T B5+=1 ms vs −=10.5 ms), (G B5+=0 ms vs −=12.5 ms), (G f: B5 +=0 ms vs −=6 ms), (G f: Bw4+=6 ms vs −=66 ms).
Conclusion: Known correlations between HLA-B5 and new among HLA-Bw4 vs clinical manifestations were observed. HLA-Bw4 could represent a new negative prognostic factor.

Board No: P-164
Ref. No: 130
Topic: Genetics

Occurrence of Behcet`s Disease in Iraqi Families
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" Occurrence of Behcet`s Disease in Iraqi Families"

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Abstract
Fourteen Arab,Iraqi family with at least two affected individuals were studied.All patients were diagnosed as having BD according to the criteria of O`Doffy and ISG.
The distributions of types of BD by sex,normal and affected family members,with HLA-B51(5) frequency in these families ara studied.The families,comprising 75(81.52%) patients 53 of 75 patients have complete type(32 males,and 21 females),while 22of 75 patients have incomplete type(9 males,and 13 females),HLA classI and ɪɪ was don for 67BD member,the frequency of HLA-B51(5) in those patients was 83.58%(44.78% males,and 38.8% in females)
Family pedigree and symbol used are illustrated in figures each family is labeled with chromosome 6 heplotype in the form of small letters and details of each haplotype are given below the family members,and to test hapiotype sharing between the affected family members,-sib,pair anaiysis was utilized.
Inspecting the families for affected sibs,all the 14 Iraqi families were informative for this purpose.The distribution of HLA-hapiotype in the sibs of BD families was not random,and associated with X^2y value of 7.782,which is significant(p

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Ref. No: 136
Topic: Genetics

Interleukin 6 gene Polymorphism in Tunisian Patients with Behçet's Disease.
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Introduction: Interleukin 6 (IL-6) is a multifunctional cytokine that regulates the immune, acute phase response and inflammation. A polymorphism in the 5′ flanking region of the IL-6 gene on chromosome 7 at position −174 has been reported corresponding to serum level and is associated with immune and inflammatory disorders. Increased IL6 plasma levels and enhanced IL6 mRNA expression have been found in patients with active Behçet's disease (BD). In this study, we investigated the possible association between BD and the IL6 gene promoter polymorphisms -174 G/C in a Tunisian population.
Patients and methods: It's a prospective study, including 43 patients with BD, fulfilling the ISGBD and 43 healthy controls. The detailed clinical characteristics were recorded for each patient. We compared the distribution of these polymorphisms between patients and healthy controls using polymerase chain reaction restriction fragment length-polymorphism (PCR–RFLP) analysis. Significance was evaluated using Fisher's exact and Pearson test ($p \leq 0.05$).
Results: There were 23 men and 20 women with a mean age of 40.63 years. No statically significance was found between the frequencies of IL6 gene polymorphisms -174 G/C in patients with BD and healthy subjects: 32.5 vs 16.26%, p=0,007. No significant associations were found between the genotypes or alleles of the IL6 -174G/C and epidemiological or clinical variables. The disease duration was significantly higher in patients with IL6 gene polymorphism.
Conclusion: The role of IL6 in BD is not genetically determined, but can be functionally explained. Further studies (other IL6 polymorphisms) in other ethnic groups are necessary.

Board No: P-166
Ref. No: 154
Topic: Genetics

Association between Tumor Necrosis Factor gene polymorphisms and Behçet's Disease in Tunisian Patients.
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Introduction: The etiopathogenesis of Behçet disease (BD) is not clearly established. However, immune dysfunction, characterized by activation in neutrophils and T cells and release of Th1-type pro-inflammatory cytokines such as tumor necrosis factor-α (TNF-α), has been considered as the major pathogenetic factor of the disease. The association between BD and alleles in the promoter region of TNF has been confirmed in Japanese patients with BD. The purpose of this study was to analyze the effect of TNF-α promoter polymorphisms at positions - 308 on the susceptibility, and clinical features of BD in Tunisian population.
Patients and methods: Forty three patients, fulfilling the ISGBD, were included in the study compared with matched 43 healthy controls. The TNF-α gene sequences were amplified by the polymerase chain reaction. Sequence analysis of the TNF-α gene locus, which contains promoter polymorphisms at position-308, was performed with a DNA sequencing kit on automated sequencer.
Results: There were of 23 men and 20 women. The mean age was 40.63 years. TNF-α-308 G/A heterozygous polymorphism were detected in 11 BD patients and 10 healthy subjects. No homozygous mutation was observed in BD patients or healthy subjects. The frequencies of the TNF-α-308G alleles were similar in the two groups.
The TNF-α -308 polymorphism did not show any association with clinical findings.
Conclusion: TNF-α gene polymorphism at position-308 is unlikely to play an important role in the pathogenesis and clinical features of BD.

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Board No: P-167
Ref. No: 163
Topic: Genetics

Mitochondrial genome association with Behçet's disease (BD)
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Only nuclear genes have been investigated thus far for BD risk, while the role of the mitochondrial DNA (mtDNA) has been completely neglected. In our expression profiling study of BD patients, we found an over-representation of genes linked to oxidative stress among those differentially expressed. Since mitochondrial function is required for normal vascular cell growth and function, and its dysfunction may underlie a multitude of clinical features in multifactorial and multisystemic diseases, we assessed whether mtDNA single nucleotide polymorphisms (SNPs) and haplogroups confer susceptibility to BD. Patients were selected as consecutive patients, according to ICBD criteria. 22 mtDNA SNPs sufficient for classifying our samples into the most prevalent haplogroups in our Iranian sample (West Eurasian H, V, pre-HV/HV, J, T, U, K, I and W, Eastern Eurasian M, N and R, African L, and Asian D haplogroups) were genotyped in 550 BD patients and 436 controls, and their association with BD tested individually or in combination into haplogroups, adjusting for sex and ethnicity. mtDNA SNP 709G/A in the 12S rRNA was found associated with BD (adjusted p=0.048, and unadjusted p=0.031). None of the haplogroups tested were associated with BD.
Since 12S RNA molecules participate in assemble of amino acids into functioning proteins that carry out oxidative phosphorylation inside the mitochondria, this association supports a role of oxidative stress in BD pathogenesis. mtDNA SNP 709G/A association with BD requires further validation and investigation.

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Board No: P-168
Ref. No: 187
Topic: Genetics

Contribution of the HLA-A region to genetic predisposition in Behcet's disease

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Objective: The etiology of Behçet's Disease (BD) is still uncertain, but likely both genetic and environmental factors play important roles in its development. Since the early identification of HLA-B51 as a susceptibility gene for BD, the identification of additional genetic locus/loci has clearly stalled. Our recent two genome-wide association studies (GWASs) using 23,465 microsatellite markers or 500,568 SNPs have identified the strong association between HLA-A26 and BD. The aim of this study was to clarify the contribution of HLA-A26 to genetic predisposition in BD.
Methods: We investigated the association of clinical characteristics with HLA-A26 in the Japanese BD patients according to the criteria proposed by the Japan Behçet's Disease Research Committee. Additionally, we performed a meta-analysis of HLA-A26 from several different population-based cohorts including the ones used in previous studies.
Results: HLA-A26 positive patients had significantly higher frequency of ocular involvement compared to HLA-A26 negative patients; some other lesions were also associated with HLA-A26. Association of HLA-A26 with female patients with BD was significantly strong (OR = 3.8) while it was less with male patients with BD (OR = 1.9). There were some populations showing no association with HLA-A26, however the overall meta-analysis including several different populations showed a strong contribution of HLA-A26 in the development of BD.
Conclusions: This study highlights two independent loci, HLA-B and HLA-A, as the premier genetic susceptibility locus for BD, and suggests HLA-A26 may play an important role in the development of BD.

Board No: P-169
Ref. No: 190
Topic: Genetics

Copy number variation of beta-defensin gene in Behcet's disease

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Background:
Immune dysregulation triggered by infectious agents in genetically susceptible patients has been suggested as pathogenetic mechanism for Behcet's disease (BD). Beta-defensin is a peptide which forms immunologic barrier to infectious agents. Its expression has been recently reported to be dependent on the copy number variation (CNV) in the genome. Thus, we sought to investigate the relationship between beta-defensin gene CNV and development of BD.
Methods
A total of 200 patients with BD and 200 healthy blood donors were enrolled from Seoul National University Hospital to measure beta-defensin gene copy number (CN) using paralogue ratio test (PRT). CNV was compared between patients with BD and controls. The CNV was also analyzed according to the clinical manifestations among patient with BD.
Results
The results of 166 patients and 174 controls finally passed the validity test and were used in the following analysis. The beta-defensin gene CN was normally distributed in patients and controls. Overall most samples had 4 or 5 CN (57.1%) with mean CN of 4.57± 1.28. There was no difference of beta-defensin gene CNV between patients with BD and healthy controls (4.49±1.20 vs. 4.65±1.35, p=0.25). There was no difference of the CNV in patients with BD, depending on the clinical manifestations (genital ulcer, erythema nodosum, acne-like skin lesion, uveitis or pathergy test)
Conclusion
We found no significant difference of beta-defensin gene CNV between patients with BD and healthy controls. Beta-defensin gene CNV may not contribute a major role in the pathogenesis of BD.

Board No: P-170
Ref. No: 193
Topic: Genetics

Functional Polymorphisms and Haplotypes of Human Beta Defensin 1 (DEFB1) Gene in Behçet's Disease Patients
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Functional Polymorphisms and Haplotypes of Human Beta Defensin 1 (DEFB1) Gene in Behçet's Disease Patients
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Behçet's Disease (BD) is a systemic, inflammatory disorder of unknown cause. Its ethiology is still unknown, but host genetic factors as well as microbial infections triggering autoimmune responses may have a role. Human beta defensin 1 (DEFB1) is a constitutively expressed antimicrobial peptide produced at a variety of epithelial surfaces, including the oral mucosa. Its expression can be altered by the functional single nucleotide polymorphisms (SNPs) located at the promoter region. DEFB1 polymorphisms are shown to be associated with immune disorders and susceptibility to various infectious agents. In this study we aimed to investigate the prevalence of DEFB1 promoter polymorphisms and haplotypes in 119 BD patients and 103 healthy control subjects by PCR+restriction endonuclease analysis. The results showed that -52GA ($p<0.001$) and -44GG ($p=0.005$) genotypes were more prevalent among the BD patients. When the allele frequencies were compared, -52A ($p=0.039$) and -44G ($p<0.001$) alleles were found to be more prevalent in the BD patients. For the -20G/A SNP, no significant difference was found between the groups. Haplotype analysis revelaed that GG/GG/GG haplotype (subjects carrying the GG genotype for all SNPs) was significantly more prevalent among the controls (pz0.001, OR =9.31), while GA/GC/GA (subjects carrying the GA, GC and GA genotypes for -52, -44, and -20 SNPs, respectively) haplotype was significantly more prevalent in the BD patients ($p=0.01$, OR=3.11).For the rest of the haplotypes determined, no statistically significant association was found between the groups. Our results show that, there may be a correlation between HBD-1 polymorphisms and haplotypes and BD.

Board No: P-171
Ref. No: 205
Topic: Genetics

What are the differences between Behcet's disease and Sweet's Syndrome?
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OBJECTIVES: To describe the clinical features of four Japanese patients with Sweet's syndrome (SS) and to study the relationship between SS and HLA-B loci. METHODS: Four Japanese patients (2 male; mean age: 38.8±9.3y/o) with SS visited our hospital, from 2004 to 2008, and whose HLA were measured. Stastical analyzing used Fisher's exact test. RESULTS: All patients were fulfilled the criteria of SS and all were detected having HLA-B54 ($p = 7.0 \times 10^{-6}$). Two of the all 4 patients were fulfilled the Japanese BD's criteria. One of the 2 was fulfilled the international BD's criteria with recurrente oral aphtous ulcers: ROA, erythema nodosum: EN, arthritis, and ileocecal ulcer. Although, another was not fulfilled the

international BD's criteria with uveitis and EN but absenting from ROA. The other 2 were fulfilled the international BD criteria with ROA and EN, however, not fulfilled the Japanese BD criteria. Pathological findings of skin biopsy from EN in each 4 patients typically showed an upper dermal infiltrate of mature neutrophils. All 4 patients were tried to treate with some antibiotics, but all agents were ineffective against each symptoms and findings of SS, and were prompted improvement of both symptoms and lesions as SS which was a multisystem inflammatory disorder after the initiation of treatment with systemic corticosteroids. CONCLUSIONS: The present findings supported that the references as SS were reported. Skin biopsy and analyzing HLA loci were very useful to differentiate between BD and SS. GRANT: No.

Board No: P-172
Ref. No: 241
Topic: Genetics

Replication of association in KIAA1529 gene with Behçet's disease

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Very few biological candidate genes have been so far investigated for their potential involvement in BD susceptibility in case-control association studies, and positive findings were not replicated. A recent genomewide association study for Behçet's disease (BD) demonstrated an association of KIAA1529, LOC100129342, CPVL and BASH3B genes with BD (Fei et al. 2009). Since replication in independent samples remains the gold-standard in association findings, we investigated the reported associations with BD of single nucleotide polymorphisms (SNPs) in LOC100129342, CPVL and BASH3B, and of all tagging SNPs in KIAA1529.
A total of nine SNPs were genotyped in 550 Iranian BD patients and 436 controls. Patients were selected as consecutive patients, according to ICBD criteria. We tested the association of alleles, genotypes and haplotypes with BD, unadjusted and adjusted for sex and ethnicity.
SNP rs7038496 in KIAA1529 showed a significant association with BD (p=0.040). This SNP belongs to a 20kb haplotype block (from rs7038496 to rs725229) that also shows a significant association (p=5.6E-3).
We have found a different polymorphism in KIAA1529 associated to BD that the one previously described (rs2061634). This finding further supports the involvement of KIAA1529 in BD susceptibility, but the exact marker within this genes remains to be pinpointed.

Grants from the Tehran University of Medical Sciences and Fundação para a Ciência e a Tecnologia.

Board No: P-173
Ref. No: 243
Topic: Genetics

Homozygous 3.1kb deletion in LEPREL1 intron1 protects from Behcet's disease

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Accumulating evidence has shown involvement of copy number variation (CNV) in inflammatory diseases including systemic lupus erythematosus and psoriasis. To ask whether CNV is associated with Behçet's disease (BD), we analyzed SNP intensity data from our Turkish BD genome-wide association study (1217 cases and 1278 controls). We found a common 3.1kb deletion in the first intron of LEPREL1, for which homozygous deletion appeared to protect individuals from disease development (OR=0.62, $p=6.5\times10^{-4}$). This association of homozygous LEPREL1 deletion replicated in a Japanese BD collection (384 cases and 415 controls, OR=0.64, p=0.011).
LEPREL1, a prolyl 3-hydroxylase which is also called P3H2, is involved in type IV collagen modification. This gene may have additional functions. Although the protein is expressed in lymph nodes, there has been no direct evidence linking

LEPREL1 to inflammation. We found that LEPREL1 is primarily expressed in dendritic cells. Furthermore, peripheral blood mononuclear cells from individuals homozygous for this deleted region produce less LEPREL1 and inflammatory cytokines following exposure to Toll-like receptor ligands. Additionally, stimulation of a dendritic cell line with lipopolysaccharide with or without interferon-gamma induced LEPREL1 expression. These results suggest that LEPREL1 plays an important role in dendritic cells, and has a significant effect on BD susceptibility.
Our data offer new insights into the role copy number variation plays in repressing development of BD and reveal a novel modulatory mechanism for inflammation.

Board No: P-174
Ref. No: 245
Topic: Genetics

Common variants within the UBAC2 gene are associated with increased risk of Behçet's disease
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Objectives: Using a genome-wide association scan and DNA pooling, we previously identified 5 novel genetic susceptibility loci for Behçet's disease. Herein, we fine-map the genetic effect within the UBAC2 gene and replicate this genetic association in an independent cohort of Behçet's disease patients and controls.
Methods: Two independent cohorts of Behçet's disease patients and controls from Turkey were studied. The discovery and replication cohorts included 152 patients and 172 controls, and 376 patients and 369 controls, respectively. Genotyping of 14 SNPs within and around UBAC2 was performed using TaqMan SNP genotyping assays.
Results: The genetic association between Behçet's disease and UBAC2 was established and confirmed in two independent cohorts of patients and controls (meta-analysis odds ratio= 2.05, meta-analysis P= 1.75X10-7). Haplotype analysis identified both a disease risk and a protective haplotype (P= 0.00014 and 0.0075, respectively). Using conditional haplotype analysis we provide suggestive evidence that the SNP rs7999348 within UBAC2 confers an independent genetic effect and likely explains the genetic effect in this locus.
Conclusion: We establish and validate the genetic association between UBAC2 and Behçet's disease in two independent cohorts of patients and controls. Fine mapping of this genetic effect and conditional analysis suggest that, among the genotyped SNPs, rs7999348 is the most likely causal SNP in this locus.

Board No: P-175
Ref. No: 252
Topic: Genetics

The CC chemokine receptor polymorphism CCR5delta32 is not a genetic susceptibility factor for Behçet disease in the Portuguese population.
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Background: Behçet's disease (BD) is a Th1 mediated recurrent multi-system inflammatory disorder of unknown aetiology. The CC chemokine receptor 5 (CCR5) is a chemokine receptor expressed by peripheral blood leucocytes, and is involved in the recruitment of inflammatory cells. Elevated expression of this receptor in the tissue of oral and genital ulcers in BD has been demonstrated.
A 32-basepair deletion (CCR5d32) in the coding region of CCR5 gene originates a truncated non-functional receptor with

reduced expression on the cell surface.
Aim: To investigate whether the CCR5 deletion is associated with susceptibility to BD in the Portuguese population.
Patients and Methods: A total of 91 BD patients (60 females and 31 males) and 227 ethnically-matched controls were studied. Genotyping of the CCR5d32 polymorphisms was performed using polymerase chain reaction product sizing.
Results: No significant difference was observed in the allelic frequency of CCR5delta32 between patients and controls (6.6% in BD vs. 8.4% in controls, OR=0.773; p=0.452). Gender effects were not seen. Also, stratification for the presence of HLA-B*51 did not reveal any differences.
Discussion: In agreement with previous reports (Yang, 2004 and Mojtahedi, 2006), our results indicate that, despite the role of chemokines and their receptors in the activation of cellular chemotaxis in the course of inflammation, the CCR5d32 polymorphism is unlikely to contribute to susceptibility to BD. This may be explained by the known functional redundancy of this signalling system.

Board No: P-176
Ref. No: 257
Topic: Genetics

Toll-like receptor 3 and 8 genes polymorphisms in Tunisian patients with Behçet's disease.
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Background:
TLRs 3 and 8 participate in the generation of protective immunity against some viral infections. As virus infections are also considered to be aetiopathogenically associated with BD any abnormalities affecting the function of these TLRs could explain the aberrant immune response in BD.
Objectives:
To investigate possible associations between TLR 3 gene and TLR8 gene polymorphisms and susceptibility to BD.
Patients and methods:
100 BD patients and 152 healthy controls were genotyped for SNPs for TLR3 gene (-7 C/A and 1377 C/T) and TLR8 gene (-129 C/G) using PCR-RFLP.
Results:
Distribution of the different allele and genotype frequencies didn't differ significantly between BD patients and controls (Table 1).
Meningitis was significantly more frequent in patients with the (–7 CA/AA) mutant allele of the TLR3 gene than those with the wild gene.
No other significant correlation was observed between TLR3 and TLR8 polymorphisms and any BD clinical manifestations or disease severity.
HLA B51 was more frequent in patients with TLR3 -7 C/C and TLR8 -129 G/G, but the difference was not statistically significant. HLA B51 frequency was similar in patients with and without SNP 1377 C/T
Comments:
Our data suggest that in Tunisian patients, the TLR 3 and 8 gene polymorphisms are not associated with the susceptibility to BD, neither its major clinical manifestations nor its severity. This is could be explained by the fact that TLR3 and 8 recognize RNA viruses while DNA virus (e.g. HSV) is mostly implicated in BD pathogenesis.

		BDn=100	Controlsn=1	p- value
SNP -7 C/AGenotypes	C/CC/AA/A	60/100 (60%)36/100 (36%)4/100 (4%)	95/139 (68.3%)36/139 (25.9%)8/139 (5.8%)	0.22
SNP -7 C/ACarriage rate	C/C C/A+A/A	60/100 (60%) 40/100 (40%)	95/139 (68.3%) 44/139 (31.7%)	0.18

SNP 1377 C/TGenotypes	C/CC/TT/T	45/97 (46.4%)43/97 (44.3%)9/97 (9.3%)	55/139 (39.6%)68/139 (48.9%)16/139 (11.5%)	0.56
SNP 1377 C/TCarriage rate	C/C C/T+T/T	45/97 (46.4%) 52/97 (53.6%)	55/139 (39.6%) 84/139 (60.4%)	0.29
SNP -129 C/G	G/GG/C	48/98 (49%)50/98 (51%)	75/152 (44.4%)94/152 (55.6%)	0.21

Board No: P-177
Ref. No: 270
Topic: Genetics

Expression and association analyses implicates the ErbB signalling pathway in Behçet's disease (BD)
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To better understand the pathogenesis of BD, we applied an integrative approach that combines expression profiling analysis with association studies.
We first compared the gene expression profiles in PBMCs of 15 patients and 14 matched controls using Affymetrix microarrays. We identified 508 differentially expressed genes that allow a good separation between cases and controls. Pathway analysis revealed that several genes in the ErbB signalling pathway were significantly differentially expressed. The ErbB family of receptor tyrosine kinases couples binding of extracellular growth factor ligands to intracellular signalling pathways regulating diverse biologic responses, including proliferation, differentiation, cell motility and survival, and vascular permeability.
Three differentially expressed genes (EREG, AREG, and NRG1) belonging to the epidermal growth factor family of ligands for ErbBs were tested for association with BD. Genotyping of 43 tagging single nucleotide polymorphisms (SNPs) was done in 550 Iranian BD patients and 436 controls. Patients were selected as consecutive patients, according to ICBD criteria. Unadjusted and adjusted (sex and ethnicity) tests of association were performed.
We found an association ($p<0.05$) in all loci studied: two SNPs in EREG, one SNP in the EREG-AREG intergenic region, and five SNPs in NRG1.
These association findings support a role for the ErbB signalling pathway in BD pathogenesis that warrants further investigation. This study highlights the importance of using integrative approaches in dissecting the genetic architecture of complex diseases.

Grants from the Fundação Calouste Gulbenkian, Fundação para a Ciência e a Tecnologia, Instituto do Emprego e Formação Profissional, and Tehran University of Medical Sciences.

Board No: P-178
Ref. No: 272
Topic: Genetics

V617F Jak2 Mutations Are Not Associated With Thrombosis Among Patients With Behcet's Syndrome
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OBJECTIVE: V617F Jak2 mutations are associated with an increased risk of thrombosis both on the arterial and the venous side among patients with chronic myeloproliferative diseases. The exact mechanism by which this mutation causes thrombosis is not clear. Thrombosis is a well known feature of Behcet's syndrome (BS) and is thought to be caused by inflammation of the vessel wall rather than the presence of thrombophilic factors. In this cross sectional survey we aimed to determine whether Jak2 mutations play a role in thrombosis of Behcet's syndrome patients.

METHODS: 62 BS patients with active or chronic arterial and/or venous thrombosis, 114 BS patients without thrombosis, 107 patients with polycytemia vera and 104 healthy controls were included. DNA was isolated from venous blood samples and the presence of V617F Jak2 mutations was evaluated using PCR.

RESULTS: V617F Jak2 mutation was not detected among any of the BS patients, with or without thrombosis, and healthy controls while the mutation was present among 56/74 (79%) of patients with polycytemia vera (PV).
CONCLUSION: V617F Jak2 mutation is not associated with thrombosis in BS patients. Further studies with other inflammatory and non-inflammatory conditions associated with thrombosis are needed to determine whether V617F Jak2 is a mutation specific to chronic myeloproliferative diseases which is related to clonal progenitor cell proliferation.

Board No: P-179
Ref. No: 273
Topic: Genetics

THE PAPULOPUSTULAR LESION/ARTHRITIS CLUSTER OF BEHÇET'S SYNDROME IS INDEPENDENTLY FAMILIAL
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Objective: The presence of symptom clusters, as we have previously shown suggests that pathogenesis of BS may involve different mechanisms. Increases in frequency of such clusters in familial BS cases would back up the notion that some pathogenetic mechanisms may have a heritable component. We aimed to compare the frequency of symptom clusters between familial and non-familial cases of BS.
Methods: We identified 380 patients with BS with a first and/or second degree relative among the 6031 patient charts reviewed. 186/380 had attended the clinic within the previous 3 months (Group F). Seventeen first-degree relative pairs were identified in group F. From the same initial pool of 6031, 500 were randomly selected. 221/500 who had attended within the previous 3 months and had no family formed Group NF. We constructed 110 random, unrelated pairs among Group NF. Data were analyzed using factor analysis.
Results: Frequency of a family history of BS was 6.8%. Clusters we identified were similar to those previously reported from our unit. The frequency of the "papulopustular lesions and joint involvement" symptom cluster was significantly higher in group F compared to group NF (39.2% vs. 21.5%, $p<0.001$). Furthermore, this cluster was shared in 5/17 pairs (29%) from group F and only in 5/110 pairs (4.5%) from group NF ($p=0.004$; OR=8.75, 95%CI2.2-34.6).
Conclusions: The "papulopustular lesions and arthritis" cluster in BS appears to cluster in familial BS as well. This further supports the notion that the pathogenesis of BS may entail several distinct mechanisms producing separate phenotype clusters.

Board No: P-180
Ref. No: 282
Topic: Genetics

Autoantibody Responses Against PINK1 and SWAP70 Antigens In Behçet's Disease
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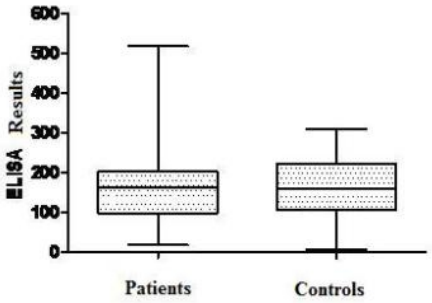
Aim: The etiopathology of Behçet's Disease (BD) remains unclear. In individuals who are genetically susceptible, immunological abnormalities which are triggered by microbial pathogens are thought to have an important part in pathogenesis of BD. Autoantibody responses against PINK1 and SWAP70 antigens was identified in sera of BD patients by our previous studies done by SEREX method. In this study we aimed to determine antibody responses to full length expressed and purified PINK1 and SWAP70 antigens in BD and healthy sera by ELISA.
Method: PINK1 and SWAP70 genes were cloned in E.coli M15 cells using pQE-30 vector for his-tagged protein expression

and proteins were purified by Ni-NTA in a metal-affinity chromatography matrix. PINK1 and SWAP70 proteins were analysed by SDS-PAGE. ELISA method was performed in BD and healthy control sera by using purified PINK1 and SWAP70 proteins as antigens.
Results and Discussion: Antibody response to SWAP70 antigen was not significant (p= 0,9293) between the groups of BD patients and healthy controls. When the cutoff value was estimated as mean±2SD there was found a significancy when comparing to controls (p= 0,014). PINK1 seroreactivity could not be found significant in BD and control groups by ELISA compared to previous studies done by SEREX. This is the first study evaluating antibody responses against PINK1 and SWAP70 antigens in BD sera by ELISA. SWAP70 seroreactivity was found significant between BD and healthy sera but further studies should be done in large BD populations.

	Patients (n)	Controls (n)	P value
Seropositivity (n)	7	0	0.0140*
Seronegativity (n)	93	100	
Total	100	100	

Graph of ELISA Results



ELISA results are higher in BD patients than controls.

In this study plasma ADMA concentration was significantly higher in BD patients and associated with neurological manifestations. In our best knowledge, such results were not reported in other ethnic groups.

Board No: P-181
Ref. No: 286
Topic: Genetics

HLA-B27-associated HLA factor with BD patient in Japanese
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[Background]
Behcet's disease(BD) is systemic inflammatory disease with uveitis, genital ulcer, oral aphtha and skin lesions such as erythema nodosum and acne. Human leukocyte antigen (HLA)-B51 and A26 are well-known genetic factors associated with BD. But all patients with BD do not have those.
Furthermore, it was reported that HLA-B27-associated HLA factor was related with reactive arthritis and ankylosing spondylitis. There are uveitis and arthritis in one of symptoms with reactive arthritis and ankylosing spondylitis.
[Objectives]
it was intended to survey HLA typing to assist a diagnosis except them.
[Methods]
Seventy-one Japanese patients with suspected BD and forty-six Japanese patients fulfilling the criteria of 1990 for BD were recruited. HLA-A and -B typing were done by a PCR-SSOP(reverse sequence specific oligonucleotide)-Luminex method.
[Results]
Among the BD patients, 14 were male and 32 were female. HLA-B51-positive patients were 30.4% and HLA-A26-positive patients were 19.6% of BD patients. 78% except those were patients with positive HLA-B27 associated HLA factor. Only four patients were negative HLA-B51, A26 and B27 associated HLA factor.
[Conclusions]
In Japanese, approximately 91% of BD patients are related to HLA-B51, A26 and B27 associated HLA factor. HLA-B27 associated HLA factor-positive patients are frequent of arthritis. The possibility that reactive arthritis and early ankylosing spondylitis without the spondylitis are included into BD patient is suggested.

Board No: P-182
Ref. No: 288
Topic: Genetics

Endothelial nitric oxide synthase gene polymorphisms in Behçet's disease
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Introduction:
Endothelial nitric oxide synthase (eNOS) dysfunction leads to reduced levels of nitric oxide (NO) which have been implicated in the development of the endothelial abnormalities occurring in Behçet's disease (BD); therefore eNOS gene might be a candidate gene for BD.
This study investigated the association of the eNOS gene polymorphisms with BD.

Patients and methods:
Eighty two unrelated Tunisian BD patients and 159 healthy controls were genotyped by PCR for two single-nucleotide polymorphisms: 894 G/T in exon 7 (Glu298Asp) and -786 T/C in the promoter region. A variable number of tandem repeats (VNTR) polymorphism in intron 4 was also investigated.

Results:
Differences in prevalence of the Glu/Glu298, Glu/Asp298 and Asp/Asp298 genotypes between BD patients and controls were statistically significant (P= 0.032). The 298Glu allele may play a protective role in Tunisian individuals with BD (odds ratio=0.5, P=0.014).
The -786 promoter region polymorphism genotypes was found to be significantly different between BD cases and controls (P<0.001). The CC genotype was associated to BD (odds ratio=2.37, P=0.005). The -786 T allele have a protective role in Tunisian individuals (P<0.001, OR=0.17), the presence of T allele decrease the risk to develop the disease by 5.35. There is no association between the -786 T/C and the Asp298 polymorphisms and specific clinical manifestations of BD. The VNTR polymorphism was not associated to BD.

Conclusion:
The -786 T/C polymorphism of the eNOS gene was associated to BD susceptibility among our Tunisian patients.

Board No: P-183
Ref. No: 289
Topic: Genetics

Apolipoprotein E polymorphisms in Portuguese patients with Behçet disease.
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Background: Behçet's disease (BD) is a multisystem disorder characterized by a relapsing inflammatory process of unknown aetiology. Evidence of immune dysfunction and association with viral and bacterial agents, such as herpes simplex virus, have been reported. Recently, a growing body of evidence has highlighted the non-lipid metabolism related properties of apolipoprotein E (apoE), including suppression of T cell proliferation, regulation of macrophage function, facilitation of lipid antigen presentation by CD1 molecules to natural killer T cells, and its role in the modulation of inflammation and oxidative stress.
Aim: The purpose of this study was to investigate a potential association between ApoE genotype and susceptibility for BD in Portuguese patients.
Patients and Methods: A total of 87 BD patients from the Centro Hospitalar do Porto/Hospital de Santo António and 242 ethnically-matched controls were studied. Genotyping of APOE was performed using a Polimerase chain reaction restriction fragment–length polymorphism (PCR-RFLP) assay.
Results: The frequency ApoE e4 allele was significantly higher in patients than in controls (13.8% vs. 6.8%, p=0.007, OR=2.19 (1.25-3.82).
Discussion: ApoE e4 seems to influence disease susceptibility in this group of patients. To our knowledge this is only the second published study addressing this issue. A previous report by Tursen and colleagues has not found any evidence of association between ApoE genotypes and BD in Turkish patients. Further studies in different ethnic populations would be helpful to clarify if these polymorphisms do indeed contribute to the susceptibility for this disease.

Board No: P-184
Ref. No: 299
Topic: Regional

Influence of HLA-B51 on clinical expression and severity of Behcet's disease in Tunisian patients.
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Objective:
To investigate the association of HLA-B51 with demographic and clinical manifestations as well as the severity of Behcet's disease (BD).
Patients and methods
HLA-B51 allele typing was determined in 177 Tunisian BD patients and 125 Tunisian healthy volunteers by either microlymphocytotoxicity test or an allele-specific PCR. All BD patients fulfilled the ISG diagnostic criteria. As only genomic DNA samples from healthy controls were available, HLA-B51 typing of both groups was performed using.
Demographic data, frequency of clinical manifestations and disease severity were compared between BD patients with and without positive HLAB51.
A severity disease score was calculated for each patient according to the method presented par Krause I (in Rheumatology 1999;38: 457-62)
Data were recorded and analyzed using SPSS 11.0.
Results
Frequency of HLAB 51 was significantly higher in BD patients (84/177 = 47.5%) than in controls (26/125 = 20.8%); $p < 0.001$
Comparison of demographic data and frequency of clinical manifestations between patients with and without positive

HLAB51 is presented in table 1.
Positive pathergy test, Retinal vasculitis, intestinal involvement were significantly more frequent in HLA B51(+) patients; While frequency of arterial aneurysms and neurological manifestations (mostly the parenchymal involvement) were significantly and clearly higher in HLA B51(-) patients

Conclusion

It seems that the presence of HLA B51 allele in Tunisian Patients predisposes to a particular form of BD that's less severe and with clearly less neurological (parenchymal) involvement and arterial aneurysm lesions. Does HLA B51 allele protect against the most serious form of BD in Tunisian patients?

	HLAB51+	HLAB51-	p
Male/female	53/31	60/34	NS
Age at BD onset (y)	29.85	28.79	NS
Diagnostic delay (y)	5.11	6.52	NS
Genital ulcers (%)	78.6	78.7	NS
Erythema nodosum (%)	20.2	17	NS
Pseudofolliculitis (%)	75	75.5	NS
Positive pathergy test (%)	71.2	50	0.011
Arthritis/Arthralgia (%)	50	45.2	NS
Ocular involvement (%)	50	36.2	0.06
Uveitis (%)	45.2	35.1	NS
Retinal vasculitis (%)	31	18.1	0.045
Neurological involvement (%)	16.7	29.8	0.04
Parenchymal (%)	9.5	24.5	0.009
Non parenchymal (%)	3.6	11.7	0.044
Vascular involvement (%)	29.8	33	NS
Deep vein thrombosis (%)	26.2	28.7	NS
Arterial aneurysms (%)	1.2	10.6	0.009
Arterial thrombosis (%)	1.2	4.3	NS
Intestinal involvement (%)	0	5.3	0.06
Severity score > 2	20.2	43.6	0.001

Board No: P-185
Ref. No: 315
Topic: Genetics

HIGH FREQUENCY OF HLA B*5101 AND *B5108 IN ITALIAN PATIENTS WITH BEHCET'S DISEASE

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Behçet's disease (BD) is strongly associated with the human leukocyte antigen (HLA) B51 in several different ethnic groups. Previous studies in Mediterranean area, including Spain, Greek and Italy, showed a strong association of BD with B*5108 as well as B*5101. However, so far only a few number of alleles has been investigated in small series. All consecutive patients with BD seen at Rheumatology Department of Lucania were subtyped using an Olerup SSP HLA DNA typing kit (Olerup SSP) with amplification patterns of 52 alleles, ranging from B*5101 to B*5152 alleles, in order to investigate the relationship between B51 subtypes and susceptibility to BD. In this study 152 patients with BD and 320 healthy Italian volunteers were enrolled. The frequency of the B51 antigen was found to be higher in the patient group as compared with the control group (64.5% vs 17%; $p<0.0001$). In the genotyping of B51 alleles, 76 out of 98 (77.6%) B51-positive patients possessed B*5101, 18 (18.4%) carried B*5108, 2 (2.0%) the B*5102 allele, 1 (1.0%) the B*5105 and the remaining 1 (1.0%) the B*5107 allele. Forty-nine out of 54 B51-positive normal control carried B*5101 (90.7%), 3 (5.5%) the B*5108, 1 (1.9%) carried the B*5107 allele and another 1 (1.9%) the B*5109. Significant differences were found for B*5101 and B*5108 alleles.
Our data obtained from a large series confirm the high prevalence of HLA B*5101 and B*5108 in Italian patients with BD underlying their role in the disease pathogenesis.

Board No: P-186
Ref. No: 317
Topic: Genetics

Correlation between clinical features and HLA-B51 subtypes in a large Italian series of patients with Behçet's disease

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Objective: To investigate the relationship between clinical features of Behçet's disease (BD) patients and HLA-B51 status or its subtypes.
Methods: HLA-B51 typing was performed by nested PCR-SSP in consecutive patients with BD seen at Rheumatology Department of Lucania. B51 positive patients were subtyped using an Olerup SSP HLA DNA typing kit (Olerup SSP) with amplification patterns of 52 alleles, ranging from B*5101 to B*5152 alleles. Medical records were retrospectively reviewed for demographic information and for all relevant clinical manifestations that had developed since the onset of BD. Analysis of the association of B51 subtypes with manifestations of BD was carried out.
Results: Among the recruited 152 BD patients (88 M, 64 F; mean age 41.5±12.9 yrs; mean disease duration 18.8±10.7 yrs), 98 (64.5%) were B51 positive. The subtypes found in the B51 positive patients were B*5101 (77.6%), B*5102 (2.0%), B*5105 (1.0%), B*5107 (1.0%), and B*5108 (18.4%).
B*5108 positive patients showed a shorter disease duration and more frequent constitutional symptoms (fever, fatigue). No significant difference was found between B51 positive and negative groups or among B51 subtypes groups for other clinical features.
Conclusions: Some clinical features of BD may be correlated with HLA-B51 status and its polymorphism.