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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7107

AB0668 TREATMENT OF THROMBOTIC EVENTS IN BEHÇET DISEASE: A SYSTEMATIC LITERATURE REVIEW

J. Janta, R.D. Gonzalez Benitez, B. Serrano Benavente, I. Monteagudo Saez. *Rheumatology, Gregorio Marañón General Hospital, Madrid, Spain*

Background: Behçet's disease (BD) is a systemic disease which etiopathogenesis is largely unknown. It is characterised by a wide variety of clinical manifestations. Venous disorder is a serious manifestation being potentially life-threatening. There is little evidence on the management of the venous complications in BD.

Objectives: To perform a systematic literature review on the treatment used in venous thrombotic events in BD.

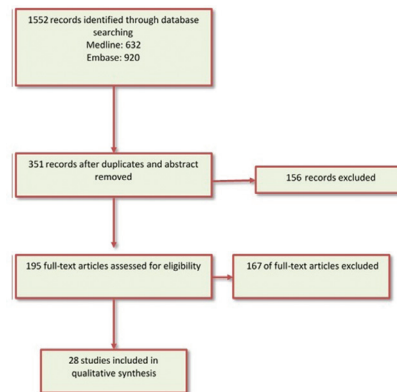
Methods: The objective was reformulated according to the PICO approach. Several synonyms for the main components (i.e. Behçet, thrombosis, treatment) were used. Search limits were applied for humans. The literature search was performed in Medline and Embase from databases inception to 1st November 2017. Only articles in English and Latin languages were retained. We excluded abstracts, reviews and letters. From the selected studies, data about the venous involvement and treatments were retired using a predefined data collection form.

Abstract AB0668 – Table 1

Treatment	Nº articles (%)
Anticoagulant	22 (78.6)
Antiplatelet	9 (33.3)
Corticosteroids	22 (78.6)
Immunosuppressive	25 (89.3)
Cyclophosphamide	16 (57.1)
Azathioprine	16 (57.1)
Cyclosporine A	5 (17.9)
Mycophenolate mofetil	2 (7.1)
Interferon alpha	2 (7.1)
Methorexate	4 (14.3)
Hydroxychloroquine	1 (3.6)
Anti-TNF alpha	4 (14.3)
Colchicine	11 (39.3)
Thalidomide	1 (3.6)
Dapsone	1 (3.6)
Fibrinolytic	3 (10.7)
Surgery	7 (25.9)

Results: The literature search resulted in 1552 articles, of which 632 were captured in Medline and 920 in Embase. Figure 1 shows the study flow-chart for article selection. The main reasons for article exclusion after full-text review were the lack venous involvement and the lack of explanation of venous involvement treatment. 28 articles reporting 1904 patients were included in qualitative analysis. The mean (range; SD) duration time between the disease onset and the vascular onset was evaluated in 15 articles and was 4.9 (1.2–9.3; 2.7) years. Superficial thrombosis was evaluated in 6 (21.4%) articles, profound thrombosis in 19 (67.9%) articles, cerebral in 7 (25%), inferior or superior cava vein in 15 (53.6%) and Budd-Chiari syndrome in 8 (28.6%) articles. Table 1 shows the treatments described in the selected articles. Treatment response was evaluated in 20 (71.4%) articles; in 7 of these treatments response was evaluated in a subjective way. In total, 52 (2.7%) deaths were reported in relation to BD. In 319 (16.7%) patients, partial efficacy or recurrence of thrombosis was reported. Considering the heterogeneity of the reported data and the variability in the measures of treatment response, predictors of mortality risk cannot be analysed. However, in the reviewed articles, a higher mortality rate was observed in patients with hepatic involvement due to Budd-Chiari syndrome. We have also observed a higher risk for the development of venous thrombosis in patients with patergia phenomenon and male sex. Two studies suggested that immunosuppressive treatment

concomitant with anticoagulant treatment is associated with a lower risk of thrombosis relapse compared with anticoagulant treatment alone.



Abstract AB0668 – Figure 1

Conclusions: There is a great variability in the treatment of venous thrombosis related to Behçet's disease. Budd-Chiari syndrome seems to be related to a worse prognosis of the disease.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5733

AB0669 MAINTENANCE TREATMENT WITH ADALIMUMAB IN REFRACTORY UVEITIS DUE TO BEHÇET'S DISEASE: OPTIMISED VS NON-OPTIMISED GROUP

J.L. Martín-Varillas¹, V. Calvo-Río¹, E. Beltrán², J. Sánchez³, M. Mesquida⁴, A. Adán⁴, M.V. Hernández⁴, M. Hernández², E. Valls⁵, L. Martínez⁵, A. Sellas⁶, M. Cordero⁷, M. Díaz⁸, R. Gallego⁸, D. Salom⁹, N. Ortego¹⁰, J.L. García¹⁰, J. L. Callejas¹⁰, J.M. Herreras¹¹, A.M. García¹², O. Maiz¹³, A. Blanco¹³, I. Torre¹⁴, D. Díaz¹⁵, E. Pato¹⁵, E. Aurrecochea¹⁶, M.A. Caracul¹⁷, F. Gamero¹⁸, E. Minguez¹⁹, C. Carrasco²⁰, A. Olive²¹, J. Vázquez²², O. Ruiz²³, J. Manero²³, S. Muñoz²⁴, M. Gandía²⁵, E. Rubio²⁶, F.J. Toyos²⁷, F.J. López²⁸, J.M. Nolla²⁹, M. Revenga³⁰, C. González-Vela¹, J. Loricera¹, B. Atienza-Mateo¹, R. Demetrio-Pablo¹, J.L. Hernández¹, M.A. González-Gay¹, R. Blanco¹. ¹Rheumatology and Ophthalmology, HUMV, IDIVAL, Santander; ²Rheumatology and Ophthalmology, H Valencia, Valencia; ³Rheumatology, H Valme, Sevilla; ⁴Rheumatology and Ophthalmology, H Clinic, Barcelona; ⁵Rheumatology and Ophthalmology, H Peset, Valencia; ⁶Rheumatology, H Vall d'Hebron, Barcelona; ⁷Ophthalmology, H León, León; ⁸Ophthalmology; ⁹Ophthalmology, H La Fe, Valencia; ¹⁰Autoimmune Diseases, H San Cecilio, Granada; ¹¹Ophthalmology, H Valladolid, Valladolid; ¹²Rheumatology, H Toledo, Toledo; ¹³Rheumatology and Ophthalmology, H Donostia, San Sebastián; ¹⁴Rheumatology, H Basurto, Bilbao; ¹⁵Rheumatology, H San Carlos, Madrid; ¹⁶Rheumatology, H Sierrallana, Torrelavega; ¹⁷Rheumatology, H Córdoba, Córdoba; ¹⁸Rheumatology, H San Pedro Alcántara, Cáceres; ¹⁹Ophthalmology, H Zaragoza, Zaragoza; ²⁰Rheumatology, H Mérida, Mérida; ²¹Rheumatology, H Germans Trias i Pujol, Badalona; ²²Rheumatology, H Ferrol, A Coruña; ²³Rheumatology and Ophthalmology, H Miguel Servet, Zaragoza; ²⁴Rheumatology, H Infanta Sofia, Madrid; ²⁵Rheumatology, H Puerta del Mar, Cádiz; ²⁶Rheumatology, H Virgen Rocío; ²⁷Rheumatology, H Virgen Macarena, Sevilla; ²⁸Rheumatology, H Gregorio Marañón, Madrid; ²⁹Rheumatology, H Bellvitge, Barcelona; ³⁰Rheumatology, H Ramón y Cajal, Madrid, Spain

Background: Uveitis is the most common ocular manifestation in Behçet's Disease (BD), which can cause irreversible blindness.^{1–2}

Objectives: To assess efficacy, safety and cost-effectiveness of adalimumab (ADA) therapy optimisation in a series of patients with uveitis due to BD.

Methods: Multicenter study of 74 ADA-treated patients with BD uveitis refractory to conventional immunosuppressants. Following remission, optimisation was performed by increasing the ADA dosing interval. Comparison between optimised and non-optimised group was performed.

Results: Ocular remission was achieved in 65 (86.6%) patients after a median ADA duration of 6^{3–12} months. ADA was optimised in 23 cases. In the remaining 42 ADA was maintained at 40 mg/sc/2 weeks. No baseline differences were found at ADA onset between the optimised and non-optimised groups. Ocular outcomes were similar after a mean ±S.D. follow-up of 34.7±13.3 and 26±21.3 months in the both groups (table 1). Adverse effects were seen in non-optimised group (lymphoma, pneumonia, local reaction and bacteremia). Mean ADA treatment costs were lower in the optimised vs non-optimised group (6101.25 €/patient/year vs 12339.48).

Abstract AB0669 – Table 1

	Optimized Group N=23	Non-Optimized Group N=42	P
Demographic features			
Age (years) (mean±S.D.)	37.2±13.4	39.1±9.3	0.5
Sex (n, ♂/♀)	15/8	19/23	0.13
Positive HLA-B51 (%)	61	74	0.26
Duration of uveitis (months) prior to ADA (median [IQR])	43 [23-74.5]	24 [6-36]	0.1
Previous immunosuppressants (n) (mean±S.D.)	2±1.1	1.7±1.1	0.3
Ocular pattern at ADA onset			
BCVA (mean±S.D.)	0.51±0.36	0.56±0.33	0.46
AC cells (median [IQR])	0 [0-2]	1 [0-2]	0.85
Vitritis (median [IQR])	1 [0-2]	1 [0-2]	0.66
OCT (mean±S.D.)	306.7±122.9	332.8±129.1	0.51
Ocular pattern at last visit			
BCVA (mean±S.D.)	0.89±0.19	0.77±0.25	<0.01
AC cells (median [IQR])	0 [0-0]	0 [0-0]	0.7
Vitritis (median [IQR])	0 [0-0]	0 [0-0]	0.3
OCT (mean±S.D.)	250.5±17.9	249±26.1	0.87
Follow-up on ADA therapy, months, (mean±S.D.)			
Relapses, n (per 100 patients/year)	2 (3.0)	4 (4.4)	0.66
Severe side-effects, n (per 100 patients/year)	0 (0)	4 (4.4)	0.19
Cost mean, euros per year	6101.25	12339.48	<0.01

Conclusions: ADA optimisation in BD uveitis refractory to conventional therapy is effective, safe and cost-effective.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3963

AB0670

SHORT AND LONG-TERM TREATMENT WITH INFILIXIMAB IN RETINAL VASCULITIS OF BEHÇET'S DISEASE. MULTICENTER STUDY OF 72 PATIENTS

J.L. Martín-Varillas¹, V. Calvo-Río¹, R. Demetrio-Pablo¹, B. Atienza-Mateo¹, J. Loricera¹, M.V. Hernández², A. Adán², M. Mesquida², D. Peiteado³, E. Pato⁴, D. Díaz⁴, L. Martínez⁵, E. Valls⁵, M.A. Caracul⁶, A.M. García⁷, J.M. Herrerías⁸, M. Cordero⁹, C. Montilla¹⁰, A. Fonollosa¹¹, A. Atanes¹², F.F. Hernández¹³, I. Torre¹⁴, O. Maiz¹⁵, A. Blanco¹⁵, C. Espartero¹⁶, N. Ortego¹⁷, E. Raya¹⁷, M. Gandía¹⁸, F.J. López¹⁹, M. Alcalde²⁰, C. Fernández²¹, Ó. Ruiz²², F. Jiménez²², R. Almodovar²³, C. Carrasco²⁴, L. Linares²⁵, F. Romero²⁶, S. Insua²⁷, S. González²⁸, M. Hernández²⁹, E. Beltrán³⁰, J. Cruz³¹, C. Fernández³¹, E. Aurecochea³², M.A. González-Gay¹, R. Blanco¹. ¹Rheumatology and Ophthalmology, HUMV, IDIVAL, Santander; ²Rheumatology and Ophthalmology, H Clinica, Barcelona; ³Rheumatology, H La Paz; ⁴Rheumatology and Ophthalmology, H San Carlos, Madrid; ⁵Rheumatology and Ophthalmology, H Peset, Valencia; ⁶Rheumatology, H Córdoba, Córdoba; ⁷Rheumatology, H Toledo, Toledo; ⁸Ophthalmology, H Valladolid, Valladolid; ⁹Ophthalmology, H León, León; ¹⁰Rheumatology, H Salamanca, Salamanca; ¹¹Ophthalmology, H Cruces, Bilbao; ¹²Rheumatology, H Coruña, Coruña; ¹³Rheumatology, H Doctor Negrín, Las Palmas; ¹⁴Rheumatology, H Basurto, Bilbao; ¹⁵Rheumatology and Ophthalmology, H Donostia, San Sebastián; ¹⁶Rheumatology, H Móstoles, Madrid; ¹⁷Autoimmune Diseases, H San Cecilio, Granada; ¹⁸Rheumatology, H Puerta del Mar, Cádiz; ¹⁹Rheumatology, H Gregorio Marañón; ²⁰Rheumatology, H Severo Ochoa, Madrid; ²¹Rheumatology, H Elda, Alicante; ²²Rheumatology and Ophthalmology, H Miguel Servet, Zaragoza; ²³Rheumatology, H Alcorcón, Madrid; ²⁴Rheumatology, H Mérida, Mérida; ²⁵Rheumatology, H Arrixaca, Murcia; ²⁶Rheumatology, Jiménez Díaz, Madrid; ²⁷Rheumatology, H Santiago Compostela, Santiago Compostela; ²⁸Rheumatology, H Cabueñes, Gijón; ²⁹Ophthalmology, H Valencia, Valencia; ³⁰Rheumatology, H del Mar, Barcelona; ³¹Rheumatology and Ophthalmology, H Pontevedra, Pontevedra; ³²Rheumatology, H Sierrallana, Torrelavega, Spain

Background: Retinal vasculitis (RV) is a serious complication of uveitis due to Behçet's disease (BD).¹⁻²

Objectives: We assess the short/long-term efficacy of Infliximab (IFX) in refractory RV of BD.

Methods: Multicenter study of patients with RV of BD refractory to corticosteroids and at least 1 conventional immunosuppressant (IS). We compared efficacy of IFX between baseline, 1st week, 1–6 months and 1–6 years.

Results: 72 patients/129 affected eyes (40♂/32♀) with mean age of 39.6±9.7 years. HLA-B51 was (+) in 63%. Before IFX onset, patients had received: oral/e.v. glucocorticoids (n=98), CyA (n=56), AZA (n=43), MTX (n=34) and other IS (n=22). IFX was used as monotherapy in 17 patients and combined with conventional IS in the remaining 55.

IFX dose was as follows: 3 mg/kg/4–8 w (n=5), 4 mg/kg/4 w (n=1), 5–5.5 mg/kg/4–8 w (n=66).

Following IFX onset, an improvement in RV was seen, as well as in the other ocular outcomes. This enhancement was maintained (table 1).

After a mean follow-up of 26.5±22.1 months, IFX was discontinued in 44: remission (n=15), primary failure (n=16), preference of another route of administration (n=8), pregnancy (n=1) and adverse effects (n=4).

Abstract AB0670 – Table 1

	Basal	1 st week	1 st month	3 rd month	6 th month	1 st year	2 nd year	3 rd year	4 th year	5 th year	6 th year
Retinal Vasculitis (n, affected eyes, %)	117 (100%)	93 (79.5%)	45 (38.5%)	21 (17.9%)	13 (11.1%)	4 (3.1%)	4 (3.1%)	3 (2.6%)	1 (0.9%)	0 (0%)	0 (0%)
BCVA (mean±SD)	0.41±0.31	0.45±0.32	0.57±0.33	0.63±0.34	0.63±0.34	0.63±0.35	0.63±0.35	0.61±0.33	0.63±0.32	0.66±0.37	0.66±0.37
Tyndall (median [IQR])	1 [0-2]	0 [0-2]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]
Vitritis (median [IQR])	1 [0-2]	1 [0-2]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]	0 [0-0]
OCT (mean±SD)	346.9±140.7	339.6±125.6	310.6±114.4	289.4±74.5	264.7±57.1	268.6±52.7	265.2±43.7	253.8±31.0	239.8±33.8	208.6±26.0	194.7±4.1

*p<0.05

Conclusions: IFX seems an effective short/long-term treatment in RV of BD.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4015

AB0671

FIRST DOCUMENTATION OF RS3PE AFFECTING THE HANDS ON 18F-FDG WHOLE BODY PET/CT IN POLYMYALGIA RHEUMATICA

C.E. Owen^{1,2}, A.M. Poon^{2,3}, L.P. Yap⁴, J.L. Leung^{1,2}, D.F. Liew^{1,2}, S.T. Lee^{2,3,5}, A.M. Scott^{2,3,5}, R.R. Buchanan^{1,2}. ¹Rheumatology, Austin Health, Heidelberg VIC; ²Medicine, University of Melbourne, Parkville VIC; ³Molecular Imaging and Therapy; ⁴Radiology, Austin Health; ⁵Olivia Newton-John Cancer Research Institute, Heidelberg VIC, Australia

Background: Remitting seronegative symmetrical synovitis with pitting oedema (RS3PE) syndrome describes a clinical entity characterised by distal synovitis with pitting oedema, the absence of rheumatoid factor (Rf) and an excellent response to glucocorticoid therapy.^[1] Most frequently associated with polymyalgia rheumatica (PMR), tenosynovial sheath inflammation represents the magnetic resonance imaging (MRI) hallmark of this condition, with concomitant joint synovitis also present in some cases.^[1] More recently, diffusely increased ¹⁸F-fluorodeoxyglucose (¹⁸F-FDG) uptake in the soft tissues around the ankles and feet has been described as the correlate of RS3PE on whole body positron emission tomography/computed tomography (PET/CT).^[2]

Objectives: To document the clinical and radiologic appearance of RS3PE syndrome affecting the hands on MRI and whole body PET/CT in PMR patients.

Methods: Patients with newly diagnosed PMR were prospectively recruited as part of the Melbourne Predictors of Relapse in PMR (MPR-PMR) study. A standard physical examination was carried out with specific focus upon the presence of peripheral synovitis and pitting oedema. In patients with findings suggestive of RS3PE, clinical photography was undertaken. All study participants underwent a whole body PET/CT scan including dedicated views of the hands using the Philips T/F machine prior to prednisolone commencement. To precisely identify anatomic correlates of abnormal ¹⁸F-FDG uptake in patients with RS3PE, MRI of the wrist and hand was performed using a 1.5 Tesla magnet.

Results: 3/35 patients (0.86%) were noted to have distal synovitis and pitting oedema of the hands at enrolment. Mean age was 70.9±10.1 years, two patients were male, and all were Caucasian. Rf and anti-citrullinated peptide autoantibodies were negative in all cases. On whole body PET/CT, intense ¹⁸F-FDG uptake was visualised at the wrist joint and hand in a distinctive volar distribution. MRI of the wrist and hand in two participants (contraindicated in the third)