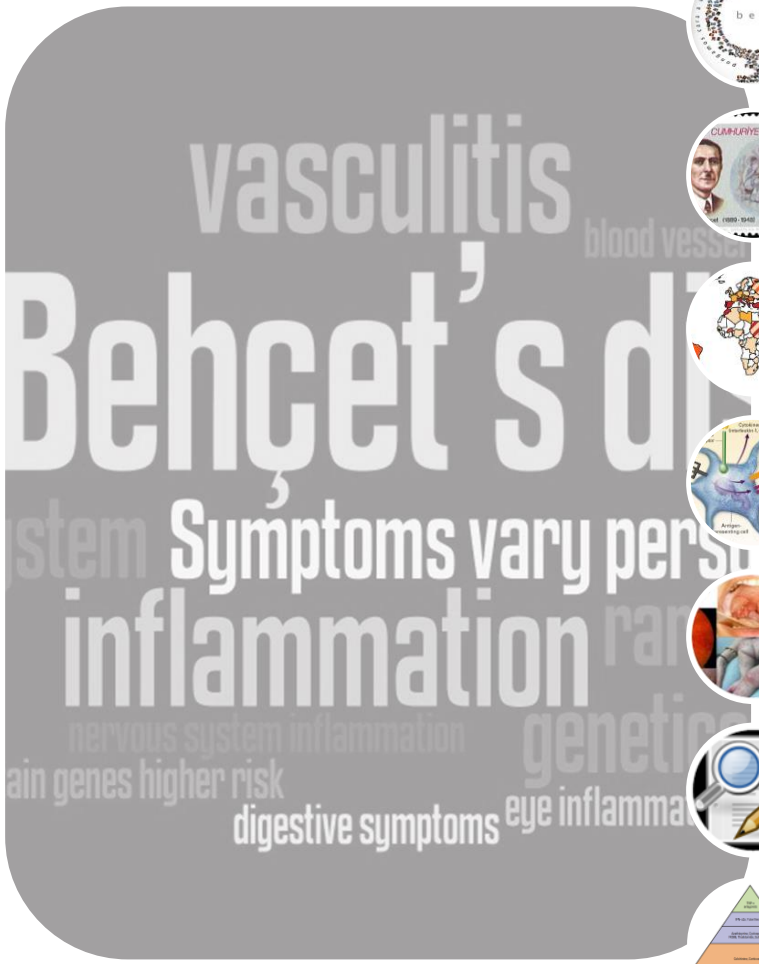
Síndrome de Behcet

Felipe Estupiñán López, MD

Residente I de dermatología y cirugía dermatológica

Universidad del Valle

Contenido



Definición

Historia

Epidemiología

Fisiopatología

Manifestaciones clínicas

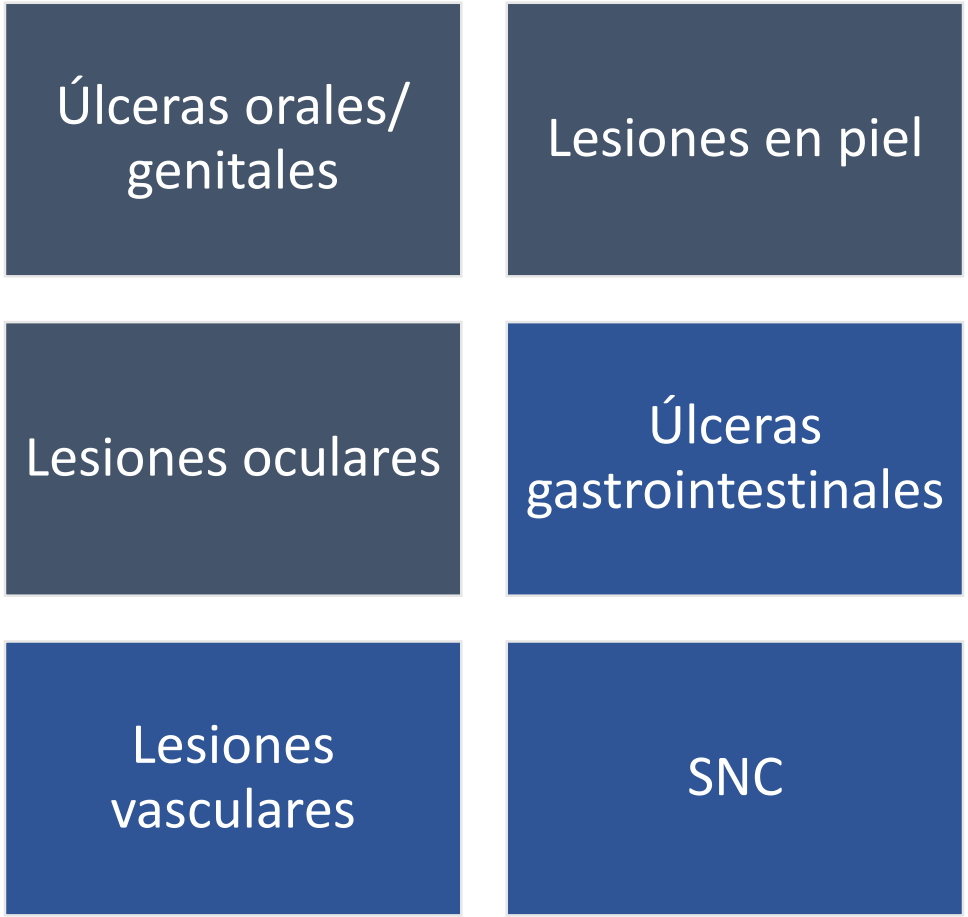
Diagnóstico

Tratamiento

Definición

La enfermedad de Behcet es un desorden inflamatorio multisistémico crónico

- Enfermedad vs. síndrome
- Etiología desconocida



Cho, S. B., Cho, S., & Bang, D. (2012). New Insights in the Clinical Understanding of Behçet's Disease. *Yonsei Medical Journal*, *53*(1), 35–42

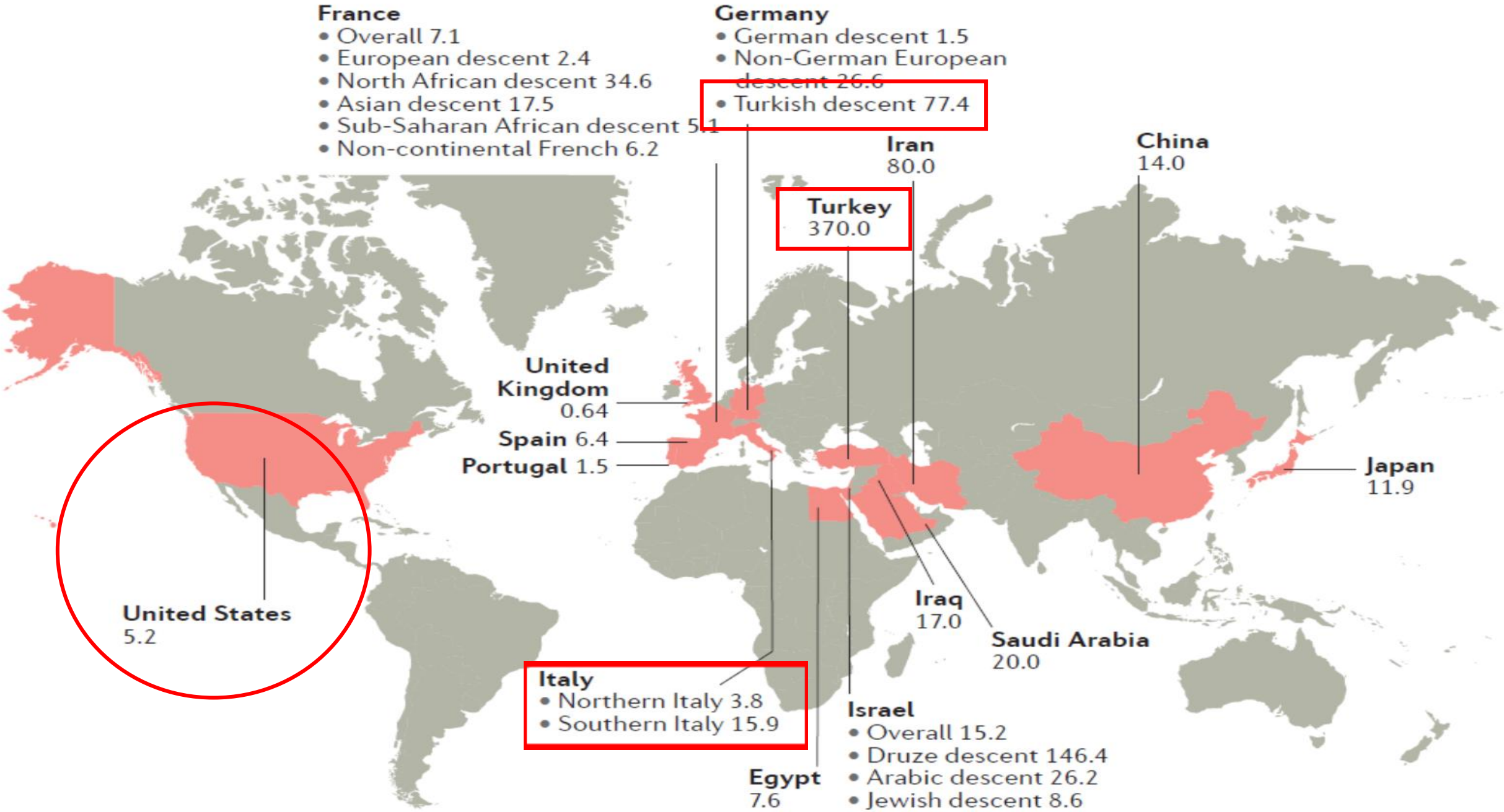
Historia

Benediktos Adamantiades

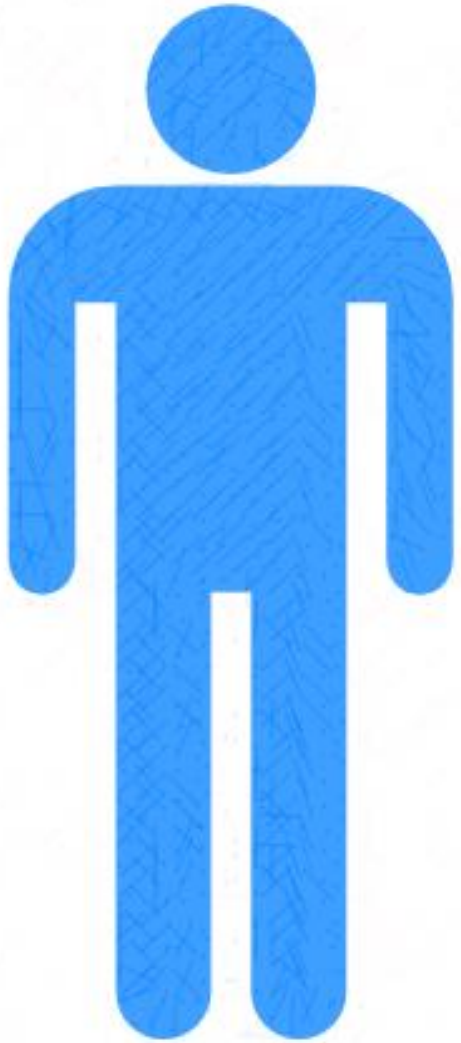
Hulusi Behcet

Epidemiología

Hasan Yazici1 et al; Behçet syndrome: a contemporary view. NATURE REVIEWS | RHEUMATOLOGY doi:10.1038/nrrheum.2017.208



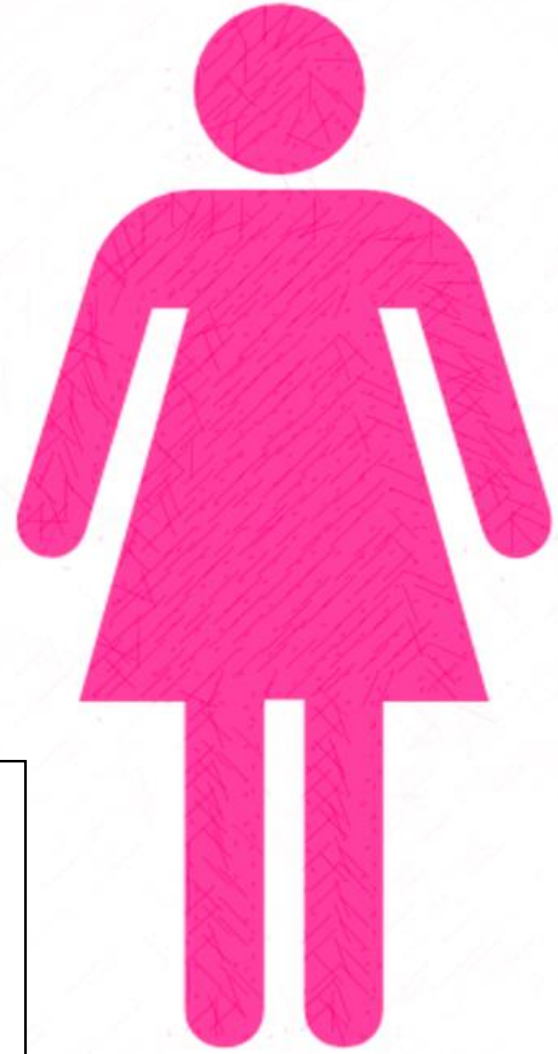
Hasan Yazici1 et al; Behçet syndrome: a contemporary view. NATURE REVIEWS | RHEUMATOLOGY



H = M

20-30 años

<200 casos (2012)



Cho, S. B., Cho, S., & Bang, D. (2012). New Insights in the Clinical Understanding of Behçet's Disease. *Yonsei Medical Journal*, *53*(1), 35–42.

Fisiopatología

Autoinflamación vs. autoinmunidad

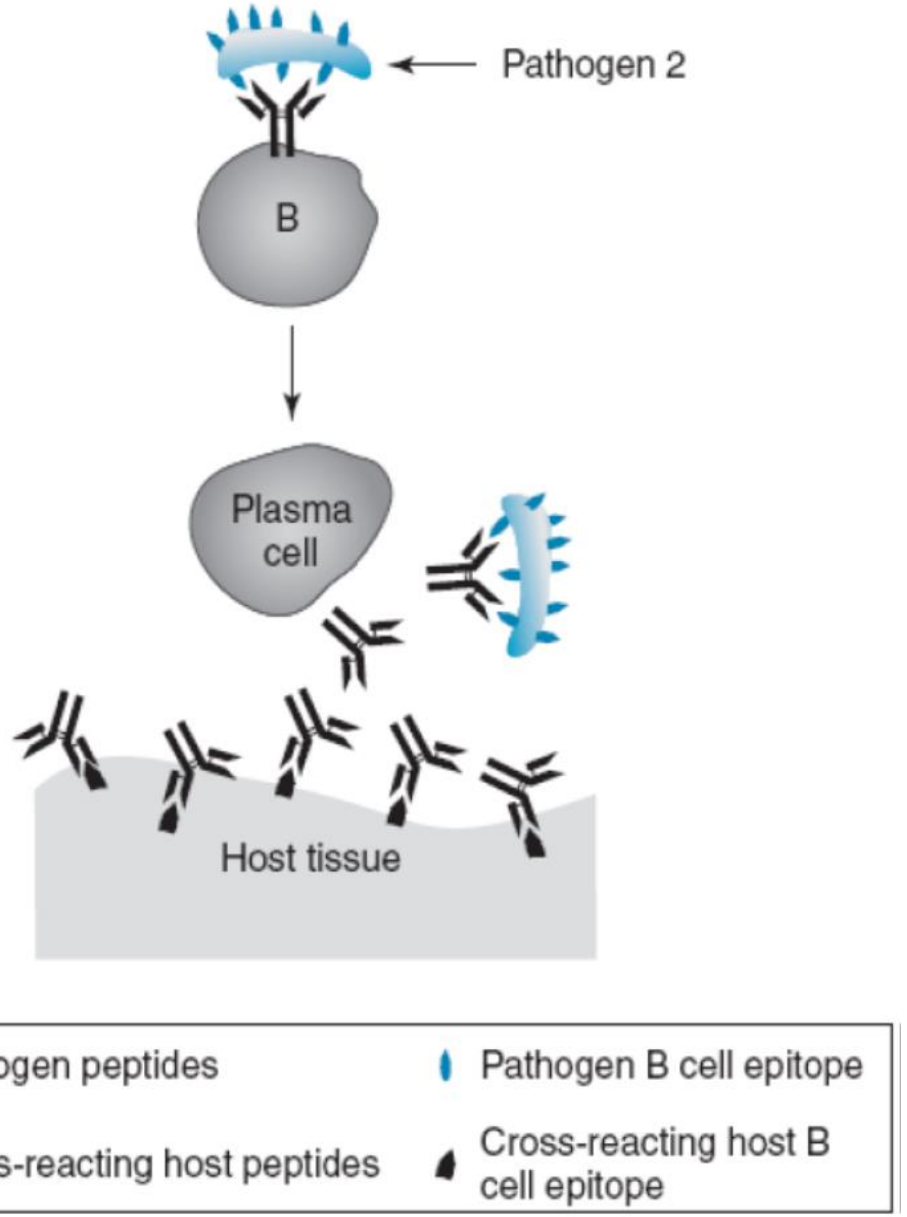
INNATE immune system	**Immune dysregulation**	ADAPTIVE immune system
Monocytes, macrophages, neutrophils	**Predominant cell types**	T cells, B cells
IL-1, TNF, IFNαβ, IL-12, IL-23, (IL-17), IL-18	**Cytokine targets used therapeutically**	IFNγ, IL-4, (IL-17), IL-6
Neutrophil- and macrophage-mediated organ damage	**Pathogenesis of organ damage**	Autoantibody- or autoantigen-specific T cell–mediated organ damage
IL-1-mediated monogenic autoinflammatory diseases	**Disease examples**	Thyroiditis, rheumatoid arthritis, SLE, ALPS

Annu. Rev. Med. 2014. 65:223–44

Is Behçet's disease a class 1-opathy? The role of HLA-B*51 in the pathogenesis of Behçet's disease M. Giza, D. Koftori, L. Chen, P. Bowness Clin Exp Immunol. 2018 Jan; 191(1): 11–18

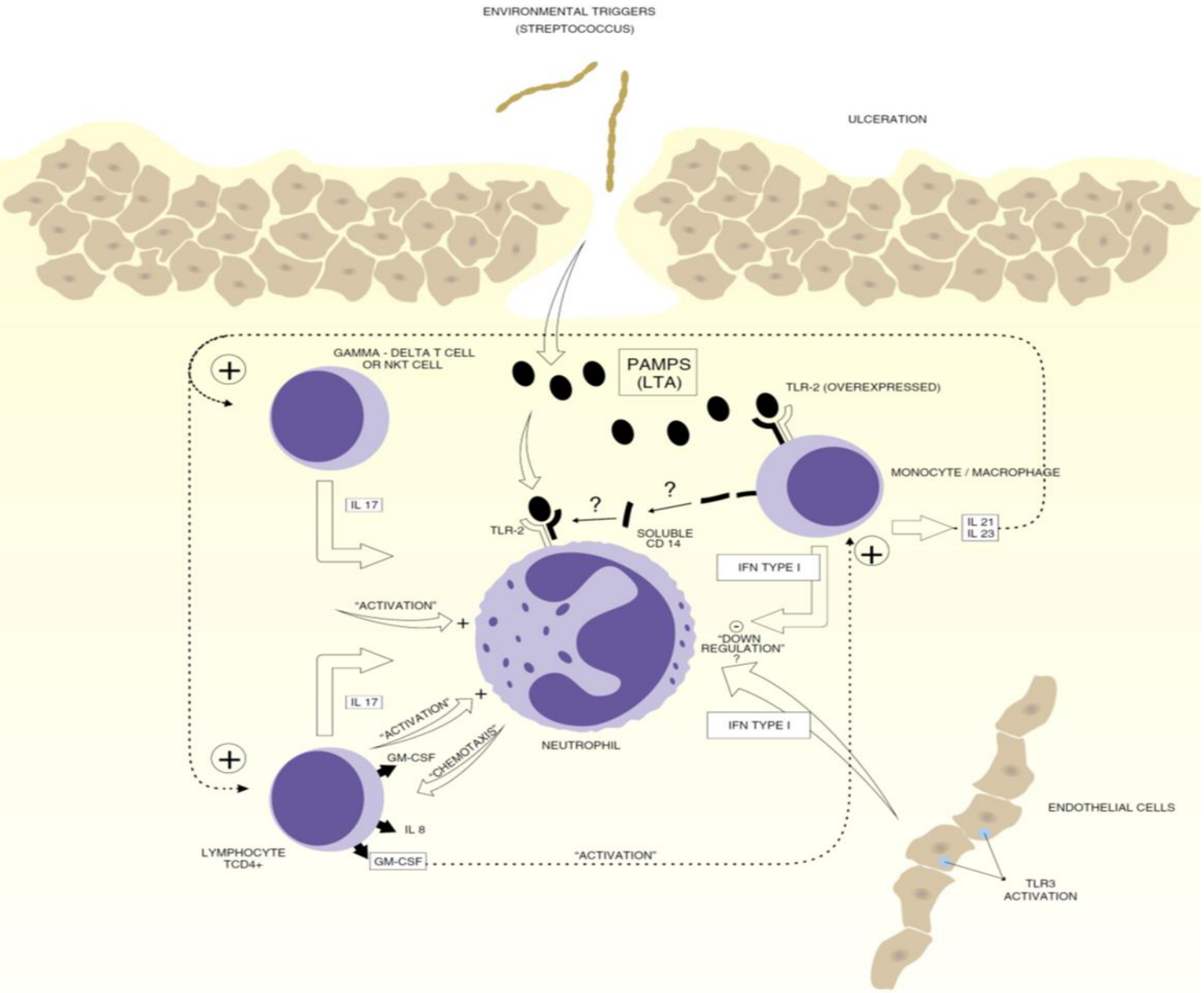
No hay elementos suficientes para autoinmunidad

- HSP – mímica molecular
- Anticuerpos contra las proteínas de choque térmico de *Streptococcus spp*. se han mostrado recientemente en Behçet



Tomado de clase: "mecanismos de autoinmunidad". Módulo inmunología

Direskeneli, H. (2013). Innate and Adaptive Responses to Heat Shock Proteins in Behcet's Disease. *Genetics Research International*, *2013*, 249157.

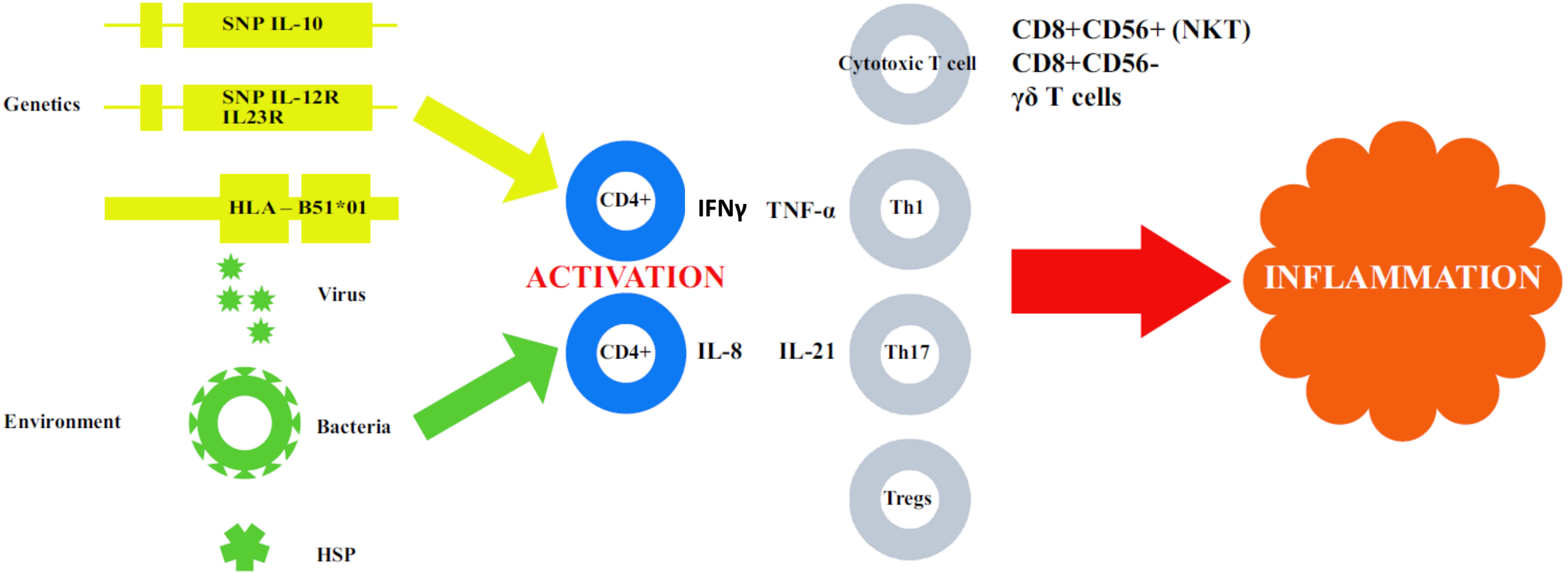


Elementos a favor de autoinflamación

- IL-1β incrementado
- Predominio de Respuesta Inmune Innata por mutaciones en TLR
- Mayor presencia de neutrófilos en lesiones
- Similitudes con MEFV

Hasan Yazici1 et al; Behçet syndrome: a contemporary view. NATURE REVIEWS | RHEUMATOLOGY doi:10.1038/nrrheum.2017.208

La confluencia de diversos factores generan inflamación



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Arthritis & Rheumatism (Arthritis Care & Research)
Vol. 61, No. 10, October 15, 2009, pp 1287–1296
DOI 10.1002/art.24642

HLA–B51/B5 and the Risk of Behçet's Disease: A Systematic Review and Meta-Analysis of Cas

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32–52%.

Conclusion. The strength of the association between BD and *HLA–B51/B5*, and its consistency across populations of various ethnicities, lends further support to this allele being a primary and causal risk determinant for BD. Variations according to sex support an interaction of this allele with BD characteristics.

Table 2. Pooled estimates for overall and subgroup meta-analyses for *HLA–B51/B5* carriage and its association with BD risk*

Subgroups	Populations, no.	Pooled prevalence for *HLA–B51/B5*† BD cases (95% CI)	Pooled prevalence for *HLA–B51/B5*† Controls (95% CI)	OR (95% CI)	I^2 (%)	P_{het}	P_{cov}
Overall	80	57.2 (53.4–60.9)	18.1 (16.1–20.3)	5.78 (5.00–6.67)	60.6	0.0001	
By geographic area							0.31
Eastern Asia	25	55.0 (49.8–60.1)	19.6 (16.0–23.7)	5.18 (4.15–6.47)	52.2	0.001	
Middle East/North Africa	27	63.5 (58.8–68.0)	21.7 (18.2–25.7)	6.25 (4.87–8.03)	70.4	0.0001	
Southern Europe	15	60.6 (51.9–68.7)	16.8 (13.3–21.0)	7.20 (4.89–10.62)	57.2	0.003	

Table 2. Pooled estimates for overall and subgroup meta-analyses for *HLA–B51/B5* carriage and its ass

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significance of corresponding covariates in the pooled genetic effect (calculated by random-effects meta-regression).
† Pooled prevalence values were calculated using random-effects normal-logistic models.
‡ Two studies combined in the North American group had distinctly different ethnicities.

De Menthon, M., Lavalley, m. P., Maldini, C., Guillevin, l., & Mahr, a. (2009). *HLA–B51/B5* and the Risk of Behçet's Disease: A Systematic Review and Meta-Analysis of Case–Control Genetic Association Studies. *Arthritis and Rheumatism*, *61*(10), 10.1002/art.24642.

LABORATORY INVESTIGATION

Association of Maj
Class I Chain-Relat
with Behç

Nobuko Mizuki[1], Akira Megu
an

[1]Department of Oral and Maxillofaci
Medicine, Yokohama, Japan; [2]Depart
University Graduate School of M
Department of Internal Medicine, I
Turkey; [4]Department of Ophthalmo
School

Purpose: Behçet's disease (BD) is kno
groups. Recently, the major histocompa
near the *HLA-B* gene, has been propo
groups. To compare the relative contribu
ethnic groups, we studied *MICA* polym

Methods: Thirty-three Turkish BD pati
morphisms in the extracellular domains

Results: The phenotype frequencies of *M*
in controls (29.2%) ($P = 0.000015$). *H*
(81.8%) than in controls (29.2%) ($P = 0$
*HLA-B**51. To assess the confounding e
analysis that showed that BD was distin

Conclusion: Our results indicate that th
association between *MICA**009 and BI
However, we suggest that *MICA**009 lik
mol 2007;51:431–436 © Japanese Ophth

Key Words: Behçet's disease, *HLA-B*

E.H. Hughes
R.W.M. Collins
E. Kondeatis
G.R. Wallace
E.M. Graham
R.W. Vaughan
M.R. Stanford

Associations of major histocompatibility
con
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Key words:
Behcet's disease; HLA-B*51; *MICA*; *MICB*; NKG2D; polymorphism

Acknowledgments:
This work was supported by grants from the British Eye Research Foundation.

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doi: 10.1111/j.1365-3083.2006.01780.x

Allelic Diversity and Affinity Variants of
Imbalanced in Spanish Patients with Bel

I. Muñoz-Saá*, A. Cambra*, L. Pallarés†, G. Espinosa‡, A. Juan§, F. Pujalte*
J. Milà* & M. R. Julià*

Abstract

The aetiology of Behçet's disease (BD) is stil
environmental factors are involved. HLA-B*51
marker and some MICA alleles have also b
lymphocytes have been suggested as responsibl
MICA through NKG2D surface molecules. In t
MICA alleles were typed by polymerase chain r
primers, in 165 healthy Spanish controls and 4
group, MICA*008 (28.48%), MICA*004 (17.58
MICA*009 (9.39%) were the predominant al
haplotype was MICA*004-B*44 (12.12%). MIC
MICA*011 (4.54%) and MICA*018 (5.15%
MICA*010 (1.81%) and MICA*008 were l
Caucasoid populations. Similar results have be
individuals and this could support the hypot
origin of both populations. The frequencies o
were significantly increased in our BD patients
22.62% versus 9.39% and 10.71% versus 1.81%
MICA*019 had not been described in other BI
the genetic heterogeneity at MICA locus in BD
alleles for NKG2D were more frequent in contr
high-affinity alleles were not found in homozyg
argue against the hypothesis of an autoaggres
through MICA–NKG2D interactions.

*Immunology Service, Hospital Universitari Son Dureta, Palma de Mallorca, Spain; †Autoimmune Diseases Unit of the Internal Medicine Service, Hospital Universitari Son Dureta, Palma de Mallorca, Spain; ‡Autoimmune Diseases Department, Institut Clínic d'Infeccions i Immunologia, Hospital Clínic, Barcelona. Spain; and §Rheumatology Service, Hospital Son Llàtzer, Palma de Mallorca, Spain

Received 15 March 2005; Accepted in revised form 19 April 2006

Correspondence to: Dr M. R. Julia, Immunology Service, Hospital Universitari Son Dureta, Palma de Mallorca, Andrea Doria, 55, 07014, Spain. E-mail: mrjulia@hsd.es

Sociedad Española de Reumatología · Colegio Mexicano de Reumatología

Reumatología Clínica

www.reumatologiaclinica.org

Original Article

The HLA-B*51 Allele Is Strongly Associated With Behçet Disease in an Argentinean Population

Sebastián Andrés Muñoz[a], Alberto Omar Orden[b], Alex Kostianovsky[c], Cecilia N. Pisoni[c], Marina Scolnik[d], Aurelia Luissi[d], Yanina Bottinelli[e], Gustavo Vijoditz[e], Mercedes Garcia[f], Claudia Pena[f], Mariana Pera[f], Oscar Rillo[g], Teresita Alvarellos[h], Luciana M. Más[h], Arturo Luis Trunzo[i], Alberto Allievi[j,*]

[a] *Hospital General de Agudos Dr. Juan A. Fernández, Ciudad Autónoma de Buenos Aires, Argentina*
[b] *Clínica San Camilo, Ciudad Autónoma de Buenos Aires, Argentina*
[c] *Hospital Universitario CEMIC, Ciudad Autónoma de Buenos Aires, Argentina*
[d] *Hospital Italiano de Buenos Aires, Ciudad Autónoma de Buenos Aires, Argentina*
[e] *Hospital Nacional Dr. Alejandro Posadas, Ciudad del Palomar, Provincia de Buenos Aires, Argentina*
[f] *Hospital General José de San Martín, Ciudad de La Plata, Provincia de Buenos Aires, Argentina*
[g] *Hospital General de Agudos Dr. Ignacio Pirovano, Ciudad Autónoma de Buenos Aires, Argentina*
[h] *Hospital Privado Universitario de Córdoba, Ciudad de Córdoba, Provincia de Córdoba, Argentina*
[i] *Instituto Nacional Central Único Coordinador de Ablación e Implante (INCUCAI), Ciudad Autónoma de Buenos Aires, Argentina*
[j] *Medicina, Universidad del Salvador, Ciudad Autónoma de Buenos Aires, Argentina*

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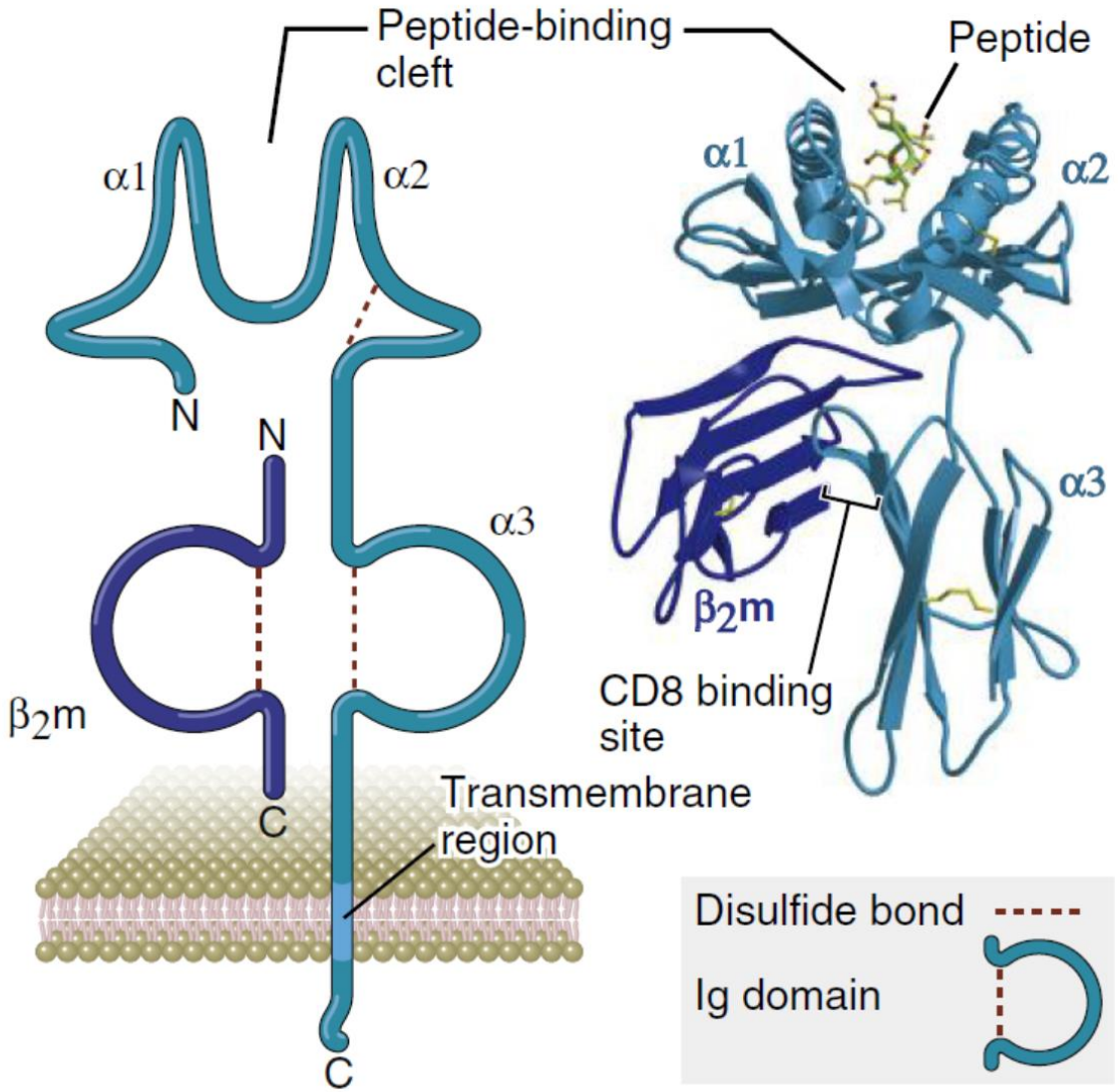
Behçet's disease
HLA-B antigen
HLA-B51 antigen
Case–control studies
Latin America

ABSTRACT

Objective: To assess the association between the HLA-B*51 allele and Behçet Disease (BD) in Argentinean patients.
Methods: We enrolled 34 consecutive Argentinean patients with definitive diagnosis of BD between October 2016 and March 2017. None of the patients had the HLA-B*51 allele determined at study entry. Unrelated controls ($n = 240$) were randomly obtained from the national cadaveric donor database. Demographic and clinical features of the patients were recorded by attending physicians through a questionnaire.
Results: Mean age of cases was 42 years old. Nineteen (55.8%) were male, and the mean age at diagnosis was 35 years old; twenty (58.8%) were Mestizos, 8 (23.5%) were Caucasian, and 6 (17.6%) were Amerindians. Thirteen (38.2%) of 34 cases were HLA-B*51 allele positive; 11 were heterozygous and 2 homozygous for the allele. Thirty-four (14.2%) of 240 controls were positive for the HLA-B*51 allele. The association between BD and HLA-B*51 allele was greater than that of control group (OR = 3.75; $p = 0.0012$).
Conclusions: The HLA-B*51 allele is strongly associated with BD in Argentinean patients. Our finding is consistent with previous studies indicating that the HLA-B*51 allele is an important susceptibility gene in BD regardless the geographical region and ethnicity.

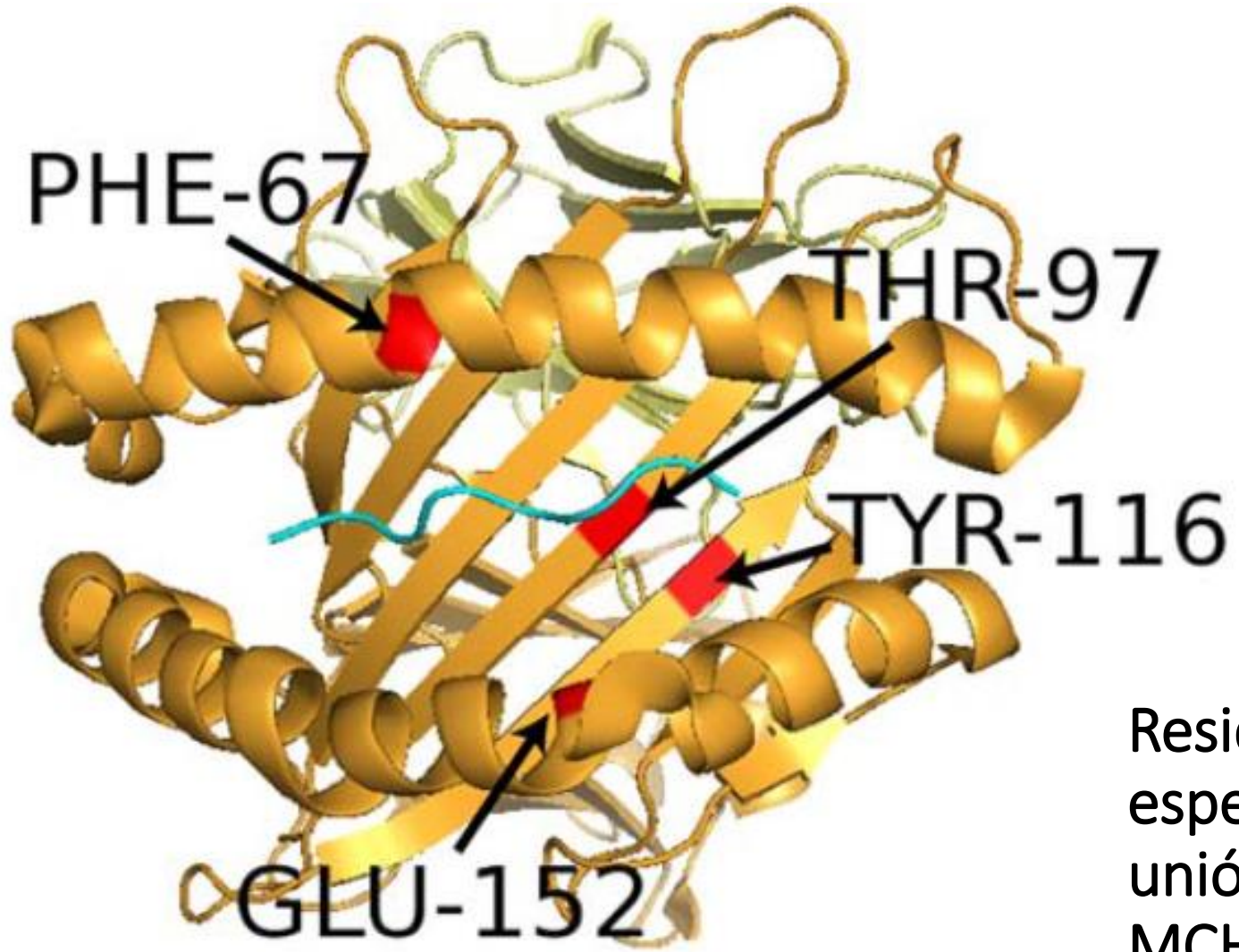
El alelo HLA-B*51 se asoció fuertemente a la Enfermedad de Behçet en la población argentina

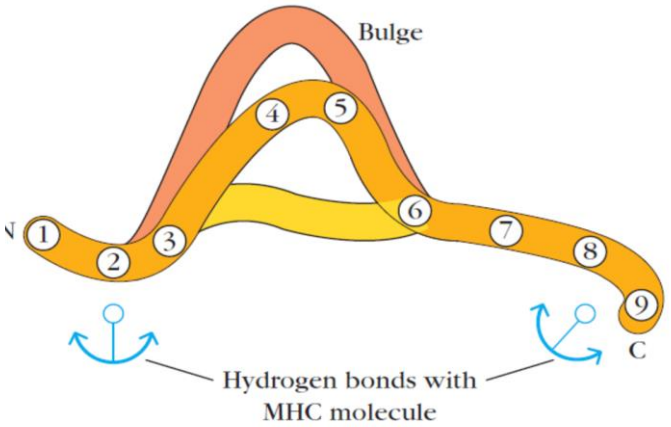


Abbas, Abul K., author. Cellular and molecular immunology / Abul K. Abbas, Andrew H. Lichtman, Shiv Pillai ; illustrations by David L. Baker, Alexandra Baker. -- Eighth edition.

Polimorfismo de HLA-B*51 se traducen en alteraciones en el bolsillo de unión al antígeno

McGonagle, D., Aydin, S.Z., Gül, A., Mahr, A., & Direskeneli, H. (2015). 'MHC-I-opathy'—unified concept for spondyloarthritis and Behçet disease. *Nature Reviews Rheumatology, 11*, 731-740.



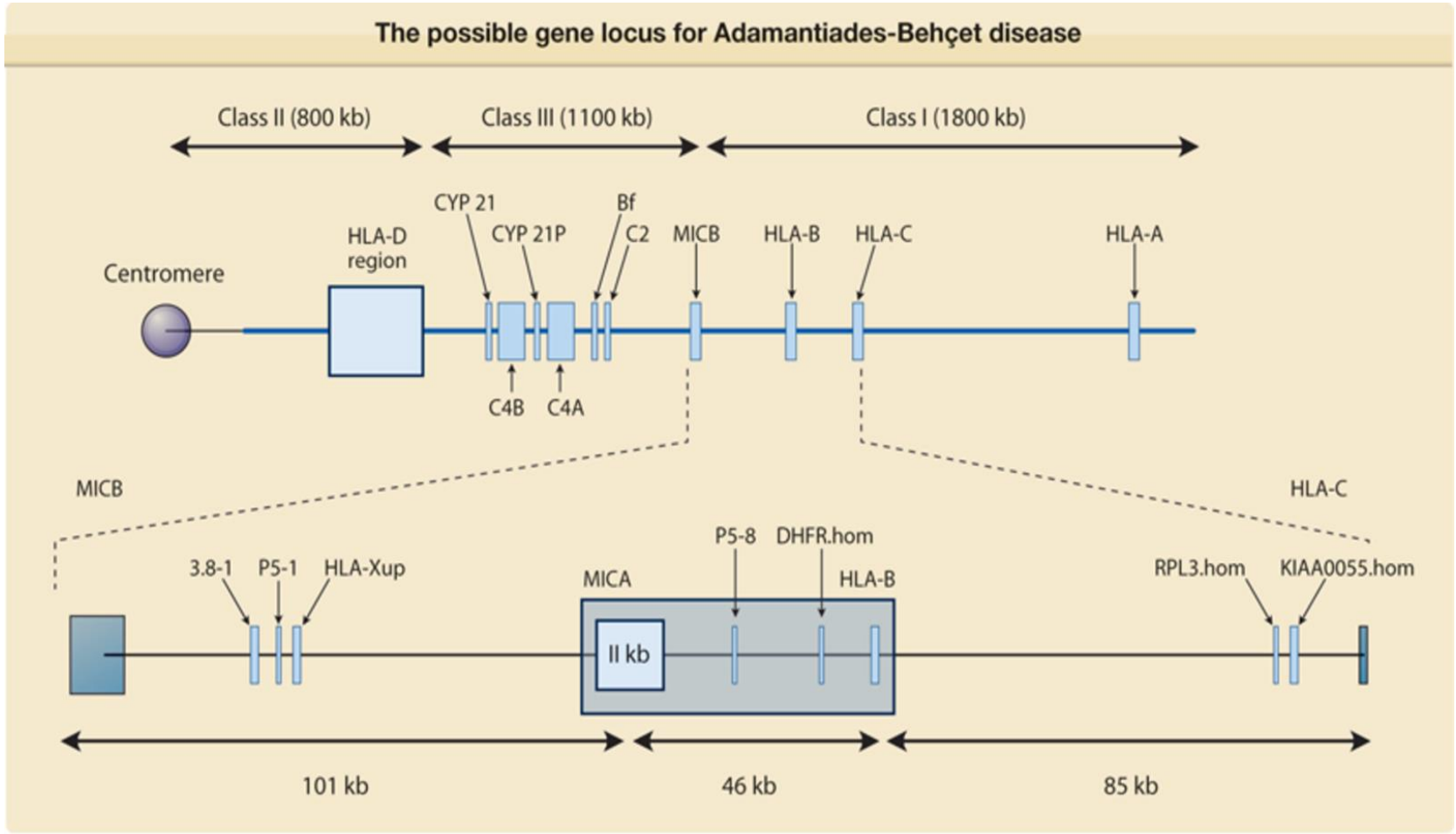


Judith A. Owen et al. Kuby Immunology, 7° e

Residuos 67 y 116 definen especificidad peptídica de unión al antígeno en molécula MCH I

Is Behçet's disease a 'class 1-opathy'? The role of HLA-B*51 in the pathogenesis of Behçet's disease M. Giza, D. Koftori, L. Chen, P. Bowness Clin Exp Immunol. 2018 Jan; 191(1): 11–18.

Susceptibilidad genética



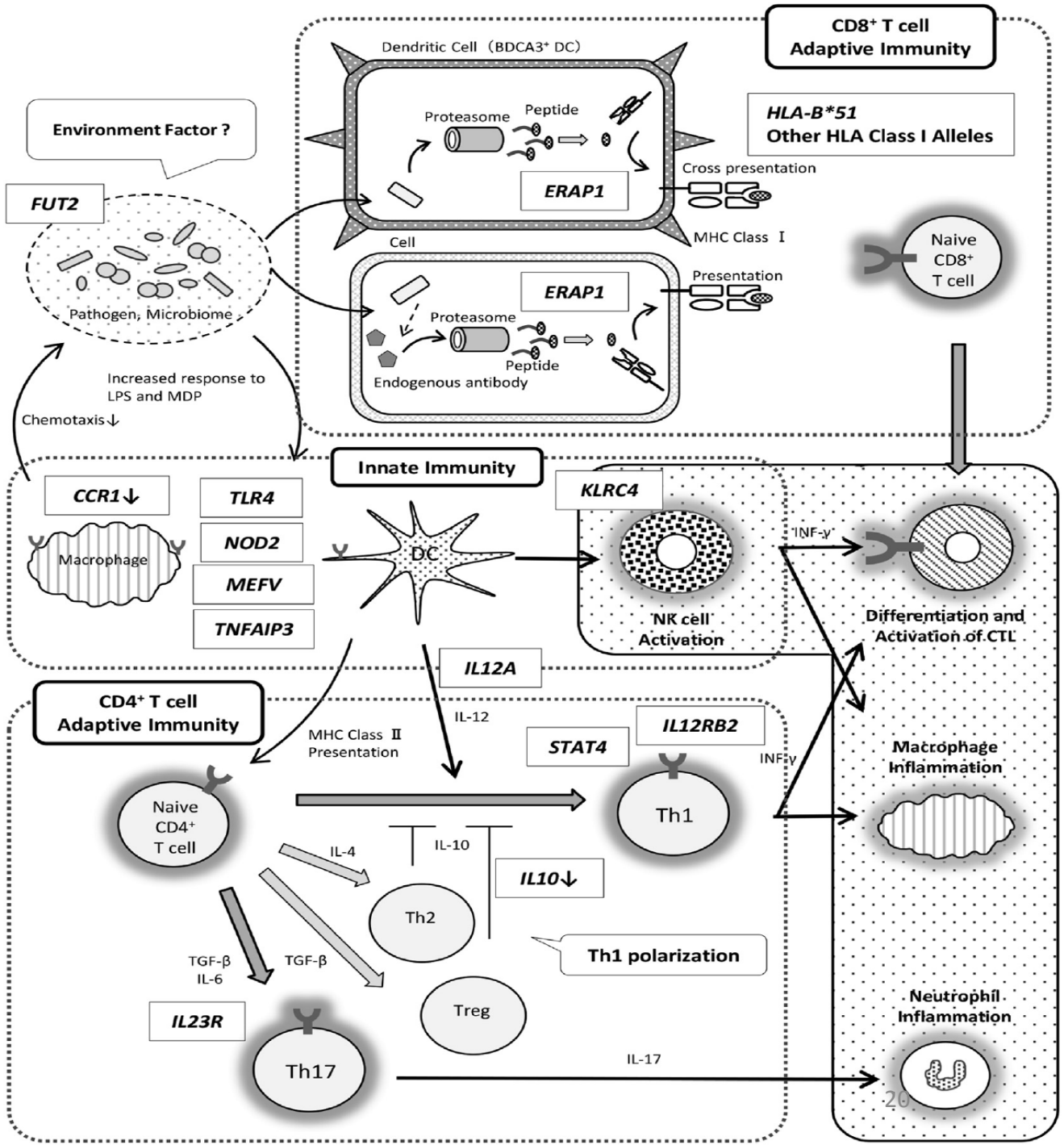
- Distribución geográfica
- Agregación familiar
- Correlación con Antígenos de clase I HLA (HLA-B51)
- Polimorfismos en genes que controlan las respuesta inmune*

Zeidan, M. J., Saadoun, D., Garrido, M., Klatzmann, D., Six, A., & Cacoub, P. (2016). Behçet's disease physiopathology: a contemporary review. *Auto-Immunity Highlights*, *7*(1), 4

- Alteración microbioma
- Presentación antígenos

- Sobreactivación respuesta inmune innata

- Inducción Th-1/Th-17
- Macrófagos/ neutrófilos

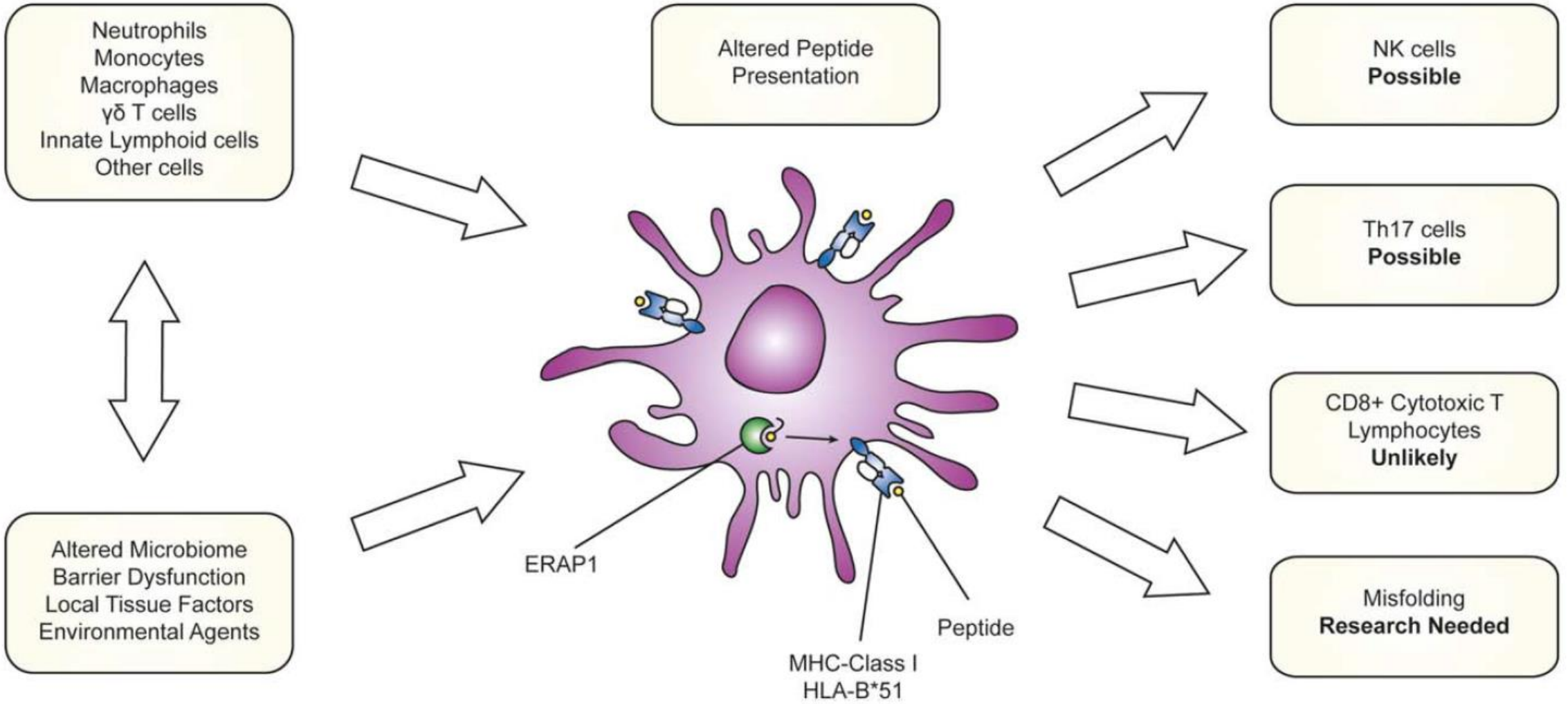


Takeuchi, M., Kastner, D. L., & Remmers, E. F. (2015). The immunogenetics of Behçet's disease: A review. *Journal of Autoimmunity*, *64*, 137–148.

Adapted from D. Mc Gonagle & M. McDermott - PLoS Medicine August 2006

McGonagle D, McDermott MF (2006) A Proposed Classification of the Immunological Diseases. PLoS Med 3(8): e297

MHC I - patía



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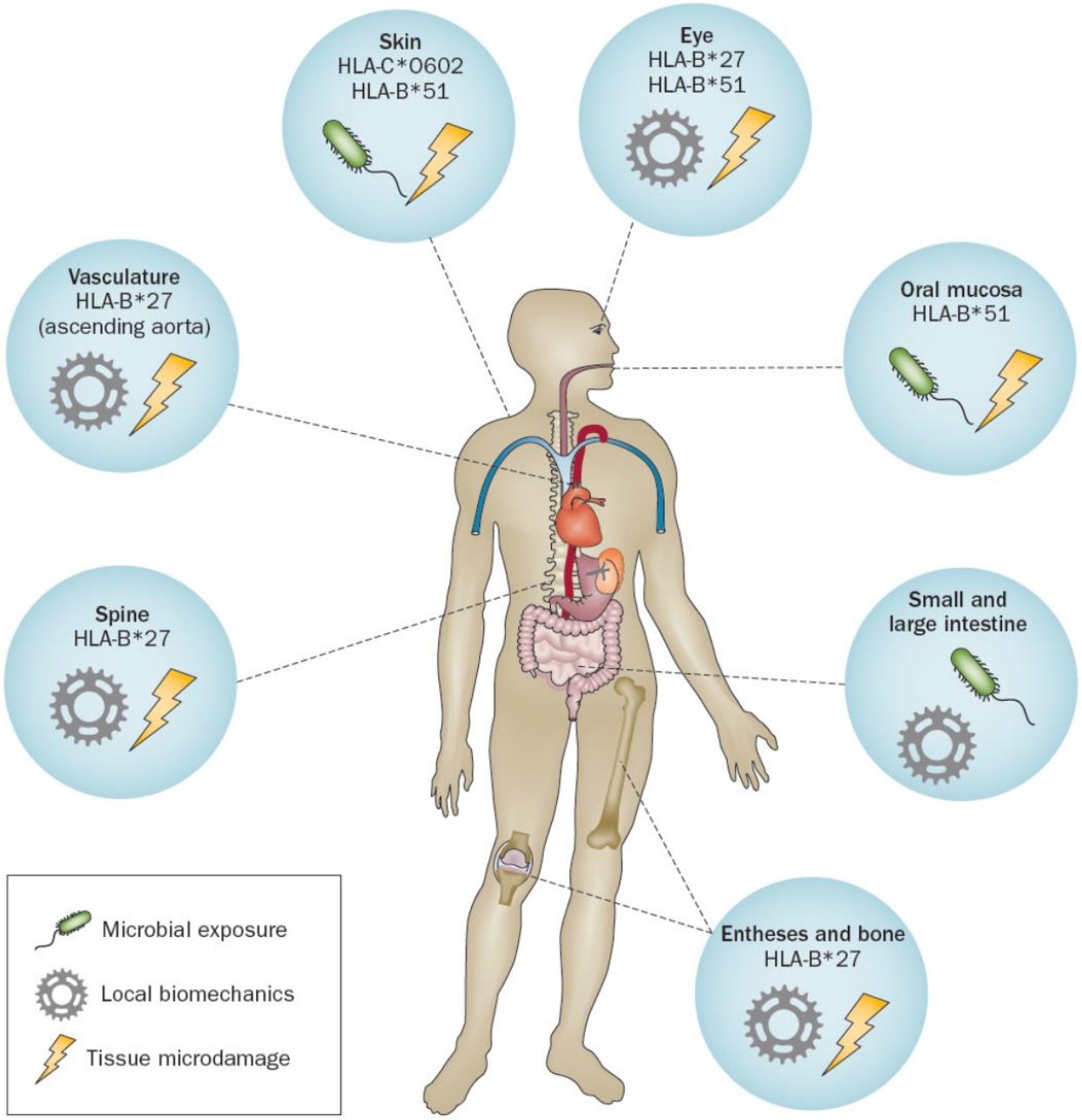
Enfermedades relacionadas con MCH I

Trauma piel:

- Patergia (BD)
- Koebner (Ps)

Áreas estrés:

- Entesis (AS)



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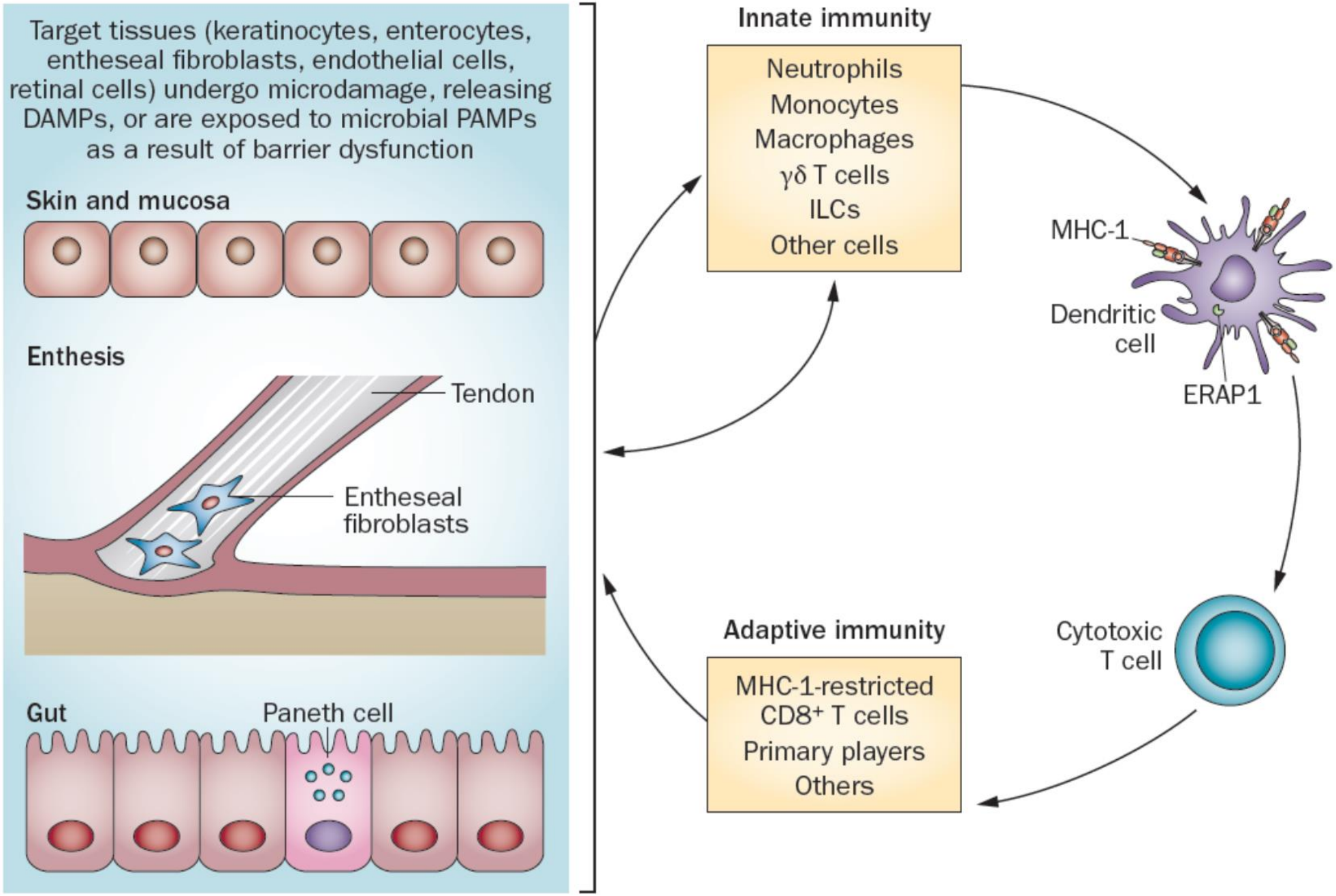
MHC –I es en enlace entre la respuesta inmune innata y adaptativa



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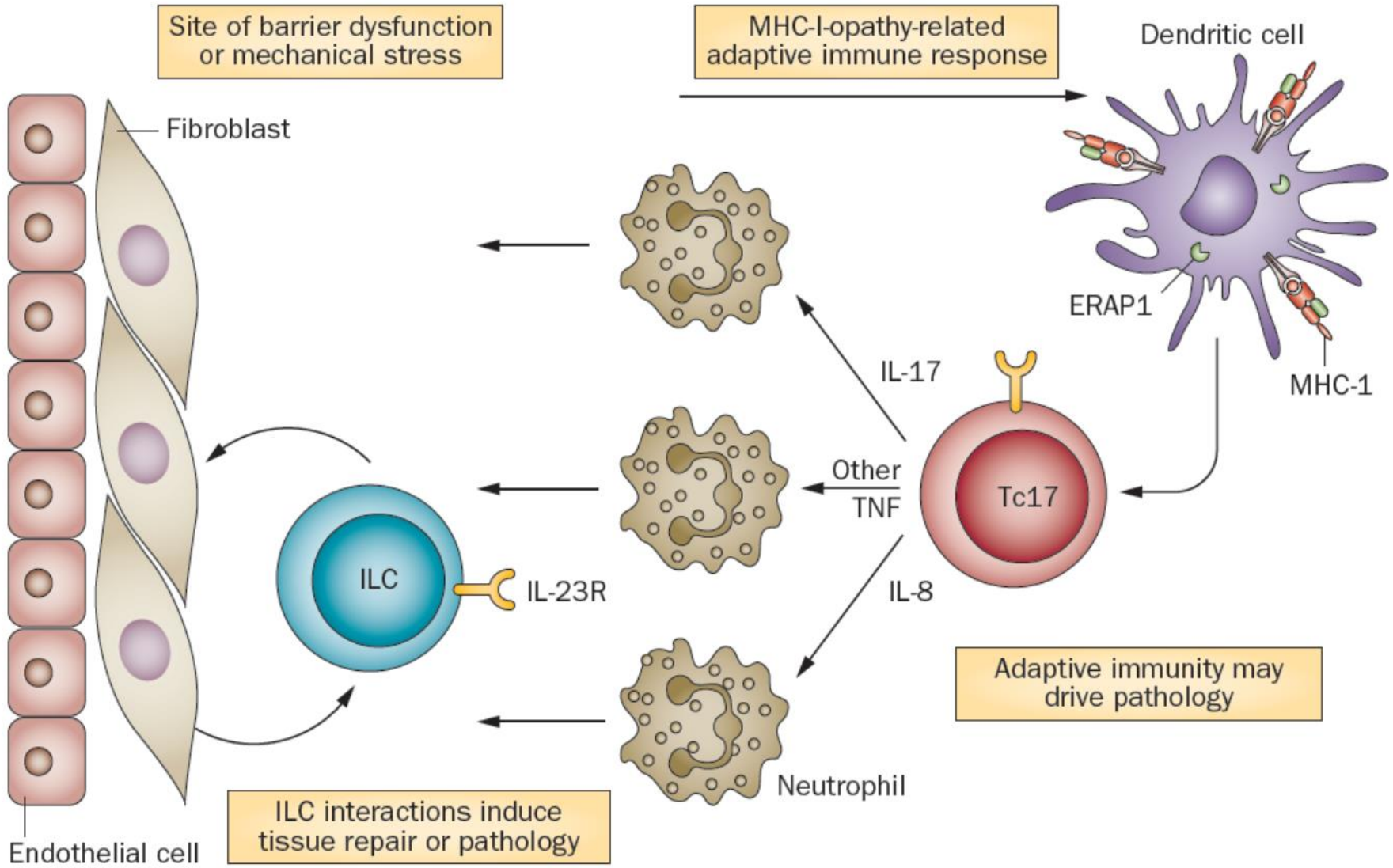
Amplificación respuesta inmune innata y adaptativa

LT CD8+ :
IL8 (CXCL8), GM-CSF

Inflamación neutrofílica



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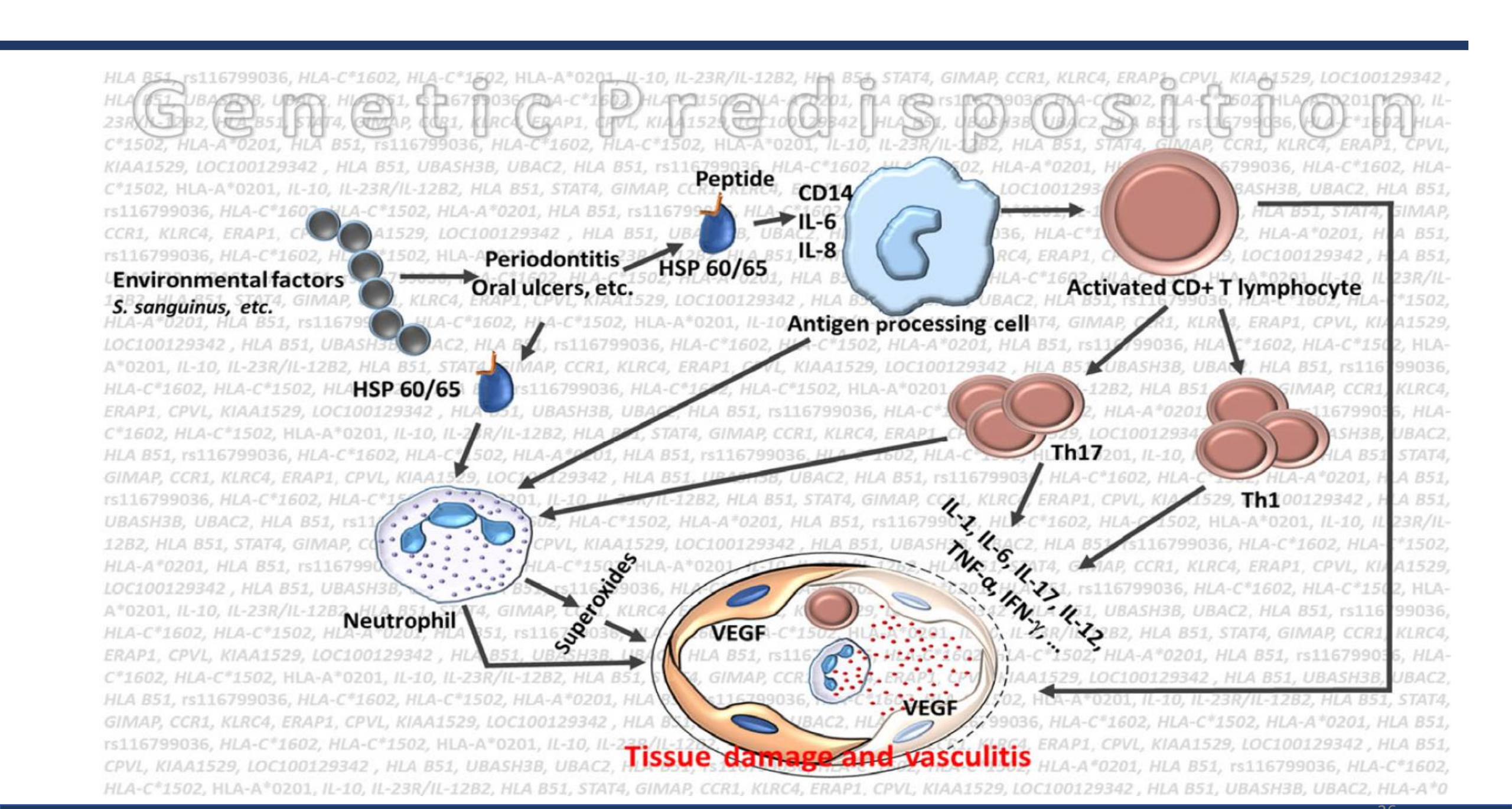
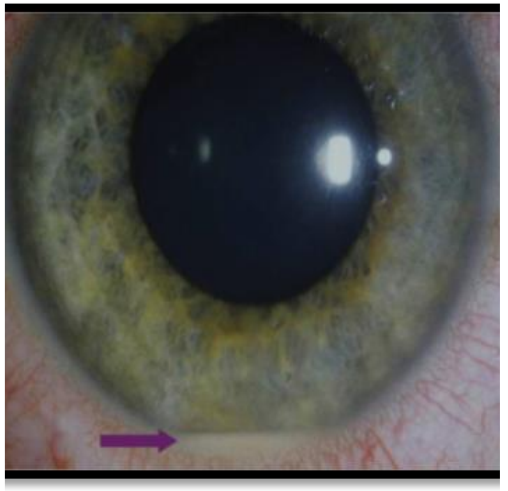


Alpsoy, E. (2016), Behçet's disease: A comprehensive review with a focus on epidemiology, etiology and clinical features, and management of mucocutaneous lesions. J Dermatol, 43: 620-632

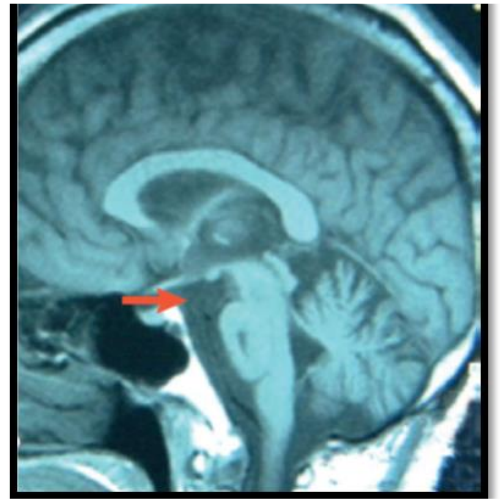
Manifestaciones clínicas

Compromiso multisistémico

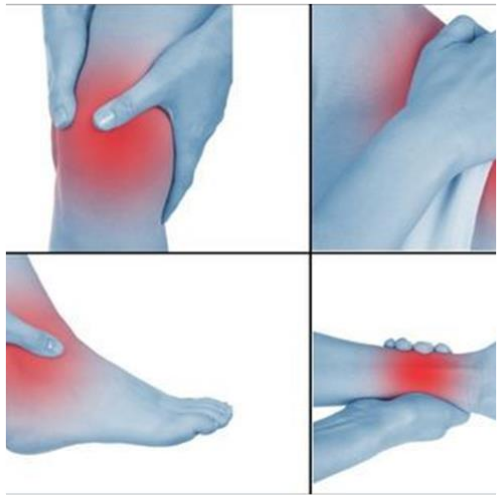
Ocular



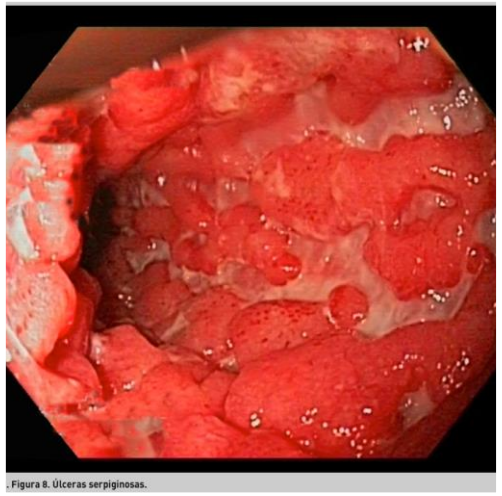
Neurológica



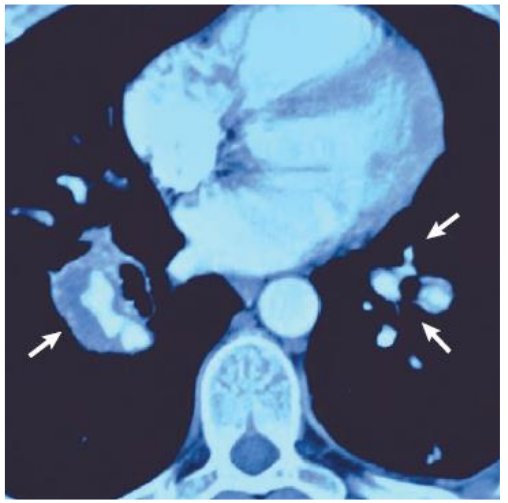
Articular



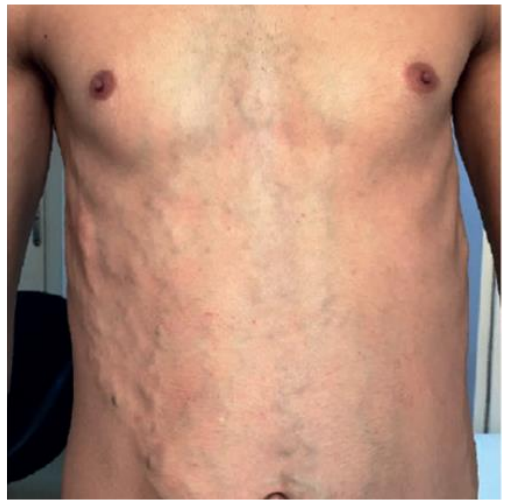
Gastrointestinal



Vascular-A

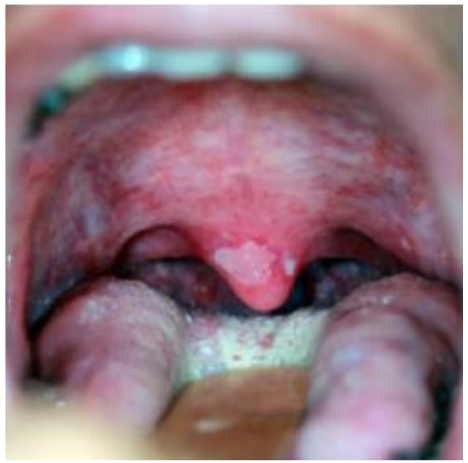
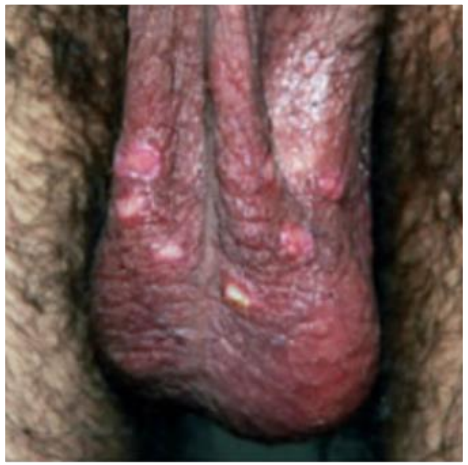
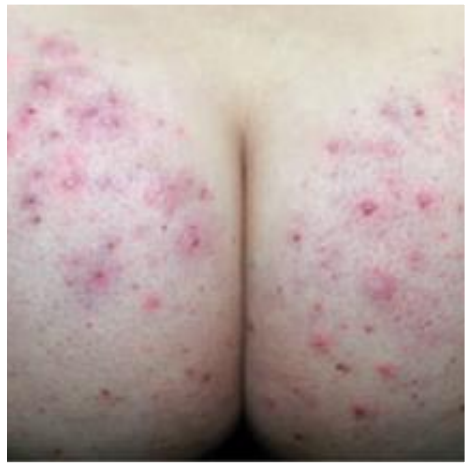
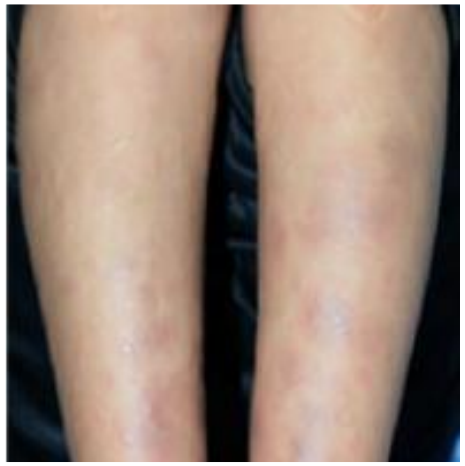
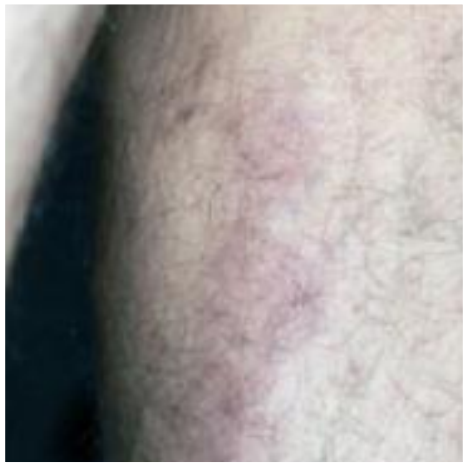
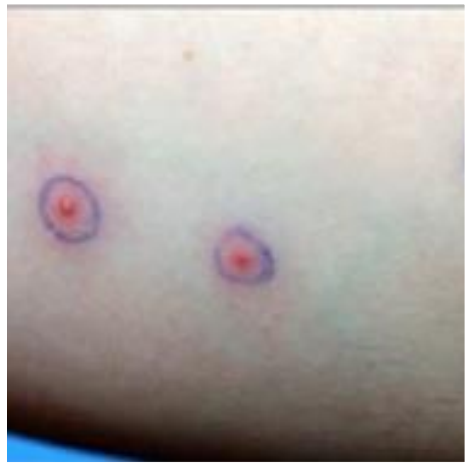


Vascular -V



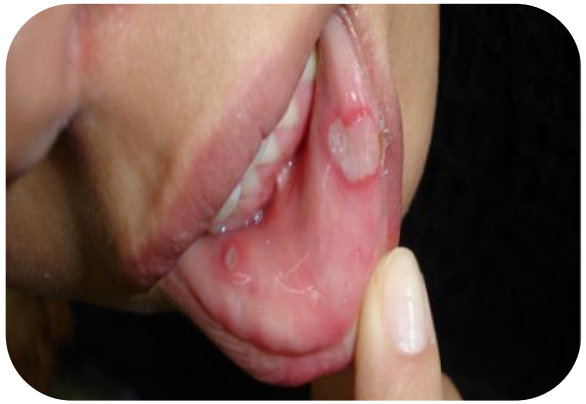
Hasan Yazici1 et al; Behçet syndrome: a contemporary view. NATURE REVIEWS | RHEUMATOLOGY

Manifestaciones mucocutáneas

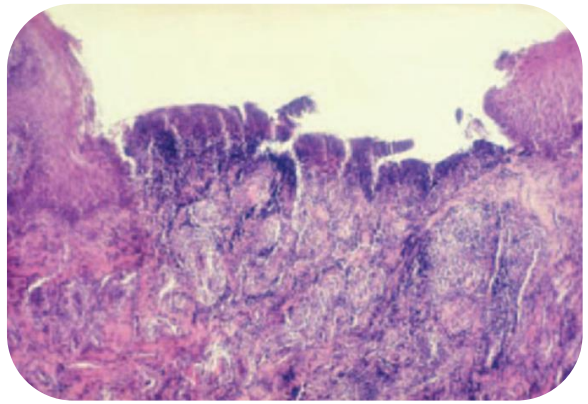







Alpsoy E. Behçet's disease: a comprehensive review with a focus on epidemiology, etiology and clinical features, and management of mucocutaneous lesions. *The Journal of Dermatology*. 2016;43(6):620–632

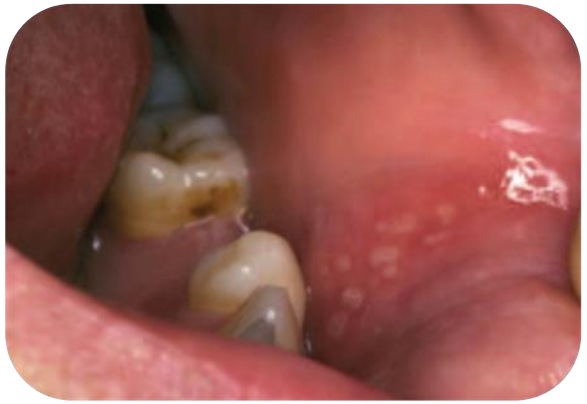
Úlceras orales



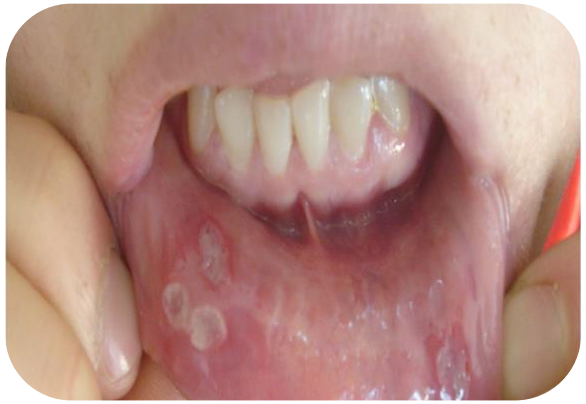
Inicial *



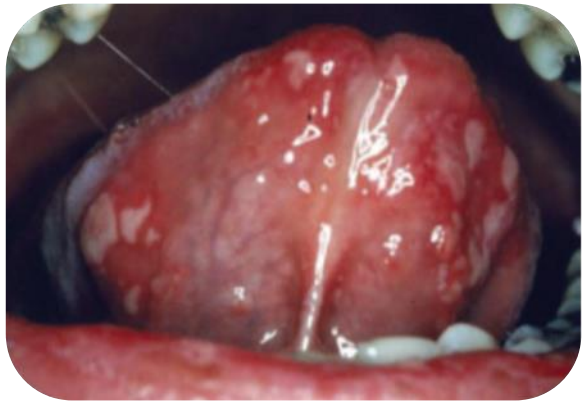
Similar a estomatitis aftosa recurrente



Múltiples y extensas / Dolorosas*



Pocos mm – 2 cm

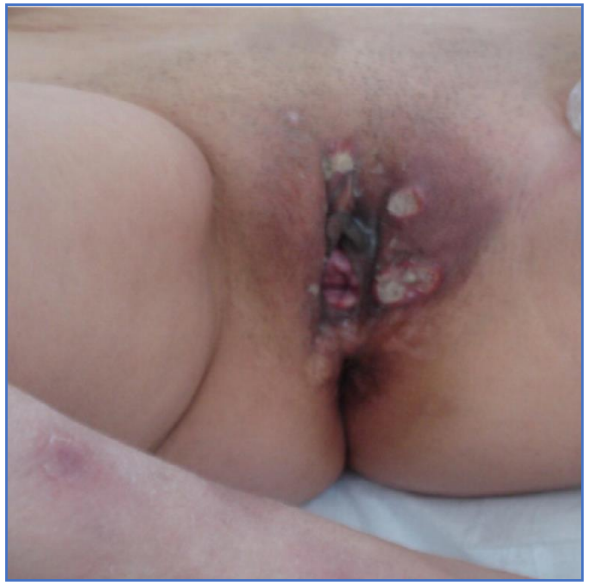


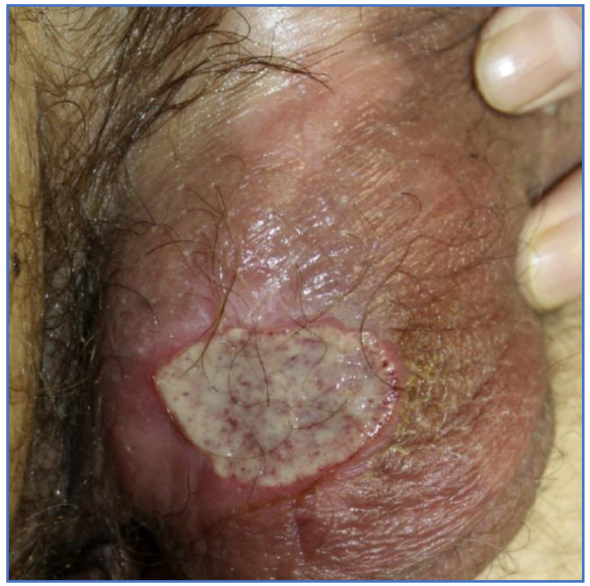
Menores < 1 cm > mayores*

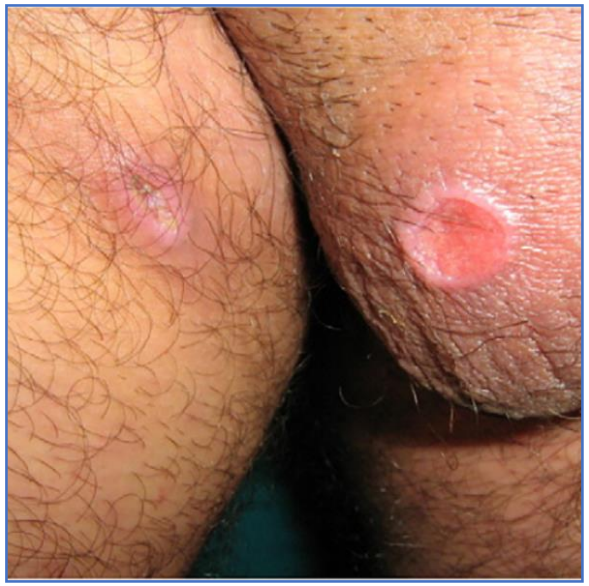
Smith EL, Yazici Y. Clinical manifestations and diagnosis of Behçet's syndrome. Uptodate 2016

Úlceras genitales

Lesión mas específica
Escroto / vulva
Dolorosas*
Testosterona-Neutrófilos
Cicatriz común
Epididimitis, salpingitis, varicocele*

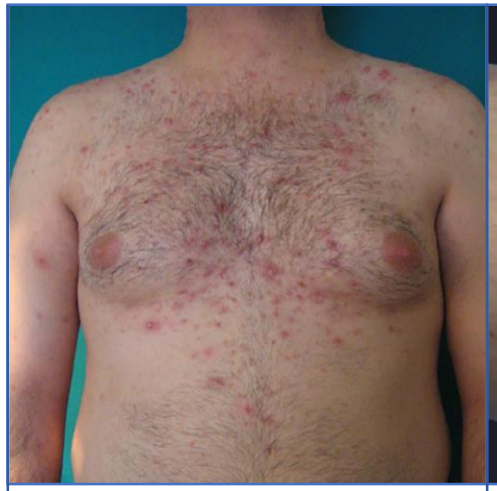
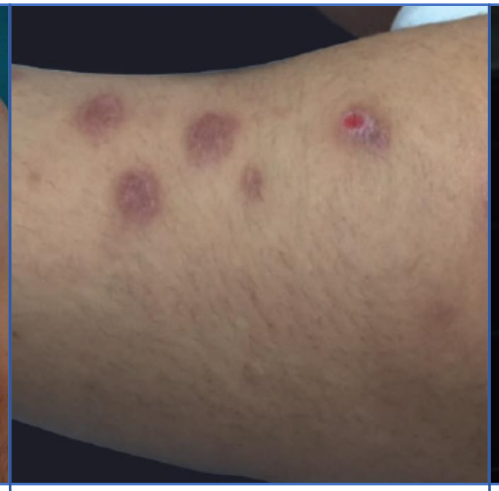
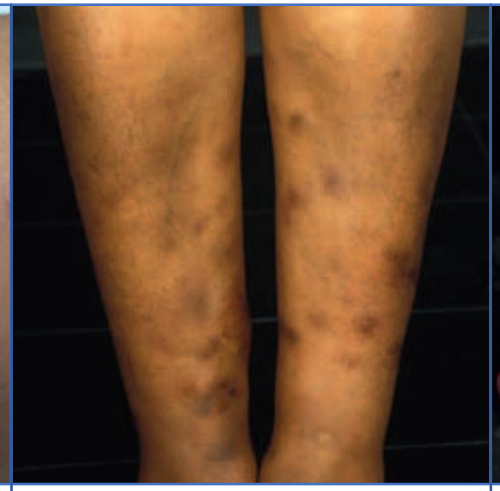
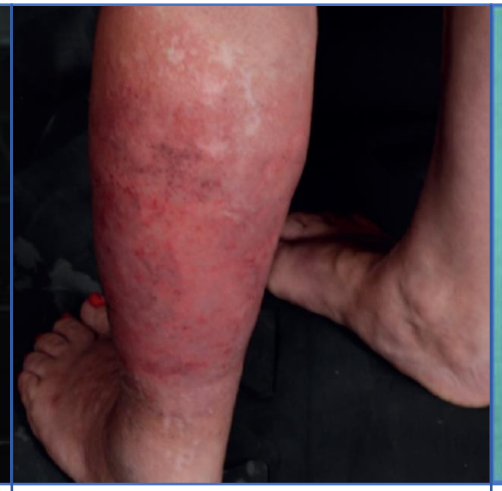
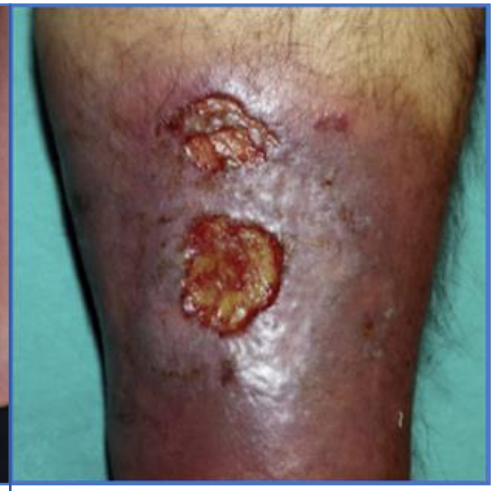






Smith EL, Yazici Y. Clinical manifestations and diagnosis of Behçet's syndrome. Uptodate 2016

Lesiones cutáneas

				
Reacciones acneiformes*	Pseudofoliculitis	Eritema nodoso**	Tromboflebitis superficial	Ulcus cruris

Erupciones pápula-vesícula-pústula

Nódulos

Eritema multiforme like

Púrpura palpable

Smith EL, Yazici Y. Clinical manifestations and diagnosis of Behçet's syndrome. Uptodate 2016

Diagnóstico

Características de mayor peso al diagnóstico

Elementos fuertes

- Úlceras orales
- Enfermedad ocular
- Úlceras genitales
- Compromiso vascular mayor
- Enfermedad neurológica parénquima

Elementos débiles

- Variación geográfica en expresión de enfermedad
- Asociación con E. Crohn
- Distinta presentación de enfermedad
- Respuesta diversa a varios medicamentos

Hasan Yazici1 et al; Behçet syndrome: a contemporary view. NATURE REVIEWS | RHEUMATOLOGY doi:10.1038/nrrheum.2017.208

Criterios diagnósticos

ISG criteria for the diagnosis of Behçet disease[80]	
Recurrent oral ulceration:	Minor aphthous, major aphthous, or herpetiform ulcers observed by the physician or patient, which have recurred at least three times over a 12-month period
Plus any 2 of the following:	
Recurrent genital ulceration:	Aphthous ulceration or scarring observed by the physician or patient
Eye lesions:	Anterior uveitis, posterior uveitis, or cells in the vitreous on slit-lamp examination; or retinal vasculitis detected by an ophthalmologist
Skin lesions:	Erythema nodosum observed by the physician or patient, pseudofolliculitis, or papulopustular lesions; or acneiform nodules observed by the physician in postadolescent
Positive pathergy test:	Test interpreted as positive by the physician at 24-48 hr
These criteria are valid in the absence of other clinical explanation	

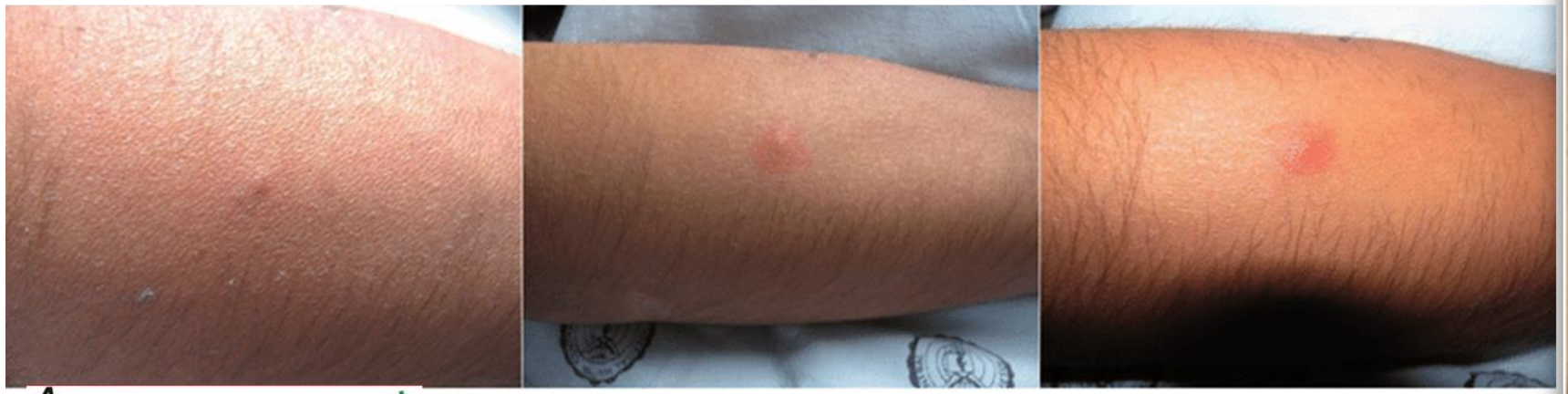
International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. Lancet. 1990;335:1078-1080

El diagnóstico es eminentemente clínico

Table 5 International Criteria for Behçet's Disease – point score system: scoring ≥ 4 indicates Behçet's diagnosis

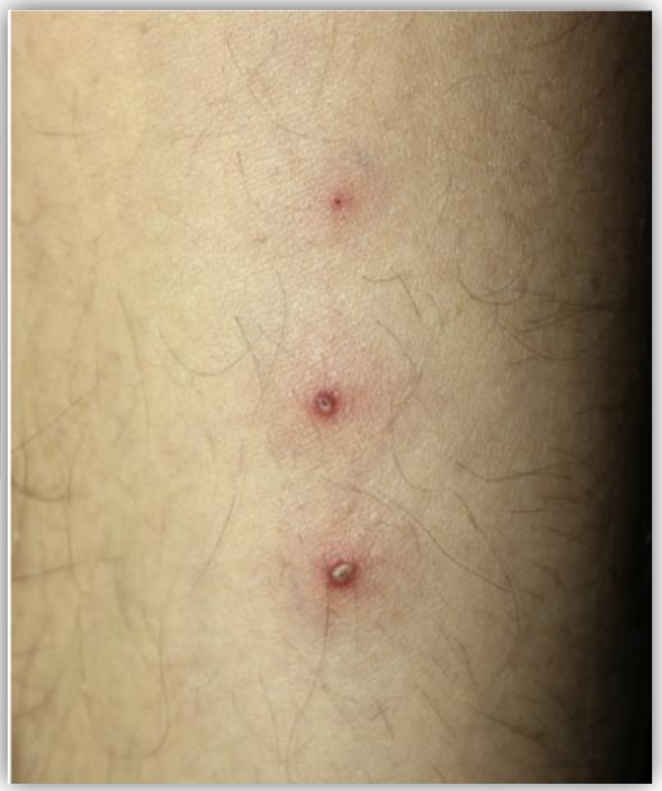
Sign/symptom	Points
Ocular lesions	2
Genital aphthosis	2
Oral aphthosis	2
Skin lesions	1
Neurological manifestations	1
Vascular manifestations	1
Positive pathergy test*	1*

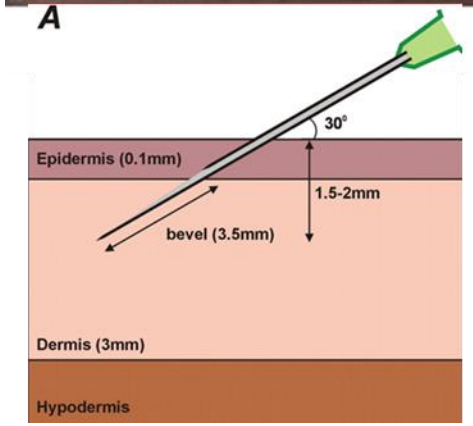
The International Criteria for Behçet's Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. J Eur Acad Dermatol Venereol. 2014 Mar; 28(3): 338–347

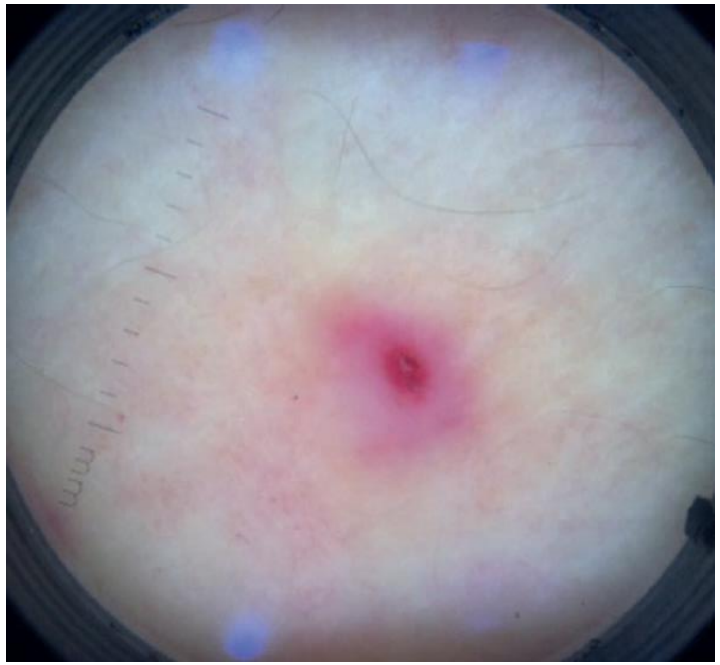


24 horas

48 horas

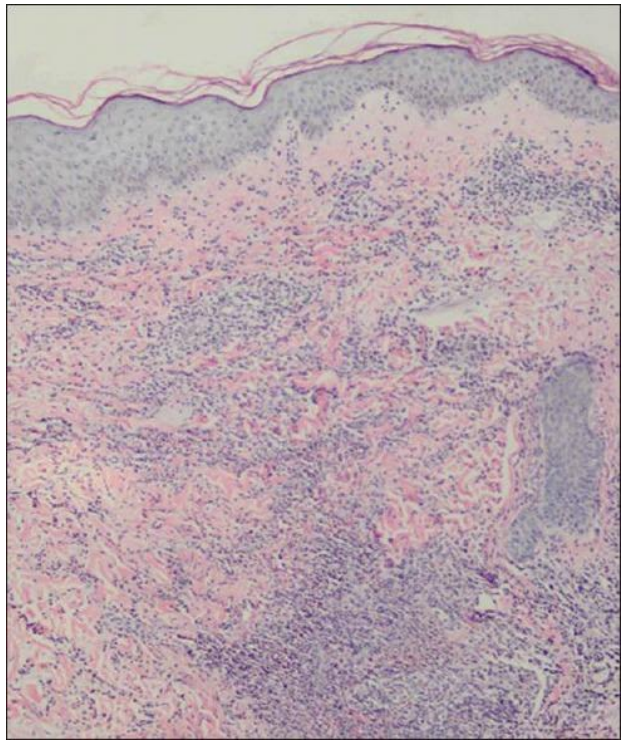
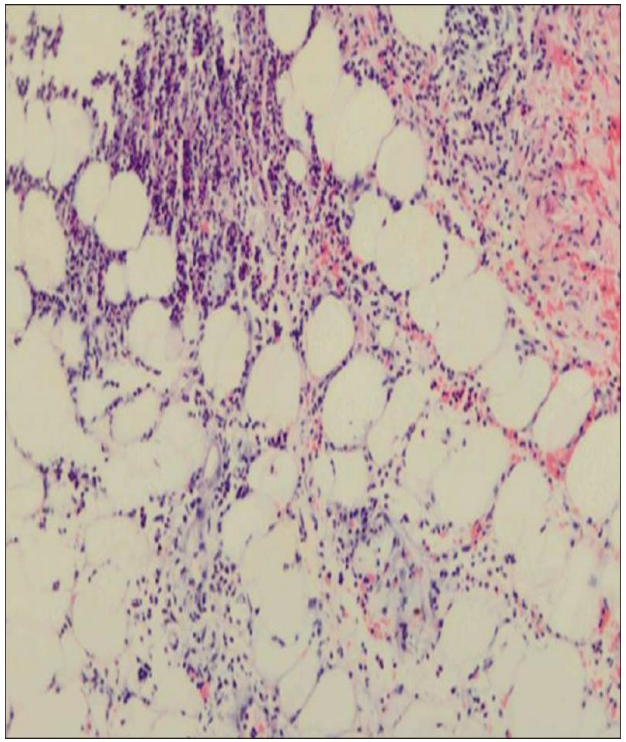
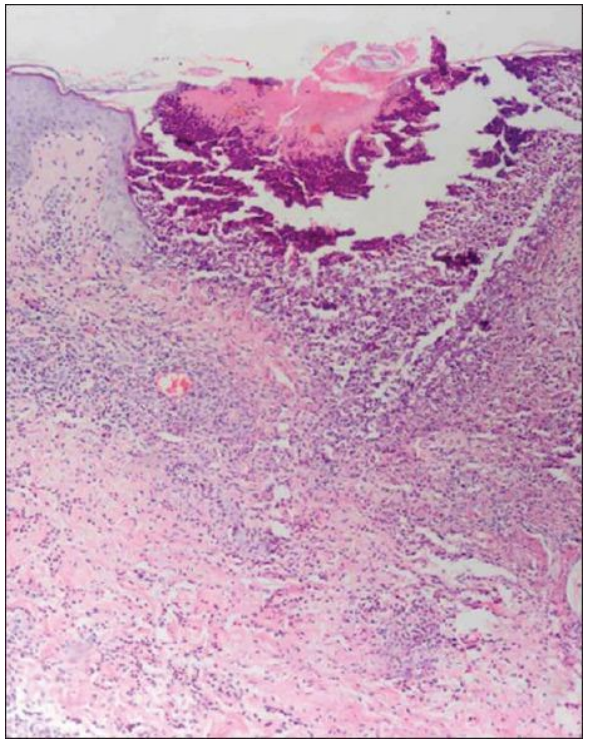
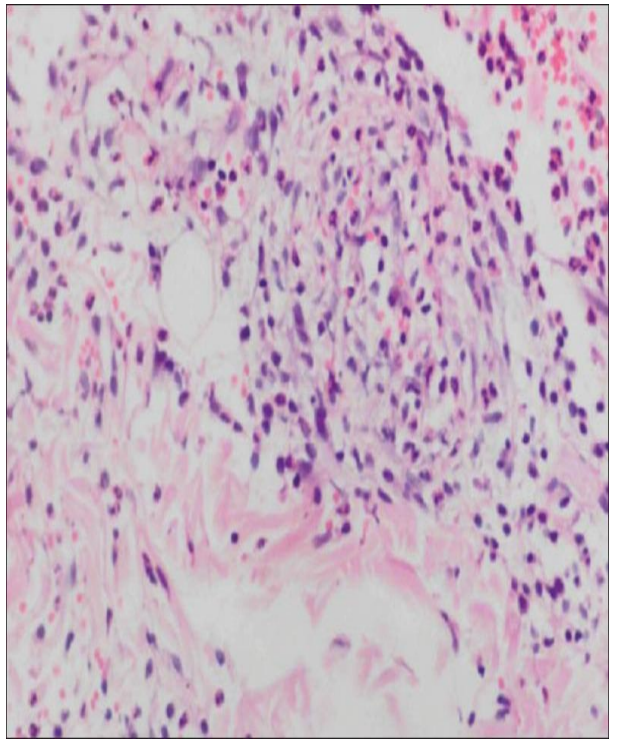






Test de patergia

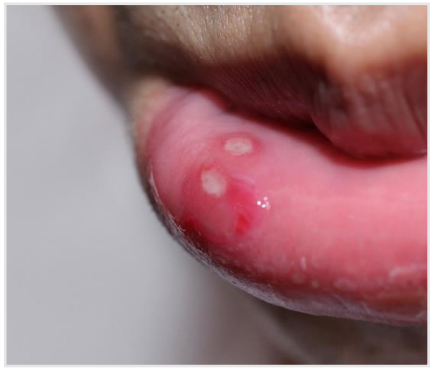
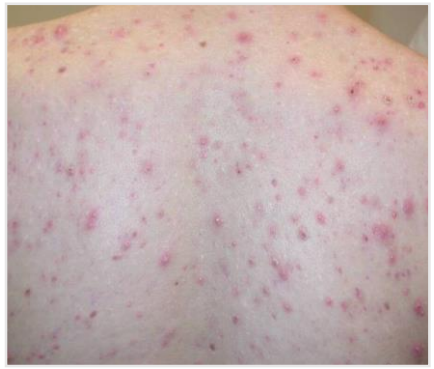
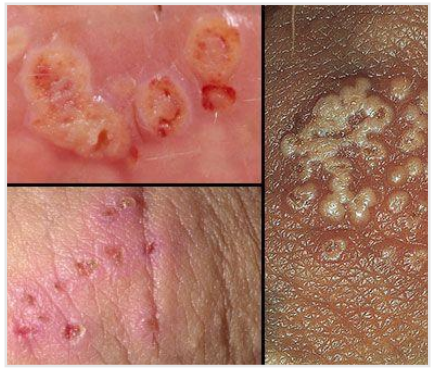
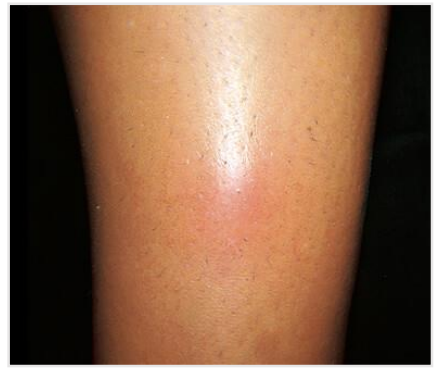
Pimienta M, Olivé A. El fenómeno de patergia. Semin Fund Esp Reumatol. 2009;10(3):87-90

Infiltrado inflamatorio mixto en dermis → Paniculitis lobular sin vasculitis → Pústula epidérmica → Vasculitis leucocitoclástica

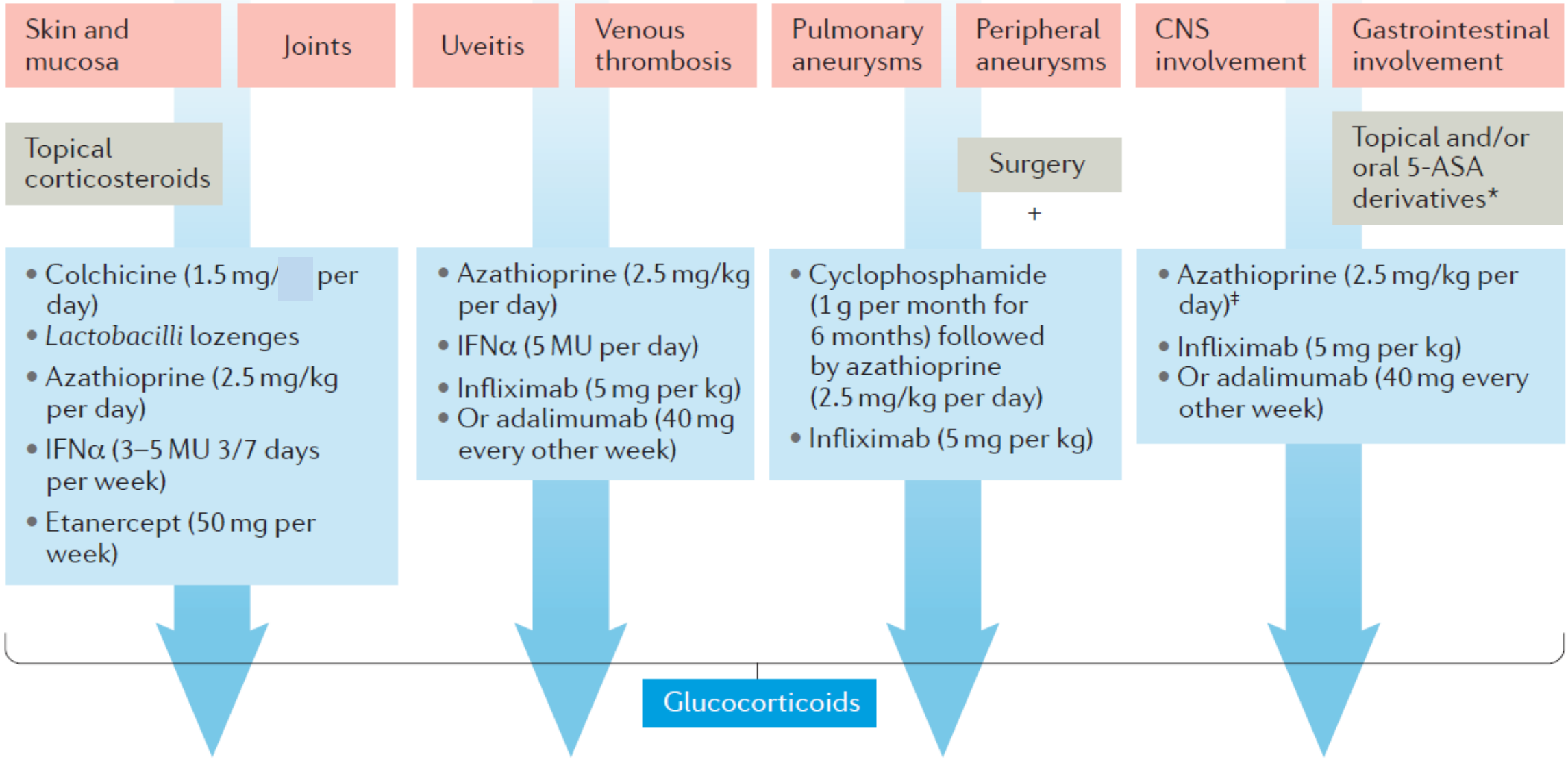
Ozluk E, Balta I, Akoguz O, et al. Histopathologic Study of Pathergy Test in Behçet's Disease. *Indian Journal of Dermatology*. 2014;59(6)

Diagnóstico diferencial

Úlceras orales	Lesiones pápulopustulares	Úlceras genitales	Eritema nodoso
			
Estomatitis aftosa recurrente	Acné vulgar	Artritis reactiva Herpes ITS	Otros
Dolor Frecuentes Múltiples	Tórax superior y extremidades > 40 años	Cicatriz Escroto-vulva	Múltiples Dolorosas Recurrentes Áreas atípicas Pigmentación residual Vasculitis en histopatología

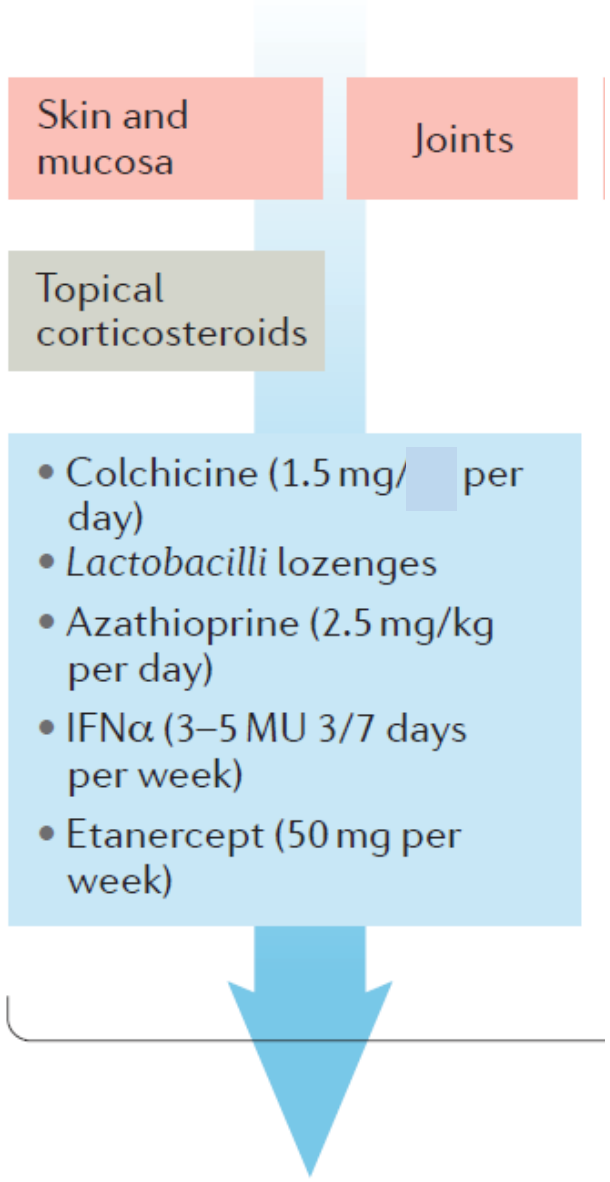
Smith EL, Yazici Y. Clinical manifestations and diagnosis of Behçet's syndrome. Uptodate 2016

Tratamiento



Hasan Yazici1 et al; Behçet syndrome: a contemporary view. NATURE REVIEWS | RHEUMATOLOGY

Tratamiento



Acetonida de triamcinolona 0,1% (orobase)

Úlceras orales

Corticoesteroides tópicos alta potencia
Úlceras genitales

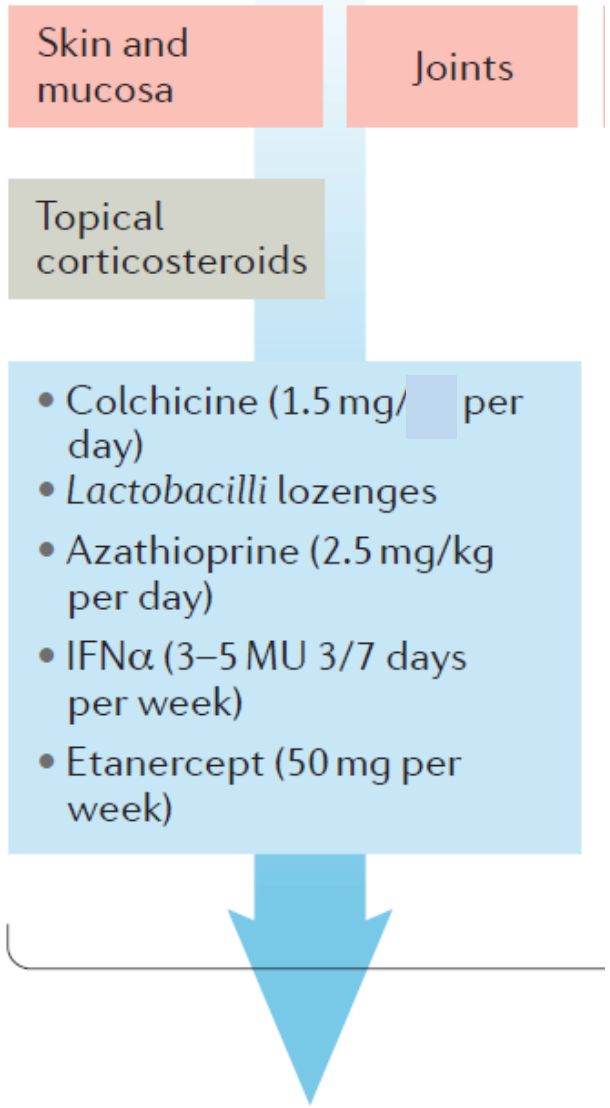
Triamcinolona intralesional (5 a 10 mg / ml)

*Úlceras grandes *

Sucralfato tópico 1g / 5mL

Smith EL, Yazici Y. Treatment of Behçet syndrome. Uptodate 2018

Tratamiento



Colchicina de 1 a 2 mg / día

Prevención de úlceras orales y genitales recurrentes

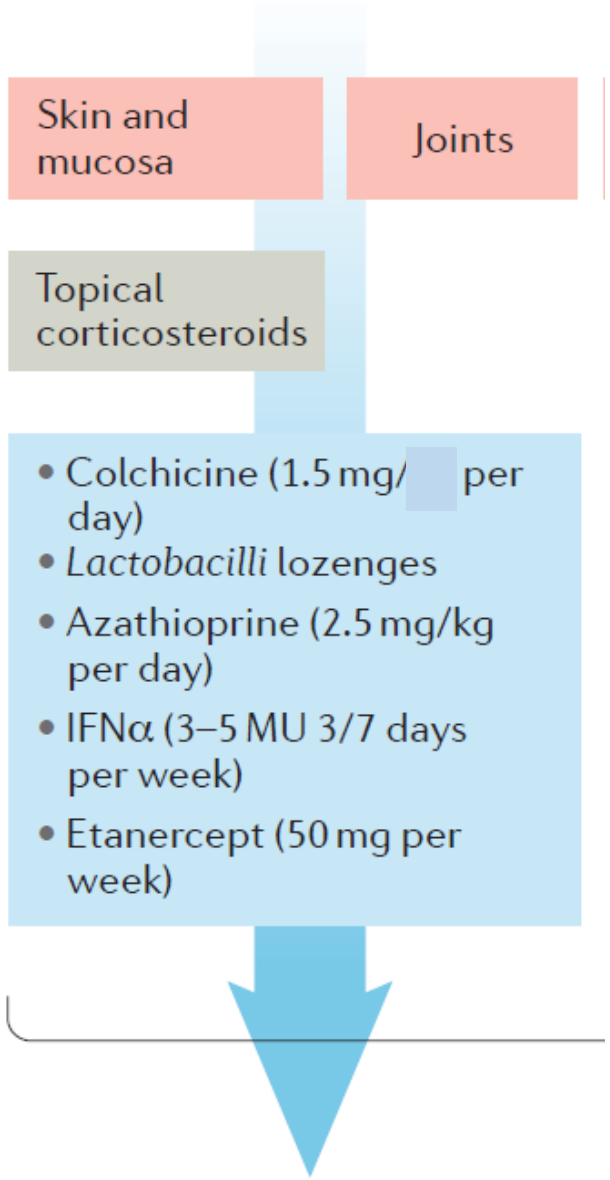
Pseudofoliculitis, eritema nodoso, artritis

Glucocorticoides sistémicos (prednisona 15 mg / día)

Aftas orales o úlceras genitales refractarias a corticosteroides tópicos y la colchicina o lesiones múltiples

Smith EL, Yazici Y. Treatment of Behçet syndrome. Uptodate 2018

Tratamiento



Azatioprina - ha demostrado mejorar la ulceración oral y genital

Interferón alfa : duración y dolor de úlceras orales, frecuencia de genitales y lesiones pápulopustulares

Anti TNF : en combinación con DMARD (azatioprina)*

Glucocorticoides sistémicos y otros medicamentos inmunosupresores: Eritema nodoso (posibilidad de vasculitis subyacente)

Smith EL, Yazici Y. Treatment of Behçet syndrome. Uptodate 2018

Conclusiones

Desorden inflamatorio multisistémico

Genética

Disparadores ambientales

Respuesta Inmune Innata y Adaptativa

Más de un mecanismo fisiopatogenético

Manifestaciones clínicas variables

Diagnostico clínico

Tratamiento multidisciplinario

Gracias