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LETTER TO THE EDITOR

Regarding the naming dilemma of Behçet disease in the 21st century

Dear Editor,

I have read the very recent review of Escudier *et al* (*Oral Dis* 2006;**12**:78–84) titled 'Number VII Behçet's disease (Adamantiades syndrome)' with great interest. The authors evaluated the history of Behçet's disease along with its epidemiology, etiopathogenesis, clinical features, complications, diagnosis and management.

However, as a 'Behçetologist' with a total of 28 published articles on 'Behçet's disease' in international peer-reviewed medical journals, I was very disappointed to see that the disease was named '(Adamantiades syndrome)' in the title and in the first line of the 'Abstract' section of the paper, as if this eponym is used as a synonym for 'Behçet syndrome' or as an alternative naming to 'BS'. The most important part of an article is, undoubtedly, its title as every investigator first reads this during a PubMed search, which is followed by the reading of the 'Abstract' section. Therefore, I think we, as researchers, should be very attentive and careful while creating article titles.

Since its first acceptance as 'Morbus Behçet' during the International Medical Congress of Geneva in Switzerland (1947) according to the suggestion of Prof. Mischner of the Zurich Medical Faculty (Dilşen, 1996; Saylan, 1997), 'Behçet disease' or 'Behçet syndrome' has been the focus of active research and, to our knowledge, there are 5929 articles and tens of thousands of citations in the medical literature up to September 15, 2006, in a simple search using the database of NLM that employed the eponym 'Behçet'. Indeed, the disorder has been well known as 'Behçet disease' for more than 65 years by every physician interested in Behçet disease care in both Eastern and Western populations from every country, as well as by medical students and the public. However, and more importantly, although there are some 88 other articles that suggested 'double naming' such as 'Adamantiades–Behçet disease' in the titles or texts of the papers, this unique disorder has never been titled since 1941 as '(Adamantiades syndrome)' alone. Therefore, I would be very appreciative if the authors would provide their evidence to the scientific community, and clearly and strongly prove it by references.

The previous publications of the authors clearly disclose the usage of 'Behçet' in at least five of their publications on the same subject during the last two decades from the beginning of 1982 (Scully, 1982, 1989; Scully *et al*, 1982, 1991; Mutlu and Scully, 1994). The act of using the name 'Adamantiades syndrome' indicates that the authors express a biased manner with deficient knowledge of the whole and true history of the origin of 'Behçet's disease', that needs extensive revision and, therefore, important historical and scientific corrections, as the review article of Escudier *et al* is probably going to be a reference paper in the coming years for us and for our students. In addition, the corresponding author has presented a previous paper with the eponym 'Behçet' indicating '*The person behind the eponym: Hulusi Behçet*' in the title of the publication in 1994 (Mutlu and Scully, 1994). But still, if the respected and experienced authors of the present review article believe and insist on the contribution of Dr Benedictos Adamantiades in the invention of this unique vasculitic disorder, I think it is time to report the whole historical evolvement of manifestations excellently enough to draw such a conclusion.

It is well known that one or several individual symptoms or signs of Behçet disease have been reported by individual cases or case reports before Dr Hulusi Behçet that spans probably to the Hippocratic writings dated 460–377 BC as Escudier *et al* also indicated (Evereklioglu, 2006).

There were other forms of fever…, many had their mouths affected with aphthous ulcerations. There were also many defluxions about the genital parts, and ulcerations, boils (phymata), externally and internally about the groins. Watery ophthalmies of a chronic character, with pains; fungous excretions of the eyelids externally and internally, called fici, which destroyed the sight of many persons.

However, let us now evaluate the publications of several faithful authors in the literature during the last 300 years from the beginning of the 18th century up to the single case of Dr Benedictos Adamantiades who presented (Adamantiades, 1930) and then reported (Adamantiades, 1931) a case with 'relapsing hypopyon iritis' associated with mucocutaneous lesions and arthritis, which focused on the 'recurrent ocular lesions' – that is the basis of the suggestion of 'Adamantiades–Behçet' by a few authors today. However, Dr Adamantiades was not the first to report such 'recurrent eye lesions', mucocutaneous symptoms and arthritis.

In the 18th century, for instance, Janin (1772) from France published a case with 'recurrent hypopyon iritis' in a male patient. One century later, Neumann (1895) from Austria and Christlieb (1895) from Würzburg independently reported a total of 12 female patients with recurrent mucocutaneous lesions consisting of orogenital aphthous ulcerations.

In the beginning of the 20th century, Reis (1906) from Germany presented a case with 'relapsing ocular inflammation' associated with cutaneous lesions (erythema

nodosum) and arthritis in a male patient. Two years later, in 1908, Blüthe (1908) from Germany reported a total of four patients from either sex with identical findings, to the single case of Dr Adamantiades, consisting of 'relapsing hypopyon–iridocyclitis', mucocutaneous lesions and arthritis with histological evidence of uveitis and optic nerve atrophy. Later Gilbert (1920, 1921, 1923, 1925) from the same country as Blüthe published patients with recurrent hypopyon iridocyclitis, recurrent arthritis, skin lesions and histological detection of uveitis with the suspicion of staphylococcal sepsis, indicating the disease as 'iritis or iridocyclitis septica' (Gilbert, 1920, 1921, 1923) and 'ophthalmia lenta' (Gilbert, 1925). It is also very clear that the symptoms and signs of Dr Gilbert's cases (1920–1925) are completely identical to the single case of Dr Adamantiades (1930–1931).

From Austria, Planner and Remenowsky (1922) reported a female case with iritis and genital lesions. One year later, Weve (1923) from the Netherlands published about a woman with recurrent hypopyon–iridocyclitis, arthritis, mucocutaneous lesions, periodontitis and neurological signs with suspicion of a staphylococcal infection: once more exactly the same symptom constellation as Dr Adamantiades' patient. Shigeta (1924) from Japan reported a man with recurrent mucocutaneous ulcers and hypopyon iritis with histological detection of uveitis and optic nerve atrophy. Similarly, Pils (1925) from Austria reported a woman with mucocutaneous lesions, thrombophlebitis and arthralgia. Grütz (1926) from Kiel and Carol and Ruys (1928) from the Netherlands published about a total of two female patients with recurrent genital lesions and arhtralgia. One year later, Samek and Fischer (1929) from the Czech Republic reported a case with recurrent mucocutaneous lesions and erythema nodosum with the first use of the pathergy test.

From Poland, Walter and Roman (1930) reported recurrent mucocutaneous lesions in two female patients with histologically demonstrated leukocytoclastic vasculitis in a genital ulcer and a skin lesion. Similarly, from Innsbruck, Kumer (1930) reported mucocutaneous lesions in a female patient with a histological detection of leukocytoclastic vasculitis in a genital ulcer and an erythema nodosum lesion with some central nervous system signs. In the following years, Dr Benedictos Adamantiades from Greece presented and then reported a case with recurrent hypopyon iritis, mucocutaneous symptoms and arthritis (*Sur un cas d'iritis à hypopyon récidivante*) (Adamantiades, 1930, 1931). In his paper, Dr Adamantiades indicated that 'recurrent hypopyon iritis' (but not the triple symptom complex of Behçet) constitutes itself as a distinct clinical entity. Indeed, Dr Adamantiades, as an ophthalmologist, further published more and more papers on 'recurrent ocular lesions', before and after the worldwide recognition and acceptance of the disease as 'Morbus Behçet' in 1947, and insistently concentrated again on 'relapsing iritis with hypopyon' during the following years. He published his cases both in French in 1946 (Adamantiades, 1946) (*La thrombophlébite comme quatrième symptôme de l'iritis récidivante à hypopyon*) and 1953 (Adamantiades, 1953) (*Le symptôme complexe de l'uvéite récidivante à hypopyon*) as well as in Greek with an English title in 1958 (Adamantiades, 1958) (*Severe complications of the central nervous system in the syndrome of relapsing iritis with hypopyon*).

However, 'relapsing ocular lesions' (in other words, 'l'iritis récidivante à hypopyon' or 'l'uvéite récidivante à hypopyon') may occur not only in many ocular or non-ocular diseases, but also in various infectious or non-infectious systemic disorders. Among these diseases, for instance, tuberculosis, syphilis, leprosy, various vasculitides and some other endogenous uveitides as well as staphylococcal bacteremia or sepsis can be listed here as some of the etiological factors. Indeed, Dr Adamantiades first thought of the role of syphilis, tuberculosis and bacteremia in the etiology and his patient received antisyphilitic treatment, although the disease was not cured, resulting in the assumption of bacterial staphylococcal infection or focal illness by Dr Adamantiades (Dilşen, 1996; Saylan, 1997). Because Reis (1906); Gilbert (1920) and Weve (1923) also reported very closely identical cases and backed the hypothesis of a bacterial, staphylococcal focal illness, Dr Adamantiades cited these papers in his article. Therefore, as the International Behçet's Society of UK indicated, the report of Dr Adamantiades was not the first to describe these manifestations as 'triple symptom complex' and, more importantly, he did not recognize the true nature of the disease, and was not able to describe it as a distinct entity or syndrome, namely as a 'classical triad' consisting of recurrent oral aphthae, genital ulcers and hypopyon uveitis (Dilşen, 1996; Saylan, 1997; Giannoukas, 2005; Evereklioglu, 2006).

Taken together, 'recurrent iritis or iridocyclitis' (a major criterion for the diagnosis of Behçet disease in both 'Japanese Behçet's Disease Research Committee Criteria' (The Behçet's Disease Research Committee of Japan, 1989) and 'International Study Group Criteria' (International Study Group for Behçet's Disease, 1990) was reported not only by Adamantiades (1931) but also by Janin, Reis, Blüthe, Gilbert, Planner–Remenowsky, Weve and Shigeta in their publications between 1772 and 1924, many of which also reported recurrent mucocutaneous lesions (oral aphthae and genital ulcers) and/or arthritis just as Adamantiades did. In addition, Dascalopoulos (1932); Whitwell (1934); Nishimura (1936); Blobner (1937); Weekers and Reginster (1938a,b) and Knapp (1938) further reported very similar cases with 'recurrent ocular lesions' between 1932 and 1938.

Similarly, if taken together, recurrent oro-mucocutaneous lesions (still major criteria for the diagnosis of Behçet disease in both classifications) (The Behçet's Disease Research Committee of Japan, 1989; International Study Group for Behçet's Disease, 1990) have been reported not only by Adamantiades (1931) but also by Neumann, Christlieb, Reis, Blüthe, Gilbert, Planner–Remenowsky, Weve, Shigeta, Pils, Grütz, Carol–Ruys, Samek–Fischer, Walter–Roman and Kumer between 1895 and 1930 and even after Dr Adamantiades by Matras (1932), Dascalopoulos, Whitwell, Nishimura, Weekers–Reginster and Knapp between 1932 and 1938.

Likewise, erythema nodosum (still a major criterion for the diagnosis of Behçet disease in both classifications) (The Behçet's Disease Research Committee of Japan, 1989; International Study Group for Behçet's Disease, 1990) was reported by Kumer and Blobner, whereas neurologic signs (a minor criterion of Japanese Behçet's disease Research Committee) (The Behçet's Disease Research Committee of Japan, 1989) with or without optic atrophy were reported by Weve, Blüthe, Shigeta, Adamantiades, Blobner and Knapp. Moreover, arthritis and/or orchitis (two minor criteria of Japanese Behçet's disease Research Committee) (The Behçet's Disease Research Committee of Japan, 1989) have been reported by Reis, Blüthe, Weve, Shigeta, Carol–Ruys, Adamantiades, Matras and Blobner. Furthermore, the pathergy test (a major criterion of International Study Group) (International Study Group for Behçet's Disease, 1990) was first used by Samek and Fisher in 1929 on a female patient and afterwards by Blobner on a male patient in 1937.

However, all these physicians cited thus far, like Adamantiades, ascribed the symptoms or findings they presented, either to another disease such as tuberculosis, syphilis, sepsis and allergy, or to a coincidence, and none of them indicated 'a new or a single syndrome' with a 'classical triad' consisting of a 'triple symptom complex'.

Having presented all the chronological and historical evidences in the development of the publications, let us see what Dr Hulusi Behçet did step by step for the invention of this novel disease. Dr Behçet was the first author who recognized the characteristic symptom and sign constellation, and grouped all the manifestations (recurrent oral aphthae, genital ulcerations, recurrent hypopyon uveitis) himself into a single disease first in 1937 *(Behçet H (1937). Über rezidivierende, Aphthöse, durch ein Virus verursachte Geschwüre am Mund, am Auge und an den Genitalien. Dermatol Wochenschr* 105: 1152–1157), and then described the results in 1939 in German as 'Tri–Symptomenkomplex' (*Behçet H (1939). Einige Bemerkungen zu meinen Beobachtungen über den Tri–Symptomenkomplex. Med Welt* 13: 1222–1227) and afterwards once more in 1940, in English, as 'triple symptom complex' (*Behçet H (1940). Some observations on the clinical picture of the so-called triple symptom complex. Dermatologica* 81: 73–83), clearly and strongly indicating an association between three unrelated manifestations unquestionably together as 'a new, specific and completely separate clinical entity or syndrome' (Dilşen, 1996; Saylan, 1997; Giannoukas, 2005; Evereklioglu, 2006).

Following these publications by Dr Hulusi Behçet, many authors from different racial and ethnic origins both in Eastern and Western populations started to report such puzzling cases with a so-called 'classical triad' of symptoms (namely dermatologic, ophthalmic and oro-genital lesions). Among these authors, for instance, Jensen from Denmark (1941) first used the eponym 'Behçet syndrome' in the title of the paper to describe the 'triple symptom complex' (*Jensen T (1941). Sur les ulcérations aphteuses de la muqueuse de la bouche et de la peau génitale combinées avec les symptômes oculaires (=Syndrome Behçet). Acta Dermatol Venereol* 22: 64–79). Three years later, we encountered two important publications. The one from Berlin used the eponym 'Behçet's syndrome' in the title of the case report to indicate the components of 'triple symptom complex' as 'mouth, genital and eye lesions' (*Berlin C (1944). Behçet's syndrome with involvement of central nervous system. Report of a case, with necropsy, of lesions of the mouth, genitalia and eyes; review of the literature. Arch Dermatol Syphil (Chicago)* 49: 227–233) and another from Ephraim (Israel) who clearly indicated again 'triple symptom complex of Behçet' in the title of the publication (*Ephraim H (1944). Triple symptom complex of Behçet. Arch Dermatol Syphil (Chicago)* 50: 37–38). Two years later in 1946, Ollendorff Curth from the USA published two different papers on American patients and reported the abortive form of Behçet's syndrome *(Ollendorff Curth H (1946). Behçet's syndrome, abortive form (?) (recurrent genital ulcerations). Arch Dermatol Syphil (Chicago)* 54: 481–483) and then published the 'classical triad' of Dr Hulusi Behçet in the title of the paper as 'orogenital ulcers with hypopyon uveitis', indicating once more 'Behçet syndrome' *(Ollendorff Curth H (1946). Recurrent genito–oral aphthosis and uveitis with hypopyon (Behçet's syndrome). Arch Dermatol Syphil (Chicago)* 54: 179–196), which resulted in the popularization of 'Behçet's syndrome' among every country that honors the first describer of the 'triple symptom complex'. Finally, the eponym 'Behçet's disease' was first used and titled by Feigenbaum and Kornblueth in the same year to report 4 such cases (*Feigenbaum A, Kornblueth W. (1946) Behçet's disease as manifestation of a chronic septic condition connected with a constitutional disorder. With a report of 4 cases. Acta Med Orient 5:139–151*).

Dr Hulusi Behçet, as a dermatologist, placed particular importance on the recurrent oral ulcerations (aphthosis) that are today the 'universal hallmark' and the only *sine qua non* symptom of this unique disorder according to the International Study Group Criteria held in the UK, for the diagnosis of Behçet's disease (but not 'recurrent iridocyclitis' of Dr Adamantiades) (International Study Group for Behçet's Disease, 1990). In turn, 'recurrent iridocyclitis with or without hypopyon uveitis', stressed by Dr Adamantiades from the ophthalmology perspective as a clinical entity, is encountered in approximately half of Behçet patients and the diagnosis of Behçet disease can still be made even if the patient has no such ocular involvement.

In light of both the knowledge of positive sciences stated above, over the centuries, with evidence-based historical articles and the statements of the reviewers of the present paper in their article as, 'it was Behçet who described the classical clinical triad of oral and genital ulceration with ocular inflammation, *(see introduction section please, second paragraph, last 3 lines)*, I completely disagree with Escudier *et al* and, therefore, strongly reject their non–referenced assumption that the other name of the disease is 'Adamantiades syndrome' as it stands in the title and in the first line of the abstract. We, as scientists, should be aware of the deeper historical realities in light of the aforesaid evidences and,

as researchers, must obey or follow the instructions of 'American Behçet's Disease Association' and 'International Behçet's Society' that acknowledge the origin of Behçet disease in their official web sites.

If credit is to be given to those who described monosymptomatic or oligosymptomatic findings of this disorder after more than 2000 years have lapsed since the Hippocratic writings, albeit attributed the manifestations to other etiologic local or systemic diseases, the entity would be fully named in that cases as 'Hippocrates–…–Janin–Neumann–Reis–Blüthe–Gilbert–Planner–Remenovsky–Weve–Shigeta–Pils–Grütz–Carol–Ruys–Samek–Fischer–Walter–Roman–Kumer–Adamantiades–Dascal opoulos–Matras–Whitwell–Nishimura–Blobner–Weekers–Reginster–Knapp–Behçet disease', although several others have remained still unnamed. Therefore, Escudier *et al* should clearly answer the most important question as to why only Dr Benedictos Adamantiades among all these aforesaid esteemed authors should be honored and term the disease as 'Adamantiades syndrome'? If Dr Adamantiades had really worried about the use of the eponym 'Behçet' in the titles of the articles in 20th century, he might himself have written at least a 'letter to the editor' not only to these international medical journals, but also to the authors who named the disease first as 'Behçet syndrome' (Jensen, Berlin, Ollendorff Curth), 'Behçet disease' (Feigenbaum, Kornblueth) or 'triple symptom complex of Behçet' (Ephraim), as well as to both the International Dermatology Society who honored the first describer of 'Tri–Symptomenkomplex/Triple Symptom Complex' and to Prof. Mischner of the Zurich Medical Faculty who suggested 'Morbus Behçet' to credit the first describer of 'classical triad' in 1947, just like I did today (Dilşen, 1996; Saylan, 1997). In direct contradiction, we know for certain today that Dr Adamantiades did not take any efforts in this regard and, therefore, did not write a letter to any physician or journal because he had already recognized that it was Dr Hulusi Behçet who had indicated a new and a separate syndrome with 'triple symptom complex'. This we can easily understand from the title of one of Dr Adamantiades' subsequent papers, published 12 years after the original report of Dr Hulusi Behçet and one year later after Dr Behçet's death (*Adamantiades B*, *Lorando N (1949)*. *Sur le syndrome complexe de uvéite récidivante ou soi–distant syndrome complexe de Behçet*. *Presse Med* 57: 501).

All the classical Textbooks of Dermatology, Rheumatology and Ophthalmology and any other sections or subheadings of medical books title this entity as 'Behçet disease' or 'Behçet syndrome', not as 'Adamantiades–Behçet disease' and, more importantly, never as 'Adamantiades syndrome' (Behçet's Disease Books and Resources: http://www.behcetsdisease.com/order.htm). Likewise, countless events like 'Korea–Turkey Behçet Days' take place and international symposiums, congresses, courses and conferences use the eponym 'Behçet', not 'Adamantiades syndrome'. Furthermore, the American Behçet's Disease Association (ABDA: http://www.behcets.com/site/pp.asp?c=bhJIJSOCJrH&b=260523), International Society for Behçet's Disease (ISBD) of UK (http://www.behcet.ws/ and http://www.behcets.org.uk/), The Behçet's Disease Research Committee of Japan (1989); International Study Group for Behçet's Disease (1990), French Behçet's Association (http://www.association_behcet.org/english.htm), Italian Behçet's Disease Association (http://www.behcet.it/), Korean Behçet's Association (http://www.behcet.co.kr/), Behçet Israel Group (BIG) (http://www.behcet.org.il/) and finally Turkish Ophthalmology Behçet Society (TOD) (http://www.tod–net.org/v3/html/todnet.asp?a=uve3|) strongly suggest and use the eponym 'Behçet', never 'Adamantiades syndrome'.

Authors and experienced reviewers of respected journals should follow the knowledge of positive sciences and should obey the suggestions and criteria of their International Research Groups as well as the Societies, Associations and the Committees, which name the disease 'Behçet disease' or 'Behçet syndrome'. Therefore, I think we should choose our terms very selectively in the titles of articles for the definition of established diseases and it is time to look ahead as emphasized in the 'Conclusion and Future Directions' section at the end of our major review paper (Evereklioglu, 2005) in order to discover much more about the etiopathogenesis by sincere efforts and to discover definitive treatment for this unique and potentially blinding disorder by novel and further investigations (Evereklioglu *et al*, 2000, 2001, 2002b,c,d; Evereklioglu and Er, 2002; Evereklioglu *et al*, 2002a, 2003a,b), because it is our common goal to cure (Evereklioglu, 2004) our patients as an ophthalmologist, rheumatologist, dermatologist, or any physician interested in oral disease care.

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References

Adamantiades B (1930). A case of recurrent hypopyon iritis. *Proc Med Soc Athens* **12:** 586–593.

Adamantiades B (1931). Sur un cas d'iritis à hypopion récidivant. *Ann Ocul (Paris)* **168:** 271–278.

Adamantiades B (1946). La thrombophlébite comme quatrième symptôme de l'iritis récidivante à hypopyon. *Ann Ocul (Paris)* **179:** 143–148.

Adamantiades B (1953). Le symptôme complexe de l'uvéite récidivante à hypopyon. *Ann Ocul (Paris)* **186:** 846–856.

Adamantiades B (1958). Severe complications of the central nervous system in the syndrome of relapsing iritis with hypopyon (in Greek). *Deltion Ellikinis Ophthalmologikis Etairias* **26:** 199–202.

Adams F (1849). *The genuine works of Hippocrates. Translated from Greek. A preliminary discourse and annotations, 1.* Epidemics III, p. 403.

Blobner F (1937). Zur rezidivierenden Hypopyoniritis. *Z Augenheilkd* **91:** 129–139.

Blüthe L (1908). *Zur Kenntnis des rezidivierenden Hypopyons*. Inaugural thesis, D Strauss, Heidelberg.

Carol WL, Ruys SC (1928). Over aphthosis en ulcus vulvae acutum. *Ned Tschr Genek* **1:** 396–406.

Christlieb O (1895). *Über Stomatitis und Vulvitis aphthosa*. Inaugural Dissertation, Universität Würzburg.

Dascalopoulos N (1932). Sur deux cas d'uvéite récidivante. *Ann Ocul (Paris)* **169:** 387–389.

Dilşen N (1996). History and development of Behçet's disease. *Rev Rhum Engl Ed* **63:** 512–519.

Evereklioglu C (2004). Managing the symptoms of Behçet's disease. *Expert Opin Pharmacother* **5:** 317–328.

Evereklioglu C (2005). Current concepts in the etiology and treatment of Behçet disease (Major Review). *Surv Ophthalmol* **50:** 277–350.

Evereklioglu C (2006). Author's response. *Surv Ophthalmol* **51:** 174–177.

Evereklioglu C, Er H (2002). Increased corneal thickness in active Behçet's disease. *Eur J Ophthalmol* **12:** 24–29.

Evereklioglu C, Yürekli M, Er H *et al* (2000). Increased plasma adrenomedullin levels in patients with Behçet's disease. *Dermatology* **201:** 312–315.

Evereklioglu C, Çokkeser Y, Doganay S *et al* (2001). Audiovestibular evaluation in patients with Behçet's syndrome. *J Laryngol Otol* **115:** 704–708.

Evereklioglu C, Özbek E, Er H *et al* (2002a). Urinary adrenomedullin levels are increased and correlated with plasma concentrations in patients with Behçet's syndrome. *Int J Urol* **9:** 296–303.

Evereklioglu C, Türköz Y, Er H *et al* (2002b). Increased nitric oxide production in patients with Behçet's disease: is it a new activity marker? *J Am Acad Dermatol* **46:** 50–54.

Evereklioglu C, Inalöz HS, Kirtak N *et al* (2002c). Serum leptin concentration is increased in patients with Behçet's syndrome and is correlated with disease activity. *Br J Dermatol* **147:** 331–336.

Evereklioglu C, Er H, Türköz Y, Çekmen M (2002d). Serum levels of TNF-α, sIL-2R, IL-6, and IL-8 are increased and associated with elevated lipid peroxidation in patients with Behçet's disease. *Mediators Inflamm* **11:** 87–93.

Evereklioglu C, Özbek E, Çekmen M *et al* (2003a). Urinary nitric oxide levels are increased and correlated with plasma concentrations in patients with Behçet's disease: is it a new urinary activity marker? *Nephrology* **8:** 246–253.

Evereklioglu C, Çekmen M, Özkiriş A, Karabaş L, Çaliş M (2003b). The pathophysiological significance of red blood cell nitric oxide concentrations in inflammatory Behçet's disease. *Mediators Inflamm* **12:** 255–256.

Giannoukas AD (2005). Regarding 'compelling nature of arterial manifestations in Behçet disease' (Letter to the editor and author reply). *J Vasc Surg* **41:** 181–182.

Gilbert W (1920). Über die rezidivierende eitrige Iridozyklitis (I. septica) und ihre Beziehungen zur septischen Allgemeinerkrankung. *Arch Augenheilkd* **86:** 29–49.

Gilbert W (1921). Über den pathologisch–anatomischen Befund bei Iridocyclitis septica (Iritis mit rezidivierendem Hypopyon). *Arch Augenheilkd* **87:** 27–34.

Gilbert W (1923). Zur Frage der Iridozyklitis mit rezidivierendem Hypopyon ('Iritis septica'). *Klin Monatsbl Augenheilkd* **71:** 409–414.

Gilbert W (1925). Über chronische Verlaufsformen der metastatischen Ophthalmie ('Ophthalmia lenta'). *Arch Augenheilkd* **96:** 119–130.

Grütz O (1926). Stomatitis et vulvitis aphthosa chronica rezidivans (blastomycetica)? *Zbl Haut* **20:** 415–416.

International Study Group for Behçet's Disease (1990). Criteria for diagnosis of Behçet's disease. *Lancet* **335:** 1078–1080.

Janin J (1772). *Mémoires et observations anatomiques, physiologiques et physiques sur l'œil, et sur les maladies qui affectent cet organe. Avec un précis des opérations et des remédies qu'on doit pratiquer pour les guerres*. Frères Périsse, Lyon-PF Didot: Paris, pp. 412–414.

Knapp P (1938). Rezidivierende Hypopyoniritis. *Ophthalmologica (Basel)* **96:** 297.

Kumer L (1930). Über Haut- und Mundschleimhauterscheinungen beim Ulcus vulvae acutum. *Dermatol Z* **57:** 401–411.

Matras A (1932). Über aphthenartige Mundschleimhautveränderungen beim Ulcus vulvae acutum mit positivem Bac. *Crassus-Befund. Arch Dermatol Syphil (Berlin)* **166:** 491–497.

Mutlu S, Scully C (1994). The person behind the eponym: Hulusi Behçet (1889–1948). *J Oral Pathol Med* **23:** 289–290.

Neumann I (1895). Die Aphthen am weiblichen Genitale. *Wien Klin Rundsch* **9:** 289–307.

Nishimura M (1936). A case of ulcus vulvae acutum with aphthae–like lesions in the mucous membranes of the mouth associated with acute iritis. *Arch Dermatol Syphil (Chicago)* **34:** 900.

Pils H (1925). Ein Beitrag zur Aphthosis. *Arch Dermatol Syphil (Berlin)* **149:** 4–8.

Planner H, Remenowsky F (1922). Beiträge zur Kenntnis der Ulcerationen am außeren weiblichen Genitale. *Arch Dermatol Syphil (Berlin)* **140:** 162–188.

Reis W (1906). Augenerkrankung und Erythema nodosum. *Klin Monatsbl Augenheilkd* **44:** 203–206.

Samek J, Fischer E (1929). Erythema nodosum als bakterielle Metastase eines Ulcus vulvae acutum. *Arch Dermatol Syphil (Berlin)* **158:** 729–733.

Saylan T (1997). Life story of Dr Hulusi Behçet. *Yonsei Med J* **38:** 327–332. http://www.hulusibehcet.net/behcet.htm [accessed on 30 April 2006].

Scully C (1982). Serum beta 2 microglobulin in recurrent aphthous stomatitis and Behçet's syndrome. *Clin Exp Dermatol* **7:** 61–64.

Scully C (1989). Orofacial herpes simplex virus infections: current concepts in the epidemiology, pathogenesis, and treatment, and disorders in which the virus may be implicated. *Oral Surg Oral Med Oral Pathol* **68:** 701–710.

Scully C, Boyle P, Yap PL (1982). Immunoglobulins G, M, A, D and E in Behçet's syndrome. *Clin Chim Acta* **120:** 237–242.

Scully C, Epstein J, Porter S, Cox M (1991). Viruses and chronic disorders involving the human oral mucosa. *Oral Surg Oral Med Oral Pathol* **72:** 537–544.

Shigeta T (1924). Recurrent iritis with hypopyon and its pathological findings. *Acta Soc OphthalmoI Jpn* **28:** 516–522.

The Behçet's Disease Research Committee of Japan (1989). Skin hypersensitivity to streptococcal antigens and the induction of systemic symptoms by the antigens in Behçet's disease: a multicenter study. *J Rheumatol* **16:** 506–511.

Walter F, Roman I (1930). Beitrag zur Kenntnis der hämatogenen Hautmetastasen bei Ulcus vulvae acutum. *Dermatol Wochenschr* **90:** 705–709.

Weekers L, Reginster H (1938a). Un nouveau syndrome: iritis, ulcères aigus de la bouche et de la vulve. Sa parenté avec l'iritis récidivante à hypopion. *Arch Ophthalmol (Paris)* **2:** 697–705.

Weekers L, Reginster H (1938b). Contribution à l'étude de l'iritis récidivante à hypopion (uvéite allergique récidivante à hypopion). *Bull Soc Belge Ophthalmol* **76:** 31–44.

Weve H (1923). Über rezidivierende allergische Staphylokokkenuveitis. *Arch Augenheilkd* **93:** 14–39.

Whitwell GP (1934). Recurrent buccal and vulvar ulcers with associated embolic phenomenon in skin and eye. *Br J Dermatol* **46:** 414–419.