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Review

Naming Dilemma of Behçet's Disease

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Abstract

Background: The disease, currently known all over the world as "Behçet's disease", "Behçet's syndrome", "Behçet's triad", "Tri-symptom Behçet", "La maladie de Behçet" or "Morbus Behçet" was first recognized by Hulusi Behçet. In 1937 he wrote clear examples of symptomatic triad, which are still used as criterias worldwide for diagnosis Behçet's disease.

Hulusi Behçet (1889-1948) was a great scientist, worked throughly in the field of dermatology (**Figure 1**). The famous German pathologist Prof *Schwartz* portrayed him as being internationally well-known and quoted "You can not find him in his country, because he reports his works internationally" [**1, 2, 3**]. His keen interest in the field of dermatology has made *Hulusi Behçet* one of the most distinguished characters of his time [**2**].

Figure 1. Prof. Dr. Hulusi Behçet

The disease, currently known all over the world as "*Behçet*'s disease", "*Behçet*'s syndrome", "*Behçet*'s triad", "Tri-symptom *Behçet*", "La maladie de *Behçet*" or "Morbus *Behçet*" was first recognized by *Hulusi Behçet* with a patient in 1924 [**3**]. This patient, who had been examined because of eye disturbances, recurrent oral and genital ulcers both in Istanbul and Vienna for 40 years, was given several diagnoses. Some doctors thought of tuberculosis or syphilis while some other doctors said a microorganism which was not present in Europe might have caused the disease. *Hulusi Behçet*, who continued to examine the patient after his loss of vision, thought that the causative

agent was a virus. In the next several years he met two more patients with similar to that was seen in the previous patient. *Hulusi Behçet* thought the symptoms of these three patients were the symptoms of a new disease and reported his ideas on this topic firstly in 1936, in the Journal of Skin and Venereal Diseases [**1, 2, 3**]. Later, in 1937 he wrote clear examples of symptomatic triad, which are still used as criterias worldwide for diagnosis *Behçet*'s disease, in Dermatologische Wochenschrift. In the same year at the meeting of the Society of Paris Dermatology he declared that several factors may cause the etiology of the disease [**1**], which still can be an acceptable statement. Later he diagnosed further patients and published in German as "Tri-Symptomenkomplex" in 1939, and in English as "Triple symptom complex" in 1940 [**4**]. In subsequent years, this unique disorder drew the attention, and the term "Behçet's syndrome" was first used by Jensen in 1941 [**5**]. The term "*Behçet*'s disease" was first used by *Fiegenbaum* and *Kornblueth* in 1946 [**4, 6**]. On 13 September 1947, international dermatologic societies came together in Zurich and named the disease as "Morbus *Behçet*", which honored the first describer of "triple symptom complex", not individual symptom [**4**]. However, in the 21th century, there are attempts to change the name of this old and established disorder well known as "*Behçet*'s disease" since 1941 [**4**]. In fact there have already been various naming efforts in the past, such as "*Gilbert*'s ophtalmia lenta" [**7, 8**], "*Gilbert-Behçet* disease" [**7, 9, 10**], "*Gilbert- Adamantiadis-Behçet* disease" [**7, 11**], and *Hippocrates- Adamantiades-Behçet* disease [**12**].

It is clear that several authors before *Hulusi Behçet* described one or several individual findings of this disorder. Among these physicians, for example, *Hippocrates* in the fifth century BC reported some individual symptoms attributed to an originally endemic and epidemic disease. But, due to sporadic appearance of the disease in the course of time, the disease became less significant and was forgotten [**13**]. Symptoms of many diseases were first reported by *Hippocrates*. If so, today many diseases should be renamed and called "*Hippocrates*-…disease or syndrome". There were also other physicians who described one or several individual findings of this disorder, for example, *Janin* (1772), *Reis* (1906), *Blüthe* (1908), *Gilbert* (1920, 1921, 1923), *Planner* and *Remenowsky* (1922), *Weve* (1923), *Shigeta* (1924), *Adamantiades* (1930), *Dascalopoulos* (1932), *Whitwell* (1934), *Nishimura* (1936), *Blobner* (1937) and probably many others remained still unnamed reported several individual findings of this unique disorder [**1, 2, 3, 4, 7, 14, 15, 16**]. However, all these papers ascribed the findings either to an other disease, such as tuberculosis, syphilis, sepsis or allergy, or to a coincidence and none of them indicated a new or a single syndrome with "classical triad" [**4**]. In addition, Dr. *Hulusi Behçet*, as a dermatologist, put the particular importance on the recurrent oral ulcerations that is today the "universal hallmark" and the only sine qua non symptom of this unique disorder according to the International Study Group Criteria for diagnosis of *Behçet*'s disease held in the UK [**7**].

If honour is to be given to all authors who reported some findings on this disorder, the disease would be entitled as "*Hippocrates-Janin- Neuman- Reis- Bluthe- Weve- Grutz-Kumer- Adamantiades- Whitwell- Nishimura-Blobner- Behçet* disease" or longer than that. However, eponyms for diseases mostly have been attributed to contemporary scientists and there are several Turkish scientists presented all historical publications with evidence-based articles on this topic which are available in English literature [**1, 2, 3, 4, 7, 14, 15, 16, 17**].

The best comment on this topic comes from Dr. *George E. Ehrlich* (University of Pennsylvania, Philadelphia, USA) in his own words "*Behçet* was not the first to describe this disorder that now bears his name. He was preceded by *Shigeta* in Japan (1924), *Adamantiades* in Greece (1931), and *Whitwell* in Great Britain (1934), probably also by *Kurosawa, Lipschhutz*, and arguable even earlier by *Hippocrates*, among extend descriptions. *Behçet* deserved to have the disease named after him, however, because he was the first modern author to group the various ophthalmological, dermatological, and orogenital lesions together as a syndrome" [**18**].

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