Value of Eponyms in Dermato-Trichological Nomenclature

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“People nowadays think that scientists exist to instruct them, poets, musicians, etc. to give them pleasures. The idea that these have something to teach them – that does not occur to them.”

Ludwig Wittgenstein, *Culture and Value*

Keywords

Eponym · Acronym · Possessive and non-possessive use of eponyms · Avoidance of derogative eponyms · Culture and value of eponyms

Abstract

Traditionally, syndromes have been named after the physician who originally identified the condition. These are referred to as eponymous syndromes. Nevertheless, the term eponym is to be regarded in a broader sense, since – by definition – an eponym is a person, place, or thing after whom or which something is named. Accordingly, some eponymous syndromes do not refer to the physician who originally reported the condition, but alternatively to the indicator patient, a geographic location, or a historical, literary, or mythological context. The recent past has seen a shift towards naming conditions descriptively by symptoms rather than eponymously. When the syndrome name is formed as an abbreviation from the initial letters of the symptoms, it is referred to as an acronym. The use of acronyms has become particularly popular and is often taught as mnemonic device or used as mental checklist. However, the use of eponyms confers historical, literary, and cultural information that reaches beyond and therefore conveys the dignity of a broader educational background and understanding.

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Introduction

A syndrome is a set of concurrent symptoms and clinical findings that form an identifiable pattern, without necessarily tying them to a single identifiable pathogenesis. In the instance of a known pathogenesis, the words syndrome, disease, or disorder end up being used interchangeably. In instances with unknown etiology, the presence of the associated symptoms with a statistically improbable correlation normally leads the researchers to hypothesize that there exists an unknown underlying cause for all the described symptoms.

In the Islamic Golden Age, Persian polymath Ibn Sīnā or Avicenna (980–1037) pioneered the idea of the syndrome in the diagnosis of specific diseases in his *Canon of Medicine* [1]. The concept of a medical syndrome was further developed by English physician Thomas Sydenham (1624–1689). Sydenham’s fundamental idea was to take diseases as they presented themselves in nature and

The paper and original proposal of Axel Munthe’s syndrome as eponym for frontal fibrosing alopecia was inspired by the author’s visit to the Island of Capri, Italy, and Axel Munthe’s Villa San Michele on the occasion of participation at the Capri Hair & Nail and Anti-Aging Practical Course 2016 by appointment of course president Antonella Tosti.

to draw up a complete picture of the objective characters of each. The conformity of type in the presentation and course of a disease was due to the uniformity of origin. The causes that he dwelt upon were the evident and congruent pathological phenomena, while he considered it useless to elucidate the remote causes. Nevertheless, Sydenham's nosological method essentially anticipated the modern one.

As yet, there is no set common convention for the naming of newly identified syndromes, leaving a considerable degree of freedom for nomenclature. Traditionally, syndromes have been named after the physician or researcher who originally identified the condition. These are referred to as eponymous syndromes. Nevertheless, the term eponym is to be regarded in a broader sense, since – by definition – an eponym is a person, place, or thing after whom or which something is named. Accordingly, some eponymous syndromes do not refer to the physician who originally reported the condition, but alternatively to the indicator patient, to a geographic location (e.g., Ambras syndrome), or to a historical (e.g., Marie Antoinette or Thomas More syndromes), literary (e.g., Rapunzel syndrome, Münchhausen syndrome, Dorian Gray syndrome), or mythological context (e.g., Thersites complex). Finally, the recent past has seen a shift towards naming conditions descriptively by symptoms rather than eponymously.

When the syndrome name is formed as an abbreviation from the initial letters of the symptoms, it is referred to as an acronym, e.g., HAIR-AN syndrome for the association of hyperandrogenism (HA), insulin resistance (IR), and acanthosis nigricans (AN). The use of acronyms has become particularly popular in the 20th century and is often taught as mnemonic device or used as mental checklist. The first acronym used in dermatology, TEN (for toxic epidermal necrolysis), appeared only 13 years after the official acceptance of the exact acronym definition, and the rise of acronyms in naming of dermatoses and syndromes has continued since [2]. According to the MEDLINE and PubMed database search, more than 90 abbreviations were recorded in dermatology [3]. In its most extreme form, acronymophilia has reduced the respective acronym to a single letter, such as the H syndrome (for hyperpigmentation, hypertrichosis, hepatosplenomegaly, heart anomalies, hearing loss, hypogonadism, low height, and hyperglycemia/diabetes mellitus) [4], despite the three simple rules recommended for creating an acronym [5]: (1) the acronym must have at least three letters and be easily pronounceable, (2) it has to make communication easier, (3) and it has to be more

readily recognized by the reader compared to the original phrase, with *familiarity* as the most important characteristic. Interestingly, the generally well-recognized acronym SAPHO (for synovitis, acne, pustulosis, hyperostosis, and osteitis) is only explained by historical connections and therefore familiarity with iconic Greek poet Sappho (630–570 BC), despite reservations of some experts with regards to its validity [6].

Eponymous Syndromes in Dermato-Trichology

Recently, the eponym Axel Munthe's syndrome has been proposed for frontal fibrosing alopecia [7], while the designation of Ambras syndrome for congenital universal hypertrichosis has been discouraged [8].

Frontal fibrosing alopecia represents a peculiar condition with a quasi-symmetrical, marginal alopecia along the frontal and temporal hairline with scarring. Since its original description credited to Steven Kossard in 1994 [9], it has been speculated as to whether the condition existed before Kossard's original report, since artistic depictions dating from the 15th and 16th century often show a high frontotemporal hairline [10–12]. Rather, these represent fashionable forms of frontal pseudo-alopecia than a genuine pathologic condition, since the hair was then shaved around the hairline or plucked at the temples and the napes of the neck, and the condition later disappeared again from the arts. Nonetheless, there is evidence that the condition may have existed well before 1994 from celebrated Swedish physician Axel Munthe's (1857–1948) description of his housekeeper Mamsell Agata in his autobiographical account *The Story of San Michele* (originally published in English language in 1929): "To describe what she looked like is beyond me. She had thin golden locks arranged in a sort of early Victorian fashion – Rosalie said it was a wig but I do not know. An exceptionally high and narrow forehead, no eyebrows..." [13]. The reliability of the literary source and the distinctive features as described by Axel Munthe are compelling to the extent that it has been proposed to eponymize frontal fibrosing alopecia Axel Munthe's syndrome in recognition of the legacy of the personal reminiscences and medical observations recorded for posterity in his timeless book.

Ambras syndrome was originally coined by Baumeister in 1993 [14] to describe a case of congenital generalized terminal hypertrichosis which was analogized to the portrait paintings of Petrus Gonsalvus (Spanish: Pedro González) and his children, exhibited in Ambras Castle

near Innsbruck, Austria. His life has been well chronicled as he became famous during his lifetime because of his condition. He and his family became an object of scientific inquiry by Italian naturalist Ulisse Aldrovandi (1522–1605) and others, including Swiss physician and professor in Basel, Felix Platter (1536–1614). Gonsalvus first came to the court of Henry II (1519–1559), King of France, in 1547 and was sent from there to the court of Margaret of Parma (1522–1586), regent of the Netherlands. He married while there and was later moved to the court of Alexander Farnese (1545–1592), Duke of Parma. Four of his seven children were also afflicted with universal hypertrichosis and were painted. The Chamber of Art and Curiosities, Ambras Castle collection, originally exhibited paintings of Petrus Gonsalvus and of his family. Ultimately, it is believed that the marriage between Petrus Gonsalvus and Lady Catherine may have partially inspired the fairy tale *Beauty and the Beast*.

The condition is characterized by excessive hair growth on the entire body, with the exception of the palms, soles, and mucous membranes. Hypertrichosis is most excessive on the face, ears, and shoulders. The forehead, eyelids, nose, cheeks, and preauricular regions are uniformly covered with hair. The fine silky hairs are of the vellus type, the hair is fine and long, reaching a length of several decimeters if not cut. The hypertrichosis of the ears is typical, with long curls protruding from the external auditory canal. Facial and dental abnormalities may also be present. Ambras syndrome has been mapped to the long (q) arm of chromosome 8, and it appears to follow an autosomal dominant pattern of inheritance.

Chen et al. [8] put forth that the designation Ambras syndrome has historically been applied to various genetic types of congenital universal hypertrichosis with potential gene loci so far identified on 8q12, 17q24.2–q24.3, or Xq24–q27.1 that differ to such a degree that the term has no specific meaning, neither in the past nor in the future, and therefore should be jettisoned as misleading. The family portrayed in Ambras probably represented a syndromic type of congenital generalized terminal hypertrichosis with abnormal dentition and autosomal dominant inheritance.

While we cannot know with certainty in the case of Petrus Gonsalvus and his family, the discussion does not seem to be relevant either, since in many instances a syndrome is not specific to only one condition: for example, toxic shock syndrome can be caused by various toxins, premotor syndrome can be caused by various brain lesions, and premenstrual syndrome is not a disease but simply a set of symptoms.

Marie Antoinette syndrome designates the condition in which scalp hair suddenly turns white. Today, the syndrome is interpreted as an acute episode of diffuse alopecia areata in which the very sudden “overnight” graying is caused by the preferential loss of pigmented hair in this supposedly immune-mediated disorder [15]. The name alludes to the wretched French Queen Marie Antoinette (1755–1793), whose hair allegedly turned white the night before her last walk to the guillotine during the French Revolution. Although the actual incidence is rare, this stigmatizing phenomenon has captured storytellers’ imagination like few other afflictions as a sign of grave sorrow. History also records that the hair of the English martyr Sir Thomas More (1478–1535) turned white overnight in the Tower of London before his execution. Sir Thomas More was an English lawyer, author, and statesman who in his lifetime gained a reputation as a leading Renaissance humanist scholar and occupied many public offices, including Lord Chancellor from 1529 to 1532. He was beheaded in 1535 after having refused to sign the Act of Supremacy that declared King Henry VIII Supreme Head of the Church of England over the Pope. In 1935, 400 years after More’s death, Pope Pius XI (papacy: 1857–1939) canonized him in the Roman Catholic Church, and in 2000 Pope John Paul II (papacy: 1978–2005) officially declared More Patron Saint of politicians. Although Marie Antoinette has originally been chosen as eponym for the syndrome, presumably out of dermatological gallantry, Saint Thomas More, who turned white in 1535, ought to have the right of seniority over the Queen of France, who succumbed to the same fate only in 1793. Moreover, it would seem more appropriate to use the term Marie Antoinette syndrome for the condition afflicting women and Thomas More syndrome for men [16].

Although early accounts of overnight whitening of hair are substantiated by more recent case reports in the scientific literature, it may be suspected that the phenomenon has been used both as a literary stylistic means in fiction, with the aim of dramatizing, and in historical accounts with the claim of authenticity. Scrutinizing the case of Henry III of Navarra (later King Henry IV of France), whose hair allegedly turned white overnight on the occasion of the horrors of St. Bartholomew’s day massacre in 1572, Navarini and Trüeb [17] found inconsistencies between Henry’s biography and the current pathophysiological understanding of the condition. Since a prerequisite for overnight whitening of hair would be that the individual in question has a salt-and-pepper pattern with pigmented hairs interspersed with white hairs and the pigmented hairs selectively shed while the white

hairs remain, it seems quite improbable that Henry III of Navarre's hair should have turned white so rapidly, as he was only 19 years old at the time in question.

Thersites complex is the term used for a subgroup of patients with body dysmorphic disorder, in which a minimal physical deformity causes excessive psychological disturbances and distress [18]. The condition is named after the ugliest soldier of the Greek army during the Trojan War. Even though he plays only a minor role in the epic, Homer described him in detail in the Iliad, Book II, as bow-legged and lame, to have shoulders that cave inward, and a head which is covered in tufts of hair and comes to a point, as vulgar, obscene, and somewhat dull-witted.

The Dorian Gray syndrome represents yet another variant of body dysmorphic disorder with reference to a literary figure. The particularity of the condition is that patients wish to remain forever young and seek lifestyle drugs and surgery to deter the natural aging process. It is believed to represent a cultural and societal phenomenon, since an estimated 3% of the total population in Western society display features of the syndrome. The Dorian Gray syndrome was first described on the occasion of a symposium about aesthetic medicine [19] and is named after Oscar Wilde's famous gothic horror novel *The Picture of Dorian Gray*, in which the protagonist, a beautiful young aesthete, exclaims in front of his portrait [20]:

Why should it keep what I must lose? Every moment that passes takes something from me, and gives something to it. Oh, if it were only the other way! If the picture could change, and I could be always what I am now! For that – for that – I would give everything! Yes, there is nothing in the whole world I would not give! I would give my soul for that!

If the defensive acting-out character of the syndrome is not understood properly and the patient incessantly uses lifestyle products without understanding the underlying psychodynamics, a chronic state of narcissistic emptiness may develop. Depressive episodes and suicidal crisis are often observed if medical and surgical lifestyle treatments as means of defense are not sufficient to preserve the patient's idea of beauty.

Munchhausen syndrome, named after Baron Munchhausen, represents yet another psychopathological disorder relevant to dermatology, wherein those affected feign disease, illness, or psychological trauma to draw attention, sympathy, or reassurance to themselves [21]. The condition fits within the subclass of factitious disorder with predominantly physical signs and symptoms, but patients also have a history of recurrent hospitalization,

traveling, and dramatic, extremely improbable tales of their past experiences. The Baron is a fictional German nobleman created by German writer Rudolf Erich Raspe (1736–1794) in his 1785 book *Baron Munchhausen's Narrative of his Marvellous Travels and Campaigns in Russia*. The fictional Baron's exploits, narrated in the first person, focus on his impossible achievements as a sportsman, soldier, and traveller, for instance riding on a cannonball, fighting a forty-foot crocodile, and travelling to the moon. Intentionally comedic, the stories play on the absurdity and inconsistency of Munchhausen's claims.

Rapunzel syndrome is the term coined for an intestinal condition resulting from ingesting hair (trichophagia) [22]. This causes a trichobezoar (hair ball) to form in the stomach, which has a long tail extending into the small intestine, in analogy to the long-haired girl Rapunzel letting down her hair locks from the tower window in the respective fairy tale by the Brothers Grimm. The syndrome is usually associated with trichotillomania and represents an extremely rare complication. Because the human gastrointestinal tract is unable to digest hair, the trichobezoar has to be treated surgically, while patients require psychiatric evaluation and treatment for disorder of impulse control. The brothers Jacob Grimm (1785–1863) and Wilhelm Grimm (1786–1859) were German academics, philologists, cultural researchers, lexicographers, and authors who together specialized in collecting folklore which they originally published in their *Children's and Household Tales* (1812), including the tale of *The Maiden in the Tower*.

A Caveat in the Use of Eponyms

The faun tail nevus describes a usually congenital, circumscribed, triangle-shaped, excessive hair growth over the lumbosacral region. Its importance to dermatologists lies in the fact that it may represent a cutaneous marker for underlying spinal dysraphism [23]. It has been named after a tailed mythological half human-half goat manifestation of forest and animal spirits (Fig. 1). Romans believed fauns inspired fear in men travelling in lonely, remote, or wild places. The French symbolist Stéphane Mallarmé's (1842–1898) famous masterpiece *L'après-midi d'un faune* (published in 1876) describes the sensual experiences of a faun who has just woken up from his afternoon sleep and discusses his encounters with nymphs during the morning in a dreamlike monologue. The composer Claude Debussy (1862–1918) based his symphonic poem *Prélude à l'après-midi d'un faune* (1894) on the

poem, which also served as the scenario for a ballet entitled *L'après-midi d'un faune* choreographed to Debussy's score in 1912 by Vaslav Nijinsky (1889–1950).

Because of their derogative connotation, zoological epithets should be omitted in the description of medical conditions, such as circumscribed hypertrichosis of the midline associated with spinal dysraphism, and others. This holds particularly true to conditions with excessive hair growth, where the coarse and dark hair already has a negative psychosocial impact on the patient's life [24].

The same applies to the term gargoylism for the condition characterized by coarsened facial surface and distorted features with thickened gums and upturned nose associated with Hurler's syndrome (mucopolysaccharidosis I-H) [25].

In architecture, the gargoyle is a carved or formed grotesque with a spout designed to convey water from a roof and away from the side of a building, thereby preventing rainwater from running down masonry walls and eroding the mortar between. The term originates from the French "gargouille," which in English is likely to mean throat, gullet, or similar words derived from the root "gar," "to swallow," which represented the gurgling sound of water. The term gargoyle is most often applied to medieval work, the most famous example being the Notre Dame de Paris. The primary use of the gargoyle was to illustrate evil through the form of the gargoyle. In the 12th century, eminent French abbot of the Cistercian Order St. Bernard of Clairvaux (1090–1153) was notorious for speaking out against gargoyles [26]:

What are these fantastic monsters doing in the cloisters before the eyes of the brothers as they read? What is the meaning of these unclean monkeys, these strange savage lions, and monsters? To what purpose are here placed these creatures, half beast, half man, or these spotted tigers? I see several bodies with one head and several heads with one body. Here is a quadruped with a serpent's head, there a fish with a quadruped's head, then again an animal half horse, half goat... Surely if we do not blush for such absurdities, we should at least regret what we have spent on them.

Ultimately, inappropriate eponymous syndromes have been rephrased when the term has been unacceptable and upsetting for affected families. An example is the Kabuki syndrome, previously known as Kabuki make-up syndrome, a pediatric disorder with multiple congenital anomalies and intellectual disabilities. It is named Kabuki syndrome because of the facial resemblance of affected individuals to the stage make-up used in Kabuki, a traditional Japanese theatrical form. Although the name Kabuki make-up syndrome has originally been used to iden-

Fig. 1. The Roman Faunus is adapted from the Greek Satyr, companion to Dionysus and Silenus in Greek mythology. Satyr with wine cup, Carlo di Cesare del Palagio (1540–1598), exhibited at the Residence Museum, Munich, Germany (photographed by the author).

tify this condition, later authors recommended that the term "make-up" be deleted from the designation as it had caused problems with some of the families [27, 28].

Accordingly, and with reference to Axel Munthe's original description of the condition in *The Story of San Michele*, the eponymous designation of frontal fibrosing alopecia as Axel Munthe's syndrome is preferred to Mamsell Agata syndrome, since Axel Munthe portrayed his housekeeper Mamsell Agata in a rather pejorative manner.

Table 1. Summary of arguments for abandoning or maintaining eponyms (modified from [32])

Arguments for abandoning eponyms

The name confers no information other than historical

There can be a national or ethnic bias to the eponym chosen for use

Accordingly, eponymic usage may vary by country (e.g., sideropenic dysphagia is Plummer-Vinson syndrome in the US and Australia, Patterson-Kelly syndrome in the UK, and Waldenstrom-Kjellberg syndrome in Scandinavia), and several eponyms may refer to the same condition

The same eponym may be used for different conditions, which creates confusion (e.g., Ambras syndrome for congenital generalized terminal hair hypertrichosis [8])

An eponym may honor an individual who has been discredited due to his associations, background, or professional conduct (e.g., Wegener's granulomatosis named after former Nazi physician Friedrich Wegener [33])

History sometimes shows credit should have gone to a different person (e.g., Graham Little syndrome, since the indicator patient was originally referred by Lassueur from Lausanne, Switzerland, to Graham Little in London [34], or Behçet's disease, since Greek ophthalmologist Benediktos Adamantiades reported the respective condition 7 years prior to Turkish dermatologist Hulusi Behçet's original publication in 1937 [35])

The scientific and medical communities regard it as bad form to attempt to eponymise oneself

Eponyms with derogative connotations should be omitted (e.g., Kabuki make-up syndrome [27, 28])

Arguments for maintaining eponyms

The name may be shorter and more memorable than the medical one (the latter requiring abbreviation to its acronym for mnemonic purpose or to circumvent problems with orthography in cultures not familiar with Latin)

Sometimes the medical name proves to be incorrect (e.g., some reports have mistakenly classified Ambras syndrome as example of hypertrichosis lanuginosa)

The syndrome may have more than one cause, yet it remains useful to consider it as a whole

Being awarded an eponym is regarded as an honor, and it continues to respect a person's legacy, which may otherwise be forgotten (e.g., Axel Munthe's syndrome [7])

The name confers historical, literary, and cultural information that reaches beyond the purely mnemonic aspects of acronyms and thus conveys the dignity of a broader educational background and understanding

An Appreciation of Culture and Value in Use of Eponyms

Eponyms have a long-standing tradition in Western medical terminology. To discuss something, it must have a name. At a time when medicine did not command the means to investigate underlying causes of many syndromes, the eponym was a convenient way to label a disease condition. Traditionally, diseases have been named after the individual who originally described the condition, typically by publishing a respective scientific article in a peer-reviewed medical journal. Being awarded an eponym is still regarded as an honor today, while the scientific community considers it as bad form to attempt to eponymise oneself.

Alternatively, eponyms are named after fictional characters who displayed characteristics attributed to the syndrome, e.g., Münchhausen syndrome and Dorian Gray syndrome, or after historical personalities, events, or locations, e.g., Marie Antoinette syndrome, Thomas More syndrome, and Ambras syndrome.

In 1975, the Canadian National Institutes of Health held a conference on the naming of medical conditions and came to the conclusion that "The possessive use of an eponym should be discontinued, since the author neither had nor owned the disorder." [29]. Nevertheless, the argument seems pointless, since it is obvious that one cannot own a medical disorder. In fact, the differentiated use of eponyms in either the genitive (or possessive) or the attributive (non-possessive) form may make sense, since it allows at first glance to understand whether the eponym relates to the first describer in its genitive (or possessive) form, or to a character from literature or history in its attributive (or non-possessive) form, e.g., Axel Munthe's syndrome points to his original description of frontal fibrosing alopecia, while alternatively Mamsell Agata syndrome would point to the individual affected with the condition in question, as described by Axel Munthe. Problematic is the use of eponyms in the genitive form for indicator patients afflicted with the respective disorder. Since only rarely an eponymous disease is named after a patient, one would have to come to a consensus to use it rather in the attributive form, in analogy to the convention for fictive or historical personalities, as in the Dorian Gray, Marie Antoinette, and Thomas More syndromes. Medical journals, dictionaries, and style guides remain divided on this issue, and the trend in possessive usage varies between countries, journals, and even diseases [30]. While European journals tend towards continued use of the possessive, US journals are largely discontinu-

ing its use [31]. Arguments for abandoning or maintaining eponyms are summarized in Table 1.

Unlike acronyms that reduce nomenclature to a purely mnemonic purpose, the conscious and conscientious use of eponyms ultimately confers historical, literary, and cultural information that conveys the dignity of a broader educational background and understanding. Dig a bit

deeper into the world of eponyms, and you will find all sorts of untold stories!

Disclosure Statement

The author has no disclosures to report.

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