

**EPONYMS IN THE DERMATOLOGY LITERATURE
LINKED TO PALMO-PLANTAR KERATODERMA**Ahmad Al Aboud¹, Khalid Al Aboud²¹*Dermatology Department, King Abdullah Medical City, Makkah, Saudi Arabia*²*Department of Public Health, King Faisal Hospital, Makkah, Saudi Arabia***Source of Support:**

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Palmoplantar keratodermas (PPKs) represent a diverse group of hereditary and acquired disorders characterized by hyperkeratosis of the skin on the palms and soles [1]. The three major patterns of involvement are diffuse, focal and punctate. There are clinical distinguishing features for each disease in this group, for example, transmigration to areas beyond the palmoplantar skin. Also the extent of associated systemic symptoms if present help in characterization of each type.

Although a number of classifications of keratodermas have been published, none unite satisfactorily clinical presentation, pathology and molecular pathogenesis.

We based our concise review of selected eponyms linked to PPK (Tabl. I) [2-36], on the classifications published in the current editions of two major textbooks in dermatology; Rook's Textbook of Dermatology and Dermatology by Jean L Bologna.

Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK)	Remarks
Bart–Pumphrey Syndrome [2]	This syndrome is characterized by knuckle pads, leukonychia, palmoplantar keratoderma (PPK) and sensorineural deafness. This syndrome is first described by Dr Schwann, from Poland and appeared later in English literature by Robert S. Bart (Dermatologist) and Robert E. Pumphrey (Otolaryngologist); both from. Dr Robert S Bart (Fig. 1) reported this syndrome, in 1967, while he was working as an assistant professor of clinical dermatology, New York University School of Medicine and Post-Graduate Medical School.
Brünauer–Fuhs–Siemens PPK [3,4]	It is a type of focal Palmoplantar Keratoderma (Isolated). Also known as, PPK striata/areata, striate PPK, focal non-epidermolytic PPK, Wachters PPK (Focal/areate/nummular keratoderma). Focal, areate or nummular, and linear or striate keratodermas have been distinguished. The occurrence of both areate and striate forms within a family led Wachters to suggest a single entity, keratoderma varians. Stefan Robert Brünauer, born in 1887, is an Austrian physician. Herbert Fuhs (1891-1960), is an Austrian dermatologist. Hermann Werner Siemens (1891-1969), is a German dermatologist.
Buschke–Fischer–Brauer type [5]	Also known as, Brauer–Buschke–Fischer keratoderma. It is a synonym for Punctate Palmoplantar Keratoderma. August Brauer (1883-1945) (Fig. 2), a German physician. Abraham Buschke (1868-1943) (Fig. 3), a German dermatologist. Heinrich Fischer (1884-1943), a German dermatologist.
Camisa's syndrome or Camisa variant of Vohwinkel syndrome [6]	Also known as Loricrin keratoderma, Mutilating keratoderma with ichthyosis, and Variant Vohwinkel's syndrome. It is a type of Transgredient keratodermas. Named for Dr. Camisa (Fig. 4), who is currently the Director of the Phototherapy Department at Riverchase Dermatology and an Affiliate Associate Professor of Dermatology at the University of South Florida in Tampa.

Table I. Selected Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK).



Figure 1. Robert Bart, Image courtesy of The Archives of the Frederick L. Ehrman Medical Library, available online at <http://archives.med.nyu.edu/resources/imagedb/detail.php?recordID=35090002763282>



Figure 2. August Brauer (1883-1945).



Figure 3. Abraham Buschke (1868-1943).



Figure 4. Charles Camisa, M.D., FAAD.



Figure 5. José María Cantú Garza (1938-2007).

Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK)	Remarks
Cantu syndrome [7,8]	Hyperkeratosis–hyperpigmentation syndrome first reported in 1978. The same name is applied to a syndrome characterized by congenital hypertrichosis, distinctive facial features, osteochondrodysplasia and cardiac defects, first reported in 1982. Both are named for José María Cantú Garza (1938-2007), (Fig. 5), who was a Mexican genetic researcher.
Carvajal syndrome [9]	Striate EPPK with woolly hair and dilated left ventricular cardiomyopathy. Carvajal-Huerta (1998) described 18 patients with a confirmation of epidermolytic palmoplantar keratoderma, woolly hair, and dilated cardiomyopathy, examined clinically and histologically in Ecuador between 1970 and 1997. CS might be a variant of Naxos disease (ND), which was first described by Protonotarios et al., in families originating from the Greek island of Naxos. ND is a rare autosomal recessive inherited association of right ventricular dysplasia/dilated cardiomyopathy with woolly hair and palmoplantar keratoderma. Any patient with a PPK and woolly hair (or alopecia) should be sent for a cardiac evaluation.
Cole disease [11]	Guttate hypopigmentation with punctate PPK. First described by Cole in 1976.
Costa/Dowd kertoderm [12]	This is an eponym for, marginal popular keratoderma. In which there will be crateriform punctate keratoses at the margin of the sole (Wallace's line). Costa reported 13 cases with cornified and umbilicated papules distributed along the borders of the hands and feet. He introduced the term acrokeratoelastoidosis. However, Dowd et al., reported 15 cases, several familial, with similar oval or polygonal crateriform papules along the borders of the hands and feet in whom there was no solar damage or elastorrhexis. To distinguish this entity, it was termed focal acral hyperkeratosis. Many patients with these disorders are of Afro-Caribbean origin.

Table I. Selected Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK) (continued).

Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK)	Remarks
Gamborg–Nielsen (Norrbotten) type [15]	A type of Diffuse ppk with no association. It is an autosomal recessive transgredient mutilating keratoderma. with knuckle pads identified by Gamborg-Nielsen in 1985. Patients with the transgredient PPK were also reported in Japan by Nakajima.
Greither syndrome [16]	A type of diffuse Palmoplantar Keratoderma (transgrediens and progrediens PPK), originally described in 1952. It is characterized by a diffuse transgredient PPK with onset in early infancy. Named for, Aloys Greither (1914-1986), a German dermatologist.
Ichthyosis hystrix Curth–Macklin [17]	This is a rare type of Ichthyoses with associated keratoderma. There are horny or velvety spikes rather than thickened scales. Named for Helene Ollendorff Curth (1899-1982), (Fig. 6) and Madge Thurlow Macklin (1893–1962), an American medical geneticist. The described the condition in 1954.
Jadassohn–Lewandowsky type of Pachyonychia congenita (PC) [18]	Type 1 PC. Type 2 is known as Jackson–Lawler type. Josef Jadassohn (1863-1936), (Fig. 7) and his assistant, Felix Lewandowsky (1879-1921), (Fig. 8), were eminent German dermatologists.
Naegeli–Franceschetti–Jadassohn syndrome (NFJS) [19]	It is a rare symptom complex out of the spectrum of ectodermal dysplasia. The main clinical findings are absence of dermatoglyphs, reticular or mottled hyperpigmentation, hypohidrosis and nail dystrophy. NFJS is named after Oskar Naegeli, Adolphe Franceschetti and Josef Jadassohn. Oskar Naegeli (1885-1959), (Fig. 9), was a Swiss dermatologist. Adolphe Franceschetti (1896-1968), (Fig. 10), was a Swiss ophthalmologist. Josef Jadassohn (1863-1936), (Fig. 7), was a German dermatologist.
Haim–Munk syndrome [20].	It is a PPK with periodontitis, arachnodactyly and acro-osteolysis. In 1965, Dr. Salim Haim (1919-1983), (Fig. 11), dermatologist and Dr. Munk, a radiologist, from Haifa reported this syndrome, which is characterized by palmoplantar keratosis, pes planus, onychogryphosis periodontitis, arachnodactyly, and acroosteolysis.
Haxthausen's disease [21]	This is another name for Keratoderma climactericum. The specificity of this syndrome described in women over the age of 45 is uncertain, as many patients are obese. Pressure areas of the heel and the forefoot are involved first. Erythema and heavy hyperkeratosis with fissuring make walking painful. It was described in 1934.
Howel–Evans Syndrome [22]	Focal non-epidermolytic PPK with carcinoma of the esophagus. It was described in 1958.
Huriez Syndrome [23]	Palmoplantar keratoderma with scleroatrophy. Named for French dermatologist, Claude Huriez (1907-1984), (Fig. 12). In 1960s, Huriez and his colleagues reported 2 families from northern France with, this syndrome, which is characterized by scleroatrophy of the hands and feet, nail hypoplasia, mild palmoplantar keratoderma and hypohidrosis.

Table I. Selected Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK) (continued).



Figure 6. Helene Ollendorff Curth (1899-1982).

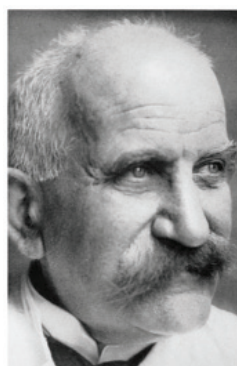


Figure 7. Josef Jadassohn (1863-1936).



Figure 8. Felix Lewandowsky (1879-1927). Reproduced with the kind permission of the Alumni Association of the Medical Faculty of the University of Basel, Switzerland.



Figure 9. Oskar Naegeli (1885-1959). A courtesy of Archives of the Institute for the History of Medicine, University of Zurich, Switzerland.



Figure 10. Adolphe Franceschetti (1896-1968). A courtesy of Library, university of Basel, Switzerland.



Figure 11. Salim Haim (1919-1983).

Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK)	Remarks
Netherton syndrome (NS) [9]	NS is characterized by the triad of trichorrhexis invaginata, ichthyosis linearis circumflexa, and an atopic diathesis. It is named after E.W. Netherton, Who described a 4-year old girl with scaly red and different hair, which he called bamboo hair, because of how it looked in the microscope. Nine years earlier, the Italian dermatologist Come described a condition in a young woman with a ring shape change in her skin, which he called itchyosis Linearis circumflex. These two descriptions were considered to be related.
Olmsted Syndrome [24]	Mutilating PPK with periorificial plaques. First described in 1927.
Papillon-Lefèvre Syndrome [1]	An autosomal recessive disorder, first described it in 1924. It is characterized by diffuse, transgredient PPK in association with destructive periodontitis (beginning in childhood) and premature loss of teeth. It is named for Papillon and Paul Lefèvre. Both are French dermatologists.
Papuloverrucous palmoplantar keratoderma of Jakac-Wolf [25]	Papuloverrucous PPK is a rare type first reported in 1975.
Refsum syndrome [26]	It is an example of occurrence of PPK in the disorders of Ichthyoses. Refsum disease is an autosomal recessive inborn error of lipid metabolism classically characterized by a tetrad of clinical abnormalities: retinitis pigmentosa, peripheral neuropathy, cerebellar ataxia, and elevated protein levels in the cerebrospinal fluid (CSF) without an increase in the number of cells. Sigvald Bernhard Refsum (1907-1991), (Fig. 13), was an outstanding Norwegian neurologist.
Richner-Hanhart Syndrome [27]	It is a type of focal Palmoplantar Keratoderma with Associated Features. It is a rare autosomal recessive disease characterized by ocular changes, painful palmoplantar hyperkeratosis, and mental retardation. Many of the reported families are of Italian origin. This syndrome is reported first by, Dr. Hermann Richner, Swiss dermatologist, born September 6, 1908, in Zürich. Ernst Hanhart (1891-1973), (Fig. 14), was Swiss internist and human geneticist.
Schöpf-Schulz-Passarge syndrome [28]	A type of ectodermal dysplasias with associated keratoderma. PPK with hidrocystomas, hypodontia and hypotrichosis. It was characterized in 1971.
Sjögren-Larsson syndrome [29]	It is an example of occurrence of PPK in the disorders of Ichthyoses. It is a rare autosomal recessive condition comprising congenital ichthyotic hyperkeratosis, spastic diplegia, mild to moderate mental retardation, and retinopathy. It is named for Karl Gustaf Torsten Sjögren (1896-1974) and Tage Konrad Leopold Larsson (1905-1998). Sjögren (Fig. 15), a Swedish psychiatrist and geneticist, was a pioneer of modern Swedish psychiatry. Tage K.L. Larsson, was a lecturer of statistics at the University of Lund.

Table I. Selected Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK) (continued).



Figure 12. Claude Huriez (1907-1984); with kind permission from; Museum Association Regional Hospital of Lille - France.



Figure 13. Sigvald Bernhard Refsum (1907-1991). This figure is reproduced with permission from the great Norwegian encyclopedia (Store norske leksikon), Available Online at; http://snl.no/Sigvald_Bernhard_Refsum



Figure 14. Ernst Hanhart (1891-1973). A courtesy of Archives of the Institute for the History of Medicine, University of Zurich, Switzerland.


Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK)	Remarks
Sybert type PPK [30-32] 	<p>It is a type of diffuse ppk with no other association. It is a severe transgredient keratoderma reported by Sybert et al. (Virginia Sybert, (Fig. 16), is a contemporary American dermatologist and medical geneticist) resembled mal de Meleda but had dominant inheritance. Onset was earlier than in Greither's syndrome. Glove and stocking hyperkeratosis, including autoamputation of toes, extended also to the elbow knees, posterior aspects of the forearms, shins, groins and natal cleft.</p> <p>Mal de Meleda is a rare autosomal recessive transgredient keratoderma named after the Croatian island of Meleda (Mljet) where it was first identified.</p> <p>Nagashima-type" keratosis is a nonprogressive, autosomal-recessive palmoplantar keratoderma that resembles a mild form of mal de Meleda.</p> <p>Lind et al, described an autosomal dominant form of diffuse nonepidermolytic PPK, designated PPK type Bothnia, which has a high prevalence of 0.3 to 0.55% in the 2 northernmost provinces of Sweden, situated to the west and the northwest of the Gulf of Bothnia.</p>
Unna-Thost PPK [33,34]	<p>Also known as, Thost-Unna keratoderma. It is a type of diffuse Palmoplantar Keratoderma. In 1880, Thost described a family with diffuse non-transgrediens PPK. This was followed by Unna's description of a clinically identical, autosomal dominant PPK in two families. Paul Gerson Unna (1850- 1929), (Fig. 17), was a German dermatologist. Herrmann Arthur Thost (1854-1937), (Fig. 18), was a German physician.</p>
Vohwinkel syndrome [35]	<p>Vohwinkel first described this autosomal dominant disorder in 1929. Honeycombed, diffuse hyperkeratosis of the palms and soles appears in infancy and then becomes transgredient. This is followed by the development of constricting bands of the digits during early childhood, which may lead to digital autoamputation, i.e. pseudoainhum. Peculiar starfish-shaped keratoses appear over the knuckles of the fingers and toes and are said to be characteristic of the disorder. Hearing loss of at least a moderate degree is seen in many patients. Additional reported findings are alopecia and ichthyosis</p>
Vörner keratoderma [36]	<p>First described by Vörner in 1901. Similar to Unna-Thost keratoderma but there epidermolysis in histology. Both are autosomal dominant and manifested as diffuse PPK without transgrediens or associated ectodermal features</p>

Table I. Selected Eponyms in the dermatology literature linked to Palmo-Plantar Keratoderma (PPK) (continued).

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Figure 15. Karl Gustaf Torsten Sjögren (1896-1974). Image is provided by the Center for History of Science, the Royal Swedish Academy of Sciences. Permission For republication is granted by Norstedts, Sweden.

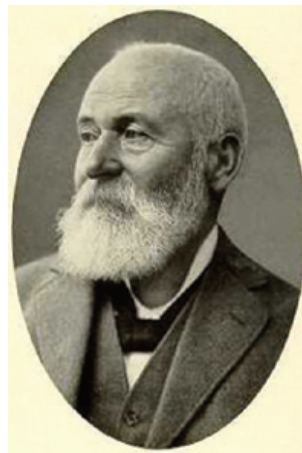


Figure 17. Paul Gerson Unna (1850-1929).



Figure 18. Herrmann Arthur Thost (1854-1937).

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