

SEPARATING "BART'S" APART IN DERMATOLOGY EPONYMS

NIEZALEŻNE WYSTĘPOWANIE „BART'S” W EPONIMACH DERMATOLOGICZNYCH

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There are 2 famous diseases in dermatology with the name "Bart", as a part of their eponyms. These are Bart syndrome and Bart-Pumphrey syndrome. This short letter is to give a brief account on these eponyms.

Bart syndrome

Bart syndrome, cited in the Online Mendelian Inheritance in Man [1], as Epidermolysis bullosa with congenital localized absence of skin and deformity of nails also as Epidermolysis bullosa dystrophica, Bart type (OMIM 132000).

Bart syndrome is a rare inherited condition characterized by epidermolysis bullosa and congenital absence of skin. It has been associated with other anomalies including pyloric atresia. The genetic abnormality has been linked to chromosome 3, with an autosomal dominant pattern of inheritance [2].

The mutant gene is COL7A1, located on 3p21.31 [1]. It is first reported by Bart and his colleagues in 1966 [3]. The disease is named after Bruce J Bart.

Dr Bruce J Bart (Fig.1) was born on April 6, 1936 in Minneapolis, Minnesota; USA. He is an academic dermatologist, working currently, as a Chair of the Dermatology department t at Hennepin County Medical Center and Professor of Dermatology at the University of Minnesota, USA.

In 1962 Dr Bart saw a 5 year old girl who had aplasia, blistering and nail abnormalities at birth and studied many members of her extended family with the same syndrome and published his work in 1966.

He subsequently studied the family and demonstrated that the syndrome was a form of dominant dystrophic EB.

In recent years many have discouraged the use of "Bart's Syndrome", favoring "a variation of DEB".

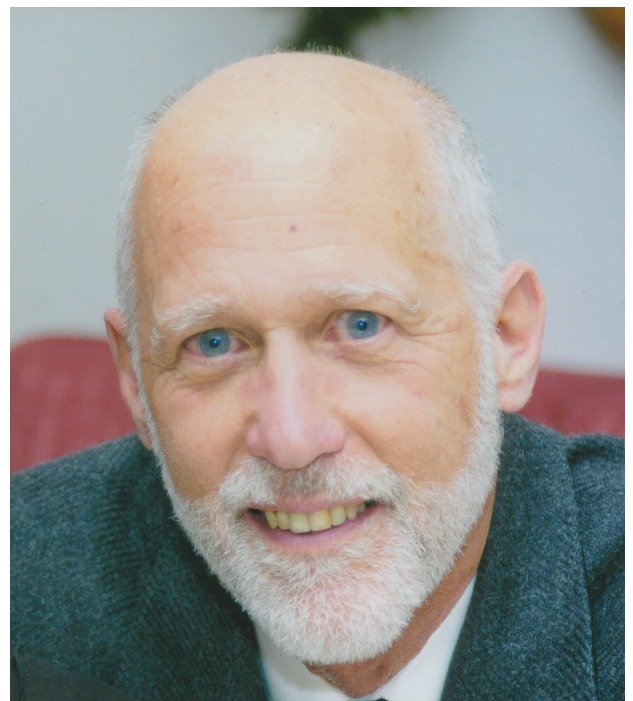


Figure 1. Dr Bruce J Bart

Bart-Pumphrey syndrome (BPS) or Schwann syndrome

This syndrome is characterized by knuckle pads, leukonychia, palmoplantar keratoderma (PPK) and sensorineural deafness. However, this syndrome has a considerable phenotypic variability. It is cited in the Online Mendelian Inheritance in Man [1], as knuckle pads, leukonychia, and sensorineural deafness (OMIM 149200) [4].

There is extensive overlap between the different forms of palmoplantar keratoderma (PPK) associated with hearing loss (HL) caused by GJB (Cx26) mutations (and they are

each quite rare). Therefore, some scientists thought that, it is worthwhile considering lumping them together under a title like 'GJB2 related PPK/HL'.

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 USA [4].

Dr Robert S Bart (Fig. 2) 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 [5].

In addition to the above syndrome, Dr Robert S Bart, had several other important publications.

In 1968, Bart et al [6] described 10 cases of an uncommon acquired skin growth that, they name it "acquired digital fibrokeratoma" (ADFK).

In their earlier description, the authors stated that the lesions resemble a "rudimentary supernumerary digit".

ADFK was once known eponymously as "Bart-Andrade acquired digital fibrokeratoma" [7], but this name is not of use any more and the disease is just known as acquired digital fibrokeratoma.

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Figure 2. 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>

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