Pachyonychia Congenita Project

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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.

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ORAL DISEASES

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http://www.blackwellmunksgaard.com

SPECIAL REVIEW

Marathon of eponyms: 10 Jadassohn-Lewandowsky syndrome (Pachyonychia congenita)

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The use of eponyms has long been contentious, but many remain in common use, as discussed elsewhere (Editorial: Oral Diseases. 2009: 15; 185). The use of eponyms in diseases of the head and neck is found mainly in specialties dealing with medically compromised individuals (paediatric dentistry, special care dentistry, oral and maxillofacial medicine, oral and maxillofacial pathology, oral and maxillofacial radiology and oral and maxillofacial surgery) and particularly by hospital-centred practitioners. This series has selected some of the more recognised relevant eponymous conditions and presents them alphabetically. The information is based largely on data available from MEDLINE and a number of internet websites as noted below: the authors would welcome any corrections. This document summarises data about Jadassohn-Lewandowsky syndrome.

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Also known as

Pachyonychia congenita

The condition

Jadassohn and Lewandowsky's original patient, reported in 1906, was a 15-year-old girl with fungating skin tuberculosis who had also had unusual keratinisation of the skin and tongue since infancy. About their patient, they wrote: 'The nail plates of all the fingers and toes are extremely thickened, and so hard that they cannot be cut with a scissors; the father has to trim them with a hammer and chisel'. Hyperhidrosis of the nose, palms and soles, scanty papular hyperkeratosis of the knees and elbows and a white plaque on the tongue were additional features. Although the parents and seven sisters had no skin problems, a younger brother was similarly affected.

Pachyonychia congenita type 1, or Jadassohn-Lewandowsky syndrome, is an autosomal dominant trait related to the keratin-16 (protein K16; gene KRT16) or keratin-6A (protein name K6A; gene name KRT6A) genes on chromosome 12. Clinical features include:

- Onychogryphosis (abnormally thick trumpet-shaped nails particularly affecting the thumb and first finger, and toe nails).
- Hyperkeratosis of palms, soles, knees and elbows appearing in the 2nd or 3rd year of life (the sole lesions occur over weight-bearing areas and make walking painful).
- Oral and anal leukoplakia with a milky appearance (usually appearing in the 2nd decade).
- Hyperhidrosis of hands and feet.
- Natal teeth.

Two other main variants of pachyonychia congenita have now been recognised, making three variants in total:

- Pachyonychia congenita Jadassohn-Lewandowsky type (type 1).
- Pachyonychia congenita Jackson Lawler type – due to mutations in the genes encoding keratin 6B (protein name K6B; gene name KRT6B) or keratin 17 (protein K17; gene KRT17) – there is no leukoplakia.
- Pachyonychia congenita; recessive type.

The persons

Josef Jadassohn was born on 10 September, 1863, in Liegnitz, Schlesien.

He studied Medicine in Göttingen, Heidelberg, Leipzig and Breslau, obtaining his doctorate in 1887 at Breslau. He was subsequently an assistant in the dermatology department of Breslau Allerheiligen-Hospital until 1892. Then in 1896, he was appointed extraordinarius and Director of the University of Bern

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dermatology clinic, where he was elevated to ordinarius in 1903. In 1917 he assumed the Chair of dermatology at Breslau, holding this position until he retired in 1931.

He was elected to honorary fellowship of the Royal Society of Medicine. Jadassohn revised and published the 14th edition of Edmund Lesser's *Lehrbuch der Haut- und Geschlechtskrankheiten* (2 volumes; Berlin, 1927–1930). He also published *Handbuch der Haut- und Geschlechtskrankheiten*, in 24 volumes (in 42), (Berlin, Springer, 1927–1937), and was also a co-publisher of *Archiv für Dermatologie und Syphilis*, *Zentralblatt für Haut- und Geschlechtskrankheiten*, and *Sammlung zwangsloser Abhandlungen aus dem Gebiete der Haut- und Geschlechtskrankheiten*. He died on 24 March, 1936.

Felix Lewandowsky was born on 1 October, 1879, in Hamburg, Germany, obtaining his doctorate in Strassburg in 1902 and then worked as an assistant at the Strassburg Institute of Hygiene. From 1903 to 1907, he worked at the dermatology clinic in Bern, Switzerland, and then returned to Hamburg. In 1917 he was invited to Basel as ordinarius and Director of the dermatology clinic. He died in Basel on 31 October, 1921.

Associated persons

Josef Jadassohn (1863–1936)
Felix Lewandowsky (1879–1921)

Source internet sites (accessed 21 February 2009) and further reading

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