

# BÖLÜM 56

## EPİDERMAL NEVUS SENDROMLARI

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### GİRİŞ

Bu bölümde öncelikle ‘epidermal nevüs’ terimi genel hatlarıyla tanımlanacak, daha sonra epidermal nevüs alt tiplerinden ve bunların eşlik ettiği sendromlardan bahsedilecektir.

### EPİDERMAL NEVÜS NEDİR?

#### Epidermal Nevüs

Epidermal nevüsler, embriyonik ektodermden gelişen hamartomatöz proliferasyonlardır. Çoğunlukla doğuştan ya da ilk bir yaş içerisinde, lineer pigmente yama veya daha az sıklıkta lineer pigmente bir plak olarak görülür. Epidermal nevüsler nadir olarak, çocukluk döneminde başlayabilir. Doğal seyri sürecinde, pubertede daha infiltrate, verrüköz ve hiperpigmente hal alır.<sup>1</sup> Genellikle sporadik olarak görülür ve epidermal göç yollarını temsil edecek şekilde, Blaschko çizgileri olarak tarif edilen yolları takip eder (Resim 1). Embriyonik ektodermden bulunan pluripotent hücreler, hem keratinositlere, hem de ekrin ve apokrin bezler, kıl folikülleri ve sebace bezler gibi deride bulunan birçok yapıya farklılaşırlar. Bu nedenle epidermal nevüsler çok çeşitli komponentleri içerisinde taşıyabilirler. Belirgin olarak sebace,

foliküler ve/veya apokrin bezler gibi adneksiyal bileşenlere sahip olan nevüsler “organoid”, sadece epidermal farklılaşma içeren nevüsler ise “organoid olmayan” veya “keratinositik” olarak adlandırılmıştır.

#### Epidermal Nevüs Epidemiyolojisi

Epidermal nevüsler, yaklaşık olarak % 0.001-0.003 oranında görülür ve her iki cinsiyeti de eşit oranda etkiler.<sup>2</sup> Tek başına izole bir bulgu olarak görülebileceği gibi, bazı sendromlara da eşlik edebilir. “Epidermal nevüs” terimi, geniş bir konu olduğu için akılda kalıcılığı artırmak adına halen kullanılmasına rağmen, son zamanlarda değişik tipte epidermal nevüslerin farklı somatik mutasyonlar sonucu ortaya çıktıkları ve farklı mozaik fenotiplerin deri bulgusu oldukları düşünülmektedir. Bu nedenle epidermal nevüsü olan bir hasta ile karşılaşıldığında, ilişkili sendromlar açısından farkında olunmalı ve gerekli durumlarda ileri incelemeler yapılmalıdır.<sup>1,2</sup> Bunun için de, tam bir anamnez ve fizik muayene gereklidir. Nadir de olsa, epidermal nevüsler tek başına yaygın deri tutulumu ile karşımıza çıkabilir. Literatürde bu durumu tanımlamak için ‘sistematize epidermal nevüs’ veya ‘nevüs unius lateralis’ terimi kullanılmaktadır.

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ya da hipoplazisi eşlik edebilir.<sup>213</sup> Yarık dudak ve umbilikal herni gibi nadir olarak bildirilen anomaliler de mevcuttur.<sup>217</sup> Nörolojik anomaliler arasında, ipsilateral hemisferlerin<sup>207,213,218</sup> ya da kranial sinirlerin hipoplazisi,<sup>213</sup> elektroensefalografik anomaliler,<sup>201</sup> entelektüel kapasitede hafif derecede bozulma,<sup>215</sup> hemiparezi,<sup>219</sup> dokunma ve ısı hassasiyetinin azalması<sup>206</sup> ve ipsilateral ya da bilateral sensorinöral işitme kaybı sayılabilir.<sup>196,215</sup>

Sınırlı sayıda CHILD sendromu vakasında malignite bildirim yapılmıştır. Bunlar, kontralateral adrenal bezde ganglionöroblastom ve CHILD nevüs zemininde gelişen bir skuamöz hücreli kanserden ibarettir.<sup>220,221</sup> Eğer CHILD nevüs çok geniş boyutta değilse, dermabrazyon ve nüksü engellemek amacıyla, kontralateral etkilenmemiş vücut yarısından elde edilen deri grefti uygulaması ile tedavisi mümkün olabilmektedir.<sup>222</sup>

## KAYNAKLAR

1. Sugarman JL. Epidermal nevus syndromes. *Semin Cutan Med Surg.* 2007 Dec;26(4):221-30. doi: 10.1016/j.sder.2008.03.006. PMID: 18395670.
2. Vujevich JJ, Mancini AJ. The epidermal nevus syndromes: multisystem disorders. *J Am Acad Dermatol.* 2004 Jun;50(6):957-61. doi: 10.1016/s0190-9622(02)61547-6. PMID: 15153903.
3. Hafner C, Toll A, Real FX. HRAS mutation mosaicism causing urothelial cancer and epidermal nevus. *N Engl J Med.* 2011 Nov 17;365(20):1940-2. doi: 10.1056/NEJMc1109381. PMID: 22087699.
4. Hafner C, López-Knowles E, Luis NM, Toll A, Baselga E, Fernández-Casado A, Hernández S, Ribé A, Mentzel T, Stoehr R, Hofstaedter F, Landthaler M, Vogt T, Pujol RM, Hartmann A, Real FX. Oncogenic PIK3CA mutations occur in epidermal nevi and seborrheic keratoses with a characteristic mutation pattern. *Proc Natl Acad Sci U S A.* 2007 Aug 14;104(33):13450-4. doi: 10.1073/pnas.0705218104. Epub 2007 Aug 2. PMID: 17673550; PMCID: PMC1948900.
5. Hafner C, van Oers JM, Vogt T, Landthaler M, Stoehr R, Blaszyk H, Hofstaedter F, Zwarthoff EC, Hartmann A. Mosaicism of activating FGFR3 mutations in human skin causes epidermal nevi. *J Clin Invest.* 2006 Aug;116(8):2201-2207. doi: 10.1172/JCI28163. PMID: 16841094; PMCID: PMC1501112.
6. Hafner C, Toll A, Gantner S, Mauerer A, Lurkin I, Acquadro F, Fernández-Casado A, Zwarthoff EC, Dietmaier W, Baselga E, Parera E, Vicente A, Casanova A, Cigudosa J, Mentzel T, Pujol RM, Landthaler M, Real FX. Keratinocytic epidermal nevi are associated with mosaic RAS mutations. *J Med Genet.* 2012 Apr;49(4):249-53. doi: 10.1136/jmedgenet-2011-100637. PMID: 22499344.
7. Paller AS, Syder AJ, Chan YM, Yu QC, Hutton E, Tadini G, Fuchs E. Genetic and clinical mosaicism in a type of epidermal nevus. *N Engl J Med.* 1994 Nov 24;331(21):1408-15. doi: 10.1056/NEJM199411243312103. PMID: 7526210.
8. Tsubota A, Akiyama M, Sakai K, Goto M, Nomura Y, Ando S, Abe M, Sawamura D, Shimizu H. Keratin 1 gene mutation detected in epidermal nevus with epidermolytic hyperkeratosis. *J Invest Dermatol.* 2007 Jun;127(6):1371-4. doi: 10.1038/sj.jid.5700712. Epub 2007 Feb 1. PMID: 17255957.
9. Akhyani M, Kiavash K, Kamyab K. Bullous ichthyosiform erythroderma in a child born to a parent with systematized linear epidermolytic hyperkeratosis. *Int J Dermatol.* 2009 Feb;48(2):215-7. doi: 10.1111/j.1365-4632.2009.03569.x. PMID: 19200214.
10. Chassaing N, Kanitakis J, Sportich S, Cordier-Alex MP, Titeux M, Calvas P, Claudy A, Berbis P, Hovnanian A. Generalized epidermolytic hyperkeratosis in two unrelated children from parents with localized linear form, and prenatal diagnosis. *J Invest Dermatol.* 2006 Dec;126(12):2715-7. doi: 10.1038/sj.jid.5700553. Epub 2006 Sep 21. PMID: 16990804.
11. Kono M, Suga Y, Akashi T, Ito Y, Takeichi T, Muro Y, Akiyama M. A Child with Epidermolytic Ichthyosis from a Parent with Epidermolytic Nevus: Risk Evaluation of Transmission from Mosaic to Germline. *J Invest Dermatol.* 2017 Sep;137(9):2024-2026. doi: 10.1016/j.jid.2017.04.036. Epub 2017 May 19. PMID: 28532675.
12. Su WP. Histopathologic varieties of epidermal nevus. A study of 160 cases. *Am J Dermatopathol.* 1982 Apr;4(2):161-70. doi: 10.1097/00000372-198204000-00011. PMID: 7048967.
13. Ersoy-Evans S, Sahin S, Mancini AJ, Paller AS, Gutart J. The acanthosis nigricans form of epidermal nevus. *J Am Acad Dermatol.* 2006 Oct;55(4):696-8. doi: 10.1016/j.jaad.2006.01.026. PMID: 17010754.
14. Brandling-Bennett HA, Morel KD. Epidermal nevi. *Pediatr Clin North Am.* 2010 Oct;57(5):1177-98. doi: 10.1016/j.pcl.2010.07.004. PMID: 20888465.
15. Dupre A, Christol B. Inflammatory linear verrucose epidermal nevus. A pathologic study. *Arch Dermatol.* 1977 Jun;113(6):767-9. PMID: 869546.
16. Hafner C, Klein A, Landthaler M, Vogt T. Clonality of basal cell carcinoma arising in an epidermal nevus. New insights provided by molecular analysis. *Dermatology.* 2009;218(3):278-81. doi: 10.1159/000189209. Epub 2008 Dec 22. PMID: 19122450.
17. Peramiquel L, Baselga E, Dalmau J, Roé E, del Mar Campos M, Alomar A. Lichen striatus: clinical and

- epidemiological review of 23 cases. *Eur J Pediatr.* 2006 Apr;165(4):267-9. doi: 10.1007/s00431-005-0032-9. Epub 2006 Jan 13. PMID: 16411095.
18. Hauber K, Rose C, Bröcker EB, Hamm H. Lichen striatus: clinical features and follow-up in 12 patients. *Eur J Dermatol.* 2000 Oct-Nov;10(7):536-9. PMID: 11056425.
  19. Saraswat A, Sandhu K, Shukla R, Handa S. Unilateral linear psoriasis with palmoplantar, nail, and scalp involvement. *Pediatr Dermatol.* 2004 Jan-Feb;21(1):70-3. doi: 10.1111/j.0736-8046.2004.21116.x. PMID: 14871332.
  20. Akelma AZ, Cizmeci MN, Kanburoglu MK, Mete E. A diagnostic dilemma: inflammatory linear verrucous epidermal nevus versus linear psoriasis. *J Pediatr.* 2013 Apr;162(4):879-879.e1. doi: 10.1016/j.jpeds.2012.10.038. Epub 2012 Nov 10. PMID: 23149170.
  21. Chien P Jr, Rosenman K, Cheung W, Wang N, Sanchez M. Linear psoriasis. *Dermatol Online J.* 2009 Aug 15;15(8):4. PMID: 19891912.
  22. Happle R. Linear psoriasis and ILVEN: is lumping or splitting appropriate? *Dermatology.* 2006;212(2):101-2. doi: 10.1159/000090647. PMID: 16484813.
  23. Lee BJ, Mancini AJ, Renucci J, Paller AS, Bauer BS. Full-thickness surgical excision for the treatment of inflammatory linear verrucous epidermal nevus. *Ann Plast Surg.* 2001 Sep;47(3):285-92. doi: 10.1097/00000637-200109000-00011. PMID: 11562033.
  24. Park JH, Hwang ES, Kim SN, Kye YC. Er:YAG laser treatment of verrucous epidermal nevi. *Dermatol Surg.* 2004 Mar;30(3):378-81. doi: 10.1111/j.1524-4725.2004.30104.x. PMID: 15008864.
  25. Thual N, Chevallier JM, Vuillamie M, Tack B, Leroy D, Dompmartin A. CO2 laser therapy of verrucous epidermal nevus. *Ann Dermatol Venereol.* 2006 Feb;133(2):131-8. doi: 10.1016/s0151-9638(06)70863-8. PMID: 16508596.
  26. Paradela S, Del Pozo J, Fernández-Jorge B, Lozano J, Martínez-González C, Fonseca E. Epidermal nevi treated by carbon dioxide laser vaporization: a series of 25 patients. *J Dermatolog Treat.* 2007;18(3):169-74. doi: 10.1080/09546630701335180. PMID: 17538806.
  27. Toyozawa S, Yamamoto Y, Kaminaka C, Kishiooka A, Yonei N, Furukawa F. Successful treatment with trichloroacetic acid peeling for inflammatory linear verrucous epidermal nevus. *J Dermatol.* 2010 Apr;37(4):384-6. doi: 10.1111/j.1346-8138.2010.00822.x. PMID: 20507413.
  28. Panagiotopoulos A, Chasapi V, Nikolaou V, Stavropoulos PG, Kafouros K, Petridis A, Katsambas A. Assessment of cryotherapy for the treatment of verrucous epidermal naevi. *Acta Derm Venereol.* 2009;89(3):292-4. doi: 10.2340/00015555-0610. PMID: 19479129.
  29. Kim JJ, Chang MW, Shwayder T. Topical tretinoin and 5-fluorouracil in the treatment of linear verrucous epidermal nevus. *J Am Acad Dermatol.* 2000 Jul;43(1 Pt 1):129-32. doi: 10.1067/mjd.2000.105563. PMID: 10863239.
  30. Koh MJ, Lee JS, Chong WS. Systematized epidermal nevus with epidermolytic hyperkeratosis improving with topical calcipotriol/beta-metasone dipropionate combination ointment. *Pediatr Dermatol.* 2013 May-Jun;30(3):370-3. doi: 10.1111/j.1525-1470.2012.01818.x. Epub 2012 Sep 3. PMID: 22938157.
  31. Herman AR, Scott RA. Systematized epidermal nevus treated with isotretinoin. *J Drugs Dermatol.* 2002 Sep;1(2):195-6. PMID: 12847745.
  32. Jaqueti G, Requena L, Sánchez Yus E. Trichoblastoma is the most common neoplasm developed in nevus sebaceus of Jadassohn: a clinicopathologic study of a series of 155 cases. *Am J Dermatopathol.* 2000 Apr;22(2):108-18. doi: 10.1097/00000372-200004000-00004. PMID: 10770429.
  33. Cribier B, Scrivener Y, Grosshans E. Tumors arising in nevus sebaceus: A study of 596 cases. *J Am Acad Dermatol.* 2000 Feb;42(2 Pt 1):263-8. doi: 10.1016/S0190-9622(00)90136-1. PMID: 10642683.
  34. Rosen H, Schmidt B, Lam HP, Meara JG, Labow BI. Management of nevus sebaceous and the risk of Basal cell carcinoma: an 18-year review. *Pediatr Dermatol.* 2009 Nov-Dec;26(6):676-81. doi: 10.1111/j.1525-1470.2009.00939.x. Epub 2009 Jul 20. PMID: 19686305.
  35. Person JR, Bentkover S, Longcope C. Androgen receptors are increased in nevus sebaceous. *J Am Acad Dermatol.* 1986 Jul;15(1):120-2. doi: 10.1016/s0190-9622(86)80153-0. PMID: 3722503.
  36. Hamilton KS, Johnson S, Smoller BR. The role of androgen receptors in the clinical course of nevus sebaceous of Jadassohn. *Mod Pathol.* 2001 Jun;14(6):539-42. doi: 10.1038/modpathol.3880346. PMID: 11406653.
  37. Mehregan AH, Pinkus H. Life History Of Organoid Nevi. Special Reference To Nevus Sebaceous Of Jadassohn. *Arch Dermatol.* 1965 Jun;91:574-88. doi: 10.1001/archderm.1965.01600120006002. PMID: 14295518.
  38. Domingo J, Helwig EB. Malignant neoplasms associated with nevus sebaceous of Jadassohn. *J Am Acad Dermatol.* 1979 Dec;1(6):545-56. doi: 10.1016/s0190-9622(79)80100-0. PMID: 528703.
  39. Chun K, Vázquez M, Sánchez JL. Nevus sebaceous: clinical outcome and considerations for prophylactic excision. *Int J Dermatol.* 1995 Aug;34(8):538-41. doi: 10.1111/j.1365-4362.1995.tb02948.x. PMID: 7591432.
  40. Duncan A, Wilson N, Leonard N. Squamous cell carcinoma developing in a naevus sebaceous of Jadassohn. *Am J Dermatopathol.* 2008 Jun;30(3):269-70. doi: 10.1097/DAD.0b013e318169cc69. PMID: 18496430.

41. Kazakov DV, Calonje E, Zelger B, Luzar B, Belousova IE, Mukensnabl P, Michal M. Sebaceous carcinoma arising in nevus sebaceus of Jadassohn: a clinicopathological study of five cases. *Am J Dermatopathol.* 2007 Jun;29(3):242-8. doi: 10.1097/DAD.0b013e3180339528. PMID: 17519621.
42. Misago N, Kodera H, Narisawa Y. Sebaceous carcinoma, trichoblastoma, and sebaceoma with features of trichoblastoma in nevus sebaceus. *Am J Dermatopathol.* 2001 Oct;23(5):456-62. doi: 10.1097/00000372-200110000-00012. PMID: 11801780.
43. Miller CJ, Ioffreda MD, Billingsley EM. Sebaceous carcinoma, basal cell carcinoma, trichoadenoma, trichoblastoma, and syringocystadenoma papilliferum arising within a nevus sebaceus. *Dermatol Surg.* 2004 Dec;30(12 Pt 2):1546-9. doi: 10.1111/j.1524-4725.2004.30552.x. PMID: 15606837.
44. Tarkhan II, Domingo J. Metastasizing eccrine porocarcinoma developing in a sebaceous nevus of Jadassohn. Report of a case. *Arch Dermatol.* 1985 Mar;121(3):413-5. PMID: 2983619.
45. Kantrow SM, Ivan D, Williams MD, Prieto VG, Lazar AJ. Metastasizing adenocarcinoma and multiple neoplastic proliferations arising in a nevus sebaceus. *Am J Dermatopathol.* 2007 Oct;29(5):462-6. doi: 10.1097/DAD.0b013e31814a50c6. PMID: 17890915.
46. Manonukul J, Omeapinyan P, Vongjirad A. Mucoepidermoid (adenosquamous) carcinoma, trichoblastoma, trichilemmoma, sebaceous adenoma, tumor of follicular infundibulum and syringocystadenoma papilliferum arising within 2 persistent lesions of nevus sebaceus: report of a case. *Am J Dermatopathol.* 2009 Oct;31(7):658-63. doi: 10.1097/DAD.0b013e31817903f5. PMID: 19652586.
47. Barkham MC, White N, Brundler MA, Richard B, Moss C. Should naevus sebaceus be excised prophylactically? A clinical audit. *J Plast Reconstr Aesthet Surg.* 2007;60(11):1269-70. doi: 10.1016/j.bjps.2007.03.038. Epub 2007 Aug 17. PMID: 17707706.
48. Saedi T, Cetas J, Chang R, Krol A, Selden NR. Newborn with sebaceous nevus of jadassohn presenting as exophytic scalp lesion. *Pediatr Neurosurg.* 2008;44(2):144-7. doi: 10.1159/000113118. Epub 2008 Jan 24. PMID: 18230930.
49. Mckee PH, Calonje E, Granter SR. Nevus sebaceus. In: Mckee PH, Calonje E, Granter SR, editors. London: Elsevier Mosby; 2005. p. 1568
50. Eisen DB, Michael DJ. Sebaceous lesions and their associated syndromes: part I. *J Am Acad Dermatol.* 2009 Oct;61(4):549-60; quiz 561-2. doi: 10.1016/j.jaad.2009.04.058. PMID: 19751879.
51. Orchard DC, Weston WL, Morelli JG. Tumors arising in nevus sebaceus. *J Am Acad Dermatol.* 2001 Nov;45(5):793-4; author reply 794. doi: 10.1067/mjd.2001.117241. PMID: 11606942.
52. Ashinoff R. Linear nevus sebaceus of Jadassohn treated with the carbon dioxide laser. *Pediatr Dermatol.* 1993 Jun;10(2):189-91. doi: 10.1111/j.1525-1470.1993.tb00053.x. PMID: 8346119.
53. Dierickx CC, Goldenhersh M, Dwyer P, Stratigos A, Mihm M, Anderson RR. Photodynamic therapy for nevus sebaceus with topical delta-aminolevulinic acid. *Arch Dermatol.* 1999 Jun;135(6):637-40. doi: 10.1001/archderm.135.6.637. PMID: 10376688.
54. Cestari TF, Rubim M, Valentini BC. Nevus comedonicus: case report and brief review of the literature. *Pediatr Dermatol.* 1991 Dec;8(4):300-5. doi: 10.1111/j.1525-1470.1991.tb00938.x. PMID: 1792203.
55. Kirtak N, Inaloz HS, Karakok M, Erguven HG, Ozgoztasi O. Extensive inflammatory nevus comedonicus involving half of the body. *Int J Dermatol.* 2004 Jun;43(6):434-6. doi: 10.1111/j.1365-4632.2004.02188.x. PMID: 15186225.
56. Lefkowitz A, Schwartz RA, Lambert WC. Nevus comedonicus. *Dermatology.* 1999;199(3):204-7. doi: 10.1159/000018275. PMID: 10592398.
57. Wakahara M, Kiyohara T, Kumakiri M, Kuwahara H, Fujita T. Bilateral nevus comedonicus: efficacy of topical tacalcitol ointment. *Acta Derm Venereol.* 2003;83(1):51. PMID: 12636024.
58. Deliduka SB, Kwong PC. Treatment of Nevus comedonicus with topical tazarotene and calcipotriene. *J Drugs Dermatol.* 2004 Nov-Dec;3(6):674-6. PMID: 15624751.
59. Vasiloudes PE, Morelli JG, Weston WL. Inflammatory nevus comedonicus in children. *J Am Acad Dermatol.* 1998 May;38(5 Pt 2):834-6. doi: 10.1016/s0190-9622(98)70469-4. PMID: 9591797.
60. Alpsoy E, Durusoy C, Ozbilim G, Karpuzoğlu G, Yilmaz E. Nevus comedonicus syndrome: a case associated with multiple basal cell carcinomas and a rudimentary toe. *Int J Dermatol.* 2005 Jun;44(6):499-501. doi: 10.1111/j.1365-4632.2005.02055.x. PMID: 15941441.
61. Walling HW, Swick BL. Squamous cell carcinoma arising in nevus comedonicus. *Dermatol Surg.* 2009 Jan;35(1):144-6. doi: 10.1111/j.1524-4725.2008.34396.x. Epub 2008 Dec 6. PMID: 19076196.
62. Milton GP, DiGiovanna JJ, Peck GL. Treatment of nevus comedonicus with ammonium lactate lotion. *J Am Acad Dermatol.* 1989 Feb;20(2 Pt 2):324-8. doi: 10.1016/s0190-9622(89)70039-6. PMID: 2915076.
63. Caers SJ, Van der Geer S, Beverdam EG, Krekels GA, Ostertag JU. Successful treatment of nevus comedonicus with the use of the Erbium Yag laser. *J Eur Acad Dermatol Venereol.* 2008 Mar;22(3):375-7. doi: 10.1111/j.1468-3083.2007.02331.x. Epub 2007 Nov 14. PMID: 18005024.
64. Tymen R, Forestier JF, Boutet B, Colomb D. Late Becker's nevus. One hundred cases (author's transl). *Ann Dermatol Venereol.* 1981;108(1):41-6. PMID: 7235503.
65. Ballone E, Fazio P, Lappa G, Di Mascio R, Di Mascio C, Schioppa F. Prevalence of Becker's nevi in a population of young men in central Italy. *J Am Acad Dermatol.* 2003 May;48(5):795. doi: 10.1067/mjd.2003.182. PMID: 12734514.

66. Ingordo V, Gentile C, Iannazzone SS, Cusano F, Naldi L. The 'EpiEnlist' project: a dermo-epidemiologic study on a representative sample of young Italian males. Prevalence of selected pigmentary lesions. *J Eur Acad Dermatol Venereol*. 2007 Sep;21(8):1091-6. doi: 10.1111/j.1468-3083.2007.02205.x. PMID: 17714131.
67. Hsu S, Chen JY, Subrt P. Becker's melanosis in a woman. *J Am Acad Dermatol*. 2001 Dec;45(6 Suppl):S195-6. doi: 10.1067/mjd.2001.101882. PMID: 11712056.
68. Happle R. Epidermal nevus syndromes. *Semin Dermatol* 1995;14(2):111-21.
69. Sood A, D'Souza P, Verma KK. Becker's naevus occurring at birth and in early childhood. *Acta Derm Venereol*. 1998 Jul;78(4):311. doi: 10.1080/000155598442034. PMID: 9689312.
70. Book SE, Glass AT, Laude TA. Congenital Becker's nevus with a familial association. *Pediatr Dermatol*. 1997 Sep-Oct;14(5):373-5. doi: 10.1111/j.1525-1470.1997.tb00985.x. PMID: 9336809.
71. Kiliç A, Kaya I, Gül U, Soylu S, Gönül M, Demiriz M. Becker nevus on face with asymmetrical growth of beard hair. *J Eur Acad Dermatol Venereol*. 2008 Feb;22(2):246-7. doi: 10.1111/j.1468-3083.2007.02301.x. PMID: 18211427.
72. de Almeida HL Jr, Happle R. Two cases of cephalic Becker nevus with asymmetrical growth of beard or scalp hair. *Dermatology*. 2003;207(3):337-8. doi: 10.1159/000073107. PMID: 14571087.
73. Alhusayan R, Kanigsberg N, Jackson R. Becker nevus on the lower limb: case report and review of the literature. *J Cutan Med Surg*. 2008 Jan-Feb;12(1):31-4. doi: 10.2310/7750.2007.00034. PMID: 18258146.
74. Khaitan BK, Manchanda Y, Mittal R, Singh MK. Multiple Becker's naevi: a rare presentation. *Acta Derm Venereol*. 2001 Oct-Nov;81(5):374-5. doi: 10.1080/000155501317140179. PMID: 11800153.
75. Khatami A, Seradj MH, Gorouhi F, Firooz A, Dowlati Y. Giant bilateral becker nevus: a rare presentation. *Pediatr Dermatol*. 2008 Jan-Feb;25(1):47-51. doi: 10.1111/j.1525-1470.2007.00581.x. PMID: 18304153.
76. Grim KD, Wasko CA. Symmetrical bilateral Becker melanosis: A rare presentation. *Dermatol Online J*. 2009 Dec 15;15(12):1. PMID: 20040251.
77. Person JR, Longcope C. Becker's nevus: an androgen-mediated hyperplasia with increased androgen receptors. *J Am Acad Dermatol*. 1984 Feb;10(2 Pt 1):235-8. doi: 10.1016/s0190-9622(84)70029-6. PMID: 6715592.
78. Grande Sarpa H, Harris R, Hansen CD, Callis Duffin KP, Florell SR, Hadley ML. Androgen receptor expression patterns in Becker's nevi: an immunohistochemical study. *J Am Acad Dermatol*. 2008 Nov;59(5):834-8. doi: 10.1016/j.jaad.2008.06.028. PMID: 19119099.
79. Nirdé P, Dereure O, Belon C, Lumbroso S, Guilhaou JJ, Sultan C. The association of Becker nevus with hypersensitivity to androgens. *Arch Dermatol*. 1999 Feb;135(2):212-4. doi: 10.1001/archderm.135.2.212-a. PMID: 10052418.
80. Mckee PH, Calonje E, Granter SR. Becker's nevus. In: Mckee PH, Calonje E, Granter SR, editors. London: Elsevier Mosby; 2005. p. 1249.
81. Fehr B, Panizzon RG, Schnyder UW. Becker's nevus and malignant melanoma. *Dermatologica*. 1991;182(2):77-80. doi: 10.1159/000247749. PMID: 2050238.
82. Honda M, Suzuki T, Kudoh K, Tagami H. Bowen's disease developing within a Becker's melanosis (Becker's naevus). *Br J Dermatol*. 1997 Oct;137(4):659-61. doi: 10.1111/j.1365-2133.1997.tb03817.x. PMID: 9390361.
83. Patrizi A, Medri M, Neri I, Fanti PA. Becker nevus associated with basal cell carcinoma, melanocytic naevus and smooth-muscle hamartoma. *J Eur Acad Dermatol Venereol*. 2007 Jan;21(1):130-2. doi: 10.1111/j.1468-3083.2006.01815.x. PMID: 17207195.
84. Trelles MA, Allones I, Moreno-Arias GA, Vélez M. Becker's naevus: a comparative study between erbium: YAG and Q-switched neodymium:YAG; clinical and histopathological findings. *Br J Dermatol*. 2005 Feb;152(2):308-13. doi: 10.1111/j.1365-2133.2004.06259.x. PMID: 15727644.
85. Choi JE, Kim JW, Seo SH, Son SW, Ahn HH, Kye YC. Treatment of Becker's nevi with a long-pulse alexandrite laser. *Dermatol Surg*. 2009 Jul;35(7):1105-8. doi: 10.1111/j.1524-4725.2009.01195.x. Epub 2009 Apr 28. PMID: 19438687.
86. Glaich AS, Goldberg LH, Dai T, Kunishige JH, Friedman PM. Fractional resurfacing: a new therapeutic modality for Becker's nevus. *Arch Dermatol*. 2007 Dec;143(12):1488-90. doi: 10.1001/archderm.143.12.1488. PMID: 18086996.
87. Morag C, Metzker A. Inflammatory linear verrucous epidermal nevus: report of seven new cases and review of the literature. *Pediatr Dermatol*. 1985 Nov;3(1):15-8. doi: 10.1111/j.1525-1470.1985.tb00479.x. PMID: 3906609.
88. Lee SH, Rogers M. Inflammatory linear verrucous epidermal naevi: a review of 23 cases. *Australas J Dermatol*. 2001 Nov;42(4):252-6. doi: 10.1046/j.1440-0960.2001.00530.x. PMID: 11903156.
89. Lee BJ, Mancini AJ, Renucci J, Paller AS, Bauer BS. Full-thickness surgical excision for the treatment of inflammatory linear verrucous epidermal nevus. *Ann Plast Surg*. 2001 Sep;47(3):285-92. doi: 10.1097/0000637-200109000-00011. PMID: 11562033.
90. Cambiaghi S, Gianotti R, Caputo R. Widespread porokeratotic eccrine ostial and dermal duct nevus along Blaschko lines. *Pediatr Dermatol*. 2007 Mar-Apr;24(2):162-7. doi: 10.1111/j.1525-1470.2007.00367.x. PMID: 17461816.

91. Goddard DS, Rogers M, Frieden IJ, Krol AL, White CR Jr, Jayaraman AG, Robinson-Bostom L, Bruckner AL, Ruben BS. Widespread porokeratotic adnexal ostial nevus: clinical features and proposal of a new name unifying porokeratotic eccrine ostial and dermal duct nevus and porokeratotic eccrine and hair follicle nevus. *J Am Acad Dermatol.* 2009 Dec;61(6):1060.e1-14. doi: 10.1016/j.jaad.2009.03.036. Epub 2009 Aug 6. PMID: 19664847.
92. Jamora MJ, Celis MA. Generalized porokeratotic eccrine ostial and dermal duct nevus associated with deafness. *J Am Acad Dermatol.* 2008 Aug;59(2 Suppl 1):S43-5. doi: 10.1016/j.jaad.2007.09.034. PMID: 18625383.
93. Valks R, Abajo P, Fraga J, Aragüés M, García-Diez A. Porokeratotic eccrine ostial and dermal duct nevus of late onset: more frequent than previously suggested? *Dermatology.* 1996;193(2):138-40. doi: 10.1159/000246230. PMID: 8884152.
94. Birol A, Erkek E, Bozdothan O, Koçak M, Atasoy P. A case of porokeratotic eccrine ostial and dermal duct naevus of late onset. *J Eur Acad Dermatol Venereol.* 2004 Sep;18(5):619-21. doi: 10.1111/j.1468-3083.2004.01019.x. PMID: 15324411.
95. van de Kerkhof PC, Steijlen PM, Happle R. Co-occurrence of linear psoriasis and porokeratotic eccrine ostial and dermal duct naevus. *Acta Derm Venereol.* 1993 Aug;73(4):311-2. doi: 10.2340/0001555573310. PMID: 7904109.
96. Yu HJ, Ko JY, Kwon HM, Kim JS. Linear psoriasis with porokeratotic eccrine ostial and dermal duct nevus. *J Am Acad Dermatol.* 2004 May;50(5 Suppl):S81-3. doi: 10.1016/s0190-9622(03)02797-x. PMID: 15097935.
97. Nassiri N, Hansen J. Diffuse squamous cell carcinoma in porokeratotic eccrine ostial and dermal duct nevus. *Plast Reconstr Surg.* 2009 Feb;123(2):87e-88e. doi: 10.1097/PRS.0b013e31819596ea. PMID: 19182594.
98. Coras B, Vogt T, Roesch A, Landthaler M, Hohenleutner U. Bowen's disease on porokeratotic eccrine ostial and dermal duct nevus. *Dermatol Surg.* 2007 Apr;33(4):496-9. doi: 10.1111/j.1524-4725.2007.33099.x. PMID: 17430387.
99. Solis J, Sau P, James WD. Puzzling palmar and plantar papules. Porokeratotic eccrine ostial and dermal duct nevus. *Arch Dermatol.* 1991 Aug;127(8):1220-1, 1223-4. doi: 10.1001/archderm.127.8.1220. PMID: 1863085.
100. Sassmannshausen J, Bogomilsky J, Chaffins M. Porokeratotic eccrine ostial and dermal duct nevus: a case report and review of the literature. *J Am Acad Dermatol.* 2000 Aug;43(2 Pt 2):364-7. doi: 10.1067/mjd.2000.100536. PMID: 10901725.
101. Masferrer E, Vicente MA, Bassas-Vila J, Rovira C, González-Enseñat MA. Porokeratotic eccrine ostial and dermal duct naevus: report of 10 cases. *J Eur Acad Dermatol Venereol.* 2010 Jul;24(7):847-51. doi: 10.1111/j.1468-3083.2009.03498.x. Epub 2009 Nov 18. PMID: 19925595.
102. Leung CS, Tang WY, Lam WY, Fung WK, Lo KK. Porokeratotic eccrine ostial and dermal duct naevus with dermatomal trunk involvement: literature review and report on the efficacy of laser treatment. *Br J Dermatol.* 1998 Apr;138(4):684-8. doi: 10.1046/j.1365-2133.1998.02186.x. PMID: 9640380.
103. Del Pozo J, Martínez W, Vereá MM, Yebra-Pimentel MT, García-Silva J, Fonseca E. Porokeratotic eccrine ostial and dermal duct naevus: treatment with carbon dioxide laser. *Br J Dermatol.* 1999 Dec;141(6):1144-5. doi: 10.1046/j.1365-2133.1999.03230.x. PMID: 10722274.
104. Davies D, Rogers M. Review of neurological manifestations in 196 patients with sebaceous naevi. *Australas J Dermatol.* 2002 Feb;43(1):20-3. doi: 10.1046/j.1440-0960.2002.00546.x. PMID: 11869203.
105. van de Warrenburg BP, van Gulik S, Renier WO, Lammens M, Doelman JC. The linear naevus sebaceus syndrome. *Clin Neurol Neurosurg.* 1998 Jun;100(2):126-32. doi: 10.1016/s0303-8467(98)00012-2. PMID: 9746301.
106. Menascu S, Donner EJ. Linear nevus sebaceus syndrome: case reports and review of the literature. *Pediatr Neurol.* 2008 Mar;38(3):207-10. doi: 10.1016/j.pediatrneurol.2007.10.012. PMID: 18279757.
107. Vidaurri-de la Cruz H, Tamayo-Sánchez L, Durán-McKinster C, de la Luz Orozco-Covarrubias M, Ruiz-Maldonado R. Epidermal nevus syndromes: clinical findings in 35 patients. *Pediatr Dermatol.* 2004 Jul-Aug;21(4):432-9. doi: 10.1111/j.0736-8046.2004.21402.x. PMID: 15283784.
108. Zutt M, Strutz F, Happle R, Habenicht EM, Emmert S, Haenssle HA, Kretschmer L, Neumann Ch. Schimmelpenning-Feuerstein-Mims syndrome with hypophosphatemic rickets. *Dermatology.* 2003;207(1):72-6. doi: 10.1159/000070948. PMID: 12835555.
109. Warnke PH, Schimmelpenning GW, Happle R, Springer IN, Hauschild A, Wiltfang J, Acil Y, Sherry E, Proksch E, Luetges J, Russo PA. Intraoral lesions associated with sebaceous nevus syndrome. *J Cutan Pathol.* 2006 Feb;33(2):175-80. doi: 10.1111/j.0303-6987.2006.00374.x. PMID: 16420315.
110. Happle R, König A. Didymosis aplasticosebacea: coexistence of aplasia cutis congenita and nevus sebaceus may be explained as a twin spot phenomenon. *Dermatology.* 2001;202(3):246-8. doi: 10.1159/000051645. PMID: 11385232.
111. Laino L, Steensel MA, Innocenzi D, Camplone G. Familial occurrence of nevus sebaceus of Jadassohn: another case of paradominant inheritance? *Eur J Dermatol.* 2001 Mar-Apr;11(2):97-8. PMID: 11275801.
112. Demerdjjeva Z, Kavaklieva S, Tsankov N. Epidermal nevus syndrome and didymosis aplasticosebacea. *Pediatr Dermatol.* 2007 Sep-Oct;24(5):514-6. doi: 10.1111/j.1525-1470.2007.00507.x. PMID: 17958800.

113. Lam J, Dohil MA, Eichenfield LF, Cunningham BB. SCALP syndrome: sebaceous nevus syndrome, CNS malformations, aplasia cutis congenita, limbal dermoid, and pigmented nevus (giant congenital melanocytic nevus) with neurocutaneous melanosis: a distinct syndromic entity. *J Am Acad Dermatol*. 2008 May;58(5):884-8. doi: 10.1016/j.jaad.2007.09.029. Epub 2008 Jan 30. PMID: 18234391.
114. Eisen DB, Michael DJ. Sebaceous lesions and their associated syndromes: part II. *J Am Acad Dermatol*. 2009 Oct;61(4):563-78; quiz 579-80. doi: 10.1016/j.jaad.2009.04.059. PMID: 19751880.
115. Happle R, Hoffmann R, Restano L, Caputo R, Tadini G. Phacomatosis pigmentokeratolica: a melanocytic-epidermal twin nevus syndrome. *Am J Med Genet*. 1996 Nov 11;65(4):363-5. doi: 10.1002/(SICI)1096-8628(19961111)65:4<363::AID-AJMG27>3.0.CO;2-R. PMID: 8923953.
116. Happle R. Speckled lentiginous nevus syndrome: delineation of a new distinct neurocutaneous phenotype. *Eur J Dermatol*. 2002 Mar-Apr;12(2):133-5. PMID: 11872407.
117. Hermes B, Cremer B, Happle R, Henz BM. Phacomatosis pigmentokeratolica: a patient with the rare melanocytic-epidermal twin nevus syndrome. *Dermatology*. 1997;194(1):77-9. doi: 10.1159/000246065. PMID: 9031800.
118. Boente MC, Pizzi de Parra N, Larralde de Luna M, Bonet HB, Santos Muñoz A, Parra V, Gramajo P, Moreno S, Asial RA. Phacomatosis pigmentokeratolica: another epidermal nevus syndrome and a distinctive type of twin spotting. *Eur J Dermatol*. 2000 Apr-May;10(3):190-4. PMID: 10725816.
119. Tadini G, Restano L, Gonzáles-Pérez R, Gonzáles-Enseñat A, Vincente-Villa MA, Cambiaghi S, Marchettini P, Mastrangelo M, Happle R. Phacomatosis pigmentokeratolica: report of new cases and further delineation of the syndrome. *Arch Dermatol*. 1998 Mar;134(3):333-7. doi: 10.1001/archderm.134.3.333. PMID: 9580120.
120. Hill VA, Felix RH, Mortimer PS, Harper JJ. Phacomatosis pigmentokeratolica. *J R Soc Med*. 2003 Jan;96(1):30-1. doi: 10.1258/jrsm.96.1.30. PMID: 12519801; PMCID: PMC539370.
121. Vente C, Neumann C, Bertsch H, Rupprecht R, Happle R. Speckled lentiginous nevus syndrome: report of a further case. *Dermatology*. 2004;209(3):228-9. doi: 10.1159/000079895. PMID: 15459538.
122. Boente Mdel C, Asial RA, Happle R. Phacomatosis pigmentokeratolica: a follow-up report documenting additional cutaneous and extracutaneous anomalies. *Pediatr Dermatol*. 2008 Jan-Feb;25(1):76-80. doi: 10.1111/j.1525-1470.2007.00588.x. PMID: 18304160.
123. Happle R. Speckled lentiginous nevus syndrome: delineation of a new distinct neurocutaneous phenotype. *Eur J Dermatol*. 2002 Mar-Apr;12(2):133-5. PMID: 11872407.
124. Piqué E, Aguilar A, Fariña MC, Gallego MA, Escalonnilla P, Requena L. Partial unilateral lentiginosis: report of seven cases and review of the literature. *Clin Exp Dermatol*. 1995 Jul;20(4):319-22. doi: 10.1111/j.1365-2230.1995.tb01333.x. PMID: 8548990.
125. Gruson LM, Orlow SJ, Schaffer JV. Phacomatosis pigmentokeratolica associated with hemihypertrophy and a rhabdomyosarcoma of the abdominal wall. *J Am Acad Dermatol*. 2006 Aug;55(2 Suppl):S16-20. doi: 10.1016/j.jaad.2005.08.045. PMID: 16843117.
126. Okada E, Tamura A, Ishikawa O. Phacomatosis pigmentokeratolica complicated with juvenile onset hypertension. *Acta Derm Venereol*. 2004;84(5):397-8. doi: 10.1080/00015550410029260. PMID: 15370709.
127. Torrelo A, Zambrano A. What syndrome is this. Phacomatosis pigmentokeratolica (Happle). *Pediatr Dermatol*. 1998 Jul-Aug;15(4):321-3. doi: 10.1046/j.1525-1470.1998.1998015321.x. PMID: 9720704.
128. Wollenberg A, Butnaru C, Oppel T. Phacomatosis pigmentokeratolica (Happle) in a 23-year-old man. *Acta Derm Venereol*. 2002;82(1):55-7. doi: 10.1080/000155502753600911. PMID: 12013201.
129. Majmudar V, Loffeld A, Happle R, Salim A. Phacomatosis pigmentokeratolica associated with a suprasellar dermoid cyst and leg hypertrophy. *Clin Exp Dermatol*. 2007 Nov;32(6):690-2. doi: 10.1111/j.1365-2230.2007.02511.x. Epub 2007 Sep 14. PMID: 17868392.
130. Martínez-Menchón T, Mahiques Santos L, Vilata Corell JJ, Febrer Bosch I, Fortea Baixauli JM. Phacomatosis pigmentokeratolica: a 20-year follow-up with malignant degeneration of both nevus components. *Pediatr Dermatol*. 2005 Jan-Feb;22(1):44-7. doi: 10.1111/j.1525-1470.2005.22110.x. PMID: 15660897.
131. Bouthors J, Vantghem MC, Manouvrier-Hanu S, Soudan B, Proust E, Happle R, Piette F. Phacomatosis pigmentokeratolica associated with hypophosphatemic rickets, pheochromocytoma and multiple basal cell carcinomas. *Br J Dermatol*. 2006 Jul;155(1):225-6. doi: 10.1111/j.1365-2133.2006.07313.x. PMID: 16792789.
132. Engber PB. The nevus comedonicus syndrome: a case report with emphasis on associated internal manifestations. *Int J Dermatol*. 1978 Nov;17(9):745-9. doi: 10.1111/ijd.1978.17.9.745. PMID: 730460.
133. Engber PB. Nevus comedonicus syndrome. *Arch Dermatol*. 1982 Jan;118(1):1. PMID: 7059193.
134. Hsu S, Nikko A. Unilateral atrophic skin lesion with features of atrophoderma vermiculatum: a variant of the epidermal nevus syndrome? *J Am Acad Dermatol*. 2000 Aug;43(2 Pt 1):310-2. doi: 10.1067/mjd.2000.106514. PMID: 10906657.
135. Martinez M, Levrero P, Bazzano C, Larre Borges A, De Anda G. Nevus comedonicus syndrome in a woman with Paget bone disease and breast cancer: a mere coincidence? *Eur J Dermatol*. 2006 Nov-Dec;16(6):697-8. PMID: 17229620.

136. Whyte HJ. Unilateral comedo nevus and cataract. *Arch Dermatol.* 1968 May;97(5):533-5. PMID: 5647028.
137. Mutton P, Lewis M, Harley J, Hertzberg R. Comedo naevus: an unusual association of infantile cataract. *Aust Paediatr J.* 1975 Mar;11(1):46-8. doi: 10.1111/j.1440-1754.1975.tb02275.x. PMID: 1137529.
138. Suite M, Mahabir R. Unilateral comedo nevus associated with ipsilateral congenital cataract. *Int J Dermatol.* 1994 May;33(5):390-1. doi: 10.1111/j.1365-4362.1994.tb01079.x. PMID: 8039983.
139. El Ghelbazouri N, Ismaili N, Ahrich N, Benarafa A, Senouci K, Mansouri F, Hassam B. Nevus comedonicus syndrome. *Ann Dermatol Venereol.* 2007 Aug-Sep;134(8-9):663-6. doi: 10.1016/s0151-9638(07)91829-3. PMID: 17925691.
140. Patrizi A, Neri I, Fiorentini C, Marzaduri S. Nevus comedonicus syndrome: a new pediatric case. *Pediatr Dermatol.* 1998 Jul-Aug;15(4):304-6. doi: 10.1046/j.1525-1470.1998.1998015304.x. PMID: 9720699.
141. Schneider C. Clinical aspects of nevus comedonicus. *Hautarzt.* 1975 Mar;26(3):153-4. PMID: 123891.
142. Seo YJ, Piao YJ, Suhr KB, Lee JH, Park JK. A case of nevus comedonicus syndrome associated with neurologic and skeletal abnormalities. *Int J Dermatol.* 2001 Oct;40(10):648-50. doi: 10.1046/j.1365-4362.2001.01287-2.x. PMID: 11737427.
143. Filosa G, Bugatti L, Ciattaglia G, Salaffi F, Carotti M. Naevus comedonicus as dermatologic hallmark of occult spinal dysraphism. *Acta Derm Venereol.* 1997 May;77(3):243. doi: 10.2340/0001555577243. PMID: 9188888.
144. Happle R, Rogers M. Epidermal nevi. *Adv Dermatol.* 2002;18:175-201. PMID: 12528406.
145. Schauder S, Hanefeld F, Noske UM, Zoll B. Depigmented hypertrichosis following Blaschko's lines associated with cerebral and ocular malformations: a new neurocutaneous, autosomal lethal gene syndrome from the group of epidermal naevus syndromes? *Br J Dermatol.* 2000 Jun;142(6):1204-7. doi: 10.1046/j.1365-2133.2000.03551.x. PMID: 10848748.
146. Primc N, Boente MC, Asial R, Happle R. Angora hair nevus: an unusual type of epidermal nevus characterized by a lanugolike depigmented hypertrichosis. Presented in poster form (5975) at the 21st World Congress of Dermatology, Buenos Aires, Argentina, September 30-October 5, 2007.
147. Happle R, Koopman RJ. Becker nevus syndrome. *Am J Med Genet.* 1997 Jan 31;68(3):357-61. PMID: 9024572.
148. Haneke E. The dermal component in melanosis naeviformis Becker. *J Cutan Pathol.* 1979 Feb;6(1):53-8. doi: 10.1111/j.1600-0560.1979.tb00305.x. PMID: 438394.
149. Al Aboud K, Al Hawsawi K. Becker nevus on the hand. *Eur J Dermatol.* 2002 Nov-Dec;12(6):588. PMID: 12506884.
150. Danarti R, König A, Salhi A, Bittar M, Happle R. Becker's nevus syndrome revisited. *J Am Acad Dermatol.* 2004 Dec;51(6):965-9. doi: 10.1016/j.jaad.2004.06.036. PMID: 15583590.
151. Hoon Jung J, Chan Kim Y, Joon Park H, Woo Cinn Y. Becker's nevus with ipsilateral breast hypoplasia: improvement with spironolactone. *J Dermatol.* 2003 Feb;30(2):154-6. doi: 10.1111/j.1346-8138.2003.tb00365.x. PMID: 12692385.
152. Sharma R, Mishra A. Becker's naevus with ipsilateral areolar hypoplasia in three males. *Br J Dermatol.* 1997 Mar;136(3):471-2. doi: 10.1111/j.1365-2133.1997.tb14976.x. PMID: 9115944.
153. Crone AM, James MP. Giant Becker's naevus with ipsilateral areolar hypoplasia and limb asymmetry. *Clin Exp Dermatol.* 1997 Sep;22(5):240-1. PMID: 9536547.
154. Alfaro A, Torrelo A, Hernández A, Zambrano A, Happle R. Becker nevus syndrome. *Actas Dermosifiliogr.* 2007 Nov;98(9):624-6. PMID: 17961453.
155. Urbani CE, Betti R. Supernumerary nipple in association with Becker nevus vs. Becker nevus syndrome: a semantic problem only. *Am J Med Genet.* 1998 Apr 28;77(1):76-7. doi: 10.1002/(sici)1096-8628(19980428)77:1<76::aid-ajmg16>3.0.co;2-l. PMID: 9557899.
156. Van Gerwen HJ, Koopman RJ, Steijlen PM, Happle R. Becker's naevus with localized lipoatrophy and ipsilateral breast hypoplasia. *Br J Dermatol.* 1993 Aug;129(2):213. PMID: 7654589.
157. Cox NH. Becker's naevus of the thigh with lipoatrophy: report of two cases. *Clin Exp Dermatol.* 2002 Jan;27(1):27-8. doi: 10.1046/j.0307-6938.2001.00961.x. PMID: 11952664.
158. Szyliet JA, Grossman ME, Luyando Y, Olarte MR, Nagler H. Becker's nevus and an accessory scrotum. A unique occurrence. *J Am Acad Dermatol.* 1986 May;14(5 Pt 2):905-7. doi: 10.1016/s0190-9622(86)70109-6. PMID: 3711399.
159. Glinick SE, Alper JC, Bogaars H, Brown JA. Becker's melanosis: associated abnormalities. *J Am Acad Dermatol.* 1983 Oct;9(4):509-14. doi: 10.1016/s0190-9622(83)70161-1. PMID: 6355214.
160. Maessen-Visch MB, Hulsmans RF, Hulsmans FJ, Neumann HA. Melanosis naeviformis of Becker and scoliosis: a coincidence? *Acta Derm Venereol.* 1997 Mar;77(2):135-6. doi: 10.2340/0001555577135136. PMID: 9111825.
161. Naranjo R, Delgado V, de Dulanto F, Herrera E, Armijo M. Becker's melanosis. *Actas Dermosifiliogr.* 1980 Sep-Oct;71(9-10):331-6. PMID: 7223521.
162. Copeman PW, Jones EW. Pigmented hairy epidermal nevus (Becker). *Arch Dermatol.* 1965 Sep;92(3):249-51. PMID: 11851244.
163. Glinick SE, Alper JA. Spectrum of Becker's melanosis changes is greater than believed. *Arch Dermatol.* 1986 Apr;122(4):375. PMID: 3954401.
164. Lucky AW, Saruk M, Lerner AB. Becker's nevus associated with limb asymmetry. *Arch Dermatol.* 1981 Apr;117(4):243. PMID: 7212749.



165. Lambert JR, Willems P, Abs R, Van Roy B. Becker's nevus associated with chromosomal mosaicism and congenital adrenal hyperplasia. *J Am Acad Dermatol.* 1994 Apr;30(4):655-7. doi: 10.1016/s0190-9622(09)80118-7. PMID: 8157797.
166. Moore JA, Schosser RH. Becker's melanosis and hypoplasia of the breast and pectoralis major muscle. *Pediatr Dermatol.* 1985 Nov;3(1):34-7. doi: 10.1111/j.1525-1470.1985.tb00482.x. PMID: 4070085.
167. Kim HJ, Kim KD, Lee MH. Becker's melanosis associated with fibrous dysplasia. *Int J Dermatol.* 2002 Jul;41(7):384-6. doi: 10.1046/j.1365-4362.2002.01439\_1.x. PMID: 12121546.
168. Jones AC, Ford MJ. Simultaneous occurrence of segmental odontomaxillary dysplasia and Becker's nevus. *J Oral Maxillofac Surg.* 1999 Oct;57(10):1251-4. doi: 10.1016/s0278-2391(99)90498-6. PMID: 10513874.
169. Fretzin DF, Whitney D. Familial Becker's nevus. *J Am Acad Dermatol.* 1985 Mar;12(3):589-90. doi: 10.1016/s0190-9622(85)80107-9. PMID: 3989022.
170. Panizzon R, Schnyder UW. Familial Becker's nevus. *Dermatologica.* 1988;176(6):275-6. PMID: 3042481.
171. Jain HC, Fisher BK. Familial Becker's nevus. *Int J Dermatol.* 1989 May;28(4):263-4. doi: 10.1111/j.1365-4362.1989.tb04818.x. PMID: 2722340.
172. Panizzon RG. Familial Becker's nevus. *Int J Dermatol.* 1990 Mar;29(2):158. doi: 10.1111/j.1365-4362.1990.tb04101.x. PMID: 2182553.
173. Book SE, Glass AT, Laude TA. Congenital Becker's nevus with a familial association. *Pediatr Dermatol.* 1997 Sep-Oct;14(5):373-5. doi: 10.1111/j.1525-1470.1997.tb00985.x. PMID: 9336809.
174. Biesecker LG, Happle R, Mulliken JB, Weksberg R, Graham JM Jr, Viljoen DL, Cohen MM Jr. Proteus syndrome: diagnostic criteria, differential diagnosis, and patient evaluation. *Am J Med Genet.* 1999 Jun 11;84(5):389-95. doi: 10.1002/(sici)1096-8628(19990611)84:5<389::aid-ajmg1>3.0.co;2-o. PMID: 10360391.
175. Biesecker L. The challenges of Proteus syndrome: diagnosis and management. *Eur J Hum Genet.* 2006 Nov;14(11):1151-7. doi: 10.1038/sj.ejhg.5201638. Epub 2006 Aug 2. PMID: 16883308.
176. Cohen MM Jr. Proteus syndrome: an update. *Am J Med Genet C Semin Med Genet.* 2005 Aug 15;137C(1):38-52. doi: 10.1002/ajmg.c.30063. PMID: 16010681.
177. Happle R. Type 2 segmental Cowden disease vs. Proteus syndrome. *Br J Dermatol.* 2007 May;156(5):1089-90. doi: 10.1111/j.1365-2133.2007.07818.x. Epub 2007 Mar 28. PMID: 17388921.
178. Schepis C, Greco D, Siragusa M, Romano C. Cerebriform plantar hyperplasia: the major cutaneous feature of Proteus syndrome. *Int J Dermatol.* 2008 Apr;47(4):374-6. doi: 10.1111/j.1365-4632.2008.03440.x. PMID: 18377602.
179. Newman B, Urbach AH, Orenstein D, Dickman PS. Proteus syndrome: emphasis on the pulmonary manifestations. *Pediatr Radiol.* 1994;24(3):189-93. doi: 10.1007/BF02012188. PMID: 7936796.
180. Nakane M, Sato M, Hattori H, Matsumoto Y, Otsubuki M, Murakawa M. Perioperative respiratory complications caused by cystic lung malformation in Proteus syndrome. *J Anesth.* 2006;20(1):26-9. doi: 10.1007/s00540-005-0353-x. PMID: 16421672.
181. Gorlin RJ, Cohen MM Jr, Hennekam RCM. *Syndromes of the head and neck.* 4th ed. Oxford: Oxford University Press; 2001. pp. 480-8
182. Ruggieri M, Pascual-Castroviejo I. Proteus syndrome. In: Ruggieri M, Pascual-Castroviejo I, Di Rocco C, editors. *Neurocutaneous diseases: the phacomatoses and hamartoneoplastic syndromes.* Vienna: Springer; 2008. pp. 527-46
183. Asahina A, Fujita H, Omori T, Kai H, Yamamoto M, Mii K. Proteus syndrome complicated by multiple spinal meningiomas. *Clin Exp Dermatol.* 2008 Nov;33(6):729-32. doi: 10.1111/j.1365-2230.2008.02846.x. Epub 2008 Aug 2. PMID: 18681877.
184. Nazzaro V, Cambiaghi S, Montagnani A, Brusasco A, Cerri A, Caputo R. Proteus syndrome. Ultrastructural study of linear verrucous and depigmented nevi. *J Am Acad Dermatol.* 1991 Aug;25(2 Pt 2):377-83. PMID: 1894776.
185. Twede JV, Turner JT, Biesecker LG, Darling TN. Evolution of skin lesions in Proteus syndrome. *J Am Acad Dermatol.* 2005 May;52(5):834-8. doi: 10.1016/j.jaad.2004.12.047. PMID: 15858474.
186. Hoey SE, Eastwood D, Monsell F, Kangesu L, Harper JI, Sebire NJ. Histopathological features of Proteus syndrome. *Clin Exp Dermatol.* 2008 May;33(3):234-8. doi: 10.1111/j.1365-2230.2007.02601.x. Epub 2008 Jan 16. PMID: 18205855.
187. Sarma N, Malakar S, Lahiri K. Unilateral proteus syndrome. *Indian J Dermatol Venereol Leprol.* 2005 Mar-Apr;71(2):122-4. doi: 10.4103/0378-6323.14000. PMID: 16394389.
188. Lindhurst MJ, Wang JA, Bloomhardt HM, Witkowski AM, Singh LN, Bick DP, Gambello MJ, Powell CM, Lee CR, Darling TN, Biesecker LG. AKT1 gene mutation levels are correlated with the type of dermatologic lesions in patients with Proteus syndrome. *J Invest Dermatol.* 2014 Feb;134(2):543-546. doi: 10.1038/jid.2013.312. Epub 2013 Jul 24. PMID: 23884311; PMCID: PMC3868633.
189. Happle R. Linear Cowden nevus: a new distinct epidermal nevus. *Eur J Dermatol.* 2007 Mar-Apr;17(2):133-6. doi: 10.1684/ejd.2007.0125. Epub 2007 Mar 2. PMID: 17337396.
190. Liaw D, Marsh DJ, Li J, Dahia PL, Wang SI, Zheng Z, Bose S, Call KM, Tsou HC, Peacocke M, Eng C, Parsons R. Germline mutations of the PTEN gene in Cowden disease, an inherited breast and thyroid cancer syndrome. *Nat Genet.* 1997 May;16(1):64-7. doi: 10.1038/ng0597-64. PMID: 9140396.
191. Happle R. A rule concerning the segmental manifestation of autosomal dominant skin disorders. Review of clinical examples providing evidence for dichotomous types of severity. *Arch Dermatol.* 1997 Dec;133(12):1505-9. PMID: 9420534.

192. García-Vargas A, Hafner C, Pérez-Rodríguez AG, Rodríguez-Rojas LX, González-Esqueda P, Stoehr R, Hernández-Torres M, Happle R. An epidermal nevus syndrome with cerebral involvement caused by a mosaic FGFR3 mutation. *Am J Med Genet A*. 2008 Sep 1;146A(17):2275-9. doi: 10.1002/ajmg.a.32429. PMID: 18642369.
193. Hafner C, van Oers JM, Vogt T, Landthaler M, Stoehr R, Blaszyk H, Hofstaedter F, Zwarthoff EC, Hartmann A. Mosaicism of activating FGFR3 mutations in human skin causes epidermal nevi. *J Clin Invest*. 2006 Aug;116(8):2201-2207. doi: 10.1172/JCI28163. PMID: 16841094; PMCID: PMC1501112.
194. Collin B, Taylor IB, Wilkie AO, Moss C. Fibroblast growth factor receptor 3 (FGFR3) mutation in a verrucous epidermal naevus associated with mild facial dysmorphism. *Br J Dermatol*. 2007 Jun;156(6):1353-6. doi: 10.1111/j.1365-2133.2007.07869.x. Epub 2007 Apr 17. PMID: 17441958.
195. Happle R. Ptychotropism as a cutaneous feature of the CHILD syndrome. *J Am Acad Dermatol*. 1990 Oct;23(4 Pt 1):763-6. doi: 10.1016/0190-9622(90)70285-p. PMID: 2229513.
196. König A, Happle R, Bornholdt D, Engel H, Grzeschik KH. Mutations in the NSDHL gene, encoding a 3beta-hydroxysteroid dehydrogenase, cause CHILD syndrome. *Am J Med Genet*. 2000 Feb 14;90(4):339-46. PMID: 10710235.
197. Bornholdt D, König A, Happle R, Leveleki L, Bittar M, Danarti R, Vahlquist A, Tilgen W, Reinhold U, Poyares Baptista A, Grosshans E, Vabres P, Niiyama S, Sasaoka K, Tanaka T, Meiss AL, Treadwell PA, Lambert D, Camacho F, Grzeschik KH. Mutational spectrum of NSDHL in CHILD syndrome. *J Med Genet*. 2005 Feb;42(2):e17. doi: 10.1136/jmg.2004.024448. PMID: 15689440; PMCID: PMC1735983.
198. Happle R, Mittag H, Küster W. The CHILD nevus: a distinct skin disorder. *Dermatology*. 1995;191(3):210-6. doi: 10.1159/000246548. PMID: 8534939.
199. Fink-Puches R, Soyer HP, Pierer G, Kerl H, Happle R. Systematized inflammatory epidermal nevus with symmetrical involvement: an unusual case of CHILD syndrome? *J Am Acad Dermatol*. 1997 May;36(5 Pt 2):823-6. doi: 10.1016/s0190-9622(97)70031-8. PMID: 9146558.
200. König A, Happle R, Fink-Puches R, Soyer HP, Bornholdt D, Engel H, Grzeschik KH. A novel missense mutation of NSDHL in an unusual case of CHILD syndrome showing bilateral, almost symmetric involvement. *J Am Acad Dermatol*. 2002 Apr;46(4):594-6. doi: 10.1067/mjd.2002.113680. PMID: 11907515.
201. Happle R, Koch H, Lenz W. The CHILD syndrome. Congenital hemidysplasia with ichthyosiform erythroderma and limb defects. *Eur J Pediatr*. 1980 Jun;134(1):27-33. doi: 10.1007/BF00442399. PMID: 7408908.
202. Bittar M, Happle R. CHILD syndrome avant la lettre. *J Am Acad Dermatol*. 2004 Feb;50(2 Suppl):S34-7. doi: 10.1016/s0190-9622(03)01827-9. PMID: 14726863.
203. Irving MD, Chitty LS, Mansour S, Hall CM. Chondrodysplasia punctata: a clinical diagnostic and radiological review. *Clin Dysmorphol*. 2008 Oct;17(4):229-41. doi: 10.1097/MCD.0b013e3282f-dcc70. PMID: 18978650.
204. Golitz LE, Weston WL. Inflammatory linear verrucous epidermal nevus. Association with epidermal nevus syndrome. *Arch Dermatol*. 1979 Oct;115(10):1208-9. PMID: 507864.
205. Hebert AA, Esterly NB, Holbrook KA, Hall JC. The CHILD syndrome. Histologic and ultrastructural studies. *Arch Dermatol*. 1987 Apr;123(4):503-9. PMID: 3827283.
206. Pereiro Miguens M, Pena Guitian J, Vieites Faya F. Psoriasiform lesions of linear distribution accompanied by congenital malformations. *Actas Dermosifiliogr*. 1960 Apr;51:213-9. PMID: 13734255.
207. Shear CS, Nyhan WL, Frost P, Weinstein GD. Syndrome of unilateral ectromelia, psoriasis and central nervous system anomalies. *Birth Defects Orig Artic Ser*. 1971 Jun;7(8):197-203. PMID: 5173260.
208. Findlay GH. Studies in morphogenetic dermatology. Annotations concerning keratoacanthoma, dyskeratosis congenita, elastosis perforans, unilateral congenital psoriasiform erythroderma with ipsilateral limb defect, cavernous haemangioma, lipid proteinosis, reticulohistiocytoma and pigmented neurofibroma. *Trans St Johns Hosp Dermatol Soc*. 1974;60(2):152-9. PMID: 4377629.
209. Peter C, Meinecke P. CHILD-Syndrom. Fallbericht einer seltenen Genodermatose [CHILD syndrome. Case report of a rare genetic dermatosis]. *Hautarzt*. 1993 Sep;44(9):590-3. German. PMID: 8407327.
210. Cullen SL, Harris DE, Carter CH, Reed WB. Congenital unilateral ichthyosiform erythroderma. *Arch Dermatol*. 1969 Jun;99(6):724-9. PMID: 5783085.
211. Falek A, Heath CW Jr, Ebbin AJ, McLean WR. Unilateral limb and skin deformities with congenital heart disease in two siblings: a lethal syndrome. *J Pediatr*. 1968 Dec;73(6):910-3. doi: 10.1016/s0022-3476(68)80247-1. PMID: 5696317.
212. Lipsitz PJ, Suser F, Weinberg S, Valderrama E. Congenital unilateral ichthyosis in a newborn. *Am J Dis Child*. 1979 Jan;133(1):76-8. doi: 10.1001/archpedi.1979.02130010082016. PMID: 760518.
213. Tang TT, McCreadie SR. Q--congenital hemidysplasia with ichthyosis. *Birth Defects Orig Artic Ser*. 1974;10(5):257-61. PMID: 4620143.
214. Saito M, Ishiko A. A novel silent mutation in the NSDHL gene causing CHILD syndrome as a result of aberrant splicing. *Br J Dermatol*. 2008 Nov;159(5):1204-6. doi: 10.1111/j.1365-2133.2008.08800.x. Epub 2008 Sep 1. PMID: 18764845.

215. Baden HP, Rex IH Jr. Linear ichthyosis associated with skeletal abnormalities. *Arch Dermatol.* 1970 Jul;102(1):126-8. doi: 10.1001/archderm.102.1.126b. PMID: 4322292.
216. Adrian RM, Baden HP. Analysis of epidermal fibrous proteins in inflammatory linear verrucous epidermal nevus. *Arch Dermatol.* 1980 Oct;116(10):1179-80. PMID: 7425666.
217. Rossman RE, Shapiro EM, Freeman RG. Unilateral Ichthyosiform Erythroderma. *Arch Dermatol.* 1963 Nov;88:567-71. doi: 10.1001/archderm.1963.01590230075012. PMID: 14060072.
218. Zellweger H, Uehlinger E. A case of half-sided bone enchondromatosis (Ollier) with ichthyosiform nevus. *Helv Paediatr Acta.* 1948 May;3(2):153-63. PMID: 18865122.
219. Giam YC, Ong BH, Tan T. Atypical erythrokeratoderma. In: Urabe H, Kimura M, Yamamoto K, Ogawa H, editors. *Proceedings of the 4th International Congress of Pediatric Dermatology*, Tokyo, June 7-10, 1986. Tokyo: University of Tokyo Press
220. Lambert D, Dalac S, Alison M, Mabile JP. Epidermal nevus associated with ganglioneuroblastoma. In: Wilkinson DS, Mascaró JM, Orfanos CE, editors. *Clinical dermatology: the CMD case collection. World Congress of Dermatology*, Berlin, May 24-29, 1987. Stuttgart: Schattauer; 1987. pp. 48-9.
221. Jacyk WK, La Cock A. Squamous cell carcinoma arising in CHILD syndrome. *J Eur Acad Dermatol Venereol.* 2006 Mar;20(3):311-3. doi: 10.1111/j.1468-3083.2006.01411.x. PMID: 16503894.
222. König A, Skrzypek J, Löffler H, Oeffner F, Grzeschik KH, Happle R. Donor dominance cures CHILD nevus. *Dermatology.* 2010;220(4):340-5. doi: 10.1159/000287253. Epub 2010 Apr 9. PMID: 20389027.