

Online Resource 4. Reported CSHS cases of resolved/improved mineral abnormalities

Age at clinical improvement (years)	Indicator of decreased skeletal disease activity	Age at latest follow-up (years)	References
4	Improvement of rickets and hypophosphatemia at age 4	39	[1]
10	Insufficiency fracture cessation since age 10	19	[2]
12	Resolution of hypophosphatemia	13	[3]
17	Resolution of hypophosphatemia at age 17	35	[4]
17	Improved biochemical abnormalities. Increased BMD	17	[5]
18	Improved response to oral treatment after age 18	18	[6]
22	Insufficiency fracture cessation since age 22	32	[7]
22	Improved biochemical profile in adulthood	22	[8]
N/A	Insufficiency fracture cessation	26	[9]
27?	Resolution of hypophosphatemia. Vitamin D stopped at age 27. Phosphorus status normal at age 52	52	[10]

REFERENCES

1. Goldblum, J.R. and J.T. Headington, *Hypophosphatemic vitamin D-resistant rickets and multiple spindle and epithelioid nevi associated with linear nevus sebaceus syndrome*. J Am Acad Dermatol, 1993. **29**(1): p. 109-11.
2. Bouwes Bavinck, J.N. and J.J.P. van de Kamp, *Organoid naevus phakomatosis: Schimmelpenning-Feuerstein-Mims Syndrome*. Br J Dermatol, 1985. **113**: p. 491-492.
3. Aschinberg, L.C., et al., *Vitamin D-resistant rickets associated with epidermal nevus syndrome: demonstration of a phosphaturic substance in the dermal lesions*. J Pediatr, 1977. **91**(1): p. 56-60.
4. Rustin, M.H., et al., *Polyostotic fibrous dysplasia associated with extensive linear epidermal naevi*. Clin Exp Dermatol, 1989. **14**(5): p. 371-5.
5. Hoffman, W.H., et al., *Elevated fibroblast growth factor-23 in hypophosphatemic linear nevus sebaceous syndrome*. Am J Med Genet A, 2005. **134**(3): p. 233-6.
6. Kishida, E.S., et al., *Epidermal nevus syndrome associated with adnexal tumors, spitz nevus, and hypophosphatemic vitamin D-resistant rickets*. Pediatr Dermatol, 2005. **22**(1): p. 48-54.
7. Muhle, C., et al., *Skeletal involvement and follow-up in linear nevus sebaceous syndrome*. Eur Radiol, 1998. **8**(4): p. 606-8.
8. Saraswat, A., et al., *Phakomatosis pigmentokeratolica associated with hypophosphataemic vitamin D-resistant rickets: improvement in phosphate homeostasis after partial laser ablation*. Br J Dermatol, 2003. **148**(5): p. 1074-6.

9. Cabanillas, M., et al., *Epidermal nevus syndrome associated with polyostotic fibrous dysplasia, CNS lipoma, and aplasia cutis*. *Dermatol Online J*, 2009. **15**(10): p. 7.
10. Zutt, M., et al., *Schimmelpenning-Feuerstein-Mims syndrome with hypophosphatemic rickets*. *Dermatology*, 2003. **207**(1): p. 72-6.