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NEUROCUTANEOUS SYNDROMES

LISCH NODULES IN NEUROFIBROMATOSIS TYPE 1

The prevalence of Lisch nodules among 167 patients with neurofibromatosis 1 is reported from the Department of Pediatrics, Genetics Division, Miami Children's Hospital, Miama, FL. Lisch nodules are dome-shaped hamartomatous lesions with a clear, yellow or brown appearance. The overall prevalence of Lisch nodules was 73.7% and similar to that of neurofibromas (68.3%). The prevalence varied directly with the age of the patients: 5% in children less than 3 years of age, 42% in those 3 to 4 years old, 55% at 5 to 6 years; and 100% in adults of 21 years of age or older. Lisch nodules appeared more frequently than neurofibromas in all but the youngest age group. Of 145 patients having radiological studies, 17 had optic gliomas (12%), 9 had other types of CNS tumor, and one had both optic glioma and another CNS tumor. The proportion of Lisch nodules was similar in those with or without CNS tumors. If Lisch nodules do not appear with increasing age, the risk of having the gene for neurofibromatosis 1 is reduced from 50% at birth to 31% at 5 to 6 years of age, 15% at 9 to 14 years of age, 8% at 15 to 20 years of age, and 0% in those over the age of 20. (Lubs MLE, Bauer MS et al. Lisch nodules in neurofibromatosis type 1. N Engl J Med May 2, 1991; 324:1264-1266).

> COMMENT. Multiple Lisch nodules, unlike cafe-au-lait spots and neurofibromas, are specific for the diagnosis of neurofibromatosis 1. Except for one case they have been absent in central neurofibromatosis 2. Lisch nodules are useful in the early diagnosis of neurofibromatosis 1 and slit-lamp examination is important in suspected cases.

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