

Self-mutilation in the Lesch–Nyhan syndrome

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Over the last few years, this 12-year-old boy had chewed off his lips and tongue, bitten his left arm, and eroded his nose by rubbing it on the floor (figures 1 and 2). He was bed bound and dependent for activities of daily living. CT head was normal. His serum uric acid level was 11.5 mg, and ultrasound of the abdomen showed multiple renal calculi. Nerve conduction studies were normal.

The Lesch–Nyhan syndrome results from deficient hypoxanthine–guanine phosphoribosyltransferase activity. Clinical features include hyperuricemia, developmental delay, movement disorders, and self-mutilation targeted to the lips, tongue, and digits.¹ The differential diagnosis includes congenital insensitivity to pain.

Reference

1. Lesch M, Nyhan WL. A familial disorder of uric acid metabolism and central nervous system function. *Am J Med* 1964;36:561–570.



Figure 1. Mutilation of the lips, tongue and nose.



Figure 2. Healed scars on his left arm following self-biting.

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