

A 15-Year-Old Boy With Snoring and Molar Tooth Sign

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A 15-year-old boy presented for evaluation of snoring and sleep-disordered breathing. The parents noted that the patient snored every night and that he had episodes when he stopped breathing, ending with gasping for air. He had no history of sleep walking, night terrors, tongue biting, or seizures. The patient had two healthy siblings, but he had a history of intellectual disability and developmental delay. The patient had a history of adenotonsillectomy.

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Physical Examination Findings

The patient was afebrile with stable vital signs. BP was 130/70 mm Hg, heart rate was 95/min, height was 185 cm, and weight was 123 kg with a calculated BMI of 35 kg/m². His general physical examination revealed intellectual disability, developmental delay, dysarthria, hypotonia, and gait disturbance. Cardiac and pulmonary examinations were within normal limits apart from periods of tachypnea precipitated by the physical examination attempts. These tachypneic episodes were sustained for a few minutes with a respiratory rate of ≥ 60 /min. The episodes were not associated with sweating, wheezing, stridor, change in color, or desaturation. Head and neck examination revealed the presence

of a high-arched palate, maxillary retrusion, and Mallampati class 4.

Diagnostic Studies

The patient underwent overnight polysomnography that showed an overall apnea-hypopnea index of 31 events/h of sleep (central apnea index, 9; obstructive apnea index, 2; obstructive hypopnea index, 20). Frequent central apnea events were noted, alternating with episodes of tachypnea (Fig 1). The patient was not receiving any narcotics, and his echocardiogram was normal. To uncover potential causes of central apnea, the patient underwent MRI of the brain that showed the molar tooth sign (MTS) (Fig 2).

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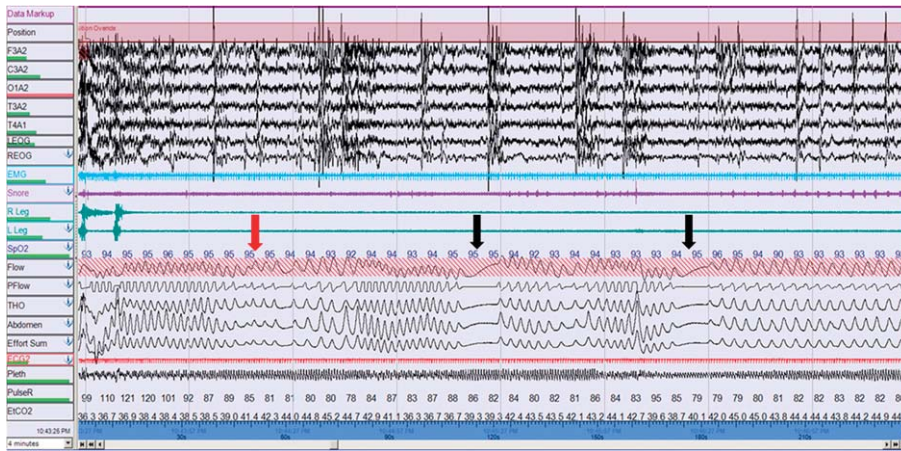


Figure 1 – A 4-min polysomnographic recording during stage N2 non-rapid eye movement shows hypopnea (red arrow) and central apneas (black arrows) alternating with periods of tachypnea.



Figure 2 – Brain MRI axial T2-weighted image shows the molar tooth sign, which results from elongated thickened superior cerebellar peduncles (arrows), deepened interpeduncular fossa (*), and cerebellar vermis hypoplasia.

What is the diagnosis?

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