Death during GH therapy in children with Prader-Willi syndrome: description of two new cases

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A few cases of death worldwide during GH treatment in pediatric patients with Prader-Willi syndrome (PWS) have been recently described. The evaluation of further cases is needed to better identify possible causal mechanism(s), as well as to suggest some additional guidelines for prevention. We report the death of 2 additional children with genetically confirmed PWS in the first months of GH therapy. Case 1: this 3.9-yr-old girl was born at 39 weeks gestation. Low GH response to two stimulation tests was observed. GH administration was started at the age of 3.5 yr (0.33 mg/kg per week), when the patient was at 130% of her ideal body weight (ibw). Hypertrophy of adenoids was previously demonstrated. Snoring and sleep apnea were present before GH treatment, and did not increase during therapy. Four months later she died at home suddenly in the morning. Case 2: this patient was a 6.3-yr-old boy. He was born at term after an uneventful pregnancy. At the age of 6 yr, his weight was at 144% of his ibw. He showed reduced GH secretion during provocation tests, and GH therapy was started (0.20 mg/kg per week). The previously reported nocturnal respiratory impairment had worsened after beginning GH administration. Tonsils and adenoids hypertrophy were noted. At the age of 6.3 yr he died at home in the morning following an acute crisis of apnea. These additional cases seem to confirm that some children with PWS may be at risk of sudden death at the beginning of GH therapy.