AUTHORS' REPLY

Dear Sir,

The authors of the letter has rightly drawn attention to the occurrence of ventilatory abnormalities in children with Prader-Willi syndrome (PWS).[1] The risk factors for these ventilatory abnormalities include obesity, restrictive lung dysfunction due to muscular weakness, micrognatia and a small nasopharynx, which contribute to upper airway obstruction and additional central autonomic impairment of ventilatory control. The authors have pointed concerns regarding the use of growth hormone (GH) in children with PWS and ventilatory abnormalities including publication of case reports documenting sudden death in children with the initiation of GH therapy. The possible mechanisms by which GH therapy can worsen ventilatory abnormalities include:

(1) Stimulation of adenotonsillar hypertrophy[2]

(2) Increase in the basal metabolic rate which increases the oxygen demand[3]

(3) Early water retention with GH therapy which augments preload.[4]

Despite these concerns, benefits of GH therapy in PWS have been well demonstrated. These include improved lean-to-fat mass ratio, improvement in linear growth, and most importantly, improved central ventilatory drive.[5] A recent review has suggested a protocol for initiation of GH therapy in obese children with PWS to minimise the risk of sudden death in children with ventilatory abnormalities, and at the same time to enable the availability of the benefits of GH therapy to a larger group of patients. The steps include a screening oximetry, in addition to a routine sleep history and upper airway examination. If significant desaturations are observed on the screening oximetry, a polysomnography should be performed. Patients with abnormal polysomnography results should only be considered for GH therapy after successful intervention for the ventilatory defect.[6]

Yours sincerely,

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