

RESPIRATORY CHANGES IN PRADER-WILLI SYNDROME

Dear Sir,

We read with great interest the article by Jacob et al, in which they appropriately emphasised the risk of foreign body aspiration and choking in patients with Prader-Willi syndrome (PWS) and stressed their effects on morbidity and mortality rates. The authors have drawn special attention to hyperphagia associated with hypotonia and muscle in-coordination as predisposing factors for aspiration and choking in these patients. Obesity was enrolled as an additional factor in the development of obstructive sleep apnoea (OSA), which would improve under growth hormone (GH) therapy and weight control.⁽¹⁾ Although GH may be useful to treat OSA in PWS, there is some concern among physicians and relatives of patients informed about the possible worsening of the respiratory abnormalities and associated fatalities.^(1,2) In fact, recent studies have cleared the pathogenesis of genetically-determined disturbances of the human respiratory control.^(3,4)

Nevertheless, we wish to highlight the role of morbid obesity on the respiratory disorders often observed in patients with PWS. Incidentally, we have just concluded a manuscript about PWS in an adolescent female which was admitted with morbid obesity (BMI > 70 kg/m²) and severe breathing changes, and needed intensive care support. Respiratory parameters improved after bi-level positive airway pressure support and hypocaloric diet to maintain the weight loss. Worthy of note, GH was not included in her treatment schedule due to aforementioned reasons.

The infancy overweight and obesity rates have grown steadily in the recent decades, and must merit greater awareness among physicians and other healthcare workers. As a whole, data from the literature and lessons from practice indicate that early diagnosis constitutes the cornerstone of the successful treatment of overweight and obese patients with PWS. Moreover, the morbid obesity prevention and weight control should constitute two of the major objectives either for the clinical management of PWS in general, or specifically for the concurrent respiratory disorders. Actually, despite the modern clinical and surgical support,^(2,5) these goals have hardly been obtained through the longstanding follow-up of children and adolescents treated for PWS.

Yours sincerely,

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