

# NEUROENDOCRINOLOGY

**P337****ROHHAD: RAPID ONSET OBESITY AND THE IDIOPATHIC HYPOTHALAMIC SYNDROME IN CHILDREN**

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**Abstract:** The terms ROHHAD and the "idiopathic hypothalamic syndrome" describe a combination of rapid-onset obesity, hypothalamic dysfunction, alveolar hypoventilation and autonomic dysfunction in children. The syndrome arises from a complex interaction between

the nervous system, the autonomic system as well as the endocrine and respiratory systems culminating in significant morbidity and mortality. A high number of patients have developed neural crest tumours over time, which indicates it is a para-neoplastic syndrome. Reports in the literature are limited largely to case descriptions and retrospective studies only. Therapy is often directed towards reversal of the presenting feature rather than to the underlying neoplasm which may not even be suspected. A lack of large-scale, well-designed studies evaluating immunologic treatments has limited the development of evidence-based treatment. This review describes three cases from Malaysia and Australia and over fifty cases from the literature and outlines the major mechanisms believed to underlie the development of this syndrome, the clinical presentations, current therapy options and outcomes as well as future research directions.