

Hyperphagic short stature: A case report and review of literature

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ABSTRACT

A 5½-year-old adopted girl was referred to us in view of short stature. After ruling out systemic illness, she was evaluated for growth hormone deficiency (GHD) by stimulation tests. The peak value was 3.47 ng/ml. She was then started on growth hormone (GH). At the end of 6 months of GH therapy, her height velocity was only 3 cm/year. There was a lack of attachment between the mother and the child. She had history of hyperphagia, stealing, and hoarding food. Psychiatry consultation confirmed that the child had appetite disorder, and hence was diagnosed as hyperphagic short stature (HSS). The girl and her parents are undergoing psychiatric therapy for the same. Psychosocial dwarfism seems to originate from serious disturbances in the mother–child relationship. These children mimic patients with GHD, but have poor response to GH therapy. This case underscores the importance of social environment in the growth of the individual.

Key words: Growth hormone deficiency, hyperphagic short stature, psychosocial short stature

INTRODUCTION

Psychosocial short stature (PSS) is characterized by growth failure in association with emotional deprivation. Hyperphagic short stature (HSS) is a variant of PSS which has short stature in association with appetite and other behavioral problems. These children demonstrate biochemical evidence of growth hormone (GH) insufficiency, but they do not respond to GH therapy. A change in social environment brings about catch-up growth and resolution of behavioral abnormalities.

CASE REPORT

A 5½-year-old girl, ER, was referred to us for short stature. She was accompanied by her father who had adopted her at

the age of 3½ years. ER was a shy, but well-behaved child. The father said that she preferred to be alone and avoided playing with other kids.

ER was thin and besides hypertrichosis did not have any significant findings on examination. Her height and weight were 91.6 cm and 10.5 kg, respectively, which were <<3rd centile. She did not have any features suggestive of malnutrition.

Her baseline routine investigations including hemogram, liver and renal function tests were normal. Her peak GH levels on clonidine and insulin-induced hypoglycemia tests were low (3.47 ng/ml and 3.24 ng/ml, respectively). The thyroid, cortisol axes were normal. Magnetic resonance imaging of the pituitary did not reveal any abnormality. Thus, she was diagnosed to have isolated growth hormone deficiency (GHD) and started on GH therapy (10 U/week).

She was followed three monthly. At the end of 6 months of GH therapy, the response in terms of height gain was very dismal (1.5 cm gain in 6 months) [Figure 1]. This made us rethink about where we had gone wrong in her management. She had history of increased food intake for the past 3 years. She had craving for food, stole food from

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lunchboxes of other children in her class, used to hoard food, and get up at night to eat. Father said that she could eat a dozen bananas at one go. In spite of this voracious appetite, she was not gaining weight and height. The other significant history was the strained relation between ER and her foster mother. The mother did not want to adopt her in the first place, and hence the child felt neglected and unwanted.

She was labeled as a probable case of HSS. Repeat GH stimulation test with clonidine done after 14 days of hospital stay (away from the psychosocial stress) showed normal peak GH of 12.5 ng/ml. They were referred to psychiatrist who confirmed this diagnosis. A change of environment was advised, but was not feasible in the current family setting. Hence, the father decided to continue only with the psychiatric therapy (which included counseling of both ER and her parents).

DISCUSSION

We describe a rare case of HSS. She was earlier misdiagnosed as GHD and treated with GH but without adequate response. Confirmation of the diagnosis of HSS would have required the demonstration of catch-up growth after removal of the stressful environment. Though this was not possible in her, we think that strong history of appetite disorder and the very poor response to GH therapy proves our diagnosis of HSS in this case.

PSS is a disorder of short stature and/or delayed puberty that is observed in association with emotional deprivation, a pathologic psychosocial environment, or both. It is also known as “psychosocial dwarfism,” “The Kaspar Hauser Syndrome,” reversible hyposomatotropism, the garbage can syndrome, and maternal deprivation syndrome. It is not

classified within psychiatric or medical diagnostic systems (DSM-IV or ICD 10) because of its rarity.

The name Kaspar Hauser syndrome comes from description of a person named Kaspar Hauser who was abandoned at the city gate of Nuremberg in 1828, after 17 years of neglect and isolation in a dungeon carrying a written note that suggested he was 16 years of age.^[1] He was stunted in growth and had incoherent speech.

Though first described by Talbot *et al.* in 1947, the link was scientifically proven by Powell *et al.*^[2-4] They described 13 children with short stature, emotional disturbances, and abnormal home environments. When placed in a convalescent hospital, these children demonstrated remarkable growth acceleration without receiving GH.^[4,5] Several characteristic behavioral features, in particular, appetite disturbance, were described as part of psychosocial dwarfism.^[3,6] Skuse *et al.* further clarified the association between appetite disturbances – hyperphagia, stealing food, eating from rubbish bins, and polydipsia – by defining a distinctive subgroup of PSS called HSS.^[7] In this condition, GH insufficiency was associated with characteristic behavioral features, especially hyperphagia and polydipsia, and a normal body mass index (BMI). Such children have the capacity of spontaneous recovery and catch-up growth after removal from the stressful environment. From their data, the authors proposed clinical algorithm for the diagnosis of HSS based solely on age, anthropometric data, and behavioral criteria [Table 1].

A classification of PSS into three groups was postulated by Blizzard and Bulatovic.^[8] If classified within this scheme, HSS will fall into type II, or possibly type IIA. Type IIA refers to hyperphagic patients in whom reversible GH insufficiency is characteristic; there is rapid catch-up growth with a change of environment and minimal response to GH treatment.

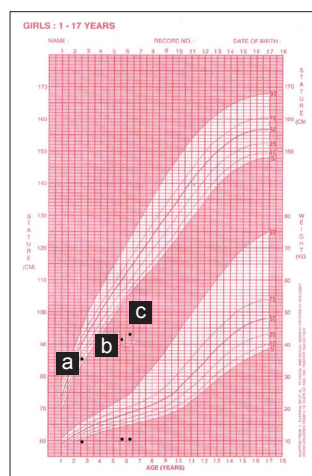


Figure 1: Growth chart. (a) Growth record at orphanage. (b) Baseline growth record at the start of growth hormone. (c) After 6 months of GH therapy

Table 1: Diagnostic criteria for hyperphagic short stature^[7]

- These features must be present
 - Height for age below third population centile with growth failure
 - Body mass index in normal range
 - Age greater than 2 years
- At least one of these symptoms must be present and should reflect a current pattern of behavior
 - Steals food at home and / or school
 - Gorges and vomits
- Two of these symptoms must be present and should reflect a persistent pattern of behavior
 - Eats excessively
 - Polydipsia
 - Hoards food
 - Wakes at night and searched for food
 - Forages for discarded food or rummages for food in bins

The physician should routinely ask whether a child with growth failure and GH deficiency eats excessively, gorges and vomits if given unlimited access to food, steals food at home and school, hoards food, has polydipsia or pica, scavenges from trash cans, or searches for food at night.

CONCLUSION

Psychosocial dwarfism seems to originate from serious disturbances in the mother–child relationship. These children mimic patients with GHD, but have poor response to GH therapy. They respond well to change in the social environment. This case underscores the importance of social environment in the growth of an individual.

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