Hyperphagic psychosocial short stature — a clinical review

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Abstract

Hyperphagic short stature (HSS) is a specific syndrome of growth failure in association with hyperphagia without a clear underlying aetiology, seen in association with a stressful environment. Environmental stressors at home are not always immediately evident however these children characteristically demonstrate reversal of growth impairment when nurtured away from their stressful environment. This review describes the clinical phenotype, underlying endocrine dysfunction and strategies for the management of this complex condition.

Keywords emotional deprivation; emotional dwarfism; hyperphagia; hyperphagic short stature; polyphagia; psychosocial dwarfism; psychosocial short stature

Introduction

Hyperphagic short stature (HSS) is a condition of growth failure in association with hyperphagia (excessive eating). Children with this condition form a distinguishable subgroup of children with psychosocial short stature (PSS). HSS has been associated with emotional and other forms of abuse. Children with HSS characteristically demonstrate significant catch up growth and reversal of biochemical markers of an underactive pituitary gland when placed in an alternative environment, making this an eminently identifiable and reversible condition. The primary challenge in the successful management of this condition is the recognition and comprehension of its significance by health and social care professionals. The prevalence of this condition is unknown, though previously estimated to account for short stature in 3% of short normal children, from data from the Wessex Growth Study and tertiary centre cases.

Growth failure

Growth is a key parameter for assessing health and well being in childhood. Short stature is defined as height less than two standard deviations (SD) below the mean for the population. Short stature and growth failure, the failure to maintain a height velocity in the normal range when accounting for pubertal development, has a wide range of organic and in-organic aetiologies.

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PSS and its classification

A proportion of children for whom no organic cause for growth failure is found will have PSS. This condition, first described by Talbot over sixty years ago, has been referred to historically by a number of synonyms including abuse dwarfism and psychosocial dwarfism.

Psychosocial deprivation impacts on growth from infancy to adolescence. Growth failure in infancy in the absence of physical disease is termed non-organic failure to thrive, also classified as PSS type I. Failure to thrive during infancy was linked to emotional deprivation in infants in institutional care by Chapin as early as 1915, and in 1957 was recognised in infants cared for in their own homes. PSS type I is usually due to inadequate calories and neglect, secondary to psychosocial circumstances. It is uncommon for this neglect to be intentionally abusive, typically reflecting inadequate parenting with poor understanding of infant feeding needs and reversing with education and adequate provision of calories.

Beyond infancy failure to grow as a result of psychosocial circumstances is referred to as PSS type II. The impact of environment and psychosocial stress on growth beyond infancy is evidenced from the earliest reports of growth failure in an orphanage in association with a sadistic teacher, despite the amount of food eaten by the children increasing, to growth studies undertaken on institutionalised children during the last decade. Following the Second World War, Widdowson, a British nutritionist, demonstrated dependency of children's growth on emotional environment in non-institutionalized settings. PSS type II can be an extension of non-organic failure to thrive in infancy, however calorie intake is not reduced. When anorexia does coexist this is termed PSS type III.

A smaller specific sub-set of children with PSS type II have the distinctive behavioural phenotype with excessive calorie intake and growth failure known as HSS. This is classified as PSS type IIA in contrast to type IIB where calorie intake is normal. Intake in HSS is not motivated by hunger but is a maladaptive hypothalamic response to negative emotions such as anxiety, depression, sadness, loneliness, boredom, fear or anger. Excessive eating in the absence of disease normally leads to obesity but a unique feature of this syndrome is that the Body Mass Index (BMI) is normal. What is striking in PSS Type IIA (HSS) in contrast to Type IIB and Type III is that the growth failure is consistently reversible when the child is removed to a nurturing environment.

Psychosocial factors determining growth failure

Risk factors for impaired growth associated with low socioeconomic status include single parenthood, illegitimacy, overcrowding, low disposable income, paternal ill health, unemployment, alcohol and drug abuse and quality of maternal care. Disturbed marital relationships increase risk, and family conflict was associated with short stature and final height in the British 1958 birth cohort.

Paternal occupation influences growth and a greater spread of height differences are observed between children from lower socioeconomic backgrounds in the lower height centiles. At the 50th centile the average height difference between children of manual and non-manual workers is 2 cm. At the third centile

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children of manual workers are 4 cm shorter on average than non-manual workers.

The behavioural phenotype of HSS

Children with HSS eat excessively and may gorge food without satiety, to the extent of vomiting. They typically hoard or steal food and wake up at night to search for food. In the extreme they may escape their home seeking food, and scavenge for food in bins. Taking food from cupboards or siblings at home, and stealing food in school is common. Pica, consumption of items that are not food, is also described. Polydipsia can be observed, including drinking water from unusual sites, such as the toilet bowl. Parents of affected children frequently take unusual measures to prevent uncontrolled eating, including locking food cupboards, fridges, freezers, and putting alarms on kitchen doors.

Behavioural problems extend beyond the relationship with food and include behaviour problems (42%), encopresis (24%) and nocturnal enuresis (18%). Disordered sleep, with or without nocturnal food scavenging, is consistently reported. Hyperactivity, pain agnosia and social isolation, including limited interactions with peers and siblings, are also described.

Clinical features of HSS

Clinical features of HSS rest with the aforementioned behavioural phenotype, together with anthropometry and biochemical findings. Height below the third centile, in children beyond 2 years of age and normal Body Mass Index (BMI) are cardinal features of HSS. Older children may have pubertal delay. Birth-weight tends to be appropriate for gestational age, however faltering growth has been reported in earlier years. The mechanism for normal, rather than increased BMI, despite excessive appetite is unclear although some children with HSS display over-activity that may offset weight gain.

There are no pathognomonic physical signs, however abdominal protuberance, lanugo hair, and disproportionately thin arms and legs are described. It is important to examine for signs of physical abuse and neglect, including general physical appearance, hygiene, dentition, previous or recent injuries including cold injury.

Bone age delay (1.9—3 years) and metaphyseal arrest lines at ends of long bones, commonly seen in chronic illnesses and malnutrition, have been demonstrated in children with HSS. Metaphyseal arrest lines are indicative of alternate periods of growth cessation and recovery. It is postulated that varying intensities of stress, over a period of time, could contribute to its occurrence in HSS.

Biochemical features in HSS

Baseline investigations are normal in HSS with the exception of thyroid function and Growth hormone (GH) testing. Diabetes Insipidus is not part of HSS and must be excluded when polydipsia is a component of the behavioural phenotype.

Abnormal thyroid function, with a picture of central hypothyroidism with low or low normal free T4 but normal thyroid stimulating hormone (TSH) is reported. Mildly raised TSH with borderline free T4 has also been observed. In HSS thyroid function tests normalise without levothyroxine therapy with removal from the detrimental environment.

Children with HSS have evidence of biochemical GHD with low levels of Insulin-like growth factor-1 (IGF-1), a surrogate marker of GH secretion. Studies of GH secretion in children with features of HSS show normal or low GH responses to GH provocation testing however HSS is resistant to treatment with exogenous GH. This raises important practical implications, as these children may receive inappropriate treatment in the form of GH if HSS is not considered.

It is striking that normalisation of previously low GH responses on provocation testing and increase in IGF-1 levels are observed upon a change in environment. This is borne out in studies of spontaneous GH secretion using serial overnight profiling on 11 children with PSS, during a three-week admission in hospital with restricted parental access where improvement and normalisation of GH pulse amplitude, and thus reversibility of GHD was apparent. Four of these children subsequently entered foster care where mean height velocity improved. Reversal of the GH axis has been demonstrated as early as within 16 days of change in environment. Normal GH secretion at admission with an increase in peak concentration following removal from stressful environment has also been demonstrated.

Thus the biochemical picture in HSS is GH insufficiency and low IGF-1 concentrations. They are considered to be two separate pathological processes contributing to the short stature; GH insufficiency, secondary to hypothalamic dysfunction, which parallels the behavioural phenotype and the additive effect of reduced IGF-1 production. While treatment with GH may improve GH concentrations it does not normalise or improve IGF-1 concentrations. Removal from the stressful environment normalises both GH and IGF-1 resulting in significant catch up growth, where growth rates more than twice the normal for age, or exceeding velocities typical of the pubertal growth spurt, are observed.

Genetic factors in HSS

A genetic predisposition to HSS has been postulated, based a degree of familial aggregation and the occurrence of similar symptoms in full siblings of 40% of children with HSS. As children with Prader—Willi Syndrome (PWS) share a number of phenotypic clinical features with HSS, methylation testing and sibling linkage analysis on children with HSS has been explored, but no abnormality in the PWS critical region was identified. It is suggested that future genetic investigations should probe candidate genes that regulate appetite and growth.

Environmental stressors in children with HSS

The strong association of a stressful psychosocial environment and HSS and clustering of symptoms within family-sibling pairs indicate that in children with a genetic predisposition, stress can provoke the manifestation of HSS.

Concerns regarding abuse in children with HSS are valid. In a large follow up study of 65 children referred over a seven-year period to Great Ormond Street Hospital (GOSH) with PSS up to 41% were found to be physically or sexually abused. This group more commonly demonstrated features of HSS including hyperphagia, bizarre eating habits, behavioural problems, soiling or nocturnal enuresis. Of this group 78% had experienced emotional abuse. Of eleven children admitted to GOSH, with restricted

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