

**52** ENDOCRINOLOGIC EVALUATION OF TWO CHILDREN WITH PSYCOSOCIAL DWARFISM

Kubo T, Furujo M, Moriwake T\*

Department of Pediatrics, National Okayama Medical Center, \*Department of Pediatrics, Iwakuni National Hospital

We report the endocrinologic evaluation of two children with psychosocial dwarfism (PSD). [Case1] A 6 year old girl was referred to our hospital because of short stature (height SDS  $-2.67$ ). Three years ago, she, her mother and her elder sister ran away from home because of her father's domestic violence against her mother. Although her growth rate has been normal, the catch-up growth has not occurred after that. Her bone age was 5y9m and the IGF-I and U-GH levels were 114.1 ng/ml and 23.4 pg/mgCr, respectively. She had inadequate anterior pituitary hormone responses to pharmacological testing (insulin, L-DOPA, clonidine, TRH, and GnRH). Consequently we concluded that she had been still in a harmful environment, though the endocrinologic environment was normal. [Case2] A 13 year old girl was referred for evaluation of short stature (SDS  $-4.30$ ). Four months ago, she was removed from the home because of physical abuse and neglect by her parents. After that the catch-up growth was observed. Her bone age was 8y10m and breast buds were not present. The IGF-I, E2 and U-GH levels were 200 ng/ml, less than 10 pg/ml and 12.2 pg/mgCr, respectively. The provocative stimulation with pharmacological agents (insulin, L-DOPA, arginine, TRH, and GnRH) demonstrated partial GH deficiency and prepubertal gonadotropin responses. The GH insufficiency and delayed puberty might be due to the prolonged problem. Otherwise, she might have constitutional delay in growth and puberty, because she has been in non-stressful environment for a long while. [Conclusion] We should consider other factors such as insufficient removal of stress or basic diseases, when we are not able to observe the catch-up growth or the reversible hormonal changes in children with PSD after removal from the poor home environment.

**54** Excessive Mid-Growth Spurt (MGS) induced by GH therapy started at early childhood in 3 male siblings with non-endocrine short stature (NESS)

Maesaka H\*, \*\*, Yamazaki S\*\*, Tokuhiro E\*\*\*, Asakura Y, Adati M, Tachibana K.

\*Odawara Health &amp; Welfare Center, \*\*Asigara-Kami Hospital, \*\*\*Odawara City Hospital, Kanagawa Children's Medical Center

The MGS is a small transient increase in growth rate occurring around the age of 7 years in healthy children. GH treatment in NESS leads to an increased growth rate in the first years of treatment, but with lesser growth rate thereafter. We observed high MGS in 3 male siblings with NESS during GH treatment started at early childhood. We studied the growth rate of these 3 siblings, 14 male patients with NESS treated with GH at the age of 5- to <9 years and 15 male patients with NESS before GH treatment. The stimulated GH peaks in 3 male siblings were in the 8.1- to 22.3 ng/ml range. Serum levels of IGF-1 and IGF-BP3 of these siblings were within normal ranges. Heights of the parent were within  $\pm 1$ SD and onset of the puberty was slightly delayed. Their sister was also diagnosed as NESS with no mutation in GH1 gene. We treated 1<sup>st</sup>, 2<sup>nd</sup> and 3<sup>rd</sup> siblings with a chronological age of 6, 4, 2 years, a bone age of 4, 3, 1 years respectively and a height SD from  $-2.3$  to  $-2$ , with GH at a dose of 0.5 IU/kg/w for more than 6 years. MGS occurred in 3 males at the age of 7.2, 7.1, 6.3 years with a high growth velocity like puberty and good growth responses thereafter in 2<sup>nd</sup> and 3<sup>rd</sup> siblings. No high MGS occurred in 14 controls during GH therapy. Bone age advanced significantly following the onset of high MGS in 2 siblings. The gonadal function of the 2 siblings was prepubertal. The latest predicted final heights by the revised Bayley-Pinneau's method in 3 siblings increased over the average adult height compared with those of before GH treatment. MGS was observed at the mean age ( $\pm$ SD) of  $6.9 \pm 1.9$  years and with the mean HVSDS ( $\pm$ SD) of  $+1.3 \pm 0.7$  in 5 of 15 controls before GH treatment. Further studies will be needed to clarify the mechanism of high MGS.

**53** PITUITARY MAGNETIC RESONANCE IMAGING IN CHILDREN WITH GROWTH HORMONE DEFICIENCY

Shimizu T, Motomura K, Kawaguchi T, Kinoshita E, Hirota T\*, Baba T\*\*, Yoshimoto M\*\*\*

Department of Pediatrics, Nagasaki University School of Medicine, Nagasaki, JAPAN. \*Children's Clinic Hirota, Nagasaki, JAPAN, \*\* Baba Children's Clinic, Saga, JAPAN. \*\*\*Children's Clinic Yoshimoto, Nagasaki, JAPAN.

In order to investigate an association between pituitary size and severity of growth hormone deficiency (GHD), we evaluated the magnetic resonance images (MRI) of 82 children (43 male, 29 female; follow up  $4.6 \pm 2.5$  years) with GHD.

The hormonal examination including GH stimulation tests led to the following subgroups: complete isolated GHD (IGHD) (peak values of GH  $< 5$  ng/ml) (n = 5); partial IGHD (peak values of GH 5-10 ng/ml) (n = 64); multiple pituitary hormone deficiency (MPHD) (n = 13). Pituitary height (PHT) was measured and small anterior pituitary gland was assumed when PHT was  $< -2$ SD. Small anterior pituitary gland was found in 2 of 5 complete IGHD cases (40%), 40 of 85 partial IGHD cases (47%) and 12 of 13 MPHD cases (92%). Pituitary size did not correlate with the severity of GHD and height velocity after GH replacement therapy. In 15 cases reexamined, 8 cases with MPHD showed no increase of PHT, while 7 with partial IGHD revealed a moderately increment of PHT.

Further examination is necessary to definite the significance of small pituitary gland in children with GHD.

**55** GROWTH RESPONSE TO GROWTH HORMONESatoh M<sup>1,2</sup>, Naiki Y<sup>1</sup>, Itoh M<sup>1</sup>, Yoshimura K<sup>1</sup>, Ikema S<sup>1</sup>, Horikawa R<sup>1</sup>, Katsumata N<sup>1</sup>, Ogata T<sup>1</sup>, Tanaka T<sup>1</sup><sup>1</sup>National Center for Child Health and Development  
<sup>2</sup>Department of Pediatrics, Toho University

Factors influencing the first year growth response were analyzed in twenty-nine growth hormone (GH)-treated short children (23 boys, 6 girls) at National Center for Child Health and Development (National Children's Hospital). Patients older than 10 years in boys and 9 years at the start of treatment were excluded to avoid the effect of pubertal spurt. Patients with Turner syndrome and achondroplasia were also excluded.

The first year growth velocity was inversely correlated with chronological age, bone age and IGF-I at the start of GH treatment, but showed no correlation with growth velocity before treatment, maximum peak GH value at provocation tests, peak GH response to GRF, mean nocturnal GH concentration, heights of parents, height SDS and weight SDS at birth, height SDS at the start of treatment, and GH dose. However, first year growth velocity SDS was only correlated with maximum peak GH value at provocation tests.  $\Delta$  growth velocity SDS was inversely correlated with growth velocity and growth velocity SDS before treatment, chronological age and bone age at the start of GH treatment. IGF-I or IGF-I SDS at the first year did not show any correlation with growth velocity, growth velocity SDS and  $\Delta$  growth velocity SDS.

This study confirmed that the younger age, younger bone age and low GH secretion capacity are the influential variables for the first year growth responses.