

Laron Type Dwarfism (Hereditary Somatomedin Deficiency): A Review

Laron type dwarfism is a syndrome of familial dwarfism that is indistinguishable from isolated growth hormone (GH) deficiency except that patients have normal or elevated GH concentrations. The syndrome was described by Laron et al in 1966.

In the current review, Laron tabulates 72 cases. Many are non-Jewish. The birth weight, known in 21 cases, was >2,500 g in 18, and the birth length was more than 2 SD below the mean in ten of 16. Pregnancies and deliveries were unremarkable. Approximately 50% had skeletal or mesenchymal anomalies, none of which was life threatening.

Development in children with Laron type dwarfism is generally slow: many sit only after the age of 1 year and walk after 18 months. Fontanel closure occurs between 3 and 7 years. Symptoms of hypoglycemia and high-pitched voice are also characteristic. With the passage of time, the acromicria and disproportion between the face, with its saddle-nose, and the cranium become more apparent. The teeth are discolored, defective, and crowded. Growth is slow, with males reaching ultimate heights of 119 to 142 cm and females, 108 to 136 cm. Surprisingly, the upper/lower ratios are more than 2 SD above the mean, indicating that the limbs are short in comparison to the trunk. After puberty, the skin assumes a prematurely aged appearance. The genitalia in affected children and adults are very small, and pubertal development is slow. Menarche occurs between 13 and 18 years of age and ejaculation between 17 and 21 (compared to a normal mean of 13½ years).

Skeletal age is delayed. By x-ray analysis, the long bones are small and delicate, the sella is of normal size, and the facial bones are small in comparison to the cranium. The head consequently appears enlarged, but it is not (on the basis of standard measurement). Glucose intolerance is present even when hypoglycemia and hypoinsulinemia

occur. Growth hormone levels often are elevated, but are suppressed normally with glucose. Serum somatomedin-C (Sm-C) concentrations are low, and do not increase after GH injections, although 50% of patients have an increase in free fatty acids. Nitrogen retention and hypercalcuria are minimal following GH administration.

No neurologic deficits were observed in these patients, and pneumoencephalograms were normal. IQ scores were strongly skewed toward the lower part of the curve (mean IQ = 82.1). Visual-motor coordination was uniformly poor. The parents regarded their own and their children's lives as ruined, since no remedial treatment exists for Laron type dwarfism. School was a

negative experience for these children.

The etiology is believed to be related to the hGH receptors, since liver cell microsomes from these patients do not bind hGH normally, although insulin binds normally. Consequently, Sm-C is not generated.

Laron Z: *Advances in Internal Medicine and Pediatrics*. Heidelberg, Springer-Verlag, 1984, p 118.

Editor's comment—Laron et al have clarified the etiology and provided additional information about the syndrome. Treatment with Sm-C (IGF-I) might be effective. Unfortunately, adequate quantities are not currently available to test this hypothesis.

Height and Weight Status of Indo-Chinese Refugee Children

Pediatricians and other health practitioners who deal with children have no guidelines with which to evaluate the growth or growth potential of refugee children. The absence of such guidelines becomes problematic when one tries to determine if growth retardation exists in a particular child (which, in itself, would indicate that a search should be made to determine a cause). This report attempts to supply the needed information, and succeeds partially.

Height and weight measurements were obtained from 1,650 children residing in Laotian and Cambodian refugee camps and in areas surrounding these camps. Reference tables that are available from China, Thailand, and the United States were also used. The mean weights and mean heights for age of the groups studied are approximately 2 SD below US means, but there is variation.

These studies are inadequate because they are not randomized, as readily stated by the authors. To what extent catch-up growth may occur in refugee children remains to be determined. Evidence that nutrition plays a role was presented in one study in which the heights and weights of children from upper-class and professional backgrounds were compared to American standards. The mean heights and weights more nearly approached the US standards than the heights and weights found in refugee children.

Olness K, Yip R, Indritz A, et al: *AJDC* 1984;138:544.

Editor's comment—While these studies are limited, they are of value. We practitioners can assume with some justification that the normal growth curves for refugee Oriental children are approximately 2 SD below US curves.

	Height (SD)	Weight (SD)
US reference	0	0
Chinese urban	-0.8	-0.8
Chinese rural	-1.4	-1.3
Thai reference	-1.3	-1.8
Khmer refugee	-1.8	-1.8
Lao refugee	-2.1	-2.2
Thai village	-2.3	-1.9