

# Letter to the Editor

## Russell-Silver Syndrome

In Vol. 2, No. 2 of *Growth, Genetics, and Hormones*, an article by Saal et al entitled "Reevaluation of Russell-Silver Syndrome" was abstracted. The Editor's Comment on that abstract prompts this letter.

One of the reasons that the Russell-Silver syndrome is heterogeneous is that *there is no such thing as the Russell-Silver syndrome*. Dr. Russell and Dr. Silver, in their original reports, described two entirely different syndromes. It is a mistake to combine the two and perpetuate the combination. I have mentioned this to Dr. Alex Russell, who agrees. Dr. Silver even described increased urinary gonadotropins in his patients. I point this out to our house staff when they refer such a patient to our clinic. In my experience, most of the patients referred to me for dwarfism, triangular facies, and intrauterine growth retardation fall into the "Russell" category. I have yet to see a patient with increased gonadotropins at a young age in the hemihypertrophy syndrome described by Silver. I continue to be a splitter instead of a lump.

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### Dr. Blizzard's Comments

Dr. Green's letter is in accord with the article written by Saal et al and published in the *Journal of Pediatrics* 1985;107:733. These authors stated that the Russell-Silver syndrome is a heterogeneous entity. Dr. Green would say it is not an entity at all. Undoubtedly, many would agree with Dr. Green. I have asked Dr. Silver to respond and his comments are listed below.

### Dr. Silver's Comments

The confusion about the Silver-Russell syndrome will un-

doubtedly continue until the specific etiology(s) of the syndrome has (have) been defined and/or a specific diagnostic laboratory test is available. Although the heterogeneity of findings suggests that multiple etiologies may be involved, there is no concrete evidence that this is so.

The Silver-Russell syndrome certainly fits the definition of a syndrome: "the sum of signs of any morbid state; a set of symptoms occurring together" (Dorland). As with most other syndromes, not every child with the Silver-Russell syndrome has every finding. However, the combination of all or most of the findings of congenital short stature continuing into childhood, asymmetry involving various parts of the body, triangular facies, clinodactyly, cafe-au-lait areas of the skin, syndactyly of the toes, and elevated gonadotropins (as first described by me in 1953, and in subsequent publications, and by Russell in 1954) occurs with sufficient frequency to be considered a specific syndrome with one or more etiologies.

Originally, the syndrome was known as the Silver syndrome in this country and the Russell syndrome in Europe. More recently, it has been termed the Silver-Russell syndrome or the Russell-Silver syndrome. Hopefully, Drs. Green, Saal, and others will soon provide us with the information that will permit us to make etiology-based diagnoses. Until then, I believe it is reasonable to continue using the names that have historically been assigned to what appears to be a single clinical syndrome with a characteristic phenotype.

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The address given for the Prader-Willi Syndrome Association in Volume 2, Number 3 was in error. The correct address is: 5515 Malibu Drive, Edina, Minnesota 55436. The phone number is 612-933-0113. Marge A. Wett is the Executive Director.