

Adolescent Growth and Pubertal Progression in the Silver-Russell Syndrome

Davies et al categorized the pattern of growth and development of 18 adolescents with Silver-Russell syndrome. All exhibited the classic features of the syndrome, including clinodactyly, triangular facies, and low-set ears. They had grown less than 1 cm in the previous year and had had their growth measured for at least 3 years prior to the onset of puberty. When attempting to describe a mean growth curve for these individuals, the authors paid special attention to the phase effect, ie, variability in timing, duration, and intensity of the adolescent growth spurt. Mathematical modeling utilizing the method of Preece and Baines was applied to the longitudinal data for each child. The age of attainment of pubertal stages was reported for each child as well.

In both males and females, the adult height was well below the third percentile for normal British

children. The standard deviation scores for adult height were -3.61 for boys and -3.58 for girls. However, subjects of both sexes exhibited height velocity curves that were well within the range of normal British children. No abnormal pattern or timing of pubertal events, including the pubertal growth peak height velocity, was observed. The actual peak height velocity was 8.3 cm per year in boys and 8.0 cm per year in girls. In addition, the age at the beginning of the adolescent growth spurt and the velocity at that time were also within normal ranges. The actual growth curve for these individuals with Silver-Russell syndrome demonstrates that there is little catch-up growth during childhood and adolescence, and that growth essentially proceeds normally in childhood. The mean age for the attainment of each stage of pubertal development and the mean age of menarche were also within normal range for British children. Thus, the authors conclude that there is normal pubertal development of sexual characteristics in those with the

Silver-Russell syndrome.

Davies P, Valley R, Preece M. *Arch Dis Child* 1988;63:130-135.

Editor's comment—*This report is one of the first that carefully evaluates a number of individuals with Silver-Russell syndrome who have reached physical maturity. The growth curves that have been constructed for these children should be reviewed by pediatric endocrinologists. They are remarkably parallel to those for normal British children and the velocity curves fall well within the normal height velocity curves for British children as well. This careful characterization of children with Silver-Russell syndrome reemphasizes their poor prognosis with regard to adult stature but reassures that puberty is essentially normal in terms of the adolescent growth spurt and the development of sexual characteristics. Future long-term trials of growth-promoting agents will be required to see if there is to be any hope of catch-up growth in these children.*

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