Limb Lengthening in Children with Silver–Russell Syndrome: A Comparison to Other Etiologies

Vladimir Goldman, MD; Thomas H. McCoy, Jr, BA; Madeleine Harbison, MD; Austin T. Fragomen, MD; Stephen Lyman PhD; S. Robert Rozbruch, MD Hospital for Special Surgery 535 East 70th St, New York, NY 10021 212–606–1415 telephone, 212–774–2744 facsimile goldmany@hss.edu, vovkin@gmail.com

What was the question?

Silver–Russell syndrome (SRS) is characterized by a primary abnormality of growth failure resulting in limb length discrepancy (LLD). Since appropriate caloric intake is a major challenge in these patients, bone healing following lengthening is a concern. We asked whether pediatric patients with SRS (treated with human growth hormone [hGH]) will have uniformly good regenerate tissue consolidation following leg lengthening.

How did you answer the question?

We retrospectively compared the Bone Healing Index (BHI) of pediatric patients with LLD resulting from SRS to similar patients with LLD resulting from tumor, trauma, or congenital etiology. All patients in the SRS group were treated with hGH. BHI was calculated based on radiographic evidence of healing of three out of four cortices. Patients treated for stature lengthening, skeletal dysplasia or those treated with combined methods of internal and external fixation were excluded from the study comparison group.

What are the results?

The SRS group included 7 lengthened segments (5 patients); the comparison group included 21 segments (19 patients). Both groups had equivalent lengthening (3.3 vs. 3.9 cm). The BHI of the SRS group was faster (lower BHI) as compared to control group (29 vs. 43 days/cm, p=0.026). Secondary analysis showed no difference between SRS and trauma patients BHI (29 vs. 31), however the BHI of SRS was significantly lower than both other congenital etiologies (29 vs. 41, p=0.032) and tumor patients (29 vs. 66, p= 0.019).

What are your conclusions?

Limb lengthening regenerate healing of SRS patients treated with hGH is faster than the healing of patients with other congenital etiologies not treated with hGH and as fast as regenerate healing of patients with post—traumatic discrepancies. hGH seems to have been a powerful enhancer of bone healing in the SRS patients and may have applications for bone healing in other difficult cases.