

Smith-Magenis Syndrome Treated with Ramelteon and Amphetamine-dextroamphetamine: Case Report and Review of the Literature

Baek WS^{1*}, Elsea SH²

¹Parkside Medical Group, 1310 San Bernardino Rd, Suite 102, Upland, CA 91786, USA

²Dept. of Molecular and Human Genetics, Baylor College of Medicine, One Baylor Plaza, NAB 2015, Houston, TX 77030, USA

*Corresponding author: Dr. Baek WS, MD. Parkside Medical Group, 1310 San Bernardino Rd, Suite 102, Upland, CA 91786, USA, Tel: 909 608 2008, Fax: 909 608 7705, E-mail: William_S_Baek@hotmail.com

Abstract

Objective: Smith-Magenis syndrome (SMS) is a monogenetic disorder caused by haploinsufficiency of the retinoic acid-induced 1 (*RAI1*) gene on 17p11.2. SMS patients are dysmorphic with developmental delay, autism, attention-deficit hyperactivity disorder (ADHD), and insomnia. Treating the insomnia, ADHD, and disruptive behavior are key in managing SMS; however, to date there are no treatment guidelines or FDA-approved medications.

Methods: We present a case of a 7-year-old girl with developmental delay, insomnia, and behavioral problems whom we had diagnosed with SMS, and treated her insomnia and ADHD.

Results: Ramelteon 4 mg at night decreased her CSHQ (Children Sleep Habits Questionnaire) score from 91 to 79, and amphetamine-dextroamphetamine salt 30 mg daily lowered her Vanderbilt ADHD parent rating scale from 70 to 54.

Conclusions: Ramelteon may be effective in treating insomnia in SMS; larger randomized studies would be beneficial in demonstrating the efficacy and safety of these medications in the future.

Keywords: SMS; *RAI1*; Ramelteon; ADHD; Amphetamine-dextroamphetamine; CSHQ; Vanderbilt ADHD parent rating scale

Abbreviations:

SMS: Smith-Magenis syndrome

RAI1: Retinoic acid-induced 1

ADHD: *Attention deficit hyperactivity disorder*

CSHQ: Children Sleep Habits Questionnaire