



Dental and Craniofacial Findings in Smith-Magenis Syndrome

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This article summarizes research conducted as part of the ongoing IRB-approved NIH protocol 01-HG-0109, Natural History of Clinical and Molecular Manifestations of Smith-Magenis Syndrome (SMS) by the NIH SMS Research Team led by Principal Investigator Ann C.M. Smith, MA, DSc(hon).

This study was published in the American Journal of Medical Genetics Part A 140A:2556-2561 (2006). It is based on the comprehensive craniofacial and dental assessment from 15 SMS subjects with confirmed diagnosis of common 17p11.2 deletions at the National Institutes of Dental and Craniofacial Research at the National Institutes of Health (NIH protocol 01-HG-0109). Children ranged in age from 4 to 19 years. Extraoral and intraoral examination, 3D photographs (3dMD®), and radiographs were taken. Our findings are summarized below with recommendations for parents and clinicians caring for children with SMS.

A strikingly high incidence of developmental dental anomalies was present in our 15 cases. The presence of missing teeth (86.67%) greatly exceeds the 3.5%-8.0% prevalence rate in the general

population. The most frequently missing teeth were premolars, with the second lower premolars most commonly missing. Because these teeth are the last to develop they may have a greater predisposition to developmental alterations.

Taurodontism (teeth with "bull-like" roots containing large, elongated pulp chambers) was also a common finding (86.57%) in both primary and permanent molars. Root dilacerations (an abnormal angulation or curve in the root or crown of a formed tooth) were present in 5 of 115 (33.3%) cases. The high prevalence of these dental anomalies may facilitate diagnosis of SMS, particularly in preschool children, when diagnosis of SMS can be problematic. Keeping regular dental visits is important, so your dentist can recognize any of these dental anomalies.

We found an increased number of caries, lesions, or restorations in our SMS cases. The number of teeth with current or past dental lesions in our 14 to 19 years old subjects was higher (range 4 to 9) than the values reported by the World

Health Organization for the US population, that is, 1.7 for ages 5-17 and 6.6 for ages 18 years and older [WHO Oral Health Country/Area Profile Programme]. Therefore there is a need for constant oral health care and maintenance with help of parents as the child with SMS gets older. Use of fluoride toothpastes and regular dental check-ups at least twice a year are recommended. A balanced diet, low in sugar and rich in fruits and vegetables is also important to preserve the oral health.

The poor oral hygiene associated with gingival inflammation observed in the older individuals with SMS suggest a need for more dental care in the adolescent years. These findings are consistent with the inverse relationship



Figure: Presence of dental plaque and gingival inflammation around teeth as a consequence of inadequate oral hygiene habits.

reported by Martin in 2002 between the child's age and performance of daily living skills. Consider getting an electrical toothbrush for your kids and monitor their oral hygiene habits not only during childhood but also in the adolescent and adult years.*

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What Is Smith-Magenis Syndrome?



Smith-Magenis syndrome (SMS) is a chromosomal disorder characterized by a specific pattern of physical, behavioral and developmental features. It is caused by a missing piece of genetic material from chromosome 17, referred to as deletion 17p11.2. The first group of children with SMS was described in the 1980's by Ann CM Smith, MA, a genetic counselor, and Ellen Magenis, MD, a physician and cytogeneticist. Although the exact incidence is not known, it is estimated that SMS occurs in 1 out of 25,000 births. SMS is underdiagnosed, but as awareness of it increases, the number of people identified grows every year.



Tips for Parents

It is very important to maintain the oral health in your children. Teeth play a role in speaking, eating, and appearance. As we found in our study, individuals with SMS seem to have more difficulty, as they get older to maintain a good oral hygiene causing an increased presence of dental caries and gingivitis in comparison to the general population. These are a few dental recommendations that can be helpful for you as SMS parent or caregiver:

Supervise how your children brush their teeth even in their adolescent or adult years. If they have trouble getting all the food in their teeth after brushing or you notice that the gum is red and swollen, you may consider getting an "electrical" toothbrush for them, this will make tooth brushing easier for them.

Make sure your children brush their teeth everyday after each meal or at least twice a day.

Use of dental floss is also important because food between the teeth cannot be removed completely by the toothbrush and this may cause the appearance of caries or gingival inflammation between teeth. This requires fine motor skills so you will probably need to help them with the dental floss at least once a week. There are different products in the market; we recommended those ones that have a plastic holder so it will make your task easier.

Diet is another important factor in dental caries. Avoid sodas, juices with high sugar, snacks such as chocolates, candies, etc. We recommend drinking water, and eating fruits, vegetables and nuts as snacks. Since it is difficult to eliminate carbohydrates completely in our diet, you can eat them with meals but avoid them between meals.

Your children should visit their dentist twice a year for regular maintenance and ask your doctor for preventive treatments such as fluoride gel application.



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