

Embracing the Inner Toddler in People with Smith-Magenis Syndrome

By *Brenda Finucane*

In my role as a genetic counselor at Elwyn, a large nonprofit agency for people with developmental disabilities, I have worked with hundreds of children and adults who have a variety of genetic conditions, including many with Smith-Magenis syndrome (SMS). My colleague, behavior specialist Barbara Haas-Givler, and I have had the opportunity to meet dozens of PRISMS families in our travels as consultants for school districts and agencies throughout the country. Barbara and I consider it a privilege to be able to work so closely with people who have SMS. We find them interesting, fun, creative, loving, poignant, and always memorable. We gain new insights every day from their dedicated parents and teachers. Like all people with developmental disabilities, those with SMS and their families face many common challenges related to their special educational, behavioral, and residential needs. Compared to people with other types of disabilities and syndromes, however, children and adults with SMS are among the most challenging we serve. We concur with the many parents who've commented that raising a child with SMS is a unique experience!

What is it about SMS that makes this condition so challenging? One obvious answer is that SMS is associated with self-injurious behaviors, such as head banging and picking at fingernails. While these behaviors are distressing, parents rarely report self-injury as the most difficult aspect of raising a child with SMS. Significant self-injury doesn't usually take place on a daily basis, and these behaviors tend to occur in cycles with varying severity. Another possibility is that having a child with intellectual impairment can be a source of stress and sadness for parents. This is true, but

no more so in SMS than for parents of other children with special needs. In fact, there is a wide range of functioning among children with SMS, with many having only mild intellectual disabilities and good communication skills.

Another potential way in which some people with SMS may pose extraordinary challenges lies in their insatiable need for individual attention. Attention-seeking behavior can be exhausting for caregivers. People with SMS are often in competition with siblings or peers for attention, and they tend to monopolize a parent's or teacher's time. Still, attention-seeking behavior by itself is not usually as problematic as the *reaction* of the person with SMS when attention is withdrawn and / or when things don't go exactly as expected. This heightened reactivity to situations and environments is almost universal among people with SMS, in our experience. The behavioral reactions of people with SMS often seem like those of a very young child, regardless of the age or intellectual level of the person. Parents report that they feel like they are "walking on eggshells", sensing that their child is smoldering under the surface and ready to erupt into a major outburst at the least provocation. By necessity, parents often become the best experts on managing their children's behaviors because they've learned to adapt their own behaviors and reactions to avoid potential outbursts. They become hyper-vigilant to potential triggers and develop an innate sense of how to phrase a request. They often know just how hard to push and when to diffuse a situation with humor or distraction. Unfortunately, such nuances are very hard to teach or even explain to people unfamiliar with SMS.

DEVELOPMENTAL ASYNCHRONY

In the special education world, the term “developmental asynchrony” is occasionally used to describe unevenness in the intellectual and socio-emotional development of gifted children. It has been observed that some children with very high IQs can at the same time be socially and emotionally immature compared to their same-aged peers. Although this observation has not previously been described in SMS, we have observed a similar phenomenon in these children and adults. In SMS, intellectual development is generally slower than that of typical children, but emotional development is even more delayed. With age, a gap develops between the intellectual attainment and emotional development of people with SMS, and this disparity poses significant behavioral and programmatic challenges. In our experience, this discrepancy is at the heart of what makes SMS so challenging.

While there is a wide range of functioning among people with SMS, most have mild to moderate intellectual disabilities and can master many skills at the 6 to 8 year old level, if not higher. By contrast, the emotional development of individuals with SMS is usually much younger, with many reacting like toddlers in the way they process information and react to their environment. Many aspects of the SMS behavioral profile could equally describe the development of typical 2 and 3 year olds, including: a low tolerance for frustration; negativity; mood swings; a need to do things for themselves; short attention span; tantrums; anxiety about separation from loved ones; resistance to changes in routine; and repeated (unrelenting!) questioning. Typically-developing toddlers often become emotionally attached and possessive of caregivers, need frequent reassurance about upcoming events, and have a poorly developed sense of time, all common SMS behaviors. Behavioral outbursts in adults with SMS can look very much like the “temper tantrums” seen in toddlers, with a person throwing herself to the floor, kicking and crying. Some of the more unusual behaviors observed in SMS may also have their roots in normal early childhood development.

For example, many typical toddlers go through a period where they insert objects into their noses or ears. This behavior is seen in about a quarter of people with SMS, usually starting in late childhood and sometimes persisting through adulthood. Another very common behavior in children and adults with SMS is the “spasmodic upper body squeeze”, a tic-like gesture that often takes the form of self-hugging or hand-squeezing. In people with SMS, this endearing behavior usually occurs in response to happiness and positive excitement. We consider this a positive aspect of the excitable SMS personality, and although it may seem unusual in an older child or an adult, a similar behavior can be observed among typical young toddlers when they are excited (fig. 1).



Figure 1. Greeting card photo of a presumably typical child engaged in a full-blown SMS self-hug

IMPLICATIONS FOR INTERVENTION

In our experience, developmental asynchrony seems to be more common among people with SMS than it is in most people with developmental disabilities, including those with other genetic conditions. A person with Down syndrome, for example, who functions intellectually at a 6 year old level also tends to react emotionally like a 6 year old. Unfortunately for people with SMS, regardless of intellectual level, their emotional development is typically at the 1 to 3 year old level which tends to be a difficult emotional age. (There’s a reason why

they call this period the “terrible twos”!) The good news is that knowing about developmental asynchrony in SMS can provide us with important insights into intervention.

It is no coincidence that parents of children with SMS frequently look back on the early school years as a “golden age” of development and learning. While parents of preschoolers are adjusting emotionally to the reality of the SMS diagnosis, their children are learning new skills and developing at a slow but steady rate. Most children with SMS seem to be in sync with the preschool and early elementary environment. The school day is filled with hands-on, high interest activities of relatively short duration, and the classroom is filled with bright visual cues, schedules, and reminders. Teachers working in such settings have backgrounds in early childhood education and are experts in the use of distraction and redirection to keep toddlers engaged and happy. They avoid known behavioral triggers and emphasize unconditional positive regard. As children with SMS age through later elementary, junior high, and beyond, the world is not quite so adapted to the very young side of their emotional style. Class periods lengthen, colorful visual organizers, charts, and schedules all but vanish, and there is an increased emphasis on verbal instruction. Few staff in the upper grades and adult programs have a background in early childhood approaches. Behavior plans generally involve consequence-based strategies which may work well for other students but frequently backfire in SMS.

As a child advances in academic areas, it often creates an expectation that the child’s overall development should match up. A 13 year old who reads well at a 4th grade level may be expected to sit through a 50 minute Language Arts class with an emphasis on auditory learning and writing. Unfortunately, if the student has SMS, her “inner toddler” is unlikely to tolerate such a format. She might do better attending half the class and working on a related, hands-on activity in another setting for the remainder of the period. Even though

many adults with SMS are quite capable of performing work tasks, they rarely adapt well to a typical 40 hour work week. Unlike many other adults with developmental disabilities who thrive in such settings, they often have difficulty staying on task without constant one-to-one support and attention. Emotionally and developmentally, adults with SMS do best with a more eclectic work schedule, one with a consistent routine of different high interest activities in a variety of settings. Variety is the spice of life for many people with SMS! For example, one successful young man’s schedule includes three afternoons a week at a traditional work setting, two mornings a week volunteering at the local animal shelter, time spent on chores and errands, and daily workout sessions at the local gym. His schedule is structured (i.e. certain tasks consistently occur at the same time in his weekly routine), but his activities are varied and motivating.

Acknowledging the young emotional level of people with SMS in no way implies that one should continue to treat them like toddlers as they age. Behavior plans that ignore a person’s “inner toddler” cannot succeed, but strategies that neglect the person’s chronological age and academic abilities do not promote growth and learning. A combined approach that incorporates fundamental practices of early childhood education, “age-progressed” to match the interests and skills of the older child or adult with SMS, is often the key to success in working with this population.

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*Brenda Finucane and
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consultants to the stars!*