



Complex Child E-Magazine

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Early Signs of Puberty in Very Young Children with Cerebral Palsy and Similar Conditions by Susan Agrawal

Occasionally I see a parent on a message board asking about early signs of puberty, such as pubic hair or body odor. I am always shocked by the numerous responses that say, “My child has this, too.” While it is well known that children with cerebral palsy, encephalopathy, and similar conditions often enter puberty early, the few studies that have evaluated this subject don’t seem to reflect the large number of families affected by this common condition.

It can be very alarming to see your one-year-old develop pubic hair or realize your four-year-old needs deodorant. It can be even more shocking when your six-year-old starts developing breasts. For some children, these changes may also make them self-conscious or uncomfortable with their bodies.

Is this normal for children with cerebral palsy and similar conditions? Is it something that needs to be treated or evaluated? This is a hard question to answer, since puberty is starting earlier and earlier in the general population, especially among black children. But many children with cerebral palsy and similar conditions start developing early signs as toddlers and preschoolers, and a large percentage have one or more signs of puberty before age nine. While common, it is still something that needs to be studied.

The Medical Literature and Patterns of Puberty in Children with Cerebral Palsy

The literature on early signs of puberty is quite limited. An early article from 1999 looked at 32 children with various neurodevelopmental disabilities, including cerebral palsy, mental retardation, and other conditions, who showed some signs of puberty.¹ This study determined that roughly half of the children experienced central precocious puberty directly caused by their neurological disorders, while the remainder simply had early thelarche (breast development) or pubarche (pubic hair growth) that was of no medical significance. This study suggests that children with neurodevelopmental disorders are up to 20 times more likely to enter puberty early as compared to typically developing children, particularly if they are girls.

One other study addresses not only precocious puberty, but also signs of all secondary sexual characteristics in children with moderate to severe cerebral palsy.² 207 children from centers across the United States were enrolled in the study, which demonstrated that

girls with cerebral palsy entered puberty earlier than boys, but tended to complete puberty later than boys. Due to demographic restrictions, much of the research focused on white girls, who developed pubic hair earlier than the general population, and tended to develop breasts either quite early or later than the average child. White boys also began developing earlier than the general population.

The most important information to glean from these studies is that children with cerebral palsy have a different pattern of sexual development as compared to typical children. Girls enter puberty early, but tend to mature over a longer period of time, menstruating later and completing puberty later. Boys followed more regular patterns, but tended to start early as well. The most common early sign of puberty in both genders is development of pubic hair.

A Survey of Children with Cerebral Palsy and Similar Conditions

What I found lacking in these studies is information on when children began developing *any* signs of puberty, whether they were medically significant or not. This is what I am concerned about as a parent—pubic hair and body odor in my five-year-old—and what I suspect concerns parents the most. To get a clearer picture of how children are affected, I conducted my own survey, recruiting parents through various lists and forums on the internet.

The survey was quite simple. I asked the child's age, sex, and level of function, using the standard GMFCS scale for motor impairment with brief descriptions of each level. Then I asked if the child had pubic hair, body odor, underarm hair, genital enlargement (boys only), or breast development (girls only). Amounts were classified as none, a small amount, a moderate amount, or a large amount like an adult. If a child had one or more of these signs, I asked when the signs first began.

Any child who developed signs of puberty before age nine was classified as an early developer. Of the 66 surveys that were completed fully and correctly, 32 (48%) children demonstrated early signs of puberty and 34 (52%) either developed signs of puberty on time (after age eight) or, for children eight and under, had no signs of puberty yet.

Girls definitely were more likely to show early signs of puberty. 22 girls (63%) showed early signs, while only 9 boys (31%) did.³ In both genders, pubic hair appeared as the first sign most of the time, with or without the development of other symptoms.

Table I: Development of early signs of puberty by gender

	Early Signs of Puberty		No Early Signs of Puberty	
	#	%	#	%
Girls	22	63%	13	37%
Boys	9	31%	20	69%
Total (all children) ⁴	32	48%	34	52%

While children with all levels of function showed early signs, those children who were most severely affected by their cerebral palsy were much more likely to develop early. Children with mild impairments, a small sample size, showed early signs 40% of the time, while children with moderate and moderately severe impairments developed early 44% and 43% of the time, respectively. The most severely affected children showed early signs of puberty 55% of the time.

Table II: Development of early signs of puberty by functional status (level of impairment)

Level of Function (GMFCS scale)	Early Signs of Puberty		No Early Signs of Puberty	
	#	%	#	%
I and II (mild)	4	40%	6	60%
III (moderate)	7	44%	9	56%
IV (moderately severe)	3	43%	4	57%
V (severe)	18	55%	15	45%

Children who did develop signs of early puberty were most likely to develop pubic hair growth first, either alone or with other symptoms, with 78% of children falling into this category. The majority (21 children, 66%) met criteria for premature adrenarche (pubic hair alone or pubic hair plus body odor and/or underarm hair, but without breast development or genital enlargement). 13 children (28%) were likely to have precocious puberty based on their development (pubic hair plus breast development or genital enlargement), and two (6%) did not easily fall into either category.

Table III: First signs of puberty in children with early puberty

sign	Number of Children	
	#	%
Pubic hair with or without other signs	25	78%
Body odor alone	4	13%
Genital enlargement alone	2	6%
Breast development alone	1	3%
Premature adrenarche	21	66%
Precocious puberty	13	13%
Other pattern	2	6%

Children with early signs of puberty developed their first signs at any age. Seven children (22%) showed signs before age two, nine children (29%) between ages two and four, nine children (29%) between ages five and six, and seven children (22%) at ages

seven or eight. Based on this information, 50% of children with early signs of puberty developed them by age four, and 78% had developed them by age six.

Table IV: Ages of children with early signs of puberty

Age	First Signs of Puberty	
	#	%
before age 2	7	22%
2-4	9	29%
5-6	9	29%
7-8	7	22%

This survey obviously has limitations. It is not scientific by any means, and responses are limited by the parent's memory and variation in assessment. There were more girls than boys and the majority of children had severe physical impairments, perhaps skewing the results since both girls and children with greater impairments tend to develop earlier. Even so, I believe this survey demonstrates a more accurate picture of just how prevalent early sexual development is in children with cerebral palsy.

Should Children be Evaluated?

The simple answer is yes, any signs of puberty before age nine should be evaluated. If your child just has isolated pubic hair growth or isolated breast development, your pediatrician might run a few simple blood tests to determine whether or not you need a referral to an endocrinologist. He or she may also order an X-ray to evaluate your child's bone age. In most cases, this is as far as the evaluation needs to progress, and the child simply needs to be watched for further signs of puberty.

If your child has multiple signs of puberty very early on, it is probably best to have a basic endocrinology work-up. This is particularly true if your child is a boy, since boys are more likely to have peripheral causes for early puberty, such as a tumor. This evaluation will attempt to determine the cause of the early puberty, which in most cases will end up as a diagnosis of central (neurologically-based) precocious puberty after excluding any other causes. It is important to get evaluated because it is possible that there may be a secondary cause for the early puberty signs. In addition, treatment is available for children who wish to have it and can benefit from it.

Any child with early signs of puberty should be closely monitored for other signs that may signify that additional evaluation is needed.

Is this a Problem?

Just because most children with cerebral palsy do have some signs of early puberty does not mean it is something that can just be thought of as normal. Traditionally, isolated early pubic hair development (premature pubarche), which is consistently the most common early sign of puberty in children with cerebral palsy, has been described as a benign condition with little medical relevance. It is thought to be caused by early activation of the adrenal glands, and is often called premature adrenarche. It may be accompanied by body odor, acne, and sometimes underarm hair. Isolated early breast development (premature thelarche), especially before age two, is also thought to be a benign condition with little medical relevance.

Recent studies have questioned this fact. Multiple studies have linked early puberty, and particularly premature adrenarche in girls, with polycystic ovary syndrome or metabolic syndromes related to obesity. Moreover, a small study used to illustrate the lack of knowledge in this area demonstrated that girls with premature adrenarche also had elevated levels of cortisol, and a large percentage had psychological disorders and lower IQs.⁵ While these studies may or may not be relevant to children with cerebral palsy, they do demonstrate that a supposedly benign condition may not actually be benign. Much further research is needed in this area to determine if treatment or intervention should be initiated for some or all of these children.

Should this be Treated?

At this time, children with isolated pubic hair growth (with or without body odor) or isolated breast development are not typically treated. Children who have multiple signs of puberty may be treated to slow down the progression of development. This may be important to some children who are self-conscious about the early development of their bodies and to help children reach their full adult height while optimizing their bone age.

Treatment also depends on the age of the child. If a child begins puberty at age two or three, her final adult height may be severely limited by the early puberty. For this child, treatment is likely to be important. But if another child begins developing at age seven or eight, his final adult height may only be moderately impacted, making treatment a tougher decision.

Some families, especially those with children who have more severe motor impairments, may choose not to treat their children. For these families, the slight reduction in height that occurs naturally with somewhat early puberty may be an asset as their children age and grow. Lifting, transferring, and transporting the child may be easier at a slightly smaller size. Smaller, lighter children are also less likely to lose motor function or develop as severe orthopedic problems as compared to larger, heavier children. I am in no way advocating intentionally making a child smaller, but when it occurs naturally, choosing not to interfere with nature may be the right choice for some families.

This decision must be made with input from the child, family, and the child's doctor. It is a decision that will be different for every family.

Where Do We Go From Here?

Despite the fact that signs of early puberty are exceptionally common in children with cerebral palsy and similar disorders, little information exists to guide parents and doctors on evaluation and treatment. It is clear that this is a subject that demands greater evaluation by medical researchers and physicians. Children who have experienced early signs of puberty need to be studied over decades as they age to determine if early puberty has any negative ramifications on their health and development. Guidelines need to be put in place to maximize growth and development.

Most importantly, parents need to be given expectations as to whether this common but not necessarily normal progression of puberty in children with cerebral palsy and similar conditions is OK.

¹ Siraj U. Siddiqi, *et al.* "Premature Sexual Development in Individuals with Neurodevelopmental Disabilities." *Developmental Medicine and Child Neurology* 1999;41:392-5.

² Gordon Worley, *et al.* "Secondary Sexual Characteristics in Children with Cerebral Palsy and Moderate to Severe Motor Impairment: A Cross-Sectional Survey." *Pediatrics* 2002;110:897-902.

³ Two children were excluded from gender calculations since they had indeterminate gender.

⁴ Includes girls, boys, and children with indeterminate gender.

⁵ Lorah D. Dorn and Deborah Rotenstein. "Early Puberty in Girls: The Case of Premature Adrenarche." *Women's Health Issues* 2004;14:177-83.