

11

Educational Considerations for Children with Prader-Willi Syndrome

Naomi Chedd, Karen Levine, and Robert H. Wharton[†]

Children with Prader-Willi syndrome (PWS) present unique characteristics, needs, and challenges to traditional educational environments. Providing an optimal educational experience requires that parents, teachers, and associated service providers be familiar with the issues and educational options for affected children and their families. This chapter provides an overview of the many and varied considerations involved in providing educational opportunities to children with PWS from infancy through adolescence. It delineates specific considerations associated with age and developmental milestones and offers practical suggestions for maximizing the educational experience of students with PWS. In addition, it provides information on current United States (U.S.) federal legislation affecting children with disabilities, including the No Child Left Behind Act of 2001, the nation's latest general education law.

U.S. Education Legislation

U.S. federal law requires that all children with a disability have a written plan that describes each area of educational need and specifies how these educational needs will be met. As of this writing, the current special education law articulating these requirements is the Individuals with Disabilities Education Improvement Act of 2004 (Public Law 108-446), known as IDEA or IDEA 2004. Although the law became effective in July 2005, amending a previous version of IDEA, new regulations have not been finalized (expected in early 2006).

For children under age 3, this plan is accomplished through an Individualized Family Service Plan (IFSP), described in Part C of IDEA. The law covering this age group calls for statewide, comprehensive, interagency service programs for all infants and toddlers with disabilities and their families. It also requires that the IFSP outline services that will be provided to meet the infant's or toddler's developmental needs

[†]Deceased.

in one or more of the following areas: physical development, cognitive development, speech and language development, psychosocial development, and self-help skills.

In contrast to the original 1986 provisions for early intervention services (specified in P.L. 99-457), the current version of the law covering infants and toddlers requires that services for children under 3 be provided in “natural environments,” which may include parks, public libraries, and other community settings as well as homes and day care facilities. If intervention services are provided in other settings, such as a medical center, the IFSP must offer justification for providing these services in a “more restrictive” environment.

Part B of IDEA focuses on children between ages 3 and 21 and requires that school systems provide a plan for identification, evaluation, and provision of special education and related services. Once a child is identified, evaluated, and found in need of special education services, an Individualized Education Plan (IEP) must be implemented. The IEP is an educational blueprint of all aspects of that child’s special education needs and the resulting services to be provided. It is, in essence, a contract between a school and a parent regarding the type and extent of educational and related services to be employed for educating the child. A main difference between Parts B and C of the IDEA law relates to the role of the family. Whereas Part C focuses on supporting the family in their capacity to provide a nurturing environment for infants and toddlers and enhancing coordination of services required by both the child and family, Part B focuses on the role of public schools, guaranteeing that children with disabilities receive a free and appropriate public education (FAPE) in the least restrictive environment (LRE). IDEA, through the amendments passed in 1997, also mandates that all students have access to the general curriculum, rather than being educated with significantly different content and programming.

In addition, differences and changes in the law reflect the needs of the children and parents at different points along the child’s developmental continuum and an increased awareness of the importance of the family as part of the team. The 1997 changes shifted the focus somewhat, from *access to education* for children with disabilities to *improving and demonstrating educational results*, and the 2004 IDEA amendments continue that emphasis, requiring higher standards for qualification of special education teachers.

Another recently enacted federal law that has considerable implications for children with special needs is the No Child Left Behind Act of 2001 (P.L. 107-110), known as “No Child Left Behind” or NCLB, enacted in 2002. According to the U.S. Department of Education, NCLB was designed to work with IDEA and is based on the assumption that all children, including children with disabilities, can meet high educational standards. Like IDEA, it states that students with disabilities must have access to the general curriculum. But it also requires that teachers in all settings—a general education classroom, integrated setting, or substantially separate setting, such as a resource room—need to be skilled in teaching all students. It differs

from IDEA in that it makes schools accountable for student performance.

It is helpful, indeed necessary, for all families engaged in educational planning for their children with PWS to familiarize themselves with the latest provisions of the federal education laws. Several excellent sources of information on these laws can be found on the Internet, including the U.S. Department of Education (www.ed.gov), the National Dissemination Center for Children with Disabilities (www.nichcy.org), and Wrightslaw (www.wrightslaw.com). In addition, getting information about individual state laws and local practices will better enable parents to secure the most comprehensive and appropriate education possible for their children.

Qualifying for Special Services

To be eligible for services under IDEA, a child must have an identifiable condition that interferes with educational progress and school performance to the extent that special education services are required. The law requires that children with a suspected disability be evaluated by an interdisciplinary team, following which an expanded team that also includes the parents designs the child's IEP based on the results of the evaluation; that children be educated in the least restrictive environment; that services be provided when deemed necessary by the IEP team; and that child and parent rights to "due process" be protected. That is, if parents disagree with team recommendations for services or placement, they have a right to a hearing in a court of law.

Program Models

There are variations in classroom options for preschool and school-age children with PWS. Which option is best depends on both the needs of the child and the support services school systems are able to provide in regular and specialized settings. Because children's needs change over time, planning and placement require periodic review and often modification. Any model must incorporate understanding and support for educational, emotional, and social development while normalizing all experiences as much as possible and optimizing quality of life.

Although terms may be defined and interpreted slightly differently, most schools recognize several program models. An "inclusion model" involves placing children with disabilities such as PWS in regular education settings while providing individualized accommodations and support as needed. Where its interpretation is flexible and a broad array of educational and behavioral supports is available, it can be very effective. One advantage is the implication that *all* children are included and valued regardless of ability or capacity to accept ongoing educational challenges. The classroom environment is altered to accommodate all children's needs. Another advantage is that children have an opportunity to develop friendships among classmates with whom they might not otherwise interact.

For children with PWS this model *must* incorporate implementation of support services when difficulties arise and, preferably, be in place before difficulties arise. For example, psychosocial/behavioral consultation may be beneficial in situations such as managing food issues, managing frustration around transitions, and facilitating development of friendships.

Computer instruction and use of a computer in class may be helpful accommodations as well. Other services (speech, physical, and occupational therapy) can either be incorporated into the classroom or take place outside class. It is often necessary to have a classroom aide or paraprofessional to assist with social, educational, and behavioral matters. Depending on the needs of the child, either a one-to-one aide or an aide who works with several children can be effective. However, in both cases, it is necessary for the aide to get appropriate, PWS-specific training in facilitating social interactions between the child with PWS and peers, as well as being able to modify work as needed and provide some behavioral interventions.

Social facilitation can occur by having the child with PWS sit next to classmates during seated group activities, by prompting conversations about topics familiar to the child with PWS, and getting to know all classmates well enough to earn their trust and gain their cooperation. When the inclusion model is not flexible, there are insufficient supports, or appropriate staff training isn't provided, children with PWS may be misunderstood, isolated from their peers, and have fewer opportunities for academic, behavioral, and social development and success.

When an inclusion model is not indicated for the child, other program formats may be pursued. For children with more significant learning and/or behavioral problems who are placed in schools that have large classes and few supports, a smaller, specialized class for most or all academic work may be beneficial. However, classrooms for children with behavioral disturbances (often termed BD classrooms) are not usually appropriate for children with PWS because their behaviors, as well as the underlying causes, tend to be very different from those of children typically placed in such an environment. When classroom teachers need to develop specific management strategies, consultation with a behavioral psychologist may be beneficial. It would be particularly helpful to work closely with a behavioral psychologist who has had experience in working with children with PWS.

Regardless of the classroom model, benefits to social-emotional development accrue from integrated experiences. Integration into a class of typically developing students can be successful for structured activities such as hands-on science or art projects and story times but poses extra problems at lunch and snack times. The concept of reverse integration also works in some instances. This involves having a child, or several children, without special needs join the special classroom to participate with the child with PWS in activities facilitated by the teacher. The child or children can subsequently "host" the child with PWS in the regular classroom.

Some children, especially older children who are experiencing significant behavioral challenges, may benefit at some point from placement in a residential school where predictable structure, a high staff-student ratio, and limited access to food create an atmosphere that greatly reduces overall stress and stress-induced behaviors. Clearly, even in this type of program, the family remains a vital emotional presence and support; close communication and cooperation between the residential program and the home are essential. Sometimes the residential model can provide “crisis intervention” until the circumstances surrounding initial placement stabilize. After a period of time, the child’s success should be reevaluated and the child may either return home and attend a public or other day school or remain in the residential program. Ongoing evaluation of the child’s needs is essential.

No matter which program is selected by the team, teachers and all other school personnel working with the child *must* be taught about food, behavior, and social issues. Such education provides information on which to base teaching methods as well as management strategies for restricting access to food, thereby reducing frustration for the child and staff. Although some behaviors may appear voluntary, such as being extremely stubborn or perseverative, it is essential to help the school/community understand that children with PWS may, at times, display behaviors over which they have little or no control. It is equally important to help school staff recognize the particular strengths of the individual with PWS and gear teaching/learning strategies toward them.

Medical Issues That Affect the Educational Process

Educators need to be aware of several medical and psychological features associated with PWS, particularly an altered level of arousal, the pronounced appetite disturbance previously referenced, and diminished muscle tone and motor planning skills. These features impact on both classroom performance and perceptions of children by teachers and classmates.

Children with PWS frequently demonstrate a diminished sense of arousal. Whereas some children with other medical conditions can be excessively active, the majority of children with PWS are underactive. This characteristic manifests itself as a general lack of dramatic affect, decreased initiation of activities, and a frequent lack of enthusiasm. For example, some children tend to fall asleep in sedentary situations, during a long lecture or silent reading period. Others demonstrate only minimal interest in classroom activities. This feature, sometimes misinterpreted as emotionally based lethargy, attention deficit, or an inability to participate, actually is part of an altered level of arousal mediated by the central nervous system and is unrelated to intelligence.

Most disabling is the inability of children with PWS to control the drive to eat. Unlike arousal, however, this appetite disorder is one of

excess rather than underactivity. This characteristic, while mild in some children and profound in others, is moderated by brain chemicals and beyond internal controls. The lack of capacity for control is not related to cognitive ability, disruptions in the home environment, or a need for comfort or emotional or physical nourishment. Children with PWS can no more control their appetite than they can control a sneeze. It is imperative that the child's environment be modified; trying to change a child's desire for food or food-seeking behavior is futile without limiting access.

Another physical feature associated with this syndrome is low muscle tone and a diminished ability to engage successfully in tasks requiring substantial motor planning skills. Children tend to appear quite weak, and even simple motor tasks such as dressing can be challenging regardless of cognitive level or weight. Moving about the classroom, changing rooms—especially where stairs are required, and engaging in exercise routines or athletic events may be arduous and trying. The inability to match the efforts of nonaffected peers in these areas must be understood on the basis of underlying problems of muscle strength and motor control.

Educational Issues Across Developmental Stages

Infancy and Toddlerhood (Birth to 3 Years)

Because infancy and toddlerhood are periods of rapid development and learning, they are crucial times for children with PWS. Infants with PWS, like other infants, develop strong attachments to parents, siblings, and other caregivers. Separation anxiety, happiness at greeting familiar people, and the particular smiles and eye contact for parents are all part of the special relationships that form between infant and family. Aside from being a delightful quality, this strong social orientation is a vital strength upon which to build a successful learning and intervention program, right from the start. In the first few months, however, sleepiness and nonresponsiveness make nurturing and early attachment to caregivers a challenge. Low muscle tone and generalized weakness cause delay in reaching gross motor milestones such as sitting, crawling, and walking. There is also lag in development of fine motor skills.

Because of developmental delays, infants can benefit from interventions beginning as early as 1 month of age. Feeding and oral motor skills, receptive and expressive communication achievements, and both gross and fine motor development should be assessed and addressed throughout infancy. Programming should focus on physical stimulation, communication, and socialization. Regular "baby" play activities are important. Singing, nursery rhymes, books, mirror play, bubbles, pictures, rough-and-tumble romping, and cuddling are vital developmental activities. However, adjustments are required to compensate for motor and speech limitations, i.e., providing more physical support, exaggerating affect and facial expression, and reinforcing any and all vocalizations and other attempts to communicate. In addition

to directly assisting with motor development, physical and occupational therapists should be consulted regarding strategies to help the child compensate for limitations (see Chapter 10).

During these first years, due to output delays in the motor and speech systems, cognitive processes tend to develop ahead of physical skills. That is, the capacity for thinking and understanding outdistances verbal expression. Expressive language (speech) generally lags behind receptive language (word comprehension). Receptive language tends to correlate highly with cognitive development. The cause of this initial speech delay is poorly understood but is thought to be due, in part, to oral motor difficulty (see Chapter 9). The delay frequently causes frustration for both the infant/toddler and the family. Further, limited expressive skills and motor skills make developmental assessment difficult and can result in underestimation of cognitive abilities.

As soon as difficulties are noticed or diagnosis is suspected, early intervention services should be ordered by the pediatrician. In some states these services are administered through the Department of Public Health, while in other states these services are administered through the Department of Education. Some early intervention programs offer speech, physical therapy, occupational therapy, and play groups, while in other locales these therapies must be funded through other sources. Whatever source is utilized, these specialized interventions should be taught to caregivers and incorporated into daily and weekly routines that encourage optimal adaptation over time. In addition to these special services, infants and toddlers can benefit from experiencing regular day care and playgroups that provide rich opportunities for enhancing social communication and play skills and offer families normalizing experiences.

Preschool (3 to 5 years)

Toddlers, with or without PWS, begin to declare themselves as individuals as they progress through the developmental milestones of walking and talking. Individual personalities emerge as youngsters learn to express themselves, make demands, and struggle to gain control of their environment. Interests, needs, and preferences may be expressed with great conviction, and adult directions may be met with opposition. During this era children *know* what they want and *demand* immediate fulfillment. When gratification is not immediate or goals of others interfere, frustration can evolve into a full-blown tantrum, especially if verbal expression is limited. Who hasn't heard of the "terrible twos"? For children with PWS, this phase may begin a little later and last a little longer.

Although preschool children with PWS exhibit delayed motor and speech abilities, they still crave independence. Delays in achieving these milestones can contribute to resistance to physical activities, frustration due to their inability to communicate, and tantrums. Difficulties adapting to change, controlling frustration, or becoming calm once upset may occur; tantrums may also be more frequent and intense.

Implementing positive behavioral strategies, speech therapy, and physical therapy, and creating a predictable environment are essential.

At the age of 3 years, children become eligible for comprehensive school programs and enter the public education system through the process of evaluation by an educational team. Cognitive, communicative, social, and behavioral skills are assessed in order to guide educational approaches, goals, and placements. In addition to including complete transitional summaries and recommendations from a child's early intervention program, supplying the school team with educational information about PWS can be beneficial. A number of such publications are available through the national and international Prader-Willi syndrome organizations (see Appendices E and F).

Part of the process for determining educational needs and developing the IEP involves standardized intelligence tests. Interpreting test results requires an awareness of the broad spectrum and characteristic patterns of learning difficulties in children with PWS. Some function in the average or borderline range of intellectual abilities, but with accompanying learning disabilities; others are in the mild-to-moderate range of mental retardation; and a small percentage are severely impaired. The numbers of individuals in these categories are not known precisely, and figures cited vary across studies.^{1,2,7,8,9} However, taken together, the literature suggests that at least half of the children with PWS test in the average-to-borderline range. Some studies suggest that there is no correlation between weight and intelligence and that intelligence remains stable throughout development.⁴ Regardless of IQ scores, children with PWS show substantial scatter in ability levels across domains. Moreover, since IQ scores are derived from averaging performances across skill areas, and because children with PWS usually have motor and speech delays, scores reported during preschool years may underestimate cognitive levels.

Measures of receptive vocabulary such as the Peabody Picture Vocabulary Test III³ are better predictors of cognitive development at this age. This test, which requires minimal motor and speech skills, has good correlation with overall intelligence in the general population but is not valid for children who have substantial attention difficulties. Intelligence test scores may become more meaningful for older school-age children with PWS, whose speech and motor skills typically catch up with their other areas of development and for whom there is less discrepancy in performance level across domains. However, when older children continue to show substantial scatter in skill levels across domains, the validity of the IQ scores remains questionable.

While IQ scores for preschoolers are not particularly useful for predictive purposes, the testing results can be quite helpful, indicating learning strengths and styles. The specific test chosen is less important than how test data are interpreted. Reports should discuss performance levels across areas of strengths and weaknesses, with particular attention to learning style and functional supports needed and should be based on assessment by a psychologist or educator experienced in evaluating children with developmental disabilities.

Children with PWS who need specific educational and behavioral intervention may benefit substantially from a specialized preschool experience, such as an integrated, special needs, language-based classroom. Others with fewer needs may benefit from enrollment in a regular preschool where, although less individualized attention is provided, there are substantial advantages gained from socialization, language, play, and group experiences. A common successful approach is enrollment in a combination of special and regular programming, or in an integrated program that includes some children without special needs. Regardless of ability, most children with the syndrome are able to handle two programs as long as approaches and daily activities remain consistent.

Preschool IEPs should include a regular, preacademic curriculum in a developmentally appropriate context. Most children will continue to benefit from continuation of earlier assistance in speech and physical/occupational therapies (see Chapters 9 and 10). These services are most effective when therapists spend part of their time in individual and/or small group interventions and some time consulting with the classroom teacher about how to incorporate therapy goals into the regular curriculum and classroom activities.

In addition to preacademic learning, preschool IEPs and programming should address behavioral and socialization needs, since social development tends to present difficulties as children get older.⁴ Early social skills training, including pragmatics (the practical use of language), is crucial and can help children develop understanding of some of the more subtle aspects of socialization. These cues for social skills are best taught through teacher facilitation of child-to-child interactions and social and dramatic play. Because social skills are so important for future success, both in and outside of school, and because children with PWS require intensive teaching and numerous repetitions, a social skills/pragmatics program should be a priority in every IEP.

Preschoolers sometimes display behaviors that can interfere with learning and acceptance in the classroom, particularly those that result in class disruption caused by schedule changes. These behaviors should be addressed through the IEP, and anticipation is the first line of defense against outbursts. A predictable daily routine is important because there is a tendency to express distress and excessive anger in response to apparently minor alterations in planned activities. A picture schedule for daily routines and a wall calendar for upcoming events can help, and providing information about changes ahead of time may prevent or reduce upsets. Bringing an anticipatory object such as a ball when going to physical therapy or a toy when going outside may ease transitions. Providing predictable "change-in-routine" signals, such as singing certain songs, also helps anticipation and adjustment. Once a child is upset, emotions may escalate rapidly and are difficult to control. When tantrums do occur, it is best to remove the child from the situation, remain nearby, ignore the behavior, and avoid scolding or even trying to reason. For children who show decreased interest and lack of arousal, interspersing motor activity with sedentary projects and seating the child near the teacher can help.

Perseveration is another behavior that challenges the patience of teachers and peers. It often takes the form of repetitive questioning and/or repeated engagement in singular play (such as tearing paper or drawing circles). When repetitive play seems to be soothing, it is beneficial to restrict the behavior to a certain place, perhaps a quiet area of the classroom, and to certain times that are predictably stressful, such as just before snack time, rather than attempting to eliminate it. Once a question has been answered, ignoring the redundancy and changing the subject are often sufficient for this age group.

Finally, at this age children not only observe that eating is unrestricted for their peers but also note that food may be accessible. Several strategies can help manage this situation. During school hours, edibles should be kept outside the classroom in cubbies or in high cabinets. Snacks should be served in child-specific portions in front of each child rather than in large serving bowls and allowing children to help themselves. Supervision is necessary whenever food is available. Children should not have to sit for long periods near others who are eating, nor should they be isolated from their peers. Teachers should be instructed not to use food as a reward for a child with PWS and, preferably, not in the classroom at all. Stickers, colorful markers, or small, safe toys are better choices. Food is an issue that requires teachers and families to function as a team, using structure and positive behavioral approaches that can be implemented at home and school. When teachers are aware of potential difficulties, the environment can be structured to minimize problems.

School-Age Children (6 to 12 Years)

Developmental goals of school-age children include concepts of mastery of tasks and pride in achievements. There is a desire to acquire knowledge, skills, and control of their environment. Providing many opportunities for success is vital to the development of healthy self-esteem during this period when awareness of differences emerges. Some children experience significant tantrums during this period in response to unexpected changes, frustrations, and limited access to food. Skin-picking behavior, common in children with PWS, may also surface during these years in response to frustration and anxiety. Social development continues, as do social challenges, especially in the development of friendships.

Parents can benefit from learning about this stage of educational growth with the assistance of a developmental psychologist, an education specialist, and/or pediatrician knowledgeable about PWS, school options, and opportunities. Psychologists play a major role in the development of IEPs that link parents and school systems. Management of food-related concerns should be included in planning, and recommendations should stress close supervision. During this age period, feeling successful at some physical activity is important in shaping lifelong attitudes toward exercise. Participation in both school-based activities as well as outside programs, such as Special Olympics, can be very rewarding, both personally and socially. Noncompetitive

activities, such as swimming, walking, and group exercise classes can be good alternatives to team sports.

Interventions

Younger school-age children with PWS usually do well in regular classrooms when provided with extra services. Another approach involves regular morning kindergarten classes and a special language-based classroom in the afternoon that allows for individualized teaching. This format can be quite successful and may be continued through the elementary grades.

Meticulous attention must be given to the learning profiles of children with PWS as they begin their school experiences. Their learning profiles generally have characteristic strengths and weaknesses. Strengths, indicated in Table 11.1, are relative to their own abilities, not to peer performance. A particular strength in many children is long-term memory for information. This strength applies to academics as well as to events and names. While initially it may be more difficult to teach new material due to evidence of learning difficulties, it is worth the effort. Functional skills should be encouraged early in the curriculum using positive approaches, such as complementing the student frequently, showing high interest, and providing individualized attention. Classroom activities should move at a brisk pace with variety to maximize motivation.

Most children with PWS become skilled readers. Verbal information is best understood when presented in brief pieces and time is allowed to process it. Much can be learned through hands-on experiences. Absorbing visually presented information is also a common strength. Teachers should be aware that visual materials such as photos, illustrations, and videos are highly motivating, useful teaching aids for most children with PWS. Recent research has indicated that “simultaneous processing” is also a relative strength for individuals with PWS.⁶ They are more adept at integrating and understanding information when it is presented as a whole and they understand the “big picture.” In contrast, they have more difficulty with tasks involving sequential, or step-by-step, processing. It is important that teachers and parents take these strengths and weaknesses into account when designing educational programs or teaching any new skill.

Other learning difficulties often present in children with PWS fall into distinct areas (Table 11.2). One area of relative weakness is difficulty with short-term auditory memory,² which makes it a struggle to remember verbally presented information. Moreover, when a series of verbal directions or a list of steps or objects is presented, the demand

Table 11.1. Characteristic Learning Strengths

Long-term memory for information

Receptive language

Visually based learning through pictures, illustrations, videos

Hands-on experiences

Reading

Table 11.2. Characteristic Learning Weaknesses

Expressive language
Short-term auditory memory
Fine motor skills, related to strength, tone, and motor planning
Interpreting subtle social cues, learning subtle social norms

for understanding and response is compounded by limited expressive ability. It may be that the children have difficulty transferring auditory information from short- to long-term memory. However, when given the opportunity to do this through “rehearsal” and attaching meaning, information can be returned and recalled from long-term memory. The difficulty with being able to remember strings of verbally presented information can be misunderstood as disobedience because the child is unable to sequentially process “pieces” of the directions. It is not uncommon to find students very effective and productive for a given period of time. However, after a while, they appear to “lose it” and need to be retaught, a very frustrating situation for educators. Performance can improve through teaching rehearsal strategies, repeating directions, writing down procedures, using visuals, and modifying verbal instructions. A speech therapist can be helpful in working with the child as well as with the classroom staff to teach these strategies.

Fine motor skills and motor planning tasks, such as writing and drawing, also present as relative difficulties, although a few children are particularly good in this area. Most can improve over time and with practice. Use of a computer should be taught in the classroom beginning as early as kindergarten, and keyboarding can begin to be taught in third or fourth grade. Minimizing writing demands, providing alternative assignments (either color a picture of a train or find one in a magazine), as well as facilitating sufficient opportunities to practice new motor tasks can reduce frustration. Learning is more effective when the right answer can be checked from a multiple-choice format rather than tracing or writing out the answer.

Physical education and therapy are useful for developing strength, coordination, and balance as well as motor planning skills. Gross motor activities such as walking, swimming, and low-impact aerobics are good choices for children with PWS. Scheduling physical activities at predictable times several days each week can positively influence many aspects of life. Children should be encouraged to participate and keep personal records of their gross motor achievements for which they can earn rewards. Physical programming should enhance opportunities for socialization as well as the development of regular, healthy exercise patterns. However, comparisons or competition with others can be discouraging. The record-keeping system should be designed to celebrate personal progress and achievements with guaranteed successes.

While the behavioral and social challenges discussed in the previous section also apply to school-age children, perseveration and obsessive behaviors may intensify. Management should include answering the question once, ignoring repetitions, and changing the subject. Writing

down the answer to the perseverative question on a card to which the child can refer can be helpful. A child who repeatedly asks, "Did I do a good job?" may proudly show an "answer card" that says, "I did a terrific job today." A child who continues to ask, "When do we go home?" can refer to a reminder card that says, "School ends at 2:45."

Home/school communication is essential. Sending a notebook back and forth across environments with information about activities, successes, strategies that work particularly well, and any special diet or behavioral issues can maximize consistency and successes for the child.

Adolescence

Adolescence is traditionally a challenging time for children, having to cope with increased pressures coming from all directions. For those with PWS, the growing awareness of the differences between themselves and their non-PWS peers occurs at the same time that being "just like" one's friends is so important. Individuals with PWS who observe changes in their peers (but none in themselves) are likely to demonstrate increased stress, which may create anger, resistance, and food-seeking behaviors that interfere with learning and adaptation.

Schools can benefit from regularly scheduled parent conferences and outside consultation from a professional familiar with PWS. For some teenagers specific behavioral programs can assist with modulating excessive behaviors (see Chapter 12), while others may require counseling or other forms of support. It should be remembered that out-of-control tantrums, excessive skin picking, and what may appear to be oppositional behavior reflect the inability to cope with stress and should be attended to promptly and comprehensively by school and outside specialists.

While challenges exist, there are positive aspects of adolescence. Many teenagers develop effective verbal skills and become active participants and contributors to school activities. This is a time during which vocational planning and work experiences should begin to be pursued (see Chapters 13 and 14). Being a good reader helps, and hobbies should be encouraged. While it is generally difficult for people with PWS to cultivate friendships independently, satisfying relationships are possible, especially if parents, teachers, and other supportive adults in the community are involved. Due to behavioral challenges—regardless of cognitive functioning—most are not yet ready for the full autonomy their same-age peers are achieving; most need continued supervision and protection. Both teachers and parents need to provide extra security in the school and home while seeking ways that adolescents with PWS can increase independence, participate in enjoyable activities, and improve their quality of life.

School programs should capitalize on strengths and teach coping skills for dealing with challenges. As with younger individuals, daily and weekly schedules should be as consistent as possible with minimal transitions. In addition to learning, behavioral, physical, and social considerations, prevocational teaching and planning become critical.

While academic work clearly continues to be a part of school programming, emphasis should be placed on practical uses of learning such as application of math skills to manage money, time, and use of public transportation. Skills necessary for community living should be part of any prework curriculum. Enhancement of social skills needs to continue, and educators should consider offering one of several excellent courses about sexuality that are designed specifically for persons with developmental disabilities.

Some adolescents may benefit from individual counseling in order to express feelings and work through frustrations associated with a desire for independence and conflicts surrounding the issue of “being different.” Providing therapeutic counseling and support during a long walk has worked well for some adolescents with PWS, as this model also supports physical well-being while promoting good mental health. A support group of other individuals who face similar challenges is usually very helpful as well. More detailed discussion of educational issues for adolescents with PWS appears in a separate chapter (see Chapter 13).

Conclusion

Children and adolescents with PWS can achieve many successes in school and community-related activities. When educators understand each child as an individual with special strengths and needs, they then can determine how the attributes of PWS can contribute to successes and can assist families, school staff, and ultimately, the children themselves to cope with the challenges associated with the presence of this syndrome and maximize their functioning, independence, and happiness.

Editor's note: This chapter is adapted from “Educational Considerations,” by Karen Levine, PhD, and Robert H. Wharton, MD, *Management of Prader-Willi: Syndrome*, 2nd Edition, Springer-Verlag, 1995.

References

1. Cassidy S. Prader-Willi syndrome. *Current Problems in Pediatrics*. 1984;14:1–55.
2. Curfs LMG, Wiegers AM, Sommers JRM, Borghgraef M, Fryns JP. Strengths and weaknesses in the cognitive profile of youngsters with Prader-Willi syndrome. *Clinical Genetics*. 1991;40:430–434.
3. Dunn L, Dunn L. *Manual for the Peabody Picture Vocabulary Test*. 3rd ed. Circle Pines, MN: American Guidance Service; 1997.
4. Dykens E, Hodapp R, Walsh K, Nash L. Adaptive and maladaptive behavior in Prader-Willi syndrome. *Journal of the American Academy of Child and Adolescent Psychiatry*. 1992;31(6):1131–1136.
5. Dykens E, Hodapp R, Walsh K, Nash L. Profiles, correlates, and trajectories of intelligence in Prader-Willi syndrome. *Journal of the American Academy of Child and Adolescent Psychiatry*. 1992;31(6):1125–1130.

6. Fidler DH, Hodapp RM, Dykens EM. Behavioral phenotypes and special education: parent report of educational issues for children with Down syndrome, Prader-Willi syndrome, and Williams syndrome. *Journal of Special Education*. 2002;36(2):80–88.
7. Gabel S, Tarter RE, Gavalier J, Golden WL, Hegedus AM, Maier B. Neuropsychological capacity of Prader-Willi children: general and specific aspects of impairment. *Applied Research in Mental Retardation*. 1986;7:459–466.
8. Taylor RL. Cognitive and behavioral characteristics. In: Caldwell ML and Taylor RL, eds. *Prader-Willi Syndrome: Selected Research and Management Issues*. New York, NY: Springer-Verlag; 1988:29–42.
9. Warren HL, Hunt E. Cognitive processing in children with Prader-Willi syndrome. In: Holm VA, Sulzbacher SJ, Pipes P, eds. *The Prader-Willi Syndrome*. Baltimore, MD: University Park Press; 1981:161–178.