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Educational and Social Issues for Adolescents with Prader-Willi Syndrome

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Adolescence—ages 13 to 21—can be a trying time for the individuals experiencing it, as well as for their caregivers. This is no less true for the child with a disability. Parents and teachers need to understand typical developmental experiences during this time in order to recognize experiences and behaviors that may be reflective of a child's particular disability.

It's All About Change

The transitions from elementary school to middle school and again to high school pose dramatic changes for any student. The safety and security of being in a single classroom in a familiar building with the same teacher for at least one, possibly several years is suddenly taken away. Classmates who have been together for many years are dispersed. For some students, this provides an exciting opportunity to try out new behaviors and put on a more grown-up persona. For many students with disabilities, it can be the beginning of a special kind of loneliness and rejection. This is especially true for students with significant cognitive and social deficits such as those with Prader-Willi syndrome (PWS).

It is not unusual for a student with PWS to be several academic grade levels behind his/her same-age peers. This may not have been a significant problem in the student's earlier class placement, where there was a single teacher who knew the student's capabilities in all areas and could adapt the environment and curriculum accordingly. The social gap between the student with Prader-Willi syndrome and his/her classmates also may have widened, as peer relationships now require greater sophistication and savvy. For many, an additional significant change involves the loss of the one-to-one paraprofessional assigned to him/her for a good part of the elementary school years.

The student placed in an inclusive program may be involved with several teachers and potentially new groups of students in each class. Each teacher has his/her own style, expectations and, perhaps, beliefs about the student with PWS. Most, if not all, will have had no prior experience working with students with PWS; further they will not have the luxury of getting to know the student every day, all day. Thus, a great deal of collaboration and contact between school and home must occur on a routine basis.

Students placed in self-contained classrooms have a greater possibility of being with at least one or two familiar classmates. Typically in this setting there is one teacher with one or more paraprofessionals. Physical transitions are fewer and a consistent routine is more likely.

Some students may transfer to a special school for students with learning disabilities at this time. Here, too, staff need to be prepared for working with the student with PWS. The student, on the other hand, may need reassurance that the school change is not the result of some kind of failure, but an opportunity to better learn and to make new friends.

In all situations, however, administrative, clinical, and support staff are new, requiring the establishment of new relationships. Further, all who will now interact with the student throughout the day have a great deal to learn. Caregivers must consider and plan for how this learning and relationship-building will occur.

Moving from classroom to classroom throughout the day poses a special challenge for many individuals with PWS. The building is probably much larger than the previous school building, and locations of the various classrooms and offices must be learned quickly. Further, changing classes means moving fairly quickly from room to room so as not to be tardy. This requires prior organization of needed materials for each class.

Larger buildings also mean more food sources, a source of increased stress. It also means bigger and more chaotic cafeterias with food service staff that are unfamiliar with the drives of a student with PWS. It means resisting the many opportunities to obtain food (e.g., open lockers, offices, snack machines). All present significant challenges for the person with Prader-Willi syndrome, challenges that should not be underestimated.

Transportation to and from school will also likely be affected. There may be a new driver and bus monitor, new children on the bus, and a new route requiring a new pick-up and drop-off time. This change alone can be very stressful for the student with Prader-Willi syndrome and should be considered when designing transition plans.

Students with PWS often experience increased stress and anxiety when facing the many changes required, feelings that are sometimes manifested in unacceptable ways. The student may be “in trouble” more often, with the result that the family is getting more and more phone calls from the school. In some instances, the school may decide that it can no longer serve the student within its existing programs and may recommend placement in a special class or school for students with behavior difficulties. Such placements have not proven optimal for the student with PWS. Schools for children and adolescents with “routine behavior disorders” focus on intervention strategies aimed at returning the student to a less restrictive program. These strategies assume that behavior can be substantially altered using traditional behavior modification approaches. Since most such strategies are ineffective for those with Prader-Willi syndrome, placement in traditional "behavior disordered" programs often results in worsened rather than improved behavior for the student with PWS. Programs utilizing environmental modifications and a variety of situation-specific preventative strategies are most effective for managing PWS-associated behaviors. Traditional rewards and consequences strategies—no matter how seemingly powerful—cannot override some of the neurological drives inherent in the syndrome.

With these background caveats in mind, let us consider some transition strategies for assisting the student with PWS in moving to middle and high school.

Transition to Middle or High School: Paving the Way

One of the most successful strategies for an educator is to "be prepared." Training and support in working with the student with Prader-Willi syndrome should be the first priority for middle or high school personnel unfamiliar with the affected student's special needs. Parents may need some additional support as well. When all people who work with the student with PWS are informed and environmental concerns have been addressed, there is a greater chance for a positive learning experience to occur.
Strategies:

- The student's multidisciplinary team should meet as early as possible in the school year prior to the planned transition in order to identify the specific program and services the student may need, and to develop a transition plan.
- If a change to a new program or school is necessitated, parents and school personnel should visit to ascertain the fit between the program's services and the student's needs. One criterion for future success is that the new program or school demonstrates openness to learning about Prader-Willi syndrome from previous teachers, parents, and experts in the field.
- Once a program and setting are determined, visits for the student to tour the building, learn where his/her primary classroom is located, and meet the teacher(s), administrators, and clinical staff should be arranged. If the student will have more than one teacher, a key teacher who will be the decision-maker and primary communicator should be identified. A regular and easily used communication system between other school personnel and the key teacher, and between the key teacher and the family or caretakers, should be established. For caregivers, e-mail may be preferable to sending a notebook back and forth in order to prevent the student from ripping out pages or "losing" the notebook.
- The daily schedule (if known), especially the snack and lunch schedule, as well as where food will be eaten, should be reviewed with the student.

- Similarly, school or classroom policies or procedures impacting the student (e.g., code of conduct, dress codes, behavior management programs) should be reviewed. If the student takes issue with any of these policies or procedures, a meeting with the principal to review the policies often resolves the problem.
- Environmental changes that need to be made prior to the school year should be determined and a plan for their implementation developed. For example, is food kept in classrooms? Where are lunches and snacks kept? Should the student eat in the cafeteria, and if so, what degree of supervision is needed? If there is a food preparation component to any of the student's classes, how will this be managed while considering the needs and abilities of the student?
- Does the student need an adapted physical education program? Will there be opportunities for the student to engage in physical activity beyond the one time per week in a physical education class?
- A meeting should be held with personnel from the transportation staff or company, especially the driver and bus assistant/monitor, to discuss the nature of PWS and the specific needs of the student.
- It is often helpful for the student to view pictures of school personnel prior to starting the school year, particularly if the student is very anxious. Therefore, caregivers may want to photograph the school, the assigned classroom, and the key teacher(s). This way, over the summer months, caregivers can use visuals to review what will be happening and with whom during the next school year.
- Finally, assuming all the basics are in place for the next school year, it is advisable to set up a time in the fall for a training session on PWS. The advantage of waiting until after the student has been in the new program for a few weeks is that school personnel will have a chance to know the student; subsequent training will then have more meaning and student-specific questions can be addressed.

Behavior Challenges

The typical characteristics associated with PWS persist throughout adolescence. Certain behavioral characteristics, such as those associated with obsessive-compulsive disorder (e.g., hoarding and skin picking) often become progressively worse as the child moves into the teen years.[2] However, this is not universal.

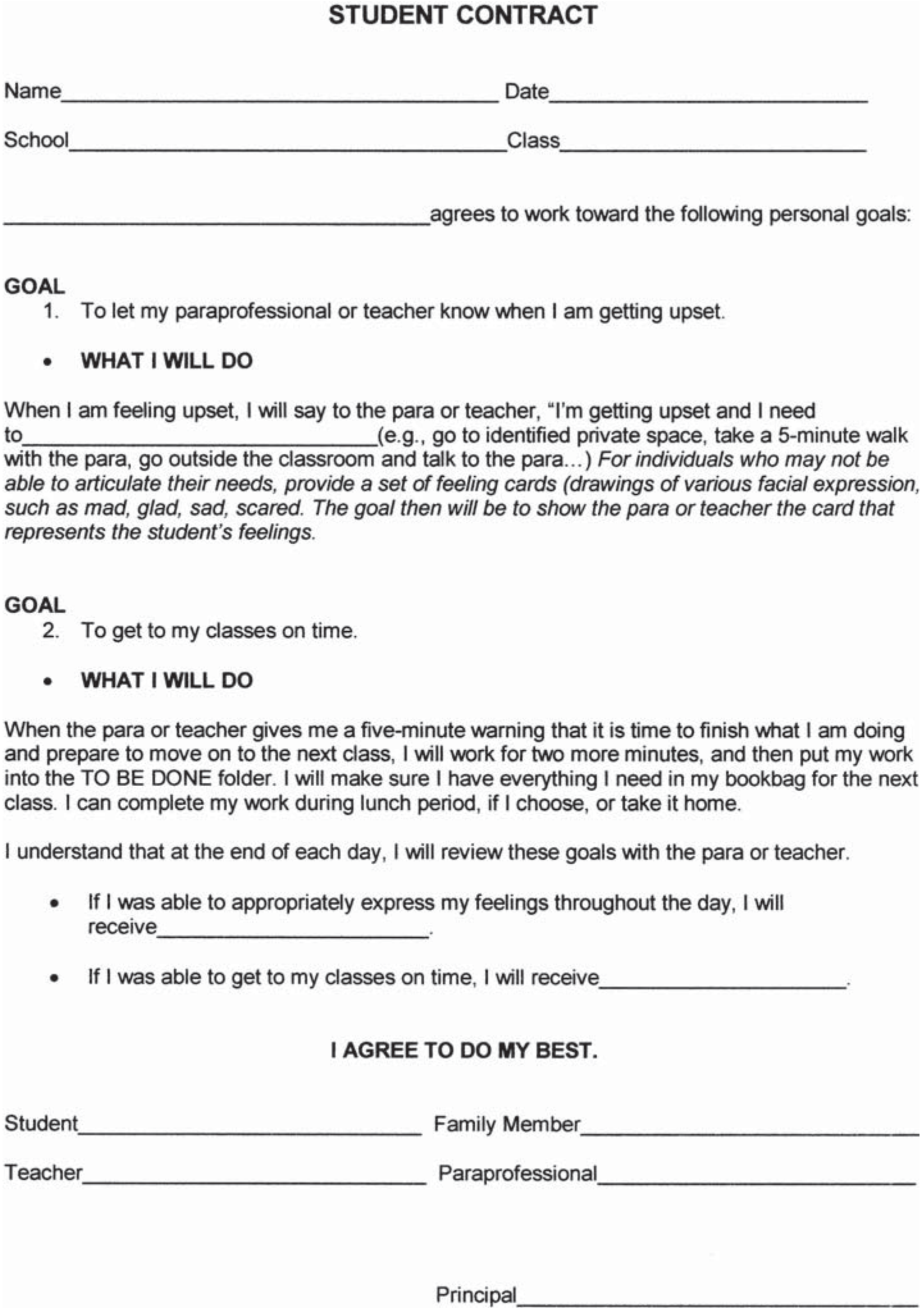
Some caretakers report an increased skill, ability, and creativity in obtaining food or other desired items or activities. It is not unusual to hear parents and teachers report that the adolescent is suddenly doing all kinds of food seeking that he/she never did before. Temper tantrums and "shutdowns" that they may have had throughout elementary school now take on a new dimension, with increased intensity and keener awareness of how to obtain desired responses. The adolescent may also experience greater and more exacting demands, which produce greater stress and confusion, resulting in an increased frequency and intensity of difficult behaviors. The majority of individuals with PWS (75% to 90%) do exhibit acting-out behaviors, including temper tantrums, impulsivity, aggression, and stubbornness to a greater degree and with greater intensity than individuals with mild or moderate mental retardation with other etiologies.[1,4] This might cause a new teacher to erroneously conclude that the behaviors of the student with PWS are worsening, when comparing them with those of children with other developmental disabilities.

Research has found that older children with PWS are more likely to experience withdrawal and depression relative to younger children, but these symptoms may abate in adulthood.[2] Weight may also affect behaviors, particularly individual, internal maladaptive behaviors such as mood disorders; those who are thinner demonstrate more maladaptive behaviors than those who are heavier.[3,9] Studies have found more symptoms of psychosis (confused thinking, delusions, and hallucinations), depression (anxiety, fearfulness, and sadness), and characteristics of dependent personality among thinner individuals with PWS.[3,9] While many perceive that individuals with higher IQs have greater and more severe maladaptive behaviors, the evidence suggests no significant differences based on IQ.[3]

Since there is significant individual variation in behavior, behavior management strategies or interventions need to be person-specific. This necessitates involving the interdisciplinary team, including the family, and the individual if appropriate. Behavior management strategies take a different shape in the adolescent and young adult years with an increasing emphasis on self-awareness and self-monitoring. Sticker charts and verbal praise are no longer adequate in providing needed supports. Behavior management strategies have far more success if the student had input in the development of the strategies. Thus, one needed focus is on the individual's recognizing his/her problematic behaviors and agreeing to change them. A common example would be skin picking. Currently there is no known cure, nor is it clear the extent to which individuals can control the urge, but it may be possible to more easily redirect an individual when that person has recognized the problem and agreed to address it. For this reason behavior contracts are generally a more effective strategy than sticker charts for the adolescent and young adult (see Figure 13.1). As with all teenagers, students with PWS react very negatively to any perceived condescension. There must be a *genuine* foundation of respect and appropriate levels of involvement to ensure the maximum effectiveness of any behavioral strategies. While for many teenagers specific behavioral programs can assist with modulating excessive behaviors, others may require ongoing counseling or other forms of support.

Food/Nutrition

Larger schools have a greater number of food sources available. Middle and high school cafeterias often provide numerous food choices in addition to a relatively wholesome school lunch. Many offer soda and bagels, chips and candy, as well as a variety of dessert options. The typically developing adolescent does not choose to spend money on the school lunch (unless it's pizza), opting *instead* for soda and snack



STUDENT CONTRACT

Name______________________ Date________________

School______________________Class________________

______________________agrees to work toward the following personal goals:

GOAL

1. To let my paraprofessional or teacher know when I am getting upset.

- **WHAT I WILL DO**

When I am feeling upset, I will say to the para or teacher, "I'm getting upset and I need to__________________(e.g., go to identified private space, take a 5-minute walk with the para, go outside the classroom and talk to the para...) *For individuals who may not be able to articulate their needs, provide a set of feeling cards (drawings of various facial expression, such as mad, glad, sad, scared. The goal then will be to show the para or teacher the card that represents the student's feelings.*

GOAL

2. To get to my classes on time.

- **WHAT I WILL DO**

When the para or teacher gives me a five-minute warning that it is time to finish what I am doing and prepare to move on to the next class, I will work for two more minutes, and then put my work into the TO BE DONE folder. I will make sure I have everything I need in my bookbag for the next class. I can complete my work during lunch period, if I choose, or take it home.

I understand that at the end of each day, I will review these goals with the para or teacher.

- If I was able to appropriately express my feelings throughout the day, I will receive______________.
- If I was able to get to my classes on time, I will receive______________.

I AGREE TO DO MY BEST.

Student____________________ Family Member____________________

Teacher____________________ Paraprofessional____________________

Principal______________________

Figure 13.1. Sample student behavior contract.

foods. Individuals with PWS will choose the school lunch *in addition to* the soda, snacks, and desserts unless closely supervised. Research suggests that individuals with PWS are more likely to choose a larger quantity of a less-preferred food over a lesser quantity of a more-preferred food.[5] This provides an opportunity for the student's caretaker or cafeteria aide to promote healthy choices, such as offering a large bowl of chef salad as an alternative to a lesser quantity of macaroni and cheese. Many parents report that as their children age and their social world expands, so too do their exposure and opportunities to access food. Therefore, it is imperative that educators work with parents in planning for any events that involve food.

Fitting In: The Adolescent Struggle

The social success of a typically developing adolescent depends, in large measure, on how well they can follow fashion, music, and other leisure time trends. Most teenagers are very self-conscious and quite critical of themselves and others. The student with Prader-Willi syndrome may have had some very good relationships with other classmates up until this point; in this period, however, the academic and social gap widens significantly. The typically developing student's tolerance for the immaturity and rigidity of the student with PWS frequently lessens, particularly for episodes of lying, stealing, and occasional aggressiveness, which are not uncommon in those with the syndrome. The student with PWS may not understand why classmates are distancing themselves both physically and psychologically from him, or even teasing or bullying him, and therefore may try even harder to be part of a social group. This lack of a social group often precipitates a transfer of the student with PWS out of inclusive situations and mainstream classes into smaller, less-integrated special education settings. Optimally, other solutions will be employed.

There are many students with social skills deficits who have disabilities other than Prader-Willi syndrome. They may be students with learning or emotional problems, sensory or neurological impairments, or students with medical disorders or physical disabilities. This constitutes a fairly large population of students who would benefit from formal social skills training. While teachers seek to take advantage of teachable moments in developing social skills in their students, this is sometimes inadequate.

It has been shown that the most effective social skills training occurs in small groups with students meeting (at least) weekly throughout the school year, facilitated by an adult proficient in social skills training.[6,7] In this setting, the group can practice many practical skills such as listening, using good manners, asking someone out for a date, expressing anger and frustration, and solving a variety of likely problems, as well as learning relaxation techniques.

Role-play is particularly useful. Students are given situations to act out, first with inappropriate behaviors to be critiqued by the group, and then with appropriate behaviors producing desired outcomes. The supplemental use of videotaping provides a powerful training tool, especially for those students with PWS who are predominantly visual learners. Students, including those who participated in the role-play, are able to see themselves as others see them and critique their own behavior, both positively and negatively. It thus makes the role-play situation more real. It is also important to create situations that mimic real life. For example, a student may want to learn how to join a club, how to ask a friend to go to a dance, or even how to dance!

Additionally, teachers and caretakers need to be up-to-date on current fashions and trends to help the student learn how to dress in style and converse about subjects of general interest. In 1960s jargon, they need to learn how to be “cool”—or, in today’s jargon, “wicked,” “sweet,” or “awesome.”

Young people with PWS want what most teens want—friends and a close relationship with another. They can carry this desire to extreme lengths in their search for a boyfriend or girlfriend, including obsessing over a particular individual (sometimes a favorite adult). Dating and relationships are another area where adolescents with PWS need education and guidance. Phone etiquette needs to be taught. Excessive and often inappropriate use of the phone can become an obsessive, problematic situation. Rules and guidelines may be needed that outline when and where public displays of affection can take place. Many need to learn what should be said and done to make sure consent is obtained. It is recommended to use this consent approach for hand-holding, hugging, and other intimate situations. Educators need to make sure they have an understanding of what their students are being told at home. If we rely on students with PWS to learn from their peers, they may see and mimic inappropriate affectionate behavior. Some parents and providers have used written contracts to outline specific dating or friendship behaviors, and when and how they are to be employed.

Sexuality is also part of social development and, while individuals with PWS may not go through a complete puberty, they are still subject to sexual feelings and desires to be romantically involved. Most schools provide some kind of health class that describes the physical changes of adolescence, so the students know what to expect. For students with PWS, these changes may be minimal or nonexistent, unless they are receiving hormone therapy. This is often a very sensitive area for individuals with PWS, who want to know why they are not growing, have an unusually small penis, are not developing facial hair or experiencing a deepening of their voice, or, for girls, are not having a period. Girls frequently want to know about their future as mothers. While reproduction is highly unlikely, it has happened in several documented cases of young women with PWS. Instructors and caregivers must be prepared to deal with these issues in an honest, respectful, and sensitive manner. Collaboration between home and school is essential to convey information and advice in a consistent fashion. See Chapter 21 for further discussion of sexuality issues.

Finally, many individuals with PWS are highly motivated to play the role of nurturer or caregiver. This drive allows for opportunities to have the student assist others who may be younger or less capable (e.g., push a wheelchair, carry someone’s knapsack, read to a young child, or be in charge of the classroom pet). There are endless opportunities to use the student’s strengths and interests to build social skills. Caring for others can be a big boost to one’s self-esteem, a critical component of self-development and socialization.

Looking Ahead

Many parents and caregivers ask, "Is there life after graduation?" The answer is "Yes, but it must be carefully designed." Federal special education laws mandate that transition planning be initiated when the student is 14 years old. This is the time to consider the specific strengths and needs of the student in light of future goals, whether they include continuing in an educational program, securing a job, or participating in a vocational or pre-vocational program. While the student is eligible to remain in school until 21 or 22 years old (varies by school district), not all will; those that do should achieve maximum benefit from available programs. Schools may provide a variety of community-based work experiences whereby the student experiences the world of work, gains a better understanding of work expectations, and determines where his/her interests lie.

Some school districts have a transition specialist who works with the multidisciplinary team in assessing the student's strengths and needs, designs a high school curriculum that addresses these areas, and identifies community resources relevant to the student's goals. Table 13.1 gives an overview of the timetable for various transition activities and events during the adolescent education years.

Making specific plans for entry into the adult world can be particularly stressful for all. The protections of IDEA no longer apply and there are no laws guaranteeing services for the student after 21 years of age. For most caregivers, and even school personnel, this comes as a surprise, often too late to have the optimal transition plan in place.

There are many things to consider in planning for the future:

- If the student is capable of further education in either a 2-year college or specialized training program, how will it be paid for and who will monitor his access to food?
- If the student has proven capable of entering the world of competitive employment, who will assist him/her in locating a job and providing the necessary job coaching? How will access to food be restricted?
- If the student requires a sheltered vocational placement, where does one exist? Is there a waiting list? Does the student meet eligibility requirements? Are the program, environment, and staffing compatible with the needs of the student with PWS? Is the program open to receiving training on the syndrome?
- Do the student and caregiver(s) desire a residential placement after high school? If so, what are the options? Is there a waiting list? Does the student meet eligibility requirements? What is the funding mechanism? If a Prader-Willi designated residence is unavailable, is the available program amenable to making significant environmental changes and to receiving training about PWS?
- Who knows the answers to these questions? Where does one go for help? Does anyone out there understand Prader-Willi syndrome as well as the school personnel?

Table 13.1. Timetable for Transition Services

This is a guide to when to expect certain activities to occur during the transition years in special education. As this is an individualized program, there may be great variation in scheduling from one student to another.

	Ages 14–18	Ages 18–20	Ages 20–22
Academic	• If mainstreamed, continue academic goals; may graduate at 18, which may cease special education services. • If in Special Day Class (SDC), include functional skills in goals.	• If mainstreamed and still in special education, continue academic goals. • If in SDC, continue mix of academic and functional skills goals.	• If mainstreamed, continue academic goals; determine use of post-secondary education and apply for funding and to post-secondary placements (junior college, trade school). • If in SDC, de-emphasize academics, increase focus on domestic and vocational functional skills.
Domestic	• Assess skills in personal hygiene, nutrition, cooking, household maintenance. • Build skills.	• Build skills.	• Continue to build skills using home, classroom, and community environments.
Community	• Assess skills in purchasing transportation, interpersonal communication and relationships, use of community services.	• In community more frequently as learning skills.	• Continue to build skills in all domains in the environment in which those skills will be used.
Vocational	• Rehab counselor assigned, application made, with proof of disability and vocational potential. • School assesses vocational knowledge, interests, aptitudes, skills.	• Begin to meet/interview vocational agency staff. • Gain a variety of work experiences and skills.	• Continue gaining work experience. • Select vocational agency, if wanted, to serve needs after leaving school.
Site of Educational Activities	• Classroom with some community-based exposure.	• Classroom. • Community. • Vocational sites.	• Classroom may be on junior college campus. • Community. • Vocational sites.
Leaving School	• Some choose to leave (drop out) with incomplete skills.	• Many of non-disabled friends have left school (graduated) and gone on to college, trade training, or work. • May graduate but continue studies.	• Will exit special education sometime during age 21, typically with certificate, not diploma.

Source: Adapted from J.A. Seguin and R.M. Hodapp, *Transition From School to Adult Services in Prader-Willi Syndrome: What Parents Need to Know.*[8] Copyright 1998, The Prader-Willi Syndrome Association (USA).

- Without early planning, the student with PWS may find him- or herself sitting at home with no work or "adult" residence. In the worst case, a parent may have to quit his/her job to provide supervision for the young adult at home.

To facilitate a smooth transition, one of the first services needed is that of case management, typically provided by a county or state agency or contracted out to a community-based agency. The school district should provide this information. Case managers assist the student in linking up with a variety of community resources for which he/she qualifies. Generally, case management services are provided to individuals with mental retardation and/or developmental disabilities. This can be a problem for individuals with PWS, since many exceed the IQ eligibility criterion (usually 70 to 75), and many states do not recognize developmental disabilities unless the person also has mental retardation. When this occurs, help can be obtained from the national Prader-Willi Syndrome Association.

Service providers for adults, once identified, should be brought together no later than when the student is 18 years of age (see Table 13.1, Timetable for Transition Services). Both vocational and residential service providers may be needed. A note of caution is in order: the language of adult service providers is quite different from that of educators, so families and school staff should ask for translations. Also, families seeking residential services for their child should be made aware that residential programs specifically designated for people with PWS are scarce; thus the student may need to be on a waiting list while looking at other residential options.

Vocational services are also difficult to secure for individuals with PWS. Most programs promote competitive employment and cannot provide the degree of supervision required by a person with PWS. Typically, the student is matched with a job and provided with a temporary job coach (someone who assists in training the individual until he/she is able to perform the job independently). In the case of individuals with PWS, the need for a job coach, in the absence of diligent and caring co-workers, is long-term, if not permanent. Parents are encouraged to tap their own resources by calling friends in business or others who can provide some kind of meaningful employment in a safe environment.

Because many individuals with PWS are nurturing and loving, common career choices include working with animals or children. Some individuals are dog walkers or work at shelters or veterinarian offices taking care of the cages. Jobs with children where there is little-to-no food are difficult to find. While many individuals could be very effective in caring for young children in a preschool setting, there would need to be supports and environmental adaptations where food is involved. Some look into volunteer positions at local libraries where they may read to young children.

For a more detailed discussion of the transition process and related issues, see Chapters 14–16.

Conclusion

In short, early planning along with a great deal of networking and creativity is required on behalf of adolescents with PWS. Schools must provide relevant programs to prepare these students for an entirely new set of challenges. Parents must continue to work collaboratively with educators while they advocate for their sons and daughters. Indeed, the adult service system is generally not as "user friendly" as the educational system and requires continued diligence in securing needed services and programs.

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