

Early Intervention Services for the Child with PWS

By definition every child age birth to three years who has received a diagnosis of Prader-Willi syndrome is eligible to receive Early Intervention Services provided by the State of California through the Regional Center system. Additionally, it continues to be successfully argued that all individuals with PWS be made eligible to receive Regional Center services for the remainder of their lives.

As soon as your child receives a diagnosis of Prader-Willi syndrome you should contact or be contacted by your local Regional Center office to begin important therapeutic services. You and your caseworker will develop a plan for your child, called an Individual & Family Services Plan (IFSP) which will list the types, amount, and duration of therapies your child will receive.

Early Intervention Services should include at a minimum occupational therapy, physical therapy, and oral motor therapy (in infancy), and speech and language therapy. Other therapies, such as assistance with feeding, music therapy or aqua therapy (physical therapy performed while in water), hippotherapy or therapeutic horseback therapy may also be fun and helpful, though your Regional Center may not always be able to provide these services. Social skills therapy will be helpful when your child reaches age three to four years. Though outside the purview of services provided by your Regional Center, don't forget to look into your local community classes such as Mommy & Me, Gymboree, My Gym, etc. for more exploration and fun with your child!

Overview of Occupational Therapy Services

Occupational therapy should begin in infancy to develop muscles and improve fine and gross motor skills. As your child grows older, gross motor skill development will be addressed by a physical therapist, while occupational therapy will continue with more focused work to improve hand strength, finger strength and dexterity, motor planning, coordination, and sensory integration issues.

Occupational Therapy for the infant will focus upon improving your baby's overall strength and endurance. Some occupational therapists specialize in feeding issues and are available to help your baby work on sucking, swallowing, and later chewing skills. The supplement Coenzyme Q-10 often helps improve this motor planning skill (see *Supplement Information Specific to PWS* inside this handbook). The therapist will place your baby in various weight-bearing positions, such as on his or her tummy to help strengthen the neck, back, and shoulder muscle groups. Therapy will utilize toys, textures, songs, movements, and more, with the emphasis always being on "fun!" As your infant grows older, Occupational Therapy will focus upon helping your child improve his or her fine motor skills, such as cutting, coloring, pincer-grasp, writing, hand-eye coordination, etc. Playing with fun toys and games, such as Easy-Grip Pegs and Pegboards, finger paint brushes, lacing beads and shapes, Playdough, Theraputty, coloring crayons, wooden puzzle pieces with grips, Light Bright (for the older child), picking up cotton balls or pom-poms with tongs or tweezers-type pinchers, etc. will help improve your child's fine motor skills.

The body's sensory integration system involves the brain's processing and organizing all of the sensory input it receives from the body. The brain processes the input received from our sense of sight; sound; touch; taste; the vestibular system which involves body awareness, movement, a sense of balance, and the pull of gravity upon the body; and the proprioceptive system which informs our brain about our body in motion from the input received from our joints and muscles.

Sensory processing disorder (SPD), also known as sensory integration disorder, is a condition that exists when the brain doesn't process input from one or more of the senses. Because of their low muscle tone, almost all children with PWS don't efficiently process the information their body's senses receive and as a result are too sensitive (hypersensitive) or less sensitive (hyposensitive) than they ought to be to move

effectively and safely in their environment. Many children are hypersensitive to certain textures on parts of their skin which may lead to their not wanting to wear gloves, tags in shirts, have difficulty with their hair being washed or cut, have aversions to certain food textures, etc. If a child's proprioceptive system doesn't work properly, the child will have difficulty negotiating or moving his body in space, for example he may have difficulty ducking one's head under a table to retrieve an object, be more "accident prone, and have difficulty grading the force to hold a crayon, pencil, or pen in order to write. Difficulties with the vestibular system will cause the child to be either hypersensitive or hyposensitive to the body's action upon or involvement with the environment. They may have difficulty crawling, walking, climbing, maneuvering objects with one's fingers or hands; have difficulty standing or walking on unstable or uneven surfaces, avoid to an extreme or seek out to an extreme certain activities such as swings, twirling around, or "wrestling-type" activities.

Sensory Integration Therapy should begin as early in life as possible to help your child better integrate his senses so that he may operate more safely and efficiently in his environment. Many Occupational and Physical Therapists are knowledgeable about Sensory Integration Therapy and incorporate integrative techniques into their therapy. It will be important for you to make certain your therapists have an understanding of and experience with sensory integration therapy.

Your Regional Center should provide you with a referral to a qualified Occupational Therapist who may even come to your home to provide services while your child is between the ages of birth to three years. As the parent and caregiver, you should participate in the therapy sessions so that you may continue the therapeutic interventions between therapy sessions.

Resources

Therapeutic Interventions for the Child with Prader-Willi Syndrome, Janice Agarwal, P.T. Published by PWSA |USA

The Out-of-Sync Child: Recognizing and Coping with Sensory Integration Dysfunction, Carol Stock Kranowitz, M.A.

Overview of Physical Therapy Services

Physical therapy should begin as early as possible, alongside occupational therapy. Physical therapy helps strengthen large muscle groups important for improving posture and gross motor skills. Children with PWS often have difficulty with motor planning, or organizing their bodies to perform fine and gross motor activities, so therapy should be designed to help the child learn how to move his or her body to change positions.

As with occupational therapy, it is important for parents and caregivers to observe your child's physical therapy session so that you can incorporate at home the interventions the therapist uses with your child. There are many therapeutic interventions you can do outside of therapy that won't feel "therapeutic" such as "tummy time," placing your child on his or her tummy with his or her toys to help strengthen the arms, neck and shoulder muscles; reaching up high for toys; rolling; swinging; and later tricycle riding.

Growth hormone therapy will help improve your child's muscle strength and abilities. Even with growth hormone therapy, however, lighter weight toys may be easier and more enjoyable for your young infant to play with.

Due to low muscle tone, some children with PWS often have excessive foot pronation where the feet "roll" inward. Corrective devices, such as dynamic ankle foot orthotics (DAFOs), or Sure Step orthotics, can assist in earlier, more stable walking.

Some children, even those treated with growth hormone therapy, have more difficulty obtaining independent walking skills. Some parents have found front rolling walkers helpful to provide additional support. Remember though, all children with PWS do walk, some just later than others. It is critical not to rush your baby to walk because crawling skills are extremely important to improve muscle strength and coordination.

Likely due in large part to low muscle tone, scoliosis and kyphosis curvatures of the spine are more common in persons with PWS than the general population. Back strengthening exercises should be incorporated into the physical therapy regimen with continuous monitoring for any developing curvatures (See *Orthopedic Issues Specific to PWS* inside this Handbook).

Just as described in the *Overview of Occupational Therapy Services* section of this Handbook, the body's sensory integration system involves the brain's processing and organizing all of the sensory input it receives from the body. The brain processes the input received from our sense of sight; sound; touch; taste; the vestibular system which involves body awareness, movement, a sense of balance, and the pull of gravity upon the body; and the proprioceptive system which informs our brain about our body in motion from the input received from our joints and muscles. Sensory processing disorder (SPD), also known as sensory integration disorder, is a condition that exists when the brain doesn't process input from one or more of the senses. Because of their low muscle tone, almost all children with PWS don't efficiently process the information their body's senses receive and as a result are too sensitive (hypersensitive) or less sensitive (hyposensitive) than they ought to be to have more difficulty navigating their environment.

If a child's proprioceptive system doesn't work properly, the child will have difficulty negotiating or moving his body in space, for example he may have difficulty ducking one's head under a table to retrieve an object, be more "accident prone, and have difficulty grading the force to hold a crayon, pencil, or pen in

order to write. Difficulties with the vestibular system will cause the child to be either hypersensitive or hyposensitive to the body's action upon or involvement with the environment. They may have difficulty crawling, walking, climbing, maneuvering objects with one's fingers or hands; have difficulty standing or walking on unstable or uneven surfaces, avoid to an extreme or seek out to an extreme certain activities such as swings, twirling around, or "wrestling-type" activities.

Physical Therapy should incorporate Sensory Integration Therapy and should begin as early in life as possible to help your child better integrate his senses. Many Physical Therapists are knowledgeable about Sensory Integration Therapy and incorporate integrative techniques into their therapy including swings, hammocks. It will be important for you to make certain your therapists have an understanding of and experience with sensory integration therapy.

Throughout childhood, physical therapy will continue to be important to assist with strength, endurance, coordination, postural stability, and motor planning development.

Resources

Therapeutic Interventions for the Child with Prader-Willi Syndrome, Janice Agarwal, P.T. Published by PWSA (USA)

The Out-of-Sync Child: Recognizing and Coping with Sensory Integration Dysfunction, Carol Stock Kranowitz, M.A.

The Out-of-Sync Child Has Fun, Carol Stock Kranowitz, M.A.

Overview of Speech Therapy Services

Most children with Prader-Willi syndrome have an underlying speech disorder, such as developmental speech delay and/or childhood apraxia of speech. Therefore, oral motor/speech therapy should begin in the first year of life, well before the child is actually able to speak. Oral motor “exercises” disguised as fun games enhance control of the muscles of the mouth and tongue, teach fundamental motor planning skills, and will help with articulation and pronunciation later on.

Some speech therapists specialize in feeding issues and are available to help your baby work on sucking, swallowing, and later chewing skills. The supplement Coenzyme Q-10 has been found to improve motor planning and coordination (See the *Supplements* section in this Handbook).

Speech and language therapy should continue throughout early childhood in order to improve verbal language development. Many therapists recommend using picture communications systems and/or sign language to augment and enhance verbal language development. These techniques can help your child communicate before he or she has the ability to form words or sentences. Using sign language does not delay speech; in fact, its use helps motivate verbal communication. Kids generally drop signs as verbal skills increase.

Songs, rhymes and finger plays are great activities to help your child develop his or her ability to coordinate oral movements, sequence sounds and learn new vocabulary words. The rhythm and repetitive nature of the songs makes it fun and easy. There is a wonderful compilation of songs on a CD titled, *Time to Sing!* produced by a speech-language pathologist especially for kids who have speech problems. The CD features traditional songs that are played more slowly making it easier for kids sing along, may be purchased through www.apraxia-kids.org.

As your child grows older, you and the speech therapist should be watchful of the characteristics associated with childhood apraxia of speech, often called speech dyspraxia. Children with PWS often have difficulties with motor planning in general, and speech dyspraxia is a problem of motor planning in the speech areas of the brain. A diagnosis of speech dyspraxia requires speech and language therapy interventions specific to this speech disorder, most often using the PROMPT method.

Some children experience intermittent dysfluencies of speech, or stuttering. Word finding or word retrieval problems may be associated with dyspraxia and may exacerbate stuttering for a period of time. Speech and language therapy interventions specific to the treatment of dysfluency will help improve “smooth talking.”

Speech therapists often use food items to motivate and reward. While it may be therapeutically appropriate to use food items on occasion, such as placing a dab of peanut butter around the outside corners of the mouth to aid with tongue movement and coordination, etc., it is generally advisable not to use *any* food items during *any* therapy. Most children with PWS are extraordinarily motivated by genuine, enthusiastic praise and tangible items such as stickers.

Individuals with PWS often have dry mouth symptoms that include thick, sticky saliva because the pituitary doesn’t release sufficient amounts of saliva. A dry mouth can interfere with the smooth movement of the tongue, cheeks and lips, interfere with swallowing, and interfere with the production of sounds and clear speech. Products designed to relieve or reduce the symptoms of dry mouth are very helpful. Many people find Biotène oral hygiene products helpful, such as Biotène toothpaste (for young children, a pea size

amount twice daily should be fine). A few parents have needed the assistance of a prescription medication to reduce dry mouth symptoms.

Appropriate speech and language therapy services will help your child communicate fully and clearly and will facilitate his or her social relationships.

Resources

Apraxia Website www.apraxia-kids.org

The PROMPT Institute www.promptinstitute.com

Don't Stop Speech Therapy, Lisa Graziano, M.A. Available from PWCF and PWSA | USA

Speech and Language and PWS. Available from PWCF and PWSA | USA

Brochure: Speech and Language and PWS. Published by PWCF and PWSA | USA

CD: Time to Sing! produced by a speech-language pathologist especially for kids who have speech problems. The CD may be purchased through the www.apraxia-kids.org website.

Dry Mouth Oral Hygiene Products. Can be purchased in toothpaste form, using a pea-size amount for toddlers.