



# **SERVING THE NEEDS OF INDIVIDUALS WITH PRADER-WILLI SYNDROME**

**Report to the  
Texas Legislature**

**As Required by:  
Section 3.03, S.B. 7,  
83<sup>rd</sup> Legislature, Regular Session, 2013**

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*The*

**Health and Human Services Commission**

*in coordination with the*

**Texas Department of Aging and Disability Services**

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## **Introduction and Charge**

S.B. 7, Section 3.03, 83<sup>rd</sup> Legislature, Regular Session, 2013, directs the Health and Human Services Commission (HHSC) and the Department of Aging and Disability Services (DADS) to “conduct a study to identify crisis intervention programs currently available to evaluate the need for appropriate housing for, and develop strategies for serving the needs of persons in this state with Prader-Willi syndrome”; and to submit a report no later than December 1, 2014, to the governor, the lieutenant governor, the speaker of the house of representatives and the chairs of the Senate Health and Human Services and House Human Services committees. HHSC delegated the preparation of this report to DADS. DADS developed this report based on information from providers, families, advocates and experts in other states, as well as a review of scholarly literature and state Medicaid data.

## **Overview of Prader-Willi Syndrome**

Prader-Willi syndrome (PWS) is a rare and incurable genetic condition.<sup>1</sup> In infancy, PWS is characterized by weak muscle tone, feeding difficulties, poor growth and developmental delays.<sup>2</sup> Beginning in childhood, individuals with PWS develop an insatiable appetite.<sup>3</sup> With the appropriate life-long supports, they are able to live productive, satisfying lives. Without these supports, individuals with PWS often develop severe medical problems (chronic overeating, obesity, type 2 diabetes, heart disease) and may die at an early age.<sup>4</sup> Individuals with PWS typically have mild to moderate intellectual impairment and learning disabilities, muscle weakness, slow growth and behavioral challenges (e.g., intense behavioral outbursts, skin picking, and anxiety).<sup>5</sup> The exact number of individuals with PWS in Texas is unknown. PWS is estimated to affect about one of every 15,000 individuals, or up to 1,750 individuals in Texas.<sup>6</sup> About 113 individuals in Texas have PWS and receive some type of long-term support services through the Department of Aging and Disability Services. Acquiring needed services and housing can prove challenging for individuals with PWS and their families.

Early diagnosis of PWS is important. In the past, many infants with PWS did not survive infancy. Identification of PWS symptoms in infants provides the opportunity for genetic testing to confirm the diagnosis and to begin treatment. Families may begin receiving education sooner as well as taking necessary safety measures, which will improve their child’s growth and quality of life and may reduce medical costs related to chronic overeating, obesity, type 2 diabetes and heart disease in adulthood.

One of the central needs of children with PWS is control of food intake. A highly structured approach to food, including serving meals at the same time each day and not making food available at any other time, appears to reduce anxiety and complex behavioral challenges.

Maintaining a strict food intake regimen for a child with PWS can prove to be a challenge for some public school systems, as school personnel may not understand PWS or the needs of a child with the condition. For example, school personnel may mistake PWS for an eating disorder, may not adhere to a child’s specialized PWS diet and may use food as an incentive for

certain behaviors, disrupting the child's diet and increasing the child's anxiety about the availability of food.

The needs of adults with PWS are similar to those of older children and include food security, consistent routines, appropriate diet and exercise, behavioral health care and specialty medical care.

Adults with PWS also need constant support and supervision. They should not be left alone and should have a support person or family member accompany them in the community. In the absence of such precautions, individuals with PWS may overeat, steal food or run away to obtain it. Because the potential availability of food causes anxiety, individuals with PWS may behave in inappropriate ways in stores, theaters, or other community locations.

### **Methodology**

Scholarly research concerning PWS is limited due to the rarity of this condition. Frequently the best source of information comes from individuals with PWS, their families and service providers who work with individuals with PWS.

Even when data is available, publication of such information may be prohibited due to privacy protections. For example, providing data on only a few individuals can reveal personal information about individuals in that group. In order to protect individual privacy, specific data about groups smaller than 10 individuals is not included in this report.

In preparing the report, DADS staff conducted the following activities:

- Interviews and discussions with parents of individuals with PWS; the staff of organizations in Texas that provide services to individuals with PWS; Texas Prader-Willi Association (TXPWA) members; and other researchers
- Interviews with staff of organizations in other states that provide services to individuals with PWS, including Massachusetts, Wisconsin, Missouri, Florida, Illinois, Michigan and Utah
- Reviews of scholarly and professional literature concerning PWS
- Analysis of data concerning Texas programs and services available to individuals with PWS
- Collaboration with stakeholders, including the SB 7 IDD System Redesign Advisory Committee, the DADS IDD Systems Improvement workgroup and DADS IDD providers

A 19-question survey was posted on the DADS website as an alert to the following provider webpages: HCS, TxHmL, DBMD, CLASS, ICF/IID, CBA, STAR+PLUS, nursing facilities and MDCP. The alert was time-limited and posted from February 26, 2014, through March 10, 2014. There were 58 valid responses. The survey was conducted through Survey Monkey.

### **Available Services**

Most individuals with PWS live with their families and require specialized care and supervision throughout their lives. Families may need a range of services such as various therapies, medical assistance, respite, special education services through the public school system and information on safe, effective ways to care for their children and provide a safe environment.

### **Crisis Intervention Programs**

Few crisis intervention programs and services are available exclusively to individuals with PWS. Many available services are designed for individuals with varying disabilities. Seventeen local intellectual disability authorities are currently developing behavioral health intervention projects that include crisis intervention teams and crisis respite funded through Medicaid.<sup>7</sup>

The HealthBridge Children's Hospital in Houston administers an inpatient interdisciplinary treatment program for children and adolescents with PWS. The program incorporates physician-led plans, which include registered dietician-developed meal plans, set daily schedules, individualized physical training programs and mental health support for patients and families.<sup>8</sup> The Children's Institute in Pittsburg, PA, is a similar program that provides inpatient services to children with PWS.<sup>9</sup> Both the Pittsburgh and HealthBridge programs address the dietary and health issues of individuals with PWS, usually involving a restricted diet and an exercise program, along with behavioral treatment and family training. The goal is to return the individual to his or her home with better health, better diet and exercise habits, and with a reduced incidence of behavioral problems. Family training and participation are essential, so that the family understands how best to manage behavior.

Families have reported difficulty obtaining Medicaid approval to use these services. They recommended that Medicaid managed care organizations include specialty hospitals in their provider networks.

Apart from these specialty resources, individuals with PWS are largely dependent on local physicians, both primary care and specialists. It can be difficult finding providers who are both willing to accept Medicaid payment and are knowledgeable about PWS. These services may be impacted by the carve-in of intellectual and developmental disability (IDD) acute care services for persons enrolled in Medicaid-funded IDD services.

Two hospitals in Texas offer specialty outpatient health care for children with PWS. Texas Children's Hospital in Houston and Children's Hospital in San Antonio have begun to offer periodic multi-disciplinary clinics for children with PWS. These clinics allow a child to be seen by specialists in a variety of areas (diet, gastroenterology, physical therapy, endocrinology and others) in a single trip, and all of these specialists are knowledgeable of the special issues associated with PWS.

## **Other Programs and Services**

Additional programs and services available to individuals with PWS include:

- Early Childhood Intervention - a statewide program for families with children birth to three years old with disabilities and developmental delays. The program is both state and federally funded, using a sliding fee scale for cost sharing when applicable
- Special education services available through the public school system
- People with Smiles Camp - a private pay, three- to five-day summer camp for individuals with PWS sponsored by the Texas Prader-Willi Association.
- Intermediate care facilities for individuals with an intellectual disability or related condition (ICFs/IID) operated in the community. An ICF/IID is licensed by DADS Regulatory Services to provide residential and direct care services to individuals with an intellectual disability or related condition.
- State Supported Living Centers (SSLCs) – large residential facilities certified as ICFs/IID operated by the state, providing campus-based direct services and supports to individuals with an intellectual or developmental disability who are medically fragile or have behavioral health needs who meet eligibility criteria
- Medicaid waiver programs that provide long-term services and supports (LTSS):
  - Home and Community-based Services (HCS),
  - Texas Home Living (TxHmL),
  - Community Living Assistance and Supports Services (CLASS),
  - Medically Dependent Children Program (MDCP),
  - Deaf-Blind Multiple Disabilities (DBMD)
  - STAR+PLUS (formerly CBA)
- Non-Medicaid funded day habilitation, behavioral support and specialized therapies available through the local IDD authorities
- Medicaid - Many individuals with PWS qualify for Medicaid
- Take Time Texas: Texas Respite Coordination Center – a DADS website that provides a database of respite providers, educational materials, information, best practices, training tools and other resources for caregivers and providers
- The National Prader-Willi Syndrome Association (<http://www.pwsausa.org/>) and the Texas Prader-Willi Association (<http://txpwa.org/>) websites - provide peer support, medical advice, telephone counseling and consultation, and resources that can help prevent, manage and respond to crises

About 113 individuals who have PWS receive Medicaid LTSS through DADS and the STAR+PLUS waiver program through HHSC. In order to protect privacy, specific data about groups smaller than 10 individuals is not included in this report.

<b>Type of Service</b>	<b># of individuals with PWS served between January 1, 2013, and June 30, 2014</b>
ICF/IID operated in the community	12
SSLC	<10
HCS	37
TxHmL	<10
CLASS	17
MDCP	20
STAR+PLUS (formerly CBA)	13
Nursing Facility	<10
Local Authorities	44
Community Care	<10

The above numbers are not unduplicated, meaning that an individual may have received services in multiple programs during this period.

### **Available Services Challenges**

The most frequently sought-after services by families of individuals with PWS include respite, information and training, assistance navigating state and local support services and peer support.

Families and residential service providers must carefully balance the importance of community integration with the safety and health needs of individuals with PWS. Common behaviors among individuals with PWS may bring them into conflict with the criminal justice system. These behaviors may include outbursts (e.g., kicking, biting), stealing food and running away.

If police and other first responders are unaware of the particular needs and challenges of individuals with PWS, the crisis may escalate. For example, the individual with PWS may be injured, or his or her behavioral outbursts may become more severe.

Access to specialized medical and residential treatment can be problematic if Medicaid or private insurers are unwilling to authorize the service. Obtaining knowledgeable specialized medical care for individuals with PWS in Texas is a challenge. Eighty-nine percent (89%) of the survey respondents reported that obtaining knowledgeable specialized medical care was difficult or very difficult. Greater flexibility of payment would make such service more accessible.

Medicaid waiver programs limit the number of enrollees and may have a waiting or interest list for persons awaiting services. Individuals must register on an interest list and could wait for years before services become available.

### **Housing Needs**

When individuals with PWS are ready to leave their family home or experience placement disruption (e.g., if the caregiver becomes ill, dies or is otherwise unable to care for the individual with PWS), they need an environment with ongoing supports that is appropriate to their unique and complex needs. In the absence of such supports, individuals with PWS will usually show increased maladaptive behaviors, become morbidly obese, develop complications from that obesity and die at an early age.

Most individuals with PWS who receive Medicaid funded residential services live in an ICF/IID operated in the community or in a small group home available in the HCS waiver program.

Many experts and families prefer residences that serve only individuals with PWS. Currently, only two Texas providers offer PWS-exclusive housing, and establishing more PWS-exclusive programs is difficult due to regulatory challenges related to protecting and promoting individual rights, as well as the cost associated with the increased staffing needed to serve this population.

In addition, residences that include individuals with PWS must practice strict food security such as locking refrigerators and cabinets. This could result in rights restrictions for those individuals living in the home who do not have PWS.

### **Available Residential Services**

Four Medicaid programs in Texas provide residential services for individuals with PWS:

- An ICF/IID operated in the community is a residential facility serving four or more individuals. Individuals in an ICF/IID receive services that may include the food security and ongoing support needed by individuals with PWS. Most ICFs/IID operated in the community serve six persons and few exclusively serve individuals with PWS.
- For persons enrolled in the HCS waiver program and receiving residential services, these services are provided in three- or four-bed group homes. Services provided in these settings may also include food security and other services related to PWS based on the specific needs of the person.
- State supported living centers (SSLCs) provide the same services as community-based ICFs/IID, with an emphasis on serving individuals with more complex medical and/or behavioral needs. These facilities house larger numbers of individuals with various diagnoses.
- Nursing facilities provide residential services to individuals whose needs are primarily medical.

<b>Type</b>	<b># of individuals with PWS served between January 1, 2013 and June 30, 2014</b>
ICF/IID operated in the community	12
HCS	37
SSLC	<10
Nursing facility	<10
Own home/ family home	60

The above numbers are not unduplicated, meaning that an individual may have moved from one residency type into another during this period.

### **Housing Challenges**

Individuals with PWS have housing needs that are unusual when compared to the general population of persons with IDD. Few facilities or homes in Texas are equipped to meet those needs, and general-purpose facilities and homes must adapt when an individual with PWS joins the household. The need for rigid scheduling and food restrictions can make it very difficult to include individuals with PWS with the general IDD population.

Safeguards need to be put in place to balance restrictions to ensure the health and safety of individuals with PWS while also protecting the rights and service quality for those individuals and other individuals who live in the same residential setting.

As previously mentioned under Housing Needs, many experts and families prefer residences that serve only individuals with PWS. Only two providers in Texas currently offer residences (in either HCS group homes or ICFs/IID) that exclusively serve individuals with PWS, and establishing more PWS exclusive programs is difficult due to regulatory challenges related to protecting individual rights and increased costs related to staffing needs. In all other cases, individuals with PWS live in residences with individuals with other types of IDD.

Access to HCS group homes and to ICFs/IID in the community is limited. Funding limits on the HCS program limit the number of enrollees, and individuals must register on an interest list and could wait years before services become available. Availability of services that can meet the unique needs of persons with PWS in an ICF/IID in the community varies, depending on the specific home.

Not all individuals with PWS will meet eligibility criteria to receive services from the various residential options available and may have to rely only on available natural supports, such as family, friends and community members with whom the individual has personal associations and relationships.

Providers also believe existing assessment instruments do not adequately identify the substantial needs of people with PWS, resulting in inadequate reimbursement.

### **Strategies for Serving the Needs of Individuals in Texas with Prader-Willi Syndrome**

As noted previously, parents of individuals with PWS and organizations that provide services to individuals with PWS face various challenges in ensuring the health and safety and meeting the unique needs of these individuals. Below are some strategies for improving services available for individuals with PWS.

1. Update and expand the Take Time Texas website to include PWS-specific information and resources, including respite options, available services for families of individuals with PWS, training opportunities, camps and caregiver and peer supports.
2. Provide education and training related to PWS to organizations that provide services and supports to individuals with PWS. Regional trainings could be hosted by the local IDD authority in conjunction with the local mental health authority and include participants from the provider network, first responders and other interested parties.
3. DADS-sponsored behavioral health trainings should include alternative behavioral techniques specifically related to individuals with PWS.
4. Continue the current crisis intervention pilots through the local authorities, such as the START program provided through Austin Travis County Integral Care (Travis County local IDD authority), and include specific strategies for supporting individuals with PWS.
5. Explore the use of existing health and human services agency resources to identify medical, dental and dietary services that can be expanded to individuals with PWS who are not currently receiving services through an enterprise agency.
6. Work with managed care organizations to increase awareness and knowledge of the specialized services that individuals with PWS require.
7. Develop mechanisms to fund residential behavioral intervention programs unique to individuals with PWS. Programs should be available in more than one area of the state and should serve individuals with PWS as well as individuals with other IDD/behavioral issues.
8. Revise the comprehensive IDD assessment instrument to better identify the unique needs of persons with PWS. Using the current assessment process, individuals with higher IQs and who are more independent with daily living skills (such as many individuals with PWS) are typically assigned lower level of need scores because they can take care of some of their own needs. For individuals with PWS, higher functioning often means greater ability to seek food and a greater need for staff support to address resulting behavior incidents. DADS and HHSC are reviewing the IDD comprehensive assessment instrument and the associated resource allocation process. The outcome of that review may result in changes to the existing assessment and resource allocation process.

**End Notes**

<sup>1</sup>See “What is Prader-Willi syndrome?” Available online at <http://ghr.nlm.nih.gov/condition/prader-willi-syndrome>, Accessed July 26, 2014

<sup>2</sup> *Ibid*

<sup>3</sup> *Ibid*

<sup>4</sup> *Ibid*

<sup>5</sup> *Ibid*

<sup>6</sup> <http://www.nichd.nih.gov/health/topics/prader-willi/conditioninfo/Pages/at-risk.aspx>.

<sup>7</sup> These projects are funded by the Delivery System Reform Incentive Payment (DSRIP) program administered by HHSC through the Medicaid 1115 Healthcare Transformation demonstration waiver. Additional information about the DSRIP program is available online at <http://www.hhsc.state.tx.us/1115-waiver.shtml>.

<sup>8</sup> Additional information about this program is available online at <http://www.healthbridgehouston.com/prader-willi-syndrome-program/>.

<sup>9</sup> Additional information about the Center for Prader Willi Syndrome at the Children’s Institute at Pittsburgh is available online at <http://www.amazingkids.org/Medical-Services/the-center-for-prader-willi-syndrome>.