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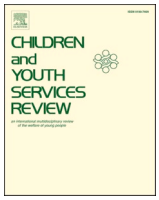

Discussion

Prader-Willi Syndrome: A primer for school psychologists

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ABSTRACT

Prader-Willi syndrome (PWS) is a rare neurogenetic syndrome that results in significant challenges in the school setting, often requiring a myriad of academic, social-emotional, and behavioral supports to maintain health and safety and academic success. This paper provides a primer for school psychologists, introducing the physiological, educational, and behavioral differences experienced by children with PWS. As the symptoms of PWS are widespread, the impacts of common physical and neurological differences on specific learning abilities, adaptive skills, and overall functioning in the classroom setting are discussed in detail. While PWS is a spectrum, and each child will experience symptoms and challenges differently, this article aims to provide a foundation for the development and facilitation of support services to assist educators in not only maintaining the health and safety of their students with PWS, but also in capitalizing on their strengths, and building a plan to help them succeed.

1. Introduction

Prader-Willi syndrome (PWS) is a rare, genetic, neurodevelopmental syndrome (Driscoll et al., 2017; Schwartz, et al., 2021). PWS has three genetic subtypes, including paternal deletion (DEL) which impacts approximately 65 % of individuals, maternal uniparental disomy (UPD) which impacts around 30 % of individuals, and imprinting center defect, which impacts between 3–5 % of individuals (Driscoll et al., 2017; Schwartz, et al., 2021). The mechanisms of each subtype leads to a loss of paternally expressed imprinted genes on chromosome 15q11.2– q13 (Driscoll et al., 2017). While population estimates range, a recent study indicated that PWS impacts around 1 in 37,000 people residing in the US (McCandless et al., 2020), with older studies indicating that approximately 1 in 25,000 live births are impacted world wide (e.g. Vogels et al., 2004; Whittington et al., 2001).

Often, the most well-known feature of PWS is the difference in appetitive and eating behavior that begins at an average age of eight years old and which often results in the development of hyperphagia, or an insatiable appetite (Miller et al., 2011). It is important to note, however, that PWS impacts multiple bodily systems, resulting in a wide variety of clinical features including growth hormone deficiency, hypotonia (low muscle tone), decreased gastrointestinal motility, disordered sleep, and reduced sensitivity to pain (Schwartz, et al., 2021). As a result, people with PWS experience a myriad of challenges unrelated to food that may impact their ability to succeed academically and socially.

People with PWS often have brain differences that result in deficits in executive functioning and other skills that are important in a classroom setting (Jauregi et al., 2007; Schwartz, et al., 2021). Additionally, PWS is associated with a set of very specific behaviors of concern, such as emotional outbursts, rigid and repetitive behaviors, and self-injury that most often presents as skin picking (Schwartz, et al., 2021). Some of these challenges (e.g., hyperphagia) can be life threatening if the appropriate supports are not in place, and many of the behaviors can result in lifelong challenges that severely impact the quality of life of a person with PWS and their family if not addressed at a young age (Schwartz, et al., 2021).

Because PWS is associated with such a diverse set of challenges, children with this rare syndrome often require several academic, social-emotional, and behavioral supports in order to maintain health and safety in the school setting and to succeed academically. Because school psychologists will often be the ones tasked with facilitating or developing the supports for children with PWS, it is imperative that they have the information and resources needed to do so successfully. To this end, this article provides an overview of the physical and behavioral features of PWS to provide a primer for school psychologists who may be unfamiliar with this rare syndrome.

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2. Physical and behavioral features of PWS

2.1. Physiological features

Hyperphagia and food-related behaviors are of the utmost concern when supporting a person with PWS (Schwartz et al., 2021). Even before hyperphagia develops, children with PWS require a very specific diet that includes portion control, healthy meal planning similar to a Mediterranean diet, and avoiding sweetened and artificially sweetened food and drinks (Miller et al., 2013; Miller & Tan, 2020). There are several nutritional phases of PWS that may impact the health and safety of children and teens in the classroom. Children three to five years of age (mean onset of 4.5 years) begin to demonstrate an increased interest in food and may gain weight without a change in caloric intake (Miller et al., 2011). During this phase children may become very anxious and rigid around food and their eating schedule.

Hyperphagia develops between the ages of five and 13 years of age, with a typical onset of around eight years of age (Miller et al., 2011). Children who are experiencing hyperphagia feel an insatiable hunger, and often fail to feel full even after consuming large amounts of food (Miller et al., 2011). This often results in aggressive food seeking and food stealing behavior (Schwartz et al., 2021). People with PWS will consume food from unsafe locations including the floor and trash cans, and may overconsume food to the point that they risk stomach rupture and death (Schwartz, et al., 2021).

Food that is visually available in the classroom or broader school setting may predispose perseveration, behaviors of concern, and may result in the inability to concentrate on academic tasks (Prader-Willi Syndrome Association USA, 2015; Schwartz et al., 2021). Some children may also become distracted by discussions of food and food locations, and the inclusion of food in educational materials. It is, therefore, of the utmost importance to the health, safety, and learning ability of a person with PWS to maintain a food secure environment in the classroom and surrounding school environment (Schwartz et al., 2021). The danger of unsecure food environments is further complicated by the fact that as many as 60–80 % of individuals with PWS have delayed gastric emptying, and between 80–90 % experience decreased vomiting (Driscoll et al., 2017). Unmonitored consumption of large quantities of food puts a child with PWS at risk of gastric necrosis, intestinal tract perforation, and death (Driscoll et al., 2017).

As PWS impacts multiple bodily systems, the physical features of PWS are widespread. Most people with PWS have low muscle tone, which results in delays in fine and gross motor skills (Driscoll et al., 2017). Having deficits in fine motor abilities means that children with PWS often struggle with many of the fine motor tasks required in the classroom, including writing, cutting, tying, and other similar skills (Hsu et al., 2018). While skills often improve with time and therapeutic intervention, older teens and young adults may still find tasks such as writing difficult and aversive (Hsu et al., 2018). Gross motor deficits complicate any activity that requires a large degree of motor planning and coordination, including jumping, running, and balance (Reus et al., 2011). This can impact the ability to participate in physical education activities and may also impact the degree to which children can participate in play that requires physical coordination and strength, such as playing sports or tag, and safely using a jungle gym. Some children may also struggle to sit at a desk or table in an upright position for an extended period of time due to a lack of core strength. Individuals with PWS often fatigue quickly during both fine and gross motor tasks, which can lead to frustration, and refusals to participate in social and learning activities.

Many people with PWS experience daytime sleepiness (Maas et al., 2010). At times this manifests as falling asleep frequently throughout the day. However, indicators of daytime sleepiness can also include increases in resistance to completing school related activities, increases in irritability and behaviors of concern, and decreases in attention and emotional regulation ability (Maas et al., 2010). While general daytime sleepiness and fatigue are common, some children may also have a diagnosis of narcolepsy. Narcolepsy is a neurological sleep disorder that occurs at a higher prevalence in PWS than in the general population (Cataldi et al., 2021). Narcolepsy is characterized by excessive daytime sleepiness, sudden and uncontrollable episodes of falling asleep, and other disturbances of sleep-wake cycles (Cataldi et al., 2021). It is important to understand that children with PWS do not have control over their degree of sleepiness, and support and accommodations may be required in the school setting either through a 504 Plan or Individual Education Plan (IEP). Educators need to recognize these limitations and may need to provide rest breaks well beyond the typical age and/or grade level when naps are considered appropriate. If necessary, these breaks should be scheduled during non-instructional times of the day.

A final physiological concern associated with PWS is the reduction in the sensation of pain (Schwartz et al., 2021). Because children with PWS may experience little pain as a result of injury, monitoring for injury is imperative (Driscoll et al., 2017). Children who become injured on the playground, during sports, or physical education activities may not report the presence of injury to an adult, even in cases of serious injury and broken bones (Driscoll et al., 2017). Close supervision during physical activities may therefore be required to ensure health and safety, and particular care needs to be taken during any type of physical crisis intervention. Any observed falls or physical injury, and any evidence of a fall or injury, such as bruising or swelling, should be brought to the attention of relevant caregivers and medical staff.

2.2. Differences in brain structure

Recent brain imaging studies indicate that individuals with PWS have differences in brain structure that may impact their learning and behavior (Brown et al., 2022, Wu et al., 2022, Yamada, 2023). For children with PWS, there are several areas of their brain that are notably smaller in comparison to the general population (Brown et al., 2022, Wu et al., 2022, Yamada et al., 2022, Yamada, 2023). These areas include the thalamus, pallidum, hippocampus, amygdala, and brainstem structures (Wu et al., 2022; Yamada et al., 2022, Yamada, 2023). Recent in-vivo neuroimaging has also shown decreased hypothalamic volume, as well as alterations in connectivity between the hypothalamus and other brain regions (Brown et al., 2022). Many of these brain regions are responsible for cognitive functioning, as well as bodily systems known to be problematic in PWS, including hunger and thirst, sleep, and the regulation of sensory input (Brown et al., 2022, Wu et al., 2022, Yamada, 2023). As a result, many children with PWS struggle with learning and cognitive delays, difficulties with regulating their emotions, and deficits in executive functioning (Schwartz, et al., 2021). These cognitive differences in conjunction with the physical challenges described above mean that even with their best effort, many school-related tasks can be both challenging and aversive for children with PWS and may put children at risk for exhibiting behaviors of concern, and for falling behind their neurotypical peers.

2.2.1. Impacts of physical features on learning and behavior

Intellectual impairment or cognitive deficits are common characteristics in individuals with PWS (Cataletto et al., 2011; Scott et al., 1999; Whittington et al., 2004; Whittington & Holland, 2017). However, there is also evidence of a scattering of cognitive abilities (splinter skills) and an uneven cognitive profile with marked strengths and weaknesses (Cataletto et al., 2011; Curfs & Fryns, 1992; Donaldson et al., 1994; Dykens et al., 1992; Whittington & Holland, 2017). Whittington et al. (2004) identified a "marked downward shift in global ability," with IQ distribution being relatively normal with an average IQ in the low 60's (p. 180). Others have estimated the average IQ of individuals with PWS is around 70, with about 5 % scoring in the "average" range of 85 and above (Gross-Tsur et al., 2001; Verdine et al., 2008). The discrepancies across studies may be attributed to the sampling methods, smaller sample sizes due to the rarity of PWS, and variations in testing measures.

2.2.2. Executive function

In addition to cognitive deficits and lower intellectual quotients, individuals with PWS typically have global deficits in executive functioning skills (Chevalère et al., 2015; Gross-Tsur et al., 2001; Jauregi et al., 2007; Whittington & Holland, 2017). Executive functioning skills include working memory, attentional control, cognitive flexibility, and inhibitory control. These skills are required daily to learn, work, and complete the simplest of tasks. In the classroom, these deficits can inhibit a child's ability to pay attention, organize and plan, initiate tasks, stay on task, and self-monitor while completing a task.

Severe deficits in working memory, as compared to typical peers, have been found in individuals with PWS (Cataletto et al., 2011; Chevalère et al., 2015). Working memory involves retaining information presented and utilizing it in order to follow steps to successfully complete a task (Chai et al., 2018). In addition, individuals with PWS score significantly lower on tests using immediate memory when requiring sequential processing (Dykens et al., 1992; Jauregi et al., 2007). Working memory is essential for students in the academic setting, as they are continuously expected to take in new information and follow teacher/adult directions. This is exacerbated by the fact that individuals with PWS have deficits in auditory processing (Curfs et al., 1991; Cataletto et al., 2011; Stauder et al., 2002) and "significant weakness in sequential processing" (Dykens et al., 1992, p. 1126), which includes understanding verbal instructions and putting information in a step-by-step manner. Due to these combined deficits, students will exhibit difficulty initiating tasks requiring the use of new information and/or verbal directions. This may appear as though they are intentionally not complying, when in reality, it is much more difficult for them to process the information presented, mentally organize the necessary sequential steps, and initiate the task.

There are also links between working memory performance and inattention in children (Lui & Tannock, 2007). Attentional skills appear weakened in regard to both global and selective attention in children with PWS (Jauregi et al., 2007). Attention-deficit/hyperactivity disorder (ADHD) is present in about 6 % of all children and is almost six times (35 %) more likely in children with PWS (Butler, 2017; Singh, 2022). Gross-Tsur et al. (2001) found that half of the children in the group studied had been diagnosed with ADHD by a treating physician. Wigren and Hansen (2005) further studied specific executive dysfunctions related to ADHD. They found that 64 % of children scored above average or higher on the ADHD Index, of which 12 % displayed clinically elevated scores and 26 % reached clinically significant scores (Wigren & Hansen, 2005). Based on experience, Singh (2022) has found that the inattentive subtype is more common in children with PWS. This may relate to the fact that people with PWS have low muscle tone and daytime sleepiness, which may reduce their overall level of activity (Singh, 2022).

Cognitive flexibility, or flexible thinking, is another executive function that refers to an individual's ability to adapt to changes in conditions by reviewing and revising their plan or way of thinking (problem-solving). Individuals with PWS exhibit rigid thinking and a preference for predictability, observed by a strong resistance to changes in their daily routines, scheduled activities, or beliefs about how things "should'' be done (Haig & Woodcock, 2017; Schwartz, et al., 2021; Woodcock et al., 2009). This may be linked to the fact that people with PWS struggle with task switching, the ability to move from one task to a different task in a timely manner, making change difficult (Chevalère et al., 2015; Whittington & Holland, 2017; Woodcock et al., 2009). Deficits in cognitive flexibility interfere with an individual's daily functioning. People with PWS can be characterized as "black and white" thinkers who are resistant to considering information that might conflict with their prior presumed knowledge. This can interfere with a child's ability to accept new strategies, skills, or information presented during instructional lessons. Although this is a known recognized deficit, it can manifest and appear as combative and argumentative behaviors.

Finally, inhibitory control is another lagging executive functioning skill in individuals with PWS. The presence of deficits in the ability to regulate emotions and subsequent reactions is a well-established feature of the syndrome (Schwartz et al., 2021). Children with PWS may become very upset at a seemingly small trigger (Rice et al., 2018; Schwartz et al., 2021). This often escalates very quickly into a heightened state of emotional distress, during which it is difficult or impossible to reason or problem-solve with the person (Rice et al., 2018; Schwartz et al., 2021). It often takes children with PWS much longer to calm down than their typical peers, and this can result in significant disruption to the classroom environment as well as the learning ability of the child (Schwartz et al., 2021).

2.2.3. Learning differences

Individuals with PWS often present with multiple severe learning disabilities and poor academic performance, greater than expected for their measured IQ (Dykens et al., 1992; Gross-Tsur et al., 2001; Semenza et al., 2008). Children with PWS typically require specialized instruction and related services through an Individualized Education Program/Plan, often referred to as an IEP (Butler et al., 2022). It is recommended that young children with PWS receive therapies and subsequent special education as soon as possible (Butler et al., 2022; Conners et al., 2000) as the gap in academic achievement increases throughout the child's educational career. In addition, the lagging executive functioning skills, prevalence of attention-deficit/hyperactivity disorder (ADHD), and deficits in emotional regulation can impede the child's overall ability to learn (Gross-Tsur et al., 2001; Jauregi et al., 2007).

Individuals with PWS often have an uneven cognitive profile with apparent strengths and weaknesses (Cataletto et al., 2011; Curfs et al., 1991; Curfs & Fryns, 1992; Donaldson et al., 1994; Dykens et al., 1992; Semenza et al., 2008; Whittington & Holland, 2017). As technology and genetic testing have advanced, researchers have also identified variability between the PWS subtypes (Deletion vs. UPD) as it relates to intelligence and academic achievement levels (Roof et al., 2000; Cataletto et al., 2011). Whittington et al. (2004) found a downward shift in global ability for both genetic subtypes, along with "subtle specific cognitive differences" between the two groups (p. 180). In general, individuals with PWS due to paternal deletion (DEL) have a higher Verbal IQ (VIQ), whereas those with PWS due to maternal uniparental disomy (UPD) tend to have a higher Performance IQ (PIQ) (Cataletto et al., 2011; Roof et al., 2000). It is important to note that it is unlikely for school psychologists or other staff to be privy to the specific genetic subtype of a student unless the parent/guardian chooses to disclose this medical information.

Children with PWS show relative strengths in academic tasks involving decoding, reading, vocabulary, and comprehension that exceed their math skills (Bertella et al., 2005; Donaldson et al., 1994; Dykens et al., 1992). Most school-age children (83.3 %) with PWS can read fluently (Lewis, 2006), and many become "skilled readers" (Butler et al., 2022, p. 281). Strengths in visual and simultaneous processing support the instructional approach of teaching sight words for reading over a phonics-based approach (Reilly, 2009). In contrast, mathematical skills have been found to be a significant weakness and the most impaired cognitive ability in individuals with PWS (Bertella et al., 2005; Dykens et al., 1992; Whittington & Holland, 2017). Bertella et al. (2005) observed specific deficits in 'syntactic' processes in number transcoding, multiplication, number facts retrieval, and calculation procedures (Bertella et al., 2005). These deficits in math skills should be taken into consideration when teachers prepare academic lessons. Specifically, math concepts should be taught in a concrete manner with manipulatives and visual supports and should be linked to real-life activities whenever possible (Reilly, 2009). Additional supports, including accommodations or modifications, will likely be required.

Another strength is that individuals with PWS have a superior skill ability in jigsaw puzzles and word searches (Donaldson et al., 1994; Dykens, 2002; Verdine et al., 2008). On visual-spatial tasks (Object Assembly, Triangles, VMI), individuals with PWS scored significantly

higher than peers who were age- and IQ-matched. Dykens (2002) found that persons with PWS outperformed both typically developing, chronological age (CA) matched controls, and CA- and IQ- matched controls with intellectual disabilities in puzzles. In fact, children with PWS scored on par with typical peers on word searches, and they "far *outperformed* them on jigsaw puzzles" (Dykens, 2002). The children with PWS placed more than twice as many pieces as their typically developing peers. The reasons for this significant strength in puzzles remain unclear, however, researchers have proposed that such skills are related to visuospatial ability (Verdine et al., 2008) or processes that involve visual and spatial awareness. This is supported by the fact that people with PWS have better simultaneous processing abilities over their sequential processing skills, suggesting strength in tasks requiring the integration of stimuli in a spatial mode (Dykens et al., 1992). However, some propose that superior puzzle skills may be due to a "practice effect" with the initial high interest in those with PWS arising from relative strengths in visuospatial activities (Whittington, p. 186).

In contrast to relative strengths in visuospatial abilities, individuals with PWS can exhibit deficits in auditory processing abilities (Curfs et al., 1991; Dykens et al., 1992; Stauder et al., 2002). Specifically, individuals with PWS typically score higher on visual motor discrimination skills than on auditory verbal processing (Curfs et al., 1991; Jauregi et al., 2007). This is further supported by a brain activity study that found that the auditory modality in individuals with PWS is more impaired than the visual modality (Stauder et al., 2002). When comparing verbal intelligence quotient (VIQ) and performance intelligence quotient (PIQ), results indicate overall strengths in performance skills (Curfs et al., 1991), especially in individuals with PWS by deletion (Cataletto et al., 2011; Roof et al., 2000). In fact, Strenilkov et al. (2020) recently investigated voice and nonvoice auditory processing in individuals with PWS. Results indicated an overall impairment with voice processing, but individuals with PWS due to UPD were more impaired than those with PWS by deletion on both voice and nonvoice perception. Researchers concluded that individuals with PWS "need to accumulate more information for decision-making [and] are slower at decision-making" (Strenilkov et al., 2020, p. 8-9).

Deficits in auditory processing are compounded by significant deficits in short-term memory and sequential processing (Bertella et al., 2005; Conners et al., 2000; Donaldson et al., 1994; Dykens et al., 1992; Jauregi et al., 2007). Individuals with PWS have severe impairments in identifying a sequence by observation only but become as efficient as typically developing children after a task including "doing" (Foti et al., 2015). In addition, individuals with PWS have improved abilities when tasks require the integration of stimuli in a spatial mode (Dykens et al., 1992). In fact, Jauregi et al. (2007) found that individuals with PWS did not differ from the standard population in terms of simultaneous processing tasks (figural memory). In summary, the PWS "overall cognitive profile at all ages" includes deficits in short-term memory, auditory processing, sequential processing, and arithmetic (Cataletto et al., 2011).

It is imperative that school psychologists and other school personnel recognize the uneven cognitive profile of children with PWS. Although PWS is a spectrum disorder and every student with PWS should be evaluated for their individual strengths and weaknesses, it is still appropriate to recognize the strengths and weaknesses that are typically associated with the syndrome. Students may appear non-compliant, uninterested, or become agitated when presented with verbal directions that require sequential ordering of steps and completion of a task in a specific time frame. Although many assessment tools do not provide flexibility in the administering protocols, results should be read with caution. Teachers should provide accommodations when instructing through lectures or giving verbal multi-step directions. In addition, they should capitalize on relative strengths, including visuospatial skills, simultaneous processing tasks, and acquiring new skills by "doing" to support learning.

2.2.4. Social cognitive deficits

PWS is marked by a variety of social cognitive deficits that vary across genetic subtypes, and often overlap with some features of Autism Spectrum Disorders (ASD) (Dimitropoulos et al., 2013; Schwartz et al., 2021; Whitman & Heithaus, 2022). These challenges impact the social interactions and interpersonal relationships of a child with PWS as they often struggle with understanding and appropriately responding to the emotions of others, perspective taking, and understanding social norms and expectations (Dimitropoulos et al., 2013; Schwartz et al., 2021; Whitman & Heithaus, 2022). Deficits in responding to social cues and emotions may be related to the fact that people with PWS struggle with recognizing the facial expressions and emotional cues of others, especially for negative emotions such as sadness or anger (Dykens et al., 2019; Whittington & Holland, 2011). These factors, in addition to behaviors of concern, can impact a child with PWS's peer relationships in the classroom, leading to isolation (Schwartz et al., 2021). As a result, children with PWS may benefit from interventions aiming to teach social skills similar to those provided to children with ASD (Dykens et al., 2019). However, even with intervention, the rigidity associated with PWS may mean that children continue to struggle to adapt to social norms, especially if the rules of social engagement are not clear or explicitly stated (Dimitropoulos et al., 2013; Whitman & Heithaus, 2022).

2.2.5. Behavioral features of PWS

Behaviors of concern are a frequent challenge for children with PWS; they can include food seeking and stealing, hyper fixation on food, tantrums or emotional outbursts, rigid and repetitive behaviors, task refusal, and self-injury (Bedard et al., 2021; Schwartz, et al., 2021). Food seeking and food stealing behaviors often include consuming food outside of a planned diet without permission, often in dangerous amounts (Schwartz et al., 2021). Children with PWS may also steal or sneak food, and sometimes obtain food from unsafe locations such as the trash or the floor (Schwartz et al., 2021). Opportunities to obtain food both inside and outside of a child's diet may become a source of anxiety and hyper fixation (Schwartz et al., 2021). For example, being denied food, having unauthorized food taken away, or delays in regularly scheduled meals may result in severe distress and tantrum behavior (Rice et al., 2018).

It is important to remember that food stealing and seeking are not an issue of poor self-control on the part of the person with PWS. Children with PWS who are experiencing hyperphagia truly feel as if they are starving. It is a biologically protective instinct to forage for and steal food if you are starving, and we therefore should not judge or lay blame on the child with PWS for food related behaviors. In addition to the potential emotional harm, reacting with judgment and blame can teach children with PWS to hide overconsumption which can be deadly if undetected.

Emotional outbursts and tantrums are often very disruptive to the child with PWS and those around them (Schwartz,et al., 2021). Tantrums tend to have a rapid onset, and slow de-escalation which can lead to a significant amount of time where the person is out of instruction (Rice et al., 2018; Schwartz,et al., 2021). If not redirected early, tantrum behavior can escalate from verbal or vocal behavior (e.g., yelling, screaming, crying) to more dangerous behavior including aggression, elopement, and disruption of property (Bedard et al., 2021, Schwartz, et al., 2021). It is again important to emphasize that tantrums often occur because of a true deficit in emotional regulation skills that occur due to brain differences, rather than an overreaction or intentional disruption of the environment. Children with PWS need help learning skills to help them regulate and accommodate for their different needs.

Children with PWS have a recognized biological predisposition towards rigid and repetitive behaviors (Schwartz et al., 2021). For many children this includes difficulties with changes in and strict adherence to schedules and routines (Schwartz et al., 2021). Children may also engage repetitive behaviors, such as completing a daily living or play

routine in a strictly structured way (Schwartz,et al., 2021). Interrupting these repetitive behaviors often results in emotional distress (Rice et al., 2018). However, this preference for routine can also benefit children with PWS, as they often enthusiastically adhere to school and classroom routines once these become well established.

Repetitive behaviors may extend to repetitive verbal behavior, or the propensity to repetitively ask questions or make statements (Schwartz, et al., 2021). Repetitive verbal behavior occurs more often when children are anxious about or perseverating on a specific topic, such as food or a change in schedule (Schwartz et al., 2021). If questions are ignored or inadequately answered from the perspective of the person with PWS, tantrum behavior may occur.

"Stubborn" and "oppositional" are words often used to describe children with PWS. However, these terms unfairly place blame on the person with PWS for their behavior without considering the immense physical and cognitive challenges associated with PWS. It is a common complaint by both caregivers and educators of children with PWS that they are noncompliant with tasks or demands (Avrahamy et al., 2015). This can include but is not limited to failing to transition from one task to another (Haig & Woodcock, 2017), and failing to initiate and complete a task either at all, or in a timely fashion. As children with PWS often struggle with fine and gross motor deficits, many school-related tasks are significantly more difficult and exhausting for them in comparison to their neurotypical peers.

This often means that children with PWS are quick to fatigue during fine and gross motor tasks, and reinforcement that is typically available in the environment (e.g., intermittent teacher praise) may be insufficient to motivate activity completion. Children may not effectively communicate the need for breaks or assistance especially if communication deficits are present or previous attempts to access breaks or attention have gone unrewarded. Executive functioning deficits may cause further difficulty as children with PWS may struggle to process direction and to plan tasks (Chevalère et al., 2015). The result is that children with PWS may resist engaging in academic activities by refusing or engaging in behaviors that prevent instruction.

A final behavior that can cause challenges in the classroom is self-injury, specifically skin picking behavior (Morgan et al., 2010; Whittington & Holland, 2020). People with PWS are believed to have a possible biological predisposition towards picking or over-grooming as evidenced by the presence of over-grooming (skin scraping) behavior in some mouse-models of PWS (Kummerfeld et al., 2021). Skin picking behavior often includes scratching and itching, removing scabs, picking at cuticles, fingernails, and toenails, continuously rubbing areas of skin, and at times, pulling out facial and head hair (Whittington & Holland, 2020). This behavior can be a challenge to manage in the classroom because it can result in the presence of blood, resulting in the need to remove the student from the classroom and treat the wound. While this behavior often co-occurs with anxiety and is believed to also have a sensory function (Hall et al., 2014; Radstaake et al., 2011; Whittington & Holland, 2020), children often learn to use picking behavior as a tool to escape unpreferred academic tasks or to gain attention from peers or adults (Radstaake et al., 2011).

2.2.6. Mental health challenges associated with PWS

Children with PWS are at risk for a myriad of mental health challenges as a result of a biological predisposition for mental illness compounded by many of the challenges described above, including physical and cognitive deficits, social emotional challenges, and behaviors of concern, all of which can exacerbate feelings of isolation, frustration, and distress (Butler et al., 2019; Feighan et al., 2020; Whitman & Heithaus, 2022). Anxiety is highly prevalent in the PWS population, and it is estimated that 40–50 % of children with PWS suffer from anxiety in some capacity, with an average onset of around eight years of age (Butler et al., 2019; Einfeld et al., 1999; Feighan et al., 2020; Shelkowitz et al., 2022). Sources of anxiety can include, but are not limited to food, especially in cases where food is unsecured, schedules and routines, and access to preferred activities, people, and items (Schwartz et al., 2021; Shelkowitz et al., 2022).

While children with PWS can be prone to engaging in obsessive behaviors, including engaging in rigid rituals, collecting, hoarding, or ordering items, and repetitive verbal behavior, these behaviors often don't meet the criteria for Obsessive Compulsive Disorder (OCD) due to differences in the types of obsessions and compulsions (e.g people with PWS often do not engage in checking, cleaning, or fixation on germs) (Ho & Dimitropoulos, 2010; Schwartz et al., 2021). However, a child's learning history associated with their patterns of repetitive and obsessive behaviors often interacts with anxiety, making it difficult to clearly differentiate symptoms of mental ill-health from behaviors of concern (Dimitropoulos et al., 2013; Schwartz et al., 2021; Woodcock et al., 2009).

While less common among children, many teens and adults with PWS also experience psychosis and mood disorders (Aman et al., 2024; Feighan et al., 2020; Ho and Dimitropoulos, 2010; Singh, 2022; Whitman & Heithaus, 2022). Symptoms of psychosis can occur during a child's school years, with the age of onset ranging between eight and 40 years of age, with teenage and early adulthood years presenting the highest risk (Aman et al., 2024; Ho and Dimitropoulos, 2010). Mood disorders in PWS are often characterized by mood instability and cyclical mood fluctuations, irritability, and episodes of persistent sadness (Singh, 2022; Whitman & Heithaus, 2022). Often psychiatric and mental health challenges can co-occur with behaviors of concern, such as emotional dysregulation, aggression, and increases in skin picking and rigid and repetitive behaviors (Whitman & Heithaus, 2022). Despite clear mental health concerns, people with PWS often do not meet criteria for DSM-V diagnoses such as generalized anxiety disorder, major depressive disorder, bipolar disorder, and schizophrenia, making diagnosis and treatment a challenge (Aman et al., 2024; Feighan et al., 2020; Schwartz et al., 2021).

3. Conclusion

PWS is a rare neurogenetic syndrome that results in significant challenges that can present direct conflict with the skills that are needed for success in the school setting. Although the number of children who will present with PWS may be small, knowledge of the physical and behavioral features of PWS is imperative, due to the high levels of risk that can accompany their unique challenges and needs. However, with appropriate academic, social-emotional, and behavioral supports, school psychologists can promote healthy and safe environments that promote academic success and allow children with PWS to thrive.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Data availability

No data was used for the research described in the article.

References

Aman, L. C. S., Lester, S. D., Holland, A. J., & Fletcher, P. C. (2024). Psychotic illness in people with Prader-Willi syndrome: A systematic review of clinical presentation, course, and phenomenology. *Orphanet Journal of Rare Diseases, 19*(1), 69. https://doi.org/10.1186/s13023-024-03026-y

Avrahamy, H., Pollak, Y., Shriki-Tal, L., Genstil, L., Hirsch, H. J., Gross-Tsur, V., & Benarroch, F. (2015). A disease-specific questionnaire for assessing behavior in individuals with Prader-Willi syndrome. *Comprehensive Psychiatry, 58*, 189–197.

Bedard, K. E., Griffith, A. K., Lister, M. A., & Swain, M. A. (2021). Behavioral and dietary management for adults with Prader-Willi syndrome in a residential setting. *Advances in Neurodevelopmental Disorders, 5*, 93–101. https://doi.org/10.1007/s41252-020-00185-4

Bertella, L., Girelli, L., Grugni, G., Marchi, S., Molinari, E., & Semenza, C. (2005). Mathematical skills in Prader-Willi Syndrome. *Journal of Intellectual Disability Research, 49*(Pt 2), 159–169. https://doi.org/10.1111/j.1365-2788.2004.00634.x

Brown, S. S. G., Manning, K. E., Fletcher, P., & Holland, A. (2022). In vivo neuroimaging evidence of hypothalamic alteration in Prader-Willi syndrome. *Brain. Communications, 4*(5). https://doi.org/10.1093/braincomms/fcac229

Butler, M. G. (2017). Clinical and genetic aspects of the 15q11.2 BP1-BP2 microdeletion disorder. *Journal of Intellectual Disability Research, 61*(6), 568–579. https://doi.org/10.1111/jir.12382

Butler, M., Lee, P., & Whitman, B. (2022). *Management of Prader-Willi Syndrome: Fourth edition.*. https://doi.org/10.1007/978-3-030-98171-6

Butler, M. G., Miller, J. L., & Forster, J. L. (2019). Prader-Willi Syndrome - Clinical Genetics, Diagnosis and Treatment Approaches: An Update. *Current Pediatric Reviews, 15*(4), 207–244. https://doi.org/10.2174/1573396315666190716120925

Cataletto, M., Angulo, M., Hertz, G., & Whitman, B. (2011). Prader-Willi syndrome: A primer for clinicians. *International Journal of Pediatric Endocrinology, 2011*(1), 12. https://doi.org/10.1186/1687-9856-2011-12

Cataldi, M., Arnaldi, D., Tucci, V., De Carli, F., Patti, G., Napoli, F., Pace, M., Maghnie, M., & Nobili, L. (2021). Sleep disorders in Prader-Willi syndrome, evidence from animal models and humans. *Sleep Medicine Reviews, 57*, Article 101432. https://doi.org/10.1016/j.smrv.2021.101432

Chai, W. J., Abd Hamid, A. I., & Abdullah, J. M. (2018). Working Memory From the Psychological and Neurosciences Perspectives: A Review. *Frontiers in Psychology, 9*, 401. https://doi.org/10.3389/fpsyg.2018.00401

Chevalère, J., Postal, V., Jauregui, J., Copet, P., Laurier, V., & Thuilleaux, D. (2015). Executive functions and Prader-Willi syndrome: Global deficit linked with intellectual level and syndrome-specific associations. *American Journal on Intellectual and Developmental Disabilities, 120*(3), 215–229. https://doi.org/10.1352/1944-7558-120.3.215

Conners, F. A., Rosenquist, C. J., Atwell, J. A., & Klinger, L. G. (2000). Cognitive Strengths and Weaknesses Associated with Prader-Willi Syndrome. *Education and Training in Mental Retardation and Developmental Disabilities, 35*(4), 441–448. http://www.jstor.org/stable/23879875.

Curfs, L. M. G., Wiegers, A. M., Sommers, J. R. M., Borghgraef, M., & Fryns, J. P. (1991). Strengths and weaknesses in the cognitive profile of youngsters with Prader-Willi syndrome. *Clinical Genetics, 40*, 430–434.

Curfs, L. M., & Fryns, J. P. (1992). Prader-Willi syndrome: A review with special attention to the cognitive and behavioral profile. *Birth Defects Original Article Series, 28*(1), 99–104.

Dimitropoulos, A., Ho, A., & Feldman, B. (2013). Social responsiveness and competence in Prader-Willi syndrome: Direct comparison to autism spectrum disorder. *Journal of Autism and Developmental Disorders, 43*(1), 103–113. https://doi.org/10.1007/s10803-012-1547-3

Donaldson, M. D., Chu, C. E., Cooke, A., Wilson, A., Greene, S. A., & Stephenson, J. B. (1994). The Prader-Willi syndrome. *Archives of Disease in Childhood, 70*, 58–63.

Driscoll, D. J., Miller, J. L., Schwartz, S., & Cassidy, S. (2017). *December 14)*. GeneReviews: Prader-Willi Syndrome. Retrieved from https://www.ncbi.nlm.nih.gov/books/NBK1330/.

Dykens, E. M., Hodapp, R. M., Walsh, K., & Nash, L. J. (1992). Profiles, correlates, and trajectories of intelligence in Prader-Willi syndrome. *Journal of the American Academy of Child and Adolescent Psychiatry, 31*(6), 1125–1130. https://doi.org/10.1097/00004583-199211000-00022

Dykens, E. M. (2002). Are jigsaw puzzle skills 'spared' in persons with Prader-Willi Syndrome? *Journal of Child Psychology and Psychiatry, and Allied Disciplines, 43*(3), 343–352. https://doi.org/10.1111/1469-7610.00025

Dykens, E. M., Roof, E., Hunt-Hawkins, H., Daniell, C., & Jurgensmeyer, S. (2019). Profiles and trajectories of impaired social cognition in people with Prader-Willi syndrome. *PloS One, 14*(10), e0223162.

Einfeld, S. L., Smith, A., Durvasula, S., Florio, T., & Tonge, B. J. (1999). Behavior and emotional disturbance in Prader-Willi syndrome. *American Journal of Medical Genetics, 82*(2), 123–127. https://doi.org/10.1002/(SICI)1096-8628(19990115)82:2<123::AID-AJMG4>3.0.CO;2-C

Feighan, S. M., Hughes, M., Maunder, K., Roche, E., & Gallagher, L. (2020). A profile of mental health and behaviour in Prader-Willi syndrome. *Journal of Intellectual Disability Research, 64*(2), 158–169. https://doi.org/10.1111/jir.12707

Foti, F., Menghini, D., Orlandi, E., Rufini, C., Crinò, A., Spera, S., Vicari, S., Petrosini, L., & Mandolesi, L. (2015). Learning by observation and learning by doing in Prader-Willi syndrome. *Journal of Neurodevelopmental Disorders, 7*(1), 6. https://doi.org/10.1186/s11689-015-9102-0

Gross-Tsur, V., Landau, Y. E., Benarroch, F., Wertman-Elad, R., & Shalev, R. S. (2001). Cognition, attention, and behavior in Prader-Willi syndrome. *Journal of Child Neurology, 16*(4), 288–290. https://doi.org/10.1177/088307380101600411

Haig, E. L., & Woodcock, K. A. (2017). Rigidity in routines and the development of resistance to change in individuals with Prader-Willi syndrome. *Journal of Intellectual Disability Research, 61*(5), 488–500. https://doi.org/10.1111/jir.12368

Hall, S. S., Hustyi, K. M., Chui, C., & Hammond, J. L. (2014). Experimental functional analysis of severe skin-picking behavior in Prader-Willi syndrome. *Research in Developmental Disabilities, 35*(10), 2284–2292. https://doi.org/10.1016/j.ridd.2014.05.025

Ho, A. Y., & Dimitropoulos, A. (2010). Clinical management of behavioral characteristics of Prader-Willi syndrome. *Neuropsychiatric Disease and Treatment, 6*, 107–118. https://doi.org/10.2147/NDT.S5560

Hsu, W. L., Chiu, V. J. Y., Chang, W. H., Lin, M. C., Wei, J. T., & Tzeng, I. S. (2018). Hand strength and dexterity in patients with Prader-Willi syndrome: A pilot intervention study. *Journal of International Medical Research, 46*(11), 4669–4677. https://doi.org/10.1177/0300060518788243

Jauregi, J., Arias, C., Vegas, O., Alén, F., Martinez, S., Copet, P., & Thuilleaux, D. (2007). A neuropsychological assessment of frontal cognitive functions in Prader-Willi syndrome. *Journal of Intellectual Disability Research, 51*(5), 350–365. https://doi.org/10.1111/j.1365-2788.2006.00883.x

Kummerfeld, D. M., Raabe, C. A., Brosius, J., Mo, D., Skryabin, B. V., & Rozhdestvensky, T. S. (2021). A comprehensive review of genetically engineered mouse models for Prader-Willi syndrome research. *International Journal of Molecular Sciences, 22*(7), 3613. https://doi.org/10.3390/ijms22073613

Lewis, B. A. (2006). Speech and language disorders associated with Prader-Willi syndrome. In M. G. Butler, P. D. K. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome*. New York, NY: Springer. https://doi.org/10.1007/978-0-387-33536-0_9.

Lui, M., & Tannock, R. (2007). Working memory and inattentive behaviour in a community sample of children. *Behavioral and Brain Functions, 3*, 12. https://doi.org/10.1186/1744-9081-3-12

Maas, A. P. H. M., Sinnema, M., Didden, R., Maaskant, M. A., Smits, M. G., Schrander-Stumpel, C. T. R. M., & Curfs, L. M. G. (2010). Sleep disturbances and behavioural problems in adults with Prader-Willi syndrome. *Journal of Intellectual Disability Research, 54*(10), 906–917. https://doi.org/10.1111/j.1365-2788.2010.01306.x

McCandless, S. E., Suh, M., Yin, D., Yeh, M., Czado, S., Aghsaei, S., Li, J. W., Francis, K., Hadker, N., & Stafford, D. E. J. (2020). SUN-604 U.S. Prevalence & mortality of Prader-Willi syndrome: A population-based study of medical claims. *Journal of the Endocrine Society, 4*(Suppl 1), SUN-604. https://doi.org/10.1210/jendso/bvaa046.993

Miller, J. L., Lynn, C. H., Driscoll, D. C., Goldstone, A. P., Gold, J.-A., Kimonis, V., Dykens, E., Butler, M. G., Shuster, J. J., & Driscoll, D. J. (2011). Nutritional phases in Prader-Willi syndrome. *American Journal of Medical Genetics. Part A, 155A*(5), 1040–1049. https://doi.org/10.1002/ajmg.a.33951

Miller, J. L., Lynn, C. H., Shuster, J., & Driscoll, D. J. (2013). A reduced-energy intake, well-balanced diet improves weight control in children with Prader-Willi syndrome. *Journal of Human Nutrition and Dietetics, 26*(1), 2–9. https://doi.org/10.1111/j.1365-277X.2012.01275.x

Miller, J. L., & Tan, M. (2020). Dietary management for adolescents with Prader-Willi syndrome. *Adolescent Health, Medicine and Therapeutics, 11*, 113–118. https://doi.org/10.2147/AHMT.S214893

Morgan, J. R., Storch, E. A., Woods, D. W., Bodzin, D., Lewin, A. B., & Murphy, T. K. (2010). A preliminary analysis of the phenomenology of skin-picking in Prader-Willi syndrome. *Child Psychiatry and Human Development, 41*(4), 448–463. https://doi.org/10.1007/s10578-010-0180-7

Prader-Willi Syndrome Association USA. (2015). Food security in the education environment for students with Prader-Willi syndrome. Retrieved from https://www.pwsausa.org/wp-content/uploads/2015/12/Food-Security-Educational-Environments-2012-ED-24.pdf.

Radstaake, M., Didden, H. C., Bolio, M., Lang, R., Lancioni, G., & Curfs, L. M. (2011). Functional assessment and behavioral treatment of skin picking in a teenage girl with Prader-Willi syndrome. *Clinical Case Studies, 10*(1), 67–78. https://doi.org/10.1177/1534650110395013

Reilly, C. (2009). Educating students with Prader-Willi syndrome. Retrieved from *REACH: Journal of Inclusive Education in Ireland, 23*(1), 13–23 https://reachjournal.ie/index.php/reach/article/view/102.

Reus, L., Zwarts, M., van Vlimmeren, L. A., Willemsen, M. A., Otten, B. J., & Nijhuis-van der Sanden, M. W. G. (2011). Motor problems in Prader-Willi syndrome: A systematic review on body composition and neuromuscular functioning. *Neuroscience and Biobehavioral Reviews, 35*(3), 956–969. https://doi.org/10.1016/j.neubiorev.2010.10.015

Rice, L. J., Woodcock, K., & Einfeld, S. L. (2018). The characteristics of temper outbursts in Prader-Willi syndrome. *American Journal of Medical Genetics. Part A, 176*(11), 2292–2300. https://doi.org/10.1002/ajmg.a.40480

Roof, E., Stone, W., MacLean, W. E., Feurer, I. D., Thompson, T. I., & Butler, M. G. (2000). Intellectual characteristics of Prader-Willi syndrome: Comparison of genetic subtypes. *Journal of Intellectual Disability Research : JIDR, 44*(Pt 1), 25–30.

Schwartz, L., Caixàs, A., Dimitropoulos, A., Dykens, E., Duis, J., Einfeld, S., Gallagher, L., Holland, A., Rice, L., Roof, E., Salehi, P., Strong, T., Taylor, B., & Woodcock, K. (2021). Behavioral features in Prader-Willi Syndrome (PWS): Consensus paper from the International PWS Clinical Trial Consortium. *Journal of Neurodevelopmental Disorders, 13*(1), 25. https://doi.org/10.1186/s11689-021-09373-2

Scott, E. M., Smith, T. E. C., Hendricks, M. D., & Polloway, E. A. (1999). Prader-Willi Syndrome: A Review and Implications for Educational Intervention. *Education and Training in Mental Retardation and Developmental Disabilities, 34*(1), 110–116. http://www.jstor.org/stable/23879246.

Semenza, C., Pignatti, R., Bertella, L., Ceriani, F., Mori, I., Molinari, E., Giardino, D., Malvestiti, F., & Grugni, G. (2008). Genetics and mathematics: Evidence from Prader-Willi Syndrome. *Neuropsychologia, 46*(1), 206–212. https://doi.org/10.1016/j.neuropsychologia.2007.07.017

Shelkowitz, E., Gantz, M. G., Ridenour, T. A., Scheimann, A. O., Strong, T., Bohonowych, J., & Duis, J. (2022). Neuropsychiatric features of Prader-Willi syndrome. *American Journal of Medical Genetics. Part A, 188*(5), 1457–1463. https://doi.org/10.1002/ajmg.a.62662

Singh, D. (2022). Neuro-behavioral Manifestations of Prader-Willi Syndrome: A Guide for Clinicians and Caregivers (1st ed.). Cambridge University Press. doi: 10.1017/9781108886727.

Stauder, J. E., Brinkman, M. J., & Curfs, L. M. (2002). Multi-modal P3 deflation of event-related brain activity in Prader-Willi syndrome. *Neuroscience Letters, 327*(2), 99–102. https://doi.org/10.1016/s0304-3940(02)00377-4

Strenilkov, K., Debladis, J., Salles, J., Valette, M., Mantoulan, C., Thuilleaux, D., Laurier, V., Molinas, C., Barone, P., & Tauber, M. (2020). A study of voice and non-

voice processing in Prader-Willi syndrome. *Orphanet Journal of Rare Diseases, 15*(1), 22. https://doi.org/10.1186/s13023-020-1298-8

Verdine, B. N., Troseth, G. L., Hodapp, R. M., & Dykens, E. M. (2008). Strategies and correlates of jigsaw puzzle and visuospatial performance by persons with Prader-Willi syndrome. *American Journal of Mental Retardation: AJMR, 113*(5), 343–355. https://doi.org/10.1352/2008.113:342-355

Vogels, A., Jenneke Van, D. E., Keymolen, K., Mortier, G., Devriendt, K., Legius, E., & Fryns, J. P. (2004). Minimum prevalence, birth incidence and cause of death for prader-willi syndrome in Flanders. European Journal of Human Genetics : EJHG, 12 (3), 238-40. doi: 10.1038/sj.ejhg.5201135.

Whitman, B. Y., & Heithaus, J. L. (2022). Neurodevelopmental and neuropsychological aspects of Prader-Willi Syndrome. In M. G. Butler, P. D. K. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (pp. 219–246). Cham: Springer. https://doi.org/10.1007/978-3-030-98171-6_8.

Whittington, J., & Holland, A. (2017). Cognition in people with Prader-Willi syndrome: Insights into genetic influences on cognitive and social development. *Neuroscience and Biobehavioral Reviews, 72*, 153–167. https://doi.org/10.1016/j.neubiorev.2016.09.013

Whittington, J., & Holland, A. (2020). Developing an understanding of skin picking in people with Prader-Willi syndrome: A structured literature review and re-analysis of existing data. *Neuroscience and Biobehavioral Reviews, 112*, 48–61.

Whittington, J. E., Holland, A. J., Webb, T., Butler, J., Clarke, D., & Boer, H. (2001). Population prevalence and estimated birth incidence and mortality rate for people with Prader-Willi syndrome in one UK Health Region. *Journal of Medical Genetics, 38* (11), 792–798. https://doi.org/10.1136/jmg.38.11.792

Whittington, J., Holland, A., Webb, T., Butler, J., Clarke, D., & Boer, H. (2004). Cognitive abilities and genotype in a population-based sample of people with Prader-Willi syndrome. *Journal of Intellectual Disability Research : JIDR, 48*(Pt 2), 172–187. https://doi.org/10.1111/j.1365-2788.2004.00556.x

Whittington, J., & Holland, T. (2011). Recognition of emotion in facial expression by people with Prader-Willi syndrome. *Journal of Intellectual Disability Research : JIDR, 55*(1), 75–84. https://doi.org/10.1111/j.1365-2788.2010.01348.x

Wigren, M., & Hansen, S. (2005). ADHD symptoms and insistence on sameness in Prader-Willi syndrome. *Journal of Intellectual Disability Research: JIDR, 49*(Pt 6), 449–456.

Woodcock, K., Oliver, C., & Humphreys, G. (2009). Associations between repetitive questioning, resistance to change, temper outbursts and anxiety in Prader-Willi and Fragile-X syndromes. *Journal of Intellectual Disability Research : JIDR, 53*(3), 265–278. https://doi.org/10.1111/j.1365-2788.2008.01122.x

Woodcock, K. A., Oliver, C., & Humphreys, G. W. (2009). Task-switching deficits and repetitive behaviour in genetic neurodevelopmental disorders: Data from children with Prader-Willi syndrome chromosome 15 q11–q13 deletion and boys with Fragile X Syndrome. *Cognitive Neuropsychology, 26*(2), 172–194.

Wu, N., Yu, H., & Xu, M. (2022). Alteration of brain nuclei in obese children with and without Prader-Willi syndrome. *Frontiers in Neuroinformatics, 16*, 1032636. https://doi.org/10.3389/fninf.2022.1032636

Yamada, K. (2023). *The Brain in Prader-Willi Syndrome. In Eating Disorders* (pp. 1261–1285). Springer International Publishing.

Yamada, K., Watanabe, M., & Suzuki, K. (2022). Differential volume reductions in the subcortical, limbic, and brainstem structures associated with behavior in Prader-Willi syndrome. *Scientific Reports, 12*(1), 4978. https://doi.org/10.1038/s41598-022-08898-3