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Prader-Willi syndrome

Understanding and support at school

By Norbert Hödebeck-Stuntebeck

“If our daughter cannot go to school in the morning, that is the greatest stress for her.”

The aim of this article is to describe the specific characteristics of Prader-Willi Syndrome and, on the basis of this knowledge, to derive accompanying and supportive actions for the educational support of children and adolescents with PWS.

Prader-Willi syndrome (PWS) was first described in 1956 by Swiss physicians Andrea Prader, Alexis Labhart, and Heinrich Willi. This multicomplex syndrome occurs when the information on paternal chromosome 15 in the region q11-13 cannot be retrieved. Causes are:

- in about 75% the deletion, the absence of the region 15q11-13 of the paternal chromosome 15,
- in about 24% of cases, disomy , in which the paternal chromosome 15 is completely missing, while the maternal chromosome 15 is present twice,
- and in about 1% the imprinting disorder, in which there is an incorrect wiring of the relevant genes of the paternal chromosome 15 in the subregion 15q11-13.

The incidence of PWS worldwide is 1:25,000, which would result in approximately 5,000 people with PWS in Germany. However, only approximately 1000 people of all ages are currently known in Germany.

Due to the increasingly challenging behaviors that occur in childhood and adolescence, PWS-specific outpatient or residential services are often necessary. The first PWS-specific residential services developed in Germany in the mid-1990s. Currently, there are only approximately 500 residential service places for people with PWS in Germany, including approximately 25 places for children and adolescents.

The following describes what the author considers to be the most significant symptoms of PWS (for a detailed description, see Driscoll et al., 2023). For each symptom, the cause and consequences for everyday school life are presented. Furthermore, options for supportive measures in school context are suggested. It is fundamentally important that the person with PWS understands why these measures are being implemented (meaningfulness) and, if possible, that they can develop and implement them independently (experience of self-efficacy).

Reduced muscle mass and balance problems

People with PWS have up to 50% reduced muscle mass. This results in life-threatening difficulty in sucking during the first few days of life. However, the muscles can be trained, so that children, adolescents, and adults with PWS can develop nearly adequate fine and gross motor skills over time.

In addition to the reduced muscle mass, the genetically determined altered interpretation of the internal body stimuli entering the brain is another special feature of PWS. This change contributes, among other things, to the fact that people with PWS often have difficulties with their sense of balance.

Impact on everyday school life

In everyday school life, it's important to remember that all motor tasks and demands can pose a significantly greater challenge and strain for people with PWS. Possible consequences include defiant behavior or suddenly emerging conflicts. These often have no connection to the current situation or appear to arise without cause. People with PWS are often unaware of the underlying muscular issues, so they often cannot identify the cause of the perceived overload.

Additionally, overcoming steps, hurdles, or walking on uneven terrain can trigger anxiety in people with PWS due to balance issues. This often results in challenging behaviors.

Measures

Promoting muscle maintenance and development reduces the sense of effort felt by the person with PWS. It contributes to improved self-esteem as they increasingly master similar tasks to other learners.

Before undertaking actions that require (particular) physical effort, the person with PWS can be involved in considering what difficulties they expect to encounter during implementation and what possible solutions they see (“What do you suggest?”).

The presence of a reduced sense of balance can be discussed with the individual and made tangible. If the individual expresses a desire for improvement, exercises to improve body awareness can be helpful, either as targeted support or integrated into everyday school life. Another simple option is regular, targeted exercise on uneven ground, such as during a walk, which can contribute to a noticeable improvement in the sense of balance.

Food addiction

People with PWS exhibit a genetically determined overeating disorder throughout their lives. However, it is increasingly evident that the severity of overeating disorder is highly dependent on the person's acquired skills in dealing with overeating (self-control). The extent of these skills depends on:

- how early parents or the environment can use sufficient information about PWS including eating disorders,
- whether the child is involved as early as possible in the need for an adjusted amount of food and experiences that family members eat different foods and quantities (transparency about the effects of the syndrome),
- whether the child is involved in the purchase and preparation of food as early as possible, can experience tasks and responsibilities in the appropriate handling of food and is proud of being able to handle food (experience of self-efficacy).

It's important to remember that eating disorders persist throughout life in people with PWS. In crisis situations, during major changes, or during other special (even positive) events, eating disorders can manifest very suddenly as a reaction, even if there were (very) long periods beforehand in which behaviors consistent with eating disorders were not observed.

Impact on everyday school life

Uncertainties or changes regarding food and drinks in school can lead to anxiety or panic in people with PWS. This often triggers a chain of thoughts that leads from insecurity and fear of loss to the fear of "never getting anything to eat again." The consequences are sometimes very severe challenging behaviors that sometimes appear "out of nowhere." The person with PWS is often unable to articulate this chain of thoughts in the situation, and outsiders cannot understand why the statement "...maybe we'll start eating a minute or two later today..." has such a serious impact.

Measures

At the start of the school year, when entering a new classroom, or during other foreseeable changes, it is very helpful to explain to the child the processes, structures, quantities, and times related to meals and beverages (consumption) in this setting and to ensure that the child understands these things. The child should be encouraged to contribute ideas or suggestions for improvement. The more successfully the child is involved in taking responsibility for these routines, the greater the chance that they will learn appropriate, self-regulated handling of food. This allows the child to once again experience a greater sense of self-efficacy, which contributes to a greater level of security and well-being.

Hypothyroidism and sleep apnea

A higher percentage of people with PWS have hypothyroidism and sleep apnea. Both disorders contribute to increased fatigue, lethargy, or decreased interest in everyday school life. If teachers lack awareness of these conditions, the child often experiences repeated requests that they cannot comply with due to their physical condition. The child feels unfairly challenged or reprimanded. The reaction is often anger, coupled with the emergence of challenging behavior.

If there is a significant increase in fatigue during everyday school life, parents or guardians should be informed so that appropriate examinations and treatment can be carried out.

Reduced cognitive flexibility and intellectual disability

Many people with PWS expect those around them to maintain existing routines and implement agreements as planned. When changes or deviations occur, the person with PWS often reacts with rejection, fear, and uncertainty, which can lead to withdrawal or (significant) challenging behavior. The frequently present mild to moderate intellectual disability certainly makes it difficult to grasp the situation or deal with it flexibly and appropriately. The impact of change is particularly severe if the person with PWS does not understand these changes.

Impact on everyday school life

School structures and procedures (start of school, break times, seating, fixed class groups, etc.) offer individuals with PWS many opportunities for orientation and predictable procedures and content. They thus create security and reliability. Illnesses of classmates, teachers, or integration assistants, a flat tire on the shuttle vehicle, new school topics, announcements of future school activities: these are just a few examples that can arise in the school context and disrupt the usual (and, from the perspective of the child with PWS, desired) routine. These changes can then contribute to uncertainty and anxiety, even in the very short term, and lead to (significant) challenging behaviors.

Measures

To avoid a child with PWS experiencing a fundamental sense of uncertainty, it is important to ensure that the child knows and understands the regular school procedures and content, and that they understand and can explain why these rules exist. If they recognize their purpose, they are much more likely to be motivated to implement them. Where the child expresses criticism, dissatisfaction, or requests for change, they should be actively involved in considering possible adjustments or changes ("What is your suggestion?"). If the child's suggestions are not feasible, this should be discussed transparently with the child. Spontaneously necessary changes (information about a teacher being ill in the morning) cannot be discussed with the child in advance. In this case, it is very helpful to develop basic strategies (Plan B) with the child: "What happens if we have planned something but it is suddenly no longer possible?" (Here, too, it is important that the proposed solution is the child's, if possible, and to ensure that the child understands this solution ("Say it again in your own words").

Setting-dependent learning

For people with PWS, recalling learned knowledge, competencies, and skills is often dependent on the setting in which they acquired this knowledge. For example, if a child has learned appropriate social interaction with other children at home, they cannot automatically recall this knowledge at school. Transferring what has been learned is possible, but often requires further instruction in the new setting. It is helpful to know *how* the person acquired the skill in the original setting.

Impact on everyday school life

Knowing about the child's abilities in non-school contexts (e.g., at home), certain expectations are often placed on the child with PWS at school ("they can already do that at home"). However, if the child is only able to transfer these abilities to the school context to a very limited extent due to the syndrome , they experience excessive demands and may display defiant or challenging behavior as a reaction.

Measures

If learners with PWS display defiant or challenging behavior in connection with tasks where experience shows that the child has already mastered the task well elsewhere, it is important to check whether the child can transfer the skills and knowledge to the current setting. This review or support of transfer cannot take place in a "crisis situation". In the debriefing, the skills they possess in the other setting (e.g. at home) should be discussed with the person with PWS and how they can successfully apply these skills in school as well. If this transfer is successful, the child with PWS will increasingly develop a feeling of self-efficacy after this successful shared process.

Reduced ability to transfer experiences

People with PWS are often involved in conflicts or (massive) arguments, and their actions can have very negative consequences. They repeatedly exhibit similar behavior in similar subsequent situations. They fail to draw conclusions, often fail to learn from unsuccessful actions and situations, or fail to transfer their experiences to subsequent situations.

It can also be observed that they are unable to transfer their own previous actions ("I took Maria's pencil away yesterday") to current situations ("I wonder why Maria is looking at me so grimly today"). The behavior of the other person (Maria) is only evaluated in the current context ("Maria is behaving incorrectly right now"). As a result, greater difficulties in social relationships can arise.

Impact on everyday school life

People with PWS react to the current perceived misconduct of teachers or classmates without considering their own role in this behavior. From the perspective of the person with PWS, it is "right" to criticize the other person's behavior because it is inappropriate in the current situation. References from the other person regarding the person with PWS's previous behavior are not accepted by the person because they do not transfer their own previous behavior to the other person's behavior.

Since, from the subjective point of view of the people involved, everyone is “in the right,” conflicts are often the result.

Measures

Because people with PWS have significantly reduced cognitive abilities, it will be impossible to convey the connections between their previous behavior and the current situation in such a tense situation. The only possible approach is to resolve and end the situation with the least possible damage. In the necessary follow-up discussion, it is then necessary to work through the aforementioned connections together with the person with PWS. The goal is for the child to make the transfer and develop and apply their own solutions for similar future situations. This also highlights the great necessity of using follow-up discussions when supporting people with PWS.

Delayed overcoming of egocentrism

In the first years of life, all children perceive connections or sensations exclusively from their own perspective. They cannot yet perceive the wishes or needs of others and therefore do not control their own behavior. Overcoming this egocentrism is usually increasingly successful during elementary school. However, in people with PWS, this happens much later due to the developmental delay. The ability to shift cognitive perspectives is often significantly better developed than the ability to shift emotional perspectives.

Impact on everyday school life

If egocentrism is not sufficiently overcome, demands on the child to take into account the wishes, feelings or actions of others usually lead to excessive demands, which in turn can result in challenging behavior.

This is especially true if, on the one hand, the child experiences that they can easily understand the processes or plans of others and behaves accordingly (cognitive shift in perspective), but, on the other hand, they do not consider the feelings or sensitivities of others in their own actions (lack of emotional shift in perspective, Hödebeck-Stuntebeck, 2012). The expectation that an emotional shift in perspective should also be possible leads to excessive demands on the child and further potential conflicts.

Measures

In order to avoid excessive demands due to possible developmental delays, it is important to check in everyday life whether the person with PWS is already pursuing common goals with others, whether it is important to them what others want and how others feel, and whether they subordinate their own goals to those of others or put them aside.

Images of faces with clear emotional expressions can be used to assess the development of emotional perspective-taking. How well does the person attest to this, how well can they describe the visible characteristics of the emotions? Promoting this skill increases the likelihood of appropriate behavior in social situations and thus promotes the development of friendships and relationships.

Striving for dominance

Every person's personality development is at its most dynamic during the puberty phase of life. The preceding phase of childhood is characterized by dictates, external control, setting boundaries, and guidance provided by the environment (e.g., the parents' home). The child bears no responsibility but also has few decision-making opportunities.

In the phase of adulthood following puberty, there is a social expectation that the person takes responsibility for themselves and others, makes decisions and is clearly recognizable to their partner with their personality traits.

The main task of puberty is therefore the development of an individual's own personality. This can only succeed if there is a (partial) rejection of the previous guidelines, rules, and norms of the environment (a drive for dominance) in order to develop one's own attitudes and behaviors through one's own experiences and their consequences, thus leading to the development of one's own personality. The cognitive developmental delays often present in people with PWS contribute to the fact that pubertal behaviors often begin later.

Impact on the school context

(In children and adolescents) In people with PWS (from around 10 years of age) in late adolescence, when defiant or challenging behaviors occur, it is always important to first consider whether the adolescent (child) understands the requirements or tasks. However, it is also important to consider whether the behavior is based on a rejection of instructions (striving for dominance) and whether the behavior ultimately serves the development of the individual's personality. Often, therefore, the content of the actions is not relevant; it is simply a question of who decides what should be done.

Measures

If, in problematic situations, the perception arises that the person with PWS is no longer oriented towards the current content in their argumentation or actions and is striving to "be right" or to assert their own opinion, the only goal can be to resolve the situation with as little damage as possible. A solution to the underlying problem is not possible in this situation. Clarification of the understanding or the causes can only be achieved in a follow-up discussion. In order to support personal development and the assumption of responsibility, it is important in the subsequent reflection discussions that the person themselves suggest solutions to the problematic situation ("How do you want to do it next time so that there is no conflict?") and experience opportunities to implement these actions. The positive implementation of their own suggested solutions then leads to a higher level of self-efficacy and self-efficacy-expectation, which overall contributes to an increase in the subjectively perceived well-being of the person with PWS.

Perspective

The support and care of people with PWS has undergone a paradigm shift in recent years. For a long time, the focus was on providing structure and security for those affected through guidelines (external control), especially regarding nutrition, not least to prevent massive excess weight and the associated health risks. These efforts are increasingly being replaced by the realization that early involvement of those affected, a high level of transparency, and the appropriate transfer of responsibility (self-determination) increasingly empowers people with PWS to take responsibility for themselves in *all* areas of life and to experience themselves as self-effective.

The support and promotion of people with PWS is increasingly based on the concept of subjectively perceived well-being (Hödebeck-Stuntebeck, 2023), the experience of:

- positive feelings of joy, pride, happiness,
- Actions in which the person “loses” himself (being in flow),
- social relationships in different settings,
- Meaningfulness in everyday actions,
- Self-efficacy in the sense of pride in the success of one’s own planning and actions.

Only a sufficiently high level of expression in all sub-areas ensures the subjective well-being of the person with PWS. This must be the goal of all those involved in the various care areas for people with PWS.

Literature

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