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How to Provide Holistic Care for Disabled People Affected by Prader-Willi Syndrome

A descriptive literature review

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Thesis abstract

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Prader-Willi Syndrome was first reported in 1956 by Prader, Labhart and Willi as a genetic multisystem disorder with eating disorders, hormones deficiency, cognitive impairment, intellectual deficits and behavioral disorders.

Prader-Willi Syndrome (PWS) is quite unknown and rare. This rarity makes it harder to understand and *de facto* harder to take care of it from a nursing point of view. Studies show that nursing care is mostly done at home by family, and for adults with PWS care is mostly provided in home care centers. Usually, home care centers are not specialized in PWS, but general home care for people with disabilities. The fact that there is not special home care specialized for individuals with PWS may also mean that there is not appropriate nursing care regarding this syndrome. Nurses and caregivers are mostly taught to provide care for people with disabilities, but not very often specialized in certain care what individuals with PWS need.

The aim of this thesis was to put forward different kinds of care for disabled people having Prader-Willi Syndrome (PWS). The purpose was to do a literature review, to regroup and organize different studies and research studies regarding this syndrome, thus permitting to caregivers to provide holistic and appropriate care for disabled people affected by Prader-Willi Syndrome. As this syndrome is quite rare and unknown, an English descriptive literature review was done. Finally, to answer to the research question, 18 articles were analysed using inductive content analysis.

The results show that individuals with PWS may face different symptoms, such as obesity and behaviour problems. However, some support and care are possible, and caregivers need to know what the special needs of individuals with PWS are, to also help them in their work and in the support they provide for them. Some methods are available to offer the food security that individuals with PWS need, but also support methods are available for caregivers, regarding the psychological side of the syndrome, such as tantrums. There is some way to avoid them, and it requires knowledge from caregivers.

Keywords: Prader-Willi Syndrome, holistic care, care management

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Terms and Abbreviations

AHNA	American Holistic Nurses Association
ASD	Autism Spectrum disorders
GH	Growth Hormone
GHD	Growth Hormonal Deficiency
GHT	Growth Hormone Therapy
GHRH	Growth Hormone Releasing Hormone
OCB	Obsessive Compulsive Behaviors
PWS	Prader-Willi Syndrome
WHO	World Health Organization

1 INTRODUCTION

Prader-Willi Syndrome (PWS) is quite an unknown, rare, and complex disorder. Its prevalence is from 1/15000 to 1/30000 cases per year worldwide (Duis et al., 2018). This rarity makes it harder to understand and *de facto* harder to take care of it from a nursing point of view. A study from Tauber et al (2015) shows that nursing care is mostly done at home by family, and mostly for adults with PWS in home care centers. Usually, home care centers are not specialized in PWS, but general home care for people with disabilities (Duis et al, 2018), even though there is some exception, such as Tauber et al (2015) demonstrate in France, with centers specialized in PWS care, but only for short-time care.

The fact that there is not special home care specialized for individuals with PWS may also mean that there is not appropriate nursing care regarding this syndrome (Duis et al, 2018). Nurses and caregivers are mostly taught to provide care for people with disabilities, but not very often specialized in certain care (Billings and Halstead, 2015), and PWS care is part of this lack of knowledge, not because of the caregivers themselves, but because of the complexity and rarity of the syndrome. Individuals with PWS need a certain specialized care according to their needs, but they also need to be understood. When caregivers can understand, they can offer better and appropriate care for individuals with PWS. In that way, we can talk about good care, which is care respecting and understanding the special need of individuals with PWS. In that way and to this thesis subject, good care means appropriate and understanding care regarding the needs of individuals with PWS.

This thesis is done in cooperation with Lehtimäki Education Centre, a special school and home care place for people with disabilities created in 1971. This place offer teaching for people according to their capacities and abilities, as well as provide them home care services (Lehtimäki Education Centre, n.d.-a). Young adults with PWS study and live in Lehtimäki Education Centre, and caregivers did not have special competence concerning their care, their physical and psychological needs, and the understanding of the illness or what it includes regarding the care. This led to this thesis subject: to deepen the competence of caregivers, but also learn more about the syndrome, and be able to provide appropriate and holistic care according to their special needs.

2 HOLISTIC CARE OF DISABLED PEOPLE

According to the research of Ngugi and Igunnuoda (2015), holistic nursing is a model that associates the concept of presence, healing and holism, and the aim of the holistic nursing is to avoid suffering.

The American Holistic Nurses Association (AHNA) holistic care and more specifically holism “involves studying and understanding the interrelationship of the bio-psychosocial-spiritual dimensions of the person recognizes that the whole is greater than the sum of its parts”, which means that all parts of a patient have to be considered by the nurse in nursing situation, not only the body and the part that is affected by an ailment. Jasemi et.al. (2017) are also writing about the consideration of the “whole” instead of the sum of its parts.

Jasemi et. al. (2017) also mention the “patient-centered care” concept, which can be considered as holistic care. In this concept, the holistic care is a behavior that “recognizes a person as a whole and acknowledges the interdependence among their biological, social, psychological, and spiritual aspects”. Holistic care also includes other aspects, such as self-care, complementary treatment, education, or drugs (Jasemi et al, 2017).

Holistic care in home care is also important, including home care for disabled people. Bamfo & Hagin (2011) are explaining the effects of holistic care in home care, such as the relationship between the nurse and the patient, and the trust that is needed between them, also to provide holistic care, but also, for the patient to receive it. For patients affected by mental and behavioral disorders, the trust is the key to establish and provide holistic care. The relationship between the caregiver and the patient is also based on respect, understanding, equality, openness, and the patient participating in the decision making regarding his/her care (Zamanzadeh et al, 2015).

Zamanzadeh et al (2015) also point out the patient’s role in the treatment process and consider that the patient should be part of the whole process, but also to encourage self-care. Having him/her take part in the process is an important aspect, especially for people with behavioral disorders (Bennett et al, 2015).

3 PRADER-WILLI SYNDROME

Prader-Willi Syndrome was first reported in 1956 by Prader, Labhart and Willi (Tauber et al. 2015). It is a genetic multisystem disorder defined during childhood by hypotonia, eating disorders with poor weight gain, lethargy and hormones deficiency. The eating disorders are characterized by feeding difficulties during infancy and turn to an excessive appetite during childhood (Paepegaey et al, 2018., Duis et al., 2018). Individuals suffering from this disorder do not have the satiety felling after a complete meal, and they face overeating that can lead to obesity (Lambert et al., 2018).

Prader-Willi syndrome is a multisystem complex disorder present in 1/15000 to 1/30000 individuals that is caused by the loss of paternally expressed genes on chromosome 15q11.2- 13 (Duis et al., 2018). This incidence varies from a study to another but is always within this range.

Individuals with PWS usually also go through features and abnormalities, such as endocrine disorders including hypogonadism with cryptorchidism (urogenital malformation) and primary amenorrhea (Paepegaey et al, 2018). In a general view, and according to Paepegaey et al, (2018), 90% of the children diagnosed with PWS have Growth Hormone (GH) deficiency, and hypothyroidism appear to be common hormonal disorders. Other features have been reported, such as moderate cognitive deficit, learning disabilities, behavioral and social skills disorders, and psychiatric disorders (Tauber et al, 2015, pp 853).

Tauber et al (2015, p 857) affirm that every patient with PWS have cognitive impairment and most have mild to moderate intellectual deficit. Still, according to Tauber et al (2015, p857), there is also mental rigidity, a lack of fluidity of thought, and difficulties in conceptualization. Neurological deficits are also present. They affect attention, memory, executive functions, and are associated with difficulties with time, space, and causality. Whatever their level, the ability of patients to adapt remains low and their integration into the ordinary environment is therefore difficult, if not impossible.

It is also quasi-systematic that individuals with PWS are presenting trouble closely connected with autism spectrum disorder, such as an absence of figurative meaning or a diffi-

culty of abstraction (Tauber et al, 2015; Duis et al, 2018). Patients with PWS are often showing psychiatric troubles, and for most cases, self-mutilation behavior (scratching) affecting the skin or all the mucous membranes (anal, vaginal, nasal) (Tauber et al, 2015; Duis et al, 2018).

3.1 Behavioral disorders in individuals with PWS

Behavioral disorders are common for individuals with PWS, and those features are different from one to another, but have common aspects (Tauber et al, 2015).

The most common behavioral feature for individuals with PWS reported is food-related behavior problem, such as seeking for food all the time, having food on the mind or any food preoccupation. This is called hyperphagic behavior (Tauber et al, 2015; Schwartz et al, 2021). According to Feighan et al (2020), hyperphagia appeared in 81% of the studied subjects with PWS. Hyperphagic behavior can also lead to food sneaking, eating left-over food from other people's plate, eating food that is normally considered unacceptable, such as food from the trash, raw food or non-food items, or getting up at night to look for food (Schwartz et al, 2021). Anxiety and agitation when food time is coming or when food is seen is also part of the hyperphagic behavior (Tauber et al, 2015; Schwartz et al, 2021). Aggressivity and temper outbursts are also behavioral problems while dealing with food, such as not having access to food can lead to aggressivity. Lying is also connected with hyperphagia, as individuals with PWS are trying anything to get food (Feighan et al, 2020).

Temper outbursts, or tantrums, are common in PWS behavior, as they are "one of the most common maladaptive behaviors reported by parents of children, adolescents, and adults with PWS" (Schwartz et al, 2021). They can be related to food (Tauber et al, 2015; Schwartz et al, 2021), or not. According to Driscoll et al (2017) temper tantrums are part of a characteristic behavior with, among other things, manipulative behavior, compulsivity, and difficulty with change in routine for 70 to 90% of individuals with PWS.

3.1.1 Examples of Autism Spectrum disorders -related symptoms

Autism Spectrum disorders (ASD) is a feature that individuals with PWS may face, and caregivers may have to deal with this issue while caring.

The World Health Organisation (WHO) defines ASD as having difficulties with social interaction and communication and atypical patterns of activities and behaviors.

According to Driscoll et al (2017), many of the behaviors of individuals with PWS are suggestive of autism, while Schwartz et al (2021) are presenting different kinds of behaviors related to ASD, such as anxiety, obsessive-compulsive behaviors, rigidity, or social cognition deficits. Anxiety and obsessive-compulsive behaviors (OCB) are often related, as they are both connected with routines and the need to know, ask, or talk about what is coming, what will happen or what is the plan. Anxiety for people with PWS are mostly related to food, or changes in the routine, which are also connected to OCB, as individuals with PWS may need repetitive questioning, which may be related to resistance to change (Schwartz et al, 2021).

Bennett et al (2015) describe ASD as a neurodevelopmental disorder characterized by repetitive or restricted behaviors. In that way, PWS also shows a behavioral characteristic that overlaps with ASD, even though a study done on individuals with PWS compare with individuals diagnosed with ASD with the Repetitive Behavior Scale-Revised (RBS-R) has shown that PWS group scored significantly lower than the ASD group.

Tauber et al (2015) also indicate the social skills and pragmatic disorders that individuals with PWS can face, such as individuals with ASD. In both groups, individuals are often not showing empathy, or pragmatism as they have a lack of figurative meaning or difficulties of abstraction justifying the importance of conveying messages in writing or visually.

3.2 Physical disorders in individuals with PWS

Physical features and problems connecting to Prader-Willi Syndrome are common for individuals having this disorder, and they also differ from a subject to another, but some features are more likely to appear in many patients. (Tauber et al, 2015)

3.2.1 Growth hormone deficiency: typical example of physical disorder

Growth hormones are hormones released from the pituitary gland, after that the hypothalamus sends a chemical messenger to it, called growth hormone releasing hormone (GHRH). Growth hormones travel through the body to get mainly to the liver, where there is most of the GH receptors. Those receptors send signals to the body to produce the insulin-like growth factors. The main protein that is produced is called insulin-like growth factor-I (IGF-I), and this protein is the one that stimulates new cells growth for muscles' tissues and cartilages (Prader-Willi Syndrome Association, 2011).

Individuals with PWS may suffer from Growth Hormone (GH) deficiency, as their body does not produce enough of certain hormones that are needed for normal growth (Prader-Willi Syndrome Association, 2011). GH deficiency characterized PWS; this deficiency includes short stature, excessive body fat, decreased muscle mass, and impaired quality of life (Grugni et al, 2017). The data regarding Growth Hormonal Deficiency (GHD) in PWS change from a study to another, the prevalence range is from 40% to 100% of individual diagnosed with it (Emerick & Vogt, 2013, Grugni et al, 2017).

Growth hormonal deficiency can also relate to hypothyroidism, as they are both connected to the endocrine system. Children with PWS having GHD do not usually have this height gain during puberty time that other children might have (Aycan & Baş, 2014).

3.2.2 Obesity: most common side effect

Obesity is a possible and common side effect of PWS, as individuals with it face an excessive appetite from childhood to the end of life (Paepegaey et al, 2018., Duis et al., 2018).

Butler et al (2019) affirms that obesity is the major cause of morbidity and mortality for individuals with PWS, and Muscogiuri et al. (2021) are talking about obesity as a hallmark of the syndrome. The excessive appetite and lack of satiety that individuals with PWS are facing lead to complications, such as obesity or diabetes mellitus (type 2 diabetes). Other morbidities connected with obesity for individuals with PWS have been observed, such as metabolic syndrome, syndrome of sleep-apnea (OSAS), respiratory insufficiency, cardiovascular disease and premature mortality. (Muscogiuri et al, 2021)

Muscogiuri et al. (2021) have established nutritional phases that an individual with PWS is usually going through in his/her life, and from their observations, it is after the phase 2, when the patient is around 2 years old, that obesity is starting, as in the phase 1 they are facing poor feeding and hypotonia, in the phase 2 the food interest and progressive increase in appetite begin and lead to weight gain. Obesity, lack of satiety and hyperphagia appear then at school age, by the median age of 8, and last at least until adulthood.

Obesity is also observed for individuals with that syndrome because of the failure of their endocrine system: excess body fat in people with PWS is essentially subcutaneous due to particular adipose tissue (tissues that store energy) in children, and little fibrosis (scar in the tissues) for adults (Tauber et al, 2015).

4 GOAL AND PURPOSE

The goal of this thesis is to offer an evidence-based literature review about health care for individuals with Prader-Willi Syndrome and to put forward different kinds of care for disabled people with Prader-Willi Syndrome. This work is mainly for the caregivers facing work and situations with people affected by PWS. This literature review regroup and organize different studies and research studies regarding this syndrome, thus permitting to caregivers to provide holistic and appropriate care for disabled people affected by Prader-Willi Syndrome, especially by focusing on mental and physical health care.

The research question is: How to provide holistic care for individuals with Prader-Willi Syndrome?

5 IMPLEMENTATION OF THE THESIS

5.1 Literature review

The meaning of this literature review is to regroup and organize other research studies, articles and other sources concerning special concepts and aspects of the care of individuals with PWS. Literature review is a unique opportunity to assess and contrast various arguments and theories, not just summarize them (Cooper et al., 2018).

There are many research studies, articles, websites, and books about PWS, so the purpose was to synthetize and reorganize works and theories about this syndrome in a literature review concerning the special care and needs for people with PWS.

According to Baumeister & Leary (1997), there are different goals in literature review, such as theory development, or description and evaluation of theory, and at the end, the literature review provides a database on the studied subject.

This present literature review is a descriptive literature review, as it contains research method information, and references more information from the abstract and introduction sections (Jaidka et al., 2013), and does not focus on the ‘results and conclusions’ sections. Descriptive literature review can also be defined as a general view of the topic, and it is not restricted with methodology. The chosen topic can be described widely and different aspects of it can be classified if necessary (Salminen, 2011).

5.2 Data collection and selection

To implement this thesis, data collection and selection were done. To search data, search engines were used, such as PubMed, Ebsco, Cinahl and SeAMK finna. Key words for this research were Prader-Willi Syndrome, care, behavior, and word combinations have been used. In addition, conditions to select articles were full-text availability, and less than 10 years old from the publication date. Most of the research were done between January and July 2022, the time that this literature review was written.

Table 1: Inclusion criteria for data collection

INCLUSION CRITERIA
Less than 10 years published articles
Articles in English, French and Finnish
Peer reviewed
Available in full text
Related to Prader-Willi Syndrome
Evidence-based guides or research articles presenting results or recommendations

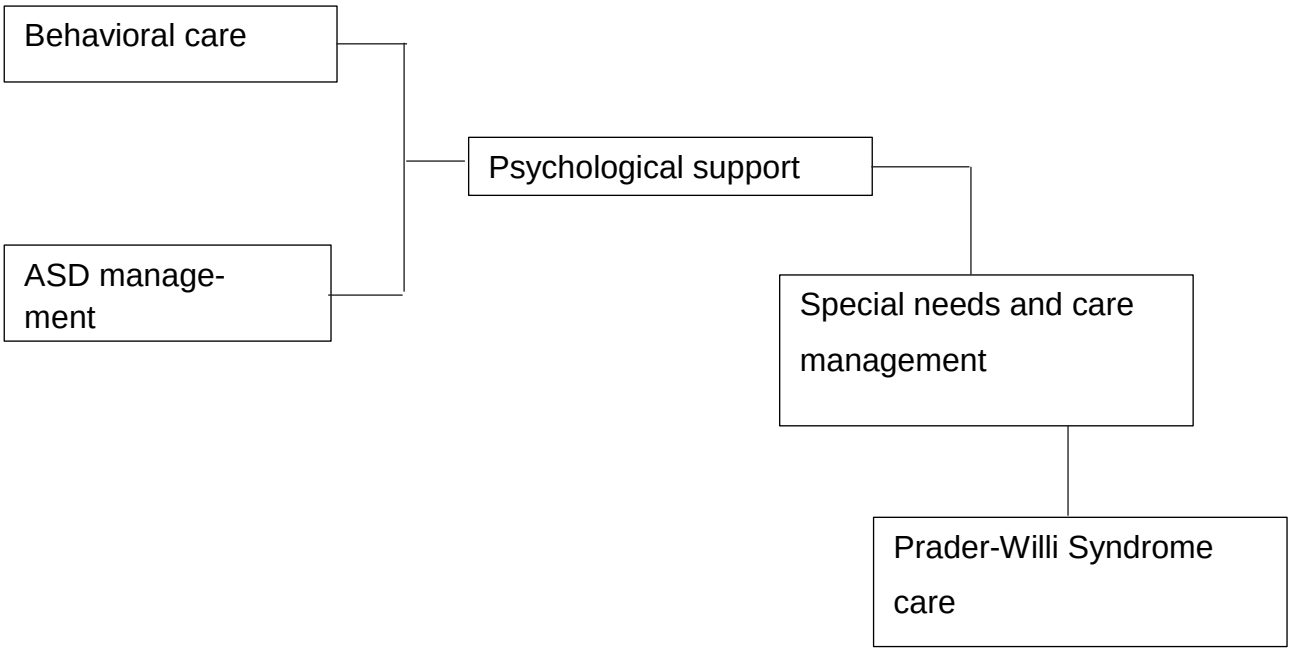
5.3 Analysis

In this thesis, inductive analysis was used as an analysis method. Kyngäs et al (2020) state that inductive analysis is done according to 3 different phases: data reduction, data grouping and the formation of concept, even though there are not accurate rules to analyze qualitative data. However, reading, organizing, forming concepts, themes and categories while analyzing differences and similarities of the selected data to answer to the research question is the aim of the content analysis method.

The chosen articles and research studies have been thoroughly read to get the main points and make the sections and key points of the subject, according to the research question. To analyze and examine the chosen material is important, as it points out the strengths but also the weaknesses of the material, and generally the chosen topic (Oliver, 2012).

After the check of the chosen material, it was important to compare it to see where the resemblances and differences were (Oliver, 2012). At this step, it permits to see if the chosen materials were bringing up the same arguments and results. This also helped to establish the themes and the plan of the literature review. This work helped to determine the relevant hits to write about, according to the research question.

Table 2: Description of analysis process for the thesis making



5.4 Ethicality and reliability

During all the process of making a literature review, the researcher must consider the ethical aspects of this work (Kyngäs et al, 2020). Kent State University (n. d) indicates the meaning of ethical writing by using reliable sources and documenting them, acknowledging many and different perspectives, writing with respect and acknowledgement of diversity and inclusion, by avoiding weaknesses of bias and exclusive language. In addition, reliability term refers to trustworthiness that comprises concepts such as quality, authenticity, and truthfulness of the findings (Kyngäs et al, 2020).

One of the main ethical issues while writing a literature review is plagiarism. As it is based on other's works, copying is a possibility, and it is important for the researcher to sustain an ethical attitude toward people's work but also her/his own work, by using proper acknowledgment and citation, as recommended by the university written instruction. Furthermore, quotation was used, and bibliography was done to ensure the referencing of the material used.

To ensure the trustworthiness of the literature review, reliable databases were used, as well as trustful other sources. In addition, criteria were set up, such as less than 10 years old research articles, full-text availability and examination of the source material references. Guidance from the university and teachers was provided to ensure that ethicality and reliability were met.

Moreover, as the audience of this literature review is caregivers and family, and the aim is to provide guidance to them, the ethicality and reliability must be taken into consideration during all the writing process. In that way, the output will be trustworthy and representative of evidence (Kyngäs et al, 2020).

6 FINDINGS OF THE LITERATURE REVIEW

PWS is a syndrome with a full variety of symptoms, which are changing and differ with the age, the sex or the abilities of the individual bearer of this syndrome. Ho & Dimitropoulos suggest in 2010 that “management of PWS is age-dependent, multidisciplinary, and utilizes a problem-based approach to cater to each individual”, which means that caregivers should work together regarding the care of individuals in PWS and include different therapies and disciplines in the care. PWS is a non-treatable syndrome, but appropriate care improves the lives of individuals with it.

6.1 Psychological support for behavioral care and ASD management

Psychological support is important for individuals with PWS. The quality of life of individuals with PWS depends a lot on the psychological care and support they receive during all their lives (Benchikhi et al., 2019). Yearwood et al (2011) even report that the prevalence of psychiatric disorders for adults with PWS varies from 5% to 15%. That shows the importance of care and support regarding psychologic and psychiatric side of this syndrome.

Behavioral features for individuals with PWS are mostly related to food, but they can also include intellectual disabilities, autism spectrum disorders -related symptoms, obsessive-compulsive behavior, aggressivity, tantrums or self-injuries (Yearwood et al, 2011). Nursing interventions can be done to prevent these behaviors but can also offer for people with PWS a structured environment known for them in advance, with clear expectations from them (Yearwood et al, 2011). Haig and Woodcock (2017) conducted a study about the flexibility level and rigidity in routines for people with PWS. This study shows that the flexibility level was changing at different stages of life, by having more flexibility when growing, but still under certain conditions, such as good reasons to switch from an activity to another (timing reason for example). The same study also shows the rigidness of individual with PWS in routines, with for example mealtimes or going to sleep times, especially during the infancy, with cases with school aged individuals. Rigidity in routines seems to be an important aspect to understand for caregivers to manage the behavior of individuals with PWS (Sethi et al, 2019).

A study conducted by Kayadjanian et al (2021) indicated the importance of behavioral care and psychological support, as the symptom with the biggest negative impact on individuals with PWS was anxiety (which include obsessive compulsive behavior) for group aged 5 to 30 years old. The Prader-Willi Association of New-Zealand has published suggestions for caregivers to help them in the management of anxiety for individuals with PWS (appendix 1), such as use of visual support, schedule to give advance warnings or remember to allow extra processing time.

Individuals with PWS are also facing self-injuries, and caregivers need to offer support but also appropriate care to prevent them. Yearwood et al (2011) and Kim et al (2021) present the skin-picking as a common behavior feature and impute it to obsessive-compulsive bias for individuals with PWS and recommend different ways for the caregiver to avoid them, such as check the skin regularly, keeping nails short and avoid excessive attention on the behavior. The Prader-Willi Syndrome Association of New-Zealand recommends the same interventions in case of skin-picking, but also recommend keeping individuals with PWS busy and use distractions when necessary (Appendix 1).

Caregivers might also face aggressivity and tantrums from individuals with PWS (Yearwood et al, 2011; Kayadjanian, 2021; Ho & Dimitropoulos, 2010). A study conducted by Rice et al (2018) points out that the most effective way to manage a person with PWS being aggressive or having outbursts was to distract them or offer them their own space to calm down. This case of misbehavior can be prevented or minimized. According to the Prader-Willi Syndrome Association of UK and the Prader-Willi Syndrome Association of New-Zealand, some situation can be avoided or managed the way that the person with PWS might accept some arrangement. It is, for example, preferable to avoid anxious situations close to the meals times or when the person is tired. Individuals with PWS are more likely to be anxious when mealtime is coming, so to be able to predict times when anxiety might be at higher level helps to avoid tantrums or aggressive situations. Expose methods for the person with PWS to relax might also help in the management of tantrums, such as to go outside or doing breathing exercises. These methods should be known by the person with PWS beforehand. Furthermore, caregivers and the person with PWS can establish together a ‘calm down plan’ (Prader-Willi Syndrome Association of New-Zealand, n.d.-a). Cooperation with occupational therapists is also useful to help them explain and put into

words their feelings (Benchikhi et al, 2019) Finally, a trustful relationship between the caregiver and the person with PWS is a key in the management of care (Prader-Willi Association of France, n.d.-a).

6.2 Physical care for obesity management and GH treatment

Obesity is the main problem that individuals with PWS are facing (Yearwood et al, 2011; Crinò et al, 2018), but there are keys and solutions to avoid this side issue of this syndrome.

Obesity management is possible by preventing it. Erhardt and Molnár (2022) published a study on possibilities of prevention of obesity for individuals with PWS. The control of the calory intake is cited as the main preventive method for weight management for individuals with PWS. The second preventive method exposed by Erhardt and Molnár (2022) is to have regular physical activity. In general, authors recommend that calory-intake control, regular physical activity, and behavioral interventions are the most effective methods for weight management of individuals with PWS. As the appetite behavior changes because of going through different nutritional phases, caregivers of individuals with PWS have to follow and make sure the calory-intake is respected (Erhardt and Molnár, 2022; Kim et al, 2021). From the first stage of life, babies and kids have poor appetite and are uninterested in food (Yearwood et al, 2011, Kim et al, 2021), so it is important to make sure that the calory-intake is enough for muscular development, as the main dietary goal of this period is to promote optimal growth and avoid obesity development (Erhardt and Molnár, 2022).

The management of physical conditions of individuals with PWS goes with the food stealing management. Gourash and Forster (n.d.-a) established a method called ‘Food Security’ for people with PWS. In this method, they consider that if a person with PWS did steal food, it would mean that there is some failure in the nursing care plan. The person with PWS who has been caught to steal food cannot be blamed for his/her act, but caregivers have to revise the nursing care plan again, to make sure that the situation will not happen anymore. For individuals with PWS, it is useless to try to teach them not to steal food, and even if it would somehow succeed, it would probably cause a certain stress that the consequences would be seen later on (tantrums, anxiety) (Gourash and Forster, n.d.-a).

The food security method of Drs Gourash and Forster consists of teaching at an early age individuals with PWS the principle of “MY food” and “YOUR food”. In 2009, Gourash and Forster published a work on how food feels for individuals with PWS, and it was egual to stress for them. Caregivers should not put a lock and prohibit people with PWS from having access to food but should put a lock on their thought of food. The ‘food security’ method of Gourash and Forster consists of 3 points: no doubt, which means that a person with PWS will know when the meals are and what they are going to eat, no hope (or no chance), which means that the person with PWS is not going to have other opportunities to eat what was planned or no other opportunities for having access to food, as it can cause stress and lead to over-eating. Finally, the strict respect of these 2 points lead to the 3rd one: no disappointment. The fact that the person got only what was known at expected time and did not get any extra expectations provide her/him security regarding food. To achieve the food security, caregivers must follow, carry and go with the person with PWS. There are simple ways to also bring this food security to them such as controlling and securing the access to food, avoiding any spontaneity related to food (no suggestions or promises of extra food, or no “surprise” related to food) and food exposure is always supervised (in markets, restaurants, or parties). Food cannot be used as a threat, and to make appropriate food choices is supported and encouraged by caregivers (Prader-Willi Syndrome Association NZ, n.d.-a; Gourash and Forster, n.d.-a, Gourash and Forster, 2009).

Growth Hormone treatment plays a role in weight management of individuals with PWS. Kim et al (2021) established a link between the use of growth hormone treatment and management of weight for individuals with PWS, as it increases muscle mass and reduces body mass index (BMI). However, GH treatment does not have any effect on food-seeking behavior or does not reduce appetite. Bridges (2014) explain that one of the main benefits of GH therapy is the improvement of physical capacity and endurance of individuals with PWS under this treatment, which can help to prevent obesity, as they are more likely to do physical exercise. Another study reports the effectiveness of GH therapy on height, body conditions and compositions, socialization and cognitive functions for children and adults with PWS (Höybye et al, 2021). The role of the caregiver regarding GH therapy is to make sure that individuals with PWS are taking their treatment and explain why it is important to

follow-up this specific therapy. In case of obesity, the use of GH is not recommended anymore (Hirsch & Gross-Tsur, 2021).

7 DISCUSSION

The main aim of this literature review was to provide a good and clear evidence-based work for caregivers of individuals with PWS. The purpose of the thesis was to make a literature review, and thanks to an evidence-based work, provide a guide for caregivers facing work with PWS individuals.

The literature review was done in cooperation with Lehtimäki Education Centre, a special school for people with disabilities and special needs, as individuals with PWS. As caregivers in Lehtimäki Education Centre might work with individuals with PWS, it felt important to provide an evidence-based work on how to provide a holistic and appropriate care for them to also implement the work of caregivers.

During all the work process, it was important to talk about the medical side of the disease, because as a caregiver, it feels important to understand a disease or a syndrome fully, and understand some special characteristics of it, and why some phenomena happen. In the case of PWS, and because this literature review was done in cooperation with Lehtimäki Education Centre, it was important to give all the keys for the caregivers there, and then for caregivers in general, to help them in their work and to provide the special care that individuals with PWS need. That is the reason why the medical aspect of the disease is taken part in this literature review, because, as the author of it and as a caregiver, it felt important to have all information about PWS to make sure I understand the disease and I understand them as a whole person, and I am able to give an appropriate answer and care for Prader-Willi Syndrome affected people.

8 CONCLUSION

Prader-Willi Syndrome is a complex disorder, from a medical point of view, but also from a nursing point of view. An early diagnosis is important in this syndrome, to offer the best appropriate care as soon as possible. After diagnosis has been established, caregivers have to face it as much as the person receiving this diagnosis, and this because individuals with PWS need a lot of help in everyday life, especially facing anxiety, tantrums or food-related behavior.

Anxiety, tantrums, food-related behaviors or self-injuries are common features for people with PWS and require appropriate care from caregivers. To provide holistic and appropriate care for people with PWS, caregivers need to have information about the disease, especially about the secure feeling that individuals with PWS need. This literature review has been done in cooperation with Lehtimäki Education Centre, a special school offering also accommodation and special-need care for disabled people. As caregivers could work with people with PWS to know about the management of this rare syndrome could help them to provide appropriate and holistic care to eventual clients.

There are some methods available for the caregivers that can help people with PWS, but also caregivers themselves, and which give secure feeling for everyone. Individuals with PWS mostly need to feel a secure environment, rigidity in routines or with timetables, and it helps to avoid anxiety and tantrums. It is important for the person taking care of them to understand that, and in that way, it makes the care easier. To know also about the food and meals related situations is important and give them the secure feeling that is needed, and help to avoid over-eating, food stealing problems or eating inappropriate food. As people with PWS have high risk of obesity, the care and management of their physical condition is important, by managing their food, but also their physical activities. An important point to keep individuals with PWS feel secure and not anxious about food or anything else is to keep them busy. Finally, to have their own space appeared to be also important in the management of PWS.

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APPENDICES

Appendix 1. Behavior management for PWS

SUPPORTING PEOPLE WITH PRADER-WILLI SYNDROME AND THEIR FAMILIES

Prader-Willi Syndrome Association (NZ) Incorporated

PO Box 258, Silverdale, North Auckland 0944 www.pws.org.nz

BEHAVIOUR MANAGEMENT FOR PWS

BEHAVIOUR	MANAGEMENT STRATEGIES
Preference for routine and predictability is typical and therefore individuals often thrive in environments where activities are timetabled. However, their need for consistency also means that they may not cope well with changes in expectation (theirs or yours).	Clear visual schedules and outlines of expectation are helpful and reassuring. Provide forewarning of possible changes; avoid sudden changes and rushing. Allow time to process and accept changes. Avoid making promises that can be broken. Try to avoid vague, ambiguous answers such as 'perhaps' or 'we'll see' which increase anxiety. Avoid open-ended questions or too much choice. Reward flexibility.
Task switching is a common area of difficulty with individuals feeling compelled to complete tasks. Support is required during task switching to reduce anxiety which may manifest in frustration.	May respond well to rituals and visual cues or timetables. Use verbal and visual countdowns. Modify tasks so that there is less to complete. Schedule activities that are typically difficult to stop before a motivating activity, i.e. lunch.

	• Use incentives but hurrying a person with PWS can make matters worse.
Obsessive thinking or perseveration Individuals with PWS can think or talk obsessively about a topic of interest. Their thoughts can become stuck on a subject or issue. Perseveration or repetitive questioning can be a sign of stress. It may be useful at times to channel obsessive behaviour into positive activities.	• Bring a topic to a resolution. • Set limits for the amount of time they can talk on a topic. Do not provide more information than is necessary and avoid grey areas which can raise anxiety. Avoid providing information too far in advance. • Answer questions specifically (not with maybe / later), check understanding by asking them to repeat answer, limit the number of times a question can be asked. Offer a range of appealing alternative activities or a variety of people to meet / spend time with. • Use diversionary tactics.
Rigidity in thinking is common. Rigid thought processes mean that information is stored in an orderly manner and there is a strong need for routine and consistency. Once something is learned it can be seen as the only 'correct' answer or method and it is difficult to have a change of mind or relearn. Rigidity can lead to stubbornness and argumentativeness.	• Activities which encourage more flexible thinking from a young age may help. • Praise flexible thinking. • Encourage an understanding that there may be differing opinions, perspectives, answers or ways of doing things. Individuals with PWS value authority figures but when they 'authorise' information, it can become set in stone and if incorrect, it can be difficult to convince a person with PWS of this. On the other hand, their response to authority figures can be useful.
Non-compliance An oppositional response can be an automatic 'flight or fight' response when an person is feeling anxious. Individuals with PWS are known for their ability to dig their heels in and be extremely stubborn. Sometimes stubbornness may be a coping device when there are genuine reasons for not wanting to do something, but they cannot find a way to tell you. They may not be consciously aware	• Provide limited preferential choices to allow a sense of control over decision making. • Create clear rules, boundaries, consistency and a calm environment. • Avoid just saying 'No' and confrontation. Negative reactions raise anxiety further. • Resolve issues with compromise, finding a new solution together. Avoid ultimatums. Offer empathy but repeat your expectation, then ignore unwanted escalation behaviour as much as possible.

of their reasoning. It may be that something has gone wrong earlier in the day which is a pre-cursor to the avoidance behaviour. Fortunately, individuals with PWS are eager to please, enjoy consistency and following rules. Routines and rules reduce anxiety. Occasionally, manipulation, lying or confabulation can be additional problems. Lying is an abstract concept and pupils with PWS may have difficulty in recognising what they say as lying.	Allow processing time for 'coming around' to a new way of thinking or an agreement. Set a limited number of clear, positive goals for expectations of compliance and reward success. In the event of extreme stubbornness that puts a person in danger, help them 'save face' by saying you need their help or try to move them on by offering a more pleasurable activity. Focus on blame or recrimination fosters an atmosphere where manipulation or lying can flourish. Ensure good team communication so that lying is identified and a behavioural pattern is prevented.
Tenuous emotional control is due to the impaired hypothalamus having poor control over emotional responses. Individuals can be volatile, becoming easily upset or frustrated. **'Meltdown'** can be caused by sudden changes in routine or expectation, increased anxiety, pressurised tasks, confusion, sensory overload, communication	Avoid false expectation and disappointment. Acknowledge feelings and encourage communication about anxieties. Use tools such as feelings cards or the feelings thermometer. Develop their understanding of how situations affect their emotions, 1. I can handle this. 2. This makes me uncomfortable. 3. This makes me nervous. 4. This can make me mad. 5.This can make me lose control. Help them to identify their feelings and teach ways of coping with stress, i.e. using stress balls, listening to music, taking a break, relaxation. Praise efforts to control emotions, especially in difficult situations. Have a 'calm down' plan where a person may remove themselves to a safe area to calm down. The plan may include the availability of prepared calming activities. Monitor for signs of an impending meltdown, learn to recognise triggers, signs and take preventative steps. Stay calm and try distraction, perhaps humour. Avoid discussion and reasoning during a meltdown; reflect afterwards.

difficulties, teasing, frustration, use of their possessions or other triggers.	
Lack of impulse control means that individuals will not self-monitor their behaviour well. They are more likely to engage in risky or dangerous behaviour.	Discuss situations and environments beforehand, what your expectations will be and what will happen if a lack of self-control occurs. Encourage self-monitoring and evaluation. Offer reminders of their evaluations so they can focus on what they need to control / improve.
Aggressive behaviour is rare but occasionally emotional outbursts can escalate to physical aggression.	List strategies a person can use when they are feeling anger, such as going for a walk, counting or rehearsed breathing exercises. Have a plan for keeping the individual and others safe in the event of an aggressive behavioural episode. Predict times when anxiety will be higher and emotional control is likely to be reduced, such as proximity to meal times, when tired, if teased.
Sensory processing difficulties Individuals with PWS may be over-sensitive to certain stimuli or under-sensitive sensory seekers, or both. Sensory processing difficulties often improve with age. They do not	Consider seating for comfort and noise distraction. Transmitters and headphones enable ability to block out other sounds and focus. Avoid talking too much and information overload. Hand fidget tools may improve concentration.

respond well to chaotic environments with too much stimuli or too little structure.	- Earplugs can reduce hypersensitivity to noise.
Common sensitivities are loud noises, smells, movement, balance, touch and clothing.	- Hypersensitivity to fluorescent lighting may exist. Pre-warn of high stimulus environments. - Provide space in crowded situations. - Water is often calming and stimulating. - A sensory diet, including sensory gyms and sensory boxes are useful developmental and management tools. - A quiet, sensory smart area is helpful. - New therapies target spacial awareness with vestib-ularvisual-auditory approaches involving listening programmes.
<u>Poor social skills</u> Individuals with PWS are friendly and need friends, although they often lack the appropriate social skills to form and maintain healthy friendships.	- Identify what is confusing about conversations, i.e. eye contact, turn taking, listening, expressing thoughts and responding to humour etc. - Teach, model, role-play conversational skills such as listening to what others say, thinking about what they say, putting thoughts into words. - Practise conversations with topics of interest. - Use social stories for situations such as making friends. - Individuals could engage with a social skill group in activities that are of interest to them.

	▯ Support individuals during activities with peers. ▯ Clear rules may be needed about social appropriateness and in particular for interactions with the opposite sex.
Self-harming behaviour can take the form of nail biting, skin picking, teeth grinding, teeth, hair or eyelash pulling and varies greatly in severity. It is usually a repetitive habit which worsens with boredom / disengagement and may be a form of self-stimulation. Sometimes it can even be to provoke a reaction or seek attention. Less frequently, it may be the result of anxiety or emotional distress. It is hard to stop due to difficulties in thought switching and lack of impulse control. Impaired neurological signalling also means pain or disgust may not be felt as intensely.	▯ Use distraction and redirection. ▯ Keep engaged and / or their hands busy. ▯ Various lotions can moisturise wounds making them less tempting to pick. ▯ Dress wounds and keep nails short. ▯ Avoid giving attention to the behaviour – positive or negative attention. ▯ If the behaviour is sensory seeking, provide more socially appropriate forms of stimulation, although avoid linking to the behaviour in case it rewards it. ▯ Although the behaviour appears compulsive, medicines targeting OCD or anxiety often prove unhelpful. A natural supplement called PharmaNAC (available in the USA) has proven beneficial.
Increased risk for mental health issues Anxiety disorder is very common. Compulsive behaviours such as skin picking, hoarding and concerns with exactness are often seen in PWS, but some individuals will have a dual diagnosis of OCD. Oppositional defiance and problems with aggression can also be present. Occasionally, extreme impulsivity or attention deficit leads to a separate diagnosis of ADHD. There are also increased rates of 'thought' problems in PWS (seeing things / hearing voices / strange ideas). In rare cases, behavioural episodes can become more extreme and evaluation for psychotic disorders may be needed.	▯ Possible interventions for psychiatric illness in PWS may involve supplements, medication and psychological strategies. ▯ Little is known about the early phase of illness and risk factors that predict the emergence of psychosis in PWS. Research is ongoing although it is known that earlier intervention leads to improved health outcomes. **(The management of food related anxiety and food seeking behaviour is covered separately.)**