

Identification of Psychosocial Practices
and Interventions that Support the
Adaptation and Rehabilitation of Adults
with Prader-Willi Syndrome

EXECUTIVE SUMMARY

Health Technology Assessment Report

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PROBLEMATICS

Prader-Willi syndrome (PWS) is a genetic disease that affects one in 15,000 to 25,000 newborns and affects children of both sexes regardless of geographic origin (*American Academy of Pediatrics*, 2011). It is a genetic defect that causes inactivity of part of chromosome 15 (*Prader-Willi Research Canada Foundation*, 2018). The mechanisms explaining the symptoms associated with PWS, including hypothalamus dysfunction that regulates hormonal functions and certain behaviours, are still unknown to date (*Whittington and Holland*, 2017). However, although the severity of PWS manifestations varies from person to person, a range of physical, neurological, developmental and behavioural characteristics can be observed. In particular, people with PWS do not receive normal signals of hunger and satiety: they are unable to control their food intake so that they develop binge-eating and they adopt behaviours related to the search for food (*International Prader-Willi Syndrome Organisation*, 2018). PWS is also associated with behavioural disorders, obsessive-compulsive disorders and psychiatric disorders (*Encyclopedia Orphanet General Public*, 2013). However, a high variability of these events is observed (*Whittington and Holland*, 2017).

CONTEXT

Since June 2011, a continuous assistance residence (CAR) at the Centre intégré universitaire en santé et services sociaux (CIUSSS) de la Capitale-Nationale located in Charlevoix has been offering rehabilitation services exclusively to adults with PWS. After six years of operation, an assessment of rehabilitation and nutrition services has highlighted the need to update the service offer with evidence-based data. For this purpose, the HTA Unit of the CIUSSS de la Capitale-Nationale, in collaboration with the HTA Unit of the CIUSSS de la Mauricie-et-du-Centre-du-Québec, conducted a HTA.

GOAL

This HTA aims to identify psychosocial practices and interventions that support adaptation and rehabilitation in adults with PWS. The results will help to update the CAR Intervention Program of the CIUSSS de la Capitale-Nationale for this clientele, as well as to improve their service offer

> For more information, see the report at:
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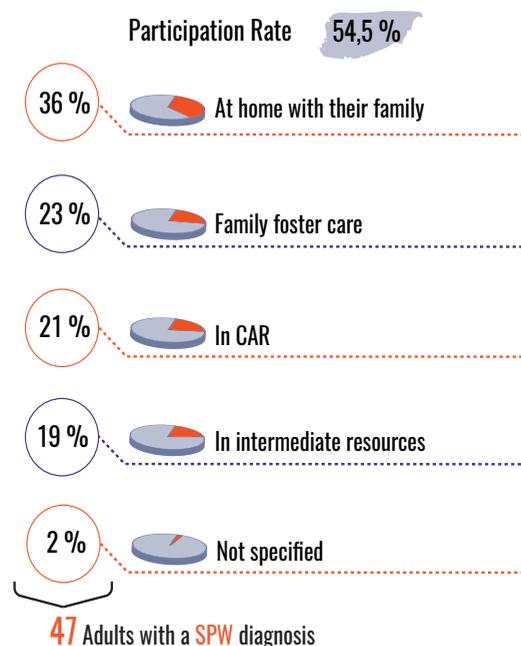
METHODOLOGY

A systematic literature review was conducted in seven bibliographic databases. In order to recreate other to identify relevant documents in the grey literature¹, 32 Websites were consulted and the *Google* search engine was used. A first selection was made from the titles and abstracts, then a second one after a thorough reading of the selected documents, according to inclusion criteria established *a priori*. The methodological quality of the studies and the credibility of the grey literature documents were assessed using validated tools. Relevant data were collected in a structured extraction grid, synthesized and analyzed. In addition, a portrait of the living environments and services offered to adults with PWS was produced in Quebec’s CISSS and CIUSSS.

RESULTS

The literature search identified 3553 eligible documents, 54 of which were selected². Among the 11 studies from the scientific literature, ten of them involve interventions and one concerns the organisation of services (risk management and promotion of autonomy). Of the 43 documents from the grey literature, considered as experiential data, 18 aim living environments and service provision; 16 involve intervention models and strategies; five concern professional integration; two target users’ rights and the use of restrictive approaches; one concerns the relationship between members of the working team and parents/caregivers and one involves the adults with PWS in France

Data obtained in Quebec’s CISSS et CIUSSS



The data obtained in Quebec’s CISSS and CIUSSS, with a participation rate of 54.5% (12/22), identified 47 adults with a PWS diagnosis: 36% live at home with their family, 21% in family-type resources, 21% in CAR, 19% in intermediate resources and 2% in an unspecified environment. Only the CIUSSS de la Capitale-Nationale has a residential environment specifically designed for adult with PWS

Data show that 76% of the adults listed with PWS receive services in their living environment. Among them, more than 50% receive services from a specialized educator, a social worker or a nurse. Finally, 67% of them participate in professional and residential integration programs.

1. Grey literature is “any type of document produced by government, administration, education and research, trade and industry, in paper or digital format, protected by intellectual property rights, of sufficient quality to be collected and maintained by a library or institutional archive, and which is not controlled by commercial publishing.” (Schöpfel, 2012)

2. The list of bibliographic references of the selected documents is available in the report.

FINDINGS

Psychosocial Interventions and Practices

- › Of the 10 evaluated interventions listed in the scientific literature, the majority use a cognitive-behavioural approach.
- › The approaches evaluated are different (cognitive-behavioural, reinforcement, motivational, behavioural analysis with support plan) and the components worked on are multiple (challenging behaviours, anger crisis, PICA, food theft, self-harm, social skills and pro-social behaviours, activities of daily living).
- › The results of the 10 interventions evaluated are variable and inconclusive.
- › To achieve and maintain the desired behaviours, it seems that the reinforcements used (token system, visual cards, etc.) cannot be removed.
- › Positive behavioural strategies are an interesting way to intervene with people with PWS.
- › Interventions should aim to prevent and avoid behavioural disorders
- › It is important to analyze and understand behaviours and their context.
- › Food has a highly reinforcing effect and is a source of stress and anxiety.
- › Programs and interventions must be adapted to the needs and particularities of each individual with PWS: the application of a single program or intervention does not seem possible for this population, since individuals differ greatly from each other.
- › The acquisition of full autonomy (not receiving help or support) by a person with PWS does not seem possible, regardless of his profile. Although the situation may vary from one individual to another, the person will always need services. The capacities of people with PWS may be overestimated to the detriment of their health and well-being.

Clinical, Organizational and Architectural Requirements

- › Members of work and management teams working with people with PWS must be highly qualified in the specific issue of PWS (not only in intellectual disability).
- › Residential staff must receive specific training from the PWS (not only in intellectual disability).
- › Experienced professionals must be available 24 hours a day, 7 days a week to provide direct assistance to residents in case of need.
- › Accommodation facilities must be adapted to the condition of people with PWS and ensure food security.
- › Accommodation facilities should encourage and promote recreation and activities in small groups and individually.
- › Several associations on PWS are widely cited around the world: there is a willingness to network, document and share information about this rare genetic disease through the creation of associations

CIUSSS DE LA CAPITALE-NATIONALE CAR RECOMMENDATIONS FOR ADULTS WITH PWS

Psychosocial Interventions and Practices

- › According to the grey literature and the opinion of the experts on the monitoring committee, the multimodal analysis and intervention approach for intervention strategies and crisis prevention (MMAI) and behaviour management in crisis situations (Therapeutic Intervention in Aggressive Behaviour (TIAB), OMEGA-plus) are to be preferred for adults with PWS.
- › In order to prevent and avoid behavioural problems, it is necessary to analyze and try to understand the residents' behaviours.
- › For the well-being of the clientele and given the highly reinforcing effect of food in people with PWS, the use of food rewards should be phased out.

However, the special snack, which is not a reward, could be maintained for its social nature.

- › The use of a non-food reward system to motivate and maintain good behaviour should be promoted and maintained.
- › Although the results from the literature are not generalizable and that there is not a single program, some of the interventions and strategies identified could be attempted with consultation with stakeholders, taking into account the characteristics, needs and capacities of each individual with PWS.

Clinical, Organizational and Architectural Requirements

- › The members of the working team should be highly qualified in the specific issue of PWS. An intellectual disability training would not be sufficient.
- › All staff (clinical, management and support) should receive initial training in PWS before working with clients, as well as ongoing training. These trainings should include:
 - › Multimodal analysis and intervention approach for intervention strategies and crisis prevention (MMAI);
 - › Behaviour management in crisis situation (TIAB, OMEGA-plus);
 - › Nutrition, diets and meal preparation;
 - › Rights, responsibilities and restrictions;

- › Health and medical problems specific to people with PWS;
- › Specialized medical needs and supports.

- › Maintain the availability of an experienced professional (member of a professional order) 24 hours a day, 7 days a week to provide direct assistance to residents when needed.
- › Maintain the following professional services: dietitian, psychoeducator, social worker, nurse, occupational therapist and physiotherapist.
- › Maintain and increase the number of hours allocated to the nutritionist.
- › Residents may receive the services of a kinesiologist to review exercise programs according to their needs and particularities.

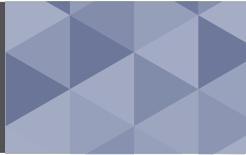
- › Maintain opportunities and formalized forums for exchange and clinical support for CAR workers, namely monthly clinical-administrative meetings.
- › Maintain exchanges between CAR educators when changing shifts.
- › Maintain adequate and safe food facilities in the CAR (with secure or inaccessible kitchen and storage areas).
- › Set up interprofessional collaboration meetings.
- › Given the limited literature on the subject of psychosocial interventions and the scarcity of residential facilities only for people with PWS in Quebec and elsewhere: document and assess the practices of the CIUSSS's CAR de la Capitale-Nationale for adults with PWS.
- › Update the CAR Intervention Program of the CIUSSS de la Capitale-Nationale for adults with PWS and develop a leading innovation practice.
- › Develop training content supported by an up-to-date «Client Program», documentation, practice evaluation and knowledge of expert practitioners in the community.
- › Given the existence of several associations on PWS around the world and considering that it is a rare genetic disease: these groups could be used to network, to share information and to promote local practices

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