

Preface

The suggestion to address the topics of wellbeing and quality of life for people with Prader-Willi syndrome took place as part of the International PWS Conference of the International PWS Organization (IPWSO) in Havana, Cuba in 2019 where aspects and perspectives of the topic were discussed from different professional perspectives. Thereafter, it increasingly became more and more the focus of discussions within the international “PWS world”.

The concept not only enables the subjective wellbeing of people with PWS to be depicted as criteria for the current quality of life of the person with PWS, but it also offers the opportunity to evaluate the quality of support services for people with PWS and to compare specific PWS services with one another.

From the author's point of view, however, the criteria can only be useful and reliable if the concept is described sufficiently clearly and comprehensibly and if possible, applications for the everyday care of people with PWS can be derived and described. The aim was to present the explanations in an understandable manner for the group of people who are involved in the everyday care of people with PWS. These are primarily relatives of people with PWS (parents) and employees (caregivers) in residential and work facilities for people with PWS.

Therefore, in addition to describing the individual elements that make up “wellbeing” and “quality of life”, the second and perhaps even more important aim of this book is to present possibilities and methods for improving the subjectively perceived wellbeing of people with PWS.

The present text attempts to achieve both goals, a description of the concept - “subjectively perceived wellbeing of people with PWS”, and the presentation of possibilities and methods to improve the level of “subjectively perceived wellbeing of people with PWS”.

I am very interested to learn about the experiences of people with PWS, their parents, relatives and caregivers for people with PWS in relation to the contents of this book and look forward to comments and feedback.

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1 Introduction

In caring for and supporting people with Prader-Willi syndrome, Driscoll, DJ (2023), Soyer, H. (2003), Hödebeck-Stuntebeck, N. (2012), Angulo, MA, et.al. (2015) it is the common goal of everyone involved to want to achieve “the best” for this group of people and for the individual person with PWS. It doesn't matter whether they live at home or in a supported living arrangement.

If you ask for a specification of this very general objective, it is often described that you want to enable people with PWS to have a high quality of life and to “feel good” where they live and the way they live.

This goal is stated by people with PWS, but also by their relatives, legal representatives or employees who work in the care of people with PWS.

Improving the quality of life is also a goal among governments, whether they are responsible for financing services in residential care homes or supporting living situations, in the work sector, in outpatient support settings, in supporting in the parents' home or in the own home of people with PWS.

Within the framework of this general objective, there is a great deal of agreement among all representatives listed here.

However, when it comes to specifically defining or describing what is meant by wellbeing or quality of life, the problem is often that each party or person involved has a very subjective idea and definition of the concept of wellbeing or quality of life. These definitions or interpretations are often driven by self-interest.

However, it is also the case that some of those involved have no or only a very vague idea of what is meant by quality of life or subjectively wellbeing.

In everyday life, these different ideas or the lack of concrete ideas often lead to very different and often contradictory actions or goals being pursued in order to achieve the goal of improving wellbeing or quality of life, for example: cost effectiveness - subjective wishes and demands for faster implementation - clinging to the status quo out of fear of change - demanding funding for all ideas -...

Many interpretations are certainly understandable from the respective perspectives, positions or given circumstances. In practice, their sometimes

contradictory approaches lead to massive conflicts and thus to the loss of resources in the care and support of people with PWS.

Unfortunately, it is not always ensured that the person with PWS understands or can understand the meaning or goal of the actions of the actors in the environment “to improve their quality of life, their wellbeing”. Often the suggestions put forward are too abstract and lead to the person with PWS not understanding the content.

In addition, people with PWS are often not included in considerations and planning.

Not understanding and not being included often results in very helpful and positive measures and supportive actions by providers, parents or other people being rejected by people with PWS, sometimes with severely challenging behaviour.

The statements in the following text are based on the assumption that improving the “subjectively perceived wellbeing of people with PWS” or quality of life for people with PWS should be the focus of care and support - and that “subjectively perceived wellbeing of people with PWS” are the criteria for providing good quality of care.

In order to be able to implement this goal, it is necessary to clearly define the concept of “subjectively perceived wellbeing of people with PWS” or quality of life of people with PWS” and make it applicable to everyday care so that everyone involved can carry out their activities in this regard with a fuller understanding of the reasons why they are doing them.

The existence of a definition of the concept of “subjectively perceived wellbeing of people with PWS” makes it possible to examine the quality of support for people with PWS, as well as comparing existing services for the care and support of people with PWS.

2 “Subjectively perceived wellbeing of people with PWS” and “quality of life”

So far, the two terms “subjectively perceived wellbeing of people with PWS” have been used interchangeably.

However, the term “quality of life” is usually broader than the term “wellbeing”.

In addition to personal aspects, “quality of life” also includes socio-economic factors such as: living environment, financial situation, school education, etc.

The term “subjectively perceived wellbeing of people with PWS”, on the other hand, focuses on the individual person and their subjective perceptions.

How a person is acting and feeling is derived from their subjective perception of a situation. This is a very important element in determining a person's behaviour.

The components of the term “subjectively perceived wellbeing of people with PWS” determine the orientation of the care and support of people with PWS:

- **Subjectively**

Ultimately, the only thing that is relevant and effective for action is what level of wellbeing the person with PWS derives and experiences from their subjective perception of the current situation and not what people in the environment assume about how the person is currently feeling.

- **Perception**

Viewed from the outside, every situation has many stimuli or components. What is crucial, however, is which of these stimuli the person with PWS actually perceives from the current situation. This can differ significantly from the perception of the actors in the environment.

- **Wellbeing**

Wanting to feel good (wellbeing) is the fundamental goal of all human endeavors, including all of the aspirations of people with PWS.

Understanding, elevating and improving this “subjectively perceived wellbeing of people with PWS” is the intention of this book.

3 Theoretical derivations of the concept of “Subjectively perceived wellbeing of people with PWS”

The definition of “subjectively perceived wellbeing of people with PWS” used in this text was first put forward in the theories of Martin Seligman (2015) and Barbara Fredrickson (2001).

Seligman and Fredrickson are co-founders and relevant representatives of Positive Psychology and consequently derive their assumptions from this approach, which found its origins in 1954 with Abraham Maslow, who coined the term.

The core and driving force of positive psychology represents the “turn away” from the orientation of (traditional) psychology, which was strongly oriented towards the pathological parts and deficiencies of the individual person .

Until then, a lot of psychological resources had to be invested in changing or "healing" a person's negative, abnormal or deviant behaviour and the focus was on this. Since the acceptance and establishment of positive psychology, the focus has become the identification and awareness of the person's abilities.

Resources are used to develop and expand a person's positive skills and characteristics, combined with the expectation and experience that the development of these areas will lead to higher levels of competency for the person to deal with existing challenges and to a higher level of subjectively perceived wellbeing of people with PWS.

Following these basic assumptions, Seligman (2015) formulates five elements (which he combined to form the PERMA model) that represent the subjectively perceived wellbeing of people with PWS and ultimately allow the subjectively perceived wellbeing of people with PWS to be recorded and evaluated.

According to Seligman, each of these elements meets the following three criteria in order to be viewed as an independent element:

1. The implementation and achievement of the goal of the respective element contributes to the (subjective) wellbeing of the person.

2. The element is achieved for the sake of the thing itself and not in order to thereby achieve or enable the goals of other elements. Each element has an independence for the sake of which it wants to be achieved.
3. Each element can be defined without the help of the other elements.

From the perspective of the author of this text there is another relevant criterion:

4. Every element can be captured (mapped).

The practical application of the five elements presented below will make it possible in the future :

- to orientate the development of concepts for the care of people with PWS on the 5 elements,
- to carry out a qualitative assessment of existing PWS (housing) services based on the 5 elements,
- to carry out a comparison of PWS support situations (living, working, leisure, ...) based on the 5 elements.

4 Description of the five elements

4.1 Element 1: Positive feelings

Example

A mother visits a zoo with her 14-year-old son J. with PWS, at his request.

The mother knows from previous visits to the zoo that her son loves visiting the llamas in the zoo. He beams “from ear to ear” when he can watch them and then pet them too. He then often waits for a long time until the llamas come close enough to the fence of the enclosure so that he can pet them.

During today's visit, the mother can see that her son hardly enjoys the animals they see while walking through the zoo.

But as they approach the llamas' enclosure, the mother can see the increasing anticipation on her son's face. Then mother and son sit in front of the enclosure for half an hour until one of the llamas approaches the boundary of the enclosure. The son approaches the llama and can finally pet it. The mother observes how the son only has eyes for the llama and how he strokes the llama for five minutes with a smiling face. The mother describes her son as “blessed” in these moments.

The human body reacts to certain situations and stimuli in the environment with an “automatic” physical reaction that the person is usually not aware of. This reaction often arises from the interplay between the person's genetic makeup and environmental factors. It can also be learned.

The boy's reaction described above involves physical changes, for example in blood pressure, pulse rate, hormonal balance and the excitation of certain areas of the brain when he sees and strokes the llama.

Only when the person (here: the boy) consciously perceives these physical reactions (emotion) does the body's reaction become a feeling: here joy (in other situations, the other basic feelings: sadness, anger, fear, disgust can occur).

Physical reactions are the unconscious part, feelings are the consciously perceived part of the reaction to certain stimuli in the environment.

If you now look at **positive feelings**, they are, according to the psychological - dictionary defined as pleasurable, pleasant sensations. (Häcker, Stapf, 2004, p. 721).

According to Frederickson (2001), positive feelings can be expressed in the experience and conscious perception of: joy - cheerfulness - hope - gratitude - awe - inspiration - interest - love - fun - pride - desire - delight - ecstasy - warmth - comfort,

What is crucial is that a positive feeling is actually consciously perceived by the person. In relation to the example above, the boy actually notices the joy of being petted by the llama. There may be situations in which people assume that the person “must have a positive feeling” because they can pet the llama, as in the example above. In fact, the person is not aware of the feeling.

Only if the person consciously experiences this positive feeling in the moment, the element “positive feeling” is realized as one of the five elements in the person and is the “positive feeling” effective as an element in the sense of part of the person's wellbeing.

These statements once again underline why the term “subjectively perceived wellbeing of people with PWS” was chosen.