

THE DEVELOPMENT OF A THERAPEUTIC APPROACH FOR THE TREATMENT
OF INDIVIDUALS WITH PRADER-WILLI SYNDROME AND THEIR PRIMARY
CAREGIVERS

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INTRODUCTION

- The Lack of knowledge, inadequate literature on the psychological impact of PWS, and limited studies conducted in South Africa – motivated this study

BACKGROUND INFORMATION

- Prader-Willi Syndrome (PWS) has been described as a neurodevelopmental genetic disorder which results from a genetic mutation involving chromosome 15
- The estimated incidence: 1 in 15000 to 1 in 25000 live births worldwide (Ho & Dimitropoulos, 2010).
- PWS was estimated to have affected 400000 people worldwide (Hurren & Flack, 2016).

BACKGROUND INFORMATION

PWS affects multiple areas in the body of the individual, which leads to:

- dysmorphic features, poor muscle tone (hypotonia)
- feeding difficulties with sucking deficit
- satiety (hyperphagia)
- under-developed sex organs (hypogonadism)
- cognitive impairment
- impaired social cognition, behavioural problems and sometimes psychiatric disturbances.

OBJECTIVES OF THE STUDY

- Learn about the experiences and challenges of individuals diagnosed with PWS and those of the primary caregiver.
- Identify the most effective interventions, by generating more strategies and by using and modifying, where necessary, existing therapeutic methods.

RESEARCH METHODOLOGY

- Qualitative research
- Collective instrumental case studies were used, making use of participatory action research, ethnography and auto-ethnography elements
- Purposive sampling
- Semi-structured interviews and observations
- Thematic analysis

FINDINGS

Experiences and challenges of the diagnosed individuals

- Food stealing, stealing money to buy food and accepting food from other learners
- An unexpected finding was that with one of the participants, the food-stealing behaviour was resolved, and she is now disciplined and has self-control around food even when she is unsupervised.

FINDINGS

Experiences and challenges of the diagnosed individuals

- Multiple behaviour problems were reported throughout the literature reviewed (Butler et al., 2006; Cassidy & Driscoll 2009; Ho & Dimitropoulos, 2010; McCandless et al., 2011). The participants in this study also presented with different behaviour problems :

FINDINGS

Experiences and challenges of the diagnosed individuals

- frustration and anxiety
- anger outbursts
- stubbornness
- struggle to take orders from certain individuals in their lives
- manipulative tendencies
- argumentative
- aggressive tendencies.

FINDINGS

Experiences and challenges of the diagnosed individuals

- swearing and lying
- being disruptive
- refusal to be in class and taking instructions from teachers
- wandering around the school premises
- undressing in front of other learners and also displaying inappropriate sexual behaviour
- obsessive-compulsive behaviours
- perseveration

FINDINGS

Experiences and challenges of the diagnosed individuals

- Encopresis and enuresis
- Equit, Piro-Hussong, Niemczyk, Curfs, & von Gontard (2013) investigated the rate of elimination disorders and behavioural symptoms in people with Prader-Willi and Fragile-X syndromes, and they found that 29.3% of people with PWS had at least one elimination disorder
- Need for psychological and psychiatric intervention.
- Medication was prescribed for better behaviour management

FINDINGS

Experiences and challenges of the primary caregiver

- Disruptions of studies, job terminations and the inability to secure full-time employment
- The caregivers dedicated most of their lives to taking care of their children with PWS.
- The primary caregivers found their sense of self being engulfed by the role of caring, and they lost touch with themselves and their main focus was taking care of their child with PWS, and also constantly monitoring them. This caring role proved to be strenuous, time-consuming, overwhelming and almost unbearable and led to stress and struggles with coping.

FINDINGS

Experiences and challenges of the primary caregiver

- The findings of this study also revealed issues around stigma. This was evident in how “others” blamed some primary caregivers for not disciplining their children, and how the primary caregivers took responsibility and blame for their children’s behaviour.

PSYCHOLOGICAL INSIGHTS

- Psychotherapy for the primary caregiver

In addition to the stress associated with caring for the individual with PWS, primary caregivers had other personal stressors, such as grief, marital problems, divorce and unemployment. The stressors influenced their ability to take care of their child and to manage some of their behaviours. Therefore, psychotherapy provided to the primary caregivers helped to enhance their quality of life and their ability to provide care.

PSYCHOLOGICAL INSIGHTS

- Anticipatory planning

Anticipatory planning was proven to be effective when the primary caregiver plans and prepares activities in advance to avoid sudden changes and delays, and negotiates with the individual if there are anticipated changes. This was also done by pre-empting the undesired behaviour before it happened, as well as by proactively reducing opportunities for an undesired behaviour to occur.

PSYCHOLOGICAL INSIGHTS

- Routine and structure

Establishing a structured environment with a scheduled daily routine proved to be effective for the PWS individuals in the study, even though it required the primary caregiver to work hard at maintaining it.

- Rule-governed behaviour

The primary caregivers gave clear rules, guidelines and were not negotiated with the individual when they present with undesired behaviour but firmly giving instructions. This also included telling them in advance of the consequences should they misbehave.

PSYCHOLOGICAL INSIGHTS

- Pivot tool

The pivot tool, proved to be effective in managing swearing. Swearing in PWS individuals was minimized by ignoring them when they swear and then shifting the focus to other activities. Doing this immobilized the individual and the duration of the episodes was reduced.

- Patient participation

Involving individuals with PWS in decision making also proved to be effective. This intervention leaves the individual with PWS feeling in control of his/her own treatment plan, encouraged and empowered in the process.

PSYCHOLOGICAL INSIGHTS

- Psycho-education

Psycho-education was used extensively in this research study and proved to be an effective method. It was used to give information to families and teachers about PWS. Secondly, it was used to help families and teachers devise coping strategies and solutions for the problems they were experiencing with PWS individuals.

Following the psycho-education, the primary caregivers and teachers exhibited empathy towards the individual, and were educated and empowered. Their perceptions regarding the displayed behaviours changed. They became aware that managing and controlling the environment was crucial, and even put structures in place to prevent undesired behaviour from happening.

RECOMMENDATIONS

- The Department of Health needs to invest more in awareness campaigns for healthcare professionals.
- During genetic counselling, parents should be informed about support systems that are available to them including special schools, availability of growth hormone therapy in government hospitals and the option of a disability grant / care dependency grant.
- Schools and teachers need to be fully equipped to deal with learners with PWS, and these learners should be placed in a school where the structure is predictable, and with a high teacher-student ratio.
- It would also benefit families if there was a specialized group home to serve individuals with PWS.



THANK YOU!!