

# INDIVIDUALS WITH PRADER-WILLI SYNDROME AND THEIR PERCEPTIONS OF SKIN-PICKING BEHAVIOUR

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## Introduction

Prader-Willi syndrome (PWS) is a genetic disorder associated with abnormal or absent expression of the paternal copy of as yet unidentified maternally imprinted gene(s) at the genetic locus 15q11-13. Three genetic subtypes are involved. The two main genetic subtypes are those with a deletion at the locus 15q11-13, involving chromosome 15 of paternal deletion (deletion 15 genetic subtype; approximately 70% of cases), and those with maternal chromosome 15 uniparental disomy (disomy 15 genetic

subtype; approximately 25% of cases). The remaining 5% have a chromosomal re-arrangement or a chromosome-15 imprinting centre defect (Butler *et al.*, 2006). PWS is characterized by many medical and psychological features, and individuals usually function in the mild to moderate range of intellectual disability (see *e.g.*, Butler *et al.*, 2002; Curfs and Fryns, 1992; Holland *et al.*, 2003).

Individuals with PWS are at increased risk for mental health and behavioural problems. A severe and distinctive type of behaviour problem often observed in individuals with PWS is skin-picking.

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Skin-picking is a self-injurious behaviour that may cause severe tissue damage and other health problems (e.g., infections). Several studies have been published that show that skin-picking is a prevalent phenomenon in PWS, with rates ranging from 58% to 95% in children and adults (e.g., Didden *et al.*, 2007; Dykens and Kasari, 1997; Symons *et al.*, 1999; Wigren and Hansen, 2003). The cause of skin-picking in PWS remains largely unknown. It has been conceptualized as a symptom of an obsessive-compulsive disorder (e.g., Wigren and Hansen, 2003). Recent results of a functional assessment in a large sample of individuals with PWS (n = 119) suggest that in most cases skin-picking may be a learned operant maintained by negative reinforcement (i.e., escape from and avoidance of high levels of arousal) (Didden *et al.*, 2007). In a minority of this sample, results suggest that skin picking has a social functioning. Skin picking is thus maintained by negative and/or positive reinforcement. The specific source of the reinforcement and the antecedents that elicit it, should be individually assessed as the sources of stress and events that elicit skin-picking may be highly idiosyncratic (Didden, 2007). The relationship between (idiosyncratic) events and skin-picking behavior should be explored before a treatment programme may be designed (see e.g. Lynne Lane *et al.*, 2006). The client's perception of this behaviour may be an important source information in this process of functional analysis and treatment.

Although our knowledge on prevalence, associated variables and causes of skin-picking in PWS has increased, no studies have been conducted on the individual's perception of the impact of skin-picking on their own lives. In other words, although stress and high arousal levels may increase the probability of skin-

picking, the sources of the stress are often unknown to staff, parents and others. In a small sample of non-disabled individuals (n = 28), the psychosocial consequences of chronic skin-picking was investigated using the Skin-Picking Scale (Keuthen *et al.*, 2001). Results show that skin-picking was associated with self-reported intensity of distress before, during and after skin-picking. Individuals with skin-picking reported severe adverse social impact of their skin-picking. Individuals mentioned that they hated the way they looked, there are sometimes things they can't do, and their relationships have suffered. Results of a recent study by Flessner and Woods (2006) among 92 non-disabled individuals with chronic skin-picking indicate that they experienced social and occupational impairments, as well as a number of medical and mental health concerns that they attributed to skin-picking. In both studies, relationships were found between skin-picking severity and levels of self-reported depression and anxiety.

Results of the above studies indicate that skin-picking may have a serious and adverse impact on the lives of the individuals involved. Data on the impact and perception of skin-picking in the above studies were all collected using questionnaires and self-reports. Due to cognitive impairments, questionnaires may not be suitable for individuals with PWS. In the present study, therefore, structured open-ended interviews were conducted with 10 individuals with PWS to explore the impact and perception of their skin-picking behaviour. We aimed to explore what these individuals thought about this behaviour in general, which adverse consequences (if any) this behaviour has and what they thought about why they show this behaviour and what elicits it.

## Method

### *Participants*

We aimed at establishing a heterogeneous sample of individuals that varied with respect to age, gender, genetic subtype and living setting (purposive sampling). In this way, we expected to get a more varied and broad picture with respect to the individual's perceptions about their skin-picking. We contacted the Dutch Prader-Willi Parent Organisation and several centres for the care of people with disabilities. Ten individuals with a confirmed diagnosis PWS were selected and they as well as their parents and/or caregivers were explained the aim of the study. Participants functioned in the mild to borderline range of intellectual disability (total IQ ranged from 57 to 77). Their mean age was 41 (range: 29 – 54) years and five of them were men. The genetic subtype was known in six individuals. Four individuals had chromosomal deletion, and two had two copies of the chromosome (disomy). Five individuals lived in a group home in the community, one person lived at home with his parents, one person lived independently in an apartment in the community (albeit with daily support from a centre), and three participants lived in a residential facility. Participants showed chronic skin-picking and were verbally competent. None of the participants were receiving a specific treatment for their skin-picking. Written informed consent was obtained for each participant and his caretaker.

### *Procedure*

A qualitative research method based on grounded theory (see Strauss and

Corbin, 1998) was used to explore the impact and perceptions of self-injurious skin-picking in the 10 participants with Prader-Willi syndrome. First, a topic-list was established that contained questions pertaining to: (a) general picture of skin-picking (e.g. "On which body part do you pick your skin?"), (b) reasons for skin-picking (e.g. "Which events usually elicit skin-picking?"), (c) prevention of skin-picking (e.g. "How can you prevent that you start skin-picking?"), (d) how skin-picking is perceived by participants ("How do you feel about your own skin-picking?"), and (e) other people's responses to skin-picking (e.g. "How do other people respond to your skin-picking?"). During the semi-structured interviews that were conducted by a trained interviewer (i.e. the fourth author) the topic-list served as a guide. However, topics were added to the list if participants mentioned new topics during an interview. The interviewer refrained from directing participants towards answers as much as possible, but she prompted participants when necessary for eliciting a response. Each interview was audio taped (after consent) and transcribed.

### *Data analysis*

The transcripts of the interviews were coded, grouped and categorized. The analysis was split in two phases, open coding and axial coding. To explore the phenomenon of skin picking, open coding was applied to five interviews. Axial coding (to explore the variety within the categories found in the open coding phase) was applied to the other five interviews. Saturation was reached in both phases. Two researchers were involved in the coding process. The fourth author coded

all interviews. Five interviews (50%) were also coded independently by the second author (peer review). Meetings were held to discuss the differences in codes given, so that the analysis could be broadened with new insights and to reduce the risk of the researcher 'being blinkered'.

## Results

### General picture of skin-picking

Body parts most often involved in skin-picking were face, legs, arms and feet. Next to this, biting and picking nails were often mentioned. One participant mentioned that she scratched open her skin and underlying varicose veins.

*"And each time it itches or if there is a crust, I'll scratch my skin open. Last time I scratched I had an itch on my leg because of a burst blood vessel."*

All participants pick their skin with their nails, they did not use other means. Frequency of their skin-picking varied from daily to only now and then. Severity of skin-picking also varied and one participant had consulted a physician because of wounds. All participants mentioned that they started skin-picking very early in their lives but most did not know when and why they began skin-picking. One individual mentioned that he began skin-picking following a visit to the hospital.

*"When the doctor used a thick needle for taking blood samples. At first I did not scratch open the wound, but later it started to itch and then I scratched it open and a crust appeared. But as soon as the crust starts itching I remove it and also scratch the wound until it begins to bleed."*

### Reasons for skin-picking

The reasons given by participants for their skin-picking can be categorised in medical/physical and psychological reasons.

**Medical and physical reasons.** According to seven participants, the most important reason for skin-picking is itchiness.

*"I scratch when it itches. Yes, then I scratch open my skin, I know that this is not a good thing to do, but I cannot help picking my skin. It starts to itch and then I start to scratch and pick my skin."*

Several participants mentioned that the itch worsens after swimming and, as a consequence, also the skin-picking gets worse. One person mentioned that he often picks at his nails because he likes it when his nails are very short. Another reason that was often mentioned was eczema. Most participants stated that skin-picking was a feature of the Prader-Willi syndrome.

*"This behaviour belongs to my Prader-Willi syndrome, it will not disappear. It has something to do with the syndrome, they cannot point that out to me."*

**Psychological reasons.** Most often psychological reasons for skin-picking were "nerves", "being teased", "being nervous", and "brooding". One participant mentioned that he skin-picked whenever he thought about the loss of his beloved father. After this, his skin-picking had worsened.

*"Yes, normally, yes, I have no father anymore... I often see my father standing in my bedroom, then I feel very nervous and stressed. Then I start picking at my skin...It's just nerves."*

Feeling bored was also often mentioned as a psychological reason for skin-picking. Participants start skin-picking when they

are alone in their room, when they lie on their bed or if they have nothing to do.

*"When I have nothing to do, I start scratching and picking at my skin. Then I am feeling bored to death. I sit and watch and am not busy with my hands. Then I start to scratch at my skin."*

One participant thought that his skin-picking "was automatic behaviour" and that "it had become a routine".

### *Prevention of skin-picking*

Eight participants were convinced that there is no remedy or treatment for their skin-picking and that their skin-picking may not be prevented. However, two participants mentioned several factors that could prevent or reduce skin-picking, such as, "cutting the nails very short", "being busy" and "being rewarded for not showing skin-picking".

*"I never scratch and pick at my skin when I am smoking in front of the television. Then I am doing something, I hold a cigarette and cannot scratch or pick at my skin."*

### *Own perception of skin-picking*

In general, participants' perception of their own skin-picking is a negative one. Three participants mentioned that they strongly disapproved of their skin-picking. Three others thought that this behaviour is "filthy". Four participants mentioned that they felt bad following skin-picking. They felt angry and guilty about it and they thought that it is painful. One participant mentioned that he felt very guilty towards other people because of his skin-picking.

*"I feel bad about doing it. Sometimes I think that the behaviour is really deep inside*

*me. I feel very guilty each time I have skin-picked."*

By contrast, two participants mentioned that they felt relieved after skin-picking because the itch had disappeared. Eight participants experiences feelings of shame, they did not like other people seeing their wounds and scars. As a result of this, participants were restricted in their choice of clothes and activities. For example, they never went to a public swimming pool or never wore shirts with short sleeves, even during the summer when the weather is hot. One participant mentioned that he wanted to have an operation on the lips and mouth area in order to remove the scar tissue.

Seven (n = 7) participants mentioned that they liked to discuss their skin-picking with other people, such as their parents and staff members. Others, however, did not like the comments from others about their skin-picking. They preferred not to discuss their skin-picking with other people.

*"When I am upstairs and I have scratched or picked at my skin, I ask my caregiver how I can make amends to him for this, how I can be of assistance to him, or whether I can buy something for him. I cannot help my skin-picking, but I want to amend to other people for my skin-picking."*

Several participants mentioned that "there are many things that other people do not understand about PWS" and that "there is much ignorance about this". They find it hard to explain to others what it means to have PWS, and to have the symptoms and behaviours.

*"I think that other people do not understand Prader-Willi syndrome! They do not know what it is! They do not understand that some behaviours [skin-picking] belong to the syndrome!"*

### *Other people's responses*

Other people's response to skin-picking is mostly negative and one of disapproval. One participant mentioned that he was given a slap on his hand during skin-picking.

*"Other people say that skin-picking is not allowed and that it is filthy."*

## **Discussion**

The present study is the first that investigated viewpoints of individuals with PWS of their skin-picking behaviour. Information was gathered through semi-structured interviews. Our findings show that people with PWS who participated were able to think about the topic of skin picking. Results of the interviews suggest that skin-picking may have some seriously adverse impacts on the lives of those involved. Furthermore, individuals with PWS may provide clues about events that may elicit and maintain their skin-picking. Results of this study broadly support results of the Curfs and co-workers study (Didden *et al.*, 2007).

Information given by individuals with PWS that were interviewed in our study is in agreement with results from large scale epidemiological studies. For example, skin-picking usually starts very early in the lives of individuals with PWS although it remains unclear what causes it. Also, skin-picking is directed towards the face, arms and feet which is in agreement with results from other studies (see e.g. Didden *et al.*, 2007; Symons *et al.*, 1999).

Results also show that skin-picking has some seriously adverse consequences for the individuals involved. Other people respond negatively towards these individuals whose ranges of activities

are restricted. Research on effective treatments for skin-picking is warranted to allow individuals with PWS to lead a life as normal as possible in this respect. Unfortunately, our knowledge on effective and humane treatment of skin-picking is very limited although several suggestions on possible effective approaches have been given (see e.g. Didden *et al.*, 2007; Dykens and Kasari, 1997; Symons *et al.*, 1999; Wigren and Hansen, 2003).

Most individuals in our study reported that an increased arousal level (e.g., nerves, being tense) often elicits skin-picking. This is in agreement with the findings from other studies suggesting that skin-picking is evoked by increased levels of arousal and/or physical sensation such as an itch (Didden *et al.*, 2007). In these individuals skin-picking is probably maintained by non-social negative reinforcement, in that skin-picking results in the decrease of elimination of the increased arousal and/or physical sensation. In other cases, skin-picking may be maintained by non-social positive reinforcement. Skin-picking may be elicited by feelings of boredom or under stimulation and skin-picking may result in a pleasant sensorical reinforcement.

There are clear clinical implications. Prior to treatment, a functional assessment of skin-picking should be conducted in which possible relationships between this behaviour and other events are explored. At present, there are relatively many procedures with which such an assessment can be carried out (Didden, 2007). Next, to identifying sources of increased stress and anxiety, events that directly elicit and maintain skin-picking should be explored. During this assessment, information of the client himself/herself may be important and valid in identification of such events. Also, caretakers (e.g. staff, parents, teachers) of these clients should be aware that this

individual's skin-picking may have one or more (idiosyncratic) function(s). They should refrain from ascribing a function of skin-picking prior to a function-based treatment. This function may be different across individuals with PWS and may even differ across time.

There are some limitations to this study. A qualitative design like grounded theory is an adequate approach for exploration and description, but cannot be used to test assumptions. Furthermore, the study was limited to interviews. No observations were made to verify if the clients's information was valid. That is, we did not experimentally assess whether skin-picking was indeed related to events that were mentioned during the interviews. Nevertheless, qualitative interview techniques provide the possibility to verify the respondent's answers, for example by asking further questions, and by asking for illustrating examples and situations. The limited size sample only permits open and axial coding. More interviews (phase of selective coding) are needed to support the 'preliminary' theory developed. Despite the particular context and limited sample size results demonstrate that people with PWS are capable of giving their views on their skin-picking and how this self-injurious behaviour affects their quality of life. More research is clearly warranted in the important area of assessment and treatment of skin picking. First, an innovative feature of the literature would be to include an individual's perception into the process of functional analysis and treatment. It remains to be assessed whether his/her information has validity in identifying setting events and discriminative stimuli that elicit skin-picking. This holds also true for the identification of events that maintain the behaviour. Usually, such information is presented by significant others and

not by the participant. Furthermore, functional assessments of skin-picking in individuals with PWS are lacking and should be conducted. For example in a study by Lane *et al.* (2006) functional assessment consisted of asking the teacher to collect data on the occurrence of skin-picking of a 9-year-old boy with mild intellectual disability who visited a classroom in a special school. His teacher provided information during a functional assessment interview and research staff collected data in the classroom. Functional assessment data showed that skin-picking was most probable during reading instruction and least probable during math instruction during which the boy's hands were occupied. High levels of skin picking were reduced by providing the boy with 'manipulatives' (e.g. a small ball, pencil, counter) each time he entered the class, especially during reading instruction. Using functional assessment methodology, treatment programmes may be designed with which skin-picking behaviour in individuals with PWS may be effectively reduced.

## Summary

Self-injurious skin-picking is a distinctive feature of Prader-Willi syndrome. Until present, studies have addressed its prevalence, characteristics and causes. However, little is known about how individuals experience this type of behaviour and what impact skin-picking has on their lives. This paper reports on a study using a qualitative research method based on grounded theory to elicit the views of people with PWS on the sensitive issue of skin-picking behaviour. Research into the perspectives of individuals with PWS pertaining to

their skin-picking behaviour is lacking. The interviews show that this behaviour may be elicited by a variety of events and that it may have adverse consequences for the individuals involved. In general, the interviews indicate that individuals with PWS are capable of providing clues on why they exhibit this behaviour and what impact it has on their quality of life. Knowledge on such clues may help in designing programmes aimed at supporting people with PWS to help them reduce skin picking.

## Authors' Note

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