

Copyright is owned by the Author of the thesis. Permission is given for a copy to be downloaded by an individual for the purpose of research and private study only. The thesis may not be reproduced elsewhere without the permission of the Author.

**Supporting adults who have Prader-Willi Syndrome:
Caregivers' Perspectives on the Ethical and Practical
Dilemmas.**

A thesis presented in partial fulfillment of the
requirements for the degree of

Doctor of Philosophy

In

Psychology

At Massey University, Wellington

New Zealand

Richard John Ford

2018

This work is dedicated to Christine, Zeta and Harry, with love and in
recognition of their many sacrifices.

Five smooth stones.

Abstract

Prader-Willi Syndrome (PWS) is a complex genetic condition that typically results in learning impairment and hyperphagia, which is the drive to consume excessive quantities of food. Obesity often develops very rapidly and is life-threatening, presenting professional and familial support people with a distinctive set of ethical and practical dilemmas. These unique challenges can be characterised as a microcosm of a wider “safety versus autonomy” debate within human services. Little is known about the nature or efficacy of publically funded supports to adults who have PWS in Aotearoa/New Zealand. The aim of this thesis was to investigate the ethos of current services and some of the outcomes associated with them. In particular, it sought to illuminate the ways in which professional caregivers address the challenges they face in their work, according to their own reported perceptions of their role.

The thesis begins with a review of the relevant international research literature, noting the dominance of clinical experience and anecdotes over research studies within it. This review has informed three investigations. First, a scoping study comprised of semi-structured interviews was conducted with professional caregivers in workplace-based focus groups. Second, an attempt was made to identify and survey all adults within Aotearoa/New Zealand who have PWS regarding the supports they received. Finally, a thematic analysis of the transcribed texts of interviews with individual caregivers was undertaken. These interviews concerned the ways in which caregivers of adults with PWS experienced, conceptualised and addressed the dilemmas inherent in their role.

Findings indicated that there are grounds for serious concern about both the wellbeing of people with PWS in Aotearoa/New Zealand, and that of their caregivers. Significant levels of obesity were reported in almost all service users with PWS, despite the ecological controls

over access to calories imposed on them by caregivers. It seems likely that some adults with PWS are at risk of serious harm while having their civil rights infringed to little or no effect.

Caregiver participants characterised their role in terms of the challenges it presented them. The management of service users' access to food was identified as only one of the sources of such challenge. Challenging behaviour among people with PWS was reported to generate the most significant and pressing difficulties. Caregivers reported responding to these challenges by imposing substitute decisions, especially in relation to food, despite awareness that such measures seemed to constitute a breach of ethical norms. They regarded compulsory ecological controls as necessary for the service users' safety, but also as a compassionate response which prevented uncertainty in relation to food and hence spared service users with PWS from subsequent anxiety.

Caregivers seemed in agreement with a view of client autonomy as relational and seemed to employ a dialogical process featuring stories and responses to stories in collective decision making. Despite the emphasis on individualism in contemporary services to people with disabilities, interviewees often seemed to prioritise corporate wellbeing, even when making decisions regarding access to food for a single individual with PWS.

The conclusions which may be drawn from these studies are limited by a number of factors, notably small sample size. Nevertheless, they offer insights into the nature of the caregiving task and the concept of autonomy within human services. Findings form the basis of suggested innovations, both in caregiving practice and for future research.

Acknowledgements

The community of those who work in support of people with PWS is a small and supportive one, both locally and around the world. I would like to offer my thanks to the wide variety of people whose support has enabled me to complete this project, first to my patient supervisors from the School of Psychology at Massey University's Wellington campus, Dr Ian de Terte, Emeritus Professor Ian Evans and Dr Stephen Humphries. I would also like to acknowledge the seemingly limitless help I have received from PWSA (NZ), and from its leading lights, Linda Thornton, Cindy Adams-Vining and their families. The Association helped me contact people with PWS and their support people, lent me books on a quasi-permanent basis, talked with me about problems and solutions, and enabled me to meet a raft of skilled and inspiring people.

I would also like to acknowledge my colleagues from the International Prader-Willi Syndrome Organisation (IPWSO), especially Dr Norbert Hödebeck-Stuntebeck and the other members of the Caregivers and Professionals' Advisory Board. Dr Janice Forster spent many hours discussing this project with me in its early stages and was an important influence on its development. Mary-Kaye Ziccardi, Jackie Mallow, Dr Hubert Soyer and Dr Larry Gentsil introduced me to their various services and Professor Barbara Whitman was also very supportive, encouraging and generous with her time.

The challenge of finding and contacting adults who have PWS was a significant impediment in the early phase of this project. It was only overcome by the voluntary help of a number of agencies, including PWSA (NZ) and many of the NASC (Needs Assessment and

Service Coordination) agencies, along with their national body, the NASC Association (NASCA) and leading service providers. I was particularly grateful for the support of staff from IHC New Zealand, Support Works Nelson/Marlborough and Auckland's Spectrum Care Trust. The staff at IHC's excellent library provided valuable assistance in sourcing some hard to find resources. John Taylor of Community Connections offered invaluable advice during the writing process and Matt Frost generously proof read an early draft of this manuscript.

Both of my employers, Tautoko Services of Paremata in Wellington and the New Zealand Ministry of Education, have generously allowed me time and office resources to work on these studies. Colleagues in both organisations have also shared a passion for the development of human services and the empowerment of children and adults with PWS. My long-term professional supervisor, clinical psychologist Helen Inkster, encouraged me many times to continue in the face of difficulty.

Since beginning this project, I have also received a steady stream of help and encouragement from people with PWS, their families and their professional support people. Many of their ideas and priorities were incorporated into the design of the final project. Caregivers and people with PWS also took considerable care and effort to participate in one of the three studies that make up this project. Many of them shared intimate parts of themselves and their lives in the process. I am grateful for their support, and share their hope that through research such as this we can increase the range of choices and opportunities that will be available to those who come after us in this area.

I would also like to thank my faithful band of study companions; Caramel, Ron, Sebastian, Inigo and Rosie. They came and went but were always encouraging, and above all quiet. I am most indebted, however, to Christine, to Zeta and to Harry; my family whose unfailing support and many sacrifices have allowed me to pursue the work I love.

Table of Contents

Dedication	ii
Abstract	iii
Acknowledgements	v
Table of Contents	vii
List of Appendices	xix
List of Tables	xx
List of Figures	xx
Foreword	xxi
The Author's Perspective	xxiv
Chapter One	
PWS and Contemporary Support Services	1
PWS in a food rich environment	1
What is PWS?	1
An overview of the project	2
PWS and Maori	4
The relevance of this project to the lives of people with PWS	4

PWS and development	6
Cognition and communication	7
PWS and challenging behavior	7
Hyperphagia	8
Two models of hyperphagia in PWS	9
A disorder of satiety	9
Reward dysfunction: hyperphagia as addictive behavior	10
Managing hyperphagia: the present research literature	11
Why are some aspects of PWS so well studied?	11
Medical interventions	12
Behavioural interventions	13
Other therapeutic interventions	15
Environmental controls over access to food	17
The central dilemma facing caregivers	17
Holland and Wong's resolution	18
Further benefits claimed for restricted access to food	19
"Food security"	20
Some problems with the present consensus	20
The views of caregivers	21

Can the imposition of controls be considered empowering?	22
The limits of this justification	23
A way forward	23
Conclusion	23
Chapter Two	
Study One: Initial Interviews in Workplace Focus Groups	26
Ethics	26
Method	27
Semi-structured interviews in workplace-based focus groups	27
Participants and recruitment	29
Procedure	29
Analysis	30
Results	30
Food management	31
Experiences identified by caregivers as most challenging	33
Challenging behavior	33
Service users' relationships with food	35
Service features identified by caregivers as most important for service quality	36
Discussion	37

Limitations	41
Conclusions	42
Chapter Three	
Study Two: A Description of Services to a Group of Adults with PWS	43
Ethics	44
Method	44
A population survey	44
Participants and recruitment	45
Measures	45
The Hyperphagia questionnaire (HQ)	45
The New Zealand PWS services questionnaire (NZPSQ)	46
SF-36	46
Procedure and Analysis	47
Results	47
HQ	47
SF-36	48
NZPSQ	48
Body-mass index (BMI)	48
Living situation	49

Support from professional caregivers	49
Food management regimes	49
Discussion	50
Study limitations	53
Chapter Four	
Study Three: Interviews with professional caregivers	55
The significance of the caregiving role	55
The present focus on the individual in residential service provision	55
The goals of Study 3	56
The role of the researcher	56
Ethical review	57
Method	58
Semi-structured interviews with individual caregivers	58
Participants	59
Procedure	60
Analysis	61
Results	62
Themes: The dominant narrative	62
A caveat	62

A counter narrative	62
The relationship between these narratives; challenge and reward ...	63
The first theme: Challenge	65
Subtheme: Service users' challenging behaviour	65
Foraging behavior	65
Aggressive behaviour and assaults in pursuit of food	65
Foraging behaviour outside the residential service	66
Other foraging behaviours	67
The tone of discussions on foraging behaviour	67
Nonfood related challenging behaviour	68
Inflexibility and excess; the PWS personality	68
Interpersonal conflict	70
Subtheme: Relationship dissonance	70
Subtheme: Threats to safety	72
The experience of a client's premature death	73
Responsibility without power	74
The safety and well-being of others	74
Subtheme: Vulnerability to criticism	75
Caregivers' experience of isolation	75

Subtheme: Challenge as dynamic yet unrelenting	76
The second theme: Caregivers' response to challenge	77
Subtheme: Authoritarianism	77
Justifications for substitute decision making	79
An apparent exception	80
Substitute decisions, empowerment and compassion	82
Subtheme: Creativity	83
The promotion of safe autonomy	83
Tolerance for a degree of risk	83
The limits of tolerance; safety remains paramount	84
Subtheme: Exercise	85
Subtheme: Practical wisdom	85
The dialogical process within the present study	86
Caregivers and resilience	88
The inevitability of risk	89
The importance of managing anxiety among people with PWS	90
Food security as a tool with which to manage anxiety	91
Empathy	92
Tricks	93

Subtheme: Consistency	95
Another dissenting voice	97
Families and consistency	97
The third theme: Outcomes valued and sought by caregivers	98
Subtheme: The search for a livable life	98
Subtheme: Safety remains paramount	101
Subtheme: Other priorities; happiness, fairness, autonomy	102
Discussion	102
The goals of the present study	102
How do caregivers conceptualize and experience their role?	103
Thrownness and being between	103
Parallels with challenges faced by other caregivers	103
The pervasive impact of hyperphagia	104
The dominant narrative	104
Challenge and response.....	104
Relationship dissonance	105
Challenge as both dynamic and unrelenting.....	107
Isolation and vulnerability	107
The counter narrative; caregiving as a rewarding task	107

Authoritarianism	107
Creativity	108
Practical wisdom	108
The outcomes sought by caregivers	108
Physical safety	108
The livable life	109
How do caregivers make difficult decisions concerning food security?	110
Caregivers follow instructions	110
Whose dilemma is this anyway?	110
Corporate decision making	110
Caregivers balance multiple priorities on behalf of multiple agents	110
Caregivers accept a degree of risk	110
Caregivers regard physical safety as paramount	111
Caregivers seem to employ a dialogical process	111
Conclusions	111
Chapter Five	
The dominant narrative among caregivers: challenge and response	114
Challenge	114
Challenging behaviour in generic disability services	115

Food related challenging behaviour.....	116
Interpersonal conflict	116
Relationship dissonance.....	117
Isolation	118
Response	118
Maintaining consistency	118
Caregiving as a corporate activity	119
The importance of managing anxiety	120
Substitute decision making	120
The primacy of service users' physical safety	121
Fairness and corporate wellbeing	121
Mutually valued relationships	121
Desired outcomes: The central dilemma facing caregivers	122
The central dilemma in the context of generic services.....	123
The concept of client autonomy within disability services	123
Autonomy and health related choices within disability services	123
Further questions about the nature of autonomy in health services	124
The relevance of this debate to services to people with PWS	126
The ethics of controlling access to food	126

The central dilemma reframed	127
How do caregivers make difficult decisions?	127
Making use of the dialogical process	128
The State of Services to Adults with PWS in Aotearoa/New Zealand	129
The limitations of these studies	131
Directions for future research	132
The relevance of research into PWS to mainstream services	132
Challenges Facing Researchers	133
The lack of instruments to measure key variables	133
Difficulty recruiting research participants	134
A way forward	134
Overcoming the problem of small sample size	134
The development of a wider range of measures	135
HQ, NZPSQ	135
Measures of service outcomes: QoL and FQoL	135
Priorities for future research	137
References	139
Appendices	158

List of Appendices

Appendix A: Information sheet, Study 2.....	159
Appendix B: Dual consent form, Study 2	163
Appendix C: The New Zealand PWS services questionnaire	165
Appendix D: Information sheet, Study 3	168
Appendix E: Interview questions, Study 3	172
Appendix F: Interview questions, Study 1	173

List of Tables

Table 1.	<i>Factor means, SD's and ranges on the Hyperphagia Questionnaire for a group of adults who have PWS.</i>	48
Table 2.	<i>BMI scores of a group of Aotearoa/New Zealand adults with PWS</i>	50

List of Figures

Figure 1.	<i>The Dominant Narrative: Challenge and Response</i>	63
-----------	---	----

Foreword

“What ultimately determines how a person or a group will be treated, and what others will afford to such a party in life, is what is in the mind of those who do the treating and affording...”

Wolfensberger (2002)

People whose behaviour challenges us show up weaknesses in systems.

Truism

Prader-Willi Syndrome (PWS) is a rare, genetically determined condition that typically results in learning impairment and a defined set of other physical and cognitive characteristics (Butler, Hanchett, & Thompson, 2006; Butler et al., 2018; Whittington & Holland, 2017). Hyperphagia, the drive to consume excessive quantities of food, seems to be regarded as the definitive feature of PWS, both by many investigators and by experienced caregivers who have written about their work (Holland, 2015; James, 2010; Waters, 2009; Ziccardi, 2006).

PWS is also associated with lowered muscle tone, commonly referred to as hypotonia. Those who have the syndrome are often less active than their peers and typically require fewer calories to gain weight (Butler, Lee, & Whitman, 2006). Unless access to food is limited by ecological constraints severe obesity often results, and may arise very rapidly. Common consequences include ill health, restricted mobility and sudden or premature death (Butler, Manzardo, Heinemann, Loker, & Loker, 2017; Hedgeman et al., 2017).

The risks associated with hyperphagia mean that those who provide support to adults who have PWS face a distinctive set of practical and ethical challenges (Butler et al., 2006; Ho & Dimitropoulos, 2010; Whitman, 2013). An international consensus exists which maintains that it is the responsibility of caregivers to impose ecological and other controls on access to food in order to protect their clients or family members who have PWS from harm (Forster, 2010; Holland, 2015). However, the great majority of publically funded supports to people who have PWS and their families are provided within the context of generic services to people with intellectual disability (ID). The promotion of independence and civil rights among service users have become core values within this industry (Office for Disability Issues, 2001, 2014). On superficial analysis at least, these values would seem to preclude caregivers imposing controls over unwilling clients, especially in relation to such fundamental matters as choices over food and personal finances.

The control of access to food for people who have PWS does not only represent an ethical challenge for caregivers, it is often also difficult to achieve in practice. Hyperphagia in PWS is associated with a range of uniquely challenging behaviours (Whitman, 2013) and published reports of caregivers record attempts at foraging for food which have often been determined and creative (James, 2010; Waters, 2009; Whitman, 2013; Ziccardi, 2006).

The current scientific literature offers little guidance to caregivers as they address these challenges. Very little is known about the nature or effectiveness of contemporary support services to adults with PWS, either within Aotearoa/New Zealand (Thornton & Dawson, 1990) or overseas (Stanley, 2014). In particular, only a small group of studies have included descriptions of the measures by which paid or familial caregivers have sought to manage the risks associated with unfettered access to food (Stanley, 2014).

Some of the people who first promoted the inclusion of people with ID and challenging behaviour into wider society ventured the opinion that everybody should be regarded as gifted (Pearpoint, O'Brien, & Forest, 1992). In this way, these authors have suggested not only that each individual has some unique perspective or experience with which they might enrich the lives of those around them, but also that it is the wider community that will likely be impoverished if any individual is prevented from making their contribution. If we accept the view of Pearpoint et al. (1992), we might speculate on the nature of the gifts contributed by adults who have PWS. It seems likely that at least some may lie in the very complexities inherent in the task of supporting these citizens in relation to food. The needs of service users who have PWS oblige both professional caregivers and members of the research community to confront difficult issues, including some which we might otherwise be tempted to minimise or ignore. People who have PWS show little tolerance for shortcuts on the part of those who are paid to support them. They do not allow us to evade controversy or to fudge difficult choices but require us to design and deliver supports that are practical, sustainable over time and able to withstand scrutiny.

The study of support services to people who have PWS is an important end in itself. However, as Schwartz et al. (2016) have noted, PWS has often also proven a convenient analogue for issues of more general concern. It seems likely that the solutions developed by caregivers supporting people who have PWS in relation to food may also have useful things to teach us about questions of client autonomy and self-determination in the face of unequal power, as they arise across human services.

The Author's Perspective

For more than 25 years, the author has worked in a variety of roles supporting people with developmental disabilities, especially those whose behaviour presented challenges to themselves or others. This thesis is the product of a caregiver studying caregivers. It is also the work of a practised advocate, both for people with PWS and for those who care for and about them. I serve as consultant on challenging behaviour to the Prader-Willi Syndrome Association of New Zealand [PWSA (NZ)] and as the Australasian representative on the International Prader-Willi Syndrome Organisation's (IPWSO) Carers' and Professionals' Advisory Board. These roles have inevitably contributed to attitudes and priorities that have influenced the way in which I have conducted this research and the conclusions I have drawn from its outcomes.

In this present project, I can make no pretence at being able to consider the subject matter dispassionately. When I engage professional caregivers and people with PWS in the research process I cannot help but be influenced by the joys and intense challenges that led to my enthusiasm for the field in the first place. Nor would I choose to. There are a number of reasons why the provision of support to people with PWS has proven a challenging field for investigators. It seems foolish as well as futile to try to exclude the outcomes of long experience in a field where all evidence is valuable.

Through the research process I have become more aware of some of my own biases, for example I believe that the paid provision of care to New Zealanders who have developmental disabilities and whose behaviour is labelled as challenging has often been a poorly resourced task, I have considerable sympathy with those who choose to make it their

career. In my experience, this work is typically undertaken by paraprofessionals operating in isolation, with little training and often little experience. Yet their role routinely makes demands which any professional would find challenging to meet. I have also found that those who persist in this work often undertake it with conspicuous professionalism and creativity, working towards outcomes that they believe are most beneficial for their clients. It is also my belief that the lived experience of these caregivers represents an under-exploited resource, both for the advancement of our understanding of the psychology of disability and for the development of future supports.

Chapter One

PWS and Contemporary Support Services

PWS in a Food-Rich Environment

PWS is a genetic condition that typically results in learning disability and hyperphagia or overeating. Obesity often arises very rapidly among people with PWS, typically beginning early in childhood unless access to food is limited (Butler et al., 2018). Consequences have been shown to be negative and severe. They include ill health and the loss of mobility, while sudden or premature death has been reported in some cases (Butler et al., 2006, 2017; Hedgeman et al., 2017; Hoybye, 2015).

The support of adults with PWS presents unique challenges to paid caregivers. The promotion of client autonomy and the rights of people with disabilities as citizens are core values within the disability support industry. Caregivers of people with PWS must not only find ways to ameliorate the risks associated with unrestricted access to food for their clients, they must also reconcile the resultant interventions with their other ethical responsibilities. This thesis will explore the ways in which the caregivers of New Zealand adults with PWS experience, conceptualise and address the challenges inherent in their work, especially those associated with their clients' access to food.

What is PWS?

PWS is the most common genetic cause of obesity in humans (Buiting & Horsthemke, 2006). It arises from any one of a number of genetic differences, each of which leads to the absence of expression of one or more paternal genes on chromosome 15, locus q11-q13 (Whittington & Holland, 2004). More than 90% of cases result from either maternal uniparental dysomy (mUPD subtype) or paternal deletion, the deletion subtype being further divided into Type 1 and Type 2 deletions (T1 and T2 deletion subtypes). This distinction depends on where exactly the break occurs. A small minority of cases (<5%) are due to an imprinting defect in the same region (Cassidy & Driscoll, 2009).

Estimates of prevalence vary and are controversial, but a population survey in the Midlands region of the United Kingdom (UK) reported a birth incidence of 1:20,000 live births (Whittington & Holland, 2004). PWS is a multisystem disorder. It results in a range of biological, cognitive and behavioural differences that comprise the syndrome's phenotype. Each genetic subtype also has its own sub-phenotype (Cassidy & Driscoll, 2009; Hoybye, 2015).

In addition to learning disabilities and hyperphagia, PWS is associated with a number of other consequences including an increased incidence of several categories of challenging behaviour, as compared to those with diagnoses of intellectual disability (ID) arising from other aetiologies (Clarke, Boer, Chung, Sturmey & Webb, 1996; Whitman 2013). Some of this behaviour seems unrelated to food (Thuilleaux et al., 2018), but various forms of food seeking behaviour are commonly reported by investigators and by caregivers who have written about their experiences (Goff, 2008; James, 2010; Stanley, 2014; Waters, 2009; Whitman, 2013; Ziccardi, 2006). These behaviours are referred to collectively as foraging. Caregivers typically describe foraging attempts among people with PWS as persistent and often as notably well planned and creative (Goff, 2008; James, 2010; Waters 2009; Ziccardi, 2006).

An Overview of the Project

The present project uses both qualitative and quantitative data to investigate services to people with PWS in Aotearoa/New Zealand. It especially seeks to describe the ways in which access to food is managed. It is based on a review of the existing research literature, which is described later in the present chapter. This review includes a brief summary of the nature and natural history of the syndrome. It also includes an exploration of the central dilemma faced by caregivers who support people with PWS, and a summary of published advice which has been offered to them by researchers or by experienced caregivers. This literature review concludes with an examination of a small group of studies that have directly investigated the experience, beliefs or practices of caregivers.

This literature review has informed three studies. First, a scoping study employing workplace based focus groups made up of professional caregivers (Study 1). Study 1 is described in Chapter 2. A list of open

ended questions was used to generate wide-ranging, relatively unstructured discussions. Results were applied to the design of subsequent studies, particularly by identifying issues which caregivers themselves regarded as priorities for investigation.

Chapter 3 of this thesis describes Study 2, a quantitative study in which an attempt was made to identify and survey every person over 16 years of age diagnosed with PWS within Aotearoa/New Zealand. A range of questionnaires were completed by pairs of respondents, each pair being made up of an adult service user with PWS and a caregiver nominated by that person to support them in the research process. Participant pairs completed a number of instruments. The New Zealand PWS Services Questionnaire (NZPSQ) asked participants to describe their current food management regime. Measures of two key service outcomes were also taken. The NZPSQ included a simple actuarial questionnaire from which body-mass index (BMI) could be calculated, and participants completed a measure of service users' generalised wellbeing known as SF-36. Finally, participants completed an instrument that generated a description of the nature and severity of the individual service user's difficulties in relation to food – the Hyperphagia Questionnaire (HQ).

It was expected that the primary value of Study 2 would be descriptive, but this investigation also afforded an opportunity to pursue two secondary goals. First, it trialled the use of the HQ as a tool for individual needs assessment in the local context. Study 2 also offered an opportunity to begin investigation into relationships between key service or service user characteristics and service outcomes such as BMI and generalised wellbeing.

Study 3 comprised a series of semi-structured interviews with individual professional caregivers or their supervisory managers. These conversations were recorded, transcribed and the resulting text analysed for themes. Interview questions examined the ways in which participants conceptualised the problem of managing access to food, the practical steps by which they addressed this problem, and the processes by which these interventions had come to be selected and implemented. This study is described in Chapter 4.

The results of all three studies are summarised together in Chapter 5. Taken together, they generate a picture of contemporary services which is disturbing in many respects. They also offer insight into the nature of the caregiving role, and of client autonomy within human services. This thesis concludes by suggesting a number of avenues for future investigation and for innovations in caregiving practice.

PWS and Maori

Although a small number of adults who have PWS are known to be supported within kaupapa Maori residential services, many more receive support through mainstream service provision agencies. Choices in relation to these services could be said to be limited for all those who have PWS and their whanau but they are especially so for anyone who seeks a specifically Maori service. It seems reasonable to assume that the challenges associated with the support of service users who have PWS may present differently in different cultural contexts. For caregivers who are Maori, and for others, facilitating access to food is an important part of an array of tikanga and kawa, principles and practices that are central to the expression of cultural values.

This project has not included the gathering of information regarding the cultural identities of either the caregivers or care recipients who have contributed to it. This decision was made to reduce complexity in the data gathering process, given that research in this field is at such an early stage and little is known about the experiences of either group. However, issues of culture remain relevant and important to any understanding of the caregiving task, including the support to people with PWS. It is hoped that these studies will facilitate future investigations.

The Relevance of this Project to the Lives of People with PWS

Despite the fact that PWS has become the subject of a substantial scientific literature many gaps in our understanding remain, particularly in relation to the support of those who have the syndrome. Several groups of investigators have made use of PWS as a means to study issues of more generic concern (Tauber et al., 2014) but few studies have addressed the complex challenges presented by the management of hyperphagia itself (Stanley, 2014).

Decisions concerning services to people with PWS continue to be based largely on clinical consensus and anecdote and it remains very difficult to evaluate the effectiveness of existing interventions. It is known, for example, that adults who have PWS vary in the nature and severity of their difficulties with food, and that these difficulties change over their lifespan (Miller et al., 2011). However, the dearth of tools available for the measurement of hyperphagic behaviour in individuals (Fehnel et al., 2015) means that the pattern of food seeking behaviour across the population of people with PWS has not been described, and that the limits of variations between individuals have yet to be defined. In my experience, this lack of key measures also has implications for individuals. It means that clinical judgement and trial and error generally seem to be the best strategies available for fitting existing services to individual need.

We do not know how many people with PWS there are in Aotearoa/New Zealand. We have only anecdotal information about what services they receive, how effective those services are, or the processes by which they have come to be selected or designed. Very little is understood about the relationships between the characteristics of individuals who have PWS, the various interventions by which caregivers try to protect these service users from the consequences of hyperphagia, and the outcomes associated with those interventions.

It is not known whether some people with PWS remain unsafe in relation to food, nor whether others may be having their rights infringed by being subject to unnecessary or ineffective controls. What is known is that a number of New Zealanders with PWS have suffered severe negative outcomes as the result of unrestricted access, while for many others such access continues to be managed by measures that would be considered unethical and illegal were they applied to other groups. These conclusions are evidenced by the anecdotal reports of local caregivers and by articles in the media (Heather, 2015). They are also supported by the findings of a single local study (Thornton & Dawson; 1990), which concluded that the pattern of expression of the syndrome among adults in Aotearoa/New Zealand seemed similar to that recorded in other countries.

An Australian population survey has noted that only 32% of the adults with PWS who participated were living in supported accommodation outside the family home (Akindola, Kentish, & McColl, 2015).

One explanation for this pattern may be that some adults with PWS simply avoid entering services because of the lack of evidence available about the likely costs and benefits that could be expected to accrue from their doing so.

PWS and Development

From birth, infants with PWS tend to be quiet and inactive. They have low muscle tone, a condition referred to as hypotonia, and so are often described as floppy babies (Butler et al., 2006). It is this feature which usually triggers the diagnostic process. Children with PWS generally remain shorter in stature than their peers as they develop. Typically, they have distinctive facial features, pear shaped bodies and relatively small hands, feet and heads. Physical challenges may arise with development, often from ongoing hypotonia, scoliosis of the spine and endocrine differences.

Feeding difficulties and failure to thrive are common early in life due, at least in part, to diminished sucking and swallowing reflexes (Butler et al., 2006). Difficulties in feeding generally end between two and four years of age, but are replaced by the chronic drive to eat (Miller et al., 2011). This hyperphagia not only seems to be commonly regarded as the defining characteristic of the syndrome, but as the one assumed to present the greatest challenge to parents and caregivers (Whittington & Holland, 2010). The characteristic hypotonia and a tendency to inactivity in many individuals, mean that people with PWS generally require fewer calories to gain weight than do their non-PWS peers (Holland, Treasure, Coskeran, & Dallow, 1995). This means that obesity can arise very rapidly. In the absence of ecological limits on access to food the consequences of obesity may lead to reduced mobility and health effects which are frequently life threatening, even among young people (Butler et al., 2006; Stanley, 2014).

Neither medical nor psychological interventions have proven consistently useful in reducing hyperphagia or in managing the resultant obesity. There is strong, international consensus among authorities that best practice remains the strict control over access to food, enforced by caregivers (Forster, 2010; Holland, 2015; Stanley, 2014; Whittington & Holland, 2010). The effectiveness of compulsory supervision and ecological controls in protecting the health and safety of people with PWS has been confirmed in

several investigations (Cassidy & Driscoll, 2009; Hauber, Stratmann, Hödebeck-Stuntebeck, & Tschoepe, 2013; Kazemi & Hodapp, 2006; Messersmith, Slife, Pulbrook-Vetter, & Bellipanni, 2008; Miller, Lynn, Shuster, & Driscoll, 2013).

Cognition and Communication

Learning and other cognitive abilities are also affected. One large population survey has charted the distribution of full scale IQ (FSIQ) scores among people with PWS and found it to broadly follow the expected normal curve (Whittington, Holland, & Webb, 2009), with the mean IQ around 70 and with significant differences in the profile of subtest scores, compared to non-PWS controls. Several studies have noted that language tends to be impaired. This impairment seems often to be reduced among those with the mUPD subtype for whom the use of language may appear as a conspicuous strength, although Dimitropoulos, Ferranti, and Lemler (2013) have found that relatively strong expressive language skills among this group were not reflected in measures of either receptive language or functional abilities. Dimitropoulos et al. concluded that the relatively strong expressive language skills of people with the mUPD subtype of PWS may mask deficits in other areas, leading caregivers and others to overestimate the cognitive abilities of this group, perhaps exacerbating the risk of misunderstandings leading to interpersonal conflict and challenging behaviour (Dimitropoulos et al., 2013). In fact, anecdotal reports of argumentativeness and a distinctive, inflexible, black-and-white thinking style seem to be common for all those who have the syndrome (James, 2010; Whitman & Jackson, 2006).

PWS and Challenging Behaviour

In adult life, people with PWS show a higher rate of certain medical and psychiatric conditions (Boyall, 2013; Eiholzer & Lee, 2006; Sinnema et al., 2011b; Soni et al., 2007; Thuilleaux et al., 2018). Challenging behaviour is also reported more frequently than among people with ID due to other aetiology (Jauregi, Laurier, Copet, Tauber, & Thuilleaux, 2013; Ogata et al., 2018), and has been reported to be more disruptive and to have a greater impact on caregivers and others associated with the person who has PWS (Sinnema et al., 2011a).

Skin picking is common and often results in lesions which are prone to infection and resistant to healing, in part because the picking behaviour itself is often quite persistent. This behaviour seems to be of organic origin (Miller & Angulo, 2014). Other common behaviours or traits include temper outbursts, stubbornness, a marked resistance to change, and repetitive or compulsive behaviours. Haig and Woodcock (2017) have identified cognitive rigidity as a significant issue for those with PWS, and suggested that it is likely to be the result of relative weaknesses in various aspects of executive functioning, especially task switching. Whitman (2013) has summarised a range of challenges sometimes referred to as the constellation of food related behaviours associated with PWS.

A growing literature has investigated the overlap of phenotypic features between PWS and autism spectrum disorders (Dykens, Lee, & Roof, 2011; Dykens et al., 2017; Ogata et al., 2018). However, there are also many reports of strong sociability among people with PWS. Dykens and Rosner (1999) have found that people with this condition show just as strong a tendency towards helping others as do people who have Williams' syndrome.

Hyperphagia

Hyperphagia is prevalent within PWS, variously reported as occurring in up to 95% (Gratton, Amar, & Watson, 2013) and 98% (Dykens & Shah, 2003) of cases where access to food is not controlled. Reports of experienced caregivers seem to imply that even these statistics may be underestimates, and that at least some difficulty around food should be regarded as close to universal (James, 2010; Waters, 2009).

Hyperphagia is one of a raft of phenotypic features that suggest the involvement of hypothalamic and pituitary dysfunction in PWS; others include low growth hormone secretion, hypogonadism, increased pain threshold, temper outbursts and sleep disturbances.

A wide range of foraging and hoarding behaviours are common in those with PWS. Although true pica (the consumption of non-food objects) arises only rarely, inappropriate food items, such as pet food, frozen food, spoiled or discarded food, are likely to be consumed by some individuals (Whitman & Jackson, 2006). Foraging is often covert, surprisingly well planned and very creative (Boyall, 2013; Goff, 2008;

James, 2010; Stanley, 2014, Whitman, 2013). For many individuals it includes behaviours that might be regarded as dishonest or manipulative, including theft, the telling of untruths (confabulation), and threatened or actual aggression. One experienced caregiver (James, 2010) has used terms such as brilliance and genius to reflect the greater level of insight and planning that many people with PWS seem to have available for the task of seeking extra calories, compared to the pursuit of goals in other areas of life.

Hyperphagia is not only a prevalent problem but also a serious one. Mortality among people with PWS has been estimated as occurring at six times the rate of people who have ID from other aetiologies, complications from obesity being identified as the primary cause (Butler et al., 2018; Einfeld et al., 2006; Hedgeman et al., 2017). Dykens, Maxwell, Pantino, Kossler, and Roof (2007) have pointed out that hyperphagia represents a danger even to slim people who have PWS, as both choking and gastric rupture leading to necrosis are recognised dangers.

Two models of hyperphagia in PWS. The eating behaviour of people with PWS has many similarities with that of the non-PWS population. For example, preferences for certain foods are common and stable over time (Hinton, Holland, Gellatly, Soni, & Owen, 2006). There are also important differences, both in the topography of food related behaviour itself and in its associated endocrinology (McAllister, Whittington, & Holland, 2011).

A disorder of satiety. People who have PWS may seem at first glance to be chronically hungry. However, studies have suggested that their hyperphagia is likely to be more strongly motivated by what happens after food is encountered and consumed than by a continual drive to seek it out. Zipf and Berntson (1987) allowed children with PWS and a control group of children of normal weight free access to sandwich quarters and measured both groups' eating behaviour over time. The investigators found that, although the two groups began eating at the same rate, the children of the PWS group continued for significantly longer before they chose to stop. A subsequent study following a similar design reported that participants with PWS ate an average of six times the number of sandwiches consumed by members of a control group, which was made up of non-PWS children who were obese (Fieldstone, Zipf, Sarter, & Berntson, 1998). Holland et al. (1995) also used the free access to sandwich quarters design, but asked participants to rate their feelings

of hunger or fullness, and their desire to eat, throughout the experiment. They found that satiation was delayed rather than absent in those who had PWS. Participants with PWS ate for longer periods of time than did controls, they consumed on average three times more calories and reported feeling hungry again significantly more quickly.

These results are consistent with those of neuroimaging studies (Hinton et al., 2006; Holsen et al., 2006; McAllister et al., 2011), which have shown abnormalities in the pattern of neural activation in the brains of people with PWS who have been exposed to food. Many of these differences arose in brain structures usually associated with satiety, but most were noted to arise after rather than before eating. A similar study (Holsen et al., 2012) investigated neural activation in response to images of food before and after a standard meal. This activation was noted in two sets of brain structures, one sub-cortical group associated with hunger/satiety and another in the prefrontal cortex associated with self-control. The investigators found that, compared to obese controls, participants with PWS showed post meal hyper-activation in sub-cortical structures associated with food motivation and hypo-activation in cortical regions associated with self-control. This suggests that hyperphagia in PWS is due, at least in part, to the reduced and delayed experience of feeling full after eating. Studies such as these have lent weight to the description of PWS as a disorder of satiety (Cassidy & Driscoll, 2009).

Reward dysfunction: hyperphagia as addictive behaviour. It also seems that the act of consuming food may yield greater reward or hedonic value for those who have PWS. This is suggested by other differences shown in imaging studies, particularly in the hyper-activation of circuitry usually associated with reward after eating (Hinton et al., 2006; Millar et al., 2007). In addition, Hinton, Isles, Williams, and Parkinson (2010) have shown that people with PWS are more motivated by cues associated with food than are the controls. McAllister et al. (2011) noted that the hyper-activation of the reward system after eating in PWS invited a comparison with addictive behaviour and that addiction was a contemporary metaphor in the study of generic obesity. They also record anecdotal reports that once access to food has been controlled, many people with PWS become focused on smoking or some other hedonic behaviour, just as they had been on eating.

Of course, these two models are not necessarily competing; they have a number of features in common. Together they seem to shed light on the findings of Kinash (2007), who conducted interviews with a group of parents and recorded reports that family members with PWS routinely showed no interest in food for several hours at times, usually when otherwise occupied and not exposed to food or cues associated with eating. These models are also consistent with the apparently paradoxical reports of caregivers that their clients who have PWS have often seemed happier after strict controls have been placed on access to food. Caregivers also report that, even though service users who have PWS may be quick to exploit any potential breach in food security, such opportunities often also seem to cause them considerable emotional distress (James, 2010; Prader-Willi Syndrome Association of New Zealand [PWSA-NZ], n.d.).

Managing hyperphagia: the present research literature.

Why are some aspects of PWS so well studied? PWS offers certain unique opportunities for the investigation of a number of issues that are of interest to the general population (Schwartz et al., 2016), hence the literature concerning the condition is quite substantial. Tauber et al. (2014) represent one such research tradition when they describe PWS as a model for human (i.e. generic) hyperphagia. This group investigated unusual levels of the hormones ghrelin (sometimes known as the hunger hormone) and oxytocin in PWS. They concluded that although ghrelin levels do appear abnormal, neither ghrelin antagonists nor oxytocin have any effect on appetite or weight among those with PWS. Other contributions in this vein include von Deneen, Gold, and Lui (2009) and Farooqi and O’Rahilly (2014).

The link between genotype and phenotype has also been investigated through the lens of PWS. This relationship is always complex, but it seems somewhat less so within this syndrome, which also offers a discrete population that is relatively easily defined and observed. Studies using mouse models of the PWS sub-genotypes feature prominently within this literature (Buiting & Horsthemke, 2006; Kubota, Miyake, Hariya, Tran Nguyen Quoc, & Mochizuki, 2016).

Finally, PWS has been seen as a microcosm of certain pervasive problems within human services, particularly the ethical question of how caregivers should respond when a conflict arises between their stated

goal of promoting service users' autonomy and the duty of care which compels them to protect those service users from harm. This has generated a third, albeit much smaller, body of research (van Hooren, Widdershoven, van der Bruggen, van den Borne, & Curfs, 2005; Wullink, Widdershoven, van Schrojenstein Lantman-de Valk, Metsemakers, & Dinant, 2009).

The question of the management of hyperphagia itself has received less attention. In a review of relevant studies, Stanley (2014) concluded that their small number and heterogeneity limited the conclusions that could be drawn from them, and that both the causes and management of hyperphagia in PWS had yet to be determined.

Medical interventions. Sheimann, Butler, Gourash, Cuffari, and Klish (2008) reviewed 60 case studies of bariatric surgical procedures undertaken on patients who have PWS. They noted that some patients reported weight loss, but concluded that these occurred inconsistently and were often minor. The reviewers also noted risks of serious side effects, including increased risk of death from gastric rupture and necrosis.

Whitman and Jackson (2006) provide a summary of early attempts to find a pharmacological intervention to manage obesity in people with PWS. They report that during the 1960s and 1970s patients with the syndrome were often prescribed amphetamines as appetite suppressants. These not only proved ineffective but were also associated with behavioural side effects. More recent trials of generic weight loss medications have yielded mixed results at best, and researchers have often found that negative side effects have proven prohibitive. Investigations into abnormalities in levels of hormones such as ghrelin (Tauber et al., 2014) and the neurotransmitter oxytocin (Einfeld et al., 2014; Kuppens, Donze, & Hokken-Koelega, 2016) have already been noted.

No effective medical tool for the reduction of either obesity or hyperphagia has been found (Driscoll, Miller, Schwartz, & Cassidy, 2014), although investigations continue (Danglas, Reidy, Korner, & Alagarsamy, 2017). Current lines of research include the use of vagus nerve stimulation (Manning et al.,

2016) and manipulations of gut microbiota, which have been reported to yield weight reduction in young people with PWS as well as in others (Zhang et al., 2015).

Behavioural interventions. A number of reviewers report the successful modification of hyperphagic behaviour in people who have PWS using applied behaviour analysis techniques (Gratton et al., 2013; Ho & Dimitropoulos, 2010; Rone, 2010; Spendelow, 2011). However, these reviewers draw on only a small number of original studies. Closer inspection of these source articles suggests that the usefulness of behavioural interventions in practice may in fact be quite limited. Some reviewers have recommend the continued imposition of environmental controls over access to food in order to maintain safety (Dozier, Dracobly, & Payne, 2013) and none overtly contradict this advice.

Page, Finney, Parrish, and Iwata (1983) used a one-way mirror in a paediatric institution to measure food stealing in two children who had PWS. The investigators then applied an escalating interval, differential reinforcement of other behaviour (DRO) procedure to reduce it. Food stealing did indeed reduce, the two study participants both lost weight and the interval for effective reinforcement grew from 10 seconds to a full hour. However, food stealing returned and the weight losses reversed when the participants returned home, even though participants' parents had been trained in the study's procedure and despite the fact that foraging did not reoccur during outpatient visits to the investigators' laboratory.

Page et al. (1983) concluded that an effective lack of supervision at home had been responsible for this regression. There is evidence that factors such as increased supervision and the knowledge among people who have PWS that food is being monitored can reduce food stealing behaviour in themselves. James (2010) records these beliefs among a large group of parents of children with PWS, and Thompson, Kodluboy, and Heston (1980) concluded that the simple acts of monitoring food intake and providing feedback to study participants with PWS were important factors in the reduction of the overeating behaviour which they recorded in their study. It might be argued that what both they and Page et al. (1983) have in fact demonstrated is a surprisingly strong ability on the part of people with PWS to discern when their eating behaviour is or is not being monitored.

Banzett, Marshall, Bowen, Glynn, and Mosk (1991) employed a complex, four-phase intervention in which 10 people with PWS were taught to divide each of their meals into 2 halves and then only to consume one of the resulting portions. After follow up measures averaging 5.3 years, the investigators concluded that although dramatic gains were made by some participants in both food related behaviour and weight loss, the key factors in maintaining those gains were the specification of a goal weight and frequent monitoring by caregivers, who would immediately intervene to reduce food intake if this weight was exceeded.

Maglieri, De Leon, Rodriguez-Catter, and Sevin (2000) were able to significantly reduce both food stealing behaviour and bodyweight for a young woman with PWS by monitoring her behaviour and delivering verbal reprimands whenever she took food from forbidden containers, which investigators had marked with distinctive orange stickers. The subject learned this discrimination task quickly. It was generalised to a refrigerator and other food containers that were marked with the same stickers, and the reduction in food stealing was maintained even after the punishment contingency had been removed. However, it should be noted that researchers seemed to have checked the food inside the refrigerator in the participant's presence. Once again it seems likely that the fact that the illicit food was being monitored was apparent to the participant with PWS throughout the experiment. It cannot be known what food stealing behaviour would have occurred had the fridge been monitored less overtly, and this study could be seen as simply another demonstration of the effectiveness of social cues indicating caregiver supervision.

Two behavioural studies do show a reduction in food stealing behaviour maintained over time and allowing at least some reduction in either supervision or ecological measures for food control. Altman, Bondy, and Hirsch (1978) taught two young women with PWS to monitor their own weight in addition to monitoring already being undertaken by caregivers. The investigators then imposed a regime of close monitoring of eating behaviour combined with contingencies, including reinforcement for reduced calorific intake and weight loss and punishments for illicit foraging. After a time the researchers' monitoring of eating behaviour ceased and only weight was measured, still by the women themselves along with their caregivers. Weight loss and behavioural gains were achieved and one family was able to take the significant

step of removing the locks they had placed on food cupboards, apparently leaving their daughter supported only by the regular weight checks.

Page, Stanley, Richman, Deal, and Iwata (1983) used an escalating schedule DRO procedure to reinforce exercise, along with a response cost contingency for food stealing and hoarding, in support of a 28-year-old woman with PWS who had been hospitalised as the result of obesity. Behaviour change and weight loss were both achieved. When the participant's discharge was planned the researchers modified their intervention and provided training to the staff of her residential home. Weight loss continued in the new environment and even accelerated after the woman moved to an apartment of her own where her programme was described as less structured.

Such single case studies should be interpreted with care, both because individual differences in hyperphagic behaviour seem to be significant among people with PWS (Boyall, 2013) and because there is evidence of spontaneous amelioration during adulthood for at least some individuals (Miller et al., 2011). However, this study is significant in that it shows gains in behaviour and bodyweight maintained by a person with PWS even in the face of a real reduction in caregiver supervision.

Other therapeutic interventions. In a replication of an earlier, single case study Singh et al. (2008) supported three adolescent males with PWS to modify their eating behaviour and reduce weight. This study took place in participants' own homes and a follow up investigation showed that gains were maintained in these settings for several years (Singh et al., 2011). The programme was made up of five components: increased physical exercise, education in food awareness, mindful eating to slow rapid consumption, visualising and labelling hunger, and education in a mindfulness based meditation technique for use when participants were confronted with the temptation to overeat or to steal food. Singh et al. (2008, 2011) were not able to assess the relative effects of the individual components of their programme, but they emphasised the importance of the engagement of parents and family members as its mediators. Unfortunately, Singh et al. (2008, 2011) reported little information about the participants' food environment before, during or after their programme's implementation. It may be that their success in engaging parents led to the more consistent application of existing food control measures.

The conclusions drawn by Singh et al. (2008, 2011) are especially interesting in the light of a study by Benarroch et al. (2012), which also featured adults with PWS successfully limiting the amount they ate while working towards corporate goals. Benarroch et al. (2012) described 22 out of 29 (76%) Israeli adult and adolescent volunteers with PWS successfully completing the traditional 25 hour fast for Yom Kippur, the Jewish Day of Atonement. Many of these volunteers achieved this feat despite living in secular settings, where fasting was not the norm and standard meals were available at any time without sanction for both non-fasting residents and those who wanted to break their fast.

Investigators found that successful fasting was not predicted by any of the medical or genetic variables they measured, including genetic subtype, growth hormone status or BMI. Nor was it correlated with participants' degree of religiosity or their susceptibility to behave in socially desirable ways. Success was positively correlated with a higher pain threshold and, to a lesser extent, with participants' confidence in their own self-control. Success was also predicted by caregivers' subjective ratings of how important the fast was to each participant. Clearly some factor or factors motivated a group of young people with PWS to deny themselves food, and a large proportion of them proved able to do so for more than a full day. Moreover, whatever these motivating factors were, experienced caregivers were apparently able to detect them and judge their strength.

As noted, Kinash (2007) has recorded the belief within a large sample of familial caregivers that food was sometimes apparently forgotten while their family members with PWS were busy and free from food related cues. There are also some interesting similarities between Benarroch et al.'s (2012) report and the programmes applied by Singh et al. (2008, 2011). Both involved considerable preparation and both engaged people with PWS in goal oriented activity alongside those around them. Notwithstanding Benarroch et al.'s conclusion that the tendency to make socially desirable responses was not predictive of successful fasting, it may be that this kind of social engagement (membership within a group and the opportunity to participate in activities towards corporate goals) may carry particularly strong intrinsic motivation for some people with PWS. Benarroch et al. point out that the fact that they were able to exclude

participants' degree of religiosity as a determining factor bodes well for this study's application to the lives of people with PWS in other settings.

Environmental controls over access to food. There is evidence which supports the consensus view that the control of access to food, achieved through high levels of supervision and/or the instillation of physical barriers, is effective in the prevention of obesity and the improvement of health and safety outcomes for people with PWS (Cassidy & Driscoll, 2009; Kazemi & Hodapp, 2006; Messersmith et al., 2008; Miller et al., 2013). Several authors also suggest that once access to food is completely controlled emotional wellbeing and behaviour may also be improved (James, 2010; Jauregi et al., 2013; Whitman & Jackson, 2006). What is more, it seems that caregivers have few options available to them. Reviews of alternative medical, behavioural or therapeutic interventions have concluded that, while some may have yielded benefit for individuals, none have been shown to be consistently effective across the syndrome (Goldstone, Holland, Hauffa, Hokken-Koelega, & Tauber, 2008; Stanley, 2014; Whittington & Holland, 2010).

The Central Dilemma Facing Caregivers

The promotion of self-determination and the recognition of service users' rights as citizens are key values in the provision of support to adults who have developmental disabilities (United Nations Office of the High Commissioner for Human Rights, 2014). In New Zealand these values are reflected in the New Zealand Disability Strategy (Office for Disability Issues, 2001), and in plans for this strategy's implementation (Office for Disability Issues, 2014), as well as in the guiding documents of service agencies and advocacy groups (Convention Coalition Monitoring Group, 2012; IHC New Zealand [IHC], 2011). Inherent in this ethic is the risk that service users may make choices that seem to others to be to their detriment. Whenever this occurs, both familial and professional caregivers face a conflict between their commitment to the promotion of their family member or clients' autonomy and to the duty of care which requires them to protect those they support from harm and to act in their best interests (Curryer, Stancliffe, & Dew, 2015; Evans, 1999; Gill & Fazil, 2013).

This dilemma is particularly stark in the support of adults who have PWS. The promotion of autonomy and choice making may be foundational ethical principles for caregivers supporting adults with disabilities, but for people with PWS such self-determination can be expected to result in outcomes that are negative and severe. The question of how to balance service users' safety with the commitment to promote their independence represents the central dilemma facing both familial and professional caregivers of people with PWS. This dilemma has been characterised in a number of ways, but primarily as a conflict between two opposing sets of values, autonomy versus safety, or the rights of people with PWS as citizens versus paternalism (Dykens et al., 1997; Hawkins, Redley, & Holland, 2011; van Hooren, Widdershoven, van den Borne, & Curfs, 2002).

Many people who have PWS are committed and articulate self-advocates (Dimitropoulos et al., 2013, James 2010), they are well equipped to protest any perceived unfairness or breach of their rights. People with PWS often also go to considerable lengths to obtain food. Their attempts at foraging can be very creative and include a variety of challenging behaviours (McAllister et al., 2011; Whitman, 2013). Any caregiver who supports an adult with PWS must not only address this central ethical dilemma, but must find a resolution that is practical, sustainable over time, and able to bear the scrutiny of service users and society at large.

To date, studies investigating the outcomes of services to people with PWS have been limited to those that measure service users' bodyweight and certain aspects of their physical health, such as diabetic status. This means that caregivers, advocates, and of course people with PWS themselves, have only limited evidence on which to base their decisions regarding their engagement with these services.

Holland and Wong's resolution. Several groups of researchers have offered guidance to caregivers facing this central dilemma (Dykens et al., 1997; Holland & Wong, 1999). Holland and Wong's resolution was typical. They argued that since hyperphagia in PWS seems to arise from organic causes, and since decision-making around food also appears to be compromised, caregivers are not only justified in imposing restrictions on their clients' access to food, but doing so in fact represents an ethical imperative, the neglect of which is itself unacceptable.

Today, Holland and Wong's resolution represents the dominant view worldwide. This is reflected in the writing of leading researchers (Holland, 2015; Hoybye, 2015; Whitman & Jackson, 2006), and in the views of clinicians, service provision agencies (Pittsburgh Partnership, n.d.; Prader-Willi Homes of Oconomowoc, n.d.), and even advocacy groups (IPWSO, n.d.; IPWSO, 2013). Holland and Wong's view is supported by caregivers and clinicians who have written about their experiences and offered advice to their colleagues (Forster, 2010; Goff, 2008; James, 2010; Waters, 2009; Ziccardi, 2006), and its influence is reflected in the work of Hawkins et al. (2011) who completed an ethnographic study among British caregivers of adults with PWS. They found that the concept of risk, and in particular risk arising from access to food, was a central determining factor in both caregivers' practice and the policy framework within which they worked. The consensus is supported in the local context by the PWS Association of New Zealand (PWSA (NZ), n.d., *Dietary management*; PWSA (NZ), n.d., *Guidelines for residential caregivers*).

There is strong international agreement that the support of persons with PWS should include the imposition of a calorie controlled diet and compulsory restrictions on access to additional food, or to financial resources which are regarded as likely to be used to access food. (Holland, 2015; Jauregi et al., 2013; Wyatt, 2006). In practice, highly intrusive measures are often required to achieve this kind of control. Those measures commonly advocated and employed include locks placed on refrigerators and food storage areas, the exclusion of people with PWS from food related activities like shopping and cooking, compulsory participation in exercise programmes and weight checks, prescribed diets, the appointment of financial agents to control personal finances, and the imposition of close supervision even during outings and social events (Goff, 2008; IPWSO, n.d.; 2013; James, 2010; Waters, 2009; Ziccardi, 2006).

Further claims for the benefits of restricted access to food. As already noted, there is evidence that environmental controls over access to food are effective in reducing bodyweight and improving health and safety outcomes for people with PWS, as long as they are able to be maintained consistently over time (Cassidy & Driscoll, 2009; Hauber et al., 2013; Jauregi et al., 2013; Kazemi & Hodapp, 2006; Messersmith et al., 2008; Miller et al., 2013).

Such restrictive practices might be expected to lead to increased challenging behaviour and to have a detrimental effect on individual and corporate wellbeing. In fact, the opposite effects are often claimed by caregivers who report improvements in a number of quality of life (QoL) domains following the implementation of such controls. Improvements in emotional wellbeing, behaviour and/or independence are often said to arise once access to food is completely controlled (Goff, 2008; James, 2010; Jauregi et al., 2013; Whitman & Jackson, 2006). This belief is reflected in a popular motto regarding the support of people with PWS in relation to food, “No doubt. No hope. No disappointment” (Gourash, Hanchett, & Forster, 2006; Prader-Willi Homes of Oconomowoc, n.d.).

“Food security”. The term food security is used to describe the objective of these controls. It often seems to refer to an emotional state within service users as much as to characteristics of the service environment itself. Food security is seen as the antithesis of uncertainty about access to food, which is believed to contribute to a range of undesirable outcomes. The view that food security leads to QoL gains for those with PWS, as well as being a prerequisite for safety and adequate care, is central to the practice of most agencies and is prominent in publications by clinicians, researchers and associations of parents and caregivers of people who have PWS (Butler et al., 2006; Forster, 2010; Hoybye, 2015; IPWSO, n.d.).

There is no credible voice in the current scientific, professional or advocacy literature which promotes, or even permits, unrestricted access to food for adults or children who have PWS. If the central ethical dilemma facing the caregivers of people with PWS can indeed be represented as a simple choice between self-determination and paternalism it would seem that it has been resolved firmly in favour of the latter.

Some problems with the present consensus. Despite the strength of this agreement some difficulties remain. Some of these stem from Holland and Wong’s (1999) apparent reduction of the central dilemma to a simple, dichotomous choice: safety versus autonomy. This not only seems an over simplification but it ignores the likelihood that care recipients who have PWS and their caregivers may value a broader range of outcomes. It fails to reflect caregivers’ own descriptions of the complex processes by which decisions concerning access to food are made, and the range of valued outcomes that drive those

decisions (van Hooren, Widdershoven, Candel, van den Borne, & Curfs, 2006; van Hooren et al., 2002; van Hooren et al., 2005). There is also a lack of research evidence for many of the secondary benefits claimed to accrue from the establishment of food security. Finally, the suggestion that some people with disabilities may best be empowered by having certain choices taken away from them is a serious one and should not be allowed to pass without comment.

The views of caregivers. Van Hooren et al. (2005) conducted interviews with a large group of caregivers who supported children or adults with PWS. Both familial and professional caregivers were included in their sample. Van Hooren et al. asked caregivers to describe both the interventions that they had undertaken to manage care recipients' access to food and the processes by which they came to plan and implement those measures. These caregivers reported views that were in agreement with Holland and Wong (1999) on a number of important points; none advocated that people with PWS have unrestricted access to food and all reported imposing at least some form of compulsory control over diet. However, these caregivers did not report simply choosing between safety and autonomy. They described decision making processes that were complex, individualised and informed by a much broader range of values.

The caregivers interviewed by van Hooren et al. (2005) reported seeking to balance multiple desired outcomes. In particular, they noted that it was not only their priorities in relation to the person with PWS that played a role in the decisions they made about access to food. These choices were also influenced by outcomes being sought for others in the caregiving environment and for the wellbeing of the family or group as a whole. Van Hooren et al. (2005) summarised the range of values that they found to be important in guiding caregivers' decisions around food under four headings. Three of these referred to outcomes sought for the person with PWS who was receiving care (physical health, wellbeing or emotional health, and the preservation of a sense of balance between personal responsibility and freedom of choice). The fourth, which van Hooren et al. labelled as the search for "A liveable life" (p. 309), described caregivers' quest for a support regime that was sustainable over time and led to a life regarded as meaningful and valued, not only for the person with PWS receiving support but for all of the people in the social context within which that care took place.

Caregivers' search for a liveable life reflected their concern for a sense of corporate, as well as individual, wellbeing. This concept seems to have much in common with that which is referred to in the research literature concerning caregivers of children with developmental disability as Family Quality of Life (FQoL) (Boelsma, Caubo-Damen, Schippers, Dane, & Abma, 2017).

Can the imposition of controls be considered empowering? Perhaps the most immediate challenge arising from the consensus view concerns the implication that some people with developmental disabilities are best empowered, and may have their wellbeing enhanced, by having certain rights and choices taken away from them. Two groups of studies have been cited to support the consensus position. They employ very different research methods but both investigate differences in the drive to eat between people with PWS and others. Together they provide a rationale for the view that, for people with PWS, unrestricted access to food might well be seen as a threat from which affected individuals should be protected (Holland, 2015).

As already noted, Holland et al. (1995) allowed children with PWS and a group of non-PWS controls free access to food (sandwich quarters). They recorded participants' eating behaviour and asked them to rate the strength of their feelings of hunger over time. Their results confirmed earlier work by Zipf and Berntson (1987), which showed that although children with PWS and non-PWS controls began eating at the same rate, participants with PWS ate for much longer before they achieved satiety. Those with PWS also reported feeling hungry again and returned to eating much more quickly. Subsequent neuroimaging studies (Hinton et al., 2006; Holsen et al., 2012; McAlister et al., 2011) have noted several differences between the brain activity of people with PWS and others in relation to food. The majority of these differences have been noted only after the person with PWS has become aware of the food or begun to eat. These findings have led to the view that it is the presence of food itself, and in particular the sensations associated with beginning to eat, which play the most important role in motivating excessive consumption.

Holland (2015) has argued that these two sets of findings support the anecdotal reports of caregivers and clinicians (Forster, 2010; Gourash et al., 2006; James, 2010; Waters, 2009) that external social and ecological controls over access to food tend to reduce anxiety. This anxiety could otherwise be expected to arise, both from exposure to food itself and from uncertainty about the likelihood of future access to it.

Holland frames control over access to food as a compassionate response by caregivers to a group of service users who have unique needs. Holland has suggested that ecological restrictions are not only necessary for safety, but are also likely to reduce challenging behaviour and lead to a net increase in the range of choices and opportunities available to the person with PWS who is subject to them.

The limits of this justification. Holland (2015) has framed this question of support to people with PWS around food as an ethical special case. He argued that controls were not only necessary to protect physical safety and enhance wellbeing, but also that the apparently free choices of those who have PWS in relation to food were, in reality, largely determined by the neurological and endocrinological consequences of the syndrome. Holland and his colleagues have consistently taken care to emphasise that their position cannot be used to negate respect for individual service users' legal rights (Holland, 2015; Holland & Wong, 1999) and there are clear limits to the range of interventions that their argument can be used to sustain. For example, it cannot be generalised to justify limits over the choices of adults with PWS in areas unrelated to food or to abrogate caregivers' responsibilities to promote self-determination as a global aspiration. Nor does Holland's work provide justification for the proposal that external controls over access to food should be imposed on others without PWS, but who have ID and face serious health risks mediated by diet and obesity, as has sometimes been suggested (Grondhuis & Aman, 2014).

A way forward. Holland's work (2015) has provided an ethical justification and a theoretical basis for the present consensus, despite the weaknesses exposed by the work of van Hooren et al. (2002, 2005, 2006). It also suggests a number of testable predictions. If Holland and others are correct we should expect to see changes in measures of mood, behaviour and individual and corporate QoL when people who have PWS and significant difficulties in relation to food move between environments that feature different levels of food security.

Conclusion

There is a strong and widely-held consensus that limits should be imposed on the freedom of people with PWS to access food. The research literature supports the view that the risks posed by unfettered access

for many people who have PWS are real and significant, even though questions remain about variations in the nature and severity of those risks among individuals. It has also been established that food security can indeed ameliorate risk and lead to a lowered BMI and improved health outcomes for people who have PWS.

Clinical consensus and anecdotal reports claim additional social and emotional benefits for the imposition of food security. Holland (2015) has made the point that these benefits seem consistent with the results of neuroimaging and behavioural studies which suggest that many of the behavioural and neurophysiological differences between people with PWS and others in relation to food seem to arise only after an individual is exposed to food related cues or has begun to eat. Restrictions placed on those cues by caregivers may then be regarded as both a compassionate and an empowering response to the unique needs of a person who has PWS, as well as one which promotes that person's health and safety.

Holland's (2015) analysis adds new information to the ethical equation referred to here as the central dilemma facing caregivers of people who have PWS. It frames unrestricted access to food as an emotional and social threat to the wellbeing of service users, as well as a physical one. This claim seems consistent with reported caregiver experience, but remains largely untested because studies that have evaluated the outcomes of food secure environments have tended to collect only measures of service users' bodyweight and health status. Other service outcomes have been neglected even though some may well be regarded as important questions for caregivers and for those who have PWS themselves. The impact of imposed food security on service users' emotional experience, challenging behaviour, community engagement, functional skills, relationships and corporate or individual QoL have received little attention.

The work of van Hooren et al. (2002, 2005, 2006) has suggested that the characterisation of the central dilemma facing caregivers as a simple, bi-polar choice between opposing sets of values is at best an over simplification, as well as being at variance to the reports of caregivers themselves on a number of points. Neither the reports of van Hooren et al. nor the narrow range of outcome data used to evaluate the effectiveness of food secure environments in other investigations constitute grounds for the rejection of the consensus view; the caregivers interviewed by van Hooren et al. (2005) also reported that they imposed compulsory controls over food. However, these challenges do highlight the need to develop a more

sophisticated model of the relationship between caregivers and people with PWS, one that is better grounded in an understanding of existing services and their outcomes.

Chapter Two

Study 1: Initial Interviews in Workplace Focus Groups

Little is known about the experiences and habitual practices of paid caregivers who support people with PWS. Study 1 took the form of a series of semi-structured interviews held with workplace based focus groups, each made up of caregivers who worked together in support of one or more individuals with PWS.

The primary purpose of this investigation was to inform the design of subsequent studies and for this reason it has been labelled a consultation with caregivers. The rationale for Study 1 was similar to that offered by Davis, Drey and Gould (2009) who described scoping studies as reconnaissance towards the identification of synthesising concepts. In this case, such reconnaissance was deemed necessary for a number of reasons. First, even though the genetics and medical implications of PWS have each generated a substantial body of research, little is known about services to people with PWS in Aotearoa/New Zealand. Second, the gathering of information concerning the issues which caregivers themselves regarded as research priorities seemed sensible. Finally, Study 1 allowed the investigator an opportunity to explore and challenge existing beliefs concerning research priorities in the field of the provision of residential support services before the designs of Studies 2 and 3 were finally determined. This consultation particularly informed the design of Study 3. It afforded the opportunity to trial a number of specific interview questions in order to test their usefulness in generating discussion.

Ethics

Ethical considerations in the design of Study 1 were complicated by the need to protect the interests of service users with PWS and service agencies, as well those of the professional caregivers who took part in the study. Copies of the study's information sheet was first distributed to service agencies which employed eligible caregivers, and support for the study was sought from these organisations senior managers or governance boards. It was agreed that no agency would be identified in any report of the study's results. Separate consent was obtained from participant caregivers. Information supplied to potential participants

stated explicitly that, despite their employers support for the study, their participation remained entirely voluntary and no sanction for non-participation would be imposed.

No identifying information regarding any service user or individual caregiver was recorded in this study, and responses were prepared by which the interviewer would use to decline any requests to provide impromptu supervision or service advice to participating caregivers, either during the course of the focus groups or at any other time during the study. Approval for Study 1 had been sought and gained through the Massey University Human Ethics Committee as a low risk activity.

Participants' responses were unexpected in a number of ways, the degree of enthusiasm with which individual participants, and the agencies that employed them, responded to the opportunity to talk about their work being a good example. Participants also showed a surprising degree of consensus on key issues, and although no participant became overtly distressed during our discussions, the depth and diversity of emotion generated for participant caregivers when prompted to reflect on their experiences in support of people with PWS was also unexpected. The responses of participants in Study 1 informed the ethical review process in planning for Study 3 (Human Ethics Committee (HEC): Southern A Application 15/56).

Method

Semi-structured Interviews in Workplace-based Focus Groups

Study 1 is based on a number of semi-structured interviews. Because of the seminal nature of research into the experiences and practices of caregivers to people with PWS, it was considered important to adopt a methodology which allowed the researcher to meet face-to-face with participants, and one which facilitated wide-ranging dialogue and the co-construction of content. DiCicco-Bloom and Crabtree (2006) have written a summary of the usefulness of interviews within qualitative research. They identified exactly these strengths within the semi-structured interview format whereby a relatively small number of open-ended questions are used to generate discussion which is subsequently guided by both participant and interviewer.

The decision was also made to canvass the views of caregivers through discussions within workplace-based focus groups. Several researchers have noted that the element of interaction among participants offers a number of advantages over methods which rely on individual interviews. Wilkinson (1998, 1999) and others (Barbour, 2018; Kidd & Parshall, 2000; Wilkinson & Silverman, 2004) have made the point that these group dynamics often offer insight into the co-construction of meaning within a group. Wilkinson (1998, 1999) has made the additional point that focus groups may also go some way to addressing the power imbalance between researcher and participants, and hence yield a better description of participants' meaning, including aspects of language and culture which participants might otherwise feel obliged to modify if they were interviewed alone. Within the context of research into services to people who have disabilities, it has been noted that focus groups are useful for capturing the voice of groups usually excluded from discussions about service development or evaluation (Kroll, Barbour, & Harris, 2007). Such comments are usually made with reference to groups of service users but they could equally be applied to direct caregivers.

Barbour (2018), who has written a guide to research using focus groups, has emphasised the importance of participant sampling to successful outcomes. In the case of Study 1, the use of existing groups of caregivers which were largely isolated from each other could be seen as a factor limiting the usefulness of the results. However, as Barbour (2018) has also noted, such naturally occurring focus groups also offer a number of advantages for researchers, including familiarity among participants and an existing group culture and history which may facilitate discussion and analysis. The choice to make use of existing groups of caregivers was also important for Study 1 because it overcame difficulties arising from the need to respect clients' personal information, as well as logistical challenges arising from geographical spread of participants and the nature of professional caregiving as shift-work. The limitations inherent in this choice have informed the subsequent analysis of results. The use of existing groups of carers may have also contributed to the free-flowing and focused nature of the discussions which resulted, and to the subsequent, richness of the data generated.

Participants and Recruitment

A group of support agencies were identified through the PWSA (NZ) database. Each of these organisations provided care to at least one adult diagnosed with PWS within the North Island of New Zealand. Five such agencies were invited to select a group of direct care staff and/or their first level managers who were then approached by the researcher. No agency or individual caregiver declined to take part in this study, although the logistics of travel and time were important factors in determining which agencies were selected.

All participants regularly provided direct support to one or more adults with PWS. They worked in a variety of residential service settings that included individualised services in which a person with PWS was supported to live alone, PWS specific group homes in which a number of adults with the syndrome lived together, and mixed group homes in which residents with PWS lived alongside others with ID who did not have PWS. The length of participants' support to people with PWS ranged from 12 months to more than 40 years, and the majority of interviewees reported supporting multiple service users with the syndrome. The number of caregivers participating in each discussion ranged from 1 to 4. A total of 12 caregivers participated in this study, 4 of whom also carried service management responsibilities.

Procedure

A single list of questions was prepared (Appendix F) and used for each group discussion. It included some questions that concerned basic demographic information, such as participants' present role and length of service, as well as others which were open ended and designed to encourage discussion. Questions were asked about the aspects of participants' work that they associated with the greatest challenge and with the greatest personal reward. Other questions focused on the interventions through which caregivers managed their clients' access to food and the processes through which decisions about these measures had been made. The service features that they regarded as most important for service quality were also investigated. Follow-up questions were prepared to encourage discussion, but these were seldom required because the initial questions proved quite successful in initiating conversation.

Discussions took place at venues arranged by each employing agency. They ranged in length between 30 and 90 minutes and were generally free-flowing and wide-ranging. This was especially the case in those that featured larger groups of caregivers. Discussions were recorded via hand-written notes taken by the researcher in full view of participants. No video or sound recordings were made and so only short quotations were documented verbatim. Group members were offered the opportunity to read and modify the investigator's notes; none chose to do so. Care was taken not to record personal or identifying information about any service user.

The tone of discussion within these focus groups was invariably positive, calm and professional, even when caregivers' responses also seemed to provoke emotion. Discussions were often quite animated, with stories and laughter featuring prominently in all. Caregivers' language was generally strength based and notably respectful when referring to service users.

Analysis

These discussions yielded a rich data set. The goals of this study were achieved while its outcomes and the experience gained in conducting it contributed significantly to subsequent investigations. Analysis was limited by the way in which conversations within focus groups were recorded, the use of hand written notes rather than an audio or digital recording meant that no transcript could be generated.

Analysis was limited to the semantic level and was largely comprised of the collation of participants' responses. This was especially true for those service features that participants regarded as most important for service quality and as the most significant sources of reward or challenge. The results of these interviews also allowed the usefulness of individual questions to be assessed; most were deemed effective in provoking relevant discussion. The usefulness of the group discussion format was also critiqued.

Results

Each workplace based focus group was convened independently without reference to the others. Different types of support agencies were represented, working in very different community contexts: urban, rural, small towns and an intentional community. Despite this diversity, conspicuous agreement arose on

each of the key points we discussed. Several conclusions could be described as universal in that they featured in every discussion and were never contradicted.

Food Management

Participants' responses showed particularly strong consensus in this area, they were also broadly consistent with those reported in overseas studies (Hawkins et al., 2011; van Hooren et al., 2002, 2005, 2006) and by caregivers who have described their own experiences (James, 2010; Ziccardi, 2006). No participant advocated free access to food for any service user who had PWS; all reported imposing controls.

Specific measures described by participants included environmental restrictions such as locks on the doors to certain rooms and to food storage cupboards and refrigerators. Some groups of participants reported imposing bans on residents with PWS entering the kitchen in their homes without supervision, or from them participating in food related activities like cooking or shopping. Some residents were required to participate in being weighed regularly. Diets and menus were generally planned by caregivers, often with the support of a professional dietician. All services encouraged regular exercise and a minority regarded it as compulsory for residents. There was universal agreement that although many of these measures breached the common ethic of support for service users' autonomy, they were nevertheless an essential part of support to residents with PWS.

Most participants reported that they imposed additional controls when residents with PWS left their homes. Caregivers typically exerted control over residents' personal finances and often reported that they continued to supervise residents while they shopped or attended social outings in order to prevent food stealing or the seeking of food from others.

A significant minority of participants (four caregivers representing two agencies) reported ethical and practical barriers which they believed prevented them from exerting control over their clients' access to food outside of the residential service. These groups of caregivers reported the belief that in order to impose such restrictions they would need the permission of the adult residents with PWS themselves, and of their parents. They also reported that the fact that they were unable to control service users' finances and to

supervise their behaviour in public represented significant weaknesses in the support they provided. These participants reported that such weaknesses had resulted not only in undesirable weight gain and safety risks for their clients, but also in intense disagreement between their clients with PWS and themselves. These caregivers attributed the frequency of these conflicts in part to their inability to establish firm food security.

Other caregivers reported that some of their clients with PWS had developed significantly increased self-control around food. For example, one caregiver described a person with PWS having learned to attend social events without direct supervision and without overeating. Adequate support was reportedly provided through the combination of regular compulsory weight checks and by that service user meeting with the caregiver before and after each social event. The purpose of these meetings was to plan what the person with PWS would eat during the social event, and then to debrief regarding the effectiveness of that plan. Participants always described this greater degree of independence as a goal which had been achieved through careful teaching over several months or years, and as one which remained dependent on a context of consistent support by familiar support people. It was considered especially important that residents remained aware that both their weight and their access to food at other times would continue to be closely monitored.

Universal, or near universal, consensus also arose among participants in two other areas related to the management of access to food. Participants emphasised the need for such measures to be individually designed in order to maximise their efficacy and to minimise the degree of intervention required. Where participants had supported more than one person with the syndrome the differences in the nature and severity of those individuals' difficulties with food were often emphasised. Most participants also reported a firm belief that such measures were not only effective but necessary. This conclusion was based on the outcomes that these caregivers had observed as arising from variations in the level of food security in the past. Several caregivers described experiences in which they had seen residents with PWS suffer rapid weight gain as the result of the neglect of food security. Hospitalisation, increased disability, and premature death were all noted as consequences which participants had witnessed personally.

Other examples were described in which the successful imposition of controls over access to food was associated, not only with weight loss, but with increases in functional abilities, emotional wellbeing,

and independence in areas of life not related to food. Participants often associated effective food control with a reduction in the frequency and intensity of challenging behaviour. Many responses in this area were articulated clearly, calmly, but also with notable firmness.

Experiences Identified by Caregivers as Most Challenging

Challenging behaviour. Participants were asked to identify the aspects of their work that they found most challenging. Once again, their responses showed a strong consensus. All participants made reference to their clients' behaviour in response to this question but the management of hyperphagia and obesity were only seldom identified specifically. This result was surprising given the strong emphasis on the issue of the management of hyperphagia within the professional literature (Forster, 2010; Holland, 2015).

A variety of specific behaviours were identified as particularly challenging, including physical aggression and damage to property. Lying or the telling of untrue stories was the most commonly reported. This behaviour among people with PWS is often referred to as confabulation, in this study it was reported in a variety of contexts. In many accounts confabulation seemed related to the search for food, functioning either as a means by which residents with PWS sought to gain food by deception or to cover up past attempts at foraging. In other accounts, this behaviour was described as a feature of interpersonal conflict that seemed to have been unrelated to food. In still other accounts, confabulation seemed to have been used by service users with PWS simply as a means to start or maintain a conversation.

Outbursts of anger, typically referred to as tantrums or meltdowns, were also commonly identified as a source of significant challenge for caregivers. Accounts of specific incidents sometimes featured descriptions of verbal and physical aggression. Tantrums were often interpreted as being related to residents' attempts to seek food, but on other occasions they seemed to have been precipitated by other factors, such as interpersonal conflict, difficulty dealing with change, or a desire by a service user with PWS to generally exert control.

A third category of behaviours was referred to by participants as particularly challenging. These could broadly be termed behaviours of excess and they included the collection or hoarding of various items

and long periods of time spent on favourite activities to the exclusion of others. Excessive use of internet chat rooms and special interest community groups were reported, as well as excessive gambling, smoking and drinking water. Accounts of these behaviours often included parallels drawn by participants to their clients' excesses in relation to food.

Participants commonly emphasised the intense emotional impact that the behaviour of their clients with PWS had on themselves as caregivers, and on others associated with the residential service. This remained true even when the specific behaviour described was essentially verbal in nature, such as in accounts of confabulation, perseveration or simply arguing. Participants sometimes described a single incident of this challenging behaviour as particularly stressful, but more frequently they emphasised the behaviour's ongoing nature, its high frequency and its resistance to their efforts to change or prevent it, as factors that intensified the behaviour's effect on fellow residents and on caregivers themselves. One participant who had several years' experience in the support of both people with PWS and those with ID arising from other causes described PWS as "*The most challenging syndrome*", while another reported being "*Shit scared*" for much of her early career. A third commented:

"I used to think, 'I don't like this syndrome, give me a different syndrome to work with.'"

Narratives about the successful resolution of challenging behaviours also arose in every interview, at least to some extent. They were often described as the fruition of a process whereby the resident with PWS had come to trust their caregivers and/or to accept limitations imposed on them in relation to food. This process was itself usually framed as the product of diligent efforts by a team of caregivers working together over a considerable period of time. "Consistent" and "fair" were adjectives commonly used to describe successful support.

Not all stories of challenging behaviour featured positive outcomes generated by caregivers. In some accounts, challenging behaviour was only reduced after changes were made to the residential service itself, often by a resident with PWS moving to a new service or being supported to live alone. Participants related several stories in which two or more residents with PWS came into conflict with each other and were

unwilling to continue living together; the view among participants seemed to be that these residents communicated their displeasure through their behaviour. On other occasions resource limitations such as deficits in the physical environment of a particular residential home, or in the technical advice or level of funding available to caregivers, were described as having made a particular service untenable.

Within these narratives it was nearly always the challenging behaviour of one or more residents with PWS that seemed finally to provide the catalyst for change. Such systemic changes often took a significant time to accomplish. Caregivers who reported these delays also reported feelings of frustration in the face of the continued challenging behaviour and/or weight gain that resulted in the meantime.

Service users' relationships with food. Caregivers identified the management of access to food for residents with PWS as a significant challenge in itself, although it was generally characterised as a relatively concrete and straightforward one when compared with the challenges associated with other aspects of service users' behaviour. One caregiver described food management as "*The easiest challenge*" and several reported that although establishing new measures for the control of access to food was often difficult, once the appropriate regime was in place its maintenance could be expected to prove relatively straightforward.

Achieving the necessary consistency of practice across a team of caregivers was identified as an important contributor to positive service outcomes, as well as being a significant challenge in itself, both for direct caregivers and for service managers. These views were reported in a number of different ways but they also featured in every interview. Sometimes this challenge of maintaining consistency was expressed as a difficulty in establishing effective communication between carers who were rostered to different shifts, and so seldom had contact with their colleagues. It also featured in descriptions of pastoral care provided to colleagues under stress and of efforts to resolve disagreements between caregivers, as well as in the strong emphasis placed on processes for recruiting and indoctrinating new team members. A struggle for consistency between study participants and residents' families was also noted several times.

Service Features Identified as Most Important for Service Quality

Greater variation was shown in participants' responses to this question, although a strong element of agreement was still evident. All caregivers identified the management of access to food as important, but this was seldom their first response, nor was it the factor identified as of primary importance for service quality. Caregivers seemed to regard the management of food as a necessary but not sufficient condition for desired service outcomes, one that had to be met as a means to achieving other, more sophisticated and meaningful ends – *"Food is a given"* as one caregiver put it.

Most caregivers identified some feature of the relationship between caregiver and care recipient to be of primary importance. Several participants described a sense of mutual valuing or positive regard in this context. Participants also identified respect for residents' autonomy on the part of caregivers and the establishment of trust in caregivers on the part of residents who have PWS.

As noted, almost all caregivers also made reference to the importance of consistency as a feature of effective support, some described it as the single most important factor in determining service quality. Consistent support seemed to be regarded as that which was predictable for service users and not subject to unplanned change. To constitute consistent support this predictability was required both in the work of each individual caregiver over time, and between those caregivers who worked different shifts to support the same clients. Some caregivers reported that consistency was particularly important for the management of residents' anxiety. Inconsistencies such as unexpected changes in routines, in personnel or in the expectations placed on residents were believed to increase the risk of uncertainty for service users and to contribute to subsequent anxiety, which in turn was believed to increase the risk of challenging behaviour. Such inconsistency was regarded as likely to be especially dangerous if service users believed that it implied either a threat to their expected access to food or the possibility of increased access.

All participants seemed to regard consistency in the application of rules around the management of food as playing an essential role in moderating risks to health, but also in reducing the frequency and intensity of challenging behaviour. Several recounted stories of negative outcomes that had arisen from the

neglect of consistency. The presence of team members who did not act consistently was generally regarded as an unacceptable liability because of the likely severity of these consequences. There also seemed to be a consensus among participants that both a certain quality of temperament and a unique body of knowledge were prerequisites for anyone seeking to become a safe and effective support person for clients with PWS. Participants were clear that even those of their colleagues who had considerable experience in the support of people with developmental disabilities arising from other aetiologies were not necessarily able to support people with PWS. *“Not everybody can work here”* was a statement made overtly by one participant but paraphrased several times.

Discussion

The caregivers who participated in this study showed considerable consensus on all its key questions. This consensus was evident despite the diverse services and communities in which they worked. Their responses were also largely consistent with other accounts by caregivers (Goff, 2008; James, 2011; Waters, 2009; Ziccardi, 2006) and with those described in studies of practice among comparable groups (Hawkins et al., 2011; van Hooren et al., 2002, 2005, 2006).

All participants in the present study acknowledged their awareness of the dilemma arising from the provision of support to adults with PWS within the culture of generic disability services where the promotion of clients' civil rights and autonomy were highly valued. In spite of this awareness none advocated unrestricted access to food. Measures taken to control access to calories were not always described as effective, but all participants regarded them as an essential feature of the residential service in which they worked and as an inescapable part of their job. This pattern is consistent with the results of the small group of overseas studies which have canvassed the view of familial or paid caregivers supporting adults who have PWS (Hawkins et al., 2011; van Hooren et al., 2002, 2005, 2006). As already noted, this view also broadly describes the position taken by those clinicians and caregivers who are experienced in the support of people with PWS and who have written to advise their colleagues (Forster, 2010; Goff, 2008; James, 2010; Pittsburgh Partnerships, n.d.; Waters, 2009; Ziccardi, 2006).

Nevertheless, several participants also reported that the adults with PWS to whom they provided support had been able to learn to safely exercise an increased degree of independence around food. The gains described seemed to be situation specific rather than generalised. They invariably required that some type of supervision still be present, even if only in the form of regular, compulsory weight checks accompanied by discussions with staff. Most gains in independence around food were reported to have occurred only after a calorie controlled diet and a regime of ecological measures for the management of access to food had been established and accepted by the service user.

These reports are broadly consistent with some accounts published by adults with PWS themselves (Stanley, 2013). These authors sometimes report gaining increased independence by learning to accept and manage the limits imposed by the condition, and their own subsequent need for support. Miller et al., (2011) have tracked the difficulties with food faced by a large sample of adults with PWS and concluded that the severity of the drive to eat reduces spontaneously in adulthood for a significant number of individuals. It is possible that the gains described by New Zealand caregivers may have arisen by chance, but this did not seem to be the position taken by the caregivers who participated in the present study. When they described gains in independence made by their clients with PWS participants invariably ascribed them to changes in the relationship between caregivers and their clients: increased food security leading to lowered anxiety, increased trust in caregivers, and/or the acceptance of ecological controls.

Not all attempts by caregivers to promote such independence were reported to have been successful. Only a minority of participants reported that those people with PWS to whom they provided support had made gains in their independence in relation to food, and no caregiver reported the belief that any of their present or past clients with PWS could ever achieve complete independence in this regard. This apparent pessimism and emphasis on control could be viewed as support for the views of Wullink et al. (2009), who reviewed the literature concerning support services to adults with ID and concluded that, although agencies providing such services typically claimed that the promotion of their clients' autonomy was a key value guiding their practice, this ethic was not generally reflected in either published descriptions of the services

delivered or in the priorities of researchers. Wullink et al. concluded that such claims were largely lip-service and that there is little evidence that disability support services prioritise or promote autonomy.

Close analysis of the responses of caregivers interviewed for the present study reveals a different view. While these participants were clear that their work did not necessarily promote increased autonomy in relation to food, they characterised this aspect of their work as an exception that arose in response to a unique need among residents with PWS. Some caregivers stated overtly that the controls they imposed over access to food did not diminish their commitment to the promotion of independence. Nearly all described making considerable efforts to empower residents in other areas of life and to moderate the effect of food control measures on their clients. Many described striving to achieve what they referred to as “*fairness*” around food.

These responses are similar to those described by the caregivers interviewed by van Hooren et al. (2002, 2005, 2006). There are further parallels in the processes by which the two groups reported planning for the complexities of limiting access to food in practice. Participants in both studies emphasised the effort and time required for planning and decision-making concerning measures for the control of access to food. Both also described trying to achieve this difficult goal at the same time as trying to make progress towards a number of other desired outcomes. Both groups described having to revisit this planning process often, either in response to changing circumstances or to innovations in their clients’ reactions to existing measures.

In the present study, caregivers described seeking food management regimes that were practical and sustainable over time and which were also compatible with outcomes being sought for others in the caregiving environment. Even in the design of measures to which only the resident with PWS would be subject, the perceived needs of other residents, of caregivers and others were also considered. These priorities of sustainability and concern for the welfare of multiple persons at the same time seem very much like the set of caregiver values summarised by van Hooren et al. (2005) as caregivers’ search for “A liveable life” (van Hooren et al., 2002, p. 309)

The present study provides support for the view of van Hooren et al. (2005) that the characterisation of the central dilemma facing caregivers of people with PWS as a simple choice between polar opposite sets of values is likely to be regarded by caregivers as an unhelpful over-simplification. Instead, members of both groups of caregivers describe decision making within the complexities of a caregiver/care recipient relationship embedded in the context of a much wider social system.

Caregivers who participated in the present study often seemed to characterise the promotion of clients' safety and autonomy as complementary priorities, rather than as contradictory ones. Several participants reported that the control they exercised over access to food not only promoted the health and safety of their clients, but also led to reductions in anxiety and challenging behaviour, and thereby empowered service users to greater achievement and independence in other areas. Examples were given in which residents had achieved goals, such as voluntary employment or vocational training and participation in community organisations, which caregivers believed would have been impossible without the prior establishment of an effective food management regime. Once again, similar accounts feature in reports from overseas caregivers (James, 2010; van Hooren et al., 2002, 2005, 2006).

The assumption has been made by some researchers that the control of hyperphagia, and the subsequent management of obesity, represent the definitive features of the behavioural phenotype of PWS and of the challenges faced by caregivers (Holland, 2015). Caregivers who participated in the present study contradicted this view. Without exception they referred to aspects of their clients' challenging behaviour when asked to identify the most significant challenges they faced in their work. Of course, many of the behaviours referenced in this way were interpreted as attempts to secure illicit food, or to avoid detection after the fact, but it is equally true that caregivers often also identified behaviours that seemed unrelated to food. They described interpersonal conflicts, confabulation, inflexibility and dependence on routine, as well as various other behaviours of excess. Participants emphasised the emotional impact and disruption typically associated with challenging behaviour undertaken by their clients who have PWS. Their accounts seemed to reflect a view that the support of people with PWS is regarded especially stressful and challenging, even in the context of support services to people with ID. Once again, these views are consistent with the reports of

overseas investigators and of caregivers themselves (Boyall, 2013; James, 2010, Sinnema et al., 2011a; van Hooren et al., 2002, 2005; Whitman, 2013).

When asked what features of their service they regarded as the most important for the achievement of desired outcomes, the majority of participants identified consistency in support, both in the work of individual caregivers over time and across caregivers working on different shifts. By this, they seemed to mean that the key features of day-to-day interactions between caregivers and care recipients should be predictable to residents. Threats to this consistency were seen to arise either from variations between caregivers or from changes to the mood and priorities of individuals. Hence, both staff turnover and the presence of colleagues under stress or inadequately skilled were identified as potential sources of inconsistency. Many participants emphasised the seriousness of consequences that could arise from these threats. Accounts of such consequences included weight gain, increased challenging behaviour, and emotional distress for the person with PWS and for others.

Limitations

It could be said that this study represented something of a missed opportunity in the context of the wider project. Interviews were recorded only via contemporaneous notes made by the interviewer which limited the depth of analysis that could be supported. This limitation arose because the richness and complexity of conversations ultimately generated within focus groups had not been anticipated during the study's design.

Limitations also arose from the formation of focus groups from existing groups of caregivers. This has meant that interviews were conducted in groups of various sizes and sometimes included caregivers alongside their immediate line managers. It may be that some of the discussions generated were wider ranging for having taken place in such a group, but it is also possible that some participants felt constrained from speaking as freely as they may have done in other settings. At the same time, only very limited demographic data was gathered regarding focus group members in the course of this study, which has meant that the sample of participating caregivers cannot be described in detail. The views of people with PWS

themselves were not canvassed and the literature concerning the lived experience of those in receipt of care was not reviewed. Future research should address these issues.

Conclusions

This study suggests that it is not the management of access to food *per se* that is the most challenging feature of the role of caregivers supporting those with PWS. Participants consistently identified the challenging behaviour of service users in this regard.

Caregivers in Aotearoa/New Zealand seemed to concur with Holland and Wong's (1999) contention that the imposition of controls over access to food is an essential feature of support to adults who have PWS. This is despite the apparent conflict with ethical statements endorsed by these same caregivers and by the agencies that employ them. Far from reflecting a lack of regard for the promotion of their clients' autonomy, several participants reported the belief that by limiting choices around food they in fact promoted their clients' emotional wellbeing and empowered them in other areas, at the same time as protecting their health and immediate safety.

The food control regimes described by participants arose as the result of complex decision-making processes: dynamic over time, informed by a range of values, and highly individualised. All caregivers described some degree of tolerance for managed risk, and they sought solutions that would be sustainable and which would balance desired outcomes for a range of stakeholders, rather than for the person who has PWS alone.

Chapter Three

Study 2: A Description of Services to a Group of Adults with PWS

Caregivers, whether familial or professional, must address a distinctive set of challenges when they support adults who have PWS. The risks presented to this group of service users by unrestricted access to food are well known but caregivers must also address the range of challenging behaviours to which adults with PWS are also prone. These behaviours have been found to generate particularly negative outcomes for caregivers and others who share the residential environment (Hodapp, Dykens, & Massino, 1997; Jurgensmeyer, 2016; Lanfranchi & Vianello, 2012; Mazaheri et al., 2013; Shivers, Leonczyk, & Dykens, 2016; Whitman, 2013).

The present research literature offers caregivers little help in addressing the unique range of challenges associated with the support of people who have PWS, beyond the assurance that improved health and safety outcomes are associated with ecological controls imposed over access to food (Cassidy & Driscoll, 2009; Hauber et al., 2013; Kazemi & Hodapp, 2006; Messersmith et al., 2008; Miller et al., 2013). For example, investigators have yet to determine which measures are most effective in the establishment of food security, or the effects of those controls on other aspects of the lives of people with PWS. The pattern of variations in hyperphagic behaviour between individuals with PWS has not yet been described, probably in part because few tools have been proposed for the assessment of this behaviour (Dykens et al., 2007; Key & Dykens, 2018).

It seems likely that clinical observation and trial-and-error remain common strategies for matching particular services to the characteristics of individual service users. Claims have also been made for additional benefits in emotional wellbeing and independence in other areas said to accrue from the control of access to food. However, to date studies evaluating food secure environments have generally limited their investigation to the effects on bodyweight and variables related to physical health.

The present study was undertaken in order to describe the supports currently being received by a group of Aotearoa/New Zealand adults who have PWS in relation to food. In addition, it afforded an

opportunity to trial a number of related instruments that may prove useful to future researchers in the local context, as well as to service planners and providers.

Ethics

Study 2 received approval from the Massey University Human Ethics Committee (Human Ethics Committee (HEC) Southern-A 14/37). Gaining properly informed consent is a common challenge in any research which includes the participation of people who have PWS. At the same time, the support of service users to make decisions about whether or not to give their consent is a common role for caregivers who work within disability support services. Each letter of invitation to participate in Study 2 was addressed to both the person with PWS and their caregivers. Each person with PWS who chose to participate in this study was asked to nominate a single support person who would agree to partner with them in completing its requirements. The role of this nominated support person was to assist the participant with PWS in the consent process, in interpreting study information, and in completing the questionnaires that constituted the study itself. Each of these nominated support people was asked to sign the study's dual consent form (Appendix B), alongside the person with PWS to whom they were to provide support. A plain language version of the study's consent form was made available but was not requested by any potential participants. In order to protect the privacy of service users and others, no identifying information was reported regarding any person with PWS, any caregiver or any service support agency. No personal information was gathered from supporting caregivers except that required to complete the consent form.

Method

A Population Survey

A number of surveys of people who have PWS have been undertaken in other jurisdictions (Grugni et al., 2008; Hiraiwa, Maegaki, Oka, & Ohno, 2007; Thomson, Glasson, & Bittles, 2006; Whittington & Holland, 2004). However, most of these studies have focused on gathering health related and actuarial data. Unlike them, Study 2 has also gathered information concerning the supports and services being received by people with PWS. This information is important for a number of reasons but particularly

as a first-step towards the investigation of relationships between service features, the characteristics of individual service users, and service outcomes.

Consideration was given to a proposal to undertake a pilot study to test the usefulness of the measures which made up Study 2. This proposal was rejected however because of the limited number of potential study participants.

Participants and Recruitment

Several agencies known to maintain databases of adults who have disabilities within Aotearoa/New Zealand were contacted. Information regarding the study was provided and agencies were invited to partner with the researchers. Partnering agencies included PWSA (NZ), the national association of agencies contracted to the Ministry of Health (MOH) to provide needs assessment and service coordination to people with disabilities (NASCA), and most of the NASCA's constituent regional NASC agencies.

Partnering agencies were invited to address envelopes containing copies of the study's information sheet (Appendix A) and a generic letter inviting adults with PWS and their nominated support people to participate in the study. Each partnering agency distributed these envelopes to individuals listed on their database as having PWS and as being over 16 years of age. Slightly fewer than 100 such envelopes were distributed. However, the number of people with PWS who were finally invited to participate remains unknown because some potential participants could be expected to be represented on both the PWSA (NZ) database and that of their local NASC; these people probably received two invitations.

Measures

Hyperphagia Questionnaire (HQ). The HQ is a caregiver completed instrument designed to identify food related behaviours and preoccupations in people who have PWS, and rates their severity. Dykens et al. (2007) developed this measure in response to a growing recognition that clinical trials up to that time had been hampered by the lack of PWS specific measures of food related behaviour. Each item is scored on a 5 point Likert scale; a rating of '5' indicating a severe or pervasive problem. Factorial analysis has resulted in 3 factors: hyperphagic behaviour, hyperphagic drive and hyperphagic severity (Dykens et al., 2007). As the authors note, this set of factors make conceptual sense in the context of our understanding of food related behaviour and PWS. Dykens et al. have also reported values for Cronbach's α for each these

factors which showed acceptable levels of internal consistency: hyperphagic drive .76, hyperphagic behaviour .80, and hyperphagic severity .60. The HQ has subsequently been adapted for use in clinical trials (Danglas et al., 2017; Key & Dykens, 2018), and has been used in a population study in the USA (Dykens et al., 2007) but has not been trialled in Aotearoa/New Zealand.

The New Zealand PWS Services Questionnaire (NZPSQ). The NZPSQ (Appendix C) is an instrument for the description of the caregiver initiated, ecological interventions by which an adult who has PWS is supported in relation to food. The NZPSQ was developed for Study 2 and designed to be completed by adults who have PWS with the assistance of a nominated family member or other support person. It delineates those who reside with family members from those who are supported by paid carers within generic services for people with developmental disabilities, and from those who live in a specialist residential service specifically designed for people who have PWS.

The NZPSQ asks respondents to identify the extent of support they receive from professional caregivers each week, as well as the measures by which access to food is managed. Four common interventions are specified: the implementation of a planned diet specific to the person with PWS, the presence of locks on food storage areas, supervision over the person with PWS's use of discretionary income and supervision outside the residential home.

This instrument also records respondents' height and weight by which a standard measure of BMI may be calculated. The concept of BMI is not without its critics (Müller, Braun, Enderle, & Bosy-Westphal, 2016). The advantages of using this measure in the present study are that it is easily gathered and calculated, and can be expected to be readily legible to study participants. Its limitations are also well known and, in the case of people with PWS, only likely to result in bias in one direction. BMI can generally be expected to yield an underestimation of obesity among people with PWS due to the distinctive body composition and low muscle tone that are features of the syndrome.

SF-36. SF-36 is a commonly used measure of health related quality of life (QoL) which can be completed by caregivers (Ware & Sherbourne, 1992). It was chosen above similar measures principally because of its common use in studies of QoL among people with PWS (Lopez-Bastida et al., 2016; Mori, Downs, Wong, Heyworth & Leonard, 2018; Vogt & Emerick, 2015), notably one longitudinal population

survey completed by Bertella et al. (2007). SF-36 was designed to be completed by caregivers but when these questionnaires were returned by participants in the present study many questionnaires proved to be incomplete or otherwise unable to be scored; it was subsequently withdrawn from analysis.

Procedure and Analysis

Student-t tests were undertaken concerning the relationship between BMI and the presence of paid caregivers, the implementation of each of the four food management interventions identified in the NZPWSQ, and the implementation of all four together. The use of the HQ was trialled in the local context and a minimum range of BMI was described, as was a range of services by which access to food was managed.

Results

Nineteen completed sets of consent forms signed by both a person with PWS and their nominated caregiver were received. Of these, 18 pairs of participants completed the study. All participants with PWS were 16 years of age or older, 9 were female and 9 male.

HQ

No significant relationship was found between the scores on the HQ and either BMI or the presence of any of the measures for food control listed in the NZPSQ. Table 1 summarises the results of the HQ. In general, these results demonstrated a pattern of responses similar to that found in populations investigated in the USA study (Dykens et al., 2007). In the present study, Cronbach's α for hyperphagic behaviour and hyperphagic drive were .69 and .77 respectively, broadly similar to those recorded by Dykens et al. (.76 and .80). The present study recorded a relatively lowered Cronbach value for the third factor, hyperphagic severity, at .40 compared to Dykens et al.'s .60. However, only two items of the HQ code to this factor. This result should be interpreted with care given the small sample size of the present study.

The HQ was initially designed for use in research, notably clinical trials of pharmacological and psychological interventions for hyperphagia or obesity. The results generated in this study suggest it may also have value as a checklist to inform the assessment of an individual's need for support in relation to food.

Table 1

Factor means, SDs and ranges on the Hyperphagia Questionnaire for a group of New Zealand adults who have PWS, n =18

Factor	Mean	SD	Range
Hyperphagic behaviour	12.18	4.25	5-20
Hyperphagic drive	11.11	3.41	6-18
Hyperphagic severity	4.76	1.66	2-8

SF-36

The SF-36 questionnaires were returned by 18 pairs of participants; however, none proved to have been fully completed. This result seemed to have been due to difficulties among participants in following the instructions which accompanied the test. This measure was not able to be reliably scored and was excluded from subsequent analysis.

NZPSQ

BMI. Table 2 describes the pattern of BMI scores among respondents with PWS. Of the 18 pairs of participants who returned the NZPSQ, 17 completed the items regarding the service user's height and weight from which BMI could be calculated. Of these 47% (8 individuals) recorded BMI over 40, within the range labelled extremely obese. None fell within the normal weight band (< 25 BMI). Mean BMI was 39.3 and those participants whose BMI fell in the overweight range represented the lowest scores.

Dykens et al. (2007) also calculated the BMI of the participants in their study, and as a result concluded that excessive body weight remained a significant concern for people with PWS in the USA. The mean value for BMI found in this Aotearoa/New Zealand study was higher than that of each of the age groups means recorded by Dykens et al., although not significantly so.

Living situation. The NZPSQ included a number of questions which asked pairs of respondents to describe the household in which the person with PWS lived and received support. No participant in the present study reported living alone. Seven participants (39%) described living with family members while the remaining (11 individuals or 61%) lived in a group home. Eight of these participants shared their home with three or four other service users and three reported receiving support in residential units shared with just one other person. Of these three respondents, two reported that their co-resident also had PWS; consequently they represented the only participants who lived in a residential service specifically designed for the support of people with the syndrome.

Support from professional caregivers. The NZPSQ also asked respondents to describe the support provided by professional caregivers. Of the 11 individuals not living with family members all reported receiving regular support from paid caregivers. Of these, 4 reported that there were times during the day in an average week when they were at home with no caregiver present, and 2 reported not having staff in their homes at night.

Food management regimes. The NZPSQ also generates a summary of measures taken to manage access to food. All respondents reported an awareness of the dangers associated with unlimited access to food and all reported that at least some food control measures were in place. Considerable variation was reported in the specific measures employed.

Only 10 (56%) of participant pairs reported that caregivers had both identified how many calories the participant with PWS could eat in a day without gaining weight, and that a diet had been planned accordingly. It is interesting to note that this figure is lower than the number of respondents who reported that locks were in use on food storage areas in their home: 11 or 61%.

The measures most commonly employed to manage access to food were reported to be the supervision of residents with PWS by caregivers, locks placed on food storage areas, and the use of a planned diet. Other interventions such as portion control, the installation of alarms, and keeping high calorie foods out of reach were also reported.

Table 2

BMI scores of a group of Aotearoa/New Zealand adults with PWS, n =17

<u>BMI</u>	<u>Number of respondents</u>
25-30	3
31-35	0
36-40	6
41-45	3
46-50	5
<i>Range: 26.5 - 49.0 (n = 17)</i>	

Several participants reported that they had replaced some environmental controls with attempts to influence the food choices of those with PWS. These caregivers described holding discussions about food, planning diets alongside the person with PWS to whom they provided support, and encouraging self-management. No significant relationships were found between the presence of any one of these measures and BMI, hyperphagic behaviour, drive or severity (Dykens et al., 2007).

It seemed unlikely that, for the majority of participants, the measures described were implemented with sufficient rigour to constitute what authorities such as Gourash et al. (2006) would regard as food security. Opportunities to seek extra calories seemed inherent in many of the regimes described. For example, half of the participants with PWS (9) reported that they were without supervision at least once a week, and 6 (33%) were able to spend discretionary income without supervision. Several participant caregivers commented that they kept track of service users' whereabouts or access to food "*as often*" or "*as much as possible*".

Discussion

The goals of the present study were to describe services to a group of New Zealand adults with PWS in relation to food, and to investigate a number of outcomes associated with those services. The usefulness of the HQ was also evaluated in a local context. The low response rate generated by this study, coupled with

the lack of useable data from one of the study's measures, has meant that some of these goals cannot be addressed. However, a number of interesting conclusions may still be drawn.

Dykens et al. (2007) recorded concern at the high BMI scores of those people who participated in their study. The two samples are not directly comparable, that of Dykens et al., included children as well as adults with PWS; however, that study found a mean BMI of 33.64, whereas Study 2 has found mean BMI of 39.3.

The individual food management plans which participants and their caregivers described in the present study often seemed piecemeal and opportunistic rather than comprehensive, especially when compared to those recommended by advocacy groups or described in the professional literature (Forster, 2010; Goff, 2008; James, 2010; Waters, 2009; Ziccardi, 2006). It is possible that such informal controls may be quite effective for some individuals with PWS, but the pattern of measures of BMI among participants in the present study renders even this speculation doubtful, except perhaps for a small minority.

This pattern of BMI and caregiver supports may have arisen for a number of reasons. It may indicate that the control of access to food is not a primary concern among local caregivers, perhaps because of the strength of other priorities or because the risks posed by hyperphagia for people with PWS have been underestimated. However, it seems more likely that this pattern may reflect the reports of overseas caregivers concerning how demanding the goal of food security is to achieve and maintain in practice (Goff, 2008; Hawkins et al., 2011; James, 2010, van Hooren 2002, 2005, 2006; Waters, 2009).

Taken as a whole, this data gives grounds for serious concerns. It seems that New Zealanders with PWS are likely to be at increased risk of premature and sudden death, as well as other consequences of severe obesity: reduced mobility, exacerbated poverty and social isolation. The reports of clinicians and caregivers also suggest an increased risk of mood and behavioural difficulties arising from uncertainty about access to food (Forster, 2010; Goff, 2008; James, 2010; Waters, 2009; Ziccardi, 2006). This finding is especially concerning given that all of the participants who contributed to this study reported that caregivers exercised at least some measure of control over service users' access to food, either through targeted supervision or environmental controls such as locks and the imposition of financial agents. This pattern of

high BMI scores suggests that at least some of these controls are ineffective, and that the adults with PWS who are subject to them are having their rights infringed in return for no useful outcome.

A significant proportion of adult participants with PWS continue to live with family members. This conclusion is consistent with one of the findings of a recent Australian population survey (Akindola et al., 2015) and might be indicative of a greater willingness among familial caregivers to participate in research, compared to their professional counterparts. This pattern may also reflect scepticism among adults with PWS and their families regarding the effectiveness of publically funded support services, perhaps born of the lack of evidence concerning the outcomes associated with them.

This study featured the use of the HQ in the Aotearoa/New Zealand context for what is believed to be the first time. This measure yielded broadly similar results to those generated in a previous North American study (Dykens et al., 2007). In addition to its usefulness in future research the HQ might also play a role as a checklist for the description of the difficulties with food faced by individuals with PWS, and so prove a valuable addition to assessment of their support needs.

The NZPWSQ also seemed to generate useful information about the food ecology of individuals and may be of use in the process of assessing support needs. However, its potential as a measure of food security is somewhat limited. None of the common interventions for the management of access to food predicted BMI, neither did the implementation of all four of them together. This is perhaps unsurprising as the NZPWSQ contains no measure of the fidelity with which these measures are practiced, or of their effectiveness in limiting access to food. Further development of this questionnaire may be possible through the inclusion of additional interventions and through the inclusion of questions asking caregivers and service users to rate the frequency with which additional food is able to be obtained.

Alternative research strategies should be adopted to better define the interventions necessary for food security and to assess the outcomes associated with this state. Closer investigation of those individuals who have PWS and the lowest BMI might be useful in this vein, as would any longitudinal study which tracks outcomes among participants with PWS as they move between residential environments featuring different levels of food security.

Study Limitations

The present survey was characterised by a high rate of incomplete or unusable questionnaires. The use of pairs of participants, in which each person with PWS was supported by their nominated caregiver, was intended to facilitate the engagement of service users in research and to make the research process more legible for those participants with PWS and associated ID. This model may be useful as a way for future investigators to address the former goal, but it has not proven effective in the latter. It seems more accessible measures need to be employed or more support must be made available for participants to complete them. The use of an online format with an email helpline might address both needs in future studies. These difficulties may have been avoided if a pilot study had been undertaken prior to Study 2. This idea was rejected because the pool of potential study participants was considered too small. Future studies should consider testing study instruments in this way, and seek to overcome the problem of small sample size by seeking participation from a wider, international pool of service providers and advocacy organisations.

No data were gathered regarding genetic subtypes of PWS. Dykens et al. (2007) did ask for this type of data, but expressed doubts as to the accuracy with which it was reported by caregivers. They found little difference in scores on the HQ between those with different sub-phenotypes.

The results of the present study may also have been limited by the lack of data gathered concerning those caregivers who supported participants with PWS. In particular, no information was gathered about the literacy skills of these participating caregivers. This study also had a number of other weaknesses, most obviously the low recruitment rate and subsequent small number of participants. PWS is a relatively rare condition but several communities have recently established comprehensive databases of those who are diagnosed with the syndrome (Butler et al., 2018), for example, both the Prader-Willi Association of Australia and those of several Australian states (Akindola et al., 2015). A considerable degree of international cooperation already exists among researchers who share an interest in PWS, often facilitated through IPWSO. Cooperation in this respect could increase the pool of potential participants. An Australasian population survey might generate sufficient data to allow investigators to test the relationships between service features, service user characteristics and service outcomes. The results of the present study

give grounds for concern about the health and wellbeing of adults who have PWS in Aotearoa/New Zealand. They also point to a pressing need for further investigation.

Chapter Four

Study 3: Interviews with Experienced Caregivers

The Significance of the Caregiving Role

Study 3 reports the results of a process of thematic analysis conducted on the texts of interviews with individual caregivers who work in support of one or more adults with PWS. The importance of professional caregivers in the lives of people who have developmental disabilities has been demonstrated in a number of ways. For example, Schuengel, Clegg, de Schipper, and Kef (2016) have examined the impact of certain caregiver characteristics such as attachment style on both their professional practices and their relationships with service users. Likewise, caregiver attitudes and behaviour are among topics of interest for those who have explored the development of culture within residential services (Bigby & Beadle-Brown, 2016; Bigby, Knox, Beadle-Brown, Clement, & Mansell, 2012). Banks (2016) has written about the specific issue of caregivers' mediation of sexuality and romantic relationships among those they support. Whatever policies, technologies or resources are applied to the task of supporting people with PWS, the outcomes that result are likely to be mediated by the work of professional caregivers. The values, beliefs and habitual behaviours of those caregivers will inevitably be a significant influence on the lives of those who use the services in which they are employed.

The present focus on the individual in residential service provision. The experiences of professional caregivers also offer a unique way of investigating the services in which these caregivers work. A great deal has been written about the importance of the individual within contemporary services to people with developmental disabilities in Aotearoa/New Zealand. This emphasis is reflected in key policy documents (Office for Disability Issues, 2001, 2014; United Nations Office of the High Commissioner for Human Rights, 2014). This focus on the individual is also reflected in common practice, service users' support needs are individually assessed, these assessments lead to individual support plans, which are individually reviewed and sometimes supported by individualised funding packages. Service provision

agencies are contracted to compensate for these individually assessed deficits in much the same way as a pharmacy might fill a medical prescription.

Duryan, Nikolik, van Merode, and Curfs (2012) have demonstrated that the facilities in which paid caregivers work can be viewed as service systems, rather than simply as responses to the needs of individuals. Within a service system, interactions between caregivers and care recipients are not expected to be unidirectional but reciprocal, with all agents subject to feedback in various forms. The study of the experiences of professional caregivers is also important because it presents the services in which these workers are employed as complex and embedded service systems.

The Goals of Study 3

Little is known about the beliefs, habitual practices or experience of those professional caregivers who support adults with PWS, either in Aotearoa/New Zealand or overseas. This study's goals were two fold. It sought to explore the ways in which local caregivers conceptualise and experience their role, especially the sources of challenges and rewards that they find within it. It also investigated the ways in which study participants have come to conceptualise and address the central dilemma inherent in that work: that of supporting their clients who have PWS safely in relation to food within a professional context in which the promotion of clients' independence and rights as citizens are core ethics.

The Role of the Researcher

The present study was not designed as action research, but it became clear at an early stage that the researcher's past experiences were likely to influence both the data set itself and its subsequent analysis and interpretation. It was first planned as a collection of semi-structured interviews embedded within a scientific and academic context. It could equally be described as a series of conversations between present caregivers and their former colleague about issues of importance to them both.

The interviewer's potential status as an insider to the world of PWS presented opportunities as well as risks. For example it may have contributed to the participants' belief they were likely to be safe and

understood in discussing controversial topics and hence to their candour and willingness to share their stories.

Steps were taken to acknowledge and manage these effects. No caregiver was considered for the present study if either they themselves or a person with PWS to whom they provided support had had a clinical (or other) relationship with the researcher within the past 10 years, although one caregiver whose interview was included in the present study reported that she had attended training and participated in the implementation of a behaviour change programme that the interviewer had developed for a man with PWS during the 1990s. The semi-structured interview format was also adopted in part so that, where the influence of the researcher's own views could not be limited, it could at least be rendered more apparent and able to be included in subsequent analysis and review. Despite these steps this study retains an element of participatory research that must be acknowledged.

Ethical Review

A review of the present study's design was sought through the Massey University Human Ethics Committee (HEC: Southern A Application 15/56). This review process was informed by the results of Study 1. It was noted that participants may be at risk of emotional distress as they described some incidents which had occurred in the course of their work. To ameliorate this risk, it was first agreed that, although no therapy or professional advice would be offered during any interview, the time allocated would be flexible so that each participant would be allowed extra time to talk should they require it. In addition, information was gathered concerning the relevant policies and procedures instituted by each participant's employer, including employee assistance programmes and service standards, which could be made available to interviewees if required.

Threats to the privacy and reputations of both service users who had PWS and the organisations which supported them were also noted. In response, no identifying information was recorded or reported regarding any person with PWS, or any other service user. Service provision agencies were informed of the project before the caregivers in their employ were invited to participate, and invitations to potential

participants not issued without the permission of senior managers. No agency was identified in reports of the study or its results.

As has been noted, the provision of support to people with PWS typically entails a number of significant challenges. Concern was also raised that interviewees may seek professional advice from the interviewer. A response was prepared which allowed the researcher to politely decline such requests while referring participating caregivers to relevant resources available, either through their own employing agency, or through PWSA(NZ).

Method

Semi-structured Interviews with Individual Caregivers

Study 3 was based on interviews with individuals using a semi-structured format. A small number of open-ended questions were used to generate wide ranging discussion between the researcher and participant caregiver. This study's design broadly followed that adopted by van Hooren et al., (2002) in their investigation of values and practices among caregivers of people with PWS. As DiCicco-Bloom and Crabtree (2006) have noted in their discussion of the use of interviews in qualitative research, less structured interview formats allow a more equal partnership in meaning making rather than regarding the person interviewed as merely a source from whom information may be extracted.

The questions on which Study 3 was based were largely informed by those tested in Study 1, which was made up of discussions in workplace-based focus groups. Group dynamics are an important part of any research which features the use of focus group, and interactions between participants seemed to contribute to the richness and depth of insight which featured in the data gathered during Study 1. Several researchers have noted however that such group dynamics may also influence subsequent data in other ways (Duggleby, 2005; Farnsworth & Boon, 2010). In particular, the responses of some participants may be inhibited by the group context and dissenting voices may be weakened or modified. Study 3 aimed to generate in-depth discussion of caregivers' experiences, beliefs and practices. This seemed a goal best addressed through

interviews with individual caregivers. It was also noted that professional caregivers in Aotearoa/New Zealand generally work alone, in isolation from other caregivers.

Study 3 inevitably included a strong exploratory component. A process of thematic analysis was selected because of its usefulness in identifying patterns within such datasets (Braun, Clarke, Heyfield & Terry, 2019).

Participants

Interviews were conducted with 10 individuals: five caregivers and five supervisors who also provided direct support to at least one person with PWS. Nine were female and one male. With the assistance of the PWSA (NZ), a number of organisations were identified as being contracted by the MOH to provide residential support services to one or more adults with PWS. This list was somewhat reduced by the exclusion of those residential services whose staff had already participated in Study 1. Senior managers of the remaining eligible services were contacted by the lead researcher and provided with an information sheet describing the study (Appendix D), along with an invitation for those employees who met qualifying criteria to participate.

All of the service managers contacted expressed support for the project and willingness to allow their employees to contribute to it. However, certain logistical constraints prevented some agencies from being involved. These included scheduling conflicts, constraints on travel for either caregivers themselves or for the researcher, and difficulties in communication between service managers and their Chief Executive Officers or governing boards which meant that consent could not be formalised within the available time.

Any employees of the participating agencies who provided direct residential support to one or more people with PWS, at least once a week on a rostered basis, were deemed eligible to participate. Their first level managers were also deemed eligible if they also had regular, direct contact with at least one service user with PWS. One participant was included in the study who did not then meet these criteria but who had done so in the recent past.

Participants represented four residential services in four different urban centres and collectively described the support of five current and five former service users with PWS. Several participants described having known or met others with the syndrome in addition to those for whom they had provided paid support.

Most participants were employed in group homes (residential services in which a number of individuals are supported by a team of caregivers working rostered shifts). Three worked in either a supported living service (in which support is provided to an individual's own home for a finite number of hours each week), or in a respite care facility for those who otherwise resided with family members. The number of eventual participants was perhaps a little smaller than expected, especially given the high level of interest in the study. The majority of professional caregivers are of course shift workers and several individuals reported that logistical difficulties of time and travel precluded their participation.

Many of those who were interviewed expressed and demonstrated a longstanding interest in PWS. It is acknowledged that this sample should not be considered a representative one. In particular, those caregivers holding especially strong interests or opinions could be assumed to have had a greater tendency to volunteer to be interviewed.

Procedure

Interviews were based on a list of open ended questions (Appendix E). They were recorded with each participant's consent using a device in plain sight. On rare occasions some participants asked for this recording to be interrupted for a short time, invariably to allow that person to discuss contextual information not directly relevant to the research topic. Such requests were always complied with and the subsequent gaps in the data set noted in the transcript.

Interviews were conducted individually and in private, except for one in which the caregiver's client, a man who has PWS, also participated. This occurred both at his suggestion and at the request of his participating caregiver. This man reviewed the study information sheet with the support of his caregiver and completed the study's consent process. This service user's contributions to the subsequent conversation

presented a challenge. His comments clearly constituted valid and important information, especially in a field in which the voices of service users are largely absent. However, their relevance was open to question given that the goals of Study 3 concerned the experiences and practices of caregivers. This service user's contributions to the study were documented in the same manner as those of the interviewer. They were recorded, transcribed and reviewed, but not included in the subsequent analysis by being coded to any theme or subtheme. This data has been saved and may be reported separately.

Most interview venues were provided by the agencies that employed participants, although one took place in an interviewee's home at her request and another was hosted by a person with PWS in his own home, which was also the workplace of the relevant interviewee.

Analysis

Recordings were transcribed by the researcher and subjected to analysis for themes, broadly following Braun and Clarke's (2006, 2013) model. The data set was read multiple times and then interviewees' statements were divided into discrete, meaningful units, each one reflecting a single statement relevant to the research topic. These text units were then grouped into categories by topic using the cut and paste function of a word processing programme. This process resulted in a list of 103 initial codes, a small number of text units being assigned to more than one code. Each of these initial codes was printed in hard copy, re-read and further grouped for subthemes and themes. At this stage, several of the initial codes were abandoned and the units of text within them reassigned by cutting the printed copies with scissors and re-categorising the pieces.

At this point it began to become clear that the emerging pattern of themes and subthemes appeared to be dominated by a single narrative. After some consideration the decision was made to allow the dominance of this narrative to be expressed in the presentation of the study's results. The plan to summarise the data set in a thematic map was modified in favour of the presentation of the relationships between themes and subthemes within the format of a single, quite straightforward narrative, characterised here as "Challenge and Response".

Results

Themes: The Dominant Narrative

The results of study 3 are summarised in Figure 1. Without exception interviews were dominated by caregivers' descriptions of the challenges they faced in their work, their responses to those challenges, and the outcomes they sought to achieve through those responses. These three themes featured strongly in every interview and formed a simple narrative in three parts, with each of the subthemes to which codes had been assigned fitting somewhere along its axis. Figure 1 is a visual representation of this dominant narrative.

The challenges that defined the caregiving role were described as chronic and unrelenting, but at the same time as arising unpredictably from multiple sources. They required caregivers to generate responses both individually and in workplace based teams. Through their responses caregivers sought certain common outcomes, reflecting their values and priorities in relation to their work.

A caveat. An important caveat should be recorded before this dominant narrative is examined in detail. This conceptualisation of the caregiving role as one dominated by challenge cannot be described as entirely emergent from the data because it featured in several of the questions on which interviews were based, what was surprising was the degree to which this narrative dominated the subsequent conversations. On rare occasions, participants reported that they had been able to turn to published programmes or to advice given by experts in order to find ways of addressing the challenges they faced in their work. Much more frequently participants framed descriptions of their work in terms of a seemingly constant quest to maintain vigilance and to generate creative responses of their own.

A counter narrative. The strength of the dominant narrative notwithstanding, it did not describe the data set in its entirety. A counter narrative was also present in which participants described their work as personally rewarding and as important and effective in the lives of service users, albeit still in the context of significant ongoing challenge. This view also seemed to be quite commonly held among participants. Even when it was not stated overtly it was expressed in many of the stories they told. Such anecdotes featured

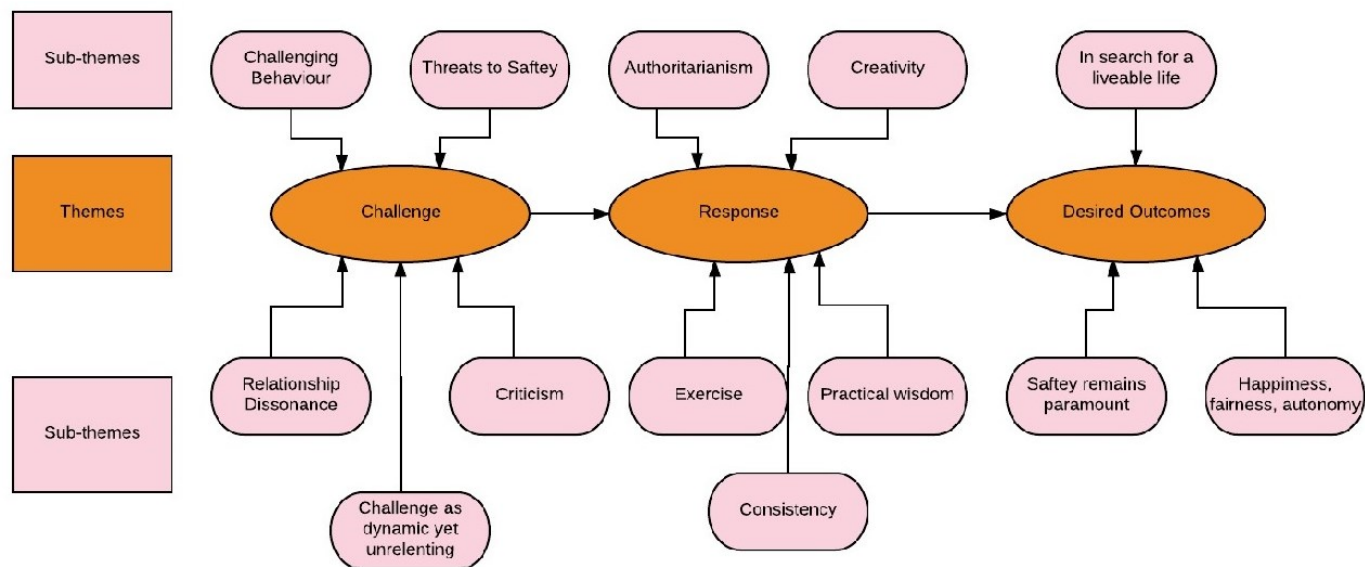


Figure 1. The dominant narrative.

accounts of goals achieved or of mutually valued and rewarding interactions between caregiver and care recipients with PWS. Typically these accounts were related with smiles, which were prominent in all interviews; laughter was also common.

Whitman (2013) has made the point that many features of the PWS personality phenotype are in fact highly valued and pro-social traits. People who have PWS tend to be socially oriented and are often especially interested in the patterns of relationships within groups. Creativity, humour and a passion for what the individual perceives to be fairness and justice are common features. People with PWS are often excellent conversationalists (Dimitropoulis et al., 2013) and several participants in the present study reported simply enjoying their company:

“Yeah, yeah it’s that interaction ... He’s really personable, and he’ll con a cup of coffee out of you (both laugh). You know, he’s just, yeah. He, he really is. I enjoy his company.”

“Absolutely a joy of a man, absolute joy of a man!”

The relationship between these narratives: challenge and reward. For several participants these two narratives seemed to be linked together, caregivers reported that they found opportunities for personal

growth and learning within their role as they addressed its demands. One supervisory manager described some of the opportunities that she believed were open to her staff:

“... it’s about growing as a person too.”

“ It’s not just about the job it’s about how, you know, they want to be better people as well, ... so, there’s big opportunities for this group of people. They’re looking at themselves all the time, they’re looking at their own conduct or ‘Hell, you know I barked a bit then’ you know or ‘How can I do that better next time?’ ”

As one caregiver said:

“... but then you learn, and you just learn the coping skills and things like that.”

Participants also related several anecdotes that reflected their experience of personal reward arising from the knowledge that they had understood or responded to a challenge effectively. These stories typically featured descriptions of unexpected challenging behaviour on the part of service users, and of how that behaviour had ultimately been rendered explicable by the application of caregivers’ understanding of the syndrome or of the individual concerned. Interviewees often seemed to take pleasure in describing these insights as one might from solving a mystery or exercising an uncommon skill. Once again, short bursts of laughter, shared between the interviewee/story teller and the interviewer, were a common feature of these accounts.

This link between the dominant narrative and counter narrative was especially clear in two participants’ accounts, each of which included a description of the caregiving role as a relatively easy and even undemanding one. These caregivers each described their work as having presented them with very difficult challenges in the past. In addressing these challenges successfully, each participant reported that they and their colleagues had garnered experience and learned lessons which continued to inform their present work.

These participants described their present role as undemanding, relative to those challenges that they had faced and overcome in the past. Even these two caregivers, who described their present work as

relatively free of challenge, still employed the underlying narrative of challenge and response in order to characterise their role.

The First Theme: Challenge

Interviewees described the challenges they faced in their work as arising from multiple sources. Challenges came not only from their clients' difficulties in relation to food, but from a range of factors, including some which were also known consequences of PWS. In addition, participants identified challenges arising from aspects of their relationships with their colleagues, with other caregivers working in generic services, with employers, service users' families, service funders, the Police, media and the courts. Conflicts within the body of values and beliefs which caregivers themselves brought to their task were also identified as a source of challenge.

Subtheme: Service users' challenging behaviour. Discussions related to this subtheme of service users' challenging behaviour were a strong focus in all interviews. This topic occupied more than half of the time taken for most conversations.

Foraging behaviour. As expected, many challenging behaviours described by participants were interpreted as illicit attempts to find and consume food. Participants described a range of manifestations of foraging behaviour, including the theft of food and money, the telling of untruths, flattery, bargaining, violence and threats of violence, outbursts of anger, damage to property, absconding, and an ability to pick locks.

Aggressive behaviour and assaults in pursuit of food. Violent behaviour, whether actual or threatened, was reported only with regard to a minority of service users. However, when this behaviour was described participants usually emphasised its seriousness and the risks it presented to their own safety and that of others. One interviewee described the way in which kitchen knives were controlled in her workplace and reported that this was because these knives had been used as weapons in the past.

Descriptions of aggressive behaviour often also featured accounts of caregivers compromising their own safety in return for that of others. One supervisory manager reported that angry, aggressive behaviour

had sometimes accompanied demands for food on the part of one man with PWS. This participant reported that she would first call the Police when this behaviour arose, and then send her staff out of the house along with the other residents. She reported implementing this plan on multiple occasions; whenever “*it got too bad*”. She said:

“I’m the last call. ‘You have hit and abused me, I ring the Police’, because I have to protect the staff.”

This participant described her experience of staying beside her client until the Police arrived:

“I was gonna sit in the lounge because ‘You’re not going to hit staff. I’m going to be here’ ... He’d hate me ... when he saw me in the street, there’d be two weeks of that but we’d get used to that.”

Foraging outside the residential service. Some foraging attempts occurred outside the residential service. These often targeted local merchants or social groups and public events. Many featured considerable creativity as well as some planning. Some could perhaps be regarded as examples of what James (2010) has referred to as food genius:

“He’s done some pretty cool things when he was on the walk. One of the things he did when he was on the walk is he walked past the local cricket club, he’s never played cricket before in his life, but he started stopping and watching the cricket because after every sports game ... what do you do? ... You go into the clubrooms and you have supper ... And so (resident) got to know a few people and he’s looking around, started chatting away and he’s very friendly, very likeable. Invited in, ‘Would you like to come in?’ ‘Oh that would be lovely. Thank you so much’, in he goes and before you know it he’s having supper there every Sunday.”

“... so if he knows he’s going out, um then we’ll say ‘Okay so you know you’re going to church tonight’, um it’s all for supper because he’s not religious in any way, shape or form.”

In another such anecdote, a person with PWS was described as being able to parlay an existing, longstanding interest in handcrafts into an opportunity to access food:

“... he goes, ‘Oh you’re the most wonderful lady in our knitting club, aw I just love ...’ and he goes ‘... and do you think I could have a sausage square?’ And if the staff are not watching ... I know they feed him up when he goes there.”

Other accounts featured sham medical crises, such as seizures or catatonic symptoms, which resulted in contact with hospital services and increased access to food. The fact that the caregivers who described these circumstances invariably felt obliged to seek medical assistance until they were entirely sure that the symptoms were not genuine reflected the effectiveness of the pretence on the part of the person with PWS, but also the high priority placed by on residents’ physical safety as a service outcome:

“So he’s very difficult to work with. You have to take what he says as the truth, that he really is doing those things ... ‘Aw, I have to get a ride in the ambulance’ and ‘I need to take my inhaler and I need to have something to drink and eat afterwards.’”

Other foraging behaviours. Hiding and otherwise hoarding food were commonly reported, but the taking and eating of frozen, raw or spoiled food was described only in a small minority. Participants’ responses often seemed predicated on the understanding that significant variation existed among clients with PWS in the range and intensity of hyperphagic behaviours to which each person was prone. As already noted, these variations have been reported before (Dykens et al., 2007; Stanley, 2014; Whitman, 2013). To some extent they are measured in the item scores of the HQ (Dykens et al., 2007), although their pattern has yet to be fully described or their limits defined, as noted in Chapter 3.

The tone of discussions on foraging behaviour. The tone of all interviews was generally calm and professional. However, in lifting these narrative accounts of foraging behaviour from their context and placing them alongside each other, it was interesting to note how often stories featuring attempts at foraging were accompanied by expressions of emotion on the part of caregivers. Participants sometimes showed apparent frustration, but invariably also warmth, humour, and a sense of respect for the creativity or tenacity of service users. At the same time, it was also clear that each participant regarded any incident of successful foraging as serious, or at least potentially so. The obvious risks to health and safety as the result of this

behaviour were referred to many times, as were risks to service users' dignity, their key relationships and even their place in the community. For example, one participant described intervening to protect her client's relationship with a schoolmate:

"He also had a best friend that he would attempt to fleece money, from his pocket money."

On another occasion, a participant described reluctantly placing limits on a service user's independence in order to protect that person's place in the community:

"... he would argue with McDonalds and the Police would be called, that they didn't put a big enough burger in his um ... if the Police were involved, which they were a couple of times, then we had to make a decision on whether or not he did go the following Thursday ..."

Non-food related challenging behaviours.

Inflexibility and excess: the Prader-Willi personality. A set of psychological and behavioural characteristics has been identified as associated with PWS. This list has been referred to by Whitman (2013) as the PWS personality phenotype (p. 217). For example, Whitman and others have noted that those who have PWS often pursue any rewarding activity to excess: gambling, collecting, participation in favourite activities or relationships have all been recorded.

One participant in the present study reported that a man with PWS to whom she provided support often engaged in challenging behaviour as a way to seek and hoard materials for handcrafts:

"... or every time we go to The Warehouse he wants to purchase things. Like it's not just food with him ... (it's also) wool, 'cos he knits."

For another man this difficulty setting limits on a pleasurable activity, combined with the relative insensibility to changes in temperature that is also common among people who have PWS, was reported to have contributed to his tinkering with the controls of an air conditioner:

"... he's now in a flat that's got air conditioning ... we tried to turn the main switch off because he would go and turn the air conditioning on because he couldn't relate to ... and then of course he'd be

sweating and he'd lose salt again so for his wellbeing we had it disconnected ... because he'd figured out how to go back and turn it on so we had to have an electrician come in and dismantle it."

Participants in the present study often also described individual services users with PWS as being inflexible; stubborn, oppositional, argumentative, controlling or otherwise unreasonable:

"... if he really wants something you can't really say 'No', that's not a good word to use, you have to think about other things and you've got to ask him something else to try and get him off track but, when he's ... stubborn and in that mood ..."

"... new rules, new house, his bedding gets washed once a week. Whereas before it wasn't getting washed, he wouldn't allow staff to wash anything."

One participant noted that her clients who have PWS tended to form very firm opinions very quickly, often based on incomplete information. Misunderstandings and conflict often resulted. She said:

"Yeah you pick your time, you pick your time. Double staffing used to be tricky ... They would overhear little bits of conversations that could actually make them anxious because they get the wrong end of the stick. You know how? With the cognitive rigidity?"

This caregiver also reported that managing such inflexibility was complicated to a surprising degree by her clients' cognitive strengths, especially their relatively strong language skills. Likewise, another caregiver simply expressed her frustration at the difficulties she often encountered in communicating with her client, especially in presenting him new information and trying to encourage him to change his mind:

(Long pause ... sigh) It's ... it's trying to ... explain to him and get across that it's better for him."

The tendency to inactivity among some people with PWS was also described in the accounts of some participants, for example:

"... because he's so hard to get to do exercise."

“... he knows that he’ll lose his one-to-one funding if doesn’t get in the community and he understands all that ... So we try and do his programme that he does get out and do stuff, but he doesn’t want to ...”

Interpersonal conflict: Most respondents identified frequent disagreements between their clients who have PWS and caregivers, fellow residents or family members. The disputes that resulted were frequently characterised by inflexibility and intense emotions on the part of the person with PWS.

“It’s the personal interactions you know, they are difficult because they’re not black and white, they’re not equipment, they’re not property, they’re not food; you can’t take it away.”

Subtheme: Relationship dissonance. Initial analysis suggested that the various accounts of interpersonal conflict between service users with PWS and others seemed diverse, without obvious common features or relationships between them. Some featured intense expressions of anger on the part of a service user others seemed to be examples of perseverance, confabulation, tactlessness, or simple conflicts of interest between residents. However, deeper analysis showed commonalities between these accounts. Many included an element of surprise, incongruity between the expectations that caregivers had formed about their relationship with a person with PWS and that person’s subsequent behaviour. This dissonance often seemed to intensify the negative emotional impact of the challenging behaviour. For example, several caregivers reported feeling that they had been fooled or manipulated, even betrayed, by their clients’ challenging behaviour, and some seemed uncertain about how best to assess future risk. The term relationship dissonance has been coined here to describe this incongruity between expected and actual social behaviour, which seems to be a feature of supporting adults with PWS.

Sometimes this dissonance seemed to arise after participant caregivers or their colleagues had formed an unrealistically high impression of their client’s cognitive ability, often based on the service user’s relatively strong language skills. This risk has already been identified by Dimitropoulos et al. (2013) in their investigation into differences in the use of language associated with the two most common subtypes of

PWS. One supervisory manager who participated in Study 3 made exactly this point when she described a man to whom she and her staff provided support:

“Because he converses like you or I so people will naturally assume that he is willingly being defiant, when he’s not willingly being defiant he can’t help it you know. So whilst for me it’s like the light bulb’s on and I can see clearly now, um for staff it is so immensely hard because he, he constantly, they feel like they’re negotiating with him, being what they say, ‘defiant’.”

Another participant, who also works as a supervisory manager, also referred to the risk of this kind of misunderstanding among her staff, noting that:

“The communication ... knowing the level of understanding that the person you’re supporting has, because they don’t understand everything, they may be really well versed and they can speak to you as if they do, but they don’t really sometimes.”

Relationship dissonance also seemed to arise in other ways. Sometimes even the descriptions that caregivers gave of individual service users’ personalities included contradictory elements. For example, individuals with PWS were often referred to as being notably sociable and interested in others – as “friendly”, “very good company” and as “a gentleman”. One man was referred to as “a generous man” and was described giving gifts. At the same time, the conceptualisation of people with PWS as egocentric pervaded all of these conversations. Individuals were also described as “self-absorbed”, “very selfish” and “money oriented”, adjectives that might have denoted real empathy towards the needs of others, such as “thoughtful”, “considerate”, “understanding” or “good house-mate”, were never used by any participant in describing a person with the syndrome.

This sense of relationship dissonance seemed to be particularly intense for caregivers whenever they described behaviour that contradicted their beliefs about the nature of the relationships that they had formed with service users, especially concerning the level of reciprocity and honesty that caregivers had come to expect. One participant laughed briefly as she described a common expression of surprise among new colleagues when they first experienced an episode of overeating or other seriously challenging behaviour:

“‘We had an agreement’ ... ‘He promised me!’”

Others seemed to resolve the uncomfortable dissonance by interpreting incidents as the person with PWS being very clever, dishonest and manipulative, and as having fooled their caregivers in a kind of long running confidence trick. This also led to individual service users commonly being described in contradictory ways; as Machiavellian at the same time as their significant ID was also acknowledged:

“Because he’s very, very clever and he’ll often try and trip you up.”

Some participating caregivers seemed to have come to expect these kinds of surprises and to regard them with something approaching fatalism, as a measure of the strength of the drive to seek food and as a fact of life for their clients and themselves. For others this sense of dissonance seemed impossible to reconcile. Several accounts of service users’ challenging behaviour were related with an air of ongoing confusion and disappointment. One experienced caregiver related the story of catching a service user returning to the residential service after he had absconded overnight, she still seemed to feel a sense of surprise when she described this man’s subsequent insistence that he had not been out:

“... and I was looking and I saw him come strolling down the driveway, and he was going to climb back in the window ... and he looked me in the eye and he said he hadn’t been anywhere.”

Another equally experienced carer expressed her feelings in the face of this relationship dissonance in this way:

“... possibly the hardest thing is if I believe he has eaten something and I ask and ... he will lie to me. That would be the hardest ... I know that it goes with the, with the Prader-Willi but to me I still think, you know, you’re still lying to me ... and I don’t like it, I don’t like it.”

Subtheme: Threats to safety. Without exception, participants expressed both a sense of personal responsibility for service users’ safety, and an awareness that harm could arise rapidly and be very severe if their clients who had PWS were exposed to unrestricted access to food. The majority of participants described being worried and sometimes feeling powerless in the face of this responsibility:

“I guess as a manager it’s the worry that something’s gonna happen, um, that he ... is gonna overeat and then something will happen to him ... That’s a challenge for me because I can’t control that ... Well I just worry that he’ll overeat and you know ... off to hospital and bowel obstructions and all the things that go along with that.”

“I get paranoid ... like when I do sleepover I lock it. Go ‘round, make sure everything’s locked, and then I think ‘Oh’, and sometimes I get back up; ‘Did I lock it?’, which I have.”

Participants also commonly reported feelings of sympathy for the effects of the restrictions that they themselves placed on service users to protect their safety. They sometimes also gave accounts of unpleasant emotions, such as guilt or concern for their own reputation, which arose for them when they believed that they had been obliged to compromise other values in order to prioritise safety:

“... and it can look mean and really hard and unfair ... I feel like I play the bad guy quite a bit.”

The experience of a client’s premature death. For at least half of participants these feelings were complicated by memories of serious negative outcomes that had arisen in the past for clients who had had PWS, either as the result of severe obesity or after a single incident of hyperphagia. Participants gave accounts of the premature deaths of two young adults. They made reference to the feelings that had accompanied these experiences, and some described the processes by which they had been able to continue to practice following them.

Interviewees’ responses tended to be more terse and less obviously emotional when they described these experiences, their accounts were delivered in quieter voices. This seemed to reflect a sense of stoicism or professionalism in the face of adversity. For example, one caregiver discussed the sudden death of a man with PWS following a single episode of overeating. She was asked if the experience had been hard on herself and her colleagues. “Well,” she replied, “*it was tough on everybody.*” Later in the same interview this participant caregiver reported that the experience had:

“... kind of scared me about working with people with Prader-Willi ... I shouldn’t say that because I’m still working with someone.”

Responsibility without power. One other participant, also a supervisory manager, described witnessing the rapid decline and death of a young person with PWS due to the consequences of severe obesity over which caregivers could exert no control:

“Three months after ... his size went up. He doubled his size ... He doubled, he was always big.”

“And that’s when we knew that he had diabetes, so from that time on for the past two years there was a definite decline.”

This caregiver described the intense, and often very creative, efforts that she and others had made to intervene, in the belief that they were trying to save this man’s life. She talked only briefly about her sadness and frustration at their inability to do so:

“In my case notes, which I’m waiting to get told off about, I put that (name) was going to die because we were unable to support him in the community.”

“Okay, ‘What was the most challenging or difficult thing about your work?’ It was the food and, and knowing ... legally there was nothing we could do.”

This caregiver’s account reflected the cardinal importance ascribed to client safety by caregivers. It was also one of a number of stories in which caregivers described themselves as powerless to mitigate the risks associated with uncontrolled access to food, either because these participants had found themselves unable to influence the behaviour of a person with PWS or because they had been unable to convince others who held the socially legitimate authority to compel the service user towards a safer option. The experience of having responsibility and accountability without power seemed to be a common feature of the caregiving experience for many participants.

The safety and wellbeing of others. Almost all participants also gave accounts that reflected their concern for the ways in which the wellbeing of other service users might be affected by the support needs and behaviour of their clients with PWS. The only interviewee who did not was a caregiver who supported a man with PWS to live on his own. Most of the negative effects that challenged caregivers were framed as

chronic rather than acute. For example, one participant reported that she believed it was unfair that her clients who did not have PWS had to “*unchain the fridge*” if they wanted to make a cup of tea.

Several participants described their concerns about the way in which the increased risk of expressions of anger by a client with PWS, or of interpersonal conflict involving that person, impacted the lives of fellow service users and others, including themselves:

“Because behaviours ... can be violent with Prader-Willi because the need for food is there, and that’s a real concern of mine, of people getting hurt.”

Subtheme: Vulnerability to criticism. Interviewees not only reported being challenged by the responsibilities they bore in their work, but also by the ways in which they might be held accountable for its outcomes. Participants described their experience of being accountable to, or vulnerable to criticism from, a wide range of different stakeholders including their clients, their employers, their team-mates and colleagues working in generic services, the families of their clients with PWS and those of other clients:

“It’s the family ... Yeah because ... like I say with the weight ... they’re constantly on our back.”

“... and someone will walk into that component of a conversation without the history of it, you know? ... I often have reports about ‘Your staff are really unfair to him.’”

“I don’t know if people think you exaggerate or what.”

Some participants also described experiences in which they and/or their clients had come under the scrutiny of certain societal agencies such as the Police, the media, or the courts.

Caregivers’ experience of isolation. Most contemporary services to people with disabilities are deliberately decentralised. As a result, many of the caregivers employed to operate them spend the majority of their time working alone. Interviewees who contributed to this study frequently reported that they regarded themselves as especially isolated, even from the community of other caregivers who could otherwise be expected to be their colleagues. This sense of isolation seemed related to a belief that the task of supporting a person with PWS required a unique set of skills and knowledge, and was often

misunderstood by those employed in generic support services. This view was reflected in participants' often stated belief that *"Not everyone can work here."* This belief seemed to increase the participants' sense of isolation and vulnerability to criticism. It also contributed to the belief that, when such judgements arose, they were likely to be ill informed and hence difficult to rebut.

Subtheme: Challenge as dynamic yet unrelenting. All of the caregivers who participated in the present study had also provided care to people with learning disabilities arising from other causes in addition to their work in support of adults with PWS. Most reported still doing so at the time of these interviews. Differences in caregivers' experience of the two roles were raised several times. In particular, interviewees emphasised the vigilance that had to be maintained around the support of service users with PWS, regardless of other demands on their time and resources:

"He still tries his luck."

"It's actually a little bit like being on the stage with them 'cos they're watching every, every move you make ... to see if you're going to slip up with locking the doors."

The creativity, and hence unpredictability, of foraging behaviour among clients with PWS was noted frequently. Sometimes this unpredictability was reportedly exacerbated by emotional volatility on the part of individuals with PWS:

"... sometimes that's hard because sometimes he's all happy or smiley or hops in the van and then, I don't know if something set him off, um ... two weeks ago, two minutes or whatever, and he can just flick a switch and become really angry."

This experience of risk as relentless but at the same time as unpredictable and dynamic featured implicitly or explicitly in every interview. The negative impact of these twin pressures on caregivers' own wellbeing was noted more than once. In describing the experiences of her staff, one service manager reported:

"They're constantly being worn down, you know."

Another said:

“... actually it’s difficult to support someone with Prader-Willi all the time! ... I think you almost need twice the annual leave, you know? ... but there isn’t twice the annual leave ... You’ve just got to cope, and suppose sometimes you’re having a bad day we can say ‘Well you’ve just got to be professional’, but we’re working with people, you know?”

The Second Theme: Response to Challenge

Subtheme: Authoritarianism. No participant advocated uncontrolled access to food for any service user with PWS. All reported imposing at least some measures to limit food seeking. All were also aware of at least the appearance of conflict between these controls and the values of independence and client choice espoused by their employers and the bulk of their colleagues:

“Well basically I was ... protecting him from himself, but (employer) policy, or their policies, state that we don’t protect them, we support them.”

In the face of this dilemma participants invariably described choosing to prioritise client safety. Most also described making compromises and tolerating a degree of risk to reflect other priorities, but no participant reported or demonstrated a belief that threats posed to clients’ safety should be tolerated or ignored. This belief seemed to represent a kind of bottom line among caregivers; it was implied even when it was not stated overtly and it was never contradicted. For example:

“We do promote you know, independence and stuff but that, for his own safety that part has to be taken away to a point where you can’t just say ‘Okay you make yourself a cup of coffee, I’m just going to be over there.’ You can’t do that.”

“No you’ve got to, you’ve got to safeguard them from, you know, from gorging themselves to death, or ... from rushing over a road because they can see somebody’s discarding some buns ... You’ve got to put those safeguards in place, those environmental safeguards.”

“.... and I’ve said, ‘You don’t have to like it but it does have to be that way.’ Yeah.”

Interviewees were clear that they sometimes adopted frankly authoritarian measures to maintain client safety in relation to food. This was often reflected in participants' descriptions of situations in which negative service outcomes had arisen. Such accounts were typically couched in terms of caregivers having lost power:

"But ... we lost his control; we lost him listening to us."

Few, if any, of the resultant interventions were described as clients' choices:

"The staff that work there, they go and buy his food separately."

"... but we're looking at changing that ... we're gonna do that on Tuesday, have a good look, and take that temptation away from him even more so."

"We have locks on fridges, pantries, rubbish bins."

"We've got a lock on the pantry door and the civil defence cupboard's locked as well. We've got a freezer in the pantry and that's locked."

In fact, many restrictions seemed to have been planned in such a way as to preclude dissent or even discussion. In many accounts it seemed that steps were taken to ensure that residents with PWS remained either unaware that restrictions had been imposed, or were otherwise presented with them as *fait accompli*:

"Ah this food management plan's been set for ... years. It's been reviewed, all the calories and that have been reviewed so that it's been, it's set. It's in a proper menu folder there, it's set for staff."

"... because I didn't want to enter into a, for lack of a better word, a negotiation period, into something that I would have to say 'no' to him about, ..."

"Because what I did was, when I come to work here um, I used the slow cooker. And I went to cook the food in there because it contained the smell so that when the men came home and they had to go and get ready they wouldn't be constantly, on my back ... 'What's for tea', because the smell, the senses."

“... Ah, I learned a lot about the syndrome then and we had locks on a pantry, and also the vegetable compartment but they just weren’t successful because they would just pick the locks ... We had to do something permanent so we got um ... the builders ‘round and we got a solid door, and we had um ... a small walk-in pantry.”

Sometimes participants seemed to regard themselves as almost equally powerless in the face of the need for such controls. They generally reported that they took authoritarian measures reluctantly. The language used to describe the processes by which caregivers made decisions around service users’ access to food was often the language of compulsion: *“We were forced to ...”* and *“We had to ...”*

There were also rare accounts of interviewees choosing to adopt an authoritarian approach even at times when their clients’ safety was not immediately threatened. One participant reported subjecting non-PWS service users to the same measures for food security that had been developed for a fellow resident who had PWS. This decision represented an effort to cause all members of the group to lose weight and to improve their health. On rare occasions, participants also reported imposing limits on the non-food related choices of service users’ with PWS.

Justifications for substitute decision making. Restrictions imposed by caregivers on service users’ who are not believed to be competent to decide for themselves are examples of what has been termed substitute decisions by those who have written about services to people with generic learning disabilities (Dunn, Clare, & Holland, 2010; Gill & Fazil, 2013). Like the caregivers interviewed by Gill and Fazil (2013), some participants in the present study expressed their reluctance to make these substitute decisions, but justified doing so by referring to the duration and quality of their relationships with the service users who would be subject to them. These relationships were regarded as lending a kind of legitimacy to substitute decision making, both because they demonstrated caregivers’ commitment to service users’ wellbeing and because the caregiver was believed to have developed specialist knowledge concerning service users’ needs:

“And he knows that when I start getting involved ... he knows authority ... Yeah, and that I care, I care as well ... Yeah and he knows that I know, he comes and he says ‘Oh you and (caregiver) just, you just pick on me’ or he’ll not say ‘pick on me’ but ‘You gang up on me’ and he’ll laugh, he always laughs so he knows that we know everything. He tried to pull the wool over our eyes but we know.”

Some participants in the present study described trying to avoid the need for substitute decision making by seeking consent and negotiating with service users:

“‘You agree with that?’ and then well they might say ‘Yes’, and you say, you know, ‘well these are the reasons why.’ ... There’s still a little bit of leeway to get through, to get through to them. It’s about picking your time when you’re going to talk to them.”

However, these negotiations invariably seemed an unequal contest. They took place only within limits already set by caregivers:

“Within this range there is flexibility yeah but this is the range, and he knows that, he’s lived with that his whole life.”

At other times, such discussions seemed to be a means to convince the person with PWS that the substitute decision had already been made, and that the restrictions that were already in place were so comprehensive that any attempt to subvert them would prove futile. For example, one caregiver described a conversation between a colleague and a man with PWS concerning some local businesses that the service user had been visiting covertly:

(Caregiver) said: “I also know the owners and the workers there, so you better tell me what you’re gonna do so that when they ring I can say that I know you’re there.”

An apparent exception. Here it is important to note a significant contrary experience. One caregiver described arriving at a successful food management regime entirely through open negotiation with a man who has PWS:

“Ah, at (resident’s) request he has locks on his cupboards ... he has locks on his fridge and his cupboard. Staff come in and, he can manage his own breakfast, and staff come in around lunchtime and then they also do meals.”

This negotiation process reportedly took considerable time and required both the acceptance of an increased degree of risk and some trial and error:

“... but when we know that the food has been disappearing or something like that, we ask directly ... Sometimes ... we don’t get the truth (long pause) and ... we work on those issues.”

The resultant food management regime seemed to have been quite successful. The man in question reportedly kept excellent health and had maintained stable BMI in the normal range.

In considering this example we should note that this service user was also regarded by caregivers as being both more intellectually able than others who have PWS, and as having inherently less severe difficulties with food. His story seems to represent an illustration of the (as yet undefined) variation in the nature and severity of difficulties with food among people who have PWS. Even this relatively liberal food management regime was essentially the product of substitute decisions made about the management of risk by paid caregivers.

Other participants also described food management plans designed, at least in part, by negotiation with service users. However, these caregivers reported that each of the agreed plans had ultimately been overturned unilaterally by caregivers after the service user concerned gained weight:

“We tried it without the keys ... You know, to give (service user) that chance.”

One participant described a regime that allowed a service user to choose aspects of their menu within a calorie controlled limit:

“... we started calorie counting with him. He really enjoyed that because there was some flexibility in that you know, and he could, he could count that, but then that didn’t really work so well because the

nature of Prader-Willi Syndrome is that, even with the calories, it really still needs to be good food. If it's high in carbohydrate and the weight gain, they can't lose the weight."

This plan was also discontinued despite the opportunity to choose being valued by the person with PWS.

Substitute decisions, empowerment and compassion. Interviewees often described the reduction of negative emotion in service users as an important goal in itself. They often framed descriptions of substitute decision making as compassionate responses to difficult circumstances. The food management regimes which resulted were often described as empowering service users with PWS in that they settled all question of access to food and so reduced anxiety and challenging behaviour:

"Um, because the guys, if the guys even see food their anxiety levels are elevated, so 'out of sight out of mind' and, you know, we're being quite kind to the guys, doing that."

"I don't want him worrying about that so I told him, 'We're gonna make it so that you always talk with us and we'll make it work so that you don't stress.' He's very anxious you know, he hurts himself and does things. So, 'what can we do so that you can actually go and have fun then, and don't have to feel guilty?'"

"... so we put an alarm there so he could get up and go to the loo you know, and he didn't have to worry ... what we also do at night is we set the alarm off deliberately when we go to bed, we put our arm through and call out, 'Yeah, just checking it's working', and so that's so he doesn't have to get up and check that it's working."

"So because you're doing that, what do you think? What do you think we need to cut back on so you can go and do that and you don't need to be anxious about it?"

These views are consistent with those of Holland (2015), and reflect the experiences of some overseas caregivers who have written about their work (Forster, 2010; James, 2010; Waters, 2009; Ziccardi, 2006).

Subtheme: Creativity. Examples of authoritarianism in the form of substitute decision making arose in every interview. However, most participants also described seeking ways to avoid, minimise or mitigate the effects of the substitute decisions they felt obliged to impose:

“You can’t be a, a dictator because then he is constantly going to fight against you.”

“... it’s about him maintaining his independence, not treating him like he’s a child ...”

“... for me I don’t even see it as being valuing but like it’s his right, to say something about it you know.”

These attempts seemed notably creative and effortful; they consumed considerable caregiver time, energy and other resources.

The promotion of safe autonomy. Many caregivers described seeking ways to promote an element of choice or independence for service users within the bounds of the food management regime, which remained sacrosanct. One common way of doing this was by promoting food choices within the imposed calorie restrictions:

“You can still work within that model to give them some rights. They can choose the meat and what, what curly lettuce they’re gonna have that night.”

Choices deemed safe were also promoted in areas unrelated to food:

“Well I thought, ‘If he wants to sleep on the couch, you know? ... I just made sure we bought him a new couch.’”

Tolerance for a degree of risk. The acceptance of a degree of managed risk was another tactic commonly described as a way in which participants sought to promote autonomy while still prioritising service users’ safety in relation to food. This was illustrated by several stories in which caregivers supported service users’ efforts to access community groups or activities. In one account a man with PWS developed an interest in greyhound racing. Another outlined the way in which a service user simply enjoyed regular visits to a local park by himself; ostensibly to go for a walk but in practice to sit down and look for members

of the public with whom to have a conversation. One participant even described supporting her client with PWS to drive and maintain his own car. The stories of service users who were supported to join a cricket club, a local church and a knitting circle have already been described.

Each of these activities carried risks. In almost all accounts participants reported at least the suspicion that their clients' initial interest in these activities was generated by the search for new foraging opportunities. Participants described accepting and assessing these risks consciously and then actively managing them in order to maintain their client's safety while respecting their choice.

These activities also generated other costs for caregivers. For example, each represented not just increased risk but also additional demands for caregivers' time and other resources. In the context of interviewees' stories, however, participants invariably framed these activities as opportunities, not as problems or liabilities. Caregivers spoke with satisfaction about the degree to which their clients enjoyed the social interaction that arose from them. They also claimed other benefits such as an increased sense of independence, the chance to fill a valued role, or simply to be free from the company of paid support people for a time.

The limits of tolerance: safety remains paramount. Even within these anecdotes of fruitful community engagement, the assumption that caregivers' primary responsibility was to ensure client safety remained constant. In facilitating service users' community activities participants typically described making substitute decisions in order to limit their choices and so reduce risk. Some participants described seeking control over service users' personal finances, and many reported forming alliances with families and members of the community in conspiracies to subvert attempts at foraging. Several of these accounts featured an element of trial and error or of improvisation. One participant related the story of a colleague receiving a phone call from a local cafe owner saying that his client, who had PWS and who was believed to have been attending a public event at that time, had entered the cafe in the absence of a support person and ordered multiple meals. The caregiver on duty reportedly went to the cafe, sat down at the resident's table and said:

“Oh I see you’ve ordered for me. Thanks very much, I’ll have the steak.”

Caregivers often reported tolerating a degree of risk and making strenuous and creative efforts to empower those they supported. Clients’ physical safety remained paramount however. Empowerment was only promoted within limits and these limits were determined by caregivers.

Subtheme: Exercise. Many participants described using exercise as a tool to manage weight with the purpose of allowing greater flexibility around food. The exercise regimes that resulted were idiosyncratic with no particular mode of exercise being generally applicable. No caregiver who participated in the present study employed compulsory exercise, as advocated by some authorities (Pittsburgh Partnerships, n.d.) but several of them described implementing plans which resulted in long-term increases in service users’ level of physical activity. This was often apparently aided by the PWS preference for routine and by a willingness on the part of caregivers to listen to aspirations of the individual they supported:

“... we made a plan with him that he scooted to school which was 3 k’s away, and scooted home ... Yeah, we worked out the exercise. The scooter, which he worked very hard for, was absolutely brilliant ... the, the 6 k’s a day, he never gained from then on.”

In contrast, several interviewees reported the belief that their clients were simply unable to lose weight through exercise. Other caregivers had also found that they were effectively unable to motivate those they supported to engage in it in sufficient quantities to be worthwhile:

“They try to get him out go for a walk, that’s a bit hard. (Staff person) she’s quite good, she’ll say, ‘Well how about we walk into (suburban shopping centre).’ Well she says, ‘We’ll get a little treat when we get into (shopping centre).’ So they do walk but he wants to know, ‘Are we there yet?’ You know, ‘Have we got our ...? Can I get my treat?’ or ‘Have you got my treat?’”

Subtheme: Practical wisdom. This term is a very old one in the study of ethics (Broadie, 1991). Used here, it refers to the body of specialist knowledge and beliefs built up over time among a group of caregivers. Data elements coded to this subtheme were quite varied but they included descriptions of either

attitudes or practices which caregivers believed were important for good practice; all were intensely practical.

The use of the term in this way follows Abma et al. (2008). They described the process by which specialist nurses who were engaged in the care of people with ID developed an ethical or moral resource which they subsequently used to address the problems they faced in their work. For Abma et al. (2008), this practical wisdom arose from experiences, and was hence distinct from either ethical first principles or abstract notions of right and wrong.

Abma et al. (2008) have particularly emphasised the importance of the stories which nurses told each other about those experiences. They cite the nurses' use of these stories as an example of the dialogical process of ethical development, whereby ethical decisions are made through a social learning process constructed out of the dialogue of stories and responses to stories shared within a community of initiates.

The dialogical process within the present study. It was notable throughout these interviews that participants generally seemed to enjoy the opportunity to discuss their work and to describe the lessons they had drawn from it. It often seemed that this was due at least in part to the interviewer having shared similar experiences.

Every participant related anecdotes and gave examples of the different kinds of support they had provided and the outcomes achieved for service users. Many of these accounts were success stories, generally about food management or weight loss:

"... we have some input from the ah ... dietician ... and he's lost a ... lovely lot of weight ... he's much better for it, you know, able to move around a lot more."

Some concerned independence achieved in other areas of life, such as the aforementioned man with PWS who had recently passed his driving test. Stories such as these were usually accompanied by smiles and often short periods of laughter. This sometimes also occurred, although perhaps to a lesser extent, even when participants described outcomes that had proven less positive. When they gave reports of mistakes made by themselves or their colleagues, or described innovations that had not proven successful.

Not only did participants seem to enjoy and find value in telling their stories, but these narratives seemed to act as vehicles for a dialogical process which took place even within the interviews on which the present study is based. The anecdotes related by participants were employed purposefully and each seemed to carry a subtext, a symbolic meaning like the moral within a parable or a folk tale. Each narrative illustrated some aspect of the interviewee's position on an ethical or practical issue, and in each of these stories this subtext seemed to invite controversy. It challenged the listener to agreement or disagreement and so invited the extension of the discussion.

For example, one participant described how a man with PWS who she supported had managed a situation in which an inexperienced caregiver had given him a slice from a chocolate cake that she had baked in his presence, but that he also knew was not on his diet plan for that day:

“Um, so here he is, his poor face, with this little wee slice. She said, ‘Aw it’s only a little slice.’ Wouldn’t have mattered if it was one little chocolate button, it’s cruel. And here he is, he took it all the way to the day base, in the van, and he gave it to the day staff and he says, ‘I’m not supposed to have this’ ... You know, that’s what I mean by, occasionally he will make some really courageous decisions ... He was really proud of himself. Really proud of himself.”

This account celebrates the man's success. It expresses sympathy for his circumstances and admiration for his response in the face of difficulty, but it also carries a strong message about the importance of solidarity and fidelity to the team plan among members of the group of caregivers involved in his support. The interviewee's frustration at her colleague's behaviour in giving an unplanned piece of cake is palpable in her choice of such an emotive adjective to describe that behaviour; one which carried such a strong moral component: *“it’s cruel”*.

The participant's bold assertion seemed to invite discussion or debate. She even began the discourse herself by rebutting her absent colleague's moral justification:

“She said, ‘Aw it was only a little slice’, wouldn’t have mattered if it was one little chocolate button.”

The position taken by this caregiver was a very definite one, and it would have been difficult for the interviewer to respond without taking a similarly polarised stance, either to agree or disagree. One option open to the interviewer in framing that response would have been to couch it in terms of a counter story; either “yes but” or “yes and”. In any case, the subsequent discussion would have required both parties to consider the proposition advanced by the interviewee in a symbolic yet intensely practical way, using this vehicle of stories and responses to stories.

This seems very like the dialogical process described by Abma et al. (2008). It also seems likely that at least one of the reasons why interviewees seemed to enjoy the interview process was that it afforded the opportunity for each of them, and for the interviewer, to discuss their stories. This in turn presented us with a chance to grow our own stores of practical wisdom by challenging or confirming our beliefs about the complex and ill-defined task of professional caregiving.

Caregivers and resilience. Many interviewees made reference to the importance of caregivers maintaining certain attitudes and core beliefs in the face of challenge or apparent failure. For example:

“No ... You can’t get offended, you can’t get offended by the abuse ... You’ve got to be able to think straight, you know, when there is a raid on your cupboard and you think ... it’s not the end of the world. You start fresh.”

“You know we don’t make mountains out of molehills or sweat the small stuff.”

“So ... once again you just gotta let it go over your head!”

“You don’t get upset about that sort of thing ... you just learned, ‘Okay (team leader’s name) is going to be that one that will deal with him.’ So we learned, we learned.”

These comments also reflect participants’ views about the importance of self-care among caregivers. Many of them were also accounts of caregivers learning by experience. They are among the quite numerous accounts of caregivers tolerating the use of simple trial-and-error as a means to eventually solve complex problems.

The inevitability of risk. In their ethnographic study among caregivers working in a British residential service for adults with PWS, Hawkins et al. (2011) noted the fundamental importance of the concept of risk in shaping both agency policies and caregivers' day to day practice. In the present study, caregivers' attitudes to risk seemed to link the themes of challenge and response within the dominant narrative.

As noted, participants were universally clear that risks to the physical safety of their clients were of primary concern:

"But ... at the same time being very aware that if he had free rein that he could eat himself to death ... Absolutely."

Participants' responses also reflected a view of risk as pervasive and difficult to control entirely. The risks associated with food often seemed to be regarded as a perennial problem, one which had to be accepted and managed because it could not be entirely eliminated. This view of risk provides an important context for our interpretation of caregivers' willingness to tolerate it in small measures in return for other benefits to service users. If, from the caregivers' point of view, some degree of risk is ultimately inescapable then neither safety nor autonomy for their clients are really available as absolutes and planning will necessarily imply compromise. For example:

"... then we have to remind ourselves that we're only in the house a certain, a certain amount of the day ..."

"... we support him to be as independent as he can be, so he has this walk that he's allowed to do without staff, because he's very high functioning, he can ask for help, so the kind of safety risk in terms of the community wasn't an issue, the only safety risk was in what he might do unsupervised because of course he's going to be seeking food."

"... and while he does have slip ups and stuff they don't seem to be um major, touch wood um and (laughs), you know he just sort of rocks along you know."

The importance of managing anxiety in people with PWS. A high level of chronic anxiety is a common problem for many people with ID (Kortisas & Iacono, 2015). Participants in the present study often made reference to their efforts to prevent stress or anxiety among those with PWS. Such anxiety was regarded as being associated with negative outcomes such as emotional distress, interpersonal conflict, and the increased frequency and intensity of challenging behaviour. Events or practices which were identified by interviewees as likely to exacerbate anxiety tended to be those which acted to reduce service users' ability to predict events around them; unexpected variations in routine or inconsistent practices among caregivers for example.

When caregivers described their responses to challenging behaviour their accounts often included measures apparently intended for the reduction of anxiety. For example:

"The strategies I use on (name of resident), like he was ... um elevating and kicking and going like that (mimes repetitive kicking at an object), and he went to his room, I'd go in there and I'd sit down, get him to talk about it ... I'd do anything to de-stress him, 'cos I know he's stressed ... even if it's for nothing, he's stressed."

Likewise, two participants independently described helping clients to plan ways of behaving appropriately at impending social gatherings. They reported paying particular attention to helping their clients to plan how they would respond to the food that would be available at these events. Both caregivers reported that these interventions had proven very successful:

"And think ahead you know, like, 'We're going to such-and-such's funeral, now I know it's gonna be hard for you because there'll be food there.' You know, so you have to get inside how they're gonna be thinking, and, 'So I know this is gonna be a hard decision but would you like the staff there to sit (you) down and they go and get you three things, not cream or pastry because you know, we've talked about that, or would you like to take your lunch and eat it there?' And because they're stuck in routine, I had one young chap, 'No, I'll take my lunch.'"

And:

“I don’t want him worrying about that so I told him, ‘We’re gonna make it so that you always talk with us and we’ll make it work so that you don’t stress.’ He’s very anxious you know ... So, ‘What can we do so that you can actually go then and have fun, and don’t have to feel guilty?’ ... He’s less anxious because like, it’s been approved.”

These examples reflect a belief among caregivers that although service users’ anxiety was likely to be increased at these events by the prospect of unsecured food, this anxiety, and hence the increased risks, could be ameliorated by reducing uncertainty and increasing predictability. Allen (2011) found a similar view amongst the parents of children with PWS in the United Kingdom. These parents emphasised the practical importance of the establishment of predictable routines and of encouraging access to non-food related activities. These same strategies were often identified as useful tools for the management of anxiety by the paid caregivers who participated in the present study:

“Because I think that, like his routines are basically the same everywhere, it’s quite consistent and he’s happy in his world ... I think he’s happier because, having less behaviours.”

“... all the staff have to be on board and be consistent, because it will be a huge breakdown.”

“You just need to ... follow what everyone else is doing.”

“Yes. Definitely. You can ... you find you can keep them occupied in other activities. ... Like we used to have them doing little projects because they loved dogs, you know? ... Projects on dogs, all types of dogs ... Get them busy in other things. Um, by minimising the effects of anything to do with food ah, you know on them ... so they’re not exposed. ... Just the thoughts of food all the time, because they’re all ... they’re up against it anyway.”

Food security as a tool with which to manage anxiety. The control of access to food in particular was regarded as important for the management of anxiety among service users. As already noted, the belief that uncertainty about future access to food was likely to contribute to service users’ anxiety was a key element in framing substitute decisions in this area as compassionate responses, rather than simply as authoritarian ones.

Once again, for participants in the present study, predictability seemed to be a key to managing service users' anxiety. Any change or threat of change which reduced service users' ability to predict future access to food was regarded as also likely to exacerbate anxiety and to increase the risk of challenging behaviour. For example:

"We've had a little, few mistakes. If (resident)'s tomato is 1 cm smaller... um, it will throw him and he doesn't cope with that change, or if instead of buying the little carrots we buy a big carrot and cut it ... He doesn't cope with that either because it's different."

"So, so that's what I mean, working with him there's a lot of difficulties, and that's when the consistency's not in place so people think that they're doing (current resident) a favour by giving him extras ... Then he tries to build that into the routine and you have to work and start again all the time."

This reaction was not just described at times when a likely reduction in available food was implied. In fact interviewees reported that even the possibility of unexpected access to extra calories could trigger distress and challenging behaviour, as demonstrated by the caregiver who described the provision of one small piece of chocolate cake as "*cruel*".

Empathy. Many participants emphasised the importance of learning about those they supported and of understanding their worldview:

"This is a very close team that works with him so ... they know him extremely well and know what he's like."

"You've really got to be in their heads, you've gotta be in their heads."

Empathy for service users was generally regarded as an important characteristic of any effective caregiver, both in that it facilitated better decision making and it allowed caregivers to rationalise and predict their clients' behaviour. This in turn allowed caregivers to better protect themselves emotionally. As one supervisor put it when she described training new team members:

“You know I try and find a common denominator that I can liken his anxiety so they know he lives constantly at that level you know, and that before he even wakes up in the morning you know, and then he’s got to deal with life’s pressures and having a disability you know, and he does have an intellectual disability so there’s that, there’s that, he doesn’t reason, doesn’t rationalise, doesn’t have that next level thinking that we have, ... you know, so I try and get them to understand before I even go into day-to-day support. It is the most important thing for everybody.”

In this vein, some caregivers used metaphors from their own experience to approximate the experience of having PWS:

“I akin Prader-Willi to, I used to be a smoker many years ago, I akin it to um, day I coming off cigarettes ... that constant crave, that constant anxiety, and for me that’s how I kind of ... for me, get to understand how he, or possibly understand.”

Tricks. Some of the tools for the management of negative emotion among service users could best be characterised as benevolent deceptions, or more simply as tricks. Participants described some strategies that seemed clearly designed to give service users with PWS the impression that they had greater access to food or greater autonomy in other areas than was in fact the case. Tricks were also employed to help service users to feel better about their circumstances. They were sometimes also used to subvert the likelihood of protest.

Some of the most straightforward tricks centred on the way in which meals were presented. These exploited difficulties which services users who have PWS were believed to have in judging proportions by sight. The tactics of presenting meals on smaller plates and spreading food in such a way as to make the meal look larger were both described more than once, as was the practice of bulking small meals up with salad greens. One caregiver reported that using a steamer for vegetables had proven popular in her workplace because it made the food expand rather than shrink in cooking.

There was a universal assumption among participants that a service was improved if it was perceived to have become more fair or empowering towards those who used it:

“... otherwise it’s a dictatorship and life isn’t about being in a dictatorship ...”

“... within that ... like we try and give him as much control as we can. Like he can have two or three eggs per week and he chooses when he has those, ... and that kind of thing, so try and give some control, yeah.”

However, as already noted, participants were often quite frank in reporting that they sometimes sought to undermine dissent and help service users feel better about their circumstances by giving them the impression of shared power, in the absence of its actuality:

“And so you’ll have a little contract book, ‘Do you remember that contract’, you know ... They get a copy and we keep a copy, because quite often their copy will get ripped up. But if you can try and keep them, you know, on the straight and narrow ... and they’ve had a say in the matter and they know it’s for their own good, you’ve got more chance of it succeeding.”

The perception of fairness, choice, empowerment and mutual regard within the caregiver/service user relationship were often described as important ends in themselves. At the same time, for some participants at least, they were also tools which could be used to manage the complex challenges of the caregiving task:

“And ... really being empathetic, like with (current resident) you’re really listening, you really care, you don’t like seeing him angry and sad, and a good phrase we have in the house is, ‘(Current resident), how can I help you?’ ‘Please don’t be angry and frustrated. Can you ask the staff? Can you ask us so we can help you? ... And we say that a lot, and then, that redirects him. ... Because if he’s angry and you try to talk to him ... Growling ... just makes him angrier, but if you be empathetic and fair, ... and that’s just like with anybody, respect people, that goes a long way.”

“... they’re being very fair ... if you’re reasonable with him and he’s like, doesn’t have to like it but he’s like, ‘Well okay.’”

“... they know they’re up against it ... if they know you know they’re up against it, and you relay that to them, then they’re not going to feel so alone.”

“When I took over it was quite rigid but without ... no mention really as to why ... well I’m very fair and reasonable and I needed to be able to sell something, if I can’t sell it ...”

Subtheme: Consistency. The quality of support labelled here as consistency was the response to challenge which seemed to pervade all others. Whatever measures were undertaken to prevent or respond to challenge, caregivers were clear that success was likely to depend on the consistency of their application. Consistency was identified with a variety of specific caregiver practices including participation in workplace communication systems, adherence to daily and weekly routines, and resistance to inconsistent behaviour on the part of others. Even the effort made by individual caregivers to present an unchanging demeanour over time was expressed as a means towards greater consistency. This subtheme of consistency featured prominently in all interviews and was often raised when caregivers were asked to identify the interventions that they regarded as most important for service quality:

“Like it doesn’t matter where you work and what industry ... the team work. And for everyone to be on the same page and working towards the same goals with consistency and communication ... is the key to success I find.”

“For our service ... is everyone working off the same page, and that’s a consistency thing.”

“Once everybody’s on board you’re pretty sweet.”

Maintaining consistency was often identified as a major source of challenge in itself, especially by interviewees who had management responsibilities:

“It’s um ... the greatest challenge would have been ... it’s just keeping staff on track.”

“The biggest thing for me that I find is supporting the team to stay above the line.”

“... because if you go in there with ‘you’ve had a rough morning’ you’ve got to put all that aside. It’s like actually being on stage a little bit with them, because they’re watching every, every move you make.”

Several factors were identified as threats to consistency. All of the caregivers who participated in this study either worked rostered shifts in residential services or supervised others who did. Participants frequently made reference to being isolated from other caregivers and often working under resource constraints that limited opportunities for communication between colleagues.

The importance of consistency was often expressed in the negative, when interviewees related stories that illustrated their frustration at negative outcomes which had arisen from inconsistency on the part of nonconforming team members. There was general agreement that any individual caregiver's failure to maintain established routines and boundaries, or to otherwise display an acceptable level of consistency, should probably preclude that caregiver from employment in the support of any person with PWS.

One participant seemed to regard inconsistent practice as due to ignorance on the part of inexperienced colleagues:

"... sometimes if other staff came on and they didn't know they had to stick to it, that caused big problems."

A motive more commonly ascribed to account for staff inconsistency was the desire to avoid challenging behaviour for the duration of a given shift:

"Ah some people can't be firm on him because he'll just have a big, whopping behaviour."

"Yeah, when their usual staff aren't with them, they ... people don't want the behaviours ... So it's like what I said before, so they give in."

"Yeah because he's now been put in a situation where there's a wider variety of staff, and he'll know the weak link ... He, he knows the weak link, yeah he knows."

Such inconsistent behaviour being perpetrated for short-term gain engendered frustration on the part of many participants. Sometimes caregivers employed on short-term or casual contracts were regarded as being more prone to such practices because their temporary status meant that they lacked the same level of vested interest in long-term service outcomes.

Another dissenting voice. One participant's accounts of her caregiving practice seemed to subvert this narrative. This caregiver reported having developed her own set of support strategies that were different from both those of her colleagues, and from those which were already in place when she began to work in her present role. She illustrated her individual approach when she answered a question about the process by which she had learned to support an individual service user who has PWS:

"Ahhhh, I dunno. I guess just winging and trial-and-error ... Wouldn't you do that? ... Just wing it you know, to see what works, you know, what is workable."

In apparent contrast with the general emphasis on the importance of consistent practice, this caregiver did not seem to regard having an idiosyncratic approach as a problem that needed to be solved, but rather as an inevitability which arose from caregivers' individual relationships with those they supported:

"... because every person that works with (resident) has a different rapport with him."

However, even in this interview the concept of consistency as a desirable trait was not entirely absent. This dissenting caregiver recognised the value of consistency and predictability within the practice of individual carers, even if not between them. She also reported the belief that she experienced a lower rate of challenging behaviours involving her client with PWS because of her unique support plan. She implied that the adoption of her strategies by other caregivers, thereby restoring consistency, would have likely yielded benefits for all. In essence, this participant agreed with the consensus that consistency of practice across caregivers was a desirable feature of a support service for adults with PWS.

Families and consistency. Interviewees' interactions with the families of service users were also prominent when discussing this issue of consistency. Family members were often described as supportive team-mates and as valued resources for information and training. As one caregiver put it:

"... when I first started here he coned me left, right and centre. Until I got in with his Mum, and that was it, he's never, ever been able to con me again."

Participants related several anecdotes that characterised clients' family members and others as integrated into a wider support team, working in a kind of benevolent conspiracy against dangerous choices that might be made by the person with PWS:

"... he always says, 'Have you told Mum?' I said, 'I talk to your Mum every day, I've got her on speed dial.'"

In contrast, several participants described negative outcomes that had arisen through what they regarded as inconsistency on the part of family members. Some of these accounts featured severe consequences for the person with PWS or others, and many were related by participants with what seemed to be a sense of frustration and perceived unfairness. Participants described frustration at the negative consequences that had ensued, but also at their own impotence in addressing practices among family members that they regarded as problematic:

"... but sometimes when they go back the families also give them treats and then it's up to us ... a lot of things they impose on us they don't do it themselves."

"We went to (family member) ... We went three times, I went with my boss. ... We, we were not, we weren't going to get anywhere."

The Third Theme: Outcomes Valued and Sought by Caregivers

Given caregivers' conceptualisation of their role as one dominated by challenges, the question of what outcomes they sought to advance in selecting a response to those challenges was an intriguing one.

Subtheme: The search for a liveable life. Like the caregivers interviewed by van Hooren et al. (2002, 2005) participants generally defined the outcomes they sought in corporate terms, rather than as goals sought for an individual alone. This meant that even in choosing controls to which only those service users who had PWS would be subject, caregivers described taking the vulnerabilities and aspirations of other service users into account, along with those of a variety of other stakeholders. The goals described by

participants in designing a food management regime included outcomes sought for all those associated with the residential service, rather than for the person with PWS in isolation.

Van Hooren et al. (2005) described this framing of goals in corporate rather than individual terms as a key feature in what they termed caregivers' search for a liveable life. This search represents the pressing practicality of the caregiving task. If it is to be regarded as useful, any solution to the central dilemma must not only be workable in practice and sustainable over time, but must yield net benefits across the small community associated with that residential service, or at least not come at too high a cost to any of its other members. As one participant commented:

"I know, I know that there's only four men here but I think of the other three."

One very experienced caregiver provided an interesting example of this search in practice. She told the story of a trip to a local restaurant taken by four residents of the group home in which she worked. Only one of these residents was diagnosed with PWS and the evening had been planned as a celebration of the birthday of one of the residents who did not have the syndrome.

The caregiver described holding a preparatory discussion in which she and her client who had PWS agreed a plan for the evening. This plan centred on an agreement by which the service user would order the lowest calorie item on the menu – an omelette with a salad. On arrival at the restaurant, however, this man began to behave in a way which attracted a great deal of unwelcome attention and disrupted the evening for his friends and other diners.

The caregiver reported that, as a response to this behaviour, she sat very close to the man and told him that she believed he was behaving in this way because he wanted to order a burger and fries for dinner instead of the lower calorie meal to which he had previously agreed. Because the restaurant was crowded with other diners, and because the evening was a special one for the man whose birthday it was, the caregiver decided to order the burger with fries for the resident who had PWS. The caregiver reported making this decision despite being aware that in doing so she would breach both her prior agreement with her client and the plan for food security to which she and her colleagues had committed. Following her

decision the challenging behaviour ceased quickly and the meal continued uneventfully. The caregiver sought ways to compensate for the excess calories over succeeding days. At the time of the interview the caregiver regarded this incident as unfortunate and as a failure in their planning processes, but she was also clear that she believed that she had managed it in the best way possible.

This story illustrated one caregiver's search, under difficult circumstances, for a liveable life on behalf of the group of individuals to whom she provided support. The man with PWS who featured in it was the subject of an individualised diet and a behaviour management plan that the caregiver generally upheld consistently. However, from that caregiver's point of view, desired service outcomes such as safety, fairness, dignity, and community integration were goals defined corporately. Faced with a difficult choice it did not seem unreasonable to her to prioritise the outcomes being sought for other members of the group. In this case, she was particularly aware of the importance of the evening to the man whose birthday it was.

Another participant linked the achievement of desired outcomes for a particular resident with PWS to improved outcomes for other residents:

"Oh. Definitely! If he's happy then ... if he's happy then the house is happy."

This response reflects both that caregiver's search for a liveable life and the disproportionate effect which the behaviour of people with PWS have been found to have on others around them (Kayadjanian, Schwartz, Farrar, Comtois, & Strong, 2018; Tvrdik et al., 2015; van Lieshout et al., 1998).

Some participants extended this search for a liveable life to include consideration of goals sought for their client's family members:

"I used to feed back to them because it's not just about the guys; it's about alleviating the anxieties of the families as well. You know especially the ones, the ones who are really good at trying to get the food ... because they've got a standing in the community ... and you've got to keep allaying their fears you know? Sometimes they're business people, they don't want to hear that their kid's got out the blimmin' door and rushed down to (fast food franchise outlet) and been scavenging in the, the bins."

And, on rare occasions, for themselves:

“I said ... ‘I’m not gonna clean up this mess.’ I said, ‘Do you think it’s fair that I should clean this when I didn’t do this?’ ... I said, ‘That’s, that’s not fair’. ... And um so ... we worked it in and had a bit of a hissy fit but hey ... the trashing the room cut down by I’d say 85% ... or 90%.”

Subtheme: Safety remains paramount. As has already been noted, caregivers generally described prioritising service users’ safety above other goals. In describing priorities for her work, one respondent described her primary concern as:

“.... to maintain that he is healthy. You know, that his, that he’s, that he’s getting a regular doctor check-up. Regular blood tests for his sodium levels. Um that ... prompt action when he’s skin picking for ... all that’s sort of wellbeing. That he doesn’t have that access to food because that hinders his wellbeing, it hinders his being alive! You know?”

Other caregivers commented:

“... his health is really well monitored, that’s very important to us.”

“But we do promote you know, independence and stuff but that, for his own safety, that part has to be taken away.”

“And then once again you kind of feel, you feel a bit mean but I try to get past that because ultimately it’s all about keeping him well and keeping him safe.”

“The service users we support in the community are extremely vulnerable.”

“And the bottom line is, they got Prader-Willi and they need help.”

The prioritisation of safety was present in most interviews and was never contradicted. One participant who had experienced the death of a client with PWS after a single incident of hyperphagia described controls over access to food in this way:

“It needs to be done; it needs to be for the future, so that things that what happened to (former resident) won’t ever happen to anybody else.”

Subtheme: Other priorities; happiness, fairness, autonomy. Although client safety may have been described as the top priority among the goals which shaped caregivers’ decisions, several other desired outcomes were also identified as important, both for service users with PWS and for others. Descriptions of these secondary priorities were often framed as emotional states which caregivers sought to inspire in service users; feelings of equity and independence were key, as was freedom from anxiety and other negative emotions:

“I mean it’s enough that he knows he has to be in a service you know, so we try and give as much power and control.”

“It, it’s about him maintaining his independence, not treating him like he’s a child, um ... because, at the end of the day he’s old enough to make his own decisions.”

“Yes. No, no I mean it is all about fairness.”

“Yeah he ... But he doesn’t have to worry about it. I try to make like his home a safety net at least.”

Discussion

The Goals of the Present Study

The present study sought to address two issues. First, to investigate the ways in which a group of professional caregivers conceptualise and experience their work in support of adults with PWS. Second, the practical questions of how and why existing measures for the control of access to food come to be selected and established.

How Do Caregivers Conceptualise and Experience Their Role?

Participants primarily describe their role in terms of its challenges. They describe these as arising continuously, but also unpredictably from multiple sources. Caregivers conceptualise their role in large part as generating responses to this range of challenges.

Thrownness and being between. This conceptualisation of the caregiving role seems consistent with the conclusions drawn by Buckles (2016), who interviewed direct care professionals employed in the support of people with learning disabilities who engaged in what appeared to be sexually motivated challenging behaviour. Buckles used the term being between to summarise a sense among those caregivers that they found themselves continuously required to navigate between competing imperatives generated by factors largely outside their control. Metaphors similar to Buckles' (2016) have been a consistent theme in recent investigations into the challenges faced by caregivers of people with developmental disability. Solvoll, Hall, and Brinchmann (2015), for example, described caregivers as being squeezed between different courses of action, each representing different sets of values and desired outcomes. Tsungu (2014) has borrowed the term thrownness to describe the experience of direct caregivers as they found themselves presented with circumstances under which they were obliged to provide care but over which they had minimal control.

Parallels with the challenges faced by other carers. In the light of these conclusions it could be argued that many of the challenges identified by participants in the present work are not unique to those providing services to people with PWS, but are common to all those supporting adults labelled as having ID and challenging behaviour. Some of these commonalities will be explored further in Chapter 5.

The responses of those caregivers who have contributed to the present study do show some similarities with the challenges described by Buckles (2016), Solvoll et al. (2015) and Tsungu (2014), but their accounts are complicated by factors unique to PWS. Even difficulties that might be considered generic or common to the caregiving task are experienced and reported by participants in uncommon ways. For example, others have noted that both paid and familial caregivers may experience vicarious stigma and

isolation by association with those to whom they provide support (Ejaz, Bukach, Dawson, Gitter, & Judge, 2015; Selman et al., 2017). This phenomenon has been identified across a range of human services (Hewitt et al., 2008); however, among participants in the present study it is aggravated by the sense that their work is poorly understood and stigmatised, even within the disability community.

The pervasive impact of hyperphagia. Only one participant in the present study identified the management of access to food *per se* as the most significant of the challenges they faced in their work. This was a pattern broadly consistent with the reports of caregivers who contributed to Study 1 and arose despite the fact that all participants in both studies expressed an acute awareness of the risks associated with hyperphagia and obesity.

The drive to seek food among people with PWS seemed to act as another complicating factor which pervaded the work of the caregivers who contributed to this study. Common challenges, such as an incident of interpersonal conflict or the concern that might arise when a service user undertook an activity independently for the first time, were described as complicated by a pervasive sense of risk associated with food in a way that is unique to these services.

The dominant narrative. The three themes of challenge, responses to challenge, and the valued outcomes which caregivers sought to achieve through those responses, summarised caregivers' descriptions of their work and formed the dominant narrative within these interviews.

Challenge and response to challenge. When asked to identify the challenges they found most intense or difficult to deal with caregivers invariably made reference to some aspect of their clients' behaviour. As expected behaviours related to foraging and hyperphagia featured prominently, but participants also reported that it was often the interpersonal behaviour of service users with PWS that generated the greatest difficulties for caregivers and others. Participants from different services tended to identify similar features in the challenging behaviour of their clients with PWS: inflexibility; egocentricity, interpersonal conflict and confabulation. These reports are broadly consistent with those researchers who have described what Whitman (2013) has termed the PWS personality, and with the experiences of

European and North American caregivers who have written about their work (James, 2010; Waters, 2009; Ziccardi, 2006).

It might seem surprising that caregivers report such intense negative impact arising from behaviours that are largely verbal in nature but this conclusion is consistent with the findings of other researchers (Jurgensmeyer, 2016; Kayadjanian et al., 2018; Lanfranchi & Vianello, 2012; Shivers et al., 2016; Tverdik et al., 2015). For example, the work of van Lieshout et al. (1998), who found levels of parental conflict and anger among the families of children with PWS that were greater than those which existed in families caring for children diagnosed with comparable syndromes. These investigators also found evidence of a feedback loop between increases in parental anger and the frequency of challenging behaviour involving the child with PWS.

Kayadjanian et al. (2018) investigated the increased burden of care faced by families of children with PWS through the use of an online instrument developed for those supporting family members with dementia. They have speculated that challenging behaviours associated with PWS are likely to be a significant contributor to elevated levels of caregiver burden and have concluded that a critical and unmet need exists for the support of familial caregivers of people with PWS. The results of the present study suggest that there is likely to be a degree of commonality between the experiences of familial and professional caregivers. The caregivers who have contributed to this study have also reported challenges which equate to an elevated burden of care, and have identified the challenging behaviour of service users who have PWS as a significant contributing factor.

Relationship dissonance. The term relationship dissonance has been used here to describe the sense of contradiction or surprise present in many accounts of service users' challenging behaviour described by participants in Study 3. This incongruity may go some way to explaining the intensity with which challenging behaviour among people with PWS seems to impact caregivers and others. It may also explain the apparent vulnerability of people with PWS to getting into conflict with others. Relationship dissonance seems to arise when certain beliefs or expectations that caregivers hold about their clients who have PWS

are contradicted by that person's subsequent behaviour. Even experienced caregivers of people with PWS seem to be vulnerable to its effects.

There are a number of possible causal mechanisms. Many stem from the unique pattern of strengths and weaknesses that seems to be a feature of the PWS cognitive phenotype. For example, there are a number of reasons why the extent of the learning difficulties faced by some people with PWS might be either over or underestimated by caregivers. Whittington et al. (2004) have suggested that relatively poor social or functional skills among some school students with PWS may lead carers to underestimate the cognitive abilities of those young people. Conversely, Dimitropoulos et al. (2013) have recorded a discrepancy between measures of expressive language and both receptive language and functional abilities among people with one of the subtypes of PWS. They conclude that such a pattern could contribute to interpersonal conflict by leading caregivers to underestimate the degree of learning disability to which service users were subject.

Haig and Woodcock (2017) are among those who have emphasised the impact of cognitive rigidity as a result of relatively weak executive functioning in PWS. They note that in particular, task switching ability often seems to be impaired. It could be speculated that this difficulty may be especially exacerbated in the face of some social stressor, such as a minor disagreement. If this were to be the case for an individual with PWS it could lead to a marked increase in cognitive rigidity in response to a relatively minor trigger which in turn could lead to behaviour which caregivers might find surprising and incongruous. The phenomenon which James (2010) has labelled food genius might also lead to relationship dissonance. Holland's (2015) conclusion that people who have PWS may not be chronically hungry in the conventional sense, but are likely to experience increased motivation to seek food when they are confronted by the possibility of access, may also contribute. Any or all of these common consequences of PWS could lead caregivers to make errors in predicting their clients' behaviour.

For participants in the present study, the most challenging instances of relationship dissonance seemed to arise when a client's behaviour contradicted beliefs that caregivers had formed about the nature of their mutual relationship. Several described feelings of having been fooled, or even betrayed, when they or

their colleagues were faced with evidence of an apparently duplicitous incident of foraging, or witnessed an argument featuring unexpected levels of anger and stubbornness.

Challenge as both dynamic and unrelenting. Interviewees often emphasised the chronic, unrelenting nature of the challenges they faced in their work. At the same time, they also characterised many of those same challenges as dynamic and unpredictable. This apparent paradox seems to reflect the origins of foraging behaviour in PWS. Hyperphagia is genetically based; pervasive and enduring, but the PWS cognitive phenotype is also associated with certain conspicuous cognitive and psychological strengths. Participants' experience of challenge in their work also reflects the varied and unpredictable range of the circumstances under which community based caregivers provide support. Many participants gave accounts of their carefully planned supports being undermined by circumstances that they had not foreseen.

Isolation and vulnerability. Participants regarded themselves as isolated. They also found themselves accountable to, or vulnerable to criticism from, a wide range of individuals and agencies. In general, they did not expect likely critics to have a comprehensive appreciation of the support needs of people with PWS.

The counter narrative: Caregiving as a rewarding task: Despite the dominance of this narrative of challenge and response, participants were generally also clear that they enjoyed their work, and in particular that they found value and reward in their interactions with people who have PWS. Interviewees especially seemed to enjoy describing their clients' successes and to take satisfaction in the part they had played in them. It often seemed that they simply liked the people with PWS for whom they provided support, and enjoyed their company.

Authoritarianism. All respondents described implementing some measure of control over service users' access to food, and all but one reported that these measures were largely chosen and imposed with little real reference to clients' desires or wishes. They reflected the priority assigned by caregivers to the maintenance of clients' safety. The one apparent dissenting experience, that of a caregiver who described successfully implementing a food management regime based on negotiation with a man who has PWS,

seemed only to have arisen because the risks posed by access to food were perceived to be relatively lower for the service user to whom that caregiver provided support.

Creativity. Interviewees were universally aware that compulsory restrictions on service users' access to food at least gave the appearance of being at variance with the stated values of their employers and their profession. While it is true that each participant described planning and implementing authoritarian measures for the control of service users' access to food, each also sought ways to minimise these controls and to moderate or compensate for their effects. Caregivers' efforts in this regard often seemed to have demanded significant resources and the acceptance of a degree of managed risk.

Participants in the present study seemed to reject the idea that risk could ever be completely controlled. This distinction significantly changes the risk management equation for caregivers, if it is acknowledged that a degree of risk is inevitable then less is lost in tolerating a degree of that risk to achieve other gains. The support plans described by participants seemed to focus on the management of risk, rather than its elimination.

Practical wisdom. The beliefs and practices which constituted practical wisdom for participants often reflected participants' concern with the management of anxiety among service users, both as an end in itself and as a means to reduce the frequency and intensity of challenging behaviour. The establishment of an effective, consistent food management regime, under which service users would be able to accurately predict their future access to food, was prominent among the interventions listed in this context. Several participants reported the belief that taking steps to remove any potential for uncertainty around access to food was a compassionate response to the needs of their clients, as well as one that reduced risk. It seems that in general local caregivers are in agreement with the motto: "No doubt, no hope, no disappoint" (Pittsburgh Partnership, n.d.; Prader-Willi Homes of Oconomowoc, n.d.).

The outcomes sought by caregivers

Physical safety. The physical safety of service users seemed to be regarded as of primary importance throughout participants' accounts of their work. Caregivers took considerable pains, and were prepared to

compromise other values, in order to protect it. Once again, food seeking and hyperphagia were prominent in conversations concerning such risks.

The liveable life. On a semantic level at least, it might seem that this prioritisation of service users' safety among participants represents support for Holland and Wong's (1999) characterisation of the central dilemma facing caregivers as essentially a simple, dichotomous choice: clients' safety versus their autonomy. A deeper analysis reveals a more nuanced view.

Caregivers did describe prioritising safety by implementing compulsory controls over access to food, but they sometimes also described situations in which they chose to take risks, thereby compromising client safety in ways that they believed could be managed, in exchange for relatively greater returns in other areas. In addition, decisions regarding access to food for individual service users with PWS were described as being informed by a much wider range of values than the ones which Holland and Wong's (1999) analysis allowed. Interviewees often described the outcomes they sought in corporate terms, including goals that they valued for other individuals within the caregiving environment or for the group as a whole. In this sense, the desired outcomes that influenced local caregivers' decisions seem very similar to the values set summarised by van Hooren et al. (2002, 2005, 2006) as the search for a liveable life.

It seems that for at least some of those employed to operate services to people with PWS the basic unit to which support is delivered is not the individual but the group. This is an important insight for any investigator who seeks to understand decisions made within human services. It also carries implications for anyone who wishes to improve service outcomes for New Zealanders who have disabilities by way of modifying caregiver practice.

It could be speculated that the potential impact of challenging behaviour by service users who have PWS might be an important factor in determining caregivers' focus on the needs of the group rather than on those of the individual alone. During these interviews several caregivers made reference to a goal which could be paraphrased as keeping the peace. These participants generally seemed to ascribe this goal a high priority.

How Do Caregivers Make Difficult Decisions Concerning Food Security?

Caregivers follow instructions. Participants often seemed to perceive themselves as strikingly powerless, especially as individuals. All paid caregivers, and even their first level managers, are employees obliged to implement the decisions of others. Participants in Study 3 described themselves as under obligation to multiple agents, many of whom they do not expect to have a comprehensive understanding of their work.

Whose dilemma is it anyway? Viewed from this perspective, the key ethical questions associated with support to those with PWS are not primarily addressed by caregivers at all. The priorities governing their work are set by policy makers and the agencies that employ them. They are then interpreted by line managers and those who control resources. In practice, the central dilemma facing caregivers of adults with PWS is only partially within the power of those caregivers to address.

Corporate decision making. As previously noted, participants also valued consistency in decision making among their fellow caregivers' even though such consistency was generally regarded as difficult to achieve. The emphasis on corporate practice further limited the power of individual caregivers.

Caregivers balance priorities on behalf of multiple agents. Like the caregivers interviewed by van Hooren et al. (2002, 2005, 2006), those who participated in this study considered the outcomes being sought for a range of service users and others, even when they made decisions about a food management regime for an individual with PWS.

Caregivers accept a degree of risk. Like the caregivers employed within the service studied by Hawkins et al. (2011), local caregivers described sometimes departing from expected practice. Hawkins et al., described these departures as exchanges, transactions in which caregivers traded a small increase in risk to their clients and themselves in return for some other valued outcome, often an increased sense of client autonomy. However, participants in the present study seemed prepared to tolerate a more significant level of risk in general. This distinction may have been the result of a relatively greater level of resources being

available to the caregivers who featured in the work of Hawkins et al. (2011). It may also be that the needs of the service users supported by those caregivers, all of whom had PWS, were more compatible.

Caregivers regard physical safety as paramount. When participants described making decisions around access to food the protection of service users' immediate physical safety was the key imperative. These caregivers often described feeling reluctant to limit the choices of service users but reported that they were obliged to do so because of perceived risks to clients' physical safety.

Caregivers seem to employ a dialogical process. As noted, others have identified a dialogical process as one of the ways in which groups of caregivers make and review complex decisions (Abma et al., 2008). This process was also detected in the present study. Participants often told stories about their work in the course of these interviews and those stories were employed for their symbolic as well as their literal meaning. The nature and limits of this dialogical process among groups of caregivers would seem to warrant further investigation.

Conclusions

Most caregivers who participated in this study supported only one service user with PWS, and most did so in the context of work in a generic group home. Participants who had known more than one person with PWS often emphasised the differences between those individuals and hence the need for individualised needs assessment and service planning. At the same time, participants' reports showed seemingly constant awareness of the needs of a small group composed primarily of those for whom the caregiver carried responsibility. Even decisions made about the management of access to food for an individual with PWS could be expected to be influenced by the perceived needs of other members of the group. It seems that, from the point of view of paid caregivers, the caregiving task is fundamentally a corporate activity. The indivisible, quantum unit of residential support is not the individual but the group.

The results of this study characterise caregivers of adults with PWS as isolated and subject to multiple challenges. However, it also suggests that these caregivers respond to these challenges in part through the use of a dialogical process which may allow them to develop and refine resources to address

future challenges. If this is the case, it follows that stories may prove an effective way to influence caregiver culture; it may prove useful to investigate the most effective ways to participate in this dialogue. An opportunity exists to develop and test a dialogical approach to professional development among paid caregivers.

Caregivers' conceptualisation of their work as a corporate activity, coupled with this identification of a dialogical process, also adds weight to the work of those researchers who have challenged individualism as a basis for publically funded supports to people with disabilities (Johnson, 1998; Wills 2018). These findings also provide support for the work of those who have investigated the outcomes of services at the facility level; the work of Bigby and colleagues on the development of culture within group homes provides an example (Bigby et al., 2012; Bigby, Knox, Beadle-Brown, & Clement, 2015).

The present study has a number of limitations. First, it should be noted that the voices of service users themselves are largely absent and this picture of the effectiveness of services to this group remains incomplete until their views are added. The number of respondents who contributed to the present study was limited by the small, hidden and geographically diverse population from which they were drawn. This problem was exacerbated by the exclusion of caregivers who had participated in a previous study. In future studies these difficulties could be overcome by consideration being given to combining data from Aotearoa/New Zealand and Australian databases.

Study 3 would also have been strengthened if more detailed demographic data had been gathered regarding participants. This would have allowed a more comprehensive description of the sample on which the study is based. Information about culture, ethnicity, age and professional background are lacking and so the impact of these factors on participants' experience of the caregiving task cannot be considered.

The interviewer's own interest and experience in the support of people with PWS could be described as either a weakness or a conspicuous strength. The addition of research partners able to take other perspectives would strengthen future research. In particular, the provision of additional coders could allow for a moderation process to strengthen future thematic analyses. Finally, there would be value in testing and

replicating the results of the present study by repeating it with larger numbers of caregivers and by incorporating interviews with other stakeholders.

Chapter Five

The Dominant Narrative among Caregivers: Challenge and Response

The results of Studies 1 and 3 have suggested that caregivers find meaning and reward in their role and in their relationships with service users who have PWS. They also seem to find value in the expertise they have developed in their work. However, whenever participants in Studies 1 or 3 described the caregiving task itself they invoked a list of conflicting responsibilities and problems, challenges which they described as having to be resolved each day to the satisfaction of a range of authorities, many of whom were perceived as having competing priorities and an incomplete understanding of caregivers' responsibilities. This narrative of challenge and response seemed to define participants' experience of caring for a person who has PWS.

Challenge

The management of access to food has often seemed to have been assumed to be the defining challenge facing caregivers who work in support of people with PWS. However, while those who participated in these studies recognised the ethical and practical difficulties associated with this challenge, it was only occasionally identified as the one that participants found most difficult to manage. *"Food is a given"*, as one participant in Study 1 commented, another reported that managing diet was *"the easiest challenge"*.

Sometimes this problem seemed very salient indeed, for example, when participants found their efforts to establish food security hampered by legal or resource constraints. Under these circumstances, participants seemed to report that seeking a solution occupied much of their time, consumed significant resources, and generated worry well outside caregivers' hours of work. Once food security had been established, or at least once foraging had reduced to a level at which its associated risks were regarded as manageable, caregivers generally seemed to find this as a relatively concrete and straightforward problem.

Challenge may define the work of caregivers supporting people with PWS, but this challenge arises from multiple sources, of which the danger posed by access to food is only one.

Challenges were also reported to stem from other common aspects of the behaviour of service users who have PWS, such as the tendencies to cognitive inflexibility, to excess in other hedonic activities and to interpersonal conflict. Study participants also identified certain organisational factors as a common source of challenge. They reported a lack of understanding of their clients' unique needs among their colleagues and others, and also that they often found themselves isolated, stigmatised and vulnerable to criticism, even within the community of other human service workers.

The prominence given to challenges associated with aspects of their clients' behaviour has received support from the results of a recent survey of mental health needs within a relatively large cohort of people with PWS (Shriki-Tal et al., 2017). Shriki-Tal et al. reported that 89% of individuals within that group had at least one psychiatric diagnosis. Disruptive behaviour disorders were by far the most common category of diagnosis reported (68% of respondents), followed by obsessive-compulsive disorders (45%). Investigators also reported that these conditions severely affected quality of life of people with PWS themselves and of those around them, and were often the most debilitating problems faced by individuals.

Challenging behaviour in generic disability services. Many other human service workers also support clients whose behaviour presents challenges to themselves or others. Challenging behaviour among children and adults with developmental disability has often been shown to impact negatively on caregivers (Etheridge & Senior, 2017; Griffith & Hastings, 2014; Lecavalier, Leone, & Wiltz; 2006). Presumably these caregivers also experience their role as complicated by the variability of work in a community context. Caregivers working in generic services supporting people with a developmental disability have also been found to experience frustration, isolation and stigma by association with their clients (Hewitt et al., 2008). However, for participants in Studies 1 and 3 these generic challenges were complicated by consequences of the PWS behavioural phenotype such as behaviour related to the search for food, the increased tendency to interpersonal conflict, relationship dissonance and an increased sense of isolation.

Food related challenging behaviour. Participants often reported feelings of anxiety derived from their awareness of the severe consequences that could arise for service users from uncontrolled access to food. For several of them these feelings were based on bitter experience. The premature deaths of former clients with PWS were described in both Studies 1 and 3. These deaths had arisen either as the result of profound obesity or from the consequences of a single incident of hyperphagia.

Caregivers reported a number of features in service users' quest for food that seemed to intensify the anxiety experienced by support people. These included the creativity and persistence of service users' foraging behaviour, the inherent complexity and unpredictability of life in a community settings, the ubiquitous presence of food within those communities, and the misunderstandings of others regarding the risks associated with the syndrome. Each of these factors contributed to caregivers' experience of the challenges associated with their work as relentless, yet at the same time dynamic and unpredictable.

Interpersonal conflict. Several participants in Studies 1 and 3 reported that their clients with PWS had a reputation for readily becoming involved in conflicts with others. These conflicts sometimes revolved around access to food, as in the case of the young man whose conflict over the size of his burger at a local McDonald's became so intense that Police were called. However, it was also common for participants to describe conflicts that seemed to have nothing to do with food. Such incidents seem associated with other aspects of the PWS personality such as those identified in Whitman's (2013) summary of the PWS behavioural phenotype. Cognitive rigidity, social skills deficits, difficulty setting limits on other hedonic activities and the strong preference for routine all feature prominently.

Several other groups of investigators have summarised the behavioural characteristics associated with PWS by collating data gathered from relatively large samples of individuals, usually gathered through participants' contact with a specialist medical service (Jauregi et al., 2013; Miller et al., 2011; Thuilleaux et al., 2018). These researchers do not always identify an increased risk of interpersonal conflict specifically, but they do emphasise characteristics such as outbursts of anger, rigidity and social skills deficits, consistent with the reports of participants in Studies 1 and 3. They also support the conclusions of researchers who have investigated the effects on families of supporting a child or adult with PWS (Allen, 2011; Mazaheri et

al., 2013; Kayadjanian et al., 2018; Tverdik et al., 2015; van Lieshout et al., 1998) who have emphasised the relatively severe impact of challenging behaviour on other family members.

Relationship dissonance. People with PWS are not unique among users of human services in showing a strong preference for sameness or in having a reputation for becoming more stubborn and less able to listen when they come into conflict with others. However, what does seem quite specific to PWS is the frequency and intensity of these conflicts, as well as the extent to which experienced caregivers seem to describe being surprised or confused by them. This element of unpredictability or internal contradiction regarding incidents of challenging behaviour has been labelled here as relationship dissonance.

This relationship dissonance was described by caregivers in a variety of ways. Some reported feelings of surprise at the vehemence with which a service user engaged in an argument or a physical confrontation, or at the intensity of reaction that followed a triggering event which had seemed relatively minor. Others reported that their client's defence of some obviously flawed opinion seemed to conflict with caregivers' beliefs about that person's general intellectual ability. Still others found the telling of untruths contradicted beliefs which they had formed about their relationship with a person with PWS. Relationship dissonance arose whenever caregivers' beliefs about an individual with PWS, or about the relationship which they shared, appeared to be contradicted by that person's subsequent behaviour.

This element of dissonance within their relationship with service users seemed to be particularly challenging for interviewees, perhaps because it rendered stressful events such as episodes of challenging behaviour less easy to rationalise and predict. The caregiver/care recipient relationship does seem to be especially important to caregivers, it was identified in Studies 1 and 3 as a key source of reward which motivated caregivers in the face of challenge. The quality of these relationships was also identified as a source of justification for the substitute decisions on which much support depends. One supervisory manager who contributed to Study 3 reported that she had noted a common tendency among caregivers to misinterpret incidents of foraging behaviour as evidence that they have been cheated or taken in by their client. She reported that a common reaction, especially among inexperienced caregivers faced with an unexpected incident of hyperphagia, was "*He promised me*".

The work of those researchers who have examined the unique pattern of cognitive strengths and weakness associated with PWS suggests a number of ways in which such dissonance might arise. These researchers offer caregivers a potentially helpful alternative rationale (Dimitropoulos et al., 2013; Haig & Woodcock, 2017; Hinton et al., 2010; Kinash, 2007; McAllister et al., 2011; Whitman, 2013; Whittington & Holland, 2017).

Isolation. Many caregivers described themselves as isolated in their work, and as vulnerable to critique from a variety of sources. Feelings of isolation and stigma have been identified among other groups of human services workers, but those who participated in studies 1 and 3 described these feelings as complicated by the unique needs of service users with PWS and by general misunderstanding of those needs among their colleagues. Many participants in Study 3 made reference to their belief that their work was both misunderstood and often judged harshly by caregivers working in other services.

Response

Maintaining consistency. It seems likely that these experiences may lead to a sense of vulnerability among caregivers of people with PWS and so contribute to the high value placed on generating a corporate response to challenge. This was expressed by participants variously as team work, solidarity among caregivers, and as consistency of support.

The degree of planning and resource which participants described expending in pursuit of consistency among caregivers, and the time that participants spent discussing it during Studies 1 and 3, are both evidence of the value placed on it. Caregivers described carefully constructing training and orientation programmes for new colleagues, holding regular staff meetings and handover conversations between shifts, keeping written records of daily events, and sometimes spending their time off duty in phone calls or face to face conversations, all in an effort to maintain consistent practice.

Several participants in Study 1 identified the maintenance of this consistency as the greatest challenge they faced in their work. Failure to maintain consistency was blamed for a variety of negative

outcomes. The most obvious examples were breaches in food security and weight gain among clients but others included increased anxiety, emotional distress and challenging behaviours.

Gill and Fazil (2013) recorded that among the nurse/caregivers who they interviewed, the metaphor of “saboteurs” was used to describe the behaviour of new caregivers in relation to the food choices of residents who had developmental disabilities and diabetes (p.259). Similar accounts of casual or inexperienced staff members behaving inconsistently were related by participants in Studies 1 and 3. Sometimes sabotage was believed to have been undertaken knowingly, especially by caregivers employed on a temporary or casual basis for whom the fear of challenging behaviour was commonly identified as the most likely motivation. One interviewee described the inconsistent behaviour of casually employed colleagues who she believed had contributed to challenging behaviour which had subsequently arisen on her own shift. She said *“They give in ... because they don’t want the behaviours.”*

These accounts carry the sense that consistency among members of caregiver teams was regarded as so important, and was often so hard won, that any variation from the expected corporate response constituted a lack of solidarity with the group; a moral and social failing rather than a professional one alone. Inconsistency was in essence a kind of betrayal; it threatened the wellbeing of service users and created unacceptable difficulties for other caregivers.

In the context of Study 3, which featured interviews conducted individually, it is interesting to note that participants did not always report following the procedures proscribed by their manager and endorsed by their peers. These participants typically justified their actions either via reference to unique circumstances or by contending that it would be useful for the rest of the group to conform to their practice. The fact that such disclosures were much less a feature of Study 1, in which caregivers participated in groups, suggests that they may have been concerned about possible censure by other group members. It also emphasises an important difference in the way data from the two studies should be interpreted.

Caregiving as a corporate activity. The process of providing support to citizens with disability within Aotearoa/New Zealand emphasises the needs and choices of the individual at every stage. However,

it seems that for these caregivers the basic unit to which support is delivered is more likely to be the small group with whom caregivers have regular contact and for whom they carry responsibility, rather than any individual. As noted, this view is consistent with the key features of the values set which van Hooren et al. (2005) described caregivers' search for a liveable life.

This not a new finding, the results of Studies 1 and 3 seem to replicate those of van Hooren et al., (2002, 2005). However, it does carry significant implications for researchers and service planners. It supports the views of those who have framed services as social systems (Duryan et al., 2012) and those who have critiqued the dominance of individualism within them (Wills, 2018).

It is not known whether this same view exists in other groups of caregivers. In Chapter 1 the argument was made that caregivers of people with PWS seem to experience services in much the same way as service users usually do; not as individuals, but as members of a small group. It seems likely that this may be equally true of caregivers employed in generic services.

The importance of managing anxiety. Participants in Studies 1 and 3 often seemed to equate the reduction of anxiety in service users with PWS with a reduction in the risk of challenging behaviour. Given the view among caregivers that unexpected or unpredictable access to food is likely to increase anxiety among people with PWS, this has been employed as additional rationale for the imposition of measures towards food security (Holland, 2015).

Substitute decision making. All caregiver participants in this thesis expressed the willingness to make substitute decisions on behalf of the people with PWS for whom they provided support. However, they often also expressed disquiet about this issue. Some participants identified areas of their clients' lives in which they believed they were prevented from making such decisions by that client's civil rights. Members of one of the focus groups within Study 1, for example, reported the belief that they were unable to exert control over their client's private income or activities outside of their residential service. They regarded this inability as a gap in food security which increased risk to their client, but also reported that they would need consent from both he and his parents to be able to address it.

It seemed that caregivers generally made substitute decisions quite consciously, often after a process of generating and evaluating options. A number of justifications were identified as grounds in specific cases, for example, the need to prioritise service users' physical safety, the wellbeing of the group, the desire for a sense of fairness towards other residents, and the quality of the relationships established between caregivers and those in receipt of care.

The primacy of service users' physical safety. A threat to service users' physical safety was the most commonly used and overt justification for substitute decision making in both studies 1 and 3. Participants often reported that their role required them to balance a complex mix of priorities but reference to physical danger generally seemed to be regarded as a decisive justification for action, regardless of service users' consent. Even in this context substitute decisions were generally reported as having been undertaken reluctantly. The language associated with them was often the language of compulsion, as if the decision had been imposed on caregivers as it would be on care recipients: "*I had to ...*" and "*We were forced to ...*" were among the phrases used in this context.

Fairness and corporate wellbeing. Sometimes substitute decisions were justified on the basis of fairness towards other service users who shared a service with a person with PWS. This is not to say that the caregivers interviewed for Study 3 regarded themselves as judicial or paternalistic figures. A more apt analogy might be that of umpires or mediators, agents seeking corporate outcomes and obliged to make decisions in order to allow multiple parties to live well in an environment of limited resources.

Mutually valued relationships. Participants sometimes referred to either the quality of the relationships that they had established with individual service users, and/or the specialist knowledge gained in the process, as legitimising their substitute decisions, especially around access to food. The view was often expressed that service users themselves had learned from experience that participant caregivers had their best interests at heart.

Desired Outcomes: The Central Dilemma Facing Caregivers

All participants expressed awareness of the tension that existed between their twin obligations to promote both the safety of service users with PWS in relation to food and those clients' rights and autonomy. This is a problem which has been described here as the central dilemma facing caregivers of adults with PWS. Others have investigated this same dilemma, safety versus autonomy, in the wider context of generic services to people with developmental disabilities. Rouse and Finlay (2016) described it in caregivers supporting people with ID to manage diabetic risk and reported caregivers holding contradictory models of the nature of responsibility, based on caregivers' conflicting views of clients themselves. Rouse and Finlay reported that service users with diabetes were regarded as competent and personally responsible, but at the same time as "Lacking competence, dependent on others and incapable of overall accountability" (Rouse & Finlay, 2016; p.1243).

The caregivers who contributed to Studies 1 and 3 seemed to hold much less contradictory views about their clients' competence in the face of food. There was close to universal agreement that limits placed on access to food were not only necessary for safety, but were compassionate and empowering interventions which increased the choices and opportunities available to services users with PWS and to others around them. This dilemma seems to be more straightforward for caregivers who support people with PWS than for their colleagues employed in generic disability services. The problem of managing access to food for service users who have PWS may often be difficult to address in practice but the associated risks are relatively clear, concrete and immediate.

Holland (2015) has argued that while the civil rights of adults with PWS must be respected, decisions made by members of this group in relation to food are likely to be so compromised by the effects of PWS as to be considered invalid. Caregivers who have participated in Studies 1 and 3 seem quite clear that they agree with Holland's conclusion that the imposition of compulsory controls over access to food is not only justified but represents an ethical imperative in itself.

The central dilemma in the context of generic services Those interviewees who contributed to Study 3 reported that the management of clients' access to food generally constituted an ethical distinction between themselves and their colleagues who worked in generic support services. Several reported that this distinction was the source of additional challenge in their work in that it increased their sense of isolation and vulnerability to criticism. However, the work of some investigators who have studied generic services to people with disabilities suggest that this distinction is less clear cut than it might appear.

Substitute decisions have also been identified in generic services to people with disabilities (Dunn et al., 2010) and several investigators have lamented the apparent lack of progress in the promotion of self-determination within these services. Jingree (2015) has concluded that disempowering discourses are deeply entrenched within generic services and that they are often used to justify disempowering practice. Brown (2014) identified a number of ethical conflicts facing frontline caregivers and recorded confusion among those caregivers about how best to respond to them. Likewise, some caregivers participating in Study 3 described imposing substitute decisions on service users with PWS for reasons other than the reduction of foraging and hyperphagia, and at least one reported applying the dietary regime of one such client to other residents, who did not have PWS. Viewed in this context, this central dilemma facing caregivers supporting people with PWS in relation to food appears to be just one manifestation of the much larger issue of the prevalence of substitute decision making by caregivers within human services.

The concept of client autonomy within disability services. Curryer et al. (2015) have described self-determination as the fundamental right of people with developmental disabilities. They have investigated this issue in the context of people with disabilities' relationships with their families. Curryer et al. collected and critiqued the attitudes of familial carers who regarded themselves as responsible for the lives of disabled family members and found attitudes and beliefs which they described as paternalistic and based on anxiety.

Autonomy and health related choices within disability services. Other groups have taken a different view of autonomy among people with ID in relation to health related choices. Several investigators have raised this issue in the context of sounding an alarm about the burgeoning rate of obesity and diabetes

within this group (Flynn, Keywood, & Fovargue, 2003; Mann, Zhou, McDermott, & Poston, 2006; Smyth & Bell 2006; Trip, Conder, Hale, & Whitehead, 2016). Wullink et al. (2009) conducted a meta-analysis of this field. They have reported one study which linked increasing independence to greater bodyweight among adults with ID (Gabre, Martinsson, & Gahnberg, 2002). Wullink et al. described this finding as a positive result in terms of independence, although a bad one in terms of health. Like Curryer et al. (2015), they concluded that autonomy was a basic human right and proposed greater education regarding healthy choices targeted at people with ID.

Authorities such as Curryer et al. (2015) and Wullink et al. (2009) make well founded statements about the ease with which the rights and autonomy of people with developmental disabilities may be disregarded by those mandated to provide them support. However, from the point of view of professional caregivers supporting adults with PWS in Aotearoa/New Zealand, their views might seem rather vague and unhelpful; more likely to generate criticism of caregiver practices than guidance towards concrete options. Curryer et al. (2015) in particular seem to be avoiding an important issue. They neither support nor reject the proposition that circumstances might arise whereby a degree of compromise to the ethic of self-determination might be expedient. They write that people with ID, presumably including those who have PWS, should have the final say wherever possible. The difficult matters of how parents or paid caregivers should judge whether it has become *impossible*, and how they should proceed under those circumstances, are not addressed. The question of risk is not discussed at all, except as one of the concerns held by anxious family members.

Further questions about the nature of autonomy in human services. Some earlier authors, such as Smyth and Bell (2006) and Flynn et al. (2003) have offered an alternative view. They have argued that the expression of client autonomy within human services is a complex and multifaceted issue, and that caregivers typically underestimate both the degree of that complexity in practice and the full ramifications of their clients' choices. Flynn et al. (2003) have investigated caregivers' understanding of their clients' choices. They concluded that caregivers often seemed to over-generalise clients' situation-specific choices, treating them as if they were irrevocable and nonnegotiable. These authors also predicted that the increasing

prevalence of the supported living model of residential support was likely to lead to greater choice for service users, and could in turn be expected to lead to increased risks and to an increased need for tools and guidelines to provide caregivers and service users with guidance more sophisticated than that which Flynn et al. (2003) have labelled the “elusive mantra of choice” (p. 30).

In a local context, Whitehead, Trip, Hale, and Conder (2016) have investigated the way in which caregivers and care recipients with developmental disability (but without PWS) manage the implications of long-term illness, specifically diabetes. They have described a process of negotiated autonomy, which they regard as the product of the relationship between caregiver and care recipient. This negotiated autonomy addresses some of the concerns raised by Flynn et al. (2003) in that it generates outcomes that are at least flexible over time.

The conclusions of Whitehead et al. (2016) seem to have much in common with the responses of participants in Studies 1 and 3. Both present client autonomy as one important driver in a continuous process of negotiation that takes place within a context of measured risks and the prioritisation of client safety. By framing the problem in these way caregivers supporting people with PWS are able to continue to regard it as their ethical and professional responsibility to find ways to promote the autonomy of those they support, while also taking steps to ensure their safety.

Risjord (2014) identified a similar dilemma within the context of the nursing studies literature, and proposed a resolution which she termed relational autonomy. Risjord drew on notions of positive and negative freedom, sometimes summarised as the freedom to and the freedom from, and argued that nurses should include both in their concept of patient autonomy. For Whitehead et al. (2016) and for Risjord service users are able to contribute to decisions about their own autonomy. This capacity may be increased by training in decision making and by systems that allow the voices of service users to be heard, but the decision itself remains a product of the caregiver/recipient relationship over time, and not simply the result of a single choice made by an individual.

The relevance of this debate to services for people with PWS. The problem which has been labelled here as the central dilemma facing caregivers who support people with PWS is not unique to this group of carers, it is part of a larger debate within human services concerning the nature of autonomy and responsibility within the caregiver/recipient relationship. Smyth and Bell (2006) and Flynn et al. (2003) have shown that the complexity of clients' choices and the risks associated with them are sometimes underestimated by caregivers. Investigators such as Whitehead et al. (2016) and Risjord (2014) suggest that, for those in need of care, the concept of self-determination is not a simple matter of one person's choice but one which emerges from negotiation in the context of a relationship.

The ideas of relational (Risjord, 2014) or negotiated autonomy (Whitehead et al., 2016) seems very similar to that found by van Hooren et al. (2002, 2005, 2006) in their interviews of caregivers supporting people with PWS, and in the responses of New Zealand caregivers who contributed to these studies. All of these groups of caregivers described striving to maintain the autonomy and promote the choices of those to whom they provided support, even at the same time as they limited some choices to prioritise safety. All described the resultant support regime as a kind of dynamic compromise, facilitated and legitimised by the quality of the relationship within which it occurred.

The Ethics of Controlling Access to Food

The case for the support of adults with PWS in relation to food as an ethical special case has been adequately made. The exercise of compulsory control over access to calories should be regarded as a valid and important part of caregivers' role, and as essential to service users' safety and wellbeing. The caregivers whose views are recorded in studies 1 and 3 of this thesis regarded the net effect of such controls as both compassionate and empowering. Their imposition does not seem inconsistent with contemporary standards in other human services but equates to the approach taken by those who support the members of similarly vulnerable groups (Buckles, 2016; Risjord, 2014; Tsungu, 2014; Whitehead et al., 2016).

The pattern of differences in hyperphagic behaviour among individuals who have PWS has yet to be described in detail, and so the question of whether or not some individuals may be able to manage access to

food without support is difficult to address. It cannot be known whether a proportion of people with PWS might face such mild difficulties with food that compulsory controls cannot be justified, although it seems likely that many caregivers would argue that this question might best be answered in the context of their care; in the meantime it seems the only safe option is to establish food security.

The Central Dilemma Reframed

It also seems that for the caregivers who participated in Studies 1 and 3, safety versus autonomy is not just an inadequate model for the central dilemma which faces them in their work, it is fundamentally flawed and participants have rejected it on a number of grounds. First, caregivers do not regard either safety or autonomy as entirely achievable in practice; the management of risk is an inevitable part of the caregiving task. Local caregivers also agree with the conclusions of van Hooren et al. (2005), in that they view their work and the goals they seek through it in corporate rather than individualistic terms. Decisions which affect the safety or autonomy of a service user who has PWS are likely to be considered in an environment in which a much wider range of valued outcomes, for a much wider range of people, are also being pursued.

Finally, caregivers often seemed to regard the promotion of service users' safety and their autonomy as complementary goals, rather than contradictory ones. They described the imposition of food security as a compassionate response, not only because it protected their clients' health and safety, but also because control exerted over access to food reduced the uncertainty to which their clients were exposed, and hence their vulnerability to negative emotions and challenging behaviour. By the same rationale, participants regarded the establishment of food security as empowering for their clients with PWS. Safety *and* autonomy might be a more apt metaphor from their perspective, or even autonomy *through* safety.

How Do Caregivers Make Difficult Decisions?

Many of the caregivers who contributed to these studies told stories to illustrate points about their work, especially to describe the reasons why certain decisions had been made and to present a justification for these choices on the basis of the outcomes achieved. By contrast, no participant referred to any set of

written guidelines in these contexts; no code of ethics, no summary of law, no moral or religious text, no philosophical or political declaration, nor any statement of agency policy.

As has already been noted, many of the policy and practice decisions which paid caregivers implement are in fact chosen by others, but when they are faced with a difficult ethical or practical decision it seems caregivers draw primarily on a resource made up of their own experience and that of others, often expressed and summarised in the stories they tell about their work.

Making use of the dialogical process. Gill and Fazil (2013) have suggested that it is important to make space for caregivers to engage in ethical conversations. In particular they recommend that employers should provide forums for caregivers to tell and respond to stories about their work. It also seems likely that an understanding of this dialogical process may offer opportunities to influence caregiver attitudes and practices. It may be useful for researchers to initiate a dialogue of their own by preparing stories and planning responses to the narratives of others. Such an approach might eventually prove useful in the context of traditional professional development events such as training seminars and conference workshops. However, the dialogical process seems to arise naturally among groups of caregivers. It might also be useful for researchers to examine the effects of facilitating or simply joining in the conversations that already take place among caregivers and others. This dialogical process might also lend itself to adaptation for social media.

The dialogical process depends on the symbolic value of stories. It seems reasonable to assume that it can only take place effectively where those participating in the conversation have access to the necessary background of shared experience or culture to allow them to decode the stories' meanings. Any researcher or service manager seeking to adapt the dialogical process as a tool for investigation or service development would first need to have a practical understanding of the caregiving task. It should also be noted that many people who themselves have PWS have proven to be skilled and effective in telling their own stories (Stanley, 2013). The narrative format may prove an effective way to include the voices of people with PWS in caregiver training and in future investigations.

The State of Services to Adults with PWS in Aotearoa/New Zealand

Care must be taken in drawing conclusions based on the present work alone. Study 2, although the most relevant to the question of the state of local services, generated limited data and employed a small and self-selected sample. However, even this limited information is sufficient to give grounds for concern, both about the effectiveness of supports provided to people with PWS within Aotearoa/New Zealand and about the welfare of those service users.

Those with PWS who participated in Study 2 generally recorded high measures of BMI, higher even than the levels recorded by Dykens et al. (2007) among their North American counterparts. Caregiver participants in Studies 1 and 3 did refer to the support of some individuals with PWS who had a lower BMI but participants in both studies also described the premature deaths of young adult service users as the result of uncontrolled access to food.

It does seem that there are grounds for concern regarding the efficacy of the supports provided to many New Zealanders who have PWS in relation to food. Throughout all three studies, efforts to limit access to food were commonly described. Caregivers reported a willingness to impose restrictions using ecological interventions and supervision, all service users seemed to be subject to at least some such controls. This suggests the pattern of service outcomes referred to in Chapter 1 as “The worst of both worlds”, whereby the civil rights of citizens who have PWS are indeed being compromised in order to protect their safety but the measures undertaken seem not to be proving reliably effective; a response to the central dilemma which could be characterised as neither safety nor autonomy.

Evidence based best practice for the support of a person who has PWS requires the individualised assessment of need, leading to the provision of a calorie controlled diet supported by ecological controls which limit access to extra food. Exceptional individuals may exist who can manage their diet safely without support but the characteristics of this group, if it exists, have yet to be defined. At present caregiver experience, garnered over time, seems to be the most important source of information on which the assessment of individual need in relation to food can be based. Until that experience is developed regarding

each individual service user it seems that the only safe option for caregivers is to accept the complex, ongoing challenge of establishing and maintaining food security.

Participants in this thesis only rarely referred to the needs assessment and service planning processes by which residents with PWS had entered their present residential support service. Services to individuals often seemed to have been developed largely through trial and error. Only rare and fleeting references were made to the assessments of local Needs Assessment and Service Coordination (NASC) services, even though professional needs assessors and service coordinators employed by one of these NASCs can be assumed to have completed individual processes for every person with PWS who is in receipt of services. Instead, service managers and caregivers reported seeking out their own information about PWS, either through PWSA(NZ) publications, through personal contacts, or generically online. In the absence of any summary of evidence based practice regarding the support of people with PWS, most reported that the greater part of the learning that they had completed about the syndrome, and about their individual client's needs, had been undertaken on the job.

Stories of staff turnover and of service users moving between residential homes were common. It seems reasonable to speculate that the rate of service breakdown may be high, and the results of Study 2 suggest that a significant proportion of the adults with PWS remain living with family members well into adulthood, even though state funded residential services are available.

It is also true that many participants in Studies 1, 2 and 3 also contributed stories of conspicuous success. Some described weight loss among their clients and others reported a BMI that at least seemed to have been stable over time. This may suggest a more comprehensive level of food security in the services in which those caregivers worked, or it may be a reflection of variations in the intensity and nature of the drive to seek food. It may also reflect the findings of Miller et al. (2011) who have reported that a proportion of people with PWS seem to experience a reduction in the drive to seek food during adulthood. Participants often also reported success for those they supported in areas apart from their bodyweight. Many individuals with PWS seem to enjoy rich relationships, maintain a good quality of life and to have achieved a wide variety of goals.

These successes notwithstanding, the responses of participants in Studies 1 and 3 seem to describe a service system in which relies heavily on the commitment and creativity of direct caregivers and their supervisors, rather than on any summary of evidence based practice to ensure service quality. The reports of caregivers suggest that this system is also characterised by chronically high levels of stress and a culture of trial and error. Key decisions are often left to teams of direct caregivers or their managers in an environment of few guidelines, little research and muddled ethics. As Gill and Fazil (2013) noted, caregivers are not in a position to wait for lawmakers, policy analysts and ethicists to catch up, they must address complex issues every day, using whatever information and resources they have available.

Limitations

The three studies which comprise this project are each subject to a number of weaknesses which limit the conclusions that can be drawn from them. First, all three are based on quite small samples of either adults who have PWS or their caregivers. Finding and recruiting eligible participants proved a challenge in each case. This was due to the relatively small and widespread nature of the population, exacerbated by the fragmented nature of service provision, the logistics of conducting interviews with shift workers in diverse locations, the decision to preclude caregivers who had participated in Study 1 from Study 3, and the decision to limit the role of service provision agencies as employers in identifying potential participants.

With the benefit of hindsight it seems that approaching these agencies in the first instance may have been a more effective way to identify people with PWS who were in receipt of services. The strategy of providing letters of invitation for these agencies to pass on to potential participants, instead of seeking this help through local NASC agencies, would probably have provided greater coverage and would have preserved recipients' anonymity equally well, without necessarily placing undue pressure on people with PWS or their caregivers to consent to participate.

Study 2 was also hampered by the lack of instruments for the measurement of key variables, and by the fact that some of those which were selected proved difficult for participants to use. Replication of this study in an online format would likely yield improvements in both response rate and accuracy. The trialling

of measures of FQoL and/or QoL might also be useful. These measures should be used in preference to the SF-36 instrument which participants seemed to find so difficult to complete.

It has already been noted that my status as former caregiver to people with PWS could be viewed as both a strength and weakness of these studies. Outcomes would have been strengthened by the provision of a wider panel of co-researchers at various points, it would have allowed for independent moderation of the coding process during thematic analysis for example. The voices of people with PWS themselves are present in a limited way in Study 2 but the project would have been strengthened by the participation of co-researchers who have PWS and/or by the addition of an advisory panel of service users.

It should also be noted that this research does not address PWS within a Maori context. No data regarding participant caregivers' ethnicity or cultural identity was gathered and questions of tikanga and kaupapa Maori in relation to either food or the provision of care were not raised. Any future replication might address these weaknesses by partnering with Maori co-researchers, by modifying interview questions and making research materials available in Te reo Maori. However, it may be more useful to consider a separate research project in the first instance, one aimed at capturing the experiences of adults with PWS, their families and their caregivers, who are themselves Maori; the goal being to describe the syndrome itself from the perspective of Maori and in a kaupapa Maori context.

Directions for Future Research

The Relevance of Research into PWS to Mainstream Services

In 2015, a multi-disciplinary workshop was convened to review the state of knowledge regarding mental health and maladaptive behaviour among people with PWS, and to set directions for future research. This meeting was called The PWS Research Strategy Workshop, its members included prominent academics, healthcare professionals and family members. The minutes of this workshop (Schwartz et al., 2016) record Dykens' view that research into the effects of PWS has untapped allegorical value relevant to the study of a number of issues of pressing concern outside the PWS community. Dykens' comment summarises those of other authors, such as Gill and Fazil (2013) and Solvoll et al. (2015), who have

identified a range of ethical and practical problems within human services that seem analogous to those facing adults with PWS. For example, obesity and rates of diabetes have been raised as pressing issues for many of those in receipt of generic disability services (Rouse & Finlay, 2016). The support of service users with various forms of challenging behaviour has raised familiar questions of rights, autonomy and the management of risk (Buckles, 2016; Gooding, Arstein-Kerslake, Andrews, & McSherry, 2016; McSherry, Murphy, Turkmendag, & Roberts, 2017).

People who have PWS represent a relatively easily defined group, many of whose needs are well known. Researchers in several fields have found that the study of those who have PWS has presented opportunities to simplify complex problems. It seems likely that those interested in advancing the ethics and practice of services to people with developmental disabilities, especially to those whose behaviour is labelled as challenging, might find similar opportunities in further studies of services to New Zealanders with PWS.

Challenges Facing Researchers

The lack of instruments to measure key variables. This opportunity notwithstanding, services to people with PWS remain poorly understood and a number of factors hamper attempts to address that gap. In particular, research is limited by the lack of tools for the measurement or description of key factors. The NZPSQ, trialled here in Study 2, seems to represent the first attempt to summarise ecological measures instituted by caregivers towards food security. Likewise, we know that the nature and intensity of the drive to seek food varies among people who have the syndrome, but our tools for describing these differences are largely limited to the HQ.

Although useful instruments exist for the measurement of service outcomes beyond BMI and diabetic status, these have seldom been trialled by researchers. Caregivers who were interviewed for Studies 1 and 3 emphasised the importance of individualised service delivery based on the assessment of individual need. Recently developed measures of QoL and FQoL (Boelsma et al., 2017) would seem to be potentially useful in this context. It should be noted that measures of generalised wellbeing have already been used to

evaluate the effects of medical interventions, particularly the usefulness of human growth hormone replacement, in people with PWS (Bertella et al., 2007).

Difficulty in recruiting research participants. PWS is a rare condition and the population of Aotearoa/New Zealand is relatively small and widespread. Identifying and recruiting sufficient potential study participants is likely to continue to be a particular challenge to future studies.

Estimates of the prevalence of PWS have varied quite widely (Lionti, Reid, White, & Rowell, 2015; Whittington & Holland, 2004), but consensus seems to have settled on a rate of about 1:15,000. This is a figure which is close to the 1:15,830 estimated by Lionti et al. (2015) from their review of births in the Australian state of Victoria, and one which suggests that there should be considerably more people with PWS in Aotearoa/New Zealand than are known to PWSA (NZ). Lionti et al. have also concluded that greater awareness of PWS and modern diagnostic techniques have allowed for earlier and more accurate diagnosis. PWSA (NZ) report that testing for PWS is now routinely undertaken for all hypotonic neonates; however, it may still be that some people with PWS are hidden within the population of adult New Zealanders with ID.

A Way Forward

Overcoming the problem of small sample size. Certain resources are available to researchers interested in the study of people with PWS. In particular, a network of productive relationships among the international research community has been fostered through national PWS associations under the IPWSO banner. At the same time, record keeping practices have been improved and new databases established in several jurisdictions (Butler et al., 2018). It seems likely that international cooperation could allow more researchers to combine databases and so reduce the problem of small sample size.

Cooperation among researchers might also facilitate the development of a new literature of single case studies in which adults with PWS and their caregivers might be invited to tell their own stories. The usefulness of this literature would be enhanced if investigators were first able to agree on a set of

conventions for each case study in order to facilitate meta-analysis, these might include a standard format and a pool of compatible measures.

Analysis of the resultant literature over time could circumvent the problem of small sample size and provide a way to investigate research questions which are difficult to address at present, the pattern of variations in the nature and severity of difficulties with food among adults with PWS for example. Individual case studies might also provide base-line data, allowing a useful longitudinal element to be introduced into the new literature by caregivers and people with PWS revisiting and revising their personal narratives following key life events, such as the move to a residential support service which featured a different food environment. Such a literature might provide insight into the key questions of the effects of food security on factors beyond bodyweight and diabetic status. It would also provide opportunities for the empowerment of people with PWS as co-researchers, and it would lend itself to engagement of families and/or professional caregivers, either as facilitators alongside the people to whom they provide support or as contributors to their own, parallel set of narratives.

The development of a wider range of measures.

HQ and NZPSQ. It seems that the HQ yields a useful description of food seeking behaviour in individuals with PWS, recent work by Key and Dykens' (2018) on the measurement of eye tracking in people with PWS may also offer promise in this regard. The usefulness of any such instrument could be enhanced if it were to be accompanied by a standardised description of food ecology. The NZPSQ for example, includes indications of the presence or absence of a number of common interventions concerning access to food. The inclusion of these instruments in future population surveys, or in a literature of single case studies, would allow the limits and distribution of food seeking behaviour among people with PWS to be defined.

Measures of service outcomes; QoL and FQoL. The development of a more comprehensive range of outcome measures, beyond measures of bodyweight and diabetic status, would allow better evaluation of existing services and facilitate investigation into the key relationships between service user characteristics,

interventions implemented by caregivers, and service outcomes. The use of measures of QoL and FQoL seem promising in this role. Schalock et al. (2002) has described QoL as the state in which basic needs are met and certain life enhancers such as choice, control, personal development and social inclusion are also present. A number of instruments exist for the measurement of QoL and there is an established tradition of their use to evaluate the outcomes of services to people with disabilities (Samuel, Rillotta, & Brown, 2012; Schalock, Gardner, & Bradley, 2007).

The role of empirical data in ethics scholarship has been the subject of debate in recent years (Gorea, 2015; Stretch 2010). However, in the field of support to people with PWS the development of measures of QoL and FQoL seem to offer opportunities to reanimate discussion about longstanding ethical problems. Data alone may not be sufficient to decide normative questions (Gorea, 2015), but these instruments allow the effects of multiple, inter-related factors to be summarised in a single statistic, one that is relevant to the service user and meaningful to advocates and service planners. Measures of QoL and FQoL may not be able to tell caregivers whether they should choose to promote their clients' safety over autonomy, but they can help researchers to investigate the effects of interventions employed to promote either ethic, and hence demonstrate what each of these choices is likely to mean in practice.

Families are embedded social systems, as are residential support services. Samuel et al. (2012) have traced the development of the concept of FQoL to the growing appreciation of the importance of families in human services and to the application of QoL concepts at a systemic level. They also note that measures of FQoL have certain advantages over QoL in addressing ethical problems. They point out that because choice and self-determination are important components in the construction of QoL, an ethical dilemma arises whenever clients' choices are at variance with caregivers' judgement or the policies of service agencies. However, the concept of FQoL includes the assumption that all parties deserve lives of quality. The pattern of relationships between members is also considered important and it is acknowledged that members will often seek solutions through compromise.

Like van Hooren et al.'s (2002, 2005, 2006) description of caregivers' search for a liveable life, QoL and FQoL are ideas which subvert the view that ethical decisions should be regarded as binary. In addition,

FQoL implies that caregivers should not be regarded as bearing sole responsibility for the outcomes of the care they provide. FQoL is based on a view of family members as creative agents within a system. It has been described as strengths based, and as shifting the focus of caregivers' support "from fixing to supporting, from deficits to strengths and from child to family" (Samuel et al., 2012, p. 3).

There are similarities between Samuel et al.'s view of relationships within families and those of participants in Studies 1 and 3 concerning their relationships with service users who have PWS. Paid caregivers also seem to describe supporting rather than fixing their clients and they conceptualise their work in corporate rather than individual terms. As has been noted, this is consistent with overseas findings regarding support to people with PWS (van Hooren et al., 2005). The development of the concept of FQoL might allow its application beyond the context of literal families. The investigation of corporate QoL within other, similar social systems seems a promising avenue for researchers who seek to progress ethical debate within social services. In the meantime, consideration should be given to the application of existing measures of both QoL and FQoL in studies evaluating the outcomes associated with the establishment of food security within services to people with PWS.

Priorities for Future Research

The published proceedings of PWS Research Strategy Workshop (Schwartz et al., 2016) included summaries of several topical presentations emphasising the challenges associated with the PWS behavioural phenotype. Workshop members also suggested a list of 10 research priorities to advance understanding of mental health concerns and maladaptive behaviour among people with PWS. The top two priorities listed by this group were the need for longitudinal studies and the development of tools to allow the measurement of key variables. Both of these needs have also been identified in the present study.

Another of the group's recommendation was the engagement of familial and professional caregivers in research. Schwartz et al. (2016) noted that, "Workshop attendees felt that their vast experience and engagement will be critical to advancing knowledge and treatment" (p. 136). This acknowledgement notwithstanding, it seems that the workshop's position represents something of a missed opportunity. It is

important to understand the perspective of caregivers of people with PWS, not just because of the body of knowledge that this group holds, but because of their key role as mediators of service outcomes.

This thesis has sought to investigate PWS by examining contemporary support services and the experiences of professional caregivers. From this, subtly different, perspective a number of additional priorities for future research might be suggested, for example the definition of the limits and distribution of difficulties with food among individuals with PWS, and the development of a standardised way to describe the food ecology of a given environment, including a record of interventions used to establish food security and the opportunities for access to calories which remain. Investigation into the effect of goal-oriented social activities on eating behaviour in PWS also seems promising, as suggested by the work of Bennaroch et al. (2012) and Singh et al. (2011).

A need also exists for a description of the nature and consequences of PWS from a specifically Maori perspective. Such a description might contribute to the development of tools and supports suitable for use on marae and in other environments in which kaupapa Maori is taken seriously. It seems likely that work to this end would prove a useful precedent for others who live or work within services to people with disabilities.

Other research priorities identified in this thesis that may also generate benefits well beyond the PWS community include the establishment of a literature of single case studies to capture the responses of individuals to changing food environments, the exploration of the dialogical process of ethical development among caregivers as a resource for professional development and problem solving, and studies trialling the use of measures of QoL and FQoL as evaluations of service outcomes. Finally, there is a need to continue to explore ways in which the voices of people with PWS themselves can be better represented in research priorities and practice, and that both they and those who support them may participate as co-researchers in the resulting investigations.

References

- Abma, T. A., Widdershoven, G. A. M., Frederiks, B. J. M., van Hooren, R. H., van Wijmen, F. & Curfs, P. L. M. G. (2008). Dialogical nursing ethics: The quality of freedom restrictions. *Nursing Ethics*, 15(6), 789-802.
- Akindola, A., Kentish, J., & McColl, E. (2015). *Prader-Willi Syndrome and the national insurance scheme: A report prepared by the University of Melbourne on behalf of the PWS Better Living Foundation*. Melbourne, Vic.: University of Melbourne.
- Allen, K. (2011). Managing Prader-Willi Syndrome in families: An embodied exploration. *Social Science & Medicine*, 72(4), 460-468.
- Altman, K., Bondy, A., & Hirsch, G. (1978). Behavioural treatment of obesity in patients with Prader-Willi Syndrome. *Journal of Behavioural Medicine*, 1(4), 403-412.
- Banks, S. (2016). 'Knowing me, knowing you': Disability support worker as emotional mediator? *Sexualities*, 19(5-6), 659-676.
- Banzett, L. K., Marshall, B. D., Bowen Jr, L., Glynn, S. M., & Mosk, M. M. (1991). Weight loss in the Prader-Willi Syndrome: Treatment and long-term follow-up. *Journal of Developmental and Physical Disabilities*, 3(1), 47-57.
- Barbour, R. (2018). *Doing focus groups* (Vol. 4). Los Angeles, CA: Sage.
- Benarroch, F., Pollak, Y., Wertman, M., Hirsch, H. J., Genstil, L., & Gross-Tsur, V. (2012). Individuals with hyperphagia can voluntarily fast: Experience from Prader-Willi Syndrome. *Health*, 4(12A), 1551-1555.

- Bertella, L., Mori, I., Grugni, G., Pignatti, R., Ceriani, F., Molinari, E.,...& Semenza, C. (2007). Quality of life and psychological well-being in GH-treated, adult PWS patients: A longitudinal study. *Journal of Intellectual Disability Research*, 51(4), 302-311.
- Bigby, C., & Beadle-Brown, J. (2016). Culture in better group homes for people with intellectual disability at severe levels. *Intellectual and Developmental Disabilities*, 54(5), 316-331.
- Bigby, C., Knox, M., Beadle-Brown, J., & Clement, T. (2015). 'We just call them people': Positive regard as a dimension of culture in group homes for people with severe intellectual disability. *Journal of Applied Research in Intellectual Disabilities*, 28(4), 283-295.
- Bigby, C., Knox, M., Beadle-Brown, J., Clement, T. & Mansell, T. (2012). Uncovering dimensions of culture in underperforming group homes for people with severe intellectual disability. *Journal of Intellectual and Developmental Disabilities*, 50(6), 452-467.
- Boelsma, F., Caubo-Damen, I., Schippers, A., Dane, M., & Abma, T. A. (2017). Rethinking FQoL: The dynamic interplay between individual and family quality of life. *Journal of Policy and Practice in Intellectual Disabilities*, 14(1), 31-38.
- Boyall, S. W. (2013). Diagnosis and management of Prader-Willi Syndrome: Sandie Walker Boyall offers a personal account of her experiences supporting people diagnosed with a genetic disorder that can manifest behaviours that challenge. *Learning Disability Practice*, 16(5), 26-28.
- Braun, V., & Clarke, V. (2006) Using thematic analysis in psychology. *Qualitative Research in Psychology*, 3(2), 77-101.
- Braun, V., & Clarke, V. (2013). *Successful qualitative research: A practical guide for beginners*. London, Sage.
- Braun V., Clarke V., Hayfield N., Terry G. (2019) Thematic Analysis. In: Liamputtong P. (Ed). *Handbook of Research Methods in Health Social Sciences*, (pp. 843-860) Singapore, Springer.
- Broadie, S. (1991). *Ethics with Aristotle*. Oxford, UK: Oxford University Press.

- Brown, P. H. (2014). *People with learning disabilities and the interpersonal construction of self-determination* (Doctoral dissertation, University of Leeds, UK). Retrieved from <http://etheses.whiterose.ac.uk/8055/1/Phillipa%20H%20Brown%20Submission%20July.pdf>
- Buckles, J. (2016). *"We walk that boundary. It's a tightrope": A grounded theory approach to the experiences of direct support professionals who work with people with intellectual disability and possibly sexualized challenging behavior* (Doctoral dissertation, University of New Mexico, Albuquerque, USA). Retrieved from https://digitalrepository.unm.edu/cgi/viewcontent.cgi?article=1022&context=educ_spcd_etds
- Buiting, K. & Horsthemke, B. (2006). Molecular-genetic findings in Prader-Willi Syndrome. In M. G. Butler, P. D. K. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (3rd Ed.), pp. 58-73. New York, NY: Springer.
- Butler, M. G., Hanchett, J. M. & Thompson, T. (2006). Clinical findings and natural history of Prader-Willi Syndrome. In M. G. Butler, P. D. K. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (3rd Ed.) pp. 3-48. New York, NY: Springer.
- Butler, M. G., Kimonis, V., Dykens, E., Gold, J. A., Miller, J., Tamura, R., & Driscoll, D. J. (2018). Prader-Willi Syndrome and early-onset morbid obesity NIH rare disease consortium: A review of natural history study. *American Journal of Medical Genetics, Part A*, 176(2), 368-375.
- Butler, M., Lee, P. D., & Whitman, B. Y. (Eds.). (2006). *Management of Prader-Willi Syndrome*. New York, NY: Springer.
- Butler, M. G., Manzardo, A. M., Heinemann, J., Loker, C., & Loker, J. (2017). Causes of death in Prader-Willi Syndrome: Prader-Willi Syndrome Association (USA) 40-year mortality survey. *Genetics in Medicine*, 19(6), 635.
- Cassidy, S. B., & Driscoll, D. J. (2009). Prader-Willi Syndrome. *European Journal of Human Genetics*, 17(1), 3-13.

- Clarke, D. J., Boer, H., Chung, M. C., Sturmey, P., & Webb, T. (1996). Maladaptive behaviour in Prader-Willi syndrome in adult life. *Journal of Intellectual Disability Research*, 40(2), 159-165.
- Clarke, V., & Braun, V. (2013) *Successful qualitative research: A practical guide for beginners*. London, UK: Sage.
- Convention Coalition Monitoring Group. (2012). *Disability rights in Aotearoa New Zealand: A report on the human rights of disabled people in Aotearoa New Zealand (Article 33)*. Wellington, NZ.
- Retrieved from:
<http://www.dpa.org.nz/store/doc/Disability%20Rights%20in%20Aotearoa%20NZ%202012%2012pt>.
- Curryer, B., Stancliffe, R. J., & Dew, A. (2015). Self-determination: Adults with intellectual disability and their family. *Journal of Intellectual and Developmental Disability*, 40(4), 394-399.
- Danglas, P., Reidy, M., Korner, P., & Alagarsamy, S. (2017). *U.S. Patent Application No. 15/510,781*.
- Davis, K., Drey, N., & Gould, D. (2009). What are scoping studies? A review of the nursing literature. *International journal of nursing studies*, 46(10), 1386-1400.
- DiCicco-Bloom, B., & Crabtree, B. F. (2006). The qualitative research interview. *Medical education*, 40(4), 314-321.
- Dimitropoulos, A., Ferranti, A., & Lemler, M. (2013). Expressive and receptive language in Prader-Willi Syndrome: Report on genetic subtype differences. *Journal of Communication Disorders*, 46(2), 193-201.
- Dozier, C. L., Dracobly, J. D., & Payne, S. W. (2013). Unique considerations of Prader-Willi Syndrome. In D. Reed, F. Reed & J. Luiselli (Ed.s) *Handbook of Crisis Intervention and Developmental Disabilities*. pp. 183-198. New York, NY: Springer Science and Business Media.
- Driscoll, D. J., Miller, J. L., Schwartz, S., & Cassidy, S. (2014). Prader-Willi Syndrome. In R. A. Pagon, M. P. Adam, H. H. Ardinger, et al., (Eds.), *GeneReviews* (Internet). Seattle, WA: University of Washington.
- Duggleby, W. (2005). What about focus group interaction data?. *Qualitative health research*, 15(6), 832-840.
- Dunn, M. C., Clare, I. C. H., & Holland, A. J. (2010). Living 'a life like ours': Support workers'

- accounts of substitute decision-making in residential care homes for adults with intellectual disabilities. *Journal of Intellectual Disability Research*, 54(2), 144-160.
- Duryan, M., Nikolik, D., van Merode, G., & Curfs, L. (2012). System dynamics modeling for intellectual disability services: A case study. *Journal of Policy and Practice in Intellectual Disabilities*, 9(2), 112-119.
- Dykens, E. M., Goff, B. J., Hodapp, R. M., Davis, L., Devanzo, P., Moss, F., ... & King, B. (1997). Eating themselves to death: Have “personal rights” gone too far in treating people with Prader-Willi Syndrome? *Mental Retardation*, 35(4), 312-314.
- Dykens, E. M., Lee, E., & Roof, E. (2011). Prader-Willi Syndrome and autism spectrum disorders: An evolving story. *Journal of Neurodevelopmental Disorders*, 3(3), 225.
- Dykens, E. M., Maxwell, M. A., Pantino, E., Kossler, R., & Roof, E. (2007). Assessment of hyperphagia in Prader-Willi Syndrome. *Obesity*, 15(7), 1816-1826.
- Dykens, E. M., Roof, E., Hunt-Hawkins, H., Dankner, N., Lee, E. B., Shivers, C. M., ... & Kim, S. J. (2017). Diagnoses and characteristics of autism spectrum disorders in children with Prader-Willi Syndrome. *Journal of Neurodevelopmental Disorders*, 9(1), 18.
- Dykens, E. M., & Rosner, B. A. (1999). Refining behavioural phenotypes: Personality-motivation in Williams and Prader-Willi Syndromes. *American Journal on Mental Retardation*, 104(2), 158-169.
- Dykens, E., & Shah, B. (2003). Psychiatric disorders in Prader-Willi Syndrome. *CNS Drugs*, 17(3), 167-178.
- Eiholzer, U., & Lee, P. D. K. (2006). Medical considerations in Prader-Willi Syndrome. In M. G. Butler, P. D. K. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (3rd Ed.), pp. 97-152. New York, NY: Springer.
- Einfeld, S. L., Kavanagh, S. J., Smith, A., Evans, E. J., Tonge, B. J., & Taffe, J. (2006). Mortality in Prader-Willi Syndrome. *American Journal of Mental Retardation*, 111(30), 193-198.
- Einfeld, S. L., Smith, E., McGregor, I. S., Steinbeck, K., Taffe, J., Rice, L. J., ... & Guastella, A. J. (2014). A double-blind randomized controlled trial of oxytocin nasal spray in Prader-Will Syndrome. *American Journal of Medical Genetics, Part A*, 164(9), 2232-2239.

- Ejaz, F. K., Bukach, A. M., Dawson, N., Gitter, R., & Judge, K. S. (2015). Examining direct service worker turnover in three long-term care industries in Ohio. *Journal of Aging & Social Policy*, 27(2), 139-155.
- Etheridge, L., & Senior, H. (2017). 'It's a matter of your personality more than anything else': The experiences of seasonal workers regarding challenging behaviour in children. *Journal of Intellectual Disabilities*, 21(1), 40-52.
- Evans, I. M. (1999). Staff development, caring, and community. In J. R. Scotti & L. H. Meyer (Eds.), *Behavioural intervention: Principles, models, and practices*, pp. 413-431. Baltimore, MD: Paul H. Brookes.
- Farnsworth, J., & Boon, B. (2010). Analysing group dynamics within the focus group. *Qualitative Research*, 10(5), 605-624.
- Farooqi, I. S., & O'Rahilly, S. (2014). Genetic obesity syndromes. In S. Grant (Ed.) *The Genetics of Obesity* (pp. 23-32). New York, NY: Springer.
- Fehnel, S., Brown, T. M., Nelson, L., Chen, A., Kim, D. D., Roof, E., & Dykens, E. M. (2015). Development of the Hyperphagia Questionnaire for use in Prader-Willi Syndrome clinical trials. *Race/Ethnicity*, 6, 100-0.
- Fieldstone, A., Zipf, W. B., Sarter, M. F., & Berntson, G. G. (1998). Food intake in Prader-Willi Syndrome and controls with obesity after administration of a benzodiazepine receptor agonist. *Obesity Research*, 6(1), 29-33.
- Flynn, M., Keywood, K., & Fovargue, S. (2003). Warning: Health 'choices' can kill. *The Journal of Adult Protection*, 5(1), 30-34.
- Forster, J. (Ed.) (2010). *Standards of care and best practice guidelines*. Munich, Germany: IPWSO.
- Gabre, P., Martinsson, T., & Gahnberg, L. (2002). Move of adults with intellectual disability from institutions to community-based living: Changes of food arrangements and oral health. *Swedish Dental Journal*, 26(2), 81-88.
- Gill, J. & Fazil, Q. (2013). Derogation of "duty of care" in favour of "choice"? *The Journal of Adult Protection*, 15(5), 258-270. Retrieved from: <http://dx.doi.org/10.1108/JAP-12-2012-0028>

- Goff, B.J. (2008). *Supporting adults with Prader-Willi Syndrome in residential settings: An expert's recommendations*. Sarasota, FL: Prader-Willi Syndrome Association (USA).
- Goldstone, A. P., Holland, A. P., Hauffa, B. P., Hokken-Koelega, A. C., & Tauber, M. (2008). Recommendations for the diagnosis and management of Prader-Willi Syndrome. *The Journal of Clinical Endocrinology and Metabolism*, 93(11), 4183-4197.
- Gooding, P., Arstein-Kerslake, A., Andrews, L., & McSherry, B. (2016). Unfitness to stand trial and the indefinite detention of persons with cognitive disabilities in Australia: Human rights challenges and proposals for change. *Melb. UL Rev.*, 40, 816.
- Gorea, R. K. (2015). Evidence based medical ethics: A critical evaluation. *International Journal of Ethics, Trauma & Victimology*, 1(1), 5-7.
- Gourash, L. M., Hanchett J. E., & Forster J. L. (2006). Inpatient crisis intervention for persons with Prader-Willi Syndrome. In M. G. Butler, P. D. K. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (3rd Ed.), pp. 395-426. New York, NY: Springer.
- Gratton, C., Amar, A. & Watson, S. L. (2013). Prader-Willi Syndrome: Implications for applied behaviour analysis. In D. Griffiths, R. Condillac, & M. Legree, *Genetic syndromes and applied behaviour analysis: A handbook for applied behaviour analysis practitioners*. (pp. 225-256). London, UK: Jessica Kingsley Publishers.
- Griffith, G. M., & Hastings, R. P. (2014). 'He's hard work, but he's worth it' – The experience of caregivers of individuals with intellectual disabilities and challenging behaviour: A meta-synthesis of qualitative research. *Journal of Applied Research in Intellectual Disabilities*, 27(5), 401-419.
- Grondhuis, S.N., & Aman, M. G. (2014). Overweight and obesity in youth with developmental disabilities: A call to action. *Journal of Intellectual Disability Research*, 58(9), 787-99.
- Grugni, G., Crino, A., Bosio, L., Corrias, A., Cuttini, M., De Toni, T., ... & Iughetti, L. (2008). The Italian national survey for Prader-Willi Syndrome: An epidemiologic study. *American Journal of Medical Genetics, Part A*, 146(7), 861-872.
- Haig, E. L., & Woodcock, K. A. (2017). Rigidity in routines and the development of resistance to change in individuals with Prader-Willi Syndrome. *Journal of Intellectual Disability Research*, 61(5), 488-500.

- Hauber, M., Stratmann, B., Hoedebeck-Stuntebeck, N., & Tschoepe, D. (2013). Medical management for adults with Prader-Willi Syndrome. *Metabolic Syndrome and Related Disorders*, 11, 392-396.
- Hawkins, R., Redley, M., & Holland, A. J. (2011). Duty of care and autonomy: How support workers manage the tension between protecting service users from risk and promoting their independence in a specialist group home. *Journal of Intellectual Disability Research*, 55(3), 873-884.
- Heather, B. (2015). Coroner criticizes care after disabled Hawke's Bay man eats himself to death. *Stuff.co.nz*, August 2015.
- Retrieved from: <https://www.stuff.co.nz/national/health/71222478/Coroner-criticises-care-after-disabled-Hawkes-Bay-man-eats-himself-to-death>
- Hedgeman, E., Ulrichsen, S. P., Carter, S., Kreher, N. C., Malobisky, K. P., Braun, M. M., ... & Olsen, M. S. (2017). Long-term health outcomes in patients with Prader-Willi Syndrome: A nationwide cohort study in Denmark. *International Journal of Obesity*, 41(10), 1531.
- Hewitt, A., Larson, S., Edelstein, S., Seavey, D., Hoge, M. A., & Morris, J. (2008). *A synthesis of direct service workforce demographics and challenges across intellectual/developmental disabilities, aging, physical disabilities, and behavioral health*. Minneapolis, MN: University of Minnesota, Institute on Community Integration, Research and Training Center on Community Living.
- Hinton E. C., Holland A. J., Gellatly M. S. N., Soni S., Patterson M., Ghatei M. A., & Owen A. M. (2006). Neural representations of hunger and satiety in Prader-Willi Syndrome. *International Journal of Obesity*, 30(2), 313-321.
- Hinton, E. C., Holland, A. J., Gellatly, M. S. N., Soni, S., & Owen, A. M. (2006). An investigation into food preferences and the neural basis of food-related incentive motivation in Prader-Willi Syndrome. *Journal of Intellectual Disability Research*, 50(9), 633-642.
- Hinton, E. C., Isles, A. R., Williams, N. M., & Parkinson, J. A. (2010). Excessive appetitive arousal in Prader-Willi Syndrome. *Appetite*, 54(1), 225-228.
- Hiraiwa, R., Maegaki, Y., Oka, A., & Ohno, K. (2007). Behavioral and psychiatric disorders in Prader-Willi Syndrome: A population study in Japan. *Brain and Development*, 29(9), 535-542.

- Ho, A. Y., & Dimitropoulos, A. (2010). Clinical management of behavioural characteristics of Prader-Willi Syndrome. *Journal of Neuropsychiatric Disease and Treatment*, 6, 107-118.
- Hodapp, R. M., Dykens, E. M., & Masino, L. L. (1997). Families of children with Prader-Willi Syndrome: Stress-support and relations to child characteristics. *Journal of Autism and Developmental Disorders*, 27(1), 11-24.
- Holland, A. J. (2015). *The Mental Capacity Act (2005) and its significance for the support of people with PWS*. Retrieved from: www.praderwillisyndrome.org.uk/prader-willi-syndrome/the-mental-capacity-act-prader-willi-syndrome/
- Holland, A. J., Treasure, J., Coskeran, P., & Dallow, J. (1995). Characteristics of eating disorder in Prader-Willi Syndrome: Implications for treatment. *Journal of International Disability Research*, 39(5), 373-381.
- Holland, A. J., & Wong, J. (1999). Genetically determined obesity in Prader-Willi Syndrome: The ethics and legality of treatment. *Journal of Medical Ethics*, 25(3), 230-236.
- Holsen, L. M., Savage, C. R., Martin, L. E., Bruce, A. S., Lepping, R. J., Ko, E., Brooks, W. M., Butler, M. G., Zarcone, J. R., & Goldstein, J. M. (2012). Importance of reward and pre-frontal circuitry in hunger and satiety: Prader-Willi Syndrome vs simple obesity. *International Journal of Obesity*, 36(5), 368-347.
- Holsen, L. M., Zarcone, J. R., Brooks, W. M., Butler, M. G., Thompson, T. I., Ahluwalia, J. S., Nollen, N. L., & Savage, C. R. (2006). Neural mechanisms underlying hyperphagia in Prader-Willi Syndrome. *Obesity*, 14(6), 1028-1037.
- Hoybye, C. (2013). Current status of growth hormone therapy in Prader-Willi Syndrome. *Expert Review of Endocrinology & Metabolism*, 8(6), 529-536.
- Hoybye, C. (Ed). (2015). *Prader-Willi Syndrome*. Stockholm, Sweden: Nova Science.
- IHC New Zealand. (2011). *What we believe and why we believe it and what it means for people with disabilities and their families*. Wellington, NZ. Retrieved from: www.ihc.org.nz

- International Prader-Willi Syndrome Organisation [IPWSO]. (nd). *Dietary management*. Retrieved from: www.ipwso.org/#!/dietary-management/c1igx
- International Prader-Willi Syndrome Organisation [IPWSO]. (2013). *Famcare*. Retrieved from: www.org/#!/famcare/canb
- James, T. (2010). *Prader-Willi Syndrome: Quality of life*. Courtney, British Columbia: Poplar Publishing.
- Jauregi, J., Laurier, V., Copet, P., Tauber, M., & Thuilleaux, D. (2013). Behavioural profile of adults with Prader-Willi Syndrome: Correlations with individual and environmental variables. *Journal of Neurodevelopmental Disorders*, 5(1), 18.
- Jingree, T. (2015). Duty of care, safety, normalisation and the Mental Capacity Act: A discourse analysis of staff arguments about facilitating choices for people with learning disabilities in UK services. *Journal of Community & Applied Social Psychology*, 25(2), 138-152.
- Johnson, K. (1998). Deinstitutionalisation: The management of rights. *Disability & Society*, 13(3), 375-387.
- Jurgensmeyer, S. (2016). *Child and parental factors affecting coping strategies and psychological outcomes for parents with children with Prader-Willi Syndrome*. (Doctoral dissertation, Vanderbilt University; USA). Retrieved from: <https://ir.vanderbilt.edu/bitstream/handle/1803/7546/Jurgensmeyer%20Thesis.pdf?sequence=1>
- Kayadjanian, N., Schwartz, L., Farrar, E., Comtois, K. A., & Strong, T. V. (2018). High levels of caregiver burden in Prader-Willi Syndrome. *PloS one*, 13(3). Retrieved from <https://doi.org/10.1371/journal.pone.0194655>
- Kazemi, E., & Hodapp, R. M. (2006). Transition from adolescence to young adulthood: The special case of Prader-Willi Syndrome. In M. G. Butler, P. D. K. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (3rd Ed.), pp. 356-370. New York, NY: Springer.
- Key, A. P., & Dykens, E. M. (2018). Eye tracking as a marker of hyperphagia in Prader-Willi Syndrome. *Developmental Neuropsychology*, 43(2), 152-161.
- Kidd, P. S., & Parshall, M. B. (2000). Getting the focus and the group: enhancing analytical rigor in focus group research. *Qualitative health research*, 10(3), 293-308.
- Kinash, S. (2007). *A recipe for success: What you can learn about coping in a food-bombarded world from people with Prader-Willi Syndrome, an extreme eating disorder*. Charlotte, NC; IAP.

- Kortisas, S. & Iacono, T. (2015). Predictors of challenging behaviour in adults with intellectual disability. *Advances in Mental Health and Intellectual Disability*, 9(6), 312-316.
- Kroll, T., Barbour, R., & Harris, J. (2007). Using focus groups in disability research. *Qualitative health research*, 17(5), 690-698.
- Kubota, T., Miyake, K., Hariya, N., Tran Nguyen Quoc, V., & Mochizuki, K. (2016). Prader-Willi Syndrome: The disease that opened up epigenomic-based pre-emptive medicine. *Diseases*, 4(1), 15.
- Kuppens, R. J., Donze, S. H., & Hokken-Koelega, A. C. S. (2016). Promising effects of oxytocin on social and food-related behaviour in young children with Prader-Willi Syndrome: A randomized, double-blind, controlled crossover trial. *Clinical Endocrinology*, 85(6), 979-987.
- Lanfranchi, S., & Vianello, R. (2012). Stress, locus of control, and family cohesion and adaptability in parents of children with Down, Williams, Fragile X, and Prader-Willi Syndromes. *American Journal on Intellectual and Developmental Disabilities*, 117(3), 207-224.
- Lecavalier, L., Leone, S., & Wiltz, J. (2006). The impact of behaviour problems on caregiver stress in young people with autism spectrum disorders. *Journal of Intellectual Disability Research*, 50(3), 172-183.
- Lionti, T., Reid, S. M., White, S. M., & Rowell, M. M. (2015). A population-based profile of 160 Australians with Prader-Willi Syndrome: Trends in diagnosis, birth prevalence and birth characteristics. *American Journal of Medical Genetics, Part A*, 167(2), 371-378.
- López-Bastida, J., Linertová, R., Oliva-Moreno, J., Posada-de-la-Paz, M., Serrano-Aguilar, P., Kanavos, P., ... & Delgado, C. (2016). Social/economic costs and health-related quality of life in patients with Prader-Willi syndrome in Europe. *The European Journal of Health Economics*, 17(1), 99-108.
- McAllister, C. J., Whittington, J. E., & Holland, A. J. (2011). Development of the eating behaviour in Prader-Willi Syndrome: Advances in our understanding. *International Journal of Obesity*, 35(2), 188-197.
- McSherry, B., Murphy, T., Turkmendag, I., & Roberts, P. (2017). Regulating seclusion and restraint in health care settings: The promise of the Convention on the Rights of Persons with Disabilities. *International journal of law and psychiatry*, 53, 39-44.

- Maglieri, K. A., DeLeon, I. G., Rodriguez-Catter, V., & Sevin, B. M. (2000). Treatment of covert food stealing in an individual with Prader-Willi Syndrome. *Journal of Applied Behavior Analysis*, 33(4), 615-618.
- Mann, J., Zhou, H., McDermott, S., & Poston, M. B. (2006). Healthy behavior change of adults with mental retardation: Attendance in a health promotion program. *American Journal on Mental Retardation*, 111(1), 62-73.
- Manning, K. E., McAllister, C. J., Ring, H. A., Finer, N., Kelly, C. L., Sylvester, K. P., ... & Holland, A. J. (2016). Novel insights into maladaptive behaviours in Prader-Willi Syndrome: Serendipitous findings from an open trial of vagus nerve stimulation. *Journal of Intellectual Disability Research*, 60(2), 149-155.
- Mazaheri, M. M., Rae-Seebach, R. D., Preston, H. E., Schmidt, M., Kountz-Edwards, S., Field, N., ... & Packman, W. (2013). The impact of Prader-Willi Syndrome on the family's quality of life and caregiving, and the unaffected siblings' psychosocial adjustment. *Journal of Intellectual Disability Research*, 57(9), 861-873.
- Messersmith, N. V., Slifer, K. J., Pulbrook-Vetter, V., & Bellipanni, K. (2008). Interdisciplinary behavioural intervention for life-threatening obesity in an adolescent with Prader-Willi Syndrome – a case report. *Journal of Developmental and Behavioural Pediatrics*, 29(2), 129-134.
- Miller, J. L., & Angulo, M. (2014). An open-label pilot study of N-acetylcysteine for skin-picking in Prader-Willi Syndrome. *American Journal of Medical Genetics, Part A*, 164(2), 421-424.
- Miller, J. L., James, G. A., Goldstone, A. P., Couch, J. A., He, G., Driscoll, D. J., & Liu, Y. (2007). Enhanced activation of reward mediating prefrontal regions in response to food stimuli in Prader-Willi Syndrome. *Journal of Neurology, Neurosurgery & Psychiatry*, 78(6), 615-619.
- Miller, J. L., Lynn, C. H., Driscoll, D. C., Goldstone, A. P., Gold, J. A., Kimonis, V., ... & Driscoll, D. J. (2011). Nutritional phases in Prader-Willi Syndrome. *American Journal of Medical Genetics, Part A*, 155(5), 1040-1049.

- Miller, J. L., Lynn, C. H., Shuster, J., & Driscoll, D. J. (2013). A reduced-energy intake, well balanced diet improves weight control in children with Prader-Willi Syndrome. *Journal of Human Nutrition and Dietetics*, 26(1), 2-9.
- Mori, Y., Downs, J., Wong, K., Heyworth, J., & Leonard, H. (2018). Comparing parental well-being and its determinants across three different genetic disorders causing intellectual disability. *Journal of autism and developmental disorders*, 48(5), 1651-1665.
- Müller, M. J., Braun, W., Enderle, J., & Bosy-Westphal, A. (2016). Beyond BMI: conceptual issues related to overweight and obese patients. *Obesity facts*, 9(3), 193-205.
- Office for Disability Issues. (2001). *The New Zealand Disability Strategy: Making a world of difference. Whakanui oranga*. Wellington, NZ: Ministry of Health. Retrieved from: <http://www.odi.govt.nz>.
- Office for Disability Issues. (2014). *Disability Action Plan 2014-2018*. Wellington, NZ: Office for Disability Issues. Retrieved from: <http://www.odi.govt.nz>
- Ogata, H., Ihara, H., Gito, M., Sayama, M., Murakami, N., Ayabe, T., ... & Shimoda, K. (2018). Aberrant, autistic, and food-related behaviors in adults with Prader-Willi Syndrome: The comparison between young adults and adults. *Research in Developmental Disabilities*, 73, 126-134.
- Page, T. J., Finney, J. W., Parrish, J. M., & Iwata, B. A. (1983). Assessment and reduction of food stealing in Prader-Willi children. *Applied Research in Mental Retardation*, 4(3), 219-228.
- Page, T. J., Stanley, A. E., Richman, G. S., Deal, R. M., & Iwata, B. A. (1983). Reduction of food theft and long-term maintenance of weight loss in a Prader-Willi adult. *Journal of Behavior Therapy and Experimental Psychiatry*, 14(3), 261-268.
- Pearpoint, J., O'Brien, J., & Forest, M. (1992). *PATH*. Toronto: Inclusion Press.
- Pittsburgh Partnership. (n.d.). Pennsylvania, USA. Retrieved from: <http://www.pittsburghpartnership.com/>
- Prader-Willi Homes of Oconomowoc. (n.d.). Wisconsin, USA. Retrieved from: www.pwho.com/what-is-pws/
- Prader-Willi Syndrome Association of New Zealand. (nd). *Dietary management*. Retrieved from: www.pws.org.nz/dietary-management

- Prader-Willi Syndrome Association of New Zealand. (nd). *Guidelines for residential caregivers*. Retrieved from: <http://www.pws.org.nz/guidelines-for-residential-caregivers/>
- Risjord, M. (2014). Nursing and human freedom. *Nursing Philosophy*, 15(1), 35-45.
- Rone, A. B. (2010). *Treatment of food stealing in individuals with Prader-Willi Syndrome*. (Doctoral dissertation, University of Florida, Gainesville, USA). Retrieved from http://etd.fcla.edu/UF/UFE0041867/rone_a.pdf
- Rouse, L., & Finlay, W. M. L. (2016). Repertoires of responsibility for diabetes management by adults with intellectual disabilities and those who support them. *Sociology of Health & Illness*, 38(8), 1243-1257.
- Samuel, P. S., Rillotta, F., & Brown, I. (2012). Review: The development of family quality of life concepts and measures. *Journal of Intellectual Disability Research*, 56(1), 1-16.
- Schalock, R. L., Brown, I., Brown, R., Cummins, R. A., Felce, D., Matikka, L., ... & Parmenter, T. (2002). Conceptualization, measurement, and application of quality of life for persons with intellectual disabilities: Report of an international panel of experts. *Mental Retardation*, 40(6), 457-470.
- Schalock, R. L., Gardner, J. F., & Bradley, V. J. (2007). *Quality of life for people with intellectual and other developmental disabilities: Applications across individuals, organizations, communities, and systems*. Washington DC; American Association on Intellectual and Developmental Disabilities.
- Scheimann, A.O., Butler, M.G., Gourash, L., Cuffari, C. & Klish, W. (2008). Critical analysis of bariatric procedures in Prader-Willi Syndrome. *Journal of Pediatric Gastroenterology and Nutrition*, 46(1), 80-83.
- Schuengel, C., Clegg, J., de Schipper, J. C., & Kef, S. (2016). Adult attachment and care staff functioning. In H. Fletcher, A. Flood, & D. Hare (Eds.), *Attachment in Intellectual and Developmental Disability: A Clinician's Guide to Practice and Research* (151-171). Oxford, UK: John Wiley.
- Schwartz, L., Holland, A., Dykens, E., Strong, T., Roof, E., & Bohonowych, J. (2016). Prader-Willi Syndrome mental health research strategy workshop proceedings: The state of the science and future directions. *Orphanet Journal of Rare Diseases*, 11(1), 131.

- Selman, L. E., Fox, F., Aabe, N., Turner, K., Rai, D., & Redwood, S. (2017). 'You are labelled by your children's disability' – A community-based, participatory study of stigma among Somali parents of children with autism living in the United Kingdom. *Ethnicity & Health*, 23(7), 781-796.
- Shivers, C. M., Leonczyk, C. L., & Dykens, E. M. (2016). Life satisfaction among mothers of individuals with Prader-Willi Syndrome. *Journal of Autism and Developmental Disorders*, 46(6), 2126-2137.
- Shriki-Tal, L., Avrahamy, H., Pollak, Y., Gross-Tsur, V., Genstil, L., Hirsch, H. J., & Benarroch, F. (2017). Psychiatric disorders in a cohort of individuals with Prader-Willi Syndrome. *European Psychiatry*, 44, 47-52.
- Singh, N. N., Lancioni, G. E., Singh, A. N., Winton, A. S., Singh, J., McAleavey, K. M., & Adkins, A. D. (2008). A mindfulness-based health wellness program for an adolescent with Prader-Willi Syndrome. *Behavior Modification*, 32(2), 167-181.
- Singh, N. N., Lancioni, G. E., Singh, A. N., Winton, A. S., Singh, A. D., & Singh, J. (2011). A mindfulness-based health wellness program for individuals with Prader-Willi Syndrome. *Journal of Mental Health Research in Intellectual Disabilities*, 4(2), 90-106.
- Sinnema, M., Einfeld, S. L., Schrandt-Stumpel, C. T., Maaskant, M. A., Boer, H., & Curfs, L. M. (2011a). Behavioural phenotype in adults with Prader-Willi Syndrome. *Research in Developmental Disabilities*, 32(2), 604-612.
- Sinnema, M., Boer, H., Collin, P., Maaskant, M. A., van Roozendaal, K. E., Schrandt-Stumpel, C. T., & Curfs, L. M. (2011b). Psychiatric illness in a cohort of adults with Prader-Willi Syndrome. *Research in Developmental Disabilities*, 32(5), 1729-1735.
- Smyth, C. M., & Bell, D. (2006). From biscuits to boyfriends: The ramifications of choice for people with learning disabilities. *British Journal of Learning Disabilities*, 34(4), 227-236.
- Solvoll, B. A., Hall, E. O., & Brinchmann, B. S. (2015). Ethical challenges in everyday work with adults with learning disabilities. *Nursing Ethics*, 22(4), 417-427.
- Soni, S., Whittington, J., Holland, A. J., Webb, T., Maina, E., Boer, H., & Clarke, D. (2007). The course and outcome of psychiatric illness in people with Prader-Willi Syndrome: Implications for management and treatment. *Journal of Intellectual Disability Research*, 51(1), 32-42.

- Spendelow, J. S. (2011). Assessment of behavioural and psychiatric problems in people with Prader-Willi Syndrome: A review of the literature. *Journal of Policy and Practice in Intellectual Disabilities*, 8(2), 104-112.
- Stanley, Katherine (2013). Benefit of the doubt: The gift that has helped me thrive with Prader-Willi Syndrome. *Neurology Now*, 9(5), 38.
- Stanley, K. (2014). Prader-Willi Syndrome and hyperphagia: An update. *Advances in Mental Health and Intellectual Disabilities*, 8(5), 321-330. Retrieved from: <http://dx.doi.org/10.1108/AMHID-07-2013-0048>
- Strech, D. (2010). How factual do we want the facts? Criteria for a critical appraisal of empirical research for use in ethics. *Journal of Medical Ethics*, 36(4), 222-225.
- Tauber, M., Diene, G., Mimoun, E., Cabal-Berthoumieu, S., Mantoulan, C., Molinas, C. & Salles, J. P. (2014). Prader-Willi Syndrome as a model of human hyperphagia. In P. J. D. Delhanty & A. J. van der Lely (Eds.) *How Gut and Brain Control Metabolism* (Vol. 42, pp. 93-106). Rotterdam: Karger Publishers.
- Thompson, T., Kodluboy, S., & Heston, L. (1980). Behavioural treatment of obesity in Prader-Willi Syndrome. *Behavior Therapy*, 11(4), 588-593.
- Thomson, A. K., Glasson, E. J., & Bittles, A. H. (2006). A long-term population-based clinical and morbidity review of Prader-Willi Syndrome in Western Australia. *Journal of Intellectual Disability Research*, 50(1), 69-78.
- Thornton, L., & Dawson, K. P. (1990). Prader-Willi Syndrome in New Zealand: A survey of 36 affected people. *The New Zealand Medical Journal*, 103, 97-98.
- Thuilleaux, D., Laurier, V., Copet, P., Tricot, J., Demeer, G., Mourre, F., ... & Jauregi, J. (2018). A model to characterize psychopathological features in adults with Prader-Willi Syndrome. *American Journal of Medical Genetics*, Part A, 176(1), 41-47.
- Trip, H., Conder, J., Hale, L., & Whitehead, L. (2016). The role of key workers in supporting people with intellectual disability in the self-management of their diabetes: A qualitative New Zealand study. *Health & Social Care in the Community*, 24(6), 789-798.

- Tsungu, Pedzai. (2014). *Learning disability nurses' experiences of promoting autonomy in adults with profound and multiple learning disabilities: A phenomenological study*. (Masters thesis, University of Huddersfield, UK). Retrieved from <http://eprints.hud.ac.uk/23489>
- Tvrdek, T., Mason, D., Dent, K M., Thornton, L., Viskochil, D.H., & Stevenson, D.A. (2015). Stress and coping in parents of children with Prader-Willi Syndrome: Assessment of the impact of a structured plan of care. *American Journal of Medical Genetics, Part A*, 167(5), 974-982.
- United Nations Office of the High Commissioner for Human Rights. (2014). *The Convention on the Rights of Persons with Disabilities: Training Guide (No. 19)*. Geneva: United Nations Publications.
- Van Hooren, R. H., Widdershoven, G. A. M., Candel, M. J. J. M., Van den Borne, B. W., & Curfs, L. M. G. (2006). Between control and freedom in the care for persons with Prader-Willi Syndrome: An analysis of preferred interventions by caregivers. *Parent Education and Counselling*, 63(1-2), 223-221.
- Van Hooren, R. H., Widdershoven, G. A. M., Van den Borne, H. W., & Curfs, L. M. G. (2002). Autonomy and intellectual disability: The case of prevention of obesity in Prader-Willi Syndrome. *Journal of Intellectual Disability Research*, 46(7), 560-568.
- Van Hooren, R. H., Widdershoven, G. A. M., Van der Bruggen, H., Van den Borne, H. W., & Curfs, L. M. G. (2005). Values in the care for young persons with Prader-Willi Syndrome: Creating a meaningful life together. *Child: Care, Health and Development*, 31(3), 309-319.
- Van Lieshout, C. F., De Meyer, R. E., Curfs, L. M., & Fryns, J. P. (1998). Family contexts, parental behaviour, and personality profiles of children and adolescents with Prader-Willi, Fragile-X, or Williams syndrome. *The Journal of Child Psychology and Psychiatry and Allied Disciplines*, 39(5), 699-710.
- Vogt, K., & Emerick, J. (2015). Growth hormone therapy in adults with Prader-Willi syndrome. *Diseases*, 3(2), 56-67.
- Von Deneen, K. M., Gold, M. S., & Liu, Y. (2009). Food addiction and cues in Prader-Willi Syndrome. *Journal of Addiction Medicine*, 3(1), 19-25.

- Ware Jr, J. E., & Sherbourne, C. D. (1992). The MOS 36-item short-form health survey (SF-36): I. Conceptual framework and item selection. *Medical care*, 30(6), 473-483.
- Waters, J. (2009). *A handbook for parents and carers of adults with Prader-Willi Syndrome* (2nd revised Ed.). Prader-Willi Syndrome Association (UK).
- Whitehead, L. C., Trip, H. T., Hale, L. A., & Conder, J. (2016). Negotiated autonomy in diabetes self-management: The experiences of adults with intellectual disability and their support workers. *Journal of Intellectual Disability Research*, 60(4), 389-397.
- Whitman, B. Y. (2013) Understanding and managing behaviour in PWS. In C. Hoybye (Ed.), *Prader-Willi Syndrome*. Stockholm, Sweden: Nova Science Publishers, Inc.
- Whitman, B. Y., & Jackson, K. (2006). Tools for psychological and behavioural management. In M. G. Butler, P. D. K. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (3rd Ed.), 317-343. New York, NY: Springer.
- Whittington, J., & Holland, A. (2004). *Prader-Willi Syndrome: Development and Manifestations*. Cambridge, UK: Cambridge University Press.
- Whittington, J., & Holland, A. (2010). Neurobehavioural phenotype in Prader-Willi Syndrome. *American Journal of Medical Genetics, Part C: Seminars in Medical Genetics*, 154(4), 438-447.
- Whittington, J., & Holland, A. (2017). Cognition in people with Prader-Willi Syndrome: Insights into genetic influences on cognitive and social development. *Neuroscience & Biobehavioral Reviews*, 72, 153-167.
- Whittington, J., Holland, A., & Webb, T. (2009). Relationship between the IQ of people with Prader-Willi Syndrome and that of their siblings: Evidence for imprinted gene effects. *Journal of Intellectual Disability Research*, 53(5), 411-418.
- Whittington, J., Holland, A., Webb, T., Butler, J., Clarke, D., & Boer, H. (2004). Academic underachievement by people with Prader-Willi Syndrome. *Journal of Intellectual Disability Research*, 48(2), 188-200.

- Wills, R. (2018). Easy targets: Seen and not heard – the silencing and invisibility of disabled children and parents in post-reform Aotearoa New Zealand. In *The Palgrave Handbook of Disabled Children's Childhood Studies*, (575-594). London, UK: Palgrave Macmillan.
- Wilkinson, S. (1998). Focus groups in health research: Exploring the meanings of health and illness. *Journal of Health Psychology*, 3(3), 329-348.
- Wilkinson, S. (1999). Focus groups: A feminist method. *Psychology of women quarterly*, 23(2), 221-244.
- Wilkinson, S. (2004). Focus Group Research. In D. Silverman (Ed.) *Qualitative research: Theory, method and practice*, (177-199). London: Sage.
- Wolfensberger, W. (2002). Social role valorization and/or versus, “empowerment”. *Mental Retardation*, 42, 252-258.
- Wullink, M., Widdershoven, G., Van Schrojenstein Lantman-de Valk, H., Metsemakers, J., & Dinant, G.-J. (2009). Autonomy in relation to health among people with intellectual disability: A literature review. *Journal of Intellectual Disability Research*, 53, 816-826.
- Wyatt, D. A. (2006). A national approach to crisis intervention and advocacy. In M. G. Butler, P. K. D. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (3rd Ed.), pp. 440-447. New York, NY: Springer.
- Zhang, C., Yin, A., Li, H., Wang, R., Wu, G., Shen, J., ... & Zhang, Y. (2015). Dietary modulation of gut microbiota contributes to alleviation of both genetic and simple obesity in children. *EBioMedicine*, 2(8), 966-982.
- Ziccardi, M. K. (2006). Residential care for people with Prader-Willi Syndrome. In M.G. Butler, P. K. D. Lee, & B. Y. Whitman (Eds.), *Management of Prader-Willi Syndrome* (3rd Ed.), pp. 381-394. New York, NY: Springer.
- Zipf, W.B., & Berntson, G.G. (1987). Characteristics of abnormal food-intake patterns in children with Prader-Willi Syndrome and a study of the effects of naloxone. *American Journal of Clinical Nutrition*, 46(2), 277-281.
-

Appendices

Appendix A



A study investigating the residential services that people who have Prader-Willi syndrome receive in New Zealand -

Information for people who have PWS and their support people.

You are invited to take part in a research project to learn more about New Zealanders who have Prader-Willi syndrome and the kinds of services they receive.

I will be the principal investigator; I am doctoral student at Massey University's School of Psychology and this study will form part of the requirement for completing this degree. I am also a Registered Psychologist; I have worked in behaviour support to people who have Intellectual Disabilities for many years and I have been interested in supporting people who have Prader Willi Syndrome for most of that time. My university supervisors are Dr Ian deTerte and Prof. Ian Evans; our project has the support of The Prader-Willi syndrome Association of New Zealand, PWSA(NZ).

What are we studying exactly?

Firstly we want to be able to make an estimate of how many adults there are in New Zealand who have PWS and then we want to identify the kinds of support services they are currently receiving, either from family members, volunteers or paid support people. We would especially like to know what steps caregivers are taking to help people with PWS cope around food. We will also ask people who have Prader Willi

Syndrome about how happy and healthy they are. We plan to compare the answers of people whose access to food is strictly controlled with the information supplied by those who have freer access to food.

Why is this project important?

People who have Prader Willi Syndrome have some unique support needs; they experience a higher rate of service failure than other people who have intellectual disabilities and they are over-represented in forensic and behaviour support services. There are also serious ethical and practical challenges for caregivers supporting people with PWS.

It is important that we find out as much as we can about the best way to support this group of New Zealanders, our study will be a part of a wider project that aims to develop a handbook that will include guidelines and resources for people planning a new service for anyone who has PWS.

Who are we looking for?

We would welcome the help of every person living in New Zealand who has Prader Willi Syndrome and is at least 16 years old.

Each study participant will also need a support person who is willing to help; either a family member, a friend, or a person who is employed as a caregiver. They'll need to be someone who the person with PWS is happy to work with and who knows them well. We need the support people to help the people with PWS participate in the process safely and to make sure their rights are respected, but also to complete one of the questionnaires that is designed to be filled out by a caregiver.

What is going to happen?

If you are a person with PWS and you would like to participate in this study please discuss it with a support person and then fill out the enclosed consent form and return it in the stamped, addressed envelope provided; we will not proceed any further unless both the person who has PWS and a caregiver are happy to sign the consent form.

Then I will send you a set of three questionnaires to fill out together. These will include a questionnaire which asks the person with PWS about age, height and weight and about the kinds of support being

received. Next there is the Hyperphagia Questionnaire which asks questions about plans to help the person with PWS to cope around food, and finally a questionnaire which asks questions about their health and happiness. All together they should not take more than an hour to complete.

These questionnaires will have your names on them and the information you give will be combined with everyone else's before anyone is allowed to see it. We will not record the name of the support person's employer either, unless that organisation would like to have their support acknowledged in the study's final report.

I'll keep the questionnaires securely until I have finished my PhD. Massey University may continue to do so afterwards but will keep any information that identifies either any person who has PWS or any of the support people who assisted them in this study. We will either destroy the questionnaires or store them in a secure archive by the time five years is up.

What are the benefits and risks of taking part in this study?

By helping with this project the people who participate will have the opportunity to influence future services and to improve the lives of people who have PWS and the working environment of those who care about them; the organisations who employ these support people will have the opportunity to have their support acknowledged in the final report. We will also link you with the New Zealand and international community of caregivers who support people with PWS by sending you a list of web addresses and blog sites for The Prader Willi Syndrome Association (NZ) and The International Prader Willi Syndrome Organisation (IPWSO).

You'll also be able to contact me about the study using the email address or phone number at the top of this letter and I will make sure a copy of the final report about the study is available for you to read; including a 'plain language' version if you'd prefer.

The answers you give us will not be rated or judged; they will just be recorded and grouped together to give us a statistical picture. We will not provide anyone else with any of the information you give us.

Participant's Rights

You are under no obligation to accept this invitation. If you decide to participate, you have the right to:

- decline to answer any particular question;
- withdraw from the study at any time before you return your questionnaires.
- ask any questions about the study at any time during participation;
- provide information on the understanding that your name will not be used unless you give permission to the researcher.
- be given access to a summary of the project findings when it is concluded.

Ethics committee approval and funding for this research.

This project has been reviewed and approved by the Massey University Human Ethics Committee: Southern A, Application 14/37. If you have any concerns about the conduct of this research, please contact Dr Brian Finch, Chair, Massey University Human Ethics Committee: Southern A, telephone 06 350 5799 x 84459, email humanethicsoutha@massey.ac.nz.

How to contact the research team

If you have any questions or concerns please contact;

John Ford: [REDACTED]; [REDACTED].

Dr Ian deTerte, Senior Lecturer, School of Psychology, Massey University, Wellington:
I.deTerte@massey.ac.nz; 04 801 5799 extn: 63603.

Prof. Ian Evans, Professor Emeritus, School of Psychology, Massey University, Wellington.
I.M.Evans@massey.ac.nz.

Thank you for considering participating in this study

Appendix B



A study investigating the residential services that people who have Prader Willi syndrome receive in New Zealand.

PARTICIPANT CONSENT FORM

I agree to participate in this study under the conditions set out in the Information Sheet. I have read the Information Sheet, or my support person has read it and explained it to me, and my questions have been answered to my satisfaction. I understand that either I or my support person may ask further questions at any time by contacting either the principal researcher (John Ford; [REDACTED]; [REDACTED]) or the study supervisors at the School of Psychology, Massey University, Wellington; Dr Ian deTerte; I.deTerte@massey.ac.nz and Emmeritus Professor Ian Evans; I.M.Evans@massey.ac.nz

I am 16 or older, I have Prader-Willi syndrome and I agree to be supported in this project by the support person who has also signed this form.

Signature:

.....

Date:

.....

Full Name - printed

.....

For the support person:

I agree to help the above named person to participate in this study as laid out in the information sheet.

Signature:

Date:

Full Name - printed

What is the best address for me to send you the study questionnaires?

Thank-you both for agreeing to participate in this study

Appendix C



A Questionnaire for New Zealanders who have Prader Willi Syndrome (PWS) and their support people

Questions for the person who has Prader-Willi

1. Do you live with members of your family?

☐ Yes

☐ No

2. How many other people live in your house? Do not count people who are paid to stay there.

☐ No one else; I live by myself.

☐ 1 other person.

☐ 2 other people.

☐ 3 other people.

☐ 4 other people.

☐ More. How many? -----

3. Do any of these people also have Prader Willi Syndrome?

- ☐ No.
- ☐ Yes. How many? -----

4. Do you have staff who are paid to help at your house during the day?

- ☐ No.
- ☐ Yes; staff are there all day or whenever a person with PWS is at home.
- ☐ Staff are there for most of the day but at least one person with PWS also spends some time there without staff at least once a week.
- ☐ Staff are there less than half of the time.

5. Do you have staff who are paid to stay at your house over night?

- ☐ No.
- ☐ Yes; we have staff who 'sleep over'.
- ☐ Yes; we have staff who stay awake.

6. What is your height in meters and centimetres? _____

7. What is your weight in kilograms? _____

Questions for the support person

8. Measures that are in place to prevent overeating?

Do you know how many calories the person who has PWS can eat each day without gaining weight? Do you have a special diet planned to help that person keep within that limit?

- ☐ Yes to both questions.
- ☐ No to one or both questions.

Are either the fridge or food cupboards locked?

☐ Yes

☐ No

Is the person who has PWS able to spend their spare money without supervision?

☐ Yes

☐ No

☐ Sometimes; please explain

.....

.....

.....

.....

.....

In an average week, does the person who has PWS go out without being supervised?

☐ Yes

☐ No

Please write down any other things you or the other support people do to help prevent overeating.

Thanks for your help.

Appendix D



A study identifying key issues in providing residential services to New Zealanders who have Prader Willi Syndrome (PWS)

- Information for professional caregivers and service managers.

Dear Colleague

My name is John Ford, I am completing a research project looking at New Zealand services to people who have PWS and the way that professional caregivers have dealt with the challenges they face in their work. I am looking for a small group of support people to participate in a brief, anonymous interview about the issues they believe are most important and most challenging in their work, and about the way in which they have managed the issue of helping the people with PWS who they support manage around food.

I am doctoral student at Massey University's School of Psychology and this study will form part of the requirement for completing this degree. I am also a Registered Psychologist; I have worked in behaviour support to people who have Intellectual Disabilities for many years and been interested in supporting people who have Prader Willi Syndrome for most of that time. My university supervisors are Dr Ian deTerte and Professor Emeritus Ian Evans, our project has the support of the New Zealand Prader Willi Syndrome Association.

Why is this project important?

As you probably know there are some ethical and practical challenges that often come with supporting a people who have PWS; health and behavioural challenges are the most common and these are often

associated with the drive to overeat that people who have this syndrome often experience. Despite these challenges we actually know very little about the services this group of New Zealanders with PWS is currently receiving.

This study will be part of a project which will identify the range of residential services currently being provided to adults who have PWS in New Zealand, and will then look at the challenges faced by caregivers and some of the outcomes being achieved.

Who are we looking for?

We are looking to recruit a small group (about 12) of support staff from different parts of New Zealand who are employed by a residential support service and who have supported at least one adult who has PWS; they may be direct care workers or supervisory (first level) service managers.

What is going to happen?

If you agree to participate in this study I will send you a consent form and give you the chance to ask any questions you might have, we will only proceed if you are happy to sign the form and even then you can pull out or ask more questions at any stage. If you would like to participate I will meet with you briefly in the meeting room of a local community organisation. I will ask you a short list of questions about the things that you think are important in supporting a person with PWS; you might just want to answer these questions or they might lead to a short conversation between us, either way I expect the whole process to take between 15 minutes and an hour.

I will record our interview on a digital voice recorder and transcribe it into written form. I will send you a copy of that transcript so that you can review it for accuracy and have the chance to change anything you are not comfortable with. We will not record your name or the name of your employer, although we will offer your employer the opportunity to have their support acknowledged in the final report. We will not record any personal information about your clients. The transcript of our interview will be identified only by a number; they will be stored securely or destroyed when the project is finished. The digital recordings will be destroyed as soon as the transcripts are complete and we've agreed that they accurately reflect what you want to say.

What are the benefits and risks of taking part in this study?

By helping with this project you will have opportunity to use the experience and knowledge you have gained in your work to help influence future services and to improve the lives of people who have PWS, and of those who support them.

Your employer will have the opportunity to be acknowledged in the final report and I will link you with the New Zealand and international community of caregivers who support people with PWS by providing you with a list of web addresses and blog sites for the work of the New Zealand Prader Willi Syndrome Association (PWSA-NZ) and the International Prader Willi Syndrome Organisation (IPWSO).

I am also happy to discuss this study with the people you support who have PWS or their families, especially if you feel they are likely to be concerned about it. Everyone who has PWS is different however and so before I meet with any resident I would want to talk to you or your colleagues and to plan the best way to communicate with the person you support, and to help them express their opinions. Please contact me if you would like to discuss this further.

The information you provide us will not be rated or judged; they will be combined with information from other caregivers and grouped according to themes. We will not provide your employer or anyone else outside of the research team with any of the information you give us; unless of course you tell us something that means that someone is at risk of serious harm and have to take action to keep that person safe.

Participant's Rights

You are under no obligation to accept this invitation. If you decide to participate, you have the right to:

- decline to answer any particular question;
- withdraw from the study at any time before you return your questionnaires.
- ask any questions about the study at any time during participation;
- provide information on the understanding that your name will not be used unless you give permission to the researcher.
- be given access to a summary of the project findings when it is concluded.

Ethics committee approval and contact details for more information.

This project has been reviewed and approved by the Massey University Human Ethics Committee: Southern A, Application 15/56. If you have any concerns about the conduct of this research, please contact

Mr Jeremy Hubbard, Chair, Massey University Human Ethics Committee: Southern A, telephone 04 801 5799 x 63487, email humanethicsoutha@massey.ac.nz.

If you have any questions or concerns please contact John Ford on [REDACTED], [REDACTED] or [REDACTED]. You can also contact Dr Ian de Terte at I.deTerte@massey.ac.nz, (04) 801 5799 ext. 63603 or Massey University PO Box 756 Wellington 6140.

Thank you for considering taking part in this study.

Appendix E



MASSEY UNIVERSITY
COLLEGE OF HUMANITIES
AND SOCIAL SCIENCES
TE KURA PŪKENGĀ TANGATA

How do New Zealand caregivers of people with Prader-Willi Syndrome (PWS) conceptualise the dilemmas inherent in their work?

- Questions for semi-structured interviews with caregivers.

1. How long have you supported people with PWS?
2. What is your present role?
3. How is access to food managed for your clients who have PWS?
4. How was that food management plan decided on?
5. What is the most challenging or the most difficult thing about your work?
6. What do you think are the most important things a residential service needs to have or to do if it is going to provide good support to someone with PWS?

Appendix F

Interview questions.

Study 1: Interviews with caregivers of adults who have Prader-Willi syndrome in workplace based focus groups.

1. What is your present role?
2. How long have you supported someone with PWS?
3. What are the most challenging or difficult aspects of your present work?
4. What aspects are most rewarding?
5. How is access to food managed in your service?
6. What are the most important things a service has to do or to have in order to provide good quality support to people with PWS?
7. Do you have any other comments?