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An analysis of use of equipment and respite services by families with a daughter with Rett Syndrome

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An analysis of use of equipment and respite services by families with a daughter with Rett syndrome.

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A report submitted in Partial Fulfilment of the Requirements for the Award of Bachelor of Science (Occupational Therapy) (Honours), Faculty of Computing, Health and Science

Edith Cowan University

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Literature Review

The use of supportive resources and the well-being of parents with a daughter with Rett syndrome.

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The use of supportive resources and the well-being of parents with a daughter with Rett syndrome.

Abstract

Purpose: This literature review examines the body of knowledge concerning the use of respite services and assistive products and technology, child and family characteristics, and parental well-being among families with a daughter with Rett syndrome or with a child with another developmental disability.

Method: Literature published in the time period 1983-2009 was searched. Studies were included in this review if they reported use of respite services or assistive products and technology among families with a daughter with Rett syndrome or with a child with another developmental disability. Child and family characteristics and parental well-being were also considered.

Results and conclusions: Respite services have the potential to decrease parental stress and therefore may provide an important resource for families with a child with a disability. Additionally a number of characteristics including the age and clinical severity of a child with a disability and where their family lives may impact on the use of supportive resources. However there is a clear need for research to examine the relationships between child and family characteristics, the use of supportive resources and parental well-being in families with Rett syndrome.

Relationships between child and family characteristics, the use of supportive resources and parental well-being in Rett syndrome and other developmental disabilities.

Introduction

Rett syndrome is a rare neurodevelopmental disorder affecting 1:8500 females by the age of 15, making it a leading cause of severe intellectual disability in females [1]. The clinical features of Rett syndrome were first identified by Dr. Andreas Rett in 1966. However not until Hagberg and his colleagues described the disorder in a case series of 35 patients did Rett syndrome become widely recognised [2]. Features of Rett syndrome typically manifest following a period of apparently normal development in the first six months of life [3]. Common clinical features include loss of purposeful hand use and the development of hand stereotypies, loss of communication skills, cognitive impairment, impaired mobility and social withdrawal [3]. In addition breathing abnormalities, epilepsy, growth retardation and scoliosis may develop [3,4].

The progression of Rett syndrome usually follows four clinical stages [5]. The first stage occurs between the ages of 6 and 18 months. During this stage girls experience developmental arrest, decreased interest in social activities and, unspecific, episodic hand waving may occur. Following is a stage of rapid developmental regression where intellectual disability, hand stereotypies, and apraxic and ataxic gross motor movements become evident. Some stabilisation in clinical features may occur once girls are of school age, although severe physical and intellectual limitations are still present [5]. During the final stage, older girls and women experience a further deterioration in motor abilities [6], with many previously mobile girls/women loosing the ability to walk [5]. Older girls/women may also experience further reductions in hand function [7] and an improvement in emotional contact at this stage [5]. Although changes in the presentation of Rett syndrome occur with age, the clinical features observed in Rett syndrome result in severe physical and intellectual disability throughout the lifespan.

Diagnostic criteria for Rett syndrome were first developed in 1988 [8] and have since been updated to better cater for the inclusion of atypical cases, that meet some but not all of the criteria [3]. The discovery of the causal gene for Rett syndrome, methyl-CpG-binding protein 2 *(MECP2)* in 1999 has allowed clinical diagnosis to be verified with genetic testing in many cases [9]. *MECP2* mutations have been identified in up to 95% of genetically tested cases [10,11], with seven commonly occurring mutations (p.R133C, p.T158X, p.R168X, p.R255X, p.R294X and R306C) accounting for approximately 80% of pathogenic mutations among an Australian cohort of girls/women with Rett syndrome [12]. Despite this, Rett syndrome remains clinically defined as girls and women may fulfil the diagnostic criteria in the absence of a mutation [3].

Many recent studies have investigated the relationship between specific mutations and phenotype. Consensus in numerous studies is that the p.R270X mutation is associated with a more severe phenotype. Girls and women with this mutation may be expected to lose skills such as motor function, hand use and social interaction earlier and overall function more poorly [12-14] In contrast, girls and women with a p.R294X mutation have been associated with a milder phenotype [12-14]. Still much clinical variation between girls/women with the same mutation and especially between girls/women with different mutations is present [15]. It is suggested that this may be due to X inactivation status and other genetic influences and continues to be the focus of present research [12,16].

The management of Rett syndrome is often complex due to the various physical and intellectual impairments that result from the condition. As a result, numerous resources including medical, hospital, therapy, respite and alternative accommodation services are utilised by families caring for a girl/women with Rett syndrome [17]. An understanding of the need for these resources is crucial for effective planning and organisation of the care and management of girls/women with Rett syndrome. Also, little is known about the relationships between the use of resources, child and family characteristics and parental well-being among families with a daughter with Rett syndrome.

The purpose of this review was to examine the body of knowledge concerning the use of respite services and assistive products and technology in Rett syndrome and other developmental disabilities. The relationships between the use of these supportive resources, child characteristics (age, clinical severity and behaviour), family characteristics (socio-economic status, family size and geographical location) and parental well-being were also considered. To facilitate an understanding of the complex interactions between these variables, the International Classification of Functioning, Disability and Health (ICF), a widely recognised framework used to describe the complex relationships between health and health related factors [18], was used to guide the review of literature.

Methods

Search strategy

Literature searches were conducted using four electronic databases CINALH, Medline PyscInfo and ISI Web of Science. The search was confined to the time period 1983-2009, as Rett syndrome was not widely recognised prior to 1983. The main search terms included disability (Rett syndrome, autism, developmental disabilities, and cerebral palsy), participant (child, adolescent, girls and women), intervention (health resources, respite care, assistive technology devices, self-help devices, augmentative communication), and outcome terms (well-being, family, and parents). With the assistance of a librarian, search terms were exploded and adjusted to the terminology of each database. Additionally, reference lists of all retrieved relevant studies were manually searched to identify further studies for possible inclusion.

Inclusion and exclusion criteria

A *priori* criteria for inclusion of studies were applied to abstracts and then to full text articles. Studies were included in this review if they reported use of respite services or assistive products and technology among families with a daughter with Rett syndrome. However, due to the paucity of research in Rett syndrome, other disability

groups were also included. Respite was defined as any organised service that provided the primary carer of the person with a disability short-term relief from caring duties [19]. Assistive products and technology were defined as any adapted or specially designed product or technology aimed at improving the functioning of a person with a disability and included assistive technology for use in daily, mobility, communication and recreation activities [18]. Both qualitative and quantitative studies were included and no restriction on the level of evidence was imposed. The search was confined to peer-reviewed literature reported in the English language.

International Classification of Functioning, Disability and Health (ICF)

The ICF provides a framework to describe the complex relationships between health and health related factors and consists of four inter-related components, body functions and structures, activities and participation, environmental factors, and personal factors [18]. Body functions are "the physiological functions of body systems" [18] and include psychomotor, emotional and intellectual functions and body structures are "the anatomical parts of the body" [18]. Impairments in either body functions or body structures often results in activity limitations and participation restrictions. Environmental factors, including the use of respite services and assistive products and technology have the ability to influence body functions and structures and help overcome activity limitations and participation restrictions [18]. The results of the literature review will be presented in relation to the components of the ICF.

Results

Included in this review were twenty one studies, eleven included information on the use of respite services and ten included information on the use of assistive products and technology. Only seven articles specifically studied girls/women with Rett syndrome.

Methodological quality of studies

The majority of respite studies were observational and involved parent questionnaires or interviews or a combination of the both. Questionnaires and type of interview

varied amongst the studies and included a mix of validated and non-validated measures and additional questions. This variability in data collection methods may reflect the need for a standardised measure to study the use and efficacy of respite services among children with disabilities. Only one study investigating the effects of respite services on parental stress levels employed a pre-test post-test design [20]. Although in most cases an observational study design was warranted, future studies that examine the efficacy or effects of respite should include more rigorous study designs.

Studies investigating assistive products and technology employed a variety of designs, with the majority of the studies employing quasi-experimental designs such as time series [21-23] and multiple baselines [24-26]. Two observational studies were included that involved parent questionnaires [27,28]. Descriptive designs were employed by another two studies, one that involved interviews with children with cerebral palsy, their parents and teachers to describe the child's perception of using assistive devices [29] and another described the use of assistive technology in two case studies with girls with Rett syndrome [30].

The use of supportive resources

Respite services were used by families with children with a range of disabilities including autism spectrum disorder [31], cerebral palsy [32], intellectual disability [33] and severe learning disability [34]. As much as 68% of a sample of parents of young children with autism spectrum disorder and/or severe learning disability (n = 66) had used respite services at some point in time [34]. Other studies report that approximately half of parents with a child with autism spectrum disorder [31,35], cerebral palsy [32], or severe intellectual disability [36] had used respite services. This research demonstrates that respite services are accessed by a large number of families with a child with a disability, yet few studies have investigated respite use in families with a daughter with Rett syndrome.

An international study identified that approximately half of the sample of girls and women with Rett syndrome (n = 86) lived at home with the support of help or respite care [17]. On the other hand a study of Dutch girls/women with Rett syndrome, aged 16 years and over (n = 53), reported that only 12% of the sample had used respite services [37]. These findings suggest that a proportion of parents caring for a daughter with Rett syndrome do not access respite services. However the limitations of this research, which include diverse geographical locations of study cases, limited sample sizes and wide age ranges, need to be considered when interpreting these results. Clearly, further research is required to determine the use of respite services at a population level by families with a daughter with Rett syndrome.

Approximately 49% of children with a range of disabilities use assistive technology in daily, mobility, communication or recreation activities [28]. Products, equipment and technology that are used to overcome activity limitations and increase participation in daily activities [18]. Although it is well documented that the majority of girls and women with Rett syndrome experience restricted participation in daily activities as a result of cognitive, hand function and mobility limitations [3,4], the use of assistive products and technology in their daily life is poorly described in the literature. The only identified study described the effect of elbow restraints and hand splints on hand stereotypies during a self feeding task in four girls with Rett syndrome [21]. The results of the study varied markedly, as girls experienced both decreases and increases in the presence of different hand stereotypies with the use of the restraints and splints. Several factors including limited sample size, differences in clinical severity between participants and lack of a rigorous intervention protocol may account for this variation. Nevertheless the findings of this study highlight the need for empirical research with larger sample sizes to describe the use and effect of elbow restraints and hand splints on hand stereotypies in a variety of daily activities.

The wider disability research describes the use of assistive products and technology in a variety of daily activities including eating, bathing and toileting. Adapted cutlery and cups, adjustable seating systems and non-skid mats were commonly used with

children and adolescents with cerebral palsy when eating [27], yet few studies report on the effectiveness of such interventions. The first known systematic study of the efficacy of adapted spoons in cerebral palsy reported that spoons with thickened handles were effective in decreasing the amount of time required for an eating activity and had a positive impact on the fluency of movement [23]. Other cerebral palsy research reported that adaptive cutlery, and non-skid mats [27] and adaptive seating devices [22,27] were associated with improvements in the child's level of independence when eating [22,27] and overall sitting ability [22]. Understanding the role of assistive products and technology in minimising the disability which results from Rett syndrome is particularly important given that these girls/women experience severe impairments in hand function which impact on their ability to perform fine motor activities such as eating [7].

Among children with disabilities, a number of assistive products and technology were also used when bathing and toileting. Bath seats, height adjustable bathtubs and, shower and changing tables were commonly used for bathing young children with cerebral palsy [27]. Additionally, toilet chairs and seats were commonly used for toileting [27]. In the only identified study evaluating the use of adaptive seating systems for use on the toilet, it was reported that these systems had a positive impact on the independence of young children with cerebral palsy [22]. Girls/women with Rett syndrome experience a number of movement disorders including stereotypies, tremors and ataxia that may impact on their ability to safely perform bathing and toileting activities [6]. Therefore girls/women with Rett syndrome may require the use of bath seats, height adjustable bathtubs, and shower and changing tables and, other assistive products and technology not described here, when bathing and toileting.

Assistive products and technology for mobility and transportation refer to products, technology and equipment used to overcome activity limitations in moving inside or outside buildings [18]. Pushchairs, car seats, ankle foot orthoses, orthotic walking systems, walkers and walking chairs, and manual wheelchairs were commonly used for mobility and transportation activities by children with cerebral palsy [27,29]. Other

mobility and transportation technology, including hoists and portable ramps were used irregularly as families perceived them to be less practical and more time-consuming than lifting the child [27]. According to parent report, the use of walking systems, powered mobility and adapted tricycles improved the child's independence in mobility activities among children with cerebral palsy [27]. In a qualitative study of children's perceptions of their use of assistive products and technology, one child with cerebral palsy attributed better walking posture and better performance of some gross motor activities to wearing ankle foot orthoses. Overall the child felt that the mobility device was *'quite helpful'* [29]. This research clearly demonstrates that specific mobility and transportation devices can offer children with severe mobility limitations, such as girls/women with Rett syndrome, a means of increasing independent participation in mobility and transportation activities.

Assistive products and technology for communication refer to products, technology and equipment that assist people to send and receive information [18]. Children with severe communication limitations commonly use alternative or augmentative methods to support sending and receiving messages including picture communication symbols, portable dialogue units and, sign and language gestures [27]. It is recognised that the vast majority of girls and women with Rett syndrome experience severe communication limitations [4,38] and as such a number of studies have examined the effectiveness of assistive products and technology for communication among girls with Rett syndrome [24-26,30,39]. Computer-based communication technology has been used with girls with Rett syndrome for requesting wanted items [24] and selecting particular words [25]. Van Acker and Grant (1995) conducted a study investigating the use of a computer and touch screen to request food and drink items among three girls with Rett syndrome. During the intervention period each girl demonstrated improvements in their ability to request food/drink items [24]. In the other study, Hetzroni and colleagues examined the effectiveness of a specially developed computer program for teaching symbol identification in three girls with Rett syndrome [25]. The girls were required to match a spoken word to a symbol displayed on the computer screen using eye gaze, body posture and nose/forehead movements

to indicate their selection. Although during intervention all girls demonstrated a trend towards increasing number of correct responses, these findings were inconclusive as improving trends were also evident at baseline. This literature highlights that further research is required clarify the effectiveness of computer-based communication technology among girls with Rett syndrome.

Other assistive products and technology including BigMack switches, picture communication symbols, multi message communication devices and communication boards have also been used to enhance communication in girls with Rett syndrome [26,30,39]. The use of such devices was found to have a positive impact on symbolic communication during storybook reading interactions with six girls with Rett syndrome [39]. However, the effect of these devices was studied in conjunction with other interventions; therefore the observed increase in symbolic communication may have been due to other interventions such as mother training.

Another study with four girls with Rett syndrome investigated the use of a "want" symbol to request food, drink and toy items [26]. Girls were required to touch the "want" symbol to request an item. However after an initial baseline and intervention phase, two girls received a modified intervention, either touching a flattened potato chip bag to request chips or pressing a switch to activate music. The number of correct requests made varied between the girls and evidence for the effectiveness of the initial and modified interventions was inconclusive. Clearly it is important to provide girls and women with Rett syndrome a variety of opportunities to communicate in a variety of environments. However, there is also a clear need for research with larger sample sizes and more rigorous intervention protocols and data collection methods to determine the most appropriate assistive products and technology for communication among girls and women with Rett syndrome.

Assistive products and technology for culture, recreation and sport refer to products, technology and equipment used to enhance participation in cultural, recreational and sporting activities [18]. A variety of recreational assistive products and technology

including adapted toys and games, switches, sitting furniture for use on the floor and tables were commonly used by a sample of children with cerebral palsy [27]. In particular, adapted toys and computer games enhanced participation in play activities in some of the children [27]. This finding was supported by a qualitative study in which a child with cerebral palsy reported that they were better able to play with other children due to the use of their assistive devices [29]. This research suggests that assistive products and technology have the ability to enhance participation in recreational activities such as play. However, further research is required to determine the use and effectiveness of such assistive products and technology among girls/women with Rett syndrome.

Child characteristics related to the use of supportive resources

The use of supportive resources by families with a child with a disability is influenced by factors such as the age, severity and behaviour of the child. Currently no relationships between age and the use of assistive products and technology have been described in the identified literature. However, age has been reported to relate to respite use among children with autism spectrum disorder and severe intellectual disability [35,36]. Parents of children aged nine years or over with autism spectrum disorder [31,35] and parents of older children with severe intellectual disability were more likely to be using respite services [36]. In contrast, another study reported that age was not a predictor of respite service use among parents of children with cerebral palsy [32]. In general, the above research suggests a relationship between the age of a child and the use of respite services, with parents of older children being more likely to use respite services [31,35,36]. Therefore as girls and women with Rett syndrome and their parents' age, they may require the more respite services.

At present there is a paucity of literature describing the relationships between the use of assistive products and technology and the severity of a child's disability. One identified study reported that the use of assistive products and technology for mobility, self-care and social activities increased with increasing severity in a sample of children with cerebral palsy [27]. Children with cerebral palsy, with more severe

levels of disability [19,32] and additional health conditions [32] and, children with autism spectrum disorder with more severe language developmental delay [40] were more likely to access respite services. Although severity was not related to whether families with a child with a severe intellectual disability received respite services, the severity of the child's condition was related to whether families wanted respite services [36]. Despite considerable evidence supporting a relationship between increasing severity of disability and more frequent use of respite, one study reported that children with more severe levels of intellectual disability were not more likely to use respite services [33]. Collectively, research seems to suggest that children with more severe levels of disability are more likely to use respite services, thus girls/women with severe phenotypes of Rett syndrome may require and use respite services more.

Girls and women with Rett syndrome display a variety of behavioural disturbances. These may include breathing abnormalities such as breath holding, general mood disturbances such as spells of screaming and crying for no apparent reason, hand behaviours such as uniform and monotonous hand movements, repetitive facial movements including mouth grimacing and repetitive tongue movements, among many other anxiety, walking/standing, body rocking and night time behaviours [41]. Research describing the relationships between specific Rett syndrome behaviours and the use of support resources has not been found. However children with developmental disabilities who display more serious challenging behaviours were reported to be more likely to be excluded from respite [34]. Despite this finding, no significant relationships were found between the presence of challenging behaviour and respite use among children with intellectual disabilities [33]. The above research highlights the need for further research to clarify the impact behavioural difficulties has on the use of assistive products and technology and respite services among parents with a daughter with Rett syndrome and parents of children with other developmental disabilities.

Family characteristics related to the use of supportive resources

The use of supportive resources may also be influenced by family characteristics including socio-economic status and family size. However no identified studies reported on the relationships between the use of assistive products and technology and socio-economic status and family size. Families with a child with a disability on average have lower income and lower rates of employment than families without a child with a disability [42]. A study investigating the use of general health services among families with Rett syndrome reported that families with lower socioeconomic status and lower levels of maternal education utilised heath services less [43]. Also where a family lives may impact the use of resources, with families with a child with autism living in non-metropolitan areas having lower odds of using respite services [31]. Despite these findings, a study investigating the use of respite services among a representative sample of children with cerebral palsy in Ontario, Canada found that the level of household income and the education level of the carer was not associated with respite use [32].

The use of respite also appears to be related to family size. In a study investigating the characteristics associated with the use and non-use of respite services among children with severe intellectual disability, those who came from large families were more likely to receive respite services [36]. In contrast in another study also exploring the use of respite in such children, as well as in those with cerebral palsy, this relationship was not found [32,33]. The impact of family size on the use of respite services for families with a daughter with Rett syndrome is unknown. Clearly additional research is required to gain a greater understanding of the relationships between other socio-economic factors and the use of assistive products and technology and respite services among families with a daughter with Rett syndrome.

Parental well-being

Parents of children with a range of disabilities experience higher levels of stress than parents caring for typically developing children [44-46]. This stress contributes negatively to the well-being of parents of children with various diagnoses including

cerebral palsy [47], Down syndrome [48] and Rett syndrome [49]. An early study investigating the level of family stress among families with a daughter with Rett syndrome reported that parents of girls with Rett syndrome experienced higher levels of parenting stress in comparison to parents in a normative sample [46].In a study of mothers of children with Rett syndrome in the Australian Rett Syndrome Database, Laurvick and colleagues found that these mothers experienced significantly lower physical and mental health outcomes than a population comparison group [49]. This literature suggests that parents of a child with a disability, including Rett syndrome, experience higher levels of stress and lower levels of well-being than parents of typically developing children.

The impact of supportive resources on parental well-being

A myriad of factors including the behaviour [47-49] and severity of the child [47,48], education status of the mother [49], levels of social support [45], family functioning [47] and financial stress [50] have been associated with the stress and well-being of parents of a child with a disability. However, no identified studies have investigated the direct relationships between the use of assistive products and technology and parental well-being, and only a few have investigated the impact of respite services on the well-being of parents of a child with a disability. Among parents with a child with a disability the use of respite services was associated with significant decreases in parental stress [20,51,52], and a trend towards reduced family stress and financial worry [51]. Over 90% of caregivers of a child with cerebral palsy (n = 468) indicated that the use of respite was beneficial for both their family and child [32], suggesting positive effects on parental well-being. This research demonstrates that respite is a valuable service for families with a child with a disability and as such should be made available to parents with a daughter with Rett syndrome.

Discussion

Rett syndrome is associated with severe physical and intellectual disability throughout the lifespan [4,5]. As a result of the additional demands associated with the care and management of Rett syndrome, parents are at risk of reduced well-being

[46,49]. Research has demonstrated that respite services have the potential to decrease stress levels in parents of a child with a disability and therefore may provide an important resource for families [20,51]. However no identified literature examined the relationships between readily available assistive products and technology and parental well-being.

It is important to understand the child and family characteristics that mediate the use of respite services and assistive products and technology to help plan and organise the care and management of girls/women with Rett syndrome. Research suggests that among children with autism spectrum disorder [31,35] and severe intellectual disability [36], older children and children with more severe levels of disability use respite services more. Relationships between socioeconomic status and maternal education level and the use of general health services among families with a daughter with Rett syndrome have been identified [43], although respite services or the use of assistive products and technology were not examined in this study. The wider disability research suggests that families living in non-metropolitan areas with a child with autism spectrum disorder had lower odds of using respite services [31]. The impact of family size on the use of resources was inconclusive, with different studies producing varied results [32,33,36]. There is a clear need for research to clarify the impact child and family characteristics have on the use of respite services and assistive products and technology among families with a daughter with Rett syndrome.

The majority of the literature included in this review was observational; highlighting the need for more detailed observational studies that collect data over time and experimental studies to understand the relationships between child and family characteristics, the use of resources and parental well-being. Additionally few studies included rigorous intervention and assessment protocols and large sample sizes. Research employing rigorous methodology and larger sample sizes that examines the relationships between child and family characteristics and the impact supportive resources have on the well-being of parents with a daughter with Rett syndrome or child with a disability is required.

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Research Report

An analysis of use of equipment and respite services by families with a daughter with Rett syndrome.

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An analysis of use of equipment and respite services by families with a daughter with Rett syndrome.

Abstract

Purpose: To assess factors that could influence use of equipment and respite services among families with a daughter with Rett syndrome and to examine the relationships between the use of these resources and the health of female caregivers.

Method: Parent questionnaire data from 2004 and 2006 in the population-based Australian Rett Syndrome Database was the source of data. Logistic regression was used to analyse relationships between child factors (age, mobility, clinical severity and behaviour), family factors (accessibility and socioeconomic factors) and the use of equipment and respite services in 2004. Linear regression was used to analyse the relationship between the use of these resources in 2004 and the health of female caregivers in 2006.

Results: Data from 170 families with girls and women aged 2-28 years was used in this study. The majority (88.3%) of families used at least one piece of equipment in 2004. Increasing mobility restrictions were associated with the use of more equipment. Most (80.1%) of the families had also used some type of respite services in the past. The use of more home respite was associated with severely restricted levels of mobility and mothers having a vocational or university qualification. The use of more overnight respite was associated with increasing age and presence of behaviours and, mothers being employed in full-time or part-time work. Female caregivers had significantly lower mental health outcomes than the Australian female norm ($p<0.001$), yet surprisingly no relationship between resource use and mental health was identified.

Conclusions: Understanding the relationships between child and family factors, use of equipment and respite services and caregiver health can influence the care and management of girls and women with Rett syndrome.

Introduction

Rett syndrome is a rare neurodevelopmental disorder usually associated with mutations on the methyl-CpG-binding protein 2 gene *(MECP2)* [1-3]. The disorder mainly affects females and results in severe physical and intellectual disability [4,5]. Features of Rett syndrome typically manifest following a period of apparently normal development in the first six months of life and commonly include loss of purposeful hand use and the development of stereotypies, loss of communication skills, cognitive impairment, impaired mobility, and social withdrawal [6]. The care and management of girls and women with Rett syndrome is often complex due to the numerous and varied impairments associated with the disorder.

Evidence suggests that parents caring for a child with a developmental disability experience higher levels of stress than parents caring for typically developing children [7-9]. This stress has been found to contribute negatively to the physical and mental health of parents [10-12]. High levels of parental stress have been reported in Rett syndrome,[9] with mothers experiencing lower physical and mental health outcomes compared with a norm population [12]. Factors such as child behaviour [11,12] clinical severity [10,11], social support [8], family functioning [10], financial stress [13], levels of education, maternal employment [12] and the burden of caring [10] have been found to be related to levels of stress and health of parents of a child with a disability.

Families with a child with a disability often use adapted or specially designed equipment [14]. Benefits associated with equipment use may include better performance in play [15,16], eating [17] and communication activities, and increased independence in eating [15,18], mobility [15,16] and toileting activities [18]. Although the impact of equipment use on caregiver health has not been studied to date, the burden of care placed on caregivers could potentially be reduced on account of the children's increased functional performance and independence facilitated by equipment use.

Respite services are organised services that provide the primary carer of a person with a disability short-term relief from caring duties [19]. They are often accessed by caregivers of a child with a disability [20-23] and their use has been associated with decreases in stress and worry [24,25]. In families with a daughter with Rett syndrome an international study identified that approximately half of 86 girls and women in the sample lived at home with the support of help or respite services [26] whilst a Dutch study (n=53) of those age 16 years and over, reported that only 12% of the sample had used respite services at some point in time [27]. This suggests that many families caring for a daughter with Rett syndrome have not accessed respite services despite the potential benefits for caregiver health.

Child factors such as age, severity of the disability and behaviour could influence the use of equipment and respite services. Parents of children aged nine years or over with autism [20,28] and parents of older children with severe intellectual disability [29] were more likely to be using respite services in comparison to younger children, although to date this relationship with age has not been found in children with cerebral palsy [21]. Overall children with autism, with more severe language developmental delay [30] and children with cerebral palsy, with more severe levels of disability or additional health conditions [21] were more likely to use respite services. The use of equipment was also found to increase with increasing severity in children with cerebral palsy [15]. However mixed findings have been found in relation to the presence of challenging behaviours and use of respite services [22,23]. Additionally families with a child with autism who lived in non-metropolitan areas were less likely to use respite services [20] and the influence of family size on the use of respite services remains unclear [21,22,29]. There has been no research on the impact of child and family factors on the use of equipment and respite services in Rett syndrome.

The purpose of this study was therefore to assess factors that could influence use of equipment and respite services among families with a daughter with Rett syndrome and to examine the relationships between caregiver health and the use of equipment and respite services. We hypothesised that families with a child with greater clinical

severity, who was older or had less behavioural difficulties, and who had a higher level of socio-economic status and lived in urban areas would use more equipment and respite services. Finally we proposed that the use of equipment and respite would have a beneficial influence on the health of female caregivers.

Materials and Methods

The Australian Rett Syndrome Database has collected longitudinal data about girls and women with Rett syndrome since its establishment in 1993 [31]. Upon enrolment into the database questionnaires are administered to the child's paediatrician and family. Follow-up questionnaires have also been distributed to participating families every two years since 2000 [12]. These questionnaires were developed and piloted with families with a child with Rett syndrome to ensure that the content was relevant and captured the range of complex issues that a person with Rett syndrome and their family experience. For this study data from the 2004 and 2006 questionnaires were used. The questionnaires primarily collected information on the current health and functioning of the person with Rett syndrome, and their development and use of services over the past two years. Data from the 2004 questionnaire were used to ascertain child (age, mobility, behaviour and severity) and family factors (accessibility and socioeconomic factors), and the use of equipment and respite services. This questionnaire was mailed out to 226 families with a child with a verified diagnosis of Rett syndrome, with a response rate of 89.4% (202 out of 226). Questionnaires were excluded if they did not include information on the outcomes of interest such as those where the person with Rett syndrome was cared for in a residential setting. As a result 32 questionnaires were excluded, leaving a total of 170 questionnaires with data from 2004. Families (n=119) where a female caregiver (natural mother, foster mother or grandmother) had also completed the SF-12® Health Survey (Version 1.0) in the 2006 follow-up questionnaire were included in final analysis.

The mobility status of the individual with Rett syndrome was determined by scoring responses to the question *'How is your daughter's walking ability compared to other*

girls her age?' Answers we categorised as either: normal or mildly restricted, severely restricted, able to support weight briefly or confined to a wheelchair, or totally dependent on carer.

The Rett Syndrome Behaviour Questionnaire (RSBQ) [32] is a measure that uses a three-point scale to score the presence of specific behaviours, with increasing scores indicating a greater presence of behaviours. The general mood and night behaviours subscales were used to determine the extent of specific behaviours in this analysis. Scale and subscale internal consistency and test retest reliability has been found to be satisfactory.

The Kerr scale [33] was used to measure the clinical severity of the sample. The scale contains 20 items, each one associated with a common feature of Rett syndrome. Items are scored according to severity, with increasing scores indicating greater clinical severity.

A variety of indicators were utilised to evaluate demographic characteristics, socio-economic status and the health of female caregivers including the number of children in the family, family income and the education level of parents and their working status. Additional measures utilised for this purpose included the following:

The Accessibility/Remoteness Index of Australia (ARIA+) was used to measure the degree of accessibility to services. Based on road distances to service centers, the ARIA+ categorises areas as either major cities of Australia, inner regional Australia, outer regional Australia, remote Australia or very remote Australia [34]

The Socio-Economic Indexes for Area (SEIFA) Index of Relative Disadvantage and the *SEIFA Index of Education and Occupation* were used to measure aspects of socio-economic conditions. SEIFA Indexes are assigned to geographical areas not individuals, therefore they provide a general measure of disadvantage and education and occupation for the studied families [35].

The Australian and New Zealand Standard Classification of Occupations (ANZSCO) was used to measure the occupations of both parents. ANZSCO assigns occupations to one of five skill levels. Skill levels refer to the level of skill typically required to competently perform the tasks of a particular occupation and range from the highest skill level of 1 to the lowest skill level of 5 [36].

The SF-12® Health Survey (Version 1) was included in the 2006 questionnaire and was used to measure the health of the female caregiver. The SF-12® is a general measure of health related quality of life and is comprised of a physical component summary (PCS) and a mental component summary (MCS). Test-retest reliability and validity for both the physical and mental component summaries are satisfactory in the general US population [37] and the Australian population [38].

Data analysis

Missing values were imputed from other information contained in the 2004 questionnaires where appropriate. For analysis the remote and very remote ARIA+ categories were combined due to small numbers. Kerr scale scores for the sample were calculated using 16 items, as information on four items was not able to be collected. Any missing Kerr scale items were imputed using the STATA programme ice, which implements a multiple imputation using chained equations from the non-missing values [39]. For families with one or two missing values on the SF-12®, missing values were imputed using regression [39].

The analysis was conducted in two phases, the first analyses involved univariate logistic regression. Analyses were conducted separately for the use of equipment, home respite, overnight respite, other respite, and respite at some point in time, as the outcome variables of interest. Outcome variables were coded as binary measures. Equipment was coded as above and below the mean cost of equipment for the sample. Cost was measured using the annualised cost of all purchased, borrowed and rented equipment that had been previously calculated for the sample using cost data

collected in the 2004 questionnaire [40]. Home respite was coded as above and below the mean hours of home respite used and overnight respite was coded as above and below the mean number of times overnight respite was used in 2004. Other respite and the use of respite at some point in time were coded as either yes or no. Child and family factors were used as predictors in this analysis.

The second analyses involved univariate linear regression. Analyses were conducted with the female caregiver SF-12® (Version 1) PCS and MCS as outcome variables and use of equipment and respite services as predictor variables. Equipment use remained binary but the use of respite services was recoded as a categorical variable. Child and family characteristics that had a significant relationship with PCS and MCS ($p<0.1$) were considered as possible confounders in final multivariate analysis. The STATA 10 statistical package was used for this analysis [39].

Results

A hundred and seventy families were included in the first analyses using data collected in the 2004 questionnaire only. Kerr scale items were imputed for 72 cases and 5 cases had imputed SF-12 scores. The characteristics of these families and the average use of equipment, home and overnight respite services are described in Table I. The girls and women with Rett syndrome ranged from the age of 2 to 28 years with a mean age of 13.45 ± 6.04 years. About one third (32.4%) were totally dependent on their carer for movement and another third (31.8%) had normal or mildly restricted levels of mobility. The average general mood subscale score was 7.58 ± 3.88 out of a possible score of 16 and the average night behaviours subscale score was 1.77 ± 1.69 out of a possible 6. The average Kerr scale score was 18.62 (95% CI 17.96–19.28) out of a possible score of 32. Most commonly families lived in major cities of Australia (61.2%). Over half the families were either in the low or moderate disadvantage SEIFA quartiles (59.5%) or in the very high or high levels of education and occupation SEIFA quartiles (51.3%). More fathers (88.9%) participated in full-time or part-time employment (54.8%). Of those fathers employed, 38.2% had

occupations at the highest ANZSCO skill level of one. Of those mothers employed, 28.6% had occupations at skill level one.

Most (88.3%) families had used at least one piece of equipment at the time of the 2004 questionnaire. Wheelchairs were most commonly used with 83.2% of families reporting having used one or more wheelchairs during 2004. Other mobility equipment used included hoists (38.5%), car seats or travelling restraints (22.4%) and ankle foot orthoses (16.8%). Daily living equipment such as shower chairs (30.1%), special beds (22.4%) and bed adaptations including bed rails (18.9%) were also used. Communication devices were used by a quarter (25.9%) of families but few families reported that their daughter used recreation equipment. The use of equipment according to child and family factors is presented in Table II. Girls/women with severely restricted levels of mobility (OR 16.00, CI 1.89-135.39), who were able to weight bear briefly or confined to a wheelchair (OR 37.71, CI 4.47-317.90) and those who were totally dependent on their carer (OR 50.00, CI 6.39-391.40) had greater odds of using above the average cost of equipment in comparison to girls/women with normal or mildly restricted levels of walking. The use of equipment was also related to the severity of Rett syndrome as measured by the Kerr scale. With increasing severity, an increase in the odds of using above the average cost of equipment was observed (OR 1.20, CI 1.09-1.32). However, when the effect of mobility was taken into account, severity was no longer a significant predictor of the use of equipment (OR 1.05, CI 0.93-1.18). Families with the father employed in full-time or part-time work had lower odds of using above the average cost of equipment (OR 0.40, CI 0.14-1.14) and families with three children also had lower odds of using more equipment (OR 0.29, CI 0.07–1.13).

Most (80.1%) families had used some type of respite service in the past. At the time of the 2004 questionnaire, 54.9% of families had used in home respite services, 47.6% had used overnight respite services and 36.9% had used other forms of respite. The use of home respite services according to child and family factors is presented in Table III. Mothers with a vocational qualification had about 4 times the odds of using

above the average hours of home respite in comparison to mothers who had only primary or some high school education (OR 4.05, CI 1.45-11.36), whereas mothers with a university qualification had about 3 times the odds of using above the average hours of home respite (OR 2.80, CI 0.96-8.06). Girls/women who had severely restricted levels of mobility had about 3 times the odds of using above the average hours of home respite in comparison to girls/women with normal or mildly restricted levels of walking (OR 2.83, CI 0.96-8.30). Families with two children, including their daughter with Rett syndrome, had lower odds of using above the average hours of home respite in comparison to families with only one child (OR 0.36, CI 0.12-1.20).

The use of overnight respite services according to child and family factors is presented in Table IV. Caregivers of girls/women aged 13 $\leq$ 19 years had about nine times the odds of using above the average times of overnight respite than girls under the age of eight years (OR 9.30, CI 1.99-43.43). Caregivers of women over the age of 19 years (OR 7.37, 1.42-38.25) and girls aged 8$\leq$13 years (OR 5.15, 1.05-25.23) also had greater odds of using more overnight respite than caregivers of girls under the age of eight years. An increase of one point in the night behaviours subscale, suggesting a greater presence of difficult behaviours at night-time, was associated with caregivers having lower odds of using above the average times of overnight respite (OR 0.72, CI 0.54-0.95). A similar picture was observed with the general mood subscale (OR 0.90, CI 0.81–1.00). Families where the mother was employed in full-time or part-time work had twice the odds of using more overnight respite in comparison to families where the mother was not employed (OR 2.06, CI 0.92-4.59). Families with an income between $52,000 and $79,999 had lower odds of using more overnight respite in comparison to families with an income of less than $20,800 (OR 0.23, CI 0.04-1.20). Also families with mothers who had a vocational qualification had lower odds of using more overnight respite than families with a mother who had primary or some high school education (OR 0.36, 0.12-1.12).

A hundred and nineteen female caregivers were included in the final analysis. The average MCS for female caregivers was 41.07 $\pm$ 12.15 which was significantly lower

than the Australian female norm of 51.4 (CI 38.87-43.28, p<0.001). However, MCS did not vary with use the of equipment or respite services (Table V). The average PCS for female caregivers was 48.66 ± 9.97 which was slightly higher than the Australian female norm of 48.4 (p=0.387). The relationships between the use of equipment and respite services and PCS are presented in Appendix F. The use of above the mean cost of equipment was associated with a lower PCS or poorer physical health (coefficient -4.92, CI -8.80 - -1.03). This relationship remained significant after separately adjusting for child age (coefficient -3.60, CI -7.50 - -0.31), paternal working status (coefficient -3.98, CI -7.76 - -0.20, p=0.04), maternal working status (coefficient -4.70, CI -7.87 - -0.27) and the SEIFA Index of Education and Occupation (coefficient -5.33, CI -9.45 - - 1.21). The use of overnight respite services was also associated with the PCS, with female caregivers accessing these services having lower PCS (coefficient -8.05, CI -14.72 - -1.39). Caregivers who used all overnight, home and other respite services also had a poorer PCS (coefficient -6.6-, CI -13.42 – 0.22).

Discussion

The results showed that a high proportion of families used equipment to support participation in mobility and other daily activities. This was to be expected as it is well documented that girls and women with Rett syndrome commonly experience restricted participation in a variety of daily activities [5,6]. In particular, the majority of girls and women in this sample experienced either severely restricted levels of mobility or worse levels of mobility, so it is not surprising that wheelchairs were the most commonly used piece of equipment. Also the majority of families had accessed respite services at some point in time, but only about half had used home respite or overnight respite services in 2004. This finding is consistent with some previous work among families with a child with Rett syndrome[19] and autism [26,28], although among children with other developmental [23] and intellectual disabilities [22] the use of respite services was reportedly higher. Collectively, these findings suggest that families with a daughter with Rett syndrome require a large quantity of equipment to care for their daughter and that respite services are an important resource for some families.

In addition it was shown that a number of child and family factors may influence the use of equipment and respite services among families with a daughter with Rett syndrome. The hypothesis that greater use of equipment would be associated with more mobility restrictions was supported. This may have been because the majority of equipment used by the sample was related to functional mobility. These findings support previous research that identified that increasing levels of mobility impairment was related to an increased use of equipment in children with cerebral palsy [15]. Additionally we expected that families with a child with greater clinical severity would use more equipment. We found that clinical severity was only a significant predictor of equipment use when analysed on its own, without adjusting for mobility. This suggests that the mobility of a girl or women with Rett syndrome is a more important predictor of equipment use than clinical severity. Therefore we can expect that as girls and women with Rett syndrome experience increasing mobility restrictions, as they often do with increasing age [41,42] they will need and use more equipment, in particular mobility equipment.

It was also hypothesised that families who had a higher level of socio-economic status and lived in urban areas would use more equipment. Surprisingly this hypothesis was not supported as the majority of socio-economic measures were not related to the use of equipment. Although families with three children were less likely to use more equipment, no clear trend between the number of children in a family and the use of equipment was identified. It was also expected that families with unemployed fathers would have less income to purchase equipment and therefore use less equipment. However we found that families with unemployed fathers were more likely to use more equipment, possibly because these fathers have more time to devote to getting equipment. These findings suggest that the family's level of income, education, occupation, and accessibility to service may not necessary influence the use of equipment which is contrary to previous research suggesting that families of lower socio-economic status utilise less health services [43].

A further hypothesis was that caregivers of girls/women who were older would use more respite. It was found that caregivers of girls/women aged 13 ≤ 19 years were the most likely to use more overnight respite and caregivers of girls under the age of 8 were the least likely. These findings are consistent with previous studies conducted in children with severe intellectual disability [29] and children with autism [20,28]. Caregivers of girls under the age of 8 may be less likely to use overnight respite services because they may be less willing to have their child spend nights away from them [29]. Also caregivers of older girls may require more overnight respite services due to the presence of more behaviours and increasing mobility restrictions that typically occur with age in Rett syndrome [41,42]. In this study no relationship was identified between clinical severity and the use of respite services, contrary to previous research [21,30]. However we may expect that as girls/women with Rett syndrome age, families may require the use of more overnight respite services.

It was also hypothesised that caregivers of girls/women presenting with fewer behavioural problems were more likely to use more overnight respite services. This hypothesis was supported with caregivers of girl/women presenting with better scores on the general mood subscale and the night behaviours subscales of the Rett Syndrome Behaviour Questionnaire, being more likely to use more overnight respite services. In particular, the presence of night time behaviours such as screaming and crying for no apparent reason at night significantly reduced the likelihood of using overnight respite. Previous literature reports that the presence of challenging behaviours among children with developmental disabilities was associated with a reduced likelihood of using respite services [23] but among children with intellectual disability this relationship was not found [22]. Although the literature presents mixed findings, our results highlight the fact that families who may require more overnight respite services due to the presence of challenging behaviours are less likely to use more overnight respite services. A variety of factors may contribute to this relationship including the characteristics of the respite service and the family or caregiver. Research to explore these relationships in greater detail is necessary to

ensure that families who may require more overnight respite services due to their daughter's behaviour are able to access more of these services.

In this study we also anticipated that families that had higher levels of socio-economic status and lived in urban areas would use more respite services. Families with mothers with a vocational qualification had the highest odds of using more home respite services but the lowest odds of using more overnight respite services. However these results are hard to interpret as it is likely that a variety of other factors such as knowledge of respite services and time availability play a role in these relationships. Also if the mother was employed in full-time or part-time work the family was more likely to use more overnight respite services. This may be because mothers are typically the primary caregivers [12] so if they are busy working they may have less time to care for their child and require more frequent breaks from caregiving. Surprising no relationship between the use of respite services and accessibility was identified in contrast to previous research that reported that parents with a child with autism living in non-metro areas had lower odds of using respite services [20].

Our hypothesis that the use of equipment and respite services would have a beneficial influence on the health of caregivers was not supported. In fact the use of equipment and respite services was associated with poorer physical health. However this does not necessarily indicate that poorer physical health is a direct result of the use of equipment and respite services. Poorer physical health may result from a number of factors such as the cumulative effect of physical caring duties on the human body, therefore making it difficult to interpret the direction of the relationship between physical health and the use of resources. Although there is a clear relationship between caregiver physical health and the use of equipment and respite services, the use of these resources was not associated with caregiver mental health contrary to previous research that reported respite use to have beneficial effects on parental health [24,25] and the use of equipment to decrease the burden of care placed on caregivers [15-17]. This suggests that other factors that were not considered in analysis,

such as the use of other supportive resources, may have a greater impact on the mental health of caregivers.

This is the first known study of the relationships between specific child and family factors, the use of equipment and respite services and caregiver health in a large sample of girls and women with Rett syndrome. Although a previous study has been conducted into the health of mothers with a child with Rett syndrome, the use of resources was not considered as a contributing factor [12]. The major strength of this study is that data was obtained through a population-based registry of girls and women with Rett syndrome. Additionally the use of data from 2004 to predict the longitudinal relationship with health of female caregivers in 2006 provides a basis for establishing causal relationships [44]. Although it is unlikely that findings from this single study will result in accurate estimate of causal relationships, it does provide the basis for a series of valid studies in this area of research that collectively may infer a causal relationship in the future [45,46]. Future research should examine the causal relationships between specific child and family factors and the use of equipment and respite services and other resources that may have a greater influence of the health of caregivers of girls and women with Rett syndrome.

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Appendix A

Number of (%) child and family categorical characteristics with mean annual (SD) use of equipment and respite in 2004 (n=170).

Characteristic	N (%)	Equipment ($AUD) (n=162)	Home respite (hours) (n=164)	Overnight respite (frequency) (n=164)
Age group (yrs)				
≤ 8	37 (21.8)	357.2 (402.6)	83.7 (139.0)	1.9 (7.3)
8 ≤ 13	45 (26.5)	630.9 (722.1)	64.0 (111.0)	5.5 (10.4)
13 ≤19	56 (32.9)	609.0 (845.0)	51.8 (125.9)	10.2 (16.6)
19 <	32 (18.8)	670.7 (968.2)	118.8 (283.7)	6.7 (12.9)
Mobility				
Normal or mildly restricted	54 (31.8)	143.1 (211.4)	37.3 (69.6)	7.0 (13.5)
Severely restricted	34 (20.0)	480.7 (384.0)	102.7 (179.5)	5.5 (9.0)
Able to support weight briefly or confined to a wheelchair	27 (15.9)	806.4 (1056.8)	101.3 (281.9)	5.0 (13.5)
Totally dependent on carer	55 (32.4)	932.1 (898.6)	82.9 (141.3)	6.9 (13.9)
Number of children in family				
1	17 (10.0)	446.1 (388.0)	80.8 (124.2)	8.9 (18.0)
2	58 (34.1)	681.7 (775.0)	94.1 (229.2)	7.0 (12.9)
3	51 (30.0)	449.6 (760.8)	60.9 (94.2)	6.0 (12.9)
4	28 (16.5)	693.5 (1010.0)	83.8 (181.9)	3.1 (4.4)
5 or more	16 (9.4)	490.0 (446.4)	30.7 (66.7)	7.7 (15.4)
ARIA+ (n=165)				
Major cities of Australia	101 (61.2)	615.2 (797.2)	51.8 (86.6)	7.1 (14.0)
Inner regional Australia	37 (22.4)	543.0 (865.8)	107.8 (187.9)	6.7 (13.4)
Outer regional Australia	18 (10.9)	401.1 (455.1)	63.2 (143.4)	1.0 (1.8)
Remote or very remote Australia	9 (5.5)	622.3 (508.1)	186.5 (459.0)	5.2 (8.4)
Maternal working status (n=166)				
Unemployed	75 (45.2)	639.1 (896.4)	77.9 (189.1)	4.5 (10.3)
Employed in full-time or part-time work	91 (54.8)	512.9 (638.7)	71.5 (144.6)	7.1 (13.5)
Paternal working status (n=153)				
Unemployed	17 (11.1)	705.1 (754.5)	115.3 (347.1)	9.2 (18.0)
Employed in full-time or part-time work	136 (88.9)	576.5 (804.2)	75.6 (134.8)	7.1 (13.5)
Family income (n=165)				
Less than $20,800	29 (17.6)	458.4 (431.1)	52.9 (111.0)	10.3 (18.3)
$20,800 - $31,199	25 (15.2)	500.3 (547.2)	137.1 (321.4)	3.7 (6.3)
$32, 000 - $51,999	30 (18.2)	478.8 (569.5)	46.9 (75.6)	11.4 (17.3)
$52,000 - $77,999	25 (15.2)	874.8 (1143.8)	39.5 (64.0)	2.1 (3.8)
$78,000 or more	30 (18.2)	585.3 (697.9)	87.9 (141.8)	5.2 (10.2)
I prefer not to answer	26 (15.8)	558.6 (999.6)	100.4 (183.5)	3.0 (7.4)
Maternal education (n=166)				

Primary or some high school	52 (31.3)	482.9 (518.6)	42.9 (94.1)	8.9 (15.4)
High school year 12	34 (20.5)	431.5 (381.1)	19.1 (34.0)	3.5 (5.3)
Vocational qualification	40 (24.1)	735.1 (1123.3)	130.4 (245.2)	4.9 (12.9)
University degree	40 (24.1)	634.8 (788.1)	105.1 (181.0)	5.2 (10.3)
Paternal education (n=160)				
Primary or some high school	37 (23.1)	508.0 (554.2)	121.1 (278.3)	5.3 (10.1)
High school year 12	25 (15.6)	678.0 (729.9)	56.8 (159.5)	4.8 (11.9)
Vocational qualification	60 (37.5)	611.3 (953.0)	63.4 (104.2)	8.6 (16.4)
University degree	38 (23.8)	561.6 (738.5)	70.2 (128.7)	5.3 (10.4)
Maternal ANZSCO skill level (n=91)				
1	26 (28.6)	702.0 (933.1)	89.0 (161.2)	10.4 (17.0)
2	21 (23.1)	347.0 (271.3)	99.8 (188.4)	4.3 (9.4)
3	9 (9.9)	520.2 (360.2)	104.9 (187.1)	1.1 (1.6)
4	25 (27.5)	407.9 (553.6)	30.3 (66.5)	8.0 (13.8)
5	10 (11.0)	632.1 (589.3)	31.6 (53.0)	8.1 (15.2)
Paternal ANZSCO skill level (n=136)				
1	52 (38.2)	734.1 (963.9)	73.7 (132.4)	5.5 (11.7)
2	22 (16.2)	304.0 (334.5)	105.4 (179.1)	6.9 (11.7)
3	26 (19.1)	580.8 (1009.3)	100.1 (146.3)	7.8 (12.5)
4	20 (14.7)	634.7 (650.3)	18.4 (35.1)	10.3 (19.2)
5	16 (11.8)	361.3 (212.0)	55.6 (100.3)	1.0 (2.4)
SEIFA Index of relative disadvantage (n=158)				
Very high disadvantage	27 (17.1)	556.7 (503.4)	40.6 (70.2)	6.5 (12.8)
High disadvantage	37 (23.4)	573.8 (634.4)	107.3 (265.0)	5.9 (12.5)
Moderate disadvantage	51 (32.3)	542.0 (786.9)	76.4 (117.0)	7.4 (14.8)
Low disadvantage	43 (27.2)	713.5 (1019.8)	82.0 (154.5)	6.6 (12.5)
SEIFA index of education and occupation (n=158)				
Low levels of education and Occupation	31 (19.6)	674.9 (979.1)	83.3 (151.2)	7.3 (12.9)
Moderate levels of education and occupation	46 (29.1)	494.7 (594.9)	77.7 (130.7)	6.1 (13.9)
High levels of education and Occupation	41 (26.0)	658.6 (939.1)	45.7 (76.3)	7.4 (13.9)
Very high levels of education and occupation	40 (25.3)	568.7 (450.6)	131.8 (296.6)	5.6 (12.1)

Appendix B

The use of equipment according to child and family factors in 2004.

Characteristic (N)	OR* (95% CI)	P value
Age (155)		
≤ 8	Baseline	
8 ≤ 13	1.60 (0.55-4.64)	0.387
13 ≤19	1.71 (0.61-4.78)	0.303
19 <	2.59 (0.84-7.96)	0.097
Mobility (155)		
Normal or mildly restricted	Baseline	
Severely restricted	16.00 (1.89-135.39)	0.011
Able to support weight briefly or confined to a wheelchair	37.71 (4.47-317.90)	0.001
Totally dependent on carer	50.00 (6.39-391.40)	<0.001
Kerr scale (154)	1.20 (1.09-1.32)	<0.001
Number of children in family (155)		
1	Baseline	
2	1.12 (0.33-3.81)	0.850
3	0.29 (0.07-1.13)	0.074
4	1.47 (0.39-5.53)	0.571
5 or more	0.91 (0.20-4.10)	0.901
ARIA+ (153)		
Major cities of Australia	Baseline	
Inner regional Australia	0.51 (0.20-1.30)	0.159
Outer Regional Australia	0.56 (0.17-1.85)	0.344
Remote or very remote Australia	0.98 (0.23-4.20)	0.982
Maternal working status (152)		
Unemployed	Baseline	
Employed in full-time or part-time work	0.84 (0.42-1.70)	0.636
Paternal working status (139)		
Unemployed	Baseline	
Employed in full-time or part-time work	0.40 (0.14-1.14)	0.087
Family income (126)		
Less than $20,800	Baseline	
$20,800 - $31,199	1.01 (0.28-3.58)	0.990
$32,000 - $51,999	0.82 (0.23-2.85)	0.750
$52,000 - $79,999	1.90 (0.59-6.17)	0.283
$78,000 or more	1.71 (0.52-5.65)	0.376
Maternal education (153)		
Primary or some high school	Baseline	
High school year 12	0.85 (0.30-2.36)	0.750
Vocational qualification	1.09 (0.43-2.76)	0.859
University degree	1.32 (0.51-3.42)	0.561
Paternal education (146)		
Primary or some high school	baseline	
High school year 12	1.30 (0.42-3.99)	0.648

Vocational qualification	0.90 (0.35-2.27)	0.819
University degree	0.91 (0.33-2.52)	0.855
Maternal ANZSCO skill level (84)		
1	baseline	
2	0.80 (0.19-3.37)	0.761
3	3.00 (0.57-15.87)	0.196
4	0.79 (0.20-3.05)	0.732
5	2.40 (0.48-11.97)	0.286
Paternal ANZSCO skill level (123)		
1	baseline	
2	0.43 (0.12-1.48)	0.181
3	0.57 (0.19-1.71)	0.315
4	0.79 (0.25-2.46)	0.680
5	0.28 (0.06-1.43)	0.126
SEIFA Index of relative disadvantage (143)		
Very high disadvantage	baseline	
High disadvantage	1.37 (0.43-4.42)	0.593
Moderate disadvantage	1.10 (0.35-3.40)	0.881
Low disadvantage	1.68 (0.54-5.21)	0.369
SEIFA Index of Education and Occupation (143)		
Low levels of education and occupation	baseline	
Moderate levels of education and occupation	0.58 (0.20-1.66)	0.311
High levels of education and occupation	0.52 (0.18-1.52)	0.235
Very high levels of education and occupation	1.26 (0.45-3.53)	0.661

* The odds of using above the mean cost of equipment.

Appendix C

The use of home respite according to child and family factors in 2004.

Characteristics (N)	OR* (95% CI)	P value
Age (153)		
≤ 8	baseline	
8 ≤ 13	1.06 (0.38-2.95)	0.904
13 ≤19	0.69 (0.24-1.97)	0.491
19 <	1.50 (0.50-4.50)	0.470
Mobility (153)		
Normal or mildly restricted	baseline	
Severely restricted	2.83 (0.96-8.30)	0.058
Able to support weight briefly or confined to a wheelchair	1.70 (0.52-5.57)	0.383
Totally dependent on carer	2.21 (0.83-5.89)	0.111
Rett syndrome behaviour questionnaire		
General mood subscale (147)	0.96 (0.87-1.06)	0.480
Night behaviours subscale (152)	0.91 (0.72-1.15)	0.426
Kerr scale (153)	1.03 (0.95-1.12)	0.466
Number of children in the family (153)		
1	baseline	
2	0.36 (0.12-1.20)	0.096
3	0.43 (0.13-1.40)	0.161
4	0.34 (0.08-1.36)	0.128
5 or more	0.32 (0.06-1.60)	0.166
ARIA + (150)		
Major cities of Australia	baseline	
Inner regional Australia	1.59 (0.67-3.80)	0.293
Outer regional Australia	0.77 (0.20-2.96)	0.703
Remote or very remote Australia	0.95 (0.18-4.94)	0.954
Maternal working status (150)		
Unemployed	baseline	
Employed in full-time or part-time work	1.00 (0.48-2.11)	0.994
Paternal working status (138)		
Unemployed	baseline	
Employed in full-time or part-time work	1.54 (0.41-5.76)	0.521
Family income (126)		
Less than $20,800	baseline	
$20,800 - $31,199	2.10 (0.50-8.76)	0.309
$32,000 - $51,999	1.43 (0.35-5.80)	0.615
$52,000 - $79,999	1.05 (0.23-4.78)	0.950
$78,000 or more	2.49 (0.66-9.41)	0.180
Maternal education (151)		
Primary or some high school	baseline	
High school year 12	0.38 (0.07-1.99)	0.254
Vocational qualification	4.05 (1.45-11.36)	0.008
University degree	2.80 (0.96-8.06)	0.059

Paternal education qualification (145)		
Primary or some high school	baseline	
High school year 12	0.35 (0.08-1.48)	0.154
Vocational qualification	0.69 (0.25-1.87)	0.464
University degree	0.93 (0.32-2.72)	0.900
Maternal ANZSCO skill level (81)		
1	baseline	
2	1.75 (0.47-6.50)	0.403
3	1.00 (0.16-6.35)	1.000
4	0.53 (0.11-2.46)	0.417
5	0.75 (0.12-4.56)	0.755
Paternal ANZSCO skill level (122)		
1	baseline	
2	1.25 (0.42-3.75)	0.691
3	1.03 (0.35-3.02)	0.958
4	0.18 (0.02-1.50)	0.111
5	0.75 (0.18-3.14)	0.694
SEIFA Index of relative disadvantage (142)		
Very high disadvantage	baseline	
High disadvantage	1.90 (0.52-7.00)	0.335
Moderate disadvantage	2.01 (0.58-7.01)	0.271
Low disadvantage	1.76 (0.48-6.45)	0.394
SEFIA Index of education and occupation (142)		
Low levels of education and occupation	baseline	
Moderate levels of education and occupation	0.72 (0.24-2.13)	0.553
High levels of education and occupation	0.88 (0.29-2.64)	0.819
Very high levels of education and occupation	0.95 (0.31-2.87)	0.927

* The odds of using above the mean hours of home respite.

Appendix D

The use of overnight respite according to child and family factors in 2004.

Characteristics (N)	OR* (95% CI)	P value
Age (157)		
≤ 8	baseline	
8 ≤ 13	5.15 (1.05-25.23)	0.043
13 ≤19	9.30 (1.99-43.43)	0.005
19 <	7.37 (1.42-38.25)	0.017
Mobility (157)		
Normal or mildly restricted	baseline	
Severely restricted	0.89 (0.31-2.55)	0.828
Able to support weight briefly or confined to a wheelchair	0.53 (0.15-1.83)	0.317
Totally dependent on carer	1.03 (0.42-2.51)	0.953
Rett syndrome behaviour Questionnaire		
General mood subscale (151)	0.90 (0.81-1.00)	0.058
Night behaviours subscale (155)	0.72 (0.54-0.95)	0.021
Kerr scale (157)	1.05 (0.96-1.14)	0.297
Number of children in the family (157)		
1	baseline	
2	1.32 (0.37-4.70)	0.671
3	0.77 (0.20-2.93)	0.701
4	0.77 (0.17-3.42)	0.735
5 or more	1.20 (0.24-5.86)	0.838
ARIA (153)		
Major cities of Australia	baseline	
Inner or outer regional Australia**	0.51 (0.21-1.22)	0.131
Remote or very remote Australia	0.78 (0.15-3.99)	0.763
Maternal working status (153)		
Unemployed	1 baseline	
Employed in full-time or part-time work	2.06 (0.92-4.59)	0.079
Paternal working status (142)		
Unemployed	baseline	
Employed in full-time or part-time work	1.03 (0.31-3.38)	0.966
Family income (132)		
Less than $20,800	baseline	
$20,800 - $31,199	0.83 (0.24-2.87)	0.772
$32,000 - $51,999	1.62 (0.53-4.94)	0.399
$52,000 - $79,999	0.23 (0.04-1.20)	0.081
$78,000 or more	0.68 (0.20-2.31)	0.538
Maternal education (155)		
Primary or some high school	baseline	
High school year 12	0.54 (0.18-1.58)	0.259
Vocational qualification	0.36 (0.12-1.12)	0.077
University degree	0.78 (0.30-2.04)	0.610
Paternal education (148)		

Primary or some high school	baseline	
High school year 12	0.74 (0.19-2.89)	0.668
Vocational qualification	1.39 (0.50-3.88)	0.526
University degree	1.24 (0.40-3.81)	0.710
Maternal ANZSCO skill level (83)		
1	1 baseline	
2	0.35 (0.09-1.37)	0.131
3 or 4**	0.44 (0.14-1.44)	0.178
5	0.35 (0.06-2.01)	0.239
Paternal ANZSCO skill level (125)		
1	baseline	
2	1.42 (0.44-4.58)	0.552
3	1.90 (0.63-5.70)	0.252
4	1.58 (0.45-5.55)	0.473
5	0.29 (0.03-2.51)	0.262
SEIFA Index of relative disadvantage (145)		
Very high disadvantage	baseline	
High disadvantage	0.76 (0.23-2.47)	0.651
Moderate disadvantage	0.62 (0.20-1.95)	0.419
Low disadvantage	1.01 (0.33-3.09)	0.986
SEIFA Index of education and occupation (145)		
Low levels of education and occupation	baseline	
Moderate levels of education and occupation	1.22 (0.39-3.79)	0.728
High levels of education and occupation	0.71 (0.20-2.49)	0.593
Very high levels of education and occupation	1.83 (0.59-5.72)	0.297

*The odds of using above the mean frequency of overnight respite.

** Categories combined for analysis due to small numbers.

Appendix E

2006 Mental component summary (MCS) scores according to the use of equipment and respite in 2004, adjusted for significant child and family factors.

Factor	Unadjusted coefficient* (95% CI) P value	Adjusted coefficient** (95% CI) P value			
		Child age	Family income	Paternal ANZSCO skill level	SEIFA Index of Occupation and Education
Equipment					
Below the mean	Baseline				
Above the mean	1.84	2.28	-1.16	-0.26	1.93
	(-3.07 – 6.75)	(-2.74 - 7.30)	(-4.29 – 6.60)	(-5.92 – 5.41)	(-3.26 – 7.13)
	p=0.459	p=0.370	p=0.674	p=0.928	p=0.462
Respite					
No respite	Baseline				
Other	-4.51	-3.29	-3.03	-0.22	-4.94
	(-16.63 – 7.61)	(-15.88 – 9.30)	(-15.24 – 9.19)	(-15.56 – 15.12)	(-17.75 – 7.88)
	p=0.463	p=0.606	p=0.623	p=0.977	p=0.446
Home	-4.10	-3.82	-0.71	-8.44	-6.01
	(-11.48 – 3.27)	(-11.20 – 3.56)	(-9.03 – 7.60)	(-16.40 – -0.48)	(-14.00 – 1.98)
	p=0.273	p=0.31	p=0.864	p=0.038	p=0.138
Overnight	-5.16	-4.84	-7.36	-5.69	-8.67
	(-13.53 – 3.20)	(-13.74 – 4.05)	(-16.36 – 1.64)	(-14.96 – 3.58)	(-17.72 - 0.37)
	p=0.224	p=0.283	p=0.108	p=0.225	p=0.060
Other & home	-7.58	-7.91	-11.09	-7.97	-7.23
	(-16.91 – 1.75)	(-17.40 – 1.58)	(-21.94 – 0.23)	(-17.61 – 1.67)	(-16.91 – 2.46)
	p=0.110	p=0.101	p=0.045	p=0.104	p=0.142
Other & overnight	-2.42	-0.70	0.79	-4.02	-4.26
	(-13.03 – 8.20)	(-11.67 – 10.26)	(-10.80 – 12.38)	(16.39 – 8.35)	(-15.26 – 6.73)
	p=0.653	p=0.899	p=0.892	p=0.519	p=0.443
Home & overnight	-6.16	-4.57	-5.29	-7.15	-7.56
	(-14.20 – 1.87)	(-13.26 – 4.11)	(-14.36 – 3.78)	(-15.94 – 1.65)	(-16.03 – 0.91)
	p=0.131	p=0.299	p=0.249	p=0.110	p=0.080
Home & overnight & other	-3.93	-2.46	-5.68	-2.58	-4.76
	(-12.48 – 4.63)	(-11.56 – 6.64)	(-14.56 – 3.21)	(-11.89 – 6.72)	(-13.58 – 4.06)
	p=0.365	p=0.592	p=0.207	p=0.582	p=0.286

* The mean fitted change in MCS associated with increased use of equipment and respite.

** The mean fitted change in MCS associated with increased use of equipment and respite when adjusted for specific characteristics

Appendix F

2006 Physical component summary (PCS) scores according to the use of equipment and respite in 2004, adjusted for significant child and family factors.

Factor	Unadjusted coefficient* (95% CI) P value	Adjusted coefficient** (95% CI) P value			
		Child age	Paternal working status	Maternal working status	SEIFA Index of Occupation and Education
Equipment					
Below the mean cost	Baseline				
Above the mean cost	-4.92	-3.60	-3.98	-4.70	-5.33
	(-8.80 - -1.03)	(-7.50 - 0.31)	(-7.76 – - 0.20)	(-7.87 - - 0.27)	(-9.45 - -1.21)
	p=0.014	p=0.071	p=0.039	p=0.036	p=0.012
Respite					
No respite	Baseline				
Other	-3.03	-2.57	-0.30	-3.96	0.10
	(-12.69 - 6.63)	(-12.52 - 7.38)	(-11.64 – 11.04)	(-13.29 – 5.38)	(-9.98 – 10.19)
	p=0.535	p=0.680	p=0.958	p=0.402	p=0.984
Home	1.26	1.21	1.49	2.67	2.16
	(-4.62 – 7.13)	(-4.62 – 7.04)	(-4.21 – 7.19)	(-3.12 – 8.46)	(-4.13 – 8.45)
	p=0.673	p=0.680	p=0.606	p=0.363	p=0.497
Overnight	-8.05	-6.57	-5.81	-7.97	-10.20
	(-14.72 - -1.39)	(-13.60 – 0.46)	(-12.41 – 0.79)	(-14.38 - -1.55)	(-17.32 - -3.08)
	p=0.018	p=0.067	p=0.084	p=0.016	p=0.005
Other & home	-5.40	-4.29	-6.53	-5.44	-5.54
	(-12.84 – 2.04)	(-11.79 – 3.21)	(-13.58 – 0.53)	(-12.60 – 1.72)	(-13.17 – 2.08)
	p=0.153	p=0.259	p=0.070	p=0.135	p=0.152
Other & overnight	-6.28	-5.73	-5.05	-4.30	-6.31
	(-14.74 – 2.18)	(-14.40 – 2.93)	(-13.08 – 2.98)	(-12.26 – 4.03)	(-14.96 – 2.35)
	p=0.144	p=0.192	p=0.215	p=0.308	p=0.151
Home & overnight	-0.47	0.08	-0.76	-0.35	-1.10
	(-6.88– 5.93)	(-6.78 – 6.95)	(-6.91 – 5.39)	(-6.52 – 5.82)	(-7.76 – 5.57)
	p=0.884	p=0.981	p=0.807	p=0.911	p=0.745
Home & overnight & other	-6.60	-5.56	-6.08	-3.74	-6.84
	(-13.42 – 0.22)	(-12.76 – 1.63)	(-12.68 – 0.52)	(-10.68 – 3.19)	(-13.78 – 0.10)
	p=0.058	p=0.128	p=0.070	p=0.287	p=0.053

* The mean fitted change in PCS associated with increased use of equipment and respite.

** The mean fitted change in PCS associated with increased use of equipment and respite when adjusted for specific characteristics