An Investigation into Parental Well-Being and Child Behaviour in Phenylketonuria (PKU)

Thesis submitted in partial fulfillment of the award of

Doctorate in Clinical Psychology

South Wales Doctoral Programme in Clinical Psychology

School of Psychology

Cardiff University

By

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Supervised by Dr Dougal Hare

May 2017

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Word counts

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Full text	5,494
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Paper three: Critical review	**9,273**
Full text	8,095
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NB: All word counts exclude tables, figures and appendices.

List of tables and figures

Paper one

Paper two

An Investigation into Parental Well-Being and Child Behaviour in Phenylketonuria (PKU)

Olivia Ambler

Thesis abstract

The current thesis was submitted on the 26th May 2017 in partial fulfillment of the award of Doctor in Clinical Psychology (DClinPsy) at Cardiff University. The thesis is comprised of an abstract and three papers; paper one is a systematic review and paper two is an empirical study, both of which have been written in preparation for submission to the *Journal of Inherited Metabolic Disease*. Paper three is a critical review of the process of carrying out this work, with reflections on the challenges that arose and how these were resolved.

The systematic review aimed to identify the factors that are associated with well-being in parents who care for a child with PKU. Six electronic databases were searched (Scopus, PsycINFO, Medline, Embase, EBSCO Cinahl and Web of Science) of papers published between 1965 and November 2016. The search yielded 189 articles; 15 were included in the final review. Quality ratings revealed six studies scored within the 'moderate' range and nine within the 'high' range. Demographic variables were the most widely reported factor associated with parental well-being, as identified by seven studies. Social support was the next most reproducible factor associated with well-being, as identified by six studies. The clinical implications of these findings are discussed and recommendations are made for future research.

The aims of the empirical study were to identify what factors predict distress for parents who care for a child with PKU and to examine the incidence of behaviour problems in children with the condition. Thirty-eight parents of children and adolescents (up to and including 18 years old) with PKU and 32 parents in the general population participated in the study. Parents in both groups completed self-report measures of psychological resilience, child behaviour, perceived social support and psychological distress. Parents of children with PKU also completed measures of their child's care dependency and behaviour related to developmental or intellectual disabilities. Findings from a multiple regression analysis showed that child behaviour related to anxiety and psychological resilience predicted 35% of the variance in distress scores for parents of children with PKU, whereas child behaviour and resilience predicted 19% of the variance in distress for parents in the general population. The implications of these findings are discussed with reference to further research and clinical practice.

Paper one

A Systematic Review of the Factors Associated with Well-Being in Parents of Children with Phenylketonuria (PKU)

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This systematic review has been written in preparation for submission to the *Journal of Inherited Metabolic Disease*. Please see Appendix 7 for a copy of the author guidelines. The word count in this version has been extended to provide additional context.

Word count abstract: 250

Word count excluding abstract, tables and references: 4,573

Number of tables and figures: 4

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Abstract

Phenylketonuria (PKU) is an inherited metabolic condition that is diagnosed in infancy and requires life-long management. Parents of children with PKU are required to implement a strict diet and treatment regime during their offspring's childhood and adolescence. If this is not adhered to, children with PKU can develop intellectual disabilities and other serious medical problems.

This systematic review is the first to explore parents' experiences of caring for a child with PKU, with the aim of identifying what factors are associated with psychological well-being. Six electronic databases were searched, yielding 189 articles. Inclusion criteria were studies that used a formal measure of psychological functioning with parents of children and adolescents with PKU. Fifteen articles were selected for inclusion in the review. Quality ratings were generally good; six studies scored in the 'moderate' range and nine scored in the 'high' range. The findings revealed demographic variables were collectively the most widely reported factor associated with parental well-being, as identified by seven studies. Social support was the next most reproducible predictor of well-being, as identified by six studies. This has implications for the role of social networks in supporting families affected by PKU, particularly mothers and parents of young children. Future research should focus on building on this evidence base using longitudinal study designs and larger, more diverse samples. This may help gain a better understanding of the challenges parents face at different stages of their child's development and how to tailor support appropriately.

Keywords*: Phenylketonuria, parents, well-being, social, support, demographic.*

Synopsis: This systematic review identified that demographic information and social support were the two most widely reported factors associated with the well-being of parents who care for a child with PKU.

Introduction

Phenylketonuria (PKU) is a rare inherited genetic disorder that affects approximately 1 in every 10,000 people in the UK (Cleary, 2015). The condition is caused by a deficiency of enzyme phenylalanine hydroxylase, which is needed to metabolise amino acid phenylalanine (Phe) into tyrosine (Blau et al 2010). In the absence of this enzyme, toxic levels of Phe can accumulate in the body causing neurological damage and the development of intellectual disabilities (Blau et al 2010). As Phe is found in many protein-rich foods, children with PKU are immediately started on a low-protein diet with amino acid supplements following diagnosis (Fidika et al 2013). Parents are responsible for closely supervising their child's daily nutritional intake, attending regular clinic appointments and submitting the child's blood samples to measure Phe levels against a target range (Medford et al under review). Providing this is managed well, children with PKU can develop intellectual abilities that are within the normal range for their age (MacLeod and Ney 2010).

In light of the high responsibility placed on parents to manage the treatment regime, it is likely that caring for a child with PKU impacts on psychological well-being. Studies suggest that parents of children with PKU are more vulnerable to mental health difficulties and have poorer quality of life compared to those in the general population (Gunduz et al 2015; Mahmoudi-Gharaei et al 2011). Possible reasons for this include a sense of loss from not having given birth to a healthy child and varying

expectations around treatment, which are some of the major challenges highlighted for new parents of children with PKU (Awiszus and Unger 1990). In line with this, a significant proportion of parents report experiencing a trauma reaction upon learning their child had PKU, which can persist for years following the diagnosis (Lord et al 2005). Given the chronic nature of the condition, parents are also likely to experience recurrent concerns about their child's health and well-being (Streisand and Tercyak 2004).

To date, few studies have examined the predictors of parental well-being in PKU. Extant findings highlight that parental well-being is closely related to child age (Fidika et al 2013), with children's older age significantly predicting parents' quality of life (Ten Hoedt et al 2011). Other predictors of well-being include the parents' higher educational attainment level (Gunduz et al 2015), larger social network size (Lord et al 2005) and less difficulty meeting the child's health care needs (Waisbren et al 2004). In addition to this, some studies suggest psychological resilience, as measured by levels of personal hopefulness and resolution, predicts psychological distress in mothers of children with PKU (Lord et al 2008).

Despite the range of factors found to impact on parents' well-being, to date, the majority of meta-analyses and systematic reviews have focused on individuals with the PKU, as opposed to the wider family (Moyle et al 2007; Smith and Knowles 2000). Moreover, there is a lack of consensus regarding what helps support parents with the emotional impact of caring for a child with PKU due to the diverse findings in the current literature. Therefore, in order to best support families affected by PKU, an understanding of the factors associated with parental well-being is warranted. The

aim of this paper is to provide the first systematic review of the available evidence to identify the factors associated with well-being in parents of children and adolescents with PKU.

Method

Literature search strategy

A search was run on the 3rd November 2016 of databases Scopus, PsycINFO, Medline, Embase, EBSCO Cinahl and Web of Science. The following search terms were used (with MeSH terms included): Phenylketonuria* or PKU or Phenylalanine* or hyperphenylalaninaemia and parent* or mother* or father* or matern* or patern* or guardian* or caretaker* or caregiver* and stress* or anxi* or depress* or well*being or distress*. The initial search yielded 353 articles, which was reduced to 164 after duplicates were removed. Inclusion criteria were studies with parents or carers of children and adolescents (up to and including 18 years old) with PKU, the use of a formal psychological functioning measure and publication in a peer-reviewed, English language journal between 1965 and 3rd November 2016. Exclusion criteria were: meta-analyses; reviews; conference or dissertation abstracts; editorials; case reports and periodicals.

An overview of the search strategy is presented within the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) framework (Moher et al 2009) in Figure 1.

---------- ***Insert Figure 1 here*** ----------

The first author (OA) screened titles and abstracts and selected relevant articles (N= 32) for full text eligibility review. An independent researcher replicated this process and any discrepancies around eligibility were discussed and resolved. Inclusion and exclusion criteria were applied, resulting in the exclusion of 19 studies: eight conference and dissertation abstracts; two reviews; one article published in German language; one case study; one periodical; five studies that did not use a formal measure of psychological functioning and one study that did not include parents.

The reference lists of relevant studies were searched, resulting in two additional articles. A final 15 studies were included in the review. Due to the heterogeneity in outcome measures and the lack of randomised controlled trials represented, it was not deemed appropriate to conduct a meta-analysis (Centre for Reviews and Dissemination 2009). Therefore, a systematic review of the studies was carried out.

Quality assessment tool

Study quality was assessed using the Quality Assessment Tool for Studies with Diverse Designs (QATSDD; Sirriyeh et al 2012). The QATSDD was chosen due to its applicability to a range of research designs and methodologies and good reliability and validity (Sirriyeh et al 2012). The first author assessed the studies against a 4-point scale ranging from 0 (‘not at all’) to 3 (‘complete’) using the 14 QATSDD criteria for quantitative designs (Appendix 8). Studies were interpreted according to their quality ratings. For ease of reference, studies scoring between 0 and 14 were referred to as falling within a ‘low’ range, 15 and 28 within a ‘moderate’ range and 29 and 42 within a ‘high’ range.

An independent researcher replicated the scoring process, rating five randomly selected studies. This demonstrated moderate inter-rater reliability (k= .55), with all scores within the same rating categories.

Results

Characteristics of studies

Fifteen studies were identified through the search process, all of which utilised cross-sectional designs (see Table 1). Five studies compared a sample of parents of children with PKU to a control group only (Bosch et al 2015; Gunduz et al 2015; Jusiene and Kučinskas 2004; Kazak et al 1988a; Kazak et al 1988b), six used no control or comparison group (Fidika et al 2013; Lord et al 2005; Lord et al 2008; Mahmoudi-Gharaei et al 2011; Read 2004; Reber et al 1987), two compared the experimental group to parents of children with other genetic biochemical disorders (Read 2003; Waisbren et al 2004) and two compared the experimental group to parents of children with other neurodevelopmental and genetic biochemical disorders, in addition to controls (Kazak 1987; Ten Hoedt et al 2011). Three studies shared the same dataset (Kazak, Reber & Carter, 1988; Kazak, Reber & Snitzer, 1987; Reber, Kazak & Himmelberg, 1987). Studies were conducted in several countries including: Australia; Iran; France; Germany; Lithuania; Italy; Spain; the Netherlands; Turkey; the United Kingdom and the majority in the United States of America. Sample sizes ranged from 29 to 253, representing a total of 1,125 parents of children and adolescents with PKU. The measure of psychological functioning varied across the studies; the majority examined psychological distress, some assessed parenting stress and others examined quality of life.

---------- Insert Table 1 here ----------

Quality ratings

Quality ratings ranged from 20 to 34, representing 48% to 81% of the maximum possible score of 42. The mean score was 28.6, which represented 68% of the maximum possible score. None of the studies scored within the 'low' range, six scored within the 'moderate' range (Gunduz et al 2015; Jusiene and Kučinskas 2004; Kazak 1987; Kazak et al 1988a; Mahmoudi-Gharaei et al 2011; Read 2003) and nine within the 'high' range (Bosch et al 2015; Fidika et al 2013; Kazak et al 1988b; Lord et al 2005; Lord et al 2008; Read 2004; Ten Hoedt et al 2011; Waisbren et al 2004; Reber et al 1987). No observable relationship emerged between the year of publication and the quality of studies reviewed.

Reasons for lower ratings included limited or no justification for the choice of analytic method selected, incomplete recruitment data and little or no assessment of the quality of the measurement tools used (see Table 2). A few studies recruited large participant samples of >100 parents from a range of settings and several recruited small sample sizes of <50 parents, often from a single. Finally, only one study (Bosch et al 2015) provided evidence of user involvement in the study design and evidence that the sample size had been considered in terms of the analysis. As all the studies scored within the 'moderate' and 'high' categories, none were excluded on the basis of the quality ratings. Rather, the quality ratings were used to give due consideration to the robustness of the findings.

Research findings

Study findings were organised according to the measure of psychological well-being employed, which included quality of life, parenting stress and psychological distress (see Table 3).

---------- Insert Tables 2 and 3 here ----------

Quality of life

Quality of life was measured in four studies. Quality of life in parents of children with PKU was comparable to the general population (Bosch et al 2015) and, in some cases, superior to control groups and parents of children with other biochemical genetic disorders (Ten Hoedt et al 2011). Both of these studies used large, representative samples and scored within the 'high' quality range of the QATSDD. One study reported worse quality of life in parents of children with PKU compared to the general population (Mahmoudi-Gharaei et al 2011). However, this study used a smaller, less representative sample than the other studies, and scored within the 'moderate' quality range of the QATSDD.

Three studies examined the predictors of quality of life using regression and linear mixed model analysis. A number of psychological variables were reported in the findings. Family stress was found to significantly predict parents' quality of life in one study, which scored within the 'high' quality range of the QATSDD (Fidika et al 2013) and depression was shown to significantly predict this is in another scoring within the 'moderate' quality range (Mahmoudi-Gharaei et al 2011). However, as

neither study used a control or comparison group, it is not possible to discern whether these trends are unique to caring for a child with PKU.

Significant negative correlations were found between the psychological subscale of quality of life and stress, anxiety and depression and family history of substance misuse was significantly negatively related to total quality of life scores in one study (Mahmoudi-Gharaei et al 2011). However, 16.3% and 6.1% of the participants reported other chronic illnesses and psychological disorders within the family that were not controlled for by the researchers (Mahmoudi-Gharaei et al 2011). Therefore, these factors could have impacted on the findings reported.

Children's older age was significantly correlated with quality of life (Fidika et al 2013; Mahmoudi-Gharaei et al 2011) and this was a significant predictor of mental health-related quality of life (Ten Hoedt et al 2011). Moreover, anxiety about blood Phe levels and guilt around poor adherence to the diet regime were identified as contributing to worse quality of life for parents (Bosch et al 2015). However, this was based on mean questionnaire scores as opposed to more formal methods of statistical analysis, which limits the conclusions that can be drawn from these findings (Bosch et al 2015).

Social support predicted quality of life in two studies. Emotional support and loss of friendship were found to significantly predict mental health-related quality of life (Ten Hoedt et al 2011). Perceived social support was also shown to significantly predict parental quality of life and mediated the impact of family stress on this (Fidika

et al 2013). This trend was more pronounced for parents of younger children with PKU (Fidika et al 2013).

Parenting stress

Parenting stress was measured by four studies in total, all of which used the Parenting Stress Index. In general, parenting stress was low for parents of children with PKU, being reported as non-significantly different to a control group (Kazak et al 1988b) low in comparison to parents of children with clinically identified genetic disorders (Waisbren et al 2004) and low in comparison to mothers of children with mitochondrial diseases (Read 2003). Parents of children with PKU reported fewer worries about their child's health and future, less difficulty meeting their child's extra health care needs and a lower impact of the condition on all aspects of their personal lives compared to parents of children with mitochondrial diseases (Read 2003). However, the study participants were recruited through an online listserv (Read 2003). This could have yielded a less representative sample by recruiting parents who have previous experience of participating in studies and are therefore likely to be highly motivated.

The total number of child behaviour problems was significantly correlated with parenting stress (Reber et al 1987). However, Phe control, cognitive development and child IQ showed no significant association with parenting stress (Reber et al 1987). These findings contrast to those reported in another study, which used multiple regression analysis to identify the potential predictors of parenting stress in families affected by PKU compared to a range of other genetic biochemical disorders (Waisbren et al 2004). Child developmental level (as measured by adaptive

behaviour), satisfaction with social support and difficulty meeting the child's health care needs significantly predicted parenting stress, together explaining 51% of the variance in scores (Waisbren et al 2004). Although these studies both scored within the 'high' quality range of QATSDD, they yielded different findings regarding the demands of managing treatment for PKU and parenting stress (Reber et al 1987; Waisbren et al 2004). This discrepancy could reflect the different times that two studies were carried out, as a number of refinements have made to the low-Phe diet over the past 60 years (Singh et al 2014). Therefore, it is possible that findings from earlier studies are less sensitive to the emotional impact of the treatment for PKU, as this was less restrictive at that time (Singh et al 2014).

Psychological distress

Ten studies measuring parents' psychological distress were identified, with six measuring anxiety and depression, one assessing adjustment to the diagnosis of PKU and emotional stress, two examining trauma reactions and one assessing coping. Levels of anxiety and depression were elevated for parents of children with PKU compared to a control group (Gunduz et al 2015). However, distress levels for parents of children with PKU did not significantly differ to controls in two studies (Kazak 1987; Kazak et al 1988b). In fact, fathers of children with PKU were reported to have the lowest levels of distress across all groups (Kazak 1987; Kazak et al 1988b). It bears consideration that these two studies emanated from the same research group, which used the same research methods and outcome measures. This could account for the similar findings reported (Kazak 1987; Kazak et al 1988b).

Trauma scores were generally low across the research findings, with only 12% of mothers and 5% of fathers scoring above the clinical cut-off for trauma (Lord et al 2005). Fathers also reported better adjustment to their child's diagnosis of PKU and lower distress levels compared to mothers (Lord et al 2008). The findings highlighted numerous demographic factors associated with distress. For example, family language background, specifically when the family's first language was not English, significantly predicted trauma for fathers and distress for mothers (Lord et al 2005; Lord et al 2008).

Mothers of children with PKU had significantly higher depression and anxiety scores compared to fathers and parents in control groups (Gunduz et al 2015) and fathers' younger age significantly predicted their trauma scores (Lord et al 2005). Children's younger age was significantly correlated with distress for both parents (Kazak 1987) and approached significance as a predictor of mothers' trauma reactions (Lord et al 2005). In line with this, one study reported trauma in response to being a PKU gene carrier significantly decreased over time, which may also reflect the impact of increasing child age (Read 2004). However, by contrast one study found no significant correlations between child age and parents' anxiety and depression scores (Gunduz et al 2015). In addition to this, an ANCOVA for mothers' stress and marital satisfaction, controlling for child age, found stress levels were still significantly higher than those reported for the control group (Kazak 1987). However, both of these studies scored within the 'moderate' quality range of the QATSDD and yielded low scores for the representativeness of the participant samples (Gunduz et al 2015; Kazak 1987). By contrast, the former study scored within the 'high' quality range and used a

larger, more representative sample, which could account for the different trends reported (Read 2004).

Families who experienced financial difficulties providing low-protein products for their child had significantly higher depression and anxiety scores compared to families who experienced little or no difficulties (Gunduz et al 2015). Belonging to a less skilled occupation group was a non-significant predictor of fathers' distress, accounting for 10% of the variance in scores (Lord et al 2008). Similarly, lower academic attainment level significantly predicted anxiety and depression in parents of children with PKU (Gunduz et al 2015). In contrast to these findings, non-significant correlations were reported between parents' distress levels and a range of demographic variables, including children's age and gender, mothers' age and education and fathers' occupation (Gunduz et al 2015; Lord et al 2005; Lord et al 2008).

Aspects of parents' social support networks were associated with psychological distress. Perception of their partner being less caring, smaller social networks and less satisfaction with social support were each significantly correlated with mothers' trauma scores (Lord et al 2005). Smaller support networks and perception of their partner being less caring also significantly predicted trauma, accounting for 19% of the variance in mothers' scores (Lord et al 2005). However, as these findings emanate from the same research group, it is possible that they represent the specific expectations of the authors or funding body, leading to circular lines of investigation. Additionally, the low prevalence of PKU in the general population could give rise to the same participants being recruited for studies in this field as a result of convenience

sampling as opposed to purposeful sampling. This too, could lead to repetitive findings being reported in the research literature.

Personal stress and marital satisfaction were highly significantly negatively correlated for both mothers and fathers (Kazak 1987). One study found main effects for distress with smaller social networks and greater network density (Kazak et al 1988a). This trend was more pronounced for mothers of children with PKU than fathers, for whom there were main effects of network size but not density (Kazak et al 1988a). A possible reason for this trend is due to differences in the way mothers and fathers perceive and utilise social support (Kazak et al 1988a).

Managing the treatment demands of PKU was associated with parents' distress levels. For example, concerns about PKU, specifically around treatment adherence and the impact of PKU on the child's health and well-being, were significantly correlated with trauma scores in parents (Lord et al 2005). However, one study found no significant correlation between psychological distress and markers of treatment adherence, including child IQ, Phe control and cognitive development (Reber et al 1987). As previously stated, it is possible that these findings are influenced by changes to the low-Phe dietary guidelines over the past 60 years (Singh et al 2014).

Psychological variables included low levels of personal hopefulness and lack of resolution to the diagnosis of PKU, which significantly predicted parents' distress (Lord et al 2008). In addition, escape-avoidance coping was significantly correlated with reported distress (Lord et al 2008). Other findings show anxiety and depression were highly significantly correlated with each other (Gunduz et al 2015). Finally,

parents' emotional coping was significantly correlated with children's anxiety, depression, somatic complaints and internalising behaviours (Jusiene and Kučinskas 2004). However, this study yielded a low quality score on the QATSDD and used a stress coping strategies questionnaire as opposed to a standardised measure of distress. Therefore, the findings are not directly comparable with those reported in the other studies.

Discussion

The aim of this systematic review was to identify the factors associated with psychological well-being in parents of children with PKU. Fifteen studies measuring quality of life, parenting stress and psychological distress were examined. The findings highlighted a range of factors associated with parents' well-being, including: demographic variables; social support networks; psychological variables; child behaviour and the demands of managing treatment. Overall, the distribution of quality ratings was good. All studies were above the 'low' score category, six scored within the 'moderate' category and nine studies scored within the 'high' category.

Demographic variables were the most widely reported factor associated with parents' well-being, identified by seven of the reviewed studies. Four studies revealed demographic variables significantly predicted parental well-being and three reported statistically significant correlations. Demographic variables spanned a range of factors, including the family's language background, socioeconomic status, occupation level, gender and age. A trend emerged whereby mothers were found to be more vulnerable to distress than fathers, which may reflect their greater involvement in caring for the child with PKU (Gunduz et al 2015). There was also a trend for

increasing child age to be associated with improved parental well-being. A potential explanation for this is that parents of younger children are still adapting to the protein-restricted diet and may feel more uncertain about their child's health and development (Ten Hoedt et al 2011). These findings are supported by studies with parents of children with developmental delay and type 1 diabetes, which highlight the role of the demographic variables in parenting stress and coping (Barak-Levy and Atzaba-Poria 2013; Streisand et al 2005). However, it should be noted that three of the reviewed studies reported non-significant correlations between demographic factors and parental well-being. These inconsistent findings suggest that collectively, demographic variables may not be a robust determinant of parents' well-being, but further research is needed to clarify this.

Social support was the next most reproducible factor related to parental well-being, identified in six of the reviewed studies. Four studies used regression analysis and mixed model analysis, one used correlation analysis and one used an ANOVA. Larger, more dispersed social networks were associated with reduced psychological distress in parents, particularly mothers, and perceived social support mediated the impact of family stress on quality of life. This indicates that social support systems are protective for parents' well-being and may enhance psychological resilience. Similar findings are reported in the literature on children with long-term conditions (Horton and Wallander 2001), including cancer and congenital heart disease (Speechley and Noh 1992; Tak and McCubbin 2002). A recent study by Thomas et al (2017) also found fewer people in the social network and poor satisfaction with social support were related to worse health related quality of life in parents of children with a range of inherited metabolic conditions.

The third most reproducible factor associated with parental well-being was psychological variables. Three studies found psychological variables significantly predicted well-being and one study reported a significant correlation. Following this, child behaviour and the demands of managing the diet and treatment regime for PKU were identified by three studies in relation to parental well-being. Two studies found a significant correlation between child behaviour and parental well-being and one study reported child developmental level (as measured by adaptive behaviour) was a significant predictor of well-being. In addition to this, one study found the demands of managing treatment for PKU significantly predicted well-being, another reported a significant correlation with well-being and one study reported guilt regarding poor treatment adherence and anxiety about blood Phe levels had the greatest impact on parents' quality of life. Due to the low numbers of studies represented here, further research is needed to clarify the implications of these findings in relation to parental well-being.

Limitations

Firm conclusions cannot be drawn from the research findings due to heterogeneity in the study methodologies, outcome measures and statistical analyses used. For example, some studies examined data from parents of children with PKU only, whilst other studies pooled data from parents of children with a range of health conditions, including PKU, and compared this to controls. Overall, sample sizes were small and few studies provided detailed information on the sample characteristics. Moreover, many studies recruited families from a single clinic. This meant it was difficult to discern the representativeness of the samples included. However, it should be

acknowledged that the low prevalence of PKU limits the potential recruitment of large sample sizes with parents of children and young people. Finally, as was identified in the QATSDD ratings, very few studies considered the sample size in terms of the analysis, which may have limited the power of some statistical analyses.

Recommendations for clinical practice

There are a number of recommendations for professionals working in clinical settings based on the findings reviewed. For example, it may be beneficial for professionals to routinely assess parental well-being to identify individuals who require additional support. Health care staff should also be aware of the higher vulnerability of mothers and parents of younger children when supporting families affected by PKU. Families may benefit from support programmes that focus on empowering parents to actively seek out a range of social relationships and broaden their social networks (Fidika et al 2013). It may also be beneficial to develop interventions that are tailored to managing the specific challenges parents face at different stages of their child's development, however further research is warranted to clarify this.

Recommendations for further research

Further research in this area is needed to develop a better understanding of the different factors associated with well-being in parents of children with PKU. Studies with large, diverse sample sizes would help clarify the relationship between demographic variables and parental well-being in PKU. Longitudinal study designs would also be valuable for measuring changes to parental well-being over the course of the child's condition, in order to identify the stages where additional support is needed (Fidika et al 2013). A small number of studies identified a link between

psychological variables, child behaviour, the demands of managing treatment and parental well-being. Further research into these factors is warranted to gain a more holistic understanding of how to support families affected by PKU.

Conclusion

This systematic review was the first to examine the factors associated with well-being in parents who care for a child with PKU. In summary, demographic variables and social support were the two most widely reported factors associated with parental well-being. There was a trend for parents' well-being to improve with increasing child age and mothers were highlighted as more vulnerable to distress than fathers. This has implications for developing interventions in clinical practice that are tailored to the child's developmental level and promote social support for parents. However, further studies are needed to build on this evidence base, to overcome the methodological variability and generate further recommendations around how best to support families affected by PKU.

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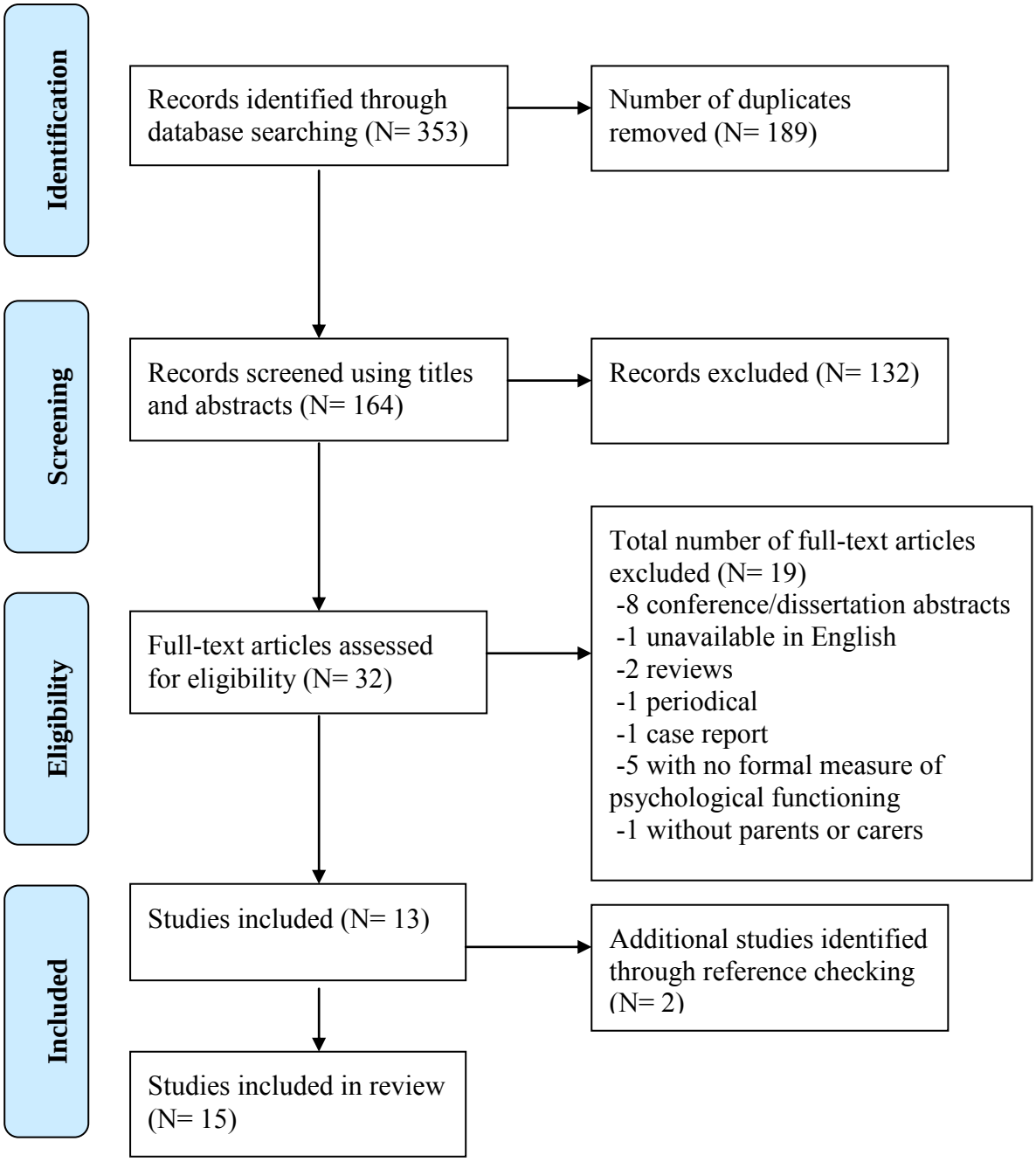

Figure 1. PRISMA diagram demonstrating the search strategy (Moher et al 2009)

Table 1. Methodological characteristics of the reviewed studies

No.	Study	Design	Comparison group	Country	Primary aim	Cohort characteristics	Total score/ quality range
1	Bosch AM, Burlina A, Cunningham A et al (2015)	Cross-sectional	Un-matched control group	7 European countries control group recruited in USA	To describe the health-related quality of individuals with PKU and their families.	N= 253 children with PKU N= 253 parents 71% of children had mild-moderate PKU, 75% had classic PKU. 27.3% of parents were male 72.3% of parents were female	**32** **High**
2	Fidika A, Salewski C, Goldbeck L (2013)	Cross-sectional	None	Germany	To describe predictors of quality of life in parents of children with PKU.	N= 89 children with PKU N=89 parents 51% of children female, 49% male. Age range of children was 0.8-19.2 years. Parent sample was 85.6% female, 14.6% male, 84.3% of parents had one child with PKU, 14.6% had more than one child with PKU, 98.9% of parents were Caucasian, 87.6% were married, 74.2% were employed, 64.1% had 8-10 years education, 35.9% had >10 years.	**31** **High**
3	Gunduz M, Arslan NM, Unal O, Cakar S, Kuyum P, Bulbul SF (2015)	Cross-sectional	Un-matched control group	Turkey	To investigate the incidence of and risk factors for depression and anxiety in parents of children with PKU.	N= 61 children with PKU N= 61 parents N= 36 control group parents Children were 1-11 years old, 32 male and 29 female. 36 healthy children in control group. Parents were 28-40 years old, 18 were male, 43 female. 7 families had more than one child with PKU.	**28** **Moderate**

No.	Study	Design	Comparison group	Country	Primary aim	Cohort characteristics	Total score/ quality range
4	Jusiene R, Kučinskas V (2004)	Cross-sectional	Matched control group	Lithuania	To investigate emotional and behavioural problems in children with PKU and parents' adjustment to the child's illness.	N= 37 parents of children with PKU N= 37 control group parents Children were 4-14 years old. 20 female, 17 male. Treatment for PKU was started early and continued without interruption. Limited demographic information provided for parents.	**20** **Moderate**
5	Kazak AE (1987)	Cross-sectional	Matched control group, parents of institutionalised individuals with 'mental retardation' and spina bifida	USA	To explore personal stress, marital satisfaction and social networks in children with disabilities.	N=43 PKU families maintained on a low Phe diet. N= 43 matched control group N= 36 'mentally retarded' group N= 46 parents of children with spina bifida Age range of PKU children= 1-8 years, mean= 3 years.	**22** **Moderate**
6	Kazak AE, Reber M, Snitzer L (1988b)	Cross-sectional	Matched control group	USA	To explore the relationships between psychological distress, parenting stress, marital satisfaction, child behaviour and family cohesion in parents of children with PKU and controls.	N=45 children with PKU N= 45 parents (both parents completed the scales) N= 49 control parents and children Children were ages 6 and younger, mean age= 3. Average length of marriage= 7.5 years, number of children= 2.1, mother's age= 28.8 years, father's age= 32.7 years, mother's education= 12.9 years, father's education= 13.1 years. Average family income= $25,575 USD	**29** **High**

No.	Study	Design	Comparison group	Country	Primary aim	Cohort characteristics	Total score/ quality range
7	Kazak AE, Reber M, Carter A (1988a)	Cross-sectional	Matched control group	USA	To explore characteristics of social networks in parents of children with PKU against a control group and to explore the relationships between distress and social networks.	N=45 children, N= 45 parents, N= 49 controls Child ages= up to 6 years old, mean age= 3. Average age of mothers= 28.8 years, fathers= 32.7 years. Average length of marriage= 7.5 years, number of children= 2.1 Mother's education= 12.9 years, father's education= 13.1 years. Average family income= $25,575 USD	**28** **Moderate**
8	Lord B, Wastell C, Ungerer J (2005)	Cross-sectional	None	Australia	To investigate parent trauma reactions to PKU and the nature of their concerns about PKU.	N= 67 parents (65 mothers, 61 fathers) Age range of mothers = 19-51 years old, fathers= 25-51 years old. Child ages= 3 months-12 years old. All children were on a treatment plan for PKU. 25% of mothers and 33% of fathers had migrated to Australia from overseas. 95% of fathers and 49% of mothers were in the paid workforce.	**34** **High**
9	Lord B, Ungerer J, Wastell C (2008)	Cross-sectional	None	Australia	To examine parents' resolution of the diagnosis of PKU, personal hopefulness, stress coping strategies and child and parent outcomes.	N= 55 children, N= 55 parents (52 mothers, 47 fathers). Child ages= 2-12 years old, parent ages= 24-51 years old. 93% of children had classic PKU. 31% mothers and 26% fathers had migrated from overseas, 94% of fathers and 56% of mothers were in the paid workforce, 60% of fathers and 50% of mothers had completed tertiary education.	**33** **High**

No.	Study	Design	Comparison group	Country	Primary aim	Cohort characteristics	Total score/ quality range
10	Mahmoudi-Gharaei J, Mostafavi S, Alirezaei N (2011)	Cross-sectional	None	Iran	To investigate quality of life, anxiety and depression in parents of children with PKU.	N=49 parents, mean age 35.63 years old (59.2% were mothers, 40.8% were fathers). Mean age of children = 9.84 years old. 51% were female, 49% male (number of children not stated). 16.3% and 6.1% of participants reported another chronic illness and severe psychiatric disorder in their first-degree family members. 22.4% of participant family members had a history of substance misuse.	**23 Moderate**
11	Read CY (2003)	Cross sectional	Mothers of children with mitochondrial disease	USA	To compare the impact of disease and dependence on health services in mothers of children with mitochondrial disease and PKU.	N= 29 mothers of children with PKU, age range 20-58 years old, 76% were married. N= 29 mothers of children with mitochondrial disease. Children age range= 0-18 years old (55% girls, 45% boys), 90% Caucasian. 57% of mothers Catholic, 29% Protestant, 14% other.	**25 Moderate**
12	Read CY (2004)	Cross-sectional	None	USA	To investigate the psychological impact of being a PKU gene carrier	N= 83 parents, mean age was 38 years old, mean time since diagnosis was 9 years (range= 1-42 years).	**34 High**

No.	Study	Design	Comparison group	Country	Primary aim	Cohort characteristics	Total score/ quality range
13	Ten Hoedt AE, Maurice-Stam H, Boelen CCA et al (2011)	Cross-sectional	Parents of children with galactosemia, un-matched control group, parents of children with lysosomal storage diseases, organic acidurias or mitochondrial respiratory chain defects.	The Netherlan-ds	To investigate health-related quality of life in parents of children with PKU and galactosemia compared to parents of healthy children and children with other metabolic diseases.	N= 116 parents of children with PKU N= 69 parents of children with galactosemia N= 434 control parents N= 108 parents of children with other metabolic diseases PKU parent sample: 43.1% male, 56.9% female, 35.3% high education level, 39.7% middle, 25% low. 94% of parents were born in the Netherlands. 7.8% of parents had another chronic disease. Child ages= 1-19 years old.	**30 High**
14	Waisbren SE, Noel K, Fahrbach K et al (2004)	Cross-sectional	Parents of children with 38 biochemical genetic disorders, including: galactosemia; arginosuccinic academia; glutaric acidemia types I and II; chain acyl-CoA dehydrogenase deficiency and maple syrup urine disease.	USA	To examine the predictors of parenting stress in parents of children with biochemical genetic disorders.	N= 112 parents (89% mothers, 10% fathers, 1% grandparents). Median family size= 2 children, 54% of children boys, 46% girls, age range= 6 months-18 years old. 91% of sample families were white, 6% Hispanic, and 2% African American and middle class. 75% were married, with a median family size of 2 children.	**30 High**
15	Reber M, Kazak AE, Himmelberg P (1987)	Cross-sectional	None	USA	To investigate the impact of caring for a chronically ill child on family functioning.	N= 41 children, < 8 years old, classic or atypical PKU, N= 41 parents Mean family income = $25,000, maternal/ paternal education = 12.9/ 13.1 years.	**30 High**

Table 2. QATSDD item scores

	Study number														
QATSDD criteria item	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**	**13**	**14**	**15**
1. Explicit theoretical framework	2	3	1	3	3	3	3	3	3	2	1	3	2	3	3
2. Statement of aims/objectives in main body of the report	3	3	3	2	3	3	3	3	3	2	3	3	2	3	3
3. Clear description of research setting	2	3	3	2	1	3	3	3	3	3	2	3	3	3	3
4. Evidence of sample size considered in terms of analysis	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0
5. Representative sample of target group of a reasonable size	3	2	2	1	1	1	1	2	2	1	1	2	3	3	1
6. Description of procedure for data collection	1	3	2	1	2	2	2	3	2	2	2	3	3	2	3
7. Rationale for choice of data collection tool(s)	3	2	3	2	3	3	3	3	3	2	2	3	3	3	3
8. Detailed recruitment data	2	3	2	1	1	2	2	2	2	1	1	3	2	2	2
9. Statistical assessment of reliability and validity of measurement tools	1	0	1	0	1	3	2	3	3	1	3	3	1	2	3
10. Fit between stated research question and method of data collection	3	3	3	2	3	3	3	3	3	3	3	2	3	2	3
11. Fit between research question and method of analysis	3	3	3	3	3	3	3	3	3	3	3	3	3	3	3
12. Good justification for analytical method selected	3	3	2	1	0	1	1	3	3	1	2	3	3	1	0
13. Evidence of user involvement in design	2	0	0	0	0	0	0	0	0	0	0	0	0	0	0
14. Strengths and limitations critically discussed	3	3	3	2	1	2	2	3	3	2	2	3	2	3	3
Total score	**32**	**31**	**28**	**20**	**22**	**29**	**28**	**34**	**33**	**23**	**25**	**34**	**30**	**30**	**30**

Table 3. Overview of the study findings

No.	Cohort size of parents	Age range of children	Measure of psychological functioning	Measurement tool	Analysis	Overview of the factors associated with psychological functioning
1	253	9-17+ years old	Quality of life	PKU-Quality of Life (PKU-QOL)	Descriptive statistics were used to document mean quality of life scores across samples. Mean domain scores were interpreted according to reference values from general population samples for PedsQL, the SF-36 and CHQ-PF28.	The highest impact scores on PKU-QOL for parents were on for the following domains: -Guilt if low-protein diet is not followed -Guilt if poor adherence to supplements -Anxiety about blood Phe levels
2	89	0.8-19.2 years old	Quality of life	Ulm Quality of Life Inventory for Parents of Chronically Ill Children	ANOVAs and Pearson's correlations were used to assess impact of socio-demographic variables on quality of life. Multiple regression analysis measured the predictors of quality of life. Simple mediation analysis to examine the relationship between social support, perceived stress and quality of life. Significance was set at $p < .05$ level.	Factors linked to quality of life: -Parent age and child age -Perceived social support -Family stress Perceived social support mediated the impact of family stress on quality of life.
3	61	1-11 years old	Anxiety and depression	Beck Depression Inventory and the State Trait Anxiety Inventory	Pearson's correlation, Student t-test and Mann-Whitney U-test were used to compare mean depression and anxiety scores. Multiple regression analysis was used to examine the predictors of anxiety and depression. Scores were interpreted at the $p < .05$ level.	Factors linked to anxiety and depression: -Parents with a lower academic attainment level -Difficulty providing low protein products -Parenting a child with 'mental retardation' -Depression and anxiety

No.	Cohort size of parents	Age range of children	Measure of psychological functioning	Measurement tool	Analysis	Overview of the factors associated with psychological functioning
4	37	4-14 years old	Coping	Coping Strategies Questionnaire	The Student t-test and Mann-Whitney U-test were used to compare mean scores on the questionnaire domains between the samples. Pearson's correlation was used to examine the relationships between scores on the coping strategies questionnaire and child behaviour checklist. Chi square analysis was used to explore the association between variables; multiple regression analysis was used to examine the predictors of psychological adjustment in children with PKU. Scores were interpreted at the $p < .05$ level.	Parents who reported feeling anger and guilt also reported significantly more child behavior problems on the following domains: -Withdrawn behaviour -Anxious or depressed behaviour -Social problems -Attention problems -Internalising behaviours -Total number of behavior problems Child anxiety, depression, somatic complaints and internalising behaviour were significantly correlated with emotional coping in parents.
5	43	1-8 years old	Anxiety and depression	The Langner Symptom Checklist	Pearson correlations and Hotelling's T were used to examine the relationships between child age, personal stress, marital satisfaction and social network variables. An ANCOVA was used to explore the relationship between mothers' stress and marital satisfaction, controlling for child age. Scores were interpreted at the $p < .05$ level.	Factors linked to anxiety, depression and personal stress: -Mothers -Marital satisfaction -Child age No significant differences between the groups were found for marital satisfaction, social network size or density or fathers' personal stress.
6	45	0-6 years old	Anxiety, depression and parenting stress.	The Langner Symptom Checklist and the Parenting Stress Index	Pearson correlations and Hotelling's T were used to compare distress, marital satisfaction, parenting stress and family cohesion. Student t-test was used to compare mean scores. Statistical significance was set at the $p < .05$ level.	No significant differences were found between groups in terms of distress, marital satisfaction, parenting stress and family cohesion. Mothers were more distress than fathers in PKU sample. PKU fathers had lowest distress levels across all groups. Parenting stress was within normal limits for all groups.

No.	Cohort size of parents	Age range of children	Measure of psychological functioning	Measurement tool	Analysis	Overview of the factors associated with psychological functioning
7	45	0-6 years old	Anxiety and depression	The Langner Symptom Checklist	Hotelling's T was used to compare social network size and density between the groups. ANOVAs compared the relationships between distress and social network characteristics. Student t-tests were used to compare group differences in social network characteristics, $p < .05$.	No differences between social network structure characteristics between PKU families and controls. Main effects found for total network size and density for mothers' distress. For fathers' distress, main effects were found for total network size but not density.
8	67	3 months -12 years old	Trauma	Impact of Events Scale	Pearson correlations were used to look at the relationships between trauma, demographic variables and concerns about PKU. Multiple regression analysis was used to examine the predictors of trauma reactions. Scores were interpreted at the $p < .05$ level for statistical significance.	Factors linked to trauma: -Concerns about PKU -Less satisfaction with social support -Perceptions of partner as less caring -Smaller social networks -Children's younger age -Severity of PKU -Fathers' younger age -Fathers' family language background
9	55	2-12 years old	Resolution/ adjustment to diagnosis of PKU & emotional stress	Interviews and the Malaise Inventory	Hierarchical regression was used to examine the predictors of distress. Student t-tests, ANOVAs, Chi square and Spearman's correlation analyses were used to compare relationships between variables. Scores were interpreted at the $p < .05$ level for statistical significance.	Factors linked to distress: -Mothers -Family language background -Low personal hopefulness -Low resolution to diagnosis -Occupation level -Escape-avoidance coping Personal hopefulness was significantly related to total child behaviour problems.

No.	Cohort size of parents	Age range of children	Measure of psychological functioning	Measurement tool	Analysis	Overview of the factors associated with psychological functioning
10	49	Not stated	Quality of life, anxiety and depression	WHOQOL-BREF and the Depression and Anxiety Stress Scale	Pearson and Spearman correlation analysis was used to compare findings between different groups. Stepwise linear regression analysis was used to examine the predictors of quality of life. Scores were interpreted at the $p < .05$ level for statistical significance.	Factors linked to lower quality of life levels: -PKU families -History of substance misuse -Child age -Depression and anxiety
11	29	0-18 years old	Parenting stress	Parenting Stress Index Short-Form	Pearson correlation analysis and two sample t-tests were used to compare scores on parenting stress, child adjustment and the demands of caring for the child between parents of children with PKU and parents of children with mitochondrial disease. Statistical significance was interpreted at the $p < .00001$ level.	Factors linked to reduced parenting stress: -Use of fewer health care services -Less worry about the child -Little difficulty meeting their child's extra care needs -Lower impact of the disease on aspects of personal life -Fewer psychoaffective and socioeconomic strains
12	83	Not stated	Trauma	Impact of Events Scale	Student's t-test was used to compare trauma scores between two time points. Paired samples t-tests were also used to compare subscale scores with the change in trauma levels over time. Statistical significance was interpreted at the $p < .01$ level.	Trauma scores around the impact of being a PKU gene carrier significantly decreased over time, particularly scores on the intrusion and avoidance subscales. This relationship was not significantly correlated with age of the parent, number of years since the diagnosis or the health or development of the child.

No.	Cohort size of parents	Age range of children	Measure of psychological functioning	Measurement tool	Analysis	Overview of the factors associated with psychological functioning
13	116	1-19 years old	Quality of life	TNO-AZL Questionnaire for Adult's Health related Quality of Life	Chi square and ANOVAs were used to test group differences on socio-demographic characteristics, with significance at $p < .01$ level. ANOVAs by group and gender were conducted to test group differences on health-related quality of life scores, with significance set at $p < .001$. Linear mixed model analysis was used to examine the predictors of mental health-related quality of life for parents of children with PKU and galactosemia. Statistical significance was set at the $p < .05$ level.	Parents of children with PKU reported higher quality of life scores than controls in 8 out of 12 of the questionnaire domains ($p<0.001$). Mental health-related quality of life was predicted by: -Children's older age -Emotional support -Loss of friendship -Perception of the disease development as stable over the past year
14	112	6 months -18 years old	Parenting stress	Parenting Stress Index-Short Form	Student t-tests were used to compare child developmental level, parenting stress, timing of diagnosis and treatment for PKU. Pearson correlations were used to measure associations between difficulties meeting the child's health care needs, social support and parenting stress. Multiple regression analysis was used to determine the predictors of parenting stress, with $p < .001$ set for statistical significance.	Parenting stress predicted by: -Child developmental level (as measured by adaptive behaviour) -Satisfaction with social support -Difficulty meeting the child's health care needs Parents of children with PKU had significantly lower levels of parenting stress, fewer difficulties meeting their child's health care needs and higher levels of social support compared to parents of clinically identified children.
15	41	<8 years old	Anxiety and depression and parenting stress.	The Langner Symptom Checklist and the Parenting Stress Index	Pearson's correlation was used to examine mothers' and fathers' scores for: distress; parenting stress; family cohesion and adaptability; marital satisfaction; child behaviour; demographic variables and child cognitive development scales. Significance was set at the $p < .05$ level.	Significant positive correlation between total child behaviour problems and parenting stress. No significant correlations between Phe control and distress, parenting stress, marital adjustment, family cohesion and adaptability. Psychological distress and parenting stress was not correlated with children's IQ, cognitive development or disease management.

Paper two

Parenting a Child with Phenylketonuria (PKU): An Investigation into the Factors that Contribute to Parents' Distress

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This paper has been written for submission to the *Journal of Inherited Metabolic Disease*. Please see Appendix 7 for the author guidelines. The word count in this version has been extended to provide additional information.

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Abstract

Phenylketonuria (PKU) is an inherited metabolic condition that can lead to the onset of intellectual disabilities if not strictly managed through a low-protein diet. Parents are responsible for supervising their child's treatment for PKU, which may impact on their experience of distress (Gunduz et al 2015). This cross sectional study aimed to identify the factors that contribute to distress in parents who care for a child with PKU, distinct from parents in the general population. Thirty-eight parents of children and adolescents with PKU and 32 parents in the general population completed questionnaires measuring psychological resilience, child behaviour problems, perceived social support and distress. Parents of children with PKU also completed measures of their child's care dependency and behaviour related to developmental and intellectual disabilities. The findings revealed no statistically significant differences in distress between the groups, but parents of children with PKU reported more child behaviour problems. Multiple regression analysis identified that psychological resilience and child anxious behaviour explained 35% of the variance in distress for parents of children with PKU. By comparison, psychological resilience and generic child behaviour only accounted for 19% of the variance in distress for parents in the general population. This has implications for developing interventions in clinical settings that aim to reduce parents' distress by enhancing their psychological resilience and supporting them to manage child behaviour difficulties, particularly anxious behaviour. Future research should include larger, more diverse samples and use longitudinal study designs.

Keywords: *Phenylketonuria, parents, child, distress, behaviour, resilience.*

Synopsis: This research study identified that psychological resilience and child anxious behaviour predict 35% of the variance in distress experienced by parents of children with PKU, whereas generic child behaviour problems and resilience account for 19% of the variance in distress for parents in the general population.

Introduction

Phenylketonuria (PKU) is a rare metabolic condition that is diagnosed in infancy and requires life-long management (Cleary 2015). The condition is caused by a deficiency or absence of phenylalanine hydroxylase, an enzyme needed to convert amino acid phenylalanine (Phe) into tyrosine (Moyle et al 2007). In the absence of this enzyme, toxic levels of Phe accumulate in the central nervous system, leading to irreversible damage to brain structures and cognitive function (Blau et al 2010). As Phe is found in protein-rich foods, the main form of treatment for PKU is a protein-restricted diet with amino acid supplements (Campbell and Ross 2003). Parents are responsible for managing their child's treatment for PKU, which involves supervising their diet, regularly meeting with health professionals and submitting blood samples to measure Phe concentrations (Campbell and Ross 2003). Due to this high level of responsibility and the adverse consequences of poor compliance, caring for a child with PKU is likely to impact on levels of psychological distress experienced by parents.

'Psychological distress' is defined as 'a state of emotional suffering characterised by symptoms of depression (lost interest; sadness; hopelessness) and anxiety (restlessness; feeling tense)' (Drapeau et al 2012; Mirowsky and Ross 2002). This concept is widely used as an indicator of mental health status in clinical, population and intervention studies (Drapeau et al 2012). Research into psychological distress

has encompassed a range of outcome measures, such as measures of stress, quality of life, well-being and depression (Massé et al 1998; Ridner 2004).

To date, few studies have examined distress in parents of children and young people with PKU. Moreover, the extant research has yielded inconsistent findings. Parents of children with PKU report higher levels of depression and anxiety compared to the general population (Mahmoudi-Gharaei et al 2011), but other studies suggest parents' well-being is comparable to control groups (Kazak et al 1987), and in some cases, superior to parents of children with other biochemical disorders (Ten Hoedt et al 2011). These inconsistent findings could reflect methodological issues such as the diverse range of outcome measures used across studies, some of which may not be sensitive to the specific impact of PKU on psychological functioning (Bosch et al 2015). To overcome this limitation, a recent large-scale international study developed a PKU-specific questionnaire measuring quality of life in patients and their parents (Bosch et al 2015). The findings revealed that mean scores on generic quality of life domains were comparable to the general population. However, parents of children with PKU reported a high emotional impact of the condition, anxiety around blood Phe concentrations and guilt if the diet was not adhered to (Bosch et al 2015).

In order to support parents with the specific challenges of caring for a child with PKU, first it is necessary to understand the different factors that contribute to distress. Findings from research into long-term conditions, such as asthma, indicate that a sense of coherence and family hardiness predicts parental well-being (Svavarsdottir et al 2000). High levels of parenting stress have also been reported in parents of young children with type 1 diabetes (Streisand et al 2005). Satisfaction with social support

was associated with parental quality of life in research on a range of inherited metabolic conditions (Thomas et al 2017). Similar findings are reported in the research literature on PKU, with younger child age (Ten Hoedt et al 2011) and levels of personal hopefulness predicting parents' distress (Lord et al 2008). Moreover, child adaptive functioning, satisfaction with social support and difficulty meeting the child's health care needs were found to account for 50% of the variance in parenting stress (Waisbren et al 2004). However, these findings are not consistent across studies, as some report non-significant relationships between demographic variables, the demands of managing treatment and parental distress (Gunduz et al 2015; Reber et al 1987). Other findings suggest that although psychological resilience is a significant predictor of parents' distress, social support and child dependency are not (Medford et al under review). A recent systematic review also identified a number of limitations in the current literature, including heterogeneity in study methodologies, small sample sizes and an overall lack of studies (Ambler and Hare in press). This highlights a need for more rigorous research to identify the factors that contribute to distress in parents of children and young people with PKU.

Another factor that could affect distress in parents of children who have long-term conditions is the presence of child emotional and behavioural problems (Craig et al 2016). Children with PKU are reported to present 1.5-1.9 times the rate of behaviour problems observed in the general population (Smith and Knowles 2000). Findings across studies indicate high rates of internalising problems, such as depression and anxiety (Jahja et al 2013), reduced social competence and lack of autonomy in this clinical population (Brumm et al 2010). Moreover, individuals with PKU were found to be twice as likely to exhibit repetitive movements (Klaverboer et al 1994) and

impulsive behaviour compared to control groups (Christ et al 2006; Smith et al 1988). This could be due to a number of reasons. Elevated Phe levels have been linked to a reduction in the concentration of neurotransmitters dopamine and serotonin, which are involved in mood regulation and executive functioning (Kolb and Whishaw 2003). High blood Phe has also been associated with increased rates of anxiety and depression in children with PKU (Ten Hoedt et al 2011) and executive functioning deficits, such as reduced inhibitory control (Albrecht et al 2009). Alternatively, other studies report more internalising problems for adolescents with good dietary adherence, suggesting that the restrictive nature of the diet itself could impact on behaviour (Cappelletti et al 2013).

However, the implications of these findings are limited due to poor experimental control in many of the studies (Christ et al 2006) and an overall lack of research in this area (Smith and Knowles 2000). Moreover, few studies have examined the impact of child behaviour on parents' distress and many of the extant findings are correlational, making it difficult to infer a causational relationship (Reber et al 1987). Therefore, in order to identify how best to support families affected by PKU, further research into child behaviour and the impact of this on parents' distress is warranted.

The aims of the current study were to: *(i)* determine whether parents of children with PKU are more vulnerable to distress compared to those in the general population *(ii)* to examine behaviour problems experienced by children with PKU in comparison to their control peers and *(iii)* to identify what factors predict distress in parents of children with PKU, distinct from parents in the general population. It was hypothesised that parents of children with PKU would report higher levels of distress

compared to parents in the general population and a higher incidence of child behaviour problems. It was also hypothesised that child behaviour problems and psychological resilience would predict distress in parents of children with PKU.

The current study was carried out in conjunction with two other research studies at the University of Manchester investigating treatment adherence, psychological well-being and parents' experiences of caring for a child with in PKU (Medford et al under review; Carpenter et al in press).

Method

Participants

Experimental group

Parents of children and adolescents were invited to participate in the study if their child was between 0 and 18 years old and had a formal diagnosis of PKU. Exclusion criteria were the presence of any other physical or mental health difficulties or significant caring responsibilities that could impact on parents' distress levels.

Control group

Parents of children and adolescents were invited to participate if their child was between 0 and 18 years old and did not have a diagnosis of PKU, or any other physical health condition. Parents were also invited to participate if they were not affected by any other significant caring responsibilities, or any physical or mental health problems that could impact on distress levels.

Measures

Demographic information

A demographic questionnaire (Medford et al under review) was used to obtain information such as parent age, child age, average family income and highest qualification (see Appendix 1 for questionnaires).

Psychological distress

The *General Health Questionnaire* (GHQ-12) was used to measure psychological distress (Goldberg and Williams 1998). Higher scores on this self-report measure indicate a greater severity of distress (anxiety and depression), with scores above 12 indicating distress within the clinical range (Goldberg et al 1997). The GHQ-12 has shown good reliability and validity (Missen et al 2012), with an internal consistency of $\alpha = 0.70$ (Kim et al 2013).

Psychological resilience

The *Resilience Scale for Adults* (RSA) is a self-report measure of psychological resilience. It has six subscales: perception of self; social competence; structured style; planned future; social resources and family cohesion (Friborg et al 2005). Higher scores indicate higher levels of resilience. Scores were summed across the subscales to provide a total score for resilience in this study. Studies show good internal consistency for total scores on the RSA ($\alpha = 0.93$; Friborg et al 2003).

Perceived social support

The *Multidimensional Scale of Perceived Social Support* (MSPSS) was used to measure levels of perceived social support (Zimet et al 1988). The MSPSS has three

subscales: significant other, friends and family. It is scored using a 1-7 point likert scale; the sum of subscale scores provides a total score for perceived social support. The MSPSS has demonstrated good psychometric properties and internal consistency (α= 0.88; Zimet et al 1988).

Child behaviour

The *Eyberg Child Behavior Inventory* (ECBI) is a parent-report questionnaire measuring disruptive behaviours in children ages 2-16 (Eyberg and Ross 1978). The ECBI has two subscales: an intensity scale to measure the frequency of behaviour problems and a problem scale to assess whether parents perceive their child's behaviour as a problem. Higher scores on the questionnaire indicate higher levels of behaviour problems, with a cut-off score of 131 for disruptive behaviour on the intensity scale and 15 on the problem scale. High internal consistency scores are reported for the intensity subscale (α= 0.94) and the problem subscale (α= 0.92; Axberg et al 2008).

The *Developmental Behaviour Checklist Parent/Carer Version* (DBC) was used to assess behaviour related to developmental disorders in families affected by PKU. This questionnaire measures the incidence of child behaviour and emotional problems over a six-month period, for children ages 4-18 (Einfeld and Tonge 1995). There are five subscales: disruptive/ antisocial; self-absorbed; communication disturbance; anxiety and social relating. A total score can also be calculated, with a clinical cut-off of 46 for behaviour relating to intellectual or developmental disabilities. The questionnaire has demonstrated good psychometric properties, with internal consistencies ranging between α= 0.66 and α= 0.91 for individual subscales (Einfeld and Tonge 2002).

Child dependency

A child dependency questionnaire was developed by Medford et al (under review) to measure how much a child depends on their caregiver to adhere to a low protein diet. Scores on this questionnaire range from 1-7, with higher scores indicating higher levels of the child's care dependency.

Procedure

Recruitment

To recruit parents of children with PKU, an advert for the study was posted on the National Society for Phenylketonuria (NSPKU) website, newsletter and online social media sites (Twitter and Facebook; see Appendix 2). The NSPKU is a UK-based third sector organisation that provides support to parents, professionals and adults affected by PKU. The researcher (OA) also attended three events organised by the NSPKU, including two conferences and a community event, to raise awareness of the study. Study adverts were displayed on the Cardiff University premises to recruit parents in the general population (Appendix 3). Any parents seeking further information were encouraged to contact the researcher using the email address displayed on study adverts.

All eligible parents were posted a questionnaire pack containing the research questionnaires, participant information sheets and a consent form (see Appendix 4 and 5). For parents in the general population, questionnaires included the GHQ-12, RSA, MSPSS and the ECBI. Parents of children with PKU were asked to complete the same questionnaires as those in the general population, with the addition of the DBC and the child dependency questionnaire. All parents were provided with a pre-

stamped envelope to return the completed questionnaires and consent forms to the researcher. An opt-out form was also provided for parents to fill in and return if they no longer wished to participate in the study. An additional questionnaire pack was sent with a reminder letter (Appendix 5.8) if the initial questionnaire pack was not responded to.

Once data collection was complete, all parents were entered into a prize draw to win a £100 shopping voucher.

Approval for the study was gained from Cardiff University School of Psychology Ethics Committee (EC.16.07.12.4554A2; see Appendix 3).

Statistical Analysis

Scores were non-normally distributed for all parent-report measures in the experimental group except for ECBI intensity subscale and RSA total score, which were normally distributed (see Appendix 6 for all SPSS output). Subscale scores for the MSPSS and the child dependency scale were positively skewed. Scores on all subscales of the DBC were negatively skewed, as were scores for the GHQ-12 and the ECBI problem subscale.

In the control group, all scores were non-normally distributed except for the ECBI intensity subscale, the RSA and the GHQ-12, which were normally distributed. Subscales of the MSPSS were all positively skewed and scores on the ECBI problem subscale were negatively skewed.

Due to the high number of non-normally distributed variables and the small sample sizes, bootstrapping approaches were used to provide a non-parametric alternative for t-tests, Pearson's correlations and multiple regressions based on 1,000 samples. This process was selected due to its higher statistical power than other approaches, particularly for studies with small sample sizes (Williams and MacKinnon 2008).

An independent samples t-test was used to compare levels of distress and child behaviour problems reported by parents of children with PKU to the general population. Pearson's correlations were used to measure the association between variables. Bonferroni corrections were not used in the analysis based on studies suggesting these are inappropriately conservative and can increase Type 2 error to unacceptable levels in studies with small samples (Bland, 1996; Nakagawa 2004).

A multiple regression analysis was selected to examine the predictors of psychological distress, as measured by scores on the GHQ-12. Predictor variables were selected based on the correlations between variables and GHQ-12 scores.

All data analysis was carried out using IBM SPSS Statistics 20 software package, with the p value set at 0.05.

Results

Participants

Experimental group

Forty-nine parents contacted the researcher regarding taking part in the study. Four parents responded to the NSPKU website advert, 18 responded to adverts posted on

social media sites and 27 following the NSPKU conferences and community event. Forty parents returned completed forms and questionnaires, with two parents opting out. Therefore, 38 eligible parents were included, representing a 78% participation rate. Thirty-four children met the age criteria for the ECBI and 28 for the DBC.

Six fathers and 32 mothers were represented. Four parents did not state their household income; the mean value for the remaining 34 parents was £56,029.41 (range= £10,000-150,000, SD= 32,976.33) and parents' mean age was 40 years and 6 months (range= 25-59, SD= 8.12). Parents reported a range of qualifications; five (14%) had a Master's degree; 17 (46%) had Undergraduate degrees; two (5%) had Doctorates; four (11%) had Diplomas; eight (22%) had GCSE's; one had A-levels (3%) and one parent did not state their highest qualification. Child mean age was 8 years and 1 month (range 1-17 years, SD= 5.36). Twenty-one (55%) of the children were female and 17 (45%) were male. The majority of families lived in the UK, with 30 (76%) from England and Wales (79%), three (8%) from Northern Ireland, one (3%) from the Republic of Ireland, three (8%) from Scotland and one family from New Zealand (3%).

Control group

Fifty-six parents contacted the researcher about taking part in the research in response to the study adverts. Thirty-two parents returned completed questionnaires, representing a 57% participation rate. All children met the age criteria for the ECBI. Thirty families met the age criteria for the ECBI.

The sample represented eight (25%) fathers and 24 (75%) mothers. The mean parent age was 38 years (range= 27-54, SD= 7.12) and the mean child age was 5 years and 6 months (range= 3 months-14 years, SD= 3.85). Eighteen (56%) of the children were male and 14 (44%) were female. Two parents did not state their average income but the mean household income for the remaining 29 parents was £73,333.33 (range= £18,000-150,000, SD= 33,777.50). A range of qualifications were represented: Twenty-two (69%) parents reported Undergraduate degrees; six (19%) had Master's degrees; two (6%) reported Diplomas; one (3%) had A-levels and one (3%) had GCSE's. Nineteen of the parents lived in Wales (59%) and 13 lived in England (41%). English was the first language for all parents in both groups.

Descriptive statistics for scores on parent-report measures

Mean scores for distress were comparable across the two groups (see Table 1), with nine parents scoring above the cut-off for distress in the experimental group and six in the control group. Scores on measures of resilience and social support were lower for parents of children with PKU compared to those in the general population. Parents of children with PKU also reported more child behaviour problems, with six parents scoring above the threshold on the ECBI intensity subscale and five on the problem subscale. By comparison, three parents from the general population scored above the threshold on the problems subscale of the ECBI. The incidence of behaviour related to developmental disorders was reported as low, with three parents scoring above the clinical cut-off for behaviour related to developmental and intellectual disabilities.

---------- ***Insert Table 1 here*** ----------

Independent samples tests for distress and child behaviour problems

An independent samples t-test with bootstrapping was carried out to compare distress and child behaviour problems between the two samples (see Appendix 6.3). Reported levels of distress, as measured by the GHQ-12, did not significantly differ across the two groups $t(68)= -.84$, $p_b= .40$. The extent to which behaviour was considered a problem also did not significantly differ between the samples, $t(64)= -1.732$, $p_b= .08$. However, the intensity of child behaviour problems approached statistical significance $t(64)= -1.911$, $p_b= .05$.

Correlations between parent-report measures and distress

Experimental group

Scores on the GHQ-12 were positively correlated with all measures of child behaviour, with a significant relationship emerging between anxious subscale of the DBC and the GHQ-12, $p< .05$ (see Table 2). A highly significant negatively correlation was found between scores on the GHQ-12 and the RSA, $p< .01$. Negative correlations emerged between the GHQ-12 and subscales of the MSPSS, except for the friend subscale, which was positively correlated with the GHQ-12. The child dependency scale scores were positively correlated with the GHQ-12, and significantly positively correlated with subscales of the DBC $p< .05$; $p< .01$.

---------- Insert Table 2 here ----------

Additional correlation analyses were run to examine the associations between child age and measures of behaviour problems (see Appendix 6.4). The findings revealed significant negative associations between child age and the self-absorbed subscale of

the DBC (Pearson's r= -.45, $p< .05$) and the intensity subscale of the ECBI (Pearson's r= -.49, $p< .05$). All other non-significant relationships were negative, except for the social-relating subscale of the DBC, which was positive (Pearson's r= .12, $p= .55$).

Control group

The GHQ-12 was negatively correlated with all subscales of the MSPSS, although these were non-significant, $p> .05$ (see Table 3). A small positive correlation emerged between the intensity subscale of the ECBI and scores on the GHQ-12, $p> .05$. By contrast, a negative correlation was found for scores on the GHQ-12 and the problem subscale of the ECBI, $p> .05$. Subscales of the MSPSS were highly significantly positively correlated with each other, indicating a degree of overlap between these constructs, $p< .01$. Scores on the RSA were positively correlated with subscales of the MSPSS, with a significant correlation found for the friend subscale. RSA scores were positively correlated with the problem subscale of the ECBI and negatively correlated with the intensity subscale, although these were non-significant, $p> .05$.

Further correlation analysis revealed a positive correlation between child age and the problem subscale of the ECBI (Pearson's r= .09, $p= .66$), indicating that behaviour difficulties in older children were considered to be more problematic for parents in this group. By contrast, a negative relationship emerged between child age and the intensity subscale of the ECBI, suggesting that younger children experience a higher incidence of behaviour problems (Pearson's r= - .24, $p= .21$).

---------- ***Insert Table 3 here*** ----------

Predictors of psychological distress

A multiple regression analysis was carried out to identify the predictors of distress in parents (Table 4). The anxiety subscale of the DBC and RSA total scores were entered into the regression model based on their significant correlations with the GHQ-12 ($p< .01$ and $p< .05$ respectively) and similar findings reported in the literature (Medford et al under review). Together, these variables explained 35% of the variance in GHQ-12 scores. RSA total scores was a significant predictor variable (Finalβ= -.45, $p< .05$), but the anxiety subscale of the DBC was not (Finalβ= .36, p= 18).

---------- ***Insert Table 4 here*** ----------

No significant correlations were found between GHQ-12 scores and scores on measures of resilience, social support and child behaviour for parents of children without PKU. Therefore, the intensity and problem subscales of the ECBI and RSA total scores were entered into the regression model to provide a control comparison model of distress to parents of children with PKU. These variables explained 19% of the variance in parents' GHQ-12 scores. However, whilst RSA total scores was a significant predictor variable (Finalβ= -.50, $p< .01$), the intensity and problem subscales of the ECBI were not (Finalβs= -.13 and -.25; p= .55 and .15 respectively).

Discussion

This study had three aims; to examine levels of parental distress, child behaviour problems and determine what factors contribute to parents' distress in families affected by PKU. Contrary to the first research hypothesis, levels of distress were

generally low for parents of children with PKU and were non-significantly different to parents in the general population. This indicates good parental well-being in this clinical population, which is consistent with studies demonstrating comparable levels of distress to control groups and parents of children with other biochemical conditions (Kazak et al 1987; Ten Hoedt et al 2011). However, contrasting findings are also reported (Gunduz et al 2015) and research suggests that many outcome measures are not sensitive to the unique stressors associated with caring for a child with PKU (Bosch et al 2015). In light of this, parental distress could have been masked by the low specificity of the GHQ-12 in the current study.

In accordance with the second hypothesis, parents of children with PKU reported a higher intensity of child behaviour problems compared to those in the general population, which approached statistical significance. Parents of children with PKU also reported more problematic behaviours than those in the general population, although this difference was non-significant. These findings are consistent with the relevant research (Smith and Knowles 2000). Interestingly, for parents of children with PKU, psychological distress was positively correlated with greater satisfaction with social support from friends. One possible explanation for this is that parents of children with PKU who seek support from other parents may draw negative comparisons between their children and feel more distressed as a result.

For parents in the general population, a trend emerged whereby parents who considered their child's behaviour to be more problematic reported lower levels of psychological distress. This could reflect high rates of support-seeking behaviour in this group or the protective value of resilience on parents' distress levels.

Although scores on measures of child behaviour were generally low, mean scores on the DBC were highest for antisocial/disruptive behaviour, indicating a relatively higher prevalence of this form of behaviour. Problems with social relating were positively correlated with child age, which is in line with research indicating lower social competence in adolescents with PKU (Brumm et al 2010). Negative trends emerged between child age and all other subscales of the behaviour measures; suggesting younger children experience a higher rate of behaviour problems generally, particularly self-absorbed behaviour. This is consistent with research demonstrating the predictive value of younger child age on parents' distress (Ten Hoedt et al 2011). Behaviour related to anxiety was significantly correlated with parents' distress, in line with the relevant research (Craig et al 2016; Jahja et al 2013).

Taken together, these findings suggest that children with PKU present with different behaviour problems at different developmental stages, which have unique implications for distress in parents. However, behaviour related to anxiety can be considered as more troublesome for parents, especially parents of younger children. Of note, no significant relationships were found between subscale scores on the ECBI and psychological distress, whereas the anxiety subscale of the DBC significantly predicted distress. This indicates that generic child behaviour scales may not be sensitive to the specific difficulties experienced by children and young people with PKU.

To address the third hypothesis, a multiple regression analysis was carried out to determine the predictors of parents' distress. Consistent with the hypothesis,

behaviour problems and psychological resilience predicted distress in parents of children with PKU, with psychological resilience as a significant predictor. These findings suggest that whilst anxious behaviour in children is associated with higher levels of distress for parents, psychological resilience is a more power predictor of this. Similar findings are reported in the literature, with research demonstrating that psychological resilience is a stronger predictor of parents' distress than social support and child care dependency (Medford et al under review). By comparison, psychological resilience was the only predictor of distress identified for parents in the general population, suggesting this trend is not unique to parents of children with PKU. However, no significant correlations emerged between child behaviour and distress for parents in this group. This suggests the association anxious child behaviour and parents' distress could be a unique trend for families affected by PKU, however further research is needed to draw firm conclusions about this.

Critical evaluation

Whilst the current study is important in adding to the evidence regarding parents' distress, there are several limitations that bear consideration. A potential constraint was the use of parent-report measures. Issues of reciprocal causation have been highlighted in the research with parents of children with health conditions (Hilliard et al 2010; King et al 1999; Shemesh et al 2005), whereby parents' own distress can influence the over-reporting of their child's distress. Other studies report high rates of child emotional and behavioural problems in families with poor psychosocial functioning, making it difficult to infer the direction of the relationship between child behaviour and parental distress (Lange et al 2005). Including an objective measure of child behaviour in the current study could have controlled for these potential biases.

The size of the sample could have limited the validity of the research findings. An a priori power calculation was conducted using G* Power, which suggested a minimum sample size of N=100 was needed for a multiple regression analysis. This indicates that the study design was underpowered to detect a significant effect where one existed. However, a range of minimum sample sizes for multiple regression are reported in the literature, with some as small as N=7, depending on the number of predictor variables (Knofczynski and Mundfrom 2007). Moreover, given the low prevalence of PKU in the general population, sample sizes in the literature have tended to be small (Ambler and Hare in press).

A further limitation was the sampling method used. It could be speculated that recruiting members of a national charity skewed the data by representing parents who are currently accessing support and therefore experience lower levels of distress. Moreover, a large proportion of the participants were recruited through NSPKU events, including two conferences and a community event, which were more likely to have been attended by highly motivated families. Furthermore, few participants from lower socioeconomic backgrounds were represented in the current study sample. This could have influenced the findings by sampling families who are exposed to fewer socioeconomic pressures, thereby representing parents who are less distressed and children who experience fewer behaviour problems.

By contrast, findings from a recent study, which recruited parents and carers through NHS Trusts in England, revealed higher mean scores for psychological distress (Medford et al under review). It could therefore be argued that recruiting through

clinical services provides a more representative sample of families affected by PKU. Moreover, it is also possible that parents who volunteered to participate in the study experienced lower levels of distress than those who did not. However, it was not possible to discern this, as reasons for non-participation were not provided.

Higher rates of inherited metabolic conditions are reported in groups with consanguineous marriages, such as in travelling communities (Thomas et al 2017). It could be speculated that families from such groups experience difficulty gaining regular access to services due to lifestyle factors and may experience higher levels of distress as a result. Moreover, there may be differences in how illness and diet are perceived in such minority groups, which could also impact on child behaviour problems and parents' distress. Therefore, future research should focus on measuring parents' distress in larger, more diverse samples to gain a deeper understanding about parents' distress in PKU.

The cross sectional design of the study may have limited the validity of the results due to the lack of control over extraneous variables. Moreover, it was not possible to infer a causational relationship based on the design of the study. By contrast, a longitudinal design could have enabled greater accuracy for observing external influences on parents' distress, such as developmental trends. This would have allowed firmer conclusions to be drawn around a causal relationship. Due to the small number of parents recruited in the current study, it could be argued that interviews may have provided more enriched data regarding the factors associated with distress. However, a relative strength in the design of the current study was the use of a control group to account for the factors associated with parenting a healthy child.

Implications for clinical practice

Based on the findings in the current study, it is likely to be beneficial if support is tailored to enhance parents' psychological resilience and the management child behaviour problems, particularly anxious behaviour. Approaches such as Acceptance and Commitment Therapy (ACT) are shown to reduce distress in parents of children with autism (Blackledge and Hayes 2006) and could help build psychological resilience in parents of children with PKU through managing negative, yet accurate thoughts about the condition. Alternatively, family-based interventions, such as systemic Family Therapy (Farrell et al 2002) and child-focused Cognitive Behaviour Therapy (CBT) with parental involvement (Barmish and Kendall 2005) could help parents to manage anxious child behaviour. However, it should be noted that few families from lower socioeconomic backgrounds were represented in the current study. This may limit the generalisability of the findings to clinical services, which are likely to be accessed by families from more diverse backgrounds. Therefore, these findings should be corroborated with clinical studies to inform further recommendations in this field.

When assessing for anxious behaviour in clinical settings, it should be borne in mind that child behaviour problems were not identified by the ECBI. This suggests routine screening using the DBC could help identify an unmet need for families affected by PKU and highlights an area for service development.

Recommendations for future research

Longitudinal studies could help identify developmental trends in children and young people with PKU and the challenges for parents associated with this. This could help

further tailor support for families. To understand the potential causes of anxious behaviour in children and young people with PKU, it would be beneficial to measure this in relation to Phe levels, parenting practices and siblings' behaviour. Future research with families from more diverse backgrounds could also help identify additional vulnerability factors for families.

Conclusion

Findings from this study suggest that levels of psychological distress are comparable between parents of children with PKU and those in the general population. However, children with PKU were reported to experience a higher incidence of behaviour problems than those in the general population. When building a model of distress for parents of children with PKU, multiple regression analysis showed anxious behaviour in children and psychological resilience in parents explained for 35% of the variance in distress. By comparison, generic child behaviour problems and psychological resilience accounted for only 19% of the variance in distress for parents in the general population. These findings build on existing research and further highlights the need to support families affected by PKU.

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Table 1 Descriptive statistics for parent-report measures for parents in the experimental group (Group 1) and control group (Group 2)

Measure	Group	N	Mean (SD)	Range (Scale range)
GHQ-12	1	38	10.29 (2.96)	4-18 (0-36)
	2	32	9.63 (3.66)	1-17 (0-36)
RSA	1	38	179.32 (24.00)	123-216 (33-231)
	2	32	191.06 (20.15)	135-220 (33-231)
MSPSS friend	1	38	21.39 (6.34)	5-28 (4-28)
	2	32	24.16 (6.00)	4-28 (4-28)
MSPSS family	1	38	22.26 (5.57)	4-28 (4-28)
	2	32	23.38 (5.75)	4-28 (4-28)
MSPSS sig. other	1	38	24.18 (5.62)	4-28 (4-28)
	2	32	24.66 (6.42)	4-28 (4-28)
MSPSS total	1	38	67.84 (14.08)	16-84 (12-84)
	2	32	72.16 (17.39)	12-84 (12-84)
ECBI intensity	1	34	103.56 (31.03)	42-168 (38-266)
	2	30	88.63 (21.48)	48-126 (38-266)
ECBI problem	1	34	6.56 (7.79)	0-29 (0-36)
	2	30	3.60 (4.46)	0-17 (0-36)
Child dependency	1	38	5.89 (1.72)	1-7 (1-7)
DBC antisocial/ disrup.	1	28	7.18 (6.86)	0-27 (0-54)
DBC self-absorbed	1	28	3.89 (4.46)	0-21 (0-62)
DBC communication	1	28	1.79 (2.44)	0-10 (0-26)
DBC anxiety	1	28	2.89 (2.87)	0-11 (0-18)
DBC social	1	28	1.54 (2.22)	0-9 (0-20)
DBC total score	1	28	18.82 (16.34)	0-60 (0-192)

Table 2. Correlations between demographic data, parent-report measures and child behaviour for parents of children with PKU

	1	2	3	4	5	6	7	8	9	10	11	12	13	14
1. MSPSS sig. other														
2. MSPSS family	.61**													
3. MSPSS friend	.49*	.30												
4. MSPSS total score	.86**	.77**	.77**											
5. ECBI problem	-.14	-.35	.24	-.08										
6. ECBI intensity	-.32	-.49*	-.08	-.36	.72**									
7. GHQ-12 total score	-.08	-.31	.07	-.12	.11	.18								
8. RSA total score	.00	.17	.03	.08	-.04	-.18	-.52**							
9. DBC disruptive	-.19	-.29	.08	-.15	.65**	.67**	.23	-.10						
10. DBC self-absorbed	-.60**	-.73**	-.35	-.68**	.55**	.73**	.11	-.10	.59**					
11. DBC communicate	-.38*	-.66**	-.17	-.48*	.37	.56**	.27	-.22	.61**	.67**				
12. DBC anxiety	-.28	-.36	-.11	-.30	.26	.38	.45*	-.19	.54**	.38	.53**			
13. DBC social relating	-.15	-.33	.20	-.10	.48*	.38	.11	.00	.71**	.45*	.62**	.56**		
14. DBC total score	-.40*	-.55**	-.09	-.41*	.59**	.71**	.28	-.15	.90**	.77**	.82**	.72**	.79**	
15. Child dependency	-.10	-.28	.11	-.10	.40*	.50**	.17	-.13	.33	.30	.28	.48**	.43*	.42*

*p< .05; **p< .01

Table 3. Correlations between demographic data and parent-report measures for parents in the general population

	1	2	3	4	5	6	7
1. MSPSS sig. other							
2. MSPSS family	.91**						
3. MSPSS friend	.87**	.84**					
4. MSPSS total	.97**	.96**	.95**				
5. ECBI problem	-.08	-.02	-.09	-.07			
6. ECBI intensity	-.24	-.22	-.21	-.24	.22		
7. GHQ-12	-.03	-.10	-.06	-.06	-.24	.02	
8. RSA total score	.28	.21	.37*	.30	.09	-.08	-.20

*p< .05; **p< .01

Table 4. Multiple regression analysis to examine the predictors of parental distress

Criterion variable: GHQ-12 total score for parents from the general population						
Enter	B	SE B	Finalβ	*p*	R^2	F change
RSA total score	-.06	.02	-.45	.04*		
DBC anxiety	.36	.23	.36	.18		
					.35	8.32**
Criterion variable: GHQ-12 total score for parents from the general population						
Enter	B	SE B	Finalβ	*p*	R^2	F change
RSA total score	-.09	.03	-.50	.01		
ECBI problem	-.02	.04	-.13	.55		
ECBI intensity	-.20	.15	-.25	.15		
					.19	3.25*

F change *p< .05, **p< .01

Paper three

Critical Review

Introduction

Evidence-based practice requires clinicians to integrate clinical knowledge and expertise with the best available research to inform their practice (Gopalakrishnan and Ganeshkumar 2013). However, the vast number of studies published across health disciplines presents a challenge for clinicians to remain informed with the most up-to-date research. Further, the varying methodological quality of published studies can make it difficult to interpret and apply the findings across clinical populations. Systematic reviews have become an increasingly important tool for overcoming these barriers, by enabling efficient access to the best quality evidence (Gopalakrishnan and Ganeshkumar 2013). By providing a synthesis of the body of evidence in the context of its scientific rigour, systematic reviews help establish the applicability of findings to different clinical populations (Mulrow 1994).

As scientist-practitioners, clinical psychologists are trained at doctorate level to be critical consumers of research and to contribute to the knowledge base through further study and evaluation (British Psychological Society 2014). Applying these research competencies is therefore integral to the professional role. Moreover, contributing to

the evidence base through further research is valuable for informing policy, service development and clinical practice across disciplines. As part of the requirements for the Doctorate in Clinical Psychology, the systematic review and empirical paper in the current thesis were carried out to offer a contribution to the field of inherited metabolic conditions.

This paper aims to provide a critical review of the research presented in the current thesis *'An investigation into parental well-being and child behaviour in Phenylketonuria (PKU)'*. The paper will be divided into three parts: part one will focus on the development and interpretation of the empirical study, part two will explore the systematic review, part three will focus on the dissemination of the findings and part four will provide my personal reflections.

Part one. Empirical study

This section will provide a reflection on the process of designing, implementing and analysing the research that was carried out for the current thesis. The rationale for the decisions that informed this work and the challenges that arose will be discussed. A critical evaluation of the research findings and their implications for theory, further research, clinical practice and service development will also be provided.

Rationale for the research topic

Prior to starting clinical training, my previous roles in the NHS involved working in medical settings with individuals who were experiencing cognitive, behavioural and physical health difficulties related to dementia and acquired/traumatic brain injury. Although the majority of my clinical work was delivered in a one-to-one format, I

was struck by the impact of these conditions on the whole family system. For example, families were often required to adjust to new treatment regimes, significant lifestyle changes and had to cope with the medical uncertainties associated with their loved one's condition. In addition to this, I observed the ways in which family functioning could in turn impact on the person. From these experiences, I developed an interest in the way that services are configured and the potential missed opportunity to work alongside families and other members of the system, following the onset of a serious physical or mental health problem.

During clinical psychology training, I have been eager to pursue my interest in systemic working as much of my previous experience has involved working with individuals. Therefore, when developing a topic for my thesis, I was motivated to carry out research in this area. In addition to this, much of my previous research experience had involved carrying out qualitative methods. Hence, discussions with my appraisal tutor highlighted competency needs in quantitative design, methodology and analysis and working with families.

Initial considerations

An initial idea I had for my research topic was to investigate the needs of families following the onset of maternal post-partum psychosis. I was particularly interested in a potential unmet need amongst fathers. Research in this area might have produced recommendations for clinical practice and service development. However, following a meeting with a consultant psychiatrist working in the field and correspondence with a national third sector organisation, it transpired that a similar research project was already being conducted with fathers at that time. I reflected on the potential practical

challenges this could bring in discussions with a member of the DClinPsy programme staff. This highlighted that, given the small sample population, recent involvement in a similar study could negatively impact on recruitment due to 'participant burnout'. I therefore discussed alternative research opportunities with the consultant psychiatrist working in the field, but these were focused on the individual as opposed to the wider system. Hence, I chose not to pursue that research topic for the thesis. Instead, I carried out scoping searches of the literature on psychological functioning in families affected by physical health conditions. This revealed a heightened vulnerability to distress in parents of children and adolescents across a range of long-term conditions, such as neurodevelopmental disorders and type 1 diabetes (Craig et al 2016; Driscoll et al 2010). Several studies also demonstrated a link between parental well-being and the physical and psychological functioning of children. For example, family-based interventions were shown to reduce parenting stress and improve physical health outcomes in children and young people with chronic pain and diabetes (Eccleston et al 2003; Wysocki et al 2005). This highlighted the potential value of investigating parental well-being in supporting children and young people with physical health conditions.

When considering this research topic I also consulted the relevant policy. The link between parental well-being and children's emotional and physical development was widely documented, as was the need to support the whole family in services for children and young people (Public Health Wales 2013; Royal College of Paediatricians and Child Health Wales 2016). In addition to this, the NHS Outcomes Framework for 2016-2017 outlines that improving health-related quality of life for individuals and their carers is a key indicator for families affected by long-term

conditions (Department of Health 2016). With these policies and with my competency needs in mind, I decided to carry out my research in the field of long-term conditions.

Rationale for studying parental well-being in PKU

When examining research conducted across different health conditions, it was evident that only a small number of family-based studies had been carried out in inherited metabolic conditions (Read 2003; Siddiq 2016; Ten Hoedt et al 2011). I therefore discussed potential opportunities for carrying out research in this field with a member of staff on the DClinPsy programme who had clinical and research expertise in this area. From these discussions and my own additional reading, I became aware of the increased vulnerability to distress in parents who have a high level of responsibility for managing their child's condition (Cousino and Hazen 2013). Moreover, I learned about the high onus placed on parents of children with PKU to manage their child's daily diet and treatment regime (Gunduz et al 2015). In particular, I was struck by the need for parents to gain control over their child's Phe levels imminently following diagnosis, to adapt family routines accordingly and cope the adverse risk of cognitive impairment if treatment was not strictly adhered to. This resonated with me as growing up a close family friend was affected by cystic fibrosis and I observed the ways that her parents adapted to cope with the demands of the treatment on their everyday lives. Therefore, I felt there was value in examining the different factors that affect parental well-being in families affected by PKU.

UK guidelines for the management of PKU highlight the need for multidisciplinary working with children, young people and their parents. For example, it is recommended that clinical psychologists 'assist patient and parent understanding of the need for dietary treatment' and 'foster parent and patient motivation to comply

with treatment' (NSPKU 2014). European guidelines also make recommendations for routine assessment of psychosocial functioning at distinct stages of the young person's development, to reflect challenges such as the transition of treatment management from parents to patients (van Spronsen et al 2017).

With these different factors in mind, I decided that further research into the well-being of parents who care for a child with PKU was important for informing clinical practice, service development and potentially contributing to future policy in PKU. I therefore chose to study this topic for my doctorate research.

Developing the research design

Given the paucity of studies investigating parental well-being in PKU, I consulted the relevant evidence for other long-term conditions. A paper by King et al (1999) described a model of well-being for parents of children with disabilities. The factors that were shown to be associated with reduced parental distress included: lower levels of child disability; family-centred caregiving; social support; effective coping strategies; fewer child behaviour problems and higher satisfaction with support from services (King et al 1999). Findings from this research showed that family-centred caregiving, social support and child behaviour problems each significantly predicted parental well-being, with child behaviour problems as the strongest predictor (King et al 1999). I also researched other models of parental well-being in the literature, such as the risk and resilience model (Wallander et al 1989). This model suggests that 'risk factors', such as psychosocial stress and child disability, can be ameliorated by 'resistance factors', such as social support and psychological resilience (Wallander et al 1989). Moreover, the systematic review I was simultaneously working on (see Part

two) highlighted a need for further research into child behaviour problems, psychological factors (such as resilience) and the demands of managing treatment for PKU. I was therefore motivated to investigate support within the social network, child behaviour difficulties, psychological resilience and the child's level of dependency on their parent, in relation to parental well-being for the empirical study.

Discussions with my research supervisor highlighted a similar study that was being conducted as part of the clinical psychology doctoral programme at the University of Manchester. I subsequently met with two trainee clinical psychologists in Manchester to discuss this work, which included a qualitative study investigating parents' experiences of caring for a child with PKU and a quantitative study measuring parenting stress, treatment adherence and psychological well-being in maternal carers of children and adolescents with PKU. Given the overlap between these projects and my own in terms of studying parental well-being, we met with their respective supervisors and discussed the potential value of collaborating on this work.

A component of the quantitative study involved analysing parenting stress, psychological resilience, the child's care dependency and social support using multiple regression analysis, to identify the potential predictors of parental well-being. The data for this project had already been collected and we discussed the option of my building on this work by measuring child behaviour problems. The aim would be to extend the regression model to gain a broader understanding of the predictors of parental well-being in PKU. We also discussed the option of my administering the same parent-report measures (i.e. parenting stress, psychological resilience, child dependency and social support) and a measure of child behaviour

with a control group. The aim of this would be to identify the factors that predict parental well-being in families affected by PKU, distinct from families in the general population.

As the data for the parent-report measures (except child behaviour) had already been collected for the Manchester-based research, we agreed to submit an amendment to NHS ethics to enable me to access this data. I planned to contact the families from the database to ask them to fill in additional measures of child behaviour and recruit a control group independently of this. However, when I contacted the University of Manchester and Cardiff University ethics departments to discuss this amendment, a number of challenges were highlighted to me. For example, the participants who had been recruited for the existing research had not provided informed consent to share their data or to be contacted about future research. This precluded me from accessing the existing database or contacting these parents about my additional research without approval via a new NHS ethics application.

It was also highlighted to me that the two trainee clinical psychologists would need to obtain written approval from the three NHS Research and Development departments involved in the recruitment of parents for their research. Unfortunately, the process of ascertaining whether or not I could submit an amendment had taken until June 2016 and the trainee clinical psychologists were due to graduate from the University of Manchester doctoral programme in September. I felt that this was not enough time to process a new NHS ethics application and receive approval from the three NHS Research and Development departments. I therefore decided to recruit participants independently of the Manchester-based projects.

When collating the questionnaires for the study, I chose to use the same measures of demographic information, psychological resilience, child dependency, social support and psychological distress as were used in the Manchester-based research, as these were well-validated questionnaires which addressed the aims of my study. Moreover, using these questionnaires would enable the findings from my study to be compared with the existing Manchester-based research. When deciding on a measure of child behaviour, I chose to use the Eyberg Child Behavior Inventory as was a well-validated measure of behaviour problems in the general population, enabling me to use this scale with both the experimental and control groups. In view of the limited research into child behaviour difficulties in PKU to date (Smith and Knowles 2000), I also decided to administer an additional measure of behaviour related to developmental and intellectual disabilities in the PKU sample, to identify any behaviour difficulties related to the condition itself.

I discussed the option of excluding the measure of parenting stress (the Pediatric Inventory for Parents) with my research supervisor, as this did not align with my reading of the relevant literature on parental well-being in PKU. Moreover, I was wary of the potential overlap between parenting stress, the child's care dependency and psychological distress. Therefore, we decided to exclude this measure from the study design.

Recruitment

As I planned to carry out a multiple regression analysis, I aimed to recruit a minimum of 40 participants in the experimental group and 40 in the control group, based on a

rule-of-thumb indicating 10 participants are needed per predictor variable to ensure adequate statistical power (Field 2009). However, given the low prevalence of PKU in the general population (Cleary 2015), I was wary of the potential challenges associated with recruiting a sample of this size. I approached local clinicians to discuss recruiting families through NHS settings in south Wales. Unfortunately, the clinicians working in this area were unable to collaborate on the project due to competing service demands at that time. Therefore, my supervisor and I discussed alternative methods of recruitment and the associated strengths and limitations. We decided that recruiting through national charity could pose some challenges, such as potentially accessing a smaller, more homogenous sample. However, a potential benefit was reaching families from a wider geographical area who may also be more motivated to participate in research.

I therefore approached the National Society for Phenylketonuria (NSPKU) about recruiting through the charity and we agreed that I could recruit parents of children with PKU through adverts on the charity website and the online social media sites. I gained ethical approval from Cardiff University to proceed with this and to recruit parents from the general population through adverts posted on the Cardiff University premises.

After several months, I had recruited a number of parents in the general population but very few parents of children with PKU. One of my hypotheses about this trend was that parents of children with PKU receive a lot of information about charity events, conferences and workshops run by the NSPKU and therefore may not attend to the study advert. A further hypothesis was that parents of children with PKU might

be less motivated to participate in the research given that there was no immediate benefit to themselves or their family. I also wondered whether high levels of stress in parents of children with PKU could mean they lack the time or energy to participate in research studies.

To discuss ways of recruiting more families for the study, I had several email conversations with the staff from the NSPKU. One option that was discussed was attending events organised by the society. I felt that this would enable me to talk to families face-to-face about the research, my rationale for doing it and the potential benefits for the PKU community. As the study relied on parents' willingness to give up their personal time with no direct benefit to themselves, I also thought that these discussions might increase their interest in contributing to the research. Therefore, before attending these events, I constructed a brief summary of the study and my rationale for carrying this out, to convey this in conversation with families. I also submitted an amendment to my ethics application enabling me to hand out questionnaire packs at these events, should families express an interest in completing these.

Although these changes enabled me to recruit more participants, I was still significantly below the target number of families by December 2016. I therefore thought about other ways of recruiting parents, such as utilising resources within the NSPKU staff team. Initiating more discussions with NSPKU staff members led to greater activity on social media sites, with more staff posting about the study. This helped increase awareness of my research within the PKU community and had a knock-on effect on the number of parents volunteering to participate. In addition to

this, I arranged to attend a national three-day conference organised by the NSPKU to present a poster of my provisional research findings (see Appendix 9). I felt that this offered a valuable opportunity to give something back to the society and allowed me to discuss my research with the conference attendees. Several families volunteered to participate in my research during the conference. It is possible that talking with families face-to-face and showing some of the emerging findings of my study encouraged them to volunteer, as did the multiple other research presentations that took place over the weekend.

Overall, I was able to recruit 38 participants in the experimental group and 32 in the control group. Although small, this sample size was deemed sufficient to run my statistical analyses based on the literature (Knofczynski and Mundfrom 2007). On reflection, I could have attended more events held by the NSPKU and other organisations to present my research, which could have increased participation rates in the study. I could also have tried to recruit families through NHS trusts in the south west of England. This may have enabled me to access a wider range of participants, potentially yielding a more representative sample. Any future studies should take this into consideration.

Critical evaluation of the empirical study findings

Overall, the findings from the empirical paper highlighted a higher incidence of behaviour problems reported by parents who care for a child with PKU compared to parents in the general population. Child behaviour difficulties and lower levels of perceived social support from family predicted distress in parents of children with PKU, a trend that was not found for parents in the general population. Although these

findings are consistent with the relevant literature (Fidika et al 2013), there were some unexpected results and limitations of the findings that warrant further discussion.

The findings revealed a non-significant difference in the reported intensity of child behaviour problems between parents of children with PKU and parents in the general population sample. However, the extent to which parents considered their child's behaviour to be problematic did significantly differ between the two samples, with parents of children with PKU reporting more problematic behaviour. One possible explanation for this finding might be due to a difference in the way parents interpret and label their child's behaviour. Parents of children with PKU might interpret behaviour changes as being the result of their child's condition and this being problematic. They may also be more vigilant of changes in their child's behaviour, given that this could be related to elevated Phe levels (Smith and Knowles 2000).

For families in the general population sample, a negative correlation emerged between parental distress and reported problematic behaviour in children. This indicates that parents who considered their child's behaviour to be more problematic were, in fact, less distressed. This could reflect a higher incidence of help-seeking behaviour for parents in this group or the protective impact of psychological resilience. However, this trend is counterintuitive to what might have been expected and could reflect an anomaly in the findings, especially given the small sample size of this group.

By contrast, psychological distress was correlated with all measures of child behaviour for parents of children with PKU, suggesting more child behaviour

difficulties could lead to higher levels of distress in parents in this sample. However, as these findings were correlational, an alternative explanation could be that parents' own distress impacts on their perceptions of their child's behaviour as problematic. In line with this, findings from studies with parents of children with type 1 diabetes suggest higher levels of stress and anxiety are associated parents' perceptions of their child's behaviour as being more problematic (Hilliard et al 2010).

Overall, the empirical findings indicated that the younger the child, the higher the number of behaviour problems that were identified on the parent-report measures. This could be explained by the relative inexperience of parents in managing the condition and the overall greater demands for care earlier in children's lives. However, an exception to this concerned social relating difficulties and the extent to which parents perceived their child's behaviour as problematic, as reflected in the social relating subscale of the DBC and problem subscale of the ECBI. Both of these variables were correlated with children's older age. One potential explanation for these trends is the implications of the treatment regime for adolescents with PKU. Adolescence is often associated with striving for autonomy and independence from authority figures (Spear and Kulbok 2004). This conflicts with the close involvement of health professionals and parents in managing the treatment and diet regime in PKU (Smith & Knowles 2000). Furthermore, adolescents with PKU have been shown to experience low autonomy and poor integration with their peer group due to the need to adhere to a strict diet (Brumm et al. 2010; Weglage et al. 1992). These difficulties could therefore account for the higher rates of social relating problems reported by parents of older children in the study and the extent to which parents perceived these behaviours as problematic. However, a similar relationship emerged between

increasing child age and parent reports of problematic behaviour in the control group, suggesting that this trend reflects challenges associated with normal development.

To assess the ecological validity of my findings and their relevance to clinical practice, I met with a clinical psychologist with experience of working in the field of inherited metabolic conditions. This highlighted a different demographic of families presenting to services compared to those represented in the empirical study. In particular, a higher incidence of families from lower socioeconomic backgrounds, and many from travelling communities, were presenting to paediatric health services. This indicates that the sample in the empirical study may not be representative of the PKU population in the UK, possibly due to the method of recruitment. Therefore, it is possible that a sampling bias in the current study skewed the data towards families from higher socioeconomic backgrounds, who are arguably less distressed (Mistry et al 2002). However, sampling through a clinical service could also introduce a selection bias in accessing a disproportionately distressed sample, as families who are coping well might not attend clinical appointments. A more accurate sampling strategy in future might be recruiting through a PKU case register.

Implications for theory and further research

The final regression model revealed perceived social support from family, behaviour related to anxiety, psychological resilience, disruptive/antisocial behaviour and the total number of behaviour problems related to developmental and intellectual disabilities, predicted psychological distress in parents of children with PKU. These findings are consistent with the relevant literature (Fidika et al 2013) and other models of well-being in parents of children with long-term conditions (King et al

1999). However, to date, few studies have measured the impact of child behaviour problems on parental well-being in PKU. Therefore, it could be argued that these findings offer a unique contribution to the field of inherited metabolic conditions by highlighting the impact of perceived child behaviour problems on parental well-being, informing further research in this area.

Future research could corroborate the empirical findings in the current thesis with other family-based studies in PKU. With further research in this area, it may also be beneficial to construct a conceptual model of parental well-being in PKU, for example using structural equation modeling. This could help develop an understanding of the interactions between child behaviour, distress, coping and caregiving in families and provide a framework for future lines of enquiry. In addition to this, the impact of support from third-sector organisations and NHS services could be measured and incorporated into a theoretical model of parental well-being. Given the limited literature on psychological interventions with this clinical population, future studies could also incorporate practice-based evidence to build on the existing knowledge base through publishing service-based research.

Investigation of why some children with PKU present with behaviour problems was beyond the scope of this study. However, possible explanations include the impact of elevated Phe levels, poor treatment adherence, cultural and family values around diet and illness or parenting practices. For example, children with PKU could present with more rigid, obsessive behaviour because this is how they have learned to manage the strict nature of the diet. Future studies could help identify the possible causes of behaviour problems in PKU, for example by measuring the behaviour of siblings

without the condition. Furthermore, involving families from diverse backgrounds, such as travelling communities, in future research could help identify potential vulnerability factors in relation to child behaviour problems, as higher rates of inherited metabolic conditions are reported in communities with consanguineous marriages (Thomas et al 2017).

Implications for clinical practice

The findings in the empirical study have a number of implications for clinical psychologists working in NHS services. As there are multiple potential explanations for why children might present with behaviour difficulties, this highlights the need for a detailed assessment and formulation of the family's needs, incorporating the biological, psychological and social factors that could impact on the child's behaviour. Gaining a more in-depth understanding of the family's needs would enable interventions to be tailored more appropriately, which would arguably be more effective than offering a 'one-size-fits' all approach, such as a behavioural parent training programmes (McCart et al 2006). Further, by understanding the factors that contribute to a family's distress, clinical psychologists can work at several levels of the system, as opposed to offering one-to-one interventions only (Barmish and Kendall 2005; Brestan and Eyberg 2010). This work could involve offering consultation to nursery staff, health visitors, GPs and pharmacists who supply low-protein products to families on prescription. Similarly, working alongside school staff could help develop creative ways of adapting the child's diet within the school environment.

Implications for professional practice and service development

As providing detailed assessment, formulation and interventions at multiple levels of the system are part of the core competencies of a clinical psychologist, this highlights the benefits of increasing the number of clinical psychologists in services working with children and young people affected by rare conditions, such as PKU. However, given the current climate of austerity within the NHS, clinical psychologists could also be positioned further up-stream within services, to influence preventative work with children and families. For example, providing psychoeducation, consultation and training to staff in community programmes such as Flying Start could help build psychological resilience and strengthen social support within families, especially those from lower socioeconomic backgrounds who are arguably more vulnerable to distress (Mistry 2002). Working alongside multi-disciplinary agencies aligns with current guidelines for the management of PKU, which emphasise the role of a clinical psychologist in liaising with other services 'regarding aspects of learning and behaviour that might be related to PKU or its treatment' (NSPKU 2014). This form of preventative working could in turn reduce the volume of referrals to secondary care and specialist health services in the long-term, in line with the principles of prudent healthcare (Welsh Government 2015). Given the research competencies of clinical psychologists, they could also be positioned to involve families in service-based research to help identify gaps in the current system and inform the development of more family-centred services. This would align with findings in the literature demonstrating a link between satisfaction with support from services and better parental well-being in long-term conditions (King et al 1999).

However, the potential barriers to implementing this work should also be considered. Discussions with a clinical psychologist working in the field highlighted that due to the low prevalence of PKU, it is difficult to gain the recognition and funding required for developing services. Such barriers could be addressed and overcome at a political level. Interestingly, Rare Disease UK, a campaign run by Genetic Alliance UK, has worked with health departments to raise awareness of the challenges facing families affected by rare diseases. Similarly, the NSPKU is hosting an event at the Houses of Parliament in June 2017 as an opportunity to increase public understanding of the condition. Activities such as these are therefore essential to improve the care and well-being of children affected by PKU and their families.

Part two. Systematic review

The following section will focus on the stages of conducting the systematic review, including: the rationale for the review topic, the process of developing the search strategy and the application of the quality assessment tool. A discussion of the challenges that arose when carrying out this work will be provided with an explanation of how these difficulties were resolved. A critical evaluation of the systematic review findings and the wider implications for future research, clinical practice and service development will also be discussed.

Rationale for choosing the systematic review topic

In choosing a topic for the systematic review, it was important to consider the relevance and potential value of this work for service users, carers and families. It was also important to consider how the work would contribute to clinical practice, service development and the wider evidence base. I gathered ideas for the systematic review

topic by examining the existing research into PKU. Initial scoping searches failed to identify any randomised controlled trials, meta-analyses or evaluation studies of family-based psychological interventions in PKU. No systematic reviews emerged of studies that investigated the psychological impact of PKU on the wider family. I therefore consulted the wider literature on long-term conditions in children and young people. I was interested in the documented links between parental well-being and children's physical and mental health outcomes in other research studies, such as in diabetes (Wysocki et al 2005). This highlighted the importance of supporting parents when working with families in clinical settings. A small number of studies in the literature on PKU investigated parental well-being, however many of the findings were inconsistent across studies. Therefore, to inform ways of supporting families affected by PKU, I chose to conduct a systematic review of the factors associated with parental well-being.

In choosing this systematic review topic, I hoped to *(i)* identify the evidence regarding what factors are helpful for supporting parental well-being; *(ii)* highlight areas warranting further investigation and *(iii)* make recommendations for developing family-based interventions in clinical practice. As no systematic reviews on this topic were identified, a further aim was to address this gap in the research literature.

Developing the search terms

Given the small body of research in the area, I revised the systematic review title several times through discussions with my research supervisor, in order to access a broad range of studies. Examples of early drafts include 'A review of the predictors of distress in parents of children with PKU'. However, this was revised to 'The factors

associated with well-being in parents of children with PKU' in order to include more diverse study methodologies and statistical analyses, such as ANOVAs and correlation analysis. The key search terms were developed using synonyms, which I expanded using MeSh terms via the online databases. This enabled me to select the most appropriate combination of search terms to access the relevant literature. I also searched several different online databases to maximise the identification of potentially relevant studies published across disciplines.

As only 15 studies were included in the final review, on reflection I could have broadened the search terms further. Scoping searches identified numerous studies published on other metabolic conditions, such as galactosemia and type 1 diabetes. Broadening my search terms to include these conditions might have enabled a wider body of research to be reviewed. Moreover, more in-depth recommendations for clinical practice and further research with families affected by PKU could have been generated based on these findings. However, due to the different trajectories of these conditions and their different treatment modalities (Ten Hoedt et al 2011), it would have potentially been difficult to interpret the research findings specifically for families affected by PKU. Furthermore, given the limited data on parental well-being in PKU, I decided that carrying out a systematic review in this field would help synthesise the extant findings and make clear recommendations for further lines of enquiry.

Another potential limitation of the search strategy was the inclusion of full-text articles published in peer-reviewed journals only. This could have excluded the most up-to-date evidence in the 'grey literature', such as conference abstracts and

dissertations. However, it is also the case that including a vast range of studies, abstracts and reports could have negatively impacted on the quality of the findings reviewed. This may have resulted in less valid conclusions and recommendations for clinical practice. Finally, including data from interviews and case studies could have highlighted additional relevant themes to the findings in the systematic review. However, given that few relevant qualitative studies were identified in the initial literature searches, I decided that it would be more rigorous to collate and synthesise findings from a single research modality. Hence, only quantitative studies were included in the review.

Quality assessment tool

The Quality Assessment Tool for Studies with Diverse Designs (QATSDD) was used to assess the studies included in the review. I chose to use this tool based on its applicability to studies with diverse methodologies and good validity and reliability (Sirriyeh et al 2012). However, numerous challenges arose when I was applying this tool to the studies in the systematic review. Namely, the lack of examples provided meant that the interpretation of the individual items could be somewhat subjective. For example, it was unclear whether item 2 'Statement of aims/ objectives in main body of report' was applicable to papers that did not explicitly state aims and objectives but provided comparable information, such as detailed research hypotheses. Similarly, item 5 'Representative sample of target group of a reasonable size' could also have been interpreted in a number of ways, such as in the context of PKU only, which, given its low prevalence (Cleary 2015), is a small population for researchers to draw from. Alternatively, the sample size could have been rated against the statistical power and type of analysis used in the study. Moreover, the

'representativeness' of study samples could have represented a number of constructs, such as having an equal number of men and women, participants from different ethnic backgrounds, or children and young people with different classifications of PKU. As no published demographic norms for this clinical population were available, it was not possible to compare the samples to a 'representative' standard.

To resolve the dilemmas that arose around the interpretation of the QATSDD, I had several conversations with my research supervisor to gain a shared understanding of the items and their applicability to the individual studies. It was also helpful to meet with other colleagues who had previously used the tool and discuss similar dilemmas and how these were resolved. Given the complexities of interpreting the items, I also consulted published reviews of the QATSDD. Interestingly, similar concerns were raised regarding items 2 and 5, including the broad definitions of the constructs and their subjectivity (Fenton et al 2015). Moreover, a general limitation of the tool was the equal weighting assigned to different items despite their varying implications for the overall study quality (Fenton et al 2015). Therefore, an independent rater reviewed five randomly selected studies to establish inter-rater reliability using the tool.

On reflection, it might have been useful to pilot a selection of quality assessment tools during an early phase of the systematic review. This would have allowed me to critically review and compare different tools, considering the potential limitations of each, hence, enabling a more informed decision about the most appropriate tool to use in the systematic review. To my knowledge, there are no quality assessment tools that are designed specifically for research into PKU. Therefore, I might also have

designed a new tool to assess the quality of family-based studies in PKU. However, based on the moderate level of inter-rater reliability with the independent rater (k= .55), the QATSDD was deemed appropriate for use in the systematic review.

Critical evaluation of the systematic review findings

Socioeconomic status

The findings from the systematic review highlighted that demographic variables were the most widely reported factor associated with parental well-being in PKU. A more disadvantaged socioeconomic background, lower education level and belonging to a less skilled occupation group were found to be linked to parental distress (Gunduz et al 2015; Lord et al 2008). These findings are consistent with the diabetes literature, with lower socioeconomic status linked to higher levels of distress in parents (Streisand et al 2005). However, similar findings are also reported in the general population, with studies suggesting that parents and children from more disadvantaged socioeconomic backgrounds are more likely to report unhealthy behaviours and poor psychological well-being (Huurre et al 2003). Similarly, high rates of behaviour problems and intellectual disabilities are reported in studies with children and young people from more disadvantaged socioeconomic backgrounds (Morgan et al 2009). Lower level of academic attainment, feelings of not belonging in school and higher rates of distress were also found for adolescents and young adults in this group (Langhout et al 2009; Mistry et al 2009). Based on this research, it is possible that the findings in the systematic review reflect a trend present in the general population. However, as many of the reviewed studies did not employ a control group, it is not possible to determine whether this trend is unique to parents of children with PKU or is applicable more broadly.

Alternatively, findings in the literature indicate that distressed parents from more disadvantaged socioeconomic backgrounds report feeling less capable and effective in their parenting practices (Mistry et al 2002). This might lead some parents to over-report distress when participating in family-based research, due to negative bias when appraising their own parenting abilities.

Language and cultural factors

Individuals whose first language was not English were found to have lower reported well-being in the systematic review (Lord et al 2005; Lord et al 2008). However, given that a proportion of the participants had immigrated from overseas, it is possible that these findings reflect other stressors, such as those associated with emigrating to a new country or integrating into the new community (Lord et al 2005; Lord et al 2008). Furthermore, different cultural perceptions of disease and illness might have influenced the results reported in the studies, as these were not controlled for. Trauma was found to be significantly associated with perceptions of the partner being less caring, smaller social networks and less satisfaction with social support in one of the studies (Lord et al 2005). Based on this trend, another interpretation of the findings is that family language background presents a barrier to accessing support. However, due to the lack of an experimental control in this research, it was not possible to discern in what way language background impacted on parental well-being.

Gender differences

A further demographic factor identified in the systematic review was mothers' higher vulnerability to distress (Gunduz et al 2015; Kazak et al 1988; Lord et al 2008). This finding could reflect gender differences in the experience or the reporting of distress

in the general population. For example, a study of 2,816 adults found women reported significantly more stress symptoms and daily stressors than men (Matud 2004). Moreover, despite a comparable number of life events experienced over the previous two years, women rated these events as being more negative and less controllable compared to men (Matud 2004). Therefore, it is possible that a similar gender difference in the reporting of distress was represented in the findings reviewed. Other findings in the literature highlight different coping styles in men and women. For example, studies indicate women are more likely to seek social support and use emotion-focused coping compared to men (Ptacek et al 1994). By contrast, men are more likely to use problem-focused coping in response to stress (Ptacek et al 1994). These different coping styles suggest women may be more likely to self-report feeling distressed than men, which too could contribute to the gender differences observed in the systematic review findings. Societal views of men as 'rational', 'independent' and 'instrumental', and women as 'emotional' and 'supportive' (Williams and Best 1982) may have introduced demand characteristics in self-reports of distress in the reviewed research.

Child age

Children's older age was another demographic variable highlighted as contributing to parental well-being in the systematic review findings (Ten Hoedt et al 2011). There are several possible explanations for this. As children grow older, they are likely to gain more autonomy and independence in managing their diet and lifestyle. For parents, this shift in responsibility may alleviate some of the stress associated with the strict diet and treatment regime for PKU (Ten Hoedt et al 2011; Lord et al 2005). In addition to this, as more time passes, parents are likely to adjust to the distress, and in

some cases the trauma, associated with receiving the diagnosis of PKU (Read 2004). They are also more likely to develop confidence around managing the diet and build larger support networks, which is shown to have a positive impact on well-being (Lord et al 2005). Alternatively, these findings could also be indicative of the factors associated with increasing age in 'healthy' children, such as higher levels of independence and maturity. However, without a normative sample or comparison group, it is not possible to control for this trend in the findings reviewed.

Social support

Social support was highlighted as the next most reproducible factor associated with parental well-being in the systematic review. In particular, larger, more dispersed social networks were associated with an increased sense of well-being in parents and perceived social support was found to buffer the impact of stress on parents' quality of life. This suggests that social support could enhance parents' psychological resilience and protect against the distress associated with caring for a child with PKU. This finding is consistent with much of research on families affected by other conditions, such as neurodevelopmental disorders (Craig et al 2016). However, due to the cross-sectional design of the study, it is difficult to determine the direction of the relationship between distress and social support. Moreover, other studies have found positive parental coping predicted better family functioning and satisfaction with social support (King et al 1999). This indicates that parents who are less distressed are more likely to access and benefit from support within their family and social network.

Overall limitations of the systematic review findings

A general limitation of the studies included in the systematic review is that many did not use a matched control group. Therefore, it was not possible to control for the factors associated with distress related to caring for a child without a long-term condition. Furthermore, many of the studies did not measure demographic variables, such as religion and ethnicity, which could impact on perceptions of illness, distress and help-seeking behaviour. Reasons for participant attrition rates were reported in some of the reviewed studies. However, the majority of the studies provided no explanation for drop-outs or non-participation. Therefore, a sampling bias could be speculated, whereby families who had a greater sense of well-being were more likely to volunteer to participate in the research. Moreover, some of the sampling methods (e.g. recruiting through a listserv) may have yielded a less representative sample, by recruiting families who are likely to have previous experience in participating in studies and are therefore highly motivated (Read 2003). Based on these limitations, the findings of the current systematic review should be interpreted with caution.

Recommendations for further research

It may be beneficial for future research in this field to include wider, more diverse samples. For example, it would be beneficial to investigate the impact of cultural perceptions of illness and help-seeking on parental well-being and family functioning. Greater experimental control over demographic variables, such as the child's classification of PKU number of siblings with the condition could also lead to a better understanding of the factors that impact on parental well-being, as these were not controlled for in many of the reviewed studies. The use of more sophisticated forms of analysis, such as mediation and regression analysis and the inclusion of a control

group would also enable more firm conclusions to be drawn around the factors important for well-being in parents of children with PKU.

Given the demonstrated link between lower socioeconomic status and psychological distress, it may be valuable for future studies to investigate the resources available for families living in areas of social deprivation. For example, statistics on the family's access to and use of professional support, other resources available within the community and access to third sector organisations may help identify families who are at greater risk of distress. Given the link between perceived social support and parental well-being, it may also be useful to investigate the benefit of different forms of social support. Many psychometric measures do not assess support from online organisations, charities, or other parents who care for a child with a serious health condition. Further research into this could generate recommendations for interventions such as peer-mentoring schemes or conferences and social events held by organisations, such as the National Society for Phenylketonuria (NSPKU).

Few qualitative studies were identified in the initial scoping searches, as such it may be beneficial for future research to include interview data to address this gap in the literature. Furthermore, all of the studies included in the systematic review utilised cross-sectional designs. Longitudinal research would help determine a more causal relationship between demographic variables and parental well-being and could help identify useful forms of support for families at different stages of the child's development.

Implications for service development

Given the paucity of family-based intervention studies represented in the current literature for PKU, practice-based research could offer a valuable contribution to the evidence base. This could be beneficial for representing more complex difficulties facing families and the experience of health care professionals. Service audits and evaluations could also help identify unmet needs for families and locate where in the system clinical psychologists could have the greatest impact for reducing distress. Involving service users and staff in research promotes the principles of co-production and could help shape services to meet the needs of children and families.

Implications for clinical practice

The demonstrated link between lower socioeconomic status and psychological distress in parents of children with PKU could reflect several constraints on the family's ability to gain the support they need to manage their child's condition. This highlights the importance of reaching families from more socially disadvantaged backgrounds who have limited access to resources. Clinical psychologists have a role in widening access to psychological support for hard-to-reach communities. This could be achieved through providing supervision, consultation and training to staff across settings, such as GPs in primary care, school staff and health visitors. A potential aim of work could be to build competences in assessing for psychological distress, identifying high-risk families and referring for appropriate psychological support.

Similarly, given the identified positive relationships between well-being, social support and children's older age, clinical psychologists could work alongside third

sector organisations, such as the NSPKU, to co-produce parenting support groups in the community. This work could draw from evidence-based interventions such as Family-Based Cognitive Behaviour Therapy, which is associated with reduced anxiety in children and young people aged 7-18 (Barmish and Kendall 2005).

Part three. Dissemination of the findings

A number of steps were taken to disseminate the findings in the current thesis to professionals working in the field and families affected by PKU. A summary of the findings will be sent to participants who have requested this. An overview of the study and the key findings will also published in the NSPKU newsletter, which is circulated to society members. I have presented a research poster of the initial findings from the study at the NSPKU annual conference in April 2017 and have liaised with the staff about presenting the completed research at the next annual conference in 2018, which is attended by families and professionals working in the field.

I am submitting the systematic review and empirical paper to the Journal of Inherited Metabolic Disease for publication. I chose this international medical journal based on its high impact factor (3.541) and wide audience, which includes nurses, dieticians, clinical psychologists and medical professionals who are likely to have contact with families affected by PKU in clinical settings.

In addition to this, to ensure the findings are disseminated more widely to clinical psychologists working in a range of settings, I have submitted a poster presentation of

the study to the Division of Clinical Psychology conference, which is scheduled for January 2018.

I have also discussed the findings of the research and their implications for services with three clinical psychologists working in the field of child health and inherited metabolic conditions in Cardiff, Manchester and Birmingham. I hope that these varying means of dissemination will enable the findings to impact on different levels of the system, including parents who care for a child with PKU, professionals working in the area and service managers.

Part four. Reflection

Recruitment to the study proved a challenging component of completing the thesis. On reflection, I think the study would have benefitted from being co-designed with a small group of parent volunteers. This may have helped recruit larger numbers of participants during the initial months. A potential barrier to recruitment could have been the length of time required to complete the study questionnaires. A co-design process might have helped produce a more concise set of questionnaires. Despite this, I was struck by parents' generosity for giving their time to the research without any immediate or guaranteed result.

Balancing the demands of the thesis with my clinical placement during the final year of the doctorate programme required me to prioritise my workload, have good organisational skills and time management skills. Moreover, working alongside the NSPKU staff to engage participants prompted me to be resourceful in my methods of recruiting parents. It was clear to me that written adverts would not be sufficient to

recruit the numbers I needed for this research to be viable and personal attendance, meeting and speaking with parents was more effective. This is something I will carry with me into my qualified role if I conduct service-based research that requires recruitment of participants, such as staff or service users.

Considering working in a qualified clinical role, I am aware of the need to balance competing demands and prioritise my time effectively. Moreover, there may be challenges in initiating research within services due to resistance, for example, from staff teams. I feel that the experience of carrying out this research has prepared me well to manage these challenges and think creatively about ways of working alongside staff and other stakeholders in clinical settings. Similarly, the engagement skills I developed to recruit participants for the research are likely to be useful when working engaging service users in assessment and therapeutic interventions, particularly if they are ambivalent.

Carrying out this research has also made me reflect on the value of understanding carers' experiences and considering ways of building resilience when supporting the whole family system. Moreover, this has made me reflect on past clinical experiences, such as working with families affected by dementia and acute neurological injuries, strengthening my belief in the value of working with the family system.

The process of doing this research has prompted me reflect on the challenges ahead regarding how to secure protected research time within a busy NHS environment. This is important because one of the unique roles of a clinical psychologist is contribution to and drawing from the evidence base to ensure that interventions

offered are likely to be the most effective to service users and families. This is particularly relevant during the current period of public sector austerity when financial constraints require prudent use of resources.

Finally, I feel that completing this work has increased my confidence in using quantitative research methodology, prior to starting the course I had experience of using qualitative approaches with service users only. I feel this experience has rounded my competence and skills in both approaches and I plan to take this forward in adopting an evaluative approach to clinical work in the future.

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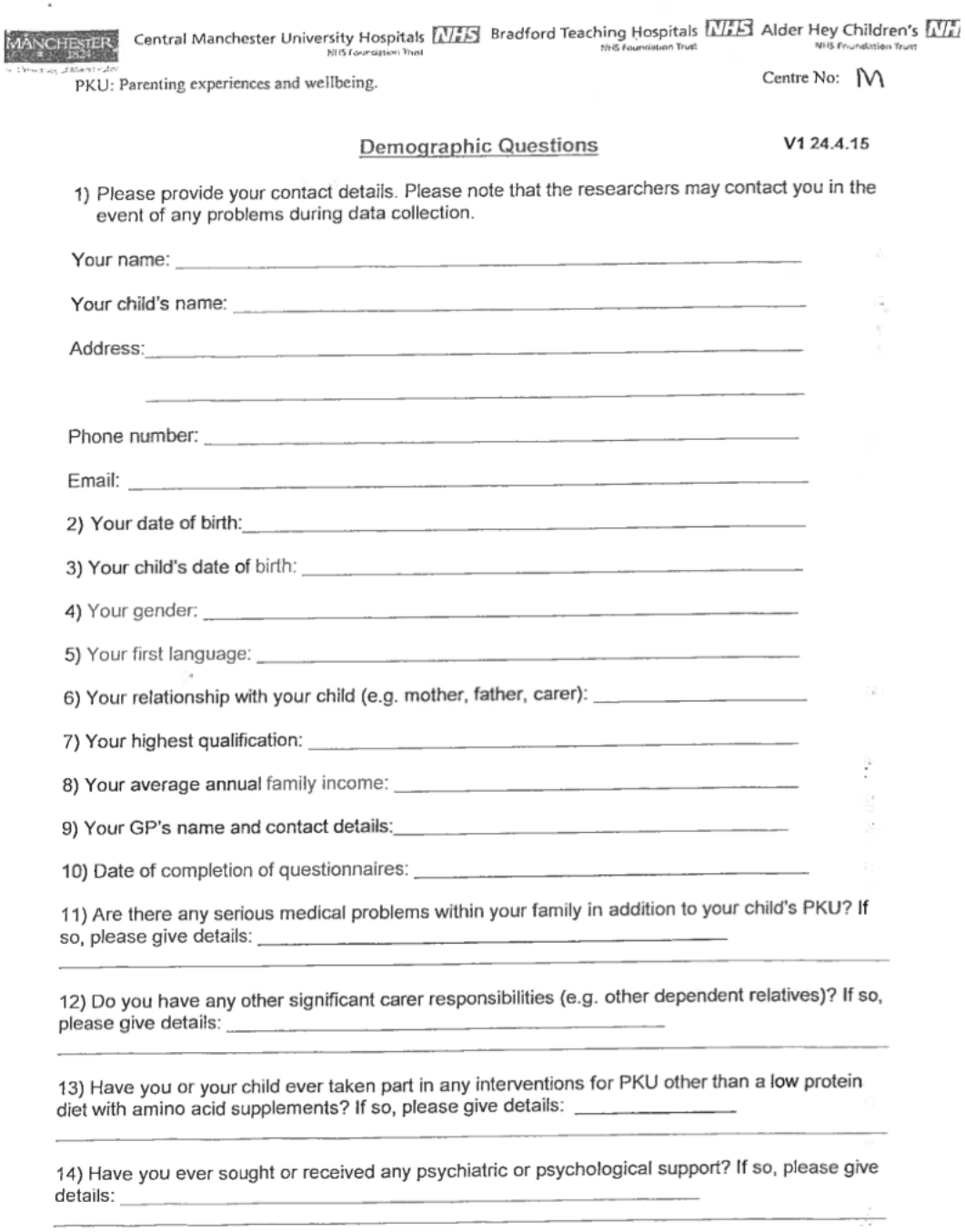
Appendices

Appendix 1: Research questionnaires

1.1 Demographics Questionnaire



Central Manchester University Hospitals NHS Bradford Teaching Hospitals NHS Alder Hey Children's NHS

PKU: Parenting experiences and wellbeing. Centre No: M

Demographic Questions V1 24.4.15

1) Please provide your contact details. Please note that the researchers may contact you in the event of any problems during data collection.

Your name: _____

Your child's name: _____

Address: _____

_____

Phone number: _____

Email: _____

2) Your date of birth: _____

3) Your child's date of birth: _____

4) Your gender: _____

5) Your first language: _____

6) Your relationship with your child (e.g. mother, father, carer): _____

7) Your highest qualification: _____

8) Your average annual family income: _____

9) Your GP's name and contact details: _____

10) Date of completion of questionnaires: _____

11) Are there any serious medical problems within your family in addition to your child's PKU? If so, please give details: _____

_____

12) Do you have any other significant carer responsibilities (e.g. other dependent relatives)? If so, please give details: _____

_____

13) Have you or your child ever taken part in any interventions for PKU other than a low protein diet with amino acid supplements? If so, please give details: _____

_____

14) Have you ever sought or received any psychiatric or psychological support? If so, please give details: _____

_____

1.2 The General Health Questionnaire-12

GENERAL HEALTH QUESTIONNAIRE

GHQ-12

Please read this carefully:

We should like to know if you have had any medical complaints, and how your health has been in general, *over the past few weeks*. Please answer ALL the questions simply by underlining the answer which you think most nearly applies to you. Remember that we want to know about present and recent complaints, not those you had in the past. It is important that you try to answer ALL the questions.

Thank you very much for your co-operation.

HAVE YOU RECENTLY:

1	**been able to concentrate on whatever you're doing?**	Better than usual	Same as usual	Less than usual	Much less than usual
2	**lost much sleep over worry?**	Not at all	No more than usual	Rather more than usual	Much more than usual
3	**felt that you are playing a useful part in things?**	More so than usual	Same as usual	Less useful than usual	Much less useful
4	**felt capable of making decisions about things?**	More so than usual	Same as usual	Less so than usual	Much less capable
5	**felt constantly under strain?**	Not at all	No more than usual	Rather more than usual	Much more than usual
6	**felt you couldn't overcome your difficulties?**	Not at all	No more than usual	Rather more than usual	Much more than usual
7	**been able to enjoy your normal day-to-day activities?**	More so than usual	Same as usual	Less so than usual	Much less than usual
8	**been able to face up to your problems?**	More so than usual	Same as usual	Less able than usual	Much less able
9	**been feeling unhappy and depressed?**	Not at all	No more than usual	Rather more than usual	Much more than usual
10	**been losing confidence in yourself?**	Not at all	No more than usual	Rather more than usual	Much more than usual
11	**been thinking of yourself as a worthless person?**	Not at all	No more than usual	Rather more than usual	Much more than usual
12	**been feeling reasonably happy, all things considered?**	More so than usual	About same as usual	Less so than usual	Much less than usual

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1.3 The Resilience Scale for Adults (page 1)

M

Resilience Scale for Adults

Please think of how you usually are, or how you have been the last month, how you think and feel about yourself, and about important people surrounding you. Please check the option box that is closest to the end statement that describes you best.

Name: _____ Todays date: _____

Age: _____ Gender: _____ female/male

1. When something unforeseen happens	I often feel bewildered	☐ ☐ ☐ ☐ ☐ ☐ ☐	I always find a solution
2. My plans for the future are	difficult to accomplish	☐ ☐ ☐ ☐ ☐ ☐ ☐	possible to accomplish
3. I enjoy being	together with other people	☐ ☐ ☐ ☐ ☐ ☐ ☐	by myself
4. My family's understanding of what is important in life is	quite different	☐ ☐ ☐ ☐ ☐ ☐ ☐	very similar
5. I can discuss personal issues with	no one	☐ ☐ ☐ ☐ ☐ ☐ ☐	friends/family-members
6. I am at my best when I	have a goal to strive for	☐ ☐ ☐ ☐ ☐ ☐ ☐	can take one day at a time
7. My personal problems	I know how to solve	☐ ☐ ☐ ☐ ☐ ☐ ☐	I can not find any solutions for
8. I feel that my future looks	very promising	☐ ☐ ☐ ☐ ☐ ☐ ☐	uncertain
9. To be flexible in social settings	is not important to me	☐ ☐ ☐ ☐ ☐ ☐ ☐	is really important to me
10. I feel	very happy with my family	☐ ☐ ☐ ☐ ☐ ☐ ☐	very unhappy with my family
11. Those who are good at encouraging me are	some close friends/family members	☐ ☐ ☐ ☐ ☐ ☐ ☐	no one
12. When I start on new things/projects	I rarely plan ahead, just get on with it	☐ ☐ ☐ ☐ ☐ ☐ ☐	I prefer to have a plan
13. My judgements and decisions	I often doubt	☐ ☐ ☐ ☐ ☐ ☐ ☐	I trust completely
14. My goals	I know how to accomplish	☐ ☐ ☐ ☐ ☐ ☐ ☐	I am unsure how to accomplish
15. New friendships are something	I make easily	☐ ☐ ☐ ☐ ☐ ☐ ☐	I have difficulty making
16. My family is characterized by	disconnection	☐ ☐ ☐ ☐ ☐ ☐ ☐	healthy cohesion
17. The bonds among my friends is	weak	☐ ☐ ☐ ☐ ☐ ☐ ☐	strong
18. I am good at	organizing my time	☐ ☐ ☐ ☐ ☐ ☐ ☐	wasting my time
19. Belief in myself	gets me through difficult periods	☐ ☐ ☐ ☐ ☐ ☐ ☐	is of little help in difficult periods
20. My goals for the future are	unclear	☐ ☐ ☐ ☐ ☐ ☐ ☐	well thought through
21. Meeting new people is	difficult for me	☐ ☐ ☐ ☐ ☐ ☐ ☐	something I am good at

The Resilience Scale for Adults (page 2)

22. In difficult periods my family	keeps a positive outlook on the future	☐	☐	☐	☐	☐	☐	☐	views the future as gloomy
23. When a family member experiences a crisis/emergency	I am informed right away	☐	☐	☐	☐	☐	☐	☐	it takes quite a while before I am told
24. Rules and regular routines	are absent in my everyday life	☐	☐	☐	☐	☐	☐	☐	are a part of my everyday life
25. In difficult periods I have a tendency to	view everything gloomily	☐	☐	☐	☐	☐	☐	☐	find something good that help me thrive/ prosper
26. When I am with others	I easily laugh	☐	☐	☐	☐	☐	☐	☐	I seldom laugh
27. Facing other people, our family acts	unsupportive of one another	☐	☐	☐	☐	☐	☐	☐	loyally towards one another
28. I get support from	friends/ family members	☐	☐	☐	☐	☐	☐	☐	no one
29. Events in my life that I cannot influence	I manage to come to terms with	☐	☐	☐	☐	☐	☐	☐	are a constant source of worry/concern
30. For me, thinking of good topics for conversation is	difficult	☐	☐	☐	☐	☐	☐	☐	easy
31. In my family we like to	do things together	☐	☐	☐	☐	☐	☐	☐	do things on our own
32. When needed, I have	no one who can help me	☐	☐	☐	☐	☐	☐	☐	always someone who can help me
33. My close friends/ family members	appreciate my qualities	☐	☐	☐	☐	☐	☐	☐	dislike my qualities

1.4 The Multidimensional Scale of Perceived Social Support

M

Multidimensional Scale of Perceived Social Support (Zimet, Dahlem, Zimet & Farley, 1988)

Instructions: We are interested in how you feel about the following statements. Read each statement carefully. Indicate how you feel about each statement.

Circle the "1" if you **Very Strongly Disagree**
Circle the "2" if you **Strongly Disagree**
Circle the "3" if you **Mildly Disagree**
Circle the "4" if you are **Neutral**
Circle the "5" if you **Mildly Agree**
Circle the "6" if you **Strongly Agree**
Circle the "7" if you **Very Strongly Agree**

1.	There is a special person who is around when I am in need.	1	2	3	4	5	6	7	SO
2.	There is a special person with whom I can share my joys and sorrows.	1	2	3	4	5	6	7	SO
3.	My family really tries to help me.	1	2	3	4	5	6	7	Fam
4.	I get the emotional help and support I need from my family.	1	2	3	4	5	6	7	Fam
5.	I have a special person who is a real source of comfort to me.	1	2	3	4	5	6	7	SO
6.	My friends really try to help me.	1	2	3	4	5	6	7	Fri
7.	I can count on my friends when things go wrong.	1	2	3	4	5	6	7	Fri
8.	I can talk about my problems with my family.	1	2	3	4	5	6	7	Fam
9.	I have friends with whom I can share my joys and sorrows.	1	2	3	4	5	6	7	Fri
10.	There is a special person in my life who cares about my feelings.	1	2	3	4	5	6	7	SO
11.	My family is willing to help me make decisions.	1	2	3	4	5	6	7	Fam
12.	I can talk about my problems with my friends.	1	2	3	4	5	6	7	Fri

The items tended to divide into factor groups relating to the source of the social support, namely family (Fam), friends (Fri) or significant other (SO).

1.5 The Child Dependency Questionnaire

Central Manchester University Hospitals NHS Bradford Teaching Hospitals NHS Alder Hey Children's NHS

M

Child Dependency Questionnaire V1 24.4.15

Child dependency

Please think about how much your child has relied on you to help them to stick to a protein-restricted diet over the last few weeks. Please check the option box that is closest to the end statement that describes your child best.

My child has managed their diet on their own. ☐ ☐ ☐ ☐ ☐ ☐ ☐ My child has relied on on me to help them stick to a protein-restricted diet.

1.6 The Eyberg Child Behavior Inventory (page 1)

ECBI™ Eyberg Child Behavior Inventory™

Parent Rating Form by Sheila Eyberg, PhD

Your Name _____ Relationship to Child _____ Today's Date ___/___/___

Child's Name _____ Child's Gender _____ Child's Date of Birth ___/___/___

Directions: Below are a series of phrases that describe children's behavior. Please (1) circle the number describing **how often** the behavior **currently** occurs with your child, and (2) circle either "yes" **or** "no" to indicate whether the behavior is **currently a problem for you**.

For example, if seldom, you would circle the 2 in response to the following statement:

	Never	Seldom	Sometimes	Often		Always	Is this a problem for you?
1. Refuses to eat vegetables	1	(2)	3	4	5	6	7	YES (NO)

Circle only one response for each statement, and respond to all statements. **DO NOT ERASE!** If you need to change an answer, make an "X" through the incorrect answer and circle the correct response. For example:

1. Refuses to eat vegetables	1	(2)	(X)	4	5	6	7	YES (NO)

	How often does this occur with your child?							Is this a problem for you?	
	Never	Seldom	Sometimes	Often			Always		
1. Dawdles in getting dressed	1	2	3	4	5	6	7	YES	NO
2. Dawdles or lingers at mealtime	1	2	3	4	5	6	7	YES	NO
3. Has poor table manners	1	2	3	4	5	6	7	YES	NO
4. Refuses to eat food presented	1	2	3	4	5	6	7	YES	NO
5. Refuses to do chores when asked	1	2	3	4	5	6	7	YES	NO
6. Slow in getting ready for bed	1	2	3	4	5	6	7	YES	NO
7. Refuses to go to bed on time	1	2	3	4	5	6	7	YES	NO
8. Does not obey house rules on own	1	2	3	4	5	6	7	YES	NO
9. Refuses to obey until threatened with punishment	1	2	3	4	5	6	7	YES	NO
10. Acts defiant when told to do something	1	2	3	4	5	6	7	YES	NO
11. Argues with parents about rules	1	2	3	4	5	6	7	YES	NO
12. Gets angry when doesn't get own way	1	2	3	4	5	6	7	YES	NO
13. Has temper tantrums	1	2	3	4	5	6	7	YES	NO
14. Sasses adults	1	2	3	4	5	6	7	YES	NO
15. Whines	1	2	3	4	5	6	7	YES	NO
						Page 1 subtotals			

OVER →

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The Eyberg Child Behavior Inventory (page 2)

	How often does this occur with your child?							Is this a problem for you?	
	Never	Seldom	Sometimes		Often		Always		
16. Cries easily	1	2	3	4	5	6	7	YES	NO
17. Yells or screams	1	2	3	4	5	6	7	YES	NO
18. Hits parents	1	2	3	4	5	6	7	YES	NO
19. Destroys toys and other objects	1	2	3	4	5	6	7	YES	NO
20. Is careless with toys and other objects	1	2	3	4	5	6	7	YES	NO
21. Steals	1	2	3	4	5	6	7	YES	NO
22. Lies	1	2	3	4	5	6	7	YES	NO
23. Teases or provokes other children	1	2	3	4	5	6	7	YES	NO
24. Verbally fights with friends own age	1	2	3	4	5	6	7	YES	NO
25. Verbally fights with sisters and brothers	1	2	3	4	5	6	7	YES	NO
26. Physically fights with friends own age	1	2	3	4	5	6	7	YES	NO
27. Physically fights with sisters and brothers	1	2	3	4	5	6	7	YES	NO
28. Constantly seeks attention	1	2	3	4	5	6	7	YES	NO
29. Interrupts	1	2	3	4	5	6	7	YES	NO
30. Is easily distracted	1	2	3	4	5	6	7	YES	NO
31. Has short attention span	1	2	3	4	5	6	7	YES	NO
32. Fails to finish tasks or projects	1	2	3	4	5	6	7	YES	NO
33. Has difficulty entertaining self alone	1	2	3	4	5	6	7	YES	NO
34. Has difficulty concentrating on one thing	1	2	3	4	5	6	7	YES	NO
35. Is overactive or restless	1	2	3	4	5	6	7	YES	NO
36. Wets the bed	1	2	3	4	5	6	7	YES	NO
					Page 2 subtotals				
					Subtotals from page 1				

Scores	Raw score	*T* score	Exceeds Cutoff (✔)
Intensity			
Problem			

Comments:

1.7 The Developmental Behaviour Checklist-Parent/ Carer Version (front page)

DEVELOPMENTAL BEHAVIOUR CHECKLIST
(DBC-P)

Some children with developmental delay have problems with their emotions and behaviour. These can sometimes be a problem for their carers.

By completing this checklist, you will help us learn more about these problems. This will assist us to know how the person might respond to help.

Name of Child or Teenager: _____

Date of Birth/Age:. _____

Sex: _____

Person Completing Form: _____

Relationship to Child: _____

Date Completed: _____

Is the Child: (please circle) Unable to see / unable to hear Unable to speak/ speaks very little

Unable to use arms / legs Subject to other serious medical condition

Please describe: _____

What does he/she do best?

_____

What do other people like about him/her?

_____

What are his/her favourite activities?

_____

Is there anything you feel he/she does as well or better than others?

_____

Have you sought help for any behaviour or emotional problems, apart from slow development, of the child or teenager in your care? Yes / No

If so, from whom?

_____

Please continue over the page →

Office Use Only

Code Number: _____

Developmental Level (circle one only)

Profound Severe Moderate Mild Unknown Contact Person: _____

	TBPS
Page 2	
Page 3	
Page 4	
Total	

①	②	③	④	⑤

Revised subscales 2002

The Developmental Behaviour Checklist-Parent/ Carer Version (page 1)

Many of the following behaviours may not apply to the child or teenager in your care. For each item that does describe the person in your care, now or within the past six months, please circle the **2** if the item is **very true** or **often true**. Circle **1** if the item is **somewhat or sometimes true** of your child. If the item is **not true** of your child circle the **0**.

0 = not true as far as you know 1 = somewhat or sometimes true 2 = very true or often true

If your child is unable to perform an item, circle the **0.** For example, if your child has no speech, then for the item "Talks too much or too fast" circle the **0**

Underline any you are particularly concerned about

Office Use Only	Please Circle			
1. ⑤	0	1	2	Appears depressed, downcast or unhappy
2. ⑤	0	1	2	Avoids eye contact. Won't look you straight in the eye.
3. ②⑤	0	1	2	Aloof, in his/her own world.
4. ①	0	1	2	Abusive. Swears at others.
5. ③	0	1	2	Arranges objects or routine in a strict order. Please describe: _____
6. ②	0	1	2	Bangs head.
7. ②	0	1	2	Becomes over-excited.
8. ②	0	1	2	Bites others.
9. ②	0	1	2	Cannot attend to one activity for any length of time, poor attention span.
10. ②	0	1	2	Chews or mouths objects, or body parts.
11. ④	0	1	2	Cries easily for no reason, or over small upsets.
12.	0	1	2	Covers ears or is distressed when hears particular sounds. Please describe: _____
13. ③	0	1	2	Confuses the use of pronouns e.g. uses "you" instead of "I".
14. ①②	0	1	2	Deliberately runs away.
15.	0	1	2	Delusions: has a firmly held belief or idea that can't possibly be true. Please describe: _____
16. ④	0	1	2	Distressed about being alone.
17. ⑤	0	1	2	Doesn't show affection.
18. ⑤	0	1	2	Doesn't respond to others' feelings, e.g. shows no response if a family member is crying.
19.	0	1	2	Easily distracted from his/her task, e.g. by noises.
20. ①	0	1	2	Easily led by others.
21. ②	0	1	2	Eats non-food items e.g. dirt, grass, soap.
22. ④	0	1	2	Excessively distressed if separated from familiar person.
23. ④	0	1	2	Fears particular things or situations, e.g. the dark or insects. Please describe: _____
24. ②	0	1	2	Facial twitches or grimaces.
25. ②	0	1	2	Flicks, taps, twirls objects repeatedly.
26. ④	0	1	2	Fussy eater or has food fads.
27. ②	0	1	2	Gorges food. Will do anything to get food e.g. takes food out of garbage bins or steals food.
28. ③	0	1	2	Gets obsessed with an idea or activity. Please describe: _____
29. ②	0	1	2	Grinds teeth.
30. ④	0	1	2	Has nightmares, night terrors or walks in sleep.

Please be sure you have answered all items

Continue next page →

Office Use Only

TBPS

Subscales

①	②	③	④	⑤

The Developmental Behaviour Checklist-Parent/ Carer Version (page 2)

0 = not true as far as you know 1 = somewhat or sometimes true 2 = very true or often true
Underline any you are particularly concerned about

Office Use Only	Please Circle			
31. ①	0	1	2	Has temper tantrums, e.g. stamps feet, slams doors.
32. ①	0	1	2	Hides things.
33. ②	0	1	2	Hits self or bites self.
34. ②	0	1	2	Hums, whines, grunts, squeals or makes other non-speech noises.
35. ①	0	1	2	Impatient.
36.	0	1	2	Inappropriate sexual activity with another.
37. ①	0	1	2	Impulsive, acts before thinking.
38. ①	0	1	2	Irritable.
39. ①	0	1	2	Jealous.
40. ①	0	1	2	Kicks, hits others.
41. ①	0	1	2	Lacks self-confidence, poor self-esteem.
42. ②	0	1	2	Laughs or giggles for no obvious reason.
43. ①	0	1	2	Lights fires.
44. ②	0	1	2	Likes to hold or play with an unusual object, e.g. string, twigs; overly fascinated with something, e.g. water. Please describe: _____
45. ④	0	1	2	Loss of appetite.
46. ②	0	1	2	Masturbates or exposes self in public.
47. ①	0	1	2	Mood changes rapidly for no apparent reason.
48. ⑤	0	1	2	Moves slowly, underactive, does little, e.g. only sits and watches others.
49. ①	0	1	2	Noisy or boisterous.
50. ①②	0	1	2	Overactive, restless, unable to sit still.
51. ③	0	1	2	Overaffectionate.
52. ⑤	0	1	2	Overbreathes, vomits, has headaches or complains of being sick for no physical reason.
53. ①	0	1	2	Overly attention-seeking.
54. ③	0	1	2	Overly interested in looking at, listening to or dismantling mechanical things e.g. lawnmower, vacuum cleaner.
55. ②	0	1	2	Poor sense of danger.
56. ③	0	1	2	Prefers the company of adults or younger children. Doesn't mix with his/her own age group.
57. ⑤	0	1	2	Prefers to do things on his/her own. Tends to be a loner.
58. ③	0	1	2	Preoccupied with only one or two particular interests. Please describe: _____
59. ①	0	1	2	Refuses to go to school, activity centre or workplace.
60. ②	0	1	2	Repeated movements of hands, body, head or face e.g. handflapping or rocking.
61. ⑤	0	1	2	Resists being cuddled, touched or held.
62. ③	0	1	2	Repeats back what others say like an echo.
63. ③	0	1	2	Repeats the same word or phrase over and over.
64. ②	0	1	2	Smells, tastes, or licks objects.
65.	0	1	2	Scratches or picks his/her skin.
66. ②	0	1	2	Screams a lot.

Please be sure you have answered all items
Continue over the page ➔

Office Use Only

TBPS

Subscales

①	②	③	④	⑤

The Developmental Behaviour Checklist- Parent/ Carer Version (page 3)

0 = not true as far as you know 1 = somewhat or sometimes true 2 = very true or often true
Underline any you are particularly concerned about

Office Use Only	Please Circle	
67.	0 1 2	Sleeps too little. Disrupted sleep.
68. ②	0 1 2	Stares at lights or spinning objects.
69. ⑤	0 1 2	Sleeps too much.
70. ②	0 1 2	Soils outside toilet though toilet trained. Smears or plays with faeces.
71. ③	0 1 2	Speaks in whispers, high pitched voice, or other unusual tone or rhythm.
72. ②	0 1 2	Switches lights on and off, pours water over and over; or similar repetitive activity. Please describe: _____
73. ①	0 1 2	Steals.
74. ①	0 1 2	Stubborn, disobedient or unco-operative.
75. ④	0 1 2	Shy.
76. ②	0 1 2	Strips off clothes or throws away clothes.
77. ①	0 1 2	Says he/she can do things that he/she is not capable of.
78. ③	0 1 2	Stands too close to others.
79.	0 1 2	Sees, hears, something which isn't there. Hallucinations. Please describe: _____
80.	0 1 2	Talks about suicide.
81. ①	0 1 2	Talks too much or too fast.
82. ③	0 1 2	Talks to self or imaginary people or objects
83. ①	0 1 2	Tells lies.
84.	0 1 2	Thoughts are unconnected. Different ideas are jumbled together with meaning difficult to follow.
85. ①	0 1 2	Tense, anxious, worried.
86. ①②	0 1 2	Throws or breaks objects.
87. ①	0 1 2	Tries to manipulate or provoke others.
88. ②	0 1 2	Underreacts to pain.
89. ③	0 1 2	Unrealistically happy or elated.
90. ②	0 1 2	Unusual body movements, posture, or way of walking. Please describe: _____
91. ④	0 1 2	Upset and distressed over small changes in routine or environment. Please describe: _____
92. ②	0 1 2	Urinates outside toilet, although toilet trained.
93. ①	0 1 2	Very bossy.
94. ②	0 1 2	Wanders aimlessly.
95. ①	0 1 2	Whines or complains a lot.
		Please write in any problems your child has that were not listed above
	0 1 2	_____
	0 1 2	_____
	0 1 2	_____
96.	0 1 2	Overall, do you feel your child has problems with feelings or behaviour, in addition to problems with development? If not, please circle the **0**. If so, but they're minor, please circle the **1.** If they're major problems, please circle the **2.**

Please be sure you have answered all items

Are there any other comments you would like to make?

_____

_____

THANK YOU

Office Use Only

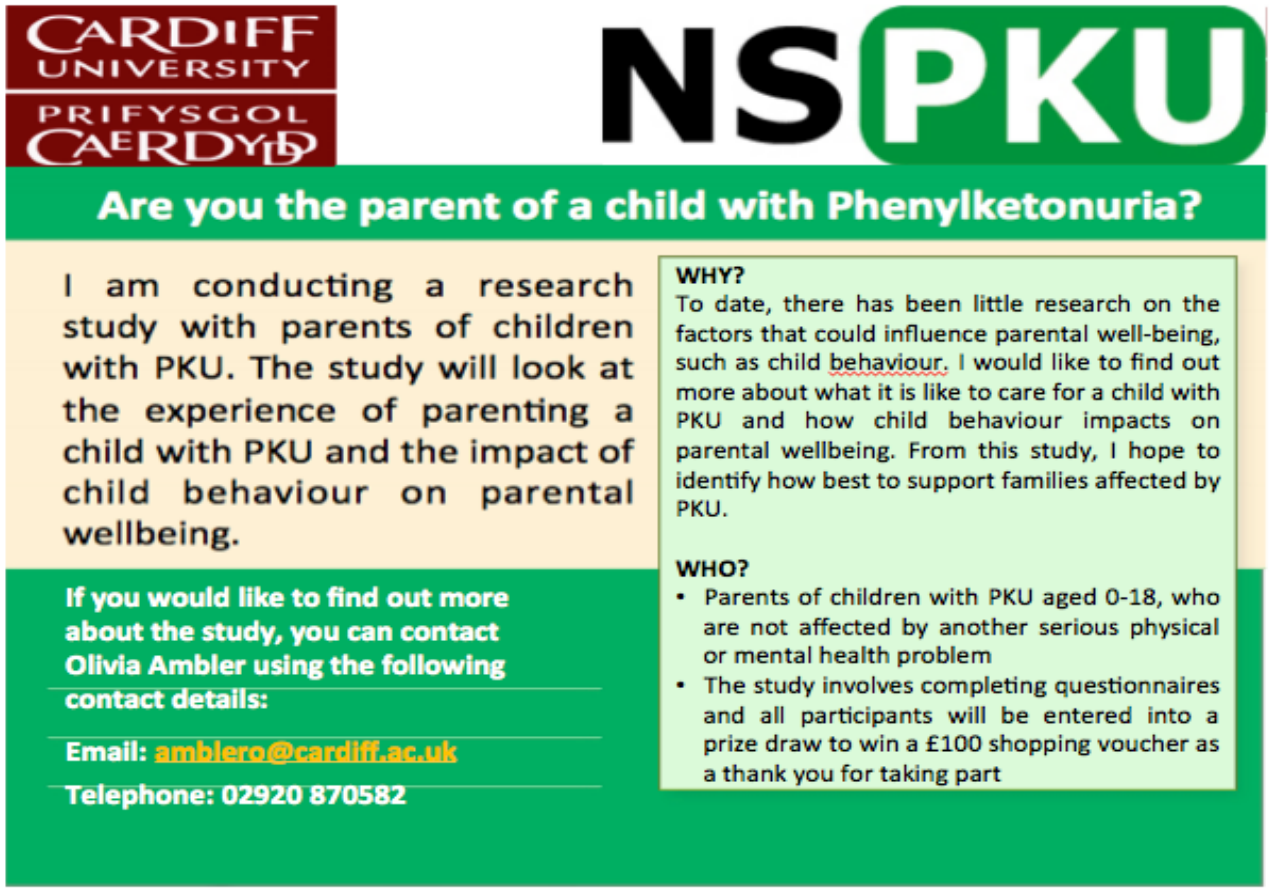
TBPS

Subscales

①	②	③	④	⑤

Appendix 2: Study adverts

2.1 Experimental group



2.2 Control group

CARDIFF UNIVERSITY
PRIFYSGOL CAERDYDD

Are you the parent of a child who is between 0-18 years old?

WHAT?

- I am conducting a research study with parents of children with **Phenylketonuria** (PKU) to understand what impacts on parent's well-being and what helps.
- As part of this work, I am also looking at what impacts on the well-being of parents who are for **healthy** children.

WHY?

- Currently, little is known about the factors that contribute to parents' well-being in PKU. The findings from this study may help clinicians to develop support packages for families in the future.

WHO?

- Parents/carers of children between 0-18 years old who **do not** have a serious health condition and are not affected by another significnt physical or mental health problem.
- The study involves completing questionnaires and all participants will be entered into a prize draw to win a £100 shopping voucher as a thank you for taking part.

If you would like to find out more about the study, please email Olivia on:
amblero@cardiff.ac.uk
THANK YOU

Appendix 3: Approval for the study

3.1 Ethical Approval (correspondence with Cardiff University Ethics Committee)

Fri 15/07/2016, 09:47
Dear Olivia,

The Ethics Committee has considered your PG project proposal: Parenting Experiences and Child Behaviour in PKU (EC.16.07.12.4554).

The project has been approved.

Please note that if any changes are made to the above project then you must notify the Ethics Committee.

Best wishes,
Mark Jones

3.2 Ethics amendment approval (correspondence with Cardiff University Ethics Committee)

Tue 04/04, 13:31
Olivia Ambler;
Dougal Hare

Action Items
Dear Olivia,

The Ethics Committee has considered the amendment to your PG project proposal: Parenting Experiences and Child Behaviour in PKU (EC.16.07.12.4554A2).

The amendment has been approved.

Please note that if any changes are made to the above project then you must notify the Ethics Committee.

Best wishes,
Mark

Appendix 4: Participant information sheets for the experimental group

4.1 Consent form

Participant ID:_____

CONSENT FORM

Title of Project: Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

Name of Researcher: Miss Olivia Ambler

1	I confirm that I have read the information sheet for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily.	
2	Other than caring for my child with PKU, I confirm that I do not have any other significant caring responsibilities that may impact on my wellbeing.	
3	Other than caring for my child with PKU, I confirm that I am not affected by any other serious physical or mental health problems that may impact upon my wellbeing.	
4	I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason, without my medical care or legal rights being affected, up until the research data has been analysed.	
5	I understand that data collected during the study may be looked at by individuals from Cardiff University, regulatory authorities and members of the National Society for Phenylketonuria. I give permission for these individuals to have access to my data.	
6	I understand that data from the study will be stored for a minimum of five years after its completion, or at least two years post-publication to allow for any further analysis or review to be conducted. After this time period, all paper copies of data will be destroyed and electronic copies will be deleted. I give permission for my data to be stored for this time.	
7	I agree to take part in the above study.	

Name of child _____ Name of Participant _____

Participant Signature _____ Date _____

4.2 Invitation letter

Invitation Letter

Dear

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

I am writing to invite you to take part in a research study being conducted at Cardiff University with parents of children with and without Phenylketonuria (PKU).

To date, there has been little research into the factors that contribute to the well-being of parents who care for a child with PKU. It would be useful to find out more information about this to help support families affected by the condition.

This study will investigate the different factors that affect the wellbeing of parents who care for a child with PKU, compared to parents who care for a child without PKU. The study will also investigate child behaviour in PKU and examine whether this too, impacts on parent's wellbeing.

As you have a child with PKU who is between the ages of 0 and 18 years old, you are invited to take part in this study. You will find enclosed with this letter a participant information sheet, consent form, a list of support services, an opt-out form and some questionnaires.

If you would like to find out more about this project, please read the participant information sheet. If you would like to find out any further information, please email me on amblero@cardiff.ac.uk or telephone me on 02920 870582.

If you would like to take part after reading the participant information sheet, please complete the enclosed consent form and questionnaires. If you would like any help with completing the questionnaires, please contact me using the above contact details. Please take short breaks when filling in the questionnaires if you feel tired.

Please return the consent form and questionnaires in the addressed pre-paid envelope (no stamp is required), or hand them into the Department of Clinical Psychology administration team on the 11th Floor, Tower Building, Park Place, Cardiff, CF10 3AT.

If you would prefer NOT to take part in this project please complete and return the attached opt-out form in the pre-paid envelope along with the blank questionnaires, so that I do not contact you again.

I look forward to hearing from you.

Yours sincerely,

Miss Olivia Ambler

4.3 Participant information sheet

Participant Information Sheet

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

Research Team: Miss Olivia Ambler (Cardiff University), Dr Emma Medford (University of Manchester) and Dr Dougal Hare (Cardiff University).

I would like to invite you to take part in my research study. Joining the study is entirely up to you. Before you decide I would like you to understand why the research is being done and what it would involve for you.
Part 1 tells you the purpose of this study and what it will involve if you take part.
Part 2 gives you more detailed information about the conduct of the study.
I recommend that you take a minimum of 24 hours to consider the information below before deciding whether to take part.

Part 1

1.1 What is the purpose of the study?

There has been little research on what it is like to look after a child with Phenylketonuria (PKU). This study will investigate child behaviour in PKU and how this affects parent's well-being.

1.2 Why have I been invited to take part in this study?

You have been invited to take part because you have a child with PKU who is between the ages of 0-18. You have also been invited because your family is not currently affected by any other serious physical or mental health problems that may impact upon your wellbeing.

1.3 Do I have to take part?

No, you do not have to take part in the study if you do not want to. Taking part in the research is voluntary; this means it is completely up to you to decide whether or not to take part. Your decision to participate in this study will not be connected to the care you and your family are receiving now or in the future. If you decide to take part and sign the consent form but change your mind later, you are free to withdraw at any point during the study without giving a reason and without any consequence to your current or future treatment.

1.4 What will participation involve?

Parents/ carers will complete a set of questionnaires, which ask about:

- Demographic information
- Levels of psychological distress
- Levels of resilience
- The care dependency of the child
- Support from family and friends
- Any behavioural problems experienced by the child

- Any behaviours related to developmental difficulties

Together, these questionnaires will take about 45 minutes to complete. You will be provided with a pre-paid envelope to return the questionnaires.

1.5 What are the possible disadvantages and risks of taking part?

It is possible that the questionnaires might raise issues that could be distressing to think about. A list of agencies and people you can contact is provided should you need any additional information/support.

1.6 What are the possible benefits of taking part?

The information gained will help services to fully understand the demands of caring for a child with PKU. It will also help identify some of the most effective ways to support parents. This will enable clinicians to develop appropriate support packages, which may help other families in the future.

1.7 Will my taking part in the study be kept confidential?

Yes. Your data will be handled sensitively and in confidence; all legal and ethical guidelines will be followed. More details are given in Part 2.

Part 2

2.1 What will happen if I do not want to carry on with the study?

You can withdraw from the study at any time without giving a reason and without any consequence to your family's current or future treatment, up until the data has been analysed. When the data is analysed it will not be personally identifiable.

2.2 What if there is a problem?

It is unlikely that anything would go wrong, but if you have a concern about any aspect of the study, you should contact one of the researchers or the School of Psychology Research Ethics Committee (contact information is provided in section 2.9). If you are not satisfied and wish to make a formal complaint, you can do so through the Cardiff University complaints procedure. Details can be obtained from the University.
In the event that something does go wrong and you are harmed during the study and this is due to somebody's negligence, then you may have grounds for a legal action for compensation against Cardiff University, but you might have to pay your legal costs.

2.3 Will my data be confidential?

All data collected about you and your child will be kept strictly confidential and only viewed by members of the research team. It will be stored securely in a locked filing cabinet at the University.

Data will be entered onto a computer database which will be password protected and encrypted. Each participant will be assigned a number, thus names will not be entered onto the database.

You will be asked to provide contact details for your GP, but they will not be routinely contacted. During the study if any concerns arise about risk of harm to anyone, then I will have to contact the appropriate agency/person to provide support. If possible, I would speak to you first about this.

I plan to publish the research and names of participants will **not** be used.

2.4 Will I receive any payment for taking part in the study?

No, participants will not receive any payment for taking part. However, when participants return their consent form and questionnaires, they will be entered into a prize draw, with the opportunity of winning a £100 shopping voucher as a thank you for taking part in the study.

2.6 Who is organising the research?

This research is being conducted as part of the Doctorate in Clinical Psychology at Cardiff University for Trainee Clinical Psychologist/postgraduate student Miss Olivia Ambler. This study will be carried out under the guidance of Dr Dougal Hare (Academic Supervisor). It is funded by Cardiff University.

2.7 Where will the findings be published?

I intend to publish the results in peer-reviewed journals

I intend to present the results at scientific conferences

I may put a summary of the findings in an NSPKU (The National Society for Phenylketonuria) newsletter.

I will provide participants with a summary of the findings if they would like this.

2.8 Who has reviewed the study?

This study has been reviewed and given a favourable opinion by the Cardiff University School of Psychology Research Ethics Committee.

2.9 Who can I contact for further information?

If you would like to discuss the study or have any questions or concerns, please do not hesitate to contact Miss Olivia Ambler at amblero@cardiff.ac.uk or tel. 02920 870582.

You can also contact Dr Dougal Hare using the following address:
Department of Clinical Psychology
11th Floor Tower Building
70 Park Place
Cardiff University
Cardiff
CF10 3AT
Tel: +44 (0)2920 870582
Email: HareD@cardiff.ac.uk

Alternatively, you can contact the School of Psychology Research Ethics Committee at Cardiff University using the address below:
School of Psychology
Cardiff University
Tower Building
70 Park Place
Cardiff
CF10 3AT
Tel: +44 (0)2920 870360
Email psychethics@cardiff.ac.uk

You can keep this copy of the information sheet.

4.4 Opt out form

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

Opt out form

I would NOT like to take part in the above study.	

You do not have to give a reason, but if you feel able to tell us why, it will help us to understand why some people choose not to take part in this type of project.

_____
_____

Name of child _____

Name of Parent / Carer _____

Date _____

Thank you for taking the time to complete and return this form

4.5 Debrief letter

Dear

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

This study had three aims:

1. To compare levels of psychological distress between parents of children and young people with PKU and parents in the general population
2. To compare the incidence of child behaviour problems experienced by children and young people with PKU and compare this to the general population
3. To examine the factors that contribute towards psychological distress in parents of children with PKU, distinct from parents in the general population

Two groups of participants were recruited for this study: An experimental group with parents of children **with** PKU and a control group with parents of children **without** PKU. As the parent of a child **with** PKU, you contributed to the **experimental group** in the study.

Parents in both groups were asked to complete the same set of questionnaires. These measured:

- Psychological distress
- Psychological resilience
- Perceived social support
- Child behaviour
- Socio-demographic information

In addition to this, parents in the **experimental group** were asked to complete two more questionnaires. These measured:

- Child behaviour related to developmental difficulties
- The care dependency of the child

The responses from these questionnaires will be analysed in four stages, outlined below:

1. For the first part of the analysis, levels of psychological distress in parents of children **with** PKU will be compared to parents of children **without** PKU.

2. For the second part of the analysis, child behaviour problems will be compared between parents of children **with** PKU and parents of children **without** PKU.

3. For the third part of the analysis, the factors contribute to psychological distress will be examined in both groups of parents

All the data we collected for this study is confidential, all personal and identifiable information will be kept anonymous and can only be accessed by the researcher and relevant members of the research team. Your participation in this study is greatly appreciated.

If you have any questions or queries about this project, please phone me on 02920870582 or email me at amblero@cardiff.ac.uk. Alternatively, you can contact my supervisor, Dr Dougal Julian Hare on the above telephone number or email address HareD@cardiff.ac.uk.

Thank you again for your participation.

Yours sincerely,

Miss Olivia Ambler
Trainee Clinical Psychologist

4.6 Thank you letter

Dear

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

I am writing to express my thanks to you for taking part in the above study. Thank you very much for completing and returning your consent form and questionnaires.

Your participation is very important, as it will help contribute toward our understanding of how best to support parents of children with PKU.

You have now been entered into prize draw for a £100 shopping voucher as a thank you for taking part.

If you would like to receive a summary of the study findings, please phone me on **02920 870582** or email me at **amblero@cardiff.ac.uk**.

Yours sincerely,

Miss Olivia Ambler
Trainee Clinical Psychologist

4.7 List of support services

LIST OF SUPPORT SERVICES

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

Some of the questionnaires used in this study covered potentially sensitive material. If you feel affected from contributing to this research and wish to seek additional support or advice, we recommend that you contact one of the following services:

- Contact details for Dr Dougal Julian Hare, Clinical Psychologist in the field of Intellectual Disabilities:

  Address: South Wales Doctoral Programme in Clinical Psychology
  11th Floor
  Tower Building
  70 Park Place
  Cardiff
  CF11 3AT
  Email: HareD@cardiff.ac.uk Telephone: 02920870582

- 'Contact a family: For parents of children with disabilities'. Online advice and support available via www.cafamily.org.uk/medical-information/conditions/p/phenylketonuria/

  Telephone support available Monday-Friday, 9.30am to 5.00pm on

  0808 808 3555 or email helpline@cafamily.org.uk

- Online information and support about PKU and its management is available from:

  www.nhs.uk/conditions/phenylketonuria/Pages/Introduction.aspx

  Online support and advice for parents of children with PKU:

  www.pkuconnect.co.uk/overview/parents/

  A comprehensive list of other online support service is also available at www.pku.com/resources/related-websites

- National Society for Phenylketonuria (NSPKU) www.nspku.org/contact/general

  Helpline 030 3040 1090

  Email info@nspku.org

- Young minds parents, support for parents of children with or without physical/mental health difficulties. Advice is offered to parents who may be worried about the behaviour or wellbeing of their child
  www.youngminds.org.uk/for_parents/parent_helpline

  Helpline 0808 802 5544, Email ymenquiries@youngminds.org.uk
  Address Suit 11, Baden Place, Crosby Row, London SE1 1YW

Appendix 5: Participant information sheets for control group
5.1 Consent form

CONSENT FORM

Participant ID:_____

Title of Project: Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

Name of Researcher: Miss Olivia Ambler

1	I confirm that I have read the information for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily.	
2	I confirm that I **do not** care for a child with a serious health condition, nor do I have any other significant caring responsibilities that may impact on my wellbeing (for example, caring for an elderly relative).	
3	I confirm that I am **not** affected by any serious physical or mental health problems that may impact upon my wellbeing (for example, chronic pain).	
4	I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason, without my medical care or legal rights being affected, up until the research data has been analysed.	
5	I understand that data collected during the study may be looked at by individuals from Cardiff University, regulatory authorities and members of the National Society for Phenylketonuria. I give permission for these individuals to have access to my data.	
6	I understand that data from the study will be stored for a minimum of five years after its completion, or at least two years post-publication to allow for any further analysis or review to be conducted. After this time period, all paper copies of data will be destroyed and electronic copies will be deleted. I give permission for my data to be stored for this time.	
7	I agree to take part in the above study.	

Name of child _____ Name of Participant_____

Participant Signature _____ Date _____

5.2 Invitation letter

Invitation Letter

Dear

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

I am writing to invite you to take part in a research study being conducted at Cardiff University with parents of children with and without Phenylketonuria (PKU).

To date, there has been little research into the factors that contribute to parental well-being in PKU. It would be useful to find out more information about this to help support families affected by the condition.

This study will investigate the different factors that affect the wellbeing of parents who care for a child with PKU, compared to parents who care for a child without PKU. It will also investigate child behaviour in PKU and examine whether this too, impacts on parent's wellbeing.

As you have a child who is between the ages of 0 and 18 years old and who **does not** have PKU, you are invited to take part in this study. You will find enclosed with this letter a participant information sheet, consent form, a list of support services, an opt-out form and some questionnaires.

If you would like to find out more about this project, please read the participant information sheet. If you would like to find out any further information, please email me on amblero@cardiff.ac.uk or telephone me on 02920 870582.

If you would like to take part after reading the participant information sheet, please complete the enclosed consent form and questionnaires. If you would like any help with completing the questionnaires, please contact me using the above contact details. Please take short breaks when filling in the questionnaires if you feel tired.

Please return the consent form and questionnaires in the addressed pre-paid envelope (no stamp is required), or hand them into the Department of Clinical Psychology administration team on the 11th Floor, Tower Building, Park Place, Cardiff, CF10 3AT.

If you would prefer NOT to take part in this project please complete and return the attached opt-out form in the pre-paid envelope along with the blank questionnaires, so that I do not contact you again.

I look forward to hearing from you.

Yours sincerely,

Miss Olivia Ambler

5.3 Participant information sheet

Participant Information Sheet

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

Research Team: Miss Olivia Ambler (Cardiff Univeristy), Dr Emma Meford (University of Manchester) and Dr Dougal Hare (Cardiff University).

I would like to invite you to take part in my research study. Joining the study is entirely up to you. Before you decide I would like you to understand why the research is being done and what it would involve for you.
Part 1 tells you the purpose of this study and what it will involve if you take part.
Part 2 gives you more detailed information about the conduct of the study.
I recommend that you take a minimum of 24 hours to consider the information below before deciding whether to take part.

Part 1

1.1 What is the purpose of the study?

There has been little research on what it is like to look after a child with Phenylketonuria (PKU). This study will investigate child behaviour in PKU and how this affects parent's wellbeing.

1.2 Why have I been invited to take part in this study?

You have been invited to take part because you have a child who is between the ages of 0-18 and who **does not** have a serious health condition. You have also been invited because your family is not currently affected by any other serious physical or mental health problems that may impact upon your wellbeing. In total, we will need 99 other people to also take part in this project.

1.3 Do I have to take part?

No, you do not have to take part in the study if you do not want to. Taking part in the research is voluntary; this means it is completely up to you to decide whether or not to take part. Your decision to participate in this study will not be connected to the care you and your family are receiving now or in the future. If you decide to take part and sign the consent form but change your mind later, you are free to withdraw at any point during the study without giving a reason and without any consequence to your current or future treatment.

1.4 What will participation involve?

Parents/ carers will complete a set of questionnaires, which ask about:

- Demographic information
- Levels of psychological distress

- Levels of resilience
- Support from family and friends
- Any behavioural problems experienced by the child

Together, these questionnaires will take about 30 minutes to complete. You will be provided with a pre-paid envelope to return the questionnaires.

1.5 What are the possible disadvantages and risks of taking part?

It is possible that the questionnaires might raise issues that could be distressing to think about. A list of agencies and people you can contact is provided should you need any additional information/support.

1.6 What are the possible benefits of taking part?

The information gained will help services to fully understand the demands of caring for a child with PKU. It will also help identify some of the most effective ways to support parents. This will enable clinicians to develop appropriate support packages, which may help other families in the future.

1.7 Will my taking part in the study be kept confidential?

Yes. Your data will be handled sensitively and in confidence; all legal and ethical guidelines will be followed. More details are given in Part 2.

Part 2

2.1 What will happen if I do not want to carry on with the study?

You can withdraw from the study at any time without giving a reason and without any consequence to your family's current or future treatment, up until the data has been analysed. When the data is analysed it will not be personally identifiable.

2.2 What if there is a problem?

It is unlikely that anything would go wrong, but if you have a concern about any aspect of the study, you should contact one of the researchers or the School of Psychology Research Ethics Committee (contact information is provided in section 2.9). If you are not satisfied and wish to make a formal complaint, you can do so through the Cardiff University complaints procedure. Details can be obtained from the University.

In the event that something does go wrong and you are harmed during the study and this is due to somebody's negligence, then you may have grounds for a legal action for compensation against Cardiff University, but you might have to pay your legal costs.

2.3 Will my data be confidential?

All data collected about you and your child will be kept strictly confidential and only viewed by members of the research team. It will be stored securely in a locked filing cabinet at the University.

Data will be entered onto a computer database which will be password protected and encrypted. Each participant will be assigned a number, thus names will not be entered onto the database.

You will be asked to provide contact details for your GP, but they will not be routinely contacted. During the study if any concerns arise about risk of harm to anyone, then I will have to contact the appropriate agency/person to provide support. If possible, I would speak to you first about this.

I plan to publish the research and names of participants will **not** be used.

2.4 Will I receive any payment for taking part in the study?

No, participants will not receive any payment for taking part. However, when participants return their consent form and questionnaires, they will be entered into a prize draw, with the opportunity of winning a £100 shopping voucher as a thank you for taking part in the study.

2.6 Who is organising the research?

This research is being conducted as part of the Doctorate in Clinical Psychology at Cardiff University for Trainee Clinical Psychologist/postgraduate student Miss Olivia Ambler. This study will be carried out under the guidance of Dr Dougal Hare (Academic Supervisor). It is funded by Cardiff University.

2.7 Where will the findings be published?

I intend to publish the results in peer-reviewed journals

I intend to present the results at scientific conferences

I may put a summary of the findings in an NSPKU (The National Society for Phenylketonuria) newsletter.

I will provide participants with a summary of the findings if they would like this.

2.8 Who has reviewed the study?

This study has been reviewed and given a favourable opinion by the Cardiff University School of Psychology Research Ethics Committee.

2.9 Who can I contact for further information?

If you would like to discuss the study or have any questions or concerns, please do not hesitate to contact Miss Olivia Ambler at amblero@cardiff.ac.uk or tel. 02920 870582.

You can also contact Dr Dougal Hare using the following address:
Department of Clinical Psychology
11th Floor Tower Building
Park Place
Cardiff University
Cardiff
CF10 3AT
Tel: +44 (0)2920 870582
Email: HareD@cardiff.ac.uk

Alternatively, you can contact the School of Psychology Research Ethics Committee at Cardiff University using the address below:
School of Psychology
Cardiff University
Tower Building
70 Park Place
Cardiff

CF10 3AT
Tel: +44 (0)2920 870360
Email psychethics@cardiff.ac.uk

You can keep this copy of the information sheet.

5.4 Opt out form

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

Opt out form

I would NOT like to take part in the above study.	

You do not have to give a reason, but if you feel able to tell us why, it will help us to understand why some people choose not to take part in this type of project.

_____
_____

Name of child _____

Name of Parent / Carer _____

Date _____

Thank you for taking the time to complete and return this form

5.5 Debrief letter

Debrief Letter

Dear

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

This study had three aims:

4. To compare levels of psychological distress between parents of children and young people with PKU and parents in the general population
5. To compare the incidence of child behaviour problems experienced by children and young people with PKU and compare this to the general population
6. To examine the factors that contribute towards psychological distress in parents of children with PKU, distinct from parents in the general population

Two groups of participants were recruited for this study: An experimental group with parents of children **with** PKU and a control group with parents of children **without** PKU. As the parent of a child **without** PKU or any other health condition, you contributed to the **control group** in the study.

Parents in both groups were asked to complete the same set of questionnaires. These measured:

- Psychological distress
- Psychological resilience
- Perceived social support
- Child behaviour
- Socio-demographic information

In addition to this, parents in the **experimental group** were asked to complete two more questionnaires. These measured:

- Child behaviour related to developmental difficulties
- The care dependency of the child

The responses from these questionnaires will be analysed in four stages, outlined below:

1. For the first part of the analysis, levels of psychological distress in parents of children **with** PKU will be compared to parents of children **without** PKU.

2. For the second part of the analysis, child behaviour problems will be compared between parents of children **with** PKU and parents of children **without** PKU.

3. For the third part of the analysis, the factors contribute to psychological distress will be examined in both groups of parents

All the data we collected for this study is confidential, all personal and identifiable information will be kept anonymous and can only be accessed by the researcher and relevant members of the research team. Your participation in this study is greatly appreciated.

If you have any questions or queries about this project, please phone me on 02920870582 or email me at amblero@cardiff.ac.uk. Alternatively, you can contact my supervisor, Dr Dougal Julian Hare on the above telephone number or email address HareD@cardiff.ac.uk.

Thank you again for your participation.

Yours sincerely,

Miss Olivia Ambler
Trainee Clinical Psychologist

5.6 Thank you letter

Dear

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

I am writing to express my thanks to you for taking part in the above study. Thank you very much for completing and returning your consent form and questionnaires.

Your participation is very important, as it will help contribute toward our understanding of how best to support parents of children with PKU.

You have now been entered into prize draw for a £100 shopping voucher as a thank you for taking part.

If you would like to receive a summary of the study findings, please phone me on **02920 870582** or email me at **amblero@cardiff.ac.uk**.

Yours sincerely,

Miss Olivia Ambler
Trainee Clinical Psychologist

5.7 List of support services

LIST OF SUPPORT SERVICES

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

Some of the questionnaires used in this study covered potentially sensitive material. If you feel affected from contributing to this research and wish to seek additional support or advice, we recommend that you contact one of the following services:

- Contact details for Dr Dougal Julian Hare, Clinical Psychologist in the field of Intellectual Disabilities:

  Address: South Wales Doctoral Programme in Clinical Psychology
  11th Floor
  Tower Building
  70 Park Place
  Cardiff
  CF11 3AT
  Email: HareD@cardiff.ac.uk Telephone: 02920870582

- 'Contact a family: For parents of children with disabilities'. Online advice and support available via www.cafamily.org.uk/medical-information/conditions/p/phenylketonuria/

  Telephone support available Monday-Friday, 9.30am to 5.00pm on 0808 808 3555 or email helpline@cafamily.org.uk

- Online information and support about PKU and its management is available from:

  www.nhs.uk/conditions/phenylketonuria/Pages/Introduction.aspx

  Online support and advice for parents of children with PKU:

  www.pkuconnect.co.uk/overview/parents/

  A comprehensive list of other online support service is also available at www.pku.com/resources/related-websites

- National Society for Phenylketonuria (NSPKU) www.nspku.org/contact/general

  Helpline 030 3040 1090

  Email info@nspku.org

- Young minds parents, support for parents of children with or without physical/mental health difficulties. Advice is offered to parents who may be worried about the behaviour or wellbeing of their child
  www.youngminds.org.uk/for_parents/parent_helpline

Helpline 0808 802 5544, Email ymenquiries@youngminds.org.uk
Address Suit 11, Baden Place, Crosby Row, London SE1 1YW

5.8 Reminder letter for participants in experimental and control groups

Dear

Parenting a child with Phenylketonuria (PKU): An investigation into the factors that contribute to parental well-being

I am writing to invite you to take part in a major research study being conducted at Cardiff University with parents of children with and without Phenylketonuria (PKU). The study is under the direction of Dr Dougal Hare from Cardiff University.

There has been little research on what it is like to look after a child with PKU and how PKU affects children's behaviour. This study will investigate child behaviour in PKU and the impact of this on parent's wellbeing. It will also look at the different things that affect parent's wellbeing and to help identify what might improve this.

You will find enclosed with this letter a participant information sheet, a consent form, some questionnaires, a list of support services and an opt-out form.

If you are interested in finding out more about this project, please read the participant information sheet for further details. If you would like any further information, please phone me on **02920 870582** or email me at **amblero@cardiff.ac.uk.**

If you would like to take part in this project after reading the participant information sheet, please complete the enclosed consent form and questionnaires. If you would like any assistance with completing the questionnaires please contact me using the above contact details.

Please return the consent form and questionnaires in the addressed pre-paid envelope (no stamp is required), or hand them in to the administration team at the Department of Clinical Psychology, 11th Floor Tower Building, Park Place, Cardiff University, Cardiff CF10 3AT.

If you experience any tiredness or fatigue when completing the questionnaires, please take short breaks as necessary. If you would like any support or information regarding your emotional wellbeing, please see the list of support services and contacts.

Participation in this study is voluntary and will in no way affect the medical treatment of your family.

I look forward to hearing from you.

Yours sincerely,

Miss Olivia Ambler
Trainee Clinical Psychologist

Appendix 6: SPSS output

6.1 Tests of normality for experimental and control groups

6.1.2 Normality statistics experimental group

Tests of Normality

	Kolmogorov-Smirnov[a]			Shapiro-Wilk		
	Statistic	df	Sig.	Statistic	df	Sig.
Ex_MPSS_SO	.240	26	.000	.696	26	.000
Ex_MPSS_Fa	.213	26	.004	.781	26	.000
Ex_MPSS_Fr	.217	26	.003	.838	26	.001
Ex_MPSS_Total	.179	26	.031	.793	26	.000
Ex_ECBI_Problem	.187	26	.020	.834	26	.001
Ex_ECBI_Intensity	.094	26	.200*	.961	26	.412
Ex_GHQ12_Total	.219	26	.002	.908	26	.024
Ex_RSA_Total	.142	26	.193	.927	26	.067
Disruptive/antisocial	.198	26	.010	.887	26	.008
DBC_self_absorbed	.199	26	.009	.783	26	.000
Communication disturbance	.279	26	.000	.746	26	.000
DBC_anxiety	.247	26	.000	.862	26	.002
DBC_social_relating	.325	26	.000	.708	26	.000
Total score	.193	26	.014	.864	26	.003
Child_dependency	.296	26	.000	.795	26	.000

*. This is a lower bound of the true significance.
a. Lilliefors Significance Correction

6.1.3 Normality statistics control group

Tests of Normality

	Kolmogorov-Smirnov[a]			Shapiro-Wilk		
	Statistic	df	Sig.	Statistic	df	Sig.
Significant Other	.319	30	.000	.603	30	.000
Family	.231	30	.000	.776	30	.000
Friends	.251	30	.000	.702	30	.000
Total score	.258	30	.000	.678	30	.000
Problem t score	.223	30	.001	.802	30	.000
Intensity t score	.095	30	.200*	.968	30	.481
C_GHQ12_total	.134	30	.179	.971	30	.577
Total score	.125	30	.200*	.950	30	.166

*. This is a lower bound of the true significance.

a. Lilliefors Significance Correction

6.2 Descriptive statistics for experimental and control groups

Descriptive Statistics

	N	Range	Mean		Std. Deviation
	Statistic	Statistic	Statistic	Std. Error	Statistic
Ex_MPSS_SO	38	24.00	24.1842	.91211	5.62261
Significant Other	32	24.00	24.6563	1.13469	6.41877
Ex_MPSS_Fa	38	24.00	22.2632	.90336	5.56866
Family	32	24.00	23.3750	1.01575	5.74597
Ex_MPSS_Fr	38	23.00	21.3947	1.02912	6.34392
Friends	32	24.00	24.1563	1.06076	6.00059
Ex_MPSS_Total	38	68.00	67.8421	2.28344	14.07610
Total score	32	72.00	72.1562	3.07471	17.39319
Ex_ECBI_Problem	34	29.00	6.5588	1.33524	7.78573
Problem t score	30	17.00	3.6000	.81452	4.46133
Ex_ECBI_Intensity	34	126.00	103.5588	5.32244	31.03487
Intensity t score	30	78.00	88.6333	3.92237	21.48373
Ex_GHQ12_Total	38	14.00	10.2895	.47991	2.95834
C_GHQ12_total	32	16.00	9.6250	.64719	3.66104
Ex_RSA_Total	38	93.00	179.3158	3.89315	23.99899

Total score	32	85.00	191.0625	3.56165	20.14774
Disruptive/antisocial	28	27.00	7.1786	1.29718	6.86404
DBC_self_absorbed	28	21.00	3.8929	.84255	4.45836
Communication disturbance	28	10.00	1.7857	.46107	2.43975
DBC_anxiety	28	11.00	2.8929	.54272	2.87182
DBC_social_relating	28	9.00	1.5357	.41938	2.21914
Total score	28	60.00	18.8214	3.08738	16.33686
Child_dependency	38	6.00	5.8947	.27917	1.72093
Valid N (listwise)	18				

6.3 Independent t-test results using bootstrapping

Group Statistics

	GROUP		Statistic	Bootstrap[a]			
				Bias	Std. Error	95% Confidence Interval	
						Lower	Upper
GHQ12_total	Control	N	30				
		Mean	9.7000	-.0072	.6595	8.4287	11.0000
		Std. Deviation	3.65919	-.09415	.48420	2.49780	4.46526
		Std. Error Mean	.66807				
	Experimental	N	36				
		Mean	10.4722	-.0141	.4882	9.5627	11.4496
		Std. Deviation	2.93244	-.07436	.36855	2.11962	3.55128
		Std. Error Mean	.48874				
Int_ECBI_Raw	Control	N	30				
		Mean	88.6333	-.0656	3.8911	80.4253	96.1411
		Std. Deviation	21.48373	-.50451	2.05928	16.62855	24.80325
		Std. Error Mean	3.92237				
	Experimental	N	36				
		Mean	101.5278	.0757	5.1401	91.7634	111.9716
		Std. Deviation	31.31270	-.43494	3.35083	23.77832	37.50389
		Std. Error Mean	5.21878				
Prob_ECBI_Raw	Control	N	30				
		Mean	3.6000	.0194	.8244	2.0371	5.3124

	Experimental	Std. Deviation	4.46133	-.13584	.73952	2.84517	5.80226
		Std. Error Mean	.81452				
		N	36				
		Mean	6.3333	.0444	1.2442	4.0338	9.0000
		Std. Deviation	7.61952	-.14152	1.21510	4.83580	9.67130
		Std. Error Mean	1.26992				

a. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

		Levene's Test for Equality of Variances						
		F	Sig.	t	df	Sig. (2-tailed)	95% Confidence Interval of the Difference	
							Lower	Upper
GHQ12_total	Equal variances assumed	.965	.329	-.840	68	.404	-2.24313	.91418
	Equal variances not assumed			-.825	59.414	.413	-2.27645	.94750

Bootstrap for Independent Samples Test

		Mean Difference	Bootstrap[a]				
			Bias	Std. Error	Sig. (2-tailed)	95% Confidence Interval	
						Lower	Upper
GHQ12_total	Equal variances assumed	-.66447	.02351	.76313	.404	-2.10584	.93174
	Equal variances not assumed	-.66447	.02351	.76313	.404	-2.10584	.93174

a. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

		Levene's Test for Equality of Variances		t	df	Sig. (2-tailed)	95% Confidence Interval	
		F	Sig.				Lower	Upper
Int_ECBI_Raw	Equal variances assumed	3.190	.079	-1.911	64	.061	-26.37708	.58819
	Equal variances not assumed			-1.975	61.880	.053	-25.94514	.15625

Prob_ECBI_Raw	Equal variances assumed	4.886	.031	-1.732	64	.088	-5.88660	
	Equal variances not assumed			-1.812	57.895	.075	-5.75342	

Bootstrap for Independent Samples Test

		Mean Difference	Bootstrap[a]				
			Bias	Std. Error	Sig. (2-tailed)	95% Confidence Interval	
						Lower	Upper
Int_ECBI_Raw	Equal variances assumed	-12.89444	.26557	6.29538	.049	-24.58161	-.36088
	Equal variances not assumed	-12.89444	.26557	6.29538	.049	-24.58161	-.36088

a. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

Bootstrap for Independent Samples Test

		Mean Difference	Bootstrap[a]				
			Bias	Std. Error	Sig. (2-tailed)	95% Confidence Interval	
						Lower	Upper
Prob_ECBI_Raw	Equal variances assumed	-2.73333	-.09832	1.51507	.077	-5.95445	.03187
	Equal variances not assumed	-2.73333	-.09832	1.51507	.076	-5.95445	.03187

a. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

6.4 Correlation analysis with bootstrapping for experimental group

```
BOOTSTRAP
  /SAMPLING METHOD=SIMPLE
  /VARIABLES INPUT=Ex_MPSS_SO Ex_MPSS_Fa Ex_MPSS_Fr Ex_MPSS_Total Ex_ECBI_Problem Ex_ECBI_Intensity Ex_GHQ12_Total Ex_RSA_Total D
BC_distruptive_antisocial DBC_self_absorbed DBC_communication DBC_anxiety DBC_social_relating DBC_total Child_dependency
  /CRITERIA CILEVEL=95 CITYPE=PERCENTILE  NSAMPLES=1000
  /MISSING USERMISSING=EXCLUDE.
```

Bootstrap

```
[DataSet1] /Users/Olivia/Desktop/SPSS thesis amendments/August dataset 1 use this one.sav
```

Bootstrap Specifications

Sampling Method	Simple
Number of Samples	1000
Confidence Interval Level	95.0%
Confidence Interval Type	Percentile

```
CORRELATIONS
  /VARIABLES=Ex_MPSS_SO Ex_MPSS_Fa Ex_MPSS_Fr Ex_MPSS_Total Ex_ECBI_Problem Ex_ECBI_Intensity Ex_GHQ12_Total Ex_RSA_Total DBC_dis
truptive_antisocial DBC_self_absorbed DBC_communication DBC_anxiety DBC_social_relating DBC_total Child_dependency
  /PRINT=TWOTAIL NOSIG
  /MISSING=PAIRWISE.
```

Correlations

```
[DataSet1] /Users/Olivia/Desktop/SPSS thesis amendments/August dataset 1 use this one.sav
```

Correlations

				Ex_MPSS_SO	Ex_MPSS_Fa	Ex_MPSS_Fr	Ex_MPSS_Total	Ex_ECBI_Problem	Ex_ECBI_Intensity	Ex_GHQ12_Total
Ex_MPSS_SO	Pearson Correlation			1	.606**	.488*	.864**	-.136	-.317	-.077
	Sig. (2-tailed)				.001	.011	.000	.506	.115	.710
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		0	-.067	-.046	-.042	-.002	.004	-.035
		Std. Error		0	.287	.212	.112	.159	.272	.163
		95% Confidence Interval	Lower	1	-.093	.009	.575	-.421	-.700	-.537
			Upper	1	.937	.807	.962	.186	.292	.143
Ex_MPSS_Fa	Pearson Correlation			.606**	1	.302	.767**	-.350	-.494*	-.310
	Sig. (2-tailed)			.001		.133	.000	.080	.010	.124
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.067	0	-.050	-.058	-.007	.029	-.023
		Std. Error		.287	0	.287	.174	.187	.178	.197
		95% Confidence Interval	Lower	-.093	1	-.309	.313	-.751	-.770	-.693
			Upper	.937	1	.745	.945	-.001	-.058	.006
Ex_MPSS_Fr	Pearson Correlation			.488*	.302	1	.774**	.240	-.082	.068
	Sig. (2-tailed)			.011	.133		.000	.238	.690	.743
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.046	-.050	0	-.007	.011	.015	.015
		Std. Error		.212	.287	0	.103	.161	.211	.179
		95% Confidence Interval	Lower	.009	-.309	1	.498	-.114	-.473	-.294
			Upper	.807	.745	1	.914	.527	.316	.416
Ex_MPSS_Total	Pearson Correlation			.864**	.767**	.774**	1	-.079	-.355	-.118
	Sig. (2-tailed)			.000	.000	.000		.703	.075	.566
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.042	-.058	-.007	0	.019	.040	-.028
		Std. Error		.112	.174	.103	0	.183	.215	.172
		95% Confidence Interval	Lower	.575	.313	.498	1	-.400	-.667	-.517
			Upper	.962	.945	.914	1	.334	.155	.144
Ex_ECBI_Problem	Pearson Correlation			-.136	-.350	.240	-.079	1	.719**	.107
	Sig. (2-tailed)			.506	.080	.238	.703		.000	.603
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.002	-.007	.011	.019	0	-.014	.015
		Std. Error		.159	.187	.161	.183	0	.112	.206

Correlations

				Ex_RSA_Total	Disruptive/antisocial	DBC_self_absorbed	Communication disturbance	DBC_anxiety	DBC_social_relating	Total score
Ex_MPSS_SO	Pearson Correlation			.003	-.192	-.602**	-.380	-.283	-.153	-.398*
	Sig. (2-tailed)			.990	.348	.001	.055	.162	.457	.044
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.066	-.012	.113	.025	-.013	.007	.021
		Std. Error		.264	.152	.328	.232	.159	.186	.216
		95% Confidence Interval	Lower	-.355	-.498	-.905	-.747	-.603	-.486	-.718
			Upper	.633	.109	.232	.121	.031	.257	.107
Ex_MPSS_Fa	Pearson Correlation			.168	-.287	-.731**	-.656**	-.364	-.328	-.551**
	Sig. (2-tailed)			.412	.156	.000	.000	.068	.102	.004
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.040	-.004	.083	.005	-.008	.011	.027
		Std. Error		.290	.147	.220	.134	.125	.169	.160
		95% Confidence Interval	Lower	-.325	-.571	-.901	-.869	-.608	-.575	-.773
			Upper	.700	-.019	-.113	-.330	-.120	.115	-.158
Ex_MPSS_Fr	Pearson Correlation			.030	.083	-.347	-.166	-.108	.200	-.088
	Sig. (2-tailed)			.885	.688	.083	.417	.600	.326	.671
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.022	.006	.085	.008	-.003	.013	.024
		Std. Error		.261	.167	.289	.227	.177	.176	.221
		95% Confidence Interval	Lower	-.481	-.268	-.705	-.602	-.479	-.169	-.486
			Upper	.531	.400	.361	.245	.204	.513	.336
Ex_MPSS_Total	Pearson Correlation			.078	-.149	-.683**	-.481*	-.303	-.095	-.412*
	Sig. (2-tailed)			.703	.466	.000	.013	.132	.645	.036
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.066	.013	.132	.028	.000	.038	.049
		Std. Error		.311	.142	.295	.188	.137	.219	.204
		95% Confidence Interval	Lower	-.379	-.396	-.885	-.792	-.563	-.426	-.694
			Upper	.744	.151	.095	-.065	-.020	.432	.063
Ex_ECBI_Problem	Pearson Correlation			-.036	.648**	.551**	.368	.256	.483*	.592**
	Sig. (2-tailed)			.862	.000	.003	.064	.207	.012	.001
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.031	-.040	.027	-.001	-.001	-.003	-.018
		Std. Error		.238	.181	.122	.184	.175	.148	.141

Correlations

				Child_dependency
Ex_MPSS_SO	Pearson Correlation			-.095
	Sig. (2-tailed)			.645
	N			26
	Bootstrap[c]	Bias		.011
		Std. Error		.196
		95% Confidence Interval	Lower	-.391
			Upper	.355
Ex_MPSS_Fa	Pearson Correlation			-.282
	Sig. (2-tailed)			.163
	N			26
	Bootstrap[c]	Bias		.008
		Std. Error		.136
		95% Confidence Interval	Lower	-.519
			Upper	.033
Ex_MPSS_Fr	Pearson Correlation			.105
	Sig. (2-tailed)			.609
	N			26
	Bootstrap[c]	Bias		.007
		Std. Error		.208
		95% Confidence Interval	Lower	-.248
			Upper	.536
Ex_MPSS_Total	Pearson Correlation			-.098
	Sig. (2-tailed)			.635
	N			26
	Bootstrap[c]	Bias		.024
		Std. Error		.211
		95% Confidence Interval	Lower	-.404
			Upper	.431
Ex_ECBI_Problem	Pearson Correlation			.396*
	Sig. (2-tailed)			.045
	N			26
	Bootstrap[c]	Bias		-.002
		Std. Error		.140

Correlations

				Ex_MPSS_SO	Ex_MPSS_Fa	Ex_MPSS_Fr	Ex_MPSS_Total	Ex_ECBI_Problem	Ex_ECBI_Intensity	Ex_GHQ12_Total
		95% Confidence Interval	Lower	-.421	-.751	-.114	-.400	1	.409	-.262
			Upper	.186	-.001	.527	.334	1	.857	.529
Ex_ECBI_Intensity	Pearson Correlation			-.317	-.494*	-.082	-.355	.719**	1	.175
	Sig. (2-tailed)			.115	.010	.690	.075	.000		.394
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.004	.029	.015	.040	-.014	0	.001
		Std. Error		.272	.178	.211	.215	.112	0	.204
		95% Confidence Interval	Lower	-.700	-.770	-.473	-.667	.409	1	-.222
			Upper	.292	-.058	.316	.155	.857	1	.550
Ex_GHQ12_Total	Pearson Correlation			-.077	-.310	.068	-.118	.107	.175	1
	Sig. (2-tailed)			.710	.124	.743	.566	.603	.394	
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.035	-.023	.015	-.028	.015	.001	0
		Std. Error		.163	.197	.179	.172	.206	.204	0
		95% Confidence Interval	Lower	-.537	-.693	-.294	-.517	-.262	-.222	1
			Upper	.143	.006	.416	.144	.529	.550	1
Ex_RSA_Total	Pearson Correlation			.003	.168	.030	.078	-.036	-.187	-.515**
	Sig. (2-tailed)			.990	.412	.885	.703	.862	.362	.007
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.066	.040	-.022	.066	-.031	-.022	.007
		Std. Error		.264	.290	.261	.311	.238	.207	.185
		95% Confidence Interval	Lower	-.355	-.325	-.481	-.379	-.556	-.621	-.836
			Upper	.633	.700	.531	.744	.374	.196	-.097
Disruptive/antisocial	Pearson Correlation			-.192	-.287	.083	-.149	.648**	.665**	.232
	Sig. (2-tailed)			.348	.156	.688	.466	.000	.000	.255
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.012	-.004	.006	.013	-.040	-.018	.011
		Std. Error		.152	.147	.167	.142	.181	.144	.201
		95% Confidence Interval	Lower	-.498	-.571	-.268	-.396	.171	.323	-.138
			Upper	.109	-.019	.400	.151	.857	.851	.667
DBC_self_absorbed	Pearson Correlation			-.602**	-.731**	-.347	-.683**	.551**	.728**	.111
	Sig. (2-tailed)			.001	.000	.083	.000	.003	.000	.591
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.113	.083	.085	.132	.027	.010	.034
		Std. Error		.328	.220	.289	.295	.122	.081	.200

Correlations

				Ex_RSA_Total	Disruptive/antisocial	DBC_self_absorbed	Communication disturbance	DBC_anxiety	DBC_social_relating	Total score
		95% Confidence Interval	Lower	-.556	.171	.331	-.017	-.087	.134	.250
			Upper	.374	.857	.819	.710	.612	.727	.789
Ex_ECBI_Intensity	Pearson Correlation			-.187	.665**	.728**	.564**	.375	.379	.706**
	Sig. (2-tailed)			.362	.000	.000	.003	.059	.056	.000
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.022	-.018	.010	-.015	.003	-.017	-.019
		Std. Error		.207	.144	.081	.139	.214	.210	.136
		95% Confidence Interval	Lower	-.621	.323	.541	.216	-.069	-.113	.357
			Upper	.196	.851	.854	.779	.751	.699	.879
Ex_GHQ12_Total	Pearson Correlation			-.515**	.232	.111	.267	.452*	.105	.280
	Sig. (2-tailed)			.007	.255	.591	.187	.020	.609	.166
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.007	.011	.034	.014	-.039	-.008	.002
		Std. Error		.185	.201	.200	.149	.217	.246	.168
		95% Confidence Interval	Lower	-.836	-.138	-.197	.013	-.060	-.407	-.033
			Upper	-.097	.667	.618	.623	.746	.559	.641
Ex_RSA_Total	Pearson Correlation			1	-.099	-.096	-.217	-.185	.000	-.153
	Sig. (2-tailed)				.630	.642	.287	.365	.999	.454
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		0	-.023	-.069	-.013	-.006	.002	-.014
		Std. Error		0	.201	.305	.201	.159	.177	.205
		95% Confidence Interval	Lower	1	-.528	-.784	-.614	-.505	-.339	-.587
			Upper	1	.232	.365	.189	.125	.376	.234
Disruptive/antisocial	Pearson Correlation			-.099	1	.586**	.610**	.538**	.707**	.900**
	Sig. (2-tailed)			.630		.002	.001	.005	.000	.000
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.023	0	.031	.016	-.005	.010	.007
		Std. Error		.201	0	.127	.156	.148	.088	.031
		95% Confidence Interval	Lower	-.528	1	.362	.315	.190	.504	.833
			Upper	.232	1	.860	.885	.788	.863	.959
DBC_self_absorbed	Pearson Correlation			-.096	.586**	1	.674**	.376	.452*	.772**
	Sig. (2-tailed)			.642	.002		.000	.058	.020	.000
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.069	.031	0	.000	.008	.010	.004
		Std. Error		.305	.127	0	.116	.192	.166	.100

Correlations

				Child_dependency
		95% Confidence Interval	Lower	.072
			Upper	.643
Ex_ECBI_Intensity	Pearson Correlation			.498**
	Sig. (2-tailed)			.010
	N			26
	Bootstrap[c]	Bias		.001
		Std. Error		.119
		95% Confidence Interval	Lower	.236
			Upper	.712
Ex_GHQ12_Total	Pearson Correlation			.169
	Sig. (2-tailed)			.411
	N			26
	Bootstrap[c]	Bias		.014
		Std. Error		.182
		95% Confidence Interval	Lower	-.217
			Upper	.511
Ex_RSA_Total	Pearson Correlation			-.134
	Sig. (2-tailed)			.513
	N			26
	Bootstrap[c]	Bias		-.024
		Std. Error		.242
		95% Confidence Interval	Lower	-.581
			Upper	.329
Disruptive/antisocial	Pearson Correlation			.330
	Sig. (2-tailed)			.099
	N			26
	Bootstrap[c]	Bias		.004
		Std. Error		.132
		95% Confidence Interval	Lower	.060
			Upper	.580
DBC_self_absorbed	Pearson Correlation			.303
	Sig. (2-tailed)			.133
	N			26
	Bootstrap[c]	Bias		.009
		Std. Error		.152

Correlations

				Ex_MPSS_SO	Ex_MPSS_Fa	Ex_MPSS_Fr	Ex_MPSS_Total	Ex_ECBI_Problem	Ex_ECBI_Intensity	Ex_GHQ12_Total
Communication disturbance		95% Confidence Interval	Lower	-.905	-.901	-.705	-.885	.331	.541	-.197
			Upper	.232	-.113	.361	.095	.819	.854	.618
	Pearson Correlation			-.380	-.656**	-.166	-.481*	.368	.564**	.267
	Sig. (2-tailed)			.055	.000	.417	.013	.064	.003	.187
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.025	.005	.008	.028	-.001	-.015	.014
		Std. Error		.232	.134	.227	.188	.184	.139	.149
		95% Confidence Interval	Lower	-.747	-.869	-.602	-.792	-.017	.216	.013
			Upper	.121	-.330	.245	-.065	.710	.779	.623
DBC_anxiety	Pearson Correlation			-.283	-.364	-.108	-.303	.256	.375	.452*
	Sig. (2-tailed)			.162	.068	.600	.132	.207	.059	.020
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.013	-.008	-.003	.000	-.001	.003	-.039
		Std. Error		.159	.125	.177	.137	.175	.214	.217
		95% Confidence Interval	Lower	-.603	-.608	-.479	-.563	-.087	-.069	-.060
			Upper	.031	-.120	.204	-.020	.612	.751	.746
DBC_social_relating	Pearson Correlation			-.153	-.328	.200	-.095	.483*	.379	.105
	Sig. (2-tailed)			.457	.102	.326	.645	.012	.056	.609
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.007	.011	.013	.038	-.003	-.017	-.008
		Std. Error		.186	.169	.176	.219	.148	.210	.246
		95% Confidence Interval	Lower	-.486	-.575	-.169	-.426	.134	-.113	-.407
			Upper	.257	.115	.513	.432	.727	.699	.559
Total score	Pearson Correlation			-.398*	-.551**	-.088	-.412*	.592**	.706**	.280
	Sig. (2-tailed)			.044	.004	.671	.036	.001	.000	.166
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.021	.027	.024	.049	-.018	-.019	.002
		Std. Error		.216	.160	.221	.204	.141	.136	.168
		95% Confidence Interval	Lower	-.718	-.773	-.486	-.694	.250	.357	-.033
			Upper	.107	-.158	.336	.063	.789	.879	.641
Child_dependency	Pearson Correlation			-.095	-.282	.105	-.098	.396*	.498**	.169
	Sig. (2-tailed)			.645	.163	.609	.635	.045	.010	.411
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.011	.008	.007	.024	-.002	.001	.014
		Std. Error		.196	.136	.208	.211	.140	.119	.182

Correlations

				Ex_RSA_Total	Disruptive/antisocial	DBC_self_absorbed	Communication disturbance	DBC_anxiety	DBC_social_relating	Total score
Communication disturbance		95% Confidence Interval	Lower	-.784	.362	1	.373	.018	.075	.537
			Upper	.365	.860	1	.860	.751	.763	.924
	Pearson Correlation			-.217	.610**	.674**	1	.533**	.619**	.819**
	Sig. (2-tailed)			.287	.001	.000		.005	.001	.000
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.013	.016	.000	0	.006	-.060	-.012
		Std. Error		.201	.156	.116	0	.123	.229	.109
		95% Confidence Interval	Lower	-.614	.315	.373	1	.319	.039	.534
			Upper	.189	.885	.860	1	.790	.880	.965
DBC_anxiety	Pearson Correlation			-.185	.538**	.376	.533**	1	.561**	.717**
	Sig. (2-tailed)			.365	.005	.058	.005		.003	.000
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.006	-.005	.008	.006	0	-.009	.002
		Std. Error		.159	.148	.192	.123	0	.184	.087
		95% Confidence Interval	Lower	-.505	.190	.018	.319	1	.062	.530
			Upper	.125	.788	.751	.790	1	.810	.876
DBC_social_relating	Pearson Correlation			.000	.707**	.452*	.619**	.561**	1	.792**
	Sig. (2-tailed)			.999	.000	.020	.001	.003		.000
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		.002	.010	.010	-.060	-.009	0	-.008
		Std. Error		.177	.088	.166	.229	.184	0	.098
		95% Confidence Interval	Lower	-.339	.504	.075	.039	.062	1	.528
			Upper	.376	.863	.763	.880	.810	1	.904
Total score	Pearson Correlation			-.153	.900**	.772**	.819**	.717**	.792**	1
	Sig. (2-tailed)			.454	.000	.000	.000	.000	.000	
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.014	.007	.004	-.012	.002	-.008	0
		Std. Error		.205	.031	.100	.109	.087	.098	0
		95% Confidence Interval	Lower	-.587	.833	.537	.534	.530	.528	1
			Upper	.234	.959	.924	.965	.876	.904	1
Child_dependency	Pearson Correlation			-.134	.330	.303	.284	.480*	.425*	.417*
	Sig. (2-tailed)			.513	.099	.133	.160	.013	.030	.034
	N			26	26	26	26	26	26	26
	Bootstrap[c]	Bias		-.024	.004	.009	.000	.004	.003	.004
		Std. Error		.242	.132	.152	.123	.117	.100	.128

Correlations

				Child_dependency
Communication disturbance		95% Confidence Interval	Lower	-.025
			Upper	.580
	Pearson Correlation			.284
	Sig. (2-tailed)			.160
	N			26
	Bootstrap[c]	Bias		.000
		Std. Error		.123
		95% Confidence Interval	Lower	.019
			Upper	.525
DBC_anxiety	Pearson Correlation			.480*
	Sig. (2-tailed)			.013
	N			26
	Bootstrap[c]	Bias		.004
		Std. Error		.117
		95% Confidence Interval	Lower	.227
			Upper	.690
DBC_social_relating	Pearson Correlation			.425*
	Sig. (2-tailed)			.030
	N			26
	Bootstrap[c]	Bias		.003
		Std. Error		.100
		95% Confidence Interval	Lower	.232
			Upper	.623
Total score	Pearson Correlation			.417*
	Sig. (2-tailed)			.034
	N			26
	Bootstrap[c]	Bias		.004
		Std. Error		.128
		95% Confidence Interval	Lower	.160
			Upper	.643
Child_dependency	Pearson Correlation			1
	Sig. (2-tailed)			
	N			26
	Bootstrap[c]	Bias		0
		Std. Error		0

Correlations

			Ex_MPSS_SO	Ex_MPSS_Fa	Ex_MPSS_Fr	Ex_MPSS_Total	Ex_ECBI_Problem	Ex_ECBI_Intensity	Ex_GHQ12_Total
	95% Confidence Interval	Lower	-.391	-.519	-.248	-.404	.072	.236	-.217
		Upper	.355	.033	.536	.431	.643	.712	.511

Correlations

			Ex_RSA_Total	Disruptive/antisocial	DBC_self_absorbed	Communication disturbance	DBC_anxiety	DBC_social_relating	Total score
	95% Confidence Interval	Lower	-.581	.060	-.025	.019	.227	.232	.160
		Upper	.329	.580	.580	.525	.690	.623	.643

Correlations

			Child_dependency
	95% Confidence Interval	Lower	1
		Upper	1

**. Correlation is significant at the 0.01 level (2-tailed).

*. Correlation is significant at the 0.05 level (2-tailed).

c. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

Correlations

				Ex_Age_Child	Ex_ECBI_Problem	Ex_ECBI_Intensity	Disruptive/antisocial	DBC_self_absorbed	Communication disturbance
Ex_Age_Child	Pearson Correlation			1	-.021	-.486*	-.135	-.452*	-.307
	Sig. (2-tailed)				.917	.012	.510	.020	.127
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		0	-.002	-.003	-.013	-.008	-.013
		Std. Error		0	.156	.125	.169	.130	.161
		95% Confidence Interval	Lower	1	-.314	-.706	-.454	-.677	-.631
			Upper	1	.281	-.226	.192	-.168	-.008
Ex_ECBI_Problem	Pearson Correlation			-.021	1	.719**	.648**	.551**	.368
	Sig. (2-tailed)			.917		.000	.000	.003	.064
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		-.002	0	-.015	-.037	.031	.010
		Std. Error		.156	0	.115	.182	.124	.186
		95% Confidence Interval	Lower	-.314	1	.435	.179	.332	.003
			Upper	.281	1	.853	.861	.825	.739
Ex_ECBI_Intensity	Pearson Correlation			-.486*	.719**	1	.665**	.728**	.564**
	Sig. (2-tailed)			.012	.000		.000	.000	.003
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		-.003	-.015	0	-.013	.012	-.006
		Std. Error		.125	.115	0	.135	.083	.141
		95% Confidence Interval	Lower	-.706	.435	1	.336	.538	.210
			Upper	-.226	.853	1	.843	.875	.784
Disruptive/antisocial	Pearson Correlation			-.135	.648**	.665**	1	.586**	.610**
	Sig. (2-tailed)			.510	.000	.000		.002	.001
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		-.013	-.037	-.013	0	.039	.023
		Std. Error		.169	.182	.135	0	.123	.156
		95% Confidence Interval	Lower	-.454	.179	.336	1	.401	.330
			Upper	.192	.861	.843	1	.868	.897
DBC_self_absorbed	Pearson Correlation			-.452*	.551**	.728**	.586**	1	.674**
	Sig. (2-tailed)			.020	.003	.000	.002		.000
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		-.008	.031	.012	.039	0	.007
		Std. Error		.130	.124	.083	.123	0	.114

Correlations

				DBC_anxiety	DBC_social_relating	Total score
Ex_Age_Child	Pearson Correlation			-.222	.124	-.256
	Sig. (2-tailed)			.277	.548	.206
	N			26	26	26
	Bootstrap[c]	Bias		-.022	-.002	-.006
		Std. Error		.251	.181	.179
		95% Confidence Interval	Lower	-.716	-.243	-.576
			Upper	.212	.491	.122
Ex_ECBI_Problem	Pearson Correlation			.256	.483*	.592**
	Sig. (2-tailed)			.207	.012	.001
	N			26	26	26
	Bootstrap[c]	Bias		.003	.003	-.013
		Std. Error		.170	.145	.141
		95% Confidence Interval	Lower	-.070	.170	.227
			Upper	.575	.750	.788
Ex_ECBI_Intensity	Pearson Correlation			.375	.379	.706**
	Sig. (2-tailed)			.059	.056	.000
	N			26	26	26
	Bootstrap[c]	Bias		.009	-.015	-.014
		Std. Error		.210	.204	.133
		95% Confidence Interval	Lower	-.031	-.102	.371
			Upper	.766	.689	.883
Disruptive/antisocial	Pearson Correlation			.538**	.707**	.900**
	Sig. (2-tailed)			.005	.000	.000
	N			26	26	26
	Bootstrap[c]	Bias		-.002	.008	.008
		Std. Error		.147	.088	.031
		95% Confidence Interval	Lower	.189	.498	.840
			Upper	.778	.865	.961
DBC_self_absorbed	Pearson Correlation			.376	.452*	.772**
	Sig. (2-tailed)			.058	.020	.000
	N			26	26	26
	Bootstrap[c]	Bias		.016	.015	.010
		Std. Error		.185	.163	.096

Correlations

				Ex_Age_Child	Ex_ECBI_Problem	Ex_ECBI_Intensity	Disruptive/antisocial	DBC_self_absorbed	Communication disturbance
Communication disturbance		95% Confidence Interval	Lower	-.677	.332	.538	.401	1	.394
			Upper	-.168	.825	.875	.868	1	.876
	Pearson Correlation			-.307	.368	.564**	.610**	.674**	1
	Sig. (2-tailed)			.127	.064	.003	.001	.000	
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		-.013	.010	-.006	.023	.007	0
		Std. Error		.161	.186	.141	.156	.114	0
		95% Confidence Interval	Lower	-.631	.003	.210	.330	.394	1
			Upper	-.008	.739	.784	.897	.876	1
DBC_anxiety	Pearson Correlation			-.222	.256	.375	.538**	.376	.533**
	Sig. (2-tailed)			.277	.207	.059	.005	.058	.005
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		-.022	.003	.009	-.002	.016	.010
		Std. Error		.251	.170	.210	.147	.185	.129
		95% Confidence Interval	Lower	-.716	-.070	-.031	.189	.030	.293
			Upper	.212	.575	.766	.778	.748	.812
DBC_social_relating	Pearson Correlation			.124	.483*	.379	.707**	.452*	.619**
	Sig. (2-tailed)			.548	.012	.056	.000	.020	.001
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		-.002	.003	-.015	.008	.015	-.062
		Std. Error		.181	.145	.204	.088	.163	.231
		95% Confidence Interval	Lower	-.243	.170	-.102	.498	.070	.019
			Upper	.491	.750	.689	.865	.763	.881
Total score	Pearson Correlation			-.256	.592**	.706**	.900**	.772**	.819**
	Sig. (2-tailed)			.206	.001	.000	.000	.000	.000
	N			26	26	26	26	26	26
	Bootstrap[c]	Bias		-.006	-.013	-.014	.008	.010	-.008
		Std. Error		.179	.141	.133	.031	.096	.111
		95% Confidence Interval	Lower	-.576	.227	.371	.840	.544	.540
			Upper	.122	.788	.883	.961	.931	.967

Correlations

				DBC_anxiety	DBC_social_relating	Total score
Communication disturbance		95% Confidence Interval	Lower	.030	.070	.544
			Upper	.748	.763	.931
	Pearson Correlation			.533**	.619**	.819**
	Sig. (2-tailed)			.005	.001	.000
	N			26	26	26
	Bootstrap[c]	Bias		.010	-.062	-.008
		Std. Error		.129	.231	.111
		95% Confidence Interval	Lower	.293	.019	.540
			Upper	.812	.881	.967
DBC_anxiety	Pearson Correlation			1	.561**	.717**
	Sig. (2-tailed)				.003	.000
	N			26	26	26
	Bootstrap[c]	Bias		0	-.011	.003
		Std. Error		0	.182	.088
		95% Confidence Interval	Lower	1	.091	.518
			Upper	1	.816	.868
DBC_social_relating	Pearson Correlation			.561**	1	.792**
	Sig. (2-tailed)			.003		.000
	N			26	26	26
	Bootstrap[c]	Bias		-.011	0	-.010
		Std. Error		.182	0	.095
		95% Confidence Interval	Lower	.091	1	.545
			Upper	.816	1	.907
Total score	Pearson Correlation			.717**	.792**	1
	Sig. (2-tailed)			.000	.000	
	N			26	26	26
	Bootstrap[c]	Bias		.003	-.010	0
		Std. Error		.088	.095	0
		95% Confidence Interval	Lower	.518	.545	1
			Upper	.868	.907	1

*. Correlation is significant at the 0.05 level (2-tailed).

**. Correlation is significant at the 0.01 level (2-tailed).

c. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

Correlations between variables for control group using bootstrapping

Correlations

				Significant Other	Family	Friends	Total score	Problem t score	Intensity t score	C_GHQ12_total
Significant Other	Pearson Correlation			1	.911**	.874**	.972**	-.081	-.242	-.027
	Sig. (2-tailed)				.000	.000	.000	.671	.199	.886
	N			30	30	30	30	30	30	30
	Bootstrap[c]	Bias		0	-.018	-.032	-.008	-.021	-.005	-.009
		Std. Error		0	.075	.134	.036	.148	.099	.181
		95% Confidence Interval	Lower	1	.676	.336	.854	-.441	-.437	-.382
			Upper	1	.967	.965	.988	.156	-.053	.295
Family	Pearson Correlation			.911**	1	.840**	.956**	-.016	-.224	-.095
	Sig. (2-tailed)			.000		.000	.000	.932	.235	.618
	N			30	30	30	30	30	30	30
	Bootstrap[c]	Bias		-.018	0	-.036	-.009	-.024	-.005	-.020
		Std. Error		.075	0	.135	.037	.151	.108	.195
		95% Confidence Interval	Lower	.676	1	.387	.840	-.382	-.438	-.506
			Upper	.967	1	.944	.987	.228	-.009	.240
Friends	Pearson Correlation			.874**	.840**	1	.945**	-.085	-.209	-.060
	Sig. (2-tailed)			.000	.000		.000	.655	.268	.753
	N			30	30	30	30	30	30	30
	Bootstrap[c]	Bias		-.032	-.036	0	-.016	-.026	-.006	-.016
		Std. Error		.134	.135	0	.062	.150	.112	.189
		95% Confidence Interval	Lower	.336	.387	1	.711	-.439	-.423	-.464
			Upper	.965	.944	1	.983	.156	.032	.282
Total score	Pearson Correlation			.972**	.956**	.945**	1	-.071	-.235	-.061
	Sig. (2-tailed)			.000	.000	.000		.711	.211	.748
	N			30	30	30	30	30	30	30
	Bootstrap[c]	Bias		-.008	-.009	-.016	0	-.026	-.007	-.020
		Std. Error		.036	.037	.062	0	.147	.095	.193
		95% Confidence Interval	Lower	.854	.840	.711	1	-.436	-.429	-.472
			Upper	.988	.987	.983	1	.162	-.039	.267
Problem t score	Pearson Correlation			-.081	-.016	-.085	-.071	1	.217	-.236
	Sig. (2-tailed)			.671	.932	.655	.711		.249	.210
	N			30	30	30	30	30	30	30
	Bootstrap[c]	Bias		-.021	-.024	-.026	-.026	0	.019	.006
		Std. Error		.148	.151	.150	.147	0	.160	.162

Correlations

				Total score
Significant Other	Pearson Correlation			.235
	Sig. (2-tailed)			.212
	N			30
	Bootstrap[c]	Bias		.014
		Std. Error		.277
		95% Confidence Interval	Lower	-.257
			Upper	.776
Family	Pearson Correlation			.204
	Sig. (2-tailed)			.280
	N			30
	Bootstrap[c]	Bias		.038
		Std. Error		.264
		95% Confidence Interval	Lower	-.213
			Upper	.773
Friends	Pearson Correlation			.176
	Sig. (2-tailed)			.352
	N			30
	Bootstrap[c]	Bias		.022
		Std. Error		.264
		95% Confidence Interval	Lower	-.266
			Upper	.712
Total score	Pearson Correlation			.214
	Sig. (2-tailed)			.256
	N			30
	Bootstrap[c]	Bias		.035
		Std. Error		.287
		95% Confidence Interval	Lower	-.239
			Upper	.815
Problem t score	Pearson Correlation			-.077
	Sig. (2-tailed)			.687
	N			30
	Bootstrap[c]	Bias		-.008
		Std. Error		.193

Correlations

				Significant Other	Family	Friends	Total score	Problem t score	Intensity t score	C_GHQ12_total
Intensity t score		95% Confidence Interval	Lower	-.441	-.382	-.439	-.436	1	-.056	-.524
			Upper	.156	.228	.156	.162	1	.549	.120
	Pearson Correlation			-.242	-.224	-.209	-.235	.217	1	.017
	Sig. (2-tailed)			.199	.235	.268	.211	.249		.927
	N			30	30	30	30	30	30	30
	Bootstrap[c]	Bias		-.005	-.005	-.006	-.007	.019	0	.005
		Std. Error		.099	.108	.112	.095	.160	0	.215
		95% Confidence Interval	Lower	-.437	-.438	-.423	-.429	-.056	1	-.378
			Upper	-.053	-.009	.032	-.039	.549	1	.444
C_GHQ12_total	Pearson Correlation			-.027	-.095	-.060	-.061	-.236	.017	1
	Sig. (2-tailed)			.886	.618	.753	.748	.210	.927	
	N			30	30	30	30	30	30	30
	Bootstrap[c]	Bias		-.009	-.020	-.016	-.020	.006	.005	0
		Std. Error		.181	.195	.189	.193	.162	.215	0
		95% Confidence Interval	Lower	-.382	-.506	-.464	-.472	-.524	-.378	1
			Upper	.295	.240	.282	.267	.120	.444	1
Total score	Pearson Correlation			.235	.204	.176	.214	-.077	-.403*	-.431*
	Sig. (2-tailed)			.212	.280	.352	.256	.687	.027	.017
	N			30	30	30	30	30	30	30
	Bootstrap[c]	Bias		.014	.038	.022	.035	-.008	.007	.002
		Std. Error		.277	.264	.264	.287	.193	.133	.143
		95% Confidence Interval	Lower	-.257	-.213	-.266	-.239	-.460	-.619	-.680
			Upper	.776	.773	.712	.815	.298	-.093	-.114

Correlations

				Total score
Intensity t score		95% Confidence Interval	Lower	-.460
			Upper	.298
	Pearson Correlation			-.403*
	Sig. (2-tailed)			.027
	N			30
	Bootstrap[c]	Bias		.007
		Std. Error		.133
		95% Confidence Interval	Lower	-.619
			Upper	-.093
C_GHQ12_total	Pearson Correlation			-.431*
	Sig. (2-tailed)			.017
	N			30
	Bootstrap[c]	Bias		.002
		Std. Error		.143
		95% Confidence Interval	Lower	-.680
			Upper	-.114
Total score	Pearson Correlation			1
	Sig. (2-tailed)			
	N			30
	Bootstrap[c]	Bias		0
		Std. Error		0
		95% Confidence Interval	Lower	1
			Upper	1

**. Correlation is significant at the 0.01 level (2-tailed).

*. Correlation is significant at the 0.05 level (2-tailed).

c. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

Correlations

				Demographics	Problem t score	Intensity t score
Demographics	Pearson Correlation			1	.083	-.238
	Sig. (2-tailed)				.661	.205
	N			30	30	30
	Bootstrap[c]	Bias		0	-.006	.009
		Std. Error		0	.160	.174
		95% Confidence Interval	Lower	1	-.253	-.557
			Upper	1	.378	.139
Problem t score	Pearson Correlation			.083	1	.217
	Sig. (2-tailed)			.661		.249
	N			30	30	30
	Bootstrap[c]	Bias		-.006	0	.008
		Std. Error		.160	0	.161
		95% Confidence Interval	Lower	-.253	1	-.073
			Upper	.378	1	.535
Intensity t score	Pearson Correlation			-.238	.217	1
	Sig. (2-tailed)			.205	.249	
	N			30	30	30
	Bootstrap[c]	Bias		.009	.008	0
		Std. Error		.174	.161	0
		95% Confidence Interval	Lower	-.557	-.073	1
			Upper	.139	.535	1

*. Correlation is significant at the 0.05 level (2-tailed).

**. Correlation is significant at the 0.01 level (2-tailed).

c. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

Appendix 6.5

Multiple regression output with boostrapping for experimental group

Bootstrap

[DataSet1] /Users/Olivia/Desktop/SPSS thesis amendments/August dataset 1 use this one.sav

Bootstrap Specifications

Sampling Method	Simple
Number of Samples	1000
Confidence Interval Level	95.0%
Confidence Interval Type	Percentile

```
REGRESSION
  /MISSING LISTWISE
  /STATISTICS COEFF OUTS CI(95) R ANOVA CHANGE
  /CRITERIA=PIN(.05) POUT(.10)
  /NOORIGIN
  /DEPENDENT Ex_GHQ12_Total
  /METHOD=ENTER Ex_RSA_Total DBC_anxiety.
```

Regression

[DataSet1] /Users/Olivia/Desktop/SPSS thesis amendments/August dataset 1 use this one.sav

Variables Entered/Removed [a]

Model	Variables Entered	Variables Removed	Method
1	DBC_anxiety, Ex_RSA_Total [b]	.	Enter

a. Dependent Variable: Ex_GHQ12_Total

b. All requested variables entered.

Model Summary

Model	R	R Square	Adjusted R Square	Std. Error of the Estimate	Change Statistics				
					R Square Change	F Change	df1	df2	Sig. F Change
1	.632 [a]	.400	.351	2.30323	.400	8.317	2	25	.002

a. Predictors: (Constant), DBC_anxiety, Ex_RSA_Total

ANOVA[a]

Model		Sum of Squares	df	Mean Square	F	Sig.
1	Regression	88.236	2	44.118	8.317	.002[b]
	Residual	132.621	25	5.305		
	Total	220.857	27			

a. Dependent Variable: Ex_GHQ12_Total

b. Predictors: (Constant), DBC_anxiety, Ex_RSA_Total

Coefficients[a]

Model		Unstandardized Coefficients		Standardized Coefficients	t	Sig.	95.0% Confidence Interval for B	
		B	Std. Error	Beta			Lower Bound	Upper Bound
1	(Constant)	19.664	3.794		5.183	.000	11.850	27.478
	Ex_RSA_Total	-.056	.020	-.447	-2.819	.009	-.097	-.015
	DBC_anxiety	.362	.158	.364	2.295	.030	.037	.687

a. Dependent Variable: Ex_GHQ12_Total

Bootstrap for Coefficients

Model		B	Bootstrap[a]				
			Bias	Std. Error	Sig. (2-tailed)	95% Confidence Interval	
						Lower	Upper
1	(Constant)	19.664	.065	4.638	.003	9.263	28.499
	Ex_RSA_Total	-.056	.000	.023	.037	-.101	-.005
	DBC_anxiety	.362	-.040	.231	.178	-.128	.708

a. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

Multiple regression with bootstrapping output for control group

```
REGRESSION
  /MISSING LISTWISE
  /STATISTICS COEFF OUTS CI(95) R ANOVA CHANGE
  /CRITERIA=PIN(.05) POUT(.10)
  /NOORIGIN
  /DEPENDENT C_GHQ12_total
  /METHOD=ENTER C_RSA_Total C_ECBI_Intensity C_ECBI_Problem.
```

Regression

```
[DataSet1] /Users/Olivia/Desktop/SPSS thesis amendments/August dataset 1 use this one.sav
```

Variables Entered/Removed [a]

Model	Variables Entered	Variables Removed	Method
1	Problem t score, Total score, Intensity t score[b]	.	Enter

a. Dependent Variable: C_GHQ12_total
b. All requested variables entered.

Model Summary

Model	R	R Square	Adjusted R Square	Std. Error of the Estimate	Change Statistics				
					R Square Change	F Change	df1	df2	Sig. F Change
1	.522[a]	.273	.189	3.29577	.273	3.249	3	26	.038

a. Predictors: (Constant), Problem t score, Total score, Intensity t score
b. Predictors: (Constant), Problem t score, Intensity t score, Total score

ANOVA[a]

Model		Sum of Squares	df	Mean Square	F	Sig.
1	Regression	105.886	3	35.295	3.249	.038[b]
	Residual	282.414	26	10.862		
	Total	388.300	29			

a. Dependent Variable: C_GHQ12_total
b. Predictors: (Constant), Problem t score, Total score, Intensity t score
c. Predictors: (Constant), Problem t score, Intensity t score, Total score

Coefficients[a]

Model		Unstandardized Coefficients		Standardized Coefficients	t	Sig.	95.0% Confidence Interval for B	
		B	Std. Error	Beta			Lower Bound	Upper Bound
1	(Constant)	29.769	7.853		3.790	.001	13.626	45.912
	Total score	-.091	.033	-.503	-2.754	.011	-.159	-.023
	Intensity t score	-.022	.032	-.132	-.707	.486	-.088	.043
	Problem t score	-.202	.141	-.246	-1.434	.163	-.490	.087

a. Dependent Variable: C_GHQ12_total

Bootstrap for Coefficients

Model			Bootstrap[a]				
						95% Confidence Interval	
		B	Bias	Std. Error	Sig. (2-tailed)	Lower	Upper
1	(Constant)	29.769	-.299	8.578	.006	13.844	47.939
	Total score	-.091	.002	.031	.009	-.151	-.030
	Intensity t score	-.022	.000	.037	.552	-.104	.041
	Problem t score	-.202	-.005	.145	.146	-.536	.058

a. Unless otherwise noted, bootstrap results are based on 1000 bootstrap samples

Appendix 7: Author guidelines for the Journal of Inherited Metabolic Disease

JIMD – Journal of Inherited Metabolic Disease

Aims and Scope

The Journal of Inherited Metabolic Disease (JIMD) is the official journal of the Society for the Study of Inborn Errors of Metabolism (SSIEM). By enhancing communication between workers in the field throughout the world, the JIMD aims to improve the management and understanding of inherited metabolic disorders. It publishes results of original research and new or important observations pertaining to any aspect of inherited metabolic disease in humans and higher animals. This includes clinical (medical, dental and veterinary), biochemical, genetic (including cytogenetic, molecular and population genetic), experimental (including cell biological), methodological, theoretical, epidemiological, ethical and counselling aspects. The JIMD also reviews important new developments or controversial issues relating to metabolic disorders and publishes reviews and short reports arising from the Society's annual symposia. A distinction is made between peer-reviewed scientific material that is selected because of its significance for other professionals in the field and non-peerreviewed material that aims to be important, controversial, interesting or entertaining ("Extras").

The JIMD exists as two sister publications which are served by a single Editorial Team and a single manuscript submission and review process: the traditional print and online journal JIMD and JIMD Reports, which publishes scientifically sound research findings or clinical observations that warrant communication in the peer-reviewed literature but are of more limited interest to the readers. In addition to full

electronic publication, and print publication as book chapters, the abstract of a JIMD Reports publication is also printed in the JIMD, to each the widest possible readership. All other types of articles are published electronically and in print in the JIMD.

Instructions for submission

Manuscript submission

Material submitted to the JIMD (including JIMD Reports) must conform to the uniform requirements for manuscripts submitted to biomedical journals as outlined by the International Committee of Medical Journal Editors (ICMJE); see http://www.icmje.org/icmjerecommendations.pdf for details.

Online Submission

All scientific contributions for publication in the JIMD (including JIMD Reports) must be submitted by the web-enabled online manuscript submission and review system. As the review process is also fully web-based, this system allows editors to keep review times as short as possible and offers authors the option to track progress of the review of their manuscripts. The online manuscript submission and review system for the JIMD offers easy and straightforward log-in and submission procedures. Please refer to: www.editorialmanager.com/boli. The system supports a wide range of submission file formats for manuscripts (Word,WordPerfect, RTF, TXT and LaTex) and figures (TIFF, EPS, Microsoft® Office formats and Postscript). PDF is not an acceptable file format.

If you encounter any difficulties while submitting your manuscript online, please contact the responsible Editorial Assistant by clicking on "CONTACT US" from the tool bar.

Manuscript Structure

The first page should include:

 Title of the article

 Authors‘ names and institutional affiliations set out as in a current issue of the JIMD

 Name, email address and full postal address, including postal (ZIP) code, of the author who will be dealing with correspondence and proofs.

 Word counts for the text (excluding summary, acknowledgments, references and figure legends) and the summary.

 Number of figures and tables.

 Whether a colour picture is provided that may be used for the front cover of the issue in which the article appears.

The next page should include:

 A summary (= abstract) of not more than 250 words (Medline allows a maximum of 4096 characters and will truncate longer abstracts).

 A concise 1 sentence take-home message (synopsis) of the article, outlining what the

reader learns from the article (this is usually printed on the inside back cover of the JIMD).

Following these pages, authors are required to provide the following, which are detailed above in the section “General Rules.”

 Details of the contributions of individual authors

 The name of the corresponding author

 A competing interest statement

 Details of funding

 Details of ethics approval

 A patient consent statement

 Documentation of approval from the Institutional Committee for Care and Use of Laboratory Animals (or comparable committee)

 A list of approximately six keywords

Recommendations for Manuscript Length

Competition for publication in all scientific journals has become increasingly intense, and the JIMD is no exception. We strongly encourage prospective authors to consider brevity in their presentation, and if needed to avail themselves of the online supplementary material for those Figures and Tables that could be accommodated in that venue. In order for the Editorial Board to accommodate the broadest perspective of submissions, and to maximize the access for prospective authors to both the JIMD and JIMD Reports, the following recommendations for length have been formulated:

 Full articles: Total word count 3000, including 500 words for the Introduction and a

maximum of 4 combined figures/tables.

 Reports: Total word count 2250, including 400 words for the introduction and a maximum of 3 total figures/tables. It is expected that more comprehensive reviews will exceed these limits, but the authors of such reviews are again encouraged to work for brevity and succinctness in presentation. In all instances, literature citations should be reasonable and appropriate for the presentation, but should not exceed 30 citations for full articles and 25 citations for reports. Appropriate use of the cited literature is one way in which prospective authors can constrain the length of their submissions.

Number of authors: The number of authors needs to be limited to a maximum of 20; additional authors may be included as consortium, listed as individual contributors at the end of the manuscript and recognized in PubMed.

Covering letter

Submit a covering letter and use it to explain why your paper should be published in The Journal of Inherited Metabolic Disease.

General Rules

It is a condition of acceptance that all articles have not been and will not be published elsewhere in substantially the same form. The submitting author must have circulated the article and secured final approval of the version to be peer-reviewed from all co-authors prior to article submission. This includes confirmation of

 absence of previous similar or simultaneous publications,

 their inspection of the manuscript,

 their substantial contribution to the work (all authors should have been involved in (a) conception and design, or analysis and interpretation of data, and (b) drafting the

article or revising it critically for important intellectual content), and their agreement to submission.

It should be noted that these conditions are later confirmed in writing by the corresponding author in a copyright transfer form at the time of acceptance. Publication elsewhere, at any time, of a similar article perhaps only differing in some aspects of data, especially if the JIMD article is not cross-referenced, may justify formal retraction at a later date. Supplementary (internet-only) material may be published for all articles; we encourage or request deposition of raw data when this appears appropriate.

The following information will be required at the time of online manuscript submission and is required on the page following the details listed in the section "Manuscript Structure" (below):

 Details of the contributions of individual authors, making clear who has contributed pertinent aspects of the planning, conduct, and reporting of the work described in the article.

 Name of one author who serves as guarantor for the article, accepts full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

 A competing interest statement, i.e., either a statement describing the interests of all authors or a declaration that they have nothing to declare, based on the "Competing Interests Questions" outlined below.

 Details of funding for all research studies including a statement that "The author(s) confirm(s) independence from the sponsors; the content of the article has not been influenced by the sponsors"

 Details of ethics approval or a statement that it was not required for all research studies

 A patient consent statement for all articles or other material that contain personal information about a patient; proof that informed consent was obtained must be available upon request

 If vertebrate animals have been utilized, documentation of approval from the Institutional Committee for Care and Use of Laboratory Animals (or comparable committee).

 A list of approximately six keywords; this is of particular importance for recognition of the manuscript after publication by some search engines.

Statements, permissions, and signatures

Authors and contributors

The number of authors needs to be limited to a maximum of 20; additional authors may be included as consortium, listed as individual contributors at the end of the manuscript and recognized in PubMed.

Declaration of interests and competing Interests Conflict of interest exists when an author (or the author's institution), reviewer or editor has financial or personal relationships that inappropriately influence (bias) his or her actions (such relationships are also known as dual commitments, competing interests or competing loyalties). These relationships vary from those with negligible potential to those with great potential to influence judgment, and not all relationships represent true conflict of interest. The potential for conflict of interest can exist whether or not an individual believes that the relationship affects his or her scientific judgment. Financial

relationships (such as employment, consultancies, stock ownership, honoraria, or paid expert testimony) are the most easily identifiable conflicts of interest and the most likely to undermine the credibility of the journal, the authors and science itself. However, conflicts can occur for other reasons, such as personal relationships, academic competition and intellectual passion. All authors (co-authors) of articles, reports, reviews, editorials and other material submitted to the JIMD (including JIMD Reports) as well as reviewers of manuscripts must answer the following questions:

1. Have you in the past five years accepted the following from an organisation that may in any way gain or lose financially from the results of your study or the conclusions of your review, editorial or letter:

 Reimbursement for attending a symposium?

 A fee for speaking or for organising education?

 Funds for research or for a member of staff?

 A fee for consulting?

2. Have you in the past five years been employed by an organisation that may in any way gain or lose financially from the results of your study or the conclusions of your review, editorial or letter? Do you hold any stocks or shares in such an organisation?

3. Have you acted as an expert witness on the subject of your study, review, editorial or letter?

4. Do you have any other competing financial interests?

Authors who have answered "yes" to any of these questions may have a competing interest which should be declared at the time of submission of the article (review, editorial or other material) and which will be published in JIMD.

Other non-financial interests that authors may like to disclose include:

 A close relationship with, or a strong antipathy to, a person whose interests may be affected by publication of the article.

 An academic link or rivalry with someone whose interests may be affected by publication of the article.

 Membership in a political party or special interest group whose interests may be affected by publication of the article.

 A deep personal or religious conviction that may have affected what the author wrote and that readers should be aware of when reading the article.

Expert reviewers approached for assessment of submitted articles are also requested to declare conflicts of interest that may impede on their judgement of that article. This specifically includes competing research in the same area that could be negatively affected by publication of the submitted article. For additional information see also the ICJME's "Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly Work in Medical Journals" at http://www.icmje.org/about-icmje/faqs/icmje-recommendations/

Ethical guidelines, human and animal rights and consents

If the work involves the use of human subjects, the author should ensure that the work

described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans (Uniform Requirements for manuscripts submitted to Biomedical journals). Authors should include a statement in the manuscript that informed consent was obtained for experimentation with human subjects. The privacy rights of human subjects must always be observed. All animal experiments should comply with the ARRIVE guidelines and should be carried out in accordance with the U.K. Animals (Scientific Procedures) Act, 1986 and associated guidelines, EU Directive 2010/63/EU for animal experiments, or the National Institutes of Health guide for the care and use of Laboratory animals (NIH Publications No. 8023, revised 1978) and the authors should clearly indicate in the manuscript that such guidelines have been followed.

Types of article and manuscript requirements

Scientific contributions

Full Articles

The JIMD welcomes scientific contributions for publication as printed full articles in the following categories:

 Original Articles: Important manuscripts that may be expected to influence or change clinical or research practice with regard to inherited metabolic disorders. Original articles may include comprehensive studies on disease features in groups of patients, important novel information on a disease or relevant research findings. Case reports are unlikely to be accepted as original papers in print JIMD, unless they describe novel diseases or new aspects of the basic pathomechanism of a disease, supported by novel functional data. The editors may reject submitted manuscripts as original articles but invite revision or resubmission for publication as Reports in

“JIMD Reports”. All authors are invited to provide a colour picture that may be used for the front cover of the issue in which the article appears.

 Rapid Communications: Highly competitive and timely manuscripts; please contact theeditorial office: editor@jimd.org.

 Guidelines: Studies providing a comprehensive, representative analysis of the clinical features, treatment and outcome of inborn errors of metabolism. For detailed information, we refer to the Editorial: Zschocke J, Baumgartner MR, Morava E, Patterson MC, Peters V and Rahman S (2016) Recommendations and guidelines in the JIMD: suggested procedures and avoidance of conflicts of interest. J Inherit Metab Dis, 39:327-330

 Reviews: Concise summaries of metabolic pathways, specific disorders, methods, treatment options etc.

 Metabolic Dissertations: The JIMD invites all researchers who have completed a Ph.D. or M.D. thesis in the field of inborn errors of metabolism to submit a comprehensive review of the topic of their thesis. The article should not focus solely on the research findings but should cover all relevant information in the respective field. Such reviews preferably (but not necessarily) have a single author (other contributors should be acknowledged) and will be published with a photograph of the investigator.

Images in Metabolic Medicine

The Editors will consider clear and interesting clinical pictures or other types of images (e.g. laboratory results or observations) submitted with a descriptive paragraph of up to 250 words.

Prints, slides or electronic copy are all acceptable. Authors must obtain informed consent for publication of patient-related materials. Case reports or additional information may be added as supplementary material. Images will be fully printed; title and author(s) will be listed in bibliographical databases such as Medline.

Editorials

The JIMD invites communicating editors and reviewers of articles that have been accepted for publication in the JIMD to provide an editorial that places the article in a broader context. Editorials have no abstract, may be comprised of up to 500 words and should contain no more than two (if any) references. Additional material can be added as supplementary material online. Editorials will be fully printed; title and author(s) will be listed in bibliographical databases such as Medline.

Letters and Clinical/Research Observations

The JIMD is happy to receive comments on previously published articles in the journal which should reach the editorial office within 4 weeks of publication of the original item. Correspondence may be subjected to peer review and counter-replies are usually invited from the authors of the original publication. The concise form of a letter may also be used to report exceptionally important clinical or research observations unrelated to a previous JIMD publication that merit communication but do not fulfil the requirements for scientific articles or short reports. These items will be peer reviewed and if accepted will be published under the heading “Observation”. Letters should have no more than five authors. They have no abstract, are limited to a maximum of 500 words and should contain no more than two (if any) references.

Additional material can be added as supplementary material online. Letters will be fully printed; title and author(s) will be listed in bibliographical databases such as Medline.

JIMD Reports

Some manuscripts present scientifically sound research findings or clinical observations that are worth communicating but are of more limited interest to the readers of the JIMD and may be sufficiently summarised in an abstract of 250 words. In order to facilitate publication of these types of manuscripts, JIMD Reports has been introduced as a sister publication of the traditional JIMD. It is an independent periodical with its own ISSN number. All manuscripts submitted as Reports to the JIMD website will be considered for JIMD Reports rather than for the traditional journal. They will undergo the same review process as Original Articles (and in exceptional cases may be reassigned for publication in the JIMD). In addition, the Editorial Team (based on the advice of reviewers and Communicating Editors) may reject Original Articles for publication in the JIMD but offer publication in JIMD Reports. After acceptance, articles in JIMD Reports are professionally typeset in the same manner as articles in the JIMD, and full documents are available online to SSIEM members and institutional subscribers via the Springer website. Articles from JIMD Reports are submitted to PubMedCentral and are listed in PubMed as well as other abstracting and indexing services. After an embargo period of 12 months, all manuscripts published in JIMD Reports (in contrast to JIMD articles) are available free of charge world-wide, thereby ensuring widest possible readership. In addition, titles and abstracts of Reports are printed in the print-only “Extras” section of the JIMD. It is recommended to make use of the full allowance of 250 words for the

abstract of Reports to convey the message of the article to the readers of the print journal. Reports follow the same rules as Full Articles; they should not be used as a form of preliminary communication. They may take the form of Research Reports, with content similar to that of original articles, or Case Reports. Case Reports will only be considered when they highlight some unusual or previously unrecorded feature relevant to the disorder or serve as an important reminder of clinical or biochemical features of a Mendelian disorder. Chance associations of two conditions or sporadic cases from new geographical locations (as opposed to systematic epidemiological studies) are not in themselves of sufficient scientific merit to justify publication.

Extras in the JIMD

The Editors of the JIMD invite submission of short items that are interesting, stimulating, important or entertaining to professionals working in the field of inborn errors of metabolism. These items will not usually be reviewed outside the editorial board and usually will not be referenced in bibliographic databases. All items of this type should be submitted by email to the editorial office (editor@jimd.org); please provide full personal details for all authors of each contribution.

Garrodian

Small texts that are used to fill gaps (e.g. at the end of original articles) have been a long and cherished tradition in some journals. They usually have the added advantage of entertaining readers and stimulating thought. The Editors are happy to receive interesting stories or personal experiences of up to a few hundred words on topics such as:

 A patient / paper / experience that changed my practice

 A memorable patient / experience

 An error that proved educational or informative for lab operation or clinical care

 How I embarked on this career path and lessons learned along the way

 Any other story conveying instruction, pathos or humour

If the Garrodian refers to an identifiable person, written consent for publication from that person or an appropriate relative is required.

Book Reviews

Instructive reviews of up to 400 words are invited on new books published in the field of inborn errors of metabolism, or closely affiliated areas.

Obituaries

The Editors of the JIMD strongly encourage submission of obituary notices for all recently deceased SSIEM members or other persons in the field of inborn errors of metabolism. Obituary notices should be emailed to the editorial office. Please give your name and contact details, including a phone number and email address. Obituaries will be considered by the editorial board and may be shortened; they will be published (without proofs) with the name of the person(s) who submitted the notice.

Please provide:

1. The full name of the deceased

2. A photograph

3. A summary of important data:

a. (Last) professional position and title, place of work

b. Date and place of birth

c. Primary degree with university and year when obtained

d. Additional professional qualifications with university and year when obtained

e. Date of death, cause of death

4. The main text summarising important contributions and personal characteristics of the deceased. The last sentence should state the remaining relatives such as spouse and/or the number of children and grandchildren.

Formatting guidelines

Language

Please write your text in good English (American or British usage is accepted, but not a mixture of these).

Text formatting

Standard text formatting is recommended in word, with the preferential use of Times New Roman, 12 font letters and double spaced text documents. The submission process automatically converts text files to pdf.

Units, Symbols, and Database References

At the time of first mention, diseases, enzymes or genes should be referenced to the appropriate classification, nomenclature or database:

 Inherited diseases to the OMIM catalogue number (http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=OMIM)

- Enzymes to an Enzyme Commission (EC) number (http://www.chem.qmul.ac.uk/iubmb/enzyme/)
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Journals:

Smith AL, Smith JD (1977) Hybridisation methods. Nucl Acids Res 8: 1095–1098.

Chapter in an edited book:

Weinstein L, Swartz MN (1974) Pathologic mechanisms of invading microorganisms. In Sodeman WA Jr, Sodeman WA, eds. Pathogenic Physiology: Mechanisms of Disease. Philadelphia: WB Saunders, 457–472.

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Appendix 8: Quality Assessment Tool for Studies with Diverse Designs (QATSDD)

Instruction sheet

Eligibility criteria for papers:

1. Original research papers for inclusion into a systematic review
2. Study design must be qualitative, quantitative or mixed methods

Method:

Scoring the studies:

1. Read through the research paper carefully.
2. There are 16 quality criteria in the tool; 14 of these criteria apply to qualitative studies, 14 apply to quantitative studies and all 16 apply to any mixed methods papers. The applicable questions are indicated in brackets in the grid under the item.
3. Read each of the criteria and look at the descriptions under each score from 0-3 to find out what is required to obtain each score.
4. Using the descriptions for each score to guide your response, give the paper a score from 0-3 on each item on your scoring grid.
5. This will result in a score out of a maximum of 48 (16x3) for mixed methods papers, and 42 (14x3) for qualitative or quantitative papers.

Comparing the quality of studies:

1. In order to compare quality of the papers you should calculate what % of the maximum possible score was obtained see example below:

A **quantitative** paper scores 39 out of 42 = **92.9%** of the maximum quality score.

This could be compared to a **qualitative** paper that scores 9 out of 42 = **21.4%** of maximum quality score, suggesting that the **quantitative** work was of a higher quality.

2. In addition you can calculate a quality score for all studies using the same design as a group e.g. the qualitative studies. This allows comparisons to be drawn for example between the qualitative and quantitative papers.
3. To do this you would take an average of the quality scores calculated for each paper for each group and then compare these.

Criteria	0 = Not at all	1 = Very slightly	2 = Moderately	3 = Complete
1. Explicit theoretical framework	No mention at all.	Reference to broad theoretical basis.	Reference to a specific theoretical basis.	Explicit statement of theoretical framework and/or constructs applied to the research.
2. Statement of aims/objectives in main body of report	No mention at all.	General reference to aim/objective at some point in the report including abstract.	Reference to broad aims/objectives in main body of report.	Explicit statement of aims/objectives in main body of report.
3. Clear description of research setting	No mention at all.	General description of research area and background, e.g. 'in primary care'.	General description of research problem in the target population, e.g. 'among GPs in primary care'.	Specific description of the research problem and target population in the context of the study, e.g. nurses and doctors from GP practices in the east midlands.
4. Evidence of sample size considered in terms of analysis	No mention at all.	Basic explanation for choice of sample size. Evidence that size of the sample has been considered in study design.	Evidence of consideration of sample size in terms of saturation/information redundancy or to fit generic analytical requirements.	Explicit statement of data being gathered until information redundancy/saturation was reached or to fit exact calculations for analytical requirements.
5. Representative sample of target group of a reasonable size	No statement of target group.	Sample is limited but represents some of the target group or representative but very small.	Sample is somewhat diverse but not entirely representative, e.g. inclusive of all age groups, experience but only one workplace. Requires discussion of target population to determine what sample is required	Sample includes individuals to represent a cross section of the target population, considering factors such as experience, age and workplace.

			to be representative.	
6. Description of procedure for data collection	No mention at all.	Very basic and brief outline of data collection procedure, e.g. 'using a questionnaire distributed to staff'.	States each stage of data collection procedure but with limited detail, or states some stages in details but omits others.	Detailed description of each stage of the data collection procedure, including when, where and how data were gathered.
7. Rationale for choice of data collection tool(s)	No mention at all.	Very limited explanation for choice of data collection tool(s).	Basic explanation of rationale for choice of data collection tool(s), e.g. based on use in a prior similar study.	Detailed explanation of rationale for choice of data collection tool(s), e.g. relevance to the study aims and assessments of tool quality either statistically, e.g. for reliability & validity, or relevant qualitative assessment.
8. Detailed recruitment data	No mention at all.	Minimal recruitment data, e.g. no. of questionnaire sent and no. returned.	Some recruitment information but not complete account of the recruitment process, e.g. recruitment figures but no information on strategy used.	Complete data regarding no. approached, no. recruited, attrition data where relevant, method of recruitment.
9. Statistical assessment of reliability and validity of measurement tool(s) (Quantitative only)	No mention at all.	Reliability and validity of measurement tool(s) discussed, but not statistically assessed.	Some attempt to assess reliability and validity of measurement tool(s) but insufficient, e.g. attempt to establish test–retest reliability is unsuccessful but no action is taken.	Suitable and thorough statistical assessment of reliability and validity of measurement tool(s) with reference to the quality of evidence as a result of the measures used.
10. Fit between stated	No research	Method of data collection	Method of data collection can	Method of data collection selected is

research question and method of data collection (Quantitative)	question stated.	can only address some aspects of the research question.	address the research question but there is a more suitable alternative that could have been used or used in addition.3	the most suitable approach to attempt answer the research question
11. Fit between stated research question and format and content of data collection tool e.g. interview schedule (Qualitative)	No research question stated.	Structure and/or content only suitable to address the research question in some aspects or superficially.	Structure & content allows for data to be gathered broadly addressing the stated research question(s) but could benefit from greater detail.	Structure & content allows for detailed data to be gathered around all relevant issues required to address the stated research question(s).
12. Fit between research question and method of analysis	No mention at all.	Method of analysis can only address the research question basically or broadly.	Method of analysis can address the research question but there is a more suitable alternative that could have been used or used in addition to offer greater detail.	Method of analysis selected is the most suitable approach to attempt answer the research question in detail, e.g. for qualitative IPA preferable for experiences vs. content analysis to elicit frequency of occurrence of events, etc.
13. Good justification for analytical method selected	No mention at all.	Basic explanation for choice of analytical method	Fairly detailed explanation of choice of analytical method.	Detailed explanation for choice of analytical method based on nature of research question(s).
14. Assessment of reliability of analytical process (Qualitative only)	No mention at all.	More than one researcher involved in the analytical process but no further reliability assessment.	Limited attempt to assess reliability, e.g. reliance on one method.	Use of a range of methods to assess reliability, e.g. triangulation, multiple researchers, varying research backgrounds.

15. Evidence of user involvement in design	No mention at all.	Use of pilot study but no involvement in planning stages of study design.	Pilot study with feedback from users informing changes to the design.	Explicit consultation with steering group or statement or formal consultation with users in planning of study design.
16. Strengths and limitations critically discussed	No mention at all.	Very limited mention of strengths and limitations with omissions of many key issues.	Discussion of some of the key strengths and weaknesses of the study but not complete.	Discussion of strengths and limitations of all aspects of study including design, measures, procedure, sample & analysis.

Appendix 9: Research poster presented at the NSPKU national conference

Parenting a child with Phenylketonuria (PKU): An investigation into child behaviour and parental well-being

Olivia Ambler, Emma Medford and Dougal Hare

Background

Studies suggest that parents of children with PKU are vulnerable to psychological distress. A number of factors may contribute to this, including the demands of managing treatment, child behaviour difficulties, psychological resilience and social support. However, due to variability in the research findings, it is still unclear what influences the well-being of parents who care for a child with PKU and what helps.

Objectives

1) To compare the psychological well-being of parents of children with PKU to parents in the general population
2) To identify what factors are important for the well-being of parents who care for a child with PKU, distinct from the general population
3) To investigate whether child behaviour is different in children with PKU compared to the general population and whether this impacts on parents' well-being

Method

40 parents of children with PKU and 40 parents from the general population are currently being recruited through the NSPKU. All parents are asked to fill in questionnaires measuring:

- Levels of psychological distress (General Health Questionnaire-12)
- Psychological resilience (Resilience Scale for Adults)
- Social support (Multidimensional Scale of Perceived Social Support)
- Child behaviour (Eyberg Child Behaviour Inventory)
- Demographic information (Demographics questionnaire)

Parents of children with PKU are also asked to fill in scales measuring:

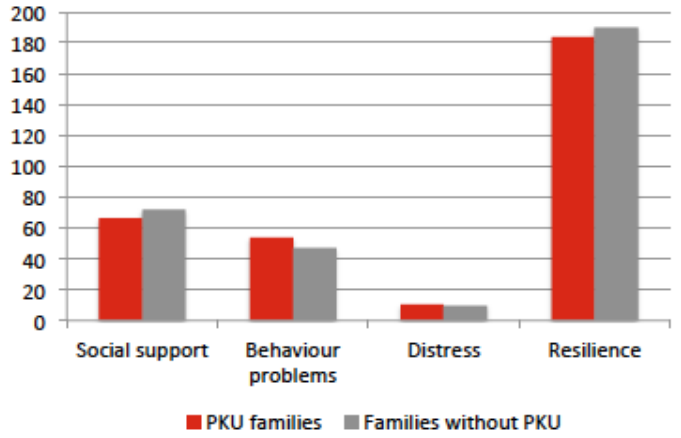
- Their child's care dependency (Child Care Dependency Scale)
- Behaviour related to developmental difficulties (Developmental Behaviour Checklist)

Results

21 parents of children with PKU and 31 parents of children from the general population have participated in the study to date. The preliminary findings show:

- Parents of children with PKU had slightly higher distress levels than parents in the general population, although this difference was non-significant (p= .6)
- Parents of children with PKU reported more child behaviour problems compared to the general population. This difference approached significance (p= .05)
- Psychological resilience was significantly correlated with well-being (p< .01)
- Fewer child behaviour problems and greater levels of social support were also correlated with well-being, although these were non-significant (p= .22; p= .06)

Mean questionnaire scores



Conclusion

- So far, the data suggests that psychological resilience and social support are important for the well-being of parents who care for a child with PKU.
- There were also some behaviour differences between children with PKU and their peers, which has implications for parents' well-being.
- We are still looking to recruit 19 more parents of children with PKU and 9 parents of children who do not have PKU to run more in depth analyses to discern which factors predict psychological well-being for parents of children with PKU.

Contact details

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