

Mothers' Experiences Breastfeeding Infants with Phenylketonuria

by

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DEDICATION

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ABSTRACT

TITLE: Mothers' Experiences Breastfeeding Infants with Phenylketonuria

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Background: Breastfeeding an infant with phenylketonuria (PKU) is possible, but requires mastering a complex management routine that combines limited breastfeeding with phenylalanine-free formula to maintain therapeutic phenylalanine levels and reflects mothers' efforts to continue breastfeeding in this challenging situation.

Purpose: This study described the prevalence and duration of breastfeeding infants with PKU in the United States and Canada, and described mothers' experiences breastfeeding in the context of PKU.

Methods: This was a cross-sectional descriptive study. Although this study was based at Oregon Health & Science University, mothers were recruited from across the United States and Canada. In phase 1, mothers ($n = 103$) with one child with PKU completed the online survey between November, 2010 and May, 2011. The online survey consisted of a questionnaire that contained sections about the demographic characteristics of mothers and infants, and infant feeding history. Response options included both forced choice and open-ended questions. Descriptive statistics were used to describe data from the internet survey. A P value of $<.05$ was considered statistically significant. In phase 2, a subset of mothers ($n = 10$) who participated in the online survey and had a child less than 36 months of age were purposively selected and completed an in-

depth, semi-structured telephone interview about their breastfeeding experiences. Following transcription, thematic analysis was used to identify key aspects of breastfeeding in the context of PKU.

Results: In phase 1 of the 103 mothers, 89 (86%) initiated breastfeeding immediately following delivery while 14 (14%) chose bottle feeding. In comparison to breastfeeding after delivery, significantly fewer mothers breastfed after diagnosis (McNemar's $\chi^2 = 30.33$, $p < .001$; $n = 72$ versus $n = 89$).

Breastfeeding duration ranged from less than one month to twenty-four months with one modal duration category ($n = 20$, 22%) at less than one month. The timing of the addition of commercial infant formula to supplement breastfeeding or expressed mothers' milk was associated with a shorter duration of breastfeeding among infants with PKU, $\chi^2 (42, n = 73) = 88.13$, $p < .001$. In phase 2, mothers ($n = 10$) reported breastfeeding an infant with PKU was more complex and labor intensive than feeding a non-PKU infant. Commitment was related to the perception that breastfeeding an infant with PKU is the healthiest choice, and therefore worth the labor. Mothers described two major components of the special effort needed to breastfeed their infant: monitoring Phe levels and adapting breastfeeding. Consequently, mothers individualized their infant's treatment in order to maintain metabolic control with appropriate Phe levels while adapting breastfeeding to their unique situation. As they continued to breastfeed their infants after diagnosis, mothers moved from viewing PKU as a disorder and that their infant was ill to a perspective that their child was healthy in spite of PKU. Normal can mean a breastfeeding infant with PKU.

Discussion: Breastfeeding infants with PKU is challenging in part because Phe intake is difficult to determine precisely. This study provided the first detailed description of mothers' perceptions of their experiences breastfeeding infants with PKU and begins an attempt to understand the central concerns of mothers' breastfeeding infants with PKU. Results provide important information for clinicians regarding the challenges mothers face and the kinds of support they need. Further research should identify the unique needs of mothers' breastfeeding infants with PKU to guide the development of interventions specific to these mothers to support their efforts to continue breastfeeding after the diagnosis of PKU.

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CHAPTER 1

INTRODUCTION

Human breast milk is the healthiest form of milk for human infants (American Academy of Pediatrics, 2005; American Association of Family Physicians, 2008; Association of Women's Health Obstetric and Neonatal Nurses, 2005; Canadian Paediatric Society, 2005; U.S. Preventive Services Task Force, 2008; World Health Organization, 2009). Multiple positive outcomes have established the benefits of human breast milk and breastfeeding for infants and mothers (Hoddinott, Tappin, & Wright, 2008; Horta, Bahl, Martines, & Victora, 2007; Ip et al., 2007). Human breast milk has advantages for infants that distinguish it from standard commercial infant formulas. Growth factors, hormones, immunological factors, and long-chain polyunsaturated fatty acids are some of these advantages.

For infants with phenylketonuria (PKU), human breast milk has a lower concentration of protein and a lower content of the amino acid, phenylalanine (Phe), than standard commercial infant formulas. Some clinics encourage mothers of infants with PKU to continue breastfeeding whereas others discourage breastfeeding after the diagnosis. Although breastfeeding and PKU have been discussed in the literature, there is a lack of research describing patterns of mothers' breastfeeding infants with PKU, including infants with PKU in the United States and Canada.

Statement of the Problem

Mothers of breastfeeding infants with PKU face an especially challenging situation. Breastfeeding in the context of PKU, include infants breastfeeding at the breast and bottle feeding expressed mothers' milk (referred to as *breastfeeding management* throughout this study), entailed the need for mothers, at each feeding, to estimate how much breast milk to feed or how long to breastfeed, as well as how much Phe-free supplementation is required to maintain acceptable blood Phe levels. In addition, they must pump and then store the expressed breast milk when supplementing with Phe-free formula to maintain their breast milk supply. In order to follow serial Phe levels, they perform routine heel stick blood testing on their infants with PKU and mail the dried blood samples to a laboratory. Mothers perform these caregiving activities for their infants with PKU in their home, which may be hundreds of miles from the pediatric metabolic clinic or from another mother who is breastfeeding an infant with PKU.

Purpose of the Study

The absence of systematic research to describe mothers' patterns of breastfeeding infants with PKU and their experiences of breastfeeding management seriously limits the evidence base for developing and testing interventions that support breastfeeding management in this unique context. Therefore, the purpose of this study was twofold: 1) to determine the prevalence and duration of breastfeeding for infants with PKU in the United States and Canada and 2) to generate knowledge about how mothers of infants with PKU

simultaneously manage breastfeeding and PKU therapy, and how they perceive family functioning.

Specific Aims

The study was a cross-sectional design with two phases: 1) an internet survey of mothers' experiences feeding infants with PKU, and 2) a qualitative approach to the exploration of a subsample's experiences with breastfeeding their infants with PKU using interview and two quantitative self-report measures. To accomplish this purpose, the following specific aims of this study were to:

1. Describe the prevalence and duration of breastfeeding among mothers who have infants with PKU in the United States and Canada.
2. Describe mothers' experiences breastfeeding in the context of PKU including their perceptions of factors that facilitate and/or interfere with breastfeeding infants with PKU and strategies to obtain the necessary skills to care for their breastfeeding infants with PKU.
3. Identify mothers' experiences breastfeeding infants with PKU and examine mothers' assessment of family functioning.

In accordance with Family Management Style Framework (FMSF), mothers' perspectives were sought to understand their definition, management, and perceived consequences of breastfeeding infants with PKU, and their incorporation and adherence to the responsibilities and activities of PKU therapy (Deatrick, Knafl, & Murphy-Moore, 1999; Knafl & Deatrick, 1990; Knafl & Deatrick, 2003). Those perspectives may be very different from those of

professionals in medicine, nursing, lactation, and nutrition who work with mothers breastfeeding their infants with PKU.

Significance to Nursing

Nurses do not have a research base for providing care regarding breastfeeding management for infants with PKU. Thus, results of this study have significance for nursing practice, research, policy, and theory. The findings from this study will contribute to establishing a scientific basis for the care of breastfeeding infants with PKU while incorporating and adhering to the responsibilities and activities of PKU therapy. Furthermore, knowledge from this study may be relevant to mothers breastfeeding infants with other inborn errors of metabolism disorders, such as other disorders of amino acid metabolism, organic acidemias, and urea cycle defects (Huner, Baykal, Demir, & Demirkol, 2005; MacDonald et al., 2006).

This study was congruent with the National Institute of Nursing Research strategic plan to support investigator-initiated research that addresses significant questions related to new knowledge and better health of vulnerable populations, such as infants with PKU. In addition, this study was consistent with the U.S. Preventive Services Task Force on *Primary Care Interventions to Promote Breastfeeding* (2008) which indicated that research is needed to promote and support breastfeeding in populations where exclusive breastfeeding is not possible, as with infants with PKU.

The exploratory descriptive nature of this study provides a basis for further research regarding mothers of children with PKU and for the investigation of

appropriate interventions with these mothers and families. In families whose everyday lives are shaped by the unique understanding of PKU and the attendant concerns and demands, these mothers require a unique breastfeeding approach that addresses the need to maintain therapeutic Phe levels. With increased understanding of mothers' experiences breastfeeding infants with PKU, we are able to better conduct further research to assist these mothers in successfully breastfeeding while simultaneously managing PKU and breastfeeding.

The practice goal for those mothers who choose to breastfeed should be a formulation of breastfeeding management strategies that maintain appropriate Phe levels. This goal can be realized if the multi-disciplinary metabolic team can knowledgeably discuss breastfeeding management of infants with PKU based upon sound research. Discussions with other healthcare providers, such as public health nurses, neonatal and pediatric nurses, and pediatric nurse practitioners, will then occur to encourage and support mothers' breastfeeding of newly diagnosed newborns with PKU. The next chapter of the dissertation presents the literature relevant to the study of breastfeeding, PKU, and the family context of breastfeeding an infant with PKU.

CHAPTER 2

REVIEW OF THE LITERATURE

As noted in Chapter 1, numerous organizations have endorsed human breast milk as the optimum nutrition for infants (American Academy of Pediatrics, 2005; American Association of Family Physicians, 2008; Association of Women's Health Obstetric and Neonatal Nurses, 2005; Canadian Paediatric Society, 2005; U.S. Preventive Services Task Force, 2008; World Health Organization, 2009). Yet despite the documented benefits of human breast milk and breastfeeding, there has been a lack of research describing the practice of breastfeeding infants with PKU. Although the limited research on breastfeeding infants with PKU addresses the incidence, duration, and various dietary management protocols, the results are difficult to interpret due to methodological issues.

There has been limited inquiry about the experience of mother' breastfeeding infants with PKU. Researchers have yet to identify the ontological aspects of mothers' experiences breastfeeding infants with PKU, knowledge that is necessary to direct practice, policy, and education. In order to provide interventions that support and educate mothers in breastfeeding these infants, more needs to be understood about this vulnerable population.

This review of the literature is drawn from three areas: research on PKU, breastfeeding, and families. First, the review provides an overview of PKU focusing on the metabolic defect. The second section of the chapter discusses breastfeeding: the benefits of human breast milk and breastfeeding, breastfeeding infants with special conditions, and breastfeeding infants with PKU.

The third section reviews research on the family context of breastfeeding an infant with PKU, drawing from the literature on family management styles when children have chronic health conditions. Family functioning is embedded within the third section.

Phenylketonuria

Phenylketonuria (PKU) is an inherited inborn error of metabolism disorder within the amino acid disorders. The disorder was discovered in 1934 by the addition of ferric chloride to the urine of two siblings with mental retardation and “musty” body odor (Folling, 1934). The resultant urine was a deep green from the presence of increased phenylpyruvic acid. Hence, the term, *phenylketonuria*, was proposed to describe the disorder (Penrose & Quastel, 1937).

Phenylketonuria is inherited in an autosomal recessive pattern. Parents of an infant with PKU are usually unaffected, healthy carriers of PKU, and they have one normal and one abnormal phenylalanine hydroxylase (PAH) gene. With each pregnancy, the carrier parents have a 25% chance of having a child with two copies of the normal gene, resulting in a healthy, unaffected non-carrier infant, a 50% chance of having an infant with one copy of the abnormal gene and one copy of the normal gene who is an unaffected carrier like themselves, and a 25% chance of having an infant with two copies of the abnormal gene, resulting in PKU.

The most common form of PKU is the deficiency of phenylalanine hydroxylase, which is responsible for the metabolism of phenylalanine (Phe) to tyrosine (Tyr), normally a non-essential amino acid, within the liver (see Figure 1)

(Jervis, 1953; Scriver, Levy, & Donlon, 2008). The defect in PAH activity results in accumulation of Phe in the blood and body tissues and a deficit of Tyr, which becomes an essential amino acid for individuals with PKU. Tyr is needed for synthesis of melanin, epinephrine, and thyroxine and, indirectly, for neurotransmitters. This is classical or typical PKU.

A less common cause of PKU is from tetrahydrobiopterin deficiency, a co-enzyme for PAH (Bartholome, 1974; Kaufman, 1958). The incidence of this rarer form of PKU is one in one million births (Scriver et al., 2008). The management for tetrahydrobiopterin is more complex than for classic PKU (Kure et al., 1999; Shintaku et al., 2004; Spaapen et al., 2001; Spaapen & Rubio-Gozalbo, 2003).

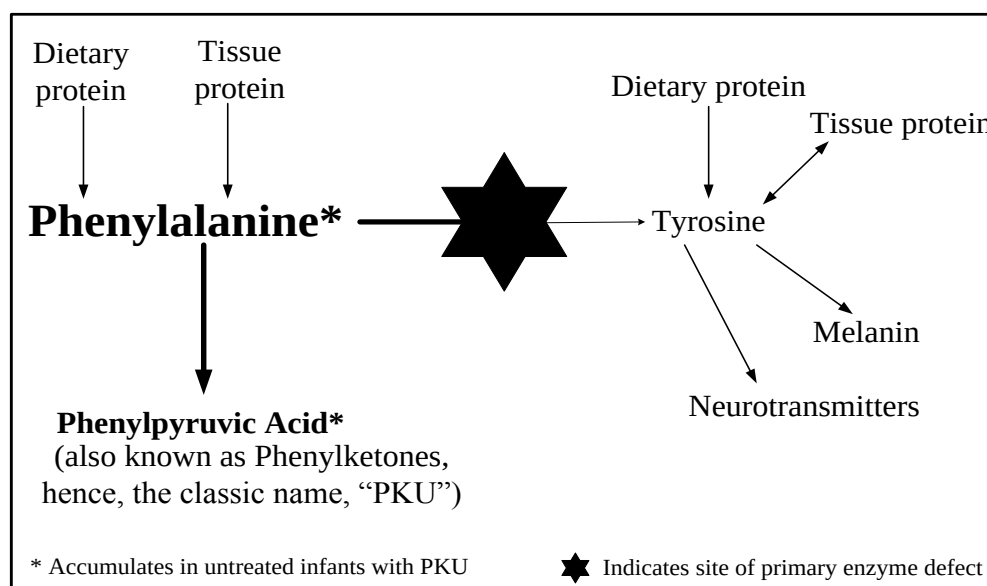


Figure 1. Simplistic Pathway of Phenylalanine Metabolism in PKU.

In the United States, approximately 160 to 400 infants per year are diagnosed with PKU (Scriver et al., 2008). Although infants with PKU appear

normal at birth, symptoms of Phe toxicity slowly develop over time and may include eczema-like skin rash, seizures, excessive restlessness, irritable behavior, and a “musty” odor of the body, urine, and perspiration (Jervis, 1953; Paine, 1957; Partington, 1961). As the toxic metabolites accumulate within the body fluids and tissues, consequent developmental delays, gait disturbances, and mental retardation become clinically present (Jervis, 1953; Paine, 1957; Smith, Beasley, & Ades, 1990).

Today, diagnosis is made soon after birth from results of newborn screening tests, and treatment for PKU begins immediately (Dhondt, 2007; Guthrie & Susi, 1963; Therrell & Adams, 2007). The treatment of PKU involves a lifelong diet low in Phe, frequent monitoring of blood Phe levels to maintain therapeutic desired levels from 120 to 360 μ mol/dl, and regular consultation with the pediatric metabolic team of physicians, nurses, and dieticians (Beasley, Costello, & Smith, 1994; Cerone, Schiaffino, Di Stefano, & Veneselli, 1999; de Baulny, Abadie, Feillet, & de Parscau, 2007; Luciana, Sullivan, & Nelson, 2001; Yap & Naughten, 2001). Normal cognitive development is expected when Phe blood levels are managed and remain within acceptable levels that range 120 to 360 μ mol/dl and the phenylalanine/tyrosine (Phe/Tyr) ratio remains low (< 2.5) (Beasley et al., 1994; Chace, Sherwin, Hillman, Lorey, & Cunningham, 1998; de Baulny et al., 2007; Luciana et al., 2001; Yap & Naughten, 2001). *Even with* acceptable Phe blood levels, many children with PKU have slightly lower cognitive and executive functioning, and more behavioral issues compared to their non-PKU siblings or age-related healthy friends (Anastasoae, Kurzius,

Forbes, & Waisbren, 2008; Luciana et al., 2001; White et al., 2009). However, in a study from Italy, school-age children with PKU who were breastfed as infants had significantly higher intelligence scores than school-age children with PKU who were exclusively formula fed as infants ($p < 0.01$) and the significance held after controlling for maternal confounding variables ($p < 0.02$) (Riva et al., 1996).

Breast Milk and Breastfeeding

Breastfeeding and human breast milk are more beneficial for the majority of infants than formula feeding with standard commercial infant formulas. Research has identified positive health outcomes from breastfeeding for full-term infants and mothers (Hoddinott et al., 2008; Horta et al., 2007; Ip et al., 2007; U.S. Department of Health and Human Services, 2003). These advantages extend not only to infants and their mothers but to their families and society as well (Riordan, 1997; Stuebe, 2009; Weimer, 2001).

Infant Feeding and Health Outcomes

Nutritional and growth benefits. Studies have revealed that human breast milk meets the human infant's nutritional requirements for growth and development better than the milk of any other species (Kramer & Kakuma, 2002; Picciano, 2001). When breastfed and formula fed infants were followed during the first 12 months, the two groups had similar weight gains during the first three months, but breastfed infants gained less weight during the remainder of the first year (Dewey, Heinig, Nommsen, Pearson, & Lonnerdal, 1992). Dewey and colleagues analyzed data from seven longitudinal studies on infant growth in North America and northern Europe, as well as reference data from the World

Health Organization (WHO) and Centers for Disease Control and Prevention (CDC) (Dewey, Peerson, et al., 1995). Breastfed infants were found to grow more rapidly during the first two months and less rapidly from three to 12 months (Dewey, Peerson, et al., 1995). An Italian study compared breastfed and formula fed infants during the first 12 months and found breastfed infants had a decline in growth rate during the second six months of life (Agostoni et al., 1999).

Consistently, the normal pattern of breastfed infants was to gain less weight and to be leaner at one year of age than formula fed infants (Agostoni et al., 1999; Dewey, 1998; Dewey et al., 1992; Dewey, Peerson, et al., 1995).

Obesity. The first indication of a significant protective effect of breastfeeding on later childhood obesity was identified in a case-control study involving adolescents for whom the relative risk for obesity was 2.35 times greater if the adolescent had not been breastfed (Kramer & Moroz, 1981). Even after controlling for family history, the protective effect of breastfeeding against later obesity remained. Reviewing published research from the 1990s, Dewey (2003) concluded that children who were breastfed had a lower risk of being overweight in later childhood. Four meta-analyses also revealed that children who were breastfed during infancy were less likely to become obese in childhood (Dewey, 2003; Harder, Bergmann, Kallischnigg, & Plagemann, 2005; Horta et al., 2007; Ip et al., 2007).

The protective effect of breastfeeding against childhood obesity may rest in the duration of breastfeeding. In a large prospective English study ($n = 1,195$), longer duration of breastfeeding was strongly associated with a lower fat mass at

age 4 years (Robinson et al., 2009). A meta-analysis examined the duration of breastfeeding and risk of obesity and yielded a dose-dependent association between longer duration of breastfeeding and decreased risk of obesity (Harder et al., 2005). However, in a critical review, Butte (2001) found breastfeeding had a reduced risk of childhood obesity in only one-fourth (four out of 16) of the studies, and those four studies had unresolved questions regarding confounding factors. Several recent studies and reviews examined the effects of breastfeeding on later obesity and were unable to identify a significant effect (Huus, Ludvigsson, Enskar, & Ludvigsson, 2008; Neutzling et al., 2009). Although the evidence remains unclear, breastfeeding may have a small protective effect from obesity.

Hypertension. Research on the influence of breastfeeding for the development of hypertension later in life is limited. In a meta-analysis, the mean systolic and the diastolic blood pressure were decreased in adulthood among those who were breastfed compared to formula fed (1.10 mm Hg systolic, 95% CI 0.42-1.78; 0.36 mm Hg diastolic, 95% CI -0.08 to 0.79) (Owen, Whincup, Gilg, & Cook, 2003). In another meta-analysis, mean systolic blood pressure was lower by 1.4mm Hg (95% CI 0.6-2.2) in adulthood for those breastfed compared to those formula fed (Martin et al., 2005). This limited research suggests there may be an association between being breastfed and a small reduction in blood pressure in later adulthood, but more studies are needed to verify this.

Total cholesterol. There has been growing evidence that cardiovascular disease begins before adulthood (Enos & Beyer, 1971; Strong et al., 1999). In

the only meta-analysis that evaluated breastfeeding and cholesterol in adulthood, lower mean total cholesterol levels were found in adults who were breastfed compared to those who were formula fed (mean difference of -6.96 mg/dl; 95% CI -2.23 to -11.6mg/dl). In addition, adults who were breastfed had lower mean LDL cholesterol levels (mean difference of -7.7 mg/dl; 95% CI -3.09 to -12.37 mg/dl) (Owen, Whincup, Odoki, Gilg, & Cook, 2002). Although this meta-analysis had intriguing findings, it is difficult to conclude an association between breastfeeding and adult cholesterol levels. More research is needed on this topic.

Developmental benefits. Studies on breastfeeding and cognitive development have revealed inconsistent findings. Whereas some studies identified no association between breastfed children and cognitive performance (Der, Batty, & Deary, 2006; Gale et al., 2010), other studies have identified an increase in cognitive development (i.e., higher IQ scores) in children who were breastfed (Anderson, Johnstone, & Remley, 1999; Der et al., 2006; Drane & Logemann, 2000; Hamosh & Salem, 1998; Horwood, Mogridge, & Darlow, 1999; Jacobson, Chiodo, & Jacobson, 1999; Jain, Concato, & Leventhal, 2002; Lucas, Morley, Cole, Lister, & Leeson-Payne, 1992). From their mega analysis, Der and colleagues (2006) found that when maternal intelligence was controlled, there was little to no evidence for an association between having been breastfed in infancy and subsequent cognitive performance. In a recent study from England, maternal or family environment and stimulation of infants were confirmed to be variables associated with increased IQ rather than having been breastfed in infancy (Gale et al., 2010). However, three earlier meta-analyses yielded

favorable results for later cognitive outcomes in infants who were breastfed (Anderson et al., 1999; Drane & Logemann, 2000; Jain et al., 2002), although the findings were less robust for studies with larger samples (Drane & Logemann, 2000; Jain et al., 2002).

A prospective longitudinal study in Australia found the benefits of breastfeeding in infancy to extend from toddlerhood to adolescence. After controlling for socioeconomic status and quality of the child's social environment, breastfed infants were advantaged with higher scores on the Bayley Mental Developmental Index at 2 years, higher scores on the McCarthy General Cognitive Index at 4 years, and higher scores on the Wechsler Full-Scale IQ at 7 years of age and again when 11 to 13 years (Wigg et al., 1998). Such longer-term benefits may be linked to duration of breastfeeding rather than just the fact of breastfeeding per se. Another prospective study assessed the duration of breastfeeding and found that longer duration predicted higher IQ and reading comprehension scores as well as higher scores on school examinations (Horwood et al., 1999).

It may be that breastfeeding provides more or higher-quality interaction between mother and infant. A study that focused on an intervention to enhance the duration of breastfeeding found infants who were breastfed longer subsequently had significantly higher verbal scores on cognitive testing (Kramer et al., 2008). It may be that breastfeeding affords more verbal interaction and other qualities that stimulate or improve cognitive development. Whatever the influence, it seems that being breastfed in infancy improves cognitive

development and contributes to positive cognitive outcomes, potentially through adolescence.

Resistance to infectious diseases. Human breast milk contains factors associated with resistance to infections. These include secretory antibodies, lactoferrin, lysozyme and bifidus factor, and other specific and innate immune factors that act against viruses and bacteria (Hamosh, 2001; Lawrence & Pane, 2007). In addition, human milk fat globule glycoproteins, mucin, and lactadherin are resistant to digestive enzymes and gastric conditions, and they are able to lyse enveloped viruses, bacteria, and protozoa (Hamosh et al., 1999). From the bronchial tree and intestine, plasma cells migrate to the mammary epithelium and produce immunoglobulin A antibodies specific to antigens in the mother-infant dyad's surroundings, thereby providing specific protection against local pathogens (Forchielli & Walker, 2005). Oligosaccharides, MUC-1 mucin, lactadherin, and glycoaminoglycans impede common pathogens, such as *Streptococcus pneumoniae*, *Escherichia coli*, and rotavirus (Lawrence & Pane, 2007). This is accomplished by preventing harmful bacteria from multiplying and causing an infection, or by damaging the outer surface of certain viruses and preventing replication (Peterson, Patton, & Hamosh, 1998). In addition to its nutritional value, breast milk has value as a protective function in the transfer of factors that provide an advantage that formula fed infants do not have.

Gastrointestinal infections. For over 30 years, studies have revealed the protective effect of breastfeeding for preventing the development of gastrointestinal infections (Bachrach, Schwarz, & Bachrach, 2003; Chen &

Rogan, 2004; Chien & Howie, 2001; Dewey, Heinig, & Nommsen-Rivers, 1995; Feachem, 1984; Indrio, Ladisa, Mautone, & Montagna, 2007; Jeppesen, Hasselbalch, Lisse, Ersboll, & Engelmann, 2004; Kramer et al., 2001; Penders et al., 2006; Quigley, Cumberland, Cowden, & Rodrigues, 2006). In one study, breastfed infant stools had a lower pH and greater colonization of non-pathogenic bacteria while the gut microbiotic compositions of formula fed infants had more pathological bacteria (Penders et al., 2006). Thymuses of breastfed infants are larger than those of formula fed infants, and a positive correlation exists between thymus size and CD8+ cells, indicating both short- and long-term immune modulating effects on the developing cellular immune system during infancy (Indrio et al., 2007; Jeppesen et al., 2004). Consequently, breastfed infants have more favorable gut colonization due to factors in breast milk that regulate colonization and the differentiation of T-cells.

Multiple meta-analyses found that the incidence and severity of bacterial and viral infections, such as gastroenteritis, are lower in breastfed infants (Bachrach et al., 2003; Chien & Howie, 2001; Ip et al., 2007). In 1984, early reviews on breastfeeding and diarrhea morbidity found breastfed infants were less likely to have diarrhea within the first six months of life (Feachem, 1984). Ten years later, Dewey and colleagues (1995) verified that the incidence of diarrheal illness in the first year was lower for breastfed infants. In a prospective study with an intervention based on the Baby-Friendly Hospital Initiative that supported initiation and maintenance of breastfeeding, mothers receiving the intervention had breastfed infants with a reduction in gastrointestinal tract

infections (Kramer et al., 2001). Consistently, breastfed infants have had a reduction in gastrointestinal infection during the first year of life (Chen & Rogan, 2004; Quigley et al., 2006).

Acute otitis media. Breast milk contains immunoglobulins with antibody activity against *Haemophilus influenza* and *Streptococcus pneumonia*, and it contains components that interfere with their attachment to the nasopharyngeal epithelial cells (Andersson, Porras, Hanson, Lagergard, & Svanborg-Eden, 1986). One study proposed breastfeeding allows for intermittent administration of breast milk with anti-adhesive substances into the nasopharyngeal of the nursing infant, thereby reducing the extent of colonization and enhancing protection from infection (Nylen, Anderson, & Aniansson, 1990). In a prospective study, acute otitis media (AOM) was decreased in breastfed infants until four months after breastfeeding was discontinued (Sassen, Brand, & Grote, 1994). Another prospective study found infants who were breastfed for at least six months had a decreased risk of AOM and otitis media effusion (Duffy, Faden, Wasielewski, Wolf, & Krystofik, 1997). Repeatedly, breastfeeding has been associated with reduction in AOM (Chung, Raman, Trikalinos, Lau, & Ip, 2008; Ip et al., 2007; Vogazianos, Vogazianos, Fiala, Janecek, & Slapak, 2007).

Lower respiratory tract infections. Studies have shown a clear relationship between breastfeeding and a decreased incidence of respiratory tract infections (Frank et al., 1982; Ip et al., 2007; Libster et al., 2009; Mathews & MacDorman, 2008; Victora et al., 1999; Woodward, Douglas, Graham, & Miles, 1990; Wright, Holberg, & Martinez, 1989; Wright, Holberg, Taussig, & Martinez,

2001; Wright, Taussig, & Ray, 1989). The absence of breastfeeding during the first six months of infancy is a risk factor for acute lower respiratory infection and subsequent hospitalization (Ip et al., 2007; Mathews & MacDorman, 2008; Victora et al., 1999). In a meta-analysis, Bachrach and colleagues (2007) found breastfed infants in developed countries were less likely to be hospitalized for lower respiratory tract infections during the first year. In a prospective cross-sectional study from Argentina, a differential gender effect for breastfeeding and breast milk was found, with term infant girls, not boys, protected against viral pneumonia and probable hospitalization (Libster et al., 2009). Overall, breastfeeding for four or more months resulted in a reduction in the risk of lower respiratory tract diseases leading to hospitalization.

Asthma. The evidence for a protective effect of breastfeeding on asthma remains a matter of controversy (Chung et al., 2008; Fergusson, Horwood, & Shannon, 1983; Gdalevich, Mimouni, & Mimouni, 2001; Ip et al., 2007; Kull, Almquist, Lilja, Pershagen, & Wickman, 2004; Wilson, Forsyth, et al., 1998; Wright, Holberg, Taussig, & Martinez, 1995). An early study concluded there was no evidence to indicate that breastfeeding had any effect on the subsequent asthma risk (Fergusson et al., 1983). Yet a more recent study reported exclusive breastfeeding for four months or more was associated with a reduced risk of asthma, although the association was strongest for children whose parents did not have a history of allergies (Kull et al., 2004). This finding is consistent with the results of an earlier meta-analysis that revealed breastfeeding for less than three months and having a family history of asthma increased the risk for asthma

(Gdalevich, Mimouni, & Mimouni, 2001). Subsequent meta-analyses have generally confirmed that breastfeeding provides for a reduced risk of asthma in children with a family history of asthma (Ip et al., 2007) and without a history of asthma (Chung et al., 2008). Further study is needed, however, to understand the role of family history of asthma in conjunction with the benefits of breastfeeding.

Atopic dermatitis. Several studies have explored the effect of breastfeeding on the development of atopic dermatitis during infancy and different conclusions have been reached (Gdalevich, Mimouni, David, & Mimouni, 2001; Gdalevich, Mimouni, & Mimouni, 2001; Kramer et al., 2001; Kull, Wickman, Lilja, Nordvall, & Pershagen, 2002; Nakamura et al., 2000). In one study, breast milk feedings elevated the risk of atopic dermatitis (Nakamura et al., 2000) whereas a meta-analysis revealed infants with a family history of atopy dermatitis and who were not breastfed for more than three months had a higher risk for developing the disease (Gdalevich, Mimouni, David, et al., 2001). In a randomized trial, mothers who breastfed longer were less likely to have an infant develop atopic dermatitis (Kramer et al., 2001; Kull et al., 2002). Thus, longer duration of breastfeeding, defined as more than three months, may have more protective benefit.

Diabetes. There have been numerous epidemiological studies regarding cow's milk formula and the development of type 1 diabetes (Gerstein & VanderMeulen, 1996; Ip et al., 2007; Norris & Scott, 1996). Meta-analyses revealed that compared to infants who breastfed for three months or more,

infants who never breastfed (Norris & Scott, 1996) and who breastfed for less than three months (Gerstein, 1994; Norris & Scott, 1996) had an increased risk of developing type 1 diabetes. Ip and colleagues (2007), in a more current meta-analysis, verified the previous findings that breastfeeding for more than three months decreased the risk of type 1 diabetes.

A limited number of studies have explored whether being breastfed in infancy may also reduce the risk of type 2 diabetes later in life (Aynsley-Green, Lucas, & Bloom, 1979; Lucas et al., 1980; Pettitt, Forman, Hanson, Knowler, & Bennett, 1997; Ravelli, van der Meulen, Osmond, Barker, & Bleker, 2000). In a cohort of Pima Indians in Arizona, infants who were exclusively breastfed during the first two months were less likely to develop type 2 diabetes (Pettitt, Narayan, Hanson, & Knowler, 1996). In a retrospective study of adults in the Netherlands, those who had been exclusively breastfed were less likely to have developed type 2 diabetes compared to those who had been partially or exclusively formula-fed (Young et al., 2002). However, a cohort study of older men (mean age of 71 years) revealed type 2 diabetes was not linked to having been breastfed (Martin et al., 2005). Two recent meta-analyses on breastfeeding and type 2 diabetes, however, found having ever been breastfed was identified with a lower risk of type 2 diabetes later in life (Horta et al., 2007; Owen, Martin, Whincup, Smith, & Cook, 2006). Overall, it seems that being breastfed in infancy may decrease the risk of type 2 diabetes, but the studies are few and further research is needed.

Childhood leukemias. Based on the hypothesis that immunoactive factors in breast milk may prevent viral infections associated with leukemia,

several studies have explored the relationship between formula feeding and childhood leukemia (Greaves, 1988). In two meta-analyses, an increased risk of acute lymphoblastic leukemia (ALL) seemed to be more common among formula fed children compared with children who had been breastfed for more than six months (Ip et al., 2007; Kwan, Buffler, Abrams, & Kiley, 2004). A case-control study reported a similar lower risk of ALL for breastfed infants (Guise, Austin, & Morris, 2005). In addition, Kwan et al. (2004) revealed formula fed infants had an increased risk of acute myeloid leukemia (AML) compared with infants who had breastfed for six months. In a study from Turkey, breastfeeding for more than six months seemed to provide protective benefit against childhood leukemias, especially ALL and AML (Altinkaynak, Selimoglu, Turgut, Kilicaslan, & Ertekin, 2006). Breastfeeding for at least six months seems to have an influence on preventing the development of ALL and AML, but more studies are needed due to limited research.

Infant mortality. There are numerous studies regarding the impact of breastfeeding on postnatal mortality (Betran, de Onis, Lauer, & Villar, 2001; Chen & Rogan, 2004; Edmond et al., 2006; Terra de Souza et al., 1999; Vennemann et al., 2009). In a case-control study in one of the poorest regions in Brazil, breastfeeding was reported to have a substantial effect in reducing infant mortality due to diarrhea (Terra de Souza et al., 1999). Similarly, exclusive breastfeeding for the first three months and partial breastfeeding for the remainder of infancy significantly reduced infant mortality in Latin America (Betran et al., 2001). Breastfed infants have also been found to have a lower risk

for neonatal mortality compared to infants who were only formula fed in the U.S. (Chen & Rogan, 2004) as well as in Ghana. In both developed and developing countries, a reduction in infant mortality occurs with breastfeeding during infancy.

Sudden infant death syndrome (SIDS) is the leading cause of infant mortality in the U.S. (Center for Disease Control and Prevention, 2009). Infants who were not breastfed were found twice as likely to die of SIDS (Alm et al., 2002; Fredrickson, 1993). Two meta-analyses of case control studies confirmed that formula feeding compared to breastfeeding was associated with an increased risk of SIDS (Ip et al., 2007; McVea, Turner, & Peppler, 2000). Recent studies in the U.S. and Germany have substantiated that a reduced risk for SIDS was identified with breastfeeding (Chung et al., 2008; Vennemann et al., 2009). Clearly, there is a relationship between breastfeeding and a reduced risk of SIDS.

Maternal Health Outcomes

Breast cancer. Several longitudinal studies have found a protective association between breastfeeding and breast cancer (Kvale & Heuch, 1987; Lee, Kim, Kim, Song, & Yoon, 2003; London et al., 1990; Michels et al., 1996; Tryggvadottir, Tulinius, Eyfjord, & Sigurvinsson, 2001). Two meta-analyses revealed that women who breastfed their infants were less likely to be diagnosed with breast cancer (Bernier, Plu-Bureau, Bossard, Ayzac, & Thalabard, 2000; Collaborative Group on Hormonal Factors in Breast Cancer, 2002). In a case-control study from India, duration of breastfeeding in premenopausal women and parity in postmenopausal women were important risk-lowering factors for breast

cancer (Gajalakshmi et al., 2009). These findings hold for women with a family history of breast cancer as well (Stuebe, Willett, Xue, & Michels, 2009). Overall, risk of breast cancer is reduced in women who breastfed their infants.

Ovarian cancer. Various studies have identified a higher risk of ovarian cancer among parous women who never breastfed (Chiaffarino et al., 2005; Danforth et al., 2007; Ip et al., 2007; Zhang, Xie, Lee, & Binns, 2004). In a case-control study from China, which has a low incidence of ovarian cancer, an inverse relation between breastfeeding duration and ovarian cancer risk was confirmed as well as a trend for lower risk with more children breastfed (Zhang et al., 2004). An Italian case-control study simply revealed an inverse relation between breastfeeding and ovarian cancer risk (Chiaffarino et al., 2005). Subsequently, the only meta-analysis that examined this relationship between breastfeeding and ovarian cancer again found that women who never breastfed had a greater risk of ovarian cancer (Ip et al., 2007). In pooled data from the Nurses' Health Studies, Danforth and colleagues (2007) verified parous women who never breastfed have an increased risk of ovarian cancer. Having breastfed seems to decrease the risk of ovarian cancer, but this is based on retrospective studies. Prospective research is needed to confirm that, indeed, women who breastfeed their infants are less likely to develop ovarian cancer.

Risk of maternal cardiovascular disease. A rather new finding is the relationship between breastfeeding and lower postmenopausal risk for cardiovascular disease. The first reported study examined the relationship between the duration that women breastfed and the subsequent postmenopausal

risk for maternal cardiovascular disease and found that an increased duration of breastfeeding was associated with a lower prevalence of cardiovascular disease (Schwarz et al., 2009). In a follow-up study on breastfeeding and maternal subclinical cardiovascular disease, mothers who had never breastfed were more likely to have coronary artery calcification, aortic calcification, carotid plaque, and larger carotid adventitial diameters compared with women who had breastfed for at least three months (Schwarz et al., 2010). In the Nurses' Health Study prospective cohort, an inverse association between breastfeeding cumulatively for two years or longer and myocardial infarction was found to be independent of risk factors for cardiovascular disease (i.e., obesity at age 18, smoking, exercise, diet, aspirin use, alcohol consumption, hormone use, and menopausal status) (Stuebe, Michels, et al., 2009). Although there is limited research, women who breastfeed their infants seem to be at a decreased risk for maternal cardiovascular disease.

Maternal diabetes. Studies have suggested a higher risk of diabetes among women who only formula fed their children (Schwarz et al., 2009; Stuebe, Rich-Edwards, Willett, Manson, & Michels, 2005). Duration of breastfeeding has been inversely associated with lower diabetes risk at 12 to 18 months postpartum independent of body adiposity (Diniz & Da Costa, 2004). Further, each additional year of breastfeeding duration was associated with decreased risk of type 2 diabetes by 15% (Stuebe et al., 2005). These findings have held up in more recent studies as well (Schwarz et al., 2009; Villegas et al., 2008). However, breastfeeding does not lower the risk of developing type 2 diabetes in women

who had gestational diabetes (Kjos et al., 1995; Kjos et al., 1998; Peters, Kjos, Xiang, & Buchanan, 1996; Pettitt et al., 1996). It seems that longer duration of breastfeeding results in a reduced risk of developing type 2 diabetes in parous women who have not had a history of gestational diabetes. Again, more studies are needed.

Maternal hypertension. Another rather new finding is the relationship between breastfeeding and maternal hypertension. In a prospective, observational study of Korean women, after adjusting for confounding factors, breastfeeding was associated with a decreased risk of hypertension in women who breastfed for more than one month (Lee, Kim, Jee, & Yang, 2005). In a study from the U.S. using data from the Women's Health Initiative, women who breastfed their infants for more than 12 months were less likely to develop hypertension (Schwarz et al., 2009). Whatever the influence, it seems having breastfed prevents later maternal hypertension, although further prospective research is needed to explore this topic.

Maternal weight loss. Several studies have explored the effect of breastfeeding on subsequent weight loss during postpartum and conflicting conclusions have been reached. In a prospective study, women who breastfed achieved their pre-pregnancy weight approximately six months earlier than women who bottle fed, yet longer duration of breastfeeding was actually the significant predictor of postpartum weight retention over time (Janney, Zhang, & Sowers, 1997). In another prospective study, the effect of breastfeeding on maternal weight loss was negligible (Janney, Zhang, & Sowers, 1997; Olson,

Strawderman, Hinton, & Pearson, 2003; Sichieri, Field, Rich-Edwards, & Willett, 2003), whereas another study found a weak association between breastfeeding at one year and less retained weight (Olson et al., 2003). The only published systematic review of research on breastfeeding for postpartum weight loss revealed inadequate evidence to support such an effect (Fraser & Grimes, 2003). A meta-analysis of eight studies revealed the effect of breastfeeding on return to pre-pregnancy weight was negligible (Ip et al., 2007). Overall, in the majority of the studies, there seem to be other factors, such as annual household income and ethnicity, which have more effect on the lack of postpartum weight loss than breastfeeding. Further research is needed.

Postpartum depression. Several studies have examined breastfeeding and postpartum depression (Chaudron et al., 2001; Chung et al., 2008; Cooper, Murray, & Stein, 1993; Henderson, Evans, Straton, Priest, & Hagan, 2003; Ip et al., 2007; O'Hara & Swain, 1996; Warner, Appleby, Whitton, & Faragher, 1996). An early study revealed that depressed mothers were less likely to continue to breastfeed beyond two to four months (Cooper et al., 1993). In fact, predictors of postpartum depression have included past history of psychopathology and psychological disturbance during pregnancy, as well as not breastfeeding (O'Hara & Swain, 1996; Warner et al., 1996). Chaudron and colleagues (2001) found that women who were breastfeeding at one month and were worried about breastfeeding were more likely to become depressed than those who did not worry. In another study, postpartum depression seemed to occur with early cessation of breastfeeding (Henderson et al., 2003), and two meta-analyses

confirmed that postpartum depression seemed to occur with either not breastfeeding or a short duration of breastfeeding (Chung et al., 2008; Ip et al., 2007). It may be that postpartum depression interferes with the decision to breastfeed or continue to breastfeed rather than breastfeeding preventing depression. Clearly, further research is needed to understand this complex topic.

Socio-Economic Benefits

Economics of breastfeeding. To the family, the healthcare system, the employer, and the country, breastfeeding offers economic benefits. Many early studies addressed the economic benefits to breastfeeding by comparing breastfeeding with formula feeding (Montgomery & Splett, 1997; Tuttle & Dewey, 1996). In these studies, breastfeeding was economically advantageous and the promotion of breastfeeding provided effective cost-containment. Families could save several hundred dollars by having mothers breastfeed. For healthcare systems, breastfeeding is less costly than purchasing standard commercial infant formula and bottle supplies. Studies identified and confirmed that breastfed infants have fewer ill child visits to healthcare providers, fewer prescriptions, and fewer hospitalizations (Avendano et al., 1993; Riordan, 1997). In a study evaluating breastfeeding in an HMO setting, total medical care costs were 20% lower for breastfed infants (Hoey & Ware, 1997).

When mothers who are breastfeeding return to work after the birth of their infants, employers benefitted as these mothers reported improved morale, better satisfaction with their jobs, and higher productivity (Galtry, 1997). As their infants were healthier, maternal absenteeism from work was significantly lower in

companies that have established breastfeeding programs for employees (Ball & Bennett, 2001; Cohen, Mrtek, & Mrtek, 1995). At one large insurance company, the retention rate of new mothers was 83% compared to the national average of 59% and was attributed to having a lactation program in the workplace (Mutual of Omaha, 2001). Lactation programs also increase the duration of breastfeeding for women who breast pumped at work and continued to breastfeed their infants for a mean duration of 9.1 months (Ortiz, McGilligan, & Kelly, 2004). Increased duration of breastfeeding likely extends all the other benefits as well.

Abandonment. The act of breastfeeding requires the engagement of both the mother and infant. The connectedness between mother and infant is encouraged through early breastfeeding and rooming-in. Whatever the influence, it seems breastfeeding results in decreased infant abandonment (Klaus, 1998). In a Thai study, a hospital had a decrease in infant abandonment from 33 per 10,000 to 1 per 10,000 births after becoming a Baby-Friendly Hospital (Buranasin, 1991). Following implementation of the Baby-Friendly Hospital Initiative that included early mother-infant contact with breastfeeding, rooming-in, and increased opportunities to breastfeed, there was also a decreased rate in the mean infant abandonment rate at a Russian hospital (Lvoff, Lvoff, & Klaus, 2000). The reduction of child abandonment could be the result of longer duration of breastfeeding, which helps enhance the spacing of children, thereby reducing the economic burden on the family (Thapa, Short, & Potts, 1988).

Environmental benefits. Breastfeeding is better for the environment given there is less trash and plastic waste compared to formula cans and bottle

supplies. In comparison to standard commercial infant formulas, breastfeeding requires no fossil fuels for its manufacture or preparation (Gilman & Skillicorn, 1985). Breastfeeding reduces pollutants, such as the release of methane gas created from by-products during the manufacture of bottles, caps, nipples, and standard commercial infant formulas, and thereby reduces the burden on landfills (Radford, 1991). Overall, breastfeeding may be considered to contribute to the health of the planet.

Breastfeeding Infants with Chronic Conditions

The care of breastfed infants differs from infants being fed standard commercial infant formula. In addition, when breastfed infants have chronic healthcare conditions, they and their families require special consideration. Any condition diagnosed at birth might have an impact on breastfeeding; hence, it is vital for healthcare providers to be knowledgeable about when and how to intervene to facilitate the breastfeeding experience of mothers with infants with chronic conditions who would benefit from the advantages of breast milk and breastfeeding.

The advantages of breastfeeding and the use of human breast milk are particularly significant for infants with chronic conditions. Human breast milk has unique nutritional properties that make it more easily digested so that the gastrointestinal tract can mature. In addition, breastfeeding promotes a very special attachment between mother and infant. Infants with chronic conditions can vary from extreme prematurity to full term, with visible congenital anomalies or invisible inborn errors of metabolism.

Thirty years ago, the research on breastfeeding premature infants was lacking. Nursing research advanced the art and science of breastfeeding premature infants by advocating for research-based interventions to facilitate breastfeeding for this vulnerable population. Today, there are over 200 papers on breastfeeding the premature infant that have provided research-based recommendations for clinical protocols. In contrast, only 71 papers have addressed breastfeeding infants with a limited number of other chronic conditions. Of these 71 papers, 33 were research studies with more than half ($n = 25$) related to cleft lip and cleft palate (CLP). The remaining papers were a combination of review articles ($n = 32$ with 22 on CLP), case reports ($n = 4$ all on CLP), and protocols ($n = 2$). Combined, these papers addressed breastfeeding with only five chronic conditions: congenital heart defects (CDH), CLP, gastroschisis, hypotonia/Trisomy21, and spina bifida (SB). The authors mainly focused on identifying breastfeeding problems and factors associated with difficulty breastfeeding (Aniansson, Svensson, Becker, & Ingvarsson, 2002; Barbas & Kelleher, 2004; Barsky & Boiteux, 2008; Biancuzzo, 1998; Combs & Marino, 1993; Crossman, 1998; da Silva Dalben, Costa, Gomide, & Teixeira das Neves, 2003; Danner, 1992; Garcez & Giugliani, 2005; Heinzl & Baltzer, 2003; Huner & Demirkol, 1996; Hurtekant & Spatz, 2007; Jamerson, 2005; Kogo et al., 1997; Lambert & Watters, 1998; Marino, O'Brien, & LoRe, 1995; Moe, Holland, & Johnson, 1998; Nejat et al., 2008; Pisacane et al., 2003; Reilly, Reid, & Skeat, 2007; Rivera, Davila Torres, Parrilla Rodriguez, de Longo, & Gorrin Peralta, 2008; Thomas, Marinelli, & Hennessy, 2007).

This rather small number of papers is disturbing as multiple chronic conditions during infancy are likely to present special challenges for mothers to breastfeed. In some cases, the challenges are a function of suckling problems, a function of metabolism, or a function of feeding intolerance and esophageal or intestinal dysmotility. Whatever the cause, the consequence can be poor infant growth with failure to thrive. On the other hand, the known benefits of human breast milk are especially important for infants born with certain chronic conditions. For example, infants with CL/P and Trisomy 21 are at increased risk for otitis media and middle ear effusion (Aniansson et al., 2002; Saarinen, 1982; Thomas et al., 2007). Infants with CHD, SB, Trisomy 21, gastroschisis, and prematurity are all at increased risk for respiratory infections (Abdullah et al., 2007; Caterino, Scheatzle, & D'Antonio, 2006; Kingsmore et al., 2008; Ma, Gauvreau, Allan, Mayer, & Jenkins, 2007). The unique immunological properties of human breast milk, whether expressed or suckled from the breast, have added value for these infants.

Clearly, there is a paucity of research on breastfeeding infants with chronic conditions even though, according to the American Academy of Pediatrics (AAP), the American College of Nurse Midwifery (ACNM), and the Association of Women's Health, Obstetrics and Neonatal Nursing (AWHONN) (Association of Women's Health Obstetric and Neonatal Nurses, 2005; Gartner et al., 2005; Nurse-Midwives, 2004), breast milk is the preferred choice for feeding all infants. These healthcare organizations recommend that mothers breastfeed their infants for the first six months of age, preferably with continuation of

breastfeeding to age 12 months as table foods are introduced. However, more research is needed regarding factors that influence breastfeeding decisions and to decrease disparities in breastfeeding initiation and duration among subgroups, such as mothers who are breastfeeding infants with chronic conditions.

Breastfeeding Infants with PKU

Dietary Management of PKU

In the early years of PKU treatment, the standard of care for infants diagnosed with PKU was to be immediately weaned from breastfeeding. To maintain appropriate Phe levels (120-360 μ mol/dl), infants were then placed on a diet of Phe-free or low-Phe formula combined with standard commercial infant formula (Ernest, McCabe, Neifert, & O'Flynn, 1980; Huner & Demirkol, 1996; McCabe et al., 1989; Motzfeldt, Lilje, & Nylander, 1999). This combination was believed to be the only effective way to monitor the infant's intake and allow for precise titration and measurement of the input of Phe, in order to protect the infant's neurological and cognitive development (Blau, Duran, & Gibson, 2008). Mothers of infants with PKU typically titrated Phe-free formula with standard commercial formula based on Phe values obtained from infant heel sticks. This management strategy thus precluded breastfeeding infants with PKU (Ernest et al., 1980; Huner & Demirkol, 1996; McCabe et al., 1989; Motzfeldt et al., 1999).

Research then determined that breast milk was lower in Phe than standard commercial infant formula (Janas, Picciano, & Hatch, 1985; Lonnerdal, Forsum, Gebre-Medhin, & Hambraeus, 1976; Nayman, Thomson, Scriver, & Clow, 1979). This meant that mothers of infants with PKU could breastfeed them.

Further, a larger volume of breast milk than standard commercial infant formula could be fed to infants with PKU.

In 1983, the first publication appeared describing breastfeeding infants with PKU (McCabe, Ernest, & McCabe, 1983). Subsequent research on combining breast milk and Phe-free formula in managing the diet of infants with PKU found that this combination (breastfeeding and Phe-free formula) did not adversely affect Phe control (Greve, Wheeler, Green-Burgeson, & Zorn, 1994; Hinrichs, Biggemann, & Wendel, 1994; Huner & Demirkol, 1996; McCabe et al., 1989). In addition, research on formula fed infants with PKU compared to breastfed infants with PKU revealed that those who had been breastfed and supplemented with Phe-free formula in the first six months had lower protein and Phe levels (van Rijn et al., 2003). More recent studies found that breastfed infants with PKU compared to formula fed infants with PKU had no significant differences in weight gain, daily Phe intake, or mean plasma Phe concentrations (Cornejo et al., 2003; Huner et al., 2005; Kanufre et al., 2007; McCabe et al., 1989; Motzfeldt et al., 1999; van Rijn et al., 2003). Although based on relatively small samples (< 50 participants), the results from these studies consistently supported the conclusion that breast milk supplemented with Phe-free formula is an acceptable dietary treatment for infants with PKU.

Guidelines were introduced in 1980 for breastfeeding infants with PKU that included a commercial formula with low amounts of Phe as part of the dietary management (Ernest et al., 1980). The document recognized that more infants with PKU were being breastfed at the time of diagnosis and their mothers

were reluctant to discontinue breastfeeding, and it concluded that breastfeeding was feasible and beneficial to both infants and mothers. It also acknowledged the absence of research-based guidelines for breastfeeding infants with PKU and for supporting their mothers. The 1980 guidelines were derived from clinical experience alone.

Phenylalanine-free formula was introduced to the dietary management of PKU in 1983 (Flannery, Hitchcock, & Mamunes, 1983). At the same time, an alternative approach was developed using estimates of the breast milk volume needed to provide energy and nutrition for infants with PKU (Acosta, 1983). However, in 1988, the original guidelines were revised with recommendations for weight checks before and after breastfeeding, to ensure correct dietary intake of breast milk (Yannicelli, Ernest, Neifert, & McCabe, 1988). In 1994, Greve and colleagues (1994) devised a dietary management plan that eliminated test weighing while breastfeeding and involved supplementation with Phe-free formula. Thus, volume-regulated breast milk could be used to control the Phe content in the diet of infants with PKU.

Motzfeldt and colleagues (1999) introduced a dietary management plan for infants with PKU in which there were no restrictions on the number of breastfeedings per day. Instead, each feeding started with a prescribed amount of Phe-free formula and finished with feeding at the breast. In 2003, a new approach for breastfeeding infants with PKU was proposed in which the numbers of breastfeedings were adapted to the plasma Phe concentrations and, whether formula feeding with Phe-free formula or breastfeeding, the infant was allowed to

drink until satiety (van Rijn et al., 2003). This approach was considered more convenient for parents and encouraged the infant to empty the breasts, thereby consuming not only foremilk but hindmilk as well, which is higher in calorie-rich fats.

In 1999, Portnoi and colleagues retrospectively collected data on the practice of feeding infants with PKU in the United Kingdom and Australia. The results revealed that feeding practices differed not only between the countries but between practitioners within the same pediatric metabolic centers for infants with PKU (Portnoi et al., 1999). A similar survey was conducted to identify the feeding practices of healthcare providers in various metabolic centers in Europe, the U.S., South America, Australia, and New Zealand (MacDonald et al., 2006). No universal approach to feeding infants with PKU was identified but there was consensus with the recommendation of Phe-free formula in combination with breastfeeding (MacDonald et al., 2006). Notably absent was any research describing the practice of breastfeeding infants with PKU from mothers' perspectives that would provide further insight.

Incidence and Duration of Breastfeeding Infants with PKU

Despite evidence that breast milk is compatible with effective dietary management of PKU and appears to enhance cognitive functioning of school-age children with PKU, the majority of research studies on breastfeeding infants with PKU have addressed only incidence and duration (Agostoni, Verduci, Fiori, Riva, & Giovannini, 2000; Banta-Wright, Shelton, Lowe, Knafl, & Houck, 2012; Cornejo et al., 2003; Demirkol et al., 2001; Greve et al., 1994; Huner & Demirkol, 1996;

Kanufre et al., 2007; MacDonald et al., 2006; Motzfeldt et al., 1999; Portnoi et al., 1999; Segev, Abraham, Anikster, & Schwartz, 2004; van Rijn et al., 2003). These studies consistently revealed that few mothers of infants with PKU persist in breastfeeding beyond an initial period. For example, in an Italian study, breastfeeding rates were significantly lower in infants with PKU than in the general Italian population (Agostoni et al., 2000). Most recently, 19 of 35 Brazilian mothers were breastfeeding their infants with PKU at 6 months and only 11 continued to breast-feed at 12 months (Kanufre et al., 2007).

In contrast, the incidence of breastfeeding infants with PKU from a pediatric metabolic clinic in the U.S. over a 25-year period was 77% at the time of diagnosis. This incidence consistently exceeded the *Healthy People 2000* and *2010* objectives for initiation of breastfeeding (Banta-Wright et al., 2012). This level of breastfeeding additionally exceeded the national and individual state breastfeeding initiation rates from 1980 to 2005. Much remains to be understood in order to replicate such rates in other countries.

The duration of breastfeeding has also been found to be shorter for infants diagnosed with PKU. In an Italian study, the median duration of breastfeeding was one month (Agostoni et al., 2000). An Israeli study on breastfed infants with PKU ($n = 39$) revealed a longer mean duration of 4 months (Segev et al., 2004) as did a Dutch study at 2.5 months (van Rijn et al., 2003). In Norway, breastfeeding is the norm with 92% of mothers exclusively breastfeeding during the first three months and 40% continuing to breastfeed their infants to 15 months (World Health Organization, 2007). Nonetheless, breastfed infants with

PKU ($n = 74$) had a mean duration of seven months (Motzfeldt et al., 1999). In the only study from the U.S., the mean duration of breastfeeding infants with PKU ($n = 75$) from 1980 to 2005 was comparable to that of Norway at 7.1 months. In this same study, the mean duration of breastfeeding infants during 1980 to 1989 was nine months. Subsequently from 1990 to 2005, the mean duration was less than six months. Longer durations have been found in Turkey where two studies had higher mean durations of breastfeeding by mothers of infants with PKU at 9.6 months ($n = 40$; Demirkol et al., 2001) and 10.8 months ($n = 13$; Huner & Demirkol, 1996).

These reports of dietary management, incidence, and duration of breastfeeding reflect the low success of breastfeeding among mothers of infants with PKU, and they provide limited insight into factors that contribute to the success or failure of breastfeeding these infants. Reasons for stopping breastfeeding are not known. Perceptions of the rewards and challenges of breastfeeding infants with PKU were not addressed in any of the studies reviewed. This lack of knowledge about how mothers who were breastfeeding infants with PKU simultaneously managed breastfeeding and the disorder of PKU prevents development of appropriate guidelines, interventions, and support.

In addition, some clinics encourage mothers to breastfeed whereas others present breastfeeding as an unacceptable option. One of the key resources used for PKU management is the *Nutrition Support Protocols* (Acosta, 2009). In the current edition, the protocols presented standard commercial infant formula rather than breast milk as the basis for oral nutrition in the sample feeding plan

for infants with PKU. This implies breastfeeding may put the infant at risk for poor developmental outcomes. On the other hand, in the *2000 National Institute of Health Consensus Statement on Phenylketonuria: Screening and Management*, breastfeeding was encouraged in conjunction with supplementation with Phe-free formula ("National Institutes of Health Consensus Development Conference Statement on Phenylketonuria: Screening and Management," 2000).

While Riva and colleagues (1996) clearly found benefits for children with PKU who were breastfed as infants, the low incidence and duration of breastfeeding success among mothers of infants with PKU clearly reflects the paucity of research on this unique topic, and little is understood about the breastfeeding needs of mothers with infants who have PKU. Given the lack of research investigating mothers' experiences breastfeeding infants with PKU, there is an absence of research-based interventions to facilitate breastfeeding success among mothers of these infants. Research-based interventions are needed to facilitate the strict dietary control and management required by this chronic condition. Hence, more research is needed regarding factors that influence breastfeeding decisions as well as to decrease disparities in breastfeeding duration among mothers breastfeeding infants with PKU.

Family Context of Breastfeeding Infants with PKU

Family has many definitions that have evolved over the decades. In the early 1980s, literature conceptualizing family nursing emerged (Feetham, 1984; Gilliss, 1983; Gilliss & Knafl, 1999; Whall & Loveland-Cherry, 1993). Gilliss (1983) examined the nature of family as a phenomenon, reviewed and critiqued

approaches that had been used to study family, identified various strategies that would be appropriate in nursing research, and concluded that family research was flawed. Feetham (1984) published her landmark paper in which she determined that a single definition of *family* was not essential; rather, it was essential that *family* within the context of the research be defined. In a review of family-focused research from 1984 to 1991, Whall and Loveland-Cherry (1993) revealed that family as the unit of analysis was not defined consistently, if at all. Six years later, Gilliss and Knafl (1999) were unable to identify in a review of the literature on family nursing the family response to chronic illness. However, in 2002, Knafl and Gilliss were able to identify two distinct avenues of knowledge: 1) describing family response to illness and 2) explaining family response to chronic illness, both of which focused on how the family responded to a member's chronic illness rather than on individual family responses. In addition, they identified evidence suggesting that stress from the daily life experiences of chronic conditions were strongly associated with impaired family functioning. The stress was related to transitions 1) from the state of child health to one of chronic health, 2) between care teams or settings, and 3) in routines of daily family life. Each of these transitions led many families to develop a sense of fear about the futures of their children and their families.

During this time, there was increasing recognition of the impact of chronic conditions on families. It became clear that families with children who had chronic conditions lacked information about the diagnosis, technical care, developmental implications, communication needs, sibling risks, support groups, respite

resources, and future outlook (Branstetter, Domian, Williams, Graff, & Piamjariyakul, 2008; Coyne, 1997; Perrin, Lewkowicz, & Young, 2000; Wertlieb & American Academy of Pediatrics Task Force on the, 2003). In addition, families verbalized that the construction of daily life could be a barrier to family functioning (Branstetter et al., 2008) while at the same time, families identified a number of facilitators of functioning for both themselves and their children with chronic conditions. Knafl and Gilliss (2002) reported that decreased family stresses were linked to increased family functioning, increased family cohesiveness, and decreased family conflicts, which were associated with increased child adjustment. When families had positive communication strategies, there was increased family functioning. Some of these strategies included family problem-solving meetings to resolve stressful situations and identifying available everyday opportunities for positive communications whenever they arose (Branstetter et al., 2008). Thus, some families seemed to engage their stressors, barriers, and facilitators with the process of adaptation, which was conceptually different from coping. Adaptation is a process rather than an outcome (Clawson, 1996). In so doing, adaptation was associated with achieving a sense of normalcy in everyday life through resilience (Branstetter et al., 2008; Clawson, 1996; Coyne, 1997; Knafl, Darney, Gallo, & Angst, 2010; Knafl & Deatrick, 2002; Knafl & Gilliss, 2002). When normalcy was present for these families, there was increased parenting self-esteem, transferable coping beliefs, and skills that could be applied in other everyday life activities, such as employment, resulting in more fulfilling and enjoyable family life (Knafl et al.,

2010). However, while some families adapted with integration of the medical treatment regimen, other families floundered into disruptions with poor adherence to recommended medical treatment therapies (Duke et al., 2002; Grey, Boland, Yu, Sullivan-Bolyai, & Tamborlane, 1998; Hanson, De Guire, Schinkel, Henggeler, & Burghen, 1992; Herzer et al., 2010). As a result, family functioning was identified as an important trait in families of children with chronic health conditions.

Family Functioning

Family functioning encompasses the different ways that family members interact with, respond to, and relate to each other in the pursuit of goals, activities the family engages in together, and acceptance of family routines (Preechawong et al., 2007). When a family is functioning well, family members are cohesive in their actions, are able to adapt to stressors, and have clear family rules and boundaries (Petrocelli, Calhoun, & Glaser, 2003). Changes within a family may occur and affect family functioning. For example, when a child has a chronic health condition (e.g., cystic fibrosis, inflammatory bowel disease, epilepsy), some studies found significantly worse family functioning (Engstrom, 1999; Janicke, Mitchell, & Stark, 2005; Rodenburg, Meijer, Dekovic, & Aldenkamp, 2005) whereas others identified no differences (e.g., cystic fibrosis, sickle cell disease, inflammatory bowel disease, epilepsy) (Ievers & Drotar, 1996; Mackner & Crandall, 2006; Noll, Swiecki, Garstein, & Vannatta, 1994; Rodenburg et al., 2005) or even better functioning (e.g., sickle cell disease) (Midence, McManus, Fuggle, & Davies, 1996). In general, family functioning across chronic pediatric

conditions varies greatly. In addition, only a few studies have addressed family functioning in the context of PKU.

Family functioning in families with PKU. Although studies have examined family functioning in families where a child has PKU (Kazak, Reber, & Carter, 1988; Kazak, Reber, & Snitzer, 1988; Keleske, Solomons, & Opiz, 1967; Lord, Wastell, & Ungerer, 2005; Read, 2003; Reber, Kazak, & Himmelberg, 1987; Shulman, Fisch, Zempel, Gadish, & Chang, 1991; Wood, Friedman, & Steisel, 1967), these studies have mainly focused on two aspects: 1) treatment adherence and metabolic control, and 2) psychosocial function. Two early studies, both from 1967, found that parental worry and restrictions inhibited the play of children with PKU (Keleske et al., 1967; Wood et al., 1967). However, many of the children in the samples were already severely cognitive impaired. The cognitive impairment, rather than the medical condition of PKU, may have affected the parent-child play interaction.

In another early study involving children with PKU under 8 years of age and their families, Reber and colleagues (1987) evaluated four areas of family functioning (parents' psychological distress, marital satisfaction, parenting stress, and family interactions (cohesion and adaptability) and found family functioning scores did not correlate with the child's Phe control or the child's cognitive performance. Rather, there was an association between cohesion and adaptability as perceived by the mothers and the children's better cognitive test performance and behavior. In a subsequent study with families who had children with PKU under 6 years of age, Kazak, Reber, and Snitzer (1988b) also found no

correlation between the children's Phe levels and the parents' psychological distress, marital satisfaction, parenting stress, or family interactions (cohesion and adaptability). However, the mothers of children with PKU perceived their families to be significantly less cohesive and less adaptable than mothers in the control group. Although Shulman and colleagues (1991) found parents of children with PKU were similar to parents with healthy children in their perception of family functioning and coping, mothers of children with PKU also perceived their families to be less cohesive and adaptable, thus confirming the results by Kazak et al. (1988b).

Kazak, Reber, and Snitzer (1988b, p.229) suggested that parents of children with PKU may have "different" rather than "deviant" patterns of family regulation and functioning. For example in another study, mothers of children with PKU were found to be significantly less likely to name their spouses, parents, and extended family members as part of their network than were mothers of healthy children (Kazak, Reber, & Carter, 1988). Rather than abnormal networks, this may suggest different support needs implicating perhaps professionals as key supporters. In 2003, Read reported that mothers of children with PKU had less worry about their child, very little difficulty meeting their child's needs, and significantly less impact of the disease on all aspects of their personal lives than mothers of children with mitochondrial disorders. This difference could be explained by the known trajectory of PKU and the many resources for families with children who have PKU, such as PKU camps, mothers

support programs, regional parent meetings, and a dedicated network of healthcare providers.

In a study by Knafl and colleagues (2007) involving families with a child who has a genetic condition including PKU, five patterns of family functioning (well-adapted, discrepant, more in satisfaction, same or more in hardness, and compromised) were identified by cluster analysis. In contrast, Lord and colleagues (2005) found that mothers of children aged 3 months to 12 years with PKU had higher levels of trauma reactions associated with younger children, a perception of their partner as less caring, and smaller support networks. Again, differences may necessarily exist and mothers' reactions seem to be generic to caring for a child with PKU. As scores were lower than those reported by parents of children surviving cancer, parental trauma may be common across a range of childhood conditions.

Research related to family functioning and breastfeeding. Only one study has evaluated the impact of breastfeeding on family functioning (Butt, 2001). In a secondary analysis, Butt (2001) found that there was a positive effect of breastfeeding on immediate family functioning. Fathers whose wives were currently breastfeeding had increased marital satisfaction and reported enhanced family functioning. The relationship between breastfeeding and family functioning was mediated by marital satisfaction and moderated by social support for fathers. Further research is needed to elucidate the impact of family functioning in the context of breastfeeding especially in regards to predictive factors of marital satisfaction and family functioning in breastfeeding mothers.

Families with Children Who Have Chronic Conditions

In the U.S., approximately 29% of children have chronic conditions that require health and related services beyond those needed by healthy children (Read, 2003; Williams, 1997). The majority of these children live at home and their condition often has implications for parenting and the family's everyday life (Hollidge, 2001; James et al., 2002; Sullivan-Bolyai, Sadler, Knafl, & Gilliss, 2003; Williams et al., 2002, 2003). Normal family functions and developmental tasks are impacted and significant adjustments must be made by families to accommodate chronic conditions (Deatrick & Knafl, 1990b; Knafl & Deatrick, 1986). Families of infants to adolescents to young adults with chronic conditions are expected to continue normal family functions, learn how to manage the chronic condition, and adapt to a state of constant change and uncertainty while incorporating additional caregiving tasks that are associated with chronic conditions (Canam, 1993; Clawson, 1996). In addition, each chronic condition has a distinct biological foundation with specific and sometimes demanding treatment management.

Despite the biological uniqueness of each condition, research across several childhood chronic conditions has revealed that the demands on families are similar regardless of the diagnosis (Barlow & Ellard, 2006; Berge & Patterson, 2004; Knafl & Gilliss, 2002; Wallander, Thompson, & Alriksson-Schmidt, 2003; Wallander & Varni, 1998). In addition, qualitative studies of families with children who have chronic conditions have found that families often come to view their child and their lives as normal (Deatrick et al., 1999; Deatrick,

Knafl, & Walsh, 1988; Knafl et al., 2010; Knafl & Deatrick, 1986; Knafl & Deatrick, 2002; Krulik, 1980; McDougal, 2002; Morse, Wilson, & Penrod, 2000; Rehm & Bradley, 2005; Robinson, 1993). This phenomenon has been labeled *normalization*.

The first study to identify normalization was done with families with a child who had polio, and two major styles of response were identified: normalization and disassociation (Davis, 1963). Two other early studies (Benoliel, 1970; Darling, 1979) emphasized parents' management behaviors and identified different typologies based on the behaviors. Twelve years later in the nursing literature, a description of normalization behaviors of parents of chronically ill children revealed that mothers used tactics to minimize their children's feelings of being different (Kruklik, 1980). A later study confirmed these results in families with children with osteogenesis imperfecta with the findings that strengthening the child's coping resources and making the environment more accommodating to the needs of the child were tactics used to minimize the child's differences (Deatrick et al., 1988). Another study revealed that some families with children with chronic conditions used normalization inconsistently, resulting in a mixed message of "you are normal" and "you are different" (Anderson, 1981). Subsequently, the concept of normalization was determined to be culturally based with Caucasian families having the goal of normalization whereas the goal of Chinese families was to keep the chronically ill child content and happy (Anderson & Chung, 1982).

Knafl and Deatrick (1986) traced the origin and development of the concept of normalization as one of the patterns that families use in response to childhood illness, and they identified four attributes: 1) acknowledgment of the impairment in the child, 2) definition of the family life as essentially normal, 3) perceptions that the social consequences as a result of the illness were minimal, and 4) engagement in behaviors that communicated the normalcy of the child and family to others. Using three case studies, normalization was differentiated from denial and disassociative coping behaviors (Knafl & Deatrick, 1986). Ten years later, the concept of normalization was reviewed, and the original attributes were refined: 1) acknowledged condition and its potential to threaten lifestyle, 2) adopted “normalcy lens” for defining child and family, 3) engaged in parenting behaviors and family routines that are consistent with “normalcy lens,” 4) developed a treatment regimen that is consistent with “normalcy lens,” and 5) interacted with others based on view of child and family as normal (Deatrick et al., 1999). Normalization was further manifested in the meaning that parents attributed to family life with children who had chronic conditions: normalization was present or there was an absence of normalization in family life (Knafl et al., 2010). Thus, families who normalize childhood chronic conditions recognized the seriousness of the condition while at the same time they continued to adopt a flexible management approach to carrying out the needs of the treatment regimen. As a result, within the family of children with chronic conditions, there was incorporation of the chronic condition into the usual family management of routine.

Family Management Style Framework

In the original study by Knafl and Deatrick (1990), the conceptualization of *family management style* (FMS) emerged from qualitative research. Family management came to be understood through the stories that families told about perceptions and meanings. This work led to the identification and definition of three interactive components of FMS: 1) definition of the situation, 2) family management behaviors, and 3) sociocultural context. Definition of the situation was each family member's subjective identification and interpretation of the situation. *Family management behaviors* were defined as the behavioral accommodations made in response to the special needs of the child with the chronic condition that each family member used to manage on a day-to-day basis and were based on each family member's perception of the situation (Deatrick & Knafl, 1990a). The sociocultural component of the model included the cultural, ethnic, and religious influences, values and beliefs, as well as the different social, political, and economic forces that shaped each family member's perception. Within the model, arrows reflect the interactive relationship among

the components (Figure 2).

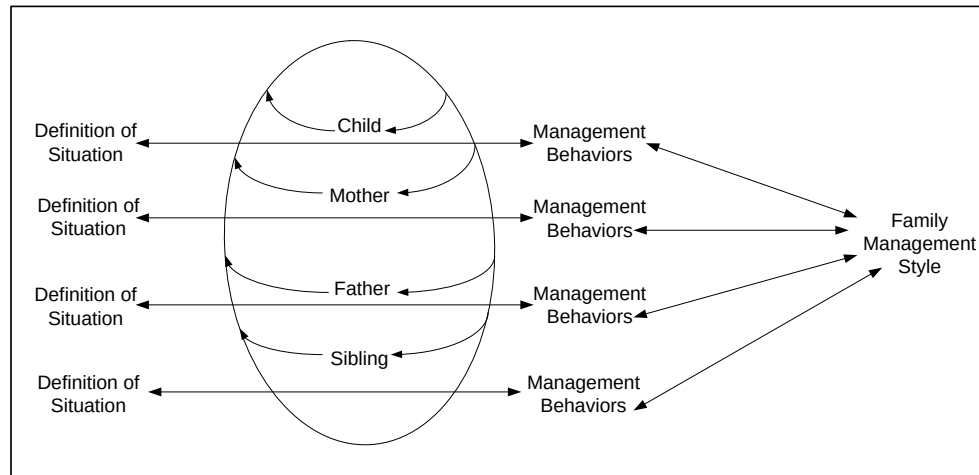


Figure 2. Original Model of the Family Management Style Framework.
Source: Knafl & Deatrick, 1990a.

Further refinement of the family management style framework (FMSF) resulted in modification of the original framework and changes to the three major components of the framework (Knafl & Deatrick, 2003). While definition of the situation and management behaviors continued to be included as major components of the framework, sociocultural context was replaced with perceived consequences (see Figure 3).

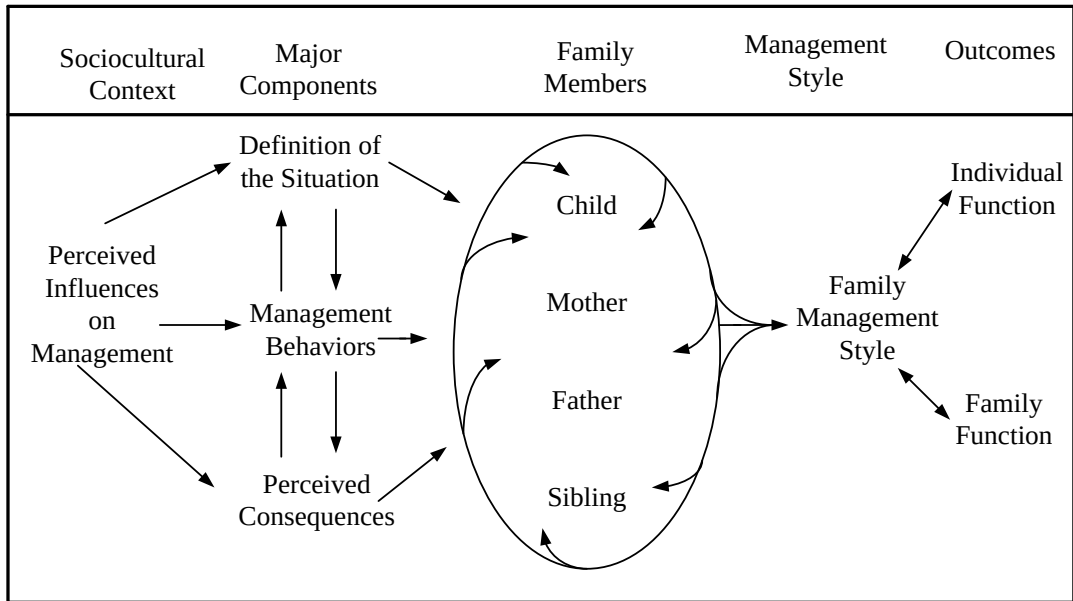


Figure 3. Second Revision, Family Management Style Framework.

Source: Knafl & Deatrick, 2003.

Definition of the situation is how the family views the child and the chronic condition. The dimensions within this component include child identity, illness view, management mindset, and parent mutuality. Within management behaviors, there are two dimensions, parenting philosophy and management approach, linked to caring for a child with a chronic condition. The final component is perceived consequences, which are defined as actual or expected perceived outcomes that shape management behaviors and affect the subsequent definition of the situation. Within perceived consequences, there are two dimensions: family focus and future expectations. Later, Knafl and colleagues (2007) articulated the interplay of concepts, data, and methods in the development of the FMSF, which represents family response to a child's chronic condition.

The FMSF explains differing patterns in which families define and manage illness-related demands and the resulting consequences for family life. It is a family typology that systematically considers patterns of family management. This includes issues common to all families within a particular situation and the ways in which the issues are typically manifested within families. The FMSF can be used either broadly or narrowly. Either way, the framework directs healthcare providers and researchers to consider how the family, both as individuals and as a unit, actively define, manage, and perceive consequences that shape management behaviors and affect the subsequent definition of the situation.

Research related to family management style framework. Several studies have focused on families who have children with chronic conditions and used FMSF as the conceptual framework (see Table 1). Using interpretative methods, Kodadek and Haylor (1990) revealed that parental perceptions of caring for a blind child resulted in four family styles: 1) the realistically accepting family, 2) the devoted parent family, 3) the perfect blind child family, and 4) the overwhelmed family. In another study, family responses to school-aged children with chronic conditions revealed five styles: 1) thriving, 2) accommodating, 3) enduring, 4) struggling, and 5) floundering (Knafl, Breitmayer, Gallo, & Zoeller, 1996). Using grounded theory, Kendall and Shelton (2003) described family responses to having a child with attention-deficit/hyperactivity disorder (ADHD) as being of four styles: 1) the chaotic family, 2) the ADHD controlled family, 3) the surviving family, and 4) the reinvested family. In addition, in ADHD controlled families, surviving families and reinvested families could be viewed as a

trajectory. As a trajectory, families progressed over time from the ADHD controlled families to the reinvested families. Chaotic families were viewed separately as over time they remained chaotic.

Table 1

Selected Research Using FMSF as a Conceptual Framework in Families with Children with Chronic Conditions

Study	Population
Kodadek & Haylor, 1990	Families of children with blindness
Knafl, Breitmayer, Gallo & Zoeller, 1996	Families of children with a variety of chronic conditions, such as diabetes, rheumatoid arthritis, renal disease, and asthma
Kendall & Shelton, 2003	Families of children with ADHD

Additional studies have used the FMSF to understand the family response to an infant/child in different care situations (see Table 2). Using grounded theory, Murphy (1990) explored interactional styles of parents following the birth of a high-risk infant, which resulted in the identification of three management styles: 1) agreeing on socially prescribed or gender-specific patterns of behavior, 2) adopting parallel styles, and 3) negotiating a mutually interdependent style. Edwards-Beckett and Cedargren (1995) described the sociocultural context and its level of supportiveness for families with a child with myelomeningoceles, and three dimensions emerged: 1) sharing information, 2) interpersonal interactions,

and 3) supports/hindrances. In another study, Gallo and Knafl (1998) revealed how parents respond and manage the challenges of childhood chronic illness:

1) strict adherence, 2) flexible adherence, and 3) selective adherence.

Naturalistic inquiry was used to describe the process that mothers raising toddlers with type 1 diabetes moved through to attain the necessary skills to care for their children (Sullivan-Bolyai, Knafl, Deatrick, & Grey, 2003). The mothers used two of the three management approaches and behaviors previously identified by Gallo and Knafl (1998): strict adherence and flexible adherence.

Table 2

Selected Research Using the FMSF as a Conceptual Framework for Families' Responses in Different Care Situations

Study	Population
Murphy, 1990	Families of critically ill infants
Edward-Beckett & Cedargren, 1995	Families of a child with myelomeningocele
Gallo & Knafl, 1998	Families of children with chronic conditions, such as diabetes, rheumatoid arthritis, and renal disease
Krouse, 2002	Families of low birth weight infants/premature
Bernaix, Schmidt, Jamerson, Seiter, & Smith, 2006	infants in the NICU who chose to breastfeed
Sullivan-Boylai, Deatrck, Knafl, & Grey, 2003	Mothers with toddlers who have type 1 diabetes
Van Riper, 2005	Ethical and social issues that emerged with genetic mutation testing in families with known genetic conditions
Wiegand, Deatrck, & Knafl, 2008	Families participating in the process of withdrawal of life-sustaining therapy from a family member due to an unexpected, life-threatening illness or injury

In another study that examined how families defined and managed the ethical and social issues that emerge with genetic mutation testing, the results revealed that one must take into account both the perspective of the individual and the family as a whole so that optimal outcomes occur for both (van Riper, 2005). Even in acute settings, similar family styles have been found. For example, in the adult intensive care unit, families that participated in the process of withdrawing life-sustaining therapy from a family member with an unexpected, life-threatening illness or injury used one of five management styles: progressing, accommodating, maintaining, struggling, and floundering (Wiegand, Detrick, & Knafl, 2008).

Recently, the FMSF has been used in two breastfeeding studies (Bernaix, Schmidt, Jamerson, Seiter, & Smith, 2006; Krouse, 2002). Using naturalist inquiry, Krouse (2002) described three management styles for families of low birth weight infants who chose to breastfeed: 1) facilitating, 2) maintaining, and 3) obstructing. During the study, some families changed styles, suggesting that management styles are fluid. Subsequently, Krouse's typology was confirmed for breastfeeding premature infants in the NICU (Bernaix et al., 2006).

Use of Family Management Style Framework in This Study

Hundreds of papers that have been published on the biochemical defect of PKU, the dietary and pharmaceutical therapies for PKU, and positive and negative psychological outcomes. However, these studies did not focus on the family as a social unit that influences and is influenced by the experience of breastfeeding an infant with PKU. Thus, in this study, the guiding framework was

the FMSF developed and refined by Knafl and colleagues to elicit mothers' construction of their experiences breastfeeding their infants with PKU (Knafl et al., 1996; Knafl & Deatrick, 1990, 1996, 2003).

Summary

PKU is a rare inborn error of metabolism disorder that affects every racial and ethnic group. There are many challenges for families with children who have PKU including life-long dietary restrictions, frequent Phe monitoring, and the added expense of dietary and pharmaceutical therapies. Although advances in understanding the defect and better treatment therapies have improved the overall outcome for PKU, these studies did not consider the perspective of the family. Little is known about mothers' experiences managing breastfeeding in the context of having an infant with PKU. Moreover, research is lacking on family functioning in families with breastfeeding infants, whether healthy or with PKU. Thus, this study drew on the experiences of mothers breastfeeding infants with PKU in response to different healthcare challenges and yielded information from which to create effective interventions. As such, the study was a cross-sectional design with two study phases. In the first phase, mothers of children with PKU were invited to participate in an Internet survey on their experiences of feeding their infants with PKU. The second phase of the study drew a subsample of mothers from phase 1 to further explore their experiences breastfeeding infants with PKU using qualitative interview and two quantitative self-report measures on family functioning. To accomplish the purpose, the following specific aims of this study were to:

1. Describe the incidence and duration of breastfeeding among mothers who have infants with PKU in the United States and Canada.
2. Describe mothers' experiences breastfeeding in the context of PKU including their perceptions of factors that facilitate and/or interfere with breastfeeding infants with PKU and strategies to obtain the necessary skills to care for their breastfeeding infants with PKU.
3. Identify mothers' experiences breastfeeding infants with PKU and examine mothers' assessment of family functioning.

CHAPTER 3

RESEARCH DESIGN AND METHODOLOGY

Broadly, three research questions framed this study. The first question concerned the prevalence and duration of breastfeeding by mothers of infants with PKU in the United States and Canada. The second question addressed factors that facilitate and/or interfere (stressors and barriers) with breastfeeding infants with PKU and strategies used by the mothers of these breastfeeding infants. The third question concerned themes of breastfeeding management in families with breastfeeding/breastfed infants with PKU and how they related to family functioning. To address these questions, this study was conducted in two phases (see Figure 4). For the first question in phase 1, a quantitative self-report Internet survey sought to identify the prevalence and duration of breastfeeding infants with PKU in the U.S. and Canada and to identify factors that facilitate and/or interfere with breastfeeding infants with PKU and strategies used by mothers of these breastfeeding infants. This survey provided a sample from which mothers who agreed to participate in Phase 2 were drawn. During Phase 2, descriptive qualitative interviews sought to elicit mothers' construction of their experiences breastfeeding their infants with PKU situated within the FMSF (Knafl et al., 1996; Knafl & Deatruck, 1990, 2003). To more fully comprehend the family functioning in the context of a family with an infant who has PKU, two quantitative measures supplemented the qualitative interviews. These allowed more specific descriptive information about the same phenomena in more than a single approach. For example, in the qualitative interview, participants were asked to

describe difficulties and how these impacted their management of breastfeeding and PKU. In contrast, the measures quantified mothers' perceived family functioning. The contribution of these factors in mothers' lives was used to expand and to complement the description described in qualitative interviews. In addition, the qualitative descriptive method allowed the exploration of overlapping and different facets of mothers' perceived family functioning in the experience of managing breastfeeding and PKU simultaneously. In this chapter, the sampling plan for each phase of the study is presented, a synopsis of the Internet surveys and telephone interviews is provided along with an overview of qualitative descriptive research, and the data collection and analysis procedures for each phase of the study are delineated.

Phases of the Study

This study was conducted in two phases (Figure 4). In phase 1, the investigator conducted the Internet survey of mothers with children who have PKU. Following analysis of the survey data, mothers were selected and recruited for qualitative interviews for phase 2.



Figure 4. Timeline for the Study.

Phase 1: Internet Survey

Sample

Data were collected from a sample of mothers with children who have PKU. In phase 1 of the study, a convenience sample was used to obtain data and trends regarding the prevalence and duration of breastfeeding infants with PKU in the U.S. and Canada. Children with PKU are born to women who represent a range of socioeconomic characteristics and types of health insurance coverage. In addition, across the U.S. and Canada, mothers of infants with PKU receive care from a variety of specialty clinics with varied feeding protocols and beliefs about the appropriateness of breastfeeding infants with PKU. As expected, the study sample represented a broad range of characteristics; variables such as education, annual household income, and marital status were examined in relationship to U.S. Census data (see Chapter 4, Results).

Approximately 64% of U.S. households use high-speed Internet (U.S. Department of Commerce, 2010). Many others use Internet services available at public libraries, at work, and at the homes of friends and family. Individuals between 24 to 55 years use high-speed Internet (68%) more than those 55 years and older (46%), but less than the 81% of those individuals who are 18 to 24 years old (U.S. Department of Commerce, 2010). In this study, mothers who participated had Internet access. There may be some mothers with children who have PKU and do not have any Internet access, and unfortunately, these mothers were excluded from participating in the Internet survey. Thus, the

sample may not be representative of the population and may reflect a sampling bias toward those mothers who have high-speed Internet.

Inclusion criteria. The sample inclusion criteria for mothers were 1) at least 21, 2) able to read and write in English, 3) have child/children with PKU, and 4) live in the U.S. or Canada. Only mothers were surveyed since they are the most involved in managing the feedings of infants and are the family member who typically assumes primary responsibility for infant care. PKU is most common in people of Northern European heritage and is rare among French Canadians (1:25,000) (Dhondt, Farriaux, Briard, Boschetti, & Frezal, 1993) and Latinos from Mexico (1:70,082) (Velazquez et al., 1994) so that translated measures were not warranted.

Mothers were 21 years of age or older to participate since there is considerable variation across states and provinces regarding laws governing age of adulthood and minor emancipation. In order to have the same inclusion criteria and consent processes for all subjects, a minimum age of 21 years was selected. Further, the developmental stage and resource needs of adolescence and young adulthood, between 14 and 21 years of age (Statistics Canada, 2008), were not accounted for in the study.

Mothers who met the inclusion criteria were invited to participate in the study regardless of their ethnic, social, racial, educational, financial, or religious characteristics. Thus, the sample reflected a diverse group of mothers who have child/children with PKU. The decision to collect data via an Internet survey allowed recruitment of mothers from various geographic locations across the

U.S. and Canada, as increasingly more and more people have access to the Internet (Dillman, 2007).

Recruitment to the Internet Survey

Although this research was based at Oregon Health & Science University (OHSU) in Portland, Oregon, mothers were recruited from across the U.S. and Canada so as to have a representative sample. Recruitment strategies were developed in collaboration with the leadership of several PKU family support groups: PKU Listserv (Kohen, 2007), *National PKU News* (Schuett, 2007), and Canadian PKU and Allied Disorders Association (Adams, 2009). In their letters of support, these leaders recommended recruiting participants through the PKU Listserv, which reaches over 1400 families in the U.S. and Canada. In addition, the Canadian PKU and Allied Disorders Association volunteered and placed a notice on their webpage regarding a description of the study and recruitment information. Several state and regional PKU advocacy groups, as well as the *National PKU News*, included a description of the study and recruitment information in their newsletters, web sites, emails, twitter, and Facebook pages (see Table 3).

After receiving approval from the university's institutional review board, the initial post on the PKU Listserv included a brief description of the purpose of the study and the inclusion criteria, with the instruction that interested participants should reply directly to the researcher (see Appendix A). After being contacted by the mother, the researcher forwarded the research information sheet and

Table 3

PKU Advocacy Organizations and Methods Used to Help Announce the Study

PKU Advocacy Groups							
	Facebook	Flyer	Email	Listserv	Newsletter	Twitter	Webpage
Canadian PKU & Allied Disorders					X		X
Florida PKU Foundation	X		X				X
Georgia PKU Connect	X		X				
Intermountain PKU & Allied Disorders							X
Michigan PKU & Allied Disorders							X
National PKU Alliance			X				
National PKU News					X		
National Society for PKU	X						
New England Connection for PKU & Allied Disorders							X
PKU & Allied Disorders of Wisconsin		X					
PKU Listserv				X			
PKU Northwest Alliance			X				
Tennessee PKU Foundation						X	X
WyoMed Campaign			X				

Instructions for the survey via email to the participant (see Appendices B and C).

Participants were informed that completion of the survey was considered evidence of their agreement to participate in the survey research. The consent assured participants that email addresses would not be used for any other purpose unless the participant agreed to be contacted in the future for other PKU research studies.

Participants who completed the survey were sent a \$10 US electronic gift certificate to a medical foods company, Cambrooke Foods, which provides low protein items popular with PKU families. The researcher sent requests via email for electronic gift certificates directly to a designated person at Cambrooke Foods and paid for them using a credit card. The company sent the electronic gift certificate to the email provided by the participant, using “survey participant” as the name to protect privacy. The results of the survey were summarized and posted on the PKU Listserv and sent to representatives of the different PKU advocacy groups at the completion of the project.

Data Collection Procedures

An Internet survey was used to obtain data from the participants to address the first and second aims of the study. Previous studies had demonstrated that there are numerous advantages to using a web-based survey including 1) recruitment of a geographically diverse sample; 2) low cost; 3) extremely fast solicitation of responses for online survey (only a few minutes to send thousands of solicitation emails to a listserv); 4) faster response time and completion return;

5) anonymity of participants, which can result in more honest answers to sensitive topics; 6) survey software that simplifies compilation and analysis of data; and 7) the ability to prompt for skipped questions to be answered (Ahern, 2005; Dillman, 2007; Gordon & McNew, 2008; Jones, Murphy, Edwards, & James, 2008; Read, 2004). By using the Internet, more participants throughout the U.S. and Canada were enrolled in the study.

The survey questions were constructed using guidelines developed by Dillman (2007). The goal was to develop questions that every possible participant would understand and interpret in the same way, and would be able to respond to accurately. In addition, the survey was designed to motivate participants to respond by making the features of the survey appear interesting and important, and making the survey easy to complete (Dillman, 2007). The Internet survey was also designed so that participants were guided through the survey to ensure full and accurate completion before they were able to submit it. The survey consisted of demographic questions, questions about how their infant with PKU was fed during the first year, and one question regarding willingness to be contacted to be interviewed about feeding infants with PKU. The answers were buttons, drop down boxes, and short fill-in-the-blanks. Three advisors knowledgeable about web-based surveys and six mothers who have children with PKU piloted and evaluated the survey design in order to refine and improve the approach and wording of the questions. It was anticipated that less than 20 minutes would be required to complete the survey, and the actual length of time to complete the survey ranged from 7 to 69 minutes with a mean of 19 minutes ($SD = 10$). Three mothers were

identified as outliers with a length of completion time greater than 70 minutes, which suggests that these participants may have started the survey, left the computer with the survey window open, and then returned to the survey later to complete. Consequently, these mothers were not included in the analysis of length of time to complete the survey.

Procedure for Analysis of Survey Data

The Internet survey enabled the investigator to 1) view summarized results, 2) view individual results, and 3) create reports, graphs, and charts that compared the results for groups of participants. Additionally, the Internet survey allowed the data to be directly downloaded into an SPSS file to view all the data and comments, and permitted further analysis of the data.

Prior to analysis, the data were cleaned to ensure accurate and valid analyses. Data from SPSS were compared and all discrepancies were resolved by checking the hardcopy of the each participant's data.

The analysis of the survey data focused first on descriptive statistics, including frequencies for each variable, item, and scale were examined prior to analyses. Frequency charts were analyzed to ensure that all data values fell into the possible values for each variable and to identify outliers. Frequency charts were also used to ensure that individual coding for missing values matched the number of total missing values. The descriptive values were examined to ensure that ranges fell within the expected range, and that means and standard deviations were such that the standard deviation did not exceed the mean. Missing data were

infrequent. Data for 103 participants were analyzed because there were missing data for less than 5% of all items (complete data for more than 95%).

Analyzing mothers' responses in the survey data. The analysis of the mothers' responses focused on describing the narrative responses to four open-ended questions regarding challenges, ease, help, and support while breastfeeding infants with PKU. Descriptive categories were identified from the narratives and defined. Categories were counted for presence versus absence (1/0) as dichotomous variables to determine frequency counts and explored linkages across categories. The description of the categories and frequency counts resulted in clusters of categories leading to an identification of major categories in the response data as well as identification of how the categories varied across responses. The final step in this stage of the analytic process was identification of facilitators, barriers, stressors, and strategies identified by the mothers who were breastfeeding infants with PKU.

Phase 2: Qualitative Interviews

Sample for Qualitative Interviews

Ten mothers of breastfeeding infants with PKU were selected from the participants in the larger Internet survey. The goal was to capture a broad spectrum of breastfeeding experiences of mothers who were currently breastfeeding or had recently breastfed their infants with PKU. The extremes of the population were included with the aim of sampling for heterogeneity from the common to unique manifestations across a broad range of demographic variety. The principle was to interview a very different selection of people so that if a group

of people is extreme in several different ways, it will contain people who are average in other ways. This type of sampling is useful in situations when researchers want to understand how a phenomenon is seen and understood among different people in different settings.

The ultimate goal was maximum variation sampling to obtain information-rich cases. The following steps were followed. First, the maximum sample size was 10 mothers selected on the basis of maximum variation. The variations for the sample population were characteristics that potentially differ widely between people in the study population including education level, annual household income, geographic location, population density, and duration of breastfeeding.

Inclusion criteria. Only mothers were interviewed since they are most involved in managing breastfeeding and are the family member who typically assumes primary responsibility for infant care. However, the investigator fully recognizes that fathers play an important role in raising children who have chronic conditions (Azar & Solomon, 2001; Baumann & Braddick, 1999; Heaman, 1995; Katz & Krulik, 1999; May, 1996; Wiener, Vasquez, & Battles, 2001). As with phase 1, mothers had to be 21 years of age or older and be English-speaking to participate in this second phase. Mothers had to have initiated breastfeeding their infant with PKU. In addition, the infant with PKU needed to be younger than 36 months. This was to ensure that mothers' experience of breastfeeding and management of PKU had been recent and not several years or decades before. Finally, the infant with PKU had to not have had other significant health problems,

such as congenital heart disease, renal disease, prematurity, gastroschisis, or cleft lip and palate.

Recruitment for Qualitative Interviews

After analysis of the phase 1 results, mothers who self-identified the willingness to be interviewed in phase 2 served as the pool from which participants were selected. Those mothers were identified and contacted by email to inquire if they remained willing to participate in the second part of the study (see Appendix D). If willing to participate, mothers contacted the investigator via an email address. After being contacted by the mother, the investigator arranged a convenient time and date for a telephone call to verify the mother's eligibility and to explain the purpose of the study and study procedures (see Appendix E). Research consent forms were read to participants over the telephone (see Appendix F) and verbal oral consent was obtained.

After verbal consent, the investigator also verified demographic information from the participants from the Internet survey for descriptive purposes. To minimize the likelihood of interruption, the importance of having a private setting was emphasized (McCormick, Workman-Daniels, Brooks-Gunn, & Peckham, 1993; Musselwhite, Cuff, McGregor, & King, 2007; Wilson, Roe, & Wright, 1998).

Data Collection Interview Procedures

Telephone interviews were used to obtain data from the participants. Previous studies have demonstrated no significant difference between face-to-face interviewing and telephone interviewing in terms of the quantity or quality of data (Barriball, Christian, While, & Bergen, 1996; Carr & Worth, 2001; Davis Kirsch &

Brandt, 2002; Evans, Kessler, Lewis, Peters, & Sharp, 2004; Musselwhite et al., 2007; Siemiatycki, 1979; Smith, 2005; Sturges & Hanrahan, 2004; Wilson, Roe, et al., 1998). In addition, there are some notable advantages to telephone interviews including an increased response rate compared to face-to-face or mail surveys, access to hard-to-reach participants due to distance or weather, more interviews achievable per day, moderate costs, and minimal missing data (Sturges & Hanrahan, 2004). By using telephone interviews, more diverse participants were enrolled in the study.

At the beginning of the scheduled telephone interview, the study purpose and procedures were explained to the participants. An opportunity was provided to ask questions about the study and what participation entailed. With the participant's permission, the investigator audio-recorded all interviews using a telephone recording device that connected to a hard-wired telephone and then to a digital recorder.

Data collection was completed using an interview guide (see Appendix E). The qualitative interviews were conducted using guidelines developed by Patton (2002). Data collection was completed using an interview guide. If a participant addressed a question on the interview guide before that question was asked, the question was asked again in sequence, acknowledging what had already been discussed and clarifying if there was anything else that the participant wanted to add. This approach communicated to the participants that the investigator was paying attention to the answers and provided an opportunity for further elaboration of the response. The investigator followed up on any important topics that the

participant raised but that were not included on the interview guide, and she probed for further clarification and detail as needed. At the completion of the interviews, mothers were reminded to complete two family assessments: the Family APGAR and the General Functioning Scale of the Family Assessment Device Questionnaires and to email their responses to the researcher. After receiving the two completed assessments, mothers were mailed their choice of a \$20 Kroger, Starbucks, or Target gift card to thank them for their participation.

Qualitative Interview

Qualitative description is a distinct method of naturalistic inquiry that uses everyday language to describe a participant's perception and experience of a phenomenon (Sandelowski, 2010). The goal of qualitative description is descriptive and interpretive validity, which means to present a precise account of the experiences, events, and processes that the researchers and participants agree is accurate (Sandelowski, 2010).

The design features for qualitative description are summarized in Table 4. These features are not exclusive or exhaustive as one of the features of qualitative description is its flexibility (Patton, 2002). Researchers conducting studies with qualitative description typically use a pre-existing theory or framework (Sandelowski, 2010). In this study, the FMSF was employed.

Table 4

Major Design Features of Qualitative Description (Adapted from Sandelowski, 2010)

Design	
Components	Design Specifics
Philosophy	Pragmatic, naturalistic approach Overtones of other qualitative approaches
Sample	Purposeful sampling Maximum variation sampling is especially appropriate
Data Collection	Slightly more structured, open-ended interviews with individuals or groups Researchers are concerned about the who, what, where and why of the experience or event Observation of specific occurrences Review of documents and other pertinent materials
Analysis	Qualitative content analysis using a modifiable coding system that corresponds to the data collected (e.g., “data near”)
Outcomes	Straight description of the data organized in a way that “fits” the data, such as chronologically by topic, by relevance, or by prevalence

Qualitative description uses the principles of naturalistic inquiry, which allows the researcher to gather information about the phenomenon through the definition and interpretation of the situation by the participants themselves (Patton, 2002). Context is important in naturalist inquiry as it gives meaning to the situation. Open-ended questions in conversation-like interviews (e.g., “Tell me about your experience breastfeeding your daughter?”) result in rich description of an experience or event. This is in contrast to focused questions (e.g., “How satisfied were you with breastfeeding your daughter?”) with specific one- or two-word response options (e.g., *very satisfied*, *somewhat satisfied*, *satisfied*, or *not at all satisfied*) that do not increase understanding of the experience and do not provide the participant’s insight and understanding.

Interview guide. The interview guide used in qualitative description is more structured than in other qualitative methods and is based on the expert knowledge of the researcher to focus on areas that are generally poorly understood. In this study, the interview guide was based on insights from the literature on managing breastfeeding in challenging situations (Barbas & Kelleher, 2004; Bernaix et al., 2006; Faridi & Dewan, 2008; Knafl & Deatrck, 1990; Knafl et al., 1996; Knafl & Deatrck, 2003; Krouse, 2002; Meier, 2003; Meier, Furman, & Degenhardt, 2007; Moe et al., 1998; Pisacane et al., 2003; Weatherley-White, Kuehn, Mirrett, Gilman, & Weatherley-White, 1987), the FMSF, and the investigator’s clinical expertise with families of infants with PKU.

The initial interview guide was based on FMSF and themes for mothers. Mothers were asked questions about how they defined their situations, their

management behaviors, and their perception of the consequences of breastfeeding and having an infant with PKU, for family life. The FMSF provided the basis for the overall structure of the interview; the focus of specific questions and probes came from the literature and the researcher's clinical experience with the target sample.

The interview guide was pilot-tested (Perry, 2002). It was anticipated that less than 45 minutes would be required to complete the telephone interview. The length of time to complete the telephone interview ranged from 56 to 125 minutes with a mean of 92 ($SD = 26.3$).

Phase 2: Quantitative Measures

Two measures assessing family functioning comprised the quantitative portion of the interview. Table 5 summarizes the concepts and quantitative measures used in the study (see Appendix G).

Family APGAR questionnaire. The Family APGAR Questionnaire provided an overall view of family functioning as perceived by the participant (Smilkstein, 1978). The Family APGAR assessed adaptability, partnership, growth, affection and resolve. The Likert-style items measured satisfaction with help received from family (adaptability), shared problem solving (partnership), encouragement of personal growth (growth), expression and response to feelings (affection), and time spent together (resolve). The response options ranged from 0 (hardly ever) to 2 (almost always); the five items were summed, yielded a total score ranging from 0 to 10. High scores, such as scores from 7 to 10, indicate a

Table 5

Quantitative Measures of Family Functioning

Variable	Measure
Family Functioning	1) Family Apgar Questionnaire (Smilkstein, G. (1978). The Family APGAR: A proposal for family function test and its use by physicians. <i>Journal of Family Practice</i> , 6(6), 1231-1239.)
	2) General functioning scale of the Family Assessment Device (Epstein, N. B., Baldwin, L. M., & Bishop, D. S. (1983). The McMaster Family Assessment Device. <i>Journal of Marital and Family Therapy</i> , 9, 171-180.)

greater degree of satisfaction with family functioning. Scores of 4 to 6 suggest moderate family dysfunction while scores of 0 to 3 reflect a highly dysfunctional family. Prior studies have reported high internal consistency demonstrating that the Family APGAR is an uni-dimensional measure of satisfaction with family life (Sawin & Harrigan, 1995). Validity of the Family APGAR has been established by extensive research showing significant correlations with other established measures of family functioning as well as clinicians' ratings of family functioning (Fink, 1995).

General functioning scale of the Family Assessment Device. The Family Assessment Device (FAD) was based on the McMaster Model of Family Functioning, which is a clinically orientated conceptualization of families. The

model identifies six dimensions of family functioning: problem-solving (ability to resolve problems), communication (exchange of clear and direct verbal information), roles (division of responsibility for completing family tasks), affective responsiveness (ability to respond with appropriate emotion), affective involvement (degree to which family members are involved and interested in one another), and behavior control (manner used to express and maintain standards of behavior). The FAD also includes an independent dimension of general functioning (overall functioning of family). Previous studies reported that the general functioning subscale (GF scale) correlated highly with the other 48 items, thus supporting its use as a single index representing overall functioning (Epstein, Baldwin, & Bishop, 1983).

The general functioning (GF) scale was designed to determine a member's overall perception of the family's functioning. This 12-item subscale measures family interaction patterns that are both healthy and unhealthy. The scale uses a 4-point Likert-type scale ranging from 1 (*strongly agree*) to 4 (*strongly disagree*). To calculate the GF score for this study, the scores for items describing unhealthy functioning were transformed by subtracting the rating of 1 to 4 from 5, in order to essentially reverse code the items. Next, the 12 ratings were summed and an average was obtained by dividing the total score by 12 to yield the family function score (Epstein, Baldwin, & Bishop, 2005). Higher scores are indicative of poorer family functioning. A 2.0 or higher suggest unhealthy family functioning (Miller, Epstein, Bishop, & Keitner, 1985; Miller et al., 1994). Several studies provide evidence of predictive, concurrent, and construct validity of the GF scale (Byles,

Byrne, Boyle, & Offord, 1988; Kabacoff, Miller, Bishop, Epstein, & Keitner, 1990; Offord et al., 1987) In addition, GF scores are able to discriminate between clinical and nonclinical samples, thus supporting the discriminate validity of the scale (Epstein et al., 1983).

Data Management and Analysis

Thematic analysis. Similar to other qualitative approaches, qualitative description uses the six analytic strategies outlined by Miles and Huberman (1994): 1) coding data from interviews, observations, and notes; 2) recording insights and reflections on the data; 3) sorting data to identify similar phrases, patterns, and themes; 4) extracting commonalities and differences for further analysis; 5) discerning generalizations from the data; and 6) examining generalizations to known scientific knowledge. The end product of qualitative description is a clear, logical, coherent, and articulate description of the participants' experiences that are easily understood by both the participants and researchers.

Thematic analysis is a systematic interpretive analysis of narrative material that involves the creation or application of codes to data (Miles & Huberman, 1994; Patton, 2002). It facilitates use of a conceptual framework from the literature, such as the FMSF, by providing a starting point for coding that allows further codes to emerge from the data. With thematic analysis, data collection and data analysis can be concurrent.

In general, there are five key stages in thematic analysis. First, one becomes familiar with the data. Transcripts are transcribed verbatim with detailed

reading afterwards. The transcripts are organized and described to maintain anonymity and summarize the data. The next step is identification of the thematic framework. This is developed from the conceptual framework and refined as analysis progresses. Once the thematic framework is identified, thematic coding proceeds. Thematic codes are defined as units derived from patterns such as vocabulary, recurring activities, meanings, or feelings. Themes that emerge from the participants' data are pieced together to form a comprehensive picture of their collective experiences. With thematic coding, one is applying codes to sections of the text that correspond to different themes from the framework. When there are no existing themes that are appropriate, a new theme is created.

Thematic charting follows thematic coding. In thematic charting, there are two different ways: theme chart and case chart. Either one tracks themes for each participant or the cases for each theme. Patterns are identified at the case level. Another aspect of thematic charting is the need to record quotations with page and line references illustrating each theme.

The last stage is mapping and interpretation. In this stage, related patterns are combined and catalogued into sub-themes. Patterns, associations, concepts, and explanations are sought in the data related to the original research questions to build a valid argument for choosing the themes. At times, it can be very helpful to use visual aids to display the ideas and interpretations (Miles & Huberman, 1994; Saldana, 2009). The aim of thematic mapping and interpretation is to illustrate how the findings fit within an existing framework, such as the FMSF: how they extend the conceptual framework; or how it explains why the conceptual

framework does or does not apply in certain contexts (Miles & Huberman, 1994; Patton, 2002; Saldana, 2009).

Data Analysis Strategies

Qualitative Interview Data Management and Analysis

The management and analysis of the qualitative interviews in this study were based upon the guidelines and techniques of Patton (2002), Miles and Huberman (1994), and Knafl and Ayres (1996). Data management and analysis began after the first two interviews were completed. Early during Phase 2, data management focused on identifying and redefining coding categories from the conceptual framework and on what the mothers said. As the study proceeded, analysis focused on identifying themes that cut across multiple codes and patterns of themes that characterize the mothers. Data management and analysis involved the following major activities: 1) processing qualitative data, 2) analyzing qualitative data to identify themes in mothers' responses, 3) identifying and verifying patterns of mothers' experiences, and 4) ensuring the quality of the overall analytic process.

Processing qualitative data. The following data processing steps were accomplished: 1) transcribed the taped interviews, 2) identified code categories, and 3) coded all interview transcripts by hand. All audiotapes were transcribed verbatim by the primary researcher. The interview transcripts were checked for accuracy against the interview tapes by the primary researcher. At first, coding categories were derived from the basic topic of a passage of qualitative data. As coding proceeded, new code categories were developed that combined codes

derived from the data and those linked to specific interview guide questions. At this stage of analysis, the coding categories were highly descriptive.

Coding categories and themes reflect the naturalistic inquiry and the data (Denzin & Lincoln, 2005; Knafl, Breitmayer, Gallo, & Zoeller, 1992; Knafl et al., 1996). This technique allowed the investigator to simultaneously collect information through interviews, read interviews as individual cases, disassemble interviews through coding, rearrange coded categories into patterns, and reintegrate the patterned categories into a conceptualization that included the experiences of all the participants. In this study, the final conceptualization was the mothers' experiences of breastfeeding infants with PKU. In order for the analysis to be systematic and rigorous, the analysis was guided by recommendations from Patton (2002), and Miles and Huberman (1994).

Analyzing, identifying, and verifying behavioral patterns. The FMSF was used to help categorize themes described by the mothers. Data were analyzed to consider the behaviors mothers used to simultaneously manage breastfeeding and PKU.

Data Analysis of Structured Measures

The third aim was to identify mothers' experiences breastfeeding infants with PKU and examine their assessment of family functioning. Data from the structured measures (Family Apgar and FAD) contributed to the beginning exploration of the relationship between simultaneously managing both breastfeeding and PKU and the mothers' perceptions of family functioning. Although the small sample size precluded a definitive test of relationships, it

provided data from the structured measures that provided additional insights into lives of families with an infant with PKU. As such, these data contribute to a more complete understanding of breastfeeding in the context of PKU.

Integration of Qualitative and Quantitative Data

In this study, quantitative analysis of the Internet survey predominated, and the qualitative description and quantitative measures from the in-depth interviews were used primarily for descriptive purposes. The qualitative findings were used to expand and to complement the description of the mothers' experiences as addressed in the quantitative analysis. Challenges arose in integrating qualitative and quantitative study findings due to the discrepancies between the two types of data. Members of the dissertation committee provided guidance to help interpret those discrepancies and put them into context.

Ensuring Quality Data

In order to gather high-quality data, the investigator carried out verbatim transcription, demonstrated data saturation, documented data analysis decisions, participated in reflexive journaling, acknowledged the researcher perspective, and maintained an audit trail and evidence that supported interpretations and yielded thick descriptions (Whittemore, Chase, & Mandle, 2001). Other strategies to ensure quality included bracketing assumptions, piloting the interview guide, modifying the interview guide during data collection as needed, and crosschecking coding with members of the dissertation committee (Tashakkori & Teddlie, 1998). Field supervision was provided by Drs. Knafl and Kodadek who facilitated reflection on the impact of the investigator's racial, ethnic, and professional

background on her interactions with participants, her interpretive lens, and her ability to maintain “empathic neutrality” (Patton, 2002).

The investigator maintained an audit trail of her efforts and used her dissertation committee members to serve as auditors who monitored the study for overall quality. Rodgers and Cowles (1993) provided concrete guidelines for maintaining an audit trail that was periodically reviewed by the dissertation committee who were in a position to judge both the quality of the research and provide ongoing advice about improving quality. In keeping with these guidelines, the investigator maintained three types of documents to track the implementation of the study: contextual, methodological, and analytical (Rodgers & Cowles, 1993). Contextual documentation focused on the interview process and the factors that might influence the quality of the participants’ responses. Methodological documentation was the basis for assessing the appropriateness and consistency of ongoing decisions regarding the research design. Analytic documentation made it possible to retrace the insights, thinking, and interpretations that led to the final results.

Protection of Human Participants

This study was submitted for approval by the Institutional Review Board at Oregon Health & Science University and was approved in September 2008 (see Appendix F). Phase 2 of this study does not include a signed consent form; rather, verbal consent was obtained and a copy of the consent was mailed to each participant. After being contacted by the mother, the investigator verified the mother’s eligibility and explained the purpose of the study and study procedures.

Research consent forms were read over the phone, and the investigator obtained verbal oral consent from the participant at this time. Each participant was assigned a unique study identifier. The investigator retained the code file separate from the data that linked the unique study identifier with the participants. This provided for the identities of the study participants to be kept strictly confidential. The data collection file with the unique study identifier was password-protected and kept in an encrypted laptop computer.

Potential human participant concerns. The purpose of the interview, its conversational format, measures taken to ensure confidentiality and the participant's rights as a research participant including the right to stop the interview at any time and not to answer any questions she chose was clearly discussed with the participant before the interview. Any questions a participant had were answered before the interview.

Risks. It was possible that the participant might have been emotionally distressed during the interview if she shared experiences that had been distressing and/or difficult for her to deal with in the past or present.

Measures to protect against risk. Participants were encouraged not to use names in the interviews. To ensure anonymity, the transcribed interview and field notes have no identifiers that can be linked to the participant's identity. The investigator forewarned the participant regarding potential study risks. Any questions from the participant was discussed and answered prior to initiation of interviews. In the event that emotional distress occurred during the interview, the participant was given the option of stopping or pausing the interview. No

professional counseling was needed by any of the participants. Following transcription, the digital recordings were destroyed.

Summary

The first purpose of this study was to determine the prevalence and duration of breastfeeding infants with PKU in the U.S. and Canada while the second and third purposes were to generate knowledge about how mothers of infants with PKU simultaneously manage PKU and breastfeeding, and how they perceived family functioning, respectively. This study was a cross-sectional design with two data collection phases. In the first phase, mothers of infants with PKU were invited to participate in an Internet survey on experiences of feeding their infants. In the second phase, a subsample of mothers from phase 1 were selected and interviewed to explore their experiences breastfeeding infants with PKU. For phase 1, using Internet survey for data collection and analysis was reviewed. For phase two, qualitative description was presented and the rationale was explained. In addition, during second phase, two quantitative measures described mothers' perceived assessment of family functioning which further elucidated patterns of family response that characterized this specific population. The sample and recruitment plans for each phase were outlined. Strategies for data collection and analysis for each phase were presented.

CHAPTER 4

RESULTS

The purpose of this study was twofold: 1) to determine the prevalence and duration of breastfeeding for infants with PKU in the U.S. and Canada and 2) to generate knowledge about how mothers of these infants simultaneously manage breastfeeding and PKU therapy, and how they perceive family functioning. The primary aim of this study was to describe the prevalence and duration of breastfeeding among mothers who have infants with PKU in the U.S. and Canada. The secondary aim was to describe mothers' experience of breastfeeding in the context of PKU, including their perceptions of factors that facilitate and/or interfere with breastfeeding infants with PKU, and to describe their strategies to care for their breastfeeding infants with PKU. The third specific aim was to identify mothers' experiences breastfeeding infants with PKU using thematic analysis and to examine mothers' assessment of family functioning. The characteristics of the mother, the child with PKU, and their family of those that constituted this sample are reported first, followed by results according to specific aims. In this study, *breastfeeding* was defined as any amount of breast milk from an infant nursing at breast and feeding on expressed mothers' milk from a bottle.

Sample

The selection of participants for the international Internet survey was convenience-based. The participants were mothers who met the following criteria: 1) at least 21 years of age, 2) able to read and write in English, 3) had child/children with PKU, and 4) lived in the U.S. or Canada. The final sample was

composed of 119 mothers in two groups: mothers with one child with PKU ($n = 103$), and mothers with more than one child with PKU ($n = 16$). Maternal data included ethnicity, age, marital status, education, and employment. In addition, data were collected on region of residence, population density of residence, distance from metabolic clinic, and forms of communication with metabolic clinic. Child data included gender, birth weight, birth difficulties, phenylalanine (Phe) level from newborn screen, and modes of feeding before and after diagnosis. In this study, data from mothers with one child who had PKU are reported.

Maternal Characteristics

The maternal sample was primarily composed of Caucasian women ($n = 95$) (see Table 6), which is consistent with the PKU profile of Northern European heritage. At time of the international Internet survey, maternal age ranged from 21 to 63 years, with a mean of 35.6 years ($SD = 6.98$) with the majority of mothers in their 30s ($n = 64$, mode = 34 years). However, three mothers were adolescents and four were in their 40s at the time of the birth of their child with PKU. Nearly all the mothers were married or partnered ($n = 94$, 91%).

Table 6

Descriptive Statistics: Maternal Age, Ethnicity, and Marital Status (N = 103)

Age at Survey			Ethnicity			Marital Status		
Years	<i>n</i>	(%)		<i>n</i>	(%)		<i>n</i>	(%)
21 -29	18	17.5	Asian/Pacific Islander	1	1.0	Never married or partnered	3	2.9
30 - 39	64	62.0	Caucasian	95	92.2	Living with partner	3	2.9
40 - 49	16	15.6	Hispanic/Latino	5	4.9	Married	91	88.3
50 - 65	5	4.9	Native American/ Alaska Native	2	1.9	Separated	2	1.9
						Divorced	4	3.9

The sample reflected a well-educated group of mothers; all mothers were high school graduates with more than two-thirds ($n = 2$, 70.1%) of them college graduates. Maternal employment varied from 30% ($n = 31$) being “stay-at-home moms,” 24% ($n = 24$) working at least half-time, and one-third ($n = 33$, 32%) working full-time. As seen in Table 7, nearly two-thirds ($n = 63$, 63.7%) of the mothers reported a household income greater than \$75,000.

Table 7

Maternal Education, Employment, and Household Income

Highest Completed Education			Employment			Gross Annual Household Income		
			Hours Worked per Week					
	<i>n</i>	(%)		<i>n</i>	(%)	\$	<i>n</i>	(%)
High School	3	2.9	0	31	30.1	< 25,000	4	4.0
Some College	13	12.6	< 8	4	4.0	25,001 – 50,000	12	12.1
Associates Degree	15	14.6	10 - 16	4	4.0	50,001 – 75,000	20	20.2
Bachelors Degree	38	36.9	20 - 38	24	24.1	75,001 – 100,000	32	32.3
Masters Degree	25	24.3	40	33	32.0	100,001 – 150,000	16	16.2
Professional Degree	4	3.9	> 40	6	5.8	> 150,000	15	15.2
Doctoral Degree	5	5.0						

Child Demographics

The proportion of males ($n = 48$, 46.6%) and females ($n = 55$, 53.4%) was comparable to that of other autosomal recessive disorders. The majority of the infants had normal birth weights ($M = 3.47\text{kg}$, $SD = 0.54$). Most of the infants were born at term ($n = 94$, 91%) from uncomplicated pregnancies ($n = 93$, 90%) via vaginal delivery ($n = 75$, 73%). In addition, 90 (77%) of the children were born between 2000 and 2011. Initial newborn screening was performed from 1 to 28 days of age; on average, the screening occurred on the fourth day ($M = 4.04$, $SD = 4.1$) and most frequently on the second day ($MO = 2.0$). Over 50% ($n = 54$) of the children had their initial newborn screening performed within 48 hours of delivery. From newborn screening results, initial Phe levels ranged from 240 – 2,520 $\mu\text{mol/dl}$ (4 – 42 $\text{mg}\%$) with a mean of 948 $\mu\text{mol/dl}$ ($SD = 601$). This resulted in two-thirds of infants ($n=69$) being seen in a metabolic clinic with treatment for PKU started by 10 days of age, which is consistent with the recommendation from the National Institutes of Health Consensus Statement on PKU Screening and Management (2000).

Family Demographics

The majority of the families who had one child with PKU ($n = 78$) were families with a single child who has PKU ($n = 38$, 36.9%) or were families with two children, one child with PKU and one without PKU ($n = 40$, 38.8%) (Table 8). The families were from various regions across the U.S. and Canada (Table 9), representing 29 states and 7 provinces, respectively. In the U.S., the majority of families were from the northeastern and southeastern regions ($n = 51$, 49.5%). In

Canada, the families were from the central and prairie provinces ($n = 9$, 8.8%). In addition, families were from a variety of communities, ranging from small rural towns to large metropolitan urban areas (Table 10). The majority ($n = 36$, 35%) of families reported living in a city with a population from 10,000 to 50,000 people. Most families reported they traveled 11 to 50 miles ($n = 48$, 46.6%) to attend a pediatric metabolic clinic, but the distance varied from less than 5 miles to more than 250 miles (Table 11).

Table 8

Total Number of Children in the Family including Child with PKU

Number of Children	<i>n</i>	(%)
One	38	36.9
Two	40	38.8
Three	22	21.4
Four	2	1.9
Five or more	1	1.0

Table 9

Region of Residence

USA ¹ (n=89)	<i>n</i>	(%)
Northeastern (ME, NH, VT, MA, RI, CT, NY, NJ, PA, DE, MD)	26	25.2
Southeastern (DC, VA, WV, NC, SC, GA, FL, KY, TN, MS, AL, TX, OK, AR, LA)	25	24.3
Midwest (WI, MI, IL, IN, OH, ND, SD, NE, KS, MN, IA, MO)	21	20.4
Mountainous (ID, MT, WY, NV, UT, CO, AZ, NM)	5	4.9
West Coast (AK, WA, OR, CA, HI)	12	11.7
Canada ² (n=14)		
Atlantic (NL, NB, NS, PE)	3	2.9
Central (ON, QC)	5	4.9
Prairies (MB, AB, SK)	4	3.9
Pacific Coast (BC)	2	1.9

¹USA: AL = Alabama, AK = Alaska, AR = Arkansas, AZ = Arizona, CA = California, CO = Colorado, CT = Connecticut, DE = Delaware, FL = Florida, GA = Georgia, HI = Hawaii, ID = Idaho, IL = Illinois, IN = Indiana, IA = Iowa, KS = Kansas, KY = Kentucky, LA = Louisiana, ME = Maine, MD = Maryland, MA = Massachusetts, MI = Michigan, MN = Minnesota, MS = Mississippi, MO = Missouri, MT = Montana, NE = Nebraska, NV = Nevada, NH = New Hampshire, NJ = New Jersey, NM = New Mexico, NY = New York, NC = North Carolina, ND = North Dakota, OH = Ohio, OK = Oklahoma, OR = Oregon, PA = Pennsylvania, RI = Rhode Island, SC = South Carolina, SD = South Dakota, TN = Tennessee, TX = Texas, UT = Utah, VT = Vermont, VA = Virginia, WA = Washington, WV = West Virginia, WI = Wisconsin, WY = Wyoming

²Canada: BC = British Columbia, MB = Manitoba, NB = New Brunswick, NL = Newfoundland and Labrador, NS = Nova Scotia, ON = Ontario, PE = Prince Edward Island, QC = Quebec, SK = Saskatchewan

Table 10

Population Density of Community of Maternal Residence

Community Population Size	<i>n</i>	(%)
A town with a population < 10,000	18	17.5
A city with a population between 10,000 – 50,000	36	35.0
A city with a population between 50,001 – 100,000	14	13.6
A metropolitan area with a population between 100,001 – 500,000	14	13.6
A metropolitan area with a population between 500,001 – 1 million	10	9.7
A metropolitan area with a population > 1 million	11	10.7

Table 11

Distance Traveled to Attend Pediatric Metabolic Clinic

Number of Miles Traveled	<i>n</i>	(%)
< 10 miles	36	35.0
10 – 50 miles	44	42.7
51 – 150 miles	19	18.4
> 150 miles	4	3.9

Over the course of the child's disorder, mothers have changed their methods of communicating with their pediatric metabolic clinic (Table 12). At the time of diagnosis, the telephone ($n = 83$, 80.6 %) was the primary method of communicating with the staff at the pediatric metabolic clinic. Currently, however, mothers reported that they are using electronic devices such as email ($n = 73$,

71%), cell phones ($n = 59$, 57%), and iPhone/Blackberry ($n = 4$, 4%) to communicate with the staff at their child's pediatric metabolic clinic.

Table 12

Modes of Communication at Diagnosis and Current (may use more than one method of communication)

Modes of Communication	At Time of Diagnosis		Currently		Change	
	<i>n</i>	(%)	<i>n</i>	(%)	<i>n</i>	(%)
Telephone	83	80.6	79	76.7	-4	3.9
Mail Service	9	8.7	35	34	+26	25.3
Fax	0	0	7	6.8	+7	6.8
Cell phones	26	25.2	59	57.3	+33	32.1
iPhone/Blackberry	4	3.9	12	11.7	+8	7.8
Email	7	6.8	73	70.9	+66	64.1

Overall, the sample is representative of highly educated, married/partnered Caucasian mothers in the U.S. and Canada who have a child with PKU. This sample represents mothers who were employed at least half time with the remainder being "stay-at-home moms."

Findings

Prevalence and Duration of Breastfeeding

The first specific aim was to describe the prevalence and duration of breastfeeding among mothers whose infants have PKU in the U.S. and Canada. A total of 103 mothers responded to an announcement on the PKU Listserv about

their experiences feeding infants with PKU. The results revealed that 89 mothers (86%) started breastfeeding and 14 mothers (14%) began bottle feeding immediately after delivery. After diagnosis, 18 mothers switched from breastfeeding to bottle feeding while one mother initiated breastfeeding. Of the 89 mothers who breastfed after delivery, 75 (84%) were from the U.S. and 14 (16%) were from Canada. After diagnosis, 60 mothers (80%) from the U.S. continued to breastfeed while 15 (20%) switched to bottle feeding while one mother switched from bottle feeding to breastfeeding. In contrast, only two Canadian mothers switched from breastfeeding to bottle feeding after the diagnosis of PKU. This reduction in breastfeeding was significant.

Analysis of this data using the McNemar's test was performed to assess the difference between the proportion of mothers breastfeeding immediately after delivery and the proportion of mothers who continued breastfeeding after the diagnosis of PKU. See Figure 5. There were significantly fewer mothers breastfeeding after diagnosis (McNemar's $\chi^2 = 30.333$, $p \leq .0001$; $n = 72$ versus $n = 89$).

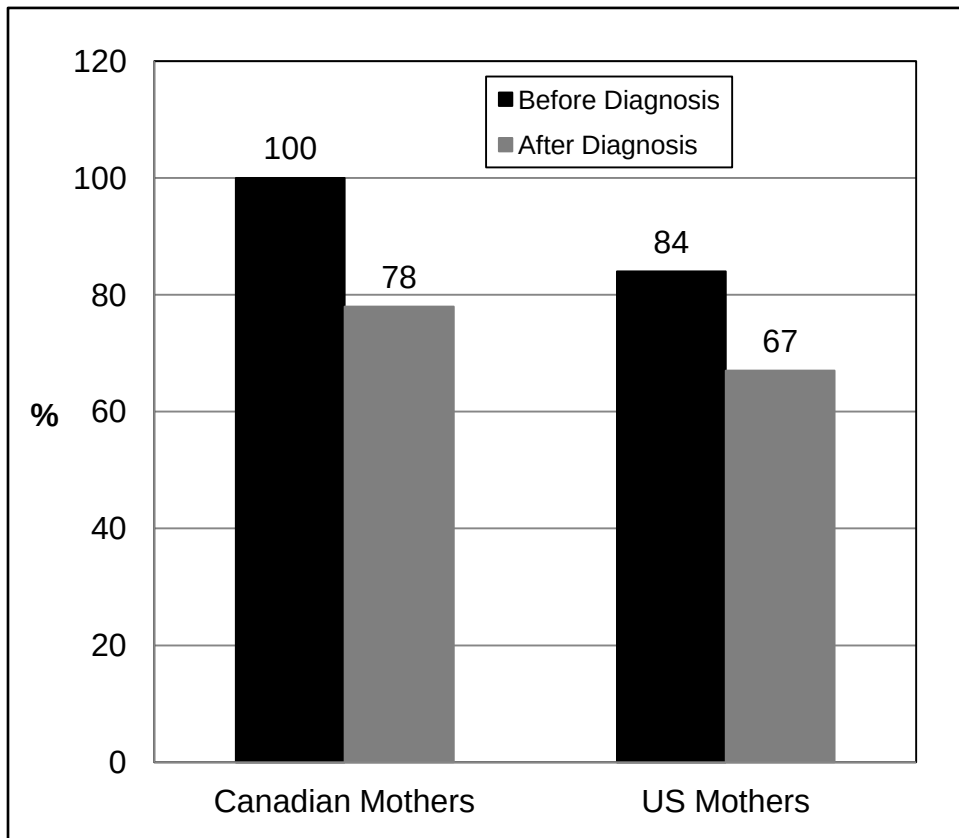


Figure 5. Breastfeeding Prevalence before and after Diagnosis of PKU

Given the significance of fewer mothers breastfeeding after diagnosis, an ad hoc analysis was conducted to ascertain whether this significance differed by country. Of the 89 mothers who after delivery, 75 (84%) were from the U.S. and 14 (16%) were from Canada. After diagnosis, 60 mothers (80%) from the United States continued to breastfeed while 15 mothers (20%) switched to bottle feeding. In contrast, only two Canadian mothers switched from breastfeeding to bottle feeding after diagnosis. Using McNemar's test with layering of the U.S. and Canada, the significant reduction in breastfeeding mothers was a function of women from the U.S. (McNemar's $\chi^2 = 27.48$ $p \leq .0001$; $n = 60$ versus $n = 75$), and not from Canada. See Figure 6.

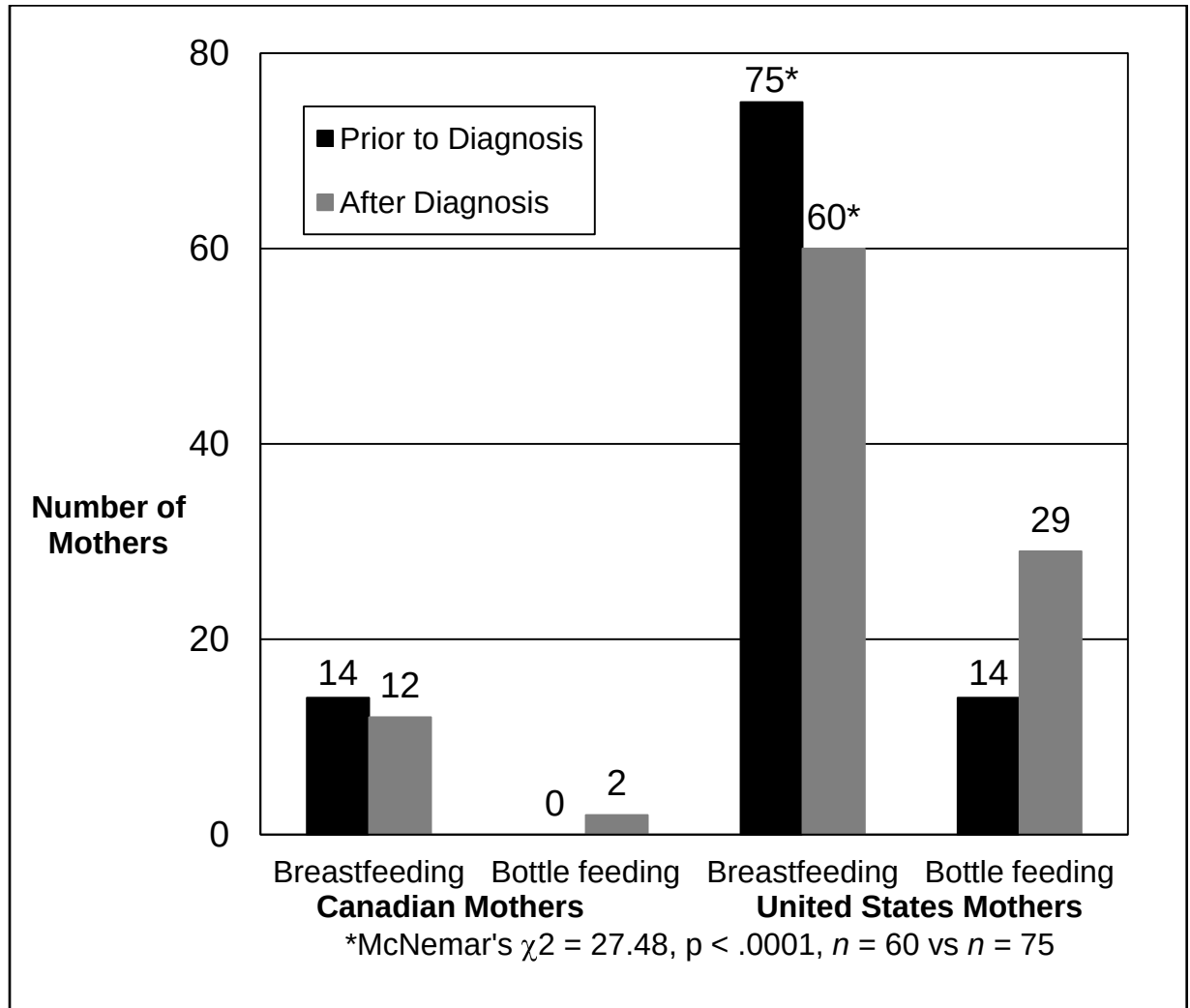


Figure 6. Comparison of Breastfeeding Prevalence before and after Diagnosis of PKU between Mothers Living in the U.S. and Canada

The duration of breastfeeding among mothers in the survey ranged from less than one month ($n = 20$, 22.2%) to 19-24 months ($n = 5$, 5.5%). There was one modal duration category ($n = 20$, 22.2%): at less than one month. See Table 13 for the distribution of duration of breastfeeding categories.

Table 13

Duration of Breastfeeding

Duration in Months	<i>N</i>	(%)
Less than one month	20	22.2
1 to 3 months	13	14.4
4 to 6 months	9	10.0
7 to 9 months	15	16.6
10 to 12 months	19	21.1
13 to 18 months	10	11.1
19 to 24 months	5	5.5

Several variables were assessed in relation to the duration of breastfeeding using chi square analysis. Only one variable, when standard commercial infant formula was added into the diet replacing breastfeeding or expressed mothers' milk, was significantly associated with decreased duration of breastfeeding for infants with PKU: $\chi^2 (42, n = 73) = 88.13, p < .0001$. Of those identifying when standard commercial formula was added to their infant's dietary plan, 30% ($n = 21$) had added it by 6 months. Thirty-three percent of the mothers ($n = 34$) never introduced standard commercial infant formula into their infant's diet; rather, they transitioned from breastfeeding to the introduction of solid foods with Phe-free formula. The following categorical variables were not associated with duration of breastfeeding: maternal age ($\chi^2 = 141.11, n = 89$; NS), marital status ($\chi^2 = 23.06$,

$n = 89$; NS), education level of mother ($\chi^2 = 34.44$, $n = 89$; NS), maternal ethnicity ($\chi^2 = 19.55$, $n = 89$; NS), hours employed per week ($\chi^2 = 96.17$, $n = 89$; NS), total number of children in the family ($\chi^2 = 22.92$, $n = 89$; NS), country of residence ($\chi^2 = 2.74$, $n = 89$; NS), mode of delivery ($\chi^2 = .963$, $n = 89$; NS), type of insurance ($\chi^2 = 22.11$, $n = 89$; NS), annual household income ($\chi^2 = 24.34$, $n = 85$; NS), infant's initial Phe level ($\chi^2 = 256.60$, $n = 81$; NS), and initiation of dietary therapy ($\chi^2 = 143.23$, $n = 86$; NS).

Mothers Experiences Breastfeeding Infants with PKU

The second specific aim was to describe mothers' experiences breastfeeding in the context of PKU including their perceptions of: 1) factors that facilitate and/or interfere with breastfeeding infants with PKU, and 2) strategies to obtain the necessary skills to care for their breastfeeding infants with PKU. Of the 89 mothers, the majority provided responses about their breastfeeding experiences to four open-ended questions on the Internet survey regarding challenges ($n = 77$), ease ($n = 67$), help ($n = 58$), and support ($n = 68$). Although all mothers did not respond to each open-ended question, many mothers provided multiple responses within their answers.

From the open-ended responses, a frequency count approach was used to begin the analysis of these responses in which mothers described: challenges, supports, factors that eased and helped, and strategies to obtain the skills for breastfeeding infants with PKU. Specific questions were asked of each set of responses to identify categories; then these were tabulated. The number for each category was divided by the number of respondents. Initially, all four sets of

responses (challenges, ease, help, and support) were double-counted by the primary researcher. In addition, members of a qualitative seminar were recruited to verify coded responses and were provided definitions and rules for coding. After discussion with them, some categories were revised and the primary researcher repeated the double counting process. Subsequent, inter-coder agreement was assessed with a faculty member. To establish inter-coder reliability, percent agreement was calculated. Percent agreement is widely used and simple to calculate as inter-coder reliability is equal to the number of agreements divided by the total number of agreements plus disagreements. All responses were double-coded and yielded the following percent agreement for each category: challenges, 91%; ease, 94%; help 90%; and support, 92%.

Quantitative Approach to Narrative Responses

Challenges to Breastfeeding Infants with PKU

The survey asked mothers to describe “What challenges did you have in breastfeeding your infant with PKU?” To develop categories for this question, three specific questions were asked of the responses: 1) What were the general breastfeeding challenges for mothers breastfeeding infants with PKU? 2) What were the breastfeeding and PKU challenges for mothers breastfeeding infants with PKU? 3) Did some mothers not have any challenges breastfeeding infants with PKU? Three categories (*Common Breastfeeding Issues*, *Breastfeeding and PKU*, and *No Challenges*) were identified; see Figures 7 and 8.

Common Breastfeeding Issues. Within the category of Common Breastfeeding Issues ($n = 56$, 74%), four subcategories were identified:

1) *breast pumping*, 2) *breast milk supply issues*, 3) *nipple confusion*, and 4) *common breastfeeding problems*. All four challenges can be identified in the breastfeeding literature, but for these mothers, the issue is heightened due to frequent Phe testing to monitor Phe levels. Of the challenges, mothers ($n = 34$, 45%) described *breast pumping* as the most frequent breastfeeding problem that challenged them while breastfeeding their infants with PKU. *Breast pumping* refers to issues using a mechanical device that provides gentle suction for the removal of breast milk from breasts when infant feeding is not possible for various reasons. Mothers wrote comments like the following: “spent hours pumping just to be able to breastfeed twice a day,” “required to pump in order to maintain supply which was exhausting,” “I had to be diligent about pumping,” and “The biggest challenge for me during this time was pumping.”

Mothers ($n = 26$, 35%) described *breast milk supply* as another common breastfeeding problem that challenged them while breastfeeding infants with PKU. *Breast milk supply* refers to the ability of the mammary glands to produce adequate amounts of breast milk due to galactopoiesis. Comments included the following: “keeping up milk production with frequent changes in how often she was breastfed,” “trouble keeping my milk supply up,” “hard time maintaining a consistent milk supply,” and “eventually my milk supply ran out.”

Another frequent challenge reported ($n = 21$, 28%) was the issue of *nipple confusion* when an infant prefers either the breast or bottle nipple over the other. One mother wrote, “My son hated taking a bottle and would scream at every

feeding.” Another mother reported, “The biggest challenge was getting my daughter to take the Phe-free formula out of a bottle! She hated the bottle!”

Despite problems with getting their infants to take the bottle, mothers revealed their successful ability to find solutions. While one mother reported, “We had problems getting my son to take a bottle after the PKU diagnosis. I had to feed him the PKU formula using a dropper because he only wanted to breastfeed,” another mother provided this description:

What made it more difficult was the confusion for the baby when he was bottle fed the formula. The way he had to suck was different and for a baby so little, it was confusing. So I stopped using standard bottle and I used syringe, the same used for giving medication for babies. It was much slower, but much easier.

Several mothers ($n = 12$, 16%) identified *common breastfeeding problems* as a challenge while breastfeeding infants with PKU. *Common breastfeeding problems* refers to breastfeeding concerns that any breastfeeding mother might experience, such as poor latch, nipple or breast pain, thrush, and sore, cracked, and bleeding nipples. For example, one mother wrote, “regular challenges of breastfeeding – plugged ducts and thrush.” Other mothers described “My baby did not have a good suction for a start, which had nothing to do with PKU,” and “I had extremely sore nipples that were constantly cracked and bled.”

Breastfeeding and PKU. Within the second category of challenges, *Breastfeeding and PKU* ($n = 49$, 65%), mothers reported three subcategories: 1) *management*, 2) “*how much*,” and 3) *time*. Frequently, mothers identified *management* ($n = 43$, 57%) as the main issue of Breastfeeding and PKU. *Management* included the work of breastfeeding, breast pumping, and feeding

Phe-free formula on a daily basis, which frequently changed due to Phe results. Mothers wrote: “difficulty with 3 different types of feeding and mixing,” “I used the pump for as long as I could to give him breast milk with Phe-free formula + formula to keep up with his feedings,” and “had to feed low protein bottle before or after breastfeeding.” One mother expressed her frustration with the frequent changes:

My son's tolerance would change on a weekly basis. Some weeks I could breastfeed 6 times a day, others only 4 times a day. Unfortunately, breast milk works on supply & demand and is not a spigot. It was hard to suddenly reduce the number of breastfeeds as I would get engorged, and even harder to suddenly increase the number of breastfeeds because I wasn't producing enough milk.

Another challenge identified by mothers ($n = 22$, 30%) was the issue of “*how much*,” which referred to the inability to determine *how much* breast milk the infant with PKU had consumed while suckling at breast leading to the concern of *how much* Phe that the infant obtained from the breast milk. Comments included: “I was always worried about knowing the exact amount of Phe that he was getting,” “difficult to know *how much* breast milk baby was getting,” and “Measuring intake. I was only able to communicate length of breastfeeding sessions” while another mother reported “To make 100% sure on *how much* Phe my son was getting, I totally gave up breastfeeding.”

Mothers ($n = 16$, 21%) reported *time* as a challenge to breastfeeding infants with PKU. *Time* referred to how long was spent and the prioritization of tasks in order to maximize personal efficiency in breastfeeding infants with PKU. Mothers revealed “spent hours pumping,” “finding the time,” and “having to spend time.”

No Challenges. There were several mothers ($n = 10$, 13%) who identified *no challenges* to breastfeeding their infants with PKU. *No challenges* referred to

the fact that nothing was a problem or issue while breastfeeding infants with PKU.

The majority of these mothers simply wrote “none” or “no challenges.”

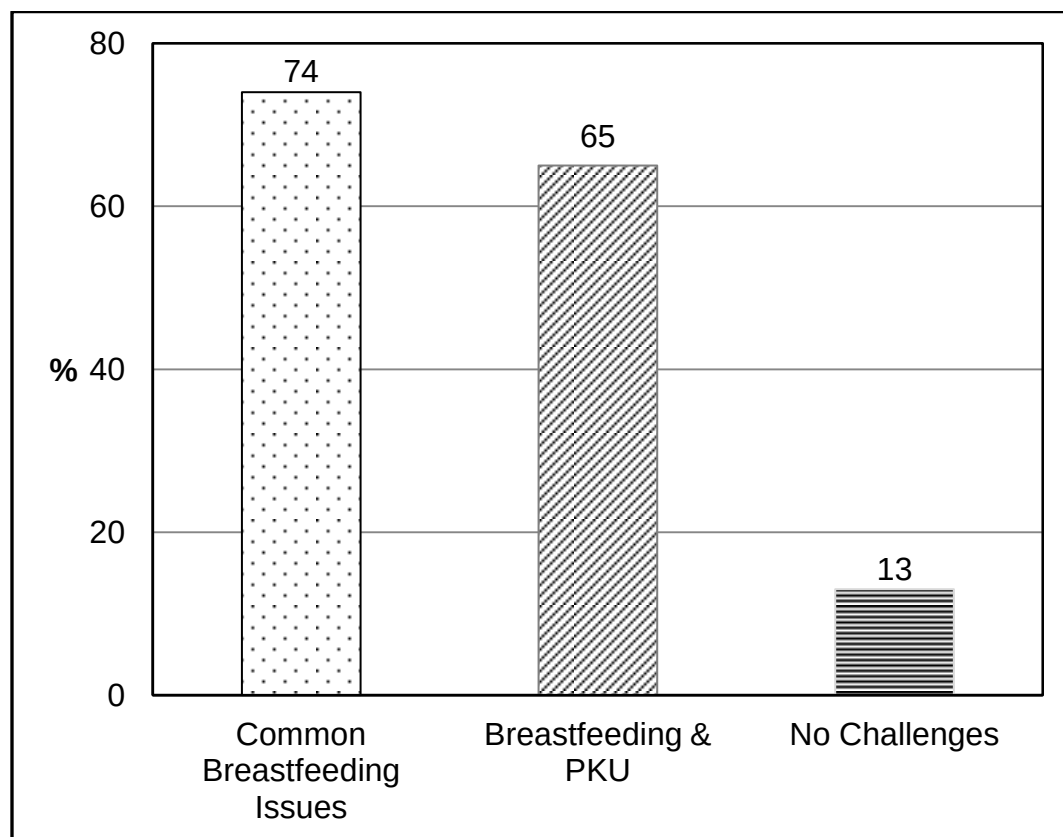


Figure 7. Challenges Identified by Mothers while Breastfeeding Infants with PKU

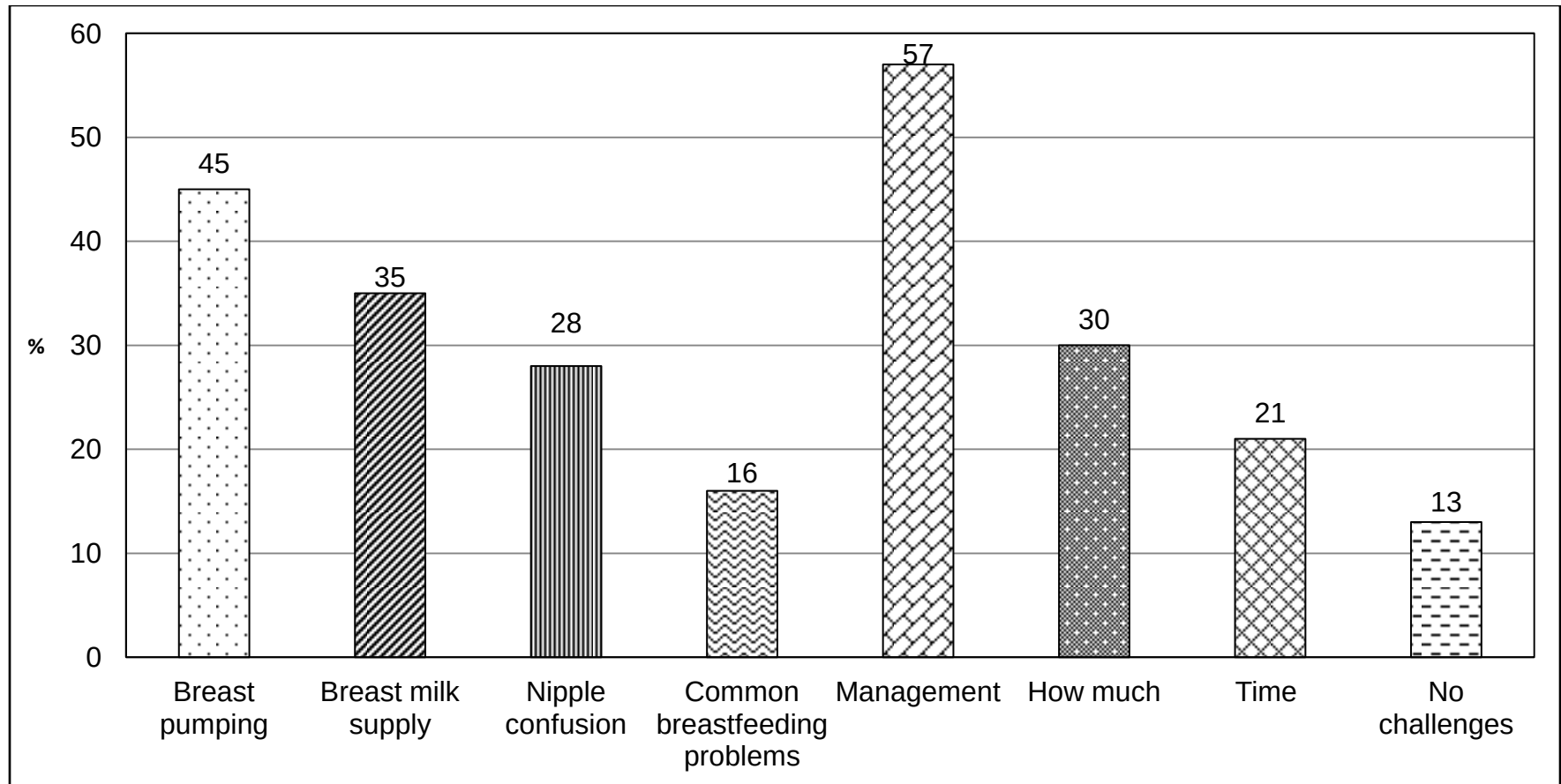


Figure 8. Subcategories of Challenges Identified by Mothers while Breastfeeding Infants with PKU. Common

Breastfeeding Issues = Breast pumping + Breast milk supply + Nipple preference + Common breastfeeding problems;

Breastfeeding & PKU = Management + How Much + Time

Ease of Breastfeeding Infants with PKU

Mothers were also asked the question, “What made it easier for you while breastfeeding your infant with PKU?” The responses were characterized as to the ease of breastfeeding infants with PKU. Four specific questions were asked of these responses: 1) What aspects of general breastfeeding made breastfeeding infants with PKU easy for mothers? 2) What aspects of breastfeeding and PKU made breastfeeding infants with PKU easy for mothers? 3) What supports made breastfeeding infants with PKU easy for mothers? 4) Did some mothers have nothing that made breastfeeding infants with PKU easy? Five categories were identified: 1) *General Breastfeeding*, 2) *Management*, 3) *Support*, 4) *Easy Child*, and 5) *Nothing Easy*. See Figures 9 and 10.

General Breastfeeding. Within ease of general breast feeding infants with PKU, General Breastfeeding was the most frequent response category ($n = 21$, 31%). For this category, three subcategories were identified: 1) *breast pump*, 2) *benefits of breast milk*, and 3) *naturalness of breastfeeding occurring between the infant and mother*. Given that the majority of mothers with infants with PKU cannot exclusively breastfeed due to the need to provide Phe-free supplementation, several responses focused on the *breast pump* ($n = 22$, 31%). *Breast pump* referred to the mechanical device that provided gentle suction for the removal of breast milk from breasts when infant feeding was not possible for various reasons. Having an appropriate electric, hospital-grade breast pump for long-term use contributed to the ease of breastfeeding infants with PKU. Remarks included the following: “An electric pump that made it quicker and easier to pump after a bottle of Phe-free formula feeding,” “I

purchased an electronic dual breast pump, which made life a lot easier. I was able to see exactly how much I was giving my daughter, plus keep up my supply when she required more Phe-free formula,” “A very good quality double electric pump. This made pumping faster and more efficient, keeping my milk supply good for the first four months,” and “Having a dual, electric, medical grade breast pump, which made life a lot easier.”

Many of these responses ($n = 16$, 25%) reflected the mothers’ knowledge of the *benefits of breast milk*, which referred to evidence of the advantages of breastfeeding and breast milk over alternatives. Comments ranged from simply “Nothing, other than knowing the benefits of breast milk” to “Knowing that breast milk was so good for him and that it is naturally lower in Phe than standard infant formula,” “Knowing I was providing her with breast milk for health and still keeping tight control over Phe intake kept me going,” and “Knowing that it was the best option for my baby to get as much of my milk as his body could tolerate.” Other mothers noted, “The benefits of breast milk outweighed the hassle of pumping my milk. Great bonding experience, health benefits, plus cost saving versus buying a separate formula,” and “I had read that those with PKU have 15 points lower IQ scores and breastfeeding on average increases IQ scores by 11 points so I thought it might help to cancel the impact of PKU.”

The responses also revealed a *naturalness of breastfeeding* occurring between the infant and mother ($n = 14$, 21%). Part of the *naturalness of breastfeeding* was from previous maternal success breastfeeding infants without PKU, resulting in confidence with breastfeeding in the context of PKU, and the intense attachment that develops between mothers and their babies. Comments included “having a

daughter without PKU first” to more detailed responses “I have an older non-PKU child and breastfed him, so I was more comfortable with the situation,” and “I’ve had experience breastfeeding my older children. If not for that experience, I doubt I’d have been able to continue.” In addition, mothers commented on the intimate relationship between infant and mother when breastfeeding. Mothers reported “Breastfeeding is a tremendous bonding experience for any mother and child,” and “It was also a wonderful bonding experience that cannot be replaced by bottle feeding.”

Management. Several mothers focused on Management ($n = 18$, 27%). *Management* referred the need to maintain Phe levels as desired by the metabolic team including modifications to frequency of breastfeeding, either increasing frequency when Phe levels were low or decreasing frequency when Phe levels were high. At times, *management* was interwoven between breastfeeding and PKU management. Mothers wrote, “My infant’s level dropped too low when he was 2 months old, and I was allowed to return to nursing. My son was much happier and I was much happier,” and “Everything was made easier once I was finally allowed to breastfeed in between metabolic formula feedings rather than having to pump during the day until all of the metabolic formula had been consumed.” One mother provided this description:

When my son was about 3 months old, I was able to breastfeed 6 times a day. Since my son was only feeding 7 times a day (including once in the middle of the night), this meant that I was exclusively breastfeeding him during the day. This lasted for 3 weeks (until his levels went up!) and it was a magical time—my milk supply really kicked in and both my son and I finally made breastfeeding work for both of us. I looked forward to feeding my son and didn’t dread the sore nipples and feeling like I wasn’t able to satisfy my son’s hunger.

Support. Another frequently identified category was Support ($n = 17$, 25%).

Mothers received support from two main sources, *professional* and *social*.

Professional support came from healthcare providers ($n = 14$, 21%), while partners, family, and friends ($n = 7$, 11%) provided *social support*. For example, some mothers wrote, “Support and encouragement from his metabolic dietitian,” “Encouraged and support from friends, and other nursing mothers,” “The support of our dietitian. She was very positive about being able to continue breastfeeding, and encouraged me to try,” and “Encouragement and support from pediatrician, genetics team, friends, mother, husband, midwife, lactation consultant, and other nursing mothers.”

Easy child and nothing easy. A few mothers described ease of breastfeeding their infant with PKU due to having an Easy Child ($n = 5$, 7%) referring to an infant who is not difficult to care for or to parent. For instance, several mothers reported, “My son made it easy. He switched from bottle to breast very easily, so I was lucky,” “My son was a very easy baby, and after the initial month I had no real problems breastfeeding him,” and “She would take either breast or bottle without fuss.”

In comparison, there were a number of mothers whose responses revealed there was not anything easy about breastfeeding infants with PKU indicating *Nothing Easy*. Several mothers wrote simply: “Nothing.” Others mothers wrote: “There was nothing easy about it” and “I don't think it was easy” ($n = 7$, 11%).

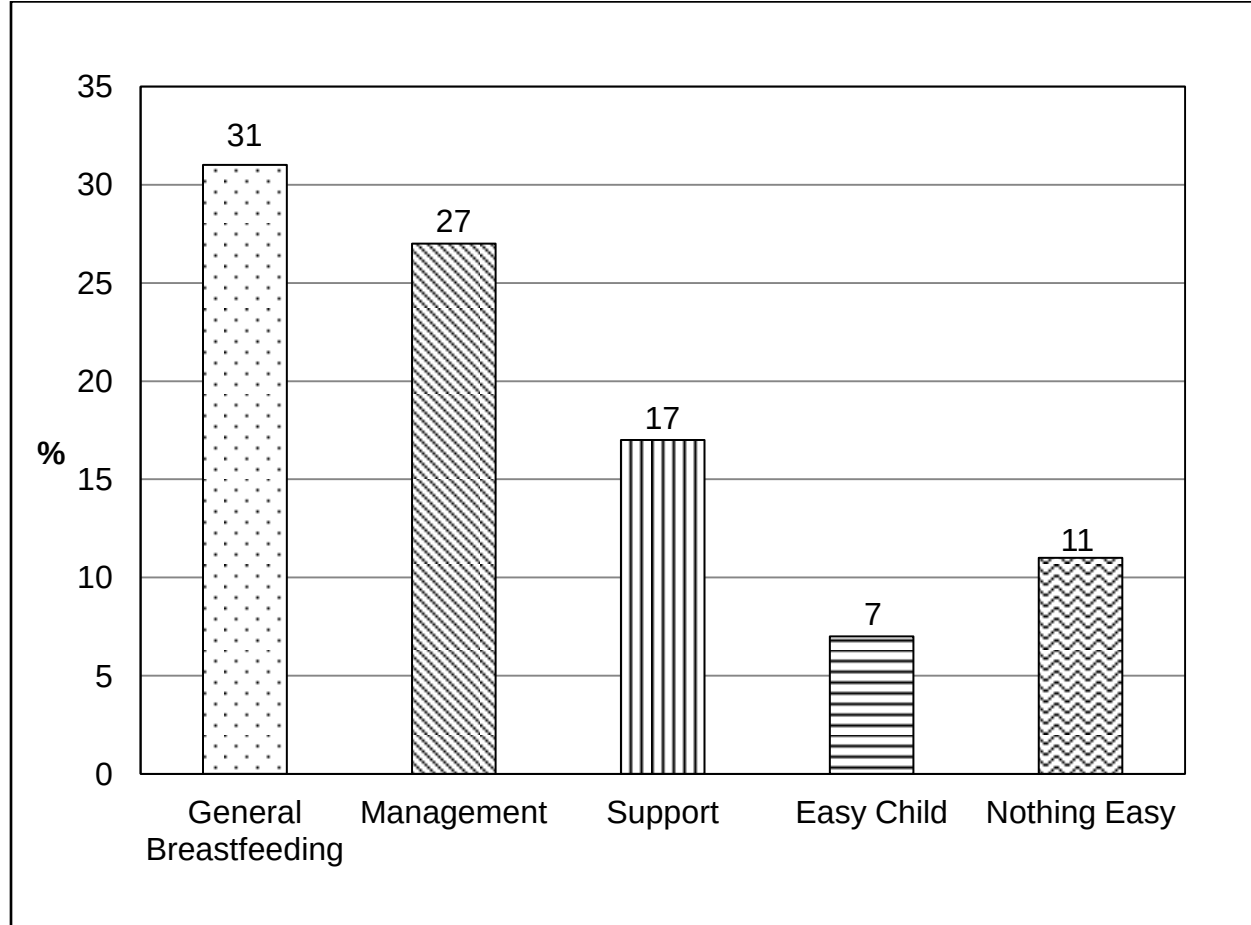


Figure 9. Ease Identified by Mothers while Breastfeeding Infants with PKU.

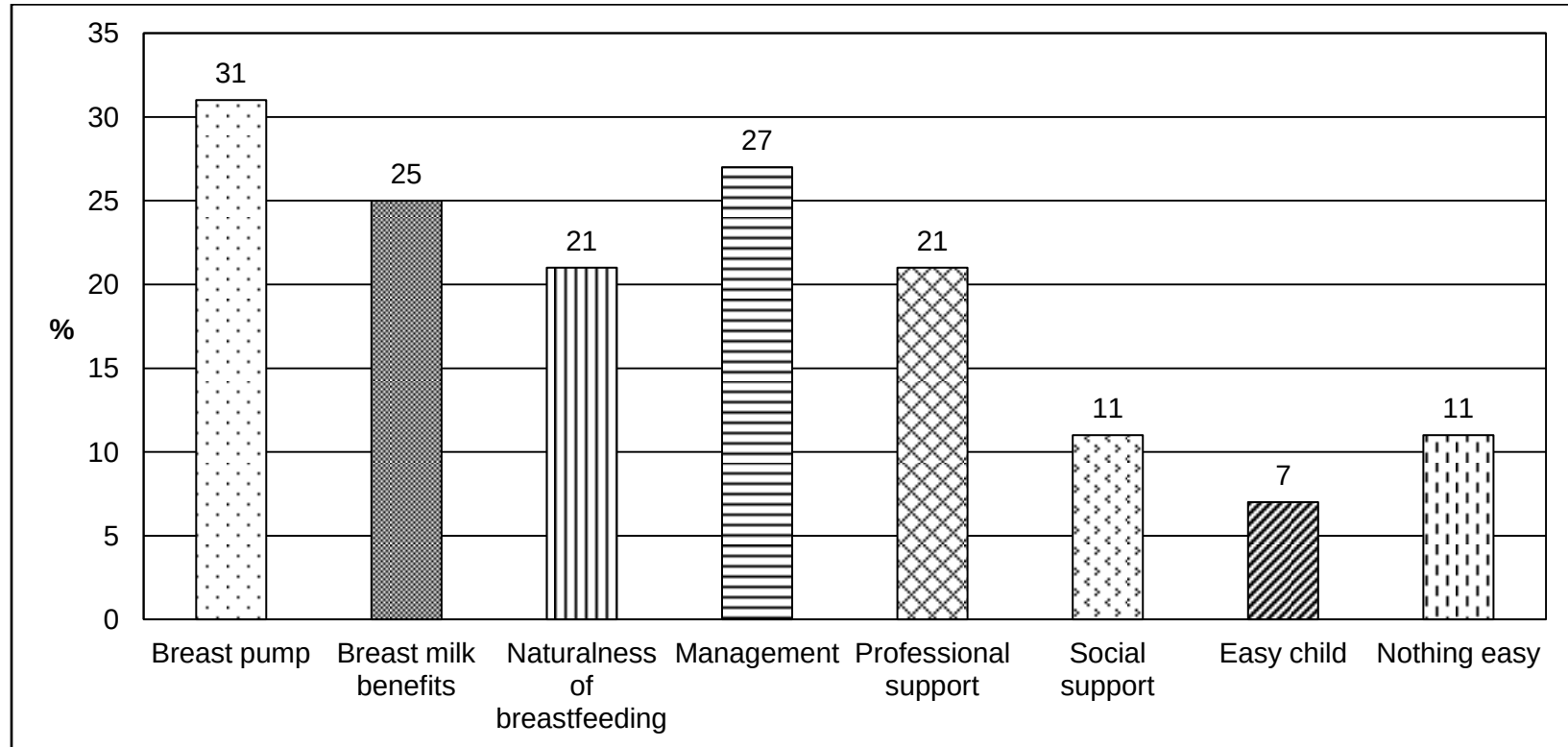


Figure 10. Subcategories of Ease Identified by Mothers Breastfeeding Infants with PKU.

Breast pump + Benefits of Breast Milk + Naturalness of Breast Feeding = General Breastfeeding;

Professional Support (healthcare providers) + Social Support (partner, family, and friends) = Support.

Help while Breastfeeding Infants with PKU

The third open-ended question on the Internet survey asked: “What would have helped you while breastfeeding your infant with PKU?” The set of responses was characterized as help while breastfeeding infants with PKU. Two specific questions were asked of these responses: 1) What forms of help did mothers’ breastfeeding infants with PKU desire? 2) Did some mothers not need any help in breastfeeding infants with PKU? From the responses about mothers’ experiences regarding Help while breastfeeding, four categories emerged: *Lactation Help, Management, More Support, and Nothing Helped*. See Figures 11 and 12.

Lactation help. Within the category of Lactation ($n = 27$, 48%), four subcategories were identified: 1) *breast milk supply*, 2) *PKU and breastfeeding resources*, 3) *breast pump issues*, and 4) *lactation consultant*. The most frequently identified subcategory within Lactation was *breast milk supply* ($n = 10$, 18%) referring to the ability of the mammary glands to produce adequate amounts of breast milk due to galactopoiesis. This was mentioned as an aspect for which mothers desired to have help while breastfeeding. Mothers wrote “if my body would have kept producing milk,” “better milk production,” and “more support with maintaining milk supply from a professional.” Another issue that mothers identified was a lack of *breastfeeding resources specific to breastfeeding infants with PKU* ($n = 9$, 16%). Mothers wanted both reading material, such as web pages, and written information, as well as the opportunity to have one-on-one communication with another mother who had breastfed her infant with PKU. Comments included “More resources are always helpful,” “A web site with FAQ or where I could ask

questions about breastfeeding with PKU,” “Being in touch with other PKU mothers and hearing what worked for their child,” “Having another mom who had a child with PKU and learning from her experiences,” “Meeting a PKU mother who had already gone through it would have helped,” and “Speaking with other moms currently or recently finished breastfeeding a PKU infant.”

The last two needs identified by mothers that would help with breastfeeding infants with PKU were *breast pump issues* ($n = 8, 15\%$), and referral to *lactation consultants* ($n = 8, 15\%$). *Breast pump issues* included an appropriate electric breast pump for long-term pumping to understanding the importance, reasons, and benefits of breast pumping when an infant is unable to exclusively breastfeed. Comments included, “getting a medical grade breast pump earlier,” “a better pump,” “knowing the importance of regular pumping at the beginning to maintain a better breast milk supply,” and “knowledge of the importance of pumping every time I didn't breastfeed to keep my milk supply high.” Last, mothers strongly voiced the specific need for a *professional lactation consultation*, who is a healthcare professional and provides education and management related to breastfeeding, after diagnosis of PKU, to provide knowledge and guidance on issues that are important to mothers breastfeeding infants with PKU. However, for these mothers, specific issues were heightened due to the management of PKU that excludes the option to exclusively breastfeed. One mother wrote, “Lactation consultant from day 1 of PKU diagnosis. This should be automatic!” Other mothers comments included, “Meeting with a lactation consultant,” “I should have talked to a lactation

consultant more. I was young and did not ask as many questions as I should have,” and “A knowledgeable lactation consultant.”

Management. Another focus within the Help responses was Management ($n = 11$, 20%) which had two subcategories, *PKU management*, and “*how much*.” *PKU management* ($n = 6$, 11%) addressed maintaining appropriate Phe levels, the impact of Phe levels, and subsequent changes in breastfeeding frequency or modification of breastfeeding plan. At times, these were interwoven as when Phe levels were elevated, breastfeeding frequency decreased and vice versa. One mother reported, “Allowing me to breastfeed throughout the day rather than having to wait until after all metabolic formula was consumed.” Other mothers described:

I often felt the changes to my son’s feeding schedule did not take into account the impact to my body. I know that my son’s treatment is all about him, but I think the clinic needs to consider the impact to the mother when they make changes to the breastfeeding schedule . . . It’s very hard to change the number of breast-feeds EVERY week. Just when you get into a routine and have the right amount of milk coming in, the instructions change. It’s very frustrating as a mother. I wanted to provide my son with the nutritional milk my body made for him, but it was a constant struggle.

If I was able get his formula in a day in during one or 2 feedings and breastfed exclusively during the other feedings, rather than having to offer a bottle first before every feeding to give formula before breast milk.

The other subcategory of Management, “*how much*,” referred to the inability to determine *how much* breast milk the infant with PKU had consumed while suckling at breast leading to the concern of *how much* Phe that the infant obtained from the breast milk. Mothers ($n = 4$, 7%) struggled being able to determine “*how much*” breast milk infants suckled at breast and to determine the amount of Phe in breast milk as illustrated by these comments, “Since it was not clear how much breast

milk she was receiving at each feeding, I was worried about her levels,” and “In hindsight, I wish I was more relaxed and not so strict about when he could or couldn't breastfeed. I'd often stop him mid-session because I worried he was taking in too much Phe.”

More support. Another focus of Help was *More Support*. During this time, mother reported a lack of support ($n = 13$, 24%) and their desire for more support. The category of More Support had three subcategories: 1) *professional support*, and 2) *social support*, and 3) *encouragement to continue breastfeeding*. Of these three subcategories, *professional support* from healthcare providers ($n = 9$, 16%) was the most frequently identified: “I really wish that the dietitian would have been more compassionate,” and simply “more support from clinic.” In comparison to needing more *professional support*, fewer mothers reported a lack of *social support* from partners, family, and friends ($n = 7$, 11%). Comments included simply “spousal support” to “I felt like it was difficult to feed her in public and even sometimes at home because my parents and in-laws did not breastfeed. They were less supportive when things were more challenging.” The third subcategory was *encouragement*, the act of inspiring that spurs mothers to continue breastfeeding or breast pumping. Mothers ($n = 4$, 6%) wanted more encouragement as illustrated by these comments: “more encouragement,” and “I felt very blessed with my experience and the support I received, but I feel others in my situation are too quickly told to bottle feed and not given the option or encouraged to breastfeed.”

Nothing helped. The last category within the Help responses was Nothing Helped ($n = 4$, 6%). However, these responses revealed positive maternal experiences breastfeeding infants with PKU with no identified problem or additional help needed. For example, mothers wrote: “Nothing. It was fulfilling and enjoyable,” and “Nothing . . . it wasn’t a problem.”

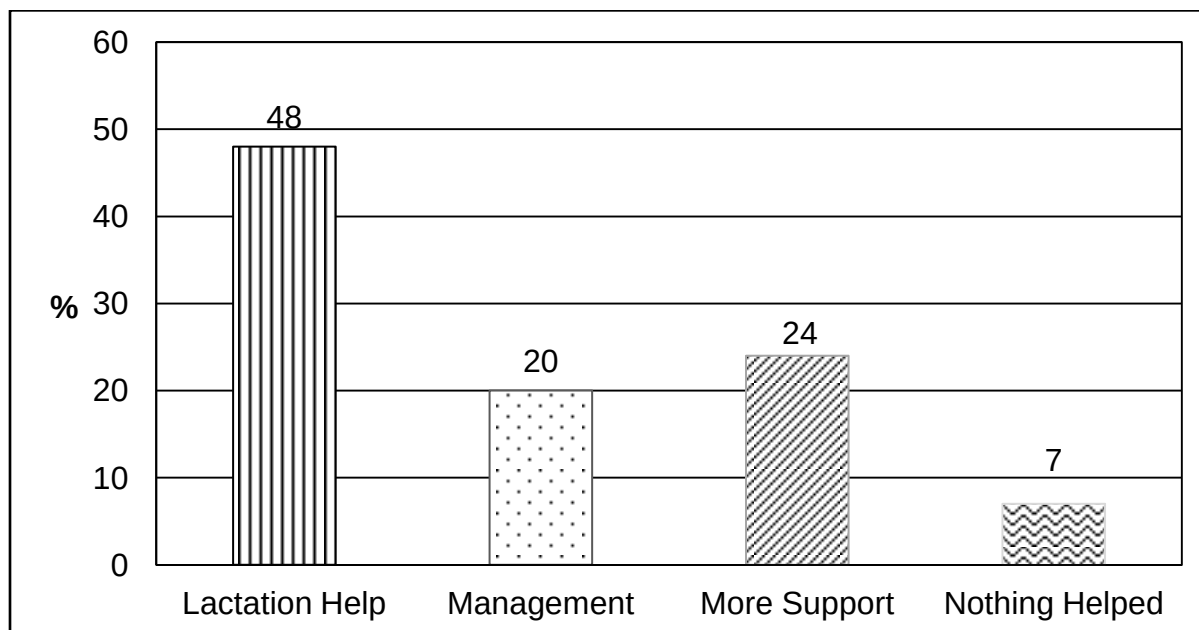


Figure 11. Help Identified by Mothers while Breastfeeding Infants with PKU.

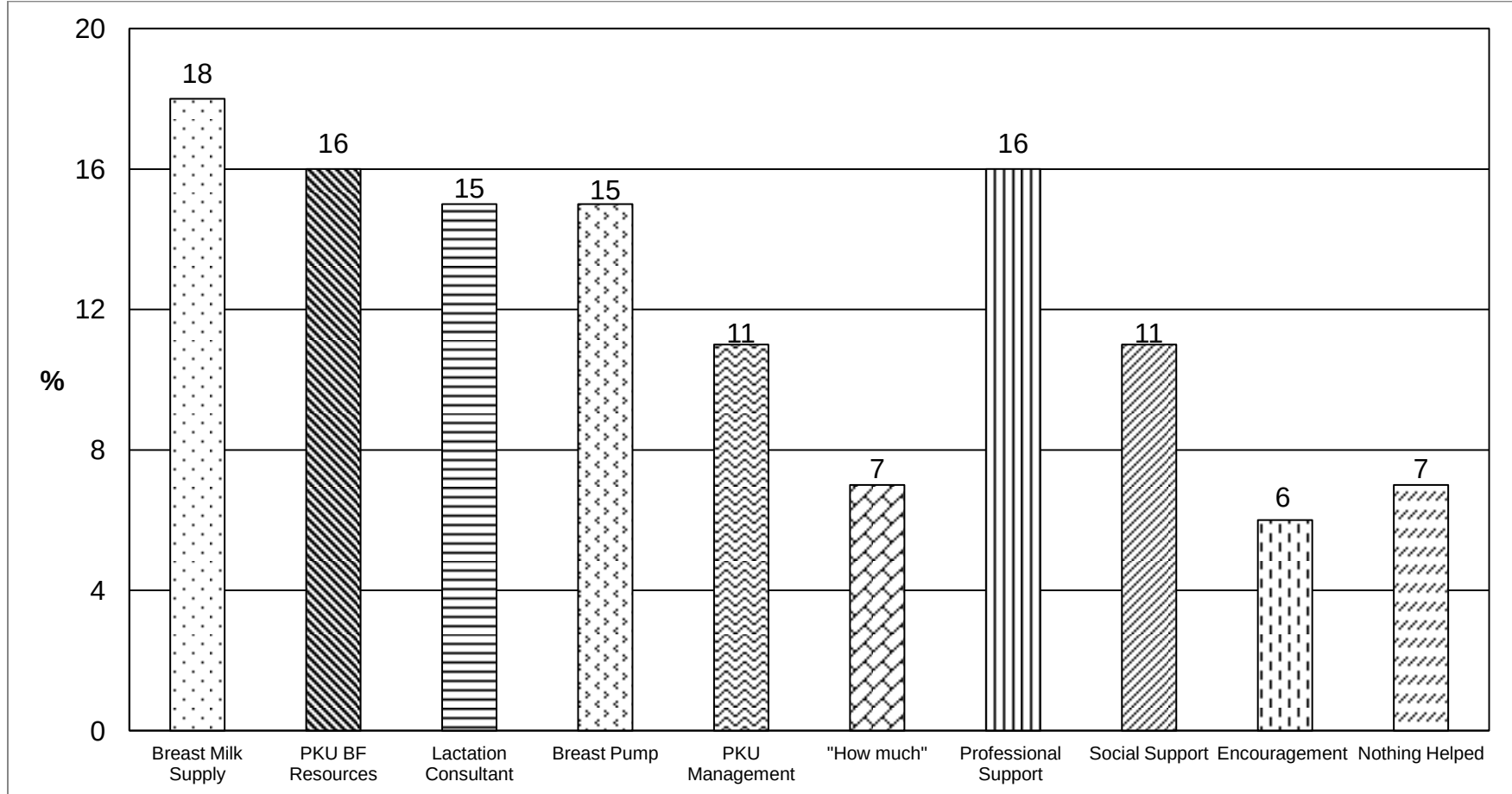


Figure 12. Subcategories of Help Identified by Mothers while Breastfeeding Infants with PKU.

Breast milk supply + PKU BF resources + Lactation consultant + Breast pump = Lactation Help. PKU Management +

"How much" = Management. Professional support + Social support + Encouragement = More Support.

Support for Breastfeeding Infants with PKU

Mothers were asked a fourth open-ended question on the Internet survey: “What support did you receive to continue breastfeeding your infant with PKU?” The responses were characterized as support for breastfeeding infants with PKU. Three specific questions were asked of this set of responses: 1) From whom did mothers breastfeeding infants with PKU receive support? 2) What form of support (emotional, esteem, informational, and tangible) did mothers receive while breastfeeding infants with PKU? 3) Did some mothers receive no support while breastfeeding infants with PKU?

After reading the set of responses addressing support for breastfeeding infants with PKU, it was not possible to clearly determine what form of support mothers received given to their answers identified who provided support. Mothers received support from many individuals including dietitians, medical providers, partners, family, and friends. Thus, three major categories of Support were identified: *Professional*, *Social*, and *No Support*; see Figures 13 and 14.

Professional. Mothers most frequently identified receiving support from healthcare professionals ($n = 59$, 58%), which included four subcategories: *dietitians*, *medical staff*, *lactation*, and *clinic*. Of those reporting positive professional support ($n = 59$, 58%), nearly one-half ($n = 27$, 46%) reported that *dietitians* specifically had been supportive while they were breastfeeding their infants with PKU. Comments included, “My dietitian continued to support me via phone and email. She explained and clarified many times what I needed to do and why. She was patient and kind,” “My dietitian continued to educate and encourage

me to breastfeed for as long as I could,” and “I spoke with our dietician weekly in the early days. She always encouraged me and told me how well my daughter was doing and how low her blood Phe levels were.”

Mothers also identified *medical staff* ($n = 10, 16\%$), such as physicians, nurse practitioners, and physician assistants, as providing support while breastfeeding infants with PKU. Mothers reported, “My nurse practitioner was very supportive. She had her children and knew how important that was for me,” “Our doctor was very supportive of breast milk feeding, due to its lower content of Phe,” and “Her geneticist, dietician and pediatrician were all strong supporters as well.” The third healthcare provider identified as being supportive of breastfeeding were *lactation consultants* ($n = 8, 14\%$), who provide education and management related to breastfeeding. Comments included, “My birth doula was a lactation consultant and coached me into an easy, near-effortless and painless breastfeeding experience,” and “I also got support from the lactation consultant at the hospital where my son was born. The lactation consultant suggested that I take some herbs, blessed thistle and fenugreek, to try to help my milk come in since I wasn’t exclusively breastfeeding.” In addition, some mothers simply reported their *clinic* ($n = 14, 24\%$) had provided them with support. Mothers stated, “At monthly visits to the clinic, I was encouraged to continue pumping,” “The metabolic clinic encouraged breastfeeding, which was fantastic as I really felt this was important in raising a newborn,” and “The clinic was very supportive of breastfeeding.”

Social. The second category of Support that mothers reported was social or lay support ($n = 31$, 31%), which combined three subcategories: *partner*, *family*, and *friends*. *Partners* referred to someone that the mother is united or associated with in an active or sphere of common interest, such as husband, spouse, domestic partner, or significant other. *Family* was defined as any group of people related biologically, emotionally, or legally to the mother who she acknowledged as significant for her well-being such as grandparents. *Friends* were a form of interpersonal relationship that may range from fleeting to enduring identified by the mother. The interpersonal relationships are formed in context of social, culture, and other influences.

Within social support, *family* support ($n = 13$, 42%) was the largest of the three subcategories identified by mothers. For example, one mother expressed delight with a gift from her parents that aided in the ability to store expressed breast milk. "My parents bought us a large freezer!" However, many responses from mothers did not identify only one social support but were a mixture of support including family along with partners ($n = 11$, 35%) and friends ($n = 7$, 23%) as illustrated by these comments, "My husband was a huge support, along with my family and friends who advocate breastfeeding," "My husband, family and friends all strongly support breastfeeding," and "My husband and maternal family were super-happy that despite her diagnosis I could still breastfeed her. Most of our friends were all long-term breastfeeders. So, great support."

No support. The third most frequently identified Support category was No Support ($n = 11$, 11%), which frequently was simply answered as "None."

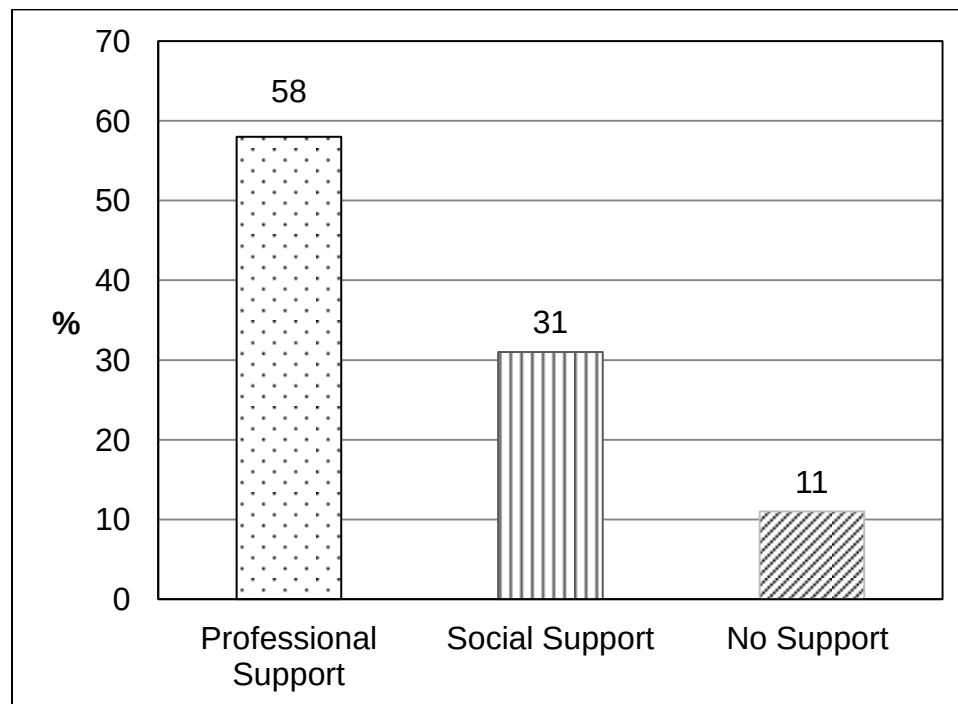


Figure 13. Support Identified by Mothers while Breastfeeding Infants with PKU.

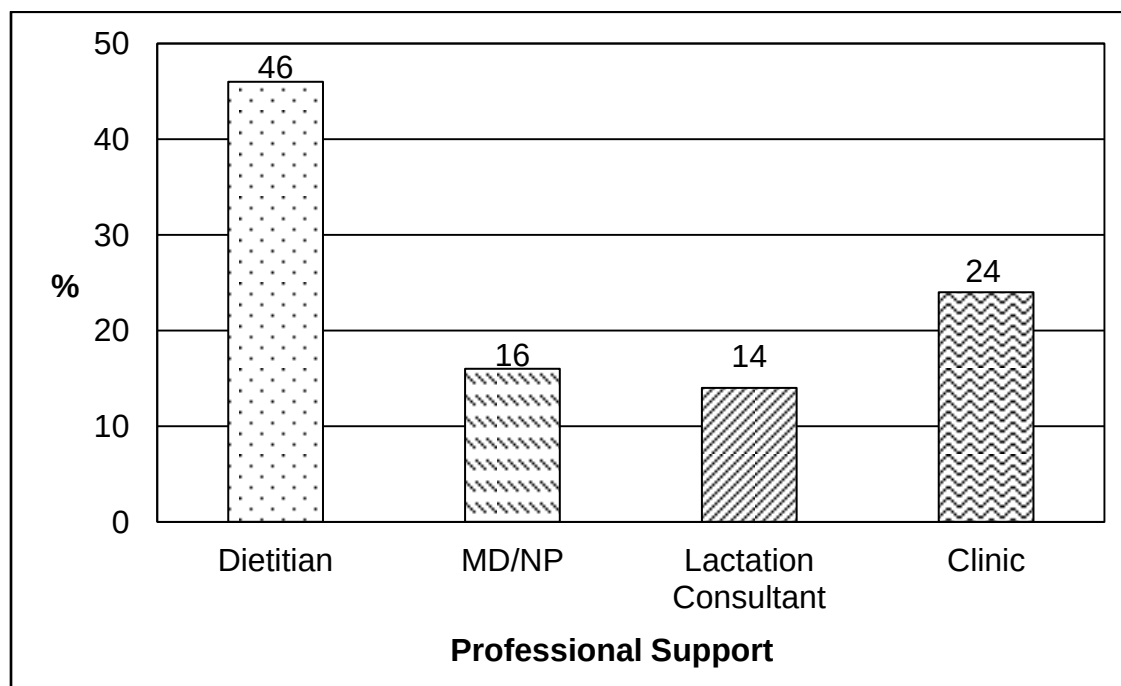


Figure 15. Professional Support Subcategories Identified by Mothers while Breastfeeding Infants with PKU. MD/NP = Medical physician/nurse practitioner

Breastfeeding and PKU: The Maternal Experience

The categories and subcategories from the four open-ended sets of responses (challenges, ease, help, and support) were examined to identify 1) factors that facilitated breastfeeding infants with PKU, 2) barriers that impeded breastfeeding infants with PKU, and 3) described mothers' strategies to obtain the necessary skills to care for their breastfeeding infants with PKU. This resulted in the development of Figure 16, Breastfeeding and PKU: The Maternal Experience.

After coding the open-ended responses from the Internet survey, the maternal experience of simultaneously managing breastfeeding and PKU began to develop and revealed three overarching constructs: Facilitators, Barriers, and Strategies. Facilitators were those identified aspects that encouraged, either directly or indirectly, mothers to breastfeed their infants with PKU and thereby enhanced their breastfeeding confidence. In contrast to facilitators, there were barriers that diminished breastfeeding confidence. Barriers were those identified factors that obstructed or impeded mothers breastfeeding. Strategies were those adaptations that mothers used to obtain the necessary skills to care for their breastfeeding infants. Under the three overarching constructs of Breastfeeding and PKU: The Maternal Experience (Figure 16), the major categories and subcategories generated from the open-ended responses from the Internet survey were developed and are described below.

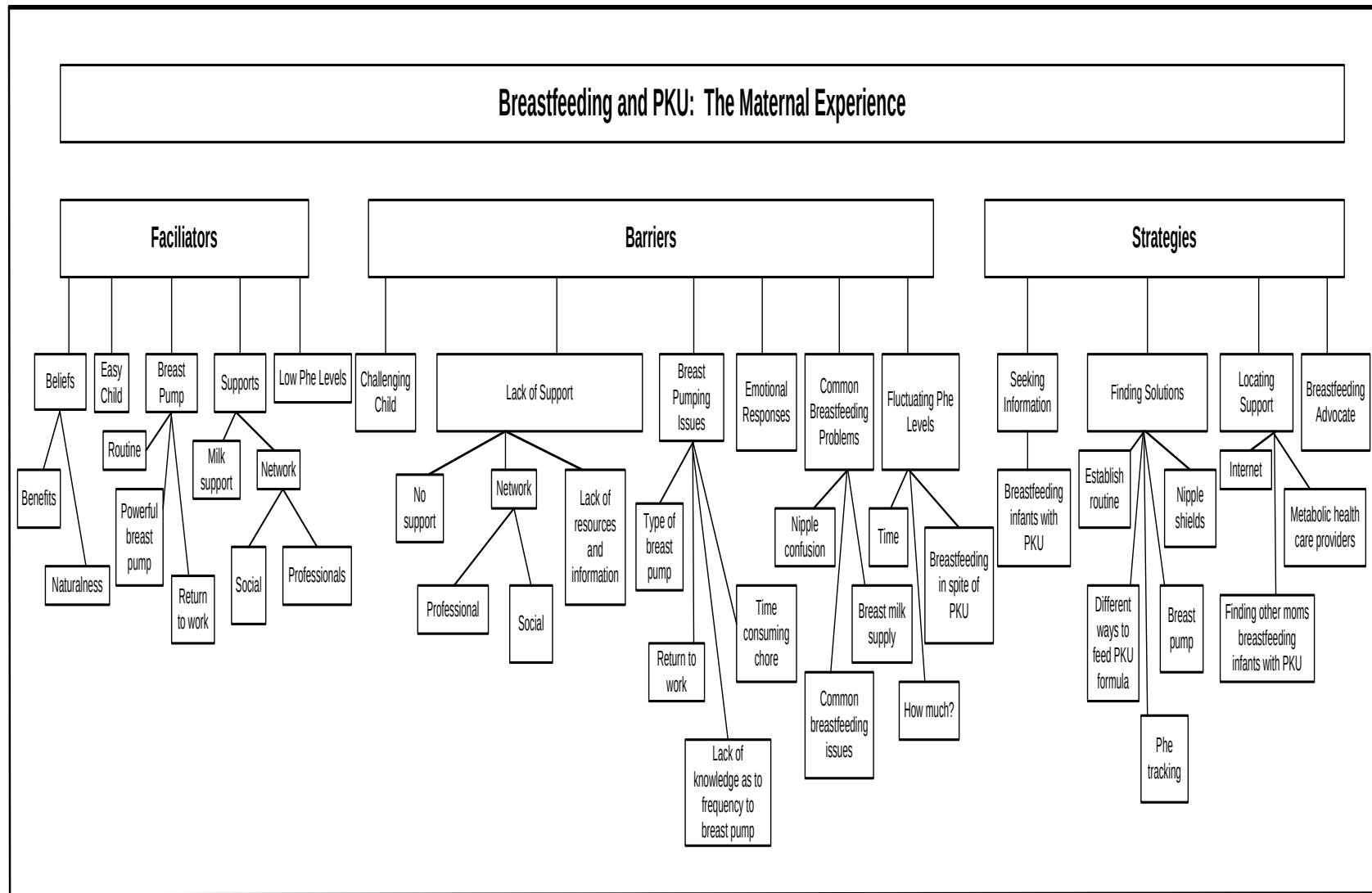


Figure16. Breastfeeding and PKU: The Maternal Experience

Facilitators of Breastfeeding Infants with PKU

A review of the literature revealed no information regarding what facilitators aid mothers in breastfeeding their infants with PKU, and therefore did not provide guidance for analysis. From the open-ended responses and subsequent interviews, specific facilitators emerged as factors that were helpful to breastfeeding infants with PKU. These were *Beliefs*, *Breast Pump*, *Easy Child*, and *Supports*.

Beliefs. Within the category of Beliefs, there were two sub-categories: 1) *benefits* and 2) *naturalness* of breastfeeding. Mothers believed there were *benefits* of breastfeeding for infants, mothers, and families. Mothers identified the benefits of breast milk for the infant most frequently: “Knowing that even though she had to drink a synthetic formula that tasted bad every day, she could still have the very best that nature has to offer especially for her,” and “Knowing that breast milk was so good for him and that it is naturally lower in Phe than regular infant formula.” Also, mothers commented on the lower content of Phe in breast milk compared to standard commercial infant formula and the higher IQ scores for children who had been breastfed. Comments included, “Statistically I have read that those with PKU have 15 points lower IQ scores and breastfeeding on average increases IQ scores by 11 points so I thought it might help to cancel out the impact of PKU,” “I also read it can help their IQ so that is motivating for me,” and “I was reading that breast milk can help with their IQ. . . and I know that is true, but because I read it, I just thought why stop, and I can keep going.”

Some of the mothers identified the cost-saving benefits of breastfeeding for their families. Mothers wrote, “It was a lot cheaper than buying formula and better for her,” “Great bonding experience, health benefits, plus cost saving versus buying a separate formula,” and “Having the money left that we didn’t have to spend on regular formula.”

In addition, mothers commented about the *naturalness* of breastfeeding such as “It was what I did with my other child. It felt normal. It was something I enjoyed and felt good about doing.” Within the naturalness of breastfeeding, mothers made several references to bonding and attachment while breastfeeding: “The bonding experience mother and child gain, particularly in the face of such tragic news, cannot be minimized. It is something you will never forget doing with your child,” “I loved breastfeeding my baby. The intimacy was incredible,” and “It was also a wonderful bonding experience that cannot be replaced by bottle feeding.”

Breast pump. Within the breast pump category of Facilitators, there were three subcategories: 1) *routine*, 2) *an appropriate electric breast pump*, and 3) *return to work*. Mothers identified breast pumping as an important component of maintaining their breast milk supply as their infants were unable to exclusively breastfeed. Establishing a breast pumping *routine* was critical to maintaining their breast milk supply as some infants never breastfeed, but received expressed mothers’ milk while others breastfed from only once or twice a day to more than six times per day. Mothers expressed a need for routine breast pumping by answering “Pumping regularly,” and “Breast pumping also made it more regular,

so it was just a habit.” Also, having an *appropriate electric breast pump* was vital to maintaining an adequate breast milk supply: “Having a medical grade breast pump made it easier to breastfeed infant,” and “I purchased an electronic dual breast pump, which made life a lot easier.” Upon *returning to work*, employer accommodations to breast pumping helped mothers to continue to breastfeed infants with PKU: “My employer was very accommodating when I would need to [breast] pump during the work day” and “Permission from my employer to pump as needed during the work day.”

Easy child. Several mothers identified their child as being “a very easy baby.” Comments included: “My son was a very easy baby, and after the initial month, I had no real problems breastfeeding him,” “My son made it easy—he switched from bottle to breast very easily so I was lucky,” and “She would take either breast or bottle without fuss.”

Support. Another category of Facilitators was Support with three subcategories: 1) *social support*, 2) *professional support*, and 3) *milk support*. *Social support* included support from partners, family, and friends. Supportive partners were viewed as assets to successfully breastfeeding as were other family members and friends. Mothers stated, “My husband was a huge support, along with my family and friends who were breastfeeding advocates,” “An extremely supportive husband and family allowed me to pump every time my son took a bottle in order to keep up with the demands of breast milk needed to add to the [Phe-free] formula,” and “My husband was a huge support, along with my family and friends who advocate breastfeeding.”

Mothers with infants with PKU necessarily interact with a large number of *professional* healthcare providers who provided support. These include dietitians and health professionals such as nurse practitioners, physician assistants, and physicians at clinics. Their support helped shape mothers' perception of the value of breastfeeding and breast milk for infants with PKU. Mothers repeatedly reported about specific healthcare providers: "Dietitian was pro-breastfeeding, and helped and encouraged me along the way with breastfeeding," "My nurse practitioner was very supportive," and "Our doctor was very supportive of breast milk feeding, due to its lower content of Phe." Other mothers reported the metabolic clinic or team was supportive: "The metabolic clinic encouraged breastfeeding which was fantastic as I really felt this was important in raising a newborn," and "My son's clinic was very supportive of my decision to breastfeed my son for as long as possible." Lactation consultants also provided support. One mother wrote, "I had a lactation consultant in town that was very helpful." Another mother described, "I got support from the lactation consultant at the hospital where my son was born. The consultant suggested that I take some herbs, blessed thistle and fenugreek, to try to help my milk come in since I wasn't exclusively breastfeeding."

Another aspect of support was the ability to provide expressed breast milk to another infant as in *milk support*. One mother reported supplying an infant who for medical reasons was unable to breastfeed and was allergic to formula. She provided her excess expressed mothers' milk for six months. She revealed:

I was often consumed with dealing with PKU. I didn't know what to do with this "white gold." I didn't want to throw it out. I couldn't admit that I was

pumping for nothing . . . but what do you “DO with it?” I had looked into donating at a milk bank, but I needed to pay my own shipping costs. They suggested it might be around \$75-100 per month based on how much I was pumping. So I called the La Leche League leaders and asked if they knew of anyone desperate. They guided me to an online yahoo group. The next day, an urgent request came in. There was a two-week old baby in the hospital—mamma couldn't produce enough and baby had a bad reaction to formula. I was standing in my kitchen reading the email and I started to cry. I started to pray. I prayed that this baby might use my milk because I NEEDED someone to. I prayed that there was a reason that my daughter was suffering that her PKU would mean that there was enough milk to help another baby in need. Within a week or so I gave my first freezer full. I continued to pump for this family for 6 months and helped try to recruit other donor mammas. They were a wonderful family and I felt compelled to try to help. After all I know how it feels to have to rely on someone else to feed your baby, it is one of the worst feelings in the world. This made it easier to breast pump because I wasn't tossing the expressed breast milk.

Barriers to Breastfeeding Infants with PKU

In the literature, little is known about what barriers affect mothers' breastfeeding infants with PKU and no guidance was found to guide this analysis. From the open-ended survey responses and interviews, the mothers identified specific barriers to breastfeeding infants with PKU: *lack of support, lack of resources and information, breast pump issues, fluctuating phe levels, common breast feeding problems, and emotional responses.*

Lack of support. Lack of support was described as a barrier to breastfeeding infants with PKU. Within the category of lack of support, there were two subcategories: 1) *social support*, and 2) *professional support*. For a few mothers, there was a lack of social support from their partners to breastfeed. One mother simply stated one challenge was “lack of spousal support” and another mother reported “My husband was frightened of taking care of our daughter with PKU.” In addition, when mothers were struggling with breastfeeding, they

perceived a lack of support from family members. At these times, family members expressed to mothers the perception that breast milk was inadequate or unnecessary for infants and that the introduction of standard commercial infant formula feeding from a bottle would meet their infant's needs: "Everyone involved told me that they thought what I was doing good, but if I ever talked about having trouble managing it all, I was quickly told that it was alright to switch to formula," "I felt like it was difficult to feed her in public and even sometimes at home because my parents and in-laws did not breast-feed. They were less supportive when things were more challenging," and "My husband's family is all formula-feeders. To them, I think, they probably thought that I was probably crazy for going to such extremes to give him breast milk." Mothers also identified a lack of other mothers who could relate to the inability to exclusively breastfeed a term healthy-looking infant. Mothers reported: "I didn't know anyone that had another child with PKU that breastfed," and "I couldn't find a lot of information from other mothers who had breastfed their newborns [with PKU] and that would have been helpful."

Another support barrier that mothers identified concerned *healthcare professionals*. As mothers asserted:

Medical professionals do not get how difficult this first year of life can be for mothers of a PKU child. Feeding a child with PKU becomes a part of everything that you do: When you can go out, where and how far. It is very, very challenging.

I was essentially ordered to stop breastfeeding immediately once my baby turned six months when I was supposed to start providing solid foods. It had been my understanding that a baby should be weaned gradually off of breastfeedings rather than such an abrupt stop. When I voiced this concern, our dietitian at the time asked me whether I was breastfeeding

for my sake or for my child's. She then seemingly reluctantly gave me a few extra days to wean him. I would have preferred to have continued breastfeeding once solids were introduced at age six months and maybe another three months at least; but, I was not given this option.

I really wish that dietician would have been more compassionate. The diagnosis is hard enough, but to add to the mental and emotional stress of a new mom who wanted to breastfeed was, in my view, just cruel.

Mothers cited conflicting advice from health professionals as a source of frustration. One mother described, "The initial clinic that we attended was very poor with breastfeeding support. Also, they had three dietitians, which was highly confusing as each had different advice." Mothers also expressed frustration with trying to locate assistance for increasing their diminished breast milk supply. One mother described the interaction between the physicians in the clinic:

During the first few months our baby's levels would fluctuate, and the doctors made it clear that this was partly because I was breastfeeding. That said, they did not openly suggest that I quit. When asked, they did say that breast is best! I had the problem of low production, which required A LOT of pumping during the first weeks. The PKU doctors told me that I did not have enough milk. It was up to me to find a lactation consultant and develop a strategy for getting my production going through pumping.

Other mothers sought assistance with lactation consultants to assist with increasing their breast milk supply but reported frustration in dealing with them due to a lack of knowledge about PKU. One mother wrote said, "I did have a lactation consultant available to me; however, she had no idea what PKU was." Another mother reported the need for "a knowledgeable lactation consultant."

Lack of resources and information. Frequently, mothers cited the *inability to locate resources and information* regarding PKU and breastfeeding as a barrier. This lack of resources pertained to both written and on-line information

breastfeeding infants with PKU. This lack of breastfeeding information was expressed by one mother as simply a need for “more information . . . handouts, list of resources and websites” and another as a need for “a web site with FAQ or where I could ask questions about breastfeeding and PKU.”

Breast pump issues. Within the category of breast pump issues, there were four subcategories: 1) *an inferior breast pump*, 2) *lack of knowledge as to frequency to breast pump*, 3) *time-consuming chore*, and 4) *return to work*. Having *an inferior breast pump* for long-term pumping was a barrier frequently identified by the mothers. The need for a hospital-grade dual electric breast pump to maintain their breast milk supply was best illustrated: “Since I had just a hand pump, this was exhausting work between his metabolic formula feedings,” “a better pump,” and “getting a medical grade pump earlier.”

Another barrier for mothers who were breast pumping was *lack of knowledge regarding the frequency of breast pumping* required to maintain an adequate breast milk supply when an infant is not exclusively breastfeeding. This lack of knowledge led to decreased breast pumping resulting in an inadequate breast milk supply and early termination of breastfeeding: “I was a young mother and did not ask as many questions as I should have. If I would have pumped more, I could have breastfed until she was a year old as I had planned,” “I did not pump like I was instructed to, so my milk supply dropped a lot,” and “knowing the importance of regular pumping at the beginning to maintain a better supply.”

Breast pumping was also a *time-consuming chore*. Mothers described spending hours breast pumping during the day and even at night while partners

bottle fed their infant. This was expressed by one mother: “Keeping motivated to pump was a big challenge.” Another mother reported:

Finding the time and emotional difficulty of pumping, and I tried to pump at feedings when I gave a bottle instead of breastfeeding, but I found that logistically hard to do. It's hard to try to take care of a newborn and pump at the same time.

Many mothers described challenges and barriers when they *returned to work* and tried to breast pump to maintain an adequate breast milk supply. One mother wrote, “I worked and had to pump. I lacked adequate space to do so.” Another mother described her work situation and breast pumping, “If I didn’t have to pump because of work, I would have been able to breastfeed for longer. It was too much working full-time, pumping, breastfeeding, and feeding the baby low-pro formula.”

Fluctuating Phe levels. Another barrier to successfully breastfeeding infants with PKU was fluctuating Phe levels, which had three subcategories: 1) *alternating breast and bottle feedings*, 2) *time*, and 3) *how much*. Frequently, mothers identified fluctuating Phe levels in reference to PKU management entailing the frequent alternating between breastfeeding and bottle feeding due to fluctuating Phe levels. One mother expressed this frustration:

While the clinic was supportive of my decision to breastfeed, I often felt the changes to my son’s feeding schedule did not take into account the impact on my body. I know that my son’s treatment is all about him, but I think the clinic needs to consider the impact to the mother when they make changes to the breastfeeding schedule. It’s very hard to change the number of breastfeeds EVERY week. Just when you get into a routine and have the right amount of milk coming in, the instructions change. It’s very frustrating as a mother. I wanted to provide my son with the nutritional milk my body made for him, but it was a constant struggle.

Another mother expressed similar feelings:

I got frustrated because my son's tolerance would change on a weekly basis—some weeks, I could breastfeed six times a day, others only four times a day. Unfortunately, breast milk works on supply and demand and is not a spigot. It was hard to suddenly reduce the number of breastfeeds as I would be engorged, and even harder to suddenly increase the number of breastfeeds because I wasn't producing enough milk. I often took it very personally that I was not able to produce enough milk to satisfy my son's hunger.

In addition to alternating between breastfeeding and bottle feeding, mothers identified the *time* consuming aspects of breastfeeding, breast pumping, bottle feeding, preparing Phe-free formula, and cleaning breast pump equipment as taxing. For example, one mother reported, "The most challenging part was spending time making formula, bottle feeding, breastfeeding, and pumping breast milk often enough to maintain a decent supply." Another mother described:

One challenge is the time and effort to continue breastfeeding increased as feeding is now done in three steps: Formula from bottle, breastfeed, and pump at each feeding. This coupled with the increased time it takes to clean and refill bottles and clean pumping paraphernalia makes the level of effort more significant than breastfeeding alone was or that formula feeding would be.

Another issue was the concern about "*how much*" breast milk the infant had suckled at breast. One mother noted, "Breastfeeding was difficult because it is not an exact science to know how much Phe is entering your baby." In contrast, mothers expressed their frustrations: "Measuring intake. I was only able to communicate length of breastfeeding sessions," "They would say breast is best, but then ask how much breast milk did she drink? How was I supposed to answer that?" "To make 100% sure on how much Phe my son was getting, I

totally gave up breastfeeding,” and “Not being able to measure how much he was getting while he was on breast and not knowing how that affected his Phe levels.”

Common breastfeeding issues. Within the category of common breastfeeding problems, there were three subcategories: 1) *common breastfeeding problems*, 2) *nipple confusion*, and 3) *breast milk supply*. *Common breastfeeding problems* included cracked and bleeding nipples, mastitis, plugged ducts, thrush, positioning, and poor latch. Mothers noted these were not unique to infants with PKU. For example, two mothers wrote: “I had all of the regular challenges of breastfeeding: plugged ducts and thrush,” and “I had extremely sore nipples that constantly cracked and bled.” Another mother said, “My baby did not have a good suction from the start which had nothing to do with PKU. It was my first baby, it was also all new to me and I didn’t know a lot of things that I now know.”

Another frequent barrier was the issue of *nipple confusion*. Mothers reported the frustration and struggle of going from breast to bottle feedings: “Sometimes she would pull away from the breast and refuse to nurse. At these times, I would give her breast milk from a bottle and she would willingly take it,” “Switching from bottle to breast each feeding . . . as my daughter got older, she only wanted to do one or the other,” and “It was stressful trying to add in the formula when he only wanted to breastfeed. As he got older, he began to refuse to breastfeed,” “My son HATED taking a bottle and would scream at every feeding,” “My son was very picky when it came to the bottle nipple. Dr. Brown’s bottles are the only type of bottle my son would take,” and “Since this was my

second child, I didn't have any real challenges breastfeeding per se. The biggest challenge was getting my daughter to take the Phe-free formula out of a bottle! She hated the bottle!"

The last subcategory under general breastfeeding problems was *breast milk supply*. The majority of the mothers were not exclusively breastfeeding because they had to bottle feed their infants a specific PKU formula that is Phe-free, so that low breast milk supply was a recurring concern: "My milk production decreased. Let-down would take longer, and my baby would become frustrated when nursing and waiting for the milk to let down," "Stress about PKU was contributing to the slowed production of my milk," "My body did not respond well to the varying demand while the clinic was trying to regulate my son's level," and "Having to weigh her to get an accurate measurement. I would free-feed her and her levels would always go up because I would underestimate how much she drank."

Emotional responses. The last category within Barriers was *emotional responses*. Mothers consistently expressed negative emotional experiences or negative feelings, usually directed toward specific aspects of breastfeeding their infants with PKU. These emotional responses included "nervous," "frustrated," "upset," "very disappointed," and "worried." Mothers wrote: "very emotional for me," "So she and I were in tears often," "I was always nervous that he wasn't getting the right amount [of Phe]," and "Since it was not clear how much breast milk she was receiving at each feeding, I worried about her levels."

Strategies Used by Mothers Breastfeeding Infants with PKU

From the open-ended survey responses and qualitative interviews, strategies used by these mothers breastfeeding infants with PKU emerged. These mothers reported several different strategies to simultaneously manage breastfeeding and PKU. These strategies were: *Seeking Information, Finding Solutions, Locating Support, and Breastfeeding Advocate*.

Seeking information. Mothers sought information not only about breastfeeding or PKU but specifically about breastfeeding infants with PKU. Some mothers were fortunate to locate another mother who had or was breastfeeding an infant with PKU and who lived within their geographical area. Many of these connections were made via clinics, but also from networking with other mothers' breastfeeding healthy, term non-PKU infants. This exchange provided mothers with ideas of how to continue breastfeeding against many barriers. For example: "Being in touch with another PKU mother and hearing what worked for her child," and "Other mothers with kids with PKU told me they breastfed for 11 months and that was my goal." Finding a lack of reading materials about breastfeeding infants with PKU, these mothers sought out online material and forums, such as the PKU Listserv, that allowed them to post questions, receive information, and provide the connection to other mothers who had previously infants with PKU to young mothers who were currently breastfeeding like themselves.

Finding solutions. Other strategies mothers used to help them care for their infants included 1) *different ways to feed Phe-free formula*, 2) *breast pump*,

and 3) *nipple shields*. Mothers struggled with bottle feeding Phe-free formula to their infants who preferred breastfeeding. Mothers were creative in coaxing infants to drink Phe-free formula rather than grapple with the infant's preference to breastfeed rather than bottle feed. Even though it took more time, mothers used droppers and syringes to administer Phe-free formula to their infants to avoid having them rail against bottles: "I had to feed him his PKU formula using a dropper because he only wanted to breastfeed. Eventually, he took to using a bottle and breastfeeding," and "I stopped using standard bottles. I used a syringe, the same used for giving medication for babies. It was much slower, but much easier." According to one mother, locating the right bottle nipple was an important solution: "My son was very picky when it came to the bottle nipple. He would only take one type."

In addition, mothers sought solutions to increase their infant's suckling at breast as with the use of a *nipple shield*: "My son didn't like drinking from my breast. I began using a nipple shield which he preferred," and "The nipple shield. He wouldn't take my breast without it."

Some mothers found options for breast pumping that would be more effective and efficient; for these mothers, the solution was to obtain a hospital-grade electric breast pump with a dual set up to simultaneously pump both breasts: "having a medical grade electric pump," "a very good quality double electric pump. This made pumping faster, and more efficient," and "an electric pump, that made it quicker, easier to pump after a bottle feeding."

Locating support. Locating support was a strategy used by mothers to provide them with support and guidance breastfeeding. A few mothers located metabolic clinics with a supportive breastfeeding environment consistent with the mother's value of breastfeeding. One mother revealed, "At the initial metabolic clinic we attended, the dietitian discouraged me from breastfeeding my daughter at all. We changed clinics and I had a much better experience with our new dietitian." Mothers also sought out other mothers who had or were breastfeeding infants during this time: "from other parents on on-line message boards," and "other nursing mothers during a weekly moms group while out on maternity leave." Many of these mothers specifically sought out mothers who had previously breastfed or were currently breastfeeding infants with PKU. Some mothers identified lactation consultants who provided support: "The lactation consultant suggested that I take some herbs, blessed thistle and fenugreek, to try to help since I wasn't exclusively breastfeeding."

Breastfeeding advocate. Valuing the benefits of breastfeeding and breast milk, several mothers were breastfeeding advocates for their infants with PKU. These mothers were adamant about continuing to breastfeed after the diagnosis of PKU, especially when healthcare providers were not supportive. For example, one mother reported, "His dietitian would have preferred I either formula or pumped and measured how much breast milk my son received. Though they allowed me to breastfeed my son as I desired." Another mother provided this description:

The concept was not a popular one with the clinic staff so they discouraged us, but I told them we were going to proceed and monitor

blood [Phe] levels closely. If there were problems, we would switch to the formula alone, but we had no problems and all went well.

Labor of Love: Commitment to Breastfeeding in the Context of PKU

The third specific aim was to identify mothers' experiences breastfeeding infants with PKU using thematic analysis and to examine the mothers' assessment of family functioning. Although a total of 28 mothers breastfeeding/breastfed children with PKU younger than 36 months were possible participants in phase 2, one-fifth of the mothers ($n = 6$) declined to be contacted after the Internet survey. Of the remaining mothers ($n = 22$) who agreed to be contacted, slightly less than one-half ($n = 10$, 45%) returned emails and agreed to be interviewed.

Sample

The selection of participants for the interviews was purposively selected. As with the Internet survey, mothers had to be 21 or older and be English-speaking to participate in this second phase. Mothers must have initiated breastfeeding their infant with PKU, but there was no restrictions on the length of time they breastfed. In addition, the infant with PKU needed to be less than 36 months old. A subset of mothers ($n = 10$) who participated in the Internet survey were interviewed via the telephone to more fully explore breastfeeding in the context of PKU. The mothers' mean age was 32 years (SD 2.8 years). One-half of the mothers ($n = 5$) had master's degrees while the others had a combination of associate's, bachelor's and doctoral degrees. The majority of the mothers

($n = 9$, 90%) were married. They ($n = 7$, 70%) were employed outside of the home, and all but one worked 40 hours or more a week. In addition, 80% ($n = 8$) of the mothers reported a household income greater than \$75,000 with an annual income ranging from \$25,000 to greater than \$150,000. Mothers breastfed for a mean duration of 11 months (SD 7.3 months) with a range from 6 to 30 months. Of the mothers, 80% of the mothers ($n = 8$) were Caucasian, which is reflective of the racial demographic distribution of PKU in the United States and Canada.

All of the infants were younger than 36 months of age ranging in age from 11 months to 35 months and were born at term with a mean weight of 3.7 kg (SD 0.48 kg). The proportion of males ($n = 5$, 50%) and females ($n = 5$, 50%) was comparable to that of other autosomal recessive disorders.

Family Functioning

When a family is functioning well, family members are cohesive in their actions, are able to adapt to stressors, and have clear family rules and boundaries (Knafl et al., 2010; Petrocelli et al., 2003). Mothers' perception of family functioning was evaluated with the Family Apgar Questionnaire and the General Functioning Scale of the Family Assessment Device (FAD). The Family APGAR yields a score ranging from 0 to 10. High scores, such as 7 to 10, indicate a greater degree of satisfaction with family functioning. Scores of 4 to 6 suggest moderate family dysfunction while scores of 0 to 3 reflect a highly dysfunctional family (Smilkstein, 1978). The FAD includes an independent dimension of general functioning (overall functioning of the family). The general functioning (GF) scale was designed to determine a family member's overall

perception of the family's functioning. Lower scores are indicative of good family functioning while higher scores are indicative of poorer family functioning. A 2.0 or higher suggests unhealthy family functioning

The mean score on the Family Apgar for these mothers was 8.3 (SD 3.3), which indicates a high degree of satisfaction with family functioning. This same finding of family functioning was revealed on the GF scale of the FAD with a mean score of 1.54 (SD 0.81) indicating satisfactory family functioning. Despite these differences, there was no variation in these mothers' experiences breastfeeding their infants with PKU.

Qualitative Analysis of Interviews

From each interview, a descriptive approach was used to complete the analysis in which mothers described experiences breastfeeding their infants with PKU (Miles & Huberman, 1994; Saldana, 2009; Wolcott, 1994). The intent was to analyze mothers' experiences breastfeeding infants within the context of PKU. Mothers were assured that all experiences would be de-identified to ensure their privacy, and any quotes used to illustrate key themes would be devoid of personally identifying elements. Rules and definitions were created to insure that coding occurred consistently throughout the narratives, in the same way every time, to give the coding process an essential level of consistency and coherence. The units of analysis included sentences, phrases, and words. Using the process of thematic analysis, subcategories were drawn from the interviews but were further refined after input and feedback from qualitative colleagues. During thematic analysis of each interview, subcategories were combined to form

categories and four overarching themes. Coding occurred manually. On completing the analysis, the interviews were found to provide powerful intentionality and implications for clinical management. Exemplar quotes were selected to illustrate the mothers' experiences breastfeeding infants with PKU and to provide thick description (Sandelowski, 2001). Quotes were corrected for grammatical errors, and at times, syntax was changed to improve readability.

Labor of Love: Commitment to Breastfeeding In the Context of PKU

In this study, prior to diagnosis, the majority of the mothers ($n = 9$) were exclusively breastfeeding their infants and one mother was breast pumping due to her infant's health condition, which improved rapidly with discharge from the NICU. In so doing, these mothers had expressed their support for the benefits of breastfeeding and breast milk. One mother described her belief about the benefits of breastfeeding "I felt strongly about the benefits of breastfeeding for baby and me, but mostly for baby." Once the diagnosis of PKU was received, these mothers decided to continue breastfeeding. With the decision, there was an increased workload that now included breastfeeding, bottle feeding Phe-free formula, and breast pumping to maintain their breast milk supply. In addition to the bottle feeding of Phe-free formula, mothers prepared the Phe-free formula as instructed by the healthcare provider (usually a dietitian), which sometimes meant the addition of expressed breast milk or standard commercial infant formula. Also, the work of breast pumping, which previously was not part of most mothers' ($n = 8$) breastfeeding routine, was added. This additional workload included the time to breast pump, cleaning pumping equipment after each use,

and storage of expressed milk in appropriate containers for freezing or refrigeration. Mothers described this increased workload frequently in their narratives.

During this time, these mothers were orchestrating the care of their infants with PKU and doing it by themselves. One mother reported, “It would be easier if I could just breastfeed him. But I can’t. It would be easier if he was exclusively formula fed, but he’s not.” These mothers wanted to provide the best nutrition to their infants and believed the extra work required to continue breastfeeding was worth the effort. Their stated goal was to provide the best nutrition, which meant breastfeeding and bottle feeding Phe-free formula while breast pumping to maintain their breast milk supply. For these mothers, breastfeeding in the context of PKU was “a labor of love.”

Common themes. Qualitative analysis yielded four themes that captured the mothers’ experiences breastfeeding infants with PKU: 1) *An Unexpected and Unwanted Diagnosis*, 2) *The Work of Managing PKU and Breastfeeding*, 3) *Accessing Support*, and 4) *Normalizing: Moving Forward*. See Figure 16.

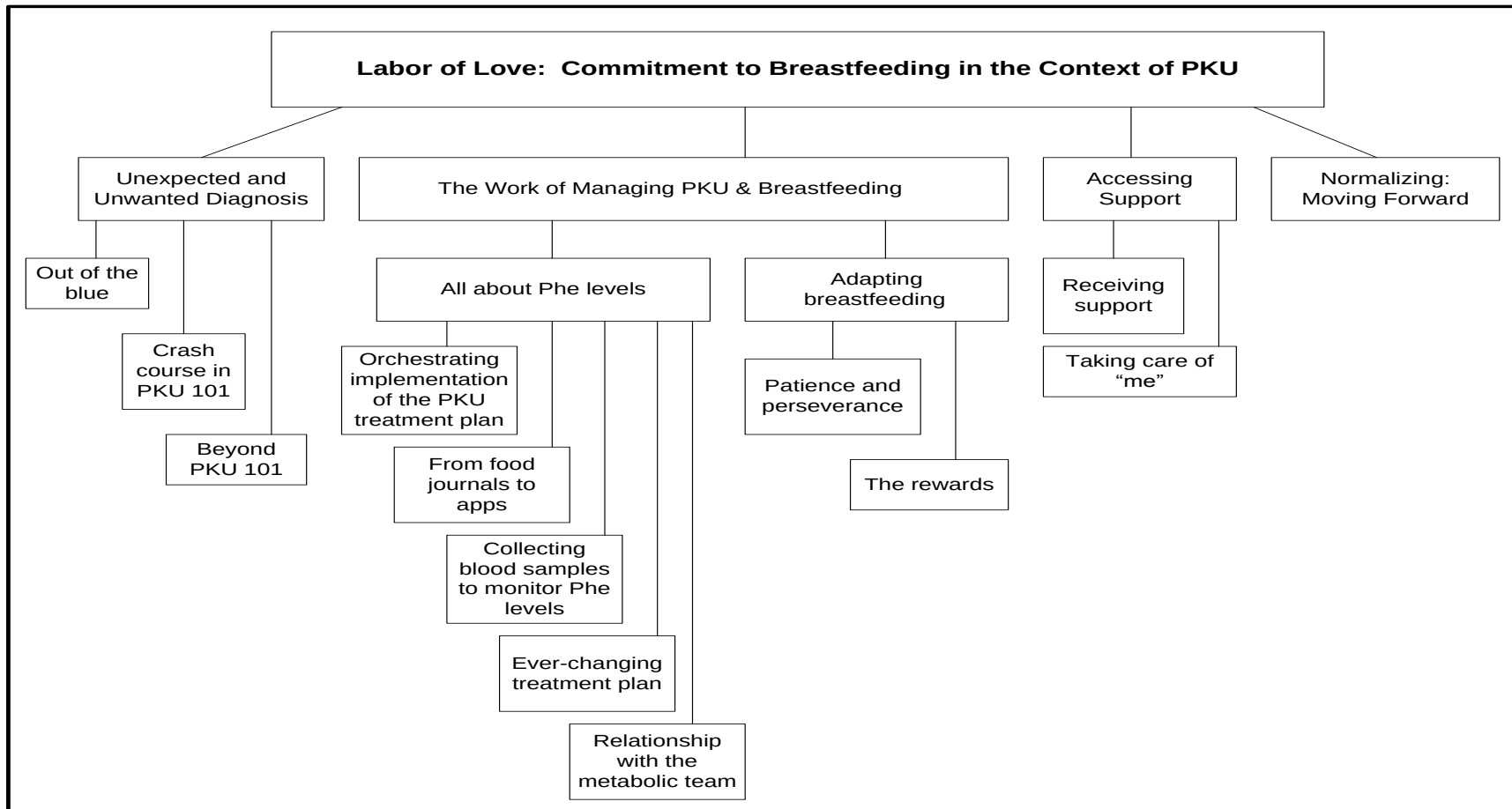


Figure 16. Labor of Love: Commitment to Breastfeeding in the Context of PKU

An Unexpected and Unwanted Diagnosis

No one can control what genes parents pass on to their children and when an unexpected PKU diagnosis was made, mothers and their families knew nothing about the condition and had no time to prepare. This first theme, an unexpected and unwanted diagnosis, describes an initial process of adjusting to having an infant with PKU and the decision to continue breastfeeding after receiving the diagnosis. Qualitative analysis yielded three sub-themes: 1) Out of the blue, 2) Crash course in PKU 101, and 3) Beyond PKU 101.

Out of the blue. In this study, these mothers reported that after nine months of pregnancy of worrying about the baby, then after giving birth to an apparent healthy full-term baby, and going home to be a family; their lives were forever changed by one telephone call from the pediatrician, or the nurse midwife, which revealed that their baby, their sweet bundle of joy, was not healthy, was not normal, was not perfect. The newborn looked normal in every regard, but the healthcare provider told the mothers the opposite, that their baby had a disorder, something totally unknown to them and their families. Mothers described their shock and disbelief saying, “I thought that everything was perfect. And suddenly, it wasn’t. Anytime during the first 6 months of her life, when someone would call her perfect, I would burst out into tears,” and “It definitely wasn’t something that we were expecting. Neither my family nor my husband’s family knew of any incident of PKU. It was a definitely a shock.” Another mother provided this description:

It was pretty emotional, I think. We were . . . you know, you worry about so many other things with your child that you don’t even think about those

genetic disorders. When she was born and appeared to be healthy, we kind of thought that we were in the clear. It was kind of out of left field for us.

During this period, all mothers struggled with the diagnosis. A few mothers ($n = 4$) described a brief period of denial.

I was like “Are you sure that this is a genetic thing? Because I was eating eggs and bacon the last part of my pregnancy, is it because I ate so much protein?” Is that the reason that his levels are so high? I was just not willing to accept it.

I think because the levels came back low that I was in denial a little bit. The levels kept coming back so low that I thought well maybe he doesn't actually have this. And then one time, it spiked after being like really, really good. And that time, I almost had to reprocess it again.

After learning of the diagnosis, mothers began a crash course to learn about PKU and what it meant for their child and their family.

Crash course in PKU 101. With the initial telephone call during the first week of life of their infants, these mothers began to rapidly learn about PKU something the majority of them ($n = 9$) had never heard of. Initially, all of them searched the Internet trying to locate information about PKU. However, the information that the mothers located discussed the old implications of untreated PKU rather than the life preserving diet and new pharmacological treatments available or under development. While meeting with the metabolic team, the mothers realized the Internet information was not applicable to their situations. From the clinics, mothers learned the basics of PKU, including that when an infant was diagnosed early (prior to two weeks of age) and treatment was started immediately, cognitive deficits could be minimized. They learned that the treatment of PKU involved reducing Phe intake through a low Phe diet, providing

a necessary alternative to natural protein through medical formula for infants, and serial monitoring of Phe level in the blood. In addition, these mothers were informed by dietitians and metabolic specialists that breastfeeding was still an option. Mothers described their decision to continue breastfeeding after the diagnosis of PKU.

I remember something that our dietitian said at the very beginning about how breast milk was actually very low in phenylalanine. Not low in phenylalanine, but lower in phenylalanine than formula. In my mind, that just meant that it would be better for me to keep trying to give him breast milk as much as I could.

It really did not make sense to me that since he still needed natural protein in this diet and the most natural protein would be breast milk. It just added to the argument of why I should continue.

Breastfeeding was really never a question for me. I was always clear that I would breastfeed. I didn't have any set notion about how long I would breastfeed, but I knew that the first few months were crucial. So that was one of my first questions actually for my dietitian. "Can we continue to breastfeed?" And she said, "Yes."

Because the clinic supported it, our doctor was very into the "breastfeeding was better." I personally thought that breastfeeding was better than formula anyway, which was what I did with my first child. So when he was going over breastfeeding, I was already pro-breastfeeding. So because my clinic was so supportive of it and was able to say to me "No, breastfeeding is better." It really wasn't a choice. It wasn't a decision. It was already our plan. Thus, we didn't have to deviate from our plan. It really wasn't a thought process. It was never a decision of should we or shouldn't we. It was more of a relief that we were able to keep doing it.

Shortly after meeting with the metabolic healthcare providers, mothers sought out various resources to learn more about the current understanding of PKU. They described the "steep learning curve" to more fully understand the disorder and the implications for their infants and families. One mother summarized this process this way:

It's been a steep learning curve, but we feel that we are there. It was a steep learning curve through with the dietitians that we worked with, through joining the online PKU Listserv, reading things, talking with other people, and meeting other moms of kids with PKU. You know you just do it; we had to do it.

Mothers' descriptions of PKU revealed their accurate understanding of the disorder:

I know that it is a genetic disorder that is recessive. Essentially, she lacks the enzyme to breakdown phenylalanine to tyrosine. She has to get just the exact amount of phenylalanine to function, but anything else can cross the CNS barrier and be toxic to her brain.

He is unable to metabolize an amino acid found in a lot of proteins. As a result, he is on a strict diet, has to follow the diet for life, and have a special metabolic formula for life to get the necessary proteins that he needs to grow.

Other mothers provided a more sophisticated, scientifically grounded description of the condition.

PKU is a metabolic disorder that is a recessive disorder. My son shares two genes. He inherited two genes: one from his mother and one from his father. We are both carriers which is why we never knew about it in our family tree. Even with treatment, there are implications for executive functioning and mental health issues. There are certainly side effects and permanent damage done when it is not controlled through diet.

My understanding in a nutshell is that obviously dietary management is essential, but that it doesn't necessarily prevent all the executive functioning and cognitive issues that can occur with PKUers, which is where they are studying right now to see if Kuvan® can help in preventing some of those things.

After learning the basics of PKU, there was a change in focus from seeking out general information to accessing more specific information about PKU and its management.

Beyond PKU 101. As these mothers became more confident of their understanding of the condition, they sought more detailed information. One mother described this change of searching.

Like now, when I am looking for information, it's not just general, basic like the definition of PKU. It's beyond that. I have search engines here at work like scholarly journals that I can look up through there, look up information through things. I can get articles from the various medical publications including the *Journal of Inherited Metabolic Disease*.

The Work of Managing PKU and Breastfeeding

While mothers continued to learn more about PKU, they were simultaneously managing PKU and breastfeeding. During the first six months following the diagnosis, these mothers managed their infants' PKU by diet therapy alone, which entailed ongoing monitoring of Phe levels and adapting breastfeeding to their special situation.

All about Phe levels. For these mothers, the management of PKU was *all about Phe levels*. Phe levels were closely monitored and adjustments were made in how much breast milk versus Phe-free formula was given to the infant. The serial Phe levels required these mothers to individualize the treatment plan as their own for their infants and families in order to maintain metabolic control with appropriate Phe levels. Qualitative analysis yielded five sub-themes:

1) Orchestrating implementation of the PKU treatment plan, 2) From food journals to apps, 3) Collecting blood samples to monitor Phe levels, 4) Ever-changing treatment plan, and 5) Relationship with the metabolic team.

Orchestrating implementation of the PKU treatment plan. These mothers were the main caregivers for their infants. As such, after taking their

infants home from the initial metabolic appointment where they received directions on how to manage breastfeeding, mothers faced being responsible for the infant's dietary adherence and managing the lifelong chronic condition. Some mothers assumed responsibility for direct management of PKU while others delegated some activities to others, but they all saw themselves as ultimately responsible for assuring adherence to the prescribed guidelines. The majority of the fathers played an important role in assuming responsibility when mothers were busy.

My husband was really great about using the gram scale and mixing the breast milk and really supportive of us doing that. And so he, when I couldn't be there to mix it, he felt comfortable weighing everything and mixing it.

My husband was always on board and up to speed with what needed to be done. He even stayed home with her when she was little over a year old when I went back to work. So he is fully versed in how to manage the diet. That is crucial.

Even when others were involved in the care, mothers assumed responsibility for orchestrating the care and making sure the special needs of their PKU infants were met. Consequently, mothers developed different methods to give daycare providers and other family members explicit information about feeding the infant.

One mother described how she communicated with her nanny on a daily basis:

I write down on a white board what he will eat that day and then how much he eats and the amount of Phe. And I need to do that because I have a nanny and she follows that, she was with me when he was diagnosed, she understands how important it is, and she follows that . . . So, I feel that I have to really, really communicate with her. And it's all the time, it's exactly what he needs to get and I have her comfortable using the gram scale and understanding it.

While other mothers reported the frustration of babysitters:

I just can't leave the kids with any babysitter and go do something. Whenever I am going anywhere and leaving the kids with anyone, we have to plan everything out. We leave them explicit instructions and also have to keep track . . . We recognize that there are some people that cannot babysit our kids. They are family members and they would be probably very hurt if we told them that to their face. We just don't feel that they truly understand the seriousness of keeping him on diet and not deviating from it.

It would have been nice if one or two other family members or close people to us really understood the diet. To the point that we could say, here is our daughter for the day. Feed her. Right. But we don't have that. My mom knows how to do it, but she is very . . . she is not very confident about it. And mistakes do get made. It would have been really nice to have someone else who really, somebody outside who really understood and could manage the diet. So that we could leave her with someone that we felt comfortable with and go out to dinner or a movie or something. And not worry about it . . . My mom knows, but I'll still phone in to see how it is going, where is she at.

We did leave her with family when she was under a year and pretty much everyone we left her with screwed the diet somehow . . . Not that we stopped doing it because of that, we did try to limit it to one or two people who could really learn how to manage the diet. Of course, it's hard. It's a hands-on type of thing. It's easy for us because we do it every day. But for those that don't, it's tricky.

From food journals to apps. While orchestrating implementation of the treatment plan, mothers also needed to record the intake of Phe and Phe-free formula. Tracking Phe intake was a daily activity for these mothers. They needed to record the amount of Phe-free formula along with breast milk intake from either breastfeeding or bottle feeding expressed mothers' milk. They used various methods to track their infant's Phe intake starting with food logs and journals, and sometimes progressing to Excel spread sheets to new "apps" available for iPhones, iPod touch, iPad, and Android phones.

Because as an infant, he would eat very sporadically like once every two hours or whatever . . . there was no telling how much he would eat. . . I logged everything and wrote “he ate 7 ounces of formula” and then how long on each breast.

I keep a daily journal of her food intake. . . . So, it’s like a little notebook and I write down the date and then I write down the time and what she has had. So that when looking at the log for a particular day such as 12:15 in the morning, she had her first feeding and it was 3 ounces of Phe-free formula. So, I wrote that down and then the next feeding was 4:45am and left or right or both sides.

Our dietitian was like he needs to be taking 21 ounces a day. It was easier for me to say he had 7 ounces at 7am, now he needs x amount of ounces for the rest of the day. I would just go into Microsoft Excel and make a little table for myself, print it out, and write it down every day. At the end of the day, I would enter it into Excel.

If his dietitian wanted me to send his food journal, I would type it up and send it as an attachment in an email. If she wanted multiple dates, I would just scan them in. I was always able to track it on paper and then I had it in a binder, but now I track it on my iPhone with an app, AccuGo for PKU®.

All mothers developed their own systematic method of tracking their infant’s daily Phe intake.

Collecting blood samples to monitor Phe levels. The majority of these mothers were taught how to collect blood Phe samples from their infants. During the first six months, they performed weekly to twice weekly in-home heel sticks to collect blood samples and sent them to laboratories to monitor the Phe levels.

The following quote reflects the nature of the work associated with such monitoring:

We did two heel pricks a week at the beginning and pretty much through the first six months to a year. After that, we backed it off to one heel prick a week. At first, we took her into the pediatrician’s office while I was off on maternity leave. . . . Then, we started to do the heel pricks at home.

In order to obtain and send routine blood Phe samples, mothers developed routines.

I was drawing the blood on Monday nights, but now I am drawing on Sunday nights because sometimes Monday is hard because I am dressed up from work. So, I do it Sunday night.

I would massage his foot during bath time. It would be warm from that. We would put a diaper on him and do a heel prick right then. So when he was an infant, we would do Sunday nights because we had all weekend to relax. Sunday night bath was the most regular bath most often.

For the blood work, they said to do the heel prick every week at about the same time. We didn't have to come in for blood work unless we were already at the hospital for an appointment.

These weekly to twice weekly routine in-home heel sticks provided the metabolic team with Phe results upon which treatment plans were constantly being revised in order to have Phe levels within the desired range.

Ever-changing treatment plan. During the first six months, these mothers waited for the Phe results from the routine in-home heel sticks to discuss with their dietitians modifying the treatment plan of breast- and bottle feedings. If Phe results were elevated, breastfeeding decreased; if Phe levels were diminished, breastfeeding would increase. One mother reported her dietitian's description of modifying the treatment plan based upon serial Phe results: "This is an art, and not a science." Treatment plan modifications were described by mothers.

I breastfed my daughter for 7 months. During that time, it fluctuated as to how much I could breast-feed because we monitored her levels every week and I would have to increase her breastfeeding if her levels were too low, and decrease breastfeeding, if they went too high.

We would watch his levels by checking him with twice weekly blood work. Initially, he was getting too much breast milk or not enough depending upon what his Phe levels were.

Mothers expressed ongoing challenges of trying to maintain optimal Phe levels and the ever-changing treatment plan as illustrated by this exemplar:

We had a book. She would go through the binder and go, “Okay, this is what the level is. This is what we have to do.” I will read you a brief note when my daughter was four months old. “Her Phe level that I got this morning was 74. A healthy level is 120 to 360. She is too low. She is not getting enough Phe into her body. She won’t be able to grow the hormones that she needs to grow and develop. The last level was 70. We increased by 8% which was pretty drastic and 10% is apparently the max. We expected it to be a few 100’s and possibly in range. We tried. But, no it is 74. This does not make sense. The last few Phe levels are as follows: 240, on target; 430 – how did that happen, let’s drop down; 72, let’s increase; 70, let’s dramatically increase; 74, now what do we do? Because we know that her Phe is going to slide, we have to push it higher, but if we push it higher – so what do we do?”

Relationship with the metabolic team. With the ever-changing treatment plans, mothers needed to frequently clarify and discuss issues and concerns with the metabolic team. Every mother interviewed stated that initially she felt a high level of trust in the metabolic team and especially their dietitian to provide them the best information. Below is one mother’s description of this:

The only thing that made it easy was the support of the dietitian. She was crucial. She was great. She supported breastfeeding in theory and in practice. It was like from the very first week, we had a lot of confidence in the dietitian that we had. She just struck us as really being on the ball, really intelligent, and really knew what she was doing. Her word is pretty much, you know, biblical to us. Whatever she says goes. So we, with her encouragement, we continued to breastfeed.

However, as time progressed, some mothers became dissatisfied with their metabolic healthcare providers and in the process began to shop for a metabolic team that would meet their needs and their infant’s needs better. Some

mothers reported being dissatisfied with lack of availability to respond to questions and concerns; others questioned the credibility of some of the information they were receiving. One mother described the frustration of the inability to discuss her questions with her original dietitian.

It wasn't that they didn't encourage breastfeeding. It was more that they weren't available as much when you needed it. PKU was brand-new to us and really scary, and I had a lot of fears and anxiety and a lot of questions throughout our first child's first few months of life and I don't know but she (the dietitian) did not just work at hospital, but someplace else. I think that she was at the hospital like 2 days a week and I could only get a hold of her those 2 days out of the week. A lot of times, I would call her and she wouldn't be available. She would call me at the very end of the day and she would miss me as I would be on my way home from work. She would leave me a message, but I wouldn't be able to reach her. I wouldn't be able to call her until the following week.

Other mothers reported doubting the accuracy of the information received from their dietitian such as "The new dietitian simply did not have the experience."

Another mother provided this description:

There were times that we questioned the dietitian's knowledge base. Was she the best person to help us with these first really crucial years of our child's life? We felt that for our child's health and for our peace of mind that we should try someone else that we trusted more and had a better relationship with.

Another mother expressed concern about the level of PKU expertise among members of the metabolic team. She believed they lacked basic knowledge needed to care for her infant with PKU:

We went in the next day to talk with the geneticist who really didn't know that much about PKU, but he had studied under a pioneer in PKU research, who was at Children's Hospital. So a few months after meeting with the geneticist, we transferred to Children's Hospital and worked with that nutritionist and nurse practitioner. That clinic is amazing. The first clinic wasn't too knowledgeable about PKU. We kept asking questions and he had to keep researching it and get back with his mentor at Children's Hospital.

Despite these frustrations, all interviewed mothers developed over time the ability to work with their original ($n = 8$) or new ($n = 2$) metabolic team. As reflected in the following quotes, their goal was to obtain the information that they needed when they needed it: “I think the dialogue with the clinic is important so that you can get your questions answered,” and “I like to communicate with the dietitian when we make changes to her diet. So we need to make sure that we’re all aware of that.” Another mother provided this description:

Our regular dietitian was unbelievable. She has a great deal of history dealing with PKU. . . . When she came back, my daughter’s numbers were able to be within range even when she had the chickenpox. This woman has a sense for it. It does not matter what the book says; she just understands that at this stage in children they are going through growth spurts or something else. She would call me and say, “I want you to do this with her. I want you to try this.” It always worked out and that was quite a relief when I got my real dietitian after she returned from her leave.

Adapting breastfeeding. As the mothers were managing PKU, they were also managing breastfeeding, which was adapted to their unique situation. These mothers reported the value of breastfeeding and breast milk for their infants and described the emotional feelings of breastfeeding: “Well, when I was pregnant, I was supplying her with all the nutrition that she needed and after I wanted to breast-feed to have that bonding experience. I never had any doubts that I didn’t want to breast-feed.”

Shortly after delivery, the majority of mothers ($n = 9$) started breastfeeding in the delivery room. After their infants’ diagnosis, their breastfeeding plans changed from exclusively breastfeeding to combining breast- and Phe-free formula feeding with ongoing adjustments based on the weekly to twice weekly,

serial, in-home heel stick Phe results. Some mothers were able to breastfeed twice a day while others were breastfeeding more frequently. Consequently, breastfeeding varied with the ever-changing treatment plan. Qualitative analysis of data related to the Adapting Breastfeeding theme yielded two sub-themes: 1) Patience and Perseverance, and 2) The Rewards.

Patience and perseverance. These mothers used a variety of approaches to combine breastfeeding and bottle feeding Phe-free formula. One mother described bottle feeding first, followed by breastfeeding “There would be a fairly small amount of formula at every feed and then breast milk afterwards. Sometimes she would take a little and other times she would take a lot.” Other mothers found options through patience and persistence that worked for their infants and families.

In the morning, he would wake up and have a bottle of the combination of the metabolic formula and breast milk. He didn’t like the bottle hot or cold. I would take the bottle out of the refrigerator for about 5 minutes and it would be just right for him. I would see if he wanted the bottle, and if he didn’t, I would just put it back in the refrigerator.

After the first day of absolutely no bottle success, I was like alright; maybe if we put half an ounce of formula in two ounces of breast milk into a bottle, we might be able to trick him into this whole bottle thing if we mix it with breast milk. So that was what I did since I was pumping so much and I had so much breast milk. Oh my goodness, it took him two weeks to kind of get the hang of formula and we were still mixing it with breast milk and he would only take it out of this one bottle. These two glass bottles were for some reason the only bottles that he would drink formula out of. I basically carried these two glass bottles everywhere for the first three months.

After diagnosis, four mothers began to exclusively breast pump on recommendation from the metabolic clinic, their child’s inability to latch onto the breast, or work constraints. Of these four mothers, two of them transitioned to

breastfeeding from exclusively breast pumping. One mother provided this description of the transition from breast pumping to breastfeeding.

He didn't latch for a month. So for a month, I was exclusively breast pumping. The good thing from that from our clinic's perspective was that we could at least measure how much breast milk that he was getting. I think that made it easier for them to adjust his formula intake based upon his blood levels . . . I think that possibly since I was pumping, [it] made the situation a little bit easier or a little bit easier to manage from our clinic's perspective. After a month though, he did start nursing and I don't think that changed things too much because we already had an idea of how much he was taking in when I was just pumping.

For the other mother who transitioned from exclusively breast pumping to breastfeeding, this transition was more complicated.

I mostly wanted to nurse her even though I was still pumping seven times a day. I called my local health clinic and I said "I need a baby scale because I need to weigh her before and after. I am going to figure out how many milliliters it is." We did that for three weeks. We timed it. At first, it started out as one minute at a time, and then I would take her off and weigh her. Then we would nurse, and we would go back and forth. That worked actually quite well. By the time that she was a month old, we had timed it and she was able to nurse. . . Every morning and every evening, I was allowed to nurse her for eight minutes.

Although the majority of these mothers ($n = 8$) were breastfeeding, they were breastfeeding less frequently than mothers who were exclusively breastfeeding healthy, term, non-PKU infants. Even with breast pumping after bottle feeding Phe-free formula, the majority of these mothers ($n = 9$) struggled to maintain a breast milk supply: "The lack of milk supply toward the end got to be a real struggle . . . The constant pumping is a little bit tedious, just to get enough milk." Another mother provided this description of her breast milk supply issues.

[Breast milk] supply issues were the biggest challenge. I would have gone for two years, if the supply was there. It was supply issues . . . Since

supply wasn't there, his desire was waning. He wanted nothing to do with the breasts . . . if I tried to put him to nurse him and there wasn't anything there, he didn't want to eat something that wasn't there to eat . . . the biggest obstacle to nursing was trying to keep him interested when supply was waning.

Shortly after the PKU diagnosis, the majority of these mothers ($n = 8$) realized the need to be proactive to maintain their breast milk supply and started breast pumping after feeding their infants Phe-free formula.

For the first feeding in the morning when we woke up, I would feed him a bottle or someone else would feed him a bottle and I would pump. Throughout the rest of the day, I would alternate between breastfeeding and bottle feeding. I think from about when he was about a month and a half to three months old, he was probably eating nine to ten times in a 24-hour period. Three during the night, then six during the day, I would alternate those six, one bottle, one breastfeeding, one bottle, one breastfeeding. Every time I bottle fed him, I pumped.

Not only did I have a lot of information about breastfeeding, I knew that if I didn't keep pumping that there was no way that I would have enough milk to maintain breastfeeding her eight minutes in the morning and at night. So, I had to keep pumping. I had a reason to keep pumping.

Basically, I think I would nurse him when I could during the day. At night though when my husband would get up to feed him bottles, I would be getting up to pump. I continued that for a long time. That was hard.

However, even using the strategy of breast pumping to maintain their breast milk supply, these mothers struggled to do so.

My [breast milk] supply changed twice with my son with PKU and that was where I had issues when I was calling the medical professionals with like "I'm drying up and it's only four months, and I'm pumping like crazy. Nothing is happening and I'm not ready to give up breastfeeding, yet.

In this study, one infant never breastfed and received expressed mothers' milk only from a bottle and another infant transitioned from breastfeeding to receiving all expressed mothers' milk only from a bottle prior to three months old. The remaining infants ($n = 8$) breastfed from twice a day to more than six times

per day, still less than mothers exclusively breastfeeding healthy, term non-PKU infants. The majority of mothers ($n = 9$) identified the need to have a breast pump to adequately empty their breasts frequently enough to maintain their breast milk supply as their infants were unable to exclusively breast-feed. Mothers described appreciating a powerful electric breast pump to maintain an adequate breast milk supply.

And having a good pump, a really good pump really did it because the issue that arises and breastfeeding and simultaneously bottle feeding are always related to supply. Because your kiddo has not nursed as frequently as an exclusively breastfed baby, your supply can wane.

So having a really good electric breast pump saved me. If I had my old pump, which was a \$25 single electric version that you stick it on there and maybe if you got an ounce in ten minutes, it was a good day. But, there is no way; there is no way that I could have kept my supply up because that pump won't have done it. I had a breast pump, the \$300 version, and I would be the first person to tell all of my friends because I pumped so much that . . . when it comes to breast pumps, you truly get what you pay for. If you are going to do it a lot, you have to have an expensive, effective version. Because if you have an efficient model, I would hook up to both sides and in five minutes, both sides would be drained of about three ounces in five minutes, and go about your business. If it took an hour even to get half of that, you're not going to stay with it because it's too much of a hassle, it's eating up too much of your time, somebody else has to manage the baby while you're trying to do all that and eventually it is going to come down to supply issues. You can't pump and you're not feeding the baby, the milk isn't going to be there.

For three of these mothers, breast pumping alone was insufficient to maintain their breast milk supply. Some mothers were prescribed domperidone, a drug that has a side effect of increasing breast milk production by increasing prolactin production by the pituitary gland, to use in combination with breast pumping and breastfeeding. One mother described using domperidone to augment breast pumping and breastfeeding: "I did need, I had a prescription for

domperidone to help with milk production She wasn't stimulating the milk production as much like a normal baby would have. So, I needed a bit of help in that regard."

Other mothers were encouraged to use herbal supplements, such as fenugreek and blessed thistle, to boost their breast milk supply. Another mother described using herbal supplements by saying: "My mom kept giving me fenugreek seeds as she was born and raised in Asia. They have a lot of herbal, all natural things that would help my breast milk supply."

Another issue for the majority of these mothers ($n = 9$) was "finding the time to breast pump, which wasn't easy" while continuing with their other household duties, childcare responsibilities, and for some, returning to their careers. Mothers described breast pumping while interacting with their children.

I usually get my pump to make sure that everything is clean and everything is put together and you know the girls are right there or occupied otherwise I sit on the floor pump while our oldest plays or I read books to her and our other daughter is there.

When I had my son, I would have to lay on the floor with him. I would pump one breast at a time so that I had one hand free for him. When my husband was home from work, I was able to pump both breasts at the same time.

In contrast, other mothers ($n = 3$) found that being home was a challenge to breast pumping because of competing responsibilities and interruptions. For these mothers returning to work provided more time to consistently breast pump: "When I went to work, I would be able to pump," and "Just having some flexibility at work." Despite the inability to exclusively breastfeed and the various challenges to breastfeeding their infants with PKU, these mothers did adapt with

patience and perseverance to successfully breastfeeding their infants.

Consequently, these mothers came to view the rewards as outweighing the work breastfeeding.

The rewards. Despite the challenges and extra work associated with breastfeeding an infant with PKU, these mothers believed the rewards outweighed the work associated with breastfeeding their infants with PKU.

I am so grateful that we were able to still breastfeed her. Even with the cracked and bleeding nipples, the thrush, and my health, it wasn't always very fun and it made me cry. I'm really grateful that we were able to breast-feed as I believe that it is the best thing and I have no regrets. I try to live my life with no regrets. And I have no regrets in how we managed her when she was an infant because we were able to breastfeed.

It's been really great because I never wanted to look back and look at his first year and say I wish that I could have done something differently. And I think when you find out that you have a child with PKU, you kind of overanalyze their development. And I think for a long time that I would get really hung up on "well, he is not looking at me" and "he crawled a lot later than his brother. He is definitely behind his brother in gross motor, but he is not at all developmentally delayed." His doctors are like "he is totally on track." And I, for a long time, I really didn't believe that, but now seeing him doing really so well and his verbal skills are great, his fine motor skills are awesome. He is so communicative. He is so smart. I think I feel so proud of myself, I have no idea if breastfeeding had any correlation to that, but I think that I hadn't done that, I might have thought that well maybe I would have done something differently. So I think that it really was a labor of love.

Accessing Support

In order for these mothers to successfully simultaneously managing PKU and breastfeeding, they required support and information from various resources: husbands, family, friends and healthcare providers along with taking care of themselves. They actively sought out others who understood what they were going through and could provide information, support, and encouragement

to help them make breastfeeding successful when one was unable to exclusively breastfeed. One mother described this time period as lonely and challenging, “So, it is very isolating feeding a PKU infant because PKU is so rare. It was difficult to find someone who had breastfed another infant with PKU. I did feel isolated.” The Accessing Support theme included two sub-themes: 1) Receiving support, and 2) Taking care of “me.”

Receiving support. Despite the feelings of loneliness, these mothers used resources from the Internet as well as local resources such as clinics and breastfeeding groups to access information about breastfeeding their infants with PKU. They were able to find information about PKU or information about breastfeeding, but not breastfeeding infants with PKU. Mothers’ breastfeeding success was dependent on their ability to integrate information from these two disparate bodies of knowledge, but there were few resources available to them. Mothers described this lack of information available on breastfeeding infants with PKU.

Specifically for breastfeeding, I relied a lot on the birth center. They provided a lot of ongoing support for nursing moms. For breastfeeding, specifically not PKU related, it was the birth center; for PKU, it was the clinic. But I didn’t have a lot of resources at all specifically related to breastfeeding a PKU baby. I had a lot of support for it through the clinic, but there wasn’t necessarily a lot of information.

I tried to go on the web to do some searches on the forums that they have online. I found a lot of information on breastfeeding in general, but not on breastfeeding and PKU.

Thus, mothers who were knowledgeable of how to breastfeed an infant with PKU were an especially valuable resource. Some mothers (n = 3) were able to locate another mother who had or was breastfeeding an infant with PKU and lived

within their geographical area. This one-on-one in-person exchange provided mothers with support to continue breastfeeding their infants with PKU.

There was another woman that lived nearby who had a second child that was diagnosed with PKU that was only a few weeks older than our first. I was introduced to her when our first was just a few weeks old and became friends and were able to talk about our experiences, and were a support for each other.

I did talk with another mom who had breastfed and she had twins and one had PKU. She said she had done it almost to 11 months. And that kind of encouraged me and was probably about two months after he was diagnosed. That definitely kept me going toward the end of things.

Many of these mothers were unable to access another mother who lived within their geographical area. These mothers sought support via the Internet. One mother described accessing Internet support in the following words:

My mom was actually the one that pointed me toward the listserv. My mom calls "Hey, do you know about this email listserv?" I'm like "No." I do email like crazy. I'm a tech guru. She gave me the email address. I joined the listserv and watched it for three or four days. After the three to four days, I was like okay and started sending out emails: Need breastfeeding advice, Having supply issues with PKU baby – How do you increase, how did you increase your supply from mothers who have done it? How did you manage? Moms who were familiar with it, what did your clinic expect you to do? It was really interesting . . . because I had 20 responses within 24 hours of women who just wanted to talk about their experiences breastfeeding a PKU baby. They were like this is what I did, this is how we managed, and this is what my clinic expected. My experience with the listserv in getting all those responses was a positive thing.

Other mothers joined the PKU Listserv, and despite varying levels of participation, reported finding it a valuable resource as indicated in the following quote: "Also joining the listserv, even though I do not post questions frequently, but reading the various conversations that people were having, were similar questions that I would have." For these mothers locating information about breastfeeding infants with PKU was a challenge. Some mothers were able to

locate another mother who had or was breastfeeding infants with PKU while others used the Internet to locate information and support.

In this study, these mothers described receiving support from a variety of sources, professional and social. Both types of support were important to them. Dietitians were identified by all mothers as the main healthcare provider who provided them with support: “The dietitian was paramount. There would be times that I would talk with the dietitian a few times a week with questions that I had,” “The dietitian was the number one support I think that she was the one that we had the most contact with on a regular base. I think that it was mostly the dietitian,” “Our dietitian was amazing. I can’t say that enough,” and “We are very happy with our dietitian. Our dietitian has been wonderful; she has been our lifeline and she has been supportive all along.” Other healthcare providers also provided mothers with support. One mother described the pro-breastfeeding belief of the metabolic specialist as being a source of support.

When our metabolic specialist heard that we wanted to breastfeed and I was all about breastfeeding for as long as possible, he was thrilled and totally encouraged that He was a huge supporter of breastfeeding so that definitely helped because he was the one in the beginning was like “Keep going, keep going. We’re stop you, when we need to stop you, but we’re never going to stop you. We will pretty much try to figure out how to work with you.”

Some mothers ($n = 5$) sought out specific support for breast milk issues as described by this exemplar: “When my milk supply started to decrease, I contacted a local lactation consultant and talked with her about keeping my milk supply up. And then I talked with her after I went back to work, with questions about what I should be doing.”

Also, the majority of these mothers ($n = 8$) in this study had husbands who supported breastfeeding. Mothers reported their support from husbands as “I had a husband who believed in it. He really believed that breastfeeding was the best thing. He understood that if I didn’t keep pumping that I won’t be able to breastfeed,” and “I think that we get a lot of support, but like my husband is the best support, honestly because he is in it with me.” Another mother reported:

Well my husband He was very supportive and wanted, when he could, to be a part of it. So, he did a lot of the bottle feedings of the Phe-free formula. I guess in a twisted way that was a blessing that he could participate a little bit more than most dads.

In addition, these mothers received support from family and friends.

Mothers described the support from family members as illustrated below.

I am definitely grateful for my support system. I think that without my parents being so close that we would have had a greater challenge in that she would have been in daycare sooner which meant that we would have to have done a lot more education. Luckily for us, my parents have been able to keep her.

My family was incredibly supportive. My sisters both nursed their children. My mom nursed us. I guess that is what made me assume that I would be breastfeeding my child. My family, I would say my family was definitely very supportive.

My husband’s parents are in the Midwest. Even though they are not with her, they have made a great effort, particularly his mother, to learn about PKU. I think that she has really made an effort even though she is not here with us every day to be part of her PKU life style.

Many of these mothers ($n = 7$) reported support from friends. One mother stated, “Even all my friends, I don’t have one friend who hasn’t breastfed. It was just the norm. In this community, we tend to be a little more open-minded.”

The majority ($n = 7$) of these mothers returned to work between three and four months postpartum. Mothers described the support from their employers to

be able to breast pump in their own offices and in breast pumping rooms.

Mothers revealed: “I was fortunate to have my own office and I could just pump in my own office, and shut the door,” and “So sometimes, if I had a lot of work to do, I would do one breast at a time. If I had more time, I would pump both breasts. My job is tied to the computer.” In addition, mothers described support to breast pump from colleagues at work.

My staff was a very young, open-minded staff and a lot of them were young moms who were breastfeeding and pumping as well. It was just kind of the expectation from the men and the women on staff that when someone had a baby, that they would be pumping.

From accessing support, these mothers were able to manage PKU and breastfeed their infants with PKU.

Taking care of “me.” In addition to outward support, these mothers also needed to take care of themselves. These mothers spent a large amount of time focusing on their infants with PKU. They reported that the first year was stressful for them as they adapted to having a new baby, returning to work, managing the unique breastfeeding challenges they faced, and managing PKU. One mother described this period of time this way: “With the birth of our daughter, a lot of stress as new parents. It was overwhelming. We were busy with her needs; sleep deprived, doctor appointments, and management of PKU. I cried a lot.” To help them deal with this stressful time, mothers reported they made an effort to take care of themselves individually and as a couple. A majority of mothers ($n = 8$) reported that their relationship with their husbands was important in taking care of themselves. One mother reported taking care meant “watching a movie with the hubby.” Another mother described the time with her husband this way:

Making sure that we take time for regular family relationships that don't have anything to do with PKU. So, we try to make more time for ourselves and nurture our relationship to talk about other things in part because there is so much that we need to talk about that deals with PKU.

In addition, mothers found time for themselves ranging from simple indulgences, such as “reading a book or having a bubble bath,” to other forms of taking care of oneself by increasing their physical activity: “I joined a moms' workout class where we took the kids in strollers to a park and worked out with other moms” and “My ‘me time’ was spent exercising and my husband was very supportive of my getting back in shape. This helped me maintain a positive outlook and keep up my energy.” However, a few mothers ($n = 2$) reported efforts directed toward “taking care of self” were more than stress relief; rather they described these efforts as essential to their capacity to be able to function and care for their infants.

I probably didn't take the best care of myself that first year. I was very stressed about learning how to manage PKU, having a first baby and all of the stress and life changes that comes with that PKU or otherwise, and returning to my career from maternity leave. All of those things overwhelmed me at the time, and I really didn't have any time for myself, unfortunately. I think the first year was more about survival, honestly.

To be completely honest, I didn't do enough the first few months and ended up in counseling because I didn't know how to handle the stress of being a first-time mother, working fulltime, and learning how to manage PKU.

From the unexpected and unwanted diagnosis, adapting breastfeeding, to maintaining Phe levels within the desired range, and accessing support, breastfeeding was a way for these mothers to view their infants with PKU as normal and thus make a transition to normalizing.

Normalizing: Moving Forward

With the diagnosis of PKU, these mothers viewed their infants as ill with PKU. Although unable to exclusively breastfeed, these mothers adapted breastfeeding and were able to meet their goal to breastfeed their infants. Consequently, as mothers perceived their breastfed infants with PKU as healthy, and they were developmental and intellectually growing like other healthy, term, non-PKU infants, mothers began to view their infants as normal. These perceptions validated the mothers' breastfeeding decision. One mother described this moment:

When our daughter was first diagnosed, one of our initial fears was: Was she going to be okay? Is she going to be like other children? Now that we see that she is we have that knowledge that is very comforting and that lets us, that gives us, the drive to do all that we are doing.

When this occurred, mothers began to view their infants not as infants with PKU but as breastfeeding infants who have PKU. Breastfeeding was a way to view their infants with PKU as normal and achieve a sense of normalcy in everyday life. Another mother described her son as follows:

He has PKU, but he isn't PKU. It is just one aspect of his life. It is not his whole life It's not a big deal.

After the initial diagnosis of PKU, these mothers grappled with the sudden unexpected and unwanted diagnosis. As they successfully integrated their adapted breastfeeding plans, the rewards outweighed the work load of managing PKU and breastfeeding even though there were reports of limited babysitting, daycare, and other respite care. Although there was support from husbands, family, friends, and colleagues, mothers emphasized the intensity of the workload

associated with simultaneously managing PKU and breastfeeding and the challenges of finding adequate information and support. Through breastfeeding, these mothers came to view their infants not as ill, but rather as healthy, term infants with PKU, thus, normalizing the outcome.

Summary

In phase 1 of the study, a convenience sample of 119 mothers who had infants with PKU participated in an online Internet survey. These mothers were recruited from the PKU Listserv. In this study, mothers ($n = 103$) who had one child with PKU were the focus of this dissertation. Mothers were English-speaking, at least 21, and lived in the U.S. or Canada. Mothers were predominately white, married, and on the average were 36.5 years of age with either one (37%) or two (39%) children in the household. Analysis revealed that significantly fewer mothers breastfeeding after diagnosis (McNemar's $\chi^2 = 28.89$, $p \leq .0001$ $n = 89$ versus $n = 72$). However, this significant reduction in breastfeeding mothers was a function of women from the U.S. (McNemar's $\chi^2 = 27.48$ $p < .0001$; $n = 60$ versus $n = 75$), and not from Canada. Of the 89 breastfeeding mothers, the majority provided responses about their breastfeeding experiences to four open-ended questions in an Internet survey. Their responses identified facilitators, barriers, and strategies to breastfeeding infants with PKU.

In phase 2, the mothers from phase 1 served as a pool for mothers ($n = 10$) who participated in telephone interviews and had a child with PKU less than 36 months of age. In the interviews, mothers described their labor of love by

committing to breastfeeding in the context of PKU. Mothers contended with the shock of an unexpected and unwanted diagnosis while simultaneously managing PKU and adapting breastfeeding to maintain appropriate Phe levels. While receiving support from husbands, family, friends, and colleagues during this time, these mothers tried to access support from other mothers who understood what they were going through and information to help them make breastfeeding successful when they were unable to exclusively breastfeed. Through breastfeeding, these mothers came to view their infants not as ill, but rather as normal healthy, term infants with PKU, not as PKU.

CHAPTER 5

DISCUSSION

The purpose of this mixed methods study was twofold: 1) to determine the prevalence and duration of breastfeeding for infants with PKU in the United States and Canada and 2) to generate knowledge about how mothers of infants with PKU simultaneously manage breastfeeding and PKU therapy, and how they perceive family functioning. This study focused on mothers' experiences breastfeeding infants who have PKU. In phase 1, quantitative analysis was used to describe the prevalence and duration of breastfeeding infants with PKU living in the United States or Canada and revealed mothers were successfully doing so. In addition, a frequency count approach was used to recognize patterns and to generalize meaning from the open-ended responses in the Internet survey and revealed facilitators, barriers, and strategies of the mothers' experiences breastfeeding infants with PKU. In phase 2, thematic analysis methodology was employed to analyze data from telephone interviews with mothers who had one child with PKU less than 36 months of age. This analysis revealed four themes in the Labor of Love: Commitment to Breast Feeding in the context of PKU. In this chapter, the methods employed will be discussed first, followed by a discussion of the findings.

Methodology

This study was primarily quantitative descriptive with a supplemental qualitative component. This approach situated the sequencing of the data collection and analysis. Findings from the first specific aim were evaluated in the

context of existing normative data from the U.S. and Canada. Subsequently, the four sets of open-ended responses were coded to identify facilitators, barriers, and strategies guided by a theoretical framework, the Family Management Style Framework (FMSF), from the mothers' perspective. These codes were described using frequency counts and percentages. This analysis provided the first account of mothers' perspective of breastfeeding infants with PKU.

In accordance with FMSF, mothers' perspectives were sought to understand their definition, management, and perceived consequences of breastfeeding infants with PKU, and their incorporation of and adherence to the responsibilities and activities of PKU therapy. The mother-infant breastfeeding dyad, a subsystem of the family, was the focus as mothers were the most involved in managing breastfeeding and typically are the family member who assumes primary responsibility for infant care. In the end, it was the analysis of the mothers' experiences that communicated their commitment of breastfeeding in the context of PKU.

Sample Characteristics

In this study, the Internet was used to recruit mothers with children who have PKU to participate in the study and to collect data. One of the main advantages was the recruitment of mothers who lived in geographically different regions, from rural communities to metropolitan cities, many who otherwise would not have been available to participate. In addition, the Internet recruitment allowed easier access to mothers who might not attend a focus group even if it was available nearby. This sample was representative of mothers who had a

range of socioeconomic characteristics and different types of healthcare insurance coverage, and who received care from a variety of specialty clinics with varied feeding protocols and beliefs about the appropriateness of breastfeeding infants with PKU. In theory, this should have provided a heterogeneous sample; however, the sample was more homogenous than heterogeneous.

Mothers who participated in this study were highly educated, married/partnered Caucasian mothers who have children with PKU in the U.S. and Canada. In comparison to the U.S. Census Data (Lofquist, Lugaila, O'Connell, & Feliz, 2012), these mothers actually were more educated, had higher annual household incomes, and were more likely to be married/partnered than those in the average U.S. household. Yet, this sample represents contemporary mothers with a mix of those who were employed and those who were “stay-at-home moms.”

In addition, this sample illustrates how electronic computer technology, such as the use of email, cell phones, iPhones, and Blackberry phones, has become a part of their everyday life from communicating with the metabolic clinic to keeping in touch to organizing the family calendar—regardless of their economic situation. This is consistent with the findings from a recent poll that revealed mothers are spending a significant amount of time using media and technology platforms (Warzel, 2012). However, there were relatively few socioeconomically disadvantaged mothers in this current study. Another recent study (Wen, Rissel, Baur, Lee, & Simpson, 2011) reported young mothers with

lower levels of education (a high school education or less), lower household incomes (< \$40,000), and lacking a computer in the home were less likely to access the Internet for health information. The recruitment of mothers who were less socioeconomically advantaged in both countries would have provided a more heterogeneous sample for the study. Future studies must attempt to address recruitment of socioeconomically disadvantaged mothers of infants with PKU.

Even though this sample was not representative of the larger population of mothers with children who have PKU, the sample characteristics were similar to those from other research involving families and individuals with PKU. In a survey of parents with children who have PKU and of young adults with PKU, two-thirds of the participants had completed some college and had annual incomes of greater than \$30,000 (Wappner, Cho, Kronmal, Schuett, & Seashore, 1999), reflecting comparable socioeconomic levels and motivation within this population to participate in research.

The findings from this study may not pertain to groups other than the participants. In this study, over 90% of those who participated in the survey and 80% of those who participated in the interviews were Caucasian. Although it is unknown if the few mothers who were from minority groups had been born and raised in the U.S., this study enabled exploration of their Western worldview, leading to the limit to which this study can shed light on the experiences of other mothers living in different places or different cultures.

Breastfeeding Success among Infants with Phenylketonuria

No previous research was located that specifically identified the prevalence and duration of breastfeeding infants with PKU from countries in North America. This study was the first to examine the prevalence and duration of breastfeeding infants with PKU from the U.S. and Canada.

Breastfeeding Infants with PKU in the United States and Canada

In this study, mothers successfully breastfed infants with PKU, and they met or exceeded the majority of breastfeeding percentages reported in both the U.S. and Canada. In comparison to U.S. national breastfeeding percentages (2011), this group of mothers exceeded the expected rate of initiation of breastfeeding (86% vs. 75%) and breastfeeding at six months (55% vs. 44%), but they did not meet the expected rate of breastfeeding at 12 months (17% vs. 24%). In addition, mothers were comparable to the Canadian breastfeeding rate at six months (55% vs. 54%) (Chalmers et al., 2009). Remarkably, these mothers almost met the Canadian initiation rate for breastfeeding (86% vs. 90%), and the three-month breastfeeding rate (65% vs. 68%). In comparison to the Healthy People 2020 Breast Feeding Objective, the mothers exceeded the goal for the initiation of breastfeeding (86% vs. 82%) but did not meet the breastfeeding objectives at six months (55% vs. 61%) or 12 months (17% vs. 34%) (U. S. Department of Health and Human Services, 2010). This drop-off over time in breastfeeding by mothers of infants with PKU perhaps reflects the ongoing demands of managing the PKU therapeutics.

This drop-off in breastfeeding was not as soon as the drop-off found by other investigators from countries in Europe, Asia, and South America who have studied the incidence and duration of breastfeeding infants with PKU (Agostoni et al., 2000; Demirkol et al., 2001; Huner & Demirkol, 1996; Motzfeldt et al., 1999; van Rijn et al., 2003). These studies consistently reported that the majority of mothers breastfeeding infants with PKU did not persist as long as mothers breastfeeding healthy non-PKU infants. Further, mothers with infants who have PKU from the U.S. and Canada breastfed longer than those from other countries. Given that statistics for the incidence and duration of breastfeeding infants with PKU from other countries are at least a decade old, current data might reveal a different comparison.

Characteristics associated with decreased duration. In this study, the initiation and duration rates of breastfeeding were remarkable despite the work involved to breastfeed. Biological and social demographic characteristics that might have influenced breastfeeding duration revealed that decreased duration of breastfeeding for infants with PKU was only linked to those incidences when standard commercial infant formula was added into the infant's diet, thereby replacing breastfeeding or expressed mothers' milk. This finding is consistent with other researchers (Branger, Cebron, Picherot, & de Cornulier, 1998; Feinstein, Berkelhamer, Gruszka, Wong, & Carey, 1986) who identified formula supplementation in breastfed infants as a strong predictor for the discontinuation of breastfeeding.

Reduction in breastfeeding after diagnosis. The overall reduction in the proportion of mothers who were breastfeeding before and after the diagnosis of PKU was significantly different between the U.S. and Canadian mothers. This illustrates the importance that is placed on breastfeeding as the norm in Canada. The provincial and territorial governments provide public health services to support and sustain all mothers in breastfeeding their infants. This support includes visits within 24 hours of hospital discharge by public health nurses who have a focus in maternal-child nursing, peer-to-peer support to sustain breastfeeding for the first six months, and providing specific infant scales to determine breast milk intake before-and-after breastfeeding for infants with specific breastfeeding needs. Although breastfeeding support is a responsibility held within the public health divisions of states and territories in the United States, the focus entails working with communities and various partners to support, protect, and promote breastfeeding within healthcare systems, work environments, and public places. Thus, in Canada, the focus is on the individual mother whereas in the United States, the focus is on systems. This difference may account for the difference in sustaining breastfeeding found between Canadian and American mothers. That said, the difference in the size of the subsamples of mothers recruited from the United States (84%) and Canada (16%) must be acknowledged. The recruitment of more comparable subsamples will be necessary to ascertain differences with any certainty.

Mothers' Experiences Breastfeeding Infants with PKU

A review of the literature revealed no information regarding mothers' experiences breastfeeding infants with PKU. This study provides the first exploration of such experiences breastfeeding. On the survey, mothers were asked to identify factors that facilitated breastfeeding, barriers that impeded breastfeeding, and to describe their strategies to manage the care for their breastfeeding infant with PKU. The following discussion will address the challenges and barriers to breastfeeding, what facilitated or helped with breastfeeding, and the strategies that eased the management of breastfeeding that mothers revealed through their survey responses.

Facilitators to Breastfeeding Infants with PKU

From the open-ended responses, these mothers identified four categories of facilitators for breastfeeding their infant with PKU: 1) beliefs, 2) breast pumping, 3) easy child, and 4) supports. All of these categories have been consistently identified in the literature as supportive for breastfeeding healthy term, non-PKU infants as well as other infants who are unable to exclusively breastfeed, such as premature and critically infants.

Beliefs. In this study, beliefs included beliefs about the *benefits* and *naturalness of breastfeeding*. Mothers believed there were *benefits* of breast milk and breastfeeding for infants, mothers, and families. One-quarter of mothers in this study described knowing the benefits of breast milk as making it easier for them to breastfeed their infants with PKU. This belief in the benefits of breast milk is similar to reports by mothers of healthy full-term infants (Chaturvedi &

Banait, 2000; Khoury, Moazzem, Jarjoura, Carothers, & Hinton, 2005; Singh, 2010). In addition, for these mothers the benefits of breast milk included the lower natural protein content of the amino acid, Phe, and the improved cognitive benefit that could compensate for some of the PKU deficits. The belief in these benefits motivated them to provide their infants with breast milk rather than standard commercial infant formula (Agostoni et al., 2000). Further research is needed to explore breastfeeding in infancy with subsequent cognitive functioning, specifically executive functioning.

The second subcategory of belief was *naturalness of breastfeeding*. One-fifth of the mothers identified that the naturalness of breastfeeding they experienced and the intense attachment that developed while breastfeeding their infants who did not have PKU contributed to their decision to breastfeed in the context of PKU. Researchers (Dewey, Nommsen-Rivers, Heinig, & Cohen, 2002; Feinstein et al., 1986; Hauck, Fenwick, Dhaliwal, & Butt, 2011; Hauck, Fenwick, Dhaliwal, Butt, & Schmied, 2011) have consistently reported that mothers who breastfed previous children were more likely to breastfeed the next child, suggesting that breastfeeding experience plays a role in subsequent breastfeeding. Several mothers described previous success breastfeeding an older child and therefore feeling comfortable with breastfeeding an infant with PKU. This previous success in breastfeeding could be interpreted as increased knowledge of breastfeeding and a positive breastfeeding belief. That mothers linked their belief in the naturalness of breastfeeding with developing a close bond with their infants is not surprising given that other researchers (Feldman,

Weller, Sirota, & Eidelman, 2003; Kim et al., 2011; Rapley, 2002; Whelan & Lupan, 1997) have emphasized the importance of breastfeeding for promoting close maternal-infant contact.

Easy child. The second category of facilitators was the easy child or the child with the easy temperament: adaptable, sociable, and easy-going (Thomas, Chess, & Birch, 1968). Although fewer than 10% of the mothers described having infants with PKU who were adaptable at such a young age, these mothers clearly described factors that facilitated their breastfeeding. Mothers of “easy children” reported being able to easily switch from breastfeeding to bottle feeding and vice versa. Although no research could be located regarding breastfeeding easy children, the literature (Niegel, Ystrom, Hagtvet, & Vollrath, 2008) revealed that difficult infants breastfeed for a shorter duration. Future research should include mothers’ perception of infant temperament and maternal stress to determine the extent to which infant temperament influences breastfeeding infants with PKU.

Breast pump. The breast pump was another important facilitator for breastfeeding. One-third of mothers reported that having a powerful breast pump, such as an electric hospital-grade breast pump, for long-term use contributed to the ease of breastfeeding their infants with PKU. This is consistent with the findings that mothers of preterm infants who had a breast pump that was efficient, effective, comfortable, and convenient were able to establish and maintain their breast milk supply until their premature infants were able to adequately suckle at breast (Meier et al., 2008; Slusher et al., 2007). Although infants with PKU are breastfeeding from one to six times per day after diagnosis

of PKU, the majority of them never exclusively breastfeed. Consequently, these mothers also need the same type of breast pump to maintain their breast milk supply long term.

Supports. The last category of facilitators identified by mothers' breastfeeding infants with PKU was supports, and 25% of these mothers reported that support played a role in making breastfeeding their infants with PKU easier. Mothers received support from three main sources: professional support, social support and other mothers who were breastfeeding or who had breastfed their infant with PKU.

Almost two-thirds of the mothers reported the importance of professional support from dietitians, medical staff, lactation consultants, and clinics. Nearly half of the professional support reported was from dietitians. Although medical staff, lactation professionals, and clinic staff also interacted with mothers, dietitians were the healthcare providers most frequently working with mothers to stabilize Phe levels within the desired appropriate therapeutic range and to determine the amount of breastfeeding each infant with PKU could tolerate due to their residual enzyme activity. The importance of dietitians' support for mothers' perception of positive support is compelling for dietitians' practice in metabolic clinics. Further evidence is needed to support the practice of healthcare providers to provide intervention for mothers who have infants with PKU to enable them to manage Phe levels and meet their breastfeeding goals.

Although mothers revealed that social support was provided, they relied more on professional support during this early time of stabilizing Phe levels. This

finding was similar to that of parents managing the care of pre-adolescent children with chronic conditions (Garwick, Patterson, Bennett, & Blum, 1998). Both professional and social supports provided emotional support. The difference was that professional support provided more helpful information whereas social support provided more practical help. Further research is needed to understand the various types of supports that these mothers need to successfully manage PKU and breastfeed their infants.

For one mother in this study, being able to provide her extra expressed breast milk to another mother to feed her infant provided the impetus to continue breast pumping and, as such, was an activity that was a support to her breastfeeding. This mother was relieved to be able to donate to another infant whose mother could not produce breast milk, and thereby live out her belief in breast milk as the “perfect food” for infants. She expressed, froze, and supplied her expressed breast milk for six months and recruited other mothers to do so as well. For this mother, although her infant could not exclusively breastfeed, she was consoled by being able to feed someone else’s child. Although a review of the literature provided no findings on why mothers donate their expressed mothers’ milk to other infants and/or to breast milk banks, this topic is frequently discussed by mothers on various breastfeeding forums on the Internet. Further research should explore this phenomenon of breast milk donation.

Barriers to Breastfeeding Infants with PKU

Barriers to breastfeeding an infant with PKU included 1) a lack of support, 2) breast pump issues, 3) fluctuating Phe levels, 4) lack of resources and

information, 5) typical breastfeeding issues, and 6) emotional responses. Many of these factors have been identified in the breastfeeding literature. The challenges and barriers are similar to those mothers have breastfeeding healthy, term, non-PKU infants and those mothers breastfeeding infants who are unable to exclusively breastfeed, such as premature infants.

Lack of support. Approximately one-fourth of the mothers surveyed identified a need for more breastfeeding support. Mothers consistently described three types of breastfeeding support needed: 1) *professional*, 2) *social*, and 3) *more resources and information*. A few mothers reported they had no support while breastfeeding their infant with PKU. Unfortunately, research (Kaufman & Hall, 1989) revealed that mothers who have no identified source of support were more likely to stop breastfeeding, thereby making this group of women especially vulnerable to discontinuing breastfeeding.

Nearly one-fifth of the mothers experienced a barrier in *professional support* from healthcare providers who lacked compassion and understanding of the work involved in managing PKU and breastfeeding. They also reported a lack of understanding from healthcare professionals who failed to share the various options available to help mothers successfully manage PKU and breastfeeding. Further research is needed on healthcare providers working with mothers who have infants with PKU to understand what inhibits them from offering these mothers the needed information and support to help them manage Phe levels and continue breastfeeding.

In addition, mothers strongly voiced the need for services from professional lactation consultants. These healthcare providers are breastfeeding specialists and provide education and management to help mothers of healthy, term infants and preterm infants with a variety of issues, from latching on difficulties, painful breastfeeding, low breast milk supply, and inadequate weight gain (Bonuck, Trombley, Freeman, & McKee, 2005; Humenick, Hill, & Spiegelberg, 1998; Thurman & Allen, 2008). They help mothers understand the fundamentals of lactation as well as support their efforts to learn how to breastfeed their infants. They also may help mothers who are returning to work or school to meet their breastfeeding goals. Mothers in this study, however, reported they had to seek out lactation consultants even when their metabolic team knew they had breastfeeding issues. For some mothers, even after locating lactation consultants, the barrier was a lack of education regarding PKU and its implications for breastfeeding. It seems, then, lactation consultants need to be more aware of the issues for breastfeeding infants with PKU in order to better support these mothers to meet their breastfeeding goals. Metabolic clinics manage PKU and, in turn, need to be more aware of the impact their management has on breastfeeding. The addition of lactation support for these mothers would enhance the team support to these unique breastfeeding mothers and alleviate this barrier.

Some mothers identified needing more *social support* from partners, family, and friends. In fact, the perception that family members thought that breast milk was inadequate or unnecessary for their infants and/or that the

introduction of standard commercial infant formula feeding from a bottle would meet their infant's needs was experienced as a lack of support and a barrier to breastfeeding. According to mothers, this barrier emerged when the mother was struggling with breastfeeding. Perhaps family members thought they were providing support by minimizing the value of breast milk and supporting the bottled formula alternative. Nonetheless, previous research (Anlar, Anlar, & Tonyali, 1988; Dormire, Strauss, & Clarke, 1989; Kessler, Gielen, Diener-West, & Paige, 1995; Kloeblen-Tarver, Thompson, & Miner, 2002; Kools, Thijs, & de Vries, 2005; Kools, Thijs, Kester, van den Brandt, & de Vries, 2005) makes it clear that the role of supportive significant others for breastfeeding cannot be over emphasized. Mothers who identify a social support system whose members affirm, aid, and affect in specific practical ways tend to breastfeed longer. A review of the literature revealed no information regarding what support that mothers of breastfeeding infants with PKU need or desire, but previous research of breastfeeding mothers of infants without PKU and the findings in the current study point up the need to better provide professional and social support to these mothers.

Lack of resources and information. In this study, nearly one in five mothers *identified a lack of resources specific to breastfeeding infants with PKU* as a barrier to breastfeeding. For mothers of healthy term infants, there are usually many options for seeking help, such as breastfeeding clinics and postpartum nurses, as well as friends and family. There are many reading sources at the library, in book stores, and articles in *Parenting* and other lay

journals. Even mothers of preterm or critically infants in NICU (level III) or Special Care Nurseries (SCN, step down, level II) have available resources in the nurses who care for their premature or critically ill infants, in-house lactation consultants, and other mothers who have or are breastfeeding/breast pumping for their special care infants. Since Meier's research on breastfeeding the premature infant (Meier, 2001, 2003; Meier et al., 2000; Meier et al., 2008; Meier, Engstrom, Janes, Jegier, & Loera, 2012; Meier et al., 2007), there has been a growing base of evidence providing knowledge to hospital and clinic staff for the smallest and most critically ill infants to receive breast milk. This evidence has also been translated into practice through publications in lay journals as well as pamphlets typically available in the NICU or SCN.

A review of resources for mothers who are breastfeeding infants with PKU revealed “Tips for Breast-Feeding a Baby with PKU” on the PKU News Home page. Unfortunately, this article was last updated in 2006. Only one article for lay audiences, “Breastfeeding the Infant with PKU” was found in the *Journal of Human Lactation* (Duncan & Elder, 1997). Even the newest edition of *Breastfeeding: A Guide for the Medical Profession* (Lawrence & Lawrence, 2010) falls short: the section on breastfeeding infants with PKU cites the Ernest et al. (1980) paper that is no longer available from the Superintendent of Documents, US Government Printing Office, Washington, DC. Consequently, there is a lack of current, evidence-based information for mothers who are breastfeeding in the context of PKU. The findings from the current study will serve to help fill this gap in resources.

Breast pump issues. In this study, nearly half (45%) of the mothers identified breast pumping issues as a barrier while breastfeeding their infants with PKU, and many of these mothers described wanting help to maintain their breast milk supply. Breast pumping is a challenge for mothers of premature and critically ill infants in the NICU and much can be learned from that research (Meier, 2001; Meier et al., 2008; Meier, Engstrom, Patel, Jegier, & Bruns, 2010; Meier et al., 2007). Yes, there is an importance difference in the pattern or trajectory of breastfeeding between mothers of preterm infants and mothers of infants with PKU. Mothers of preterm infants use the breast pump for weeks (or months) until their infants transition to and are eventually able to feed directly from the breast. In contrast, although infants with PKU are breastfeeding from one to six times per day after diagnosis, most of them never exclusively breastfeed due to Phe levels and they necessarily alternate between the breast and bottle feeding. In addition, mothers of infants with PKU must continue to breast pump in order to maintain their breast milk supply whereas mothers of preterm infants may transition to full breastfeeding and stop using a breast pump altogether. It is not surprising then that mothers in the current study consistently described four breast pump issues: 1) *having an inadequate breast pump* for breast milk expression, 2) *lack of knowledge regarding the frequency of breast pumping* required to maintain an adequate breast milk supply when an infant is not exclusively breastfeeding, 3) *the time-consuming chore* to breast pump, and 4) *returning to work*. Having an inadequate breast pump was identified by some mothers as an issue. These mothers described using a hand pump or mini-

electric breast pump, rather than a powerful, electric breast pump for long-term pumping.

Nearly two decades ago, research found that mothers who needed a breast pump for long-term milk expression required a hospital-grade electric breast pump with a double collection kit to sustain their milk supply (Auerbach, 1990; Auerbach & Walker, 1994; Hill, Aldag, & Chatterton, 1999; Johnson, 1983). More current evidence found the hospital-grade Symphony® breast pump, with a multiphase expression technology mimicking an infant at breast and weighing only seven pounds, was as efficient and effective as the classical, hospital-grade electric breast pumps that had only one expression phase and weighed over 20 pounds (Meier et al., 2012). In addition, mothers rated the Symphony® breast pump as more efficient, effective, comfortable, and convenient than the classical hospital-grade breast pump.

Consequently, an efficient and effective breast pump is fundamental to the mother's ability to produce breast milk when her infant is unable to exclusively breastfeed. Given the finding from the current study that an inadequate breast pump was a concern for nearly half of the mothers who were breastfeeding infants with PKU, metabolic clinicians can easily alleviate this barrier by providing breast pump recommendations based on the published evidence and evaluations of breast pumps.

Many mothers reported they lacked the understanding of the importance of *frequent* milk expression when an infant is unable to exclusively breastfeed. In order to maintain a breast milk supply, these mothers must rely on the breast

pump to replace the sucking stimulation and milk removal of a healthy, term breastfeeding infant (Meier et al., 2008). As such, their breast pumping needs are different from those mothers who are occasionally breast pumping and who can depend upon their infants to provide the needed stimulation and removal required for breast milk production (Meier & Engstrom, 2007). Mothers breastfeeding infants with PKU need to have an understanding of the breast pump *frequency* required in conjunction with breastfeeding in order to maintain an adequate breast milk supply.

The lack of understanding about the necessary frequency of pumping the breast may be compounded by the *time* it takes to breast pump. Certainly these barriers were identified separately but to the extent that they co-exist, breast pumping can be a significant challenge. Again, existing breastfeeding literature can inform clinicians; there are numerous papers on bilateral breast pumping in order to significantly reduce the time invested in breast pumping (Auerbach, 1990; Groh-Wargo et al., 1995). Although a review of the literature revealed no information regarding how mothers find the time to breast pump, this topic is frequently discussed by mothers among themselves on breastfeeding forums on the Internet. Metabolic clinicians can likewise be informed.

Many mothers *returned to work* and described breast pumping barriers related to finding an appropriate place to breast pump and the difficulty of finding the time to breast pump while at work. Maternal employment has been established as a barrier to breastfeeding in numerous articles (Auerbach, 1984; Auerbach & Guss, 1984; Gielen, Faden, O'Campo, Brown, & Paige, 1991; Roe,

Whittington, Fein, & Teisl, 1999). The U.S. Department of Health & Human Services Blueprint for Action on Breastfeeding (2011) singled out the workplace as one of the important barriers to extended breastfeeding for employed mothers.

For mothers of infants with PKU, the ability to breast pump is vital to maintaining their breast milk supply. Although employed mothers of healthy term infants return home to exclusively breastfeed during the remainder of their day and night, employed mothers of infants with PKU are unable to do so. Mothers of infants with PKU must provide bottles of Phe-free formula in order to maintain Phe levels within the desired range. Consequently, for these mothers who cannot compensate with exclusive breastfeeding after they return home with their infant, the lack of an appropriate place and/or time to breast pump further diminishes their breast milk supply.

Common breastfeeding issues. Another category of barriers included common breastfeeding issues, which included three dimensions: 1) *common breastfeeding problems*, 2) *nipple confusion*, and 3) *breast milk supply issues*. Of the mothers in this study, 16% identified *common breastfeeding problems* that are typical for new mothers and infants, such as poor latch on, nipple pain, thrush, and sore (cracked, bleeding) nipples as challenges to continuing to breastfeed. Most mothers realized these problems were not PKU-related, and, in fact, the frequency of their report of these concerns was less than in a study where more than one third of mothers identified one or more common breastfeeding problems (Scott, Binns, Oddy, & Graham, 2006). Although these problems are viewed by many as “common,” they represent a potential barrier to

continued breastfeeding and subsequent stopping to breastfeed (Walker, 2008). Their prompt recognition and resolution are essential to alleviating these challenges.

Another barrier for the mothers in this study breastfeeding was *nipple confusion*: 28% reported frustration with nipple confusion exhibited by their infants. Mothers described considerable frustration getting their infants to accept the introduction of bottle feeding when PKU management dictated the need. Many of these same mothers reported that after many weeks of breast and bottle feedings, their infants refused to latch on and breastfeed. At the same time, mothers also reported a waning breast milk supply. These findings are consistent with the description of two forms of nipple confusion: nipple confusion occurring during the neonatal period and nipple confusion occurring after several months of breastfeeding (Neifert, Lawrence, & Seacat, 1995; Newman, 1993). The theory of nipple confusion and its association with early weaning has both supporters and doubters. Recent scientific findings have revealed a change in sucking pattern between breastfeeding and bottle feeding (Mizuno & Ueda, 2003; Weber, Woolridge, & Baum, 1986). More current breastfeeding research investigated infants' sucking patterns across exclusively breastfeeding, exclusively bottle feeding, and mixed feeding of both breast- and bottle-feeding. The patterns were found to be indeed unique and different (Moral et al., 2010). Questions have been raised about whether infants who struggle with bottle feeding after having been breastfed may not have yet developed the mixed sucking pattern (Moral et

al., 2010). Further research is needed, particularly given that infants with PKU are necessarily expected to develop such a mixed pattern.

Although *breast milk supply* is a common breastfeeding problem, little has been reported in the literature about the challenges in maintaining breast milk supply for mothers breastfeeding infants with PKU. Whereas mothers of healthy, term non-PKU infants breastfeed exclusively, mothers of infants with PKU experience problems with providing sufficient amounts of breast milk due to the need to bottle feed Phe-free formula to maintain therapeutic Phe levels. Over one-third of the mothers surveyed revealed breast milk supply as a breastfeeding problem. This finding is similar to those reported by mothers who were breastfeeding healthy term non-PKU infants (Ahluwalia, Morrow, & Hsia, 2005; Hill, 1992; Hill & Aldag, 2007; Hill & Humenick, 1996).

The process of developing an adequate breast milk supply begins during pregnancy when the breasts undergo anatomic and physiologic changes in preparation for breastfeeding (Lawrence & Lawrence, 2010). During the second trimester of pregnancy, lactogenesis I occurs with the transformation of the mammary glands so as to be sufficiently developed and differentiated to secrete small amounts of colostrum (Neville, Morton, & Umemura, 2001). However, colostrum and breast milk secretion is suppressed throughout the remainder of pregnancy by elevated circulating levels of progesterone (Neville & Morton, 2001). After delivery of the placenta, circulating progesterone levels decline rapidly, resulting in lactogenesis II, the onset of breast milk secretion (Neville et al., 2001). With lactogenesis II, breast milk synthesis and secretion are regulated

by a combination of autocrine and endocrine mechanisms dependent on regular and effective removal of breast milk to the feedback inhibitor of lactation mechanism.

For mothers exclusively breastfeeding a healthy, term non-PKU infant, lactogenesis I to lactogenesis II occurs flawlessly. For mothers breastfeeding infants with PKU, this transition is interrupted with the need to provide Phe-free formula exclusively for 24 to 72 hours (or longer) to rapidly reduce significantly elevated Phe levels. The issue of decreasing breast milk supply is not unimportant, given that mothers have reported breast milk supply issues as the most common reason for stopping breastfeeding (Wambach et al., 2005). Further research is needed to explore methods for mothers who must alternate between breast- and bottle-feeding to maintain their breast milk supply for their infant with PKU.

Fluctuating Phe levels. Fluctuating Phe levels presented additional barriers: 1) *increased workload*, 2) *“how much,”* and 3) *time*. Again, these concerns can be identified in the breastfeeding literature with other infants who are unable to exclusively breastfeed, such as premature infants. Although fluctuating Phe levels require changes in PKU management including breastfeeding, PKU management has been described in the literature only from healthcare providers’ perspectives (Cornejo et al., 2003; Greve et al., 1994; Huner et al., 2005; Kanufre et al., 2007; McCabe et al., 1989; Motzfeldt et al., 1999; van Rijn et al., 2003; Yannicelli et al., 1988). This study provides mothers’

perceptions of managing PKU and breastfeeding, and the barriers and concerns involved in the management of PKU are startling.

Almost 60% of mothers in this study reported the *increased workload* associated with breastfeeding, breast pumping, and feeding Phe-free formula on a daily basis as a barrier. Although mothers of preterm infants have reported similar challenges of breast pumping and breastfeeding, the focus for them is on nutrition and hydration to achieve adequate growth (Hill, Ledbetter, & Kavanaugh, 1997; Hurst, Meier, Engstrom, & Myatt, 2004). For mothers breastfeeding infants with PKU, the focus includes the additional need to lower Phe levels to prevent long-term sequelae from elevated Phe levels and consequently requiring supplemental feedings of Phe-free formula to maintain appropriate Phe levels.

Nearly one-third of mothers in this study identified the concern of “*how much*” Phe that infants obtained from breastfeeding. This concern is similar to concerns for “getting enough” breast milk reported by mothers breastfeeding healthy term infants and those breastfeeding critically ill or preterm infants (Hill, Aldag, Zinaman, & Chatterton, 2007; Hurst et al., 2004; Meier et al., 1994; Riordan, Gill-Hopple, & Angeron, 2005). For healthy term non-PKU infants and infants in the NICU, the focus on “how much” concerns the amount of breast milk suckled. For infants with PKU, the focus is not only on “how much” breast milk was suckled but additionally “how much” natural Phe infants obtained from their mothers’ breast milk. Mothers used and tried various methods to determine “how much” their infants with PKU could suckle during breastfeeding and yet maintain

the Phe level within the desired range. Further research is needed to ascertain whether such methods are necessarily individual or if best practices for mothers of infants with PKU may be identified.

Mothers identified the *time*-consuming aspects of breastfeeding, breast pumping, bottle feeding, preparing Phe-free formula, and cleaning breast pump equipment as taxing. Similar aspects of time-consuming activities can be found in the literature on mothers' feeding their preterm infants after hospital discharge (Bakewell-Sachs & Gennaro, 2004; Holditch-Davis & Miles, 1997; Reyna, Pickler, & Thompson, 2006). Further research is needed to understand how mothers handle the feeding process when breastfeeding infants with PKU and, again, to ascertain if these activities are necessarily particular to the individual mother-infant dyad or if there may be some efficiencies to be had from identifying best practices.

Emotional responses. Little is known about maternal emotional responses to having a breastfeeding infant with PKU. In this study, mothers expressed many negative feelings toward specific aspects of breastfeeding their infants with PKU. Mothers of preterm infants and late-preterm infants have reported similar marked emotional responses (Callahan & Hynan, 2002; Holditch-Davis, Cox, Miles, & Belyea, 2003; Holditch-Davis et al., 2009; Miles, Holditch-Davis, Schwartz, & Scher, 2007), and this distress was associated with parenting styles (Fewell & Deutscher, 2002; Singer et al., 2003). The potential for negative emotional responses such as frustration and worry in mothers who were breastfeeding infants with PKU is a concern. More research is needed about

these emotional experiences of breastfeeding, their consequences for mother-infant interaction and parenting, and how mothers cope with these experiences.

Strategies Used by Mothers Breastfeeding Infants with PKU

Mothers identified four categories of strategies they employed to help them manage breastfeeding their infant: 1) seeking information, 2) finding solutions, 3) locating support, and 4) becoming a breastfeeding advocate. These categories are consistent with those found in the breastfeeding literature with healthy, term, non-PKU infants and other infants who are unable to exclusively breastfeed, such as premature infants.

Seeking information. In this study, mothers sought information specifically about breastfeeding infants *with PKU*. Although some mothers described being able to locate breastfeeding information and information about PKU, they were unable to locate online material and published material about breastfeeding infants with PKU. Several mothers located forums, such as the PKU Listserv, which allowed them to post questions, receive information, and provide connections to other mothers who had previously breastfed infants with PKU. This online support is similar to the Australian Breastfeeding Association's breastfeeding support to mothers other than those breastfeeding full-term and preterm infants. With the advent of the Internet, this organization reached out to provide breastfeeding support to mothers who have unusual breastfeeding circumstances. Results revealed that the project helped mothers overcome the problem of geographic isolation, lack of appropriate information and support, and a sense of being alone (Gribble, 2001). Further research should explore whether

the current Internet forum, PKU Listerv, continues to provide the needed breastfeeding support.

Finding solutions. Finding solutions included 1) *adapting bottle feeding*, 2) *use of nipple shields*, 3) *breast pump*, and 4) *Phe tracking*. Some mothers struggled with bottle feeding PKU formula to their infants who preferred to breastfeed and *adapted bottle feeding* to have their infants with PKU receive the needed Phe-free formula. They used droppers and syringes to administer PKU formula to their infants, which took more time but ensured intake of the Phe-free formula to maintain the desired Phe levels. Other mothers diluted the Phe-free formula into expressed mothers' milk to bottle feed to their reluctant infants while others searched for a bottle that their infant would suck. Research is needed to examine the various strategies for bottle feeding Phe-free formula to breastfeeding infants with PKU in order to provide mothers with guidance on how to approach this aspect of management.

Some breastfeeding infants with PKU developed a preference for bottle feeding. Mothers used *nipple shields* (placed over the nipple and areola) to change the maternal nipple presentation to encourage their infants to continue breastfeeding. The use of nipple shields has been found to prevent early weaning (Bodley & Powers, 1996; Nicholson, 1993; Powers & Tapia, 2004; Wilson-Clay, 1996). In fact, there have been successful outcomes for preterm infants transitioning from gavage to breastfeeding using the ultrathin nipple shield (Chertok, Schneider, & Blackburn, 2006; Meier et al., 2000). Healthcare providers working with mothers breastfeeding infants with PKU need to be aware

of indications for nipple shields as it may provide a means to facilitate latching and breast milk transfer. However, nipple shields cannot just be dispensed; healthcare providers must demonstrate proper use and continued follow-up.

Some mothers realized the need to obtain a better *breast pump* for long-term milk expression. For many mothers, the difference between hand and electric breast pumps for milk expression can be a perplexing decision. A hand pump is less expensive, which attracts many mothers to purchase one. Using a hand pump means manually creating the suction; this can be tiring after a few minutes. For mothers needing to pump infrequently, this type of breast pump is adequate. However, for mothers who need to pump regularly for milk expression, an electric hospital-grade electric pump is recommended (Engstrom, Meier, Jegier, Motykowski, & Zuleger, 2007; Meier & Engstrom, 2007; Meier et al., 2008; Meier et al., 2012). A hospital-grade breast pump with a double pump set-up reduces pumping time through faster let-down, milk flow, and milk removal similar to a breastfeeding infant (Kent et al., 2008; Kent, Ramsay, Doherty, Larsson, & Hartmann, 2003; Ramsay & Hartmann, 2005; Ramsay, Mitoulas, Kent, Larsson, & Hartmann, 2005). Given that mothers who have infants with PKU are not able to exclusively breastfeed, they need to be educated about the differences between hand pumps and electric breast pumps to provide them with the best breast pump for their pumping needs.

Locating support. Locating support as a strategy included two groups of people: 1) *metabolic providers*, and 2) *mothers who were breastfeeding or had breastfed infants with PKU*. Although some mothers established a long-term

relationship with their metabolic team of clinicians, several mothers became dissatisfied when they realized their needs and their infant's needs were not being met by their current metabolic providers. These mothers began to explore other metabolic clinics and healthcare providers that would provide a better fit for managing breastfeeding and PKU, and they typically did so before their infants were six months of age. This is not unexpected and is congruent with a previous finding that over two-fifths of those who change their healthcare provider did so because of personal preferences and the desire to obtain better quality care (Reed, 2000). While some mothers sought out mothers who were breastfeeding like themselves, others specifically sought out mothers who had or were breastfeeding infants with PKU. Some mothers located another breastfeeding mother with a child who has PKU within their geographical area, but many mothers in this Internet recruited sample were making Internet connections via online forums, listservs, and PKU organizations. A recent poll (Warzel, 2012) revealed that all mothers are spending a significant amount of time using media and technology platforms, with 28% likely to own a tablet. Online communities have been found to provide the means for instrumental and emotional support (Drentea & Moren-Cross, 2005; Miyata, 2002; Roberts & Fox, 1998; Sharf, 1997). It may be that Internet access to online communities created a supportive network of these mothers who were empowered through discussing issues of mothering with other mothers.

Breastfeeding advocate. The last strategy was becoming a breastfeeding advocate. Many mothers in this study valued the benefits of

breastfeeding and breast milk for their infants. For the majority of the mothers who participated in the survey, breastfeeding was a priority and their actions verified their commitment to breastfeeding their infants with PKU. Mothers who are more likely to successfully breastfeed tend to be educated (college graduate), married/partnered, Caucasian mothers who have a breastfeeding support network (Celi, Rich-Edwards, Richardson, Kleinman, & Gillman, 2005; Chezem, Friesen, & Boettcher, 2003; Heck, Braveman, Cubbin, Chavez, & Kiely, 2006; Riva et al., 1999). Besides these attributes of breastfeeding success, these mothers additionally held a strong commitment to breastfeeding, which predicts continuing (Avery, Zimmermann, Underwood, & Magnus, 2009). Mothers who successfully sustained breastfeeding have confidence in the breastfeeding process, confidence in their ability to breastfeed, and a commitment to make breastfeeding work despite barriers. Future research should include breastfeeding self-efficacy to explore how breastfeeding commitment for mothers breastfeeding infants with PKU may be different from mothers breastfeeding healthy, term, non-PKU infants and to identify strategies for enhancing self-efficacy and commitment.

Labor of Love: Commitment to Breastfeeding in the Context of PKU

Using descriptive thematic analysis, mothers in this study revealed their perceptions of breastfeeding infants with PKU. From their descriptions, themes and sub-themes that described breastfeeding infants with PKU evolved, were combined, and eliminated. These themes and sub-themes were used to describe

their qualitative experience. The following discussion incorporates the themes and sub-themes with a focus on the experience of these mothers as a whole.

As this was a retrospective study of mothers' experiences breastfeeding their infants with PKU, the mothers might have forgotten or minimized events that occurred while breastfeeding their infants, thus introducing some recall bias in the findings. However, this was the first study of what mothers said occurred and what they thought about it. Due to the exploratory nature of this study, no attempt was made in the methods to saturate themes. Further research should be directed at a more exhaustive analysis of the themes.

Sample size. This was a qualitative descriptive study with a small sample size selected from participants from the larger phase 1. Mothers were invited to participate and constituted a purposively selected sample; therefore, it is not representative of all mothers breastfeeding infants with PKU. Data collection was limited to 10 interviews with slightly more than 15 hours of interviewing. This was before saturation of themes; however, due to the nature of the research, the experience of breastfeeding in the context of PKU emerged. Additional interviews are needed to confirm the thematic analysis.

Family functioning. Despite the differences in family functioning, there was little variation in these mothers' experiences breastfeeding their infants with PKU. Although studies have examined family functioning when there is a child with PKU in the family, these studies occurred in the 1960s and 1980s when many of the children in the samples were already cognitively impaired. In those studies, the cognitive impairment, rather than the medical condition of PKU, was

thought to have affected the family functioning (Kazak, Reber, & Carter, 1988; Kazak, Reber, & Snitzer, 1988; Keleske et al., 1967; Lord et al., 2005; Reber et al., 1987; Shulman et al., 1991; Wood et al., 1967). In this study, none of the infants appeared to be cognitively impaired from their mothers' descriptions. Further studies need to include family functioning through the life-span of raising children with PKU who have been identified from newborn screening and who have adhered to the PKU diet recommendation.

Commitment

Commitment was the means for exploring mothers' experiences breastfeeding their infants with PKU. In this study, the theme of commitment characterized these mothers who successfully breastfed their infants with PKU. Although the study included a small group of mothers who were interviewed in-depth, the results provided the development of the central theme, "Labor of love: Commitment to breastfeeding in the context of PKU" to describe their breastfeeding experiences. Several studies found being committed to breastfeeding influenced mothers to persist with breastfeeding (Blyth et al., 2002; Bottorff, 1990; Humenick, Hill, & Wilhelm, 1997; Nelson & Sethi, 2005). Bottorff's (1990) phenomenological study summarized the concept of mothers' commitment to breastfeeding by stating, "For mothers, commitment to breastfeeding is an important part of their continuing to breastfeed. They are not obligated to breastfeed in that they ought to do it; rather, they feel they must because of the commitment" (p. 205).

In this study, the commitment to breastfeeding was evident with mothers' decision to continue breastfeeding even after the diagnosis of PKU. This decision meant adapting breastfeeding to be able to maintain therapeutic Phe levels while persevering to overcome common breastfeeding problems and not-so-common fluctuating Phe levels. When confronted with common breastfeeding problems such as latch-on and cracked, bleeding nipples, these mothers sought support from professional healthcare providers, family, friends, and peers who had or were breastfeeding infants. With the development of nipple confusion, mothers persevered to continue breastfeeding and having their infants' bottle feed Phe-free formula to maintain therapeutic Phe levels. When breast milk supply waned, these mothers reported the main challenge was breast milk supply, yet these mothers persevered to meet their breastfeeding goals of six months or longer.

Fluctuating Phe levels provided many challenges for mothers' breastfeeding infants with PKU including the ever-changing treatment plans, which was another challenge combined with the increased workload of breastfeeding, bottle feeding, and breast pumping. Despite weekly to twice weekly adaptations in breastfeeding plans, these mothers continued with the increased workload to supply their infants with the best natural source of nutrition and Phe even when it was not easy. At any time, these mothers could have stopped or given up breastfeeding. Instead, they made a commitment to breastfeed their infants, which tied these mothers and their infants in a special way. This aspect of laboring out of love and commitment to continue breastfeeding broadens the definition of successful breastfeeding in this unique

group of mothers. Hence, mothers' experience breastfeeding their infant with PKU is characterized by the theme of Labor of love: Commitment to breastfeeding in the context of PKU.

An unexpected and unwanted diagnosis. The mothers in this study all experienced an unexpected and unwanted diagnosis. From the interviews, mothers described three sub-themes of that diagnosis: 1) Out of the blue, 2) Crash course in PKU 101, and 3) Beyond PKU 101. These sub-themes are consistent with those identified in the pediatric literature.

Initially, all mothers described the shock, denial, and disbelief of the diagnosis of PKU in the breastfeeding infant. This finding is similar to that reported by mothers of preterm infants (Costello & Chapman, 1998; Miles, Funk, & Kasper, 1992; Nystrom & Axelsson, 2002; Shields-Poe & Pinelli, 1997), infants with other serious childhood conditions (Beresford, 1994; Shapiro, McCormick, Starfield, & Crawley, 1983), and infants with PKU (Lord, Ungerer, & Wastell, 2008; Waisbren, 1980). In a study of parental adjustment to the PKU diagnosis, the emotional impact of the diagnosis was objectively highlighted (Lord et al., 2008). This study describes mothers' subjective experience of the unexpected and unwanted diagnosis of PKU. Together, both studies point up the importance of acknowledging the emotional impact of the diagnosis on parents.

The "crash course" of intense learning to have a cursory understanding of the basics of PKU allowed mothers to be able to simultaneously manage breastfeeding and PKU immediately. After becoming confident in their understanding of PKU, they sought more detailed information specifically related

to new therapies and long-term concerns, such as executive functioning. Mothers used regional conferences and online medical journals available to them.

Parents of children who have other serious childhood conditions would also be rapidly learning about their conditions, and a review of the literature revealed information on family life (Fawcett, Baggaley, Wu, Whyte, & Martinson, 2005; Yantzi, Rosenberg, Burke, & Harrison, 2001) and managing the chronic condition (Gallo & Knafl, 1998; Gallo, 1990; Kodadek & Haylor, 1990; Sullivan-Boylai, Deatrick, Gruppuso, Tamborlane, & Grey, 2003). Yet literature was not readily identified that discussed how parents with children who have chronic conditions initially learn and continue learning about new therapies and long-term issues related to their children's chronic condition. Again, research efforts directed at learning strategies employed by parents and discerning best practices could provide them with much-needed guidance.

The work of managing PKU and breastfeeding. Breast milk has been determined to be lower in Phe than standard commercial formula (Janas et al., 1985; Lonnerdal et al., 1976; Nayman et al., 1979). Thus, a larger volume of breast milk than standard commercial infant formula can be fed to infants with PKU. In 1983, the first publication appeared describing breastfeeding infants with PKU. Subsequent research on bottle fed infants with PKU compared to breastfed infants with PKU during the first six months showed that those infants with PKU who had been breastfed and supplemented with Phe-free formula had lower protein and Phe levels (van Rijn et al., 2003). In addition, combining breast milk and Phe-free formula in managing the diet of infants with PKU did not adversely

affect Phe control (Greve et al., 1994; Huner & Demirkol, 1996; McCabe et al., 1989). In more recent studies, breastfed infants with PKU had no significant differences in weight gain, daily Phe intake, and the mean plasma Phe concentrations compared to formula fed infants with PKU (Cornejo et al., 2003; Huner et al., 2005; Kanufre et al., 2007; Motzfeldt et al., 1999; van Rijn et al., 2003). The results from these studies consistently supported the conclusion that breast milk supplemented with Phe-free formula is an acceptable dietary treatment for infants with PKU.

Yet none of those studies was from the perspective of mothers who simultaneously manage PKU and adapt breastfeeding to maintain therapeutic Phe levels. This study provides the first thematic description from mothers' perception of their experiences breastfeeding infants with PKU. Clearly this study begins to address understanding the breastfeeding needs of mothers with infants who have PKU. Given the lack of research investigating mothers' experiences breastfeeding such infants, this study provides needed knowledge upon which research-based interventions to facilitate breastfeeding success among these mothers can occur. Research-based interventions are needed to facilitate the strict dietary adherence and required PKU management while adapting breastfeeding to maintain desired Phe levels.

Mothers described the importance of the relationship with the metabolic team while managing PKU and breastfeeding. Every mother stated that she initially felt a high level of trust in the metabolic team. However, as time progressed, some mothers became dissatisfied with their metabolic healthcare

providers and, in the process, began to shop for a metabolic team that would better meet their needs and their infant's needs. Despite their frustrations, mothers developed the ability to work with their original or new metabolic healthcare team. These findings of the importance of the relationship with the metabolic team are consistent with the research concerning healthcare relationships in chronic conditions (Thorne & Robinson, 1988a; Thorne & Robinson, 1989; Thorne & Robinson, 1988b). The relationship with the metabolic team is complicated as mothers need timely information urgently, as they are managing PKU while adapting breastfeeding to their unique situation. It will be important to explore models of working with mothers who have children with PKU, rather than continuing with the standard, traditional models of healthcare that frustrate and dissatisfy mothers.

Accessing support. In this study, mothers sought out resources and supports for breastfeeding their infants. Generally, they located information about breastfeeding and PKU separately but not about breastfeeding infants with PKU. In reviewing the new PKU resource, *My PKU Binder* (National PKU Alliance, 2011), breastfeeding is briefly mentioned as breast milk contains less Phe than standard commercial infant formulas and provides a needed general resource for parents about PKU. This binder does not provide information that mothers need to successfully breastfeed their infants with PKU.

As more research explores mothers' needs and supports for breastfeeding infants with PKU, the growing scientific base will provide an evidence-based approach breastfeeding. In the future, the binder needs to

provide a more detailed section on breastfeeding to prepare mothers for and facilitate their breastfeeding in the context of PKU. In addition, healthcare provider education will be required to give clinicians the tools to support mothers to successfully breastfeed their infant with PKU. Finally, research about human milk and lactation that includes infants with inborn errors of metabolism, such as PKU, will be important to inform clinicians and parents alike.

Normalizing: moving forward. Families with children who have chronic conditions have been the subject of nursing research for over 50 years. From family-centered care that addressed the complex medical needs of children, research moved to the experiences of families with children who have special needs due to the chronic conditions (Branstetter et al., 2008; Gilliss & Knafl, 1999; Knafl & Gilliss, 2002; Litman, 1974; Perrin et al., 2000; Wertlieb & American Academy of Pediatrics Task Force on the, 2003). Subsequently, there was increased recognition of the impact of chronic conditions on families. Researchers reported on family adaptation, stressors, barriers, facilitators, and family functioning in families with children with chronic conditions (Branstetter et al., 2008; Clawson, 1996). Adaptation was viewed as achieving a sense of normalcy in everyday life (Branstetter et al., 2008; Knafl et al., 2010; Knafl et al., 1996; Knafl & Deatrick, 2002; Knafl & Gilliss, 2002). This sense of normalcy in everyday life is consistent with the findings in this study breastfeeding their infants with PKU.

As mothers breastfed their infants with PKU and perceived their infants as healthy and meeting developmental milestones, they viewed their infants as

normal, healthy breastfed infants. By adapting breastfeeding to manage PKU, these mothers began to view their infants not as infants with PKU, but as breastfeeding infants who have PKU. They were thereby normalizing the outcome.

Study Limitations

There are limitations with this study that must be considered. First, the sample was representative of highly educated, married/partnered, Caucasian mothers who may not be representative of the larger population of mothers with children who have PKU in the United States and Canada. However, this sample population was drawn from the Internet and data were collected in part through an Internet survey. The sample characteristics are consistent with those who use the Internet; researchers (Cotton & Gupta, 2004; Fogel, Albert, Schnabel, Ditkoff, & Neugut, 2002) have found that individuals who use the Internet for health and other purposes tend to be younger, more educated, and affluent than those who do not. Thus, mothers who read and post to the PKU Listserv may be more educated than the general population of mothers with children who have PKU, a population for whom demographic characteristics are not known.

In addition, this study was conducted with American and Canadian mothers who chose to breastfeed their infants with PKU. However, the clinical management of infants with PKU and the care provided to mothers breastfeeding those infants are not the same in all institutions, and therefore the maternal experience of breastfeeding infants with PKU will not necessarily be the same. However, commonalities do exist in similar, or like groups, experiencing similar

human experiences such as breastfeeding infants with PKU. Consequently, the thoughts, feelings, and practices described throughout this study, although not generalizable to all mothers of infants with PKU, are anticipated to describe commonalities and highlighted differences in mothers' experiences. Together with the interpretations, it is hoped the findings have revealed meanings of the mothers' experiences breastfeeding infants with PKU.

Another limitation of this study is that over 90% of the participants in the survey and 80% of those in the interviews were Caucasian. All of the mothers who were minorities had been born and raised in the U.S. This study thus enabled exploration of their Western worldview and limits the extent to which this study can shed light on the experiences of other mothers living in different places or different cultures. These findings may have limited relevance to other groups.

Due to the developmental stage and resource needs of adolescent and young adult mothers between 14 and 21 and the considerable variation across states and provinces regarding laws governing age of adulthood and minor emancipation, these mothers were excluded from participating. Exclusion of these adolescent mothers likely missed entire themes of the experience of breastfeeding an infant with PKU.

The survey data in this study were collected between seven months to 27 years after the birth of the child with PKU, whereas the interview data were collected on mothers whose infants were between seven to 35 months old. The long length of time after birth may have affected some mothers' ability to recall their experiences. However, a review of the literature revealed no information

regarding mothers' experiences breastfeeding infants with PKU. This study provided the first exploration of breastfeeding in the context of PKU from a maternal perspective.

Although there was a high level of self-reported willingness to be contacted about participating in mothers' experiences feeding children with PKU survey, approximately one-fifth of initial responders did not complete the survey after receiving the instructions. Another one-fifth of the mothers breastfeeding infants younger than 36 months declined to be contacted about future PKU feeding studies. Of the remaining mothers who agreed to be contacted, only one-third returned emails and agreed to be interviewed. It is unknown what these other mothers might have added to the study and the findings.

Strengths of Design

An important strength of this study was using the Internet as an opportunity for innovative recruitment. The broad geographic distribution and the socioeconomic profile of the mothers recruited from across the U.S. and Canada in a variety of communities from small rural towns to metropolitan cities would not have occurred if the study had been limited to one geographic location. Using the Internet allowed for a unique group of mothers who were waiting to be asked to participate. The success of this method has implications for future nursing research in families with children who have PKU and other rare inborn errors of metabolism disorders.

These study findings enabled the development of a descriptive model of mothers' experiences breastfeeding infants with PKU. The central theme that

emerged was *labor of love: breastfeeding in the context of PKU*. Mothers described this “labor of love” through their narratives, which revealed considerable work to manage both breastfeeding and PKU monitoring and treatment in the face of many barriers and challenges. Even so, their love and commitment to breastfeeding their infant facilitated their search for strategies and solutions to those barriers and challenges, and recognition of factors that eased the process. The findings, then, will contribute both to addressing the needs and barriers identified and to providing proactive education and intervention for mothers of infants with PKU.

The qualitative interviews included only 10 mothers who were predominately white, married, and well educated, thereby limiting the findings. Yet the interviews revealed how mothers manage the work of breastfeeding in the context of PKU as it became a labor of love. This is the first study that described mothers’ experiences breastfeeding infants with PKU and revealed that research on mothers’ experiences of breastfeeding infants in the context of PKU is sorely lacking. Although the incidence of PKU is rare, collectively the incidence of inborn errors of metabolism is common. Consequently, this study is timely in providing preliminary descriptive information regarding the experiences of mothers’ breastfeeding a unique group of infants who are unable to exclusively breastfeed.

Clinical Implications

Breastfeeding benefits infants with PKU from nutritional, gastrointestinal, immunological, developmental, and psychological perspectives. In addition to the

benefits of breastfeeding and breast milk for infants with PKU, human breast milk has a lower concentration of protein and a lower content of the amino acid, Phe, than standard commercial infant formulas making it ideal as the base Phe for infants with PKU. Research (Cornejo et al., 2003; Demirkol et al., 2001; Greve et al., 1994; Hinrichs et al., 1994; Huner & Demirkol, 1996; Kanufre et al., 2007; Motzfeldt et al., 1999; van Rijn et al., 2003) has reported that infants with PKU can breastfeed and maintain appropriate Phe levels for metabolic control and normal growth and development. Consequently, mothers desiring to breastfeed their infants with PKU after diagnosis should be encouraged and supported.

Nurses and other metabolic providers could support mothers to begin breast pumping after the diagnosis of PKU to establish a good breast milk supply. Mothers need to be educated on the early initiation of breast pumping after diagnosis of PKU and the importance of pumping frequency. Research (Freed, Clark, Cefalo, & Sorenson, 1995; Freed, Clark, Curtis, & Sorenson, 1995; Freed, Clark, Lohr, & Sorenson, 1995; Hellings & Howe, 2004; Register, Eren, Lowdermilk, Hammond, & Tully, 2000) has revealed that healthcare providers have limited knowledge and skills related to assisting mothers to breastfeed and provide breast milk to either healthy term infants and preterm infants in the NICU. Although metabolic specialists, nurses, and dietitians are experts in metabolic disorders and therapies, they are probably not experts in human lactation and breastfeeding for mothers of infants with PKU. Consequently, many mothers seek out lactation consultants whose primary training and expertise is in the management of breastfeeding for term infants or preterm infants in the NICU. As

a result, mothers are often “caught in the middle” with conflicting advice in regards to specific PKU lactation problems that they encounter, such as selecting an appropriately powerful breast pump to maintain their breast milk supply versus selecting a breast pump to be used at intervals for returning to work.

As discussed previously, the ability to locate a knowledgeable lactation consultant willing to work with mothers is a challenge. Consequently, to minimize cost while increasing efficacy, the incorporation of using breastfeeding peer counselors (BFPC) as in the NICU and in other settings could be considered as they effectively improve breastfeeding rates (Chapman, Morel, Anderson, Damio, & Perez-Escamilla, 2010; Rossman, 2007; Rossman et al., 2011). Mothers who volunteer as metabolic BFPCs would need training in order to acquire the necessary knowledge and skills to practice safely and effectively in the metabolic clinic. These BFPCs could augment the work of the lactation consultants by performing many of the basic lactation services required in the metabolic clinic. For example, they could aid mothers in locating an appropriate breast pump for long-term use, and they could assume teaching responsibilities of how to use the breast pump, how to clean the collection kit, and how to safely collect, label, and store the expressed mothers' milk. However, more importantly, the BFPC would be a peer of the mother and could help problem-solve many of the lactation and feeding problems that arise while breastfeeding in the context of PKU. Rossman et al. (2011) reported that mothers value BFPCs as “they’ve walked in my shoes.” Another advantage to using BFPCs is for mothers who live a significant distance from the metabolic clinic as mothers have reported the use of the

Internet overcame the problem of geographic isolation, lack of appropriate information and support, and a sense of feeling alone (Gribble, 2001). BFPCs could provide information, reinforcement, reassurance, and encouragement through Internet communication (Thomas & Shaikh, 2007). As BFPCs have made a significant impact on the breastfeeding success for healthy term infants and preterm infants in the NICU, it is time to consider their use in metabolic clinics to provide mothers with newly diagnosed infants with PKU support to continue to breastfeed their infants.

A better practice approach to solving these inconsistencies and barriers in the management of breastfeeding infants with PKU would be to have policies and procedures based on scientific evidence rather than individual opinions of metabolic team members. Most metabolic programs would not tolerate staff members providing information that is based on whether they are “pro” or “con” on monitoring Phe levels to determine metabolic control. Breastfeeding infants with PKU should be no different. These mothers need to have access to lactation consultants who have been educated in the science of lactation and breast milk as well as complicated metabolic situations. Consequently, lactation consultants need to be more aware of the unique breastfeeding needs of mothers whose breastfeeding infants have PKU.

Future Research Directions

New Research

There are numerous avenues for future research that builds on findings from this study. While this study only identified one variable with a negative

impact on breastfeeding duration, many challenges and barriers were identified by the mothers. In the breastfeeding literature, researchers (Dennis, Hodnett, Gallop, & Chalmers, 2002; Kearney, Cronenwett, & Barrett, 1990; Walker, 2008) have reported on the various difficulties with breastfeeding term, healthy infants: sleepy infants, latching difficulties, infants spitting up, and infants breastfeeding too frequently. Compounding these breastfeeding problems were maternal issues: sore nipples, leaking breasts, engorged breasts, mothers feeling sad and crying, sleep deprivation, finding time for self, and perceptions of being isolated and tied down. Yet the most prevalent reported reason for discontinuing or supplementing breastfeeding was insufficient breast milk supply (Hill, 1992; Hill, 1991). Further research is needed to understand 1) the variables that increase the duration of breastfeeding within this unique group of mothers who are unable to breastfeed exclusively due to their infants having PKU; 2) the variables that impact the duration of breastfeeding, especially within the first month in this vulnerable population; and 3) the variables resulting in waning breast milk supply and expression leading to early termination of breastfeeding in this unique population.

Although the focus in this study was mothers' experiences breastfeeding infants, during the interviews mothers consistently moved from breastfeeding to the introduction of foods for an infant at six months, providing a challenge with data collection, analysis, and interpretation. To build on this initial study, a larger mixed methods study could be conducted with a descriptive, cross-sectional design describing mothers' and fathers' experiences raising infants and toddlers

with PKU. Again, mothers can be recruited from across the U.S. and Canada in order to have a representative sample using the PKU Listserv, Emory University, and national and regional PKU advocacy organizations websites. The focus of this study would be to better understand how families manage and adapt to the day-to-day experiences of parenting very young children with PKU. No other study has explored the experience of parenting very young children with PKU. Consequently, this study could provide needed information for appropriate and effective parenting interventions.

The day-to-day experiences of parents raising very young children under the age of 4 years with PKU would be explored through the following ideas: 1) In what ways does family PKU management influence the mother-child relationship and management of the young child's daily routine and developmental needs in the face of emerging autonomy? 2) What resources do families of young children with PKU have access to and use to facilitate parenting this developmental transition? 3) What unique concerns and stressors do mothers have with the day-to-day routine management of PKU as their infants mature into toddlerhood and pre-school? As in the previous study, an Internet survey could be used to obtain self-report data. Data from the structured measures would provide insights into the lives of families with children who have PKU, and these data would contribute to a more complete understanding of feeding in the context of PKU.

Maternal concerns with PKU-related management and feeding could be measured with the modified Family Management Measure (FaMM) to be a FaMM PKU Breastfeeding instrument for mothers breastfeeding/breastfed infants with

PKU and the original FaMM instrument for exclusively formula fed infants and children with PKU (Knaf, Dixon, Grey, O'Malley, & Knafl, unpublished manuscript). In addition, mothers breastfeeding infants with PKU could complete the Breastfeeding Self Efficacy (Dennis, 2003). The Parental Stress Index could be used to measure parental stress as perceived by the mothers (Abidin, 1990). The Medical Time Use Questionnaire and the Parent Restriction Questionnaire could be used to measure parents' use of time and restrictions placed on them that might directly influence the management of the infant's daily routine and developmental needs (Turner-Henson, Holaday, & Swan, 1992). Two instruments, the Health Care Resource Inventory and the Coping Health Inventory for Parents, could describe types of resources used by the mothers (Arnold, O'Leary, Wolff, & Acker, 1993; McCubbin et al., 1983). The Family APGAR and the General Functioning scale of FAD would assess family functioning (Epstein et al., 1983; Smilkstein, 1978). A subset of mothers from across Canada and the U.S. could be interviewed regarding the transition to self-feeding in young children with PKU. In addition, to measure the influence PKU might have on mother-child interactions, a subset of the mothers could be asked to participate in an in-home interview during the child's snack or lunch time in order to conduct a feeding observation. If the child were less than a year of age, the Parent-Child Interaction Feeding Scales (PCI) (Summer & Spietz, 1994) could be used; if the child were between one and four years of age, the Toddler Snack Scale (TSS) (Hodges, Houck, & Kindermann, 2009) could be used to describe the mother-child interaction during the feeding process. Lastly, a subset

of fathers could be recruited to participate in telephone interviews to explore their initial response and concerns associated with the child's PKU diagnosis, describe the process of learning the management of PKU, and describe the strategies they use to incorporate the daily management of PKU into family life.

Further Analysis of the Data Set

There remains a large amount of data from the original study that has yet to be analyzed, interpreted, and presented. In terms of duration of breastfeeding in the survey, over 20% of the mothers ($n = 19$) stopped breastfeeding less than 30 days after the PKU diagnosis. Analysis of their demographics and responses to the four sets of open-ended questions might provide further insight about the challenges and barriers to breastfeeding infants with PKU.

In the original data set, 16 mothers who had two or more children with PKU responded to the survey. Analysis of their demographics, duration of breastfeeding, and open-ended responses might provide further insight about successful strategies for breastfeeding infants with PKU.

In the open-ended responses and qualitative interviews, mothers reported and described interactions with healthcare providers. Descriptive thematic analysis could be used to identify the mothers' perceptions of helpful and unhelpful behaviors from healthcare providers that they received while managing PKU and breastfeeding their infants.

Summary

This mixed methods research study was designed to describe the

1) prevalence and duration of breastfeeding infants with PKU in the U.S. and Canada and 2) to generate knowledge about how mothers of these infants simultaneously manage breastfeeding and PKU therapy and how they perceive family functioning among a sample of mothers who had breastfed infants with PKU.

In this study, mothers successfully breastfed infants with PKU. Despite the increased workload, the initiation and duration rates of breastfeeding were remarkable for these mothers. The only variable significantly associated with decreased duration of breastfeeding for infants with PKU was when standard commercial infant formula was added into the diet replacing breastfeeding or expressed mothers' milk. In addition, the overall reduction in the proportion of mothers' breastfeeding before and after diagnosis between the U.S. and Canada was significant because of the number of mothers from the U.S. who stopped breastfeeding.

Mothers' experiences breastfeeding infants with PKU resulted in the description from two perspectives, quantitative and qualitative. The quantitative analysis revealed *Breastfeeding and PKU: The maternal experience* while the qualitative analysis resulted in the *Labor of love: Commitment to breastfeeding in the context of PKU*. Both perspectives provided similar yet different perspectives of breastfeeding infants with PKU. The quantitative analysis identified the facilitators and barriers mothers encountered and the strategies they used while breastfeeding infants with PKU. The qualitative analysis described the experience breastfeeding from the perception of the mothers.

Recommendations for clinical care include supporting mothers to continue to breastfeed after the diagnosis of PKU and helping them maintain an adequate breast milk supply were identified as most serious challenges while breastfeeding infants with PKU. These mothers need to begin breast pumping after the diagnosis of PKU to establish a good breast milk supply. They need to be educated on the early initiation of breast pumping after diagnosis of PKU and the importance of pumping frequency. Finally, they need to have a powerful electric breast pump for long-term use that allows them to simultaneously pump their breasts and effectively empty them.

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LIST OF APPENDICES

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Appendix A.

Initial Email posting on PKU Listserv

Hello. I am a doctoral candidate in nursing at Oregon Health & Science University (OHSU), Portland, Oregon. For my dissertation, I am exploring "Mothers' Experience of Feeding Infants with PKU." The National Institute of Nursing Research funds this project. I am seeking mothers who have fed infants with PKU.

Are you . . .

- A mother of a child/children who have PKU?
- Live in the United States or Canada?
- Read and write in English?
- Are at least 21 years of age?

If so, you might qualify for a study to explore mothers' experience of feeding infants with PKU. It is anticipated that it will take less than 20 minutes to complete the survey. After completion of the survey, you will be emailed a \$10 (US) Cambrooke Foods electronic gift certificate.

The Principal Investigator is Gail Houck, PhD, RN, PMHNP. The OHSU IRB number is 4107. If you are interested in participating, please contact Sandra A. Banta-Wright, MN, RN, NNP by email at bantawrs@ohsu.edu.

Appendix B.
Information Sheet for Phase 1

IRB # 4107

Protocol Approval Date: 10/25/2010

OREGON HEALTH & SCIENCE UNIVERSITY**Information Sheet****TITLE:** Mothers' Experience Feeding Infants with Phenylketonuria (PKU)**PRINCIPAL INVESTIGATOR:** Gail Houck, PhD., PMNNP 503.494.3825**CO-INVESTIGATORS:**

Sandra A Banta-Wright, RMN, RN, NNP	503.703.1895
Kathleen A Knafl, PhD., FAAN	919.843.3175
Nancy Press, PhD.	503.494.2533
Robert D. Steiner, MD.	503.494.2783

SPONSOR: National Institute of Nursing Research, National Institutes of Health**PURPOSE:**

You have been invited to be in this research study because you are the mother of a child/children with phenylketonuria (PKU). The purpose of this study is to identify the methods of mothers feeding infants with PKU. This study may provide information that will help nurses and other health care professionals to better understand how mothers of infants with PKU manage feedings and PKU.

We plan to enroll about 250 participants in this study that is being conducted at Oregon Health & Science University (OHSU).

PROCEDURES:

You will complete one survey for each of your children with PKU.

RISKS AND DISCOMFORTS:

A code number will be assigned to you and to information about you. Only the investigators named on this consent form will be authorized to link the code number to you. Neither your name nor your identity will be used for publication or publicity purposes.

Although we have made every effort to protect your identity, there is a minimal risk of loss of confidentiality due to the rarity of PKU. All other parties including employers, insurance companies, personal physicians, and relatives will be refused access to the information unless you provide written permission or unless we are required by law to do so.

BENEFITS:

There will be no direct benefit to you for participating in this study. However, by participating, you may help us learn to help other mothers feeding infants with PKU in the future.

ALTERNATIVES:

You may choose not to be part of this study; your decision will be anonymous.

CONFIDENTIALITY AND PRIVACY OF YOUR PROTECTED HEALTH INFORMATION:

All information you provide will be kept confidential with two exceptions: 1) according to Oregon Law, suspected child abuse must be reported to appropriate authorities or 2) when a child, such as an infant, might be physically harmed if information were kept secret (such as significant physical illness, risk of injury, or suicidal thoughts).

Infant neglect or harm. In the event that information is discovered regarding abuse or neglect of an infant, we are legally required to report that information to the Child Protective Services in the mother's state of residence.

Research records may be reviewed and copied by the OHSU Institutional Review Board and the Office for Human Research Protections.

COSTS:

There is no cost to you for being in this study. After completion of the survey, you will receive a \$10.00 (US) Cambrooke Foods electronic gift certificate to thank you for your time and effort given to this research project.

PARTICIPATION:

If you have any questions regarding your rights as a research subject, you may contact the OHSU Research Integrity Office at (503) 494-7887.

You do not have to join this or any research study. If you do join, and later change your mind, you may quit at any time. If you refuse to join or withdraw early from the study, there will be no penalty or loss of any benefits to which you are otherwise entitled.

The participation of OHSU residents or employees in OHSU research is completely voluntary and you are free to choose not to serve as a research subject in this protocol for any reason. If you do elect to participate in this study, you may withdraw from the study at any time without affecting your relationship with OHSU, the investigator, the investigator's department, or your grade in any course.

Completion of the survey will be considered evidence of your agreement to participate in the survey research.

This Information Sheet is yours to keep.

Appendix C.

Instructions for Accessing SurveyMonkey

Thank you for your inquiry regarding the internet survey, Mothers' Experiences Feeding Infants with PKU."

Nurses, doctors, and dietitians want to learn from mothers of the infants they care for. Your experience with feeding your infant may help nurses, doctors and dietitians understand better how mothers manage feedings and PKU.

Here is a link to the survey:

<http://www.surveymonkey.com/s/feedinginfantswithpku>

This link is uniquely tied to this survey. Copy the link into your browser and you will be sent to the internet survey. Please let other mothers of infants with PKU know about this internet survey and have them contact Sandra Banta-Wright, MN, RN, NNP either by email at bantawrs@ohsu.edu or via a dedicated telephone number> 503.703.18995.

Thank you for your participation.

Appendix D.

Email to Mothers who Identify Willingness to Participate in Phase 2

Hello [potential participant's name],

My name is **Sandra Banta-Wright** and I am a doctoral candidate in nursing from Oregon Health & Science University. The reason I am emailing is that you agreed to consider to participate in a follow up study on Mothers Experience Feeding Infants with PKU. This phase of the study is **Mothers' Experience Breastfeeding Infants with PKU**. The purpose of this study is to understand how mothers manage breastfeeding an infant with the disorder of phenylketonuria (PKU). The incorporation of breastfeeding into the management of PKU has not been well studied. This study will identify and describe the day-to-day management experiences of mothers who are breastfeeding infants with PKU and identify the factors that contribute to the experiences of mothers' success in breastfeeding their infants with PKU while adhering to the PKU management plan.

I am currently seeking volunteers as participants in this study. This study phase involves participating in two interviews. The first interview is information about you and your family to provide us with a better picture of who is participating in the study. This part of the study should take less than 30 minutes. The second interview is to explore your experience of breastfeeding an infant with PKU using an interview guide. After the interview, there are two short questionnaires for you to complete. After I have read each question, I will record your answer. Participation in this part of the study will take approximately 60 minutes of your time. I will audiotape both the first and second interviews for later transcription. At the completion of the study, all audiotapes will be destroyed. After the second interview, I will send you a \$20 gift certificate for Target or Starbucks.

If you are interested in participating in this study phase, please contact me at bantawrs@ohsu.edu or via my cell telephone at 503.703.1895.

Thank you for your consideration to participate in PKU research.

APPROVED: **Oct. 25, 2010**

Appendix E.
Telephone Script

Telephone Interview Guide #1

“Hello **[potential participant's name]**, my name is **Sandra Banta-Wright** and I am a doctoral candidate in nursing from Oregon Health & Science University returning your email/call. The reason I am calling is that you contacted me regarding the study, **Mothers’ Experience of Breastfeeding Infants with PKU**. The purpose of this study is to understand how mothers manage breastfeeding an infant with phenylketonuria (PKU). The incorporation of breastfeeding into the management of PKU has not been well studied. This study will identify and describe the day-to-day management experiences of mothers who are breastfeeding infants with PKU and identify the factors that contribute to the experiences of mothers' success in breastfeeding their infants with PKU while adhering to the PKU management plan.”

I am currently seeking volunteers as participants in this study and I wondered if you are still interested in hearing more about it”.

[IF NO] “Thank you, good-bye.”

[IF YES] “This study involves participating in two interviews. In the first interview I will ask you some questions about you and your family to provide me with a better understanding of who is participating in the study. This part of the study should take less than 30 minutes. The second interview is to explore your experience of breastfeeding an infant with PKU. I will ask you a series of open-ended questions about your experiences. After we have finished talking about your experiences, I will ask you questions from two short questionnaires about satisfaction with family life. Participation in this part of the study will take approximately 45-60 minutes, depending on how much you have to say about your breastfeeding experiences. I will audiotape both the first and second interviews for later transcription. At the completion of the study, all audiotapes will be destroyed. After the second interview, I will send you a \$20 gift certificate for Kroger, Target or Starbucks, whichever you prefer.”

“Would you be interested in participating?”

[IF NO]: “Thank you, good-bye.”

[IF YES]: “Thank you; I appreciate your interest in our research. Now I will read the consent form for this study. **[Read Consent Form, pausing after each section to be sure the recruit seems to understand what has been read]**.”

[IF NO] “Thank you and good bye.”

[IF YES] [If contacted by email, participant will receive email with one attachment that will contain a copy of Consent form.] “Let me clarify your email address so that I may send to you a copy of the Consent form.” [If contacted by telephone,

will inquire if able to email forms to participant. If able to email, participant will receive email with one attachment as described previously.] “As you contacted me by telephone, I was wondering if you have access to email? If so, I will email you a copy of the Consent form.” [If unable to email forms, will obtain participant’s mailing address and mail copy of form to participant.] “What address would you like me to mail you a copy of the Consent form?”

“There are two parts to this interview. The first part of this interview is to gather some information about the people who take part in my research, so I can describe my sample when I write my final report. If there are any questions you prefer not to answer, you do not have to. **[Go over Internet Survey Form]**. The second part of this study is an interview regarding your experience of breastfeeding your infant with PKU. I have a session open on **[day and date]** at **[time, a.m. or p.m.]**. Will you be available then?”

[IF NO]: Offer another day and time until one is found that is mutually convenient.

[IF YES]: “This is great. Let me give you some important details about the study. This study is called Mothers’ Experience of Breastfeeding Infants with PKU and my name is Sandra Banta-Wright. I will call you on **[mention day and date]**.”

[IF NO]: Renegotiate date, day and time

[IF YES]: “Great! The day before your session, I will contact you by email or telephone, which is your preference, as a reminder. However, in the meantime, if you discover you will be unable to keep this appointment, please call me at **503.703.1895** and leave a message if I am not available or email me at **bantawrs@ohsu.edu**. I would appreciate knowing as soon as possible if you are unable to keep the appointment.

“I look forward to talking you on **[mention day, date and time again]**. Thank you very much again for helping us with our research”.

Telephone Interview Guide #2

“Hello **[participant's name]**, this is **Sandra Banta-Wright**. I am calling to complete the second interview for the study, **Mothers’ Experience of Breastfeeding Infants with PKU**. Is this still a convenient time to continue with this interview?”

[IF NO]: Offer another day and time until one is found that is mutually convenient.

[IF YES] “This is great. The second interview is to explore your experience of breastfeeding an infant with PKU. This part of the interview is very open-ended. I want to understand what’s it’s been like for you to breast feed your baby with PKU. After we have finished talking about your experiences, I will ask you questions from two short questionnaires about satisfaction with family life. Participation in this part of the study will take approximately 45-60 minutes, depending on how much you have to say about your breastfeeding experiences. I will audiotape both the first and second interviews for later transcription. At the completion of the study, all audiotapes will be destroyed. After the second interview, I will send you a \$20 gift certificate for Kroger, Target or Starbucks, whichever you prefer.”

“To refresh your memory, the purpose of this study is to understand how mothers manage breastfeeding an infant with the disorder of phenylketonuria (PKU). The incorporation of breastfeeding into the management of PKU has not been well studied. This study will identify and describe the day-to-day management experiences of mothers who are breastfeeding infants with PKU and identify what mothers believe contributed to their breast feeding success as well as what they thought the challenges were.

Breastfeeding the Infant with PKU (Defining the Situation)

First I would like to ask you a few questions about your infant and when you found out about the PKU. Let’s talk by talking a little about your baby....

- 1) Tell me about your infant who has PKU.
- 2) Tell me how the initial few days of breastfeeding went with this infant
- 3) Tell me about your current understanding of PKU.

Probes, if conversation is difficult to start:

- a.) Tell me about when your infant was diagnosed with PKU.
- b) What was your initial reaction to the diagnosis?

- c) Tell me how you explained PKU to members of your family and friends
- 4) Tell me how breastfeeding changed once the diagnosis of PKU was made
- 5) Open-ended conversational probes,
 - a) For example
 - b) Tell me more about that
 - c) Anything else?

Management Behaviors

Now I have some questions about how you manage breastfeeding and PKU. Let's start with your decision to continue breastfeeding

- 1) Tell me about your decision to continue breastfeeding your infant after the diagnosis.

Probes – Encouraged/discouraged from continuing?

- 2) Tell me what it has been like for you to breastfeed an infant with PKU

Probes, if conversation is difficult to start:

- a) Tell me what happens/happened on a day-to-day basis with breastfeeding your infant with PKU – schedule/routine; what is usual routine?
- b) Tell me what makes/made it easy to continue to breastfeed your infant with PKU
- c) Tell me what makes/made it a challenge to continue to breastfeed your infant with PKU
- d) Tell me about the support or lack of support you received to continue to breastfeed your infant with PKU (professionals; friends/family; sources of information)
- e) Tell me what would have made it easier for you while you were breastfeeding your infant with PKU

I'd also like to know about the special things to do to monitor and take care of the baby's PKU. You may have already talked about some of these things, but I want to make sure you have a chance to tell me more if you like.

3) Tell me what it has been like for you to manage your infant's PKU

Probes, if conversation is difficult to start:

- a) Tell me what happens on a day-to-day basis with your infant's PKU
- b) Tell me what makes/made it easy to manage your infant's PKU on a daily basis
- c) Tell me what makes/made it a challenge to manage your infant's PKU on a daily basis
- d) Tell me about the support or lack of support that you received in managing your infant's PKU
- e) Tell me what would have made it easier for you to manage your infant's PKU and continue to breastfeed
- f) Tell me how PKU affects your family's budget

4) Open-ended conversational probes,

- a) For example
- b) Tell me more about that
- c) Anything else?

Perceived Consequences

In these last questions in this part of the interview, I want to talk about how you think having an infant with PKU may have affected your family life. Of course having a new baby is a big change for a family, but I am especially interested in how the infant's PKU may have changed things even more.

1) Tell me about the experience of PKU for your family.

Probe, if conversation is difficult to start:

- a) Tell me how your family deals/dealt with having a breastfeeding infant with PKU

2) Tell me about your perception of changes in how you parent or take care of the child with PKU

Probes if conversation is difficult to start:

- a) Tell me about parenting or taking care of a child with PKU

- b) Tell me about the challenges of parenting or taking care of a child with PKU
- c) Tell me whether having a child with PKU makes parenting more stress
- d) Tell me about the rewards of parenting or taking care of a child with PKU

3) Tell me how PKU impacts or affects your family

Probe if conversation is difficult to start:

- a) Tell me about how your family functions having a child with PKU
- b) Tell me about family relationships with having a child with PKU

4) Tell me about your family's experience with PKU

Probe, if conversation is difficult to start:

- a) Tell me what happens on a day-to-day basis with your family due to breastfeeding infant with PKU
- b) Tell me about family outings with a child who has PKU
- c) Tell me about family holiday experiences with a child who has PKU

5) Tell me how you think PKU will affect your child as he/she grows up.

6) Tell me how you think PKU will affect your family as your child with PKU continues to grow and mature

7) Tell me how PKU affects your family's budget.

Probe if conversation is difficult to start:

- a) Tell me about the financial issues of raising a child with PKU
- b) Tell me about the financial drain of raising a child with PKU

8) Open-ended conversational probes,

- a) For example
- b) Tell me more about that
- c) Anything else?

Thank you for your time to participate in this interview. Is there anything else that you would like to add about your breastfeeding experience with an infant who has PKU?

Appendix F.
Consent for Phase 2

OREGON HEALTH & SCIENCE UNIVERSITY

Consent and Authorization Form

TITLE: Mothers' Experience of Breastfeeding Infants with Phenylketonuria

PRINCIPAL INVESTIGATOR: Gail Houck, PhD, PMHNP 503.494.3825

CO-INVESTIGATORS:

Sandra A. Banta-Wright, MN, RNC, NNP	503.703.1895
Nancy Press, PhD	503-494-2535
Kathleen A. Knafl, PhD, FAAN	919.843.3175
Robert D. Steiner, MD	503.494.2783

SPONSOR: National Institute of Nursing Research, National Institutes of Health

PURPOSE:

You have been invited to be in this research study because you are the mother of an infant who has phenylketonuria (PKU). The purpose of this study is to identify and describe the day-to-day experiences of mothers who are breastfeeding infants with PKU. This study may provide information that will help nurses and other health professionals better understand how mothers of infants with PKU manage breastfeeding.

The long-term goal of our research is to create guidelines for breastfeeding infants with PKU. Mothers will be recruited from across the United States and Canada to participate. Being part of the study will involve a telephone interview for approximately 1-2 hours. We expect about 50 subjects to participate in this study phase that is being conducted at OHSU.

PROCEDURES:

The study involves two or three interviews. During the first telephone call, we will ask you questions to determine if you are eligible to participate in the study. Next, we will either email or mail you a copy of the consent form and two family assessment measures. We will need to schedule a time for a second interview. We will read the consent with you. We will also ask you questions about your race and ethnicity and which clinic you use. We will schedule a time for the face-to-face interview and determine your preferred private location for the interview.

You will receive either a telephone call or an email 24 hours prior to the interview to remind you of the date and time of the interview. During the interview, you will discuss your experience of breastfeeding an infant with PKU. An interview guide will be used. After completion of the interview, you will be asked if you have any questions regarding completing the two family assessments: the Family APGAR and the General

Functioning Scale of the Family Assessment Device Questionnaires. After completion of the two family assessments, the participant will email the completed assessments back to Ms. Banta-Wright at bantawrs@ohsu.edu. It is anticipated that the two family assessments will take less than 15 minutes to complete. The interview will last 60-120 minutes, will be tape-recorded, and typed up.

The information will be kept in a locked file drawer or on a password locked computer. If you have any questions regarding this study now or in the future, contact Sandra A. Banta-Wright at 1.503.703.1895 or Dr. Gail Houck at 1.503.494.3825..

RISKS AND DISCOMFORTS:

You might find it upsetting or painful to talk about what it is or was like to breastfeed an infant with PKU. If you become sad and want to talk about your feelings with another person, such as a counselor, we will help you do so.

BENEFITS:

You may or may not personally benefit from being in this study. However, by serving as a participant, what we learn in this study may help doctors, nurses and dietitians help other mothers breastfeeding infants with PKU in the future.

ALTERNATIVES:

You may choose not to be in this study. You may stop taking part in the study at any time. Talking to the co-investigator about your breastfeeding experience will not affect your health care.

CONFIDENTIALITY AND PRIVACY OF YOUR PROTECTED HEALTH INFORMATION:

All information you provide will be kept confidential with two exceptions: 1) according to Oregon Law, suspected child abuse must be reported to appropriate authorities or 2) when a child, such as an infant, might be physically harmed if information were kept secret, such as suicidal thoughts, significant physical illness or risk of injury.

Infant neglect or harm. In the event that information is discovered regarding abuse or neglect of an infant, we are legally required to report that information to the Child Protective Services in the state of residence of the mother.

We will not use your name or your identity for publication or publicity purposes. All information you provide will be identified only with a code number and never linked to your name. Audiotapes will be destroyed at the completion of the study.

COSTS:

You do not have to pay to be part of this study. After the completion of the interviews and returning the questionnaires, you will receive a \$20.00 (US) Kroger, Target or Starbucks gift certificate to thank you for your time and effort to this research project.

LIABILITY:

If you are harmed in the study, please contact Sandra A. Banta-Wright (503.703.1895) or Dr. Gail Houck (503.494.3825).

It is not the policy of the U.S. Department of Health and Human Services to compensate or provide medical treatment for human subjects in the event the research results in physical injury.

You have not waived your legal rights by signing this form. If you are harmed by the study procedures, you will be treated. Oregon Health & Science University does not offer to pay for the cost of the treatment. Any claim you make against Oregon Health & Science University may be limited by the Oregon Tort Claims Act (ORS 30.260 through 30.300). If you have questions on this subject, please call the OHSU Research Integrity Office at (503) 494-7887.

PARTICIPATION:

You do not have to join this or any research study. If you do join, and later change your mind, you may quit at any time. If you refuse to join or withdraw early from the study, there will be no penalty or loss of any benefits to which you are otherwise entitled.

Sandra A. Banta-Wright (503.703.1895) or Dr. Gail Houck (503.494.3825)

has offered to answer any questions you may have about this study. If you have any questions about your rights as a member of this study, you can call the OHSU Research Integrity Office at 503.494.7887.

Your consent to take part in this study and your permission to let us use your protected health information are voluntary. You may refuse to this consent and authorization form. If you refuse this consent and authorization form, your health care and relationship with OHSU will not be affected. However, you will not be able to enter this research study.

We will give you a copy of this form.

SIGNATURES:

Your signature below indicates that you have read this entire form and that you agree to be in the study.

OREGON HEALTH & SCIENCE UNIVERSITY INSTITUTIONAL REVIEW BOARD PHONE NUMBER (503) 494-7887 CONSENT/AUTHORIZATION FORM APPROVAL DATE Oct. 26, 2011 Do not sign this form after the Expiration date of: <u>10/25/2012</u>

_____	_____
Date	Participant's Signature

_____	_____
Date	Person Obtaining Consent's Signature

Date copy given to participant

Appendix G.

Quantitative Measures for Phase 2

FAMILY APGAR

Please identify which response is most appropriate for each statement by marking with an X in the appropriate column. "Family" is defined in this questionnaire as the individual(s) with whom you usually live.

	Almost Always (2)	Some Of The Time (1)	Hardly Ever (0)
I am satisfied that I can turn to my family for help when something is troubling me.	_____	_____	_____

Comments:

I am satisfied with the way my family talks over things with me and shares problems with me.	_____	_____	_____
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Comments:

I am satisfied that my family accepts and supports my wishes to take on new activities or directions.	_____	_____	_____
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Comments:

I am satisfied with the way my family expresses affection and responds to my emotions, such as anger, sorrow or love.	_____	_____	_____
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Comments:

I am satisfied with the way my family and I share time together.	_____	_____	_____
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Comments:

General Functioning Scale of the Family Assessment Device

Instructions: This assessment contains a number of statements about families. Reach each statement carefully, decide how well it describes your own family, and mark one response for each statement. You should answer according to how you see your family.

For each statement there are four (4) possible responses:

Strongly Agree (SA)	Check SA if you feel that the statement describes your family very accurately
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Agree (A)	Check A if you feel that the statement describes your family for the most part.
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Disagree (D)	Check D if you feel that the statement does not describe your family for the most part.
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Strongly Disagree (SD)	Check SD if you feel that the statement does not describe your family at all.
------------------------	---

The answer spaces for the statement would look like this. For each statement, there is an answer space below.

Try not to spend too much time thinking about each statement, but respond as quickly and as honestly as you can. If you have difficulty, answer with your first reaction. Please be sure to answer every statement and mark your answers in the space provided below each statement.

1. Planning family activities is difficult because we misunderstand each other.
_____ SA _____ A _____ D _____ SD
2. In time of crisis, we can turn to each other for support.
_____ SA _____ A _____ D _____ SD
3. We cannot talk to each other about sadness we feel.
_____ SA _____ A _____ D _____ SD
4. Individuals are accepted for what they are.
_____ SA _____ A _____ D _____ SD
5. We avoid discussing our fears and concerns.
_____ SA _____ A _____ D _____ SD
6. We can express feelings to each other.
_____ SA _____ A _____ D _____ SD
7. There are lots of bad feelings in the family.
_____ SA _____ A _____ D _____ SD
8. We feel accepted for what we are.
_____ SA _____ A _____ D _____ SD
9. Making decisions is a problem for our family.
_____ SA _____ A _____ D _____ SD
10. We are able to make decisions about how to solve problems.
_____ SA _____ A _____ D _____ SD
11. We don't get along well together.
_____ SA _____ A _____ D _____ SD
12. We confide in each other.
_____ SA _____ A _____ D _____ SD