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Challenges to Breastfeeding Infants With Phenylketonuria¹

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Breastfeeding;
Challenges;
Infant–Newborn;
Phenylketonuria;
Phenylalanine

Breastfeeding duration for infants with phenylketonuria (PKU) is less than other full-term infants. However, no study has examined the challenges encountered by mothers' breastfeeding infants with PKU. In 75 mothers of a child with PKU, three categories of breastfeeding challenges were identified: common breastfeeding issues, breastfeeding and PKU, and no challenges. The common breastfeeding issues can be identified in the literature but for these mothers, the issues are heightened due to frequent phenylalanine (Phe) monitoring. Even so, many mothers adapt breastfeeding to maintain desired Phe levels. A few mothers had no issues and were the exception, not the norm.

Background

PHENYLKETONURIA (PKU) IS an autosomal recessive disorder involving amino acid metabolism. The enzyme phenylalanine hydroxylase (PAH) which is required for metabolizing phenylalanine (Phe) to tyrosine in the liver is defective in PKU. This results in an excess of Phe in the blood and eventually the brain. Accumulation of Phe and its metabolites without proper intervention in the neonatal period results in progressive cognitive–neurologic damage in the infant. The signs of untreated PKU develop gradually and are not noticed until irreversible cognitive insult has occurred. In contrast, when identified via newborn screening and treatment begun prior to 1 month of age, cognitive–neurologic development is essentially normal. The recommended treatment for PKU is a Phe restricted diet for life.

In the past, mothers with newly diagnosed infants with PKU were not allowed to breastfeed since it is difficult to precisely estimate Phe intake with breastfeeding. However, breastfeeding may be advantageous since Phe concentration is lower in breast milk (2,482 $\mu\text{mol/L}$) than standard commercial infant formula (4,419 $\mu\text{mol/L}$) making it possible to utilize a higher proportion of natural protein (Lonnerdal et al., 1976; McCabe et al., 1989). For infants with PKU, Phe restriction means breast milk (which contains protein and therefore Phe) must be limited and recommended intake based upon weekly blood Phe levels. With each Phe level, breastfeeding is adapted to maintain desired Phe levels (120–360 $\mu\text{mol/L}$) that are dependent upon the infant's Phe tolerance (Blau et al., 2008, 2010). Consequently, the addition of Phe-free medical formula is vital in order to provide adequate protein and caloric intake while maintaining Phe restriction for normal cognitive and behavioral development. Phenylalanine is an essential amino acid; therefore in addition to Phe-free medical formula, infants with PKU need natural protein, such as breast milk or standard commercial infant formula, and later in infancy, cow's milk.

Breastfeeding infants with PKU are challenging. First, there is no universal approach to breastfeeding infants with PKU and

supplementing with Phe-free medical formula. As a result, mothers offer breast milk and Phe-free medical formula by one of three methods based upon recommendations from their metabolic clinic. Mothers may 1) provide a fixed amount of Phe-free medical formula followed by breastfeeding until satiety, 2) provide a fixed amount of expressed breast milk or a fixed time limitation of breastfeeding followed by Phe-free medical formula until satiety, and 3) alternate between breastfeeding or expressed breast milk feeding and Phe-free medical formula feedings. For some mothers, breastfeeding occurs only twice a day while others breastfed more frequently such as every other feeding. This difference is due to several factors. Phe concentration significantly decreases over time in breast milk ($2806 \pm 134 \mu\text{mol/L}$ at 2 weeks to $2173 \pm 87 \mu\text{mol/L}$ at 8 weeks) (Janas & Picciano, 1986). Also even though PKU is a classic Mendelian disease, there is variable presentation and severity in the clinical severity of PKU related to differing mutations in the gene encoding PAH enzyme resulting in differing degrees of residual PAH activity level (Guldberg et al., 1994). Some infants (classical form) have almost a complete loss of enzyme activity while others have only a partial reduction in the activity of the PAH enzyme. Consequently an infant with a classical form of PKU will be able to breastfeed less often than an infant with a milder form of PKU.

Despite these challenges, mothers have attempted to continue breastfeeding after the diagnosis to provide their infants with the benefits of breast milk and breastfeeding. Nonetheless, studies have revealed that few mothers with a child with PKU persist in breastfeeding. For example an Italian study found that breastfeeding rates were significantly lower in infants with PKU than in the general Italian population (Agostoni et al., 2000). Similarly, a Brazilian study reported that slightly more than half of the mothers were breastfeeding their infants with PKU at 6 months and by 12 months less than a third continued to breastfeed (Kanufre et al., 2007). In a more recent study involving mothers from the United States and Canada, the prevalence of breastfeeding infants with PKU before and after diagnosis revealed that significantly few mothers continued to breastfeed after diagnosis (Banta-Wright et al., 2014). Overall, the incidence, prevalence, and duration of breastfeeding when infants have PKU continue to be less than when full-term infants do not have PKU (Agostoni et al., 2000; Banta-Wright et al., 2014; Kanufre et al., 2007; Motzfeldt et al., 1999; Segev et al., 2004). In addition, qualitative analysis of mothers' experiences breastfeeding infants with PKU revealed that breastfeeding an infant with PKU is more work than breastfeeding a healthy term infant who does not have PKU (Banta-Wright, Houck, Kodadek, Steiner, & Knafl, in review). However, there is no information about challenges to breastfeeding an infant with PKU. In order to better understand the issues when breastfeeding an infant with PKU, we asked mothers living in the United States and Canada to identify and describe the challenges to the continuation of breastfeeding after an infant is diagnosed with PKU.

Methods

A quantitative descriptive design was selected for this study. Data were from an international Internet survey conducted to explore mothers' current or retrospective experiences breastfeeding an infant with PKU. A convenience sample of 103 mothers with one child who has PKU participated in the study during a 6-month period from November 2010 to May 2011. Eligibility criteria included having a child with PKU, a maternal age >21 years, the ability to read and write English, and residing in the United States or Canada. The study was approved by the institutional review board of Oregon Health & Science University.

After receiving approval from the institutional review board, the study was announced on the PKU Listserv; the purpose of this Listserv is to support individuals with PKU and their families through sharing their experiences in managing PKU. Subsequently, fourteen other regional and national PKU support groups used the original or modified university approved announcement of the study to communicate its availability to their members. Postings included a brief description of the purpose of the study and the inclusion criteria with the instruction that interested mothers should reply directly to the researcher by email. After being contacted, the research information sheet and instructions for the survey were emailed to mothers. The university's institutional review board waived the requirement for a formal written consent as mothers were informed that survey completion was considered evidence of their agreement to participate. The mean duration for mothers to complete the survey was 19 minutes ($SD = 10$), and ranged from 7 to 69 minutes. Mothers who completed the survey were sent a \$10 (U.S.) electronic gift certificate to a medical foods company which provides low protein items popular with families who have individuals with PKU. Of the 149 women who received the information sheet and instructions for the Internet survey, 119 completed the survey for a return rate of approximately 80%.

The survey contained sections about the demographic characteristics of mothers and their infants and infant feeding history. Both forced-choice and open-ended questions were included. The survey was pilot-tested by six mothers and modified based upon feedback to improve the approach and wording of the questions and forced-choice responses.

After the Internet survey was closed, data were downloaded into an SPSS file, and then compared to the hardcopy of each participant's data with discrepancies resolved accordingly. The survey asked mothers "What challenges did you have while breastfeeding your infant with PKU?" Breastfeeding challenges were defined as any problems or difficulties that mothers self-reported. To develop categories for this question, three more specific questions were asked of the responses: 1) What were the general breastfeeding challenges for mothers breastfeeding an infant with PKU? 2) What were the breastfeeding and PKU challenges for mothers breastfeeding an infant with PKU? and 3) Did some mothers not have challenges breastfeeding an infant with PKU? From the open ended responses in the

Internet survey, a frequency count approach was used to begin the analysis. Intercoder reliability yielded a 91% agreement; disagreements were resolved through conferencing. Statistical analysis was performed with the SPSS Windows (version 19.0). The focus of this report is on the 75 women with one child with PKU and the challenges that they faced while breastfeeding that child.

Sample

Respondents in this analysis also were participants in the larger study examining the prevalence and duration of breastfeeding infants with PKU (Banta-Wright et al., 2014). Demographic characteristics of the mothers and infants in this report are presented in Tables 1 and 2 respectively. In this group of mothers, breastfeeding duration ranged from less than 1 month to 24 months (Table 3). In addition, this sample was consistent with the known ethnic distribution of PKU in the United States and Canada, with a higher incidence in individuals of Northern European ancestry than black, Hispanic, and Asian individuals (Borrajo, 2007; Kietduriyakul et al., 1989; Williams et al., 2008).

Results

Common Categories

Qualitative analysis yielded three categories that identified and described the challenges for mothers' breastfeeding an infant with PKU: common breastfeeding issues, breastfeeding and PKU, and no challenges (Figure 1). The categories were common to mothers who participated in the survey and did not vary by maternal age, socioeconomic status, ethnicity, or region of country, nor by prior breastfeeding experience and infant gestational age and birth weight.

Common Breastfeeding Issues

Within the category of common breastfeeding issues ($n = 56$, 75%), four subcategories were identified: 1) breast pumping, 2) breast milk supply issues, 3) nipple confusion, and 4) common breastfeeding problems (Figure 2). All four challenges have been reported in the breastfeeding literature. Yet, for these mothers, the issues were 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 in breastfeeding their infants with PKU. One mother wrote, "spent hours pumping just to be able to breastfeed twice a day." Another mother reported, "I had to be diligent about pumping" and another asserted "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

Table 1 Demographic characteristics of mothers.

Variable	Maternal characteristics	
	$n = 75$	n (%)
Maternal age, years ^a	21–29	15 (20)
	30–39	45 (60)
	40–49	10 (13)
	50–65	5 (7)
Ethnicity	Asian/Pacific Islander	1 (1)
	Caucasian	71 (95)
	Hispanic/Latino	2 (3)
	First Nation/Inuit	1 (1)
Marital status	Never married or partnered	3 (4)
	Married/Living with partner	70 (93)
	Separated/Divorced	2 (3)
Education ^b	High school	6 (8)
	Associates degree	11 (15)
	Bachelors degree	31 (41)
	Advanced degrees (masters, PhD, professional)	27 (36)
Employment ^c	0	24 (32)
	<20	6 (8)
	20–38	16 (21)
	40	25 (34)
	>40	4 (5)
Gross annual household income ^{d,e}	<\$25,000	1 (1)
	25,001–50,000	8 (11)
	50,001–75,000	16 (22)
	75,001–100,000	23 (32)
	100,001–150,000	12 (17)
	>150,000	12 (17)
Community Population Size	A town with a population <10,000	10 (13)
	A city with a population between 10,000–50,000	29 (39)
	A city with a population between 50,001–100,000	12 (16)
	A metropolitan area with a population between 100,001–500,000	11 (15)
	A metropolitan area with a population between 500,001 to 1 million	6 (8)
	A metropolitan area with a population > 1 million	7 (9)

^a Maternal age in years.

^b Highest level of education completed.

^c Hours per week.

^d US dollars.

^e Mothers, $n = 72$.

refers to the ability of the mammary glands to produce adequate amounts of breast milk due to galactopoiesis. Maintaining a breast milk supply can be difficult when an infant is unable to exclusively breastfeed. Mothers' comments included the following challenges: "keeping up milk production with frequent changes in how often she was breastfeeding," and "trouble keeping my milk supply up." One mother shared, "hard time maintaining a consistent milk supply and eventually my milk supply ran out."

Table 2 Demographic characteristics of infants with PKU ($n = 75$).

	n (%)
Gender (female)	40 (53%)
Term gestation	78 (88%)
Uncomplicated pregnancies	69 (92%)
Infants born between 2000–2011	66 (88%)
	$\bar{x} \pm SD$ (range)
Birth weight ^a (kg)	3.45 \pm 0.5 (2–5)
Phenylalanine level at diagnosis ($\mu\text{mol/L}$)	871 \pm 594 (240–2520)
Age in days at diagnosis	4 \pm 4 (1–28)

^a Infants, $n = 74$.

Another frequently reported challenge was the issue of nipple confusion ($n = 21$, 28%) which occurs 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. 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 a standard bottle and I used a 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

Table 3 Duration of breastfeeding.

Duration in months	Primipara ^a	Multipara	Total
	($n = 27$)	($n = 47$)	($n = 74$)
	n (%)	n (%)	n (%)
< 1 month	1 (4%)	4 (8.5%)	5 (7%)
1–3 months	8 (30%)	5 (10.5%)	13 (17%)
4–6 months	5 (18%)	4 (8.5%)	9 (12%)
7–9 months	5 (18%)	9 (19%)	14 (19%)
10–12 months	3 (11%)	16 (34)	19 (25%)
13–18 months	3 (11%)	6 (13)	9 (12%)
19–24 months	2 (8%)	3 (6.5)	5 (7%)

^a Total number of primipara mothers = 28.

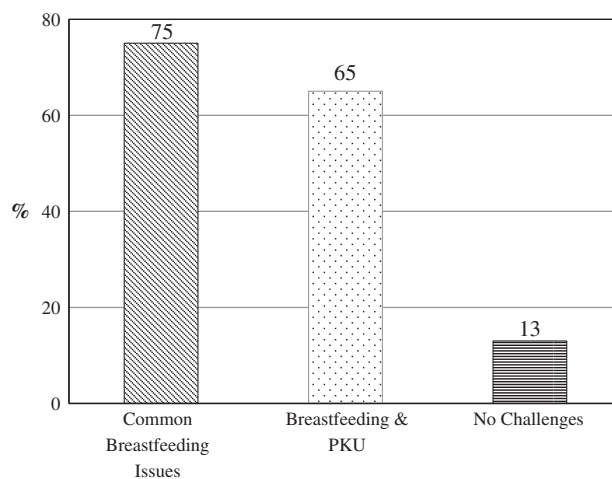


Figure 1 Challenges identified by mothers while breastfeeding infants with PKU.

mother wrote, “regular challenges of breastfeeding – plugged ducts and thrush.” Another mother described “My baby did not have a good suction [latch], which had nothing to do with PKU.”

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 (Figure 3). Mothers identified management ($n = 43$, 57%) as a central issue for Breastfeeding and PKU. Management included the work of breastfeeding, breast pumping, and feeding Phe-free medical formula on a daily basis, for which the amounts frequently changed due to Phe results. Several mothers expressed dissatisfaction with having more than one type of feeding as illustrated by a mother who wrote, “difficulty with 3 different types of feeding and mixing.” Another mother reported, “I used the pump for as long as I could to give him breast milk with Phe-free [medical] formula + formula

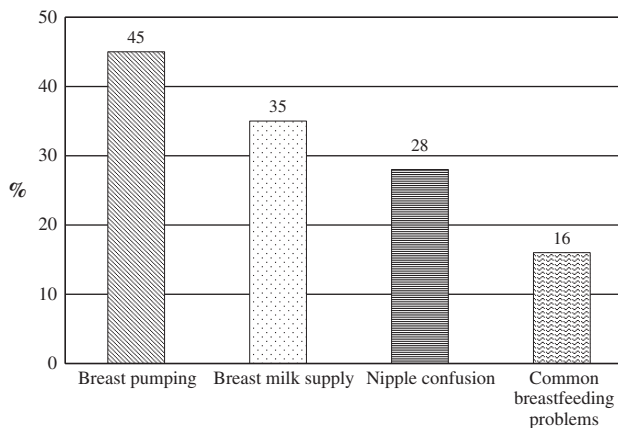


Figure 2 Subcategories of common breastfeeding issues identified by mothers breastfeeding infants with PKU.

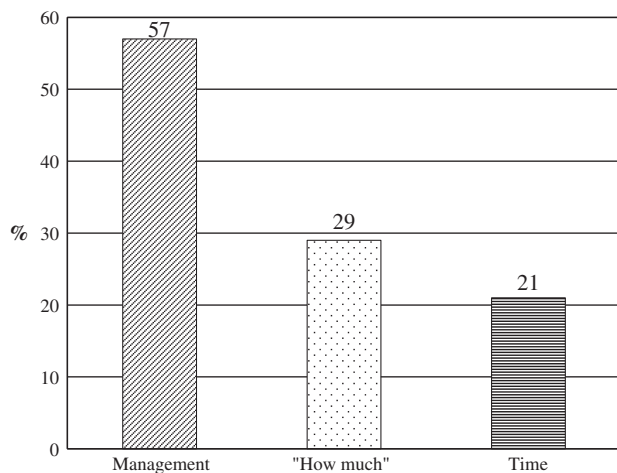


Figure 3 Subcategories of breastfeeding and PKU identified by mothers breastfeeding infants with PKU.

[regular standard commercial infant formula] to keep up with his feedings.” Several mothers wrote about the need to supplement with Phe-free medical formula and “had to feed low protein bottle before or after breastfeeding.” One mother expressed her frustration with the frequent feeding 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 and 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 ($n = 22$, 29%) 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 and thereby raising the concern of how much Phe that the infant obtained from the breast milk. Mothers were concerned about the amount of Phe that their infants received, as illustrated by mothers’ comments: “I was always worried about knowing the exact amount of Phe that he was getting” and “difficult to know *how much* breast milk baby was getting.” One mother revealed her frustration with trying to communicate with the metabolic health care team, “Measuring intake. I was only able to communicate length of breastfeeding sessions.” Another mother reported making a difficult decision: “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 the prioritization of tasks in order to maximize personal efficiency in breastfeeding as well as the amount of time involved. Mothers revealed several different aspects of time. Mothers repeatedly reported about the time to breast pump by one mother’s comment “spent hours pumping.” Several

mothers reported being challenged by “finding the time” and “having to speed the time” to breast pump, cleanse the breast pump equipment, mix the Phe-free medical formula, breastfeed, bottle feed the Phe-free medical formula with or without expressed breast milk or standard infant formula, and performing heel sticks.

No Challenges

A few mothers ($n = 10$, 13%) 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.”

Discussion

This study is unique in that no other studies have examined mothers’ reports of breastfeeding challenges for infants with PKU. In this study, challenges to breastfeeding infants with PKU yielded two categories: common breastfeeding issues and breastfeeding and PKU. These findings suggest that many of the breastfeeding challenges are interconnected.

In this study, over one-third of the mothers ($n = 28$) were first-time mothers. Among first-time mothers, breastfeeding challenges are common as breastfeeding is a learned behavior (Wagner et al., 2013; Williamson et al., 2012). Nonetheless in this study, mothers who have breastfed more than one child ($n = 47$) reported challenges breastfeeding their infant with PKU. Regardless of being a primipara or multipara mother, both groups reported maintaining a breast milk supply (29% vs 36%) and pumping challenges (36% vs 34%) to be major obstacles to breastfeeding as their infants necessarily breastfed less frequently than other full-term infants. Pumping issues and maintaining a breast milk supply were interconnected. When mothers wrote of waning breast milk supply, pumping issues were many times also noted, “I had the problem of low production, which required A LOT of pumping,” and “pumping often enough to maintain a decent supply.”

Breast pumping is a challenge for mothers of premature and critically ill infants in the neonatal intensive care unit (NICU) and much can be learned from that literature (Meier, 2001; Meier et al., 2007, 2008, 2010). However, there is an important 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 even 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 the need to maintain healthy 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.

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 full-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 medical formula to maintain desired therapeutic Phe levels. Over one-third of the mothers ($n = 26$) 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 et al., 2005; Hill, 1992; Hill & Aldag, 2007; Hill & Humenick, 1996).

For the majority of mothers exclusively breastfeeding a full-term non-PKU infant, lactogenesis occurs flawlessly with the transition from hormonal endocrine control (lactogenesis I and II) to autocrine control (lactogenesis III). Immediately after diagnosis, mothers breastfeeding infants with PKU need to provide Phe-free medical formula exclusively for 24 to 72 hours to rapidly reduce significantly elevated Phe levels. This interrupts the transition, thereby interfering with maintaining a milk supply. 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).

Another barrier for the breastfeeding mothers in this study was nipple confusion: 28% reported frustration with nipple confusion exhibited by their infants. Mothers described considerable frustration with helping 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 alternating breast and bottle feedings, their infants subsequently refused to latch on and breastfeed. At the same time, they also reported experiencing 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 et al., 1995; Newman, 1993). These challenges may be attributed to the differences in sucking pattern required for breastfeeding and bottle feeding (Mizuno & Ueda, 2003; Weber et al., 1986). Recent breastfeeding research investigated infants' sucking patterns associated with exclusive breastfeeding, exclusive bottle feeding, and mixed feeding of both breast and bottle in newborns and infants less than 6 months of age (Moral et al., 2010). The sucking patterns were found to be indeed unique and different. It may be that infants who struggle with bottle feeding after having been breastfed may not have yet developed the mixed sucking pattern.

Of the mothers in this study, 16% identified common breastfeeding problems that are typical for new mothers and infants, such as poor latch, nipple pain, thrush, and sore, cracked, or bleeding nipples as challenges to continuing to breastfeed. Most mothers realized these problems were not

PKU-related and, in fact, they reported these concerns less frequently than a study in which more than a third of mothers identified one or more common breastfeeding problems (Scott et al., 2006). Although these problems are viewed by many as "common," they are potential challenges to continued breastfeeding and contribute to subsequently stopping breastfeeding (Walker, 2008). The prompt recognition and resolution of these challenges are essential to sustaining breastfeeding.

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 United States 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 enrollment of a unique group of mothers who were waiting to be asked about their experiences breastfeeding in the context of PKU. The success of this method has positive implications for future nursing research in families with children who have rare inborn errors of metabolism disorders.

Limitations

This study was limited by its design. The survey data were collected between 7 months to 27 years after the birth of the child with PKU. However, only 9 mothers had children born prior to 2000. This length of delay may have affected their ability to recall their experiences. Nonetheless, a review of the literature revealed no information regarding challenges for mothers breastfeeding infants with PKU. This study provided the first exploration of challenges to breastfeeding in the context of PKU from mothers' perspectives.

Even though this convenience sample was drawn from the Internet, the sample was representative of highly educated, married/partnered, Caucasian mothers. Yet, they consistently identified similar challenges while breastfeeding their infants who have PKU even though the clinical management of infants with PKU and the care provided to these breastfeeding mothers were not the same in all institutions. Consequently, the challenges, although not generalizable to all mothers breastfeeding infants with PKU, are anticipated to describe commonalities and highlighted differences in mothers' experiences. Together with the interpretations, it is hoped that the findings have revealed insight to the challenges that mothers experience breastfeeding infants with PKU that will enhance their care.

Clinical Implications

These results reveal that mothers of breastfeeding infants with PKU are not receiving the support that they need to continue to breastfeed successfully in their unique situation. Mothers identified needing help with their breast milk supply and breast pumping. In order to maintain a milk supply when

nonexclusive breastfeeding, mothers need to have an appropriate breast pump for long term pumping, to know how frequent to breast pump, and to integrate breast pumping into their busy schedules. Nurses should prepare mothers for nonexclusive breastfeeding their infants with PKU and maintenance of lactation that might be breast pump dependent. Mothers need to be aware that the first 2 weeks after diagnosis will be an important time to establish a good breast milk supply. During this time, mothers must fully empty their breasts to mimic exclusive breastfeeding just like mothers who are pumping for preterm infants in the NICU. To accomplish this, mothers need to have an appropriate hospital-grade electric breast pump with a double collection that allows simultaneous pumping of both breasts. Not only does the hospital-grade electric breast pump mimic sucking rates and rhythms of healthy newborns during initiation and maintenance of lactation, but the double collection kit decreases pumping time. Some small electric and battery-operated breast pumps work well for mothers that need intermittent breast pumping, but they are not adequate for long term breast pumping needs to maintain lactation. Until mother's milk supply is established, mothers should be encouraged to breastfeed and breast pump eight to ten times in 24 hours. These sessions do not need to be equally spaced. In addition, mothers should avoid going longer than 5 to 6 hours without pumping especially during the day. Many of these sessions can occur after bottle feeding Phe-free medical formula. Mothers should be encouraged to empty the breasts as thoroughly as possible at each pumping session by pumping for 2 to 5 minutes after the last drops of milk. The addition of a pumping diary to monitor the milk supply may be helpful to identify when a decrease in milk supply has occurred so that additional interventions may be implemented as needed to maintain an adequate milk supply for the infant.

Another important aspect is nurse's collaboration with the mother on the ever-changing treatment plan based upon serial Phe levels. During these times, mothers may find breastfeeding an infant with PKU difficult and disheartening. Nurses can provide encouragement, support, and continued feedback that breast milk provides many benefits to infants that standard commercial infant formulas cannot provide. Lastly, mothers need nurses to show a sincere desire to help them succeed in breastfeeding their infant with PKU.

Future Research

This study revealed that mothers experience many challenges breastfeeding an infant with PKU. The majority of mothers persist with the challenges of simultaneously managing breastfeeding and PKU to successfully adapt breastfeeding and maintain desired Phe levels. Yet, some mothers struggle and stop breastfeeding. Further research is needed to explore breastfeeding strategies for mothers who must alternate between breast- and bottle feeding to maintain their infant's desired Phe levels while sustaining their breast milk supply.

This would include exploring the breastfeeding experience of mothers who reported no challenges breastfeeding their infant with PKU as well as mothers who stopped breastfeeding within 6 months of their infant's diagnosis to provide insight. Information from these two different sets of mothers might provide new strategies to help more mothers to continue to breastfeed their infant with PKU longer.

Summary

This study provided the first description of mothers' challenges when breastfeeding an infant with PKU: common breastfeeding issues, breastfeeding and PKU, and no challenge. These challenges provide further insight into mothers' experiences in breastfeeding an infant with PKU. While nurses can provide support, encouragement, and anticipatory guidance for mothers' breastfeeding an infant with PKU, further research is needed to explore how to best support mothers breastfeeding an infant with PKU.

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