Authentic Caring Occasions for Patients in Hairy Cell Leukemia Clinical Trials

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Background: Rare diseases present challenges for patients, healthcare providers, and researchers. Rare disease communities exist for collaboration, dissemination of information, and to promote support for all community members. Patients with a rare disease desire to be supported through a microcommunity. Hairy cell leukemia (HCL) is a rare adult B-cell lymphocytic cancer that currently has no cure.

Objectives: Patients with relapsed or refractory HCL may need to consider participation in a clinical trial. The research nurse can initiate a planned caring occasion based on Watson’s Theory of Human Caring. The purpose of the planned caring occasion for patients with HCL in clinical trials is to establish authentic intentional caring encounters between the research nurse and patients and meaningful caring encounters between patients.

Observations: Relapsed or refractory patients enrolled in an HCL clinical trial identify the trial as a microcommunity and the research nurse as an advocate and liaison. Patients seek support, empowerment, and the opportunity to connect with other patients with HCL. The planned caring occasion has the potential to provide a healing environment and facilitate shared experiences of living with HCL. The potential outcome for patients is strengthened holistic wellness.

A rare disease diagnosis is a significant life event for patients and has the potential to alter patients’ sense of security and elicits feelings of vulnerability and isolation (Haylock, 2010). Rare disease communities have formed around the world to support patients, healthcare providers, and researchers (Colledge & Solly, 2012). Patients seek participation in a rare disease community for many reasons, including increased knowledge, quality of life, holistic wellness, connection, and collaboration with other patients, as well as expert healthcare providers. Researchers and healthcare providers seek participation in rare disease communities because of the limited number of researchers dedicated to working with rare diseases (Aymé, Kole, & Graft, 2008; Budych, Helms, & Schultz, 2012; Colledge & Solly, 2012; Grever & Lozanski, 2011; Holzman, 2009; Huyard, 2009; Patos, 2001; Schieppati, Henter, Daina, & Aperia, 2008; Walker, 2013). Key members of the rare disease community include the National Organization of Rare Disorders (NORD), established in the United States, and the European Union Committee of Experts on Rare Diseases. The primary mission of NORD includes promoting translational research and advocacy, and funding the search for cures for rare disease. NORD (2013) defines rare disease in the United States as affecting less than 200,000 individuals within the total population. The European Union Committee of Experts on Rare Diseases is committed to identifying rare disease centers of expertise, as well as establishing a collaborative network for information sharing (Aymé & Rodwell, 2014; Humphreys, 2012).

Patients with a rare disease face challenges in terms of treatment. Hairy cell leukemia (HCL) is a rare adult B-cell lymphocytic...
cancer. Patients may experience relapse after treatment or be refractory to current standard of care therapies. With no cure available for HCL, patients with relapsed or refractory HCL may be asked to consider participation in a clinical trial using investigational therapies (Grever & Lozanski, 2011). Patients with HCL participating in a clinical trial may identify the trial as a microcommunity within the larger community of HCL and as an opportunity to meet other HCL patients. The clinical trial research nurse, as liaison, advocate, and educator, can initiate a planned caring occasion to meet patients' expressed desire for community (Gibbs & Lowton, 2012; Green, 2011; Poston & Buescher, 2010). The planned caring occasion is based on the core concept of the transpersonal caring transaction of Watson's Theory of Human Caring (Sitzman & Watson, 2013; Watson, 2002). The method of communicating the planned caring occasion will be dependent on individual healthcare facilities' policies related to patient privacy rights and protection of research participants. The purpose of this article is to examine the potential impact of caring encounters within the HCL community, particularly patient-to-patient encounters, and encounters between the research nurse and patients. This article also examines the potential of the encounters and planned caring occasions to facilitate connections and sharing that positively affect holistic wellness.

Hairy Cell Leukemia

HCL, also known as leukemic reticuloendotheliosis, is a rare adult chronic B-cell lymphoproliferative leukemia (Bouroncle, Wiseman, & Doan, 1958; Grever & Lozanski, 2011). HCL affects 2% of adults diagnosed with leukemia. About 600–800 new cases are diagnosed each year (NORD, 2013). Confirmation of an HCL diagnosis is based on peripheral blood and/or bone marrow aspirate examination by flow cytometry and bone marrow biopsy examination by immunohistochemical stains. More recently, testing for BRAF V600E mutations has been used to confirm diagnosis (Langabeer et al., 2012). The determination of classic or variant HCL is crucial in selecting the appropriate treatment (Grever & Lozanski, 2011). Patients begin treatment with a diagnosis of HCL coupled with symptoms that may include significant cytopenias, fatigue, splenomegaly, and active infections because of low neutrophil count (Grever & Lozanski, 2011; Naik & Saven, 2012; Robak, 2006).

In the early 1970s, the initial treatment option for HCL was splenectomy. Alpha-interferon was identified as a treatment but had limited effectiveness (Habermann, 2006). In the late 1980s, cladribine and pentostatin, known as purine nucleoside analogs (PNAs), were discovered for treating HCL. PNAs remain first-line treatment for symptomatic HCL. PNAs allow 60% of patients to achieve remission. For individuals who relapse after therapy, a second remission may be possible for as many as 52%–75% of patients when treated with PNA therapy (Hairy Cell Leukemia Foundation, 2013). Other treatments for HCL include monoclonal antibodies and immunotoxins (Grever & Lozanski, 2011; Kreitman & Pastan, 2006; Naik & Saven, 2012; Robak, 2006; Sheldon, Wisener, & Roberts, 2005; Thomas, Ravandi, & Kantarjian, 2006). Despite the available treatment options and ongoing clinical trials, currently available therapies do not cure HCL.

The Hairy Cell Leukemia Rare Disease Community

Rare disease communities exist because patients seek to meet others with the same disease or diagnosis. Patients take advantage of the opportunities for creating or engaging in community (Colledge & Solly, 2012; Huyard, 2009; Nierse, Schipper, van Zadelhoff, van de Grijndt, & Abma, 2012; Patso, 2001; Zhao, Abrahamson, Anderson, Ha, & Widdows, 2013). The HCL rare disease community consists of healthcare providers, researchers, nurses, patients, family, and friends. The patients participate in HCL organizations, online communities, such as Macmillan Cancer Support (http://community.macmillan.org.uk) or Hairy Cell Leukemia Foundation (www.hairycellleukemia.org), and blogs. Patients with HCL desire to gain knowledge from other patients and compare the expertise of healthcare providers providing treatment (Aymé et al., 2008; Budyech et al., 2012; Kvåle & Bondevik, 2008; Zhao et al., 2013). Patients with a rare disease are often motivated to become experts in their disease and treatment, as well as self-advocates. One reason for the role transformation to expert patient and self-advocate is, in part, because of limited local access to healthcare providers with expert knowledge specific to HCL and best treatment options (Budyech et al., 2012; Huyard, 2009; Kvåle & Bondevik, 2008; Patso, 2001; Schieppati et al., 2008; Walker, 2013). Patients as self-advocates, particularly those with rare diseases, have initiated support groups, social media sites, and peer online communities (Ashing-Giwa et al., 2012; Chambers et al., 2012; Colledge & Solly, 2012; Patso, 2001; Stang & Mittelmark, 2010; Walker, 2013). In an effort to establish transgeographic connections within the HCL community, the Hairy Cell Leukemia Foundation was established for patients, physicians, and researchers. The online HCL community removes the barriers in reaching other patients with HCL within the United States and across the world (Colledge & Solly, 2012; Patso, 2001; Schultz, 2002; Walker, 2013; Zhao et al., 2013).

Patients who participate in the HCL community are supported with the physical dimension of health while experiencing the benefit of wellness support in the psychosocial dimension. Qualitative studies with a focus on supportive care and patient-centered care were conducted with patients with cancer and revealed the importance of the shared experience of having cancer, surviving cancer, and maintaining quality of life (Ashing-Giwa et al., 2012; Bonito, Horowitz, McCorkle, & Chappar, 2013; Carter, Bryant-Lukosius, DiCenso, Blythe, & Neville, 2011; Kvåle & Bondevik, 2008). In addition, surveys about health-related quality of life (HRQOL) revealed the need for psychosocial quality of life among patients with cancer (Stang & Mittelmark, 2010; Zabalegui, Cabrera, Navarro, & Cebría, 2011). Efficace et al. (2012) identified the top 10 HRQOL aspects of value to patients with chronic myeloid leukemia and to healthcare providers treating chronic myeloid leukemia. The top 10 list included worries and uncertainties about health condition in the future, satisfaction with information received, and importance of social support in coping with the disease.

The HCL community at large has the potential to positively affect patients and provide benefits of social support, quality of life, disease knowledge, and advocacy. However, some patients...
with HCL may need more personal and deeper connections as they search for holistic wellness, and they may have the desire to share their lived experience with HCL (Colledge & Solly, 2012; Huyard, 2009; Nierse et al., 2012; Patsos, 2001; Zhao et al., 2013).

Caring Encounters and Occasions Within the Community

Watson’s Theory of Human Caring is a theoretical foundation for caring encounters and occasions. Watson’s theory highlights humanity and each being’s need for respect, dignity, affirmation, compassion, and connectedness. Watson’s theory identifies nursing practice as a means to provide caring, particularly transpersonal caring. Transpersonal caring is intentional authentic presence and purposeful connectedness to promote human dignity and a healing environment. The act of transpersonal caring connects the nurse and patient, being to being. Two core concepts of Watson’s theory include the transpersonal caring relationship and the caring occasion or moment. In addition, Watson’s theory contains the 10 Caritas Processes™. The caritas are the actions that occur during a transpersonal caring relationship and occasion. Of the 10 caritas, two are significant to patients diagnosed with HCL: (a) being an authentic caring presence to create a healing environment and (b) allowing positive and negative feelings to be expressed while authentically engaged with another’s story (Sitzman & Watson, 2013; Watson, 2002). Transpersonal caring encounters can occur between the nurse and patient, between patients, or during a planned caring occasion.

Nurse and patient transpersonal caring encounters or occasions are an essential support for patients with a rare disease like HCL. The focus of nurse and patient caring encounters is an intentional, individualized promotion of the patients’ holistic wellness. Patients with relapsed or refractory disease may have an even greater need for authentic and intentional caring encounters than those with HCL that is in remission. Patients with relapsed or refractory disease may be asked to consider participation in a clinical trial using an investigational therapy for HCL because standard care therapies were not effective (Grever & Lozanski, 2011; Robak, 2006). Patients must process that the disease has returned or is resistant to treatment and make a decision about screening for a clinical trial using an investigational therapy. Patients with relapsed or refractory disease may experience uncertainty and decisional conflict (Biedrzycki, 2011; Miller et al., 2013).

In the event that a patient must consider participation in a clinical trial, the research nurse or clinical trial nurse becomes an essential contact. The literature (Gibbs & Lowton, 2012; Green, 2011) identifies the research nurse as a source of information and communication between the patient and the treating team or principal investigator (PI). The research nurse is often a liaison, advocate, and patient educator. The research nurse can establish a rapport and connection with the patient (Brinkman-Denney, 2013; Gibbs & Lowton, 2012; Green, 2011; Poston & Buescher, 2010). Also, patients may see the research nurse as a pathway to connect with other patients participating in the clinical trial.

Patients may have many questions related to clinical trial data and the other participants in the clinical trial. Although the PI and the research nurse may be able to answer or address many questions and concerns, clinical trial data cannot be disclosed prior to public presentation or publication. In a quest for more information and to establish connections to those with an HCL diagnosis, patients may ask to meet individuals participating in the clinical trial. Patients with HCL recognize the clinical trial as a microcommunity within the larger HCL community. The research nurse has an opportunity to promote overall holistic wellness, particularly the psychosocial elements of social, emotional, and intellectual dimensions of health, through a planned caring occasion.

A Planned Caring Occasion and the Clinical Trial Community

As the point of contact for the patients in a clinical trial community, the research nurse can integrate Watson’s theory of transpersonal caring through a planned caring occasion (Gallagher-Lepak & Kubsch, 2009; Mullaney, 2000; Sitzman & Watson, 2013; Watson, 2002). A planned caring occasion has the potential to enable authentic caring transactions to form between patients and to meet patients’ expressed need for community and allow patients to experience the benefits of being a member of a rare disease community (Gallagher-Lepak & Kubsch, 2009; Haylock, 2010; Iranmanesh, Axelsson, Sävenstedt, & Häggeström, 2009; Mullaney, 2000). In addition, patients may experience connection through shared experiences of living with HCL, an increase in knowledge through group question-and-answer sessions, and reflection on participation in an HCL clinical trial, as well as hope, empowerment, and enhanced coping. Empowered patients may experience a decreased sense of loss of control, increased confidence, and elevated holistic wellness (Aymé et al., 2008; Stang & Mittelmark, 2010).

The planned caring occasion is the setting to facilitate patient-to-patient caring encounters. The caring encounters enable patient sharing of the mutual understanding of losses, such as living from transfusion to transfusion, maintaining treatment calendars that disrupt work and family life, accumulating medical bills and loss of income, and experiencing limited socialization because of risk for infection. Patients may understand one another’s concerns with traveling out of state to HCL expert hematologists and considering innovative treatment options and clinical trials (Miller et al., 2013). Only a patient with HCL can identify with another patient with HCL concerning the physical, emotional, and mental stress (Aymé et al., 2008; Becker, 2007; Miller et al., 2013).

The potential outcomes of the planned caring occasion may include patient experiences of positive, energy-filled, meaningful interactions; a sense of being lifted up through verbal and nonverbal cues; increased courage; decreased sense of isolation; promotion of inner holistic wellness; and affirmation of patients’ personal stories and the stories of other patients (Gallagher-Lepak & Kubsch, 2009; Iranmanesh et al., 2009; Mullaney, 2000; Sitzman & Watson, 2013). Achievement of the outcomes will be dependent on the participants and the format of the caring transaction.

The role of the research nurse at the planned caring occasion may take the form of facilitator or recorder. Planned caring occasions may occur as face-to-face meetings or by synchronous communication, such as web, video, or audio conferences. At the beginning of the caring occasion, patients are asked to agree.
to value one another’s shared experience, respond without judgment, and be respectfully present. During the planned caring occasion, the research nurse can establish a sacred space for patients’ stories and vulnerabilities by modeling intentional presence, authentic compassion, and affirmation of the beings within the patients’ bodies. The modeling of authentic presence by the research nurse may be unconsciously perceived by other patients and contribute to deeper patient-to-patient caring transactions during future caring occasions (Clark, 2003; Watson, 2002). The presence of the research nurse at the planned caring occasion can minimize sharing of misinformation, diffuse negative energy, and offer compass in emotional moments.

The method of communicating the planned caring occasion to potential participants will be dependent on the healthcare facility and policies related to patient privacy rights. The research nurse should consult the healthcare facility’s privacy officer to ensure compliance with research participants’ rights and privacy rules. As a point of contact for patients, the research nurse must maintain a delicate balance between Good Clinical Practice (CTI Program, 2015), adhering to the Health Insurance Portability and Accountability Act (2015), and using common sense about the dissemination of information related to patients participating in the clinical trial.

Communication to patients must contain the intended purpose and potential outcome, as well as a statement that patient participation is voluntary. In addition, the research nurse can confirm in advance that questions posed by patients during the caring occasion will be answered by an expert HCL clinician and communicated to participants if requested. At the conclusion of the planned caring occasion, the research nurse can assist patient attendees with identifying the form of future communication between those present at the caring occasion and reiterating future participation and communication as optional and voluntary.

The concept and potential impact of the planned caring occasion was introduced to patients and HCL experts at the second annual Hairy Cell Leukemia Foundation Patient Seminar held in Houston, Texas, in 2014. Seeking patient input and interest in planned caring occasions is important to increase the effectiveness when these are implemented.

**Implications for Nursing Practice**

- **Support the expression of positive and negative feelings related to clinical trial therapy and perceived physical response to therapy for patients with hairy cell leukemia.**
- **Offer intentional, authentic caring with active listening to support patients’ emotional and psychosocial needs during nurse-to-patient encounters and when present during patient-to-patient encounters.**
- **Advocate for patients’ expressed need to interact with other patients with rare disease through the initiation of a planned caring occasion.**

**Conclusion**

The heart of the rare disease community comprises patients who desire connectedness through the shared lived experience with a rare disease diagnosis, as well as access to knowledge from all members comprising the rare disease community. Patients diagnosed with HCL have the same needs as other patients diagnosed with a rare disease. A review of the literature supports that patients with rare disease diagnoses find meaning in coming together as a community. Benefits from being in a community include decreased isolation, enhanced psychosocial and emotional support, and a positive impact on overall holistic wellness. Watson’s Theory of Human Caring and the concept of the transpersonal caring relationship is the theoretical foundation for the intervention of a planned caring occasion. The research nurse can promote holistic wellness by planning a caring occasion for the patients. The planned caring occasion can serve as a healing environment facilitating patient-to-patient caring encounters and patient sharing of lived experiences with HCL. Potential outcomes are dependent on the patients’ participation in the caring encounters and their perception of authentic presence as they interact with one another and the research nurse.

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**References**