13 Pediatric Hematopoietic Stem Cell Transplantation: Psychosocial Assessment and Care

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Introduction to Hematopoietic Stem Cell Transplant

There are an ever growing number of successful hematopoietic stem cell transplants (HSCT) being conducted on an increasingly wide range of oncologic, hematologic, and immunologic conditions (Cairo and Heslop 2008). Despite the progress being made in curing life-threatening diseases, HSCT is still considered a high-risk procedure because of the treatment-related morbidity and mortality. There are two types of transplants, which is based on whether the patient receives her own hematopoietic stem or progenitor cells (autologous) or cells from a donor (allogeneic). Allogeneic cells are donated by another person (e.g., sibling, parent, or unrelated donor from the National Bone Marrow Registry) and attained from their bone marrow, peripheral blood, or umbilical cord.

Clinical indications for HSCT include malignancies, particularly in the case of cancer relapses or recurrences [e.g., relapsed acute lymphoblastic leukemia (ALL) or acute myeloid leukemia (AML)] as well as non-malignancies (e.g., immune deficiency disorders, bone marrow failure, severe aplastic anemia). Oncology patients who are referred for an HSCT likely have already experienced a lengthy illness and treatment course, which consisted of chemotherapy and other treatment-related side effects and hospitalizations, as is often the case with relapsed ALL and AML patients. This is in stark contrast to patients with non-malignancies referred to HSCT, such as aplastic anemia, who may not have had previous treatment experiences or hospitalizations or may present as asymptomatic prior to HSCT. Therefore, in these latter cases, the patient and family may be less well prepared or less likely to know what to expect.

Upon transplant hospitalization, the treatment includes several sequential phases beginning with the conditioning phase, followed by infusion and the engraftment process, which combined make up the acute phases of transplant hospitalization (approximately 4–6 weeks in total). The conditioning phase typically lasts 7–10 days and consists of myeloablative chemotherapy and, in some cases, is coupled with total body irradiation. The purpose of this phase is to destroy any remaining malignant cells, suppress the immune system to prevent rejection, and create marrow space. Nausea, vomiting, pain, and fatigue are common side effects that begin during this phase and continue throughout the admission. Patients also become blood and platelet transfusion dependent. Maintaining good

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nutrition is a common struggle and may require total parenteral nutrition. More recently, reduced intensity conditioning regimen transplants or "mini transplants" with lower doses of chemotherapy and radiation have been provided to more vulnerable patients who are not healthy enough to receive a myeloablative regimen prior to HSCT, in an effort to reduce toxicities that lead to morbidity and mortality.

The next phase is the bone marrow or stem cell infusion, which is infused through a central venous line over a period of 20–60 minutes, the process of which is similar to a blood transfusion. A primary potential side effect that can occur during the infusion is an anaphylactic reaction, thus requiring patients to be closely monitored during the infusion. A 2–4-week "waiting period" follows the infusion, during which the patient has no effective immune system and is very susceptible to infection. Additional symptoms during this phase include mucositis and fevers, with the primary treatment focus being symptom management. As the patient begins to demonstrate signs of engraftment (which is when the new immune system begins to grow and create new blood cells), mucositis begins to heal and infections resolve; however, for those that receive an allogeneic transplant, acute graft-versus-host disease (GVHD) can occur. GVHD occurs when the donor cells see the host cells as foreign and attack them. Skin, gut, liver, and lungs can be impacted and graded from mild to severe, with severe GVHD causing life-threatening symptoms. Chronic GVHD is defined as symptoms that last for or begin at least 100 days after transplant. GVHD is typically treated with steroids, which have a number of physical and emotional side effects. GVHD symptoms have a negative impact on the patient's health-related quality of life (HRQL), particularly in the physical and social domains (Clarke et al. 2008; Tanzi 2011). Patients with moderate to severe GVHD often require hospitalization for treatment to control their symptoms and manage their pain. Patients with GVHD often are socially isolated due to pain, debilitating diarrhea, risk for infection, and physical appearance changes related to steroid side effects (e.g., significant weight gain and swelling in the face, stomach, and limbs) and skin GVHD (severe itchiness, skin flaking, and severe body rashes).

Following discharge from transplant hospitalization, patients experience a lengthy recovery with an intensive outpatient regimen that can last from months to years. During this time, patients often receive intravenous (IV) hydration; take many medications which need to be frequently monitored and adjusted, including immunosuppressive medications; have dietary and activity restrictions and recommendations; and are socially isolated due to continued infection risk with a young immune system. Motivating good nutrition and adequate fluid intake as well as treatment adherence become primary focuses of intervention during recovery. Social isolation may last for 6 or more months post-HSCT depending upon the patient's transplant-related complications and immunosuppressed state. This isolation can be a primary stressor as patients struggle to cope with not being able to attend school or socialize with their friends in public areas despite feeling well.

Pre-HSCT Psychological Evaluation

Once a patient is recommended to receive a transplant by the HSCT team, HSCT candidates receive a comprehensive medical evaluation or "workup." Potential HSCT candidates receive a physical examination, blood tests, a bone marrow biopsy, as well as a number of tests to assess organ functioning to determine the patient's overall health status and appropriateness to undergo an HSCT. In addition to the medical workup, most pediatric HSCT centers require a psychosocial assessment prior to a patient receiving an HSCT, and, in fact, many health insurance companies will not approve coverage for an HSCT without one. A psychosocial provider (Sherman et al. 2004), with expertise in the psychosocial issues associated with pediatric HSCT, typically conducts pre-HSCT psychological evaluations. The purpose of these evaluations is not meant to make a determination as to whether a patient should have an HSCT or not but instead should

serve to assess the patient and family's psychosocial functioning, stress and coping, HSCT knowledge and preparedness, and supportive care needs in order to best prepare and support the patient and family prior to, during, and after an HSCT. Taking a sensitive, supportive approach when conducting a pre-HSCT psychological evaluation can also serve to establish a therapeutic relationship with the patient and family, thereby making it more likely for the psychosocial provider to be successful in addressing any problems or crises that may arise during the transplant and recovery. Recommendations based upon results of the evaluation should be shared in written and/or verbal format with the HSCT team prior to the HSCT admission in order for the multidisciplinary team to (1) provide more education when there is a lack of or misunderstanding of the HSCT, (2) address any potential psychosocial or economic concerns or barriers raised prior to the HSCT that may negatively impact the success of the HSCT, and/or (3) ensure that appropriate supportive care services are in place for the patient and family prior to or at HSCT admission.

Optimal timing of the pre-HSCT psychological evaluation occurs after the patient and family meet with the HSCT oncologist for education but prior to the patient or parents formally consenting and assenting, when appropriate, to HSCT. The purpose of the psychological evaluation should be discussed with the parents and patient. Whenever possible, it is important for the psychosocial provider to meet with the patient and parents separately, particularly when the HSCT candidate is an adolescent, so each has the opportunity to freely share personal thoughts, feelings, and concerns.

There are a number of salient domains to assess during the pre-HSCT psychological evaluation, which are discussed below and summarized in Table 13.1, along with examples of quantitative measures that may be used to supplement semi-structured interview questions. In a survey of pediatric HSCT centers registered in the Pediatric Blood and Marrow Transplantation Consortium, psychosocial screening was typically assessed via interview with infrequent use of quantitative measures (Sherman et al. 2004). Sherman et al. (2004) recommend taking a qualitative and quantitative assessment approach. Development of validated measures specific to pediatric HSCT is needed.

Knowledge of HSCT and Ability to Consent or Assent

A primary focus of the assessment should be to ascertain the level of understanding patients and parents have about the HSCT and recovery process. Any questions, concerns, or gaps in knowledge, including misperceptions, should be addressed with further education provided by the HSCT team so that the patient and parents can provide informed consent and, when appropriate, assent to the HSCT (see Table 13.2 for educational tools and resources). In addition, having a developmentally appropriate understanding of the HSCT process better prepares children, adolescents, and young adults for the HSCT, which may help them cope or reduce anticipatory anxiety leading up to the transplant. For the patient, the extent of HSCT knowledge is dependent upon age or developmental level, and therefore, questions assessing the level of comprehension should be tailored accordingly. For example, younger to school-aged children want to be informed about how a treatment will make them feel in the present or immediate future, how the doctors and nurses will help them to feel better when ill, and who will take care of or be with them in the hospital. Therefore, for younger children, it is expected that they may understand the very basics of HSCT, such that they will be in the hospital for a long time, may feel sick at times, but will get medicine to help them feel better. In contrast, adolescents are typically able to cognitively understand the complexity and intensity of the HSCT process at a level similar to their parents and thus should be able to provide a more detailed description of the HSCT process.

When inquiring about the HSCT procedure, the psychosocial provider should gain a sense of whether the patient and parents understand the different phases of the transplant, including a

Table 13.1 Summary of patient and family assessment domains and measurement examples for pre-HSCT psychological evaluation

Assessment domain	Salient assessment factors	Examples of interview questions	Examples of measures tailored to individual patient needs
HSCT knowledge (informed consent/ assent)	HSCT procedure Benefits vs risks Decision making/voluntariness Alternative treatment options Patient and parental HSCT responsibilities Length of hospitalization Recovery process	What is your understanding of what happens during transplant? What happens when you are first hospitalized (conditioning regimen); what happens next (HSCT infusion, engraftment)? How did you come to the decision of having a transplant? Who decided? What are some of the risks or potential side effects of transplant? Did the doctor tell you about any other treatment options besides transplant? What do you anticipate the experience will be for you during your hospitalization for your transplant? Tell me what you know about the recovery process after you are discharged from the hospital after transplant.	Open-ended interview questions
Psychosocial functioning	Current psychosocial distress (depression, anxiety, behavioral problems) HRQL Helpful vs unhelpful coping strategies Previous mental health history	Tell me about how you have been feeling emotionally. How have you been coping with your illness and treatment? Do you have any specific fears associated with medical procedures? What are you most concerned or worried about with HSCT? Have you ever seen a counselor or therapist for any reason? Have you ever been prescribed medications for your mental health?	*Internalizing and externalizing problems*: Achenbach System of Empirically Based Assessment (ASEBA; Achenbach and Rescorla 2001) Behavior Assessment System for Children – Second Edition (BASC-2; Reynolds and Kamphaus 2004) *HRQL*: Behavioral Affective and Somatic Experiences Scale (BASES; Phipps et al. 1994, 1999) Child Health Ratings Inventories (CHRIs Generic & HSCT module; Parsons et al. 2005) PedsQL™ (Varni et al. 2001) Functional Assessment of Cancer Therapy Quality of Life Measurement System (FACT-BMT; Cella et al. 1993) *Coping strategies*: Kidcope (children and adolescents; Spirito et al. 1988) Coping Strategies Inventory (Tobin et al. 1984) Ways of Coping (young adults; Folkman and Lazarus 1988)

Assessment domain	Salient assessment factors	Examples of interview questions	Examples of measures tailored to individual patient needs
Medical adherence	Past and current problems following the medical regimen Missed or errors in medication Pill-swallowing problems History of missed appointments	How difficult has it been to follow what the doctors and nurses tell you to do? Tell me how you do with swallowing pills. Do you ever have difficulty remembering when to take your medicine or which medicines to take?	BASES Compliance Scale (Phipps et al. 1994, 1999)
Social support and stressors	Quantity and quality of support Identification of primary support persons Types of support received Other stressors or barriers to HSCT (economic stressors)	Who supports you the most through treatment? Tell me how you would rate the level or quality of the support you receive. Tell me about other stressful situations in your life.	Multidimensional Scale of Perceived Social Support (Zimet et al. 1988) Life Event Scale (Adapted from the Social Readjustment Rating Scale; Holmes and Rabe 1967)
Family functioning and parental adjustment	Family structure Family expression of feelings with each other Family communication Family conflict/cohesion Past and current parental stress, coping, and psychological functioning Family mental health history Family role changes Parental employment issues	Who lives at home with you? How do you get along with your parent(s) and sibling(s)? How has your family life changed since being diagnosed? Who do you talk to in your family when you are stressed? For parent: Tell me how you are coping with your child's health and treatment. For young adult patient or parent: Is your employer or work environment supportive?	Family Environment Scale – 3rd Edition (Moos and Moos 1994) Impact on Family Scale (Stein and Riessman 1980) PAT2.0 (Pai et al. 2008) CHRIs – parent version (parent report of own health and child's health; Parsons et al. 2005) Coping Health Inventory for Parents (McCubbin et al. 1983) Sibling Relationship Questionnaire (Furman and Buhrmester 1985) Pediatric Parenting Stress Inventory (Devine et al. 2014)
Cognitive, academic, and vocational functioning	Past or current learning or cognitive problems School or vocational performance Educational or vocational services	Do you/your child or have you/your child ever received extra help in school? Do you/your child have an Individualized Education Plan or 504 Plan? What classes are you/your child really good at? What classes do you/your child have difficulty in?	Objective, standardized cognitive, and academic tests (see narrative section) Wechsler Scales of Intelligence (Wechsler 2004) or WASI-II (Wechsler 2011) Wide Range Achievement Test 4, Reading and Arithmetic Composites (academic screen; WRAT4; Wilkinson and Robertson 2006) COG ALTE07C1 abbreviated cognitive testing battery (Embry et al. 2012)

Table 13.2 Informational and supportive resources for pediatric HSCT

Support and resource programs	*Program description*	*Website address*
American Cancer Society	Provides information about pediatric and adult HSCT.	cancer.org
Blood & Marrow Transplant Clinical Trials Network	A multi-institutional clinical trials network for HSCT that offers clinical trial participation to HSCT patients in the United States.	https://web.emmes.com/study/bmt2/
Blood & Marrow Transplant Information Network	A support and informational resource for HSCT patients, survivors, and caregivers.	bmtinfonet.org
Imerman Angels	International program that provides patient to survivor and caregiver to caregiver connections for support.	imermanangels.org
Insights e-Community	Online social networking or discussion group to connect with other adolescents and young adults who are facing or have undergone an HSCT.	bethematch.org
National Bone Marrow Transplant Link	An information and resource program for patients who receive an HSCT and their families.	nbmtlink.org
National Cancer Institute	Provides information regarding pediatric HSCT using physician data queries (PDQ) or fact sheets about HSCT.	cancer.gov/cancertopics/pdq or cancer.gov/cancertopics/facts
National Marrow Donor Program; Be the Match	This program recruits potential bone marrow donors, maintains a registry of potential bone marrow donors, and facilitates the donor search and matching for a potential HSCT recipient. Be the Match is operated by the National Marrow Donor Program, which conducts research to improve HSCT outcomes and provides support and informational resources for patients and families.	bethematch.org
Pediatric Blood and Marrow Transplant Consortium	An international clinical trial group specifically focused on improving outcomes in pediatric HSCT through research and education	pbmtc.org
Educational resources	*Format*	*Available at*
Bone Marrow & Blood Stem Cell Transplants: A Guide for Patients (2012) by Susan K. Stewart	Book; Also available in Spanish.	bmtinfonet.org
Autologous Stem Cell Transplants: A Handbook for Patients (2012) by Susan K. Stewart	Book	bmtinfonet.org
Super Sam versus the Marrow Monsters	Animated DVD for children and their families. Also available in Spanish.	bethematch.org
Insights: Experiencing Transplant as a Young Adult	DVD for adolescents and young adults to learn about HSCT based upon other HSCT survivors' personal experiences.	bethematch.org
Me and My Marrow: A Kid's Guide to Bone Marrow Transplants (1999) by Karen Crowe	Children's book	www.us.astellas.com

general overview of the conditioning or preparative regimen (i.e., chemotherapy and total body irradiation versus chemotherapy only; number of days for conditioning), what occurs during the stem cell or bone marrow infusion (i.e., similar to an intravenous blood transfusion), and what happens after the infusion (i.e., waiting for engraftment, monitoring and treatment of infections or toxicities). Patients and parents typically describe the benefit of transplant to be a cure for their disease, whereas potential risks and side effects of HSCT are commonly discussed as death, symptoms such as nausea, pain, and/or fatigue, GVHD, infection, rejection or loss of graft, and/or long-term sequelae of treatment. Encouraging the patient and parents to discuss the process of arriving at the decision to choose an HSCT provides insight into the informed consent or assent process for the patient and family, such as whether the patient participated in the decision-making process, whether it was a mutual decision between the patient and parents, and what information or factors led them to the decision to proceed with HSCT. Patients and parents should understand any potential alternative options (e.g., palliative care) and therapies (e.g., chemotherapy only protocol) with associated benefits and risks of each option compared to HSCT. Patients and their parents should have awareness that they will be hospitalized for about 1 month and potentially longer based upon the patient's medical needs. To promote adherence, it is important for the patient and parents to understand what the oncologist expects of them during the HSCT hospitalization and recovery, such as taking multiple medications several times per day, bathing, mouth rinses, physical activity or therapy, and compliance with vitals. Finally, patients and parents should have an understanding of the length of the recovery process, number of outpatient clinic appointments per week and what those appointments will entail, and isolation expectations and duration. Semi-structured, open-ended patient interview questions (with parallel parent questions) aimed at assessing comprehension of each of these domains may best capture the depth of understanding (see Table 13.1 for sample interview questions).

Psychosocial Functioning of the HSCT Candidate

HSCT is an intensive treatment that is both physically and emotionally demanding. Not only do HSCT patients have to cope with painful procedures, treatment-related side effects such as nausea and vomiting, diarrhea and/or constipation, pain, fatigue, infertility, and worries about their health and survival, they also are isolated from school and friends and separated from family members during a lengthy hospitalization and recovery during which they are immunocompromised, all of which negatively impact their quality of life across physical, emotional, and social domains. Pediatric HSCT patients often experience increased distress and anxiety and lower health-related quality of life (HRQL) prior to their HSCT admission which extends through the acute phase of transplant and then begins to steadily improve 4–12 months post-HSCT before returning to baseline functioning by 1–3 years post-HSCT (Clarke et al. 2008; Packman et al. 2010; Tanzi 2011). Declines in social competence and self-esteem are also noted from pre- to 6 months post-HSCT (Phipps 2006). Risk factors for lower HRQL and negative psychosocial outcomes in HSCT recipients include older age at transplant, lower socioeconomic status, allogeneic (unrelated donor) transplant, presence of acute or chronic GVHD, pre-HSCT parental distress or maternal depressive symptoms, and pre-HSCT child distress or poor psychological functioning (Clarke et al. 2008; Packman et al. 2010; Tanzi 2011).

Given the multitude of HSCT stressors the pediatric recipient faces, it is important to assess the patient's current and past psychological functioning, stress, and coping prior to HSCT, but also intermittently about once per week (depending upon needs) during the acute and recovery phases of transplant. HSCT recipients should be screened for internalizing (i.e., depression, anxiety) and externalizing (i.e., oppositionality, inattention, hyperactivity) symptoms, which may impact coping with HSCT. Information also should be obtained regarding the patient's history of procedural distress, pain, and/or mood and

behavioral side effects secondary to steroid treatment. An assessment of the patient's current HRQL also will help the psychosocial provider ascertain the severity of the impact that the patient's disease and previous treatment history has had in physical/functional, emotional, social, and practical domains. Screening may include both open-ended interview questions and quantitative questionnaires or checklists completed by the patient (when age appropriate) and parent, particularly for younger children (see Table 13.1 for examples of measures). There are a few validated measures that have been developed for pediatric patients who undergo an HSCT, primarily to evaluate HRQL in the HSCT setting [i.e., Behavioral, Affective, and Somatic Experiences Scale (BASES), Child Health Rating Inventories – HSCT module (CHRIs-HSCT), Functional Assessment of Cancer Therapy – BMT (FACT-BMT)]. A review of the patient's preexisting mental health conditions, past or current participation in counseling services, past or current psychiatric medications or hospitalizations, and family psychiatric history should be conducted. For adolescent and young adults, it is additionally important to screen for past and current cigarette smoking, tobacco use, alcohol/illicit drug use, sexual activity, and suicidality or self-injurious behaviors (see Chapter 4 on Assessment).

Coping Strategies

There is a dearth of information related to the use of effective coping strategies in pediatric HSCT. HSCT recipients report using multiple strategies to cope with HSCT-related stressors. It is important to inquire what strategies patients have used to help them cope with illness-related stress, including prior hospitalizations, which can be promoted during transplant. Coping strategies may be cognitive, behavioral, social, or spiritual. Common coping strategies reported in clinical practice include distraction, engaging in hospital-based activities, reading, watching TV or movies, staying connected to or being with friends and family, online social networking, talking to others, and relying on faith and prayer (see Chapters 5 and 6 on Coping and Psychotherapeutic Modalities, respectively). Patients should be encouraged to discuss what activities they will be engaging in their hospital rooms on a daily basis and what items they will be bringing with them during the HSCT hospitalization to keep them busy. Similarly, it is important to inquire about any previous experiences with hospitalizations. This will help the psychosocial provider ascertain whether the patient and family have firsthand knowledge of what to expect with lengthy hospitalizations and chemotherapy side effects.

Medical Adherence

HSCT patients and their parent(s) face a challenging, complicated outpatient medication regimen once they are discharged from the HSCT hospitalization. Evaluation of medical adherence is one of the most essential domains to assess prior to an HSCT because it has the potential to directly impact treatment-related complications and HSCT outcomes. Because of the complicated daily medication regimen and lengthy recovery process, problems with medical adherence are not uncommon. A recent study found that adolescents who have undergone HSCT have difficulty adhering to the outpatient medication regimen (McGrady et al. 2014). Psychosocial providers should inquire about any past or current problems with pill swallowing, following the medical care regimen, listening to or complying with the instructions by the HSCT team, or missed clinic appointments. Communication with the referring medical provider may also be a helpful source of information to identify any adherence concerns. Any factors that may have contributed to adherence problems should be identified and intervened upon as soon as possible.

Case Vignette
Kyle, a 16 year old with relapsed acute lymphoblastic leukemia (ALL), presented for a pre-HSCT psychological evaluation. During the evaluation, he disclosed having

a complicated social and mental health history, including behavioral problems and placement in a foster care group home. Kyle was denied an HSCT at another hospital due to concerns about his mental health history. Adherence had not been reported as a problem. Utilizing a multidisciplinary team care approach, the patient was able to successfully receive an HSCT by proactively addressing his psychosocial and caregiving needs. Successful intervention approaches utilized with this patient included (1) establishing a team of consistent caregivers with whom he could develop positive, trusting relationships; (2) creating a daily schedule with age-appropriate activities; (3) providing clear, firm behavioral guidelines and expectations with rewards and consequences (i.e., privileges); and (4) participating in individual supportive psychotherapy, with goals of establishing a therapeutic relationship, utilizing cognitive-behavioral strategies to promote goal setting and positive behavioral choices as well as teaching anger and stress management techniques. Once discharged from the hospital after his HSCT, Kyle recovered for a period of time at an inpatient rehabilitation hospital.

Social Support and Stressors

Positive social support is a key factor in patients' coping with the stresses of HSCT (Bingen et al. 2012). Psychosocial providers should assess the level and quality of the support perceived by the patient and family and who is providing the most practical and emotional support to them. Support may be provided by a combination of family members, friends, school personnel, employer or coworkers, church or spiritual community, and/or neighborhood or surrounding community. Patients and families with limited support would benefit the most from consistent multidisciplinary supportive care services. Any stressors or barriers related to HSCT as well as non-HSCT stressors should be identified in order to potentially address or reduce these concerns prior to transplant whenever possible. For example, patients and their families often report financial strain related to previous medical treatment, potential loss of employment to care for the ill child, or health insurance problems, particularly for young adult patients who may be underinsured.

Family Functioning

Evaluation of family functioning should include obtaining information about the family constellation/structure, cohesiveness, conflict, and expressiveness as well as parental adjustment, parent-child relationships, and sibling relationships. In addition, the psychosocial provider should ascertain whether there have been any family role changes and disruptions in the family's daily life due to the patient's illness and treatment. For families who live a distance from the transplant center, it will be important to assess where they will be residing (e.g., Ronald McDonald House) and who will be taking care of the patient's siblings, when applicable. A qualitative assessment of pre-HSCT parental stress and coping and premorbid parental mental health history is crucial based upon previous research indicating that premorbid parental psychological functioning during the acute phase of transplant is a strong predictive factor in long-term parental adjustment after HSCT (Vrijmoet-Wiersma et al. 2009). Only parental and family history that is relevant to the child's HSCT should be sensitively documented in the child's medical records. Equally as important, both family and parental functioning prior to HSCT are predictive of child adjustment and HRQL after HSCT (Clarke et al. 2008; Packman et al. 2010; Tanzi 2011).

Cognitive, Academic, or Vocational Functioning

A comprehensive educational and cognitive assessment prior to HSCT can serve many purposes including determining the child's understanding of the HSCT, school needs during and after the HSCT, and reintegration after the

HSCT. As part of the pre-HSCT psychological evaluation and when insurance coverage permits, pediatric psychologists or neuropsychologists may also conduct cognitive and/or academic testing to directly evaluate HSCT candidates' functioning prior to HSCT, which can later serve as a comparison for future testing after HSCT to determine if there have been any cognitive changes over time. Due to HSCT workup time constraints as well as the patient's health at presentation, a comprehensive psychological or cognitive testing battery which would assess all domains that may be impacted by treatment may not be feasible. Because of this, an abbreviated testing battery is recommended to serve as a neurocognitive screen of the patient's current abilities (see Chapter 10 on Neurocognitive Issues).

Previous research has established that neurotoxic chemotherapy and radiation (particularly cranial radiation) can negatively impact cognitive functioning years after treatment, known as "cognitive late effects" (for review, see Mulhern and Butler 2006). However, due to methodological challenges such as small sample sizes from single transplant centers, it remains unclear as to whether the treatment regimen (myeloablative chemotherapy, total body irradiation, corticosteroid treatment for GVHD) for HSCT is a sole risk factor for development of cognitive late effects or an additive risk factor for patients who received chemotherapy (and is some cases radiation therapy) prior to HSCT,such as in the case of patients with ALL. In a review of longitudinal studies, Phipps (2006) concluded that there is low risk for development of cognitive or academic problems for children who receive an HSCT at or above the age of 6 years, but this risk may be increased for children who receive an HSCT when they are younger than 5 years old and, in particular, 3 years old or younger.

Research indicates that parental and child distress, HRQL, and adjustment fluctuate over the course of transplant and recovery (Clarke et al. 2008; Packman et al. 2010; Tanzi 2011; Vrijmoet-Wiersma et al. 2009) and therefore should be evaluated pre-, during, and post-HSCT. As a result, weekly assessments are recommended, even if only for a brief "check-in," during the acute and recovery phases of transplant, which can gradually become less frequent over time based upon supportive care needs and frequency of clinic appointments.

Clinical Interventions for Commonly Reported Psychosocial Issues in Pediatric HSCT

Procedural Anxiety and Pain

Children undergoing an HSCT frequently experience invasive and/or painful procedures, such as nasogastric (NG) tube placement, peripherally inserted central catheter (PICC) or central venous line dressing changes, and apheresis collection of stem cells. Managing pain and reducing procedural distress become important priorities for clinicians that provide supportive care. The following is a brief review of clinical interventions for pain and procedural anxiety; see Chapters 6 and 7 for a more comprehensive review.

Cognitive-behavioral interventions, including deep breathing and relaxation, distraction, psychoeducation, behavioral rehearsal, modeling, and imagery, have all been demonstrated to be efficacious in managing pain and anxiety during procedures (Kuppenheimer and Brown 2002; Packman et al. 2010). For more painful procedures, a combination of pharmacologic and psychological interventions may be needed to effectively reduce distress (Kazak et al. 1998).

Behavioral Concerns

Care plans are used during HSCT admissions to manage nonadherence (e.g., refusing to take medications), motivate cooperation (e.g., participating in physical therapy), and set the tone for expectations coming into a lengthy hospitalization. Ideally, care plans are created with the cooperation and input of all pertinent parties, including medical and nursing staff, psychosocial providers, parents, and patients. The care plan should be individualized for each patient's particular needs

with developmental age and cultural considerations in mind. The care plan is meant to be a more comprehensive document than a reward program or behavioral management plan alone (although these may be included in a care plan). It also may include an overview of tasks that must be completed daily by the patient, formalize the role of daily structure or schedule from wake to bedtime, and set expectations for parental involvement. Care plans should be shared with families and documented in the medical record and a copy placed in the patient's room.

Case Vignette

Suzy, a 6-year-old girl with relapsed ALL, had numerous admissions for chemotherapy prior to proceeding with HSCT. During these admissions, Suzy often refused to take her medications. When urged to comply, Suzy would hit and kick staff. Although Suzy's parents were supportive of staff efforts to garner compliance, they would also get angry with staff when Suzy was upset. In anticipation of her HSCT, Suzy's transplant psychologist in conjunction with her parents created a sample care plan to help set appropriate expectations for her transplant (see Box 13.1).

Behavioral management plans to address specific behaviors may be part of the larger care plan. These plans follow the principles of behavioral modification and include specific expectations, along with rewards for successful completion of tasks. In the case of Suzy, a "behavior bucks" program was implemented whereby she earned a "behavior buck" each time she completed an assigned medical task (e.g., taking medication within 15 min). She was able to "go shopping" with her earned behavior bucks for small prizes (similar to prizes found in the treasure box at a doctor or dentist office) and privileges that were selected and priced for the plan. Behavior bucks were withheld and privileges withdrawn for nonadherent behavior.

The success of behavioral care plans during transplant hinges on the consistency with which they are followed among care providers over time. Before beginning any plan, there should be discussion about who will implement the plan (i.e., parents and/or hospital staff). If hospital staff will carry out the plan, communicating the plan and training the many staff that may be impacted is of the utmost importance.

Coping with Lengthy Hospitalization and Isolation

The stringency of isolation restrictions during HSCT admission varies by institution. Nevertheless, at a minimum, children undergo a lengthy, physically and emotionally demanding treatment during which time they are unable to attend school or participate in typical social activities. Cognitive-behavioral and supportive therapies may be utilized in reducing sadness and anxiety during the acute phase of hospitalization. Systematic research is needed before these therapies should be applied as standard of care.

Anecdotally, consistent care provided by a small team of primary nurses increases patient and family comfort and willingness to seek support and/or ask questions. Nurses that are more familiar with the psychosocial issues of a specific patient are better able to tailor their support efforts and provide increased consistency in implementing care plans. Daily schedules can be used effectively to increase predictability and establish routines during hospitalization.

Depending on transplant unit policies, visitation from family and friends can provide much needed support to the patient during long-term hospitalization. Some patients may benefit from connecting with transplant survivors, which can be facilitated by the psychosocial provider with both parties permission or through a national program, such as Imerman Angels (see Table 13.2).

The role of technology during HSCT is an understudied area. Texting, social media websites, video-sharing websites, and video chatting all offer ways for patients to receive encouragement and psychological support, socialize, seek

information, and entertain themselves in ways that did not exist for patients a decade ago (see Chapter 19). Future research related to the uses and gratifications of technology is warranted.

Pill Swallowing

Throughout the HSCT admission, children are required to take medications orally. As the date of discharge nears, medications that were previously given intravenously are transferred to oral form. Most medications can be given in liquid form, but children complain of bad taste and large volume. Therefore, emphasis is placed on being able to swallow pills. This can be quite anxiety provoking for some children and lead to crying spells or avoidance in anticipation of having to take medication. If possible, pill swallowing should be taught well before the date of discharge as the pressure to learn a new skill under time constraints can increase distress and frustration.

The standard approach to teach pill swallowing is to use systematic desensitization utilizing placebo pills of various sizes. Candies, such as Tic Tacs and Mini M&Ms, are frequently used to mimic the different shapes and sizes of oral medications. Candies that are very light are not recommended as they have a tendency to float around the mouth rather than be swallowed. The clinician should begin by providing a brief explanation of the basic anatomy of the esophagus, including the ability to stretch around large amounts of food. Then, children should be taught to place the placebo pill on the back of their tongue. A sip of preferred liquid (i.e., water or milk; soda is discouraged as the carbonation can cause stomach upset that children can attribute to the pill) precedes swallowing the placebo. After several successes, children are gradually moved up to larger sized placebos until they are able to swallow candies that will resemble the size and shape of home medications.

There are times when the standard approach to pill swallowing proves ineffective. Pill swallowing aids, such as Pill Glide (a lubricated flavored spray) and pill cups that allow children to swallow water and a pill simultaneously, have been used with mixed results (Diamond and Lavallee 2010). Kaplan et al. (2010) demonstrated success utilizing a novel approach focused on head placement (see a training video at www.ucalgary.ca/research4kids/pillswallowing). Regardless of approach utilized, it is imperative to educate parents about the importance of keeping the environment calm during practice sessions and to handle failures with as much neutrality as possible.

Preparing for Discharge

Discharge discussions begin once children have demonstrated engraftment, are able to take oral medications, and are infection-free. Discharge is a time of ambivalence for parents and patients alike. On the one hand, families are excited to leave the hospital following a lengthy admission; on the other hand, they can be overwhelmed by the complex medical needs that they now must manage with more limited hospital support. Most institutions require parents to participate in a variety of teaching topics and demonstrate proficiency before they are able to be discharged. Having parents complete all medical care over a 24-hour period during the final days of hospitalization can increase competency and confidence. Nevertheless, parents continue to report high levels of stress as they prepare to take over their child's care. Normalization of their feelings and reassurance that they can reach someone by telephone day or night can be helpful in quelling some of the anxiety. In the early days following discharge, parents are also appreciative of the intensive follow-up that is required post-HSCT. Although difficult in certain settings, continuity of care between the inpatient and outpatient setting can also ease distress.

As families prepare to leave the hospital, it is also important to establish realistic expectations about the possibility of future hospitalizations given the immunocompromised state of the patient. This preparation can decrease feelings of guilt, frustration, and disappointment upon readmission.

Adherence at Home

The medical requirements following HSCT discharge are demanding and time consuming. There are medications to take throughout the day, dietary and activity restrictions, and caloric intake and fluid goals. Nonadherence can lower the chances of survival, increase risk of complications, and lead to poorer quality of life. Unfortunately there are no known adherence interventions created specifically for the HSCT population; however, because the demands of daily care are similar in complexity to some of the other pediatric chronic health conditions, one can extrapolate from interventions targeted at other chronic illness populations. Adolescents and young adults are particularly vulnerable to high rates of nonadherence because they are still developing independence and self-management skills, have different immediate priorities, have interrupted social support, and sometimes lack detailed knowledge of their treatment. Drotar (2013) recently proposed a comprehensive collaborative adherence promotion model that can be adapted for the adolescent and young adult population as described by Butow and colleagues (2010; see Table 13.3).

Table 13.3 HSCT adherence model

Develop a collaborative relationship and increase ownership in health by involving the patient in all HSCT-related discussions.
Discuss adherence throughout the course of treatment. Critical time points: discharge from the HSCT hospitalization, any time there is a change in treatment regimen (e.g., discontinuing intravenous fluids and increasing the need for oral intake), and when the patient takes over self-management.
Education is key! Discuss what each medication treats/prevents, how it works, and why it is important to take consistently. Highlight the importance of participating in HSCT follow-up care and continuing to follow isolation restrictions even after feeling better.
Ask how medication is managed at home and what strategies (e.g., pill box) are being utilized to facilitate adherence.
Acknowledge and normalize barriers to adherence (e.g., returning to school/work, changes in dosing, and adverse side effects). Flexibly and creatively strategize solutions (e.g., altering the timing of follow-up appointments that interfere with classes).
Establish a plan to monitor adherence (e.g., using laboratory values to test for therapeutic levels of cyclosporine and tacrolimus, frequency of medication refill requests).

Adapted from Drotar (2013) and Butow et al. (2010)

School Reintegration

Returning to school following an HSCT can be a difficult transition given the academic and social difficulties inherent in extended time away from the classroom due to medical isolation precautions. Additionally, treatment-related medical (e.g., fatigue) and neurocognitive late effects can have long-ranging impact on the successful return to school. While there are no school reintegration studies aimed at the HSCT population, interventions specific to pediatric cancer have primarily focused on improving communication between HSCT providers and school personnel, addressing academic needs through homebound education, and preserving social relationships during post-HSCT home isolation (Tadmor et al. 2012). Many patients benefit from a gradual return to school (e.g., half days) as well as supportive educational services (i.e., 504 Plan, Individualized Education Plan). Chapter 11 covers this topic in great detail.

Sibling Donors

When considering HSCT, physicians must consider the degree of human leukocyte antigen (HLA) "match" between donor and recipient; those with a strong degree of match are considered immunologically compatible. Siblings have a 25 % chance of being an HLA match with one another and, as such, are often the first to be considered for donation once the decision has been made to pursue an allogeneic HSCT as the treatment course. Matched sibling transplants also offer lower risk of GVHD, nonrelapse-related mortality, and equivalent, if not superior, rates of disease-free survival as compared to unrelated bone marrow and cord blood transplants (Zhang et al. 2012).

Donor Assessment

The decision to have one child undergo a non-therapeutic medical procedure for the benefit of another child has been discussed extensively in the literature and has been brought before the court system (Pentz et al. 2008; also see Chapter 17). Historically, parents have made the decision for a minor child to donate to his sibling because it was implied that children were unable to make such a decision for themselves. In a study examining hospital management of minor donors, Chan and colleagues (1996) found that 84 % of pediatric transplant centers allow parental consent as the only documentation necessary to permit a child to donate bone marrow. However, the method of sole parental consent has been questioned in the literature, suggesting that parents of minor children are conflicted decision makers for sibling donation (Pentz 2006; Ross and Glannon 2006).

Despite evidence to suggest that children as young as 9 years of age are able to make medical decisions on their own behalf (Weisz 1992; Weithorn and Campbell 1982), only two states – Alabama and Wisconsin – have written statutes that specifically address the issue of minor sibling donation. In Alabama, minors may consent to donation of their bone marrow once they have reached the age of 14 years or have been emancipated (ALA. CODE § 22-8-9, 2012). The Wisconsin law (WIS. STAT. ANN. §146.34, 2012) allows potential donors to assent as long as the transplant physician has informed the parent/donor about the benefits and risks to donor and recipient and alternative treatments to transplant. The minor also must be deemed the most acceptable donor, medically cleared to donate, and undergo a psychological and intellectual evaluation.

Studies have found that sibling donors do not feel as if they have a choice in the decision to donate (Packman et al. 1997; Pentz et al. 2014) and feel compelled to do it (MacLeod et al. 2003; Wiener et al. 2007). Even years after their donation, siblings may understand why they needed to donate but wish that they could have had more influence in coming to that conclusion for themselves.

Once the decision to donate has been made, many donors continue to be ambivalent toward the process; while they may feel proud to be the donor, many also report feeling very nervous about the idea of injections, pain, and possible harm during the procedure (Wiener et al. 2008). The literature focusing on the sibling donor's experience after donation is limited, but points to a number of consistent findings. In the months and years following donation, siblings retrospectively report experiencing feelings of anger, jealousy, guilt about being healthy, anxiety, sadness, and loneliness (Freund and Siegel 1986; Packman et al. 1997; Wiener et al. 2008; Wilkins et al. 2007).

The emotional impact of donation can be complicated by the health of the recipient sibling. Wiener and colleagues (2007) found that donor siblings of successful transplants without complications reported overall positive experiences, such as an increased closeness of the family and increased understanding of the recipient sibling's illness, while donor siblings of successful transplants with complications, such as GVHD, had negative experiences and felt responsible for the outcome of the transplant. Donor siblings of unsuccessful transplants resulting in the death of the recipient experienced feelings of blame, guilt, and anger.

As a result of the vague and/or nonexistent state laws governing minor donation, along with the known psychosocial risks of donation, the American Academy of Pediatrics (AAP 2010) created guidelines to direct the participation of minors as hematopoietic stem cell donors (see Table 13.4). The AAP further suggested that potential donors be assigned an advocate early in the process whom will help them understand the process of donation and protect their rights (see Table 13.5). The meeting with the donor consists of three primary goals: (1) determine ability and willingness to assent to donation, (2) minimize the emotional risks of donating, and (3) investigate the quality of the sibling relationship and potential likelihood of emotional benefits from donation. See Table 13.6 for a recommended assessment model.

Table 13.4 AAP guidelines for minor participation in hematopoietic stem cell donation

No medically equivalent histocompatible adult relative willing and able to donate
Strong personal and positive relationship between the sibling donor and recipient
Some likelihood that the recipient will benefit from transplant
Clinical, emotional, and psychosocial risks to the donor are minimized
Parental permission and donor assent are obtained

Table 13.5 Donor advocate

Role and training
Not involved in the care of the recipient
Trained in child development
Has knowledge of HSCT and donation
Process
Initial contact should be as soon as the donor begins the process of medical clearance to donate
Meetings with donor can be conducted jointly with caregiver, although separate interviews may yield more meaningful information

It is common for donors to voice misinformation or fears during the assessment. Most of these issues can be addressed by the donor advocate, although occasionally the donor may be referred back to the medical team for additional education. Very rarely, a sibling will report information that may make them an unsuitable candidate for donation, such as unwillingness to donate/coercion or a significant mental health history that places them at very high risk of emotional distress. In these cases, the transplant team, in conjunction with the donor advocate, must make a decision about whether to proceed. If available, an ethics consult can also be helpful in these cases.

Case Vignette
Megan, the 14-year-old sister of a boy with refractory ALL, was evaluated by a psychologist as a potential donor for her brother. During the evaluation, she reported feeling forced to donate her marrow because her parents said if she didn't she would be to blame if her brother died. The family history was complicated by parental divorce and reports of abuse and neglect in both households. Megan described her parents as physically abusive and emotionally unavailable. Megan had a history of depression and suicidal ideation and was being followed by a psychotherapist in the community. Based on the results of the predonation psychological evaluation, the HSCT team determined that she was at significant emotional risk and opted to proceed with a different course of treatment for her brother.

Intervention

A recently published multisite prospective study by Pentz and colleagues (2014) advocates for basic education for all potential sibling donors before HLA typing, more robust education for identified donors, and ongoing supportive follow-up in the post-transplant period. The donor assessment described above can serve as an initial therapeutic intervention because it normalizes the experiences and feelings of the donor, prepares them for the emotional experience of donation and recovery, and establishes guidelines for parents about when to seek additional psychotherapeutic support. Beyond the initial assessment, the donor advocate remains a resource for the family and ideally would provide supportive follow-up care; however, there are practical and emotional barriers to providing ongoing care to donors. Following donation, siblings return to their usual routine at home and school and become less available for intervention. Parents already busy at the bedside with the recipient may be unaware of any distress occurring at home or too emotionally taxed to manage less pressing issues. Donors, aware of parental stress, may be unwilling to burden them with additional concerns and try to manage their feelings on their own.

Table 13.6 Summary of minor sibling pre-donation psychological evaluation

Assessment goals	Salient assessment factors	Examples of interview questions	Examples of measures (time/resources permitting)
Determine ability and willingness to assent to donation	Donation procedure Decision making/voluntariness Alternative treatment options Ability to read assent/consent form	What is your understanding of the procedure to collect your stem cells/bone marrow? What do you expect for your recovery? How did you make the decision to donate? What were the most and least important factors in making this decision? When were you told that you could donate bone marrow to your sibling? What were your thoughts when you were told that you were a match for your sibling? If you decided not to be a donor for your sibling, what other options are available for your sibling?	WRAT4, Reading Composite (Wilkinson and Robertson 2006)
Minimize emotional risks of donating	Current psychosocial distress (depression, anxiety, needle phobia) Expectations for recipient's health Control and responsibility	Tell me about your previous experience with blood draws/IVs. What have you been told to expect for your sibling? What is the name of your sibling's illness? How responsible do you think you are for your sibling's health? How much control do you have over your sibling's health? Do you know what GVHD is? Explain your understanding of GVHD.	*Behavioral checklists* ASEBA Self-Report (Achenbach and Rescorla 2001) BASC-2 (Reynolds and Kamphaus 2004)
Investigate the quality of the sibling relationship and potential likelihood of emotional benefits from donation	Family functioning Sibling relationship (strength of foundation for personal connection)	What was your family like before your sibling got sick? What is it like now? What do you and your sibling like to do together? Name some of your sibling's friends.	Observation of sibling interaction
Educate caregivers	Donor psychosocial history Provide education and resources	What role has your child played in the decision to donate? How can you help your child feel more prepared for the procedure? What do you view as the psychosocial risks and benefits of donation? How can you support your child after donation knowing that your other child will be in the hospital and undergoing intensive medical treatment?	*Behavioral checklists* ASEBA Parent Proxy (Achenbach and Rescorla 2001) BASC-2 (Reynolds and Kamphaus 2004)

When a recipient dies or experiences significant complications (e.g., chronic GVHD), the likelihood of donor distress increases. In these cases, the donor advocate is encouraged to reach out to the donor to assess their need for additional support. This may include psychotherapeutic interventions by the donor advocate or referral to a local mental health provider.

Parent, Sibling, and Family Functioning

HSCT is not only stressful for the recipient, but it has a significant impact on the family as well. As a result, HSCT assessment and interventions should be family-centered throughout the HSCT care continuum. The following sections provide an overview of the impact of HSCT on parents, non-donor siblings, and the family as well as supportive care needs.

Parental Stress, Coping, and Adjustment

Parents are faced with a host of practical and emotional challenges when their child receives an HSCT. They are naturally worried about their child's health and well-being given the child's high-risk disease and the intensity of the HSCT regimen with potential severe or life-threatening treatment-related side effects or complications. They are concerned about whether the transplant "will work" to cure their child's disease. For patients who have an HLA-matched sibling, parents have the additional worry of the well-being of their child who donated. Employed parents may have to reduce their work hours or take a medical leave of absence to care for their child during and after HSCT, thereby creating a significant financial strain on the family. This is extremely burdensome in single-parent households in which there is only one income supporting the family, and a loss of employment may lead to a loss of health insurance for the HSCT recipient and family. In a two-parent household, one parent may be the "resident caregiver" during the HSCT hospitalization and outpatient recovery while the other parent may be working full-time to financially support the family. The primary breadwinner may feel guilty about not being able to care for their child as much as they would like, whereas the resident caregiving parent may feel overwhelmed with the full-time care responsibilities. While parents are trying to deal with all of these emotional and practical challenges, they may also be caring for other children and/or their own elderly parents. Parents struggle with trying to find alternative caregivers for their children when they are in the hospital with the HSCT recipient and are challenged to maintain some normalcy for their children such as attending school and participating in extracurricular activities and social events. Parents feel guilty about not being present or being less available for their other children while in the hospital caring for the HSCT recipient. This is a particular struggle for single-parent households or for families who live a long distance from the transplant center.

Given the number of HSCT-related stressors, it is important to assess parental stress and coping across the HSCT care continuum. Similar to the trajectory of child distress, a review of research of parents of children receiving HSCT found that the majority of parents reported significantly higher levels of distress (anxiety, depression, posttraumatic stress symptoms, somatic complaints) prior to and during the acute hospitalization phase of HSCT compared to normative group data, with distress levels decreasing over time especially between 3 and 6 months following transplant and returning to baseline or positive adjustment by 18 months post-HSCT (Vrijmoet-Wiersma et al. 2009). Despite this, a subset of parents is at risk for difficulty coping years after their child's HSCT. Risk factors for long-term parental distress were grouped into three categories: (1) factors related to the child's disease and treatment (transfers to the intensive care unit, higher number of hospitalizations 6 months after HSCT, and higher HSCT risk); (2) demographic factors (younger maternal age, lower socioeconomic status); and (3) parental stress and coping during the acute phase of HSCT or premorbid parental or child psychosocial func-

tioning. Protective factors of parental distress included mothers with a more optimistic personality or who used coping strategies of acceptance, humor, and "putting reason before emotion" or parents who had a supportive family environment (Vrijmoet-Wiersma et al. 2009). Unfortunately, results of the reviewed studies are mainly generalizable to mothers as very few studies included fathers or had a small number of fathers as respondents.

Sibling Stress, Coping, and Adjustment

Far less is known about sibling adjustment and supportive care needs when a child receives an HSCT. Siblings who are not donors of HSCT recipients still have to cope with one or both parents being less physically or emotionally available because they are caring for their brother or sister in the hospital or at frequent clinic appointments during recovery. Siblings may need to stay with relatives or family friends and, as a result, may feel disconnected or displaced from their family. In addition, siblings may be less able to socialize with their friends or participate in extracurricular activities as they used to before their brother or sister became ill. In a review of the scant literature, Packman et al. (2010), who have conducted the majority of studies on siblings of HSCT recipients, concluded that siblings of HSCT patients experience a range of natural feelings but some report posttraumatic stress symptoms, anxiety, and low self-esteem. Siblings of HSCT recipients also expressed interruption in their "normal" family life, a lack of parental attention, limited understanding of transplant, feelings of loneliness, and a reliance on a faith in God to help them cope, which were thematic responses in qualitative studies that were reviewed (Packman et al. 2010).

Family Functioning

When a child undergoes an HSCT, many aspects of a family's life are disrupted. Families of children who receive an HSCT experience a reliance on supportive others for assistance, disrupted daily schedules and routines, changes in familial roles, economic strain, and separation or difficulty finding time to spend together as a family. Both positive (i.e., family cohesion) and negative (i.e., family conflict) family functioning have been shown to be predictive factors for parent and child outcomes in HSCT (Clarke et al. 2008; Phipps et al. 2005; Vrijmoet-Wiersma et al. 2009). There has been limited research on the impact of a child's HSCT on the family unit, with family functioning as the primary outcome. Jobe-Shields et al. (2009) found that higher parental depressive symptoms were associated with lower levels of family cohesion and expressiveness, and lower family conflict and greater family expressiveness was related to less severe illness-related distress reported by the child prior to HSCT. Pre-HSCT family cohesion and expressiveness served as protective factors against child illness-related distress when parents reported low depressive symptoms, but this was not the case when parents reported high depressive symptoms (Jobe-Shields et al. 2009), which highlights the importance of addressing the supportive care needs of parents and families when a patient receives an HSCT.

The impact of the child's HSCT on the parents' marital relationship is not known. Anecdotally, parents face reduced or limited time together as a couple (physical separation for some due to distance) because of their child's HSCT care demands, which can make communication and support difficult. Parents often have to cope with role changes in the family, which also may strain the relationship. Extrapolating from the few studies that have investigated the impact of childhood cancer on parents' relationship suggests that there is no elevation in divorce rates (Syse et al. 2010), but negative or positive changes may occur depending on factors such as time since diagnosis (minimal or positive changes in relationship the further out from diagnosis), communication between parents (quality and quantity), gender differences in stress and coping, and parental role changes (Da Silva et al. 2010).

Family-Based Interventions

Multidisciplinary supportive care of families, based upon family systems and stress and coping models, is necessary to provide comprehensive care that helps to reduce HSCT-related stress and enhance existing positive support systems and healthy coping strategies. For example, hospital-based social workers have the expertise necessary to assist parents or young adult patients as they apply for financial assistance, including Supplemental Security Income disability benefits, address health insurance and prescription medication issues, identify or arrange medical transportation, assist in filing for family medical leave, and complete advanced directives or power of health care agent documents.

Psychosocial providers should inform and connect families to local and national support programs and resources (see Table 13.2) and can advocate for support and assistance from the family's support system such as school personnel for the siblings (i.e., teacher, school psychologist, or guidance counselor) and employers for the parents, with the family's permission. Many parents and siblings express benefiting from being connected to other parents and siblings of children who have undergone HSCT. Referrals to support groups and family-based or sibling support camps should be provided, when available. Psychosocial providers also may initiate communication with another willing HSCT family whose child has received an HSCT at the transplant center in person, by telephone and email, or via online social networking (Facebook), or this connection may be facilitated through a national program such as Imerman Angels (see Table 13.2; see Chapters 25 and 11 on Resources and Education, respectively).

Importantly, psychosocial providers should provide family-centered care by providing supportive counseling to parents, siblings, and the family unit when an immediate family member is having difficulty coping with the child's HSCT, which may decrease short- and long-term negative family outcomes. Goals of supportive counseling may consist of strengthening and teaching healthy coping, problem-solving, and stress management strategies, teaching effective family communication skills, and encouraging self-care activities, such as eating healthy, taking regular breaks from the bedside for respite whenever possible, physical activity or exercise (even if it is a walk in or around the hospital), and good sleep hygiene. Referrals to community-based mental health providers may be necessary to address and treat premorbid mental health disorders. Hospital-based chaplains should also be accessible to meet their religious or spiritual needs or to help coordinate visits from their community-based church or spiritual leaders.

The majority of supportive care provided to families of children who undergo an HSCT is based upon anecdotal evidence in clinical practice. Despite family functioning being a strong contributing factor to parental and child distress or adjustment in HSCT, to date there have been only two published intervention studies that targeted parents of children undergoing HSCT (Lindwall et al. 2014; Streisand et al. 2000). Streisand et al. (2000) evaluated a one-session stress inoculation intervention for mothers ($N=22$) of children receiving an HSCT, who found no difference in parental stress between mothers who were randomly assigned the intervention compared to those who received the standard care. A more recent multisite randomized controlled study compared the parental benefit of a child targeted complementary therapy intervention (massage and humor therapy), a combined parent (massage and relaxation) and child intervention (massage and humor therapy), and standard of care (Lindwall et al. 2014). Results suggested no significant differences between groups in the areas of acute parental distress, positive affect, depression, or posttraumatic stress. Parents in all three groups demonstrated significant improvement over time across all domains, indicating that parental functioning decreases around the time of HSCT admission but returns to baseline functioning over time (Lindwall et al. 2014). Parsons and colleagues (2013) also recently completed a 6-month randomized controlled multisite study of a web-based support and education intervention for parents of pediatric HSCT recipients ($N=102$

intervention arm; $N=99$ control arm) known as HSCT-CHESS™ (Comprehensive Health Enhancement Support System) funded by the National Cancer Institute. Preliminary results indicated no intervention effect on parental emotional functioning at 6 months post-HSCT. However, parents who used the website though the 6-month intervention period had a significantly higher emotional functioning score (6.3 points higher; effect size = 0.32) compared to parents who were randomized to the intervention but did not actually use the website (Parsons et al. 2013).

Future Directions and Conclusion

Despite the significant inherent challenges of a lengthy HSCT hospitalization and recovery with a major disruption of normalcy and social isolation, most children, adolescents, and young adults and their families demonstrate resiliency and seem to recover over time from the distress they experience prior to and during the acute phase of transplant. However, a subset of patients and parents continue to struggle years after transplant. Therefore, it is important to understand the contributing risk and protective factors for HSCT outcomes in patients and families. Pre-HSCT functioning of the patient and family is a strong predictor of post-HSCT functioning based upon existing research, which highlights the importance of conducting a comprehensive psychological assessment prior to HSCT but also throughout the different phases of HSCT given our understanding of the trajectory of distress and HRQL. Despite the limited assessment tools developed for use in pediatric HSCT, consideration should be made for the inclusion of quantitative measures of psychosocial functioning and HRQL that have been validated with children, adolescents, and young adults in cancer and other chronic illness populations. Similarly, most clinical interventions that have been implemented in the pediatric HSCT setting are based upon "what works" or "what doesn't work" in clinical practice to support the patient and family. More research is needed to develop and evaluate interventions specific to the pediatric HSCT population, their parents, siblings, and families and/or determine the efficacy of existing interventions in cancer and other chronic illness for use in pediatric HSCT. Regardless, given the intensity and complexities of HSCT care, supportive care should be multidisciplinary, with care coordination and communication among providers, and family-centered and consistently delivered from inpatient to outpatient settings across the HSCT care continuum. Special attention also should be made for siblings, both donors and non-donors, and family functioning as a primary focus of research. Longitudinal, multisite studies with larger sample sizes are essential to gain a better understanding of siblings' natural reactions to changes in the family related to HSCT versus siblings at risk for psychosocial difficulties.

Clinical Pearls

- Psychosocial providers should have expertise in the unique stressors and supportive care needs of pediatric HSCT recipients and their families.
- Assessment and intervention should be family-centered given the impact of HSCT on patients, parents, and siblings.
- Sibling donors require an assessment and supportive care approach that takes into consideration their unique experience.
- Evidence-based psychosocial assessment and treatment specific to pediatric HSCT is needed.

Box 13.1: Sample HSCT Care Plan

The following plan has been created in collaboration with the HSCT team, nursing, supportive services, and Suzy's parents in order to help Suzy have a successful and safe transplant experience.

A. Suzy will have many new demands placed on her during her transplant. It is recommended that the following expectations be incorporated into a reward system so that she is motivated to participate in her care. The reward system will be created by parents and psychosocial providers.
   (a) Oral medications must be taken within 15 minutes regardless of parent presence. A timer can be set if this would be a helpful visual reminder for Suzy.
   (b) Suzy must sleep in her own bed every night, all night.
   (c) Mineral oil baths three times daily.
   (d) Clothing changed two times daily.
   (e) Mouth care to include brushing teeth two times a day and mouthwash four times a day.
   (f) Vitals every 4 hours. May be more often if medically necessary.
   (g) Participation in bedside education.
   (h) Participation in physical therapy.

B. If Suzy refuses to cooperate or becomes disrespectful toward nursing, parents are encouraged to leave the room and allow nursing to work individually with Suzy to complete her care.

C. A sleep/wake routine will help Suzy to maintain structure in her day. It will also guarantee better cooperation with early morning demands and help facilitate her transition back home after transplant. Sleep/wake routines have been shown to improve patient and parent quality of sleep.
   (a) At 9:30 PM, parents and/or nursing will remind Suzy that she needs to be in her bed with lights out and television off by 10:00 PM. This warning will help prepare her and allow her time to complete any bedtime cares (e.g., brushing teeth).
   (b) At 10:00 PM, lights and television must be turned off. If nursing enters the room after 10:00 PM and this has not happened, they will provide a verbal reminder and then turn off lights and television.
   (c) Parents and/or nursing will wake Suzy at 7:30 AM in preparation for 8:00 AM vitals and weight.
   (d) Napping during the day is permitted. However, daytime naps will be limited, particularly late afternoon naps when Suzy feels well, in order to facilitate better sleep routines at night.

D. Suzy is encouraged to play outside of her room as long as she is not on isolation precautions.

E. Good hygiene is of the utmost importance during transplant. Parents and Suzy are encouraged to handwash or sanitize every time they reenter the room. Food and drink should be fresh. All partially consumed food and drink should be thrown away. Suzy should not eat or drink after anyone else.

F. Parents are strongly encouraged to participate in morning rounds to help ensure that they understand the plan as presented by the team.

References

Achenbach TM, Rescorla LA (2001) Manual for the ASEBA school-age forms and profiles. University of Vermont Research Center for Children, Youth and Families, Burlington

Ala. Code 22-8-9 (2012) Consent of minor to donation of bone marrow; Consent by parent or legal guardian

American Academy of Pediatrics (2010) Policy statement – children as hematopoietic stem cell donors. Pediatrics 125(2):392–404

Bingen K, Kent MW, Rodday AM, Ratichek SJ, Kupst MJ, Parsons SK (2012) Children's coping with hematopoietic stem cell transplant: results from the journeys to recovery study. Child Health Care 41(2):145–161

Butow P, Palmer S, Pai A, Goodenough B, Luckett T, King M (2010) Review of adherence-related issues in adolescents and young adults with cancer. J Clin Oncol 28(32):4800–4809

Cairo MS, Heslop H (2008) Pediatric blood and marrow transplantation: state of the science. Bone Marrow Transplant 41:97

Cella DF, Tulsky DS, Gray G, Sarafian B, Linn E, Bonomi A, Silberman M, Yellen SB, Winicour P, Brannon J, Eckberg K, Lloyd S, Purl S, Blendowski C, Goodman M, Barnicle M, Stewart I, McHale M, Bonomi P, Kaplan E, Taylor S IV, Thomas CR Jr, Harris J (1993) The functional assessment of cancer therapy (FACT) scale: development and validation of the general measure. J Clin Oncol 11(3):570–579

Chan KW, Gajewski JL, Supkis D, Pentz R, Champlin R, Bleyer WA (1996) Use of minors as bone marrow donors: current attitude and management as survey of 56 pediatric transplantation centers. J Pediatr 128(5):644–648

Clarke SA, Eiser C, Skinner R (2008) Health-related quality of life in survivors of BMT for paediatric malignancy: a systematic review of the literature. Bone Marrow Transplant 42:73–82

Da Silva FM, Jacob E, Nascimento LC (2010) Impact of childhood cancer on parents' relationship: an integrative review. J Nurs Scholarsh 42(3):250–261

Devine KA, Heckler CE, Katz ER, Fairclough DL, Phipps S, Sherman-Bien S, Dolgin MJ, Noll RB, Askins MA, Butler RW, Sahler OJ (2014) Evaluation of the psychometric properties of the pediatric parenting stress inventory (PPSI). Health Psychol 33:130–138

Diamond S, Lavallee DC (2010) Experience with a pill-swallowing enhancement aid. Clin Pediatr 49(4):391–393

Drotar D (2013) Strategies of adherence promotion in the management of pediatric chronic conditions. J Dev Behav Pediatr 34(9):716–729

Embry L, Annett RD, Kunin-Baston A, Patel SK, Sands S, Reaman G, Noll RB (2012) Implementation of multi-site neurocognitive assessments within a pediatric cooperative group: can it be done? Pediatr Blood Cancer 59:536–539

Folkman S, Lazarus R (1988) Ways of coping questionnaire. Mind Garden, Palo Alto

Freund BL, Siegel K (1986) Problems in transition following bone marrow transplantation: psychosocial aspects. Am J Orthopsychiatry 56(2):244–252

Furman W, Buhrmester D (1985) Sibling relationship questionnaire. Child Dev 56:448–461

Holmes TH, Rabe RH (1967) The social readjustment rating scale. J Psychosom Res 11:213–218

Jobe-Shields L, Alderfer MA, Barrera M, Vannatta K, Currier JM, Phipps S (2009) Parental depression and family environment predict distress in children prior to stem-cell transplantation. J Dev Behav Pediatr 30(2):140–146

Kaplan BJ, Steiger RA, Pope J, Marsh A, Sharp M, Crawford SG (2010) Successful treatment of pill-swallowing difficulties with head posture practice. Paediatr Child Health 15(5):e1–e5

Kazak AE, Penati B, Brophy P, Himelstein B (1998) Pharmacologic and psychologic interventions for procedural pain. Pediatrics 102:59–66

Kuppenheimer WG, Brown RT (2002) Painful procedures in pediatric cancer: a comparison of interventions. Clin Psychol Rev 22:753–786

Lindwall JJ, Russell K, Huang Q, Zhang H, Vannatta K, Barrera M, Alderfer M, Phipps S (2014) Adjustment in parents of children undergoing stem cell transplantation. Biol Blood Marrow Transplant 20:543–548

MacLeod KD, Whitsitt SF, Mash EJ, Pelletier W (2003) Pediatric sibling donors of successful and unsuccessful hematopoietic stem cell transplants (HSCT): a qualitative study of their psychosocial experience. J Pediatr Psychol 28(4):223–231

McCubbin HI, McCubbin MA, Patterson JM, Cauble AE, Wilson LR, Warwick W (1983) CHIP-coping health inventory for parents: an assessment of parental coping patterns in the care of the chronically ill child. J Marriage Fam 45:359–370

McGrady ME, Williams SN, Davies SM, Pai AL (2014) Adherence to outpatient oral medication regimens in adolescent hematopoietic stem cell transplant recipients. Eur J Oncol Nurs 18(2):140–144

Moos R, Moos B (1994) Family environment scale manual: development, applications, research, 3rd edn. Mind Garden, Palo Alto

Mulhern RK, Butler RW (2006) Neuropsychological late effects. In: Brown RT (ed) Comprehensive handbook of childhood cancer and sickle cell disease: a biopsychosocial approach. Oxford University Press, New York, pp 262–278

Packman WL, Crittenden MR, Rieger JB, Schaeffer E, Bongar B, Cowan MJ (1997) Siblings perceptions of the bone marrow transplantation process. J Psychosoc Oncol 15:81–105

Packman W, Weber S, Wallace J, Bugescu N (2010) Psychological effects of hematopoietic SCT on pediatric patients, siblings and parents: a review. Bone Marrow Transplant 45(7):1134–1146

Pai ALH, Patino-Fernandez AM, McSherry M, Beele D, Alderfer MA, Reilly AT, Hwang WT, Kazak AE (2008) The psychosocial assessment tool (PAT2.0): psychometric properties of a screener for psychosocial distress in families of children newly diagnosed with cancer. J Pediatr Psychol 33(2):50–62

Parsons SK, Shih MC, Mayer DK, Barlow SE, Supran SE, Levy SL, Greenfield S, Kaplan SH (2005) Preliminary psychometric evaluation of the Child Health Ratings Inventories (CHRIs) and Disease-Specific Impairment Inventory-HSCT (DSII-HSCT) in parents and children. Qual Life Res 14:1613–1625

Parsons SK, Mayer DK, Ratichek SJ, Rodday AM, Syrjala KL, Davies S, Patel SK, Schwartz L, Kupst MJ, Bingen K, Guinan EC, McTavish F, Stewart S, Terrin N, Gustafson DH (2013) An eHealth program for parents of pediatric hematopoietic stem cell transplant (HSCT) recipients enhances parents' emotional functioning with continued use: results of the HSCT Comprehensive Health Enhancement Support System (CHESSTM) Study. Psycho-Oncology 22:100–101

Pentz R (2006) Duty and altruism: alternative analyses of the ethics of siblings bone marrow donations. J Clin Ethics 17(3):227–230

Pentz RD, Haight AE, Noll RB, Barfield R, Pelletier W, Davies S, Alderfer MA, Hinds PS (2008) The ethical justification for minor sibling bone marrow donation: a case study. Oncologist 13:148–151

Pentz RD, Alderfer MA, Pelletier W, Stegenga K, Haight AE, Hendershot KA, Dixon M, Fairclough D, Hinds P (2014) Unmet needs of siblings of pediatric stem cell transplant recipients. Pediatrics 133(5):e1156–e1162

Phipps S (2006) Psychosocial and behavioral issues in stem cell transplantation. In: Brown RT (ed) Comprehensive handbook of childhood cancer and sickle cell disease: a biopsychosocial approach. Oxford University Press, New York, pp 75–99

Phipps S, Hinds PS, Channell S, Bell GL (1994) Measurement of behavioral, affective, and somatic responses to pediatric bone marrow transplantation: development of the BASES scale. J Pediatr Oncol Nurse 11:109–117

Phipps S, Dunavant M, Jayawardene D, Srivastiva DK (1999) Assessment of health- related quality of life in acute in-patient settings: use of the bases instrument in children undergoing bone marrow transplantation. Int J Cancer 12:18–24

Phipps S, Dunavant M, Lensing S, Rai SN (2005) Psychosocial predictors of distress in parents of children undergoing stem cell or bone marrow transplantation. J Pediatr Psychol 30(2):139–153

Reynolds CR, Kamphaus RW (2004) BASC-2, behavior assessment system for children, 2nd edn. AGS Publishing, Circle Pines

Ross LF, Glannon W (2006) A compounding of errors: the case of bone marrow donation between non-intimate siblings. J Clin Ethics 17:220–226

Sherman AC, Simonton S, Latif U, Nieder ML, Adams RH, Mehta P (2004) Psychosocial supportive care for children receiving stem cell transplantation: practice patterns across centers. Bone Marrow Transplant 34:169–174

Spirito A, Stark L, Williams C (1988) Development of a brief coping checklist for use with pediatric populations. J Pediatr Psychol 13(4):555–574

Stein REK, Riessman CK (1980) The development of the impact on family scale: preliminary findings. Med Care 18:465–472

Streisand R, Rodrigue JR, Houck C, Graham-Pole J, Berlant N (2000) Brief report: Parents of children undergoing bone marrow transplantation: documenting stress and piloting a psychological intervention program. J Pediatr Psychol 25(5):331–337

Syse A, Loge JH, Lyngstad TH (2010) Does childhood cancer affect parental divorce rates? A population-based study. J Clin Oncol 28(5):872–877

Tadmor CS, Rosenkranz R, Ben-Arush MW (2012) Education in pediatric oncology: learning and reintegration into school. In: Kreitler S, Ben-Arush MW, Martin A (eds) Pediatric psycho-oncology: psychosocial aspects and clinic interventions, 2nd edn. Wiley-Blackwell, Chichester

Tanzi EM (2011) Health-related quality of life of hematopoietic stem cell transplant childhood survivors: state of the science. J Pediatr Oncol Nurs 28(4):191–202

Tobin DL, Holroyd KA, Reynolds RVC (1984) User's manual for the coping strategies inventory. Ohio University, Department of Psychology, Athens

Varni JW, Seid M, Kurtin PS (2001) PedsQL 4.0: reliability and validity of the pediatric quality of life inventory version 4.0 generica core scales in health and patient populations. Med Care 39:800–812

Vrijmoet-Wiersma CMJ, Kolk AM, Grootenhuis MA, Spek EM, van Klink JMM, Egeler RM, Bredius RGM, Koopman HM (2009) Child and parental adaptation to pediatric stem cell transplantation. Support Care Cancer 17:707–714

Wechsler D (2004) The Wechsler intelligence scale for children, 4th edn. Pearson Assessment, London

Wechsler D (2011) Wechsler abbreviated scale of intelligence, 2nd edn. Pearson Assessment, London

Weisz V (1992) Psycholegal issues in sibling bone marrow donation. Ethics Behav 2:185–201

Weithorn LA, Campbell SB (1982) The competency of children and adolescents to make informed treatment decisions. Child Dev 53:1589–1598

Wiener LS, Steffen-Smith E, Wayne AS (2007) Hematopoietic stem cell donation in children: a review of the sibling donor experience. J Psychosoc Oncol 25:45–66

Wiener LS, Steffen-Smith E, Battles HB, Wayne A, Love CP, Fry T (2008) Sibling stem cell donor experiences at a single institution. Psychooncology 17:304–307

Wilkins KL, Hons BS, Woodgate RL (2007) An interruption in family life: siblings' lived experience as they transition through the pediatric bone marrow transplantation trajectory. Oncol Nurs Forum 34(2):28–35

Wilkinson GS, Robertson GJ (2006) Wide range achievement test four professional manual. Psychological Assessment Resources, Lutz

Wis. Stat. 146.34 (2012) Donation of bone marrow by a minor.

Zhang M, Davies SM, Camitta BM, Logan B, Tiedemann K, Eapen M (2012) Comparison of outcomes after HLA-matched sibling and unrelated donor transplantation for children with high-risk acute lymphoblastic leukemia. Biol Blood Marrow Transplant 18:1204–1210

Zimet GD, Dahlem NW, Zimet SG, Farley GK (1988) The multidimensional scale of perceived social support. J Pers Assess 52:30–41