exploring job satisfaction and factors contributing to job satisfaction specific to pediatric SCT.

Results:

Of the participants, >90% of the respondents categorized themselves as APSCTN, some with additional responsibilities including research and education and 85% are employed full time. 75% of the respondents provide direct patient care with additional duties in other areas.

Employing a Lickert-type scale from 1-10, 1 being very unsatisfied/very burnt out, 10 being very satisfied/not burnt out, 50% of APSCTN categorized themselves as a 7 or greater with overall job satisfaction. When characterizing current job burn out, 60% responded between 1-6, however when describing burnout about nursing as a profession, 80% responded as a 7 or greater (Table 1). Job satisfaction was associated with direct patient/family interaction, clinical care, and satisfying relationships with MD's and medical staff. Conflict with hospital administration, understaffing and difficulties with other hospital departments are cited as having negative impact on job satisfaction.

Conclusions:

Job satisfaction remains high in the SCT discipline despite difficult interactions with hospital departments, administration and chronic understaffing. However, nursing burnout remains a significant concern in pediatric SCT and should be addressed in this unique group of professionals.

References:

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Scale 1-10; 1 Very Unsatisfied/Very Burnt Out, 10 Very Satisfied/ Not Burnt out (n=20)

	≤3	4-6	7-9	10
Job Satisfaction	**2/20**	**8/20**	**10/20**	**0/20**
Burnout in Current Job	**4/20**	**8/20**	**6/20**	**2/20**
Burnout Nursing as a Profession	**1/20**	**3/20**	**9/20**	**7/20**

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FASCIITIS AS A MANIFESTATION OF CHRONIC GRAFT VERSUS HOST DISEASE FOLLOWING ALLOGENEIC HEMATOPOIETIC STEM CELL TRANSPLANTATION IN CHILDREN AND ADOLESCENTS: A SINGLE INSTITUTION EXPERIENCE

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Chronic graft versus host disease (CGVHD) remains one of the most challenging obstacles to successful outcome following allogeneic hematopoietic stem cell transplantation (HSCT). Although the incidence of CGVHD is less in children than in adults the impact is devastating as both disease and therapy can profoundly impact growth, development and functional outcome. Fasciitis is a rare manifestation of CGVHD that can occur without other organ involvement. Histologically edema and fibrosis are present in the septae separating subcutaneous fat lobules and in the muscular fascia beneath; overlying epidermis is unaffected. Patients present with painful edema that almost always progresses to fascial scarring. Ultimately the process leads to loss of tissue flexibility and a dimpled "peu d'orange " appearance of the skin associated with poor functional and cosmetic outcome. From January 1999 through December 2005 we performed 305 allogeneic HSCT; 5 patients received donor lymphocyte infusions (DLI) during this time.40 had significant CGVHD, 9 post- related SCT, 31 post-unrelated donor SCT and one following DLI. Of these patients 8 have been diagnosed with significant fasciitic CGVHD, 1 after a 3rd DLI, 3 after related donor SCT (2nd SCT for one)and 4 following matched unrelated donor(UD) SCT. 2 patients received peripheral blood stem cells as the donor source; the remaining 5 received bone marrow.3 had acute or chronic leukemias and 1 had an immunodeficiency. The children ranged from 4-17 years of age at the time of diagnosis and the diagnosis occurred 7 mos to 2 yrs after HSCT/DLI. All patients had oral GVHD at diagnosis; 7/8 had no other CGVHD. Excluding the patient developing fasciitis post-DLI, all failed to respond to steroid therapy including doses of up to 3mg/kg/day. Currently one child has died of infection with aspergillus and atypical mycobacterium, 2 are off all immunosuppression with resolution of disease after therapy with MMF and steroids, 2 have disease control on weaning immunosuppression including MMF and 3 have active disease that impacts mobility despite multiagent therapy. No patient with leukemia has had disease recurrence. Fasciitis can occur with minimal other manifestations of CGVHD and inital findings such as painful edema or fascial thickening may be subtle. Steroid therapy may prove ineffective illuminating the necessity for other therapeutic agents as significant functional limitations occur if the disease can not be controlled.

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SEVERE VENO-OCCLUSIVE DISEASE AFTER BONE MARROW TRANSPLANTATION IN PREMATURE MONOZYGOTIC TWINS WITH FAMILIAL HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS (FLH)

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Introduction: Familial hemophagocytic lymphohistiocytosis is a rare histiocytic disorder with hepatosplenomegaly, fever, infections and hemophagocytosis visible in biopsies of hematopoietic and lymphoid tissue. Hematopoietic stem cell transplantation is the only curative option. Bone marrow transplant recipients with FLH are at an increased risk for VOD as busulfan-based conditioning regimens add to liver toxicity. Here, we report two female, premature, identical twins with FLH and severe VOD following allogeneic BMT. **Case reports.** After diagnosis of FLH in the 2 months-old twins in spring 2006, the children achieved complete remission using protocol HLH-2004 (dexamethasone, etoposide, CsA) after 3 months. The girls received an unrelated matched marrow transplant from the same donor. The preparative regimen consisted of i.v. busulfan 16mg/kg BW, cyclophosphamide 200mg/kg BW and ATG 40mg/kg BW; GvHD prophylaxis was CsA and short course MTX. Twin A was transplanted two weeks prior to her sister. On day +9, she developed VOD associated with oliguria, renal and respiratory insufficiency necessitating mechanical ventilation. Further complications were disseminated intravascular coagulation (DIC) and potentially CsA-induced thrombocytopenic purpura requiring support with cellular and plasmatic blood products. Defibrotide and heparin were started with the onset of VOD and DIC. On day +49, her bilirubin varies between 20 and 30mg/dl, but otherwise she displays improved hepatic parameters, is breathing spontaneously with minimal, additional oxygen supply, has a sufficient urine output and no infections or GvHD under steroids and MMF. In twin B, the prophylactic use of defibrotide could not prevent VOD being diagnosed on day +13. However, she was not as severely affected as her sister without the need for mechanical ventilation. In other respects, she experienced the same complications. On day +35, her bilirubin varies between 15 and 25mg/dl, but she demonstrates a gradual improvement of hepatic function, her respiratory system is not impaired, the renal function slightly reduced under CsA. Both girls engrafted rapidly and exhibit full donor cell chimerism. **Discussion.** Significant liver involvement in FLH patients poses a considerable risk for developing VOD. The use of defibrotide could not prevent, but may have mitigated the severity of VOD in the second child. Children with FLH and a high risk for VOD may be considered for reduced intensity conditioning regimens.