436 Identifying locations of Merkel cell carcinoma associated with higher disease-specific mortality C. Cullinan, D. Z. Zheng1, MA Levoca1, JF Scott1 and JB Bordeaux1 1 Case Western Reserve University, Cleveland, Ohio, United States, 2 Johns Hopkins University School of Medicine, Baltimore, Maryland, United States and 3 Dermatology, University Hospitals, Cleveland, Ohio, United States

Merkel cell carcinoma (MCC) can occur anywhere on the skin surface, yet an understanding of whether tumor primary site impacts prognosis is currently incomplete within the literature. To best address this knowledge gap, we designed a study to analyze disease-specific mortality, rather than overall survival. A system survey or review, patients are often of advanced age and therefore may die of other causes prior to dying from MCC. Therefore, a death from any other cause represents a competing risk outcome. As such, we applied a competing risk analysis using the Fine-Gray model to investigate disease-specific mortality among patients within the Survive, Epidemiology, and End Results (SEER) database (1973-2016), with MCC tumor site as the primary variable of interest. With the results from this model, we calculated the 5-year cumulative mortality incidence (i.e., probability of mortality), for tumors at nine primary sites (ear, eyelid, lip, scalp/neck, other skin of the face, trunk, upper limbs, lower limbs and unknown primary site), while stratifying by stage at diagnosis. Of the 9407 MCC patients identified, 6105 (66%) had localized disease, 2397 (25.5%) had regional metastasis, and 705 (7.5%) had distant metastasis. Primary tumor site was predictive of cumulative mortality incidence (p < 0.0001), with which varied by stage at diagnosis. MCC involving the scalp/neck carried the highest cumulative mortality among localized tumors (24%), and regionally metastasized tumors (48.8%). In contrast, lower extremity metastasis, the lowest cumulative mortality (89.5%). Further, an unknown primary site was found to have a lower cumulative mortality incidence than some, but not all cutaneous tumor sites. Implications of these findings largely pertain to the prognostication of MCC outcomes. A staging guidelines may incorporate tumor primary stratification. Consideration of treatment escalation may be warranted for tumor sites with worse prognosis.

437 SMASH: Perceived stigma and social health in patients with chronic skin disease J. Tang1, A. Bruckner2, M. Chen3, DT. Woodley3, D. Keene4, M. Barriga1, K. Peoples2, R. John-Paradis3, J. Bordeaux3, 1 Dermatology, Massachusetts General Hospital, Boston, Massachusetts, United States, 2 University of Colorado, Children’s Hospital Colorado, Aurora, Colorado, United States, 3 University of Southern California, Los Angeles, California, United States, 4 Shriner’s Hospitals for Children Portland, Portland, Oregon, United States and 5 Phoenix Tissue Repair, Inc, Boston, Massachusetts, United States

SMASH is a valid, reliable measure that captures the emotional burden of chronic skin disease. Impression of Severity (PGIS) at two visits. All statistical analyses (correlations, Cronbach’s α, interclass correlation coefficient (ICC), Bland-Altman) were completed for 92 subjects. SMASH was completed by 50 subjects (AD—12; PS—12; acne—12; CL—7; AA—7). SMASH subscales exhibited strong internal consistency (Cronbach’s α = 0.815; Social Health α = 0.855), similar to the PSQ and the SQ (z = –0.914 and 0.899). SMASH subscales strongly correlated with PQ and SQ across all diagnoses (r = 0.902 and 0.858; p < 0.0001) and within diagnoses (r ≥ 0.681; p < 0.05). Acne subjects had the highest mean perceived stigma (2.51 ± 0.73) and the lowest mean social comfort (3.41 ± 0.85). A total of 92 subjects consented to Phase 2 (AD = 20; PS = 26; acne = 21; CL = 9; AA = 11). We expect good test-retest reliability (ICC = 0.700) for SMASH and strong correlation between patient-reported disease severity (r = 0.700; p < 0.05). We conclude that SMASH is a valid, reliable measure that captures the emotional burden of chronic skin disease. Utilization of SMASH in future studies would elucidate how social health is modified by treatment and disease course.

438 A phase 1/2 trial of PTR-01, a collagen 7 (C7) protein replacement therapy, in patients with recessive dystrophic epidermolysis bullosa (RDEB) J. Tang1, A. Bruckner2, M. Chen3, DT. Woodley3, D. Keene4, M. Barriga1, K. Peoples2, R. John-Paradis3, J. Bordeaux3, 1 Dermatology, Massachusetts General Hospital, Boston, Massachusetts, United States, 2 University of Colorado, Children’s Hospital Colorado, Aurora, Colorado, United States, 3 University of Southern California, Los Angeles, California, United States, 4 Shriner’s Hospitals for Children Portland, Portland, Oregon, United States and 5 Phoenix Tissue Repair, Inc, Boston, Massachusetts, United States

RDEB is a multisystem disorder affecting the skin, GI and GU tracts, eyes and immune system. Treatments in development primarily target cutaneous manifestations. PTR-01 is a human recombinant C7 intended to address both cutaneous and systemic manifestations of RDEB. In a multicenter Phase 1/2 study, we treated 10 adults with confirmed RDEB in 4 cohorts receiving 3 IV infusions of PTR-01 (0.1, 0.3, 1.0 or 3.0 mg/kg) or placebo every other week in a cross-over fashion (1:1). To allow tolerability and safety as the primary outcome. Secondary outcomes included pharmacokinetics (PK) and demonstration of C7 at the DEJ. Pharmacodynamic measures and wound healing were also assessed. All patients completed the study. There were no unexpected or drug-related serious adverse events. Milder moderate infusion-associated reactions occurred in four patients (2 each in Cohorts 3 and 4) and were managed with standard of care treatments (diphenhydramine, acetaminophen/ ibuprofen, glucocorticoids). Only two patients (1 each in Cohorts 2 and 3) developed anti-drug antibodies (transient/lower titer). PK showed dose-dependent increases. All Cohort 3 and 4 patients had increased C7 NC1 at the DEJ by direct immunofluorescence; largely absent baseline NC2 staining increased in all but one patient. Three patients had modest improvement in blisters within blisters, one of which also showed a transient increase in anchoring fibers. No consistent changes in wounds were seen. In this short-term study, PTR-01 has shown encouraging findings warranting further clinical trials. A longer Phase 2 study to assess efficacy is now enrolling.