

Supplemental Figure Legends

Supplemental Figure 1. Design of a Phase I Study of Plerixafor Treatment in Adults with WHIM Syndrome. P1 and P2 were receiving G-CSF before the study and had it discontinued 7 days before beginning plerixafor as marked by the 'X'. The last dose of IVIG prior to the 6 month plerixafor treatment period was given 45, 9, and 17 days before the first dose of plerixafor for P1, P2, and P3 respectively.

Supplemental Figure 2. Effect of Low-dose Long-term Plerixafor on Serum Immunoglobulin Levels in WHIM Patients. The last dose of IVIG prior to the 6 month plerixafor treatment period was given 45, 9, and 17 days before the first dose of plerixafor for P1, P2, and P3 respectively. IgG levels prior to treatment with IVIG was known to be 722 mg/dL for P2, but were unknown for P1 and P3 because they had been on treatment for many years prior to the study.

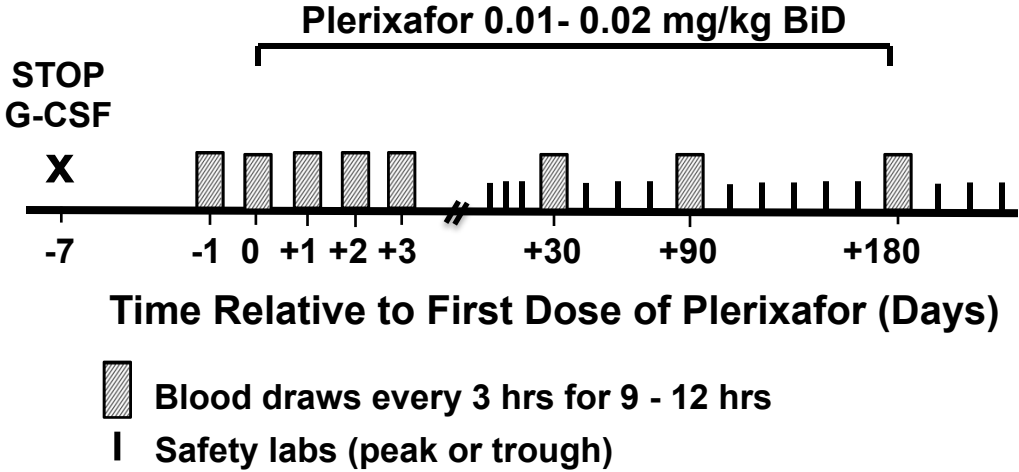
Supplemental Figure 3. Effect of Low-dose Long-term Plerixafor on Serum Chemistry Values in WHIM Patients. The factors indicated in each row were measured before the trial began (baseline, white bar) and at least twice per month during the trial (on therapy, black bar). The data are the mean +/- SEM of all values for each patient P1-P3 labeled on top of each column. Horizontal dotted lines demarcate the normal range for each factor in the NIH Laboratory of Clinical Pathology.

Supplemental Figure 4. No Effect of Low-dose Long-term Plerixafor on Blood Platelet or Erythrocyte Concentrations. The factors indicated at the top of each column were measured before the trial began (baseline, white bar) and at least twice per month during the trial (on therapy, black bar). The data are the mean +/- SEM of all values for each patient P1-P3 labelled on top of each column. Horizontal dotted lines demarcate the normal range for each factor in the NIH Laboratory of Clinical Pathology.

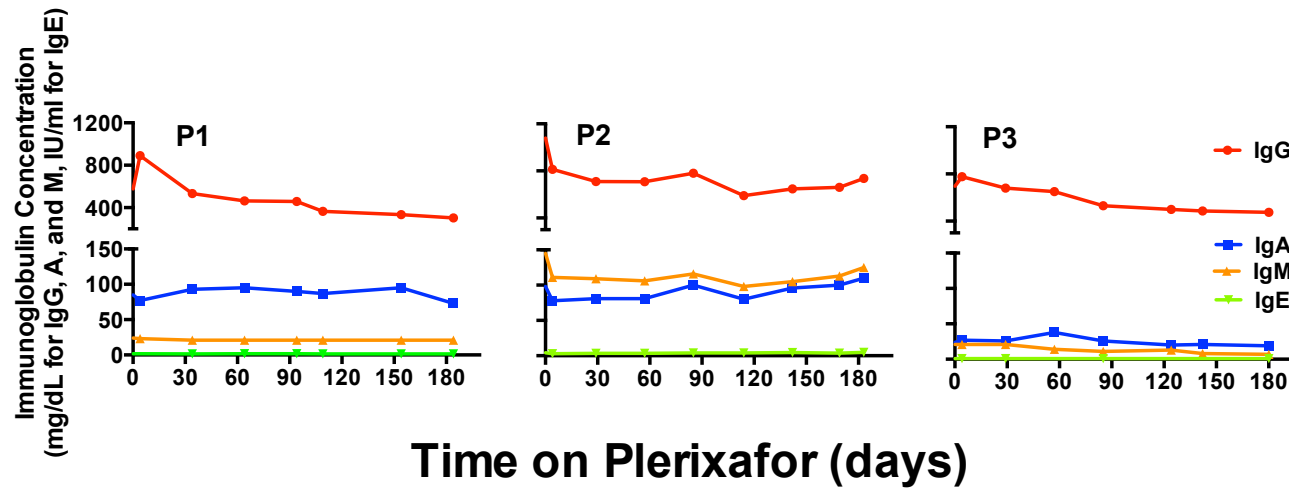
Supplemental Figure 5. Effect of Long-term Low-dose Plerixafor Therapy on Leukocyte Distribution in the Bone Marrow. Serial bone marrow aspirates were analyzed by FACS at the times indicated on the x-axis relative to beginning treatment (0, baseline before treatment started; 6W and 6M, 6 weeks and 6 months, respectively, after treatment started). Each point represents the percent of the indicated cells in total live bone marrow cells excluding red blood cells (granulocytes and CD14⁺ monocytes) or the percent of the indicated cells in the total lymphocyte gate (CD19⁺ B cells and CD3⁺ T cells). *, P<0.05 comparing baseline to 6 month values

Supplemental Figure 6. Effect of Long-term Low-dose Plerixafor Therapy on Bone Marrow Histology. Shown are representative bone marrow biopsy specimens stained with hematoxylin and eosin from immediately before treatment started (labeled Pre) and after 6 months of treatment (labeled Post) for each of the 3 patients (P1, P2, and P3). Images were obtained at 100X on an Olympus BX41 microscope with an Olympus DP72 camera.

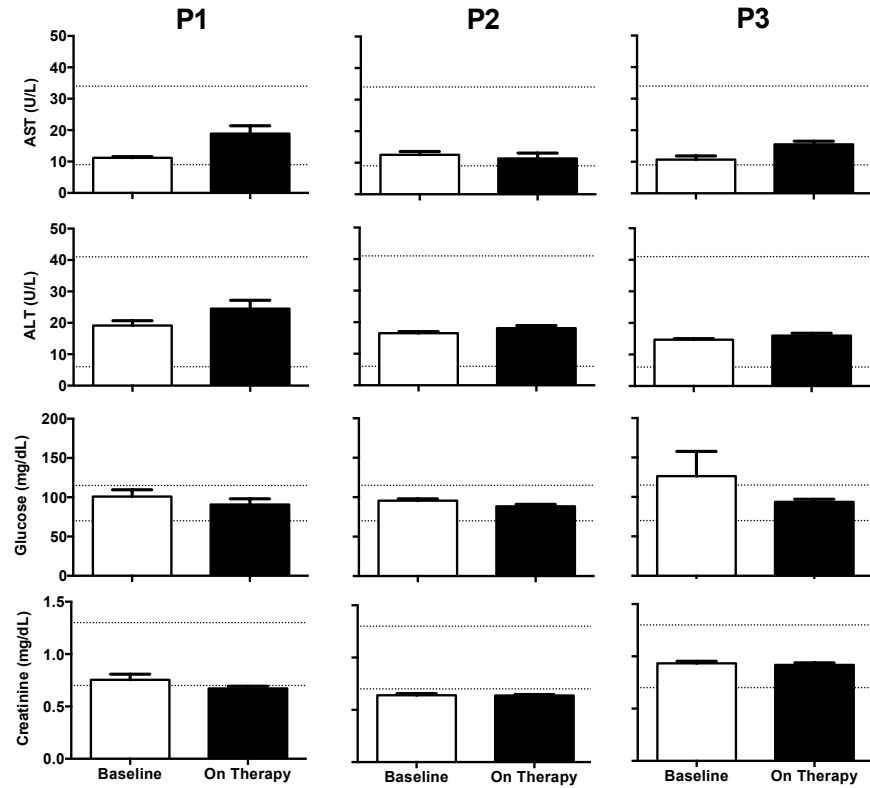
Supplemental Figure 1



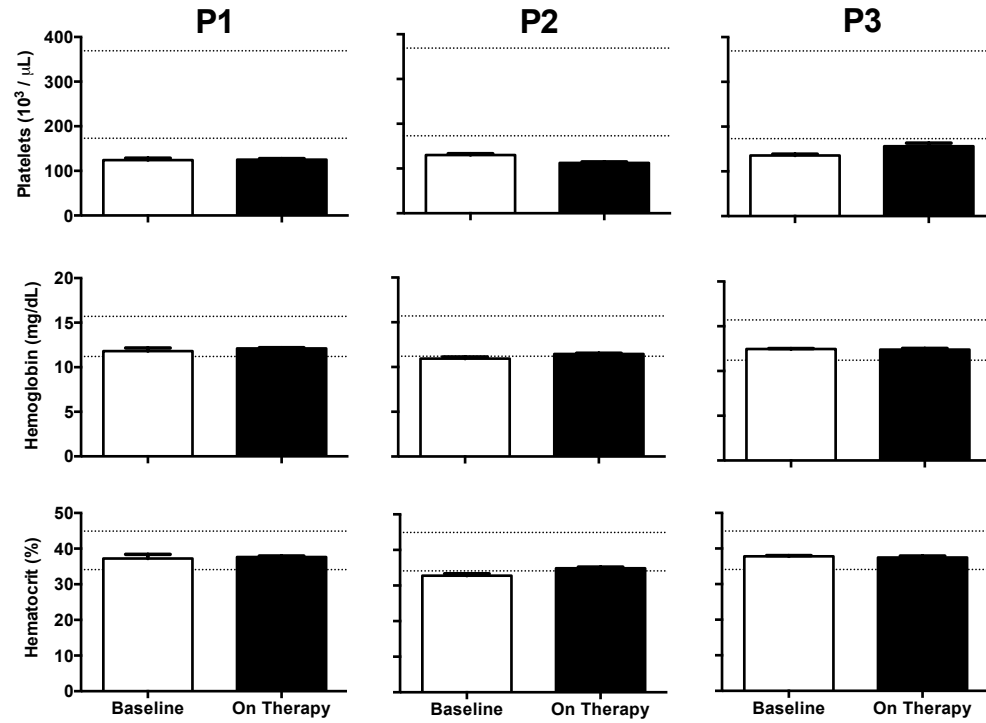
Supplemental Figure 2



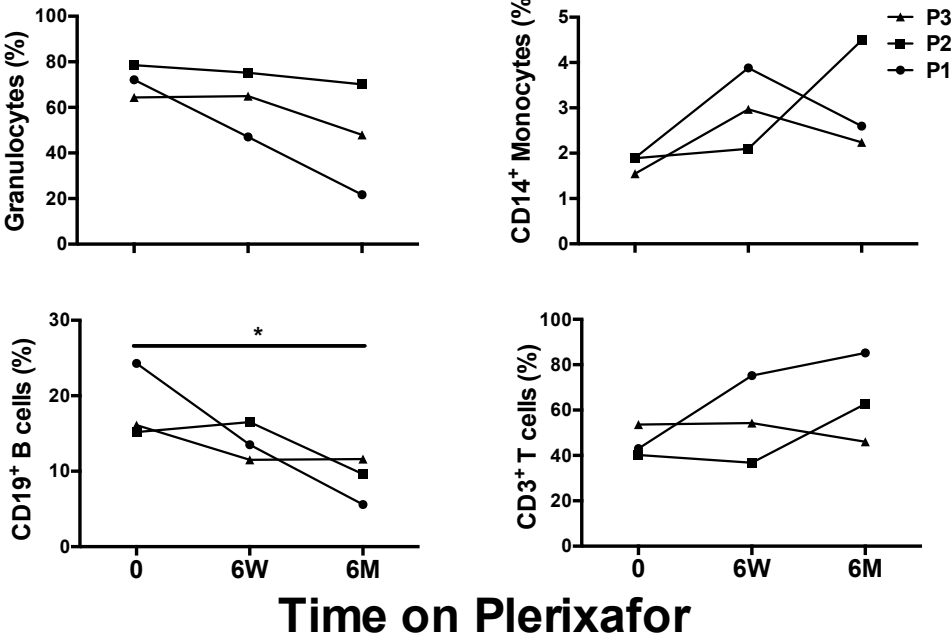
Supplemental Figure 3



Supplemental Figure 4

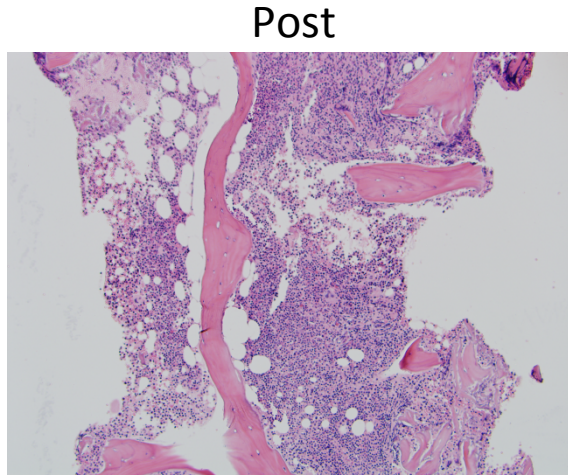
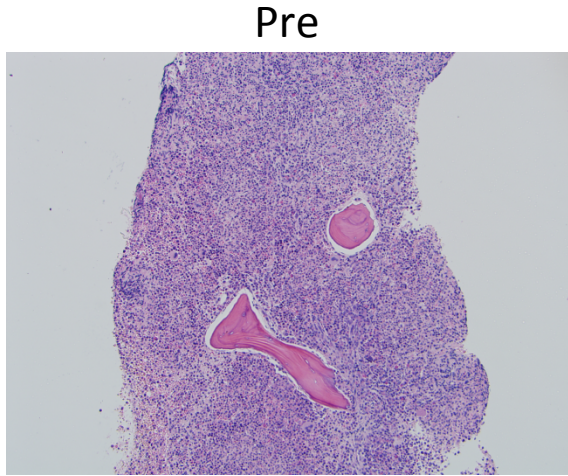


Supplemental Figure 5

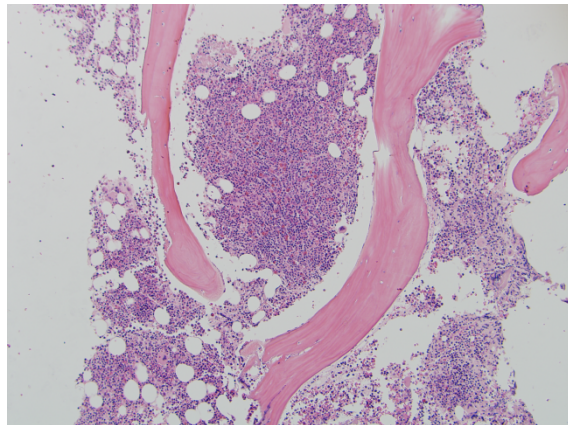
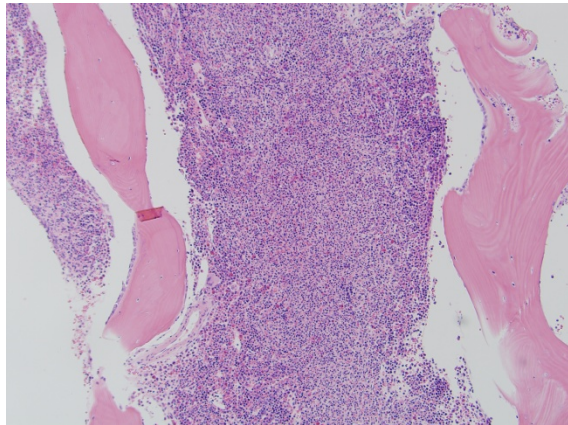


**Supplemental
Figure 6**

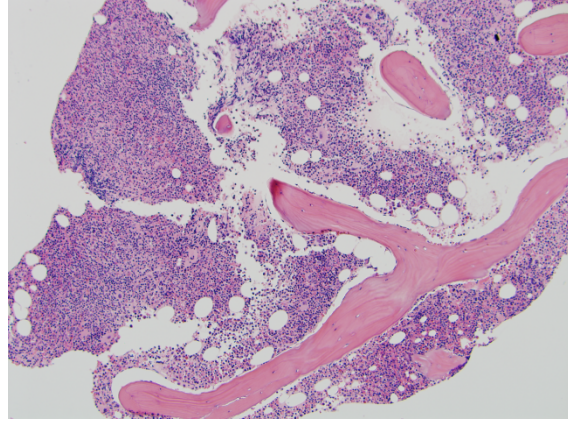
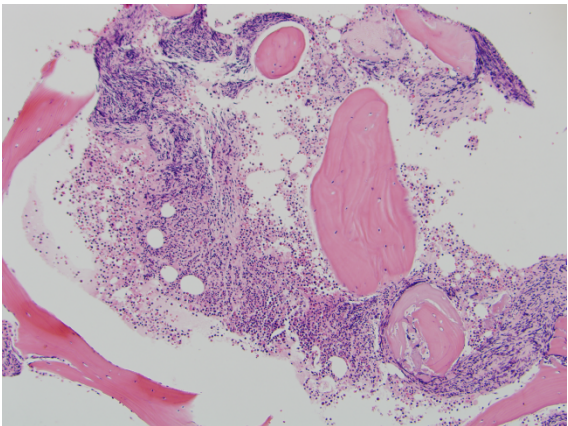
P1



P2



P3



Supplemental Table 1. Infections documented in three WHIM syndrome patients during, 3 years before and 1 year after a 6 month treatment protocol with plerixafor. The date of each infection is given in parentheses as the number of months relative to the start of chronic plerixafor treatment period and is shown graphically in Figure 5. AECB, acute exacerbation of chronic bronchitis, URI, upper respiratory infection, UTI, urinary tract infection

Patient	3 Year Period Before Treatment	6 Month Period During Treatment	1 Year Period After Treatment
P1	<ol style="list-style-type: none"> 1. Esophagitis (-2) 2. AECB (-6) 3. Vaginitis (-10) 4. AECB (-13) 5. Pneumonia (-19) 6. URI (-21) 7. AECB (-29) 8. AECB (-30) 9. AECB (-30) 10. Pneumonia/Lung abscess (-32) 		<ol style="list-style-type: none"> 1. AECB (+14) 2. Sinusitis, otitis media, AECB (+15) 3. AECB (+17)
Subtotal:	10	0	3
P2	<ol style="list-style-type: none"> 1. UTI and vaginal cuff infection (-1) 2. Endometritis (-2) 3. Gastroenteritis (-10) 4. Otitis media (-10) 5. Pharyngitis/Cervical Adenitis (-15) 6. Febrile Neutropenia (-34) 		<ol style="list-style-type: none"> 1. Dental abscess (+11) 2. Dental abscess (+14) 3. Dental abscess (+15)
Subtotal:	6	0	3
P3	<ol style="list-style-type: none"> 1. Pneumonia (-1) 2. Pneumonia (-2) 3. Bronchitis (-5) 4. Pneumonia (-6) 5. Bronchitis (-7) 6. Pneumonia (-13) 	<ol style="list-style-type: none"> 1. Pneumonia/aseptic meningitis (+0.25) 2. Localized dermatomal zoster (+6) 3. AECB (+9) 	<ol style="list-style-type: none"> 1. AECB (+12) 2. AECB (+14) 3. AECB (+15) 4. AECB (+17) 5. Pneumonia (+20)

	7. Pneumonia (-14) 8. Otitis Media, sinusitis and pneumonia (-14) 9. Pneumonia (-16) 10. Pneumonia (-17) 11. Bronchitis (-18) 12. Anitis (-20) 13. Pneumonia (-21) 14. Bronchitis (-22) 15. Pneumonia (-24) 16. Bronchitis (-25) 17. Pneumonia (-26) 18. Bronchitis (-27) 19. Pneumonia (-28) 20. Pyelonephritis (-35) 21. Pneumonia (-35)		
Subtotal	21	3	5
TOTAL	37	3	11
<i>#/patient-year</i>	4.1	2	3.7