Supplementary material to article by A. Vahlquist et al. "Recurrent Pyoderma Gangrenosum and Cystic Acne Associated with Leucocyte Adhesion Deficiency due to Novel Mutations in ITGB2: Successful Treatment with Infliximab and Adalimumab"

Table SI. Granulocyte cell surface expression of $\beta_2$ -integrins and Fcy-receptors,		
granulocyte migration, and granulocyte respiratory burst and phagocytosis tests		
performed when the patient was on lowest possible corticosteroid dose		

	Sept 2004 1 <sup>st</sup> visit	Sept 2004 2 <sup>nd</sup> visit	Reference*
Blood leucocyte count (× 10 <sup>9</sup> /l) Cell surface antigen expression (antibody clone) <sup>a</sup>	13.8	12.1	3.3-8.0
CD11b (Bear 1)	4.0/0.22	3.3/0.29	>95/>3.(
CD18 (MHM23)	49/0.72	41/0.65	>95/>3.0
CD16 (FcyRIII)	99/36.4	97/26.6	>99/>16
CD32 (FcyRII)	99/16.6	100/18.2	>99/>13
CD64 (FcyRI)	30/0.51	29/0.54	<22/<0.4
Random migration (µm/h)	Not determined	29	21-30
Chemotaxis (C5a, 10 <sup>-8</sup> mol/l) (µm/h)	Not determined	115	75-130
Chemotaxis (IL-8, 10 <sup>-9</sup> mol/l) (µm/h)	Not determined	87	85-130
Respiratory burst test (E. coli) (Index)**	* 241	184	>180
Respiratory burst test (PMA) (Index)**	422	268	>300
Phagocytosis test (E. coli) (Index)**	6,960	Not determined	>1,100

<sup>a</sup>Cell surface expression presented as relative number (%) and mean fluorescence of granulocytes labelled by the respective antibody. For methods see Appendix S1<sup>1</sup>.

\*Reference interval established at the laboratory.

\*\* Index = Positive granulocytes (%) × mean fluorescence