

## **Alveolar Macrophage Dysregulation in Hermansky-Pudlak Syndrome**

### **Type-1**

ONLINE DATA SUPPLEMENT

Farshid N. Rouhani, Mark L. Brantly, Thomas C. Markello, Amanda Helip-  
Wooley, Kevin O'Brien, Richard Hess, Marjan Huizing, William A. Gahl,  
and Bernadette R. Gochuico

**Supplemental Figure 1.** *High alveolar concentrations of cytokines and chemokines in subjects with HPS-1 pulmonary fibrosis.* Concentrations (pg/ml) of monocyte chemoattractant protein-1 (MCP-1), macrophage inflammatory protein-1 alpha (MIP-1a), granulocyte-macrophage colony-stimulating factor (GM-CSF), and macrophage colony-stimulating factor (M-CSF) were significantly higher in ELF from subjects with mild HPS-1 pulmonary fibrosis (HPSPF mild) compared to HPS-1 subjects without lung disease (HPS). Concentrations of MCP-1 and M-CSF were also significantly higher in subjects with moderate or severe HPS-1 pulmonary fibrosis (HPSPF severe) compared to HPS-1 subjects without lung disease.

Supplemental Figure 1.

