

**Supplemental Fig. 1. Longitudinal monitoring of plasma ATP levels.**

Plasma ATP levels were measured in two patients suffering from burn injuries on six or seven consecutive days using the HPLC method used to study adenylate levels in our PCAS patients. The dotted line indicates averaged ATP plasma levels of healthy controls (n=9). The study was approved by the Institutional Review Board of Juntendo University, Urayasu Hospital, Chiba, Japan, and informed written consent was obtained prior to blood draw.