Pyruvate dehydrogenase complex deficiency is linked to regulatory loop disorder in the αV138M variant of human pyruvate dehydrogenase

Matthew J. Whitley^{‡1}, Palaniappa Arjunan^{‡1}, Natalia S. Nemeria^{§1}, Lioubov G. Korotchkina[¶], Yun-Hee Park[¶], Mulchand S. Patel[¶], Frank Jordan[§], and William Furey^{‡†2}

From the [‡]Department of Pharmacology & Chemical Biology, University of Pittsburgh School of Medicine, Pittsburgh, PA 15261; [§]Department of Chemistry, Rutgers, the State University of New Jersey, Newark, NJ 07102; [¶]Department of Biochemistry, Jacobs School of Medicine and Biomedical Sciences, University at Buffalo, The State University of New York, Buffalo, NY 14203; [†]Biocrystallography Laboratory, Veterans Affairs Medical Center, Pittsburgh, PA 15240

Supplementary Figure S1

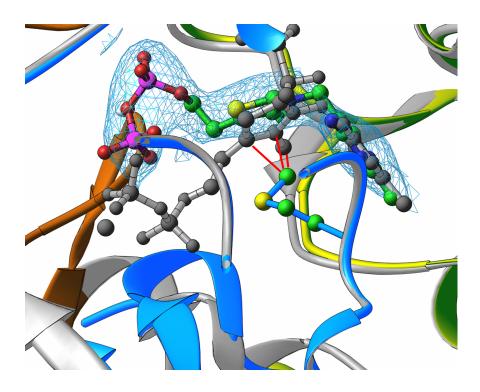


Fig. S1. An alternate orientation of Figure 3A in which the clashes between M138 and the C4, C4 α , and C5 atoms of ThDP in the WT-AcPhi structure are more readily apparent. The clashing atoms are connected by red lines.