



FIG. 3. Structure of odCPO/Hem13p. **A**, ribbon diagram. The chain is color ramped from blue (N terminus) to red (C terminus). Shown is a stereo view of odCPO/Hem13p with secondary structural elements and N/C termini labeled. **A** and Figs. 4–6 were made using PyMOL (available at www.pymol.org). **B**, amino acid sequence. The sequences of *S. cerevisiae* Hem13p and human odCPO are shown, with secondary structural elements of the Hem13p crystal structure above. Residues invariant across an alignment of 10 diverse odCPO sequences after alignment with ClustalW (55) are shown on a magenta background. The sequences used in the analysis were from *S. cerevisiae*, *Drosophila melanogaster*, *Homo sapiens*, *Aplysia californica*, *Mus musculus*, *Nicotiana tabacum*, *Synechocystis* sp. PCC6803, *E. coli*, *Ralstonia solanacearum*, and *Agrobacterium tumefaciens* strain C58. Residues that expose at least 10 Å² of accessible surface area to the active-site cavity of the form II structure are indicated with green dots. Residues that lose surface area upon dimer formation are indicated with blue squares. Black diamonds indicate positions of mutations identified in cases of coproporphyria (Table II).

collected using a rotating anode generator fitted with a chromium anode, appropriate optics, and a helium x-ray path (22). The single wavelength anomalous diffraction/solvent-flattened map (Fig. 2) was of sufficient clarity to allow building of an initial model that was subsequently refined to an R factor/ R_{free}

of 22.8/23.1% using 1.9-Å data collected from another form C crystal at a synchrotron (Table I). This refined model starts at Arg⁸ and ends at Arg²⁶⁵. A total of 219 ordered residues are included in the model, and three flexible loops and the 63 C-terminal residues have been omitted; the ordered residues are