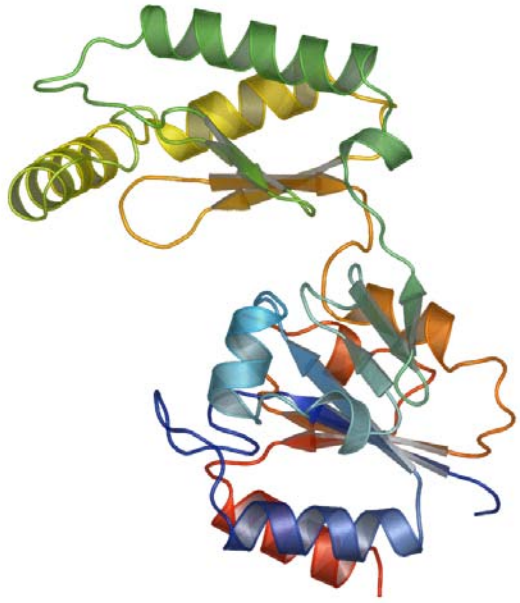
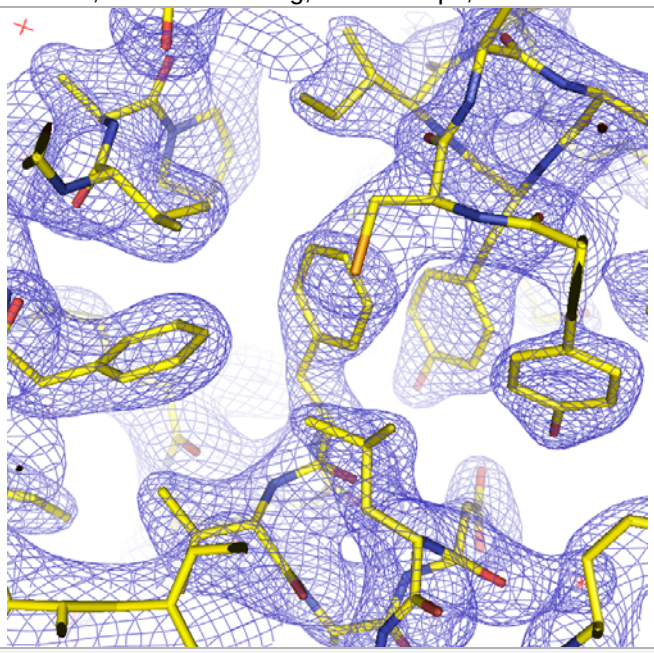
Center for Eukaryotic Structural Genomics

Protein Structure Initiative

Target ID	GO.36653	
Source Organism	*Homo sapiens*	
Target Name	BC008310	
PDB Entry	2AMY	Deposition: 10-Aug-2005
Function	phosphomannomutase 2 (PMM2) (FF/Refine: 2Q4R)	
Produced From	*E. coli* B834, pRARE-2, pVP-16	
Structure by X-ray	Resolution: 2.09 Å	R-value (R-free): 19.9% (27.2%)
	No. of Residues/ASU: 240 (245)	Complexes/ASU: 1
Data Collected At	Advanced Photon Source SBC 22-ID 13-Jun-2005	
Authors	C.A. Bingman, S.T.M. Allard, E. Bitto, G.E. Wesenberg, G.N. Phillips, Jr.	





Structural Features

The structure of phosphomannomutase-2 from *Homo sapiens* was determined by SeMet MAD at SER-CAT (APS 22-ID). This enzyme (EC 5.4.2.8) is involved in the synthesis of GDP-mannose and dolichol-phosphomannose required for a number of critical mannosyl transfer reactions. The structure reveals a two domain structure, with the larger, lower domain having a typical hydrolase fold. A DALI search reveals structural similarity to members of the haloacid dehalogenase (HAD) superfamily. PMM2 is known to be involved in a human disease state. Patients with carbohydrate-deficient glycoprotein syndrome type 1 (CDG1 or Jaeken's disease) show a deficiency in PMM activity. This inherited defect usually manifests itself as a severe neonatal disorder, with 20% mortality in the first year of life, due to infections, hepatic insufficiency and cardiomyopathy. On a molecular level, these defects are characterized by defective protein glycosylation.

Percent Identity with Nearest PDB Structure at Time Solved	16% (1NF2)
Pfam Cluster	Hydrolase_3, PMM
Sequence Cluster Size : Structures in PDB	104 NR at e<0.1

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