During N-linked glycosylation of proteins, oligosaccharide chains are assembled on the carrier molecule dolichyl pyrophosphate in the following order: 2 molecules of N-acetylglucosamine, 9 molecules of mannose, and 3 molecules of glucose. These 14-residue oligosaccharide cores are then transferred to asparagine residues on nascent polypeptide chains in the endoplasmic reticulum. As proteins progress through the Golgi apparatus, the oligosaccharide cores are modified by trimming and extension to generate a diverse array of glycosylated proteins. ALG7 encodes the dolichyl-P-dependent N-acetylglucosamine-1-P transferasethat catalyzes the first step in the synthesis of lipid-linked oligosaccharides: addition of the first N-acetylglucosamine to dolichyl phosphate on the cytosolic side of the endoplasmic reticulum. Alg7p is essential, and its activity is inhibited by tunicamycin. Expression of ALG7 is cell-cycle regulated, in coordination with ALG1 and ALG2. Upon alpha factor arrest, mutants with diminished Alg7p activity show only transient downregulation of CLN1 and CLN2 mRNAs, and therefore these alg7 mutants resume their progression through the cell cycle. An interesting hypomorphic alg7 mutant lost mitochondrial DNA. ALG7 is the most ancient and widely found of the ALG genes examined. An orthologous gene from an Archaeal speciescomplements the alg7 deletion. The human ortholog, DPAGT1, also complements a conditional deletion of ALG7. Mutations in DPAGT1 cause the congenital disorder of glycosylation CDG-Ij.