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. Alg6p, an alpha 1,3 glucosyltransferase, catalyzes the addition of the first glucose to the growing lipid-linked oligosaccharidein the lumen of the endoplasmic reticulum. Alg8pand Die2padd the second and third glucoses, respectively. In a hexokinase/glucokinase-deficient background, alg6 mutants fail to accumulate intracellular glucose, indicating that glucose accumulates by trimming of glucose from LLO's. Human ALG6rescues defective glycosylation in alg6 mutant yeast. Mutation of human ALG6 causes the congenital disorder of glycosylation CDG-Ic, and may exacerbate CDG-Ia.