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Carglumic Acid

National Cancer Institute

Source

National Cancer Institute. *Carglumic Acid*. NCI Thesaurus. Code C76884.

An orally active, synthetic structural analogue of N-acetylglutamate (NAG) and carbamoyl phosphate synthetase 1 (CPS 1) activator, with ammonia lowering activity. NAG, which is formed by the hepatic enzyme N-acetylglutamate synthase (NAGS), is an essential allosteric activator of the enzyme carbamoyl phosphate synthetase 1 (CPS 1). CPS 1 plays an essential role in the urea cycle and converts ammonia into urea. Upon oral administration, carglumic acid can replace NAG in NAGS deficient patients and activates CPS 1, which prevents hyperammonaemia.