

Title: Citrullinemia Type I *GeneReview* Scavenger Therapy

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Updated: January 2014

Note: The following information is provided by the authors listed above and has not been reviewed by *GeneReviews* staff.

Scavenger Therapy

Scavenger therapy utilizes alternative physiologic pathways that promote nitrogen excretion in the urine. Benzoate conjugates with glycine (containing 1 nitrogen) forming hippurate while phenylacetate conjugates with glutamine (containing 2 nitrogens) forming phenylacetylglutamine with both compounds excreted in the urine. Arginine supplementation in CTLN1 patients repletes the typical deficiency and allows continued excretion of citrulline in the urine. Scavenger therapy should be provided initially as a bolus run over 90 minutes followed by a repeat dose run over 24 hours.