

UDP-galactose transporter

MOGHE is defined as mild [malformation](#) of [cortical development](#) with [oligodendroglial hyperplasia](#) in [epilepsy](#). Approximately half of the patients with histopathologically confirmed MOGHE carry a brain somatic variant in the [SLC35A2](#) gene encoding a [UDP-galactose transporter](#). Previous research showed that D-[galactose](#) supplementation results in clinical improvement in patients with a congenital disorder of [glycosylation](#) due to germline variants in SLC35A2. Aledo-Serrano et al. aimed to evaluate the effects of D-galactose [supplementation](#) in patients with histopathologically confirmed MOGHE, with uncontrolled [seizures](#) or [cognitive impairment](#) and epileptiform activity at the [EEG](#) after [epilepsy surgery](#) (NCT04833322). Patients were orally supplemented with D-galactose for 6 months in doses up to 1.5 g/kg/day and monitored for seizure frequency including 24-h video-EEG recording, cognition and behavioral scores, i.e., WISC, BRIEF-2, SNAP-IV, and SCQ, and quality of life measures, before and 6 months after treatment. Global response was defined by > 50% improvement of seizure frequency and/or cognition and behavior (clinical global impression of “much improved” or better). Twelve patients (aged 5-28 years) were included from three different centers. Neurosurgical tissue samples were available in all patients and revealed a brain somatic variant in SLC35A2 in six patients (non-present in the blood). After 6 months of supplementation, D-galactose was well tolerated with just two patients presenting abdominal discomfort, solved after dose spacing or reduction. There was a 50% reduction or higher of seizure frequency in 3/6 patients, with an improvement at EEG in 2/5 patients. One patient became seizure-free. An improvement of cognitive/behavioral features encompassing impulsivity (mean SNAP-IV - 3.19 [- 0.84; - 5.6]), social communication (mean SCQ - 2.08 [- 0.63; - 4.90]), and executive function (BRIEF-2 inhibit - 5.2 [- 1.23; - 9.2]) was observed. Global responder rate was 9/12 (6/6 in SLC35A2-positive). The results suggest that supplementation with D-galactose in patients with MOGHE is safe and well tolerated and, although the [efficacy](#) data warrant larger studies, it might build a rationale for [precision medicine](#) after [epilepsy surgery](#) ¹⁾.

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Aledo-Serrano Á, Valls-Carbó A, Fenger CD, Groeppel G, Hartlieb T, Pascual I, Herraes E, Cabal B, García-Morales I, Toledano R, Budke M, Beltran-Corbellini Á, Baldassari S, Coras R, Kobow K, Herrera DM, Del Barrio A, Dahl HA, Del Pino I, Baulac S, Blumcke I, Møller RS, Gil-Nagel A. D-galactose Supplementation for the Treatment of Mild Malformation of Cortical Development with Oligodendroglial Hyperplasia in Epilepsy (MOGHE): A Pilot Trial of Precision Medicine After Epilepsy Surgery. *Neurotherapeutics*. 2023 Jun 6. doi: 10.1007/s13311-023-01395-z. Epub ahead of print. PMID: 37278968.

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