In vivo Animal Models

Gaucher Disease

Conduritol-B-Epoxide (CBE) treatment

Gaucher disease (GD) is the most common lysosomal storage disorder. The main pathological hallmark is the intracellular accumulation of glucosylceramide and glucosphingosine as a result of reduced glucocerebrosidase (GCase) enzyme activity due to mutations in the β-glucocerebrosidase gene (GBA). Conduritol-B-epoxide (CBE) is a specific inhibitor of GCase activity and can thus be used to induce Gaucher disease *in vivo*. C57Bl/6 mice were intraperitoneally injected with 100 mg/kg CBE on 9 consecutive days and brains analyzed for neuroinflammation.

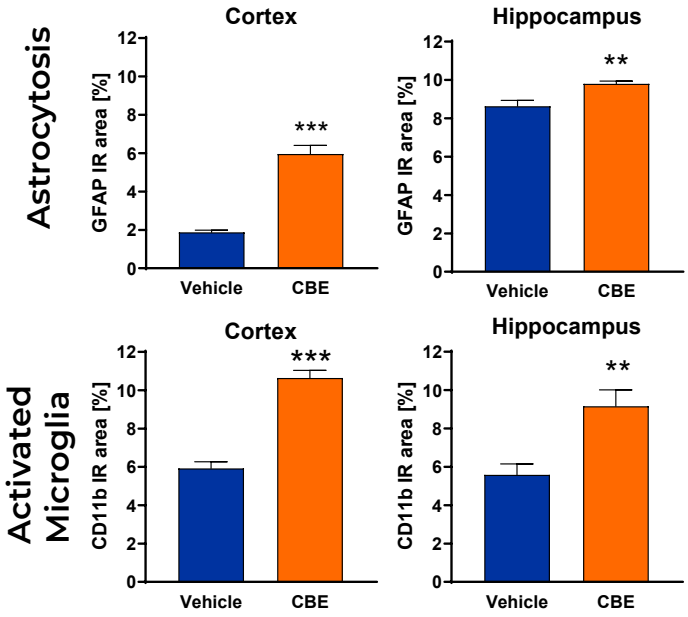

Figure 1. Quantification of cortical and hippocampal astrocytosis and activated microglia of CBE-treated mice. GFAP (A,B) and CD11b (C,D) immunoreactive (IR) area in percent. n = 9; unpaired t-test; Mean + SEM. **p<0.01; ***p<0.001.

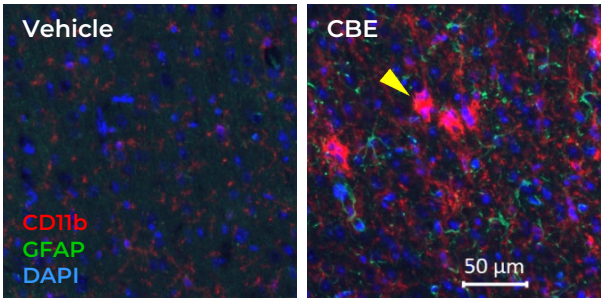

Figure 2. Representative images of cortical tissue of vehicle- or CBE-treated mice. Note the occurrence of extremely enlarged microglia (arrowhead) and increased GFAP levels in CBE-treated mice.

Grabowski GA, Osiecki-Newman K, Dinur T, Fabbro D, Legler G, Gatt S, Desnick RJ. Human acid beta-glucosidase. Use of conduritol B epoxide derivatives to investigate the catalytically active normal and Gaucher disease enzymes. J Biol Chem. 1986 Jun 25;261(18):8263-9.

QPS Austria
Parkring 12, 8074 Grambach, Austria
Email office-austria@qps.com
Website www.qpsneuro.com
Tel +43 316 258 111

QPS LLC
3 Innovation Way, Suite 240
Newark, DE 19711, USA
Email info@qps.com
Website www.qps.com