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In vivo Animal Models

Gaucher Disease

Conduritol-B-Epoxide (CBE) treatment

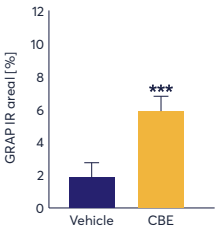
Gaucher disease (GD) is the most common lysosomal storage disorder. The main pathological hallmark is the intracellular accumulation of glucosylceramide and glucosphingosine as a result of reduced glucocerebrosidase (GCase) enzyme activity due to mutations in the β-glucocerebrosidase gene (GBA). Conduritol-B-epoxide (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.

Astrocytosis

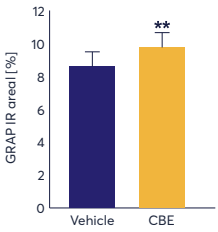
Cortex

Figure 1: A



Hippocampus

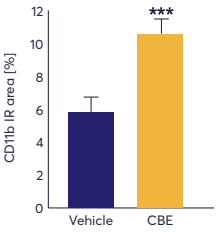
Figure 1: B



Activated Microglia

Cortex

Figure 1: C



Hippocampus

Figure 1: D

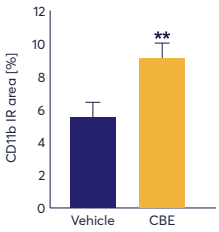
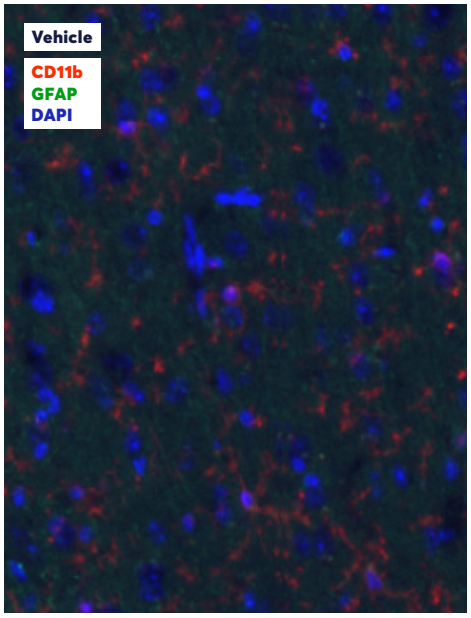
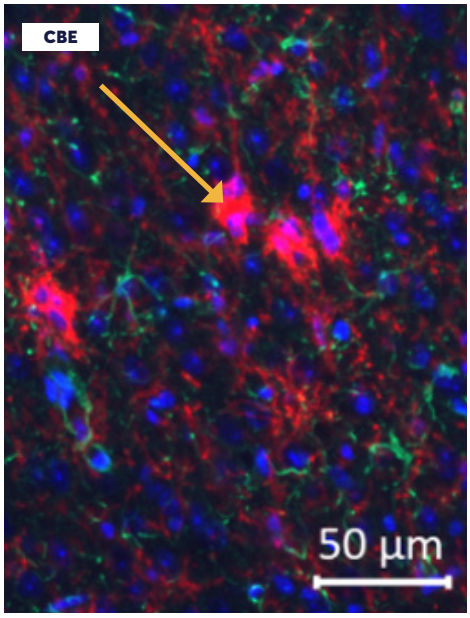


Figure 2:
Representative images of cortical tissue of vehicle- or CBE- treated mice. Note the occurrence of extremely enlarged microglia (arrowhead) and increased GFAP levels in CBE-treated mice.

Figure 2:





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.

Scantox
Discovery

Scantox Group, HQ
Hestehavevej 36A, Ejby
DK – 4623 Lille Skensved
clientservice@scantox.com
www.scantox.com
+45 5686 1500

126-38-GaucherCBE-EN (04/2024)