

Figure S1. "Immune cell infiltration in Ccdc39prh/prh brain"

- (A) CD68 staining in *Ccdc39<sup>wt/wt</sup>* and *Ccdc39<sup>prh/prh</sup>* at P5. More number of CD68 positive macrophages are seen in white matter and striatum in *Ccdc39<sup>prh/prh</sup>* mutant rats compared to wild type rats at P5 (scale bars = 500, 200, 200 µm from left to right, mean  $\pm$  SD, p = 0.046, n = 3 in each group) (WM = white matter, ST = striatum).
- (B) MCP-1 expressions in P11 Ccdc39<sup>wt/wt</sup>. Double staining of Monocyte chemoattractant protein (MCP) -1 with either NeuN, CD68, or Isolectin B4 (IB4) shows that in wild type rats, MCP-1 is weakly expressed in NeuN-positive neurons. In wild type rats, CD68 positive macrophages are not migrated on the surface of the brain, and therefore MCP-1 is observed on brain surface. MCP-1 is barely expressed in endothelial cells labeled with IB4 in deep brain parenchyma (scale bars = 100, 100, 50 µm from top to bottom).



Figure S2 "Neural cell differentiation in Ccdc39<sup>prh/prh</sup> rats"

- (A) Myelination in *Ccdc39<sup>wt/wt</sup>* and *Ccdc39<sup>prh/prh</sup>* at P5. At P5, myelination as seen at P11 is not yet started even in wild type rats. When quantifying the number of Myelin basic protein (MBP)-positive mature oligodendrocytes, the number of mature oligodendrocytes in *Ccdc39<sup>prh/prh</sup>* mutant rats was comparable to wild type rats (scale bars = 200 µm, mean ± SD, p = 0.18, n = 3 in each group).
- (B) Neurofilament in *Ccdc39<sup>wt/wt</sup>* and *Ccdc39<sup>prh/prh</sup>* at P5. There was a tendency of reduced neurofilament formation stained with neurofilament-H subunit (NF-H) in in *Ccdc39<sup>prh/prh</sup>* mutant rats at P5 compared to wild types, although not statistically significant (scale bars = 200 µm, mean ± SD, p = 0.11, n = 3 in each group).