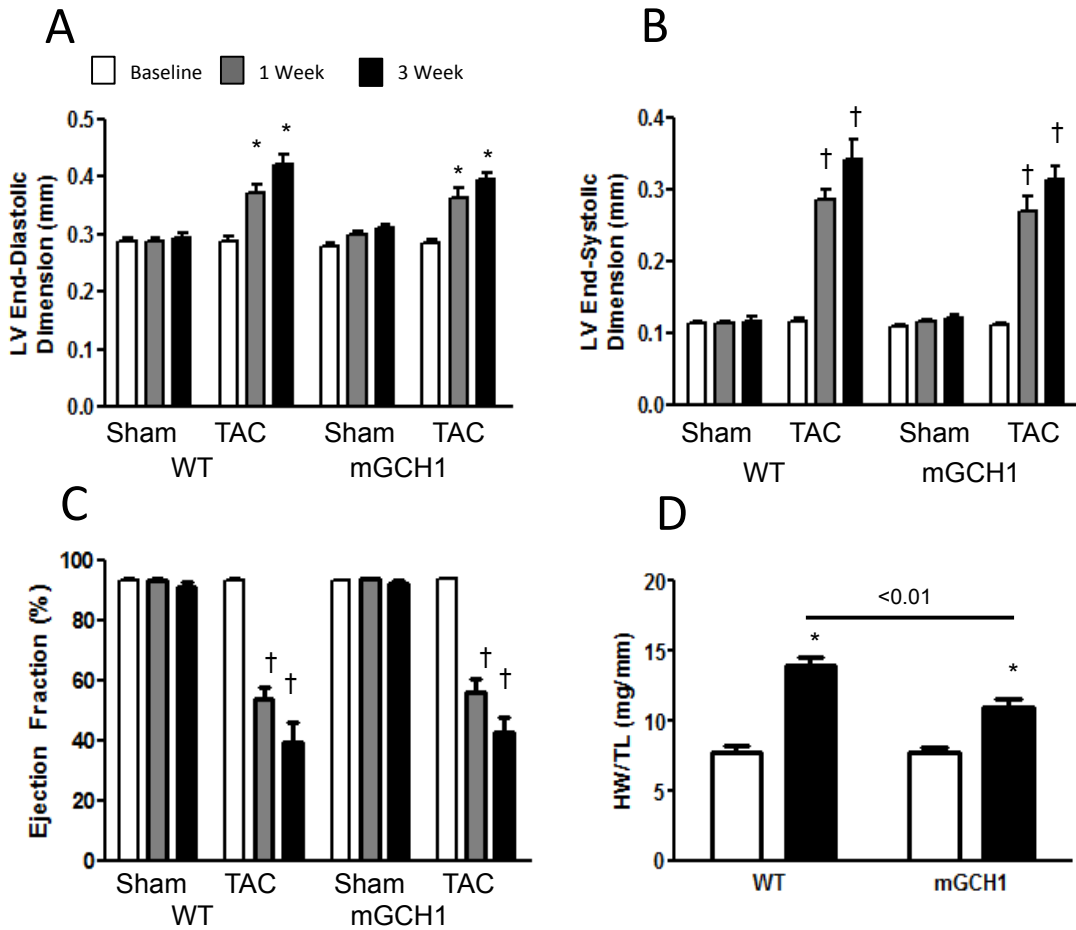


## **SUPPLEMENTAL MATERIAL**

**Figure S1:** Echocardiographic measurements of chamber (A, B) end-diastolic and end-systolic dimension, and calculated total ejection fraction (C) at each time point in WT and mGCH1 mice in both Sham operated and TAC operated groups. Volumes increased and EF declined similarly in both groups with TAC. Within group 1-way ANOVA post-hoc comparisons (Tukey) results: \*  $p < 0.005$  vs baseline; †  $p < 0.0001$  vs baseline. The responses in WT and mGCH1 groups were not significantly different. (D) Heart weight/tibia length ratio for both WT and mGCH1 groups in sham and TAC groups. \*  $p < 0.001$  versus Sham. These data analyzed by Kruskal-Wallis test due their non-normal distribution.



**Figure S2:** Echocardiographic measurements of chamber end-systolic volume and calculated ejection fraction at each time point in WT and mGCH1 mice in TAC groups administered control soft diet, or diet supplemented with BH4. BH4 treatment suppressed chamber remodeling (less rise in end-systolic dimension) and increased ejection fraction. Within group 1-way ANOVA post-hoc comparisons (Tukey) results: \*  $p < 0.005$  vs baseline; †  $p < 0.05$  vs baseline, #  $p = 0.06$  vs baseline. §  $p < 0.05$ , ¶  $p < 0.01$  vs control within same genotype in the same week (n=6-9).

