



Figure S1. IFG increases the lysosomal content of L444P GCCase in mouse liver. Twenty four-week old male L444P GCCase mice were administered IFG tartrate (20 mg/kg per day, equivalent to 10 mg/kg free base) *ad libitum* in drinking water for 24 weeks. Subsequently, liver tissue was isolated and subjected to lysosomal fractionation according to the manufacturer's instructions (Lysosomal Isolation Kit; Pierce, Rockford, IL). *Panel A.* β -Hexosaminidase activity was measured in each of the three isolated fractions as described previously (Rigat and Mahuran 2009) and is presented as the percent of total activity from all three fractions combined. The first fraction showed the highest level of β -hexosaminidase activity, indicating an enrichment of lysosomes compared to the other two fractions. *Panel B.* Western blotting of all three fractions for GCCase, the Golgi marker formiminotransferase cyclodeaminase (FTCD), and the β -actin loading control. As seen with β -hexosaminidase activity, the first fraction showed the highest level of GCCase, and the lowest level of FTCD, again indicating lysosomal enrichment. Importantly, GCCase levels were elevated in the lysosomal fractions isolated from livers of L444P GCCase mice administered IFG, confirming that IFG increases the absolute quantity and lysosomal localization of the mature 69 kD form of GCCase. These data are representative of two independent measurements.