Cystic Fibrosis (CF) is a recessive genetic disorder caused by mutations on the CF transmembrane conductance regulator (cfr) gene, which encodes for the CFTR protein. The CFTR protein plays a critical role in regulating chloride and bicarbonate passage into and out of cell membranes. Those living with CF suffer a wide range of symptoms, including gastrointestinal (GI) problems, pulmonary infections, delayed development, coughing, and fatigue. CF currently has no cure, but treatments have been developed to increase the quality of life and life expectancy of those with the condition. While pulmonary disease is the leading cause of death in adult patients, intestinal disease claims the early morbidity and mortality in those with CF. The intestinal disease can be characterized by small intestinal bacterial overgrowth, large intestine dysbiosis, and intestinal inflammation and obstruction. CF has long been considered an epithelial disease.

However, previous research has shown that the innate immune system is deeply affected by CF. The role of innate immune cells in CF intestinal disease is not very well understood. We hypothesize that innate immune cells play a role in the GI symptoms associated with CF. Mice from 4 different lines of CFTR-knockout (KO) mice, including myeloid-CFTR-KO (Mye-CF) mice, neutrophil-only CFTR-KO (Neu-CF) mice, macrophage/monocyte-only CFTR-KO (Mac-CF), and Pan-CFTR-KO (Pan-CF) mice. The three lineage-specific lines of mice and their WT siblings were given an oral gavage of carmine red dye, and each mouse’s intestinal transit time was calculated. Intestinal transit times are a good indicator of the severity of the symptoms of CF. Results show that the differences in transit times were not statistically significant in the Neu-CF and Mac-CF mice, but the transit times in the Mye-CF were marginally significant. Investigation of Pan-CF mice is still ongoing.

This research will provide data on the relationship between the CF defect in immune cells and the gastrointestinal symptoms most associated with CF.