We recently collected, deidentified, and organized a supplementary dataset including more than 150 lung clearance scans, performed as part of previous research studies, to augment efforts to develop and validate our organ level model of mucus clearance and liquid absorption in the lungs. Though the data being gathered prospectively as part of the UO1 award still forms the primary basis for our model and is the only dataset that includes matched cell and organ level data, the supplementary data set will allow for more robust testing of specific model elements under a wider range of conditions. It includes some particularly useful subsets such as repeated studies with individual subjects and studies of response to different therapies. The dataset is now in use by our primary organ level model developer, Monica Shapiro.

Primary human nasal epithelial (HNE) cells are a key source of data for our cell level model which will become a key component of our multiscale model. Our research plan involves collecting and extensively evaluating cell level physiology from HNE cultures from cystic fibrosis (CF) patients, single mutation carrier parents of these patients, and healthy controls. A good number of these assessments have already been performed and we are using this available data to compare cell level physiology in HNE cultures and human bronchial epithelial (HBE) cell cultures. HBE cultures have served as the primary model for therapeutics development in CF which lacks a readily available animal model. In our center HBE cultures are only available from lung explants and thus paired comparisons are generally not feasible. However, general comparisons of HNE and HBE physiology will allow us to better understand how HNE physiology (cell scale) will predict lung physiology (organ scale). This work is being performed by our primary cell level model developer Florencio Serrano-Castillo along with co-investigator Dr. Carol Bertrand.

We have modified our IRB to allow for re-collection of HNE cells from subjects where culturing was unsuccessful or where studies provided poor quality data. The nasal cell sampling has been well tolerated by subjects enrolled in the study and this modification will help us to ensure we obtain the most complete dataset possible for generating our models.

We have fully enrolled our control group and anticipate that data collection from that group will be complete by the end of 2018. This will be the first cohort within the study with complete cell and organ level data. The dataset now includes full or partial data from 16 healthy controls, 14 cystic fibrosis patients, and 6 parent single-mutation carriers. We have implemented a system for rolling quality assurance review of our main combined datasheet. This review is being conducted by Mr. Serrano-Castillo and Dr. Corcoran using source documents from the bench or clinical studies. The goal is to ensure continuity of experimental technique, full traceability of data, and rapid availability of a clean dataset for modeling applications.