Monitoring of ion transport in CF and control cells and tissue
- Ussing chamber analyses
- Transepithelial chloride conductance (TECC) using robots that calculate changes in conductance or equivalent current in high-throughput formats

Measurements of mucociliary transport and fluid flux
- μOCT imaging of freshly harvested trachea with or without further experimental exposures ex vivo
- Quantitation of organoid swelling derived from airway cells, intestinal organoids, and iPS-derived spermatids

Patch clamp and single-channel analyses to study changes in CFTR channel gating and regulation
- Patch clamp unitary conductance tracings to monitor open channel probability and test conductance or equivalent current in high-throughput formats
- Macropatch of cells or excised tissues or fluorescent dye-based halide efflux methods to evaluate CFTR activity in cells grown on coverslips (SPQ assays) or isolated from excised tissues

CF Clinical & Translational Assay Core
Steven Rowe, MD, MSPH and Amit Gaggar, MD, PhD

Designs and conducts in vivo measurements of CFTR activity in humans
- Nasal potential difference (NPD)
- PD measurements of the lower airway and sinus tract (by the endoscopic approach)
- Sweat chloride analysis / sweat rate
- Rectal intestinal current measurements

Conducts cardinal measures of airway epithelial cell function in vivo
- μOCT imaging for in vivo use by endoscopic probes
- Whole lung MCC by TC99 clearance approach
- Mucus rheology and solid content

Supports the execution of CF clinical studies
- Clinical trial design and regulatory support
- Collection and storage of biospecimens
- Supports key clinical outcome measures in infants, children, and adults with CF (nutritional outcomes, spirometry, lung clearance index (LCI), and infant PFTs)

CF Rat Models Core
Susan E. Birket, PharmD, PhD

Breed, genotypes, and tissues
- CFTR+/-, hG551D, and G542X rats, in addition to maintenance of WT Sprague Dawley rats

CFTR physiological outcome measures (NPD, sweat secretion)
- Assays of lung structure and function (micro-CT imaging, Fluixent)
- Collection and banking of biospecimens (survival bronchoscopic BAL, blood, tissues)

Other diseases related to CFTR defects studied in UAB CF cores
- Asthma

Cell Model and Evaluation Core
George Solomon, MD and Bradford Woodworth, MD

Procures, grows, and distributes well-differentiated primary human airway epithelial cells from CF and non-CF donors

Conducts functional anatomic imaging of airway epithelia by μOCT in vitro and ex vivo
- Well-differentiated primary epithelial cells (of human or non-human origin)
- Intact full-thickness trachea or mainstem bronchi of human origin

Performs measures of CFTR activity and expression in primary cells
- Ussing chamber analyses, high-throughput evolution by equivalent and transepithelial conductance (G0) and equivalent current (IEQ)
- Western blot, digital mRNA analysis utilizing primary cells on permeable supports

CF Animal and Preclinical Models Core
David M. Bedwell, PhD

Breed, genotypes, and tissues
- CFTR knockout mice and CF mice harboring clinically relevant mutations
- Ctrftr+/+; Ctrftr+/-.mCftrF508delEssential, CtrftrG551D; Ctrftr and others

Generates and procures relevant CF animal models
- Includes analysis and procurement of pig, ferret (and very recently, CF rabbit tissues), and maintains a colony of ferrets to evaluate acquired CFTR dysfunction and therapeutic approaches

Conducts endpoint measures to assess CFTR Function, epithelial physiology, preclinical endpoints, and biospecimen analysis in CF animal models
- Nasal and lower airway potential difference
- Short circuit current (Isc) measurements of excised trachea and intestine
- Measurement of lung function (Flexivent)
- In vivo and ex vivo μOCT imaging
- 6-Voxel resolution computed tomography
- Abdominal ultrasound
- Cough monitoring
- Glundar CFTR assay
- Electrocporation-mediated gene manipulation
- Miniaturized bronchoscopy for longitudinal lung sampling
- Anesthesia, physiologic monitoring, intubation for exposure and assessment procedures

Other diseases related to CFTR defects studied in UAB CF cores
- COPD (chronic obstructive pulmonary disease)
- APBA (allergic bronchopulmonary aspergillosis)
- IB (idiopathic bronchiolitis)
- CRS (chronic rhinosinusitis)
- Primary Ciliary Dyskinesia
- Asthma

The Gregory Fleming Jams Cystic Fibrosis (CF) Research Center was established in 1981 at UAB as a multidisciplinary Center to study CF basic research and therapy.

Also in 1981, the CF Foundation began an initiative for funding CF research known as the Research Development Program (RDP). The UAB Center was the first to receive RDP support from the Foundation, which now sponsors 10 such Centers in the United States. The Center has since maintained continuous CF Foundation and NIH funding, which has been supplemented by University Wide Interdisciplinary Research Center (UWIRC) and other important funding from the University and the State of Alabama. Together, this support has fostered the Center’s sustained legacy of advancing scientific success in CF, which was recognized through the recent renewal of fund NIDDK P30 funding (1 of only 3 Centers selected nationwide) and RDP funding. Multidisciplinary collaborations across UAB and beyond—cultivated in part by the unique and cutting-edge facilities, techniques, and resources offered through the Center’s 6 Cores—have been fundamental in driving these achievements and are a defining feature of our Center.

The CF Care Center located at the Children’s Hospital of Alabama and UAB Hospital provides state-of-the-art care for approximately 500 CF patients, generating additional partnerships across departments and specialties.

GREGORY FLEMING JAMES CYSTIC FIBROSIS RESEARCH CENTER
Director: Steven M. Rowe, MD, MSPH
https://www.uab.edu/medicine/cysticfibrosis/