Alpha-1 Antitrypsin Deficiency (AATD) iPSC Biorepository

Welcome to the CReM’s AATD iPSC Biorepository. Through this website, you can access iPSC lines generated by reprogramming somatic cells (blood or fibroblasts) obtained with informed consent from patients with a variety of SERPINA1 genotypes and known clinical phenotypes such as fibrotic liver disease or emphysema. Many of these lines are associated with detailed clinical data maintained in an IRB-approved REDCap database. AATD iPSCs are archived as frozen, shareable vials housed in the Center for Regenerative Medicine (CReM) at Boston University and Boston Medical Center. Please access the searchable catalog of these iPSC lines, where you can click on the “View all AATD iPSC lines” button and where information on how to request the lines from our iPSC Core facility is also detailed.

Our AATD biorepository includes iPSC lines from patients with the following SERPINA1 genotypes:

- PiZZ
- PiSZ
- PiMZ (gene corrected from PiZZ)
- PiMM (gene corrected from PiZZ)
- PiFZ
- PiZM_{Heerlen}
- PiQoBolton-QoBolton (“PiBB”)  

We have previously differentiated many lines from this repository and characterized lung and liver cells derived from them. Please click here for a manuscript characterizing lines in the repository.

Dilated ER in PiZZ and PiMZ iPSC-derived hepatocytes.

We thank the Alpha-1 Foundation and The Alpha-1 Project (TAP) for funds that made the launch of this repository possible, the NIH/NHLBI and NIH/NCATS for funds that have supported the maintenance and distribution of our iPSC lines, and most importantly the patient volunteers who generously donated the samples from which these iPSC lines were derived.