

Center For Cell Studies

Director: Horace M. DeLisser, M.D.

Abstract Pulmonary arterial hypertension (PAH) is an under-diagnosed disease for which there is no cure. At a structural level, remodeling of pulmonary arteries (PA) in severe PAH may result from selective death as well as proliferation of sub-sets of endothelial cells (ECs), hypertrophy and hyperplasia of vascular smooth muscle cells (SMCs), expansion of the adventitial fibroblast layer, (together with its associated vasa vasorum via neo-angiogenesis), recruitment of circulating inflammatory and EC precursors to the pulmonary vascular wall, and simultaneous catabolism and synthesis of specific components of the extracellular matrix (ECM). Despite the increase in knowledge concerning the etiology of idiopathic PAH (IPAH), (e.g. the discovery of BMP type II receptor mutations in familial and sporadic forms of IPAH), a more comprehensive characterization of normal and PAH cells and fluids is needed in order to promote development of new diagnostics and therapies.

To help accomplish this mission, the Penn CMREF PAH Cell Center Core proposes to:

- (1) Acquire control and PAH tissues and fluids;
- (2) Isolate control and PAH lung vascular, circulating cells and fluids;
- (3) Phenotype control and PAH lung vascular, circulating cells and fluids;
- (4) Test, Preserve & Bank Cells; (5) Distribute cells to the CMREF/PAH network;
- (6) Define an PAH plasma proteome and PAH cell signalome;
- (7) Prepare material for network partners and interact with other Investigators;
- (8) Generate, coordinate and manage cell- and fluid-derived data.

The schematic below summarizes our activities:

