

Cystic Fibrosis Research News

Title:

Comparative meta-analysis of cystic fibrosis cell models suggests partial endothelial-to-mesenchymal transition

Lay Title:

Bio-informatic analyses clarify how blood vessel malfunction may happen in people with cystic fibrosis

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What was your research question?

We investigated whether a process leading to the malfunction of endothelial cells (ECs), the cells lining the inside of blood vessels, was involved in cystic fibrosis (CF) and how this may contribute to different aspects of the disease, such as diabetes, cardiovascular-, lung- and liver-associated complications.

Why is this important?

ECs are important in several biological processes including the regulation of blood pressure, inflammation and blood clotting. In healthy persons these processes are tightly regulated. However, when deregulated as in subset of people with CF, it can lead to harmful cardiovascular complications where ECs have a pivotal function. However, the “endothelial conversion”, a phenomenon that contributes to vascular malfunctions, has never been investigated in people with CF. Since the life expectancy of people with CF is increasing due



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to improving health care, the risk associated with vascular malfunction during aging is likely to emerge, hence urging to study ECs in CF.

What did you do?

We combined the results of several research studies on human ECs with CF (grown artificially) and analyzed whether these cells present the typical characteristics of an endothelial conversion. Investigations using other cell types with CF (namely the epithelial cells), where a similar process has been described, were used as a comparison.

What did you find?

Despite other research studies on epithelial cells showing a clear implication of endothelial-like conversion in people with CF, we could not identify a similar signature in ECs. Rather, CF blood vessels display a limited signature of endothelial conversion, which could be induced by other underlying dysfunctions such as inflammation.

What does this mean and reasons for caution?

We hypothesize that endothelial conversion is not the primary driver of vascular dysfunction observed in people with CF, but is rather induced by other underlying phenomena occurring in CF ECs. However, this hypothesis still need to be carefully explored with in-depth investigations.

What's next?

It will be important to understand how ECs contribute to disease developing CF, and to develop new therapeutic strategies to heal malfunctioning blood vessels and improve patient's quality of life.

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