

Cystic Fibrosis Research News

Title: The role of endothelial cells in cystic fibrosis

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What was your research question?

If and how endothelial cells (ECs), the cells lining the inside of blood vessels, are involved in cystic fibrosis (CF) and how they may contribute to the different aspects of the disease.

Why is this important?

ECs control the exchange of oxygen, nutrients and cells between blood and nearby tissues. Besides controlling the exchange, ECs are important in a lot of other biological processes including blood clotting, inflammation, controlling the blood pressure and the formation of new blood vessels. All these processes are tightly regulated in normal healthy conditions, but can lead to several diseases upon EC dysfunction. That is why ECs could play a role in various elements of CF including heart and blood vessels (cardiovascular) symptoms, CF-related diabetes, excessive blood vessel formation and elevated blood pressure in the lungs and liver.

What did you do?

We extensively searched online databases containing medical and scientific publications to find all studies reporting about ECs and their functions in CF. The studies were grouped based on their subject. Afterwards all findings were summarized to conclude with their potential role in CF.

What did you find?

Despite the important role of ECs in many biological processes, they have been underinvestigated in CF although they could play a potential role in the disease. On top of



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that, as the life expectancy of CF patients has fortunately improved, one could expect new yet unknown symptoms to arise in the ageing people with CF. Since blood vessel diseases are age-related, vessel ageing could play a role in CF as it does in healthy people.

What does this mean and reasons for caution?

This review is the first one to describe the endothelial cells and their markers of dysfunction in CF and suggests that ECs could indeed be important in the disease mechanism of CF. Since blood vessels play an important role in many biological functions, it is often difficult to pinpoint to which process(es) it mainly contributes. In people with CF, it is also still unknown if some of the endothelial markers assessed are due to CFTR defects in ECs themselves, or if these ECs adapt themselves to other biological changes in people with CF.

What's next?

The current review has highlighted the important role of ECs in CF but more in depth studies are required to identify the mechanism by which ECs contribute to the disease.

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