



Cystic Fibrosis Research News

Title:

A case of self-limited drug induced liver injury under treatment with elexacaftor/tezacaftor/ivacaftor: when it is worth taking the risk.

Lay Title:

Liver toxicity with triple therapy in cystic fibrosis: when it is worth taking the risk.

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What was your research question?

Can liver malfunction induced by triple therapy recover spontaneously ?

Why is this important?

If liver abnormalities caused by triple therapy can be shown to normalize spontaneously, one could consider continuation of treatment with strict monitoring of liver function tests in cases where the benefits of treatment outweigh the risks of treatment continuation.

What did you do?

We reported a remarkable clinical case of a 58-year old female cystic fibrosis patient with severe airflow limitation and high symptom burden, developing severe liver dysfunction during treatment with triple therapy.

What did you find?

Liver toxicity can occur with triple therapy, which was proven by liver biopsy in this case. However, continued treatment led to a normalization of liver function. Triple therapy led to a significant clinical improvement of the patient's condition to the extent that she could be removed from the lung transplantation list.

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What does this mean and reasons for caution?

This case emphasizes the possibility of continuing treatment when liver function is strictly monitored and the patient is completely informed about the potential risks. One should stay vigilant for liver dysfunction under treatment with triple therapy, although a risk-benefit analysis should be done in an individual patient.

What's next?

We believe that a strict follow-up of liver function in all patients treated with triple therapy is warranted. Other such case reports with a sufficiently long follow-up period would be useful to provide guidance for those few patients faced with triple therapy induced liver malfunction.

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