

# Cystic Fibrosis Research News

**Title:**

**URSODEOXYCHOLIC ACID TREATMENT IS ASSOCIATED WITH IMPROVEMENT OF LIVER STIFFNESS IN CYSTIC FIBROSIS PATIENTS**

**Authors:**

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**What was your research question?**

Does ursodeoxycholic acid (UDCA) slow the progression of, or even improve, liver disease in people with cystic fibrosis (CF) who develop this complication.

**Why is this important?**

Up to 40% of people with CF will ultimately have some liver involvement, and in a third of these people it may progress to cirrhosis, i.e. advanced liver disease. Some might even die of the complications of liver disease. After taking UDCA signs of liver cell damage, i.e. high serum transaminases (ASAT and ALAT), improve or even normalize. However, until now the effect of UDCA on the long-term progression of liver disease has been unclear. Therefore its use is somewhat controversial, although most doctors will prescribe this drug for people with CF and (severe) liver disease.

**What did you do?**

A group of 105 people with CF were followed for an average period of almost four years. In this group, 73 people showed no signs of liver disease whatsoever and were not given any UDCA. The remaining 32 were given UDCA at an average dose of 16 mg/kg/day. These 32 people were subdivided into three groups: 11 had UDCA because of mild abnormalities of liver biochemistry, 15 had some fibrosis (scarring) of the liver by conventional criteria and



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six had advanced liver disease, i.e. cirrhosis. All participants were regularly evaluated for liver stiffness, i.e. the amount of liver scarring, by a new device (the fibroscan).

## **What did you find?**

In people with mild liver disease (“fibrosis”) who were given UDCA, liver stiffness generally improved over the four-year observation period. Conversely, in all those with advanced liver disease (“cirrhosis”) liver disease worsened.

## **What does this mean and reasons for caution?**

It appears sensible to start treatment with UDCA when the first signs of liver disease are seen, as UDCA seems to be able to reduce liver scarring. Starting this medication after cirrhosis has already developed does not seem useful. However, as this is a single-centre study, it should be repeated by others to strengthen the evidence for this recommendation. We also conclude that liver stiffness should be regularly measured in people with CF, as it allows for the non-invasive monitoring of liver scarring and liver disease.

## **What’s next?**

Even with the introduction of drug treatments which affect the genetic defect in CF (potentiators and correctors), our conclusions regarding the use of UDCA in people with early liver disease still seems sensible. Regular follow-up of liver disease with non-invasive measures, such as liver stiffness scans, will allow a future judgement on whether this remains a valid recommendation.

## **Original manuscript citation in PubMed**

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