

# Cystic Fibrosis Research News

## **Title:**

International prospective study of distal intestinal obstruction syndrome in cystic fibrosis: associated factors and outcome.

## **Authors:**

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## **What was your research question?**

The research question was to evaluate across Europe, the incidence of distal intestinal obstruction syndrome (DIOS) in children and adults, the management and the factors associated with occurrence, recurrence and resolution of DIOS episodes. It is the first prospective long-term study on DIOS.

## **Why is this important?**

It is important to better understand DIOS, a specific complication of cystic fibrosis that is responsible for incomplete or complete intestinal obstruction with significant illness. DIOS is characterized by the accumulation of viscous (thick sticky) material, combined with sticky mucous secretions located in the distal ileum and symptoms which are mainly apparent to the affected individual. Previous reports of DIOS without clear definition may have included constipation with an over estimated incidence and bias in the knowledge of this entity. We

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used the definition of DIOS based on the European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN) criteria recently published in a consensus document.

## **What did you do?**

We conducted a prospective long-term study in 10 countries in children and adults with CF. Demographic data were recorded, CF-comorbidity factors (meconium ileus, CF-liver disease, CF-related diabetes) and environmental factors (outdoor temperature, intensive sport activity, pancreatic enzyme dosage and compliance, current daily fat and fibre intake, beverage intake, dietetic recent modifications via a face-to-face interview with a dietician), severity of DIOS episodes, treatment modalities, DIOS resolution defined as the delay in tolerance for usual feeding starting from initial symptoms.

## **What did you find?**

DIOS incidence was similar in children and adults. Medical treatment failed only in cases of complete obstruction (11%). Long-term maintenance therapy was widely prescribed (86%). Children with meconium ileus had a significant higher rate of surgery for DIOS. Complete DIOS required longer hospitalisation. Delayed arrival at hospital and weight loss had a significant impact on DIOS resolution. Associated factors for DIOS included meconium ileus, pancreatic insufficiency, liver disease, diabetes. Low fibre intake and insufficient hydration were also frequent. Females had more recurrent episodes; constipation was more frequent with incomplete episodes and poor pancreatic enzyme compliance with complete episodes.

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

A weakness of our study was the lack of a 3-day nutrition and beverage record and of a control-matched CF group; however, organising this type of study at so many centres would have been unrealistic.

## **What's next?**

CF teams should be encouraged to include DIOS in educational programs, leading to earlier recognition and care. We emphasise here the complexity of factors contributing to DIOS. Our study will help gastroenterologists in understanding of this complication.

## **Original manuscript citation in PubMed**

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