

# Intestinal failure-associated liver disease—new position paper in *Journal of Pediatric Gastroenterology and Nutrition*

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Children who require long-term parenteral nutrition are at risk of a potentially devastating complication called intestinal failure-associated liver disease (IAFLD). The diagnosis, prevention, and treatment of IAFLD are discussed in a new position paper in the [\*Journal of Pediatric Gastroenterology and Nutrition\*](#), official journal of the European Society for Paediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) and the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition.

The new statement stresses the importance of early referral to specialized [intestinal failure](#) treatment centers, enabling proper multidisciplinary treatment of children with more severe or progressive IFALD. The position paper was developed by an expert panel under the leadership of Drs Florence Lacaille of Hôpital Necker-Enfants malades, Paris, and Girish Gupte of Birmingham (UK) Children's Hospital.

## IAFLD in Children—Causes and Risk Factors

Intestinal failure-associated liver disease refers to liver injury occurring in patients receiving long-term parenteral nutrition—intravenous administration of nutrients. Parenteral nutrition is required in children with various causes of intestinal failure, which makes them unable to receive gastrointestinal (enteral) feedings.

Making IAFLD particularly dangerous is the fact that it can be difficult to recognize—liver injury may develop gradually and without symptoms. The true prevalence of IAFLD is unclear; in one study of more than 300 children receiving home parenteral nutrition, 23 percent developed IAFLD. Studies of patients with established IAFLD have reported mortality rates up to 40 percent.

Risk factors for IAFLD include bowel injury in premature infants (necrotizing enterocolitis) and certain types of infections occurring in critically ill infants, including catheter-related bloodstream infections. Overgrowth of bacteria in the inactive small intestine may also play a role.

Parenteral nutrition feeding solutions themselves may also contribute to the development of IAFLD; nearly all components have been implicated as potential causative or aggravating agents. The statement provides recommendations for parenteral nutrients—including energy content, amino acids, and lipid (fat) emulsions—with an eye toward optimizing nutrition while limiting IAFLD risk.

## **Prevention and Treatment—Need for Early Specialist Referral**

A key step in preventing IAFLD is to introduce enteral feeding as early as possible. The presence of food in the gut stimulates intestinal adaptation, reducing the need for parenteral nutrition.

For children who need long-term parenteral feeding, treatment for intestinal failure and IAFLD is challenging. In some cases, surgery to lengthen the available intestine may help infants to better tolerate enteral feeding, potentially improving or preventing IAFLD.

For other patients, the best option is intestinal transplantation. Children with mild or moderate IAFLD may benefit from transplantation of the small intestine only. In those with severe IAFLD, combined liver and intestinal transplantation may be necessary.

For children with intestinal failure and complications of long-term parenteral nutrition, early contact with a transplant center is essential. "Discussion should be held with a transplantation center as soon as early signs of IFALD appear, even if transplantation is not yet indicated," the position paper states. "Waiting until IFALD is established before making a referral reduces the potential for successful outcome following transplantation."

The statement also emphasizes the benefits of treatment at a specialized intestinal failure center for infants and [children](#) on long-term PN. The authors conclude, "Optimal management requires the skills of an experienced multidisciplinary nutritional care team thoroughly familiar with this condition, aware of the potential for progression, and employing management strategies to reduce risk."

**More information:** "Intestinal Failure - Associated Liver Disease. A Position Paper By The ESPGHAN Working Group of Intestinal Failure and Intestinal Transplantation." [journals.lww.com/jpgn/Abstract...isease\\_A.98143.aspx](http://journals.lww.com/jpgn/Abstract...isease_A.98143.aspx)

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