Synopsis:

In the United States, patients dependent upon parenteral nutrition (PN) receive parenteral lipid emulsions composed of soybean oils. Lipids are necessary in PN dependent patients due to their high caloric value and essential fatty acid content. They have been implicated in predisposing patients to PN associated liver disease. Phytosterols such as those contained in soybean oils are thought to have a deleterious effect on biliary secretion. Accumulation of lipids in the hepatic Kupffer cells may further impair liver function.

Children requiring prolonged courses of PN are at risk for developing PN associated liver disease. The investigators hypothesize that although omega-6 fatty acid emulsions prevent fatty acid deficiency, they are not cleared in a manner similar to enteral chylomicrons and therefore accumulate in the liver and resulting in steatotic liver injury. It is further hypothesized that a fat emulsion comprised of omega-3 fatty acids (i.e., fish oil) such as Omegaven™ will be beneficial in the management of steatotic liver injury by its inhibition of de novo lipogenesis, the reduction of arachidonic acid-derived inflammatory mediators, prevention of essential fatty acid deficiency through the presence of small amounts of arachidonic acid, and improved clearance of lipids from the serum. Animal studies have shown that IV fat emulsions (IFE), such as fish oil that are high in eicosapentaenic and docasexaenoic acid reduce impairment of bile flow which is seen in cholestasis caused by conventional fat emulsions. Furthermore, investigators hypothesize that that intravenous omega three fatty acids will be well tolerated and might reduce the inflammatory effect in the liver of prolonged PN exposure and could potentially reverse any hepatic dysfunction due to PN/IFE use. By administering Omegaven™ in place of conventional phytosterol/soybean fat emulsions we may reverse or prevent the progression of PN associated cholestasis and thus allow the patient to be maintained on adequate PN until they are able to ingest adequate nutrition enterally.

In this study eligible patients will be given fish-oil-based emulsion, Omegaven, as part of their parenteral nutrition, rather than soybean based emulsion. This will be done under an FDA drug IND. Patients will be monitored for their tolerance of Omegaven and for resolution, stabilization, or progression of their cholestasis and liver dysfunction.