



# Perinatal Gazette

Newsletter of the Regional Perinatal Center  
Westchester Medical Center



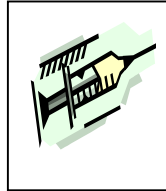
Volume 4, Issue 5

September 2004

## Liver Transplantation in Neonates

Liver transplantation in humans began with the first successful transplant of a child in 1967. After the development of cyclosporine in the late 1970s and the NIH's declaration, in 1983, that liver transplantation is "no longer an experimental procedure," liver transplantation grew to become the standard treatment for both chronic and acute forms of liver failure. Advancements in surgical technique and immunosuppression have allowed for ever increasing success rates for pediatric liver transplantation. However, transplantation in neonates (defined, for this article, as children under the age of three months) has been challenging for a number of reasons. Obviously, the small size of these children at the time of transplantation contributes to difficulty obtaining suitable donor organs, as well as increases technical difficulties when performing the transplant itself. In addition, children who need transplantation at such a young age have little ability to tolerate severe physiologic stress and are often malnourished and/or in respiratory/renal failure at the time of surgery. Consequently, complications and mortality are increased. For all of the above-stated reasons, liver transplantation in the neonate is a significant challenge

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## Vaccinations in Pregnant and Breastfeeding Women

The benefits of vaccines are great and include partial or complete protection against the consequences of infections for the vaccinated individual, as well as overall benefits to society. No vaccine is completely safe. Risks of vaccines can include common, minor ones to rare, life-threatening conditions. In deciding who should receive vaccinations and which ones, the benefits to the individual and society greatly exceed the risks.

To date, no evidence exists of a risk from vaccinating pregnant women with inactivated viral or bacterial vaccines or toxoids. The recommendations regarding vaccinating pregnant women take into consideration the risk of exposure to the infection, the potential seriousness of infection to both the mother and the fetus, and the overall safety of the vaccination.

Tetanus toxoid is indicated routinely in pregnant women. If pregnant women have not received the tetanus toxoid vaccination within the past ten years, then it should be administered in pregnancy. Women in the second and third trimesters of pregnancy should routinely receive influenza vaccinations during the flu season (usually December to March). In particular, women who have underlying medical conditions that increase their risk of complications from influenza should be vaccinated before flu season regardless of the stage of pregnancy. Inactive polio vaccine can be administered to pregnant women who are at risk for exposure to wild-type poliovirus infection. Hepatitis B vaccine is also recommended for pregnant women at risk for hepatitis B infection. Hepatitis A, pneumococcal and meningococcal polysaccharide vaccines should be given to women at increased risk for those infections. Pregnant women, who must travel to areas where the risk of yellow fever is high, should receive the yellow fever vaccine.

Pregnant women should not receive the measles, mumps, rubella, and varicella vaccines, although the risks are theoretical. Breastfeeding is not contraindicated for any vaccine.

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## Neonatal Short Bowel Syndrome

Short bowel syndrome is defined as malabsorptive state following massive loss of small intestine. Most cases of short bowel syndrome begin in the neonatal period following intestinal resection either for necrotizing enterocolitis or gastrointestinal anomalies such as intestinal atresia, gastroschisis, omphalocele or Hirschsprung's disease. Reduced absorptive surface area results in varying degrees of malabsorption, malnutrition and failure to thrive. Other factors also play a role in the development of malabsorption, including loss of the ability to secrete gastrointestinal regulatory and trophic hormones and loss of the gastrointestinal immune functions. The malabsorption is further aggravated by the presence of ostomy, bypassing large portion of the small and large intestine and by abnormal intestinal bacterial colonization (bacterial overgrowth), causing mucosal inflammation that further exacerbates the fluid, electrolytes and nutrient losses. Despite many advances in the treatment of short bowel syndrome, extended hospitalization, multitude of complications, such as sepsis and parenteral nutrition induced liver damage and high mortality remain a major concern.

The clinical management of these patients is a multi-stage process beginning with parenteral nutrition, transitioning through varying stages of enteral nutrition and pharmacotherapy, and hopefully, ending up with a patient who can eat in a relatively normal fashion without supplemental intravenous nutrition.

The primary challenge during the first stage of therapy is maintaining fluid and electrolyte balance by continuous replacement of the ostomy losses. Once fluid and electrolyte losses have diminished, enteral feeding is initiated. Breast milk, peptide or amino acid-based formulas are usually used at this stage to facilitate absorption and to reduce the risk of secondary allergic inflammation in the gut. These formulas are initiated at a very slow rate and often at a dilute concentration. The rate of infusion and the concentration are gradually increased with concurrent reduction in parenteral nutrition. Continuous enteral infusion has many advantages over bolus feeding, especially in small infants by better promoting the intestinal mucosal growth and adaptation. However, enteral nutrition bypasses the cephalic and oral phases of digestion. Some oral feedings should be initiated very early to allow the neonate to learn to suck and swallow and to prevent the development of feeding aversion. Enteral feeding formulas should be relatively high in fat and low in carbohydrate, as these create a lower osmotic load on the gut and provide less substrate for bacterial overgrowth in the small intestine. Providing aggressive enteral nutrition reduces the need for parenteral nutrition and significantly reduces the hepatic injury caused by parenteral nutrition, the primary cause of morbidity and mortality in neonates with short bowel syndrome.

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Pharmacologic therapy, in addition to enteral feeding, plays an important role in the management of patients with short bowel syndrome. Careful attention should be paid to the minerals, vitamins and bile acids deficiencies, which invariably develop depending on the resected bowel segment (duodenum, jejunum, ileum or colon). Patients should be supplemented early in the course of the disease with calcium, magnesium, iron and zinc, as well as water and fat-soluble vitamins. Ursodeoxycholic acid and cholecystokinin may have an ameliorating effect on the liver injury. Acid secretory inhibitors (Zantac) and prokinetic agents (Reglan) are also frequently used. Agents, which slow gut motility (Loperamide) and Octreotide, a long acting somatostatin analogue which has anti-secretory properties are also used. Rotating or continuous oral antibiotic therapy is often indicated when bacterial overgrowth is suspected. Several different therapeutic protocols exist, including metronidazole with trimethoprim sulfamethoxazole, amino gentamicin, penicillin's and cephalosporins. Further, probiotics, naturally occurring intestinal bacteria, such as Lactobacillus, are also being used to ameliorate the bacterial overgrowth and its attendant mucosal inflammation. Growth hormone and glutamine have shown mixed results in the augmentation of mucosal adaptation. Epidermal growth factor and glucagons like peptide 2 are showing promising results in animal studies.

Surgical interventions such as the Bianchi procedure to reduce intestinal dilatation, improvemotility and extend surface area may be helpful. Liver or intestinal transplantation alone or combined are more frequently used in refractory cases with promising results.

Short bowel syndrome is a chronic condition requiring a great deal of patience and attention to multi-stage therapy, careful screening for complications and constant re-evaluation. Therapeutic decisions should be measured in months to years rather than in days to weeks.

At Maria Fareri Children's Hospital, WMC, NYMC we have established "Neonatal Short Bowel Syndrome Team", comprising of Neonatologist, Surgeon, Neonatology Fellow, Gastro-enterologist, Dietician, Social worker and Neonatal nurse to address the complex medical and psychosocial needs of the infants with short bowel syndrome and their families and to investigate and to incorporate new modalities of treatment to continue to improve immediate and long term outcome. For the "Short Bowel Syndrome Team":

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Continues from liver

Although less than 15 neonatal transplants are performed each year, when transplantation is necessary, the main indications are giant cell hepatitis, iron storage disease, and viral hepatitis. This is in contrast to older infants and children who undergo liver transplantation most often for biliary atresia. In most instances, the etiology of giant cell hepatitis is unknown and reflects a rather nonspecific injury to hepatocytes. Unidentified peri-natal infection and inborn errors of metabolism are suspected contributory factors. When a clear etiology can be identified, hemochromatosis is the most common indication for liver transplantation. Its course is more fulminant than giant cell hepatitis and requires transplantation within days of diagnosis.

In the late 1980s and early 1990s, the increasing disparity between the number of patients on the waiting list for liver transplantation and available donor organs led to significant innovation in the techniques of liver transplantation. Initially, full size cadaveric donor livers were “reduced” in size by removing the left lateral segment of the liver for subsequent transplantation – the remaining portion was then discarded. Techniques then evolved so as to avoid “wasting” more than half of the liver to create two transplantable livers from one donor. This was achieved by separating the left lateral segment of the liver while preserving the remaining extended right lobe, which could then be utilized for another recipient. These modifications to standard techniques laid the groundwork for the development of living donor liver transplantation which began initially in children with the left lateral segment, but has since been expanded to right lobe adult to adult transplantation. Yet another innovation required for the smallest recipients was the further modification of a left lateral segment graft (segments II and III) to a “monosegment” (Segment II or III). While these modifications increased organ availability to children, they were also associated with an increased incidence of technical complications (e.g. biliary leaks/strictures and vascular thromboses).

Not surprisingly, the incidence of complications after neonatal liver transplantation is higher than in other age groups. Patient and graft survival rates range, respectively, from 55-60% and 38-56%. This contrasts with that of children older than 12 months who have graft and patient survival rates of approximately 80% at one-year post transplant. Re-operations in this population are common. One of the most common indications for re-operation is the need for delayed abdominal closure. This is a consequence of the large size of the graft relative to the patient, which prevents primary closure at the time of transplant. Closing the abdomen at this time may lead to increased intra-abdominal pressure, which can lead to respiratory and renal compromise with subsequent graft loss and mortality. In addition, hepatic artery or portal vein thrombosis occurs in up to 11% of patients, and biliary complications occur in 10-30% of patients. Yet another postoperative problem that contributes significantly to morbidity and mortality are bacterial and fungal infections, which are common in this immunosuppressed population.

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Immunosuppressive regimens are essentially the same as those of older children and rely on “triple drug therapy.” This consists of a calcineurin inhibitor (cyclosporine or tacrolimus), an antimetabolite (mycophenolate mofetil or azathioprine) and steroids. Induction therapy with monoclonal or polyclonal antibodies may also be used to minimize or eliminate the need for steroids.

Although the occurrence of the previously mentioned complications is greater in neonates, there are some advantages to operating on such extremely young patients. Neonatal recipients have a lower incidence of rejection and are not mandated to receive only ABO compatible organs. Multiple studies have shown that this group of patients is able to receive ABO incompatible grafts without the rapid rejection that attacks the grafts of older recipients. This is in large part because of the inherent immaturity of the neonatal immune system. The “holy grail” of transplantation is “tolerance,” the physiologic state whereby the graft and the patient live in harmony without rejection and without immunosuppressive medications. This small group of recipients is often able to achieve this goal.

Because of the relative paucity of neonatal liver transplants at any given center, there is little long-term data on outcomes. The few studies that do exist reflect a challenging experience fraught with the difficulties of transplanting these tiny children. Evolution of surgical techniques and immunosuppressive protocols in the future that are aimed specifically at this subset of pediatric liver transplant recipients will likely lessen the incidence of complications, improve quality of life and increase survival.

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**Annual Regional Perinatal Forum:  
Reducing Racial and Ethnic Disparities in  
Perinatal Care in the Hudson Valley**

*Sponsored by the Hudson Valley Regional Perinatal Forum  
&*

**Brains, Nutrition and Regional Perinatal Care:  
A Vision with a New Focus**

*Sponsored by the Regional Perinatal Center at Westchester  
Medical Center*

**Tuesday, October 5, 2004**

8:30am – 4:00 pm

At the Maria Fareri Children’s Hospital AuditoriumValhalla,  
New York

CME/CEU’s are available

For any questions and/or registration please contact  
(914) 493-8590

## Lower Hudson Valley Perinatal Network

Since its inception in March of 2004, the Lower Hudson Valley Perinatal Network has gained the support of numerous clinical and human services organizations in all four of its counties – Dutchess, Putnam, Rockland, and Westchester. The goal of the Network is to capture women who are at risk for delayed prenatal care or no prenatal care, poor compliance with care, and poor birth outcomes, and connect these women with organizations that best suit their needs. It also seeks to provide education and support systems to all women of childbearing age and their families, particularly those at high-risk. The Network's website – <http://www.nymc.edu/depthome/peds/rpc/perinetwork.asp> - provides information on its purpose, activities, and products. The Network is currently developing a perinatal resource inventory for use by providers and consumers that will be made available on the website in the next few months and that will have interactive capability. A county-level and municipality level perinatal health assessment of the four counties – individually and as a region – is in process and will be available for distribution early next year. The information provided in this assessment will allow healthcare organizations to identify key sociodemographic and perinatal health issues in their area. Useful perinatal data sources are also currently listed on the website and will be periodically updated, as will ongoing meetings and educational events. The creation of the Lower Hudson Valley Perinatal Network was made possible by funding from the March of Dimes.

**Audrey Waltner, MPH**

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We are interested in providing you with a newsletter that is relevant and of interest to you. Please contact us with perinatal topics you would like to see addressed. For a copy of our newsletter or to be placed on our mailing list contact us by phone or e-mail. Please see below the NYMC neonatal web site address to locate other issues of The Perinatal Gazette:

<http://www.nymc.edu/neonatology>

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