

From the Desk of the Director

This issue of our newsletter discusses gastroschisis.

We have provided information describing prenatal management, and the subsequent surgical procedure and neonatal management.

We are also pleased to announce the implementation of infant care coordination services. We appreciate those

of you who responded to our survey regarding infant services available through UNC. We feel that the infant care coordination program will enhance our existing services by the facilitation of timely communication between UNC providers and community providers, provide families with education, counseling, and support during and after their infant's hospitalization, and streamline patient appointment scheduling to decrease the number of visits families have to make to Chapel Hill.



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Neonatal management of Gastroschisis

Neonatal management of infants with gastroschisis begins in the delivery room with stabilization and preparation for surgery or transport to a tertiary care facility. With an average gestational age of 34 to 36 weeks, complications of prematurity, such as respiratory distress syndrome, may exist. As with all neonates, management of respiratory distress and hypovolemia or acidosis take priority. Bowel distention from prolonged bag-mask ventilation or nasal CPAP should be avoided, and a nasogastric tube for decompression should be inserted.

Due to the large surface area of exposed bowel, these infants experience increased heat and fluid losses. Thermoneutrality is aided by wrapping the exposed bowel in warm, saline-soaked towels or gauze, without twisting or strangulating the bowel, and enclosure of the lower half of the infant in a sterile "bowel bag" or plastic wrap. Hypovolemia is treated with crystalloids and maintenance fluids must be generous and contain electrolytes to compensate for losses.

Broad spectrum antibiotics are started and echocardiogram and chromosome studies are considered if symptoms or

other anomalies exist.

Surgery occurs within hours of birth and the type of repair dictates post-operative management. Co-morbidities such as RDS, meconium aspiration or bowel atresias may complicate recovery. After a primary repair, transient respiratory insufficiency, due to increased intra-abdominal pressure, may occur and ventilator support is determined accordingly. After a staged repair, with the placement of a silo, less respiratory insufficiency is initially present, but difficulties may arise as the bowel is serially reduced into the abdomen. High fluid requirements continue for several days, until resolution of 3rd space losses from the bowel. If the fascial closure is too tight, perfusion to the bowel may be compromised. Impaired venous return due to pressure on the inferior vena cava, and acute renal failure may also occur. A urinary catheter can monitor

rising bladder pressures and signal the need for surgical relief.

Return of bowel function precedes establishing enteral feeds. This process often takes 2 to 6 weeks, necessitating parenteral hyperalimentation via a central line. Feeding intolerance is common and malabsorption may exist. Often, elemental formulas are indicated. While most infants establish feeds prior to discharge, those with short-gut syndrome,

due to bowel removal, may require prolonged parenteral nutrition, placing them at risk for cholestatic jaundice and sepsis.



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Gastroschisis Surgical Procedure

The treatment of infants with gastroschisis improved dramatically in the late 1950's with the development of parenteral nutrition (complete IV nutrition) and the technique of creating a Silastic (plastic) silo around the exposed bowels until they could be reduced back into the abdomen. With these advances the mortality of infants with gastroschisis dropped from over 75% to less than 5%.

As surgeons and pediatricians became more experienced with prolonged IV nutrition (TPN) for babies, children born with gastroschisis were some of the first and most successful outcomes using this new therapy. TPN allowed a method to counteract the long periods of time during which these babies could not eat, the so-called "bowel inertia", so that they no longer died of malnutrition.



Tim Weiner, MD
Section of Pediatric Surgery, Department of Surgery

It still has not been discovered what causes this bowel inertia but clinicians feel that therapies that would decrease the inflammation of the exposed bowel by the amniotic fluid might be beneficial. Trials of amniotic fluid exchange for fetuses with gastroschisis have been proposed to decrease this inflammation or "peel".

About 20% of the time, the "peel" is minimal and the bowel can be returned to the abdomen at the first surgery. These patients clearly have a faster return of function and a very satisfying cosmetic result. The remaining 80% require the creation of a temporary, artificial abdominal wall to contain the intestines—the "silo". This silo protects the bowels from the risk of infection and allows the surgical team a mechanism to progressively compress them back into the belly. Much like squeezing a tube of toothpaste, the intestines are carefully pushed back into the abdomen over a 4-7 day period. As the baby pees away its extra "birth water", the swelling

and edema of the bowels diminishes and assists in the reduction. Leaving the child paralyzed and on a ventilator also helps relax the abdominal wall muscles and hastens the closure. Newer plastic silos have been developed which do not require stitching to the baby and decrease the time of the initial surgery.

Following the closure or siloing of the patient, a central venous catheter ("Broviac") is placed for the administration of IV nutrition. Feeding of the baby begins when the abdomen has been completely closed and bowel function has returned. This is the most frustrating period for parents and may take from 2-5 weeks before the child is ready to go home.

Today, most babies born with gastroschisis will do very well and have a normal life post-operatively. As dramatic and upsetting as it may be for families when the child is born, this condition is one of the simplest congenital problems to fix and should have an excellent outcome.

Prenatal Management of Gastroschisis

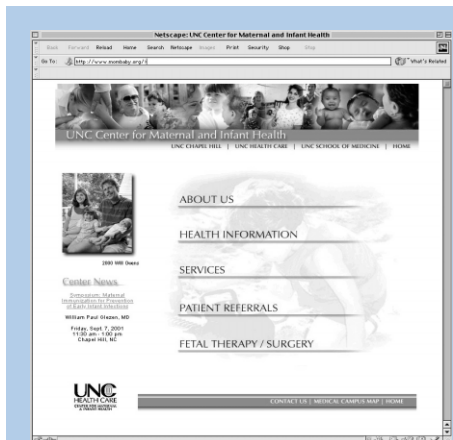
Referral to The University of North Carolina Hospitals (UNC-CH) for fetal evaluation of a possible abdominal wall defect such as gastroschisis is not uncommon.

Gastroschisis is a fetal abdominal wall abnormality in which there is evisceration of fetal abdominal contents such as bowel, stomach, or even liver through a paraumbilical wall defect. Other fetal abdominal wall defects include, but are not limited to, omphalocele, body-stalk abnormality, and Pentalogy of Cantrell. Gastroschisis occurs in approximately 1 out of every 4,000-5,000 births with males and females being equally affected. Antenatally, up to 75% may be detected by maternal serum alpha-fetoprotein screening, as the alpha-fetoprotein level is usually elevated in these pregnancies. The diagnosis is confirmed by ultrasound.



Michael J. McMahon, MD, MPH
Maternal-Fetal Medicine Division, Department of Obstetrics and Gynecology

After the gastroschisis is confirmed a thorough investigation is made for other possible birth defects. Amniocentesis is offered to evaluate whether or not there are any chromosomal abnormalities. However, the rate of aneuploidy is low (unlike omphalocele in which it is high). Pediatric surgery and neonatal consultation are requested. Ultrasonography is utilized throughout pregnancy to evaluate the fetal bowel, fetal growth pattern, and amniotic fluid. Fetal growth is frequently below normal (restricted). Evaluating the bowel for intra- or extra-abdominal dilatation and thickening is warranted and may impact the timing of delivery. There has been a long-term debate as to whether or not women whose pregnancy is complicated by fetal gastroschisis would benefit from an elective cesarean delivery versus an attempted trial of labor in order to decrease the risk of possible fetal bowel damage during the laboring process. At UNC-CH we generally allow women to labor. Finally, the survival rate for gastroschisis diagnosed prenatally, now exceed 90%.



**Visit The Center's Web Site:
www.mombaby.org**

We recently added several new fact sheets on congenital anomalies. Our patient information sections are available in both English and Spanish. We will continue to add fact sheets on prenatal conditions and caring for medically fragile infants.

Please contact us to let us know how our web site can better serve you, your patients and their families.

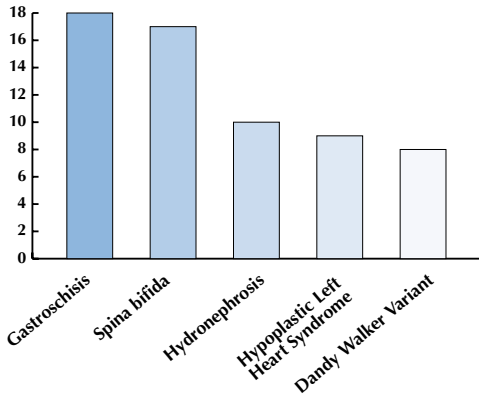
Perinatal Care Coordination Update

In November 2000, the UNC Center for Maternal and Infant Health implemented perinatal care coordination services targeting prenatally diagnosed fetal anomalies. In our first year of

service, we have diagnosed 73 different fetal conditions. We have enrolled a total of 262 patients (159 women, 80 live infants, and 23 fetal, neonatal and infant deaths). The map below depicts

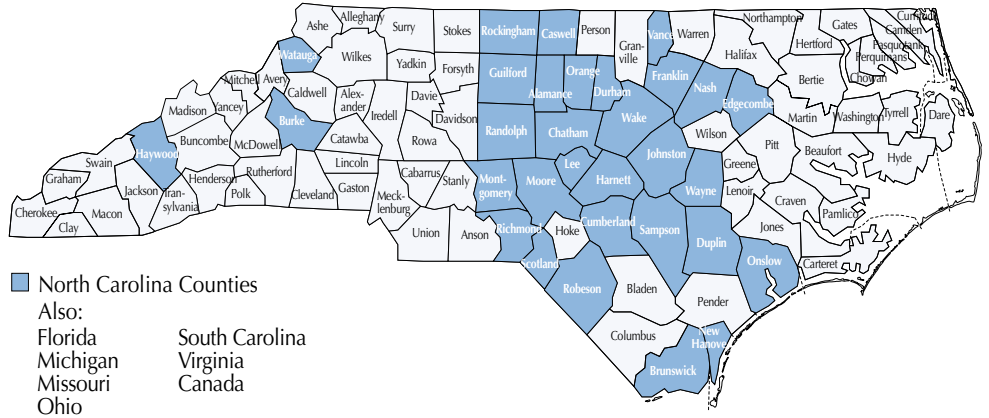
the different counties and states where our patients reside. The most common fetal anomalies seen at the UNC Center for Maternal and Infant Health are depicted in the bar graph.

Top 5 Medical Diagnoses



Patients' County of Residence

November 2000-November 2001



Infants born and referred to UNC with one or more of the following medical conditions will be enrolled in our infant care coordination program. For more information on the referral process, please contact Lori Carter, RN, BSN, at 919.843.4428 or visit our web site: www.mombaby.org.

MEDICAL CONDITIONS OF THE INFANT

Cardiac

- Atrioventricular Canal
- Complete Heart Block
- Double Outlet Right Ventricle
- Hypoplastic Left Heart Syndrome (HLHS)
- Single Ventricle
- Tetralogy of Fallot (TOF)
- Transposition of Great Vessels (TGA)
- Tricuspid Atresia
- Truncus Arteriosus

General and Thoracic

- Congenital Cystic Adenomatous Malformation
- Diaphragmatic Hernia
- Gastroschisis
- Intestinal, Duodenal or Esophageal Atresia
- Omphalocele
- Ovarian Cysts

- Sacrocoxygeal Teratoma (SCT)
- Thoracic Tumors
- Volvulus

Medical Genetics

- All chromosome abnormalities (Trisomy 13, 18, 21, Turner Syndrome, Klinefelter Syndrome)
- Microcephaly
- Multiple congenital anomalies (diagnosed and undiagnosed syndromes)
- Skeletal Dysplasias (e.g. osteogenesis imperfecta and Achondroplasia)

Miscellaneous

- Conjoined Twins
- Multiple births
- Non-Immune Hydrops
- Red Cell Alloimmunization
- Twin-Transfusion syndrome

Neurologic

- Cranial Deformities
- Craniosynostosis
- Encephalocele
- Hydrocephalus
- Microcephaly
- Neural Tube Defects

Orthopedics/Rehabilitation

- Achondroplasia (Dwarfism)
- Osteogenesis Imperfecta (OI)
- Skeletal Dysplasias
- Talipes Equinovarus (clubfoot)
- Arthrogyrosis

Urology/Nephrology

- Absent Organs/Pelvic Abnormality
- Bladder Outlet Obstruction
- Exstrophy or Cloacal Anomalies
- Hydronephrosis (including pelvic stasis)
- Intersex/Ambiguous Genitalia
- Neural Tube Defects
- Pelvic Mass
- Multicystic Kidney
- Polycystic Kidney
- Prune Belly
- Renal Mass
- Teratoma
- Ovarian Cyst

CenterNews

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Pediatric Care Coordination Services Now Available Through the UNC Center for Maternal and Infant Health

Ms. Lori A. Carter, RN, BSN, joined the UNC Center for Maternal and Infant Health as a Pediatric Care Coordinator on September 17, 2001. Ms. Carter comes to this position with 14 years of Newborn Critical Care experience and expertise in working with critically ill infants who require complex medical treatments.

Ms. Carter will provide care coordination services for infants with complex medical conditions. In an effort to assist families with streamlining care for their infants and understanding their special health needs. Her work will involve educating and supporting families while their infants are hospitalized. After hospital discharge, she will assist families with coordinating their infants' medical appointments, identifying community resources, and providing education, and counseling on caring for infants with complex medical conditions. She will work with multiple UNC pediatric subspecialists and ensure that there is timely communication with the infants' pediatricians.

This service is designed to assist families with infants who require multi-specialty follow-up after hospital discharge.

Pediatric care coordinators:

- Provide referring physicians and community providers with updates concerning the care of their patients
- Contact community providers regarding hospital discharge to discuss the potential discharge plan and date
- Are a resource for families throughout their infants' hospitalization
- Aid in the transition to home and community with a medically fragile child
- Assist in the coordination of follow-up clinic appointments



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