In Utero and Postnatal Management of Gastroschisis

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There is evidence that the incidence of gastroschisis (GS) has been increasing over the last 35 years and 1300-1500 children in the US are born with it each year. It consists of a small abdominal wall defect and usually occurs on the right side of the umbilicus. Abdominal contents are eviscerated and in amniotic fluid (AF) during gestation with no protective membrane surrounding them. The extruded viscera commonly includes small bowel, but may involve large bowel, stomach, and rarely other organs.

From the 5th to 9th week, the intestine elongates and herniates into the umbilical ring, then undergoes a series of rotations. Around the 10th week it re-enters the abdominal cavity. Diagnosis is made after this point by fetal ultrasound. An alpha-fetoprotein level is elevated in GS, and can be used to distinguish it from omphalocoele. The etiology of GS tends to be controversial and probably involves a combination of factors including a weak spot created by involution of the right umbilical vein, rupture of the umbilical stalk during mid-gut herniation, teratogenic exposures, and genetic factors.

Pregnancy with GS carries an increased risk of preterm labor and delivery, growth restriction, and stillbirth. There is a 5-25% incidence of associated defects, mostly intestinal atresias, caused by ischemic or mechanical injury to eviscerated bowel. The cause of injury is likely due to exposure to digestive enzymes, urine, and inflammatory mediators in AF, amino acids which make AF hyperosmolar, and the partial closure of the abdominal wall defect constricting the eviscerated intestines. Some hypothesize that vaginal delivery directly contributes to the development of a “peel”. These changes lead to impaired absorption and a motility disorder with the need for long-term parenteral nutrition (PN). Survival in GS is >90% and mortality is also related to prematurity; most deaths are caused by sepsis, necrotizing enterocolitis, abdominal ischemia, or late hepatic failure.

Initial problems with poor growth are usually overcome in the first few years. Psychosocial development and emotional problems are higher than in the general population, although some of these differences are related to length of stay, prematurity, and other neonatal difficulties. In complicated cases, GS may require home PN and is one of the more common reasons for discharge from the NICU on PN. In this latter group, mortality is nearly 50% in the first 2 years of life.
Treatment for stress incontinence has undergone major transformation in the last decade, from extensive surgical interventions to minimally invasive, ambulatory surgery. Currently the primary treatment for stress urinary incontinence is the tension free mid urethral sling. There are generally two approaches in placing a tension free mid urethral sling: retropubic, and trans obturator. The retropubic approach is commonly known as the TVT and the trans obturator is commonly known as the TOT. Both approaches are effective and provide cure or significant improvement in up to 80%-90% of patients. The newer innovation in stress incontinence treatment is the adjustable slings, which is indicated for the small percentage of patients who failed tension free slings. Single incision slings are the newest group of slings that are even less invasive by avoiding any external skin incisions.

The central nervous system, the brain stem, and spinal cord all play important roles in bladder function. Changes in the neuromuscular function can result in urinary urgency, and urge incontinence, more commonly known as over active bladder (OAB). The treatments for OAB are pelvic floor exercises, timed void and oral medications. Medications for OAB are generally the anticholinergic/ antimuscurenic group, such as Detrol, Ditropam. All medications can produce side effects, commonly dry mouth, dry eyes, constipation, blurred vision, and cognitive impairment. There are newer more bladder specific medications such as Vesicare, Enablex, and Sanctura. These medications target the bladder’s muscurinic receptors and thus bind less to the central nervous system and GI tract, resulting in fewer side effects. Unfortunately, despite improvement of side effect profiles, not all patients want to take medication or are able to tolerate medications for OAB.

Non-medication treatment for OAB includes neuromodulators, such as InterStim. InterStim is a device that provides electrical impulses to the third sacral nerve root. It has a testing phase, where a patient gets the stimulator placed with local anesthesia or sedation to evaluate its efficacy. Generally after having the testing electrode placed, the patient perceives benefit or lack of benefits within a few days. A permanent implant will be placed only if the patient perceives benefits. InterStims may function up to 10 years and are efficacious in up to 70-80% of patients.

The newest treatment option for OAB is Botoxin injection. Botoxin A is injected directly into the bladder detrusor muscles via cystoscopy. Botoxin paralyses the detrusor muscles and thus treating OAB symptoms. It is efficacious and efficacy may last up to 8 months.

Bladder function is complex, involves intact central, peripheral nervous system as well as musculature surrounding the urethra and bladder. Treating bladder disorder involves a thorough evaluation of the neuromuscular function. Effective treatment is available once the underlying cause is understood. Women suffering from voiding dysfunction are subject to severe emotional distress and are at risk for anxiety, depression and social isolation. Physicians and health care providers can make a difference in patient’s lives by actively inquiring about bladder function.

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The Maria Fareri Children’s Hospital at Westchester Medical Center (MFCH WMC) was a New York State Chapter Northern Metro Division Sponsor of March for Babies 2010. On April 25, 2010, at Saxon Woods Pool in White Plains, MFCH at WMC joined with half a million walkers from around the country to become a member of the most potent force of supporters and volunteers ever assembled to save babies.

The March for Babies was a success this year and the money raised for March for Babies supports the fight against premature birth in many ways by:

- Funding research to find out what causes premature birth and how it can be prevented.
- Supporting families whose babies must spend time in neonatal intensive care units (NICU’s) by developing and expanding NICU family support programs.
- Assisting health care professionals to improve prematurity risk detection and addressing risk factors with patients.
- Educating women about the signs of preterm labor.
- Advocating for expanded access of health insurance for mothers and babies.
- Providing women and men of childbearing age with the latest information about pregnancy and premature birth.

The March of Dimes is fortunate to have the support of so many dedicated friends in the community. Again, we thank The Maria Fareri Children’ Hospital at Westchester Medical Center for your participation and we look forward to working with you next year for March for Babies 2011! (April 24, 2011)
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life, although outcomes have continued to improve over the years. Up to 5% of neonates with GS end up with short gut syndrome and many of those patients will not survive. Intestinal transplantation and bowel lengthening procedures may be the only chance in a subset of children that cannot be weaned off of TPN. It is difficult to quote the success rate for these procedures as reports are confined to selective case series and patients with GS are often reported together with patients with other gastrointestinal problems.

There are several issues that are still controversial regarding GS management, and they include: amniotic infusion, timing and method of delivery, timing and method of repair, and nutritional management.

Serial amniotic infusions or oligohydramnios is present have been reported to reduce bowel injury by lessening the effects of AF contents and cushioning the extruded bowel wall, but this has not been subjected to a randomized trial and the literature consists of case series. In AF exchange, AF is replaced with an equal amount of normal saline or lactated ringers, with the expectation of decreasing intestinal damage. Definitive demonstration of the benefit of this therapy has yet to be shown; in particular, it is not clear which subgroup of patients may benefit, nor is there an unambiguous strategy for the timing and number of such amnioexchanges.

It was originally thought that early delivery and repair might improve intestinal function because of the damage caused by prolonged exposure to AF and progressive constriction of the defect. Definitive evidence that this strategy is of any benefit is lacking. Preterm delivery may simply cause morbidity, especially in later preterm infants, including neurological sequelae not previously recognized. A meta-analysis from 2009 looked at 10 studies that evaluated whether bowel dilatation was a good predictor of morbidity and mortality. It found that these infants are not at increased risk for adverse outcomes; thereby early delivery for dilatation may not be indicated.

Almost all fetuses with GS can be delivered vaginally without increased injury to the bowel, but there is a belief that c-section protects the viscera from birth trauma, edema, and ischemia, prevents exposure to vaginal bacterial flora, and eliminates the chance of dystocia. However, several retrospective reviews have examined this and have found that there was no benefit based on mode of delivery.

Closure or coverage of the defect is the highest priority to limit injury; this should be done in the DR to prevent water and heat loss by wrapping the torso in a transparent plastic film to limit inadvertent volvulus and ischemia. After stabilization of the newborn, surgical management should be initiated either primary closure or a silo, depending on the size discrepancy and infant’s status. 60-70% can be primarily closed with vigorous stretching of the abdominal wall and defect; however it makes sense to avoid intra-abdominal hypertension during the first few days of life when major hemodynamic and ventilatory changes occur. Increased abdominal pressure may limit the safety of complete closure. The introduction of the silastic silo was a critical and simple step forward in the management of GS. It establishes a nearly watertight barrier to evaporative heat loss and the intestines are gradually reduced into the abdominal cavity, which slowly expands to accommodate them. Previous studies favoring primary closure for survival may have contained a selection bias because those with the worst intestinal damage could not be primarily closed. Silo use is limited to two weeks and when reduc-

New York Premature Infant Health Network

The Premature Infant Health Network is an initiative of the Association of Perinatal Networks of New York and recognizes the unique and sometimes life-long challenges children born prematurely and their caregivers face once leaving the NICU. In an effort to better understand these challenges the Premature Infant Health Network, along with the 18 Perinatal Networks of NY State, will bring together community and health organizations, health care providers, and parents in roundtable meetings across the state to hear firsthand the many unique challenges premature infants are facing since leaving the hospital. The information will be used in efforts to increase quality healthcare access and awareness around premature infant issues.

To learn more about the NYS Premature Infant Health Network please contact: Erin White, Program Coordinator, Association of Perinatal Networks Phone: 607-772-0517 ewhite@associationofperinatalnetworks.org
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 visit http://www.worldclassmedicine.com/RPC for information about the Regional Perinatal Center at the Maria Fareri Children's Hospital at Westchester Medical Center and to locate other issues of The Perinatal Gazette.

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