

The Perinatal Gazette

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Hirschsprung Disease (Congenital Aganglionic Megacolon)

Divya Chhabra, M.D., Myra Mercado, M.D.

Hirschsprung disease is a birth defect that affects the large intestine and causes delayed passage of stool in the first 24 to 48 hours after birth in the newborn. It was first described by Harald Hirschsprung in 1886. With an incidence of 1/5000 live births, it is the main genetic cause of functional intestinal obstruction.² The disease is more common in boys and is known to be multifactorial. It may be familial or develop spontaneously.

The clinical features in the newborn period are delayed passage of meconium, distended abdomen, bilious vomiting and feeding intolerance.⁶ 99% of full term neonates pass meconium within the 48th hr of birth.¹ Hirschsprung disease is caused by abnormal innervation of the bowel, which starts at the internal anal sphincter and extends proximally to involve a variable length of the colon¹. It results when there is failure of migration of neural crest cells from the proximal to distal bowel. This results in absence of both the Meissner and Auerbach myenteric plexus,^{1,4} thus on histology an absence of ganglion cells and hypertrophy of nerve bundles with high concentration of acetylcholine can be seen. The aganglionic segment is narrowed and there is dilatation of the normal colon proximal to it.⁵ The entire colon is aganglionic in 8% of patients.

Several genes are known to be involved in Hirschsprung disease development, with the tyrosine kinase receptor RET and EDNRB being the main ones.² The disease may be associated with other congenital anomalies and syndromes such as trisomy.

Continued on page 2

Noise Management in the Neonatal Intensive Care Unit

Historically, Neonatal Intensive Care Units (NICUs) have been busy, loud and overly stimulating environments that add further challenges to the premature and critically ill newborns receiving care within them. The negative physiological and behavioral effects of over-stimulation during this crucial time in development have been an ongoing area of research. A study by Long and colleagues (1980) concluded that excessively noisy environments cause agitation and crying in preterm infants which leads to increases in intracranial pressure, heart rate, and respiratory rate as well as decreases in transcutaneous oxygen tension.

High noise levels have also been found to affect the neuroendocrine system. In animal studies, high intensity noise levels have been shown to raise cortisol levels by stimulating the adrenal cortex and sympathetic nervous system (Morris, Philbin, & Bose, 2000). Higher levels of cortisol result in an increased metabolism. This in turn may compromise already weak immune systems by diverting energy from the body's internal defense mechanism to alternate sources leaving the infant victim to environmentally induced sickness (Van Goozen et. al., 2007; Brown, 2009.)

The effect of excessive auditory stimulation on brain development is also an area of high concern. The impact of noise on the developing brain has been identified through animal studies which lend continuous, moderate-intensity noise as a contributor to a primitively organized auditory cortex (Chang & Merzenich, 2003). It is known that sensory development occurs sequentially, with the auditory system developing earlier than many other systems. By the 24th gestational week the peripheral auditory system is equipped to send information to the central nervous system. Preterm birth alters this essential sequence of sensory development by prematurely stimulating systems that would otherwise be protected by the amniotic sac. This disorganization in brain development has also led some to believe that excessive noise in the NICU may be connected to auditory attention and distraction in school aged-children. Gray and Philbin (2004) have hypothesized that distractibility increases with decreased environmental predictability as found in the NICU. Premature infants are particularly vulnerable to these unpredictable conditions.

One of the most significant long-term effects of excessive noise levels in the NICU is sensorineural hearing loss. The American

Continued on page 3

INSIDE THIS ISSUE

1. **Hirschsprung Disease**
Noise Management in the Neonatal Intensive Care Unit
2. **Hirschsprung Disease – continued**
RPC Website – New information added
CONGRATULATIONS – Dr. Heather Brumberg
3. **Welcome Ann Marie Bentsi-Addison, RN, CNM, MSN**
Perinatal Clinical Nurse Specialist/Outreach Coord.
Noise Management in the NICU – continued
4. **Late Preterm Birth, Inductions and C-sections**

Continued from Page 1. *Hirschsprung Disease*

Several genes are known to be involved in Hirschsprung disease development with the tyrosine kinase receptor RET and EDNRB being the main ones.² The disease may be associated with other congenital anomalies and syndromes such as trisomy 21, Smith-Lemli-Opitz, Waardenburg, cartilage-hair hypoplasia and congenital hypoventilation syndromes and urogenital or cardiovascular abnormalities.¹

Other intestinal causes of delayed passage of meconium include intestinal atresia, malrotation, volvulus, meconium ileus due to cystic fibrosis, meconium plug syndrome, anorectal malformation and hypoplastic left colon syndrome.⁶ Systemic causes that may result in delayed passage of meconium are narcotics, electrolyte abnormalities, hypothyroidism and sepsis. It may also be seen in very low birth weight infants, although the disease is unusual in preterm infants.¹⁶ Breast fed infants may not suffer as severe a disease as formula-fed infants.¹ Older children may present with absence of soiling or overflow incontinence, chronic progressive constipation, failure to thrive, fecal impaction, malnutrition and progressive abdominal distension.⁷

Failure to pass stool results in dilatation of the proximal bowel, causing an increase in intraluminal pressure. This decreases the blood flow and causes deterioration of the mucosal barrier. The proliferation of bacteria, which occurs due to stasis, can then result in enterocolitis.¹ Hirschsprung's associated enterocolitis (HAEC) may present with abdominal distension, explosive diarrhea, vomiting, fever, lethargy, rectal bleeding and shock.⁸

Hirschsprung disease should be differentiated from functional constipation in older children with presenting symptoms. On rectal examination, if the ampulla is empty and if the aganglionic segment is short, there may be a gush of flatus and stool as the finger is withdrawn in Hirschsprung disease.⁵

Diagnostic tests for diagnosing Hirschsprung disease include contrast enema, anorectal manometry, full thickness biopsy and rectal suction biopsy.⁶ Imaging may show a dilated proximal colon or small bowel and absence of gas in the pelvic colon. The presence of transition zone between the dilated proximal bowel and the obstructed distal colon may be visible on contrast enema.

In anorectal manometry, with Hirschsprung disease, rectal distension fails to initiate a drop in internal sphincter pressure or there is a paradoxical rise.¹

Rectal biopsy, which shows absence of ganglion cells and presence of acetyl cholinesterase positive hypertrophied nerve fibers, is confirmatory. Suction biopsy should be performed at least 2.5 cm above the anal verge.⁶

The treatment for Hirschsprung disease is surgical. However in patients that present with enterocolitis, aggressive resuscitation, rectal irrigation and antibiotics are used to manage the enterocolitis. The options are to perform a one stage definitive repair or to perform a colostomy and wait till the infant is 6-12 months old to perform the definitive repair. The most commonly used procedures are those described by Swenson and Bill (rectosigmoidectomy), Duhamel (rectorectal-transanal approach) and Soave (endorectal procedure).⁶

Postoperative problems include recurrent enterocolitis, stricture, prolapse, parianal abscesses and fecal soiling.¹ Postoperative obstruction occurs from retention of distal aganglionic

segment or from destruction of ganglion cells due to vascular impairment. Pseudo-obstruction syndrome may result from neuronal dysplasia.⁹

Most children with this disease go on to live normal lives. Hirschsprung disease can lead to malnutrition and weight loss, mainly in the young. The colon absorbs much of the water and salt the body needs, and shortening of the colon during surgery may not deliver all the fluids and nutrients the child needs.

At the MFCH, we diagnose, treat, and provide long term follow up through our G.I. and Pediatrics surgical divisions or departments for patients with Hirschsprung's disease (866-WMC-PEDS). Organizations that can provide more information and support include International Foundation for Functional Gastrointestinal Disorders (www.iffgd.org), March of Dimes (www.marchofdimes.com), National Digestive Diseases Information Clearing House (www.digestive.niddk.nih.gov) and North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (naspgan@naspgan.org). www.naspgan.org.

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Regional Perinatal Center Website

www.worldclassmedicine.com/rpc

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CONGRATULATIONS!

Dr. Heather Brumberg

We are pleased to announce the election of Dr. Heather Lynn Brumberg, MD, MPH to membership in the Society of Pediatric Research.

On May 1st, 2010 at the Society for Pediatric Research annual meeting in Vancouver, Canada, Dr. Brumberg will be inducted into the Society at the Pediatric Academic Societies' Presidential Reception honoring the newly elected members.



Welcome...

The Regional Perinatal Center at Westchester Medical Center welcomes Ann Marie Bentsi-Addison into the role of Perinatal Clinical Nurse Specialist/Outreach Coordinator. She comes to us with a vast amount of experience running the gamut of Maternal Child Health. After a year of

Medical-Surgical Nursing, Ann Marie spent ten years in the NICU at New York Hospital/Cornell Medical Center. She has also spent two years in the step-down nursery at Northern Westchester Hospital. She has practiced the full-scope of midwifery ranging from preconception and infertility evaluations, to surgical first assist for c/section surgery to the delivery of 2000 babies. Ann Marie was previously the Clinical Nurse Specialist at St. Luke Cornwall for Labor and Delivery/Mother Baby, NICU and Pediatric units and comes to us from her tenure as Clinical Nurse Manager for Ambulatory OB/GYN dept at Mount Sinai Medical Center.

Ann Marie is a proud wife, mother of three children, and Pastor of Kairos Mission Church in Port Jervis, New York. The Regional Perinatal Center is excited to have Ann Marie as a member of the RPC team at WMC and in partnering with her as she utilizes her experience and expertise to help affect evidence based change in our perinatal community.

Ann Marie Bentsi-Addison, RN, CNM, MSN

Perinatal Clinical Nurse Specialist/Outreach Coordinator

phone: 914 493 2887 fax: 914 493 2891

email: addisona@wcmc.com

Continued from page 1. Noise in the NICU

Academy of Pediatrics (AAP-1997) cautions that excessive noise in the NICU may result in permanent hearing deficiencies ranging in severity from mild to profound. McFadden and Pittman (2008) documented that children with minimal hearing loss have significantly poorer word categorization skills in noise when compared to children with normal hearing. In 1998, Bess and colleagues found that children with minimal sensorineural hearing loss experience more difficulty than children with normal hearing on educational and functional test measures. Furthermore, Moeller et al (2007) reported that even when identified early, children with hearing loss are delayed in consonant and syllable structure development, which influences the rate of word learning.

The emotional development of the neonate may also be affected by excessive noise in the NICU. Noise suppresses certain aspects of speech essential for recognizing and discriminating familiar voices. Segall (1972) has documented that while in a crying state, infants exposed to their mother's voice show significant decreases in heart rate compared to infants exposed to nursery noise. Without the ability to recognize the mother, father, or caretaker's voice, the social growth of the infant may be compromised.

It has been documented that sound levels in NICUs often times

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Continued from previous column – Noise in the NICU

exceed 93 dB. The Occupational Safety and Health Administration (OSHA) limits noise values for industries in the United States. Although there is no such regulation for NICUs, the importance of implementing such a program is highly recognized. The AAP, the Joint Commission on Infant Hearing (JCIH), and the Joint Commission on Accreditation of Healthcare Organizations (JCAHO) are amongst the organizations aware of the demand for implementing a noise management program for NICUs. The AAP and JCAHO recommend maximum noise levels of 45 dB for continuous noise and 65 dB for spontaneous noise (2007).

The education of staff and the modification of environmental aspects leading to excessive noise have been proven effective ways in reducing NICU noise levels, (Brown, 2009). Many hospitals have implemented "quiet time" where talking is reduced to whispering, doctors' rounds are prohibited directly at the bedside, and alarms and crying infants are responded to quickly. Slevin et al (2000) found reductions in infants' diastolic blood pressure, arterial pressure, and overall movement by reducing light, noise, alarm events and unnecessary staff conversation. In addition many hospitals have been using noise-monitoring systems which help to alert staff and visitors at times when noise levels exceed the AAP recommended limit.

Within the coming months, The Regional Neonatal Intensive Care Unit at Maria Ferrari Children's Hospital will be installing noise monitoring systems in each of the 14 patient care rooms. Together with the education of nursing, medical, respiratory, housekeeping and other applicable hospital staff, these monitoring systems will help maintain a healthy environment to promote the physical, emotional, and social development of the NICU population.

Kristi Adamo, AuD, CCC-A

Clinical Audiologist, Westchester Medical Center

adamok@wcmc.com

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Late Preterm Birth, Inductions and C-sections

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- Late preterm births are births that occur at 34 to 36 weeks gestation. Babies born even a few weeks early are at greater risk than full-term infants of having serious problems, such as respiratory distress, delayed brain development, sudden infant death syndrome (SIDS), jaundice, rehospitalization and feeding problems.
- In the United States among singleton preterm births from 1996 to 2004, the largest percentage increase in c-section rates occurred among late preterm births.¹ From 1996 to 2004, c-sections accounted for nearly all — 92 percent — of the increase in U.S. singleton preterm births. In 2005, the c-section rate in the United States was 30 percent of all live births.²
- Good dating of a pregnancy by early ultrasound can help ensure that the decision to induce or to have a c-section is made with the best available information. With careful dating of pregnancies and strict adherence to guidelines from the American College of Obstetricians and Gynecologists (ACOG), the March of Dimes believes we can reduce the number of non-medically indicated inductions and c-sections performed before 39 weeks gestation and, as a result, slow the increase of preterm birth.
- The March of Dimes is asking hospitals and health professionals to implement quality improvement programs that can help prevent unnecessary early (before 39 weeks gestation) inductions and c-sections and follow ACOG guidelines. These guidelines

clearly state that c-sections and inductions should not be done before 39 weeks gestation unless medically necessary. Many chapters are working on this effort with hospitals and organizations in their states.

- A woman may need a c-section because of medical complications including: she has problems with the placenta or an infection that she could pass to her baby during birth; she is pregnant with multiples; her labor is too slow; or the baby is too large, in a breech or transverse position or has problems during labor.
- C-sections can be lifesaving for women and babies, but they also pose real risks, including placental problems and the potential need for a repeat c-section in future pregnancies. Health care providers and women should weigh the medical circumstances and make a decision based on potential benefits and risks to mom and baby.
- Nationally, ACOG has a special program to help hospitals perform a "Voluntary Review of Quality of Care."³ In recent years, the most important observation from these reviews has been the problem of inappropriate early inductions that result in late preterm births.

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State Perinatal Database Team and Perinatal Gazette Editorial Board

Susan Marchwinski, R.N., C., M.S., SPDS Coordinator
Editor in Chief
(914) 493-8590 (marchwinskisa@wcmc.com)

Clare Nugent, R.N., C., BSN
Neonatal Data Collection
(914) 493-8346 (nugentc@wcmc.com)

Heather Brumberg, M.D. M.P.H., FAAP, Director of Neonatal Public Health Programs,
Coordinator, Regional Perinatal Center
(914) 493-8491 (heather_brumberg@nycmc.edu)

Donna Dozor, R.N., M.S.
Neonatal Data Collection
(914) 493-8309 (dozord@wcmc.com)

Edmund LaGamma, M.D., FAAP
Director Newborn Medicine
(914) 493-8558 (edmund_lagama@nycmc.edu)

Howard Blanchette, M.D., FCOG
Professor and Chairman of the Department of Obstetrics and Gynecology
(914) 594-4526 (howard_blanchette@nycmc.edu)



The Regional Perinatal Center
Maria Fareri Children's Hospital
At Westchester Medical Center
95 Grasslands Road
Valhalla, New York 10595

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