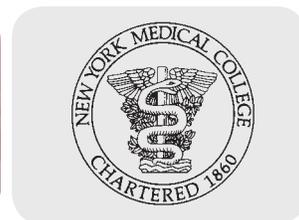


# The Perinatal Gazette

Newsletter of the Regional Perinatal Center Maria Fareri Children's Hospital at Westchester Medical Center



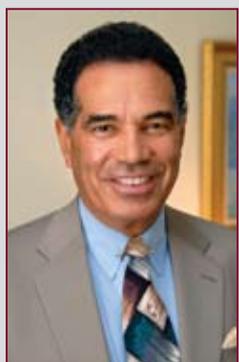
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## Warm Welcome for Newborns

### A new labor-and-delivery suite makes childbirth more family-friendly

Having a baby is one of a family's most precious moments — even when there are special medical challenges. Soon Westchester Medical Center childbirth facility will combine



the most advanced clinical resources with an atmosphere of warmth and comfort that honors that joyful family experience.

"We like to say we're going to have high tech and high touch," says Howard A. Blanchette, M.D., Director of Obstetrics and Gynecology.

Westchester Medical Center has long been the top referral center for high-risk births in the Lower Hudson

Valley. In April, when its renovated Labor and Delivery Center is expected to open, it will be known as a family-friendly place as well.

"We'll still maintain the high-tech expertise to take care of the sickest mothers in the counties we serve," says Dr. Blanchette. "But now our suite will also meet the emotional and family needs of women giving birth."

Before the renovation, there were six rooms for mothers to  
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## Congenital Diaphragmatic Hernia

Boriana Parvez, M.D. Jonathan Blau, M.D.

Congenital diaphragmatic hernia (CDH) is a developmental defect in the fetal diaphragm, leading to herniation of abdominal contents into the thorax. Significant pulmonary, cardiac, and gastrointestinal sequelae result. This is a relatively common congenital anomaly, affecting between 1 in 2000 to 1 in 5000 births. There are several subtypes of CDH. Bilateral CDH is uncommon, and thought to be lethal as most cases are diagnosed after stillbirth. Left-sided CDH is more common, approximately 80% of all cases, and is associated with a better prognosis than right-sided CDH.

Formation of CDH occurs during a specific period in the embryologic development of the fetus. During the fourth week of gestation, a lung bud develops off the caudal end of the foregut, giving rise to development of the bronchi and lungs. During the sixth week, pleuroperitoneal membranes fuse with the other diaphragmatic components. If this fusion does not occur by the tenth week, during which abdominal viscera travel from the umbilical cord into the abdomen, these abdominal contents may herniate into the thorax. In approximately 95% of cases, herniation of abdominal contents occurs through a posterolateral defect (foramen of Bochdalek). Retrosternal herniation occurs in the remaining 5% of cases through the foramen of Morgagni.<sup>1</sup>

The pathogenesis of CDH continues to be debated. Classically, the constellation of the pathological findings and clinical presentation was considered to be related to the primary diaphragmatic defect, leading to ipsilateral lung hypoplasia from compression of the herniated abdominal contents. More recently, it is hypothesized that primary pulmonary hypoplasia leads to incomplete closure of the diaphragm.<sup>2</sup> Studies have revealed that the non-hypoplastic lungs in unilateral CDH have surfactant deficiency and decreased alveolar number. Further, a dual-hit hypothesis also exists, implicating both pulmonary and diaphragmatic pathology in the development of CDH.<sup>3</sup>

As much as 40% percent of CDH patients have co-existing genetic syndromes, congenital malformations and chromosomal anomalies. Many of these disorders are ultimately lethal conditions or disorders that are associated with severe developmental delay. Amniocentesis with high

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labor in, but only two delivery rooms. “The suite was set up 1960s-style—the women labored in one room, then delivered in another,” says Dr. Blanchette.

The redesigned unit has six labor-and-delivery rooms large enough to welcome the father, partner or other friends or relatives. The rooms feature softer color schemes, indirect lighting and comfortable furniture. “They have the feel of a nice hotel,” Dr. Blanchette says.

There will also be two operating rooms in which doctors can perform cesarean sections or handle unexpected complications. Also, Dr. Blanchette has expanded the hospital-based practice, Advanced OB/GYN Associates, to 10 physicians, including specialists in several newly emerging fields.

One of these fields is oncofertility, which helps young women undergoing chemotherapy maintain their fertility. Kutluk Oktay, M.D., an expert in ovarian transplants, recently removed one ovary from a 15-year-old girl now in chemotherapy and froze it. If the chemo makes her remaining ovary sterile, Dr. Oktay can re-implant the healthy one so she’ll still be able to have children someday.

Another emerging field is urogynecology, or women’s urology. Huan-Sue Zhou, M.D., treats such conditions as uterine prolapse (fallen uterus) and urine leakage. She and the other surgeons in the practice offer minimally invasive procedures for hysterectomies and other surgeries. And surgeons will soon be using robotic technology to perform minimally invasive procedures to treat even more conditions, such as endometrial cancer.

“We can cover every specialty in ob/gyn care,” Dr. Blanchette says. “You don’t need to go into the city, as all the expertise you could ever need is right here. Our motto is, ‘You don’t have to commute to deliver.’”



**To learn more about Westchester Medical Center’s New Labor and Delivery Center and Advanced OB/GYN Associates, call 1-877-WMC-DOCS or visit [www.WorldClassMedicine.com](http://www.WorldClassMedicine.com).**

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resolution banding must be offered to parents of all fetuses diagnosed in utero with CDH. High-resolution ultrasound in the first trimester can be used to determine the extent of the associated anomalies. If the ultrasound suggests CDH, antenatal fetal lung MRI may be an important tool in providing prognostic information. Most cases of CDH in the United States are diagnosed prenatally with the advances of imaging during prenatal care. If amniocentesis was not performed antenatally, then high-resolution banding chromosomal analysis must be performed postnatally with Genetics consultation. Parents must be educated about possible associated conditions that may not be diagnosed by physical examination alone.

The clinical presentation of CDH involves respiratory distress at birth: cyanosis, tachypnea, grunting, and retractions. On physical exam, one can often appreciate a scaphoid abdomen, mediastinal shift of heart sounds, absent breath sounds on the affected side, and the presence of bowel sounds in the chest. Delivery room resuscitation of the infant with CDH differs from NRP guidelines. If the neonate is depressed, immediate intubation is required. Bag-mask ventilation must be avoided to prevent distention of abdominal contents in the thorax and worsening of already compromised lung function. Nasogastric tube placement and gastric decompression in the DR is also recommended.

Major immediate sequelae of CDH are pulmonary hypertension resulting from lung hypoplasia, increased medial thickness of pulmonary arteries, and blunted oxygen related vasodilatation. Historically, treatment of CDH included emergent surgical repair and aggressive control of PPHN. However, improved outcomes have resulted with gentler ventilation: practicing of permissive hypercapnia, avoiding high ventilation pressure, and delayed surgical repair until the patient is more stable.<sup>4</sup> Studies have shown that CDH patients who require less ventilatory support have improved mortality. The degree of ventilatory support helps neonatologists predict which patients are likely to survive surgical repair.<sup>5</sup> Patients with a smaller pre- and post-ductal differential, a marker of pulmonary hypertension, also have improved mortality. Ultimate survival depends on the degree of pulmonary hypoplasia and pulmonary hypertension.

Adjunctive therapies have included surfactant, nitric oxide, and ECMO. Surfactant therapy was used since animals with CDH were found to have surfactant deficiency. Recent reviews have found surfactant therapy is associated with increased mortality and is therefore no longer widely used. Nitric oxide, a potent vasodilator, has been used to control pulmonary hypertension. Although NO does not improve mortality, it does decrease the need for ECMO in infants with CDH. ECMO has also been found to decrease mortality in CDH. A multi-disciplinary team of pediatricians, pulmonologists, and other sub-specialists are necessary to optimize care.

Despite these advances in therapy, significant morbidity and mortality exists. A permissive hypercapnia and delayed surgical repair strategy has led to survival rates approaching 90%. CDH is often noted on autopsies of stillbirths. Therefore these 90% may represent a healthier sub-group of patients who were able to survive pulmonary hypertension and surgical repair.

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# WANTED!

## Perinatal Clinical Nurse Specialist

Westchester Medical Center Perinatal service is **seeking a highly motivated Registered Nurse** to fill the position of Perinatal Clinical Nurse Specialist. This position offers an exciting opportunity to join a dynamic, rapidly expanding high-risk obstetrical division offering world-class obstetrical medicine.

### Responsibilities Include:

- Serving as a clinical resource for L&D and Mother/baby staff focused on high-risk perinatal nursing, fetal monitoring, and well baby care.
- Developing & implementing educational programs on high-risk obstetrical and well baby nursing.
- Maintaining & improving communication between the Regional Perinatal Center (RPC) and its affiliate obstetrical services within our seven county region.
- Working collaboratively with perinatal personnel and the Regional Perinatal Center team relating to maternal transport quality improvement issues.

**Qualifications:** Masters prepared with minimum five years recent acute care obstetrical experience; Certification in Inpatient Obstetrical nursing preferred. Preference will be given to candidates with outreach experience.

**Please contact: Rose Cordella at 914-493-7808**

**\*New\***

## Regional Perinatal Center Website

[www.WorldClassMedicine.com/rpc](http://www.WorldClassMedicine.com/rpc)

What is an RPC? Who is the team? What do we do?  
Who are our affiliates? Interested in a mini-grant?

Please visit our website for answers to these questions and for more information about the RPC at Westchester Medical Center.

## Cool Cap<sup>®</sup> References

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- Shankaran S et al. for NICHD Research Network. Whole body hypothermia for neonates with hypoxic ischemic encephalopathy. *N Eng J Med*. 2005;353(15):1574-84.
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In conclusion, CDH is a relatively common congenital anomaly caused by an underlying developmental problem of the lungs and diaphragm. Most cases are diagnosed prenatally and CDH infants should be delivered in a center well-equipped to treat a critically ill neonate who is likely to develop pulmonary hypertension. Permissive hypercapnea and delayed surgical repair have resulted in improved mortality, but significant morbidity exists in survivors.<sup>6</sup>

Our Regional NICU offers ECMO, NO, and a comprehensive program to treat neonates with this disorder.

### References:

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7. Haleem et al. Can bedside Lung Mechanics Identify the Ability to Survive in near Terminal Lung Disease? *E-PAS2005* 3353

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## HELPING NICU FAMILIES IN NEED

Hailey's Hope Foundation is a family-based, non-profit organization dedicated to helping families in need with premature and seriously ill babies in the NICU.

The Foundation was established in loving memory of Hailey Zion, the infant daughter of Isaac and Donna Zion, who was born prematurely at 21 weeks and passed away shortly after birth. Together, our members have overcome difficult NICU experiences, including the tragic loss of Hailey. We are incredibly thankful for all the love and support we received during these very difficult times and we want to be there for other families.

In just one short year, we have provided financial support and resources to over 100 NICU families at the Maria Fareri Children's Hospital at Westchester Medical Center. Through our financial support program, we have helped these families pay for daily, non-medical expenses associated with a NICU hospitalization, including lodging, transportation and food.

Hailey's Hope Foundation relies on the generosity of donors and volunteers. If you would like to donate, volunteer, or find out more information about us, please visit our website at [www.haileyshopefoundation.org](http://www.haileyshopefoundation.org) or contact us at (845) 837-1182.

# Selective Head Cooling for Acute Hypoxic Ischemic Encephalopathy

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Perinatal asphyxia occurs in approximately 2-3/1000 births and may account for up to 30% of cases of cerebral palsy. Currently, there is no specific treatment for acute perinatal hypoxic-ischemic encephalopathy, except for supportive care and pharmacological treatment of seizures. A new treatment modality has recently been approved by the FDA for babies at risk for developing this condition-selective head cooling with the Cool Cap®.

The Cool Cap® covers the scalp and contains tubing connected to a refrigerated cooler, which circulates cold fluid of about 10-11 °C (50-51.8 °F). While the head is being selectively cooled, the core body temperature is carefully maintained between 34-35 °C (93.2-95 °F). Randomized controlled trials in full-term newborns suggest that treatment with mild hypothermia is effective and safe and may improve survival without disabilities up to 18 months of age, especially in newborns suffering from moderate encephalopathy.

The goal of selective head cooling is to preserve cerebral energy metabolism, reduce cerebral tissue injury and improve neurological function. A cascade of reactions follows hypoxia-ischemia. There may be primary energy failure of the brain, which could be so severe that permanent brain injury results. Alternatively, resuscitation may be successful so that limited or no injury results. Finally, limited primary energy failure may be followed by a latent phase, during which oxidative metabolism has normalized, but in which there is hyperactivity of the glutaminergic receptors (excitotoxicity), and the intracytoplasmic components of the apoptotic cascade are activated resulting in the initiation of a secondary inflammatory reaction. This may be followed by subsequent deterioration, which leads to delayed neuronal death after 72 hours. Animal studies have shown that there is a potential therapeutic window of six hours to initiate head cooling following hypoxia-ischemia, after which little therapeutic gains are realized. The head cooling is continued for 72 hours, with the goal of decreasing secondary deterioration and limiting the subsequent neuronal cell death.

## ELIGIBILITY

Three criteria all need to be satisfied (A, B, and C) to qualify for head cooling:

### A. Infants >36 weeks gestation and at least one of the following:

- Apgar score  $\leq$  5 at 10 minutes
- Continued need for resuscitation, including endotracheal or mask ventilation, at 10 minutes after birth
- Acidosis defined as either umbilical cord pH or any arterial pH  $<$  7.00 within 60 minutes of birth
- Base deficit  $\geq$  16 mmol/L in umbilical cord blood sample or any blood sample within 60 minutes of birth (arterial or venous blood)

### B. Moderate to severe encephalopathy, consisting of altered state of consciousness (lethargy, stupor, or coma) and at least one of the following:

- Hypotonia
- Abnormal reflexes including oculomotor or pupillary abnormalities
- Absent or weak suck
- Clinical seizures

### C. Infant has an amplitude-integrated encephalogram / cerebral function monitor (aEEG/CFM) recording of at least 20 minutes duration that shows either moderately/severely abnormal aEEG background activity or seizure.

#### Exclusion criteria:

- Infants > six hours of age
- Imperforate anus
- Evidence of head trauma or skull fracture causing major intracranial hemorrhage
- Birth weight  $<$  1,800 grams

If there is a possible brain insult at birth, provide basic cardio respiratory support and maintain rectal temperature between 34-35 °C by turning off the radiant warmer, and applying ice packs (in plastic bags) to the sides of the head and the chest wall. Call the Westchester Medical Center Transfer Center as soon as possible at 866-468-6962. Discuss selective head cooling with the family, but make no promises regarding the use of cooling and the outcome of the baby. These are often the sickest infants, with multi-system involvement. Most importantly, initiation of selective head cooling must begin within six hours following hypoxia-ischemia, which may have started in utero. This requires prompt notification whenever hypoxia-ischemia is suspected, whether or not full manifestations are present, and even when this scenario is expected prior to delivery. The sooner the call is made, the quicker transport can be arranged. (See page 3 for references)

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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 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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