Cystic Hygroma & Potential Airway Obstruction in a Newborn: A Case Report & Review of the Literature

Cystic Hygroma is a benign congenital malformation of the lymphatic system that has its genesis in the lack of development of communication between the lymphatic and venous systems. The cyst may be unilocular or multilocular and could be of variable size but it is characteristically brilliantly transilluminant. The incidence of cystic hygroma is approximately 1/6000 live births. 70-80% of cystic hygromas occur in the neck, usually in the posterior cervical triangle. The remainder 20-30% occurs in the axilla, superior mediastinum, chest wall, mesentry, retroperitoneal region, pelvis and lower limbs. Cystic hygroma is known to present at birth in about 50% of the affected newborns and 90% present by age 2 years. We present the case of a right neck mass which was initially asymptomatic but eventually caused stridor, and vomiting episodes after feeding.

Case Report:

A 3.3 kg full term infant was delivered by C/S secondary to fetal bradycardia and suspected macrosomia to a 37 y.o. G8P5 mother. The pregnancy was complicated by gestational diabetes mellitus & pregnancy-induced hypertension. Apgars were 8 & 9 at 1 and 5 minutes respectively. In the well baby nursery a right neck mass was noted. Due to a history of spitting up regular infant formula, soy based formula was started, but without improvement. The infant was transferred to the Regional Neonatal Center for further evaluation and management. The baby was on room air for one week when she developed stridor. The neck mass had increased in size and a pediatric ENT consult was obtained. An MRI demonstrated “a cystic...” Continued page 2.

Is Proximity to a Nuclear Power Plant Associated with Increased Rates of Congenital Malformations?

Recent radiation leaks at the Indian Point nuclear power plant have raised public concerns as to the safety of nuclear power plants and the potential consequences of exposure to ionizing radiation. Currently there are 103 active nuclear power plants in the United States. The Indian Point nuclear power plant located in Buchanan, New York has two operating units since 1976. Approximately 20 million people live within the 50-mile exposure zone of Indian point.

Animal studies have shown increases in birth defects, such as central nervous system abnormalities and growth retardation after ionizing radiation exposure. However, the effects of chronic low dose ionizing radiation exposure in humans are unclear. Most of the human data from ionizing are from Japanese atomic bomb and Chernobyl survivors. In Japan there were increased prevalence of mental retardation, microcephaly, and childhood leukemia following the atomic bomb fallout. Increased incidence of thyroid cancer was seen in surrounding populations after exposure to the nuclear plant meltdown in Chernobyl.

There have been very few studies on the associated effects of chronic low dose ionizing radiation exposure on individuals living near nuclear plants. Our study reported the rates of congenital malformations surrounding Indian Point nuclear power plant over a decade.

This epidemiologic ecologic study utilized aggregate data from the New York State Department of Health Vital Statistics and Congenital Malformations Registry to examine birth defects of children whose maternal residence was within a 20 mile radius of Indian Point from 1992 to 2001. Proximity to the nuclear power plant was used as a surrogate measure of exposure to ionizing radiation. The number of cases of congenital cancers, congenital anomalies, genetic syndromes, low birth weight (<2500g), and Prematurity (<37 weeks) were recorded. The congenital cancer category included leukemia, lymphoma and central nervous system tumors. The congenital anomalies category included neural tube defects, microcephaly, cleft lip, cleft palate, abdominal wall defects and hypothyroidism. The genetic syndromes category included trisomy 13, 18, & 21.

Over the 10 year period 702 malformations in 666 children were identified from a birth population of 328,124; yielding a rate of 2.1 malformations per 1,000 births in the Hudson Valley. This rate was lower than the... Continued page 2

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Indicated in a few asymptomatic cases as spontaneous regression. Indications for surgery in pediatric cases include significant cosmetic deformity, obstructive symptoms, bleeding and recurrent infections. Other treatment modalities include aspiration, radiation, and injection of sclerosing agents, in particular the agent OK-432, derived from a strain of streptococcus pyogenes, which has been used successfully, especially in macracystic lymphangiomas and in patients who are at increased anesthetic risk. It is not uncommon for infants to develop neural paresis or paralysis after excision of massive cervical lymphangiomas. These congenital abnormalities tend to distort normal anatomy. They may surround or displace neurovascular structures making their identification quite challenging intraoperatively. A team of experienced surgeons from varying fields including Otolaryngology, Cardiothoracic and Pediatric Surgery will help to ensure a successful surgical outcome postoperatively. Endocrinology may need to be involved to monitor surgery-related endocrine dysfunctions.

Sulaiman Sannoh, MD
Division of Newborn Medicine,
Maria Fareri Children’s Hospital at Westchester Medical Center

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NY State rate of 5.9 malformations per 1,000 births. The prevalence of defects, prematurity and low birth weight were not significantly related to proximity to the nuclear plant. We concluded that the congenital malformations identified in the areas surrounding Indian Point nuclear power plant did not substantiate an association between proximity of the reactor and the occurrence of birth defects. This study provides baseline population data for comparison of the occurrence of malformations in the event of a nuclear plant accident.

Tania Mangones, MD
Division of Newborn Medicine,
Maria Fareri Children’s Hospital at Westchester Medical Center

Web-Site Resources for Healthy Living

- www.mayoclinic.com – Healthy Lifestyle Planners
- www.foodtv.com – The Food Network has a fun website for those who love to cook.
- www.ars.usda.gov/main/site_main.htm?modecode=12354500 - On the USDA Nutrient Data Laboratory site, the nutrient composition of virtually any food can be found
- www.nal.usda.gov/fnic/Fpyr/pyramid.gif – These sites allow access to images of the USDA Food Guide Pyramid
- www.deliciousdecisions.org – This site, maintained by the American Heart Association, provides a wealth of information, including detailed recipes
- www.noah-health.org/en/search/health.html - The New York Online Access to Health (NOAH) website provides health information in both English and Spanish
- http://vm.cfsan.fda.gov/~dms/wh-nutr.html – Essays on topics in health by the US Food & Drug Administration's Center for Food Safety & Applied Nutrition

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hygroma in the nasopharynx and right neck which inferiorly followed the course of the right carotid artery probably consistent with cystic hygroma.” The mass continued to increase in size so a follow-up MRI and MRA were done. MRI showed increase in size of right posterior retropharyngeal to right neck cystic mass measuring 4.1 cm and displacing the trachea and oropharynx to the left and right carotid artery posteriorly and laterally. MRA showed the right carotid artery displaced posteriorly and laterally without definite homodynamic stenosis. Carotid duplex ultrasound was recommended, which was severely limited due to displacement of vessels, presence of the mass and small size of the neck. To reduce fluid buildup in the neck mass, the patient was started on dexamethasone after the first MRI. She was later intubated for stridor and concern for potential airway obstruction. A team of pediatric otolaryngologists and pediatric surgeons was assembled to address the mass. The otolaryngologist decided to approach the mass via a cervical incision. Imaging studies demonstrated that the mass extended inferiorly to the aortic arch and a sternotomy had been planned. Intraoperatively the mass was found to be limited to the neck, so a sternotomy was not necessary. A hemiresectiondectomy was required when the cystic hygroma was found to be inseparable from the right half of the thyroid. Dexamethasone was tapered over 2 weeks. The infant was observed over 2 weeks and for 24 hrs. after the steroid taper. Endocrinology was consulted after the hemiresectiondectomy. Initial thyroid function tests were normal except for mildly elevated T4. PTH was also normal. Genetics was consulted and chromosomal analysis showed normal karotype. The patient was noticed postoperatively to have a weakness of the lower right lip, consistent with neuropaxia involving the marginal mandibular branch of the facial nerve. This deficit is expected to correct with time.

Discussion: We report an unusual case of neonatal neck mass. The mass was investigated with US, MRI/MRA and found to be a cystic hygroma of the neck and mediastinum. Cervical cystic hygromas or lymphangiomas are believed to occur as a result of the failure of establishment of the appropriate connection to the normally present lymphatic channels. They are usually encountered at birth or in early infancy. Very few hygromas extend into the mediastinum. Lymphangiomas may be divided histologically into two major groups based on the depth and size of abnormal lymph vessels. The superficial ones are called lymphangioma circumscriptum. The more deep seated ones are cavernous lymphangioma or cystic hygroma. Cystic hygromas are deeply seated in areas of areola or loose connective tissues. They appear early in life as large soft-tissue mass on the axilla, neck or groin. They are soft, vary in size and shape, and tend to grow extensively if not surgically excised. They are multilocular cysts filled with clear or yellow lymph fluid. Usually cystic hygromas are diagnosed clinically with large size, location and translucence. Although cystic hygromas tend to enlarge progressively over months a relatively rapid increase in size has also been described. Cystic hygromas may be associated with aneuploidy, cardiac anomalies, and fetal hydrops. The management of lymphangiomas including cystic hygromas is preferably surgical, although a careful “wait & see” policy may be
The Annual Regional Perinatal Forum Conference entitled; *The Interplay of Stressors in Perinatal Health: Racism, Obesity and Violence* was held this year at the Westchester Marriott in Tarrytown, New York on November 1, 2006. It was sponsored by The Maria Fareri Children's Hospital at Westchester Medical Center, the March of Dimes, Maternal Infant Services Network and the Lower Hudson Valley Perinatal Network.

The conference drew over 240 participants from our 7 county regions and beyond to hear from our distinguished, internationally-renowned speakers which included the former United States Surgeon General David Satcher, MD, MPH, PhD. After a warm welcome from Dr. Michael Gewitz, Director of the Maria Fareri Children's Hospital followed by an overview on the state of the Regional Perinatal Center and Regional Perinatal Forum Dr. Satcher delivered an eloquent keynote address on the Elimination of Disparities in Health or Obesity: A National Epidemic. Dr. Camara Jones, MPH, PhD then presented an in-depth view on the Social Determinants of Health and Equity: The Impact of Racism on Health followed by Janet Rich-Edwards, SCD, MPH who spoke on Racism and Maternal Health: Implications for Perinatal Outcomes. Further details to follow in the next edition of The Perinatal Gazette.

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**Regional Perinatal Center (RPC)**

- Maternal Fetal Medicine (MFM) Outreach Coordinator: (20 hrs./week)
  - A Registered Nurse or similarly skilled individual who is responsible to help improve communication between Regional Perinatal Center (RPC) and affiliate obstetrical services through on-site visits, phone & e-mail communications.
  - Will act as an educational resource person assessing affiliate educational needs and assisting affiliates with developing and implementing a plan to meet these needs. (e.g. providing & coordinating educational in-services)
  - Will work collaboratively with the RPC Neonatal Intensive Care (NICU) personnel in reviewing quality issues related to maternal transports; maternal mortality and assessing affiliate and RPC quality improvement activities related to issues of high risk pregnancies/maternal transports.
  - Collection/analysis of regional maternal statistical data.
  - Participation in RPC quarterly Quality Assurance (QA) quadrant meetings and the Regional Perinatal Forum. (RPF)

**TO APPLY:** Please E-mail resume/cover letter to heather_brumberg@nymc.edu, or marchwinski@wcmc.com

**The Lower Hudson Valley Perinatal Network – LHVPN**

- Comprehensive Prenatal-Perinatal Services Network Health Education Specialist: (Full-time position available October 2006)
  - The Health Education Specialist provides expertise in planning and implementing provider and consumer health education and programming, to carry out the mission of the Lower Hudson Valley Perinatal Network and fulfill the work plan requirements of the Comprehensive Prenatal-Perinatal Services Network (CPPSN) contract and other contracts as assigned. The mission of the LHVPN’s CPPSN program is to improve perinatal health outcomes in the Lower Hudson Valley region. The program works to organize perinatal health care information, educate providers and consumers and to facilitate access to available maternal and child health care system at the local level.

**TO APPLY:** Please send resume/cover letter, salary to Cheryl Hunter-Grant, Executive Director, LHVPN, MFCH at WMC, 1 Woods Road, Valhalla NY 10595 or E-mail Hunter-grante@wcmc.com

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**The Maternal-Infant Services Perinatal Network – MISN News**

- The Maternal-Infant Services Perinatal Network (MISN) which covers Orange Ulster and Sullivan Counties is continually striving to improve perinatal services and outcomes in their communities. One of the ways they serve their regions is through community baby showers which are a means to draw pregnant and newly post-partum moms to a setting where immediate and long-term needs can be met through education and provision of available services and tangible items to assist them in caring for their infants. The goal is to improve birth outcomes and encourage healthy lifestyle living. Previous showers were held in Ellenville (Ulster County), Grahamsville (Sullivan County), Liberty (Sullivan County) and Newburgh (Orange County). Upcoming baby showers are scheduled for the spring of 2007 in Port Jervis (Orange County), Middletown (Orange County) and Newburgh (Orange County).
  - MISN also translates all community shower baby announcements in Spanish and has translated and printed the prenatal breastfeeding education brochure distributed at the showers and elsewhere into Spanish.
  - The breastfeeding committee has produced, printed & distributed over 10,000 copies of a breastfeeding/referral brochure through funding from the March of Dimes. They have developed a mother-based survey to help determine types of breastfeeding resources & services offered to families of newborns.
  - MISN continues to hold “Lactation Live Chats” quarterly. These are roundtable discussions among lactation service providers to encourage breastfeeding best practices.
  - Upcoming for 2007 is an exciting conference sponsored by the collaboration of the two existing lactation consortiums. Further details to be announced in upcoming issues of the *Perinatal Gazette*.

For questions or further information on any of MISN’s activities please contact: Stephanie Sosnowski, 845-928-7448, Stephahnie@misen.us
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