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OFFICE OF THE CHIEF RESIDENTS,
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EDITED BY:
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Pediatric Residency Newsletter

Maria Fareri Children's Hospital

Welcome to the First edition of the PRN. This newsletter will be published monthly as a method of keeping all residents and our medical community up to date on important topics discussed during the previous month at MFCH.

VOLUME 1, ISSUE 1 NOVEMBER 2007

Grand Rounds

November 7
"Use of Sports Supplements in Children"
Dr. A. Gagliardi

November 14
"Feeding, Flora and Infant Development"
Dr. E. LaGamma

November 21
"Neonatal Crohn's"
Case Conference
Multidisciplinary

November 28
Pediatric Dermatologic Emergencies
Dr. A. Schliftman

Resident As Teacher

November 7
"Bleeding & Thrombotic Disorders"
Rebekka Levis

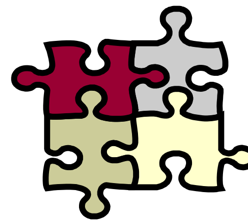
Journal Club

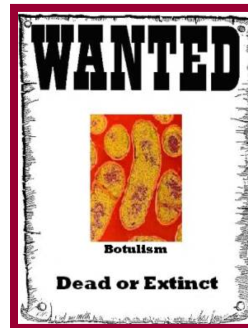
November 8
"High dose Ibuprofen in Cystic Fibrosis"
Pediatrics Sept 2007
Ramneet Gill

Baby BIG	1
Biliary Atresia	1
Picture of the Month	2
Back to the Basics	2
Inside the ICU	2
Bug of the Month	3
Director's Corner	3
Ripped from the	3
Puzzle of the Month	4
New Staff	4

Baby BIG - A case of Infant Botulism

KF is a 3mth old previously healthy female who presented to an outside hospital with 2 days of congestion, constipation and poor feeding. During evaluation, she was noted to have progressive weakness and lethargy, and was transferred to MFCH. Routine blood work and initial radiologic studies were normal. The patient's neurologic condition continued to worsen resulting in intubation on HD# 3, leading physicians to pursue the diagnosis of botulism. Baby Botulism Immunoglobulin (BIG) was administered on HD #4 with clinical improvement noted on HD#5. The diagnosis of infantile botulism was confirmed via PCR on stool for botulinum toxin type B on HD # 7.





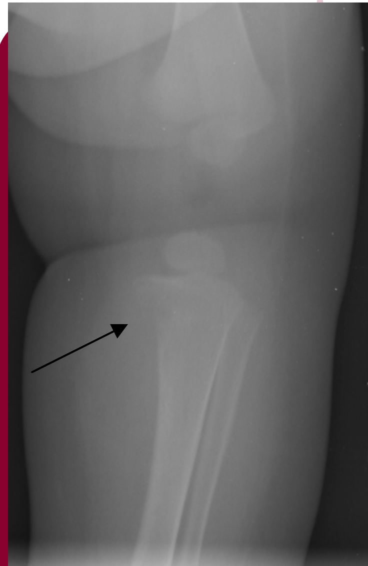
The patient was intubated for a total of 9 days, and slowly regained muscle use. She was discharged home on HD #23 with NGT feeds.

Infantile Botulism is caused by consumption of spores of botulinum bacteria which germinate in the intestines and release toxin that blocks acetylcholine at the neuromuscular junction, leading to paralysis and respiratory failure. Infant botulism accounts for 72% of all botulism cases in the US and infants present with lethargy, poor feeding, constipation, associated with weak cry and poor muscle tone. Care is mostly supportive and Baby BiG has been approved for use in infants under the age of one.

When Jaundice isn't just looking a little yellow... a case of Biliary Atresia

LM presented to MFCH at 5 weeks of life with persistently elevated bilirubin and relapsing/ recurring jaundice. Initially she had presented at 36 hours of life, with resolution of jaundice with observation. Subsequently, she was followed by her pediatrician who noted persistent elevation of T. Bili (25) with associated elevated LFT's, white acholic stools and dark urine. US of abdomen on admission demonstrated absence of a gallbladder and HIDA scan was inconclusive. Therefore, patient had intraoperative cholangiogram with liver biopsy confirming diagnosis of biliary atresia. She was discharged on Actigall and Zantac, and readmitted for elective Kasai Procedure. Postoperatively the patient demonstrated improvement in stool color. However, her post operative period was complicated by significant electrolyte abnormalities, liver failure, ascites and pseudomonas peritonitis. She is scheduled for a liver transplant from a donor portion of her father's liver.

Infants with biliary atresia typically present with icteric sclera and/or jaundice, hepatospenomegaly, and acholic stools. Laboratory tests demonstrate elevated bilirubin and serum aminotranferases. Diagnosis is made on basis of absence of gallbladder on Ultrasound, HIDA demonstrating lack of tracer excretion into small bowel from liver, and finally laparoscopy and intraoperative cholangiogram. Early diagnosis is critical because success of surgical intervention in establishing bile flow, including Kasai Procedure (hepatoportoenterostomy) is diminished after 8 weeks of age. If diagnosis is delayed, Kasai procedure may be successful at re-establishing bile flow, however, liver damage may be too severe for adequate function. Liver transplantation becomes necessary in these cases.



Picture of the Month - Osteomyelitis by Drs. Levis and Mendelsohn

CC, a 9 wk old female born via NSVD was healthy until 3 days PTA when developed fever Tm 102 F without associated symptoms. Patient was seen by PMD who sent blood cultures. On repeat examination on date of admission patient noted to be unwilling to move left leg and previous blood culture positive for Staph Aureus. Admission physical at MFCH was unremarkable except for local tenderness to palpation over the left proximal tibia. An x-ray demonstrated an area of bony destruction in the medial aspect of the proximal tibia consistent with osteomyelitis. She was admitted for prolonged IV antibiotics and her condition improved.

The most common cause of osteomyelitis in children is hematogenous spread with seeding to the bone; Other causes include direct inoculation and local invasion from a contiguous infection. In neonates, the most common organisms causing infection are group B streptococci and *Escherichia coli.* A septic joint is involved in about 50% of cases among neonates. Beyond the neonatal period, the most common organisms causing osteomyelitis are *Staphylococcus aureus*, group A streptococci, and *Haemophilus influenzae* type b. *Pseudomonas aeruginosa* is associated with osteomyelitis and osteochondritis following penetrating wounds of the foot through a tennis shoe. Hematogenous osteomyelitis usually presents acutely with pain and decreased movement of the affected limb and adjacent joint, possibly with swelling or redness over the infected area. Systemic signs (including fever, malaise, and irritability) and/or leukocytosis may or may not be present. CRP and ESR are usually elevated; Blood cultures are positive in 50% of cases. Treat with prolonged course (4 - 6 weeks) of IV antibiotics.

Announcements

☆ Dr. Newman, our esteemed chairman, has welcomed two new grandchildren to his family, Sophia LG Newman and Anabelle Francis Shotton, both cared for in our NICU.

☆ Dr. Markenson celebrates the birth of his first son, George Jacob Markenson.

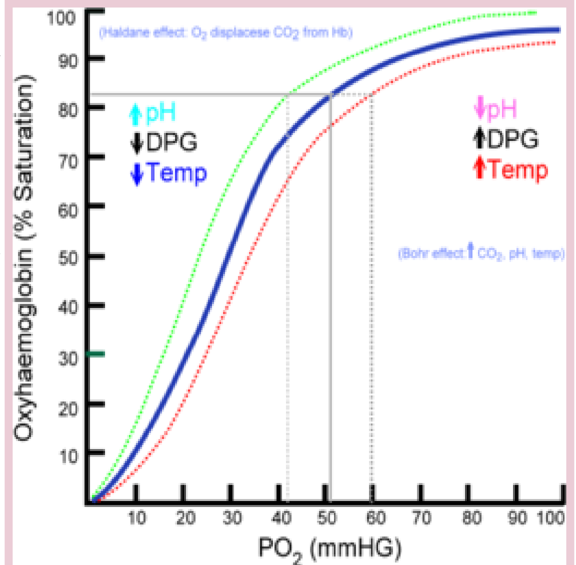
☆ Dr. Goltzman a-cappello group, The Westchester Chordsmen Chorus, will hold their holiday concert "A Barbershop Carol" on December 15 at 2 pm or 7:30 pm, @ Stepinac High School in White Plains.

Back to the Basics by Kristen Woodard

The oxygen dissociation curve shows the % saturation of hemoglobin at various partial pressures of oxygen. At high PO_2, usually in the lungs, hemoglobin binds to oxygen to form oxyhemoglobin. As the erythrocytes travel to tissues deprived of oxygen the PO_2 will decrease and oxyhemoglobin releases the O_2.

The sigmoid shape is a result of the cooperative binding of oxygen to the four polypeptide chains; showing an ↑ affinity for oxygen as each successive O_2 molecule binds to a heme group.

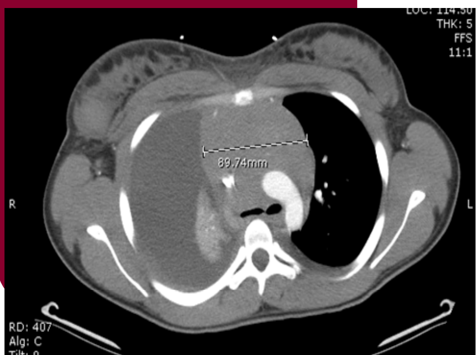
Left shift of the curve- ↑ affinity for O_2 (eg. at the lungs). Right shift - ↓affinity for O_2, found with increases in body temperature, hydrogen ion, 2,3-diphosphoglycerate or carbon dioxide concentration (the Bohr



Inside the ICU - Mediastinal Mass

16 y female presented with 10 days weakness, anorexia, SOB and orthopnea, with associated swelling of face/ neck, which improved with supine position. CT Chest and CXR demonstrated large anterior mediastinal mass with right pleural effusion. Diagnosed with T-cell NonHodgkin Lymphoma on aspiration of pleural fluid.

Don't forget 'The Terrible T's': thymoma, teratoma, 'terrible' lymphoma and thyroid carcinoma. Anatomic boundaries help delineate cause. Anterior mediastinal masses include lymphoma, teratoma, thymoma, retrasternal thyroid, pericardial cysts and aneurysms of ascending aorta. Middle mediastinal masses include bronchogenic cysts, lymph node enlargement, thyroid tumors, esophageal lesions and aneurysms of aortic arch. Posterior mediastinal masses most commonly include neurogenic tumors and paraspinal manifestations of spinal tumors. Although most mediastinal masses are asymptomatic, symptoms may include fatigue, cough, chest pain, fever, wheeze, dysphagia, weight loss and SVC syndrome.



"Bug of the Month" by Dr. Koelsch

If you have spent anytime in the PICU at Maria Fareri Children's Hospital you know there have been many children admitted with encephalitis. These children have had similar presentations but the etiology and prognosis has been vastly different. This is a difficult diagnosis for both the families and physicians taking care of these children because often no etiology is ever found and though most recover, our recent cases have resulted in severe impairment.

Encephalitis is defined as an acute CNS dysfunction (seizures, focal neurological findings, altered mental status) with radiographic or laboratory evidence of brain inflammation. There are two forms of infection-related encephalitis: primary and para/post-infectious. Primary encephalitis occurs when there is a direct CNS invasion by an offending agent and usually targets the gray matter of the brain, the prototypical example being HSV. HSV encephalitis is diagnosed by classical findings on MRI involving the temporal/frontal cortical brain tissue and EEG finding of PLEDS (paroxysmal lateral epileptiform discharges). Para/post-infectious encephalitis is an illness caused by indirect CNS infection where the neurological effects are due to the host's immune response to the infection. This type of encephalitis usually targets white matter of the brain and is prototypical of ADEM (acute disseminated encephalomyelitis). In cases of ADEM often there is a history of infection days to weeks prior to the onset of neurological symptoms. It is diagnosed by classic MRI findings of multifocal, patchy, high signal lesions on T2-weighted and FLAIR images. These patients are treated with corticosteroids and, if necessary, IVIG or plasma exchange. Clinically patients usually have a full, although slow recovery. The exception is post-measles ADEM, in which there is a 25% chance of mortality and permanent neurological deficits occurs in 25-40% of patients.

The overall incidence of hospitalizations due to encephalitis in the US is 7.3 cases/100,000 population. These patients account for 200,000 days of hospitalization and 1400 deaths. The highest incidence occurs in children < 1 year old or >65 years old. Encephalitis peaks in the late summer and fall which is consistent with our experience.

60% of encephalitis is due to an unknown etiology. When an etiology is found the great majority are viral including HSV, VZV, EBV, enterovirus and arbovirus. Non-viral infectious causes occur with *Mycoplasma pneumonia* being by far the most frequent. *M pneumonia* accounted for 5.5% of patients referred to the California Encephalitis Project, however many were diagnosed using serology, rather than CSF tests. Noninfectious etiologies of encephalitis are rare but should be in the differential and include autoimmune diseases/vasculitis, neoplasia or metabolic disorders.

Given the wide differential diagnosis for these patients a core battery of tests should be done upon arrival to the hospital and empiric treatment should be instituted. All children should have a LP and CSF sent for HSV PCR, bacterial culture, viral culture, and NYS encephalitis panel. Serum should be sent for Bartonella, EBV, Mycoplasma, West Nile Virus, and Lyme titers. Beyond blood and CSF patients should have a PPD placed, general viral cultures sent from the nasopharynx and rectum, an MRI and an EEG. Even though the etiology is not determined in more than half these cases it is important for the family and physicians to understand the diagnosis and feel satisfied with the search for a definitive cause.

DIRECTOR'S CORNER: DR. THERESA HETZLER

Welcome to the first issue of PRN. As I sat and thought about what I would write, I could not help but to think about the change of seasons. The falling leaves and dropping temperatures once again mark the beginning of recruitment season and the start of the 5th month of this academic year. Thanks to all who answered the Chiefs' survey which helped us tremendously as we revamped our daily schedule to help improve your educational needs while striving for assured compliance with the Duty Hours. We are also working on adding some more sub specialist to the Arts Teaching Attending schedule. Your comments were greatly appreciated. As we move through Autumn and look toward Winter please keep the following in mind: Flu season is approaching-GET YOUR FLU SHOT at Occupational Heath; RSV season is approaching-WASH your hands; Thanksgiving is around the corner-THANK your family and friends for all the support they provide through out the year; and, lastly, the annual Holiday Open House with the Hetzler/Denu family will take place Sunday December 16-SAVE THE DATE. Keep up

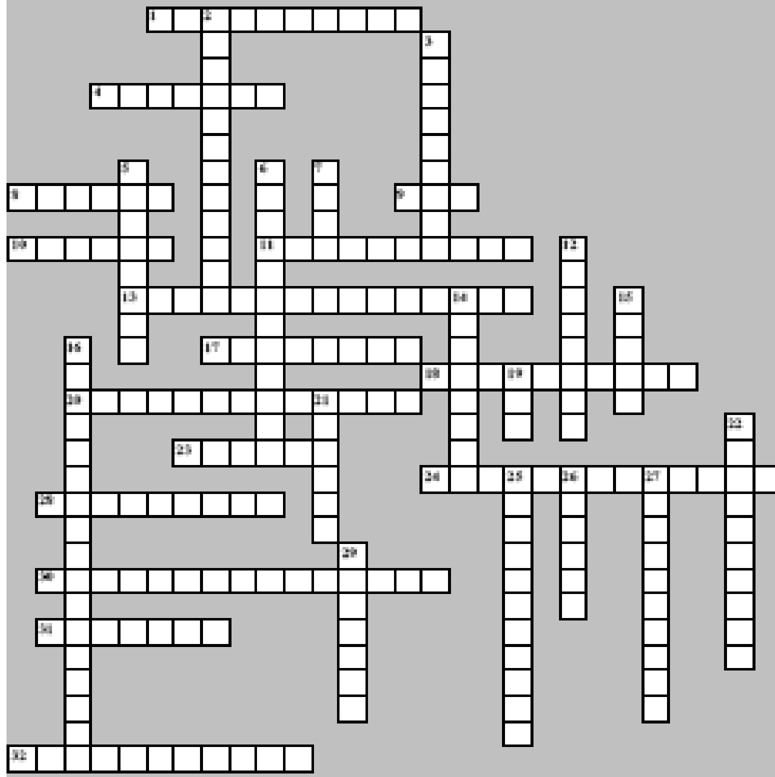
Ripped from the Headlines

In October's issue of Pediatrics, Carroll, et al. present a retrospective chart review of all children from 2 to 18 years old who were evaluated in the Connecticut Children's Medical Center Emergency Room for asthma exacerbation in 2005, excluding all of those patients with chronic medical conditions. The Modified Pulmonary Index Score (MPIS) were used to assess severity of illness, and weight criteria from National Center for Health Statistics was used to determine normal weight vs. overweight status. Patients were treated with an established protocol according to MPIS; admission criteria included MPIS greater than 7 after treatment in ED or oxygen requirement to maintain saturation greater than 94%. Of 884 ED visits, 22% of visits were from overweight children. These patients tended to be older and live in impoverished areas. Despite similar severity of illness and therapeutic interventions in the ED, overweight children were significantly more likely to be admitted to the hospital or ICU (p= 0.002). No clear explanation for this finding

"Childhood Overweight Increase Hospital Admission Rates for Asthma" Carroll, et. al *Pediatrics* 2007; 120; 734-740.

Puzzle of the Month

developed with help of Dr. Scherr, Dr. Rivera-Araujo, Dr. Garnecho and Dr. Scharbach



Across

1 Virus implicated in aplastic anemia in Sickle Cell patient
4 Inheritance pattern of Hunter's disease
8 Syndrome associated with congenital absence of pectoralis muscle
9 Number of Café-au-lait spots needed to diagnose NF
10 Sensorineural hearing loss and renal disease
11 Peg shaped upper incisors associated with syphilis
13 Disease associated with Gottron papules
17 Pierre Robin Sequence and Myopia
18 Treatment of Malignant Hyperthermia
20 #1 Cause of Cyanotic Heart Disease in First day of Life
23 Disease associated with Keiser-Fleischer Rings
24 Cells associated with Hodgkin's Disease (2 words)
28 Cofactor in Methylmalonic Acidemia (MMA)
30 Frontal Abscess associated with extension of Sinusitis (3words)
31 10 y male with newly discovered indirect hyperbilirubinemia
32 #1 Cause of Osteomyelitis in Children

Down

2 Abnormality of Feet Associated with Trisomy 18
3 Blockage of Meibomian Glands
5 Eye Findings associated with Wilm's Tumor
6 Disease associated w/ Colloidian Baby
7 Developmental Regression and Hand Wringing in Females
12 The "C" of CHARGE association
14 Vitamin whose deficiency causes Beriberi
15 Food often implicated as cause of botulism
16 Apparent Cyanosis with normal oxygenation on ABG
19 Boot-shaped heart on CXR (abb)
21 Muscle Hypertrophied in Beckwidth Wiedemann
22 Vitamin occasionally implicated in intractable seizures in Newborns
25 Treatment of RMSF
26 Widespaced Nipples, Webbed Neck, Short Stature
27 Cell on gram stain of Transient Neonatal Pustulosis
29 Pinpoint Bleeding after removing psoriatic scale

ANSWERS TO BE PUBLISHED IN DECEMBER 2007 EDITION

QUOTE OF THE MONTH

"There is something called the heart where all venous blood mixes together." - Dr. Gewitz (on new Calcium/Ceftriaxone guidelines)

Welcome to Our New Staff at MFCH-NYMC

WMC and MFCH welcome **Chrissy Schabacker, PharmD** as the new Pediatric Clinical Pharmacy Specialist. Chrissy graduated from Rutgers University with a Doctor of Pharmacy and then completed a Pediatric Specialty Residency at Al duPont Hospital for Children. As a Clinical Pharmacy Specialist, Chrissy will attend patient-care rounds and will be available to answer drug information questions. Chrissy's goal is to increase patient safety at MFCH by creating and implementing guidelines and policies that encourage appropriate medication use and monitoring.

Please contact Chrissy with any questions. She is available by phone at extension 5937 or via email at schabackerc@wcmc.com.

NYMC and MFCH welcome Dori Anchin, M.D. as a new Academic General Pediatrician. Dr. Anchin graduated from Weill Medical College of Cornell University in 2002 and then completed a residency in Pediatrics at Cornell Medical Center. Since that time Dr. Anchin has been practicing as a General Pediatrician at New England Pediatrics in Stamford, CT. She has already begun seeing patients at Bradhurst and taking service for General Pediatrics.