

BROUGHT TO YOU BY:
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Pediatric Residency News

Maria Fareri Children's Hosp



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APLS

July 9

Metabolic/Neonatal
Emergencies

July 16

Cardiovascular Emergencies

July 23

Environmental/
Toxicological Emergencies

July 30

CNS Emergencies

Journal Club

July 23

Dr. Julie Sweeney

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Case 1: PID by Dr. Melissa Cellini and Dr. Priya Sharma

BH is a 16 y.o. F with a PMH of ovarian cysts, who p/w RLO pain for 7 days. BH stated that her pain was initially intense and made worse with bearing down. She sought medical attention at an outside hospital at that time and an abdominal U/S demonstrated right hemorrhagic ovarian cysts. She was discharged but she continued to experience increasing left sided pain of the same character. She was sent to MFCH for further workup. On ROS, BH denied fever, vaginal discharge, nausea/vomiting or diarrhea. She had a decreased appetite secondary to abdominal pain. She reported having protected sex with one male partner 3 months PTA. On examination, BH's abdominal exam revealed +BS and b/l tenderness to palpation, rebound and guarding. On her pelvic exam, she was noted to have cervical motion tenderness and b/l adnexal tenderness. She did have white-gray colored discharge at the cervical os. On lab evaluation, she had a mild leukocytosis. Her urinalysis and pregnancy test were negative. Vaginal cultures and STI testing were performed. A pelvic U/S revealed a physiological cyst of the left ovary and right hemorrhagic follicle in the right ovary. BH was started on IV antibiotics for suspected PID.

Pelvic Inflammatory Disease (PID) is an ascending genital tract infection of sexually active females. The spectrum of inflammatory disorders classified as PID include endometritis, salpingitis, oophoritis, tubo-ovarian abscess (TOA), and peritonitis. The classic presentation of PID includes lower abdominal pain, abnormal vaginal discharge and cervical motion or uterine/adnexal tenderness on

exam. The specificity of the diagnosis of PID may be enhanced by fever, elevated ESR and CRP, and laboratory-documented evidence of *Neisseria gonorrhoea* or *Chlamydia trachomatis* infection. An U/S revealing thickened fluid-filled fallopian tubes may be seen. In difficult cases endometrial biopsy and laparoscopy may be aid in diagnosis. PID is a polymicrobial infection and infection with *Neisseria gonorrhoea* and *Chlamydia trachomatis*, as well as bacterial vaginosis, are the most common causes of acute PID.

Several options for effective treatment exist. Outpatient treatment may be initiated with ofloxacin or levofloxacin orally twice a day for 14 days or with ceftriaxone IM for one dose plus doxycycline orally twice a day for 14 days. If the patient has failed outpatient treatment, is clinically ill, is immunocompromised, or has a suspected TOA, inpatient treatment is the standard of care. Management includes IV antibiotics until clinical improvement and doxycycline orally for 14 days.

Adolescent cases account for 20% to 30% of all PID cases, with adolescents having a tenfold risk as compared to adults. The risk of PID increases with failure to use condoms, increased number of lifetime partners, having new partners in the last 3 months, a history of STIs, and the presence of an intrauterine device. Although the prognosis for PID is excellent if adequately treated early, there is a risk of infertility and future ectopic pregnancy which increases with recurrent episodes of PID. Therefore, early education and aggressive screening for STIs is critical.

Case 2: Klippel-Trenaunay Syndrome by Dr. Umesh Paudel

KM is a 20 year old female with a history of Klippel-Trenaunay Syndrome (KTS) who presented at an outside hospital with two weeks of worsening rectal bleeding and severe anemia. On the initial work up, she was found to have a Hb of 4.7 and she was transferred to MFCH for further management.

KM was born with a port-wine stain lesion and was diagnosed in the NICU with Klippel-Trenaunay Syndrome. She has a history of a lymphatic malformation in her bladder which required resection with augmentation of her bladder. She also has a history of an umbilical vein thrombosis and a microthrombi to her heart. She is known to have a vascular malformation of her rectum and has had multiple hospital admissions. Her mother also has significant for transmetatarsal amputation secondary to their gigantism.

KM's exam is remarkable for a port-wine stain on her right flank extending down to the right thigh, her right leg is larger than her left leg.

Klippel-Trenaunay Syndrome is a rare congenital syndrome characterized by a triad of port-wine stain, varicose veins, and bony or soft tissue hypertrophy involving an

extremity. Organs such as pleura, liver, spleen and bladder may also be affected with hemangiomas. This syndrome is often associated with other skin or skeletal anomalies.

Although Klippel-Trenaunay syndrome is rare, it is the most common form of combined vascular malformations. No specific inheritance has been established and there is no association to race or sex. The pathophysiology behind the syndrome is not well defined.

Complications may include cellulitis, chronic skin ulcers, and internal bleeding, DVTs and pulmonary embolism, chronic pain, and scoliosis. Management is mainly conservative. Nonoperative management includes compression garments for chronic venous insufficiency, laser therapy and anticoagulant therapy. Laser therapy can be used to reduce the size of hemangiomas. Surgical procedures can be used to remove bulk hemangiomatous tissue or to correct uneven limbs.



Picture of the Month: Intra-abdominal Abscess by Dr. Yanna Beniyaminov



AP is a 13 y.o. female with no significant past medical history, who presented with a 5 day history of worsening bilateral lower abdominal pain with LLQ predominance. She also had fever, nonbilious vomiting and diarrhea. After an initial physical and work up, a CT scan of her abdomen was performed. The results, as demonstrated, were consistent with a perforated appendicitis and a 9x11 cm pelvic abscess. Medical management was initiated with IV antibiotics. AP failed to improve clinically and was taken to the OR for appendectomy and abscess evacuation on HD#3. Intra-abdominal abscesses can develop in visceral intra-abdominal organs or in the intrainestinal, periappendiceal, subdiaphragmatic, subhepatic, pelvic, or retroperitoneal spaces. Most commonly, periappendiceal and pelvic abscesses arise from a perforation of the appendix. Fever, anorexia, vomiting, abdominal pain and lethargy may be present with the development of an intra-abdominal abscess. The peripheral

WBC count is usually elevated, as is the erythrocyte sedimentation rate. With an appendiceal abscess, localized tenderness and a palpable mass in the right lower quadrant may be appreciated. Abdominal distention, tenesmus with or without the passage of small-volume mucoid stools, and bladder irritability may indicate a pelvic abscess. Rectal examination may reveal a tender mass anteriorly. A psoas abscess can develop from extension of infection from a retrocecal or retroileal appendicitis and, in these cases, the presentation may include limp, hip pain, and fever. For radiographic evaluation, both ultrasound examination and CT scan can be used to localize intra-abdominal abscesses, although CT scans have superior sensitivity and can assess the retroperitoneal space. Treatment commonly includes drainage and broad-spectrum antibiotic therapy until sensitivities of the organism can be determined. Drainage can be performed surgically or under radiologic guidance. Intestinal phlegmon formation can complicate surgical resection and therefore an alternative approach to treatment is intensive antibiotic therapy for 4-6 weeks followed by an interval appendectomy provided the patient clinically improves on antibiotics.

Announcements

Welcome to our new residents:

- Nichelle Adegbite-Maravantano PL-3
- Yasamin Alwan PL-I
- Joseph Dayan PL-I
- Ellen Diamant PL-I
- Barry Diener PL-I
- Claire Elpenord PL-I
- Yonathan Elstein PL-I
- Nancy Fazzinga PL-I
- Myriam Fernandes PL-2
- Michelle Gaba PL-I
- Aditi Gupta PL-I
- Alpa Lotlikar PL-I
- Steven Horwitz PL-I
- Purnima Mandal PL-I
- Shirley Obioha PL-I
- Nisha Patel PL-I
- Cigal Shaham PL-I
- Tanzid Shams PL-I

Welcome to the new
Pulmonology, GI and NICU
fellows .



Advocacy Corner by Dr. Rebekka Levis

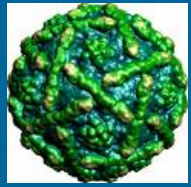
Welcome to the first edition of the Advocacy Corner for the 2008-2009 year. The pediatric RRC requires that residency programs have structured curriculae and evaluation processes to prepare residents for their roles as advocates for the health of children in the community. Our advocacy program has been divided into four "advocacy task forces". These task forces include: 1) Nutrition, including food safety, obesity and breast-feeding; 2) Legislation encompassing insurance issues and access to care; 3) Violence and abuse; 4) Adolescent and reproductive health;

Whether you are already involved in an advocacy task force, or you are just beginning your advocacy career, here are some suggestions on how to become a child advocate: Become informed. Read the paper, watch the news, and familiarize yourself with the issues at hand. Educate your patients, your community, and your peers about the importance of supporting our youth. Educate elected officials and write letters to government officials to advocate for children. Teach patients and parents to advocate for themselves. And finally, don't forget to vote!

Inside the ICU: Venous Sinus Thrombosis by Dr. Amy Lim

JC is a 4 y.o. M recently diagnosed with Pre-B cell ALL who presented after he was found unresponsive, following several days of mild headaches. He had begun chemotherapy 3 weeks prior to admission with vincristine, daunorubicin and peg-asparaginase & his last treatment was 4 days prior to admission. CT scan from an outside ER showed evidence of intracranial hemorrhage. Repeat head CT with angiography confirmed a left frontal parenchymal hemorrhage and a subarachnoid hemorrhage with associated vasogenic edema, a subdural hemorrhage along the falx anteriorly, early hydrocephalus and a superior sagittal sinus consistent with venous sinus thrombosis. JC's prothrombotic work up was significant for low antithrombin III. Initial management included mechanical ventilation, ICP monitoring and FFP transfusions. On day 3, after a repeat CT showed continued presence of thrombus with worsening hemorrhage and edema, endovascular thrombolysis was performed by interventional radiology. Thrombotic events are a side effect in about 1% of ALL patients receiving peg-asparaginase. Asparaginase depletes asparagine in leukemic cells, which inhibits protein synthesis and causes apoptosis of the cells. It can cause a decrease in the levels of prothrombin, multiple coagulation factors, fibrinogen, antithrombin, proteins C and S and plasminogen which collectively increase the coagulability state of the patient. Venous sinus thrombosis can be subdivided into septic

and nonseptic causes. Septic causes include bacterial spread from sinusitis, otitis media, mastoiditis or direct spread from facial infections. Aseptic causes include severe dehydration with thrombosis of the superior sagittal sinus and the superficial cortical veins due to hyperviscosity and sludging of blood. Prothrombotic disorders-including deficiencies of coagulation inhibitors, increased factor VIII levels, activated protein C resistance, Factor V Leiden mutation, abnormalities of lipoprotein A, MTHFR gene mutations, elevated homocysteine levels, and anticardiolipin antibodies-are found in 12-50% of cases. Focal neurologic signs may be seen in older children while neonates often present with altered MS and seizures. The immediate treatment goals are to recanalize the occluded vein, to prevent the propagation of the thrombus, to treat the underlying prothrombotic state, and to prevent the recurrence. The main treatment is anticoagulation. Direct endovascular thrombolysis has been used as an alternative treatment in cases where patients worsen despite anticoagulant therapy or anticoagulation is contraindicated, as it was in JC's case. Direct thrombolysis aims to dissolve the venous clot by delivering a thrombolytic substance within the occluded sinus through an intravenous catheter. Cerebral sinus thrombosis is associated with a good outcome in close to 80 percent of patients. Despite the overall favorable prognosis, approximately 5 percent of patients die in the acute phase of the disorder, and longer-term mortality is nearly 10 percent. The main cause of acute death is brain herniation.



Bug of the Month: Enterovirus by Dr. Narendra Dereddy

Enteroviruses (EV) are single-stranded RNA viruses, belonging to the family *Picornaviridae*. The 64 known enterovirus serotypes are divided into five subgenera: Polioviruses, group A Coxsackie viruses, group B Coxsackie viruses, Echoviruses and Enteroviruses. Poliovirus infections have been eliminated in most parts of the world and are targeted for global eradication. Humans are the only reservoirs of clinically significant EV and the most common route of transmission is fecal-oral. Coxsackie A21 infections are also transmitted via respiratory secretions. Enteroviral infections occur frequently in the summer and fall. There are several clinical manifestations of EV infections with gastroenteritis being an infrequent presentation. The most common manifestation is an acute febrile illness without an apparent source. EV infections contribute to more than 90% of non-specific febrile illnesses in infants less than 3 months of age undergoing evaluation and treatment for serious bacterial infections. Approximately half of these febrile infants have aseptic meningitis. Most infants recover within 7 days without sequelae. Enteroviruses may also present with exanthems and enanthems. Hand, foot and mouth disease (HFMD) is a distinctive illness characterized by fever, vesicular stomatitis and peripherally distributed papular and/or vesicular lesions (especially on hands and feet and sometimes on buttocks and genitalia). Most patients recover within a few days but HFMD caused by EV-71 may accompany serious neurologic disease. In fact, a recent outbreak of HFMD due to EV-71 in China has been responsible for the deaths of over 22 children. Herpangina is an intraoral vesicular enanthem. Fever and other systemic signs such as headache and myalgias are present at onset. Sore throat and difficulty swallowing precede the onset of oral lesions. EVs can cause viral meningitis and encephalitis. These patients present acutely with fevers, mild meningeal signs and increased WBCs in their CSF with a predominance of PMNs. Echoviruses and group B Coxsackieviruses cause more than 90% of all cases of viral meningitis. All age groups are susceptible, but the majority of cases are among infants younger than 12 months.

Complications such as febrile seizures, coma and movement disorders occur early in the course in 5-10% of patients. It is a self-limiting illness with complete recovery but subtle disturbance of motor function may persist for weeks. Focal cerebritis, brainstem encephalitis, and transverse myelitis are less common but serious diseases associated with a limited group of serotypes. EV-71 and Coxsackie A7 have been associated with acute motor neuron disease (MND) clinically similar to poliomyelitis. MND associated with non-polio enteroviruses has a better outcome than poliomyelitis. EVs are also the most common cause of viral myocarditis. Group B Coxsackieviruses account for one half of diagnosed myocarditis cases. As many as 15% of these children die during the acute illness and 10% develop persistent compromise from dilated cardiomyopathy (DCM) requiring cardiac transplantation. DCM is the final result of multiple infectious and non-infectious cardiac insults including clinically apparent and unrecognized past EV infections. Pleurodynia is an acute illness characterized by fever and localized myositis of the intercostal and abdominal muscles leading to localized sharp, spasmodic thoracic and abdominal pain. Acute EV myositis may localize to other muscle groups or may generalize leading to myoglobinemia and myoglobinuria. Neonatal infections are acquired perinatally from infected mothers. Neonates develop nonspecific signs of lethargy and poor feeding between the 3rd and 7th day of life before progressing to either severe myocarditis or fulminant hepatitis. Diagnosis of EV is accomplished by isolation and identification of virus in cell culture or by detection of RNA by PCR. Specimens placed in the pink viral transport media are sent to the county lab. PCR in the CSF is available through the NYS encephalitis panel. Serology is not useful in diagnosis of acute disease. The great majority of EV infections are self-limited and do not require specific therapy. Therapeutic options for more serious/life-threatening infections such as myocarditis, neonatal infections and infection in B-cell deficient hosts include IVIG and the experimental antiviral drug pleconaril.

DIRECTOR'S CORNER: DR. THERESA HETZLER

It's July and those of us in medical academia know what that means...new residents and fellows. I would like to take this opportunity to welcome our 18 new pediatric residents as well as our new Pulmonary, GI and Neonatology fellows to MFCH.

This July also marks the 20th anniversary of my joining the WMC family. Many changes have taken place over the past 20 years. The days of Q3 call with 36-hour days is a thing of the past. But the focus on education, the caliber of our residents and the strength of our faculty have improved dramatically. This is a result of the dedication of the faculty and departmental leadership who, not only admit their patients from which the residents learn, but also take the time to be teaching attendings, conference speakers, recruitment interviewers, mentors and preceptors in the clinics. Thanks to all of you who go the extra mile for the residents.

Best of luck to all the new members of the MFCH-WMC family. We are here to support you through the next three years and to help you become the best pediatricians you can be.

Terry Hetzler, MD

Back to the Basics: Portal Circulation By Dr. Rachel Lewis

Portal circulation refers to the flow of blood from the small intestine, proximal colon, and spleen to the liver prior to return to the right atrium. Blood from the capillary beds of the intestinal tract drains to the larger portal vein, which, in turn, supplies the capillary beds of the liver. Excess nutrients such as protein and glucose, that are absorbed in the small intestine, are eliminated in the liver. Ingested drugs and toxins are also filtered and metabolized in the portal circulation, often resulting in a lower concentration of active metabolite and decreased bioavailability (known as the first-pass effect). Blood from the portal vein flows through sinusoids between hepatocytes. It is then joined by blood from the hepatic artery to collect in the central vein and then into the hepatic vein, which drains to the inferior vena cava and the right heart.

Portal hypertension can be caused by cirrhosis of the liver, portal vein thrombosis, schistosomiasis (in developing nations), or other causes including idiopathic portal hypertension. It results in the development of anastomoses between the portal and systemic circulations. Blood returning towards the heart encounters elevated pressure in the portal vein and redirects into the systemic circulation to reach the heart. The resulting 4 portacaval anastomoses are esophageal, rectal, paraumbilical, and retroperitoneal. Diverted flow through portacaval anastomoses leads to venodilation of these areas and subsequent esophageal varices, hemorrhoids and caput medusae.

Medical Spanish by Dr. Doris Rivera-Araujo and Dr. Jennifer Garcia-Hoffman

Sun Safety *Seguridad contra el Sol*

< 6 months/meses:

Avoid sun exposure and dress infants in lightweight long-sleeved clothes

Evitar la exposición a la luz solar y vestir a los infantes con ropa ligera y de manga larga

Apply sunscreen to small areas

Aplicar loción para protección solar en áreas pequeñas

For Older Children/ Para niños de mayor edad:

The best way to protect against the sun is covering up
La mejor manera de protegerse del sol es cubriéndose

Stay in the shade whenever possible, especially between the 10am and 4pm

Permanezca en la sombra cuando sea posible, especialmente entre las 10am y 4pm

Use sunscreen with SPF of 15 or greater

Use loción para protección solar con un FPS de 15 o más

Reapply sunscreen every two hours, or after swimming or sweating

Vuelva a aplicar loción para protección solar cada dos horas, o después de nadar o sudar

Outdoor Safety *Seguridad en exteriores*

Never leave children alone in or near a pool

Nunca deje a los niños solos, ni cerca de la piscina

Your child needs to wear a helmet on every bike ride

Su hijo necesita usar un casco cada vez que corra en bicicleta/ patines

Bug Safety *Seguridad Contra Insectos*

Don't use scented soaps, perfumes or hair sprays on your child

No use jabones perfumados, perfumes ni aerosoles para el cabello en su hijo

Insect repellents containing DEET are most effective against ticks and mosquitoes

Los repelentes de insectos que contienen DEET son más eficaces contra garrapatas y mosquitos

DEET should not be used on children younger than 2 months of age

El DEET no debe usarse en niños menores de dos meses de edad

Puzzle of the Month: Word Search by Dr. Julie Sweeney

s p z w z k f i s m n g k a w a s a k i
 m i d g r c m e j t n f r u l u j v q b
 b a s n r e a c s g e c w m o m t t o
 t k r o a c t i f i g k s i s z y s l s
 g d u f t r m l l p s c c l s j f z k t
 y p i n a a b y e e y o x i l e p n j g
 w e f g o n m e n f c s i a r a n y g h
 b g t v e i y o l e e t g l c h c r p m
 r m j m t o t u r l g n a n o o n h m c
 t s r h t s r p d b i b i a o c g s c h
 u n d v e y k g e d i w p l o g s m v d
 j n u l r n c k e c y f n p k n i j p f
 s a i s u o i e p h s p o o c i s x j j
 t m h q o s f l s m l u t r v h e f f n
 k l g p t t j g p w e l s t u t r c o r
 y l y f s o l z b i k v i s v e u n l m
 t a b a m s h q m q c n r p u e n r j f
 r k e l l i g a l a e s o s l t e l a b
 j r s d i s h d d e m s i t u a n u e u
 b q j m w s t o d e h t u d o p l i k l

alagille
 alport
 autism
 breastfeeding
 celiac
 craniosynostosis
 croup
 digeorge
 discipline
 enuresis
 intussusception
 kallmann
 kawasaki
 klinefelter
 lice
 marfan
 meckels
 milestones
 milia
 neurofibromatosis
 noonan
 precocious
 rickets
 rsv
 scoliosis
 sids
 teething
 tourette
 vonwillebrand

ANSWERS TO BE PUBLISHED WITH AUGUST 2008 EDITION

Quote Of The Month:

"I can't take that patient on to my service. When will I have time to drink my coffee?" (Dr. McBride alluding to the graduate skit)