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Pediatric Residency News

Maria Fareri Children's Hospital



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Grand Rounds

April 2

M. Balazy—Children's
Hospital Foundation
Scholar

April 9

Communication
Dick Harris, PhD

April 15

"The Long Road to a
Very Strange Operation"
Dr. T. Kulick

April 23

Advances in Pediatric
IBD
Dr. H. Winter

April 30

Lyme Disease Guidelines
Dr. G. Wormser

Resident As Teacher

April 22

Dr. Amy Lim

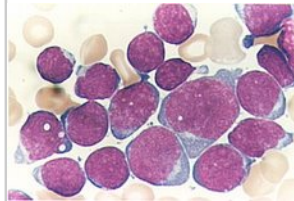
Journal Club

April 23

Dr. Alison Airall-Ryan

Case 1: Acute Lymphoblastic Leukemia by Dr. Sathya Theodore

CP is a 4 year old female who presented with one month of decreased activity and 3 days of fever and pallor. On initial presentation she was diagnosed with a UTI. However, she developed petechial rash and bruising, as well as mucosal bleeding. In MFCH ED, she was noted to have enlarged left anterior cervical and supraclavicular lymph nodes. Laboratory findings showed leukocytosis (WBC 43.5, Blasts 82%), anemia, and thrombocytopenia. Coagulation factors, UA, CMP and CXR were all normal.



She was admitted for further diagnostic testing. Bone marrow aspiration confirmed early pre-B cell acute lymphocytic leukemia (ALL) and induction chemotherapy was initiated.

ALL is the most common malignancy of childhood, accounting for nearly 1/3 of all pediatric cancers; peak incidence is between 2-6 years of age. Clinical presentation is varied and includes fever, anemia, fatigue, anorexia, irritability, petechiae, bleeding, bone pain, LAD, and HSM. ALL is suggested by CBC showing one or more cytopenias and a peripheral blood smear demonstrating blasts (seen right), however, CBC can be normal. Definitive diagnosis is made by bone marrow aspiration/biopsy demonstrating leukemic blast cells. Lumbar puncture determines the presence of CNS extension. Although there is no known etiology for ALL, predisposing genetic and environmental factors have been identified. Newly diagnosed children are stratified into risk categories: low, standard, high. Standard risk ALL consists of: female gender, age at diagnosis of 1-10yrs, initial leukocyte count

<50,000/ μ L, no CNS involvement, certain morphologic subtypes (eg. hyperploidy) and rapid response to initial treatment. Phases of therapy include induction (goal <5% blasts in bone marrow), consolidation, and maintenance. Induction chemotherapy achieves complete remission in greater than 90% of patients. Before and during initial therapy, patients are at risk for tumor lysis syndrome (TLS), metabolic derangements caused by the breakdown of leukemic blast cells. TLS consists of hyperuricemia, hyperkalemia, hyperphosphatemia, and hypocalcemia. To prevent the sequelae of TLS (including acute renal failure), patients are treated with high rates of bicarbonate-containing IVF, allopurinol and urate oxidase, to increase secretion of phosphate and uric acid. Consolidation therapy consists of variable multidrug regimens and may include CNS therapy to decrease the chance of CNS relapse. Maintenance therapy follows for 2-3 yrs and includes surveillance studies and chemotherapy. Overall prognosis has improved with long-term survival greater than 80% after 5 years.

Case 2: Neonatal HSV by Dr. Jennifer Garcia-Hoffman

LF is a 2 wk old male presenting with crusted lesions and purulent discharge on the top of a scalp nevus. One day PTA he was evaluated by a dermatologist and treated with mupirocin. The following day the mother noticed an increased number of lesions on the scalp and one on the face. He had no altered mental status, irritability, fever, decreased oral intake or urinary output. In MFCH ED a full sepsis workup was done. Clindamycin and acyclovir were started on admission with a primary differential diagnosis of herpes and impetigo. Birth history included full term C-section secondary to failure to progress with no other complications, including no fetal scalp monitoring, Maternal labs were negative. LF's mother had no apparent STI or genital lesions, but had buccal "cold sores". Laboratory studies including CBC, BMP, blood and urine cultures, and bacterial wound culture were all negative. Viral wound culture demonstrated HSV-2. Herpes-PCR of CSF showed HSV-2. Brain MRI was normal. Clindamycin was discontinued and

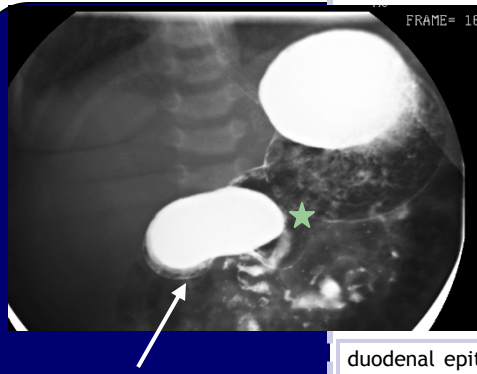
treatment was continued with IV Acyclovir for 21 days for CNS herpes infection. Infants affected with neonatal HSV are born to asymptomatic women in 50-70% of cases. Antepartum cultures are not useful in assessing risk of neonatal infection. Several risk factors have been identified including rupture of membranes > 6 hours, scalp electrode or other internal monitoring, chorioamnionitis, cervicitis, and vaginal delivery. Risk is significantly increased with primary maternal herpes versus recurrent disease. *In utero* and postnatal transmission are very uncommon, although there have been cases of infection from a nipple lesion during breast feeding and infection from a father with a cold sore. Ninety percent of cutaneous herpes presents with skin, eye, and mouth (SEM) disease, as clusters of discrete vesicles on an erythematous base. These lesions invariably recur in the first 6 months of life, with many infants demonstrating recurring lesions after a year of age. Disseminated disease involves multiple organs, most prominently the liver

and lungs, with a possible CNS component. Localized CNS herpes is diagnosed by abnormal LP results or brain MRI/CT changes in conjunction with a positive surface cultures. 70% of infants with neonatal herpes develop CNS herpes. Signs and symptoms include irritability, seizures, poor feeding, bulging fontanel, and thermal instability. CSF-HSV culture is positive in 25-40% cases with pleiocytosis and proteinosis. Patients may have CNS or disseminated herpes without mucocutaneous lesions. Continued on page 4.



Case 1 ALL	1
Case 2 Neonatal HSV	1
Picture of the Month	2
Advocacy Corner	2
ED Snapshot	2
Bug of the Month	3
Director's Corner	3
Back to the Basics	3
Homeopathy	4
Puzzle of the Month	4

Picture of the Month: Duodenal Stenosis by Dr. Rachel Lewis



Dilated early segment of duodenum.

Minimal contrast advancing through stenotic segment.

AC is a 2 wk old female with Down syndrome and AV canal defect, who presents with non-bilious vomiting and FTT since birth. UGI series at an outside hospital (seen left) demonstrated duodenal stenosis. NG tube decompression was initiated at MFCH and surgical intervention followed. Duodenal atresia and stenosis represent a spectrum of outcomes from disordered embryological development. Proliferation of the

duodenal epithelium occurs during week 6 of fetal development, obliterating the lumen of the duodenum. This proliferation is followed by vacuolization, then recanalization, in weeks 7 and 8, restoring the intestinal lumen. The leading theory on the etiology is failure of recanalization. Duodenal atresia/stenosis occur once in every 10,000 live births. Twenty to 30% of infants with atresia or stenosis have trisomy 21, and approximately 8% of infants with trisomy 21 have atresia or stenosis.

Presentation of duodenal atresia is vomiting within hours of initiation of enteral feeds, whereas duodenal stenosis presents later, depending on degree of obstruction. The vomitus is usually bilious, as the stenosis most often occurs distal to the ampulla of Vater, but can be bile-free if the obstruction occurs proximal. If duodenal obstruction is suspected, a plain abdominal film can be taken; the classic sign with atresia is the “double-bubble,” which represents both the dilated stomach and duodenum above the obstruction. Air present distal to the duodenal “bubble” can be seen when the obstruction is incomplete. An upper GI series can also aid in diagnosis of varying degrees of duodenal obstruction. Barium can be seen flowing from the stomach to a dilated segment of duodenum until the stenotic region and is seen slowly trickling out into the distal intestine. Duodenal atresia and stenosis are treated surgically. Prior to surgery, the infant’s digestive tract must be decompressed via nasogastric tube. The atretic or stenotic region is then excised, and the two segments of duodenum are anastomosed in a duodeno-duodenostomy. After surgery, feedings are advanced slowly as tolerated. When infants are able to tolerate adequate enteral nutrition, they can be discharged.

Announcements

Congratulations to all on a successful Match Day.

Come support Team Residents at Go The Distance by walking with us on Sunday, April 13th at 9:30 am. Visit our team webpage to sign up and make a donation at: mfchwalk.kintera.org.

Graduation is approaching, save the date: Wed, June 25th. Party will be at Windows on the Hudson in Dobbs Ferry from 6p to 11p.

Advocacy Corner by Dr. Emily Koelsch

The Advocacy Task Forces have been busy meeting, gathering information on the needs of our community and making connections with community programs. Angela Lumba and other members of the Adolescent Task Force visited the Children’s Village group home to discuss asthma with teenage residents. Additional presentations on topics including nutrition, exercise, STIs and sexual pressures are planned. All residents interested in participating are encouraged to speak with Angela. In February, Dr. Kapklein and I attended the AAP District II Advocacy Day in Albany, NY. Assemblyman Richard Gottfried and Senator Kemp Hannon, who both chair representative Health Committees, discussed their opinions on the possibility of Universal Health Care in New York state, as well as expansion of the Child Health Plus program. Guthrie Birkhead, the Deputy Commissioner of Health, briefed us on several preventative services the state offers, such as HPV vaccines for 19-26 year olds, adolescent pregnancy prevention, diabetes and asthma education, lead poisoning prevention and universal vaccine purchasing. Later, we hit the Legislative Office Building to meet with our state assembly-people and senators to lend our voices in support of child-health related parts of the executive budget. The budget calls for expansion of the Child Health Plus program to include families with incomes up to 400% of the poverty level, to increase payment for primary care services, and to improve school based nutrition (funding for more nutritious foods, expansion to include breakfast.) The executive budget supported by the AAP was proposed by former Governor Spitzer; we hope Governor Patterson will continue to make children’s health a priority.

ED History

CM is a healthy 33 mo F who presented to MFCH ED 6 hours following unwitnessed, presumed ingestion of “Tom Cat” rat poison (active ingredient: bromodialone). She arrived to ED without physical complaint except abdominal pain. Specifically, there were no reported signs of bleeding. Examination in the ED was within normal limits. Poison Control was contacted for assistance.

ED Snapshot: Superwarfarin Ingestion by Dr. Pamela Gonzalez

Bromodialone is a second-generation anticoagulant of the indandione group, also dubbed *superwarfarins* due to their extremely long action. This class was synthesized to combat warfarin-resistant rodents. Bromodialone acts via vitamin K antagonism, inhibiting vitamin K reductases and thereby reducing vitamin K-dependent factors. Its intended action is death via hemorrhage in its rodent target.

The clinical effects of bromodialone and other superwarfarin ingestion in humans are variable, depending on dose and chronicity of exposure. In the case of acute, non-intentional ingestion - the more common scenario in children - the dose is usually small with no or resultant anticoagulant effect or bleeding. These patients may have initial GI complaints of nausea, vomiting or non-specific abdominal pain. In severe poisoning (large dose or chronic exposure), patients may present with bleeding from any organ. Anticoagulation and clinical bleeding may persist for to 24-36 hours in humans, with the elimination half-life on the order of days to weeks. Laboratory findings in humans may include PT or INR prolongation. In severe

cases, PTT may also be prolonged. The four vitamin K-dependent factors (II, VII, IX and X) may be reduced. Current treatment recommendations depend on clinical signs and laboratory values. Although most poison centers suggest decontamination with charcoal, there is no proven benefit of doing so. For patients with evidence of bleeding (symptomatic) and laboratory evidence of coagulopathy, FFP and subcutaneous vitamin K may be indicated. For asymptomatic patients with laboratory evidence of anticoagulation, PO vitamin K is suggested. In the case of the asymptomatic patient with normal laboratory values, prophylactic vitamin K is not recommended for two main reasons: distortion of subsequent PT/PTT measurements, and likely lack of benefit of treatment due to long action of superwarfarins.

Due to delay in presentation, CM did not receive GI decontamination. Initial laboratory values included normal PT, PTT, INR, platelet, and Hb values. She was discharged home, returning to our ED in 24 hrs, with no bleeding signs and normal repeat labs.

1-800-POISONS



Adenovirus



Wash Up For Your Patient's Health!

Bug of the Month
by Dr. Carol Spagnolo-Ilye

Since its discovery in the 1950s, *adenovirus* has been isolated from virtually every human organ system. There are several "classic" clinical syndromes attributed to *adenovirus*. As pediatricians, it is important to be familiar with the wide variety of clinical manifestations and their treatment.

Adenoviruses are medium-sized, non-enveloped, icosahedral viruses containing double-stranded DNA. Forty-nine immunologically distinct types are responsible for causing infection in humans. These viruses are extremely stable, which allow for prolonged survival outside of a host, and therefore, are highly contagious. Transmission of disease is person-to-person, particularly in young children where fecal-oral contamination is common. The typical incubation period is 2-9 days. Laboratory diagnosis is made by isolation of *adenovirus* in culture, antigen detection or PCR.

Adenovirus most often manifests as an upper respiratory illness including coryza, pharyngitis/tonsillitis and fever. This is most common in the pre-school age child. More than half of infants affected with *adenovirus* URI's have an associated otitis media and large number develop diarrhea. Pharyngoconjunctival fever is a distinct disease caused by *Adenovirus* type 3. Clinical features include high fevers that persist for 4-5 days, pharyngitis, rhinitis, and periauricular and cervical lymphadenopathy. The associated conjunctivitis is non-purulent and occurs in 75% of affected patients. Physical exam shows inflammation of the bulbar and palpebral conjunctivae and is often bilateral. The conjunctivitis persists after resolution of fever and other symptoms. This syndrome is often confused with Kawasaki disease.

Adenovirus is the most common cause of follicular conjunctivitis, a mild illness, and keratoconjunctivitis, which may occur in epidemics. Epidemic keratoconjunctivitis is a severe eye disease and is frequently associated with exposure to swimming pools (termed "swimming pool

conjunctivitis.") Clinical symptoms begin as photophobia and foreign body sensation, and progress slowly to impaired vision, eyelid swelling and subconjunctival hemorrhage. Acute symptoms may last for up to 4 to 6 weeks.

Adenovirus is a cause of lower respiratory illness including bronchitis, bronchiolitis and pneumonia. There is a well described "Pertussis-like" syndrome which manifests as paroxysmal cough, URI and fever; it is frequently seen in association with *Bordetella pertussis*, but may be seen in isolation. *Adenovirus* pneumonia is responsible for 7-9% of acute pneumonias in hospitalized children. The radiographic features closely resemble those of bacterial disease, with lobar infiltrates and/or effusions. These children often have high fever, abdominal pain and diarrhea. Mortality rates as high as 10% have been reported. Survivors have residual damage to the airway including bronchiectasis, bronchiolitis obliterans (BOOP) and, rarely, pulmonary fibrosis.

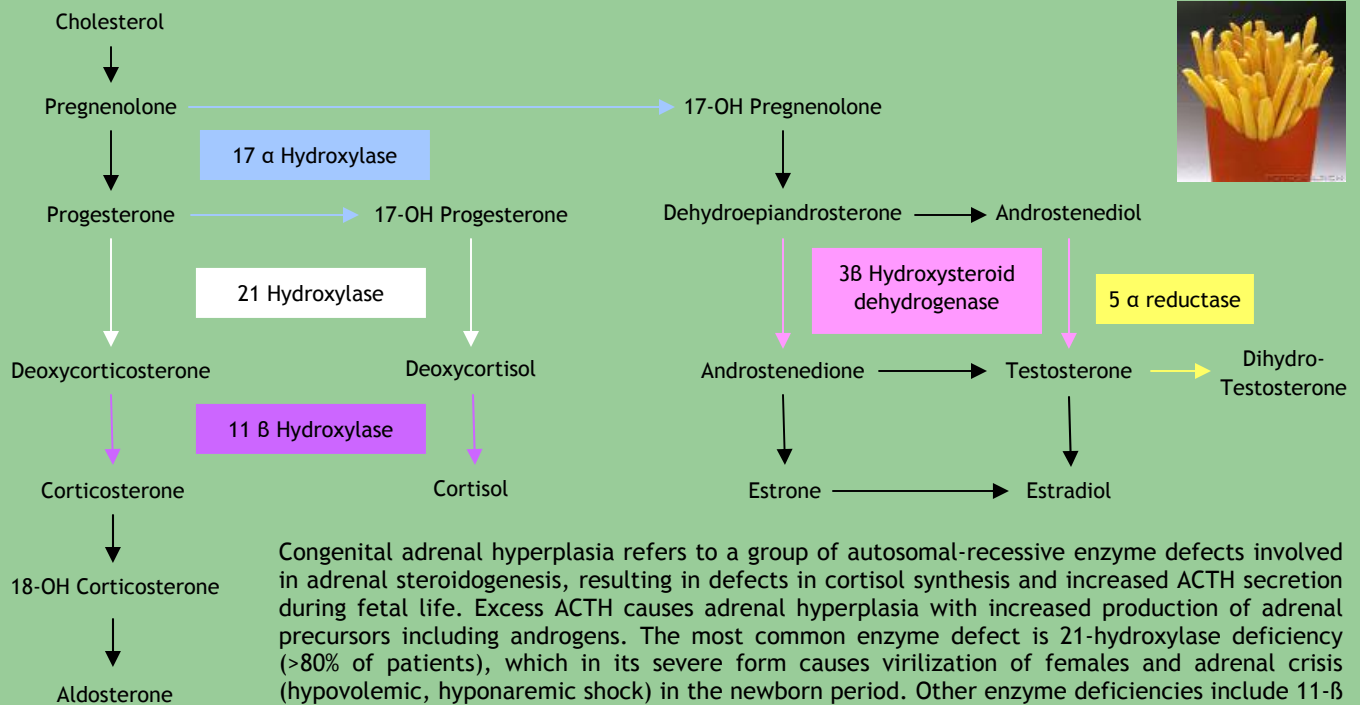
Enteric *adenoviruses* "40 and 41" are isolated in stools of 5-9% of children with acute diarrhea. Enteric *adenovirus* is often asymptomatic; however, there is a risk of intussusception secondary to enlarged lymph nodes. Infection with *adenovirus* is additionally associated with hemorrhagic cystitis. This clinically manifests as fever with sudden onset of bacteriologically-sterile hematuria, dysuria, and urgency lasting 1-2 weeks.

Clinical illnesses including CNS disease (meningitis, encephalitis) and GU disease (ulcerative genital lesions, urethritis, cervicitis) have also been attributed to *adenovirus*.

Treatment of adenovirus is largely symptomatic. Cidofovir has shown activity against *adenovirus* and may be considered for immunocompromised patients. BMT patients with hemorrhagic cystitis caused by *adenovirus* have been treated with cidofovir. In addition to standard precautions, patients should be on contact and droplet isolation.

Back to the Basics: Steroid Metabolism

By Dr. Kristen Woodard and Dr. Lauren Mendelsohn



Homeopathy by Dr. Pamela Gonzalez

Neonatal HSV, Cont. By Dr. J. Garcia-Hoffman

Complementary and alternative medicines (CAMs) are not created equal, and neither are the terms that delineate them. For example, *herbal medicine* denotes the use of medicinal herbs to promote health or healing, whereas *homeopathy* describes a treatment system and philosophy, which may or may not involve the use of medicinal herbs. In this issue, we will review homeopathy in brief, including comments on its use in children.

German **physician** Samuel Hahnemann is credited with the origin of homeopathy, beginning in the late 18th century. Homeopathy is a system of therapy based on the concept that disease can be treated with compounds, in minute doses, thought capable in larger doses of producing the same symptoms in healthy people as the disease itself. This approach is called *similia similibus curentur*, or “like cures like”. The proposed mechanism of action of these remedies is via stimulation of the body’s immune defenses. Preparation begins with a *mother tincture*, which subsequently undergoes serial dilution or *potentization* and agitation or *succussion*. These dilute amounts could putatively be used to treat the very symptoms they were known to produce. Homeopathic remedies have been regulated by the FDA since 1938, in the same manner as nonprescription, over-the-counter drugs.

Part of the appeal of homeopathy to many parents of children with chronic illness is its holistic approach, considering the whole of body and spirit, rather than just localized symptoms or disease. The effects of homeopathic remedies have been examined in several childhood illnesses including ADHD, autism, diarrhea and influenza. The evidence from rigorous clinical trials of any type of therapeutic or preventive homeopathic intervention in children is not yet convincing enough for recommendations in any condition. Similarly, systematic reviews have yet to support or refute the use of homeopathy in children. One major debate is over whether homeopathy affords any benefit beyond placebo. Also at issue is how the activity of a given substance could be retained in solutions diluted to below-zero concentration.

As with many CAMs, more and larger studies need to be performed before determining whether or not homeopathy has any place in the treatment of illnesses in children. In the meantime, it is important to explore the individual appeal of homeopathy, as well as to inquire about any CAM treatments parents may be pursuing for their children.

Useful resources:

- www.nccam.nih.gov; National Center for Complementary and Alternative Medicine
- Coulter MK and Dean ME (2007). Cochrane Database of Systematic Reviews Issue 4
- Altunc U et al. (2007). Mayo Clinic Proceedings 82: 69-75

In the absence of skin vesicles diagnosis is difficult, because other clinical signs are nonspecific and mimic neonatal sepsis. Treatment for CNS or disseminated disease includes Acyclovir 60 mg/kg/day IV divided q8h for 21 days. CT scan C+ or MRI of the brain should be done at the beginning and end of therapy. Dilated ophthalmologic examination should be completed to assess for chorioretinitis during the first week of therapy and at 6 mo. Brainstem auditory evoked potential (BAER) is necessary during the initial admission and must be repeated at 6 mo if abnormal. Developmental follow-ups at 6 and 12 months of age are recommended. For any subsequent fever, neurologic symptoms, or skin lesions repeat examination of CSF and blood PCR, and HSV culture and PCR on scrapings of skin lesions, must be completed until 6 mo of age. Consider oral acyclovir prophylaxis from day 21 to 6 mo. While on oral acyclovir, monthly CBC and serum creatinine at 3 and 6 months are necessary. Prognosis is improved significantly w/ early acyclovir treatment. Overall mortality of untreated CNS herpes is 50%, and 85% for disseminated disease; treatment reduces mortality to 4% and 29%, respectively. Despite advances in the diagnosis and treatment of neonatal HSV, it continues to be associated with significant long-term sequelae, inc. learning disabilities, CP, blindness, and persistent seizures.

Puzzle of the Month: Endocrine Anagrams by Dr. Pamela Gonzalez

Anagram	Hints	Answer
1. A Cad Oppositely Spits	Pituitary deficiency, blindness, mental retardation	-----
2. Lank Lam	Proper name, anosmia, eunuchoid habitus	-----
3. Pass Over Sin	Released from posterior pituitary	-----
4. Relief Knelt	Proper name, hypergonadotropic hypogonadism; gynecomastia, long limbs	-----
5. Disdained Oases	Adrenal crises during acute stress	-----
6. Sticker	In X-linked form, results from renal PO4 losses	-----
7. Induces Gym Horns	Exogenous glucocorticoid most common cause	-----
8. Ahoy Epic Clam	(+) Chvostek’s sign	-----
9. Vine or Nine Days Sitting	Clitoromegaly, labial fold masses XY karyotype, infertile aunts	-----

A Short History of Medicine

- 2000 B.C.—“Here, eat this root.”
- 1000 B.C.—“That root is heathen, say this prayer.”
- 1850 A.D.—“That prayer is superstition, drink this potion.”
- 1940 A.D.—“ That potion is snake oil, swallow this pill.”
- 1985 A.D.—“That pill is ineffective, take this antibiotic.”
- 2000 A.D.—“That antibiotic is artificial. Here, eat this root.”

— Author Unknown

“The doctor may also learn more about the illness from the way the patient tells the story than from the story itself.”

— James B. Herrick

Quote Of The Month: “Who’s gonna be the cute dumb one next year now that I am leaving?”

- Dr. Pamela Gonzalez regarding graduation from MFCH residency.

ANSWERS TO BE PUBLISHED WITH MAY 2008 EDITION