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# Pediatric Residency Newsletter

Maria Fareri Children's Hospital

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## Grand Rounds

May 7

"Recognition and  
Treatment of Elevated ICP"  
Dr. Ronald Jacobson

May 14

Sinusitis  
Dr. Moscatello

May 21

Telephone Medicine  
Dr. Jeffrey Brown

May 28

Research Symposium

Resident As Teacher

May 21

Dr. Sharon Nager

Journal Club

May 22

Dr. Eileen Rivera

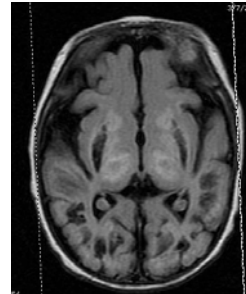
## Case 1: Leigh's Encephalopathy by Dr. Rachel Lewis

BM is a 2 month old FT NSVD female who presented with irritability, poor feeding, and inconsolable crying. Her irritability was progressively worsening over the prior 2 weeks, with recent development of vomiting, diarrhea and fever (Tmax 105 F). She had a full sepsis work-up and received one dose of Ceftriaxone prior to transfer to MFCH. Family history was significant for a sibling death at 2 months of age from "meningitis," with similar presentation. Her diet consisted of breast milk and Enfamil. On physical, she was jittery, with increased muscle tone, and notable lack of social smile. Exam was otherwise unremarkable. BM's plasma ammonia and lactate were slightly elevated. Levels of lactate and pyruvate in her CSF were normal. Head CT demonstrated extensive abnormal white matter and basal ganglia attenuation with diffuse cerebral atrophy. MRI revealed extensive T2 signal abnormality with involvement of bilateral thalami and the putamen, and diffuse cerebral and cerebellar white matter changes. She remained on antibiotics until all cultures were negative. BM was diagnosed clinically with Leigh's Encephalopathy based on brain imaging and started on Coenzyme Q10,

Levocarnitine, and Riboflavin.

Leigh's disease, a metabolic syndrome, is a subacute necrotizing encephalomyelopathy of the grey matter. Cytochrome C oxidase deficiency (autosomal recessive inheritance) and pyruvate dehydrogenase deficiency (x-linked inheritance) are two common causes; mutations of mitochondrial DNA affecting ATPase is the most common cause of maternally-inherited Leigh's

disease. Although there have been reports of late and even adult onset, it most frequently presents in the first two years of life. Clinical presentation is characterized by poor feeding, irritability, inconsolable crying, hypotonia, vomiting, ataxia, and respiratory difficulty, and commonly progresses to visual loss, hearing loss, and seizures. Death usually occurs about a year after presentation, although there are rare reports of infants living into adulthood. When associated with pyruvate dehydrogenase deficiency or mitochondrial DNA mutations, Leigh's disease is usually fatal before one year of age. Specifically, mutations in mitochondrial DNA have been linked to a constellation of symptoms (neuropathy, ataxia, and retinitis pigmentosa) known as NARP syndrome. Lab findings include lactic acidosis, elevated CSF lactate and pyruvate, and elevated CSF protein in 25% of patients. Lactate and pyruvate can also be minimally elevated in blood and urine. CT or MRI of the brain show bilateral symmetric areas of low attenuation in the basal ganglia. The most common sites are the brainstem, midbrain, medulla, pons, basal ganglia, and posterior columns of the spinal cord.



Case 1  
Leigh's  
Encephalopathy 1

Case 2  
Hirschsprung  
Disease 1

Picture of the Month/  
Inside the ICU 2

Back to the Basics 2

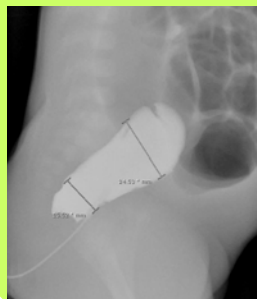
Bug of the Month 3

Director's Corner 3

Advocacy Corner 3

Medical Spanish 4

Puzzle of the Month 4



## Case 2: Hirschsprung Disease by Dr. Dayanny Langiulli

HF is a 4 wk term female who presented to MFCH ED with 1 wk of abdominal distention and infrequent stools, along with decreased activity and poor feeding. History was notable for 2 wk NICU characterized by inability to pass stool without stimulation. ED exam was notable for lethargy, abdominal distension and dehydration. Rectal exam elicited explosive, liquid stool. Barium enema showed small distal rectum and dilated proximal rectum and sigmoid colon. Definitive diagnosis of Hirschsprung disease (HD) was made by rectal biopsy. Diverting colostomy was performed with plan for definitive repair in 4 to 6 months.

HD is a developmental disorder of the enteric nervous system characterized by aganglionosis of the distal colon, leading to functional obstruction. Aganglionosis begins at the anal sphincter and continues proximally for a variable distance. HD occurs in about 1/5000 newborns (M:F, 4:1).

Nearly 20% have at least one associated anomaly, and 12% have chromosomal abnormalities (esp. Tr 21). Consider HD in any newborn with delayed passage of meconium, abdominal distension relieved by rectal stimulation/enemas, or neonatal enterocolitis. Older infants or children may have severe constipation, chronic abdominal distension, vomiting, and failure to thrive. Rectal exam reveals normal/increased tone, and may produce explosive diarrhea. Abdominal X-rays show distended bowel loops with paucity of rectal air. Small rectum and uncoordinated contractions are classic barium enema findings. Subsequent retention of contrast beyond 24 hours is also suggestive. Definitive diagnosis is made by absence of both myenteric and submucosal plexuses on full-thickness rectal biopsy. Enterocolitis is the most important complication, developing in up to 30%. Signs include explosive diarrhea, acute abdominal

distension, fever, vomiting and lethargy. As inflammation progresses, intestinal perforation risk increases. Untreated HD in infancy is associated with an 80% mortality rate, reduced to below 6% with treatment.

Initial care focuses on fluid and electrolyte management, preventing bowel overdistension and perforation, and managing septic complications. After medical stabilization, definitive treatment is surgical, removing aganglionic bowel and anastomosing normal bowel with the anus. This typically begins with diverting colostomy and, after interval growth (wt > 10 kg) definitive repair is performed. One-stage correction is possible with early diagnosis - before colonic dilatation, in short segment disease. Otherwise, a primary colostomy is warranted. Short-term post-op complications include fistula or stenosis of the anastomosis, and enterocolitis. Long-term problems include chronic constipation and

Congenital Heart Disease: Decreased Pulmonary Blood Flow

LH was a FT NSVD baby boy who presented to an outside hospital at 15 days of life with respiratory distress and cyanosis leading to cardiac arrest. Family had noted the baby seemed dusky at birth. At home, the baby was feeding well and showed no signs of respiratory distress, though he remained dusky. He was scheduled for his first PMD appointment on the day of admission. At 4 am on the day of admission, the baby woke for an uneventful feeding. At 4:30am, mom heard the baby "gasping" for breath and saw that he was cyanotic. The baby was transported by EMS to an outside hospital where he was intubated, resuscitated and transferred to MFCH. CXR done at outside hospital showed a "boot-shaped" heart. In our ED, he was cyanotic and hypotensive. PGE and dopamine were immediately started. Pt's SaO2 transiently improved, with frequent desaturations to the 40's. ABG: 7.03/55/11/14.5/-16.2/18%. An echocardiogram showed a hypoplastic right ventricle with pulmonary atresia and small pulmonary arteries. Pt was transferred to the PICU and remained desaturated despite higher doses of PGE, and he was placed on ECMO (see below). The PDA opened on Day #2 of ECMO and he was weaned off ECMO on HD#4. Echo at this time revealed a hypoplastic RV with a wide-open PDA to the RPA with a stenotic LPA. He was edematous secondary to capillary leak and weighed 2.5kg greater than admission. Pt was taken to the OR on HD#11 for a modified Blalock-Taussig shunt and a left pulmonary arterioplasty.

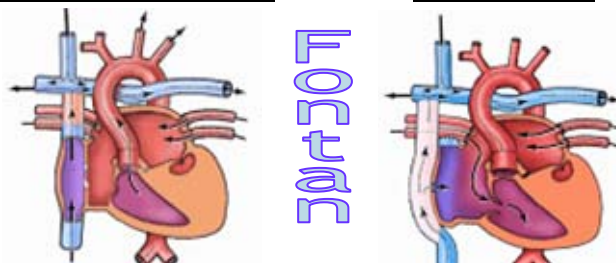
Hypoplastic right heart syndrome (HRHS) refers to underdevelopment of the right sided structures of the heart, leading to inadequate blood flow to the lungs and thus cyanosis. The major problem is a very small or hypoplastic right ventricle. Associated anatomic findings include atresia/stenosis of the tricuspid or pulmonary valve and a hypoplastic pulmonary artery. During fetal life the foramen ovale and ductus arteriosus maintain pulmonary blood flow. As these connections close, typically in the first week of life, the infant becomes cyanotic and critically ill. Diagnosis is typically made by exam including pulse oximetry and echocardiogram. Because the deoxygenated blood cannot get to the lungs through the right ventricle, it crosses into the left atrium and mixes with oxygenated blood returning from the lungs. This mixed blood is pumped out of the aorta. The only way blood gets to the lungs is through the PDA, which must be maintained open with PGE. A modified BT shunt is usually performed in the

neonatal period to maintain an aorto-pulmonary connection. Long-term the patient requires a staged "single ventricle repair", including a Glenn shunt and Fontan procedure, which does not correct the anatomic abnormality but rather allows pulmonary blood flow by bypassing the right side of the heart. At about three months of age, the child undergoes the bidirectional Glenn shunt in which the superior vena cava is attached to the pulmonary arterial system. Sometime in the next few years, the surgeon will complete the Fontan, which connects the inferior vena cava to the pulmonary artery through an intra-cardiac baffle or tunnel, or through an extra-cardiac shunt. The result is complete bypass of the right ventricle, so blood flows back from the body passively, directly into the lungs. For more info see the following website: <http://www.cincinnatichildrens.org/health/heart-encyclopedia/anomalies/sv.htm>



Intracardiac baffle or tunnel

Extracardiac shunt



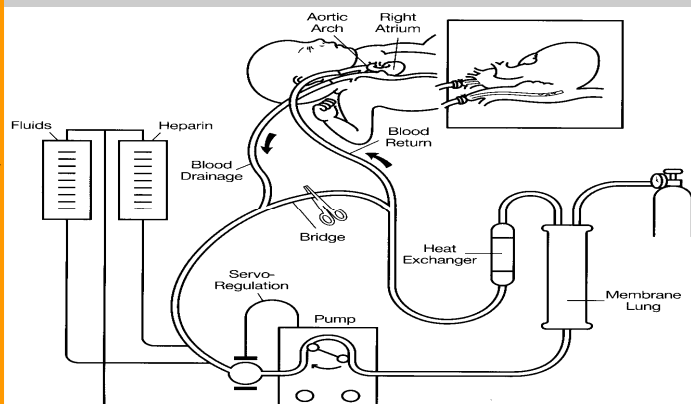
Single Ventricle Repair (Hypoplastic Right or Left Heart Syndrome)

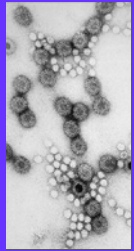
Back to the Basics: ECMO  
By Dr. Kristen Woodard  
and Dr. Lauren Mendelsohn

ECMO (extracorporeal membrane oxygenation) has its roots in cardiopulmonary bypass, where a machine acts as an artificial heart and lung during surgery, removing blood from the venous system and returning oxygenated blood via a pump to the aorta. Technology has adapted to allow for longer periods of time on ECMO, as well as for return of oxygenated blood to the venous system in patients with functioning hearts. There are several pediatric indications and criteria for consideration of ECMO.

Diagram (right) demonstrates the basic components of the ECMO circuit. Vascular cannulas are placed into a large vein and artery (often the IJ vein and carotid artery) for blood drainage and reinfusion. Blood is drawn from the venous canula into the pump by negative pressure. A small drainage reservoir (bladder) helps to ensure continuous availability of blood for the pump without relying on second-to-second flow rate. Flow (volume/time) through the pump determines the circuit's "cardiac output" and can be adjusted to allow the patient's own heart to contribute varying degrees to overall blood flow. For example, if the patient has poor cardiac function, the flow rate must be increased to permit the patient's heart to rest while maintaining adequate perfusion. Once the blood is pumped, the artificial lung (membrane oxygenator) provides gas exchange by both oxygenating and

"ventilating" for the patient. The rate of blood flow through the oxygenator determines carbon dioxide exchange and can be adjusted to allow more or less retention of carbon dioxide. The heat exchanger maintains a controlled temperature of reinfused blood. Other circuit components allow for infusion of medications, incorporation of a hemofilter for fluid control, and introduction of heparin to prevent clots within the circuit.





Bug of the Month  
by Dr. Daniel Mooney

Rotavirus (RV) is a double-stranded RNA virus belonging to the Reoviridae family. RV is ubiquitous, but strikes particularly hard at children in developing nations. Worldwide, RV accounts for 25% of all diarrheal-related deaths, or 6% of all deaths in children younger than 5 years of age. In the U.S., RV accounts for fewer than 40 pediatric deaths annually. However, even in the U.S., RV is responsible for an estimated 500,000 outpatient visits and 50,000 hospitalizations yearly. The resulting annual financial burden on the U.S. health-care system is an estimated \$200 to \$500 million.

In the U.S., RV infection commonly strikes during the winter months (November - May), first appearing in the Southwest, and spreading to the Northeast. RV is classically spread by fecal-oral transmission with nearly all children infected by 3 years of age. Infectious particles are shed in the stool of infected persons, starting before the onset of symptoms and persisting for up to 10 days after symptom appearance. Fomites serve as important vectors since RV remains infectious for a prolonged period on a number of surfaces, especially nonporous surfaces such as metal and plastic.

RV primarily infects cells of the small intestinal villi and leads to malabsorption and excessive fluid loss from the intestine. Symptoms usually begin within 2 days of exposure and include anorexia, low-grade fever, watery/non-bloody diarrhea, vomiting, and abdominal cramps. Stool output can be copious during the diarrheal phase of illness, and dehydration is a common presenting complaint. Physical exam findings are often unremarkable except for signs of dehydration and hyperactive bowel sounds. Extraintestinal manifestations, especially respiratory disorders, occur in 20% to 50% of infected children, and otitis media complicates as many as 20% of RV cases. Common complicating features of RV acute gastroenteritis (AGE) include dehydration, electrolyte disturbances, metabolic acidosis, nutritional deficiencies, and diaper rash. Rare complications include gastric rupture and central pontine myelinolysis. In immunocompromised children,

extraintestinal spread is more common; a state of chronic diarrhea may occur, and infection is more likely to result in significant morbidity and mortality. In neonates, a wide range of illness may occur, with an estimated 30-40% of necrotizing enterocolitis cases thought to be related to RV infection.

Treatment of RV AGE should address hydration, nutrition, and symptomatic relief. Routine labs may not be necessary, but assessment and correction of hypoglycemia and electrolyte disturbances, especially hyponatremia, hyponatremia, acidemia and hypocalcemia, may be indicated. Presence or absence of an elevated anion gap may help determine if acidosis is due to dehydration-related perfusion disturbances or diarrheal bicarbonate loss. In the latter case, bicarbonate or citrate-supplemented oral rehydration solution (ORS), if tolerated and clinically appropriate, should be considered. Finally, barrier creams and pastes are appropriate for the diaper rashes that commonly accompany diarrheal illness.

Pediatricians should teach parents about modes of transmission and prevention. The importance of thorough handwashing along with disinfection of fomites with commercially available disinfectant spray should be emphasized. Hospitalized children require contact precautions.

Since the withdrawal of the original RV vaccine in 1999 due to an association with intussusception, two RV vaccines have been licensed for use in North America. RotaTeq (Merck), is a live-virus, pentavalent vaccine based on the bovine RV strain, administered in three doses, typically at 2, 4, and 6 months of age. Children should receive the 1<sup>st</sup> dose between 6 and 12 weeks of age and the 3<sup>rd</sup> dose no later than 32 weeks of age. In the prelicensure clinical trial, efficacy was 74% for AGE of any severity and 98% against severe AGE. Vaccination was associated with an 86% reduction in clinic visits, and over 95% reduction in hospitalizations. Post licensure surveillance during the first two years has not revealed an increased risk of intussusception. Rotarix, a monovalent RV vaccine based on human Rotavirus, requires 2 doses and has been recently approved.

## DIRECTOR'S CORNER

Thank you to all residents, former residents, Beth, Harriet and families for joining Team Dr. Owsy Resident- at *Go the Distance 2008* on April 13. The team raised over \$7100, with special kudos to all of our fund raisers. The next step is to decide how the money can best be spent to enhance resident education. In the past we purchased equipment for the conference room, but other options include seed money for research or advocacy projects conducted by the residents. Please forward your ideas to me or one of the chiefs.

Speaking of advocacy projects, it has been several months since we started the Advocacy Task Forces and feedback from some of the projects has been excellent. I would like to invite a representative from each task force to briefly describe your project to the new PL1's at orientation. This will occur during lunch hour. You will have the opportunity to recruit new PL1's to join your task force, allowing the projects to continue for the 2008-2009 residency year.

Lastly, a reminder to please complete the *SPACER* survey. This is our program's opportunity to participate in a national multi-residency research project through CORNET. CORNET is the Academic Pediatric Association's Continuity Research Network. This is your opportunity to participate in a scholarly activity, see what types of questions can be used in educational research projects and, perhaps, stimulate ideas for future projects.

*Terry Hetzler, MCD*

## Advocacy Corner

By Dr. Emily Koelsch

Welcome to another edition of the Advocacy Corner at Maria Fareri Children's Hospital. Many of you may not know that in September 2007 the College Cost Access and Reduction Act was eliminated. This act included the "20/220 pathway" which had allowed medical residents to defer their student loans if the debt burden was greater than 20% of their income and the income minus the debt burden was not greater than 220% of the federal poverty level. Despite initial attempts to encourage the Department of Education to reinstate the 20/220 pathway into the Higher Education Act Reauthorization, it appears that Congress will eliminate the pathway completely by July 1, 2009. The alternative for medical residents would be to enter the Income-Based Repayment program which would require us to pay small monthly loan repayments. Without loan deferment, many residents may be discouraged from entering public service, medical education and research, or working in underserved populations, simply to survive financially. I have placed a letter drafted by the AAP Federal Affairs Office on the P drive. Please email, fax or call your legislators to let them know how important this issue is to all of us. The easiest way to find the contact information for your legislator is to visit [www.congress.org](http://www.congress.org) and type in your zip code.

I also wanted to congratulate Dayanny Langiulli on being elected as a delegate representing Westchester Medical Center on the CIR. We are looking forward to having our voice heard in a hospital

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

## Gastrointestinal System (Sistema Gastrointestinal)



Hello, I'm Doctor Gerd  
Hola, soy el Doctor Gerd

How is your child feeling today?  
Cómo se encuentra el niño hoy?

Is your child vomiting?  
El niño está vomitando?

When did the vomiting start and how frequent is it?  
Cuándo comenzó con los vómitos & cuán frecuente son?

Is the vomit green or bloody?  
El vómito es verde o contiene sangre?

Does your child have diarrhea?  
El niño tiene diarrea?

Since when and how often?  
Desde cuándo & cuán frecuente?

Is the diarrhea watery/ bloody/ mucousy?  
La diarrea es bien líquida o contiene sangre/moco?

Is your child eating or drinking?  
El niño está comiendo bien/ tomando líquidos?

Does your child have decreased urine output? How many wet diapers?  
El niño está orinando bien? Cuántos pañales le ha cambiado?

Does your child have abdominal pain?  
Tiene dolor de estómago?

Has your child have fever? How high?  
El niño ha tenido fiebre? Cuánto de temperatura?

Has there been any recent travel?  
Han viajado recientemente?

Is your child irritable, listless, have decreased activity?  
El niño está más irritable, decaído, hipoactivo?

Has your child had a rash?  
Ha tenido algún tipo de erupciones/rash en la piel?

Is there anyone else sick at home or daycare?  
Alguien más enfermo en la casa o en el cuidado?

What did your child eat before starting to have these symptoms?  
Que comió el niño antes de comenzar los síntomas?

What type of formula does the baby drink?  
Que fórmula toma el niño?

Have you given the child Pedialyte? How much? Is he tolerating it?  
Ha tratado de hidratar al niño con Pedialyte? Cuánto? Lo tolera?

**Quote Of The Month:** "Can the patient with the infection of the penis gland be put in a swing room?" - Question from Bed Management to Emergency Room Department.

## Puzzle of the Month: Allergy-Immunology Word Fragments by Dr. Pamela Gonzalez

In each of these puzzles, a word fragment is given, and you must think of the allergy-immunology-related term that contains the fragment -- that is, you must form a word by adding letters to the beginning and/or the end of the fragment. You may not add letters to the middle of the fragment, nor may you rearrange the letters given. Clues are provided for each.

Fragment	Clue
1. TICAR (9 letters)	pruritic wheals; type I HSR
2. PHYL A (11 letters)	severe, widespread mast cell degranulation; ↑ tryptase
3. SINOPHI (12 letters)	most likely 2° allergy/asthma; infection, drug rxn; neoplasia
4. EORG (8 letters)	syndrome; proper name; ↓ Ca; (-) 22q11; thymic hypoplasia
5. RUT (6 letters)	proper name, no B-cells; ↓ IgA, IgG, IgM; pyogenic orgs; enteroviral infection
6. CKNES (2 wds; 13 letters)	Type III HSR; immune-complex mediated vasculitis
7. RBILL (12 letters)	most common cutaneous drug eruption
8. LK (4 letters)	common cause of food allergy
9. OSTMA (8 letters)	proper name; cyclic neutropenia; stomatitis; abscess
10. PLEME (10 letters)	↓ early component → infection w/encapsulated orgs; ↓ late component → Neisseria infection

ANSWERS TO BE PUBLISHED WITH JUNE 2008 EDITION

Answers from April 2008 Edition: 1. Septo-Optic Dysplasia, 2. Kallman, 3. Vasopressin, 4. Klinefelter, 5. Addison Disease, 6. Rickets, 7. Cushing Syndrome, 8. Hypocalcemia, 9. Androgen Insensitivity