

The National MedPeds Residents' Association

Newsletter

Vol. 2, No. 2 Spring 2000

(Sponsored in part by Weatherby Health Care)

MedPeds Scholarships

The mission of the National MedPeds Residents' Association has always been to support MedPeds residents. In an attempt to recognize outstanding residents and provide some financial support for their activities we have spent the last year setting up several scholarships. The following were awarded this year. To apply for next years awards please submit a letter of interest, a cv, and a letter of recommendation to NMPRA@hotmail.com.

Howard Schubiner Award

This award honors a MedPeds resident physician who has made an extraordinary, lasting contribution to the success of NMPRA and MedPeds. It is named for Howard Schubiner, MD, for his notable, extraordinary, lasting contributions to MedPeds. In the words of Dr. Schubiner "a resident who exemplifies the highest standards for excellence in MedPeds, including exemplary clinical care of patients, compassion and humanism in relationships with patients and with colleagues in medicine and hospital co-workers, involvement in community activities, and contributions to the field of Medicine? Pediatrics."

1999-2000

William F. Grella, MD

*St Joseph's Hospital & Medical Center
Paterson, NJ*

The winner receives \$1000 sponsored by CompHealth, Inc. (www.CompHealth.com). The award is presented annually at the National MedPeds Conference/NMPRA Annual Meeting.

Gary Onady Award

This award honors a MedPeds resident physician who has made an extraordinary, lasting contribution to the success of NMPRA and MedPeds. It is named for Gary Onady, MD, for his notable, extraordinary, lasting contributions to MedPeds.

1999-2000

Heath Parker, DO

*Texas A & M Univ - Scott & White Hospital
Temple, TX*

The winner receives \$2000 sponsored by Weatherby Health Care (www.whcfirst.com). The award is presented annually at the time of the annual MedPeds Program Director's Meeting.

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Weatherby Health Care:

NMPRAâ€™s Newest Supporter

NMPRA is pleased to announce a new relationship with Weatherby Health Care. They have initiated a scholarship program, become the primary supporter of *The MedPeds Newsletter*, and will be providing our residents with much needed publications, advise, and help with job placement.

MedPeds Clinical Corner

Treatment of Hepatic Complications of Visceral Niemann-Pick Disease in a 2 year old by Orthotopic Liver Transplant

Jeff Bates MD, David Easley MD, & James Daniel MD

Department of Internal Medicine & Pediatrics
Texas A & M University - Scott & White Memorial Hospital Temple, TX

Case Report

Niemann-Pick disease (NPD) is a heterogeneous group of liposomal storage diseases caused by sphingomyelinase deficiency leading to deposits of sphingomyelin in the brain, liver, spleen, lungs, bone marrow and other visceral organs. In the rarer Type 1b form of the disorder there is lack of the devastating nervous system involvement and the disease course is often less severe. Although massive hepatosplenomegaly is often a presenting feature, significant compromise of liver function is uncommon. We report here a patient with an unremarkable prenatal, birth, and family history with no known

consanguinity who developed severe hepatic complications.

Initial presentation to clinic was with diarrhea and dehydration at 6 months of age. Development of jaundice several days later led to further investigation. Laboratory values revealed Aspartate aminotransferase (AST) 413 IU/L (normal values for our lab 10-42 IU/L), Alanine aminotransferase (ALT) 117 IU/L (nl 10-60 IU/L), Alkaline Phosphatase 414 IU/L (nl 42-360 IU/L), Total Bilirubin 10.5 mg/dL (nl 0.2-1.0 mg/dL), Direct Bilirubin 5.8 mg/dL (nl 0.0-0.2 mg/dL), Prothrombin time 16.8 sec (nl 10.0 - 12.1sec), and International Normalized Ratio 1.6 (nl 0.9 -1.1). Referral was made to pediatric gastroenterology. Hepatosplenomegaly was appreciated prompting further work up including percutaneous needle biopsy of the liver.

On light microscopy, multiple Kupffer cells were seen with increased vacuolation. Vacuoles were negative for glycogen. No obstructive changes were seen. Orcein, periodic acid-Schiff (PAS) and iron stains were unrevealing. The biopsy also demonstrated early cirrhosis. Electron microscopy revealed normal glycogen and large droplet fatty changes. Multiple dilated lysosomal cavities containing laminated myelin-like material were seen. Findings were consistent with NPD. Confirmatory assay using peripheral leukocyte sphingomyelinase activity was 0.07 micromoles/mg protein/hr (i.e. less than 10% of normal activity) compatible with the diagnosis of NPD type I. On evaluation by pediatric neurology the patient had a normal neurological evaluation and no signs of developmental delay, consistent with a diagnosis of NPD type Ib.

Subsequent clinical course included moderate growth failure, mild developmental delay, multiple episodes of anemia requiring blood transfusions, and the development of portal hypertension as demonstrated on Doppler ultrasound. With continued deterioration of hepatic function, the patient was evaluated for liver transplantation. After initial evaluation, patient demonstrated progressive somnolence and irritability with hyperammonemia of 79 UMOL/L (nl 11-35 UMOL/L) consistent with hepatic encephalopathy. Orthotopic liver transplant and splenectomy were performed at 2 years of age with subsequent marked clinical improvement. She has had complete resolution of her growth failure and developmental delay. There has been no recurrence of hepatic involvement or disease as evidenced by normal clinical exam, laboratory studies and multiple biopsies after transplantation. She is now 2 years post transplant and doing well.

Editorial:

This case is interesting from a MedPeds point of view for two reasons.

First, Niemann Pick disease is generally thought of as a pediatric zebra because the common Type A usually leads to death in the first few years of life. However, Type B is less severe and numerous cases have been reported in the literature with no diagnosis made until adulthood, and is thus referred to as the Adult type by many.

Second, this particular case is of the common "adult type" but the patient presented in the first year of life.

Highlights of Previous Issues of *The MedPeds Newsletter*

Study shows we are mostly Primary Care¹

A new survey of the MPPDA shines the light on all those questions about what we are going to do after residency. Seventy-four program directors provided information on their 708 MP graduates from the years 1987 to 1993, which showed that 68% of MP graduates are practicing as generalists. You can reassure all those doubters that 85% do practice both internal medicine and pediatrics. Another 7% found their niche in the ER and 21% entered subspecialty training. Of the subspecialists, half continue to see both adults and pediatric patients. If you were wondering how we compare with other programs, it seems that only 35-45% of internal medicine graduates and 67% of pediatric graduates go on to practice primary care. Family practice holds out with 95%, but that argument is another article in itself.

The Stress of "The Switch"²

A survey of Med/Peds programs in 1989 showed a trend in which programs that switch disciplines on a more frequent basis (i.e., every 1-4 months) are less stressful on the residents than those that make "the switch" only once or twice a year. This survey also showed that the stress of "the switch" was usually worse in the first 24 months of training and steadily improved in the 3rd and 4th years. While 35% reported no differences in switching from IM to Peds vs. switching from Peds to IM, 37% reported more difficulty in switching from Peds to Internal Medicine. (Only 18% reported more difficulty in switching from Medicine to Peds.) The most common problems associated with "the switch" were the various differences in diagnoses, treatment decisions, and drug dosages. Those few programs that still switch after 6, 8 or more months may want to reevaluate these numbers and speak with residents from other programs about their experience.

Important Program Features²

A 1989 survey looked at the features residents felt were most important in the selection of a Med/Peds program. The top three issues listed were:

Ambulatory training sites

Having a specific coordinator for the program
University affiliation

These were followed in importance by Med/Peds faculty role models, a tertiary referral center, a combined Med/Peds clinic, the presence of a Children's Hospital, and community based ambulatory training sites.

MedPeds Applicants²

MedPeds residents were asked in a 1989 survey about residency programs they had inquired about, applied to, or ranked, in addition to MedPeds. Nearly half of all those surveyed had also inquired about Internal Medicine alone, while only about one-third had also inquired about Pediatrics alone. About one-third also inquired about family practice. Interestingly another 9% inquired about other programs. When the final rank list was turned in by those who became MedPeds residents, 28% also ranked Internal Medicine alone, 18% also ranked pediatrics alone, and 14% also ranked family practice. Only 2% also ranked a program not listed above.

Perceived Competencies³

An interesting look at how practitioners view themselves and their abilities was published in a 1993 article. These perceived competencies were compared among all of the primary care fields; Med/Peds, family practice, internal medicine, and pediatrics. As is often the case in life, those who feel most competent are not necessarily the best. In fact, they can be the most dangerous as they often don't know their own limitations. All things considered and with a grain of salt, the study still provides some interesting information.

The specific differences that stood out were that family practitioners felt significantly less comfortable than Med/Peds when it came to a "complex delivery room case". FP's were also less comfortable than med/peds with specific complex internal medicine problems. Med/Peds had higher confidence ratings in areas of complex cases of neonates and adults. Interestingly enough, there were no issues in which pediatricians felt more comfortable than Med/Peds. However, this was not true for internal medicine issues.

The flip side was as follows. FP's were more comfortable than Med/Peds with the routine care and psychosocial aspects of adolescent health care. There were also several issues in intensive care management of acute MI with which internal medicine physicians were more comfortable than Med/Peds.

While it is hard to draw conclusions from this study perhaps this can help us to guide our curriculum in these areas of less perceived confidence. While the guidelines already require a full month of adolescent care, the required ICU experiences are limited to 4 months in each field. These months should be maximized.

1. Schubiner H. Current positions of graduates of internal medicine-pediatrics training programs. Arch Pediatr Adolesc Med. 1997; 151:576-579.
2. Schubiner,H, Schuster, BL, Moncrease A, Mosca C. The perspectives of current trainees in combined internal medicine-pediatrics: results of a national survey. AJDC 1993;147:885-889.
3. Biro FM, Siegel DM, et al. A comparison of self-perceived clinical competencies in primary care residency graduates. Pediatric Research. 34(5):555-9, 1993 Nov.

Visit our All New Website at: www.MedPeds.org

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(The views expressed in this newsletter are those of the authors and not necessarily those of NMPRA)