**Abstracts: Poster Sessions**

**P45**

**HYPOGAMMALGLOBULINEMIA IN DIABETICS WITH NEPHROTIC SYNDROME, S.S. LANE MD, 63 EISENBERG MD, HOUSTON, TX.**

Nephrotic syndrome (NS) results when the glomerular filtration system becomes damaged by disease leading to complications including proteinuria, hyperlipidemia, edema and hypogammaglobulinemia. We report 2 cases of diabetics with NS, low IgG and severe infections.

The 1st is a 56 y/o woman with frequent bouts of severe bronchitis with various organisms (including Pseudomonas aeruginosa) as well as sinusitis requiring endoscopic surgery. She has developed chronic bronchitis and steroid dependent asthma and is currently responding well to therapy with IVIG. Her IgG is 548 mg/dl (IPG=315, 2480, J=81, k=17) and urine protein is > 3gms.

The 2nd is a 69 y/o woman who requires prolonged courses of IV antibiotics to clear her bronchial infections. She becomes severely dyspneic with thick purulent sputum and has once required mechanical ventilation. Her IgG is 530 mg/dl (IPG=386, 2856, J=45, k=71) and urine protein is > 5gms/24 hours.

NS with low IgG occurs more frequently than reported and should be considered in diabetics and renal patients with recurrent infections. IVIG may be of benefit.

**P46**

**SEVERE COMBINED IMMUNODEFICIENCY: A CASE REPORT S. GONZALEZ MD, C. GALINDO MD, G. ALCALA MD, M. GARZA MD, J. EZQUIELO MD, H. MORENO MD, C. CANSECO MD, MONTERREY, NUEVO LEON, MEXICO.**

We report a 4 month old Latin boy with very low values of immunoglobulins and failure to thrive, since the first month of life. He had intractable diarrhea, chronic oral moniliasis and growth retardation. He had severe hospitalizations, medical history: breast fed exclusively for the first month of life, at the end of this period he had delayed separation of the umbilical cord and granuloma in this area. Chronic diarrhea with 6-8 evacuations per day and abdominal distention, that improved with soy milk formula to 3 evacuation per day.

Continued in this way for the next 3 months and at the age of 4 months he had growth retardation, (5.500 KG, 63 CM) and an enema with disintegration RASH, he was hospitalized for 18 days and his LABORATORY DATA REVEALED: PERIPHERAL EOSINOPHILIA, ABSENCE OF IgA AND IgE LEVELS, VERY LOW LEVELS OF IgG AND IgM, At CELL COUNT of 24 (65-90), CD8 22(62-32), CD4 8 (42-58), CD4/CD8 1.10/23, CD20: 42 (0-10), CD56 39/30, 7). A BIOPSY OF GASTROINTESTINAL TRACT SHOWED CHRONIC INFLAMATION - AND A SKIN BIOPSY WITH RECENT HEMORRHAGIC AREAS. HE RECEIVED IgG IV (IGAMAGARD) AND ANTIBIOTIC THERAPY FOR 14 MONTHS WHEN HE DIED.

**P47**

**LATE DIAGNOSIS OF X-LINKED AGAMMALGLOBULINEMIA WITH NEUTROPENIA IN A 7 YEAR OLD BOY, M. Garcia MD, C. Ob, MD, and C. Sone MD, Torrance, CA.**

The mean age of diagnosis of new XLA in previously unaffected families is 3½ years. We report a boy diagnosed with XLA at 7½ years of age. He presented with draining otitis media, pneumonia and neutropenia. He had history of recurrent otitis media since 8 months of age, pneumonia at 8 months and skin infection at 5 years. Laboratory results at the time of diagnosis: IgG<35mg/dl, IgM 17mg/dl, IgA<0.7mg/dl, IgE undetectable, total B cell<1%, absolute B cell<1% total T cells 91%, CD8 49%, CD4 41%, WBC 7.1 with absolute neutrophil count of 203. Patient responded to IVIG and systemic antibiotics. Neutropenia resolved within 2 weeks. The recurrent infections have led this patient to several outpatient medical services which failed to identify his primary immune problem.

We conclude that an index of suspicion regarding XLA should be maintained in boys with recurrent infections despite normal appearance and growth parameters.

**P48**

**SPECTRUM OF IMMUNODEFICIENCY IN THE FAMILY OF AN IgA DEFICIENT CHILD, A.L. HIRSCH MD, T.P. WATKINS MD, PhD, T.P., W. SCHWARTZ MD, PhD, BIRMINGHAM, AL.**

IgA deficiency (IgAD), characterized by the absence of serum IgA, is the presence of IgA- B cells, is the most common primary immunodeficiency in the United States. We have previously reported that IgAD and Common Variable Immunodeficiency (CVI), characterized by panhypogammaglobulinemia and normal numbers of peripheral B cells, represent the peripheral ends of the same spectrum of immunodeficiency.

These disorders often occur in members of the same family. In order to better define the spectrum of immunodeficiency within families of IgAD patients, we are in the process of extensive family studies. We now report a family wherein all 6 of the proband and his mother both have selective IgAD complicated by recurrent sinopulmonary infections. The proband's grandmother is clinically asymptomatic, but has borderline IgM, IgG, and IgA levels. She responded adequately to intranasal and pneumatic. The proband's maternal granuloma suffers from recurrent sinopulmonary infections, including pneumonia. Her IgG (179 mg/dl), and IgG (332 mg/dl) levels are just below the normal range, her IgG3 level is normal (123 mg/dl).

However, she failed to respond to pneumococcal and tetanus polysaccharide antibody responses but did within the spectrum of immunodeficiency characterized by familial IgAD and CVI.