AN OFFICIAL JOURNAL OF THE AMERICAN COLLEGE OF RHEUMATOLOGY

Arthritis & Rheumatism

VOLUME 54, NUMBER 9 (SUPPLEMENT) SEPTEMBER 2006

ABSTRACT SUPPLEMENT

2006 ANNUAL SCIENTIFIC MEETING

November 10–15, 2006
Washington, DC

ACR Criteria	Frequency positive		Frequency positive before age 18	
	n	%	n	%
Antinuclear Antibody	105/109	96.3	76	72.4
Lymphopenia	104/111	93.7	65	62.5
24 hr Urine for Protein >500 mg	35/42	83.3	19	54.3
Anti-DNA ab	82/109	75.2	53	64.6
Arthritis	84/114	73.7	70	83.3
Leukopenia	78/110	70.9	51	65.4
Urine Protein to Creatinine Ratio >5	31/47	66.0	24	77.4
Malar Rash	63/114	55.3	47	74.6
Proteinuria >3+	46/106	43.4	32	69.6
Anticardiolipin ab	32/77	41.6	24	75.0
Anti-Smith ab	29/74	39.2	23	79.3
Urine Cell Casts	37/99	37.4	26	70.3
Mucosal Ulcers	42/114	36.8	32	76.2
Photosensitivity	38/114	33.3	25	65.8
Pleuritis	27/114	23.7	15	55.6
Thrombocytopenia	25/112	22.3	17	68.0
Pericarditis	24/114	21.1	16	66.7
Discoid Rash	23/114	20.2	16	69.6
Seizures	23/114	20.2	12	52.2
Hemolytic Anemia	15/114	13.2	12	80.0
Serositis	13/114	11.4	9	69.2
Lupus Anticoagulant	12/114	10.5	7	58.3

Conclusions: Although all ACR criteria occurred more frequently before 18 than after 18, seizures and pleuritis occurred with almost equal frequency in both stages. The results for 24-hour urine protein, psychosis, and false positive syphilis test in particular are influenced by a small number of cases.

Disclosure: L. Rossello, None; **C. Drenkard**, None; **K. Easley**, None; **N. Shenvi**, None; **L. Vogler**, None; **S.S. Lim**, None.

1723

Childhood Systemic Lupus Erythematosus in Latin America. The GLADEL Experience in 230 Children. Luis A. Ramirez-Gomez[1], Oscar Uribe[1], Oscar Osio[1], Hugo Grisales[1], Mario H. Cardiel[2], Daniel Wojdyla[3], Donato Alarcon-Segovia[4], Bernardo A. Pons-Estel[5]. [1]Universidad de Antioquia, Medellin, Colombia; [2]Hospiatal General Dr Miguel Silva, Morelia, Mexico; [3]Escuela de Estadistica, Universidad Nacional de Rosario, Rosario, Argentina; [4]Instituto Nacional de Ciencias Medicas y Nutricion Salvador Zubiran, Mexico, Mexico; [5]Hospital Provincial de Rosario, Rosario, Argentina

Purpose: It has been described that systemic lupus erythematosus (SLE) clinical manifestations change according age of onset. Demographic, clinical, classification criteria, disease severity, organ damage and mortality in 230 children seen in GLADEL cohort (Grupo Latinoamericano De Estudio de Lupus) are presented.

Methods: This cohort study was conducted in 34 research centers in 9 Latin American countries. It comprises a careful follow up of 1214 SLE patients with an early diagnosis of SLE (< 2 years of disease duration) and followed up with a common protocol in a standarized format. Quality of information was centralized in a single center.

Results: In this cohort 230 SLE patients were younger than 18 years and were compared with 884 adults SLE patients. Malar rash (70.4 vs. 59.1%, p=0.002), fever (63.5 vs. 55.2%, p=0.022), oral ulcers (49.1 vs. 39.9%, p=0.011), thrombocytopenia (25.2 vs.17.8%, p=0.01) and hemolytic anemia (16.1 vs. 10.8%, p=0.024) were more prevalent in children with statistical significance. On the other hand, myalgias (18.9 vs. 11.7%, p=0.010), Sjögren's syndrome(9.3 vs. 3.9%, p=0.007) and cranial nerve involvement (4.2 vs. 1.3%, p=0.037) were more prevalent in adults.

Some differences were seen in children according ethnic group. Afro-Latin-American children had a higher prevalence of fever, thrombocytopenia and hemolytic anemia. White and mestizo children had higher prevalence of malar rash. Mestizo SLE children had higher prevalence of cerebrovascular disease and cranial nerve involvement.

Children had an earlier fulfillment and more SLE criteria (6+/−1.59 vs. 6+/−1.50 p=0.009). They also had higher disease activity scores (13+/−8.4 vs.11+/−8.29 p=0.011) whereas adults had higher damage (0+/−0.95 vs 0+/−1.09, p=0.021).

Conclusions: SLE in pediatric Latin American patients has more severe presentation when compared with adults and differences can be detected among ethnic groups.

Disclosure: L.A. Ramirez-Gomez, None; **O. Uribe**, None; **O. Osio**, None; **H. Grisales**, None; **M.H. Cardiel**, None; **D. Wojdyla**, None; **D. Alarcon-Segovia**, None; **B.A. Pons-Estel**, None.

1724

Utility of Cardiac Monitoring in Fetuses at Risk of Congenital Heart Block: The PR Interval and Dexamethasone Evaluation (PRIDE) Prospective Study. DM Friedman[1], MY Kim[2], JA Copel[3], C. Davis[4], JP Buyon[4]. [1]Saint Barnabas Medical Center, Livingston, NJ; [2]Albert Einstein College of Medicine, Bronx, NY; [3]Yale University School of Medicine, New Haven, CT; [4]NYU School of Medicine, New York, NY

Anti-Ro/La-associated congenital heart block (CHB) carries a 20% mortality, and nearly all surviving children require pacing by adulthood. Moreover, 3rd degree block, once established, has never been reversible. Accordingly, the PRIDE Study was initiated to identify an early marker of cardiac injury prior to permanent scarring. The protocol required fetal echocardiograms weekly from 16–26 wk gestation, and biweekly from 26–34 wk. PR interval >150 msec (normal mean + 3 SD) was considered abnormally prolonged consistent with 1st degree block. Inclusion required maternal anti-Ro Ab titer >30 EU (normal <19); mean titer was 11,000 EU and most subjects showed reactivity with recombinant protein and immunoblot. Mothers were excluded if taking >10 mg/day prednisone. 118 mothers were initially enrolled with 88 completing an evaluable course (14 voluntary drop, 3 fetal demise before first echo, 7 low titer anti-Ro, 1 prednisone >10mg/day, 5 still pregnant). 82 fetuses (93%) had normal PR intervals throughout the study. No cases of 1st degree block developed after a normal EKG at birth. Neonatal lupus (NL) developed in 9 cases, 3 of which were NL rash only. 3 fetuses had 3rd degree block. The first had normal PR intervals from 16–22 wk with 3rd degree seen at 23 wk, although tricuspid regurgitation (TR) was noted at 17 wk and an unexplained atrial density at 22 wk. Despite 4 mg/day maternal dexamethasone (dex), the fetus was terminated at 24 wk with severe hydrops. The second had normal PR intervals from 16–18 wk, missed 19-wk echo, with 3rd degree block seen at 20 wk and TR noted at 17 wk. 3rd degree block persisted despite dex and the child is currently 8 mo old. The third had normal PR at 18 wk but at 19.5 wk had 3rd degree block with severe hydrops, and was terminated at 20.5 wk despite 4 mg/day maternal dex. 3 fetuses had 1st degree block detected by Doppler echo only. The first had normal PR from 16–18 wk, with PR 165 msec at 19 wk that resolved within 7 days of 4 mg dex; EKG was normal at birth. The second had normal PR at 19 wk, missed 20-wk echo, and at 22 wk had PR 160 msec that resolved within 3 days of 4 mg dex; EKG normal at birth. The third had normal PR through 30 wk (103 msec) and at 32 wk was born with EKG showing 1st degree block that has persisted through followup at 3 yr. Overall, CHB occurred in 3 (19%) of 16 mothers with a previous CHB child and 3 (5%) of 56 with no previous NL. In sum, 1st degree block may be reversible with dex, supporting the utility of close monitoring, especially given the increased recurrence rate substantiated in this study. Advanced block and cardiomyopathy can occur within 1 wk of a normal echocardiogram, and even weekly echos may not reveal an early marker, although TR merits attention as an early sign of injury. Finally, 3rd degree block is not reversible despite early intervention. Therefore, a prophylactic treatment is needed.

Disclosure: D. Friedman, NIH-NIAMS Grant No. AR46265, 2; **M. Kim**, NIH-NIAMS Grant No. AR46265, 2; **J. Copel**, NIH-NIAMS Grant No. AR46265, 2; **C. Davis**, NIH-NIAMS Grant No. AR46265, 2; **J. Buyon**, NIH-NIAMS Grant No. AR46265, 2.

1725

Combination of Rituximab And Cyclophosphamide for the Treatment Of Chilhood Onset Systemic Lupus Erythematosus. Lilliana Barillas-Arias, Alexa B. Adams, Sheila T. Angeles, Emma J. MacDermott, Laura Barinstein, Thomas JA Lehman. Hospital For Special Surgery, New York, NY

Objective: To describe an open label prospective case series of 12 patients with childhood onset lupus treated with B-cell depletion therapy.

Background: Childhood onset lupus has significant morbidity despite aggressive immunosuppressive therapy. Newer, safer, and more effective treatment is needed. B cell lymphocytes play an active role in the pathogenesis of autoimmune diseases, and B-cell depletion therapy has been used successfully in various rheumatologic conditions. There have been promising