Mortality in Children with Juvenile Systemic Lupus Erythematosus in an Argentine Pediatric Centre.

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Background: Juvenile systemic lupus erythematosus (JSLE) is a complex multisystemic autoimmune disease, often causing irreversible damage, reduced quality of life and life expectancy. Data on causes of death and time trends in infant lupus mortality are limited.

Objectives: The aim of this study was to determine mortality in a pediatric cohort.

Methods: This retrospective study included patients with childhood onset lupus (fulfilling ACR 1997) who were diagnosed at the Paediatric centre in Santa Fe Argentina, Alasius Children Hospital from 1991 to 2018 and had 15 years of age at presentation.

Results: This study included 54 JSLE children (F:M=12.5:1) with a mean age of onset lupus 12 years (range 4-15 years). During a mean follow-up in our centre of 3 years (range 12m–10 years), 12 patients (24%) were died; 5 during the follow-up in our centre and 7 in adult centre. 4 lost to follow-up. The average age of death was 16 years (range 10-29 years) and the average since diagnosis was 17 months (range 3m-15 years).

The principal system involved was renal (100%). 10 patients with diffuse proliferative lupus nephritis (WHO class IV), 2 membranous lupus nephritis (WHO class V), 5 patients (41.6%) with hematological disorders (2 pancytopenia, 2 leukemia, 1 Macrophage Activation Syndrome). 3 patients had thrombosis. 5 patients (41.6%) with neurological involvement (4 psychosis, 1 depression). One patient with liver dysfunction. There one death related to renal biopsy. The principal cause of mortality was active disease and infection. Meningo-coccal meningitis, Klebsiella peritonthis, Pneumococcal cellullitis of the neck, peritonthis and 1 patient with disseminated tuberculosis.

1 patients (91.6%) had received intravenous steroids as pulses and Cyclophosphamide pulse therapy, azathioprine or mycophenolate mofetil, hydroxychloroquine immunoglobulins. In some patients peritondialysis and ventilation. 8 patients (86%) with bad socioeconomic environment. Four patients (33%) with poor adherence to treatment.

Conclusion: In this single-center study, childhood onset lupus was associated with a mortality of 24%. With higher frequency of aggressive renal disease and higher requirement for steroids and immunosuppressive drugs. Pediatric care should consider transition to adult's centers and ensure continuity of treatment.

REFERENCE


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Trends in Employment and Hospitalization in Patients with Sjögren’s Syndrome 1993–2016: Results from the German National Database

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Background: During the last 20 years, employment rates increased substantially in the German population and also in patients with arthritis [1]. Whether patients with primary Sjögren’s syndrome (pSS) also show this trend is less clear in the absence of new treatment options.

Objectives: To assess trends in treatment and outcomes in patients pSS, focusing on employment, hospitalization and medical treatment in the past two decades.

Methods: Data from 1996 to 2016, ~300 patients with pSS were documented annually in the National Database of the German Collaborative Arthritis Centres. Data on treatment, physician assessment of disease activity, patient-reported outcomes, hospitalization and employment were collected and compared to patients with rheumatoid arthritis (RA), matched 1:1 for age, sex and disease duration for each calendar year.

Results: Patients with pSS (~90% female, age ~44 years at disease onset, disease duration ~10 years) were more frequently assessed to be in low disease activity in 2016 (93%) than in 1996 (62%, p<0.01). Treatment with antimalarials increased (31% to 50%, p<0.01) and less patients were on glucocorticoids (50% to 34%, p<0.01). Less than 5% were treated with biologics in 2016. The percentage of employed patients (~65 years) increased by 21 percentage points (43% to 64%, p<0.001), exceeding the increase observed for RA patients (~15 percentage points). In 2016, significantly less patients compared to 1996 were on early retirement (22% and 10%, p<0.01), hospitalized/year (13 and 7%, p<0.05) or on temporary sick leave (27% compared to 39%, p=0.09). This trend is comparable to RA patients.

Conclusion: Overall, similar trends were observed for RA and pSS cohorts despite minor changes in pSS therapy. Work participation has improved significantly over two decades in pSS. A greater perception of pSS without systemic manifestations may have caused a shift towards less severely affected patient cohorts today.

REFERENCES


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Whole Blood versus Serum Hydroxychloroquine Levels for Drug Monitoring of Patients with Systemic Lupus Erythematosus: Preliminary Results of a Pharmacological Study

Benoit Blanchef,1, Moez Jalloul1, Marie Allarde1, Pascale Ghiliani Dabbín1, Lionel Galicier2, Véronique Le Guern1, Noël Zahr1, Claire Goulvestre1, Zahir Amoura2, Jean-Charles Pietto2, Noémie Jourde-Chiche3, Nathalie Costedoat-Chalumeau, Group PLUS, 4Cochin Hospital, Paris, France, 5Hôpital Pitié-Salpêtrière Hospital, Paris, France, 6Saint Louis Hospital, Paris, France, 7Aix-Marseille Univ, C2VN, INSERM, INRA, AP-HM Centre de Néphrologie, Marseille, France

Background: In order to assess the pharmacokinetic/pharmacodynamic relationship of hydroxychloroquine (HCQ) in patients with systemic lupus erythematosus (SLE), HCQ levels have been measured in whole blood as well as in serum but both methods have never been compared. In addition, cut-offs for non-adherence (classically 200ng/ml but also 100 ng/ml) have been established only in whole blood.

Results: Patients with pSS (>90% female, age ~44 years at disease onset, disease duration ~10 years) were more frequently assessed to be in low disease activity in 2016 (93%) than in 1996 (62%, p<0.01). Treatment with antimalarials increased (31% to 50%, p<0.01) and less patients were on glucocorticoids (50% to 34%, p<0.01). Less than 5% were treated with biologics in 2016. The percentage of employed patients (~65 years) increased by 21 percentage points (43% to 64%, p<0.001), exceeding the increase observed for RA patients (~15 percentage points). In 2016, significantly less patients compared to 1996 were on early retirement (22% and 10%, p<0.01), hospitalized/year (13 and 7%, p<0.05) or on temporary sick leave (27% compared to 39%, p=0.09). This trend is comparable to RA patients.

Conclusion: Overall, similar trends were observed for RA and pSS cohorts despite minor changes in pSS therapy. Work participation has improved significantly over two decades in pSS. A greater perception of pSS without systemic manifestations may have caused a shift towards less severely affected patient cohorts today.

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