Clinical features, damage accrual, and survival in patients with familial systemic lupus erythematosus: data from a multi-ethnic, multinational Latin American lupus cohort

Por: Quintana, R (Quintana, Rosana)[1,2]; Pons-Estel, GJ (Pons-Estel, Guillermo J.)[1,2]; Roberts, K (Roberts, Karen)[2]; Sacnun, M (Sacnun, Monica)[2]; Serrano, R (Serrano, Rosa)[1]; Nieto, R (Nieto, Romina)[2]; Conti, S (Conti, Silvana)[2]; Gervasoni, V (Gervasoni, Viviana)[2]; Catoggio, LJ (Catoggio, Luis J.)[3]; Soriano, ER (Soriano, Enrique R.)[3]; Scolnik, M (Scolnik, Marina)[3]; Garcia, MA (Garcia, Mercedes A.)[4]; Alvarellos, A (Alvarellos, Alejandro)[5]; Saurit, V (Saurit, Veronica)[5]; Berbotto, GA (Berbotto, Guillermo A.)[6]; Sato, EI (Sato, Emilia, I)[7]; Costallat, LTL (Lavras Costallat, Lilian T.)[8]; Borba, EF (Borba Neto, Eduardo Ferreira)[9]; Bonfa, E (Bonfa, Eloisa)[9]; Xavier, RM (Xavier, Ricardo M.)[10]; Montandon, ACDES (de Oliveira e Silva Montandon, Ana Carolina)[11]; Molina-Restrepo, JF (Molina-Restrepo, Jose Fernando)[12]; Iglesias-Gamarra, A (Iglesias-Gamarra, Antonio)[13]; Guibert-Toledano, M (Guibert-Toledano, Marlene)[14]; Reyes-Llerena, GA (Reyes-Llerena, Gil Alberto)[14]; Massardo, L (Massardo, Loreto)[15]; Neira, OJ (Neira, Oscar J.)[16]; Cardiel, MH (Cardiel, Mario H.)[17]; Barile-Fabris, LA (Barile-Fabris, Leonor A.)[18]; Amigo, MC (Amigo, Mary-Carmen)[19]; Silveira, LH (Silveira, Luis H.)[20]; De La Torre, IG (De La Torre, Ignacio Garcia)[21]; Acevedo-Vasquez, EM (Acevedo-Vasquez, Eduardo M.)[22]; Ugarte-Gil, MF (Ugarte-Gil, Manuel F.)[22,23]; Alfaro-Lozano, JL (Alfaro-Lozano, Jose Luis)[22]; Segami, MI (Segami, Maria Ines)[24]; Chacon-Diaz, R (Chacon-Diaz, Rosa)[25]; Esteva-Spinetti, MH (Esteva-Spinetti, Maria H.)[26]; Gomez-Puerta, JA (Gomez-Puerta, Jose A.)[27]; Alarcon, GS (Alarcon, Graciela S.)[28,29]; Pons-Estel, BA (Pons-Estel, Bernardo A.)[1] ...Menos

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Abstract

Objectives This study aimed to compare the clinical features, damage accrual, and survival of patients with familial and sporadic systemic lupus erythematosus (SLE). Methods A multi-ethnic, multinational Latin American SLE cohort was studied. Familial lupus was defined as patients with a first-degree SLE relative; these relatives were interviewed in person or by telephone. Clinical variables, disease activity, damage, and mortality were compared. Odds ratios (OR) and 95% confidence intervals (CI) were estimated. Hazard ratios (HR) were calculated using Cox proportional hazard adjusted for potential confounders for time to damage and mortality. Results A total of 66 (5.6%) patients had familial lupus,

and 1110 (94.4%) had sporadic lupus. Both groups were predominantly female, of comparable age, and of similar ethnic distribution. Discoid lupus (OR = 1.97; 95% CI 1.08-3.60) and neurologic disorder (OR = 1.65; 95% CI 1.00-2.73) were significantly associated with familial SLE; pericarditis was negatively associated (OR = 0.35; 95% CI 0.14-0.87). The SLE Disease Activity Index and Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index (SDI) were similar in both groups, although the neuropsychiatric (45.4% vs. 33.5%;p = 0.04) and musculoskeletal (6.1% vs. 1.9%;p = 0.02) domains of the SDI were more frequent in familial lupus. They were not retained in the Cox models (by domains). Familial lupus was not significantly associated with damage accrual (HR = 0.69; 95% CI 0.30-1.55) or mortality (HR = 1.23; 95% CI 0.26-4.81). Conclusion Familial SLE is not characterized by a more severe form of disease than sporadic lupus. We also observed that familial SLE has a higher frequency of discoid lupus and neurologic manifestations and a lower frequency of pericarditis.

Palabras clave

Palabras clave de autor: Systemic lupus erythematosus; familial lupus; sporadic lupus; disease activity; damage accrual; mortality
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Información del autor

Dirección correspondiente: Quintana, R (corresponding author)

Bv Orono 1024, Rosario, Santa Fe, Argentina.

Direcciones:

[1] Ctr Reg Enfermedades Autoinmunes & Reumat GO CREA, Rosario, Argentina

[2] Hosp Prov Rosario, Rosario, Argentina

[3] Hosp Italiano Buenos Aires, Buenos Aires, DF, Argentina

[4] Hosp Interzonal Gen Agudos Gen San Martin, La Plata, Argentina

[5] Hosp Privado, Ctr Med Cordoba, Cordoba, Argentina

[6] Hosp Escuela Eva Peron, Granadero Baigorria, Argentina

[7] Univ Fed Sao Paulo UNIFESP, Sao Paulo, SP, Brazil

[8] Univ Estadual Campinas, Campinas, Brazil

[9] Univ Sao Paulo, Sao Paulo, Brazil

[10] Univ Fed Rio Grande do Sul, Hosp Clin Porto Alegre, Porto Alegre, RS, Brazil

[11] Univ Fed Goias, Fac Med, Goiania, Go, Brazil

[12] Reumalab, Ctr Integral Reumatol, Medellin, Colombia

[13] Univ Nacl Colombia, Hosp Santa Rosa Bogota, Bogota, Colombia

[14] Ctr Invest Med Quirurg CIMEQ, Havana, Cuba
[15] Univ San Sebastian, Fac Med, Santiago, Chile
[16] Univ Chile, Hosp Salvador, Fac Med, Santiago, Chile
[17] Ctr Invest Clin Morelia SC, Morelia, Michoacan, Mexico
[18] Hosp Angeles Pedregal, Ciudad De Mexico, Mexico
[19] Ctr Med ABC, Ciudad De Mexico, Mexico
[20] Inst Nacl Cardiol Ignacio Chavez, Ciudad De Mexico, Mexico
[21] Hosp Gen Occidente Secretaria Salud, Guadalajara, Jalisco, Mexico
[22] Hosp Gen Guillermo Almenara Irigoyen, Serv Reumatol, EsSalud, Lima, Peru
[23] Univ Cient Sur, Lima, Peru
[24] Hosp Nacl Edgardo Rebagliatti Martins, ESSALUD, Lima, Peru
[25] Hosp Univ Caracas, Ctr Nacl Enfermedades Reumat, Caracas, Venezuela
[26] Hosp Cent San Cristobal, San Cristobal, Venezuela
[27] Hosp Clin Barcelona, Serv Reumatol, Barcelona, Spain
[28] Univ Alabama Birmingham, Birmingham, AL USA
[29] Univ Peruana Cayetano Heredia, Lima, Peru

Direcciones de correo electrónico:rosanaquintana@gmail.com

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