# Systemic Lupus Erythematosus in a 6 Month Old Female Child

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Systemic lupus erythematosus (SLE) is an uncommon disease, particularly in the first decade of life. Finding it before the first year of life is very rare and it entails a difficult diagnosis to document because the clinical manifestations can be very variable and can simulate a great variety of diseases.
In the pediatric age, incidence of SLE is 0.36-0.9/ 100 000 children, with greater prevalence in females 3:1 under 12 years of age and 10:1 in patients over 12 years. There is a very low incidence of SLE before 4 years of age.
We present the clinical case of a 9 month-old female in whom 4 criteria of SLE were clearly documented at 6 months of age. In a review of literature, this is the youngest patient reported with SLE.
Physicians must be very alert of the higher risk of systemic complications in children that are diagnosed with SLE at an early age.

**Key words:** Systemic lupus erythematosus. Infant. Systemic complications.

### Lupus eritematoso sistémico en una lactante de 6 meses

El lupus eritematoso sistémico (LES) es poco frecuente, particularmente en la primera década de la vida; su inicio antes de 1 año de edad es muy raro y conlleva un diagnóstico difícil de documentar, ya que las manifestaciones pueden ser muy variables y parecerse a las de una gran variedad de enfermedades.
En la edad pediátrica la incidencia de LES se estima en 0,36-0,9/100.000 niños, con prevalencia mayor en mujeres que en varones (aproximadamente 3:1 en menores de 12 años y 10:1 en niños mayores). Sin embargo, el inicio de LES antes de los 4 años ocurre con muy baja incidencia.
Presentamos el caso de una lactante de la que se documentaron claramente más de cuatro criterios diagnósticos de LES desde los 6 meses de edad. Por lo revisado en la literatura, podría ser el caso de la paciente más pequeña que se haya publicado.
Los médicos deben estar atentos al mayor riesgo de complicaciones sistémicas en niños con LES que se inicia a una edad temprana y realizar un diagnóstico oportuno de esta enfermedad.

**Palabras clave:** Lupus eritematoso sistémico. Lactante. Complicaciones sistémicas.

## Introduction

Systemic lupus erythematosus (SLE) is infrequent, particularly in the first decade of life; its onset before the patient is 1 year old is very rare and is usually a difficult diagnosis to reach because its manifestations can be variable and resemble a great number of illnesses.[1] The diagnosis is reached through clinical and laboratory findings, apart from the fact that the patient must meet certain diagnostic criteria proposed by the American College of Rheumatology.[2]
We present the case of an infant in which more than 4 diagnostic criteria for SLE were clearly documented since the child was 6 months old and which could constitute, according to our review of the literature, the earliest recorded case.

## Clinical Case

A 9 month old female child presented a history of quotidian fever of up to 40°C, predominantly nocturnal, with an increase in the volume, temperature and pain of the right wrist and left knee as well as functional limitation in their movement that had had its onset 3 months before. Physical examination showed generalized pallor, cervical adenopathy, a liver palpable 3 cm under over the normal

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limit, a spleen palpable 1.5 cm over the normal limit and inguinal adenopathy measuring 0.5 cm. A complete blood count showed anemia (hemoglobin, 6.4 g/dL); normal leukocytes and platelets; lactate-dehydrogenase, 3495 U/L; and a positive Coombs test at 3 different temperatures. Renal function tests were normal but the urinalysis showed blood cells and granulous casts in the sediment, with negative proteins. ANA were reported as 1:320 in a fine speckled pattern; anti-DNA, 262 U/m; C3, 181 mg/dL (90-180 mg/dL); C4, 29.3 mg/dL (10-40 mg/dL); IgG, 2660 mg/dL (700-1600 mg/dL); IgM, 194 mg/dL (40-230 mg/dL); IgA, 12 mg/dL (70-400 mg/dL); anti-Sm, anti-RNP, anti-Ro, and anti-La were negative. An axial tomography showed a small bilateral pleural effusion. Nine months later the patient presented skin lesions that were characterized as erythematous macula and papulae in the malar region, something very indicative of disease activity. When the child was 2 years old a renal biopsy documented class IIb lupus nephritis.

## Discussion

In pediatric patients the incidence of SLE is estimated to be 0.36-0.9/100 000 children, with a larger prevalence in women than in men (approximately 3:1 in children under 12 and 10:1 in older children).[1,3] However, the start of SLE before the age of 4 occurs with a very low incidence.[2] Of the 300 cases of pediatric lupus seen during a period of 10 years at the immune-mediated outpatient clinic of the Hospital Infantil Federico Gómez in Mexico, none started before the age of 4. In the case that we present here, symptoms started at 6 months of age and because of the age of the patient and the presentation of the symptoms, both infectious and neoplastic causes had to be excluded. The current medical literature only documents 3 cases of SLE in children under one year of age, but their onset did not show a characteristic form.[4-6] Neonatal lupus, caused by the transplacental transmission of anti-Ro/La IgG antibodies, is a well-known phenomenon. Usually, these infants present malar rash and annular lesions in addition to congenital heart block. But these lesions are seen upon birth or in the first 8 weeks of life.[7]

The case which we present might represent the youngest patient with immune and hematologic alterations, non-erosive arthritis, lupus nephritis and serositis, similar to the typical presentation of SLE in older children and adults.[8] It is generally established that children tend to have a more severe renal affection than adults, and that hepatosplenomegaly and lymphadenopathy are more characteristic of the pediatric-onset form. The autoantibody profile reveals an elevated frequency of anti-DNA antibodies seen more frequently in children than adults and ANA are present in 95% of children with SLE. In addition, SLE is less frequent in children than adults.[9] A recent study by Pluchinotta et al[10] found, upon evaluating the clinical characteristics of SLE in pediatric patients in 3 different age groups, that the prevalence of internal organ affection seems to diminish with age. Pediatric lupus is more severe and produces a higher mortality than in other age groups.[10]

Physicians must be on the lookout because of the higher risk of systemic complications in children with a younger age-onset of SLE and reach an early diagnosis of the disease.

## References


1. Faller G, Thomson PD, Kala UK, et al. Demographics and presenting clinical features of childhood systemic lupus erythematosus. S Afr Med J. 2005;95: 424-7.
2. Cassidy JT, Petty RE. Textbook of pediatric rheumatology. 5th ed. Philadelphia: Elsevier Saunders; 2005. p. 342-83.
3. Houghton KM, Page J, Cabral DA, Petty RE. Systemic lupus erythematosus in the pediatric North American Native population of British Columbia. J Rheumatol. 2006;33:161-3.
4. Cummings NP, Hansen J, Hollister R. SLE in a premature infant. Arthritis Rheum. 1985;28:573-5.
5. Dogra D, Rathi SK, Sharma NK, et al. Systemic lupus erythematosus in an infant. Indian Pediatr. 1996;33:779-81.
6. Saberi MS, Jones BA. Remission of infantile systemic lupus erythematosus with intravenous cyclophosphamide. Pediatr Nephrol. 1998;12:136-8.
7. Buyon JP, Clancy RM. Neonatal lupus: basic research and clinical perspectives. Rheum Dis Clin North Am. 2005;31:299-313.
8. Font J, Cervera R, Espinosa G, et al. Systemic lupus erythematosus in childhood: analysis of clinical and immunological findings in 34 patients and comparison with SLE characteristics in adults. Ann Rheum Dis. 1998;57: 456-9.
9. Klein-Gitelman M. Pediatric lupus versus adult lupus role of the laboratory. Clinical and applied. Immunol Rev. 2004;4:333-50.
10. Pluchinotta FR, Schiavo B, Vittadello F, Martini G, Perilongo G, Zulian F. Distinctive clinical features of pediatric systemic lupus erythematosus in three different age classes. Lupus. 2007;16:550-5.