Published in final edited form as:
Pediatr Clin North Am. 2012 April ; 59(2): 345–364. doi:10.1016/j.pcl.2012.03.007.

Systemic Lupus Erythematosus in Children and Adolescents

Deborah M. Levy, MD MS FRCPC[1] and **Sylvia Kamphuis, MD PhD**[2]

[1]Assistant Professor, Pediatrics, University of Toronto Staff Rheumatologist, Hospital for Sick Children

[2]Pediatric rheumatologist/immunologist, Sophia Children's Hospital, Erasmus University MC, SP 3429, PO Box 2060, 3000 CB Rotterdam, tel: 0031-10-7036104 (secr), fax: 0031-10-7036943, s.kamphuis@erasmusmc.nl

Synopsis

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease that can involve any organ system with a wide range of disease manifestations, and can lead to significant morbidity and even mortality. This article reviews the epidemiology, common clinical features, complications of disease, and briefly discusses the available treatment options. In addition, important medical and psychosocial issues relevant to the pediatrician caring for children and adolescents with SLE are discussed.

Keywords

pediatric; childhood; SLE; clinical features; neuropsychiatric; nephritis; diagnosis; treatment; damage; complications

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease that can involve any organ system, and may lead to significant morbidity and even mortality. In this article we review the epidemiology, common clinical features, complications of disease, and briefly address available treatment options. Further, we discuss important medical and psychosocial issues relevant to the pediatrician caring for children and adolescents with SLE.

Epidemiology

Childhood-onset SLE (cSLE) is a rare disease with an incidence of 0.3-0.9 per 100.000 children-years and a prevalence of 3.3-8.8 per 100.000 children.[1] A higher frequency of cSLE is reported in Asians, African American, Hispanics and native Americans.[2,3] When compared to two more common childhood autoimmune diseases, Juvenile Idiopathic Arthritis (JIA) and type 1 Diabetes, cSLE is around 10 to 15 times less common in white children.[4,5] However, in Asian children, cSLE is reported to be equally as common as JIA.[6] Most studies report a median age of onset of cSLE between 11-12 years; the disease is quite

Corresponding Author: Deborah M. Levy, Mailing Address: Hospital for Sick Children, Division of Rheumatology, 555 University Avenue, Toronto, ON M5G 1×8, Phone 416-813-6982, Fax 416-813-4989, deborah.levy@sickkids.ca.

rare under the age of 5 years. As in adult onset SLE, approximately 80% of patients with cSLE are female.[7,8]

Classification and Diagnosis of cSLE

SLE is called the great mimicker, as the disease shares characteristics with many other (autoimmune) diseases. Especially when the classic malar rash is absent, diagnosing SLE can be a challenge. However, the astute pediatrician who considers SLE when presented with an unusual constellation of symptoms can recognize important patterns of disease manifestations crucial for the diagnosis. Most patients who are diagnosed with cSLE fulfill 4 or more of the American College of Rheumatology classification criteria for SLE (Table 1).[9,10] The criteria were designed for use in research studies, and we caution that the diagnosis of SLE should not solely be based on fulfilling these criteria. Although not rigorously studied in cSLE, the criteria have a greater than 95% sensitivity and specificity for the diagnosis of cSLE.[11]

Clinical Features

The current review will not attempt to describe all possible clinical manifestations but instead we focus on specific features that may be crucial for immediate recognition. Table 2 summarizes the frequencies of the common manifestations of cSLE.[7,12-17] SLE can affect any organ system, and leads to glomerulonephritis and central nervous system involvement arguably more often in cSLE than in adults with SLE.

Constitutional Symptoms

Patients ultimately diagnosed with cSLE frequently recount nonspecific constitutional symptoms that include fever, fatigue, anorexia, weight loss, alopecia and arthralgias.[7,12] These and other symptoms of diffuse generalized inflammation including lymphadenopathy and hepatosplenomegaly occur both at onset and during disease flares.

Mucocutaneous

The hallmark of SLE is the malar, or butterfly rash. The rash is seen in 60 - 85% of children with SLE, is generally described as erythematous, raised, non-pruritic, and non-scarring. The rash often extends over the nasal bridge, affects the chin and ears, but spares the nasolabial folds (Figure 1). It is photosensitive in more than a third of patients, and exacerbation of the photosensitive rash frequently heralds the onset of a systemic flare. Therefore, sunscreen with a high sun protection factor, as well as hats and protective clothing are recommended year round for all individuals with SLE.

Discoid rash, unlike in adult-onset SLE, is a rare manifestation of cSLE, occurring in fewer than 10% of patients.[7] This scarring rash most frequently occurs on the forehead and scalp, and its scaly appearance may be mistaken as a tinea lesion.[18] Table 3 summarizes the spectrum of dermatologic involvement, illustrating the diverse range of skin manifestations. Children and adolescents with SLE can develop a rash of (almost) any morphology, location and distribution, often presenting a diagnostic challenge to the primary care physician. A skin biopsy for histology aids in making the correct diagnosis, although biopsies of facial skin should be avoided. Non-scarring hair loss is common, but not specific for SLE. The alopecia is most often noted as thinning of the temporal areas of the scalp, although rarely it is more global and severe enough to require systemic immunosuppressive therapy. Nevertheless, for the affected child or adolescent, even mild hair loss can be distressing.

Involvement of the oral and nasal mucosa ranges from oral and/or nasal hyperemia to painless oral ulcers of the hard palate (Figure 2) and shallow nasal septal ulcers, and rarely,