Systemic Lupus Erythematosus in Children and Adolescents

Deborah M. Levy, MD, MS, FRCPC[a,*], Sylvia Kamphuis, MD, PhD[b]

KEYWORDS

• Pediatric • Clinical features • Neuropsychiatric • Nephritis
• Diagnosis • Treatment • Complications

Key Points

- cSLE is a rare but severe autoimmune disease with multisystem involvement and wide heterogeneity of disease manifestations.
- Making the diagnosis of cSLE can be difficult, but early recognition of the disease is important to limit adverse outcomes.
- cSLE follows a more severe disease course than adult-onset SLE, with higher frequency of morbidity and lower survival rates.
- Dealing with the diagnosis of a lifelong, unpredictable, and relapsing-remitting disease in adolescence is challenging for cSLE patients, and recognition of the specific needs of this age group is important for optimal outcome.

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that can involve any organ system, and may lead to significant morbidity and even mortality. This article reviews the epidemiology, common clinical features, and complications of this disease, and briefly addresses available treatment options. Important medical and psychosocial issues relevant to the pediatrician caring for children and adolescents with SLE are discussed.

EPIDEMIOLOGY

Childhood-onset SLE (cSLE) is a rare disease with an incidence of 0.3 to 0.9 per 100,000 children-years and a prevalence of 3.3 to 8.8 per 100,000 children.[1] A higher

[a] Division of Rheumatology, Hospital for Sick Children, University of Toronto, 555 University Avenue, Toronto, ON M5G 1X8, Canada; [b] Divisions of Infectiology/Immunology/Rheumatology, Sophia Children's Hospital, Erasmus University MC, SP 3429, PO Box 2060, 3000 CB Rotterdam, The Netherlands
* Corresponding author. Division of Rheumatology, Hospital for Sick Children, 555 University Avenue, Toronto, ON M5G 1X8, Canada.
E-mail address: deborah.levy@sickkids.ca

Pediatr Clin N Am 59 (2012) 345–364
doi:10.1016/j.pcl.2012.03.007 **pediatric.theclinics.com**
0031-3955/12/$ – see front matter

frequency of cSLE is reported in Asians, African Americans, Hispanics, and Native Americans.[2,3] When compared with 2 more common childhood autoimmune diseases, juvenile idiopathic arthritis (JIA) and type 1 diabetes, cSLE is approximately 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 and 12 years; the disease is rare in children younger than 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 it must be cautioned 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

This review does not attempt to describe all possible clinical manifestations but instead focuses 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 (CNS) 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 signs of diffuse generalized inflammation, including lymphadenopathy and hepatosplenomegaly, may occur both at onset and during disease flares.

Mucocutaneous

The hallmark of SLE is the malar, or butterfly, rash. Seen in 60% to 85% of children with SLE, the rash is generally described as erythematous, raised, nonpruritic, and nonscarring. It often extends over the nasal bridge, affects the chin and ears, but spares the nasolabial folds (**Fig. 1**). It is photosensitive in more than one-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] **Box 1** 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. Nonscarring