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Childhood Onset Systemic Lupus Erythematosus: How much it is different than in Adults

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Introduction

Systemic Lupus Erythematosus (SLE) is an autoimmune connective tissue disease with multiple-organ involvement. Although the disease is more common in adults, it is more severe and has a much poorer prognosis in children. SLE begins in childhood or adolescence in approximately 15% of cases [1]. In young adult women the peak incidence of about 5 in 100 000 per year [2,3]. By contrast, in children the overall incidence is about 0.5 in 100 000 per year [4,5]. The female/male ratio increases from 2:1 in prepubertal children to 4.5:1 in adolescents and 8:1 in adults [6]. In children, most cases of lupus occur after age five, with a peak incidence in late childhood and adolescence [7].

Diagnosis and Classification

The diagnostic criteria of SLE are the same in both children and adults. Diagnosis of SLE is quite different from its classification. The diagnosis may not fulfill the American College of Rheumatology (ACR) classification criteria which were defined and validated for the purposes of clinical trials and were not specifically developed as diagnostic criteria [8]. This differentiation is highly important to avoid inappropriately delayed treatment waiting for the classification criteria to be fulfilled [9].

Diagnosing SLE in children may be difficult due to multisystem affection and heterogeneity of clinical manifestations. The initial symptoms may be vague and red flags for the pediatrician include an older child or adolescent with any combination of persistent fever, fatigue, anemia, leucopenia, lymphadenopathy, malar or other rash, alopecia, unexplained weight loss, neuropsychiatric symptoms, or unexplained microscopic hematuria or proteinuria [10].

The child patient fulfills classification criteria when presents with four of 11 criteria simultaneously or sequentially as in adults [11]. There have been few large-scale efforts to validate the ACR classification criteria for SLE in the childhood onset population. Ferraz et al. [12] in Brazil examined the sensitivity and specificity of ACR criteria in a group of 103 children with SLE and 101 pediatric patients with other rheumatic diseases. These authors found that the most common criteria were positive ANA, arthritis, immunologic disorder, hematologic disorder, malar rash and photosensitivity. Sensitivity was 96% and specificity was 100% in this analysis. These data show that the ACR classification system is accurate when applied to a pediatric population.

Distinctive Clinical Features of Childhood SLE

One method of measuring disease severity is organ involvement. Compared with adults, children with SLE have more active disease at time of diagnosis and over time. Children have higher rates of organ involvement, develop lupus nephritis at a higher frequency and renal damage more rapid than adults [13,14].

A second method for measuring the disease severity is by categorizing patients according to the amount of corticosteroids used to treat initial disease. Tucker et al. categorized patients who required corticosteroid more than 0.5 mg/kg /day as high disease severity patients while patients who didn't require corticosteroids were categorized as low disease severity patients. It was found that the higher disease severity category included 77% of children and only 16% of adults, while 46% of adults were categorized as low disease severity and only 13% of children belonged to this group [15].

A third method for describing disease severity is disease damage. The Rood study used the Systemic Lupus International Collaborating Clinic (SLICC)/ACR damage index to assess damage in a group of children with SLE. The mean index was found to be 2.6 after 4.7 years of follow-up, while an index of 0.75 after 5 years of follow-up was found in adults [16].

Mortality rate is the fourth measure of disease activity. Pediatric deaths mostly occur during the acute phase of the disease, while most adults' deaths are related to disease outcome (organ failure, myocardial infarction) or side effects (infection) [11].

Distinctive Immunologic Features of Childhood SLE

An increased prevalence of anti-DNA antibodies was reported in the childhood SLE in comparison to the adult SLE (15). In contrast, other studies did not find a significant difference [13,17]. These studies found no difference in frequency of presence of ANA and neither of these studies commented on difference in the pattern of ANA (speckled, homogenous, nucleolar or centromere) in the pediatric versus adult population.

Tucker found that anti-Sm antibodies and antibody to the 70kDa component of RNP are more common in childhood SLE [15]. However, Klein-Gitilman did not confirm this finding [11].

Anticardiolipin frequencies in childhood and adult SLE are similar as reported by most investigators, however, Font et al. [17] described an increased presence of IgG anticardiolipin Ab in childhood SLE. Finally, Tucker study [15] reported an increased frequency of low levels of C3 at onset compared to adult patients.

Treatment Difficulties in Childhood SLE

Treatment of childhood onset SLE is a challenge. The therapeutic issues, risks and balances faced by adult patients are much more

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complicated in children with SLE by an unpredictable disease course and long requirement for therapy. Non-compliance is a major obstacle to satisfactory outcome which must be recognized and dealt with in every patient to attain optimal outcome [18]. Unfortunately, there have been no randomized controlled trials in children to assess the drugs currently used in the treatment of pediatric SLE. Data therefore, come from uncontrolled studies, case reports, and extrapolation from the experience with adults with SLE.

In conclusion, the presentation, clinical symptoms and immunological findings of childhood SLE are similar to those of adult SLE patients; however children usually have a more severe disease at onset. They also have higher rates of organ involvement, and a more aggressive clinical course than adults.

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