

mm) and 186 (0.32%) children had induration within 10 to 30 mm (mean: 14.57 mm). Children with TST induration ≥ 10 mm were given treatment for tuberculosis; for those with induration of 5 to 9 mm, their environment was examined for the presence of risk factors.

CONCLUSIONS: The discovery of a high number of children with positive TST results (≥ 10 mm) in both groups indicates a remaining tuberculosis problem in Greece. The extent of induration up to 30 mm reveals the exposure of children of these age groups to high mycobacterial burden from adults with tuberculosis, especially immigrants from countries of Eastern Europe.

Rheumatology

CLINICAL COURSE AND OUTCOME IN CHILDREN WITH RARE CONNECTIVE TISSUE DISEASES: A RETROSPECTIVE REVIEW OF A 17-YEAR EXPERIENCE

Submitted by Christina Dracou-Kakava

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INTRODUCTION: Juvenile dermatomyositis/juvenile polymyositis (JDM/JPM), juvenile systemic sclerosis (JSCL-SYST), and juvenile mixed connective tissue disease (JMCTD) are rare in childhood.

OBJECTIVE: The objective of this study was to evaluate the prognosis of the rare connective tissue diseases (RCTDs) in children,

METHODS: We reviewed the medical charts of children with a diagnosis of RCTD since 1989 and a minimum follow-up of 5 years.

RESULTS: Twenty-four (16 female, 8 male) children with JDM/JPM, JSCL-SYST, and JMCTD were studied. The age at disease onset ranged from 4 to 13 years. The follow-up duration was 5 to 12 years. Sixteen children had JDM, and 2 had JPM. Four had JSCL-SYST, and 2 had JMCTD. Until now, 13 children have reached clinical remission, lasting >3 years after stopping drug therapy. Twelve children had JDM/JPM, and 1 had JMCTD. Persistent disease activity was noted in 11 children: 4 with JSCL-SYST, 6 with JDM/JPM, and 1 with JMCTD. Severe pulmonary disease developed in 3 children: 2 with JSCL-SYST and 1 with JMCTD. None of the children with JDM had pulmonary disease. Pulmonary hypertension (PH) was found in 2 children with JMCTD or JSCL-SYST. The child with JSCL-SYST and PH died. Persistent scleroderma

pattern by wide-field capillaroscopy was noted in 4 children who had JDM and had had skin ulcerations and have developed subcutaneous calcifications. One of them has also had marked muscle atrophy and severe contractures.

CONCLUSIONS: Persistent activity and/or severe pulmonary involvement may be present during the clinical course of RCTD. The presence of PH indicates very poor prognosis in JSCL-SYST/JMCTD cases. Capillaroscopy may identify children who have JDM and are candidates for aggressive therapy.

RE-TREATMENT AND RISK FACTORS OF REFRACTORY KAWASAKI DISEASE

Submitted by Zhong-Dong Du

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OBJECTIVE: The objective of this study was to evaluate the incidence and risk factors of children with refractory Kawasaki disease (KD).

METHODS: All children with KD were analyzed in Beijing from 2000 through 2004. Risk factors were analyzed by logistic regression. Refractory KD was defined as persistent fever of $\geq 38.5^{\circ}\text{C}$ 36 hours after initial intravenous immunoglobulin (IVIg) treatment.

RESULTS: A total of 1052 patients (aged 1 month to 13.8 years) with IVIg treatment were included; of them, 135 did not respond to IVIg treatment, with an incidence of 12.8%. Refractory KD occurred more frequently in children who received 1 g/kg per day IVIg for 2 days (20.9%) than in those who received a single dose of 2 g/kg (9.9%) or 400 to 600 mg/kg per day for 4 days (8.7%). Logistic regression revealed that erythrocyte sedimentation rate, alanine aminotransferase, white blood cell count, serum albumin, time from onset to IVIg treatment, and IVIg dosage were independent risk factors for refractory KD. Children with refractory KD were re-treated: 8 received 2 g/kg IVIg, with 5 (62.5%) responding; 114 received 1 g/kg IVIg, with 35 (30.7%) responding; and 11 received 400 to 600 mg/kg IVIg, with (9.1%) responding. In addition, 2 received corticosteroids, with 2 responding.

CONCLUSIONS: The incidence of refractory KD in Beijing is 12.8%. A 2-g/kg dose of IVIg is probably the best re-treatment option for refractory KD.

NEUROPSYCHIATRIC SYMPTOMS IN CHINESE CHILDREN WITH SYSTEMIC LUPUS ERYTHEMATOSUS

Submitted by Yu-Lung Lau

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INTRODUCTION: There is a paucity of clinical studies on children with neuropsychiatric systemic lupus erythematosus (NPSLE).

OBJECTIVE: The objective of this study was to define the clinical characteristics of and the potential predictors for NPSLE in Chinese children.

METHODS: Sixty-two children with SLE diagnosed between 1990 and 2006 were retrospectively reviewed. Patients were evaluated according to the American College of Rheumatology case definitions (1999) for classification of neuropsychiatric symptoms. The demographic data, clinical manifestations, laboratory parameters (complete blood count, erythrocyte sedimentation rate, C-reactive protein, complement levels, anti-cardiolipin antibodies, and autoimmune markers), treatment, and SLE disease activity index score were analyzed.

RESULTS: Nineteen (30.65%) patients with SLE and 21 neuropsychiatric events were identified. Mean age at NPSLE manifestations was 13.57 ± 4.33 years. The most common neuropsychiatric manifestations were cognitive dysfunction (47.62%), seizure disorder (42.86%), and headache (28.57%), followed by mood disorder (19.05%), myelopathy (19.05%), cerebrovascular disease (14.29%), psychosis (9.52%), cranial neuropathy (9.52%), and mononeuropathy multiplex (4.76%). Renal involvement at diagnosis of SLE was significantly less common in patients with NPSLE than in those with non-NPSLE. Apart from that, we could not identify other clinical or laboratory parameters that could predict the development of NPSLE. Six patients presented with neuropsychiatric symptoms at onset of SLE. Comparing them with patients with later neuropsychiatric development, their mean age was younger and the SLE disease activity index score was significantly higher.

CONCLUSIONS: Neuropsychiatric symptoms were common in Chinese children with SLE. Early-onset NPSLE occurred in younger patients with higher disease activity score. Neuropsychiatric development was negatively associated with renal involvement at diagnosis.

RECURRENT MAJOR INFECTIONS IN JUVENILE-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS: A CLOSE LINK WITH LONG-TERM DISEASE DAMAGE

Submitted by Pamela Pui-Wah Lee

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INTRODUCTION: Infection is a significant cause of morbidity and mortality in patients with systemic lupus erythematosus (SLE). We postulated that patients with recurrent infections are more likely to have poorer disease outcome.

OBJECTIVE: The objectives of this study were to describe the pattern of infections and disease damage that occurred in a cohort of patients with juvenile-onset SLE and to determine whether cumulative disease damage was associated with recurrent infections in these patients.

METHODS: We retrospectively reviewed (1988–2004) the clinical characteristics, infective complications, and disease damage as measured by the Systemic Lupus International Collaborating Clinics/American College of Rheumatology Damage Index Score (SDI) in 47 patients with juvenile-onset SLE. Potential risk factors for disease damage were evaluated by univariate analysis and logistic regression. The correlation between number of major infections and disease damage was determined.

RESULTS: Thirty-two (68.1%) patients had lupus nephropathy, and 16 (34.0%) patients had neuropsychiatric lupus. Sixty-one episodes of major infections, defined as infections that required more than 1 week of antimicrobial agents, occurred in 27 (57.4%) patients, and 18 (31.4%) patients had recurrent major infections (≥ 2 episodes). Organ damage (SDI ≥ 1) was documented in 21 (44.7%) patients. By logistic regression, occurrence of major infections was the only significant risk factor for disease damage. There was a positive correlation between SDI score and the number of recurrent major infections.

CONCLUSIONS: Infections and disease damage are common comorbidities in juvenile-onset SLE. Recurrent infections could predict poorer disease outcome and associated organ damage in SLE.

Surgery

TRANSLUMINAL ENDOSCOPIC TREATMENT OF FENESTRATED DUODENAL MEMBRANES

Submitted by Gerardo Blanco-Rodriguez

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INTRODUCTION: Duodenotomy and resection or lateral duodenoduodenostomy by open or endoscopic sur-

**NEUROPSYCHIATRIC SYMPTOMS IN CHINESE CHILDREN WITH
SYSTEMIC LUPUS ERYTHEMATOSUS**

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