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
Christina Dracou-Kakava, Katerina Karakontinou, Marianthi Galetselli, Maria Avgeri, Sophia Drakonaki
Pediatric Rheumatology Outpatient's Clinic, Second Department of Pediatrics, Panagiotis and Aglaia Kyriakou Children's Hospital, Athens, Greece

INTRODUCTION: Juvenile dermatomyositis/juvenile polymyositis (JDM/JPM), juvenile systemic scleroderma (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
Zhong-Dong Du[a], Di Zhao[b], Junbao Du[c]
[a]Beijing Children's Hospital, Capital Medical University, Beijing, China; [b]Capital Institute of Pediatrics, Beijing, China; [c]First Hospital of Peking University, Beijing, China

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 ≥38.5°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

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

Christina Dracou-Kakava, Katerina Karakontinou, Marianthi Galetselli, Maria Avgeri and Sophia Drakonaki

Pediatrics 2008;121;S161
DOI: 10.1542/peds.2007-2022JJJJJJ

PEDIATRICS®
OFFICIAL JOURNAL OF THE AMERICAN ACADEMY OF PEDIATRICS

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

Christina Dracou-Kakava, Katerina Karakontinou, Marianthi Galetselli, Maria Avgeri and Sophia Drakonaki

Pediatrics 2008;121;S161
DOI: 10.1542/peds.2007-2022JJJJJJ