

syndrome that has attracted a great deal of attention because of an uneven neurolinguistic profile characterized by relative strengths in language, facial processing, and social cognition in the context of poorer spatial cognition, planning, and problem solving. WS has also been used as evidence for the existence of dissociations within subsystems of the language module itself. It has been reported that individuals with WS perform better on grammatical versus lexical tasks and on regular versus irregular forms.

**OBJECTIVE:** This study addressed 2 main questions: (1) Do individuals with WS show differences between language and cognition? (2) Do individuals with WS perform differently across tasks that tap different aspects of language?

**METHODS:** We investigated nonverbal and verbal abilities of 20 Greek-speaking children with WS (aged 6–18 years with molecular definition of chromosome 7 deletions) and compared their performance to a group of 20 normal children aged 4 to 10 years. The 2 groups were matched on language ability (comprehension and expression) through the Diagnostic Verbal IQ Test. Verbal ability was measured by 3 experimental linguistic measures that assessed comprehension of pronouns and production of verbs and nouns.

**RESULTS:** Nonverbal IQ was low and ranged from 40 to 68 points. Those in the WS group, as a whole, showed unimpaired performance on pronouns but faced difficulties in using verbs and nouns. Great variation in performance was evident, which highlights the heterogeneity of the group. A subgroup of individuals with WS showed clear dissociations between language and cognition and within language.

**CONCLUSIONS:** Our results indicate that (1) there is a clear dissociation between language and cognition and (2) children with WS show strengths on some aspects of their linguistic development.

### **A NORMAL LIFE WITH AN UNHEALTHY BODY: SELF-IDENTITY IN ADOLESCENTS GROWING UP WITH CHRONIC ILLNESS**

**Submitted by AnneLoes Van Staa**

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**INTRODUCTION:** Chronic illness is often described in terms of biographic disruption. However, for those growing up with congenital disorders, an unhealthy body is the norm. An important developmental task in adolescence is the formation of self-identity. How does a

chronic disorder influence the development of self-identity in adolescents?

**OBJECTIVE:** Our aim was to investigate attitudes and preferences of adolescents living with chronic disorders.

**METHODS:** A qualitative study was conducted by using semistructured interviews that focused on daily life issues. The sample consisted of 31 adolescents (aged 12–19 years) with various chronic disorders who were randomly selected from the patient database of Erasmus Medical Center-Agia Sophia Children's Hospital. Data analysis was performed by using a qualitative analysis computer program (ATLAS.ti, Berlin, Germany).

**RESULTS:** For most adolescents with chronic disorders, living with illness is "normal." By comparing themselves with healthy peers, they recurrently stress their own normality. They strongly agreed with the statement, "I am like everyone else, my illness is something extra." Therefore, disclosure of health problems remains a sensitive issue, and contact with fellow patients is not often sought. Most held optimistic views about their futures, and only a minority told problematic accounts of the acceptance of their dysfunctional bodies.

**CONCLUSIONS:** Normalization of an unhealthy childhood seems to be an important strategy in identity-forming in adolescents. It may be interpreted as denial, but adolescents consider denial to be "dangerous" and "stupid." We view normalization as a strategy to accept reality while preventing illness to dominate their life: "I try not to think about it, not because it scares me, but because it's there."

## **Endocrinology**

### **LYMPHOCYTES IN PERIPHERAL BLOOD AND THYROID TISSUE IN CHILDREN WITH GRAVES' DISEASE**

**Submitted by Iwona Ben-Skowronek**

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**OBJECTIVE:** Our goal was to analyze interactions of lymphocytes in peripheral blood and thyroid tissue in children with Graves' disease (GD).

**METHODS:** The prospective study concerned 15 children affected with GD and 15 healthy children. The levels of autoantibodies against thyrotropin receptor, thyroid peroxidase, and thyroglobulin were assayed. Monoclonal antibodies (Ortho Diagnostic Systems, Raritan, NJ) were used to define peripheral blood lymphocyte subsets and analyzed by using a flow cytometer. After thyroidectomy, thyroid specimens were stained

with hematoxylin/eosin. T cells were detected by CD3<sup>+</sup>, CD4<sup>+</sup>, and CD8<sup>+</sup> antibodies and the antigen-presenting dendritic cells with CD1a<sup>+</sup> and CD35<sup>+</sup> antibodies (DakoCytomation, Glostrup, Denmark).

**RESULTS:** Before treatment, all children with GD had increased thyroid autoantibody levels, an increased percentage of CD4<sup>+</sup> helper cells, and decreased levels of CD8<sup>+</sup> suppressor/cytotoxic T cells, which resulted in an elevated CD4/CD8 ratio. The percentage of CD19<sup>+</sup>Cd5<sup>+</sup> B cells was increased, although the total population of CD19<sup>+</sup> B cells did not differ from that of the control group. The number of lymphocytes in the thyroid was decreased in 10 patients after long-term thiamazole treatment. In 5 patients with short-term therapy (<6 months after relapse of GD), the lymphocytes had formed lymphatic follicles: antigen-presenting dendritic cells CD1a<sup>+</sup>CD35<sup>+</sup> in the germinal center and T-helper CD4<sup>+</sup>, T-suppressor CD8<sup>+</sup>, and B cells CD79<sup>+</sup> on the edges.

**CONCLUSIONS:** The primary defect of immunoregulation in GD consists of an increase of T-helper lymphocytes with a simultaneous decrease in the number of T-cytotoxic/suppressor cells. Thiamazole therapy probably leads to reduction of the lymphocyte amount in the thyroid.

#### **HLA-DQB1\*05 ASSOCIATION WITH HASHIMOTO THYROIDITIS IN CHILDREN OF NORTHERN GREEK ORIGIN**

**Submitted by Styliani Giza**

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**INTRODUCTION:** Hashimoto thyroiditis (HT), an organ-specific autoimmune disorder of the thyroid gland, is considered to be associated with the major histocompatibility complex. Association studies of human leukocyte antigens (HLAs) with HT concern adults and have not revealed consistent results.

**OBJECTIVE:** We sought to investigate *HLA-DRB1* and *HLA-DQB1* gene polymorphisms in Greek children and adolescents with HT.

**METHODS:** We analyzed the distribution of *HLA-DRB1* and *HLA-DQB1* alleles in 17 Greek children and adolescents with HT and in 181 randomly chosen healthy subjects from northern Greece. The typing of *HLA-DRB1* and *HLA-DQB1* genes was performed by using polymer-

ase chain reaction with sequence-specific primers. Differences of frequencies for HLA alleles were tested by the  $\chi^2$  test.

**RESULTS:** There was no significant association detected between HT and *HLA-DRB1* or *HLA-DQB1* alleles. However, *HLA-DRB1\*16* was slightly significantly increased in patients with HT (41.2%) compared with that in controls (19.3%) ( $P = .057$ ; relative risk: 2.92), and *HLA-DQB1\*05* was significantly increased in patients with an age of diagnosis of >10 years (87.5%) as compared with those with an age of diagnosis of  $\leq 10$  years (33.3%) ( $P = .05$ ; relative risk: 14).

**CONCLUSIONS:** This is the first study to examine children and adolescents from northern Greece with HT and analyze the distribution of *HLA-DRB1* and *HLA-DQB1* alleles according to the age of onset of HT. However, this study needs to include a greater number of patients to ascertain the possibility of an association and avoid the result of a chance event or random variation.

#### **IMPAIRED DIURNAL BLOOD PRESSURE AND HEART RATE VARIATION AND THEIR RELATIONSHIP WITH LEFT-VENTRICULAR FUNCTION IN ADOLESCENTS WITH TYPE 1 DIABETES MELLITUS**

**Submitted by Kyriaki Karavanaki**

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**INTRODUCTION:** Diabetic cardiomyopathy is a well-defined complication of diabetes that occurs in the absence of ischemic heart disease or hypertension and has been associated with autonomic dysfunction.

**OBJECTIVE:** Our aim was to evaluate diurnal blood pressure (BP) fluctuations and autonomic function and their possible association with left-ventricular function in adolescents with type 1 diabetes mellitus.

**METHODS:** In 48 normotensive, normoalbuminuric diabetic adolescents, with a mean ( $\pm$ SD) age of 17.3 ( $\pm 4.1$ ) years and diabetes duration of 8.5 ( $\pm 3.3$ ) years, 24-hour ambulatory BP and heart rate (HR) monitoring was performed. Left-ventricular end-diastolic and end-systolic diameters were estimated by echocardiography, and left-ventricular mass index (LVMI) was calculated.

**RESULTS:** The patients were divided into 2 groups according to the absence of decrease (nondippers) or the decrease (dippers) of nocturnal diastolic BP. The nondippers presented, in comparison with the dippers, reduced mean HR during 24 hours (79.6 vs 84.0 beats/minute;  $P = .05$ ) and also during daytime (81.3 vs 86.0

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