

METHODS: Twenty-seven children known to have GSD were included in this study. Fifteen healthy age- and gender-matched children were also included as controls. Routine urine analysis and measurement of urinary β_2 -microglobulin and microalbumin levels were performed for all patients and controls. Renal-function tests, measurement of serum electrolyte, alkaline phosphatase, urinary calcium, blood, and urine pH levels, creation of a urinary and plasma aminogram, calculation of the glomerular filtration rate, bone radiography to detect rachitic manifestations, and abdominal ultrasound to measure renal size were performed for all patients.

RESULTS: Twenty-one patients had ≥ 1 renal abnormality. The most common was increased urinary β_2 -microglobulin level (15 of 21) followed by an abnormal glomerular filtration rate, whether low or high (8 of 21), and microalbuminuria (6 of 21). Sonographically, there was nephrocalcinosis in 1 case and renal stone in another. The area under the receiver operating characteristic curve for β_2 -microglobulin was 0.86 ($P = .01$) and 0.7 for the urinary microalbumin/creatinine ratio ($P = .15$). The best cutoff level for predicting renal abnormality for urinary β_2 -microglobulin was 0.22 mg/L with 70% sensitivity and 100% specificity, and the best cutoff value for the urinary microalbumin/creatinine ratio was 4.5 with 86% sensitivity and 50% specificity.

CONCLUSIONS: Renal abnormalities are common in patients with GSD. Urinary β_2 -microglobulin level can be considered the gold standard for early detection of renal dysfunction in these patients.

LEPTIN AND LEPTIN RECEPTOR IN SERUM AND URINE FROM CHILDREN WITH NEPHROTIC SYNDROME ACCOMPANYING HYPERLIPIDEMIA

Submitted by Xi Qiang Yang

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INTRODUCTION: Hyperlipidemia may cause glomerulosclerosis in children with nephrotic syndrome (NS).

OBJECTIVE: Our goal was to observe the role of soluble leptin receptor (sOBR) and leptin in serum and urine on the mechanism of hyperlipidemia in children with NS.

METHODS: Twenty-three children with untreated NS and 15 age-, gender-, and BMI-matched healthy controls were enrolled onto the study. Leptin and sOBR in serum and urine were measured by enzyme-linked immunosorbent assay, and plasma lipid and insulin levels were detected by automatic biochemistry analyzer and radioimmunoassay, respectively. sOBR messenger RNA and membrane protein expression in peripheral blood mono-

nuclear cells were detected by reverse-transcription polymerase chain reaction and immunocytochemistry.

RESULTS: Low-density lipoprotein, total cholesterol, triglyceride, and apolipoprotein A levels were increased. sOBR messenger RNA and membrane protein expression by peripheral blood mononuclear cells were significantly lower in the patient group compared with controls. The ratio of serum leptin versus sOBR (free leptin index) was significantly higher in the NS group. Urinary leptin in the patient group was higher than that in the control group. The free leptin index showed no correlation with BMI or total cholesterol, triglyceride, or apolipoprotein B levels in both groups but did show a correlation with plasma albumin, low-density lipoprotein, high-density lipoprotein, apolipoprotein A, and insulin levels in the patient group.

CONCLUSIONS: The reduced sOBR level, which enhanced the biologically active form of leptin in children with NS, might be correlated partly with serum lipid parameters, albumin, and insulin. Increased free leptin in serum might be a complementary mechanism against hyperlipidemia in children with NS.

LONG-TERM PROGNOSIS OF HENOCH-SCHÖNLEIN NEPHRITIS IN CHILDREN

Submitted by Ayse Oner

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INTRODUCTION: The long-term prognosis in Henoch-Schönlein purpura is determined principally by the development of progressive glomerulonephritis (>10% progress to end-stage renal failure).

OBJECTIVE: In this study we aimed to investigate the long-term prognosis of Henoch-Schönlein nephritis (HSN) in childhood.

METHODS: Between 1991 and 2003, 156 patients with HSN were investigated retrospectively.

RESULTS: There were 86 boys and 70 girls with a mean age of 9.6 years. They were graded according to the degree of renal involvement: grade 1, isolated microscopic hematuria ($n = 31$); grade 2, hematuria and mild proteinuria ($n = 60$); grade 3, acute nephritic syndrome ($n = 4$); grade 4, nephrotic syndrome \pm hematuria ($n = 18$); grade 5, acute nephritic and nephrotic syndrome ($n = 43$). Renal biopsy was performed on 43 patients with grade 4 or 5 disease. Twenty patients had extensive crescent formation (>50%) as shown by the renal biopsy and were given triple therapy (intravenous pulse methylprednisolone [30 mg/kg per day for 3 days] fol-

lowed by oral prednisolone [OP], oral cyclophosphamide [2 mg/kg per day for 2 to 3 months], and dipyridamole. The other 23 patients with <50% crescent formation were given methylprednisolone followed by OP and dipyridamole. The patients with grade 3 or 4 disease were given OP and dipyridamole. Those with grade 1 or 2 disease were not given any immunosuppressive agent. During the follow-up period (mean: 30 ± 3.5 months; range: 12–96 months), 23 patients with grade 1, 38 patients with grade 2, 2 patients with grade 3, 8 patients with grade 4, and 21 patients with grade 5 disease showed complete remission (59%). Of the 5 patients with extensive fibrosis shown by renal biopsy, 2 (1%) had persistent nephropathy and 3 (2%) developed end-stage renal failure. The remaining 59 patients showed near-complete recovery with minimal urinary abnormalities (38%).

CONCLUSIONS: Although initial presentation of renal involvement determines the prognosis for those with HSN, intensive treatment with triple therapy seems to be effective for severe renal disease, especially if started before the development of fibrotic changes in crescents and tubulointerstitial tissue.

Neurology

THYROID FUNCTION IN EPILEPTIC CHILDREN TREATED WITH SODIUM-VALPROATE MONOTHERAPY: A PROSPECTIVE LONG-TERM STUDY

Submitted by Achilleas Attilakos

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INTRODUCTION: Sodium valproate (VPA) is widely used for the treatment of partial and generalized epilepsy in childhood and adolescence. The results of studies that have evaluated the effect of VPA monotherapy on thyroid function in children are controversial.

OBJECTIVE: The aim of this study was to investigate, prospectively, whether treatment with VPA has an effect on serum thyroid hormone concentrations in epileptic children.

METHODS: Serum levels of triiodothyronine, thyroxine, free thyroxine, and thyrotropin were determined in 30 epileptic children (aged 2 to 14 years [mean ± SD: 9.10 ± 3.74 years]) before and after 6, 12, and 24 months of VPA monotherapy.

RESULTS: Serum levels of thyroxine and free thyroxine were significantly decreased after 6 ($P = .000$ and

$.000$, respectively), 12 ($P = .000$ and $.015$, respectively), and 24 ($P = .000$ and $.003$, respectively) months of treatment with VPA, whereas serum levels of triiodothyronine were significantly decreased only after 24 months of treatment ($P = .043$). Serum levels of thyrotropin were significantly increased after 6 ($P = .000$), 12 ($P = .000$), and 24 ($P = .000$) months of treatment with VPA. Thirteen children (43.3%) had thyrotropin values higher than the normal-range maximum after 6, 12, and 24 months of VPA monotherapy. Serum VPA concentrations remained within the therapeutic range during the period of study.

CONCLUSIONS: Our results showed that VPA monotherapy in childhood may cause early and persistent alterations in thyroid function, which suggests a need for early and careful monitoring of serum thyroid hormone concentrations in epileptic children who receive VPA.

CLINICAL MARKERS THAT ENHANCE ETIOLOGIC YIELD IN GLOBAL DEVELOPMENTAL DELAY

Submitted by B. H. Y. Chung

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INTRODUCTION: Etiology remains unknown in 30% to 50% cases of children with global developmental delay (GDD). A selective approach has been recommended to increase etiologic yield.

OBJECTIVE: Our aim was to identify clinical markers that enhance diagnostic yield of GDD at initial assessment.

METHODS: The charts of all patients with GDD ($N = 577$) followed up at the Duchess of Kent Child Assessment Centre were reviewed. GDD was defined as significant delay (<2 SD) in ≥2 developmental domains. Nine clinical items at initial assessment (gender, severity of delay, parental consanguinity, family history, behavioral disturbance, head size, facial dysmorphism, malformations, and neurologic deficits) were correlated with the likelihood of finding an etiology for GDD.

RESULTS: A significant threshold effect was found between mild and moderate GDD (positive likelihood ratio [LR⁺]: 1.9; negative likelihood ratio [LR⁻]: 0.72). Other items that significantly affected diagnostic yield were (1) female gender (LR⁺: 1.62; LR⁻: 0.79), (2) behavioral trait (LR⁺: 0.24; LR⁻: 1.67), (3) microcephaly (LR⁺: 2.78; LR⁻: 0.79), (4) presence of facial dysmorphism (LR⁺: 2.65; LR⁻: 0.65), (5) malformation (LR⁺: 1.49; LR⁻: 0.50), and (6) neurologic deficits (LR⁺: 2.86; LR⁻: 0.32). A dose-response relationship was found between LR⁺ and the number of facial dysmorphisms and malformations.

CONCLUSIONS: Most checklists used for GDD are syndrome specific (eg, fragile X syndrome checklists). These 7 clinical markers are useful in the initial assessment,

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