Glucocorticoids in Infancy Didn't Dent Bone Density

CHICAGO — Bone density was no different in children treated with oral glucocorticoids for hemangiomas of infancy than it was in healthy controls, according to a presentation at the annual meeting of the Society for Pediatric Dermatology.

Although oral glucocorticoids are considered the first choice for the treatment of infant hemangiomas, concerns persist about the risk that these children will develop osteoporosis because glucocorticoids may prevent the formation of new bone, wrote Dr. Amy J. Nopper, of the Children's Mercy Hospitals and Clinics in Kansas City, Mo.

To assess the possible impact of systemic glucocorticoids on bone density, she and her colleagues compared 35 infants (mean age 44 months) who received glucocorticoids for hemangiomas for an average of 8.5 months with 35 controls. The average treatment dose was 2.2 mg/kg per day of prednisolone. The average body mass index was approximately 16 kg/m^2 for both groups.

The researchers measured the children's bone density after they had been off treatment for at least 1 year and found that the average spinal bone mineral density was the same (0.6 g/m^2) for both the treatment and control groups. The average total bone mineral density also was the same for both groups (0.8 g/m^2), and no significant differences appeared in the tibial ultrasound measurements between the two groups.

The results support findings from other studies that show steroid use to treat hemangiomas in early childhood does not prevent children from catching up in growth and achieving normal adult height, Dr. Nopper noted.

—Heidi Splete

Transient Arthritis Accounts for a Significant Number of Apparent JIA Cases

BY DIANA MAHONEY
New England Bureau

BOSTON — Small joint involvement and symptoms that last for more than 2 weeks are among the most important early predictors of the evolution of juvenile idiopathic arthritis in children with acute recent onset of the disease, according to findings from a Norwegian study.

A significant percentage of what appears to be juvenile idiopathic arthritis (JIA) actually is transient arthritis, according to the findings.

Knee joint involvement, a neutrophile white blood cell count within the normal range, and a platelet count above the normal limit are also significant predictors of the chronic inflammatory arthritis in children with early symptoms, Dr. Kai S. Handeland reported at the annual meeting of the American College of Rheumatology.

The inability to distinguish between JIA and other types of recent-onset childhood arthritis at an early stage limits the degree to which aggressive management can be carried forth, said Dr. Handeland of Rikshospitalet-Radiumhospitalet medical center, Oslo.

To determine whether any clinical or laboratory features in children with recent-onset arthritis might predict the diagnosis of JIA, Dr. Handeland and colleagues conducted a multicenter population-based study in three Norwegian counties between May 2004 and June 2005. During this period, the investigators asked all primary care physicians in the region to refer for study inclusion all children aged younger than 16 years with symptoms consistent with recent-onset arthritis or osteomyelitis, confirmed by clinical or radiological examination. In addition to prospectively enrolled patients, children who met inclusion criteria were identified retrospectively by searching electronic medical records for relevant diagnoses.

Of the region's 255,303 children, 504 were referred for possible inclusion in the study. A total of 214 patients were enrolled in the study and followed up for a minimum of 6 months, at which point a final diagnosis was made. The investigators reevaluated the diagnoses by chart review after 2 years.

Of the 214 children with confirmed arthritis symptoms, 40 had JIA, 8 had septic arthritis, 5 had osteomyelitis with coexisting arthritis, 21 had poststreptococcal reactive arthritis, 2 had acute rheumatic fever, 4 had Borrelia-arthritis, 1 had enteropathic arthritis, 61 had transient synovitis of the hip, and 72 had other types of transient arthritis, Dr. Handeland said in a poster presentation.

To identify predictors of JIA, the investigators used multiple logistic regression analyses with JIA diagnosis as the dependent variable. Of the factors identified as statistically significant predictors of JIA, the presence of small joint involvement at presentation was the most common, with an odds ratio of 14.1. The odds ratios for symptom duration of 14 days or more, normal neutrophile count, knee-joint involvement, and elevated platelet count, were, respectively, 13.3, 6.2, 4.1, and 3.4, reported Dr. Handeland. Patient gender, monoarthritis, hip-joint involvement, and elevated temperature were not significantly predictive of a JIA diagnosis, he said.

The investigators also conducted subsequent analyses to determine predictors of JIA versus transient/postinfectious arthritis and predictors of JIA versus infectious arthritis.

With respect to the former, the same determinants that discriminate JIA from all arthritis discriminate between JIA and transient/postinfectious arthritis, according to Dr. Handeland.

The determinants that discriminate JIA from infectious arthritis at the time of symptom presentation are the absence of fever and a low occurrence of hip-joint involvement, he said.

Dr. Handeland reported having no conflicts of interest to disclose relative to this presentation. ■

Erythropoietic Protoporphyria Looked Autoimmune at First

BY DOUG BRUNK
San Diego Bureau

CORONADO, CALIF. — It is a wise rheumatologist who knows when to refer a child to a dermatologist, judging from cases reported at the annual meeting of the Pacific Dermatologic Association. Dr. Anna L. Bruckner discussed two such cases.

The first was that of a 9-year-old girl who was referred by a rheumatologist for evaluation of possible dermatomyositis.

The girl had a 4-month history of intermittent redness and swelling of the hands that worsened after prolonged outdoor activities.

The girl was healthy and described one remote episode of burning hands following a hike several years before. She was on naproxen and ranitidine, which had been prescribed by the rheumatologist as treatment for the redness and swelling.

"The work-up by the rheumatologist was negative for autoimmune disease," said Dr. Bruckner.

Clinical exam revealed a few waxy papules and plaques distributed over the knuckles. Her hands also had a slightly weather-beaten appearance. A skin biopsy showed cuffs of hyaline material around the superficial blood vessels in the upper dermis, suggesting a diagnosis of erythropoietic protoporphyria (EPP). Confirmatory studies demonstrated that the patient had elevated total red blood cell porphyrins with a predominance of free protoporphyrin.

Variable findings of TRPS type 1 include short stature, nail and/or teeth abnormalities, and a deep voice.

DR. BRUCKNER

EPP is the most common type of porphyria in children. It presents between 1 and 6 years of age and symptoms include burning, stinging, redness, and edema, all of which occur following sun exposure.

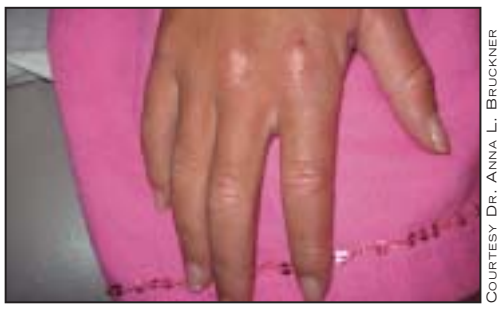
The condition is caused by a deficiency of ferrochelatase, which leads to accumulation of protoporphyrin IX.

Treatment involves sun avoidance, sunscreens, and beta-carotene 30-150 mg/day.

In the second case, a 6-year-old girl with suspected loose anagen syndrome was referred to Dr. Bruckner, who is director of pediatric dermatology at Lucile Packard Children's Hospital in Palo Alto, Calif. In this condition, the anagen hairs are loosely anchored into the scalp so that the hair will fall out with very minor trauma. The hair is short, sparse, and seldom cut. It typically is seen in blond girls aged 2-5 years, but can affect boys and brunettes as well.

Many of the girl's friends at school "had long, flowing hair and she wanted to see if there was something we could do about her hair," Dr. Bruckner said. She did a gentle hair pull test and only two hairs came out. The girl's hair was very short and had a matted appearance in the back.

Dr. Bruckner prescribed 5% minoxidil lotion and scheduled a 3-month follow-up visit. On follow-up the girl's hair was fuller but it remained short and gentle hair pull tests remained negative.



COURTESY DR. ANNA L. BRUCKNER

Waxy papules and plaques are shown on a girl with erythropoietic protoporphyria.

"We obtained some additional history," Dr. Bruckner recalled. "Her nails were thin, often peeled, and never required trimming. She had no history of dental anomalies, and she'd had a coarse, deep voice since age 2. Her mother said that she looked different than her siblings."

She also had sparse lateral eyebrows, a pear-shaped nose, and a thin upper lip.

The girl's combination of short, sparse hair and abnormal facial features led Dr. Bruckner to consider trichorhinophalangeal syndrome (TRPS) type 1 as the diagnosis.

An x-ray of the girl's hand performed after her follow-up visit revealed cone-shaped epiphyses of the phalanges, which confirmed the diagnosis. TRPS type 1 is an autosomal dominant disorder characterized by craniofacial and bony abnormalities that include sparse, slow-growing hair and thin lateral eyebrows, a pear-shaped nose, elongated philtrum and thin upper lip, prominent ears, and cone-shaped epiphyses of the phalanges.

Variable findings include short stature (the patient was in the 25th percentile for height), nail abnormalities, teeth abnormalities, and a deep voice. The condition is caused by mutations in the TRPS1 gene.

Although there is no specific treatment for TRPS type 1, the girl's parents were happy to better understand why their daughter's hair failed to grow normally. She has continued to use 5% minoxidil for 6 months with some improvement. ■