Bone Mineral Density of the Lumbar Spine and Proximal Femur Is Decreased in Children With Sickle Cell Anemia

Mark R. Brinker, MD, Kevin A. Thomas, PhD, Steven J. Meyers, MD, Terry Texada, MD, James R. Humbert, MD, Stephen D. Cook, PhD, and Richard Gitter, MD

ABSTRACT

Bone mineral density (BMD) was evaluated in the proximal femora (femoral neck, Ward's triangle, and greater trochanter) and lumbar spines of 25 black children and young adults with sickle cell anemia using dual-photon absorptiometry. Compared with normal subjects from the general population, the patients with sickle cell anemia exhibited lower bone mineral density values in all scan regions (approximately 6% to 21% lower than expected). These differences in the lumbar spine were significant for both girls and boys. When compared with normal black subjects from the general population, the girls with sickle cell anemia exhibited significantly lower lumbar spine bone mineral density, and the boys with sickle cell anemia exhibited significantly lower bone mineral density in the femoral neck and Ward's triangle. No consistent or significant correlations were found between the bone mineral density data and the patients' hematologic indices.

Sickle cell anemia is a chronic hereditary disorder commonly found in the black population. It is characterized by chronic hemolytic anemia and tissue infarction, secondary to microvascular occlusion by sickled erythrocytes. Many organ systems are affected by the vaso-occlusive process of sickle cell anemia, including the musculoskeletal system. Repeated ischemic episodes affecting the musculoskeletal system are responsible for much of the morbidity associated with this disease.

The effects of sickle cell anemia in bones are the result of two processes: bone marrow hyperplasia and vaso-occlusion. Bone marrow hyperplasia is a response to the chronic anemia associated with this disease. The normal half-life of red blood cells is approximately 60 days, but with sickled erythrocytes, it is 5 to 20 days. Bone marrow hyperplasia is most evident in the first 6 months after birth, a time when fetal hemoglobin is being replaced by sickle cell hemoglobin.

The earliest changes in sickle cell anemia are found in the metacarpals, with the classic change being squaring of the first metacarpal head. The spectrum of changes are expansion of the medullary space, connective-tissue replacement, coarsening of the trabeculation with cortical intrusion, cortical thinning, and a periosteal reaction. As the disease process continues, there is decreased osteoblastic activity and decreased bony formation, leading to an osteoporotic state. These changes have been shown in the lumbar vertebrae by many investigators.[1-6] It was found that lumbar vertebral bodies had a decrease in vertical height, an increase in width, and a biconcave or fish vertebra appearance.

The vaso-occlusive effects of sickle cell anemia are more responsible for the morbidity and disability of the disease, unlike the hyperplastic changes that are more responsible for diagnostic purposes. Vaso-occlusive changes begin early in the disease process when fetal hemoglobin is being replaced by sickle cell hemoglobin. Dactylitis or "hand-foot syndrome" is the first clinical manifestation experienced; it usually manifests during the first 6 years of age and is characterized by infarction of the medullary cavity and diaphyseal cortex of the metacarpals, metatarsals, and phalanges. Medullary infarction is due to vaso-occlusion of the nutrient

Dr. Brinker is Director, Department of Acute and Reconstructive Trauma, Fondren Orthopedic Group LLP, Texas Orthopaedic Hospital, Houston, Texas, and Clinical Associate Professor, Department of Orthopaedic Surgery, Tulane University School of Medicine, New Orleans, Louisiana.
Dr. Thomas is Associate Professor, Department of Orthopaedic Surgery, Louisiana State University School of Medicine, New Orleans, Louisiana.
Dr. Meyers is Medical Student, Department of Orthopaedic Surgery, University of Texas Medical School, Houston, Texas.
Dr. Texada is Clinical Assistant Professor, Dr. Cook is Director of Orthopaedic Research, and Lee C. Schlesinger Professor, and Dr. Glitter is Medical Student, Department of Orthopaedic Surgery, and Dr. Humbert is Professor, Department of Pediatrics, Tulane University School of Medicine, New Orleans, Louisiana.