Chronic Bone Disease in Pediatric Sickle Cell Disease

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Background

Avascular necrosis (AVN) is common in Sickle Cell Disease (SCD), frequently involving the femoral and humeral head and less commonly involving the spine. AVN leads to joint collapse, chronic pain and disability, and often requires joint replacement in early adulthood. AVN develops in childhood with 27% of children having femoral AVN (Adekile et al 2001), and 50% of adults affected by 35 years of age (Milner et al 1991). There are no medical therapies for AVN in SCD despite the high burden of disease and there are no published reports of bisphosphonate therapy in this condition.

Methods

We performed a retrospective review of our centre's cohort, evaluating for bone disease in pediatric patients with SCD. This study was conducted with approval from the Research Ethics Board of the University of Alberta.

Radiography of Chronic Bone Disease in SCD

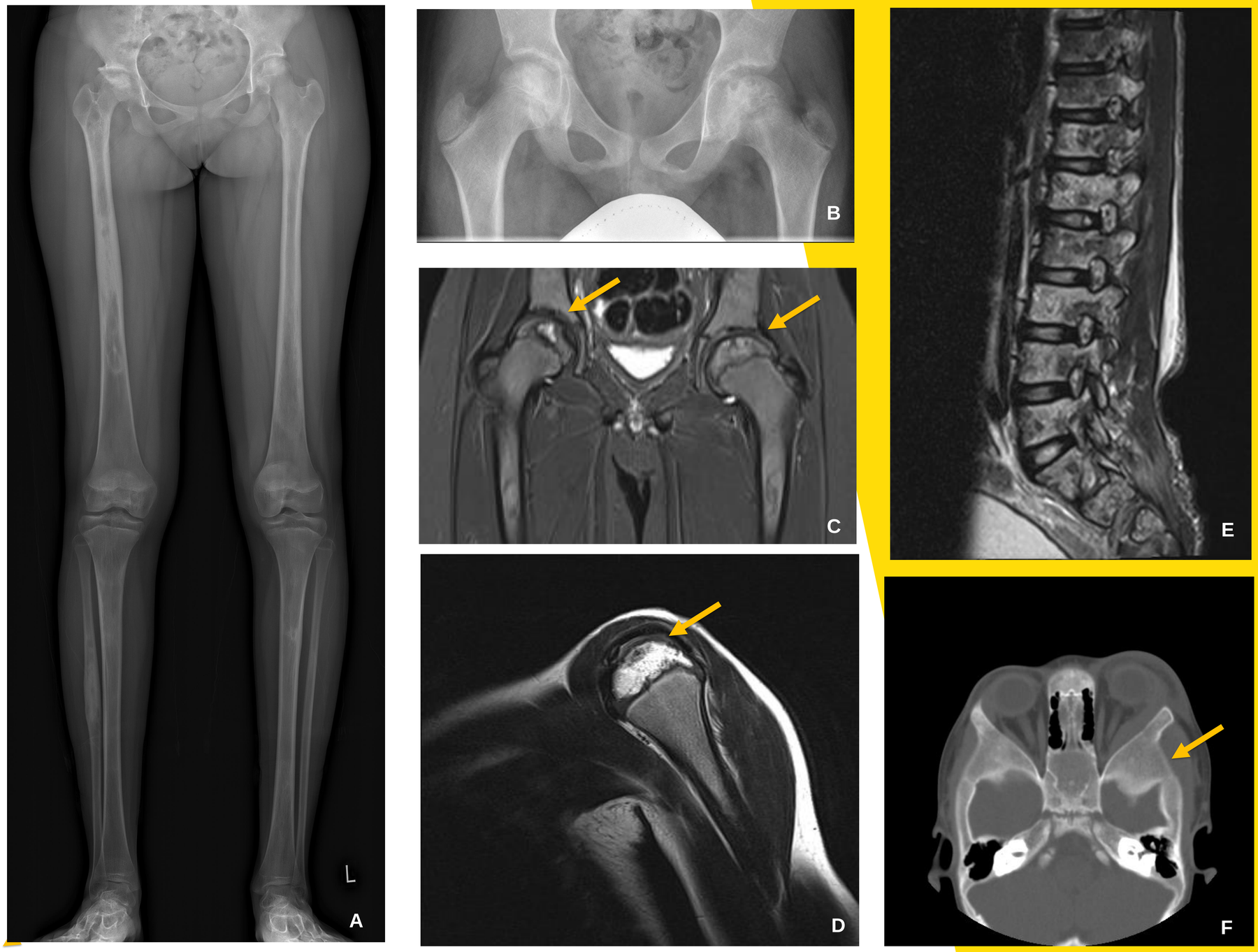

Figure 1: Diversity of Bone Disease in SCD. A) Patient with bilateral femoral head AVN and femur/tibial sclerosis due to bony infarcts. B) Possible bilateral femoral head AVN on Xray. C) MRI showing increased STIR intensity in femoral heads of the patient in (B), confirming bilateral AVN. D) Humeral head AVN. E) Bone infarcts throughout the thoracic, lumbar and sacral vertebral bodies. F) Sphenoid bone infarct with orbital hematoma.

Bisphosphonate Therapy for Vertebral AVN

An adolescent male with HgSS had a complicated course including spontaneous sphenoid infarct with orbital hematoma and AVN of the femur and humerus. The patient had chronic back pain and recurrent vaso-occlusive crises localized to the lumbar region. Imaging showed extensive vascular necrosis of the vertebral bodies (Figure 2A & 2B). Intravenous bisphosphonate improved pain and stabilized vertebral body height (Figure 2C).

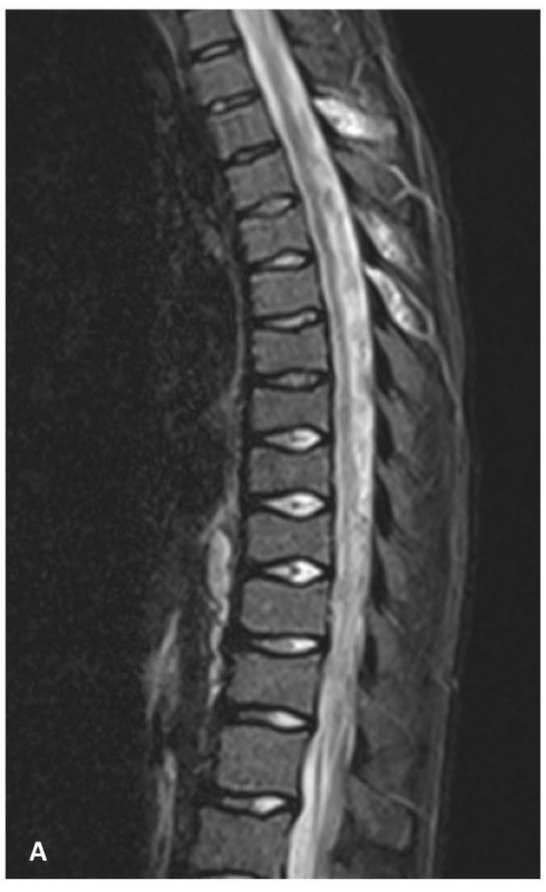

Figure 2A: STIR weighted MRI with high signal in several spinous processes, consistent with AVN

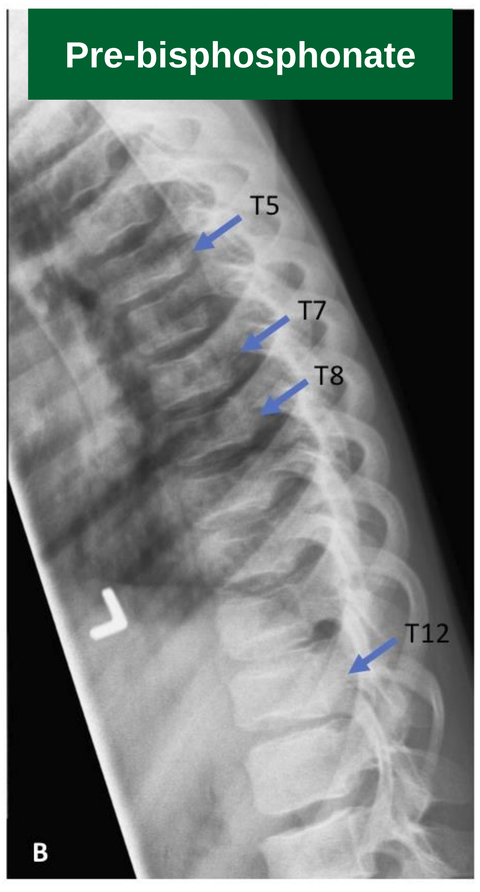

Figure 2B: Spine XR demonstrating multiple vertebral body involvement

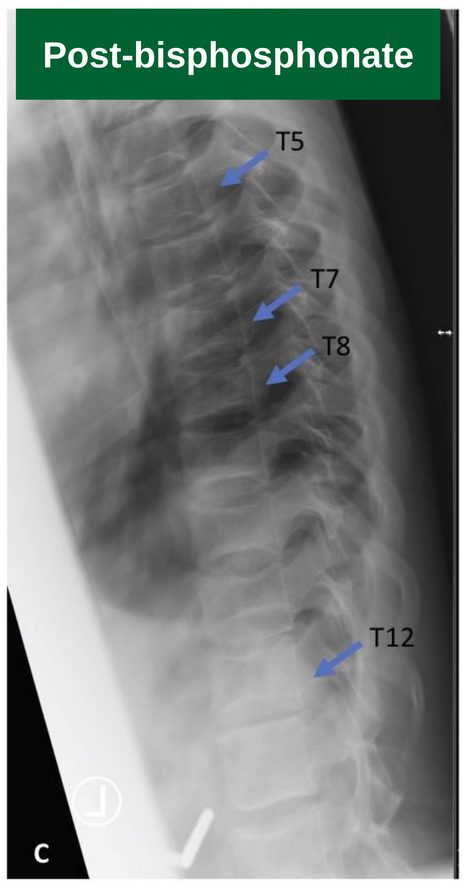

Figure 2C: Spine XR demonstrating vertebral body stabilization with improved height and less end-plate irregularity

Results

N= 14 (%)	
10 (71%)	Presented by 12 years of age
8 (57%)	Male
12 (86%)	HgSS
10 (71%)	Vertebral AVN
4 (29%)	Femoral AVN
6 (43%)	Lytic/sclerotic lesions – femur, pelvis, sphenoid bone, mandible
7 (50%)	More than one bony insult

Conclusion

Bone Disease in Sickle Cell Anemia is diverse and can present early in childhood. Our cohort had evidence of disease as early as 2 years of age. The majority of patients presented by 12 years of age. Vertebral AVN was much higher than previous reports, affecting 85% of our cohort. We recommend having a low index of suspicion and pursuing MRI early. Future research should look at bisphosphonates as a therapy option for pain and stabilization of bone necrosis

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