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.

Results

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.

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.