G51 Sickle Cell Trait Associated Deaths: A Case Series With a Spectrum of Clinical Presentations

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After attending this presentation, attendees will be educated with respect to the wide variety of clinical presentations persons with sickle cell trait manifest including clinical symptoms, laboratory abnormalities, and gross anatomic and microscopic findings. This study also seeks to show how simply the diagnosis can be made by the astute clinician or forensic pathologist if only he or she will consider it in the differential diagnosis.

This presentation will impact the forensic community by showing that sickle cell trait is a condition which is not restricted to conventional ethnic boundaries of Afro-Americans and that the diagnosis needs to be seriously considered in individuals living in geographic locations in which the natural environment plays a prominent role in the manifestation of the disease. Early recognition of the disease in such individuals can possibly result in a decline in mortality.

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This presentation will hopefully have a significant impact on not only the forensic community but also on humanity by showing that sickle cell trait is a condition which is not restricted to conventional ethnic boundaries of Afro-Americans and that the diagnosis needs to be seriously considered in individuals living in geographic locations in which the natural environment plays a prominent role in the manifestation of the disease. Early recognition of the disease in such individuals can possibly result in a decline in mortality. As many as one in three Africans living in areas where malaria is indigenous and approximately one in twelve Americans with African ancestry have sickle cell trait. The affected individuals are generally asymptomatic and many are not even aware that they carry the gene. The general consensus of the public is that sickle cell trait is a relatively benign condition and affected persons are at no increased risk of morbidity or mortality because of their condition. However, the forensic community is cognizant that under the proper set of circumstances, sickle cell trait can be potentially fatal.

This study presents a series of 11 individuals with sickle cell trait and one with hemoglobin SC disease who died during various circumstances. All of the victims were subject to the warm and humid climate of Florida. The onset and/or duration of symptoms varied from a few to several hours with many displaying a prolonged lucid interval with stable vital signs. Despite seeking medical treatment, sickle cell trait related micro-occlusive crisis was never considered in the differential diagnosis. Several cases were associated with sudden death. In those deaths which were delayed, high anion gap and uncompensated metabolic acidosis were typical. Also characteristic were large increases in creatine phosphokinase, alanine aminotransferase and aspartate aminotransferase along with myoglobinemia. Although the antemortem diagnosis of rhabdomyolysis was made, the underlying cause was never deduced by the clinicians. Of particular interest was a case of a fatal splenic crisis due to sickle cell trait in a Caucasian and a victim with hemoglobin SC who died from a combination of mild traumatic injuries and prolonged bodily inversion. In some cases, sickle cell trait was not even considered in the original death certification.

In conclusion, this study demonstrates the varying characteristics and presentations of 11 cases of sickle cell trait and one case of hemoglobin SC related deaths and shows that such deaths can be sudden or delayed. Conventional racial delineation of the sickle cell hemoglobinopathies should not deter one from considering it in the differential diagnosis especially if the patient is subjected to environmental and physical stressors which can potentiate the disease. Furthermore, failure to consider sickle cell trait related crises as a diagnosis can result in improper death certification. Greater efforts to educate the public especially athletes and coaches on the possible hazards of exercise induced sickle cell trait related micro-occlusive crisis hopefully will result in less morbidity and mortality.

Sickle Cell Trait, Exertion, Metabolic Acidosis

 ** Presenting Author*