Prevalence of venous or arterial thrombosis in hospitalized patients with Sickle cell disease

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Introduction

- Sickle cell disease (SCD) is a systemic disease, and, in itself, is a prothrombotic condition.
- Sickle cell disease is caused by a mutation in the hemoglobin-beta chain gene that is either inherited by one or both parents.
- Homozygous sickle cell anemia (HbSS) is the most severe form of sickle cell disease, and, most commonly, is caused by the inherited sickle cell genes from both parents.
- Hb SC disease and beta thalassemia are clinically milder than HbSS disease.
- Patients who are hospitalized due to pain crises are invariably immobilized, which is an additional risk factor for thrombosis.
- Yet, there has not been any standard practice and procedure to prevent thrombosis in patient with SCD.
- Even though literature has documented the increased thrombotic risk of the patients with sickle cell disease, there has not been any quantitative data to show actual concrete risk.

Objectives

The objectives of this study are to find the actual number of thrombotic episodes that patients with sickle cell disease developed during the hospital stay, and to compare it with that of control non-sickle cell patients.

Subjects

Patients of all ages with sickle cell disease (SCD)(SS, SC, & S/β^+). Patients who were diagnosed with any sickle cell genotype are *case patients*. Patients who were not diagnosed with any sickle cell genotype are *control patients*.

Methods

With institutional review board approval, investigators conducted retrospective chart review of patients admitted with sickle cell disease at Hurley Medical Center and was diagnosed with a thrombotic event between April 2012 and April 2020.

To obtain a logistic model, investigators (with the help of Hurley IT) identified all patients with the the final discharge diagnosis of thrombosis (ICD-9 or ICD-10 code indicating any thrombosis, such as DVT, arterial thrombosis, catheter-related thrombus, pulmonary embolism, and stroke), combined with the diagnosis of sickle cell disease. Repeats of case subjects were eliminated from the database and a special data extraction sheet was developed. For the control subjects, the total numbers of hospital admissions and of thrombosis admissions between April 2012 and April 2020 were obtained. Contingency table is shown in Table 1.

Contingency Table (Table 1)

	Thrombosis	No Thrombosis	Total	Prevalence
Case patients	37	100	137	.27=27%
Controls patients	7044	145,161	156,205	.045=4.5%.

Results

A total of 137 case patients (Age range 20-76 median: 36) were identified of whom 37 (27%) patients were found to have at least one thrombotic event during their hospitalization. 17 of 37 were venous thrombosis and 20 were arterial thrombosis. As predicted, patients with sickle cell disease appeared to have a very high frequency of thrombotic complications compared to the control group (4.5%) (Figure 1). The frequency of the thrombosis varied with the genotype in this study (68 Hb SS disease, 49 Hb SC disease, 20 Hb S/β^+ thalassemia). Patients with the Hb-SS disease had a much higher rate of thrombotic complication(25/68=37%) compared with those with Hb SC disease patients (12/49=24%) (p,0.01, chi square=10.47) (Figure 2). There were no thrombotic events reported in patients with Hb S/beta thalassemia+, or 0 patients or in patients with Hb SS with HPFH.

Figure 1

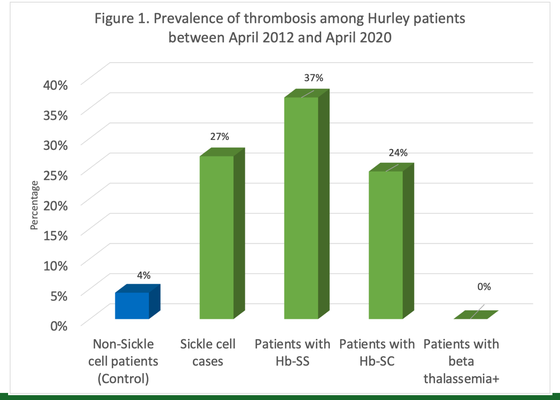
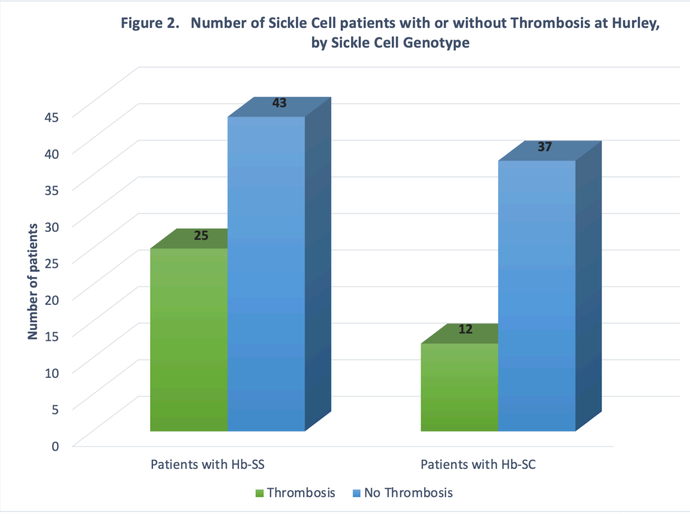


Figure 2



Discussion and Conclusion

Even though one may empirically use aspirin or low molecular weight heparin (Lovenox) in a hope to prevent a thrombotic event, this practice, however, is based on empirical data and experts' opinions. There have not been any randomized studies to systematically answer the question of what evidence-based preventive measures can be used for patients with high risk of thrombosis, such as patients with sickle cell disease. This study only addresses the risk of thrombosis in sickle cell patients and has potential limitations, such as the small number of patients, and multiple admission counted for the control number. Thus, more research should address the need of evidence-based prophylactic agents to prevent thrombosis, specifically for patients with sickle cell anemia. These preventive measures could potentially reduce the mortality rate of sickle cell disease that are associated with thrombosis.

Interpretation

The risk of a thromboembolic event in patients with Sickle cell disease is high. Sickle cell anemia is a procoagulant condition, which is a risk factor for thrombosis. Further research is required to determine if preventive modalities could reduce thrombotic risk.

Reference: SCDResearchReferences