Abstract: P1506

Title: MORTALITY AND CLINICAL COMPLICATIONS AMONG PATIENTS WITH SICKLE CELL DISEASE WITH RECURRENT VOCS IN CANADA

Abstract Type: Poster Presentation

Topic: Sickle cell disease

Background:

Sickle cell disease (SCD) is an inherited hemoglobinopathy that results in the formation of sickle hemoglobin leading to painful vaso-occlusive crises (VOCs) and end-organ damage. VOCs are a marker of disease severity and can cause premature mortality in patients with SCD.

Aims:

To describe the mortality and clinical complications among patients with SCD with recurrent VOCs in Canada.

Methods:

This longitudinal, retrospective cohort study used administrative datasets from the Institute for Clinical Evaluative Science in Ontario, Canada to identify patients with a diagnosis of SCD between 1 January 2010 and 31 December 2021. Patients with SCD who had ≥2 VOCs/year in any 2 consecutive years were eligible for inclusion. A VOC was defined as SCD with crisis, priapism, or acute chest syndrome. The index date was the second VOC in the second consecutive year. Patients were required to have ≥1 year of continuous enrollment before and after the index date. Each patient was matched to 3 controls from the general population without SCD by age, sex, and region. The pseudo-index date for controls was the same as the index date for patients. Patients and controls were followed from the index date until censoring, defined as the earliest of death, loss to follow-up, most recent data availability, receipt of hematopoietic stem cell transplant, or end of study period (31 December 2022). Demographics were assessed at index date. Mortality (proportion of total population and rate [deaths per 100 person-years]) was summarized in the follow-up period. Clinical complications (proportion of total population) were summarized descriptively during the study period. Survival among patients and matched controls was described using Kaplan-Meier analysis starting from index date. A chi-square test was used to assess statistical difference in mortality proportion between cases and matched controls (P<0.05). Subgroup analysis was conducted based on rate of VOCs in the follow-up period (<2 VOCs/year, ≥2 VOCs/year).

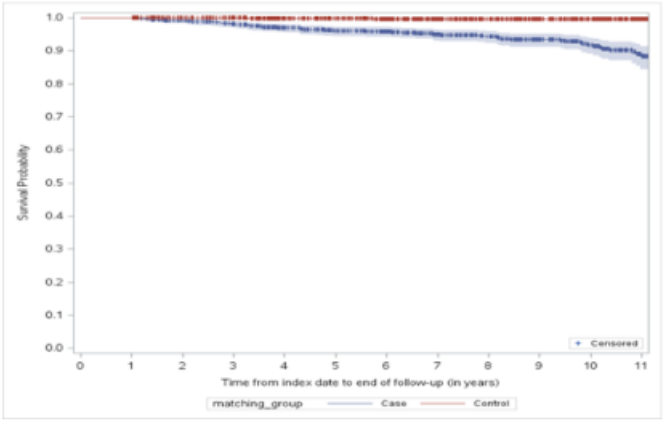
Results:

Overall, 859 patients with SCD with recurrent VOCs met inclusion criteria and were matched with 2,577 controls. The mean age of patients at index was 22.07 years and 50.87% were female; matched controls had similar demographics. Compared to controls, a higher proportion of patients with SCD were in the lowest income quintile (SCD: 38.42% vs controls: 21.03%). Mean follow-up was 7.44 years for patients and 7.66 years for controls. Mortality proportion (SCD: 55/859 [6.40%] vs controls: 9/2,577 [0.35%]; P<0.001) and rate (SCD: 0.86 vs controls: 0.05 deaths per 100 person-years) were substantially higher for patients with SCD than for controls (Figure 1). Mean age at death for patients with SCD was 39.22 years. The most prevalent complications among patients with SCD were bone and joint complications (23.2%), gallstones (20.6%), infection (19.9%), mental health complications (13.4%), acute renal failure (13.2%), chronic pain (12.6%), cardiopulmonary complications (12.2%), and pulmonary embolism (12.0%). Mortality rate was higher in the subgroup of patients with more VOCs during the follow up period (≥2 VOCs/year: 1.54 vs <2 VOCs/year: 0.26 deaths per 100 person-years). Patients with more VOCs in the follow up period generally had a higher prevalence of clinical complications.

Summary/Conclusion:

Patients with SCD with recurrent VOCs had substantially higher mortality and prevalence of clinical complications than matched controls, which worsened with increased VOC frequency. These findings highlight the need for novel therapies in this patient population.

Figure 1. Kaplan-Meier Curve on patients with SCD with recurrent VOCs and matched general population controls



Keywords: Sickle cell disease, Heme