Clinical Guidance on COVID-19 Vaccines for People with Sickle Cell Disease

This guidance is intended for health-care providers and is based on available evidence as of April 18, 2023.

Sickle Cell Disease (SCD) is an inherited blood cell disorder predominantly affecting persons of African ancestry with an estimated prevalence of 1/500 African Canadians. Patients with SCD often have underlying cardiopulmonary co-morbidities that may predispose them to poor outcomes if they become infected with SARS-CoV-2. The majority of adult sickle cell patients also have functional asplenia which contributes to infection severity and adverse outcomes.

Is COVID-19 immunization recommended for people with sickle cell disease?

COVID-19 vaccines should be encouraged for adults and youth with sickle cell disease and are not contraindicated, including those who have had COVID-19 infection. This recommendation is based on the following review:

- In a UK cohort study, persons with SCD and SARS-CoV-2 infection have increased hazard ratios of hospitalization (4.87 in men and 6.68 in women) and death (4.41 in men and 5.94 in women).¹
- In an American registry study of 784 patients with SCD and SARS-CoV-2 infection (average age 22 years), 50.5% were hospitalized, 8.2% admitted to ICU, 3.1% required ventilatory support, and the death rate was 2.4%.²
- A retrospective electronic medical record study identified 312 patients with COVID-19 and SCD between January and September 2020. Those with SCD had 2.0 times higher risk of hospitalization, 2.4 times higher risk of pneumonia, and 3.4 times higher risk of pain crisis compared to matched individuals without SCD.³
- The U.S. Centers for Disease Control (CDC) identifies SCD among the medical conditions at high risk for severe COVID-19 disease.⁴
- Children with SCD may be at higher risk of morbidity with COVID-19 infection including acute vaso-occlusive events and severe acute chest syndrome requiring exchange transfusion.⁵
- The American Society of Hematology recommends that given the high levels of efficacy for vaccines and the low rates of vaccine-related adverse reactions, providers should encourage SCD patients to receive COVID-19 vaccinations as soon as possible.⁶

There are data to suggest that the currently available COVID-19 vaccines have efficacy.⁷ While data specific to the safety and efficacy of COMIRNATY (Pfizer-BioNTech), SPIKEVAX (Moderna), and VAXZEVRIA (AstraZeneca) vaccines in people with sickle cell disease is currently limited, trials studying vaccine efficacy in people with sickle cell disease are ongoing.⁸
The authors of this guidance agree that the benefits of COVID-19 immunization with these vaccines outweigh any theoretical risks of immunization.9

**Are COVID-19 vaccines efficacious and safe for people with sickle cell disease?**

As sickle cell disease is considered to be a severe underlying medical condition, persons with sickle cell disease were excluded from the Pfizer-BioNTech and Moderna COVID-19 vaccine clinical trials. While data collection is ongoing10, it is currently unknown if COVID-19 vaccines are as efficacious for patients with sickle cell disease as they were found to be for the clinical trial participants.

Due to the functional asplenia of the majority of people with SCD, persons with SCD are immunocompromised.6 As with most vaccines, there is a potential for blunted immune response in individuals who are immunocompromised due to their disease or treatment.9,11 It is therefore possible that persons with SCD may not respond as well to COVID-19 vaccines as the general population and should continue to follow local public health guidelines and adhere to precautionary infection prevention procedures following immunization for as long as SARS-CoV-2 continues to circulate at high rates in the community.

While not specific to COVID-19 vaccines, persons with SCD may be at risk for developing vaso-occlusive pain crises within a few days of immunization with any vaccine and should be counselled to monitor for symptoms and advise their SCD care providers in the event that they develop symptoms.

Currently, there are no serious safety warnings or precautions associated with the COMIRNATY (Pfizer-BioNTech), SPIKEVAX (Moderna), and VAXZEVRIA (AstraZeneca) vaccines in persons with SCD beyond those of the general population. If vaccination with the VAXZEVRIA (AstraZeneca) vaccine is considered, clinicians should be aware of the rare potential for development of venous or arterial thrombosis accompanied by thrombocytopenia 4 to 30 days after vaccination.4

**Are there any specific contraindications or exceptions for people with sickle cell disease?**

Individuals who have had a severe allergic reaction to an ingredient of one type of COVID-19 vaccine are still able to receive future doses of the other type of vaccine.12 BCCDC has a list of the individual components and their purpose in the vaccines. For a complete list of components in the vaccine, consult the vaccine monographs found at: www.bccdc.ca/health-info/diseases-conditions/covid-19/covid-19-vaccine/vaccines-for-covid-19.

For individuals with a history of anaphylactic reaction to a previous dose of an mRNA COVID-19 vaccine, re-vaccination (i.e., administration of a subsequent dose in the series when indicated) may be offered with the same vaccine or the same mRNA platform if a risk assessment deems that the benefits outweigh the potential risks for the individual and if informed consent is provided. Prior to revaccination, consultation with an allergist or another appropriate physician
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(e.g., Medical Health Officer) is advised. If re-vaccination is going ahead, vaccine administration should be done in a controlled setting with expertise and equipment to manage anaphylaxis, with an extended period of observation of at least 30 minutes after re-vaccination.

Health Canada continues to monitor any adverse events following immunization through their post-authorization surveillance process.

Otherwise, there are no contraindications or exceptions to immunization for individuals within the SCD population beyond those for the general population.

COVID-19 vaccines can be given concomitantly with, or any time before or after any other indicated vaccine.13-16

Are there specific recommendations or considerations for safe and/or most effective administration?

SCD patients receiving chronic red blood cell exchange should be vaccinated within 10 days following exchange to minimize the potential for developing vaso-occlusive pain crises. Otherwise, there are no additional timing considerations for the administration of the COVID-19 vaccine in SCD patients.

References

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