



OBJECTIVE

To provide guidance to health care providers when they receive notification a newborn in their care is a sickle cell trait carrier. Providers will be responsible for communicating results with the family and arranging necessary follow-up in accordance with this guide.

TARGET POPULATION

Infants born in Alberta after April 1, 2019

EXCLUSIONS

Adults

RECOMMENDATIONS

See algorithm in [Appendix A](#).

BACKGROUND

Sickle cell disease (SCD) is a clinically significant hemoglobinopathy that is inherited in an autosomal recessive manner. In patients with SCD, red blood cells can acquire a crescent or 'sickle' shape, which may cause pain and tissue damage. Individuals with SCD have two sickle cell gene variants, or one sickle cell gene variant alongside a second gene variant that impacts the quantity or quality of adult hemoglobin, such as beta thalassemia.

When an individual has only one sickle cell gene variant, they are said to carry **sickle cell trait (SCT)**. SCT occurs in all ethnic groups, but it is more common in certain populations such as African, Mediterranean, Middle Eastern and Asian groups. Carrier frequency is dependent on ethnic background. Worldwide, SCT affects an estimated 300 million people, with a prevalence ranging from 2% to 30% in more than 40 countries¹. The carrier frequency is estimated to be approximately 1/100 overall in the Alberta population.

IMPLEMENTATION CONSIDERATIONS

SUMMARY

There are no immediate health concerns for an infant who carries SCT. No urgent action is required and referral to a hematologist is generally not necessary. However, there may be implications for other family members. See [Appendix B](#) for frequently asked questions to help guide your discussions with the family.

HEALTH EXPECTATIONS FOR PEOPLE WITH SICKLE CELL TRAIT

Most people who carry SCT will not have associated health problems and they will not develop SCD. In rare cases, carriers may experience some health issues such as hematuria, proteinuria or chronic renal disease, see [Centres for Disease Control and Prevention – Sickle Cell Trait Toolkit](#) for additional information. People who carry SCT may also experience exertional health illness associated with extreme strenuous activities or during periods of significant dehydration.

While SCT is usually a benign condition that does not require referral to a hematologist, if you are concerned that your patient may have active clinical symptoms associated with SCD, please contact pediatric hematology, based on the family’s location preference:

Edmonton

Stollery Children’s Hospital
8440 – 112 Street
Edmonton, AB T6G 2B7
Phone: 780-248-5415
Fax: 1-888-775-9068

Calgary

Alberta Children’s Hospital
28 Oki Dr. NW
Calgary, AB T3B 6A8
Phone: 403-955-7155
Fax: 403-955-7393

IMPLICATIONS FOR OTHER FAMILY MEMBERS

When a child is found to carry SCT, at least one parent is also expected to carry SCT. Given how common SCT is, there is a chance that both parents carry SCT. These couples have a 1 in 4 probability of having a child with SCD (see [Figure 1](#)).

If the parents of an infant carrying SCT are planning more pregnancies, the following hemoglobinopathy investigations are recommended: hemoglobin electrophoresis, CBC and ferritin. Genetic counselling is recommended if both parents are found to be carriers for any thalassemia or other hemoglobinopathy.

If your patient’s family is interested in reproductive genetic counselling, and both parents are carriers, please contact the Clinical & Metabolic Genetics Program, based on the family’s location preference:

Edmonton

8-53 Medical Sciences Building
8440 – 112 Street
Edmonton, AB T6G 2H7
Phone: 780-407-7333
Fax: 780-407-6845

Calgary

Alberta Children’s Hospital
28 Oki Dr. NW
Calgary, AB T3B 6A8
Phone: 403-955-7373
Fax: 403-476-8762

RESOURCES FOR HEALTHCARE PROVIDERS

- Centres for Disease Control and Prevention – Sickle Cell Trait Toolkit:
<https://www.cdc.gov/ncbddd/sicklecell/toolkit.html>

- Alberta Health Services, NMS Program Provider Information Sheet – Sickle Cell Disease: <https://www.albertahealthservices.ca/info/Page9080.aspx> under the ‘Provider Info’ tab

RESOURCES FOR PATIENTS AND FAMILIES

- Sickle Cell Foundation of Alberta: www.oursdfa.org
- Sickle Cell Society: www.sicklecellsociety.org
- My Health Alberta: <https://myhealth.alberta.ca>

REFERENCES

1. Naik, R. P., Smith-Whitley, K., Hassell, K. L., Umeh, N. I., de Montalembert, M., Sahota, P., et al. Clinical outcomes associated with sickle cell trait: a systematic review. *Ann Intern Med* [Internet]. 2018 November [cited 2019 March 19]; 169 (9), 619-627. Available from: <https://annals.org/aim/article-abstract/2709819/clinical-outcomes-associated-sickle-cell-trait-systematic-review>

SUGGESTED CITATION

Toward Optimized Practice (TOP) Sickle Cell Trait Working Group. 2019 April. Practice guide for sickle cell trait identified in newborn screening. Edmonton, AB: Toward Optimized Practice. Available from: <http://www.topalbertadoctors.org>

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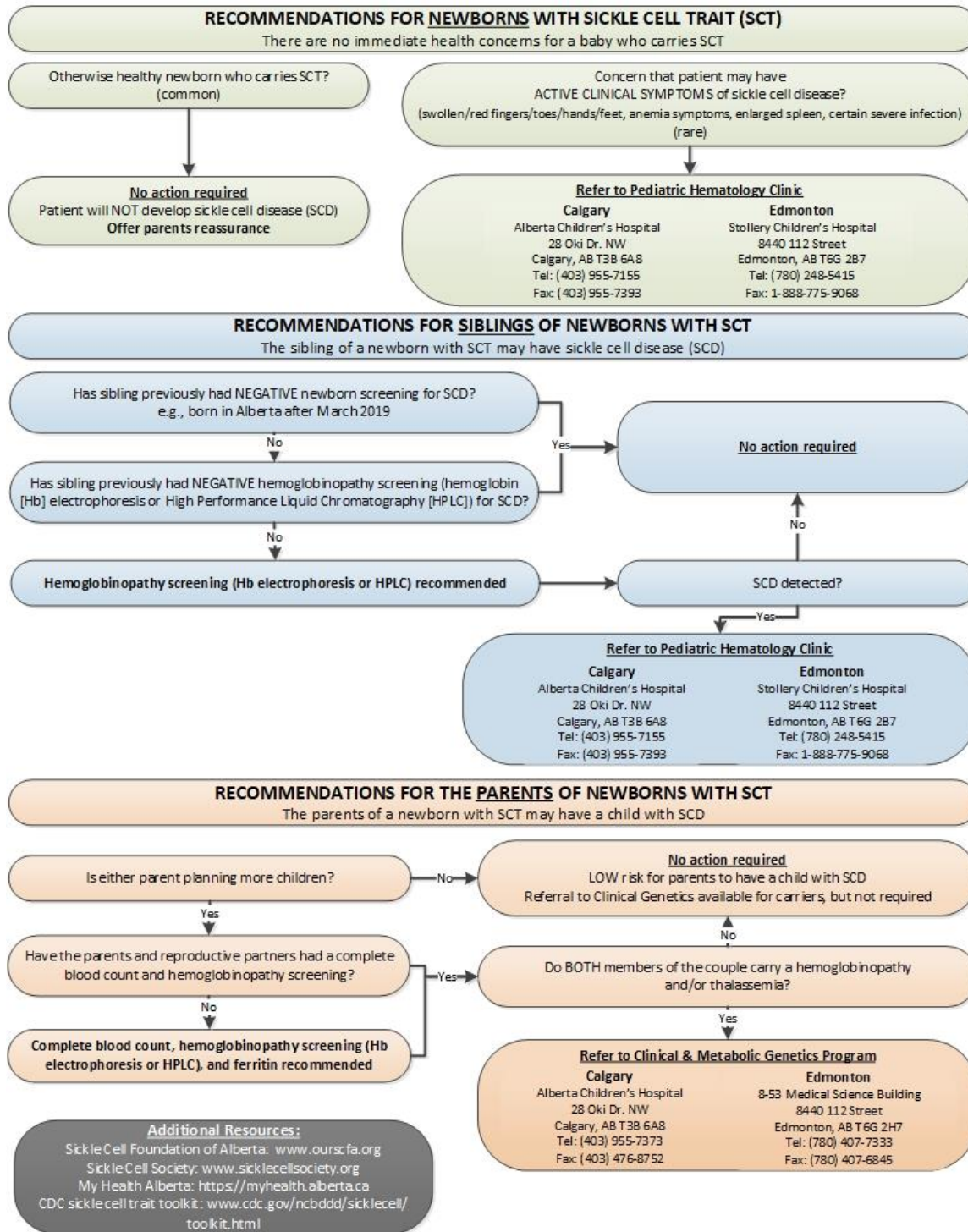
COMMITTEE

The committee consisted of representatives of family medicine, medical genetics, genetic counsellors, pediatric hematology, screening programs and public health and preventive medicine. (See [Committee Members and Conflict of Interest Summary](#).)

April 2019

APPENDIX A

SICKLE CELL TRAIT NOTIFICATION ALGORITHM



These recommendations are systematically developed statements to assist practitioner and patient decisions about appropriate health care for specific clinical circumstances. They should be used as an adjunct to sound clinical decision making.

APPENDIX B

SICKLE CELL TRAIT FREQUENTLY ASKED QUESTIONS

This resource can be used by health care providers when talking with parents about their child's sickle cell trait status.

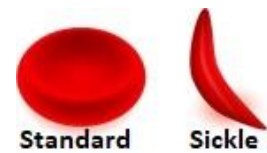
WHAT IS SICKLE CELL TRAIT?

Sickle cell trait (SCT) is not a disease, but an inherited blood condition. This occurs when a person inherits a sickle hemoglobin gene from one parent and a standard (fully working) hemoglobin gene from the other parent. Therefore, a person with one sickle hemoglobin gene is said to 'carry' SCT.

Hemoglobin is a protein in red blood cells that carries oxygen throughout the body.

WHAT IS THE DIFFERENCE BETWEEN SCT AND SICKLE CELL DISEASE (SCD)?

SCT is NOT a milder form of SCD. SCD results from the inheritance of two sickle hemoglobin genes, one from each parent, which causes normally round red blood cells to become curved or "sickle" shape. When the sickled red blood cells travel through small blood vessels, they are more likely to become stuck and clog the blood flow to organs in the body. Since individuals with SCT have only one sickle hemoglobin gene, they still make enough standard hemoglobin to prevent the cells from sickling. Without sickling, red blood cells are able to transport oxygen to tissues and organs in the body without becoming stuck in the small blood vessels. Individuals with SCT are at no higher risk for symptoms of SCD than anyone else.



WHO IS AFFECTED BY SCT?

SCT can occur in any ethnic group, but it is more common in certain populations such as African, Mediterranean, Middle Eastern and Asian groups. Worldwide, an estimated 300 million people carry SCT, with a prevalence ranging from 2% to 30% in more than 40 countries (1). The carrier frequency is estimated to be approximately 1/100 overall in the Alberta population.

WHAT HEALTH COMPLICATIONS ARE ASSOCIATED WITH SCT?

Most people with SCT will rarely have any associated health problems as a result of being a carrier and will not develop SCD. However, people with SCT are at increased chance of experiencing blocked blood vessels should they become significantly dehydrated, particularly during times of extremely strenuous activity. In rare cases, people who carry SCT may have some health issues such as blood in the urine, presence of excess proteins in the urine, or chronic kidney disease.

WHY IS IT IMPORTANT TO KNOW THAT MY CHILD HAS SCT?

Your child and their future partner may want to know this information before they plan to have children of their own. Should your child's partner also have SCT, then their children (your grandchildren) would be at an increased risk of having SCD (see [Figure 1](#)).

WHAT DOES HAVING A CHILD WITH SCT MEAN FOR ME, MY PARTNER AND FOR FUTURE PREGNANCIES?

If both members of a couple carry SCT, they can have a child who has SCD. Each child born to a couple where both parents carry SCT has a:

- 1 in 2 (50%) chance of carrying SCT.
- 1 in 4 (25%) chance of having SCD.
- 1 in 4 (25%) chance that they will neither have SCT or SCD.

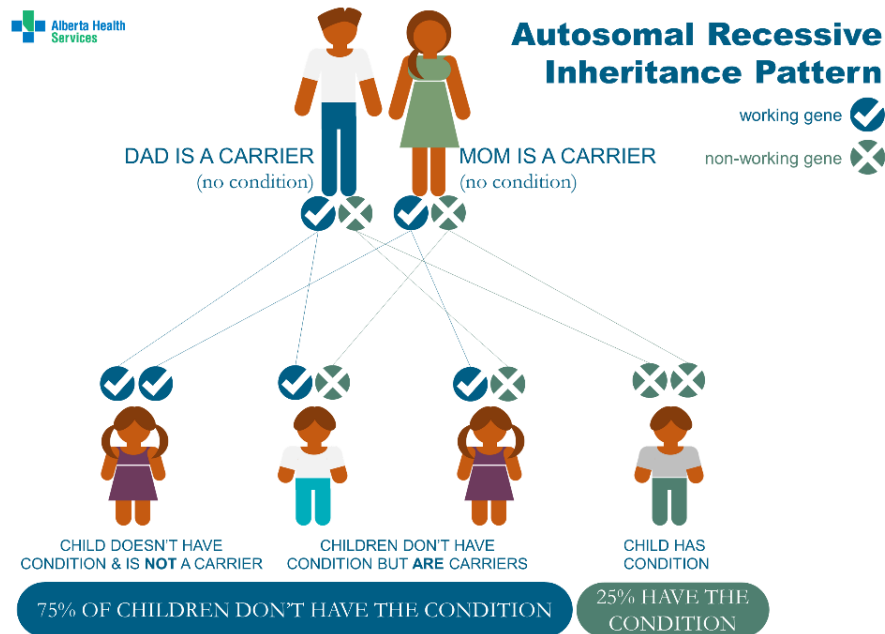


Figure 1: Autosomal Recessive Inheritance Pattern of SCT

If the parents of an infant with SCT are planning more pregnancies, carrier testing is recommended. Genetic counselling is also recommended if both parents are found to be carriers in order to review the different options available to the parents during pregnancy.

SHOULD MY OTHER CHILDREN BE TESTED?

If your other children were born outside of Alberta, or within Alberta but before April 1, 2019 they may not have been screened for SCD. Therefore, we suggest you talk with your family doctor about additional screening for SCD.

References

1. Naik, R. P., Smith-Whitley, K., Hassell, K. L., Umeh, N. I., de Montalembert, M., Sahota, P., et al. Clinical outcomes associated with sickle cell trait: a systematic review. *Ann Intern Med* [Internet]. 2018 November [cited 2019 March 19]; 169 (9), 619-627. Available from: <https://annals.org/aim/article-abstract/2709819/clinical-outcomes-associated-sickle-cell-trait-systematic-review>

Sickle Cell Trait Development Committee Members and Disclosed Conflicts of Interest Summary											
*on topics related to the CPG **from a commercial organization											
Name, credentials	Affiliation	Written articles*	Presented*	Created apps, software, tools, etc.*	On Advisory Board**	On Speakers' bureau**	Received Payment**	Grant(s) or Honorarium*	Patent	Investments	Clinical trial
Aisha Bruce, MD,FRCP	University of Alberta Alberta Health Services – Stollery Children's Hospital	No	No	No	No	No	No	No	No	No	No
Courtney Felker, BKin, MHSc, CRP	Alberta Health Services, Screening Programs	No	No	No	No	No	No	No	No	No	No
Grace Johner	Alberta Health Services, Screening Programs	No	No	No	No	No	No	No	No	No	No
Huiming Yang, MD, MSc, FRCPC (Public Health and Preventive Medicine)	Alberta Health Services University of Calgary Cumming school of Medicine	No	No	No	No	No	No	No	No	No	No
Joseph Ojedokun, MBChB, MRCSI, MICGP; MCCFP, DCH	University of Alberta, Assistant Clinical Professor Family Physician, Life Medical Clinic	No	No	No	No	No	No	No	No	No	No
Kara Patterson, MSc, MHSc, CCGC, CGC	Alberta Health Services, Screening Programs	No	No	No	No	No	No	No	No	No	No
Margaret Lilley, MSc, CCGC, CGC	Alberta Public Laboratories	No	No	No	No	No	No	No	No	No	No

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Name, credentials	Affiliation	Written articles*	Presented*	Created apps, software, tools, etc.*	On Advisory Board**	On Speakers' bureau**	Received Payment**	Grant(s) or Honorarium*	Patent	Investments	Clinical trial
Mary Ann Thomas, MD, FRCPC (Medical genetics)	Alberta Health Services, Alberta Children's Hospital University of Calgary, Associated Professor	No	No	No	No	No	No	No	No	No	No
Michael T Leaker, MD, FRCPC	Deputy Department Head, Strategic and Clinical Affairs Section Chief, Pediatric Hematology Associate Professor of Pediatrics Department of Pediatrics University of Calgary Alberta Children's Hospital Treasurer, Canadian Hemoglobinopathy Association, 2012-present	No	No	No	ApoPharma Ad Board re Deferriprone, 2015.	No	No	No	No	No	No
Nicola Wright, MSc, MD, FRCPC	Alberta Health Services – Alberta Children's Hospital University of Calgary, Cumming School of Medicine										

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Rachel Ferrier, CGC (Certified Genetic Counsellor)	Alberta Health Services - Clinical & Metabolic Genetics Clinic	No	Workshop to the Canadian Association of Genetic Counsellors on this topic about a decade ago, and have given CME to community physicians in the past	No	No	No	No	No	No	No	No
Ross Ridsdale, PhD, MSc, BSc, BA, FACMG	Alberta Public Laboratories University of Alberta, Dept. of Medical Genetics	No	No	No	No	No	No	No	No	No	No
Sajid Merchant, MS, CGC Genetic Counsellor	Alberta Health Services, Clinical & Metabolic Genetics Program University of Alberta - Dept of Medicine and Dentistry	No	No	No	No	No	No	No	No	No	No